This ready reference health guide features 240 major topics that occur regularly in clinical work with children and adolescents. It sorts out the information vital to successful management of common health problems and concerns by presentation of tables, charts, lists, criteria for diagnosis, and other useful tips. References on which the entries are based are provided so that the reader can perform a more extensive search on the topic. The entries are arranged in alphabetical order, and include: (1) abdominal pain; (2) anemias; (3) breathholding; (4) bugs; (5) cholesterol, (6) crying, (7) day care, (8) diabetes, (9) ears, (10) eyes; (11) fatigue; (12) fever; (13) genetics; (14) growth; (15) human bites; (16) hypersensitivity; (17) injuries; (18) intoeing; (19) jaundice; (20) joint pain; (21) kidneys; (22) Lyme disease; (23) meningitis; (24) milestones of development; (25) nutrition; (26) parasites; (27) poisoning; (28) quality time; (29) respiratory distress; (30) seizures; (31) sleeping patterns; (32) teeth; (33) urinary tract; (34) vision; (35) wheezing; (36) x-rays; (37) yellow nails; and (38) zoonoses, diseases transmitted by animals. (TJQ)
the pediatrician
the portable pediatrician
The authors (the big people in white coats, left to right: JAO, JAM, FAO, HM) and friends in the lobby of The Johns Hopkins Children's Center.
the portable pediatrician

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DEDICATION

We dedicate this book to our parents:
Bernice and Samuel Markel
Barbara and Frank Oski
Sara and Aram Oski
and
Dorothy and Robert McMillan,

and to all children, past, present, and future.
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The Portable Pediatrician is intended to instruct, enlighten, and entertain those who are studying or providing for the health care needs of children and adolescents. Although this book is clearly not meant to be an all-encompassing textbook of pediatrics, it is the authors’ hope that the busy practitioner, house officer, medical student, or nurse can turn to these pages in the quest for an important bit of information that solves an immediate problem or to replenish his or her reservoir of knowledge in pediatrics.

We have arranged this book in a dictionary format so that the reader can look up in alphabetical order the subject or key word at hand for quick and ready reference. A more complete index is available at the end of the volume. References on which the individual entries have been based are also provided so that the reader can perform a more extensive search on the topic in question.

As in any endeavor, there are acknowledgements to be made. We would like to express our thanks to Lori Waugh, who patiently transcribed our handwritten notes into a typewritten manuscript; to Laurel Blewett, formerly Research Librarian for the Department of Pediatrics, The Johns Hopkins Hospital; to John J. Hanley and Linda C. Belfus, our editors and publishers; to the many interns, residents, and medical students on whom we tried out much of this material in the form of clinical rounds and teaching sessions; and, of course, to the children who are our patients and who make coming to work each morning such a joyful experience.

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On some of the pages that follow are listed major signs and symptoms and their causes. The causes are classified as COMMON, UNCOMMON or RARE. The COMMON category contains those diseases that, in the aggregate, are responsible for approximately 90% of the patients who have that particular sign or symptom. The term is not meant to suggest that the entity itself is common. The designation UNCOMMON indicates that 1% to 10% of patients with the symptom or sign will be found in that category, whereas the designation RARE indicates the diseases that are responsible for less than 1% of the symptom or sign under discussion. It is common sense, when confronted with any given sign or symptom, to consider the COMMON causes first. (These entries are adapted from Dietz HC, Oski FA: Presenting signs and symptoms. In Oski FA, DeAngelis CD, Feigin FD, Warshaw JB (eds): Principles and Practice of Pediatrics. Philadelphia, Lippincott, 1990, pp 2023-2053.)
ABDOMINAL MASSES

Common Causes

Appendiceal abscess
Bladder distention
Fecal collection
Hepatomegaly (any cause)
Hydronephrosis
Multicystic dysplastic kidney
Neuroblastoma
Polycystic kidney disease
(± liver involvement)
Pregnancy (± ectopic location)
Pyloric stenosis
Splenomegaly (any cause)
Wilms' tumor

Uncommon Causes

Adrenal hemorrhage
Hernia (± incarceration)
Intestinal duplications
Intussusception
Leukemia
Lymphoma
Ovarian cyst
Renal vein thrombosis
Teratoma (abdominal/ovarian)

Rare Causes

Abscess
Anterior meningocele
Aortic aneurysm
Benign cystic causes
  Urachal cyst
  Mesenteric cyst
  Omental cyst
  Pancreatic cyst/pseudocyst
Bezoar
Hepatobiliary causes
  Cholecystitis/ascending cholangitis
  Choledochal cyst
  Hemangioendothelioma
  Hydrops of the gallbladder
Hydrometrocolpos
Intestinal causes
  Intestinal atresia (proximal dilatation)
  Mairotation with volvulus
  Meconium plug/ileus
  Regional enteritis
  Retroperitoneal lymphangioma
Solid tumors
  Granuloma-thecal cell tumor
Hepatoblastoma
  Hepatocellular carcinoma
  Lymphoma
  Mesoblastic nephroma
  Nephroblastomatosis
  Rhabdomyosarcoma

ABDOMINAL PAIN

ACUTE

Common Causes

Appendicitis
Bacterial enterocolitis
Campylobacter
Salmonella
Shigella
Yersinia

Dietary indiscretion
Food poisoning
Mesenteric lymphadenitis
Pharyngitis
Urinary tract infection
Viral gastroenteritis

Uncommon Causes

Cholecystitis/cholelithiasis
Diabetes mellitus
Hepatitis
Herpes zoster
Incarcerated hernia
Infectious mononucleosis
Intussusception
Meckel’s diverticulum
Obstruction (adhesions)
Pelvic inflammatory disease
Peritonitis
Post-trauma/instrumentation
Spontaneous

Pneumonia
Pregnancy (± ectopic location)
Sepsis
Trauma
Bowel perforation
Intramural hematoma
Intrapерitoneal blood
Liver/spleen laceration
Musculocutaneous injury
Pancreatic pseudocyst
Volvulus

Rare Causes

Abdominal abscess
Acute arrhythmia
Acute rheumatic fever
Adynamic ileus
Drugs
Metabolic
Postsurgery/trauma
Ascites
Eosinophilic gastroenteritis
Glomerulonephritis
Hemolysis
Malignancy
Leukemia/lymphoma

Solid tumor (± rupture/hemorrhage)
Mesenteric arterial insufficiency/
occlusion
Nephrolithiasis
Nephrotic syndrome
Obstructive nephropathy
Pancreatitis
Testicular torsion
Vasculitis
Henoch-Schönlein, purpura
Kawasaki’s disease
Polyarteritis nodosa
Systemic lupus erythematosus
RECURRENT

Common Causes

“Psychophysiologic”
Conversion hysteria
Depression
Idiopathic recurrent pain

Reaction anxiety
Secondary gain
Task-induced phobia (e.g., school, sports)

Uncommon Causes

Aerophagia
Constipation
Drugs
  Antibiotics
  Anticonvulsants
  Aspirin
  Bronchodilators
Dysmenorrhea
Enzymatic deficiency
  (e.g., lactose intolerance)
Food allergy
Hepatosplenomegaly (any etiology)

Hiatal hernia
Inflammatory bowel disease
Irritable bowel syndrome
Mittelschmerz syndrome
Parasitic infection
  Ascarisiasis
  Giardiasis
  Strongyloidesis
  Trichinelasis
Peptic ulcerative disease
Sickle-cell anemia
Urinary tract infection

Rare Causes

Abdominal epilepsy
Abdominal masses/malignancies
  Lymphoma
  Neuroblastoma
  Ovarian lesions
  Wilms' tumor
Abdominal migraine equivalent
Acute intermittent porphyria
Addison's disease
Angioneurotic edema
Bowel anomaly with obstruction
  Duplication
  Malrotation
  Stenosis
  Web
Choledochal cyst
Collagen vascular disease
Cystic fibrosis (meconium plug/ileus equivalent)
Endometriosis
Familial Mediterranean fever
Heavy metal intoxication
Hematocolpos
Hirschsprung's disease
Hyperlipoproteinemia
Hyperthyroidism
Hypoperfusion states
  Coarctation of the aorta
  Familial dysautonomia
  Superior mesenteric artery syndrome
Mesenteric cyst
Neurologic
  CNS mass lesion
  Radiculopathy
  Spinal cord injury/tumor
  Recurrent/chronic arrhythmia
  Recurrent pancreatitis
  Wegener's granulomatosis

4—Abdominal Pain

The Differential Diagnosis of Acute Lower Abdominal Pain in Adolescent Women

The complaint of lower abdominal pain in a sexually active adolescent female frequently points toward the work-up for acute pelvic inflammatory disease (PID). Symptoms that often accompany lower abdominal pain include urinary symptoms, nausea, vomiting, fever, malaise, and dyspareunia. Unfortunately these findings are seen in other pathologic processes involving the reproductive tract, as well as disease entities of the gastrointestinal tract and urinary tract. It is obvious, therefore, that one needs to consider a great many problems when evaluating the adolescent female complaining of lower abdominal pain.

<table>
<thead>
<tr>
<th>Urinary Tract</th>
<th>Gastrointestinal Tract</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cystitis</td>
<td>Appendicitis</td>
</tr>
<tr>
<td>Pyelonephritis</td>
<td>Constipation</td>
</tr>
<tr>
<td>Urethritis</td>
<td>Diverticulitis</td>
</tr>
<tr>
<td>Other</td>
<td>Gastroenteritis</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Reproductive Tract</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute pelvic inflammatory disease</td>
<td>Ovarian cyst (torsion/rupture)</td>
</tr>
<tr>
<td>Cervicitis</td>
<td>Pregnancy (intrauterine/ectopic)</td>
</tr>
<tr>
<td>Dysmenorrhea (primary/secondary)</td>
<td>Ruptured follicle</td>
</tr>
<tr>
<td>Ectopic pregnancy</td>
<td>Septic abortion</td>
</tr>
<tr>
<td>Endometriosis</td>
<td>Threatened abortion</td>
</tr>
<tr>
<td>Endometritis</td>
<td>Torsion of adnexa</td>
</tr>
<tr>
<td>Mittelschmerz</td>
<td>Tubo-ovarian abscess</td>
</tr>
</tbody>
</table>

The Closed-eyes Sign in Separating Nonspecific Abdominal Pain from the Acute Abdomen

The child presenting with an acute onset of abdominal pain is frequently a frustrating problem for the pediatrician. Indeed, more than 90% of these children have no organic source of pain that would be amenable to surgical intervention, and they are diagnosed as having “nonspecific abdominal pain.” A group of three surgeons at the Radcliffe Hospital of Oxford University looked for the presence or absence of the “closed eyes” sign during abdominal palpation. Specifically, the surgeons hypothesized that patients with nonspecific abdominal pain were more likely to keep their eyes closed when an examiner palpates the abdomen, whereas patients with abdominal pain of an organic source usually kept their eyes open. The surgeons reasoned that patients with genuine abdominal tenderness are more likely to keep their eyes open in order to watch the examining physician carefully and to avoid unnecessary pain. The Oxonians studied 158 consecutive patients presenting to the emergency room with a complaint of abdominal pain. The data presented below support a new version of an old adage; the eyes have it.
### Numbers of Patients Who Closed Their Eyes During Abdominal Palpation

<table>
<thead>
<tr>
<th>DIAGNOSIS</th>
<th>NO. OF PATIENTS OBSERVED</th>
<th>NO. (%) WHO CLOSED THEIR EYES</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>TOTAL</td>
<td>MALE</td>
</tr>
<tr>
<td>Appendicitis</td>
<td>53</td>
<td>31</td>
</tr>
<tr>
<td>Other disease</td>
<td>38</td>
<td>20</td>
</tr>
<tr>
<td>Non specific abdominal pain</td>
<td>67</td>
<td>25</td>
</tr>
</tbody>
</table>


### ACID-BASE

**Acid-Base Imbalance in Childhood, Which Can Lead to Coma**

Imbalances in a child's acid-base state can progress to severe metabolic derangement and coma. Systemic acidosis or alkalosis generally results from either a primary metabolic process or respiratory abnormalities; the specific causes of these derangements are many. Listed below are the more frequently occurring conditions that can alter a child's acid-base status and progress to coma.

1. **Metabolic acidosis** (increased anion gap)
   - a. Lactic acidosis (e.g., hypoxic-ischemic insult; septic shock)
   - b. Diabetic ketoacidosis
   - c. Renal failure and uremia
   - d. Organic acidurias
   - e. Ingestions (e.g., methanol, paraldehyde, ethylene glycol, acetone, etc.)
   - f. Salicylate poisoning (late)
   - g. Severe diarrhea

2. **Respiratory acidosis** (apena or hypoventilation)
   - a. Supratentorial or infratentorial lesions
   - b. Ingestions (e.g., narcotics, barbiturates, sedatives, clonidine)
   - c. Respiratory muscle fatigue; neuromuscular disease
   - d. Metabolic encephalopathies
   - e. Generalized seizure activity

3. **Respiratory alkalosis** (hyperventilation)
   - a. Intracranial hypertension
   - b. Septic shock (early)
   - c. Hepatic failure
   - d. Salicylate poisoning (early)
   - e. Reye's syndrome
   - f. Brainstem dysfunction

6—Alopecia

**Relationship of pH to PaCO₂ and Base Change**

A rapid calculation will often be of great help in interpreting the significance of blood gas values. When the child is very sick, every second saved can be of enormous importance. Listed below are two useful facts that can enable you to interpret the carbon dioxide and the pH results.

1. A change of PaCO₂ of 10 torr is associated with a decrease or increase in pH of 0.08 units:

   \[ \text{PaCO₂} \ 10 \text{ torr} \ \uparrow \text{ or } \downarrow \ \text{pH} 0.08 \]

   For example:

   \[ \text{PaCO₂} \ 40 \text{ torr} \quad \text{pH} 7.40 \quad \text{Normal} \]
   \[ \text{PaCO₂} \ 50 \text{ torr} \quad \text{pH} 7.32 \quad \text{Respiratory Acidosis—Hypoventilation} \]
   \[ \text{PaCO₂} \ 30 \text{ torr} \quad \text{pH} 7.48 \quad \text{Respiratory Alkalosis—Hyperventilation} \]

2. A base change (base excess or base deficit) of 10 mEq/L is associated with a pH change of 0.15.

   For example:

   \[ \text{PaCO₂} \ 40 \text{ torr} \quad \text{pH} 7.25 \]
   \[ \text{Normal PaCO₂} \quad \text{No respiratory component} \]
   \[ \text{Calculated pH} \ 7.40 \]
   \[ \text{Measured pH} \ 7.25 \]
   \[ \text{pH difference—0.15} \]
   \[ \text{Base deficit} = 10 \text{ mEq/L} \quad \text{—Metabolic acidosis} \]
   \[ \text{No respiratory component—Metabolic acidosis only} \]


**ALOPECIA**

**Common Causes**

- Alopecia areata
- Distal trichorrhexis nodosa
- Physiologic (newborns)
  - Temporal recession at puberty
- Tinea capitis
- Traction alopecia
- Trichotillomania (also *trichologia*)

**Uncommon Causes**

- Acute bacterial infections
  - Cellulitis
  - Folliculitis decalvans
  - Pyoderma
- Burns
- Cancer therapy
  - Antimetabolites
  - Radiation
- Chemical injury
- Kerion
- Proximal trichorrhexis nodosa
- Psoriasis
- Seborrhea
- Viral infections
  - Herpes simplex
  - Varicella
Rare Causes

Circumscribed alopecia
Androgenic alopecia
Aplasia cutis
Conradi’s disease (autosomal dominant chondrodysplasia punctata)
Epidermal nevi-organoid
Follicular aplasia
Goltz’s syndrome (focal dermal hypoplasia)
Hair follicle hamartoma
Incontinentia pigmenti
Infections
Tuberculosis
Inflammatory etiologies
Keratosis follicularis
Lichen planus
Morphea
Porokeratosis of Mibelli
Sarcoid
Systemic lupus erythematosus
Myotonic dystrophy
Diffuse alopecia
Anagen effluvium
Cytostatic agents in plant
Mimosine
Selemocystothionine
Radium
Thallium
Anhidrotic ectodermal dysplasia
Atrichia congenita
Cartilage-hair hypoplasia
Chondroectodermal dysplasia
Crouzon’s syndrome (craniofacial dystosis)
Hair shaft deformities
Monilethrix

Diffuse alopecia (Cont.)
Pili torti
Classic form
Trichopolyiodystrophy (Menkes syndrome)
Trichorrhexis invaginata
Trichorrhexis nodosa
Argininosuccinic aciduria
Hallermann-Streiff syndrome
(mandibulo-oculofacial syndrome)
Hidrotic ectodermal dysplasia
Langer-Giedion syndrome
(trichorhinophalangeal syndrome type II)
Marinesco-Sjögren syndrome
Oculodentodigital dysplasia
Progeria
Rothmund-Thomson syndrome
(congenital poikiloderma)
Telogen effluvium
Childbirth
Chronic infection/illness
Drugs
Anticoagulants
Anticonvulsants
Antikeratinizing drugs
Antithyroid drugs
Heavy metals
Hormones
Excessive dieting
High fever
Hypothyroidism
Stress
Surgery


**ALPHA-FETOPROTEIN**

**Maternal Serum—α-fetoprotein Screening**

Maternal serum α-fetoprotein (MSAFP) screening has been quite successful in identifying neural tube defects in pregnancy. Approximately 80 to 85% of all open neural tube defects can be detected by this method. There also may be a
8—Amenorrhea

relationship of the MSAFP to other birth defects such as Down syndrome and various chromosomal abnormalities.

1. Findings associated with elevated MSAFP
   More advanced gestational age
   Multiple gestation
   Fetal death
   Neural tube defects
   Ventral wall defects
   Congenital nephrosis
   Other fetal malformations
   Oligohydraminos
   Placental anomalies or insufficiency
   Fetomaternal transfusion
   Maternal liver disease or malignancy
   Normal pregnancy

2. Adverse outcomes of pregnancy associated with unexplained MSAFP elevations
   Spontaneous abortion
   Stillbirth
   Prematurity
   Intrauterine growth retardation
   Congenital anomalies
   Possibly pre-eclampsia

3. Findings associated with low MSAFP
   Less advanced gestational age
   Missed abortion
   Hydatidiform mole
   Non-pregnancy
   Fetal chromosomal anomalies
   (e.g., Down syndrome, trisomies
   13 and 18, Turner syndrome)
   Normal pregnancy


AMENORRHEA

Amenorrhea in the Adolescent

Amenorrhea is defined as the absence of normal, spontaneous menstrual periods in a woman of reproductive age. It is typically separated into two forms: primary amenorrhea (the adolescent female who has never achieved menarche), and secondary amenorrhea (the cessation of menstrual cycles, once menarche has occurred, for 3 to 6 months). The most common cause of amenorrhea is pregnancy (including missed abortion and ectopic pregnancy). It is vital to consider amenorrhea, whether primary or secondary, as a symptom and not a disease process in and of itself. Although there is a large number of disease entities that can yield amenorrhea, the basic disease process generally involves one of the following dysfunctions: (1) inadequate hormonal stimulation of the endometrium; (2) an inability of the endometrium to respond to hormonal stimulation; or (3) an obstruction to the outflow of endometrial sloughing. The following table outlines the major causes of amenorrhea.

<table>
<thead>
<tr>
<th>Etiology of Amenorrhea</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>I. CENTRAL NERVOUS SYSTEM (GENERAL)</strong></td>
</tr>
<tr>
<td>A. Infection</td>
</tr>
<tr>
<td>1. Encephalitis</td>
</tr>
<tr>
<td>2. Meningitis</td>
</tr>
<tr>
<td>B. Neoplasm</td>
</tr>
<tr>
<td>1. Craniopharyngioma</td>
</tr>
<tr>
<td><strong>B. Neoplasm (Cont.)</strong></td>
</tr>
<tr>
<td>2. Glioma</td>
</tr>
<tr>
<td>3. Pineal tumor</td>
</tr>
<tr>
<td><strong>C. Congenital anomalies</strong></td>
</tr>
<tr>
<td>1. Hydrocephaly</td>
</tr>
<tr>
<td>2. Sellar malformation</td>
</tr>
</tbody>
</table>

Table continued on next page.
Etiology of Amenorrhea (Cont.)

II. HYPOthalamic
A. Infection
1. Tuberculosis (granuloma)
2. Syphilis (gumma)
B. Inflammatory
1. Sarcoïdosis (granuloma)
C. Neoplasm
1. Cranioopharyngioma
2. Midline teratoma
D. Syndrome
1. Kallmann's
2. Fröhlich’s
3. Laurence-Moon-Bardet-Biedl
e. Kallmann’s
2. Fröhlich’s
3. Laurence-Moon-Bardet-Biedl
F. Tumor
1. Hamartoma
2. Hand-Schüller-Christian disease
C. Congenital anomaly
1. Idiopathic hypogonadotropic hypogonadism
G. Constitutional delay
H. Hypothalamic hyperprolactinemia

III. Pituitary
A. Neoplasm
1. Adenoma
a. Lactotrophic
b. Cushing's disease
c. Acromegaly
d. Chromophobe
2. Carcinoma
B. Idiopathic congenital
1. Hypopituitarism partial or complete
C. Space occupying lesion
1. Arterial aneurysm
2. Empty sella
D. Inflammatory
1. Sarcoïdosis
E. Infiltrative
1. Hemachromatosis
a. Idiopathic
b. Congenital anemia (e.g., thalassemia)
F. Trauma

IV. Gonadal
A. Gonadal dysgenesis
1. Turner’s syndrome
2. Pure gonadal dysgenesis
3. Mixed gonadal dysgenesis
4. XX gonadal dysgenesis
5. XY gonadal dysgenesis (Swell’s syndrome)
B. Insensitive ovary
1. Resistant ovary – Savage’s syndrome
2. Afollicular ovary
a. Idio-ethnic premature aging
b. Injury (e.g., radiation, chemotherapy)
c. Autoimmune disease
d. Infection (e.g., mumps oophoritis)
e. Infiltrative/mucopolysaccharidosis
F. Pregnancy (including missed abortions and ectopic pregnancies)

C. Gonadal agenesis
1. Anorchia (early, late)
2. Ovarian agenesis
a. Idiopathic
b. Surgical
D. Ovarian tumor
1. Androgen-producing
E. True hermaphroditism

V. UTERINE-VAGINAL
A. Müllerian agenesis (Rokitansky’s syndrome)
B. Vaginal agenesis – isolated
C. Cervical agenesis – isolated
D. Vaginal septum transverse
E. Imperforate hymen
F. Asherman’s syndrome – infectious

VI. GENERAL CONDITIONS
A. Endocrinopathy
1. Thyroid disease
a. Hypothyroidism
b. Hyperthyroidism
2. Adrenal disease
a. Cushing’s syndrome
b. Congenital adrenal hyperplasia
c. Adrenal androgen tumor
3. Androgen excess syndrome
a. Polycystic ovarian disease
b. Exogenous androgen therapy
4. Male pseudohermaphroditism
a. Androgen insensitivity syndrome
b. Androgen biosynthetic defects
5. Estrogen biosynthetic defects
6. Diabetes
B. Systemic disease (severe)
1. Examples
a. Crohn’s disease
b. Hepatic failure
c. Glomerulonephritis
d. Systemic lupus erythematosus
C. Nutritional problem
1. Generalized malnutrition (moderate to severe)
2. Weight fluctuations – acute
D. Psychiatric disease
1. Anorexia nervosa
2. Psychosis
E. Miscellaneous conditions
1. Exercise-induced
2. Stress-related

ANDROGENS

Clinical Causes of Androgen Excess in Adolescence

Hirsutism, the increase of sexually stimulated (i.e., androgen-mediated) terminal hair located in the midline of the body and including the face, often accompanies the other tribulations of puberty and adolescence. The presence of excessive facial hair is an unfortunate stigmata to the adolescent woman and is rife with social and psychological implications. Most commonly, hirsutism is confused with hypertrichosis, a generalized increase of vellus or lanugo hair, particularly on the limbs (lanugo on the fetus is replaced by vellus on the infant, which is then replaced by terminal hair). Excessive hair growth that is felt to be androgen-mediated warrants evaluation in order to identify those hirsute girls with Cushing's syndrome, congenital adrenal hyperplasia, or an androgen-producing neoplasm.

In evaluating the hirsute adolescent female, careful attention to the history and physical are necessary, specifically the progression and pattern of hair growth. A detailed menstrual history, appearance of secondary sexual characteristics, body habitus, and weight are also valuable. Laboratory measurements of serum testosterone (which reflects adrenal and ovarian secretion of androgens in addition to peripheral conversion of 4-androstenedione) and DHEA-sulfate (an androgen that is almost exclusively adrenal in origin) should be obtained. A pelvic examination and, depending upon one's findings, and abdominal and pelvic CT scan are useful in delineating ovarian or adrenal tumors. Finally, if congenital adrenal hyperplasia is suggested in the young patient by strong family history of hirsuitism, androgen excess, or hypertension, ACTH stimulation testing and serum 17-OH progesterone measurements are indicated.

Listed below are common causes of androgen excess in adolescence.

1. Ovarian causes
   a. Polycystic ovarian syndrome
   b. Neoplasms.
2. Adrenal causes
   a. Congenital adrenal hyperplasia
      i. 21-hydroxylase deficiency
      ii. 11β-hydroxylase deficiency
      iii. 3β-01-dehydrogenase deficiency
   b. Neoplasm
   c. Nodular hyperplasia
   d. Cushing's syndrome
3. Idiopathic (altered sensitivity and/or metabolism of androgens in the pilosebaceous unit)
4. Iatrogenic
   a. Phenytoin
   b. Danazol
   c. Androgenic steroids
5. Genetic
   a. Incomplete forms of testicular feminization
   b. Mosaic forms of gonadal dysgenesis
6. Miscellaneous
   a. Acromegaly
   b. Porphyria

ANEMIAS

Rule of 34s

The sequence of numbers 34 applies to several values in pediatric hematology. These include:

34 \( \text{mg bilirubin produced/gm Hb} \)

3.4 \( \text{mg iron/gm Hb} \)

3.42 \( \text{nuclear lobes/neutrophil, the upper limit of normal when averaging 100 or more neutrophils} \)

1.34 \( \text{cubic centimeters of oxygen carried by each gram of hemoglobin (if you don’t mind stretching the rule of 34s a little)} \)


Anemia in Early Infancy

During the first months of life there are many causes of anemia. Anemia during the first 3 months of life is rarely a result of nutritional iron deficiency. The accompanying table is intended to call your attention to the more likely causes of anemia that occur at birth and at 2 or 3 months of age, as well as to provide you with leads to establishing the diagnosis.

### Common Causes of Anemia in Early Infancy

<table>
<thead>
<tr>
<th>AGI</th>
<th>DIAGNOSIS</th>
<th>SUPPORTING DATA</th>
</tr>
</thead>
<tbody>
<tr>
<td>At birth</td>
<td>Hemorrhage</td>
<td>History and visual inspection of placenta and cord</td>
</tr>
</tbody>
</table>

*Obstetric accidents* (placenta previa, abruptio placenta, incision of placenta, rupture of cord, rupture of anomalous placental vessel)

*Occult hemorrhage*

- Fetomaternal
- Twin-to-twin

*Internal hemorrhage* (intracranial, retroperitoneal, intrahepatic, intrasplenic, cephalhematoma)

Isooimmunization

Blood groups of mother and infant; evidence of antibody on infant’s red cells

*Table continued on next page.*
### Common Causes of Anemia in Early Infancy (Cont.)

<table>
<thead>
<tr>
<th>AGE</th>
<th>DIAGNOSIS</th>
<th>SUPPORTING DATA</th>
</tr>
</thead>
<tbody>
<tr>
<td>At birth</td>
<td>Inherited defect of red cell (includes G-6-PD deficiency, pyruvate kinase deficiency, hereditary spherocytosis, elliptocytosis, stomatocytosis, etc.)</td>
<td>Red cell morphology, family history, and appropriate screening tests</td>
</tr>
<tr>
<td></td>
<td>Acquired defect (generally in association with hypoxemia, acidosis, or infection)</td>
<td>Physical findings, red cell morphology, coagulation disturbance, blood and urine cultures, and serologic studies and gamma-M determination</td>
</tr>
<tr>
<td></td>
<td>Red cell hypoplasia (Blackfan-Diamond syndrome, congenital leukemia, osteopetrosis)</td>
<td>Rare disorders; bone marrow aspirate</td>
</tr>
<tr>
<td>2-3 months</td>
<td>Iron deficiency as a consequence of previous hemorrhage</td>
<td>Obstetric history when available</td>
</tr>
<tr>
<td></td>
<td>Late manifestation of previous isoimmunization</td>
<td>Blood types of mother and infant; maternal antibody titers</td>
</tr>
<tr>
<td></td>
<td>Hereditary defects of the red cell</td>
<td>Persistence of hemolytic anemia; red cell morphology and laboratory tests</td>
</tr>
<tr>
<td></td>
<td>Thalassemia major</td>
<td>Red cell morphology, splenomegaly, persistence of fetal hemoglobin elevation, family studies</td>
</tr>
<tr>
<td></td>
<td>Sickle cell anemia</td>
<td>Red cell morphology, hemoglobin electrophoresis</td>
</tr>
<tr>
<td></td>
<td>Vitamin E deficiency</td>
<td>Infant of low birth weight; red cell morphology, low serum E level, positive hydrogen peroxide hemolysis test</td>
</tr>
<tr>
<td></td>
<td>Folic acid deficiency</td>
<td>Premature infant, history of infections or diarrhea, red cell and marrow morphology, response to folic acid</td>
</tr>
<tr>
<td></td>
<td>Persistent infection</td>
<td>Elevated titers to rubella, cytomegalovirus, toxoplasmosis</td>
</tr>
<tr>
<td></td>
<td>Renal tubular acidosis</td>
<td>Acidosis, hypochloremia, mild azotemia, urine pH of 6.0 or greater in presence of acidosis</td>
</tr>
</tbody>
</table>


### Anemias Associated with a Low MCV*

- **98% of anemias with low MCV:**
  - Iron deficiency
  - α-thalassemia
  - β-thalassemia

- **Others:**
  - Lead poisoning
  - Protein-calorie malnutrition
  - Copper deficiency
  - Sideroblastic anemia

---

*MCV = mean cell volume.
† Microcytosis most commonly results from associated iron deficiency.
ANION GAP

In these days of automated laboratory procedures, most sick patients will have their serum electrolytes measured. Obvious abnormalities are easily recognized. Hidden clues to diagnosis are also present in these numbers. Interpretation of the anion gap provides such a clue.

The principle of electroneutrality is always working and dictates that the sum of the positive charges, i.e., the mEq/L of cations, be exactly counterbalanced by the number of negative charges, i.e., the mEq/L of anions.

The principal cations in the plasma include sodium, potassium, calcium, and magnesium. The principal anions are chloride, bicarbonate, carbonic acid, dissolved carbon dioxide, albumin, globulin, sulfate, phosphate, and the organic acids, lactic and pyruvic acid.

Measurement of all the anions and cations is not required for interpretation of the patient's status. The serum sodium and potassium are representative of the extracellular fluid cations, and, in fact, account for 95% of the cations present. Chloride and bicarbonate account for 85% of the anions. Thus, the sum of the usually measured anions does not fully counterbalance the sum of the measured cations. Their difference is termed the anion gap. Because of potassium's relatively low and stable serum concentration, it has only a minor influence on the anion gap. Therefore the anion gap equation can be simplified to read as follows:

\[ \text{Anion gap} = \text{sodium} \times (\text{chloride} + \text{bicarbonate}) \]

The normal value for the anion gap is approximately 12.0 ± 2.0. The normal range for the anion gap is thus 8 to 16 mEq/L.

Causes of a high anion gap include:

- Metabolic acidosis
- Dehydration
- Therapy with sodium salts of strong acids
- Therapy with certain antibiotics (carbenicillin, large doses of sodium penicillin)
- Alkalosis

Specific causes of high anion gap metabolic acidosis include:

- Uremia
- Lactic acidosis
- Methanol intoxication
- Ketoacidosis
- Salicylate intoxication
- Paraldehyde toxicity

Specific causes of normal anion gap metabolic acidosis include:

- Gastrointestinal bicarbonate loss
- Diarrhea or pancreatic fistula
- Ureterenterostomy
- Drugs: acetazolamide, sulfamylon, cholestyramine, acidifying agents (ammonium chloride, oral calcium chloride, arginine, hydrochloride, lysine hydrochloride)
- Rapid intravenous hydration
- Hyperalimentation
- Posthypocapnia
- Renal tubular acidosis
### Causes of a low anion gap include:

- Reduced concentration of unmeasured anions
- Dilution
- Hypoalbuminemia
- Systematic underestimation of serum sodium
- Severe hypernatremia
- Hyperviscosity
- Systematic overestimation of serum chloride
- Bromism
- Retained nonsodium cations
- Paraproteinemia
- Hypercalcemia
- Hypermagnesemia


### ANOREXIA

#### Common Causes

- Acute infection
- Apparent anorexia
  - Dieting/fear of obesity
  - Manipulative behavior
  - Unrealistic expectations of caretakers

#### Uncommon Causes

- Chronic infection
- Drugs
  - Aminophylline
  - Amphetamines
  - Anticonvulsants
  - Antihistamines
  - Antimetabolites
  - Digitalis
  - Narcotics
- Esophagitis/gastroesophageal reflux
- Food aversion in athletes
- Iron deficiency
- Irritable bowel syndrome
- Pregnancy
- Psychosocial deprivation
  - (neglect/abuse)
- Psychosocial factors
  - Chronic mental/environmental stress
  - Anxiety
  - Fear
  - Loneliness/boredom
- Depression
- Grief
- Mania

#### Rare Causes

- Acquired immunodeficiency syndrome (AIDS)
- Adrenogenital syndrome
- Alcohol/drug abuse
- Anorexia nervosa
- Chronic disease
- Collagen vascular disease
- Congestive heart failure
- Cyanotic heart disease
- Electrolyte disturbances
  - Hypochloremia
  - Hypokalemia
- Endocrine disease
  - Addison’s disease
  - Diabetes insipidus
  - Hyperparathyroidism
Endocrine Disease (Cont.)
- Hypothyroidism
- Panhypopituitarism
- Hypervitaminosis A
- Inborn errors of metabolism
- Kwashiorkor
- Lead poisoning
- Liver failure

Neurologic
- Congenital degenerative disease
- Diencephalic syndrome
- Hypothalamic lesions
- Increased intracranial pressure
- Mental retardation/cerebral palsy

Pain avoidance
- Appendicitis
- Constipation
- Gastrointestinal obstruction
- Inflammatory bowel disease
- Pancreatitis
- Superior mesenteric syndrome

Kwashiorkor

Lead poisoning

Liver failure

Pain avoidance

Hypervitaminosis A

Inborn errors of metabolism

Kwashiorkor

Lead poisoning

Liver failure

Neurologic
- Congenital degenerative disease
- Diencephalic syndrome
- Hypothalamic lesions
- Increased intracranial pressure
- Mental retardation/cerebral palsy

Polycythemia

Postsurgical outcome

Pulmonary insufficiency

Renal failure

Renal tubular acidosis

Schizophrenia

Zinc deficiency


Anorexia Nervosa and Bulimia

Anorexia nervosa is characterized by excessive weight loss due to a self-inflicted starvation and a morbid, and often unrealistic, fear of becoming too fat. Amenorrhea frequently accompanies this disorder in women; a decreased libido has been noted in anorexic men. A related eating disorder, bulimia, is compulsive overeating followed by drastic attempts to avoid gaining weight as a result of the eating binge, e.g., self-induced vomiting, the ingestion of self-prescribed laxatives or diuretics, and strenuous exercise. These two eating disorders share many features and can actually evolve from one into the other; classically, anorexia nervosa evolves into bulimia. They also have distinctive features that can distinguish one from the other.

Comparison of Anorexia Nervosa (Food-Restricting) and Bulimia

<table>
<thead>
<tr>
<th>ANOREXIA NERVOSA (FOOD-RESTRICTING)</th>
<th>BULIMIA</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Similar Features</strong></td>
<td></td>
</tr>
<tr>
<td>1. Psychological</td>
<td></td>
</tr>
<tr>
<td>a. Fear of fatness</td>
<td>d. Variable degree of distortion of body size</td>
</tr>
<tr>
<td>b. Active pursuit of weight loss</td>
<td>e. Family history of affective disorder</td>
</tr>
<tr>
<td>c. Fear of loss of control of eating</td>
<td></td>
</tr>
<tr>
<td>2. Medical</td>
<td>f. Constipation</td>
</tr>
<tr>
<td>a. Orthostatic hypotension</td>
<td>g. Aerocyanosis</td>
</tr>
<tr>
<td>b. Return to prepubertal breast development</td>
<td>h. Lanugo hair</td>
</tr>
<tr>
<td>c. Amenorrhea</td>
<td>i. Pedal edema</td>
</tr>
<tr>
<td>d. Bradycardia</td>
<td>j. Loss of subcutaneous lipid layer and decreased muscle mass</td>
</tr>
<tr>
<td>e. Lowered core temperature</td>
<td></td>
</tr>
</tbody>
</table>

(All of the above medical complications are the result of starvation.)

Table continued on next page.
### Comparison of Anorexia Nervosa (Food-Restricting) and Bulimia (Cont.)

<table>
<thead>
<tr>
<th>ANOREXIA NERVOSA (FOOD-RESTRICTING)</th>
<th>BULIMIA</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Contrasting Features</strong></td>
<td></td>
</tr>
<tr>
<td>1. Food intake severely restricted</td>
<td>1. Control of intake is lost resulting in binges</td>
</tr>
<tr>
<td>2. Less vomiting, diuretic, or laxative abuse</td>
<td>2. Self-induced vomiting, laxative and diuretic abuse</td>
</tr>
<tr>
<td>3. Younger</td>
<td>3. Older</td>
</tr>
<tr>
<td>5. Denies hunger</td>
<td>5. Experiences hunger</td>
</tr>
<tr>
<td>7. Most of the medical complications stem from chronic starvation</td>
<td>7. Many medical complications may stem from starvation but there also exist a number of gastrointestinal complaints that result from self-induced vomiting and laxative and diuretic abuse (e.g., loss of dental enamel, parotid gland swelling, dry mouth, esophagitis, gastric dysrhythmia, irritable bowel syndrome, and constipation). Renal problems, hypokalemic alkalosis, cardiac arrhythmias, and tetany can also result from laxative and diuretic abuse. Scars on the dorsum of the hand from frequent, self-induced vomiting.</td>
</tr>
<tr>
<td>8. Eating behavior is a source of pride</td>
<td>8. Eating behavior is a source of shame</td>
</tr>
<tr>
<td>10. Amenorrhea or loss of sex drive</td>
<td>10. Variable amenorrhea and change in sex drive</td>
</tr>
<tr>
<td>11. Death from starvation acutely; chronically from starvation or suicide</td>
<td>11. Death from hypokalemia acutely; chronically from suicide</td>
</tr>
<tr>
<td>12. Patient is described as a &quot;model&quot; child</td>
<td>12. Patient often exhibits behavioral abnormalities</td>
</tr>
</tbody>
</table>


### ANTIBIOTICS

**Tasteful Antibiotics, or “Just a Spoonful of Sugar Helps the Medicine Go Down”**

These days, the successful marketing of antibiotic suspensions for children is as competitive as any other industry. Creating an effective and safe antibiotic is simply not enough. It also has to taste good! Listed below are the flavors, sugar content, and availability of some of the most commonly prescribed antibiotics in general pediatric practice:
Antimicrobial Suspensions Tested

<table>
<thead>
<tr>
<th>Suspension Name</th>
<th>Generic Name</th>
<th>Product</th>
<th>Strength (mg/5 ml)</th>
<th>Color</th>
<th>Flavor</th>
<th>Sweetener</th>
</tr>
</thead>
<tbody>
<tr>
<td>Augmentin</td>
<td>Amoxicillin; clavulanate</td>
<td>Beecham</td>
<td>250 mg</td>
<td>Cream</td>
<td>Orange</td>
<td>Saccharin</td>
</tr>
<tr>
<td>Trimox</td>
<td>Amoxicillin</td>
<td>Squibb</td>
<td>250 mg</td>
<td>Pink</td>
<td>Cherry</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Cefaclor</td>
<td>Cefixime</td>
<td>Lederle</td>
<td>100 mg</td>
<td>Cream</td>
<td>Strawberry</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Suprax</td>
<td>Cephalexin</td>
<td>Distacur</td>
<td>250 mg</td>
<td>Orange</td>
<td>Bubble gum</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Kellex</td>
<td>Dieloxacillin</td>
<td>Bristol</td>
<td>62.5 mg</td>
<td>Pink</td>
<td>Orange; pineapple</td>
<td>Saccharin/ sucrose</td>
</tr>
<tr>
<td>Pediazole</td>
<td>Erythromycin ES; sulfoxazole</td>
<td>Ross</td>
<td>200 mg; 600 mg</td>
<td>White</td>
<td>Strawberry; banana</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Erythromycin ES</td>
<td>Erythromycin ethylsuccinate</td>
<td>Barr</td>
<td>200 mg</td>
<td>Pink</td>
<td>Cherry</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Ilosone</td>
<td>Erythromycin estolate</td>
<td>Distacur</td>
<td>250 mg</td>
<td>Red</td>
<td>Cherry</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Grifulvin V</td>
<td>Griseofulvin microsize</td>
<td>Ortho</td>
<td>125 mg</td>
<td>Peach</td>
<td>Raspberry</td>
<td>Saccharin/ sucrose</td>
</tr>
<tr>
<td>VecTids</td>
<td>Penicillin VK</td>
<td>Squibb</td>
<td>250 mg</td>
<td>Red</td>
<td>Berry-like</td>
<td>Saccharin/ sucrose</td>
</tr>
<tr>
<td>Gantrisin</td>
<td>Sulfisoxazole</td>
<td>Roche</td>
<td>500 mg</td>
<td>White</td>
<td>Raspberry</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Achromycin-V</td>
<td>Tetracycline</td>
<td>Lederle</td>
<td>25 mg</td>
<td>Red</td>
<td>Cherry</td>
<td>Sucrose</td>
</tr>
<tr>
<td>Sulfatrim</td>
<td>Trimethoprim-sulfamethoxazole</td>
<td>Barre</td>
<td>40 mg; 200 mg</td>
<td>Pink</td>
<td>Cherry</td>
<td>Saccharin/ sucrose</td>
</tr>
</tbody>
</table>


**ANTICONVULSANTS**

The anticonvulsants often produce side-effects. The commonly used anticonvulsants and their commonly produced side-effects are described in the table below.

Side-effects of Commonly Used Anticonvulsants

<table>
<thead>
<tr>
<th>Anticonvulsant</th>
<th>Predictable</th>
<th>Idiosyncratic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Carbamazepine</td>
<td>Diplopia</td>
<td>Agranulocytosis</td>
</tr>
<tr>
<td></td>
<td>Dizziness</td>
<td>Aplastic anemia</td>
</tr>
<tr>
<td></td>
<td>Drowsiness</td>
<td>Hepatotoxicity</td>
</tr>
<tr>
<td></td>
<td>Headache</td>
<td>Photosensitivity</td>
</tr>
<tr>
<td></td>
<td>Nausea</td>
<td>Stevens-Johnson syndrome</td>
</tr>
<tr>
<td></td>
<td>Hyponatremia</td>
<td>Lupus-like syndrome</td>
</tr>
<tr>
<td></td>
<td>Hypocalcemia</td>
<td>Morbilliform rash</td>
</tr>
<tr>
<td></td>
<td>Orofacial dyskinesia</td>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td></td>
<td>Cardiac arrhythmia</td>
<td>Pseudolymphoma</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Side-effects of Commonly Used Anticonvulsants (Cont.)

<table>
<thead>
<tr>
<th>ANTICONVULSANT</th>
<th>PREDICTABLE</th>
<th>IDIOSYNCRATIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium valproate</td>
<td>Anorexia Peripheral edema</td>
<td>Acute pancreatitis</td>
</tr>
<tr>
<td></td>
<td>Dyspepsia</td>
<td>Hepatotoxicity</td>
</tr>
<tr>
<td></td>
<td>Nausea Weight gain</td>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td></td>
<td>Vomiting Drowsiness</td>
<td>Hyperammonemia</td>
</tr>
<tr>
<td></td>
<td>Hair loss Tremor</td>
<td>Stupor</td>
</tr>
<tr>
<td></td>
<td>Rash</td>
<td>Encephalopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Teratogenicity</td>
</tr>
<tr>
<td>Phenytoin</td>
<td>Anorexia Paradoxical seizures</td>
<td>Blood dyscrasias</td>
</tr>
<tr>
<td></td>
<td>Dyspepsia</td>
<td>Lupus-like syndrome</td>
</tr>
<tr>
<td></td>
<td>Nausea Gum hypertrophy</td>
<td>Reduced serum IgA</td>
</tr>
<tr>
<td></td>
<td>Vomiting Coarse facies</td>
<td>Pseudolymphoma</td>
</tr>
<tr>
<td></td>
<td>Aggression Hirsutism</td>
<td>Peripheral neuropathy</td>
</tr>
<tr>
<td></td>
<td>Ataxia Megaloblastic anemia</td>
<td>Rash</td>
</tr>
<tr>
<td></td>
<td>Cognitive impairment</td>
<td>Stevens-Johnson syndrome</td>
</tr>
<tr>
<td></td>
<td>Depression Hyperglycemia</td>
<td>Dupuytren's contracture</td>
</tr>
<tr>
<td></td>
<td>Drowsiness Osteomalacia</td>
<td>Hepatotoxicity</td>
</tr>
<tr>
<td></td>
<td>Headache Neonatal</td>
<td>Teratogenicity</td>
</tr>
<tr>
<td></td>
<td>Nystagmus hemorrhage</td>
<td></td>
</tr>
<tr>
<td>Phenytoin</td>
<td>Fatigue Decreased libido</td>
<td>Macropapular rash</td>
</tr>
<tr>
<td></td>
<td>Listlessness Impotence</td>
<td>Exfoliation</td>
</tr>
<tr>
<td></td>
<td>Tiredness Folate deficiency</td>
<td>Toxic epidermal necrolysis</td>
</tr>
<tr>
<td></td>
<td>Depression Neonatal</td>
<td>Hepatotoxicity</td>
</tr>
<tr>
<td></td>
<td>Insomnia* Hemorrhage</td>
<td>Dupuytren's contracture</td>
</tr>
<tr>
<td></td>
<td>Distractability* Hemorrhage</td>
<td>Frozen shoulder</td>
</tr>
<tr>
<td></td>
<td>Aggression* Hypocalcemia</td>
<td>Teratogenicity</td>
</tr>
<tr>
<td></td>
<td>Poor memory Osteomalacia</td>
<td></td>
</tr>
<tr>
<td>Primadone</td>
<td>Nausea Psychosis</td>
<td>Rash</td>
</tr>
<tr>
<td></td>
<td>Vomiting Neonatal</td>
<td>Agranulocytosis</td>
</tr>
<tr>
<td></td>
<td>Drowsiness Hemorrhage</td>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td></td>
<td>Weakness Decreased libido</td>
<td>Lupus-like syndrome</td>
</tr>
<tr>
<td></td>
<td>Dizziness Impotence</td>
<td>Teratogenicity</td>
</tr>
<tr>
<td></td>
<td>Diplopia Hypocalcemia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Nystagmus Osteomalacia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ataxia Megaloblastic anemia</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Personality change Neatal</td>
<td></td>
</tr>
<tr>
<td></td>
<td>change Hemorrhage</td>
<td></td>
</tr>
<tr>
<td>Ethosuximide</td>
<td>Anorexia Headache</td>
<td>Rash</td>
</tr>
<tr>
<td></td>
<td>Nausea Lethargy</td>
<td>Erythema multiforme</td>
</tr>
<tr>
<td></td>
<td>Vomiting Parkinsonism</td>
<td>Stevens-Johnson syndrome</td>
</tr>
<tr>
<td></td>
<td>Agitation Psychosis</td>
<td>Lupus-like syndrome</td>
</tr>
<tr>
<td></td>
<td>Drowsiness</td>
<td>Agranulocytosis</td>
</tr>
<tr>
<td>Clonazepam/clobazam</td>
<td>Fatigue Hyperkinesia*</td>
<td>Rash</td>
</tr>
<tr>
<td></td>
<td>Dizziness Hypersalivation*</td>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td></td>
<td>Drowsiness Bronchorrhcea*</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Ataxia Weight gain</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Irritability Muscle weakness</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Aggression Psychosis</td>
<td></td>
</tr>
</tbody>
</table>

*In children

APNEA

Common Causes

Breathholding spells
Bronchiolitis
Extrinsic suffocation
Gastroesophageal reflux/aspiration

Idiopathic (? CNS immaturity)
Prematurity
Seizure

Uncommon Causes

Asthma
Bronchopulmonary dysplasia “spells
CNS hypoperfusion
CNS trauma/bleed
Congenital airway anomaly
Hypoglycemia
Hypoxemia/hypercarbia (severe)
Infection
Croup
Meningitis/encephalitis

Infection (Cont.)
Epiglottitis
Pertussis
Pneumonia
Sepsis
Laryngospasm
Laryngo-tracheo-bronchomalacia
Obstructive sleep apnea
SIDS
Toxins/drugs

Rare Causes

Anemia
Arrhythmia
Glossoptosis
Guillain-Barré syndrome
Hypocalcemia
Increased intracranial pressure
Infantile botulism
Intraventricular hemorrhage
Macroglossia
Metabolic disease
Hyperammonemia

Metabolic disease (Cont.)
Inborn errors
Metabolic alkalosis
Micrognathia
Ondine’s curse
Spinal cord injury
Cervical spine instability
Down syndrome
Dwarfism
Trauma
Tumor (CNS, airway)


Sleep Apnea

The child with unrecognized sleep apnea may present to the pediatrician with any number of chief complaints: cardiovascular abnormalities, failure to thrive, pulmonary abnormalities, obesity, apparent mental retardation, and recurrent respiratory infections. An inadequate history may fail to reveal the culprit.

Sleep apnea can occur in infants, children and adults of any age, although the incidence is known to increase with age and be more prevalent among males than females. The diagnosis depends upon an eye for the predisposing factors and an ear for the symptoms.
20—Argyll Robertson Pupil

Sleep apnea: Predisposing factors

- Enlarged tonsils or adenoids
- Upper airway or maxillofacial abnormalities
- Hyperthyroidism
- Obesity-Pickwickian syndrome
- Down syndrome
- Hypotonic cerebral palsy
- Congenital myopathies
- Pharyngeal “sphincter”
- Dysautonomia

Symptoms of sleep apnea patients

<table>
<thead>
<tr>
<th>Symptom</th>
<th>% of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Snoring—usually all night every night; worse with respiratory infections</td>
<td>91</td>
</tr>
<tr>
<td>Apnea—observed by parents</td>
<td>81</td>
</tr>
<tr>
<td>Restless sleep and abnormal sleep positions</td>
<td>70</td>
</tr>
<tr>
<td>Awakenings from sleep at night</td>
<td>60</td>
</tr>
<tr>
<td>Nocturnal enuresis (children &gt; 4 years of age)</td>
<td>33</td>
</tr>
<tr>
<td>Daytime somnolence</td>
<td>31</td>
</tr>
<tr>
<td>Irritability, hyperactivity</td>
<td>22</td>
</tr>
<tr>
<td>Cardiomegaly</td>
<td>6</td>
</tr>
</tbody>
</table>

The most common cause of sleep apnea in infancy and childhood is tonsillar and adenoidal hypertrophy, which may require surgical intervention. Beware the symptoms and predispositions. You may avert congestive heart failure or cor pulmonale!


THE ARGYLL ROBERTSON PUPIL

Its Clinical Significance

The Argyll Robertson pupil as a sign of tabes dorsalis or neurosyphilis was described in 1868 by the eye surgeon Douglas Moray Cooper Lamb Argyll Robertson (1837–1909) of Edinburgh, Scotland. It is a miotic pupil that accommodates but fails to react to direct light. The sign is caused by lesions to the area immediately rostral to the Edinger-Westphal nucleus of the midbrain and can be found in a number of conditions that affect this area (see figure). For example, Charles Dickens, in his 1855 novel Little Dorrit, described a young girl named Maggy who was severely afflicted with “brain fever” or encephalitis and whose eyes were “very little affected by light” and stood “unnaturally still.” More important to the present day clinician is the association of the Argyll Robertson pupil with Bannwarth’s lymphocytic meningoradiculitis, a syndrome of radicular pain, cranial nerve palsies, and sensory and motor impairment secondary to infection with Borrelia burgdorferi or Lyme disease.
Lesions in the shaded area (periaqueductal gray [PG]) interrupt descending pathways from the oculomotor complex to the Edinger-Westphal nucleus (EWN). SC = superior colliculus; A = aqueduct; RN = red nucleus; SN = substantia nigra; CP = cerebral peduncle. (From Dasco CC, Bortz DL, Am J Med 86:199-202, 1989, with permission.)

Listed below are reported non-syphilitic causes of the Argyll Robertson pupil.

1. Diabetes mellitus
2. Multiple sclerosis
3. Wernicke's encephalopathy
4. Dejerine-Sottas progressive hypertrophic neuritis
5. Charcot-Marie-Tooth disease (peroneal muscular atrophy)
6. Tumors and hemorrhage affecting the Edinger-Westphal nucleus (e.g., midbrain tumors such as pinealomas, third ventricle gliomas, and pituitary stalk tumors)
7. Herpes zoster
8. Lyme disease (Bannwarth's syndrome)
9. Sarcoidosis
10. von Economo's disease (encephalitis secondary to influenza)


ARTHRITIS

Differential Diagnosis of Childhood Arthritis

Arthritis in childhood represents a special problem to the pediatrician, because both the types and etiologies cover a broad spectrum. In the child with suspected septic arthritis, an early diagnosis is especially important for the prevention of deformities and/or functional impairment.
## Clinical Criteria for Diagnosis of Childhood Arthritis

1. Swelling of a joint
2. Limitation of motion with heat, pain, or tenderness

<table>
<thead>
<tr>
<th>DIAGNOSIS</th>
<th>DEFINITION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Juvenile rheumatoid arthritis</td>
<td>See JRA entries</td>
</tr>
<tr>
<td>Enteroarthritis</td>
<td>Antecedent enterobacterial infection (Yersinia, Salmonella, Shigella, or Campylobacter species) verified by stool culture or agglutination titer ≥ 1:160.</td>
</tr>
<tr>
<td>Septic arthritis</td>
<td>Positive bacterial culture from synovial fluid</td>
</tr>
<tr>
<td>Transient synovitis of the hip</td>
<td>Acute hip effusion verified by ultrasonography, roentgenography, synovial fluid aspirate, or clinical findings</td>
</tr>
<tr>
<td>(TSH) JRA</td>
<td>Typical clinical picture with petechial rash and normal platelet count</td>
</tr>
<tr>
<td>Henoch-Schönlein purpura</td>
<td>Acute urticaria 5–12 days after vaccination</td>
</tr>
<tr>
<td>Serum sickness</td>
<td>Disease duration &lt; 3 months; diagnosis of exclusion</td>
</tr>
<tr>
<td>Acute transient arthritis</td>
<td>Joint pain without trauma; no physical signs of arthritis</td>
</tr>
<tr>
<td>Arthralgia</td>
<td>Arthroscopically or radiologically verified bone disease, or internal derangement of joint, especially knee</td>
</tr>
<tr>
<td>Orthopedic disease</td>
<td>An ESR 20 in the presence of a low CRP and no fever suggests JRA or other connective tissue disease and necessitates further immunologic workup. JRA may also present as FUO.</td>
</tr>
</tbody>
</table>

### Laboratory Tests in the Differential Diagnosis of Juvenile Arthritis

<table>
<thead>
<tr>
<th>PATIENT GROUP</th>
<th>TEST</th>
<th>SIGNIFICANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>All children with joint symptoms</td>
<td>C reactive protein (CRP), erythrocyte sedimentation rate (ESR), CBC, platelet count, urinalysis, bacterial culture of throat smear</td>
<td>CRP &gt; 20, ESR &gt; 20, WBC &gt; 1500, and T° &gt; 38.5°C suggest septic or enteroarthritis. Low CRP and absence of fever with acute limp and hip pain suggest TSH. An ESR 20 in the presence of a low CRP and no fever suggests JRA or other connective tissue disease and necessitates further immunologic workup. JRA may also present as FUO.</td>
</tr>
<tr>
<td>Arthritis lasting longer than 2 weeks</td>
<td>Anti-nuclear antibodies, serum immunoglobulins, Yersinia antibodies, Salmonella antibodies, stool bacterial culture</td>
<td>Elevated in JRA and other CT diseases. IgG elevated in JRA. Yersinia and/or salmonella Ab's are thought to be valid indicators of enteroarthritis as are positive stool cultures. EA onset generally acute while JRA normally insidious.</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
Laboratory Tests in the Differential Diagnosis of Juvenile Arthritis (Cont.)

<table>
<thead>
<tr>
<th>PATIENT GROUP</th>
<th>TEST</th>
<th>SIGNIFICANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Special indications</td>
<td>Rheumatoid factor, antistreptolysin O (ASO)</td>
<td>Rarely indicated in child &lt; 8 years of age. Both tests for suspected ARF.</td>
</tr>
<tr>
<td></td>
<td>Viral antibodies</td>
<td>Indicated when systemic onset JRA suspected.</td>
</tr>
<tr>
<td></td>
<td>Chlamydia antibodies</td>
<td>Rare in childhood reactive arthritis. More commonly seen in adults.</td>
</tr>
</tbody>
</table>


The Three Modes of Onset of Juvenile Rheumatoid Arthritis

Juvenile rheumatoid arthritis (JRA) differs from rheumatoid arthritis in adults in several ways, including types of onset. The three forms of onset of JRA are: (1) the acute febrile onset (systemic disease), (2) the monoarticular or pauciarticular onset (oligoarthritis), and (3) polyarticular onset (polyarthritis).

Systemic disease is manifested by spiking fevers on a daily basis plus the appearance of a characteristic rash.

Oligoarthritis is defined as onset in four or fewer joints, often only one, usually the knee.

Polyarthritis is defined as onset in five or more joints.

All three forms can mimic other diseases and the diagnosis is often one of exclusion. It is important to be intimately familiar with the clinical signs and symptoms of each type of onset to avoid the serious consequences of misdiagnosis.

Approximately 5% of all cases of JRA begin in childhood (by definition before 16 years, usually between 1 and 3 years). It is the most common pediatric connective tissue disease, and about a quarter of a million children in the U.S. are affected.

### Three Modes of Onset of Juvenile Rheumatoid Arthritis

<table>
<thead>
<tr>
<th></th>
<th>ACUTE FEBRILE ONSET</th>
<th>MONOARTICULAR ONSET</th>
<th>POLYARTICULAR ONSET</th>
</tr>
</thead>
<tbody>
<tr>
<td>Per cent</td>
<td>20</td>
<td>30</td>
<td>50</td>
</tr>
<tr>
<td>Joint Manifestations</td>
<td>One-half have no joint swelling at onset. The other one-half have only arthralgia. Pain may be inferred from the flexed-knee position in which these children tend to lie.</td>
<td>The knee is most common site of onset. Other sites are ankle, elbow, wrist and finger joints. Swelling, stiffness, and pain are usually minimal. Painful tendinitis or bursitis, especially of the heel, may be the presenting symptom.</td>
<td>Four or more joints are involved. May have abrupt onset with painful swelling of knees, ankles, feet, and hands. May have insidious onset with no complaint of pain. Joint involvement must be inferred from guarding movements and knee-flexed position.</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Three Modes of Onset of Juvenile Rheumatoid Arthritis (Cont.)

<table>
<thead>
<tr>
<th>Joint Manifestations (Cont.)</th>
<th>ACUTE FEBRILE ONSET</th>
<th>MONOARTICULAR ONSET</th>
<th>POLYARTICULAR ONSET</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fever</td>
<td>Daily spikes to 105°F or higher with temperature falling sometimes to subnormal levels. Fever may precede arthritis by weeks, months, or years.</td>
<td>There may be low-grade daily fever spikes.</td>
<td>Low-grade fever with daily spikes.</td>
</tr>
<tr>
<td>Rash</td>
<td>90% have macular or slightly maculopapular rash usually on the trunk and extremities, occasionally on the neck and face. Rash is rarely pruritic, is usually fleeting with macules appearing for a few hours during the day or week, usually in conjunction with fever. Rash is more florid when the skin is rubbed or scratched (Köbner phenomenon).</td>
<td>Rash is sometimes present, but is rarely of diagnostic help.</td>
<td>Maculopapular rash is sometimes present.</td>
</tr>
<tr>
<td>Iridocyclitis</td>
<td>Rarely occurs in patients presenting in this way.</td>
<td>This group is most susceptible to ocular disease. It is often asymptomatic and may smolder for weeks or months. It may be the first manifestation of the disease. If undetected and untreated, it may lead to blindness from band keratopathy and cataracts. Diagnosis may be made only by slit lamp examination.</td>
<td>Rarely occurs in patients presenting in this way.</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td>May be generalized. Splenomegaly may be present. Enlarged mesenteric nodes may lead to abdominal pain and vomiting. Lymphadenopathy may suggest lymphoma or leukemia.</td>
<td>Infrequent.</td>
<td>Infrequent.</td>
</tr>
<tr>
<td>Cardiac manifestations</td>
<td>10% have pericarditis clinically. Pericarditis may last 2 to 12 weeks and may recur years later. Myocarditis and resulting heart failure may occur.</td>
<td>Infrequent.</td>
<td>Infrequent.</td>
</tr>
</tbody>
</table>

Table continued on next page.
Three Modes of Onset of Juvenile Rheumatoid Arthritis (Cont.)

<table>
<thead>
<tr>
<th>General appearance</th>
<th>Laboratory</th>
<th>Differential diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>ACUTE FEBRILE ONSET</strong></td>
<td><strong>MONOARTICULAR ONSET</strong></td>
<td><strong>POLYARTICULAR ONSET</strong></td>
</tr>
<tr>
<td>Patient is usually irritable, listless, anorectic, and suffers from weight loss.</td>
<td>May have generalized symptoms.</td>
<td>Patient is usually listless, anorectic, and underweight.</td>
</tr>
<tr>
<td><strong>Laboratory</strong></td>
<td>CBC and ESR may be normal.</td>
<td>WBC may be elevated, but is rarely higher than 20,000. ESR is elevated, usually corresponding roughly to the intensity of the arthritis.</td>
</tr>
<tr>
<td>Neutrophilic leukocytosis with WBC of 15,000 to 50,000/mm³</td>
<td>X-ray examination may reveal accelerated maturation or early closure of epiphyses, periosteal proliferation, metaphyscal overgrowth of long bones, especially about the knee.</td>
<td></td>
</tr>
<tr>
<td>There may be a moderate normocytic, normochromic anemia</td>
<td>Synovial fluid aspiration reveals clear to opalescent fluid with good to poor mucin clot. 15,000 to 25,000 WBC/mm³ with 50 to 90% neutrophils.</td>
<td></td>
</tr>
<tr>
<td>The ESR is usually elevated.</td>
<td>Glucose of synovial fluid is about 25 mg/100 ml less than the serum glucose.</td>
<td></td>
</tr>
<tr>
<td>Must be differentiated from other connective tissue diseases by absence of antinuclear antibody, difference in the nature of the rash, and age of onset (peak onset of JRA is 1 to 3 years of age, while SLE is rare in children under 5 years of age).</td>
<td>Must be differentiated from traumatic injury and from infectious arthritis by synovial fluid analysis. (Onset of symptoms commonly follows trauma.)</td>
<td>Must be differentiated from rheumatic fever by difference in fever pattern (fever of rheumatic fever is remittent or sustained), by x-ray findings, and by arthritis persisting longer than a few weeks.</td>
</tr>
<tr>
<td>Differentiation from the arthritis sometimes accompanying rubella is made by detection of an increase in the HI antibody to rubella in acute and convalescent sera. The synovial fluid of rubella arthritis has a predominance of mononuclear cells.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


**ATAXIA**

**Ataxia, Muscle Weakness, Extrapyramidal Disorders**

The following tables cover a broad range of neuromuscular disorders that comprise an area of difficult differential diagnosis for the clinician. A child with ataxia, muscle weakness, or extrapyramidal signs and symptoms should be
examined with particular care because identification of the clinical disorder can often indicate the site of the lesion.

**Differential Diagnosis of Chronic Progressive Ataxia**

<table>
<thead>
<tr>
<th>CLINICAL DISORDER</th>
<th>PRECEDING HISTORY</th>
<th>USUAL YEAR OF ONSET IN CHILDREN</th>
<th>EXAMINATION</th>
<th>USUAL LABORATORY EXAMINATION</th>
<th>USUAL PROGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arnold-Chiari malformation</td>
<td>Headache, dysphagia</td>
<td></td>
<td>Palatal and tongue weakness, pyramidal signs, ataxia</td>
<td>May have hydrocephalus, spina bifida</td>
<td>Slowly progressive; stationary after surgery</td>
</tr>
<tr>
<td>Hereditary spinoocerebellar ataxia</td>
<td>Stumbling, dizziness, familial incidence</td>
<td>7 10</td>
<td>Ataxia, loss of position sense, extensor plantar responses, kyphoscoliosis, pes cavus</td>
<td>Frequent associated ECG changes</td>
<td>Progressive, with death usually by 30 years of age</td>
</tr>
<tr>
<td>Abetalipoproteinemia</td>
<td>Fatty diarrhea at 6 weeks to 2 years of age</td>
<td>2 17</td>
<td>Cerebellar ataxia, posterior column signs, retinitis pigmentosa, scoliosis, pes cavus</td>
<td>Acanthoeytosis, lack of β-lipoprotein in serum</td>
<td>Slowly progressive</td>
</tr>
<tr>
<td>Dentate cerebellar ataxia</td>
<td>Myoclonus, convulsions</td>
<td>7 17</td>
<td>Ataxia with severe intention tremor</td>
<td></td>
<td>Slowly progressive</td>
</tr>
<tr>
<td>Hereditary cerebellar ataxia</td>
<td>Familial incidence</td>
<td>3 17</td>
<td>Ataxia, optic atrophy, occasionally associated posterior column and pyramidal tract signs</td>
<td>Pneumocerehalogram: small cerebellar folia</td>
<td>Slowly progressive</td>
</tr>
<tr>
<td>Ataxia telangiectasia</td>
<td>Recurrent sinusopulmonary infections in two-thirds of cases; familial incidence</td>
<td>1 3</td>
<td>Oculocutaneous telangiectasia at 4 to 6 years; ataxia, choreoathetosis, dysarthria</td>
<td>Chest roentgenogram: bronchiectasis, absence of IgA in serum</td>
<td>Death before 25 years of age</td>
</tr>
<tr>
<td>Cerebellar tumors</td>
<td>Headache, vomiting</td>
<td></td>
<td>Papilledema, ataxia, nystagmus</td>
<td>Skull roentgenogram: separation of sutures</td>
<td>Progressive until operated</td>
</tr>
<tr>
<td>Heredopathia atactica polynueurtiformis</td>
<td>Anorexia, failing vision, unsteady, familial incidence</td>
<td>4 7</td>
<td>Retinitis pigmentosa, ataxia, deafness, polyneuropathy, ophthalmoplegia, dysthesis</td>
<td>Elevated phytic acid in blood, increased spinal fluid protein</td>
<td>Slowly progressive with death</td>
</tr>
<tr>
<td>Multiple sclerosis</td>
<td>Preceding neurologic symptoms</td>
<td>14 17</td>
<td>Optic neuritis: brain stem, cerebellar, pyramidal, or sensory signs</td>
<td>Spinal fluid may reveal increased cells, protein, or γ-globin</td>
<td>Exacerbations and remissions</td>
</tr>
<tr>
<td>Spinal cord tumor</td>
<td>May have numbness or bladder disorder</td>
<td></td>
<td>Ataxia with weakness or sensory loss</td>
<td>Defect on myelography</td>
<td>Progressive until operated</td>
</tr>
</tbody>
</table>
### Differential Diagnosis of Acute Ataxia

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Receding History</th>
<th>Examination</th>
<th>Laboratory Examination</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute cerebellar ataxia</td>
<td>Half have had a prodromal systemic illness, occasionally exanthems</td>
<td>Cerebellar ataxia</td>
<td>Spinal fluid usually r</td>
<td>Recovery</td>
</tr>
<tr>
<td>Dilantin intoxication</td>
<td>Convulsions treated with phenytoin</td>
<td>Cerebellar ataxia, nystagmus</td>
<td>High serum phenytoin level</td>
<td>Recovery</td>
</tr>
<tr>
<td>Cerebellar tumor or abscess</td>
<td>Headache, vomiting</td>
<td>Papilledema, ataxia, nystagmus</td>
<td>Separation of cranial sutures</td>
<td>Progressive until operated</td>
</tr>
<tr>
<td>Hartnup syndrome</td>
<td>Skin eruptions on exposure to sun; familial incidence</td>
<td>Skin lesions, ataxia, nystagmus, mental disturbances</td>
<td>Aminoaciduria, increased indole in urine</td>
<td>Recurrent ataxia</td>
</tr>
<tr>
<td>Multiple sclerosis</td>
<td>Preceding neurologic symptoms</td>
<td>Optic neuritis; brain stem, cerebellar, pyramidal or sensory signs</td>
<td>Spinal fluid may reveal increased cells, protein or γ-globulin</td>
<td>Exacerbations and remissions</td>
</tr>
<tr>
<td>Encephalitides</td>
<td>Headache, stiff neck, fever</td>
<td>Cerebral and brain stem signs; also may have ataxia</td>
<td>Spinal fluid: lymphocytosis; possible virus isolation or rise in antibody titer</td>
<td>May be fatal, or slow recovery with or without residual</td>
</tr>
<tr>
<td>Spinal cord tumor</td>
<td>May have numbness or bladder disorder</td>
<td>Ataxia with weakness or sensory loss</td>
<td>Defect on myelography</td>
<td>Progressive until operated</td>
</tr>
<tr>
<td>Infectious polyneuropathy</td>
<td>Half have a prodromal systemic illness</td>
<td>Ataxia with motor and sensory loss</td>
<td>Spinal fluid: normal cells, increased protein</td>
<td>May be fatal, but recovery usually complete</td>
</tr>
</tbody>
</table>

### Differential Diagnosis of Disorders of Muscle, Anterior Horn Cell, and Peripheral Nerves

<table>
<thead>
<tr>
<th>Clinical and Laboratory Features</th>
<th>Muscle</th>
<th>Anterior Horn Cell</th>
<th>Peripheral Nerves</th>
</tr>
</thead>
<tbody>
<tr>
<td>Site of predisposition</td>
<td>Usually proximal and axial musculature</td>
<td>Proximal and/or distal extremity musculature</td>
<td>Usually distal extremity musculature</td>
</tr>
<tr>
<td>Deep tendon reflexes</td>
<td>Preserved until late in course</td>
<td>Reduced to absent early in course</td>
<td>Reduced to absent early in course</td>
</tr>
<tr>
<td>Sensation deficit</td>
<td>Rarely observed</td>
<td>Not observed</td>
<td>Usually present</td>
</tr>
<tr>
<td>Fasciculations</td>
<td>Usually absent</td>
<td>Frequently present</td>
<td>Occasionally present</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
### Differential Diagnosis of Disorders of Muscle, Anterior Horn Cell, and Peripheral Nerves (Cont.)

<table>
<thead>
<tr>
<th>Clinical and Laboratory Features</th>
<th>Muscle</th>
<th>Anterior Horn Cell</th>
<th>Peripheral Nerves</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF protein</td>
<td>Normal</td>
<td>Normal or elevated</td>
<td>Elevated or normal</td>
</tr>
<tr>
<td>Electromyography</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Interference pattern</td>
<td>Normal until late in disease</td>
<td>Reduced</td>
<td>Reduced</td>
</tr>
<tr>
<td>Fibrillation potentials</td>
<td>Not usually present</td>
<td>Usually present</td>
<td>Present</td>
</tr>
<tr>
<td>Action potentials</td>
<td>Short duration</td>
<td>Prolonged with occasional giant potentials</td>
<td>Prolonged with normal or polyphasic potentials</td>
</tr>
<tr>
<td>Evoked sensory and mixed nerve potentials</td>
<td>Normal</td>
<td>Normal</td>
<td>Absent, diminished amplitude, or prolonged conduction time</td>
</tr>
</tbody>
</table>

### Differential Diagnosis of Extrapyramidal Disorders

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Familial</th>
<th>Signs</th>
<th>Associated Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hepatolenticular degeneration</td>
<td>Autosomal recessive</td>
<td>Rigidity, tremor, dystonia, dementia, corneal ring, jaundice</td>
<td>Increased urinary and hepatic copper, low serum ceruloplasmin</td>
</tr>
<tr>
<td>Juvenile parkinsonism</td>
<td>Rarely</td>
<td>Resting tremor, rigidity, bradykinesia</td>
<td>Decreased dopamine level in substantia nigra</td>
</tr>
<tr>
<td>Kernicterus</td>
<td>No</td>
<td>Athetosis, deafness, occasional intellectual impairment</td>
<td>Neonatal hyperbilirubinemia</td>
</tr>
<tr>
<td>Huntington's disease</td>
<td>Autosomal dominant</td>
<td>Rigidity, chorea, convulsions, dementia</td>
<td></td>
</tr>
<tr>
<td>Torsion dystonia</td>
<td>Autosomal dominant or recessive</td>
<td>Dystonia, involuntary movements, normal intellect</td>
<td></td>
</tr>
<tr>
<td>Chorea minor (Sydenham's)</td>
<td>No</td>
<td>Involuntary choreic movements, possible carditis</td>
<td>Group A streptococcal infections</td>
</tr>
<tr>
<td>Absence of hypoxanthine-</td>
<td>X-linked recessive</td>
<td>Choreoathetosis, mental retardation, self-mutilation</td>
<td>Increased urinary and blood uric acid</td>
</tr>
<tr>
<td>guanine phosphoribosyl</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>transferase (Lesch-Nyhan</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>syndrome)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

B

BACK PAIN

Common Causes

Mechanical derangement (muscle strain or poor posture)
Scheuermann's kyphosis

Scoliosis
Spondylolysis/spondylolisthesis

Uncommon Causes

Disc space infection (discitis)
Rheumatic disorders
Sacroiliac joint infections
Spina bifida occulta

Spinal cord tumors (lipomas, teratomas)
Vertebral osteomyelitis

Rare Causes

Aneurysmal bone cyst
Aseptic necrosis of vertebrae
Benign osteoblastoma
Eosinophilic granuloma of vertebrae
Hemangioma of bone
Herniated nucleus pulposus
Malignancy involving bone
(neuroblastoma, leukemia)

Osteomalacia of the spine
Paraspinal tumor or infection
Secondary hyperparathyroidism
Tuberculosis of the spine
Vertebral osteoid osteoma

A Pain in the Back

The differential diagnosis of back pain in infants and children may not be as lengthy as that of chest pain, but the possibilities are equally perplexing. Unlike back pain in adults, which frequently defies identification of an etiology, nearly 75% of children with back pain have a definable cause. Because the presentation can be variable, an understanding of the potential etiologies and a rational approach to the work-up can save time and money in needless examinations and tests.

When a child presents with sudden-onset refusal to walk (or sit), irritability, elevated temperature, abdominal pain and/or nausea, vomiting, and anorexia—and laboratory studies consistent with inflammation—the physician must immediately differentiate between infectious and noninfectious etiologies. Although the distinction between the generally benign entity of discitis and...
30—Back Pain

Inevitably destructive osteomyelitis is relatively simple (see table), one must also consider meningitis, appendicitis, peritonitis, septic arthritis, and urinary tract infections. With the help of the flow chart and tables below, differentiation will be simpler and the oftentimes delayed diagnosis of discitis will not evade the pediatrician.
## Discitis vs. Intervertebral Infection

<table>
<thead>
<tr>
<th></th>
<th>DISCITIS</th>
<th>INFECTIOUS VERTEBRAL OSTEOMYELITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age</td>
<td>4 yr</td>
<td>9.8 yr</td>
</tr>
<tr>
<td>Sex (M:F)</td>
<td>0.6:1</td>
<td>2.4:1</td>
</tr>
<tr>
<td>Complaint cited most often</td>
<td>Gait refusal</td>
<td>Severe pain, even at rest</td>
</tr>
<tr>
<td>History of trauma</td>
<td>20%</td>
<td>4%</td>
</tr>
<tr>
<td>Vertebral site</td>
<td>Lumbar</td>
<td>Thoracic or lumbar</td>
</tr>
<tr>
<td>Mean maximum temperature</td>
<td>&lt; 100°F</td>
<td>&gt; 101°F</td>
</tr>
<tr>
<td>Mean maximum WBC count</td>
<td>&lt; 8,000/µL</td>
<td>&gt; 15,000/µL</td>
</tr>
<tr>
<td>Mean maximum ESR</td>
<td>&lt; 35 mm</td>
<td>&gt; 50 mm</td>
</tr>
</tbody>
</table>

**Plain radiographs**
- At outset: Normal
- At 4 wk: Disc space narrowing, Bony destruction
- Tc-99m bone scan at outset: Positive, Positive
- Gallium scan at outset: Negative, Positive
- Indium-labeled WBC scan at outset: Negative, Positive

**Blood or local tissue culture**
- 2% positive, 60% positive

**Fate of disc**
- Regenerates; often narrow, Destroyed

**Fate of vertebral body**
- Unaffected, Destroyed

**Fate of neural arch**
- Unaffected, Often destroyed

**Fatality**
- 0%, 6%

**Chronic persistent illness**
- 0%, 8%

**Clinical duration**
- 2-5 wk; always < 12 wk Many months

## Vertebral Disorders in Children

<table>
<thead>
<tr>
<th>ENTITY</th>
<th>USUAL SITE</th>
<th>ETIOLOGY</th>
<th>PEAK AGE</th>
<th>BEST TEST</th>
<th>LABS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infection</td>
<td>Thoracic or lumbar spine</td>
<td>Staphylococcus, TB or abscess formation</td>
<td>8 yr</td>
<td>Plain x-ray; look for vertebral destruction</td>
<td>Blood and tissue culture for WBC, ↑ ESR and ↑ platelets</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tumors</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Malignant</td>
<td>Low back, pelvis</td>
<td>Pelvic invasion with marrow tumors or chondrosarcoma</td>
<td>Adolescence</td>
<td>Plain x-ray; look for bony lesion w/soft tissue mass</td>
<td>Histology</td>
</tr>
<tr>
<td>Nonmalignant</td>
<td>Neural arches</td>
<td>Osteoblastoma</td>
<td>Adolescence</td>
<td>Plain x-ray</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>Posterior</td>
<td>Osteoid</td>
<td>Adolescence</td>
<td>Look for sclerotic nidus w/lucent halo</td>
<td></td>
</tr>
</tbody>
</table>

*Table continued on next page.*
Vertebral Disorders in Children (Cont.)

<table>
<thead>
<tr>
<th>ENTITY</th>
<th>USUAL SITE</th>
<th>ETIOLOGY</th>
<th>PEAK AGE</th>
<th>BEST TEST</th>
<th>LABS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discitis</td>
<td>Lumbar anterior</td>
<td>Avascular necrosis of epiphyseal end plates and discs</td>
<td>4 yr</td>
<td>Early: Tc-99m uptake. After 4 wk: disc-space narrowing on plain x-ray</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>aspect</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spondylolysis</td>
<td>L 4, L 5</td>
<td>Traumatic defect in posterior aspect of pars articularis</td>
<td>Early teens</td>
<td>Oblique plain x-ray</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>vertebrae</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spondylolisthesis</td>
<td>L 5, S 1</td>
<td>Forward slippage (L 5 moves anterior to S-1) in pt w spondylolysis</td>
<td>During growth spurt</td>
<td>Standing lateral plain x-ray</td>
<td>None</td>
</tr>
<tr>
<td></td>
<td>vertebrae</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Scheuermann's kyphosis</td>
<td>Lower thoracic vertebral</td>
<td>Osteochondrosis w/herniation of disc into vertebral bodies; w/anterior narrowing of disc space; disc walled off in vertebra (Schmorl's node)</td>
<td>Adolescence</td>
<td>Standing lateral plain x-ray shows Schmorl's node</td>
<td>Node</td>
</tr>
</tbody>
</table>


BACTERIAL ENDOCARDITIS

Extracardiac Manifestations of Bacterial Endocarditis

The patient with bacterial endocarditis presents both a diagnostic and a therapeutic challenge. The myriad manifestations of the disease result from the hemodynamic, embolic, and immunologic sequelae of the endovascular infection.

The following review of the more common extracardiac manifestations may serve as an aid in diagnosis and management of this disease.

Extracardiac Manifestations of Bacterial Endocarditis

<table>
<thead>
<tr>
<th>MANIFESTATION</th>
<th>COMMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Renal</td>
<td></td>
</tr>
<tr>
<td>2. Occasionally azotemia</td>
<td></td>
</tr>
<tr>
<td>3. Abnormalities usually resolve with effective antimicrobial therapy</td>
<td></td>
</tr>
</tbody>
</table>

Table continued on next page.
Bacterial Endocarditis—33

Extracardiac Manifestations of Bacterial Endocarditis (Cont.)

<table>
<thead>
<tr>
<th>MANIFESTATION</th>
<th>COMMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>II. Neurologic</strong></td>
<td></td>
</tr>
<tr>
<td>1. Major neurologic complications are:</td>
<td></td>
</tr>
<tr>
<td>a. Cerebral infarction in region of middle cerebral arteries secondary to emboli (most common neurologic complication)</td>
<td></td>
</tr>
<tr>
<td>b. Meningeal signs and symptoms</td>
<td></td>
</tr>
<tr>
<td>c. Seizures</td>
<td></td>
</tr>
<tr>
<td>d. Intracranial hemorrhage</td>
<td></td>
</tr>
<tr>
<td>e. Large macroscopic brain abscesses are uncommon</td>
<td></td>
</tr>
<tr>
<td>f. Microscopic brain abscesses are common and reflect multiple microemboli</td>
<td></td>
</tr>
<tr>
<td><strong>Note:</strong> Neurologic complications occur in 25-40% of patients with bacterial endocarditis. The mortality of patients with neurologic complications is &gt; 50%. Embolic phenomenon are usually seen in endocarditis due to S. aureus, Pneumococcus, Enterobacteriaceae, and anaerobic streptococci. Mitral valve endocarditis produces major cerebral emboli more frequently than aortic valve endocarditis. Mycotic aneurysms occur more frequently in the early course of acute endocarditis than late in the course of subacute endocarditis. CSF exam tends to reflect the nature of the infecting organisms; i.e. virulent organisms are more likely to produce meningitis with a purulent CSF than are less virulent organisms, which are likely to produce a sterile “aseptic” CSF.</td>
<td></td>
</tr>
</tbody>
</table>

**III. Musculoskeletal**

1. Arthralgia—usually in shoulder, knee, hip
2. True synovitis
   a. Ankle, knee, wrist most frequent
   b. Usually sterile
   c. Biopsy shows acute inflammatory changes
3. Low back pain
   a. Often severe
   b. Often demonstrates spinal tenderness and decreased range of motion
   c. X-rays usually normal
   d. Usually not secondary to disc space infection
4. Myalgias—often localized to thighs and calves
5. Miscellaneous
   a. Clubbing of the digits
   b. Hypertrophic osteoarthropathy
   c. Avascular necrosis of hip

**IV. Skin**

1. Petechiae
2. Osler’s nodes
3. Janeway lesions
4. Periungual erythema
5. Subungual “splinter” hemorrhages

Table continued on next page.
Recognition of Basal Skull Fractures

Basal fractures through the floor of the skull are usually linear. They are difficult to recognize in x-ray studies and are diagnosed clinically. The clues to the diagnosis include:

1. Bleeding from nose, eyes, or ears, or discoloration in the mastoid area (ecchymosis behind the ear, or Battle's sign). The “raccoon eye” may be seen, with a hematoma in the upper lid.
2. Blood and CSF behind the eardrum, causing a bulging of the membrane. Otorrhea occurs when the tympanic membrane is ruptured.
3. Cerebrospinal rhinorrhea. Some believe that testing the nasal discharge for the presence of glucose is an indication of CSF leak. However, approximately 75 to 90% of normal children will give a positive glucose oxidase test strip in their nasal secretions, which makes the use of such a test for CSF leak valueless.
4. Cranial nerve palsies, involving cranial nerves I, III, and VIII.
5. Appearance of “sinusitis.”

A basal skull fracture can lead to meningitis by spread of organisms from the nose or ear, and prophylactic use of penicillins is justifiable.

**BEHAVIOR**

**Behavioral Concerns of Parents**

If you were to provide a behavioral checklist to middle-class parents of children 1.5 to 6 years of age, which behaviors would they note as being of greatest concern to them? Listed below are these behaviors listed in order of frequency.

Are you prepared to discuss these topics with parents?

<table>
<thead>
<tr>
<th>BEHAVIOR</th>
<th>PERCENTAGE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stubbornness</td>
<td>29%</td>
</tr>
<tr>
<td>Poor appetite</td>
<td>23%</td>
</tr>
<tr>
<td>Getting child to sleep</td>
<td>22%</td>
</tr>
<tr>
<td>Effects of both parents working</td>
<td>22%</td>
</tr>
<tr>
<td>Day care</td>
<td>19%</td>
</tr>
<tr>
<td>Restless sleep</td>
<td>18%</td>
</tr>
<tr>
<td>Temper tantrums</td>
<td>16%</td>
</tr>
<tr>
<td>Feelings hurt too easily</td>
<td>16%</td>
</tr>
<tr>
<td>Problems at meals</td>
<td>15%</td>
</tr>
<tr>
<td>“High strung”/easily upset</td>
<td>15%</td>
</tr>
<tr>
<td>Wanting too much attention</td>
<td>12%</td>
</tr>
<tr>
<td>Disobedient</td>
<td>12%</td>
</tr>
<tr>
<td>Hyperactive</td>
<td>12%</td>
</tr>
<tr>
<td>New sibling</td>
<td>11%</td>
</tr>
<tr>
<td>Moving</td>
<td>10%</td>
</tr>
</tbody>
</table>


**BLADDER**

**Bladder Capacity in Children**

Bladder capacity correlates linearly with age from birth to the 11th year. *The bladder capacity in ounces equals age in years plus 2, with a standard deviation of 2 ounces.* Knowledge of the functional bladder capacity, with a detailed history, may suggest a diagnosis of large or small bladder capacity. Children with infrequent voiding tend to have larger bladder capacities, whereas those with frequency or enuresis have smaller than predicted capacities.

Neonatal Blistering Disorders

A number of disorders can give rise to neonatal blisters, ranging from the benign suction blister (which is presumably caused by thumb, finger, or distal forearm sucking in utero) to epidermolysis bullosa, a heterogeneous group of inherited skin disorders notable for marked skin fragility. Listed below is the differential diagnosis for blistering disorders of the neonate.

### Differential Diagnosis for Blistering Disorders

<table>
<thead>
<tr>
<th>CONDITION</th>
<th>ONSET</th>
<th>PATHOLOGIC FEATURES</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Epidermolysis bullosa (EB)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1. EB simplex (usually autosomal</td>
<td>Usually at</td>
<td>Intraepidermal blisters</td>
<td>Trauma causes blisters; patients have mild involvement without scarring in the absence of infection. Mucous membranes are usually spared. Teeth develop normally. Typically a benign course with normal life-span and no significant functional impairment.</td>
</tr>
<tr>
<td>dominant inheritance)</td>
<td>birth</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Junctional EB (autosomal</td>
<td>At birth</td>
<td>Blistering occurs in the lamina lucida between the</td>
<td>Trauma causes extensive blistering on the skin and any mucosal membrane. In one type (junctional EB letalis of Herlitz-Pearson), GI involvement is frequent, leading to perforation, sepsis, and death in early infancy. In other subtypes, patients follow a more indolent course and survive to adulthood, although nonhealing cutaneous wounds yield significant morbidity.</td>
</tr>
<tr>
<td>recessive inheritance)</td>
<td></td>
<td>epidermis and dermis</td>
<td></td>
</tr>
<tr>
<td>3. Dystrophic EB (there exist</td>
<td>At birth</td>
<td>Blistering occurs in the dermis, below the lamina densa</td>
<td>Lesions heal with milia formation and marked scarring that can lead to crippling deformities.</td>
</tr>
<tr>
<td>both autosomal recessive and</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>autosomal dominant forms)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bullosous congenital ichthyosiform erythroderma (autosomal dominant)</td>
<td>At birth</td>
<td>Vacuolization of cells of granular and upper spinous layers</td>
<td>Red scaly skin; secondary bacterial infection; thick, grayish brown scales after age 3 mo.</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
Differential Diagnosis for Blistering Disorders (Cont.)

<table>
<thead>
<tr>
<th>CONDITION</th>
<th>ONSET</th>
<th>PATHOLOGIC FEATURES</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Congenital herpes simplex virus infection</td>
<td>In first 20 days; mean = 6 days</td>
<td>Intraepidermal blisters with multiple thin-walled vesicles on an erythematous base</td>
<td>Blisters and bullae; positive Tzanck smear and viral culture; fever, poor feeding, hypothermia, and lethargy.</td>
</tr>
<tr>
<td>Aplasia cutis congenita (usually autosomal dominant, but autosomal recessive also reported)</td>
<td>At birth</td>
<td>Ulcer down to subcutaneous tissue</td>
<td>Absence of skin on scalp; similar cutaneous defects may be present elsewhere; limb abnormalities; some cases associated with epidermolysis bullosa.</td>
</tr>
<tr>
<td>Staphylococcal scalded skin syndrome</td>
<td>2-30 days</td>
<td>Blisters below or within granular layer</td>
<td>Abrupt onset of erythema, followed by blistering and exfoliation; responds to antibiotics.</td>
</tr>
<tr>
<td>Suction blisters</td>
<td>At birth</td>
<td></td>
<td>One or two blisters on thumb, finger, radial aspect of forearm, presumably due to sucking in utero; spontaneous resolution.</td>
</tr>
</tbody>
</table>


Bullous Eruptions in the Newborn

Eruptions of vesicles (raised, fluid-filled lesions < 1 cm) and bullae (raised, fluid-filled lesions > 1 cm) in the neonatal period are due to a variety of mostly unrelated conditions, with different treatments and prognoses in each category. The following table lists the principal criteria for the differential diagnosis of bullous eruptions in the nursery.

Principal Criteria for Differential Diagnosis of Bullous Eruptions

<table>
<thead>
<tr>
<th>DISEASE OR CONDITION</th>
<th>CHARACTER OF LESIONS</th>
<th>DISTRIBUTION</th>
<th>MUCOSAL IN Volvement</th>
<th>OTHER ECTODERMAL DEFECTS</th>
<th>SCARRING</th>
<th>COURSE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epidermolysis bullosa</td>
<td>Clear blisters; sometimes hemorrhagic noninflammatory base</td>
<td>Sites of trauma or friction</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes or no</td>
<td>Chronic or fatal</td>
</tr>
<tr>
<td>Bullous impetigo</td>
<td>Blister, clear, opaque, purulent</td>
<td>General, particularly flexures</td>
<td>Possible</td>
<td>No</td>
<td>Yes</td>
<td>Short</td>
</tr>
<tr>
<td>Congenital syphilis</td>
<td>Bullae and maculopapules</td>
<td>Palms, soles, trunk, and limbs</td>
<td>Yes</td>
<td>Yes</td>
<td>No (other than rhagades)</td>
<td>Short</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Principal Criteria for Differential Diagnosis of Bullous Eruptions (Cont.)

<table>
<thead>
<tr>
<th>Disease or Condition</th>
<th>Character of Lesions</th>
<th>Distribution</th>
<th>Mucosal Involvement</th>
<th>Other Cutaneous Effects</th>
<th>Scarring</th>
<th>Course</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dermatitis herpetiformis</td>
<td>Vesicles and bullae in crops; also urticariiform lesions</td>
<td>In infants, face and limbs chiefly involved</td>
<td>Sometimes</td>
<td>No</td>
<td>Minimal in long-standing cases</td>
<td>One-third curable. Chronic or recurrent</td>
</tr>
<tr>
<td>Burns</td>
<td>Erythema, bullae, desquamation</td>
<td>Anywhere</td>
<td>No</td>
<td>No</td>
<td>Yes, if deep</td>
<td>Depends on type, depth, and therapy</td>
</tr>
<tr>
<td>Congenital porphyria</td>
<td>Red urine, photosensitivity of skin, erythema, bullae</td>
<td>Areas exposed to sunlight</td>
<td>No</td>
<td>Pigmented teeth</td>
<td>Pigmented scars</td>
<td>Chronic</td>
</tr>
<tr>
<td>Erythema multiforme bullosa</td>
<td>Dusky red circinate plaques, papules, bullae</td>
<td>Trunk, limbs, face</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
<td>Short or recurrent</td>
</tr>
<tr>
<td>Dermatitis medica-mentosa</td>
<td>May be vesicular</td>
<td>No particular site</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Short</td>
</tr>
<tr>
<td>Papular urticaria</td>
<td>Papules, bullae, vesicles, pustules</td>
<td>Trunk only or limbs</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Short or recurrent</td>
</tr>
<tr>
<td>Chickenpox</td>
<td>Vesicles, pustules</td>
<td>Trunk, face, limbs</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Short</td>
</tr>
<tr>
<td>Smallpox</td>
<td>Vesicles, pustules</td>
<td>Limbs, trunk, face</td>
<td>Yes</td>
<td>No</td>
<td>Yes</td>
<td>Short</td>
</tr>
<tr>
<td>Kaposi's varicelliform eruption</td>
<td>Vesicles, pustules</td>
<td>Exposed parts</td>
<td>No</td>
<td>Pre-existing skin disease of infantile eczema or Besnier's prurigo</td>
<td>No</td>
<td>May be fatal</td>
</tr>
<tr>
<td>Herpes zoster</td>
<td>Vesicles</td>
<td>Classical girdle</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Short</td>
</tr>
<tr>
<td>Bullous erysipelas</td>
<td>Raised tender erythema, bullae</td>
<td>Periungual, bilical, limbs, face, trunk</td>
<td>Rarely</td>
<td>Nil</td>
<td>Nil</td>
<td>Short with therapy</td>
</tr>
<tr>
<td>Benign familial pemphigus (Hailey's disease)</td>
<td>Vesicles and bullae</td>
<td>Anywhere</td>
<td>No</td>
<td>No</td>
<td>Nil</td>
<td>Benign chronic</td>
</tr>
</tbody>
</table>

Table continued on next page.
Blood Cultures—39

Principal Criteria for Differential Diagnosis of Bullous Eruptions (Cont.)

<table>
<thead>
<tr>
<th>DISEASE OR CONDITION</th>
<th>CHARACTERISTICS OF LESIONS</th>
<th>DISTRIBUTION</th>
<th>MUCOSAL INVOLVEMENT</th>
<th>OTHER EPIDERMAL DEFECTS</th>
<th>SCARRING</th>
<th>COURSE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Contact dermatitis</td>
<td>Often vesicles and bullae</td>
<td>Anywhere</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Short</td>
</tr>
<tr>
<td>Phytophotodermatitis</td>
<td>Vesicles and bullae</td>
<td>Areas exposed to sunlight</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Short</td>
</tr>
<tr>
<td>Acrodermatitis enteropathica</td>
<td>Crops scaling, vesiculo-bullous</td>
<td>Near orifices, around eyes, elbows, knees, hands, feet</td>
<td>Yes</td>
<td>Hair scanty</td>
<td>No</td>
<td>May be fatal</td>
</tr>
</tbody>
</table>


BLOOD CULTURES

Changing the Needle When Inoculating Blood Cultures: A No-Benefit and High-Risk Procedure

For some time now, in response to the high prevalence of HIV and hepatitis B infections, the U.S. Centers for Disease Control has recommended that needles should never be recapped in order to prevent unnecessary needle-stick injuries. Yet, many phlebotomists and physicians routinely recap and change needles before blood-culture inoculation. A group of pediatricians and pathologists at the University of Virginia were concerned about this clinical paradigm and designed a study to compare the extrinsic contamination rate in blood cultures when the needle was and was not changed.

The investigators had 108 medical students obtain 182 blood specimens from each other using standard methods. Each blood sample was inoculated into two culture bottles. The first bottle was inoculated with the needle used for phlebotomy, and the second was inoculated after a needle change. Of the 182 culture bottles, 4 (2.2%) were contaminated when the needle was not changed and 1 (0.6%) was contaminated when the needle was changed. This small difference was found to be statistically insignificant, and the possibility of having failed to detect a 5% difference in contamination rate was small.

The conclusion of this study, therefore, was that the risk of needle-stick injury incurred by changing the needle before inoculation of blood culture bottles seems to be unjustified.

BODY TEMPERATURE

A Comparison of Rectal, Axillary, and Inguinal Temperatures in Full-Term Newborn Infants

What is the mean temperature in the various sites commonly employed for the measurement of a newborn's temperature? In the study described below, rectal thermometers were inserted 2.5 cm into the rectum. Inguinal temperatures were measured by abducting the infant's leg, locating the femoral pulse, placing the bulb of the thermometer lateral to the pulse site, and adducting the leg to create a seal. Readings were recorded every 30 seconds after placement and were discontinued when no change occurred for 90 seconds.

<table>
<thead>
<tr>
<th>SITE</th>
<th>MEAN TEMPERATURE (°F)</th>
<th>RANGE (°F)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Rectal</td>
<td>98.7</td>
<td>97.6 - 100.4</td>
</tr>
<tr>
<td>Axillary</td>
<td>98.5</td>
<td>97.2 - 100.2</td>
</tr>
<tr>
<td>Inguinal</td>
<td>97.9</td>
<td>96.4 - 99.2</td>
</tr>
</tbody>
</table>


BONE MARROW EXAMINATIONS

Is There a Role for Bone Marrow Examinations in the Child with Prolonged Fever?

When evaluating a child with prolonged fever, the question of whether or not to perform a bone marrow examination is often posed. Bone marrow examinations have been shown to be of great use for aid in the diagnosis of malignancy, but their usefulness for detecting occult infections has been the source of a long and, excuse the pun, heated debate. Recently, a group of physicians at the Texas Children's Hospital reviewed 414 cases of children with prolonged fevers in order to assess this situation.

In their retrospective series, noninfectious causes of prolonged fever were revealed by the bone marrow examination in 34 (8.2%) of the 414 study patients (e.g., malignant conditions such as acute leukemia, both newly diagnosed and relapsed, lymphoma, solid tumors, and chronic myelocytic leukemia, in addition to nonmalignant illness such as virus-associated hemophagocytic syndrome, histiocytosis, and hypoplastic anemia). In the majority of these cases, a diagnosis of myelopthisis was clinically suspected before the bone marrow was obtained.

An infectious etiology of prolonged fever was uncovered in 15 (3.6%) of the febrile episodes. It should be noted that only one patient of the 414 children studied had a positive marrow culture (for Salmonella, group D) without concurrent positive cultures from any other source. In patients who were immunocompetent, the yield of positive marrow cultures was rather low (1.9%), whereas in immunocompromised children (particularly those with AIDS) the yield was 8.7%.
With these data in mind, the following conclusions were reached.

1. Bone marrow examination is indicated in the child with prolonged fever and clinical or laboratory evidence consistent with malignancy.
2. Bone marrow examination may be helpful in ascertaining the diagnosis of opportunistic infection in the febrile, immunocompromised patient, especially a child with AIDS.
3. Bone marrow examination in the child with prolonged fever but no findings suggestive of malignancy or immunodeficiency is probably not warranted as a means of detecting occult infection.


**BREAST MASSES**

**Breast Masses and Lesions in the Infant, Child, and Adolescent**

Lesions and masses of the breast generate a great deal of concern upon discovery. This holds particularly true for such lesions among the pediatric age group, despite the fact that the overwhelming majority of lesions in this population are benign. There do, however, exist some lesions that require immediate attention (e.g., mastitis in the newborn or developing breast and the rarely occurring malignancy). Clinicians, therefore, need to be able to recognize and assess these lesions in order to offer appropriate treatment and reassurance to their patients. As each age group seen in pediatrics (i.e., neonatal, prepubertal, and adolescent) has its own set of breast masses or lesions, we present the differential diagnosis in a chronologic manner:

**Infancy**

1. **Neonatal hypertrophy** presents as a palpable, tender mass with or without a milky nipple secretion ("witch's milk"), which is most likely due to a low prolactin level in a premature infant that rises postnatally to that of a normal-term infant. The milky discharge should abate within 4 to 6 weeks of life, although it may take up to 1 year for the breast enlargement to recede.
2. **Mastitis and resultant abscess** present as a tender, erythematous breast mass, usually with fever. This is, perhaps, the only breast lesion in the pediatric population that requires immediate intervention. Mastitis and resultant abscesses tend to occur in the infant aged 2 weeks to 6 months, but can occur at any time. Both gram-positive organisms (e.g., *Staphylococcus aureus*) and gram negative organisms (e.g., *E. coli*) are common culprits; antibiotic therapy, therefore, should be broad in its spectrum. Warm compresses are also useful. Septicemia is a concern in the young infant. Increased pressure and inflammation from the infection itself and surgical drainage by an overzealous surgeon can result in future deformity of the breast in an infant or prepubertal girl.
Breast Masses

3. **Polythelia** is the presence of one or more supernumerary nipples along the "milk-line," which extends from the axilla to the symphysis pubis. About 50% of the patients with polythelia have some other congenital anomaly; renal anomalies lead the list.

**Prepubertal**

1. **Premature thelarche** refers to the onset of bilateral or unilateral breast development before the age of 7½. It is most likely a disorder of hypothalamic hormone receptor sensitivity, as opposed to hypothalamic-pituitary tract tumors or primary ovarian neoplasms.
2. **Precocious puberty**
3. **Unsustained puberty**
4. **Pseudopuberty**
5. **Gynecomastia** (breast enlargement in males)
6. **Polythelia/polymastia** (the presence of more than 2 breasts)

**Adolescence**

1. **Thelarche**: normal development of the female breast which may begin as early as age 8 but, on average, occurs at age 11.
2. **Gynecomastia**
3. **Fibroadenoma** is the most common breast lesion of adolescent females. It presents as a unilateral, mobile, slowly growing, isolated, rubbery mass (1-8 cm in size). These lesions are benign.
4. **Juvenile giant fibroadenoma** is a rapidly growing fibroadenoma seen most commonly in adolescent black females. It is a benign tumor usually treated by surgical excision.
5. **Cystosarcoma phyllodes** is a rapidly growing, large breast mass seen commonly in adolescent black females. It is firm to palpation, has discrete mass borders, and can cause skin or nipple retraction, necrosis, and discharge. Conversion to a malignant tumor has been reported and surgical excision is recommended.
6. **Cystic breast disease** involves cystic lesions in the breast that become painful during the perimenstrual period. Because the disease is self-limiting and usually resolves at the close of adolescence, hormonal manipulation is ill-advised.
7. **Intraductal papilloma** is a benign, subareolar, cylindrical mass with or without a brown to frankly bloody discharge.
8. **Virginal breast hypertrophy** refers to the symmetric enlargement of all breast tissue after puberty. No specific hormonal imbalance has been identified. The breasts can be quite painful and their size embarrassing.
9. **Trauma-induced mass** (fat necrosis) can result in subsequent scarring with a firm, palpable nodular mass.
10. **Polythelia/polymastia**
11. **Carcinoma**
12. **Metastatic sarcoma**

Breathholding Spell or Idiopathic Epilepsy?

Breathholding is a common occurrence in infants and children 6 months to 4 years of age, with about 5% having at least one breathholding spell and some losing consciousness during a prolonged attack. The onset is always with crying, and the characteristic picture of crying and breathholding distinguishes benign episodes from convulsions. The behavior almost always disappears before school age. Some advise parents to leave the room during breathholding to discourage the behavior, but this may be difficult to carry out.

Features of breathholding that distinguish it from grand mal seizures are shown in the accompanying table.

<table>
<thead>
<tr>
<th>Distinguishing Features of Breathholding and Grand Mal Seizures</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
</tr>
<tr>
<td>Age of onset</td>
</tr>
<tr>
<td>Family history</td>
</tr>
<tr>
<td>Precipitating factors</td>
</tr>
<tr>
<td>Occurrence during sleep</td>
</tr>
<tr>
<td>Posture</td>
</tr>
<tr>
<td>Sequence and patterns</td>
</tr>
<tr>
<td>Perspiration</td>
</tr>
<tr>
<td>Heart rate</td>
</tr>
<tr>
<td>Duration</td>
</tr>
<tr>
<td>Incontinence and tongue biting</td>
</tr>
<tr>
<td>Postictal state</td>
</tr>
<tr>
<td>Interictal EEG</td>
</tr>
<tr>
<td>Oculocardiac activation</td>
</tr>
<tr>
<td>Ictal EEG</td>
</tr>
</tbody>
</table>


Bugs in the Band-Aid Box

Wide-eyed and frightened, they appear with white knuckles clutching the metal Band-Aid box. Their gaze is intense and expectant.
You suspect what is in the box without having them tell.
"Is it alive?" you ask phlegmatically.
Frequently, residents of the Band-Aid box include the following:

**Crab Lice (Phthirus pubis)**
This small (1 mm), round, reddish-brown louse causes itching. Transmission is by close personal contact. On close examination, the crab louse is found in the pubic area with its head buried in a hair follicle or clutching two adjacent hairs. The dark nits are frequently difficult to find. Crabs may infest the chest and axillary hair as well as the eyelashes. Treatment: 25 per cent benzyl benzoate or gamma benzene hexachloride on two successive days. Infested eyelashes are treated with daily application of yellow oxide of mercury.

Crab louse (Phthirus pubis)

**Scalp Lice (Pediculus humanus var. capitis)**
This long (up to 4 mm), slender, white louse causes pruritus and excoriations with frequent secondary infection. The densest involvement is posteriorly, behind the ears. There may be tender occipital nodes as well as excoriated bites on the neck and shoulders. You may not find the adult louse, but the small white nits glued to hair shafts are obvious. Nits fluoresce under Wood's light. Treatment: Gamma benzene hexachloride shampoo for two days, repeated in a week. Comb out nits with a fine-toothed comb.

**Body Lice (Pediculus humanus var. corporis)**
The adult louse is 1 to 4 mm long and lives, loves, and lays eggs (nits) in the seams of clothing. This louse feeds on the body, leaving an urticarial wheal with a hemorrhagic central punctum.
Examination of the skin reveals parallel linear excoriations that often are secondarily infected. Treatment: Thorough laundering of clothes and bedding. Iron all seams. Bedding and clothing may be dusted with 10 per cent DDT powder. 1 per cent gamma benzene hexachloride may be applied topically once.
Pinworms (*Enterobius vermicularis*)

The patient may find small, white worms at the anal orifice in the early morning hours. Infestation produces intense perianal pruritus, which leads to excoriation, lichenification, and infection. Bruxism and nightmares are common. The diagnosis is usually made by identifying ova on transparent tape that has been pressed to perianal skin at bedtime. *Treatment*: The Medical Letter has recommended pyrantel pamoate (Banminth) (11 mg/kg) as a single oral dose. Mebendazole, 100 × one dose, regardless of weight, may also be used. The treatment should be repeated in two weeks.

---

![Female pinworm (*E. vermicularis*)](image)

Maggots (Fly Larvae)

Rarely, maggots will be picked from an open sore, the nose, the ear canal, or from the stool.

---

![Maggot](image)

Fish Tapeworm (*Diphyllolothrium latum*)

This is a very large cestode that produces enormous numbers of yellowish eggs. It has been an occupational disease of Jewish housewives who taste raw ground fish to check seasoning when making gefilte fish. Thus, its incidence may be decreasing (at least in this population). Immobile, white, flat segments may be found in the stool. Treatment is with niclosamide. 1 gm for children under 35 kg and 1.5 gm for children over 35 kg. The tablets should be chewed thoroughly.

---

![Fish tapeworm (*D. latum*)](image)

Beef Tapeworm (*Taenia saginata*)

Gravid, white, mobile segments of this worm may be passed in the stool. *Treatment*: Quinacrine, 200 mg every 5-10 min for four dosages, on an empty stomach, followed by a magnesium sulfate purge 2 to 4 hr later. Niclosamide may also be employed in the same dose as for fish tapeworm.
Roundworm (Ascaris lumbricoides)

Ascaris lumbricoides is characterized by an elongated, cylindric, nonsegmented, translucent, flesh-colored body 15–35 cm long. A cosmopolitan worm, ascaris infects 25 per cent of the world’s population. One or more worms may be passed in the stool or, less frequently, vomited. Worms have been known to crawl out of the nose, ear, and umbilical fissures! Treatment: Piperazine citrate syrup 75 mg/kg daily × 2. Pyrantel pamoate may also be employed as a single-dose therapy (11 mg/kg with a 1-gm maximal dose).

Debris

Vegetable particles, such as seeds (corn), stems, and celery, and other debris, like dirt, gravel, stringy fuzz, and cellophane, can be swollen and discolored by passage through the alimentary canal. Even a normal person would be alarmed, and the person with parasitophobia will be in panic. Treatment: Show the patient the characteristics of the debris by hand lens or dissecting microscope.

Miscellaneous

Products of conception, menstrual blood clots thought to be products of conception, “grape-like bodics” of hydatidiform mole, fragments of tampons, and clotted mucus and blood from cystitis have all made it to the Band-Aid box.


Keeping Bugs at Bay

For those of you who hate bugs of all kinds, including mosquitoes, chiggers, flies, and ticks, the following advice from The New York Times ought to come in handy. We hope this makes your summer more pleasant and a bit less itchy.
Keeping Bugs at Bay (Cont.)

Entomologists predict clouds of mosquitos this summer, a result of heavy spring rains. Drain all stagnant water on roofs and in yards. Chiggers, flies and other insects will also be out looking for food. Here are ways to keep flying pests, particularly mosquitos, from assaulting you.

**Repellents**  
Repellents containing DEET are most effective, but apply with care. Do not apply to hands or face or use in concentrations stronger than 50 percent; 15 percent is recommended.

**Clothing**  
Don't look like a flower. Many bugs are attracted to bright colors.

**Perspiration**  
Wash off perspiration. Sweat produces a scent that attracts bugs.

**Chlorine**  
Add a capful or two of chlorinated bleach to your bath water or take a swim in a chlorinated pool. The smell repels most insects.

**Sun oil**  
Oily sun screens make skin too slippery for insects to get a grip.

**Food and fragrance**  
Avoid alcohol and foods high in serotonin, like bananas and nuts. Mosquitoes are attracted to those scents. Avoid perfume, cologne and hair spray.

Sources: Dr. Roger Grothaus, Raid Center for Insect Control, Racine, Wis., and Dr. Jeffrey D. Bernhart, New York City Department of Health.

Oh, for boyhood's painless play,
Sleep that wakes in laughing day,
Health that mocks the doctor's rules,
Knowledge never learned of schools,
Of the wild bee's morning chase,
Of the wild flower's time and place,
Flight of fowl and habitude
Of the tenants of the wood;
How the tortoise bears his shell,
How the woodchuck digs his cell,
And the groundmole sinks his well;
How the robin feeds her young,
How the oriole's nest is hung,
Where the whitest lilies blow,
Where the freshest berries grow,
Where the ground-nut trails its vine,
Where the wood-grape's clusters shine,
Of the black wasp's cunning way,—
Mason of his walls of clay,—
And the architectural plans
Of gray-hornet artisans!—
For, eschewing books and tasks,
Nature answers all he asks,
Hand in hand with her he walks,
Face to face with her he talks,
Part and parcel of her joy,—
Blessings on the barefoot boy!

John Greenleaf Whittier
The Barefoot Boy
CALCIUM

Idiopathic Hypercalciuria in Childhood

The most common metabolic cause of renal calculi in adults is idiopathic hypercalciuria (IH). Children with IH have been noted with increasing frequency to present with a myriad of lower urinary tract signs and symptoms, including calculi, renal colic, hematuria (gross and microscopic), dysuria, frequency-urgency syndrome, pyuria, proteinuria (<100 mg/day), enuresis, osteopenia, failure to thrive, and recurrent urinary tract infections. Given the rather significant manifestations of IH in childhood, the potential that these children will become stone-formers in adult life, and the well-known morbidity of urolithiasis in adults, the following review of idiopathic hypercalciuria in childhood is offered.

1. Definition: Hypercalciuria is an abnormally high urine calcium excretion rate (>2 mg/kg body weight/day) and is either primary (idiopathic) or secondary to a variety of pathologic entities. Investigators have identified two types of IH: absorptive IH, which involves a defect of the intestinal absorption of dietary calcium; and renal leak IH, which is related to a defect of incomplete renal tubular reabsorption of filtered calcium.

2. Disorders Associated with Hypercalciuria

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Immobilization*</td>
<td>Hyperparathyroidism</td>
</tr>
<tr>
<td>Diuretic therapy (e.g., furosemide)*</td>
<td>Cushing's syndrome</td>
</tr>
<tr>
<td>Corticosteroid therapy*</td>
<td>Medullary sponge kidney</td>
</tr>
<tr>
<td>Type 1 renal tubular acidosis</td>
<td>Lead poisoning</td>
</tr>
<tr>
<td>High dietary calcium</td>
<td>Tubular dysfunction</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>(e.g., Fanconi’s syndrome, Wilson’s disease)</td>
</tr>
<tr>
<td>Syndrome of inappropriate ADH secretion</td>
<td>Juvenile rheumatoid arthritis</td>
</tr>
</tbody>
</table>

3. Factors Affecting Renal Calcium Excretion

<table>
<thead>
<tr>
<th>Factor</th>
<th>Cause</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glomerular filtration rate</td>
<td>Vitamin D metabolism (both at the intestinai and renal tubular levels)</td>
</tr>
<tr>
<td>Extracellular volume</td>
<td>Parathyroid hormone</td>
</tr>
<tr>
<td>Serum calcium</td>
<td>Calcitonin</td>
</tr>
<tr>
<td>Serum phosphate</td>
<td>Glucocorticoids</td>
</tr>
<tr>
<td>Blood acid-base status</td>
<td>Mineralocorticoids</td>
</tr>
<tr>
<td>Dietary intake of calcium and sodium</td>
<td>Diuretics (e.g., furosemide)</td>
</tr>
</tbody>
</table>

*The most common causes of hypercalciuria in childhood are indicated by the asterisk.
4. The laboratory approach to evaluating a child with suspected hypercalciuria

Random urine calcium/creatinine ratio
a. If the urine calcium/creatinine ratio is <0.18 mg/mg, quantify with a 24-hour urine
b. If the 24-hour urine [Ca] > 2 mg/kg body weight, RULE OUT:
   i. Secondary causes of hypercalciuria (obtain serum calcium, phosphate, magnesium, bicarbonate, alkaline phosphatase, and blood pH
   ii. Urinary tract infections (perform urine culture).

5. Therapeutic approaches to children with noncalculi urinary tract disorders due to IH.

a. General treatment measures include a fluid intake large enough to allow a high urine flow rate but not large enough to complicate disorders such as enuresis. Excess salt intake should be avoided because increased dietary sodium and the subsequent increased renal filtered sodium load can lead to hypercalciuria. Dietary oxalate (e.g., fruit juices, chocolate, tea) should be avoided because urinary oxalate can serve as the nidus for early urinary calcium crystalization.

b. Dietary calcium. Although restricting dietary calcium in adults with renal stones secondary to IH is frequently recommended, such restriction is usually not indicated in the growing child. A positive calcium balance is optimal for normal development and bone and tissue growth. However, the restriction of dietary calcium is probably indicated in children who develop kidney stones due to absorptive IH and are at risk for destructive renal disease—but, again, the dietary calcium restriction should be limited as much as possible. Children who experience recurrent gross hematuria and severe frequency-urgency syndrome due to absorptive IH are the only patients who should be prescribed moderate-to-severe dietary calcium restrictions.

c. Pharmacologic therapy. The physician should carefully weigh the risks of the disease's morbidity against the potential side-effects of pharmacologic agents. Children presenting with recurrent gross hematuria, dehydration or severe frequency-urgency syndrome, severe dysuria, persistent urinary incontinence, severe abdominal pain, or recurrent urinary tract infections are all possible candidates for drug intervention. Thiazides are the most commonly prescribed agent for IH. These drugs presumably enhance calcium reabsorption in the ascending loop of Henle.


Nondairy Foods Rich in Calcium

Consider caring for a lactase-deficient child who required a high dietary calcium intake. Which calcium-rich foods could you use rather than resorting to medicinal calcium supplements? A few examples of other-than-cow-in-origin calcium are as follows:
Candidal Infection—51

Penile Plaque—An Early Sign of Neonatal Candida Infection

Systemic candidal infections occur in immunosuppressed and immunocompromised patients, particularly those whose normal bacterial flora are suppressed by the use of broad-spectrum antibiotics. Diagnosis of candidal infection is often delayed by the failure to suspect fungal disease as well as by delayed growth of yeast from patient specimens.

The premature neonate is both immunodeficient and likely to be treated with multiple antibiotics. The use of indwelling catheters provides an additional risk factor for the development of candidal infection.

The development of a white plaque adherent to the tip of the penis but beneath the foreskin of male premature infants may provide early evidence of candiduria and disseminated candidal infection. Urine, blood, and cerebrospinal fluid and/or endotracheal aspirate cultures should be used to confirm the suspicion provoked by this finding. The detection of the penile plaque allows early initiation of antifungal therapy. It is hypothesized that the plaque is formed by *Candida* that originates in the perineum or the urine and then forms visible growth in the moist area underlying the foreskin.
52—Carotenemia

Careful examination of the genitalia is important even in the intensive care nursery.


**CAROTENEMIA**

**Carotenemia or, Better Expressed, Hypercarotenemia**

Carotenemia is a common condition characterized by a yellow-orange discoloration of the skin and concomitant elevated serum carotene levels. The majority of these cases are harmless and due to the ingestion of large amounts of carotene-rich foods over a long period of time (e.g., carrots, sweet potatoes, squash). The syndrome is associated rarely with disease entities such as diabetes mellitus and hypothyroidism.

**Conditions Associated with Carotenemia**

| Excessive dietary intake (the most common cause of carotenemia) | Liver disease |
| Diabetes mellitus | Renal disease (e.g., chronic glomerulonephritis and nephrotic syndrome) |
| Hypothyroidism | Inborn errors of metabolism |
| Simmonds’ disease (panhypopituitarism) | Familial conditions |
| Hypothalamic amenorrhea | Malaria |
| Anorexia nervosa | |
| Human castrates | |

**Clinical Manifestations**

1. Yellow pigmentation of the skin, most prominently on palms, soles, and nasolabial folds
2. Carotene is excreted by the sebaceous sweat glands; thus, the discoloration of the skin is most prominent where sweating is most profuse.
3. It gradually extends over the body.
4. Sclera are always spared from carotene staining. (Corneum of the skin has a high lipid content with an affinity for carotene; the mucosa has no affinity to carotene.)
5. Elevated serum carotene level.
6. Patients rarely complain of constitutional symptoms, such as loss of appetite, malaise, itching, and right upper quadrant abdominal pain.

**Differential Diagnosis**

- Jaundice
- Lycopenemia (orange-reddish discoloration of the skin due to an increased consumption of lycopene-rich foods, especially tomatoes)
- Excessive ingestion or percutaneous absorption of chemicals such as quinacrine, mepacrine, dinitrophenol, saffron, picric acid, and canthaxanthin, the major coloring constituent in “tanning capsules.”
Treatment

Avoid carotene-rich foods but consume a well-balanced diet. The yellow discoloration resolves over several weeks to months, although serum carotene levels drop severely after only 1 week of a carotene-poor diet. If carotenemia does not resolve or an underlying etiology is suspected, an appropriate investigation should ensue.

Foods High in Carotene Content

<table>
<thead>
<tr>
<th>Vegetables</th>
<th>Fruits</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alfalfa</td>
<td>Rutabagas</td>
<td>Butter</td>
</tr>
<tr>
<td>Asparagus</td>
<td>Spinach</td>
<td>Egg yolks</td>
</tr>
<tr>
<td>Beans</td>
<td>Squash</td>
<td>Milk</td>
</tr>
<tr>
<td>Beet greens</td>
<td>Sweet potatoes</td>
<td>Palm Oil</td>
</tr>
<tr>
<td>Broccoli</td>
<td>Yellow turnips</td>
<td>Yellow corn</td>
</tr>
<tr>
<td>Carrots</td>
<td>Watercress</td>
<td>Yellow fat</td>
</tr>
<tr>
<td>Chard</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Collard greens</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cucumbers</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Endive</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Escarole</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kale</td>
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</tr>
</tbody>
</table>


CAT SCRATCH DISEASE

Diagnostic Criteria for Cat Scratch Disease

Cat scratch disease is a self-limited bacterial infection that is usually transmitted to humans by felines, although other animals have been implicated. It presents primarily in children with a single lymph node enlargement or regional lymphadenitis and an ocular, skin, or mucous membrane lesion in the region of the adenitis. Other manifestations include fever, malaise, headache, anorexia, rash, sore throat, splenomegaly, and, rarely, a severe, chronic, systemic form of the illness. The symptoms can last for weeks to months.

The most important historical information in confirming a diagnosis of cat scratch disease is whether or not the patient has had contact with a cat; 99% of cat scratch disease patients have had such contact, and 78% of that population have had contact with a kitten. An inoculation site is also vital to the diagnosis. Of the criteria listed below, at least 3 of the 4 noted are required for diagnosis of cat scratch disease. To these criteria most cat scratch aficionados would add pertinent laboratory data to rule out other causes of lymphadenopathy.

1. Single or regional lymphadenopathy.
2. Animal contact, with a scratch or inoculation lesion.
4. Node or inoculum site with compatible histopathology or Warthin-Starry stain positive organisms.
**54—Cervicitis**

**Differential Diagnosis**

**Other infections causing adenopathy:**
- Infectious mononucleosis
- Mycobacterial infection
- Staphylococcal infection

**Other fungi**
- Noninfectious disorders:
  - Streptococcal infection
  - Tularemia
  - Syphilis
  - Toxoplasmosis
  - Sporotrichosis

**Cervicitis and Vulvovaginitis in the Adolescent**

There exists much overlap in the clinical presentations of cervical, vaginal, uterine, fallopian tube, and urinary tract infections. Most of these entities share...
the same symptoms such as dysuria, vulvar pruritis, dyspareunia, and increased or altered vaginal discharge. It is essential, therefore, when evaluating adolescent women with such complaints to (1) exclude the diagnosis of upper tract disease, such as endometritis, salpingitis, and pyelonephritis; (2) differentiate among vaginitis, cervicitis, urethritis, and cystitis; and (3) identify the specific etiologic agent that is causing the infection so that the proper treatment can be prescribed. Listed below are the various causes of vulvovaginitis and cervicitis:

1. Vulvovaginitis
   a. Physiologic leukorrhea (normal vaginal discharge that increases in volume with estrogen stimulation)
   b. Candidiasis (e.g., Candida albicans and Torulopsis glabrata)
   c. Trichomoniasis (Trichomonas vaginalis)
   d. Bacterial vaginosis (this entity occurs in 30 to 50% of women with vaginitis, making it the most common cause of abnormal vaginal discharge. It is probably the result of an interplay between the overgrowth of Gardnerella vaginalis and various anaerobes, and the subsequent decrease in the presence of lactobacilli that normally inhabit the vagina)
   e. Foreign body (e.g., tampons, IUDs, etc.)
   f. Allergic or contact vulvovaginitis (e.g., contact with soaps and other cleaning agents, spermicides, lubricants, douches, sanitary napkins, nylon or rayon underwear, obesity, hot weather, poor hygiene, etc.)
   g. Allergic seminal vulvovaginitis
   h. Psychosomatic illness should be considered when an adolescent frequently presents with vaginal symptoms but without objective evidence of vulvar or vaginal inflammation or discharge.

2. Other Causes of Vulvovaginal Complaints
   a. Systemic conditions.
      i. Fistulas from the bladder or rectum (e.g, Crohn’s disease, Stevens-Johnson syndrome, Behçet’s syndrome)
      ii. Tropical ulcerations (e.g., amebiasis, filariasis, tuberculosis, schistosomiasis)
      iii. Systemic illnesses (e.g., typhoid, smallpox, varicella, measles, scarlet fever)
      iv. Dermatologic complaints (e.g., atopic dermatitis, seborrheic dermatitis, psoriasis, lichen sclerosus)
      v. Anatomic anomalies (e.g., aberrant urethral orifice, labial agglutination, urethral prolapse)
   b. Vulvar lesions
      i. Condyloma acuminatum
      ii. Genital herpes
      iii. Syphilis
      iv. Chancroid
      v. Lymphogranuloma venereum
      vi. Granuloma inguinale
      vii. Pediculosis
      viii. Scabies
      ix. Bartholinitis
      x. Skenitis
      xi. Tumors (e.g., carcinoma, sarcoma, botryoides, vaginal polyps)
      xii. Pemphigus
      xiii. Acute ulcerative vulvitis
      xiv. Lipschütz ulcer
3. Cervicitis

Whatever its etiology, cervicitis should be considered a sexually transmitted disease with great potential to spread to other sexual partners as well as to extend, in the case of contact, from the cervix to the endometrium and salpinx. Etiologies include:

a. *Chlamydia trachomatis*

b. *Neisseria gonorrhoeae*

c. Herpes simplex virus

d. Possible etiologic agents include *Mycoplasma hominis, Ureaplasma urealyticum*, and group B streptococci.


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**CHEST PAIN**

**Common Causes**

- Costochondritis
- Arthritis
- Infectious costochondritis
- Tietze’s syndrome
- Cough
- Herpes zoster
- Idiopathic
- Indigestion (heartburn, esophagitis)
- Mitral valve prolapse
- Musculoskeletal (strain, occult trauma)
- Pneumonitis
- Psychogenic
- Reactive airway disease
- Sickle-cell disease
- Trauma

**Uncommon Causes**

- Arrhythmia
- Congenital heart disease
- Congestive heart failure
- Esophageal (trauma associated with vomiting, foreign body)
- Pleuritis/pleurisy
- Pneumothorax
- Precordial catch

**Rare Causes**

- Cholecystitis
- Diaphragmatic irritation
- Abscess
- Fitz-Hugh-Curtis syndrome
- Peritonitis
- Ruptured viscus
- Tumor
- Endocarditis
- Juvenile rheumatoid arthritis
- Myocardial ischemia (e.g., anomalous coronary artery)
- Myocarditis
- Osteomyelitis (vertebrae, ribs)
- Peptic ulcerative disease
- Pericarditis
- Pneumoniediastinum
- Pulmonary embolism
- Rheumatic fever
**Recommendations for Screening and Managing Cholesterol in Children**

The following table summarizes the American Academy of Pediatrics (AAP)/American Heart Association's (AHA) and the National Cholesterol Education Program's (NCEP) recommendations for testing and managing cholesterol in children. The American Health Foundation's (AHF) recommended universal, population-based approach to cholesterol control is also presented in outline.

### High-risk, patient-based approach

<table>
<thead>
<tr>
<th>AAP/AHA Recommendations</th>
<th>NCEP Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Screening strategy</strong></td>
<td>Screen children over 2 yr who have a parental history of hyperlipidemia (&gt;240 mg/dL total cholesterol) or a family history of early CAD (under 55 yr for males and females). Also consider screening if family history is not obtainable or patient has several risk factors for CAD independent of history (obesity, hypertension, smoking).</td>
</tr>
<tr>
<td><strong>Screening method</strong></td>
<td>If child has family history of early CAD, screen initially with two lipid profiles. In other high-risk patients, screen initially with nonfasting total cholesterol. If total cholesterol is borderline (170-199 mg/dL), repeat and average results. If initial screening measurement is 200 mg/dL or more, or the average of two measurements is &gt;170 mg/dL, perform two lipid profiles.</td>
</tr>
<tr>
<td><strong>Management</strong></td>
<td>Total cholesterol is normal (&lt;170 mg/dL): Routine care, repeat cholesterol testing 5 yr later. Low-risk lipid profile (LDL-C &lt;110 mg/dL, less than 75th percentile): Routine care, repeat lipid profile every 5 yr. Moderate-risk lipid profile (LDL-C 110-129 mg/dL, 75th to 95th percentile): Dietary counseling, follow-up lipid profile in 1 yr. High-risk lipid profile (LDL-C ≥130 mg/dL, 95th percentile): Dietary counseling, perform lipid profiles on parents, initiate step 1 diet. Repeat lipid profile in 6 wk. If unchanged, intensify step 1 diet and repeat lipid profile in 3 mo. If still unchanged, initiate step 2 diet and repeat lipid profile in 3 mo. Refer to lipid specialist if diet therapy ineffective.</td>
</tr>
</tbody>
</table>

* The NCEP defines a family history of early CAD as having parents or grand parents 55 yr of age or under who have had one or more of the following: coronary atherosclerosis diagnosed by coronary arteriography; balloon angioplasty or coronary artery bypass surgery; documented myocardial infarction, angina pectoris, peripheral vascular disease, cerebrovascular disease, or sudden cardiac death. A repeat nonfasting total cholesterol measurement or second fasting lipid profile should be done no sooner than 1 wk after initial test and no later than 8 wk after initial test. The second measurement is unacceptable if the total cholesterol or LDL-C level is within 30 mg/dL of the initial measurement. If unacceptable, obtain a third measurement.

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Table continued on next page.
Recommendations for Screening and Managing Cholesterol in Children (Cont.)

Universal, population-based approach (recommended by AHF)

Screening strategy
Screen all children over 2 yr of age. Consider repeat screening every 5 yr thereafter.

Screening method
Screen initially with nonfasting total cholesterol, and proceed to fasting lipid profile if levels are elevated as described above under NCEP recommendations.

Management See recommendations above.


CLINICAL JUDGMENT

Quantifying Judgment in the Evaluation of Sick Children

We all use judgment and experience to distinguish the “sick” child from the “not-so-sick” child. No amount of wisdom and no laboratory test will make this distinction accurate in every case, but the scale below will help you quantify your judgment, or at least it will help you recognize how you reach your own conclusions.

Six Observation Items and Their Scales*

<table>
<thead>
<tr>
<th>OBSERVATION ITEM</th>
<th>1 NORMAL</th>
<th>3 MODERATE IMPAIRMENT</th>
<th>5 SEVERE IMPAIRMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Quality of cry</td>
<td>Strong with normal tone or content and not crying</td>
<td>Whimpering or sobbing</td>
<td>Weak or moaning or high pitched</td>
</tr>
<tr>
<td>Reaction to parental stimulation</td>
<td>Cries briefly then stops or content and not crying</td>
<td>Cries off and on</td>
<td>Continual cry or hardly responds</td>
</tr>
<tr>
<td>State variation</td>
<td>If aware, stays awake; or if asleep and stimulated, wakes up quickly</td>
<td>Eyes close briefly awake; awakes with prolonged stimulation</td>
<td>Fails to sleep or will not rise</td>
</tr>
<tr>
<td>Color</td>
<td>Pink</td>
<td>Pale extremities or acrocyanosis</td>
<td>Pale, cyanotic, mottled, or ashen</td>
</tr>
<tr>
<td>Hydration</td>
<td>Skin normal, eyes normal, and mucous membranes moist</td>
<td>Skin, eyes normal and mouth slightly dry</td>
<td>Skin doughy or tented, and dry mucous membranes and/or sunken eyes</td>
</tr>
<tr>
<td>Response (talk, smile) to social overtures</td>
<td>Smiles or alerts (≤2 mo)</td>
<td>Brief smile, alerts briefly (≤2 mo)</td>
<td>No smile; face anxious, dull, and expressionless or no alerting (≤2 mo)</td>
</tr>
</tbody>
</table>

*Source see reference below; with permission.
To use this system, assign a score of 1-3 for each of the observation items. When the investigators used the scale to evaluate children less than 24 months of age who had fever of 38.3°C or more, only 2.7% of children with a score of less than 10 had a serious illness, whereas 92.3% of those with a score greater than or equal to 16 were seriously ill. Among those children with scores of 11 to 15, serious illness was found in 26.2%. A child was defined as having a serious illness if a bacterial pathogen was isolated from blood, CSF, urine, stool, joint fluid, or deep soft tissue aspirate, or if electrolyte abnormality, pulmonary infiltrates on chest x-ray, abnormal blood gases, or CSF pleocytosis was found.

It might be helpful to compare your own instincts to this scale to find out why you conclude what you do.


COCAINE

Differential Diagnosis of Cocaine Intoxication

The acute manifestations of severe cocaine ingestion are many. Manifestations may be divided systematically:

1. Autonomic nervous system overactivity
   a. Dilated pupils
   b. Tachycardia
   c. Diaphoresis
   d. Pallor secondary to vasoconstriction
2. Central nervous system
   a. Dysphoric agitation or stimulation
   b. Tremor
   c. Convulsions
   d. Coma
   e. Hyperthermia
3. Cardiovascular
   a. Small doses slow the heart rate
   b. High doses increase the heart rate and elevate the blood pressure
   c. Dysrhythmias
4. Respiratory (stimulation followed by depression of the respiratory center)

It is important when entertaining the diagnosis of cocaine intoxication to consider other clinical entities that may mimic it:

Medical differential of cocaine ingestion:
- Thyroid storm
- Hypoglycemia
- Thiamine deficiency
- Acute psychosis
- Sedative/hypnotic withdrawal
- Seizure disorders
- Sepsis
- Pheochromocytoma
- Head injury

Chemical differential of cocaine ingestion:
- Stimulants/sympathomimetics
- Phenylpropanolamine
- Phencyclidine (smoked)
- Amphetamines
- Anticholinergic agent
- Hallucinogens
- Cyclic antidepressants
- Strychnine

COMA

Common Causes

CNS trauma
  - Cerebral edema
  - Concussion
  - Hemorrhage
    - Epidural
    - Subarachnoid
    - Subdural
  - Increased intracranial pressure

Drug intoxication
  - Analgesics
  - Anticonvulsants
  - Antihistamines
  - Benzodiazepines
  - Digoxin
  - Ethanol
  - Heavy metals
  - Hydrocarbons

Drug intoxication (Cont.)
  - Hypnotics
  - Barbiturates
  - Insulin
  - Lithium
  - Organophosphates
  - Phencyclidine
  - Phenothiazines
  - Salicylate
  - Tricyclic antidepressants

Uncommon Causes

Cardiorespiratory
  - Cardiopulmonary arrest
  - Hypercapnea
  - Hypoxemia

Infections
  - Abscess
  - Encephalitis
  - Meningitis

Metabolic
  - Hyper/hypocalcemia
  - Hyper/hypomagnesemia

Uncommon Causes (Cont.)
  - Metabolic
    - Hypernatremia
    - Hypoglycemia
    - Hyponatremia
    - Water intoxication
    - Metabolic acidosis
    - Metabolic alkalosis
    - Postictal state
    - Postoperative
    - General anesthesia
    - Hypotension/hypoxemia
    - Sepsis

Rare Causes

Cardiac
  - Arrhythmia
  - Hypertension
  - Hypoperfusion
    - Aortic stenosis
    - Coarctation of the aorta

Cerebrovascular
  - Hemorrhage
  - Thrombophlebitis
  - Vasculitis
  - Venous thrombosis

Dehydration

Diabetic ketoacidosis

Endocrine disorders
  - Addison's disease
  - Congenital adrenal hyperplasia
  - Cushing's disease

Inborn errors of metabolism
  - Hyperammonemia
  - Hypoglycemia

Heat stroke

Hepatic failure

Hypothermia

Malignant hyperthermia

Porphyria

Postinfectious encephalomyelitis

Measles

Other viral infections

Psychiatric disturbances
  - Fugue state
  - Hysteria

Reye's syndrome

Sudden infant death syndrome (SIDS)

Uremia
Evaluation of the Comatose Child

There exist three important categories of central nervous system lesions that can cause alterations in one's level of consciousness: (1) supratentorial mass lesions, (2) infratentorial mass lesions, and (3) metabolic abnormalities. In evaluating the comatose child at the time of presentation, careful notice of the neurologic findings and determination of the type of CNS lesion incurred are very useful in dictating acute medical management.

1. Supratentorial Mass Lesions
   a. Important causes of supratentorial lesions that yield progressive deterioration in children:
      i. Cerebral hyperemia secondary to head trauma
      ii. Epidural and subdural hematomas
      iii. Intracerebral hemorrhages
      iv. Acute hydrocephalus
      v. Subdural hemorrhages
      vi. Severe systemic hypertension
      vii. Obstruction of an existing ventricular-peritoneal shunt
      viii. Bleeding arteriovenous malformation
   b. Neurologic findings of supratentorial lesions
      i. Initial signs and symptoms suggest focal hemispheric disease
      ii. Signs progress in a rostral to caudal direction
      iii. Pupillary reflexes are usually depressed
      iv. Motor signs are often symmetrical

2. Infratentorial Mass Lesions (Posterior Fossa)
   These lesions can yield coma either by destroying the ascending reticular activating system or by compression of that system by a mass or tumor.
   a. Important causes of infratentorial lesions:
      i. Brainstem contusions associated with trauma
      ii. Cerebellar hemorrhage or tumor with secondary hydrocephalus
      iii. Brainstem encephalitis
      iv. Basilar artery thrombosis
   b. Neurologic findings of infratentorial lesions
      i. Brainstem signs and symptoms are common.
      ii. Signs are not rostral to caudal in evolution.
      iii. Cranial nerve palsies are common.
      iv. Abnormalities of the respiratory pattern are common and appear at the onset of coma.

3. Metabolic Disorders
   Metabolic disorders make up the majority of nontraumatic processes that cause acute coma in the pediatric patient.
   a. Important causes of metabolic coma:
      i. Hypoxic-ischemic coma (e.g., respiratory failure, shock, severe anemia, apnea of infancy, carbon monoxide poisoning, cerebral vasculitis)
ii. Infections (e.g., encephalitis, meningitis, botulism)
iii. Postictal state
iv. Hypoglycemia
v. Nonendocrine organ failure (e.g., hepatic and renal)
vi. Endocrine organ failure (e.g., pancreas, adrenal, thyroid, pituitary)
vii. Poisonings (e.g., narcotics, barbiturates, sedatives, etc.)
viii. Miscellaneous (e.g., Reye's syndrome, electrolyte abnormalities, and hypothermia or hyperthermia)

b. Neurologic findings of metabolic lesions
i. Stupor or coma precede motor signs.
ii. Motor signs are usually symmetrically depressed.
iii. Pupillary reactions are preserved.
iv. Acid-base imbalance is common.
v. Seizures or abnormal motor movements are common findings.


Ingestions and the Pupillary Examination in a Comatose Child

When confronted with a comatose child, the possibility of ingestion of a toxic or poisonous substance should always be considered. A history should be obtained addressing which medications and poisonous substances are at home, how they are kept separate from the child, and if there is any suspicion on the parents’ part that the child may have ingested something. Modern advances in technology and pharmacology have made the serum and urine toxicologic screening tests most useful to the diagnosis of toxic ingestion. The examination of a comatose child’s pupillary size and reactivity, however, remains a useful bedside exam that can be performed with a minimum of difficulty and time. Although the pupillary exam is not nearly as specific as the toxicologic screening tests, it can point the way toward considering a diagnosis of ingestion of poison.

The Pupillary Examination and Coma

<table>
<thead>
<tr>
<th>MYDRIASIS (DILATION OF THE PUPIL)</th>
<th>MIOSIS (CONTRACTION OF THE PUPIL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amphetamines</td>
<td>Cocaine</td>
</tr>
<tr>
<td>Antihistamines</td>
<td>Ephedrine</td>
</tr>
<tr>
<td>Atropine</td>
<td>Ethyl alcohol</td>
</tr>
<tr>
<td>Botulism</td>
<td>Snake venom</td>
</tr>
<tr>
<td></td>
<td>Opiates</td>
</tr>
<tr>
<td></td>
<td>Barbiturates</td>
</tr>
<tr>
<td></td>
<td>Propoxyphen</td>
</tr>
<tr>
<td></td>
<td>Meperidine</td>
</tr>
<tr>
<td></td>
<td>Methadone</td>
</tr>
<tr>
<td></td>
<td>Carbon monoxide</td>
</tr>
<tr>
<td></td>
<td>Organophosphates</td>
</tr>
<tr>
<td></td>
<td>Clonidine</td>
</tr>
</tbody>
</table>


CONGENITAL HEART DISEASE

The First Manifestation of Congenital Heart Disease

Depending on the nature of the anatomic lesion, the first sign or symptom of congenital heart disease may vary. The table below is designed to serve as a guide to the most likely lesions, given the clinical presentation.
Marked Cyanosis

Transposition of the great arteries
Pulmonary atresia and stenosis with intact ventricular septum
Tetralogy of Fallot with severe pulmonary stenosis
Complex pulmonary atresias
Tricuspid atresia
Ebstein's malformation of the tricuspid valve

Congestive Heart Failure

Aortic atresia
Coarctation of the aorta
Double outlet right ventricle syndrome
Patent ductus arteriosus
Truncus arteriosus
Ventricular septal defect
Arteriovenous fistulas

Abnormal Heart Rate

Supraventricular tachycardia
Heart block

Heart Murmurs

Patent ductus arteriosus
Pulmonary stenosis
Aortic stenosis
Pulmonary artery stenosis
Ventricular septal defect
Arteriovenous fistulas
Atrioventricular valve regurgitations


Recurrence Risks for Congenital Heart Disease

Congenital cardiac lesions are thought to recur in three patterns of inheritance:

1. As part of a single gene defect syndrome (2%).
2. Chromosomal abnormalities (4%).
3. Multifactorial inheritance (94%).

Some single gene syndromes that often include cardiac defects are listed in the following table.

Selected Single Mutant Gene Syndromes with Cardiovascular Disease Other than Coronary Artery

<table>
<thead>
<tr>
<th>Autosomal Dominant</th>
<th>Autosomal Recessive</th>
<th>X-Linked</th>
</tr>
</thead>
<tbody>
<tr>
<td>Apert</td>
<td>Adrenogenital syndrome</td>
<td>Incontinentia pigmenti</td>
</tr>
<tr>
<td>Crouzon</td>
<td>Alkaptonuria</td>
<td>Mucopolysaccharidosis II</td>
</tr>
<tr>
<td>Ehlers-Danlos</td>
<td>Carpenter</td>
<td>Muscular dystrophy</td>
</tr>
<tr>
<td>Forney</td>
<td>Conradi</td>
<td></td>
</tr>
<tr>
<td>Holt-Oram</td>
<td>Cutis laxa</td>
<td></td>
</tr>
<tr>
<td>IHSS (not strictly</td>
<td>Ellis-van Creveld</td>
<td></td>
</tr>
<tr>
<td>a syndrome)</td>
<td>Friedreich's ataxis</td>
<td></td>
</tr>
<tr>
<td>Leopard</td>
<td>Glycogenosis IIa, IIIa, IV</td>
<td></td>
</tr>
<tr>
<td>Marfan</td>
<td>Jervell and Lange-Nielsen</td>
<td></td>
</tr>
<tr>
<td>Myotonic dystrophy</td>
<td>Laurence-Moon-Biedl</td>
<td></td>
</tr>
<tr>
<td>Neurofibromatosis</td>
<td>Mucolipidosis III</td>
<td></td>
</tr>
<tr>
<td>Osteogenesis imperfecta</td>
<td>Mucopolysaccharidosis II, IV, V, VI</td>
<td></td>
</tr>
<tr>
<td>Romano-Ward</td>
<td>Réfsum</td>
<td></td>
</tr>
<tr>
<td>Treacher Collins</td>
<td>Seckel</td>
<td></td>
</tr>
<tr>
<td>Tuberous sclerosis</td>
<td>Smith-Lemli-Opitz</td>
<td></td>
</tr>
<tr>
<td>Ullrich-Noonan</td>
<td>Thrombocytopenia with absent radius (TAR)</td>
<td>Weill-Marchesani</td>
</tr>
</tbody>
</table>

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The recurrence risk for congenital heart disease in families in which one member has one of these syndromes depends on the recurrence risk for the syndrome (generally 25 to 50%) and the frequency with which congenital heart disease is encountered in the syndrome. The recurrence risk for congenital heart disease due to a chromosomal abnormality depends on the risk of recurrence of the chromosomal defect. A familial tendency for nondisjunction and the presence of a translocation in the chromosomal pattern of one parent may increase the likelihood of recurrence. Some chromosomal defects are associated with particular cardiac abnormalities. The more common chromosomal aberrations and their associated cardiac defects are listed in the following table.

### Congenital Heart Disease in Selected Chromosomal Aberrations

<table>
<thead>
<tr>
<th>Population Studied</th>
<th>Percentage Incidence of CHD</th>
<th>Most Common Lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>General population</td>
<td>1</td>
<td>VSD, PDA, ASD</td>
</tr>
<tr>
<td>4p</td>
<td>40</td>
<td>VSD, ASD, PDA</td>
</tr>
<tr>
<td>5p (cri du chat)</td>
<td>25</td>
<td>VSD, ASD, PDA</td>
</tr>
<tr>
<td>C mosaic</td>
<td>50</td>
<td>VSD</td>
</tr>
<tr>
<td>13 trisomy</td>
<td>90</td>
<td>VSD, PDA</td>
</tr>
<tr>
<td>13q</td>
<td>50</td>
<td>VSD</td>
</tr>
<tr>
<td>18 trisomy</td>
<td>90+</td>
<td>VSD, PDA, Dex</td>
</tr>
<tr>
<td>18q</td>
<td>50</td>
<td>VSD, AV, ASD</td>
</tr>
<tr>
<td>21 trisomy</td>
<td>50</td>
<td>VSD, AV canal, ASD</td>
</tr>
<tr>
<td>XO Turner</td>
<td>35</td>
<td>Coarc, AS, ASD</td>
</tr>
<tr>
<td>XXXXY</td>
<td>14</td>
<td>PDA, ASD</td>
</tr>
</tbody>
</table>


The essential components of multifactorial inheritance of congenital heart defects include: (1) a genetic predisposition to cardiovascular maldevelopment, (2) a genetic predisposition to be adversely affected by environmental teratogens, and (3) an environmental insult occurring at a vulnerable period of cardiac development (i.e., very early in pregnancy). Since there is no method of quantitating the presence of these three risks for a given offspring, a few percentages may be kept in mind:

1. In general, the risk of recurrence of congenital heart disease in a given family is 1 to 5%.
2. The more common the heart defect in the affected family member, the more likely that defect is to recur.
3. The risk to subsequent offspring of two parents triples if two existing family members are affected. (For example, the risk of recurrence of a ventriculo-septal defect is about 5% if one previous sibling is affected; however, it increases to approximately 15% if one sibling and one parent or two siblings are affected.)
4. If the majority of family members have some form of congenital cardiac defect, the risk to subsequent offspring approaches 100%. More specifically, if three first-degree family members are affected, the risk in future pregnancies is 60 to 100%.
Recurrence risks for siblings of family members with congenital heart defects are shown in the following table:

<table>
<thead>
<tr>
<th>Defect</th>
<th>% Sibling at Risk</th>
<th>1 Sibling Affected</th>
<th>2 Siblings Affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ventricular septal defect</td>
<td>3</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Patent ductus arteriosus</td>
<td>3</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Atrial septal defect</td>
<td>2.5</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2.5</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Pulmonary stenosis</td>
<td>2</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>2</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>2</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Transposition</td>
<td>1.5</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Endocardial cushion defects</td>
<td>3</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Fibroelastosis</td>
<td>4</td>
<td>12</td>
<td></td>
</tr>
<tr>
<td>Hypoplastic left heart</td>
<td>2</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Tricuspid atresia</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Ebstein anomaly</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
</tbody>
</table>

Based on combined data published during two decades from European and North American populations. (From Nora JJ, Nora AH: Update on counseling the family with a first-degree relative with a congenital heart defect. Am J Med Genet 29:137-142, 1988, with permission.)


**CONSTIPATION**

**Common Causes**

- Appendicitis
- Breastfeeding (begins around 6 weeks of age)
- Cow's milk ingestion
- Drugs
  - Anticholinergics
  - Antihistamines
  - Narcotics
  - Phenothiazines
- Dysfunctional toilet training
- Emotional disturbances
- Functional ileus
- Immobility
- Inappropriate expectations of the caretaker
- Intentional withholding
- Intestinal abnormalities
  - Atresia
  - Hirschsprung's disease
  - Microcolon
  - Volvulus
  - Web
- Low dietary fiber
- Meconium plug/ileus
- Meningomyelocoele
- Mental retardation/cerebral palsy
- Painful defecation (hemorrhoids, fissure, skin irritation)
Uncommon Causes

Diabetes mellitus
Electrolyte disturbances
Hyper/hypocalcemia
Hyperkalemia
Hypothyroidism
Imperforate anus/anal stenosis

Intestinal pseudo-obstruction
Lead poisoning
Salmonellosis
Spinal cord injury/tumor
Starvation

Rare Causes

Amyloidosis
Botulism
Dolichocolon
M, opathies/myotonias
Pheochromocytoma

Sacral malformations
Scleroderma
Tetanus
Tethered cord

CONTRACEPTION

Side Effects of Hormonal Contraception

The mnemonic ACHES for recalling the dangerous side-effects of oral contraceptive use is well known (A: abdominal pain; C: chest pain, cough, and shortness of breath; H: headaches; E: eye problems such as blurred vision; S: severe leg pain in the calves and thighs). There exist, however, a great many other untoward effects from oral contraceptive use that may play a role in an adolescent woman’s decision not to comply with this method of birth control. Arranged in terms of the hormonal cause, these untoward effects are presented below:

Hormonal Side Effects of Oral Contraceptives

<table>
<thead>
<tr>
<th>ESTROGEN EXCESS</th>
<th>PROGESTIN EXCESS</th>
<th>ESTROGEN DEFICIENCY</th>
<th>PROGESTIN DEFICIENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea, vomiting</td>
<td>Fatigue, depression</td>
<td>Irritability, nervousness</td>
<td>Late breakthrough bleeding and spotting</td>
</tr>
<tr>
<td>Edema bloating</td>
<td>Acne, oily skin, hirsutism</td>
<td>Hot flashes, motor symptoms</td>
<td>Heavy menstrual flow and clots</td>
</tr>
<tr>
<td>Cyclic weight gain</td>
<td>Alopecia</td>
<td>Early midecycle spotting</td>
<td>Delayed onset of menses, dysmenorrhea, weight loss</td>
</tr>
<tr>
<td>Dysmenorrhea, uterine cramps</td>
<td>Increased appetite, shortened menses</td>
<td>Decreased amount of early menstrual flow</td>
<td></td>
</tr>
<tr>
<td>Breast tenderness, increased breast size, vascular headaches</td>
<td>Decreased libido</td>
<td>No withdrawal bleeding</td>
<td></td>
</tr>
<tr>
<td>Chloasma</td>
<td>Headaches between pill packages</td>
<td>Dry vaginal mucosa, atrophic vaginitis</td>
<td></td>
</tr>
<tr>
<td>Lactation supression</td>
<td>Cholestatic</td>
<td>Headaches</td>
<td></td>
</tr>
<tr>
<td>Irritability, depression</td>
<td>jaundice</td>
<td>Depression</td>
<td></td>
</tr>
</tbody>
</table>


CORTISOL

Cortisol Replacement During Febrile Episodes

Daily cortisol production rates among normal children have previously been estimated at 12 mg/m²/day. The detection of clinical evidence of cortisol excess among children with adrenal insufficiency treated with this dose suggests that it may be too high. A new study using recently developed, more accurate methods of determining cortisol production rates in normal children suggests that 7 mg/m²/day may be a more appropriate dose.

Whatever the baseline dose of cortisol used to treat patients with adrenal insufficiency, an increase in that dose is recommended during periods of stress. The most common form of generalized stress during childhood is febrile illness. The usual recommendation is that the daily dose of steroid should be increased 2- to 3-fold during febrile illnesses. In fact, when 105 normal children 1 month to 12 years of age were studied, those children with upper respiratory infection, streptococcal pharyngitis, and otitis media experienced serum cortisol increases of 2- to 3-fold, whereas those children with pneumonia, fever of unknown origin, and bacterial meningitis demonstrated a 5- to 6-fold rise. It seems prudent to attempt to reproduce these apparently physiologic stress levels of serum cortisol for children with inadequate intrinsic production during severe physiologic stress. It is also important to remember that oral replacement should be approximately twice the above recommendations because of poor oral absorption and hepatic biodegradation.


COUGH

Common Causes

Allergic disease
Aspiration (direct or indirect)
Atelectasis
Bacterial infection
  Bronchiectasis
  Bronchitis
  Pneumonia
Sinusitis
Trachitis
Congestive heart failure
Environmental pollution
Foreign body
Gastroesophageal reflux
Infections, other
  Chlamydia
  Mycoplasma
  Pertussis
  Postnasal drip
  Reactive airway disease
  Smoking/passive smoking
Viral infection
  Bronchiolitis
  Croup
  Pneumonitis
  Upper respiratory infection
Uncommon Causes

Cystic fibrosis
Malformation of the airway
Malignancy (primary or metastatic)
Mediastinal adenopathy
Psychogenic

Tracheobronchomalacia
Tracheoesophageal fistula
Tuberculosis
Vascular ring

Rare

Allergic bronchopulmonary aspergillosis
Auricular nerve stimulation
Bronchogenic cyst
Congenital lobar emphysema
Immotile cilia syndrome
Lymphocytic interstitial pneumonitis

Opportunistic infections
(PCM, CMV, MAI, fungal)
Parasitic infection
Pulmonary embolism
Pulmonary hemosiderosis
Pulmonary sequestration
Sarcoidosis

Habit (Psychogenic) Cough

Habit cough is a transient tic disorder that may be seen in early adolescence, especially in puberty, and, by definition (DSM-III-R), lasts more than 1 month and less than 1 year. The behavior may be seen as part of Gilles de la Tourette syndrome but is usually an independent entity. The clinical features were described by Berman:

... persistent violent spasms of harky, harsh, nonproductive cough, occurring almost always during the waking hours and unaccompanied by systemic signs and symptoms of chronic disease: a paucity or complete absence of abnormal findings in the chest; lack of response to most potent cough preparations; and a cough that remains unchanged after exertion, laughter, infection, dampness, and extremes of temperature.

The diagnosis requires exclusion of organic causes. Habit cough usually disappears with sleep, which is not the case with most organic diseases.

Other motor tics include throat clearing, eye blinking, neck stretching, sniffing, grimacing, shrugging, and grunting. Hyper- and hypoventilation syndromes are also possibilities.


CRAYONS

"Classic"Crayons

PHILADELPHIA, Oct. 2--This week Binney & Smith Inc., the manufacturer of Crayola crayons, brought back eight colors from retirement, albeit temporarily. Last year, with much hoopla, the company stopped using maize, violet blue, raw umber, orange yellow, blue gray, green blue, orange red and lemon yellow and replaced them with neon colors.
The crayons, now called "classics," will be packaged separately from a box of 64 in a special commemorative tin with notes about the crayons and their retirement, said J. O'Brien, a company spokesman.

"The response has been amazing," said Mr. O'Brien, speaking of the withdrawal of the eight. "You just have to watch an adult open that box and sniff it. It's like a time machine."

For some, opening the green and yellow box is like Proust discovering 64 madeleines. Mr. O'Brien, 32 years old, said he gets the feeling each morning in Binney & Smith's headquarters in Easton, Pa. As he opens his car door, he smells a billion crayons melting. Some who protested the withdrawal of the eight said it was like removing a favorite baby blanket or toy.

Thousands were heartened by the crayons' temporary comeback. Others were not.

"It's a limited victory," said Kenneth E. Lang, the founder of Rumps, the Raw Umber and Maize Preservation Society. Mr. Lang, from Locust Valley, L.I., said he felt cheated.

"Raw umber and maize represent a bygone time in America," he said. "You can't draw a picture of Nebraska or Kansas or South Dakota without using these colors."


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CREAMATOCRIT

Now that more than 50% of women are initially breastfeeding their infants, questions commonly arise about the adequacy of the milk being produced. Abnormalities in milk composition are quite rare and are overdiagnosed. The misinterpretation of a spot creamatocrit to determine the percentage of milk fat contributes to this overdiagnosis. The creamatocrit can be a reliable reflection of milk fat content when a 24-hour sample or an entire expressed feeding from one or both breasts is used and when the procedure has been standardized by comparison with gravimetric tests of milk fat content.

The creamatocrit takes advantage of the fact that fat is the major determinant of the energy value of the milk sample.

A sample of human milk is placed in a hematocrit tube and spun in a microcentrifuge at full speed for 15 minutes. The fat rises to the top of the column. The cream layer, easily visible, is read from the hematocrit capillary tube, and, like a hematocrit, is expressed as a percentage of the milk column in the tube. This is the "creamatocrit."

This number can then be employed in a formula which will provide you with the energy content of the milk expressed as kcal per liter. The formula is:

\[ \text{kcal/liter} = 290 + 66.8 \times \text{creamatocrit} \]

For example: A human milk sample is found to have a creamatocrit of 5%. Its caloric value is:

\[ 290 + 66.8 \times 5 \]

or

625 calories per liter.
Note: Creamatocrits should be read within one hour of centrifugation, because after that time the cream column begins to "unpack," and falsely elevated values are obtained.


**CRYING**

_The Crying Infant_

Unexplained, excessive crying in the afebrile infant usually achieves just what it is meant to achieve: every adult around wants to find its cause and make it stop. In investigating the cause of excessive, prolonged crying in 56 infants, aged 4 days to 245 months, who were brought to the emergency room of the Children's Hospital of Denver, a reason for the infant's distress was found for 46. The history provided a clue to the cause for 11 of the 56 infants, a careful physical examination resulted in a diagnosis for 23 patients, and a variety of laboratory tests revealed the cause for 11. The final diagnosis included a broad array of conditions, of which 61% were considered serious. The table below lists the diagnoses that explained the reason for excessive crying for these patients.

### Diagnosis in 56 Infants with Unexplained, Excessive Crying

<table>
<thead>
<tr>
<th>Diagnosis Description</th>
<th>No. with Diagnosis</th>
<th>Diagnosis</th>
<th>No. with Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic</td>
<td>10</td>
<td>Gastrointestinal tract</td>
<td></td>
</tr>
<tr>
<td>Colic</td>
<td>6</td>
<td>Constipation*</td>
<td>3</td>
</tr>
<tr>
<td>Infectious causes</td>
<td></td>
<td>Intussusception*</td>
<td>1</td>
</tr>
<tr>
<td>Otitis media*</td>
<td>10</td>
<td>Gastroesophageal reflux* with esophagitis*</td>
<td>1</td>
</tr>
<tr>
<td>Viral illness with anorexia, dehydration*</td>
<td>2</td>
<td>Central nervous system</td>
<td></td>
</tr>
<tr>
<td>Urinary tract infection*</td>
<td>1</td>
<td>Subdural hematoma*</td>
<td>1</td>
</tr>
<tr>
<td>Mild prodrome of gastroenteritis</td>
<td>1</td>
<td>Encephalitis*</td>
<td>1</td>
</tr>
<tr>
<td>Herpangina*</td>
<td>1</td>
<td>Pseudotumor cerebri*</td>
<td>1</td>
</tr>
<tr>
<td>Herpes stomatitis*</td>
<td>1</td>
<td>Drug reaction: overdose*</td>
<td>1</td>
</tr>
<tr>
<td>Trauma</td>
<td></td>
<td>Inadvertent pseudoephedrine overdose*</td>
<td>1</td>
</tr>
<tr>
<td>Corneal abrasion*</td>
<td>3</td>
<td>Behavior</td>
<td></td>
</tr>
<tr>
<td>Foreign body in eye*</td>
<td>1</td>
<td>Night terrors</td>
<td>1</td>
</tr>
<tr>
<td>Foreign body in oropharynx*</td>
<td>1</td>
<td>Overstimulation</td>
<td>1</td>
</tr>
<tr>
<td>Tibial fracture*</td>
<td>1</td>
<td>Cardiovascular</td>
<td></td>
</tr>
<tr>
<td>Clavicular fracture*</td>
<td>1</td>
<td>Supraventricular tachycardia*</td>
<td>2</td>
</tr>
<tr>
<td>Brown recluse spider bite*</td>
<td>1</td>
<td>Metabolic</td>
<td></td>
</tr>
<tr>
<td>Hair tourniquet syndrome (toe)*</td>
<td>1</td>
<td>Glutaric aciduria, type I*</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>56</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* Indicates condition considered serious.

1. Diphtheria-tetanus-pertussis vaccine.
The components of the physical examination that were important in providing the diagnosis are listed below.

<table>
<thead>
<tr>
<th>COMPONENT OF PHYSICAL EXAM</th>
<th>NO. OF PATIENTS WHOH COMPONENT PROVED USEFUL (n = 30)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Otoscopy</td>
<td>10</td>
</tr>
<tr>
<td>Rectal exam</td>
<td>4</td>
</tr>
<tr>
<td>Fluorescein staining of cornea</td>
<td>3</td>
</tr>
<tr>
<td>Inspection underneath clothing</td>
<td>2</td>
</tr>
<tr>
<td>Palpation of bones</td>
<td>2</td>
</tr>
<tr>
<td>Oral exam</td>
<td>2</td>
</tr>
<tr>
<td>Auscultation of heart (tachyarrhythmia)</td>
<td>2</td>
</tr>
<tr>
<td>Laryngoscopic exam of hypopharynx</td>
<td>1</td>
</tr>
<tr>
<td>Eversion of eyelid</td>
<td>1</td>
</tr>
<tr>
<td>Palpation of anterior fontanelle</td>
<td>1</td>
</tr>
<tr>
<td>Retinal exam</td>
<td>1</td>
</tr>
<tr>
<td>Neurologic exam</td>
<td>1</td>
</tr>
</tbody>
</table>

Laboratory studies that identified the diagnosis are listed in the following table.

<table>
<thead>
<tr>
<th>DIAGNOSTIC STUDY</th>
<th>NO. OF PATIENTS (n = 11)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Skeletal roentgenography</td>
<td>2</td>
</tr>
<tr>
<td>Lumbar puncture cerebrospinal fluid analysis</td>
<td>2*</td>
</tr>
<tr>
<td>Electrocardiography</td>
<td>2</td>
</tr>
<tr>
<td>Computed tomographic scan of the head</td>
<td>2*</td>
</tr>
<tr>
<td>Barium enema</td>
<td>1</td>
</tr>
<tr>
<td>Esophagram</td>
<td>1</td>
</tr>
<tr>
<td>Amino and organic acid studies</td>
<td>1</td>
</tr>
<tr>
<td>Urinalysis</td>
<td>1</td>
</tr>
</tbody>
</table>

* One patient, with pseudotumor cerebri, required lumbar puncture and computed tomographic scan of the head to make the diagnosis.


Colic, or Excessive Crying, in Young Infants

Colic, from the Latin *colicus*, strictly means related to or associated with the colon (and was originally an adjective). With respect to excessive crying in young infants, it has a poorly defined meaning that is connected to crying substantially more than the mean amount for age in an infant under 3 months. Usually this is thought to be a result of an intestinal (or "colic") disorder of paroxysmal pattern.
**Normal Crying Time**

In a 1962 study of 80 middle-class infants, Brazelton found the normal crying times, usually concentrated in the evening, to be as follows:

- 2 hr/day at 2 weeks of age
- 3 hr/day at 6 weeks of age
- 1 hr/day at 3 months of age

The clinical pattern of infant colic (also called paroxysmal fussing, infantile colic, evening colic, and 3-month colic) is well described:

1. Attacks occur suddenly, usually in the evening.
2. They are characterized by a loud, almost continuous cry.
3. They last several hours.
4. The face of the infant is flushed, with occasional circumoral pallor.
5. The abdomen is distended and tense.
6. Legs are drawn up on the abdomen and the feet often cold. Legs may extend periodically during forceful cries.
7. The fingers are clenched.
8. Relief is often noted from passage of flatus or feces.
9. The attack is not quelled for long by feeding, even though the infant may appear hungry and eats normally.
10. The attack usually terminates from apparent exhaustion.

No consistent etiology has been identified.


**Inconsolable Crying**

Nothing is as troublesome both to parent and physician as an infant who cries inconsolably. Diagnoses to consider when confronted with an infant who cries continuously, particularly if it is shrill or high-pitched, include:

- A corneal abrasion
- An eyelash, or other foreign body, in the eye
- Glaucoma
- Colic and intussusception
- Shaken baby syndrome
- Meningitis
- Fractures
- A DPT reaction
- An open diaper pin in the skin
- Strand of hair wrapped around finger or penis

CYANOSIS

Common Causes

Acrocyanosis (especially cold stress)
Apnea of prematurity
Aspiration
- Direct (swallowing disorders, neuromuscular disease)
- Indirect (gastroesophageal reflux, emesis)
Atelectasis
Breath holding
Bronchiolitis
Congenital heart disease
- Decreased pulmonary blood flow (no pulmonary hypertension)
  - Anomalous systemic venous return
  - Ebstein's anomaly
  - Hypoplastic right ventricle
  - Pulmonary stenosis atresia
  - Tetralogy of Fallot
  - Tricuspid stenosis/atrophia insufficient
  - Eisenmenger's syndrome
  - Increased pulmonary blood flow
  - Arteriovenous (AV) canal
  - Coarctation (postductal)
  - Hypoplastic left heart
  - Total anomalous pulmonary venous return (TAPVR)

Congenital heart disease (Cont.)
  - Increased pulmonary blood flow (Cont.)
    - Transposition
    - Truncus arteriosus
    - Ventricular septal defect (VSD), large

Pump failure
  - Aortic stenosis (severe)
  - Coarctation (postductal)
  - Patent ductus arteriosus (PDA)
  - VSD

Croup
Crying
Drugs -- respiratory depressants (e.g., narcotics, benzodiazepines)
Hyaline membrane disease
Mucous plug
Nasal obstruction
Pneumonia
Pulmonary edema
Reactive airway disease
Seizures
Sepsis
Sleep apnea (tonsillar adenoidal hypertrophy)

Uncommon Causes

Abdominal distention
Arterial thrombosis
Bronchopulmonary dysplasia
Chest wall abnormalities
  - Congenital bone/cartilage abnormalities
    - Pectus
    - Flail chest
  - Cystic fibrosis
  - Epiglottitis
  - Foreign body
  - Hypovolemia
  - Mediastinal mass
  - Persistent fetal circulation
  - Pickwickian syndrome

Pleural effusion
Pneumothorax
Polycythemia
Pulmonary hemorrhage
Retropharyngeal/peritonsillar abscess
Scoliosis
Tracheal compression
  - Abscess
  - Adenopathy
  - Hemorrhage
  - Tumor
  - Vascular ring
Tracheobronchomalacia, stenosis
Venous stasis
Rare Causes

Angioedema
Bronchogenic cyst
Central nervous system disease
- Edema
- Hemorrhage
- Infection
- Trauma
Chylothorax
Diaphragmatic hernia
Factitious (blue paint/dyes; makeup)
Glossoptosis
Hemoglobinopathy
- (M, low oxygen affinity)
Hypoplastic lungs
Laryngeal web
Lobar emphysema
Methemoglobinemia
- Methemoglobin reductase deficiency
Oxidant stress
- Acetophenetidin
- Antimalarials
- Benzocaine
- Crayons
- Disinfectants
- EDTA
- Hydralazine
- Marking dyes
- Naphthalene
- Nitrites
- Amyl; butyl nitrate
- Nitrate-contaminated well water
- Nitrate food additive
- Nitroglycerin
- Plant nitrates (e.g., carrots grown in contaminated soil)

Methemoglobinemia (Cont.)
- Oxidant stress (Cont.)
- Nitroprusside
- Prilocaine
- Pyridium
- Sulfonamides
- Vitamin K analogs

Ondine's curse
Primary pulmonary hypertension
Pulmonary AV malformation/fistula
Pulmonary embolism/thrombosis
Pulmonary hemosiderosis
Pulmonary sequestration
Pulmonary tumor (primary or metastatic)
Respiratory muscle dysfunction
- Botulism
- Muscular dystrophy
- Myasthenia gravis
- Neuromuscular blockade
- Phrenic nerve damage
- Werdnig-Hoffmann disease
Superior vena cava (SVC) syndrome
Tracheoesophageal fistula
Tumor
Vocal cord paralysis

Cystic Fibrosis

The Thirty Faces of Cystic Fibrosis

Cystic fibrosis is the great imitator. It may first present in utero with a picture of meconium peritonitis or its first manifestation may be sterility in the adult male. Listed below are 30 ways the disease may first manifest itself. Be suspicious and perform a sweat test when these problems are encountered without a plausible alternative explanation.
Meconium ileus and meconium peritonitis
Pancreatic insufficiency and growth failure
Recurrent pulmonary infections
Intestinal impaction and obstruction
Hypoproteinemia in infancy, with edema and anemia
Rectal prolapse
Cholestatic jaundice in neonates
Cirrhosis of the liver
Portal hypertension
Glucose intolerance
Diabetes
Acute or recurrent pancreatitis
Vitamin K deficiency and bleeding
Vitamin A deficiency
Vitamin E deficiency with neurologic abnormalities
Night cramps
Lactase deficiency
Duodenal ulcer
Cholelithiasis and cholecystitis
Chronic obstructive airway disease
Cor pulmonale
Recurrent episodes of asthma
Hypertrophic pulmonary osteoarthropathy
Nasal polyps
Optic neuritis
Salty taste of infant noted by the mother
Hyponatremic dehydration in warm weather
Hypochloremia metabolic alkalosis
Heat stroke
Infertility in males

CYSTITIS

Hemicystitis: Think Zoster!

There exists a fascinating clinical entity known as hemicystitis, which is an infection limited to one half of the bladder. It occurs with herpes zoster infection of the bladder in which vesicles are unilateral, conforming to the affected nerve supply.


CYTOMEGALOVIRUS

Cytomegalovirus Infection in the Newborn

Cytomegalovirus (CMV) is the most common herpes infection that occurs during the neonatal period. Exposure to CMV can occur from a congenital source (e.g., either a primary infection in a seronegative mother or reactivation of latent virus in a seropositive mother), a natal source such as vaginal delivery in a mother shedding virus from the cervix, or postnatal sources (e.g., breast milk from a seropositive mother, blood transfusion from a seropositive donor, or close contact with individuals actively shedding CMV).

CMV is especially dangerous to the developing fetus when the pregnant woman is experiencing primary infection; in such cases there is a 30% mortality rate among those CMV-infected infants who were symptomatic at birth. Surviving infants who were symptomatic at birth encounter the following sequelae: microcephaly (70%), moderate-to-severe mental retardation (61%).
hearing loss (30%), and chorioretinitis (22%). Symptomatic infants without neurologic signs at birth remain at risk for such sequelae as failure to thrive, microcephaly, spastic quadriplegia, and deafness during the first year of life. Asymptomatic infants of mothers experiencing a primary CMV infection are at risk for hearing defects and moderate-to-severe brain damage. Conversely, infants and fetuses who become infected with CMV as a result of recurrent viremia or perinatal exposure to a seropositive mother are generally protected from symptomatic infection of any kind. A minority of such infants, however, may be at risk for hearing defects and learning problems.

Clinical Manifestations of CMV Infection
(among infants born to previously seronegative mothers)

Prenatal Infection
Hepatomegaly
Hyperbilirubinemia
Petechiae/thrombocytopenia
Microcephaly
Hydrocephalus
Periventricular calcifications

Chorioretinitis
Optic atrophy
Strabismus
Microphthalmia
Cataracts
Deafness

Infection During Vaginal Delivery
Protracted pneumonitis

Postnatal Infection
Healthy infants and children who acquire CMV infection suffer no permanent sequelae and usually remain entirely asymptomatic. Symptomatic infection does occur rarely, however, and may include the following:

Fever
Hepatomegaly
Pneumonia

DAY CARE

What You Ought to Know About Your Day Care Center

With more and more parents relying on day care centers, clinicians need to be knowledgeable about what to look for in selecting such a center. One source of information both for parents and clinicians is the National Association for Child Care Resource and Referral Agency (NACCRRA), 2116 Campus Drive SE, Rochester, MN 55904 (507) 287-2220. This clearinghouse advises parents and others on how to select a safe and reliable day care center.

Clinicians should urge parents to visit their day care center frequently (and unannounced) in order to make sure their children are cared for in a loving and positive manner.

Parents also need to observe their children closely for behavioral changes such as depression, aggressiveness, fear of the day care center, and signs of physical abuse (e.g., bruises, abrasions) or neglect (e.g., diaper rash, bald spots on the back of the head indicating that the infant has been left supine all day, extreme hunger, etc.).

In the case of older toddlers who are left in day care, parents should ask the child what activities are offered during the day. Are constructive games and teaching employed or is the child merely parked in front of a television set?


Pathogens Transmitted in Day Care Centers

Approximately 11 million children under the age of 6 years spend all or part of their day in a day care center. Some experts have predicted that by the year 2000, 80% of all American mothers will be a part of the nation's workforce. The result, of course, is an ever-increasing reliance upon day care. One of the most frequent questions that comes up regarding day care has to do with parents worrying about the risk of infections in the day care setting. The table below summarizes the most common pathogens you need to be aware of:

Pathogens Transmitted in Day Care Centers*

<table>
<thead>
<tr>
<th>MODE OF TRANSMISSIONS</th>
<th>BACTERIA</th>
<th>VIRUSES</th>
<th>PARASITES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Direct</td>
<td>Group A streptococci</td>
<td>Herpes simplex</td>
<td>Pediculosis</td>
</tr>
<tr>
<td></td>
<td><em>Staphylococcus aureus</em></td>
<td>Herpes zoster</td>
<td>Scabies</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Pathogens Transmitted in Day Care Centers* (Cont.)

<table>
<thead>
<tr>
<th>MODE OF TRANSMISSIONS</th>
<th>BACTERIA</th>
<th>VIRUSES</th>
<th>PARASITES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Respiratory</td>
<td><em>Haemophilus influenzae</em>&lt;br&gt;<em>Neisseria meningitidis</em>&lt;br&gt;<em>Bordetella pertussis</em>&lt;br&gt;<em>Mycobacterium tuberculosis</em></td>
<td>Adenovirus&lt;br&gt;Coxsackie A16 (hand-foot-mouth disease)&lt;br&gt;Epstein-Barr virus&lt;br&gt;HHV6 (roseola)&lt;br&gt;Influenza&lt;br&gt;Measles&lt;br&gt;Mumps&lt;br&gt;Parainfluenza&lt;br&gt;Parvovirus B19 (fifth disease)&lt;br&gt;Respiratory syncytial virus&lt;br&gt;Rhinovirus&lt;br&gt;Rubella&lt;br&gt;Varicella</td>
<td></td>
</tr>
<tr>
<td>Fecal-oral</td>
<td><em>Campylobacter spp</em>&lt;br&gt;<em>Escherichia coli</em>&lt;br&gt;<em>Salmonella</em>&lt;br&gt;<em>Shigella</em>&lt;br&gt;<em>Yersinia</em></td>
<td>Enteroviruses&lt;br&gt;Hepatitis A&lt;br&gt;Rotavirus</td>
<td><em>Cryptosporidium</em>&lt;br&gt;<em>Entamoeba histolytica</em>&lt;br&gt;<em>Giardia lamblia</em>&lt;br&gt;<em>Hymenolepis nana</em> (dwarf tapeworm)&lt;br&gt;Pinworms</td>
</tr>
</tbody>
</table>

Contact with infected blood and secretions (urine, saliva)


* To date no reported cases of HIV infection are known to have resulted from transmission in day care centers.


### DEHYDRATION

**Predictable Fall of the Blood Urea Nitrogen (BUN) in a Dehydrated Child**

Is the increased BUN in a child with diarrhea and vomiting always the result of simple dehydration or can it reflect the presence of associated renal disease? Brill and coworkers found that the rate of fall of BUN in a dehydrated child with normal renal function was predictable. They plotted BUN levels against time on semilogarithmic graph paper. BUN had fallen to one-half the admission level in 24 hours or less in all children with uncomplicated dehydration and diarrhea.

Line A in the accompanying figure represents the slope along which the BUN should fall in a child without renal disease or excess nitrogen load (e.g., gastrointestinal bleed). Lines B and C represent 2½ standard deviations on either side of that rate of fall.
Complicating disease should be investigated in the dehydrated child whose BUN does not fall at a rate parallel to line A or within 2½ standard deviations from that rate.


**DERMATOLOGY**

**Skin Lesions**

Can you speak “dermatologese”? Or is it “dermaterminology”? In any event, understanding dermatology is impossible without a working knowledge of the sometimes exotic vocabulary of the specialty. The following list defines some (but by no means all) of the more commonly used terms for skin lesions and related structures and conditions.

Abscess A localized accumulation of purulent material so deep in the dermis or subcutaneous tissue that pus is usually not visible on the surface of the skin.

Atrophy An acquired loss of underlying tissue causing skin depression with intact epidermis.

Bulla A relatively large vesicle (diameter 0.5 cm).

Carbuncle Coalescence of several furuncles (see below).

Comedo (pl., comedones) A greasy plug in a sebaceous follicle capped by a layer of melanin, hence its black appearance (blackhead).

Crust Dried exudate of body fluids (serous and or hemorrhagic).

Cyst A sac that contains liquid or semisolid material.
Dermatology

Erosion--A superficial deficit of epithelium.
Erythema--Increased redness of skin from capillary dilatation.
Excioriations--Linear, angular erosions caused usually by scratching.
Furuncle--A deep necrotizing form of folliculitis with pus accumulation.
Figurate lesions--Lesions forming rings and arcs, usually erythematous.
Hyperpigmentation--Excessive pigmentation of any origin.
Hypopigmentation--Loss of pigmentation of any origin.
Keratosis--Benign horny lesion (also called keratoma).
Lichenification--A proliferation of keratinocytes and stratum corneum forming a plaque-like structure. The skin appears thickened, and the skin markings appear accentuated. The process results from repeated rubbing.
Macule--A circumscribed area of change (less than 2 cm in diameter) in normal skin color without elevation or depression of the surface in relation to the surrounding skin.
Milia--Small, firm, white papules filled with keratin.
Nodule--A palpable, solid, round, or ellipsoid lesion; it can be located in the epidermis or extend into the dermis or subcutaneous tissue.
Papule--A solid elevated lesion generally understood to be less than 1 cm in diameter.
Patch--A large, flat lesion (greater than 2 cm in diameter) with color different from surrounding skin. Differs from macule only in size.
Plaque--Elevation above the skin surface that occupies a relatively large surface area in comparison with its height above the skin.
Pustule--A circumscribed elevation of skin that contains a purulent exudate. (Follicular pustules are conical and usually contain a hair in the center.)
Rash--An inflammatory skin eruption.
Scale--A thin, platelike, external layer of horny epidermis.
Scar--Change in skin character--a mark secondary to trauma or inflammation.
Sclerosis--Circumscribed or diffuse hardening or induration in the skin.
Tag--Small, sessile protuberance of skin.
Telangiectases--Permanent dilatations of blood capillaries that may or may not disappear with the pressure of a glass slide.
Tumor--A firm, solid, raised growth greater than 5 cm in diameter.
Ulcer--A deep, local deficit or excavation of skin and underlying tissue.
Vesicle--A small (less than 0.5 cm in diameter) fluid-filled lesion. A "dew-drop."
Wart--A benign keratotic tumor.
Wheal--A rounded or flat-topped elevation in the skin that is characteristically evanescent, disappearing within hours. Lesions are the result of edema in the upper layers of the dermis.

Dermatologic Manifestations of Viral and Bacterial Infections

There are a number of cutaneous manifestations associated with bacterial and viral infections common among children. The specific skin lesions that appear are usually the result of various pathways involved in inflammation and necrosis, such as the complement cascade, localized or generalized Schwartzman reactions, factors that yield hypotension and disseminated intravascular coagulation, and a host of yet undiscovered factors. The table lists the dermatologic manifestations of common pediatric bacterial and viral infections and should be useful in the bedside evaluation of the patient presenting with "fever and a rash."

## Differential Diagnosis of Pediatric Infections with Dermatologic Manifestations

<table>
<thead>
<tr>
<th>Clinical Entity</th>
<th>CAUSATIVE AGENT</th>
<th>AGE</th>
<th>CLINICAL SYNDROMES</th>
<th>TYPE OF RASH</th>
<th>DISTRIBUTION</th>
<th>SIMILAR ENTITIES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Roseola infantum (exanthem subitum)</td>
<td>Herpesvirus-6*</td>
<td>6 months - 4 years</td>
<td>Fever, irritability; rapid lysis of fever with appearance of rash</td>
<td>Discrete macular or papular rash</td>
<td>Trunk with extension to neck, extremities, face</td>
<td>Scarlet fever, Rubella</td>
</tr>
<tr>
<td>Frythema infectious (fifth disease)</td>
<td>Parvovirus B-19</td>
<td>School-age children, infants, adults less common</td>
<td>Flu-like illness</td>
<td>Bilateral erythema of cheeks: &quot;slapped cheeks&quot;</td>
<td>Face, trunk, extremities, palms soles spared</td>
<td>Scarlet fever, Rubella</td>
</tr>
<tr>
<td>Measles</td>
<td>Measles virus</td>
<td>All ages</td>
<td>Fever, cough; conjunctis</td>
<td>Flu-like illness</td>
<td>Silent; Starts on face, moves downward</td>
<td>Enteroviral infection, Mycoplasma, Drug eruption</td>
</tr>
<tr>
<td>Hand-foot-and-mouth disease</td>
<td><em>Primary:</em> Coxsackie A viruses</td>
<td>&lt;10 years</td>
<td>Fever, anorexia; oral pain</td>
<td>Oral: discrete, ulcerative</td>
<td>Anterior mouth, hands, feet; occasionally trunk, face</td>
<td>Aphthous stomatitis, Varicella, Herpes simplex</td>
</tr>
<tr>
<td></td>
<td><em>Secondary:</em> Coxsackie B viruses; enterovirus 71</td>
<td></td>
<td></td>
<td>Skin: maculopapular, vesicular</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Varicella</td>
<td>Varicella-zoster virus</td>
<td>90% of cases &lt; 15 years</td>
<td>Fever, pruritus; malaise</td>
<td>Maculopapular, then vesicles on erythematous base, which rupture</td>
<td>Diffuse, includes scalp, oral mucosa</td>
<td>Insect bites, Herpes simplex</td>
</tr>
<tr>
<td>Periorbital buccal cellulitis</td>
<td><em>Primary:</em> H influenza type B</td>
<td>3 - 36 months</td>
<td>Fever, hætemeria</td>
<td>Unilateral indurated cellulitis</td>
<td>Periorbital, cheek</td>
<td>Orbital cellulitis, Parotitis</td>
</tr>
<tr>
<td></td>
<td><em>Secondary:</em> S. pneumonae, S. aureus, β-hemolytic streptococci</td>
<td></td>
<td></td>
<td>Indistinct borders, Violaceous hue</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Staphylococcal scalded-skin syndrome</td>
<td>S. aureus</td>
<td>Infants</td>
<td>Fever, irritability; septicemia (rare); eye, nasal discharge</td>
<td>Tender, diffuse erythematous rash progressing to bullae</td>
<td>Diffuse</td>
<td>Bullous impetigo, E. multiforme, Toxic epidermal necrolysis, Pemphigus, Epidermolysis bullosa, Kawasaki disease</td>
</tr>
</tbody>
</table>

Seborrheic Dermatitis or Atopic Dermatitis?

Many clinicians have difficulty distinguishing these two common entities. Seborrhea is the excretion by the sebaceous glands of abnormally copious amounts of grease-like sebum. There is usually no underlying disorder. Atopy is a form of immediate hypersensitivity reaction to certain common allergens that produce the IgE antibody, reagin, and atopic dermatitis is the dermal manifestation of the allergic reaction. The following table should help you further to differentiate between the two conditions.

<table>
<thead>
<tr>
<th>Seborrheic Dermatitis vs. Atopic Dermatitis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>SEBORRHEIC DERMATITIS</strong></td>
</tr>
<tr>
<td>Family history of allergy</td>
</tr>
<tr>
<td>Character of individual lesions</td>
</tr>
<tr>
<td>Color of lesion</td>
</tr>
<tr>
<td>Feature of lesion</td>
</tr>
<tr>
<td>Vesicles</td>
</tr>
<tr>
<td>Weeping and edema</td>
</tr>
<tr>
<td>Lichenification</td>
</tr>
<tr>
<td>Pruritus</td>
</tr>
</tbody>
</table>


**DEVELOPMENT**

Developmental Delay: Seeking the Etiology

As we learn from medical school on, the physician's best diagnostic tools are the history and physical exam. This dictum certainly holds true in the work-up of developmental delay.

Regardless of the age of the child at presentation, there are several strategies that the pediatrician can employ in the search for an etiology. The What, When, and How are invariably asked. The answers will often guide the parents in the decision to have another child and how to come to terms with their child's disability. The following guidelines and accompanying table should prove quite useful when confronted with a developmentally delayed patient.
1. Analyze the family's pedigree with attention paid to physical appearance, "birth defects," inheritance patterns of any disease, disabilities or dysmorphisms, consanguinity, and parental age. When possible, obtain photographs of the extended family.

2. Obtain a thorough prenatal history. Look for:
   a. First-trimester febrile illnesses; possible teratogenic exposures, including alcohol, cigarette, and drug use.
   b. First through second trimesters: viral illnesses, maternal diabetes mellitus, maternal medications, cardiorespiratory disease, and maternal metabolic abnormalities.
   c. First through third trimesters: see table.

### Temporal Approach to the Etiology of Developmental Delay

<table>
<thead>
<tr>
<th>CLASSIFICATION</th>
<th>ETIOLOGIC PERIOD</th>
<th>TYPICAL CAUSES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Genetic (Mendelian)</td>
<td>Preconceptional</td>
<td>Autosomal dominant</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neurofibromatosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tuberculous sclerosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Autosomal recessive</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Phenylketonuria</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Galactosemia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Tay-Sachs disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sex-linked recessive</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sex-linked nonspecific mental retardation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lesch-Nyhan syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sex-linked dominant</td>
</tr>
<tr>
<td>Chromosomal</td>
<td>Preconception or early mitotic phase</td>
<td>Albright's hereditary osteodystrophy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Polysomy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Trisomy 21 (Down syndrome)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Autosomal deletion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cri du chat syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td>(deletion of short arm 5)</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sex chromosome aberrations</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Multiple-X syndromes</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Turner's syndrome</td>
</tr>
<tr>
<td>Multifactorial (genetic and environmental)</td>
<td>Preconception and first 12 weeks of gestation</td>
<td>Neural tube defects</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Meningomyelocele</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Encephalocele</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hydrocephaly</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cleft lip: cleft palate</td>
</tr>
<tr>
<td>Environmental</td>
<td>Prenatal</td>
<td>Teratogenic agents (suspected)</td>
</tr>
<tr>
<td></td>
<td>First trimester (period of CNS morphogenesis and neuroblast proliferation)</td>
<td>Phenytoin</td>
</tr>
<tr>
<td></td>
<td>Midpregnancy (period of neuroblast proliferation)</td>
<td>Maternal alcoholism</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intrauterine infection</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Maternal irradiation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Teratogens</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
Temporal Approach to the Etiology of Developmental Delay (Cont.)

<table>
<thead>
<tr>
<th>CLASSIFICATION</th>
<th>ETIOLOGIC PERIOD</th>
<th>TYPICAL CAUSES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Environmental</td>
<td>Prenatal (Cont.)</td>
<td>Intrauterine infection</td>
</tr>
<tr>
<td>(Cont.)</td>
<td>Midpregnancy to perinatal period (rapid brain growth, glial cell proliferation, myelinization, dendritic tree formation)</td>
<td>Preeclampsia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hemorrhage</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hormonal disturbances</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malnutrition</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prematurity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Teratogens</td>
</tr>
<tr>
<td></td>
<td>Perinatal</td>
<td>Intrapartum hemorrhage and anoxemia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Trauma:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Breech delivery</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Difficult delivery</td>
</tr>
<tr>
<td></td>
<td>Postnatal</td>
<td>Meningitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Encephalitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malnutrition</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lead poisoning</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Head injury</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Near drowning</td>
</tr>
</tbody>
</table>

3. Obtain a birth history. Approximately 30% of the cases of developmental delay can be traced to difficulties in the prenatal period. It is wise, however, to avoid the temptation to overinterpret problems in labor and delivery. As often as possible, a hospital record of the birth and nursery stay should be procured.

4. Continue your detective work with a postnatal history. In addition to the causes listed in the table, inquire about any known metabolic diseases and their treatment, diarrheal illnesses with possible hypernatremia, failure to thrive, history of seizures, loss of milestones, and apparent receptive or expressive delays.

5. Examination of the patient: The physical exam should be undertaken with a fine-toothed comb.
   a. Height, weight, and head circumference are invaluable in the setting of developmental delay. [Refer to “Diagnosing Dysmorphism” (p. 95) or Smith’s Recognizable Patterns of Human Malformation: Genetic, Embryologic and Clinical Aspects (Philadelphia, W.B. Saunders, 1988). Microcephaly, macrocephaly, growth retardation, and growth acceleration may steer you toward a recognized syndrome and/or an etiology.
   b. Transilluminate the head if hydrocephaly, hydranencephaly, porencephaly, or cerebral cortical atrophy are in your differential. You may obviate the need for a CT scan with this simple test.
   c. Pay attention to the eye exam (see “Leukocoria: A Differential Diagnosis” [p. 204]). Look for a cherry red spot, chorioretinitis, retinitis pigmentosa, or retrolental fibroplasia when examining the retina.
   d. Do not neglect to assess hearing. Unidentified hearing loss can lead to profound developmental delay.
   e. Proceed with the examination of the skin, facies, and body as if you were uncovering buried treasure. Remember that both major and minor abnormalities are more common in patients with developmental delay or frank mental retardation. Compile a list of your findings and consult the sources noted above, beginning with the least common abnormality.
6. Laboratory studies will often help secure a diagnosis, thus comforting the parents. They may also reveal chromosomal or metabolic abnormalities that will influence a couple's plans for other children. Consider pertinent laboratory studies, chromosomal analyses, viral cultures, computed tomography, and ultrasonography as you and the parents see the need.


The Draw-A-Person Test

Goodenough called her test the “Draw-a-man” test when she first introduced this superb simple screening test for intelligence. The name of the test has changed, but the test and its scoring have remained the same.

Basal age = 3 years. For each four criteria, add 1 year to arrive at mental age, between ages 3 and 10 years. Instruct child to draw a complete person; no further instructions.

\[
\frac{\text{Maturation age}}{\text{Chronological age}} \times \frac{100}{1} = \text{IQ}
\]

Twenty-eight criteria for scoring:
1. Head present
2. Legs present
3. Arms present
4. Trunk present
5. Length of trunk greater than breadth
6. Shoulder indicated
7. Both arms and legs attached to trunk
8. Legs attached to trunk and arms to trunk at correct point
9. Neck present
10. Outline of neck continuous with that of head or trunk, or both
11. Eyes present
12. Nose present
13. Mouth present
14. Both nose and mouth in two dimensions, two lips shown
15. Nostrils indicated
16. Hair shown
17. Hair on more than circumference of head, nontransparent, better than scribble
18. Clothing present
19. Two articles of clothing, nontransparent
20. Entire drawing, with sleeves and trousers shown, free from transparency
21. Four or more articles of clothing definitely indicated
22. Costume complete without incongruities
23. Fingers shown
24. Correct number of fingers shown
25. Fingers in two dimensions, length greater than breadth, angle subtended not greater than 180 degrees
26. Opposition of thumbs shown
27. Hand shown as distinct from fingers or arms
28. Arm joint shown; either elbow, shoulder, or both

Watching for Developmental Lags

Parents often ask, "When should I expect my child to do ______?" When should you, as a clinician, begin to worry about the possibility of developmental delay? The following table includes many of the warning signs indicative of abnormal patterns of development. In many cases, the presence or absence of any one sign may mean nothing if the rest of development is normal, but certain signs, in and of themselves, are very important (e.g., no social smile at age 6 months).

### Indications for Further Evaluation for Developmental Delay

#### At 3 mo.
- Does not react to sudden noises.
- Does not appear to listen to a speaker's voice.
- Does not try to find the speaker's face with his or her eyes.
- Has not begun to vocalize sounds.
- Has been left to lie in a crib for hours without visual or auditory stimulation.
- Does not raise the head when lying on the stomach.

#### At 6 mo.
- Does not turn to the speaking person.
- Does not respond to being played with.
- Is not visually alert.
- Never laughs or smiles.
- Is not babbling.
- Does not reach for or try to pick up a toy.
- Is not learning to sit up.
- Does not appear to be gaining weight.
- Does not arched the back when lying on the stomach and raising the head.

#### At 1 yr.
- Has not been responding to "Pat-a-Cake," "Peek-A-Boo," or other baby games.
- Is not imitating a variety of speech sounds.
- Is not saying two or three words such as "bye-bye, mama, dada."
- Is not pulling up to a standing position.

#### At 18 mo.
- Is not yet beginning to feed itself with a spoon.
- Does not imitate speech or vocalize in jargon.
- Is not moving about to explore.
- Does not give eye contact.
- Has not or does not spontaneously squat when picking up objects.

#### At 2 yrs.
- Is not naming a few familiar objects and using a few two- or three-word phrases.
- Is not noticing animals, cars, trucks, trains.
- Is not beginning to play symbolically with housekeeping toys, little cars.
- Is not moving about vigorously, running, climbing, exploring.
- Avoids eye contact.
- Does not seem to focus eyes on a large picture.
- Engages in rocking or head banging for extensive periods of time.
- Is not walking up stairs.

#### At 3 yrs.
- Does not seem aware of other children, of adults, of the weather, traffic, and so forth.
- Uses little or no speech.
- Does not engage in imitative play symbolic of adult activities.
- Avoids looking at pictures or pointing to pictures of familiar objects
- Does not follow simple directions.

*Table continued on next page.*
### Indications for Further Evaluation for Developmental Delay (Cont.)

<table>
<thead>
<tr>
<th>At 3 yrs. (Cont.)</th>
<th>Engages for long periods of time in repetitive behaviors like flipping pages of a magazine, or spinning a wheel on a little truck, head banging, and so forth. Cannot ride a tricycle if given plenty of opportunity to do so.</th>
</tr>
</thead>
<tbody>
<tr>
<td>At 4 yrs.</td>
<td>Does not have at least partially understandable speech with sentences. Uses echolalic speech or frequent, bizarre, meaningless sounds. Does not seem interested in listening to a simple story about his or her experiences. Repeatedly tests all limits. Is so quiet and conforming that he or she never tests or tries anything new. Has pronounced fears and phobias. Frequently engages in flapping of the arms or flapping of the hands to express excitement. Runs about from one thing to another every minute or so without getting fully involved in an activity. Is still untrained in toileting (occasional slips do occur at this age). Does not draw some sort of representation of human beings (at least a head and a few features), if crayons or pencils have been available to the child. Stays on the periphery of the playroom, paying no attention to other children for some weeks, after most children have overcome shyness and begun to play with or near other children. Avoids eye contact. Engages in head banging or rocking. Cannot tolerate change or frustration without frequent 2-year-old tantrums.</td>
</tr>
</tbody>
</table>


### DIABETES

#### Classification and Etiology of Diabetes Mellitus

**Classification of Diabetes Mellitus**

<table>
<thead>
<tr>
<th>IDIOPATHIC</th>
<th>SECONDARY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insulin-dependent (type I)</td>
<td>Pancreatic trauma, disease, or resection</td>
</tr>
<tr>
<td>Non-insulin dependent (type II)</td>
<td>Hormone-induced</td>
</tr>
<tr>
<td>Maturity onset diabetes of youth (MODY)</td>
<td>Drugs and chemical agents</td>
</tr>
<tr>
<td></td>
<td>Genetic syndromes</td>
</tr>
<tr>
<td></td>
<td>Insulin receptor abnormalities</td>
</tr>
<tr>
<td></td>
<td>Other types</td>
</tr>
</tbody>
</table>


Although the majority of cases of pediatric diabetes mellitus will be of the type 1 classification, the remaining idiopathic and secondary etiologies cannot be overlooked. Some clues in establishing a diagnosis between type I and type II are provided in the following table:
**Insulin-Dependent vs. Non-Insulin-Dependent Diabetes**

<table>
<thead>
<tr>
<th></th>
<th>Insulin-Dependent</th>
<th>Non-Insulin-Dependent</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Association with HLA</td>
<td>2.5 x expected</td>
<td>Same frequency as</td>
</tr>
<tr>
<td>B8/D3 or HLA B15/D4</td>
<td>frequency</td>
<td>normal population</td>
</tr>
<tr>
<td>2. Pancreatic insulin content</td>
<td>0</td>
<td>&gt;50% of normal</td>
</tr>
<tr>
<td>3. Anti-islet antibodies</td>
<td>85%</td>
<td>&lt;5%</td>
</tr>
<tr>
<td>4. Primary insulin resistance</td>
<td>Minimal</td>
<td>Marked</td>
</tr>
<tr>
<td>5. Concordance rate of identical twins for diabetes mellitus</td>
<td>25 to 50%</td>
<td>~100%</td>
</tr>
</tbody>
</table>


The secondary causes of diabetes mellitus are too numerous to cover adequately in this text. There are, however, a number of hormonal and chemical causes that deserve mention, principally because treatment of the primary disorder in hormonal abnormalities and removal of the offending agent in chemically induced diabetes mellitus frequently reverse the disease process.

**Hormonally Induced Diabetes Mellitus**

<table>
<thead>
<tr>
<th>Abnormality</th>
<th>Prevalence (where known)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acromegaly</td>
<td>20%</td>
</tr>
<tr>
<td>Cushing's syndrome</td>
<td>20%</td>
</tr>
<tr>
<td>Primary aldosteronism</td>
<td></td>
</tr>
<tr>
<td>Pheochromocytomas</td>
<td></td>
</tr>
<tr>
<td>Glucagonoma</td>
<td></td>
</tr>
</tbody>
</table>

**Diabeticogenic Drugs**

<table>
<thead>
<tr>
<th>Diuretics and Antihypertensives</th>
<th>Psychoactive Agents</th>
<th>Hormonally Active Agents</th>
<th>Active Agents</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chlorthalidone</td>
<td>Chlorprothixene</td>
<td>ACTH</td>
<td>Diphenhydantoin</td>
</tr>
<tr>
<td>Clonidine</td>
<td>Haloperidol</td>
<td>Glucagon</td>
<td>Epinephrine</td>
</tr>
<tr>
<td>Diazoxide</td>
<td>Lithium carbonate</td>
<td>Glucocorticoids</td>
<td>Isoproterenol</td>
</tr>
<tr>
<td>Furosemide</td>
<td>Phenothiazines</td>
<td>Oral contraceptives</td>
<td>Levodopa</td>
</tr>
<tr>
<td>Metalazone</td>
<td>Tricyclic antidepressants</td>
<td>Growth hormones</td>
<td>Norepinephrine</td>
</tr>
<tr>
<td>Thiazides</td>
<td>Catecholamine and Other Neurologically</td>
<td>Thyroid hormones (thyrotoxic doses)</td>
<td></td>
</tr>
</tbody>
</table>


**Diagnosis of Diabetes Mellitus**

In the presence of symptoms, the diagnosis of diabetes mellitus is an uncomplicated task: a child presenting with polydipsia, polyuria, polyphagia, and weight loss with an accompanying elevation of blood glucose and/or ketonemia leads the
pediatrician to a rapid answer. In the absence of symptoms or the presence of mild symptoms, however, the diagnosis of diabetes mellitus is much more difficult. In children, the diagnostic criteria for diabetes mellitus are as follows:

1. Presence of symptoms of diabetes, such as polydipsia, polyuria, ketonuria, and weight loss, together with a random plasma glucose of 200 mg/dl

or

2. In asymptomatic children, both an elevated fasting glucose concentration and a sustained elevated glucose concentration during an oral glucose tolerance test (1.75 g/kg up to maximum of 75 g) on two or more occasions.

Fasting value: 2-hour OGTT value and an intervening value:

<table>
<thead>
<tr>
<th>Maintenance Requirements*</th>
<th>Losses*</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Water</strong></td>
<td>1500 ml/m²</td>
</tr>
<tr>
<td><strong>Sodium</strong></td>
<td>45 mEq/m²</td>
</tr>
<tr>
<td><strong>Potassium</strong></td>
<td>35 mEq/m²</td>
</tr>
<tr>
<td><strong>Chloride</strong></td>
<td>30 mEq/m²</td>
</tr>
<tr>
<td><strong>Phosphate</strong></td>
<td>~10 mEq/m²</td>
</tr>
</tbody>
</table>

* Maintenance is expressed in surface area to permit uniformity because fluid requirements change as weight increases.

Diabetic Ketoacidosis

Despite continued advances in control of the diabetic child, diabetic ketoacidosis remains an acute medical emergency. In the known and presenting diabetic child, ketoacidosis is defined as “hyperglycemia with a blood glucose exceeding 300 mg/dl, ketonemia with total ketones (β hydroxybutyrate and acetoacetate) in serum exceeding 3 mmol/L or positive at a 1:2 dilution in serum or undiluted urine with the sodium nitroprusside reaction (Acetest; Ketostix; Chemstrip UGK), and acidosi s with pH reduced to less than 7.30 or reduced serum bicarbonate to less than 15 mEq/L.” The maintenance requirements for fluid and electrolyte therapy for diabetic ketoacidosis are outlined below. The clinician should keep in mind that these values represent averages and that the extent of dehydration and electrolyte imbalances vary with the duration of symptoms, the possible presence of vomiting, and prior insulin administration.


Glycosylated Hemoglobin Assay

The glycosylated hemoglobin assay provides the clinician with a profile of glycemia during the previous 60 to 120 days. This is helpful in ascertaining inter-visit glycemia control and to determine the relationship between complications and compliance. The issue of compliance is particularly important in early adolescence, when responsibility for glycemic control shifts from the parents to the teenager.

<table>
<thead>
<tr>
<th>GLYCOsyLATED HEMOGLOBIN (%)</th>
<th>PROBABLE BLOOD GLUCOSE RANGE (mg/dL)</th>
<th>ESTIMATE OF &quot;CONTROL&quot;</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.4-7.4</td>
<td>60-120</td>
<td>Normal range</td>
</tr>
<tr>
<td>8</td>
<td>120-150</td>
<td>Excellent</td>
</tr>
<tr>
<td>9-10</td>
<td>150-180</td>
<td>Good</td>
</tr>
<tr>
<td>10-11</td>
<td>180-220</td>
<td>Fair</td>
</tr>
<tr>
<td>11-12</td>
<td>220-260</td>
<td>Fair</td>
</tr>
<tr>
<td>12-13</td>
<td>260-300</td>
<td>Fair to poor</td>
</tr>
<tr>
<td>&gt;13</td>
<td>&gt;300</td>
<td>Very poor</td>
</tr>
</tbody>
</table>


The Diabetic Patient with Concomitant Systemic Disease

The following guidelines are intended to assist the clinician or other caregiver in managing the diabetic patient with additional illness:

**Guidelines to Sick Day Management**

1. Never skip insulin administration.
2. Check blood glucose and urinary ketones every 4-6 hours.
3. Give supplemental short-acting insulin every 4 hours for elevated blood sugar and additional dose amounts for hyperglycemia with ketonuria.
4. Evaluate and treat the underlying illness.
5. If blood glucose levels are low (less than 120 mg/dL), reduce short-acting insulin and give glucose-containing fluids.
6. If adequate fluid intake cannot be maintained or vomiting persists for more than 2 hours, intravenous hydration is necessary.
7. Notify a clinician if blood glucose is more than 400 mg/dL with moderate to large acetone or change in patient’s level of alertness or signs of dehydration (weighing every 6 hours may be a useful guide).

Diarrhea—Chronic

Common Causes

Antibiotic-induced
Carbohydrate malabsorption, hereditary
  Lactose
Chemotherapy-induced
Cystic fibrosis
Dietary
  Allergy (milk, soy, other)
  Overfeeding

Infection
  Bacterial
  Human immunodeficiency virus (HIV)
  Parasitic
  Postinfectious
  Carbohydrate malabsorption

Uncommon Causes

Anatomic lesions
  Hirschsprung’s disease
  Malrotation
Celiac disease
Irritable bowel syndrome
Malnutrition, starvation

Necrotizing enterocolitis
  Parenteral infections
  Otitis media
  Urinary tract infections
  Regional enteritis
  Ulcerative colitis

Rare Causes

Abeta- and hypobetalipoproteinemia
Adrenal insufficiency
Biliary atresia
Blind loop syndrome
Carbohydrate malabsorption
  Sucrose, isomaltose, glucose, galactose
Chronic hepatitis
Enterokinase deficiency
Familial chloride diarrhea
Ganglioneuroma
Hyperthyroidism
Immune deficiency
  Combined immune deficiency
  Hypogammaglobulinemia
  IgA deficiency

Intestinal ischemia
  Intestinal lymphangiectasia
  Intestinal pseudo-obstruction
  Mesenteric artery insufficiency
  Neuroblastoma
  Pancreatic insufficiency and neutropenia (Schwachman-Diamond-Oski syndrome)
  Pancreatic tumors
  Radiation-induced
  Short gut syndrome
  Small bowel tumors;
  lymphosarcoma
  Wolman’s disease

The Common Bacterial Causes of Bloody Diarrhea

The association of diarrhea and the passage of small amounts of blood in the stool (hematochezia) can be due simply to hemorrhoids or mucosal tears caused by spasm, hypermotility, or irritation at the mucocutaneous junction. The persistent case of hematochezia associated with diarrhea, however, is most likely indicative of an infectious or inflammatory etiology. In the pediatric patient, bloody diarrhea is usually a result of an infectious enteric pathogen.

The following table summarizes the types and manifestations of bloody diarrhea caused by bacterial pathogens.
# Bloody Diarrhea of Bacterial Origin

<table>
<thead>
<tr>
<th>INFECTIOUS AGENT</th>
<th>AGE CHILD MOST AFFECTED</th>
<th>USUAL INCUBATION PERIOD</th>
<th>USUAL SOURCE OF ACQUISITION</th>
<th>SEASON</th>
<th>PRESENTING SIGNS &amp; SYMPTOMS</th>
<th>MAIN PATHOPHYSIOLOGICAL MECHANISM</th>
</tr>
</thead>
<tbody>
<tr>
<td><em>Shigella</em></td>
<td>6 mo-3 yr</td>
<td>36-72°</td>
<td>Fecal-oral; contaminated food or water</td>
<td>Warm months</td>
<td>Fever, abdominal pain, watery diarrhea becoming bloody</td>
<td>Enteroinvasive; cytopathic</td>
</tr>
<tr>
<td><em>Salmonella</em></td>
<td>Infants &lt;2 yr sometimes older</td>
<td>24-48°</td>
<td>Oral via contaminated food or water</td>
<td>Warm months</td>
<td>Vomiting, abdominal pain, diarrhea—dysentery-like</td>
<td>Enteroinvasive</td>
</tr>
<tr>
<td><em>Campylobacter</em></td>
<td>Infants and children &lt;6 yr</td>
<td>2-7 d</td>
<td>Oral via contaminated food or water; infected pets</td>
<td>Warm months</td>
<td>Severe abdominal pain, bloody diarrhea</td>
<td>Cytolytic exotoxin</td>
</tr>
<tr>
<td><em>Yersinia</em></td>
<td>Toddlers to teenagers</td>
<td>3-4 d</td>
<td>Oral via contaminated food or water; person-to-person</td>
<td>Cooler months</td>
<td>Fever, abdominal pain, vomiting, diarrhea</td>
<td>Enteroinvasive; enterotoxigenic</td>
</tr>
<tr>
<td><em>C. difficile</em></td>
<td>All ages after neonatal period</td>
<td>1-6 wk</td>
<td>Almost always req. res prior exposure to antibiotics, especially ampicillin, clindamycin, cephalosporins</td>
<td>All year</td>
<td>Abdominal pain, diarrhea, distention, blood in stool</td>
<td>Cytotoxin</td>
</tr>
<tr>
<td><em>Escherichia coli</em></td>
<td>a. Enteroinvasive</td>
<td>All ages</td>
<td>24-72°</td>
<td>Fecal-oral; contaminated food or water</td>
<td>Warm months</td>
<td>Fever, chills, abdominal pain, watery diarrhea becoming dysentery-like</td>
</tr>
<tr>
<td></td>
<td>b. Entero-pathogenic</td>
<td>All ages</td>
<td>1-3 d</td>
<td>Fecal-oral; contaminated food or water</td>
<td>Warm months</td>
<td>Fever, nausea, cramps watery or bloody diarrhea in some serotypes</td>
</tr>
<tr>
<td><em>N. gonorrhoeae</em></td>
<td>Teenagers</td>
<td>2-7 d</td>
<td>Anal intercourse</td>
<td>All year</td>
<td>Dysentery, odynochezia</td>
<td>Enteroinvasive; cytotoxic</td>
</tr>
<tr>
<td><em>C. trachomatis</em></td>
<td>Teenagers</td>
<td>2-3 wk</td>
<td>Anal intercourse</td>
<td>All year</td>
<td>Dysentery, odynochezia</td>
<td>Enteroinvasive; cytotoxic</td>
</tr>
</tbody>
</table>
### Bloody Diarrhea of Bacterial Origin (Cont.)

<table>
<thead>
<tr>
<th>INFECTIOUS AGENT</th>
<th>STOOL FINDINGS</th>
<th>LABORATORY</th>
<th>ENDOSCOPY</th>
<th>HISTOLOGY</th>
<th>BARIUM ENEMA</th>
<th>RX</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Shigella</strong></td>
<td>Blood, WBCs</td>
<td>Band forms &gt; segmented forms</td>
<td>Mild-to-severe colitis</td>
<td>Acute inflammation</td>
<td>Normal to colitis-like, mucocutaneous ulcers</td>
<td>Trimethoprim-sulfamethoxazole</td>
</tr>
<tr>
<td><strong>Salmonella</strong></td>
<td>Blood, WBCs</td>
<td>Normal to 1 WBC L shift</td>
<td>Mild colitis</td>
<td>Acute focal inflammation</td>
<td>Normal to colitis-like</td>
<td>None except in sick infant &lt;1 yr; chloramphenicol, ampicillin, trimethoprim-sulfamethoxazole</td>
</tr>
<tr>
<td><strong>Campylobacter</strong></td>
<td>Blood, WBCs</td>
<td>Normal to 1 WBC</td>
<td>Colitis-like</td>
<td>Focal or diffuse acute inflammation</td>
<td>Colitis-like</td>
<td>Erythromycin stearate</td>
</tr>
<tr>
<td><strong>Yersinia</strong></td>
<td>Rare PMNs, blood</td>
<td>1 WBC and sed rate</td>
<td>Crohn's-like aphthous lesions</td>
<td>Focal inflammation; monos &gt; PMNs</td>
<td>Spasm cecum, abnormal terminal ileum</td>
<td>Seldom needed: tetracycline, ampicillin, chloramphenicol, trimethoprim-sulfamethoxazole</td>
</tr>
<tr>
<td><strong>C. difficile</strong></td>
<td>Blood, few WBCs</td>
<td>Normal to slight 1 WBC</td>
<td>Hyperemia; plaques, focal or diffuse</td>
<td>Pseudomembranes, distended glands, acute inflammation locally</td>
<td>“Dirty”-colon, mucosal irregularities, nodular</td>
<td>Vancomycin, Bacitracin, metronidazole, cholestyramine</td>
</tr>
<tr>
<td><strong>Escherichia coli</strong></td>
<td><strong>a. Enteroinvasive</strong></td>
<td>Blood, few WBCs</td>
<td>Normal or 1 WBC</td>
<td>Lesion in R colon; hyperemic exudate, sm ulcers</td>
<td>Mild inflammatory erosions</td>
<td>--</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Ampicillin, gentamicin, trimethoprim-sulfamethoxazole</td>
</tr>
<tr>
<td></td>
<td><strong>b. Enteropathogenic</strong></td>
<td>Blood, few WBCs</td>
<td>1 WBC L shift</td>
<td>Hyperemia, hemorrhage, superficial ulceration</td>
<td>Nonspecific colitis</td>
<td>“Thumb printing” R colon</td>
</tr>
<tr>
<td><strong>N. gonorrhoeae</strong></td>
<td>Mucopus, blood</td>
<td>1 WBC L shift</td>
<td>Severe proctitis to 10-15 cm</td>
<td>Acute diffuse inflammation</td>
<td>Normal or proctitis</td>
<td>Penicillin, tetracycline</td>
</tr>
<tr>
<td><strong>C. trachomatis</strong></td>
<td>Blood, WBCs</td>
<td>Slight 1 WBC</td>
<td>Proctitis to 15 cm</td>
<td>Acute inflammation giant cells</td>
<td>Proctitis, stricture rectum</td>
<td>Tetracycline, trimethoprim-sulfamethoxazole</td>
</tr>
</tbody>
</table>
The stool culture, of course, yields the most definitive information in the consideration of a patient with bloody diarrhea of an infectious source. Other useful tests that can be performed immediately include a microscopic examination of the stool for the presence of mucus, red blood cells, and white blood cells, using either methylene blue or Wright's stain, a Gram stain, and visual examination for ova and parasites. Immunoassays for the diagnosis of possible viral etiologies of diarrhea (e.g., rotavirus, adenovirus, Norwalk virus, etc.) can also be helpful.


Management Plan for an Infant Less Than 1 Year of Age with Diarrhea Who Does Not Require Hospitalization at the Initial Evaluation

1. First evaluation
   a. Colitis (fecal leukocytes) 0 12 mo Stool culture; blood culture if <3 mo
   b. No colitis; diarrhea <5 days 0 12 mo No stool culture
   c. History of exposure to Salmonella 0 3 mo Stool culture

2. Follow-up evaluation
   a. Diarrhea >5 days 0 12 mo Stool culture
   b. Stool culture positive Blood culture positive
   c. Stool culture positive; blood culture negative
      i. Toxic or immuno-compromised 0 12 mo Admit; look for focal infection in meninges
         bone, urinary tract
      ii. Febrile <3 mo Admit; do blood culture and give antibiotics
      iii. Febrile >3 mo Admit; do blood culture; withhold antibiotics pending culture results
   iv. Afebrile, improving 0 12 mo Reexamine, observe at home
   d. Stool culture positive; blood culture not obtained at first visit.

Antibiotics of choice: cefotaxime or ceftriaxone.

DYSMENORRHEA

Primary and Secondary Dysmenorrhea

Dysmenorrhea is a cramping pain in the lower abdomen and lower back that is temporally associated with menstrual blood flow. It may also be accompanied by headache, nausea, or diarrhea. Epidemiologic studies reveal, with great consistency, the high prevalence rate of dysmenorrhea among adolescent girls. During adolescence, dysmenorrhea becomes more common as age increases. This is probably because primary dysmenorrhea, the more common of the two types, is associated with ovulatory menstrual cycles, and most girls are anovulatory 18 to 24 months after menarche.

**Primary dysmenorrhea** is associated with no clinically detectable pelvic disease or other disorder. It usually begins 18 months after menarche (with the onset of ovulation). The pain starts on the same day as blood flow, lasting a few hours to 3 days, and is frequently accompanied by diarrhea in moderate or severe cases. The etiology of primary dysmenorrhea remains unclear, although it is known that women who suffer from this disorder produce increased amounts of prostaglandins F₂ and F₃α in their menstrual fluids. These prostaglandins cause the myometrium to increase its resting muscle tone, which yields excessive uterine contractions. The result is uterine ischemia and painful cramping.

**Secondary dysmenorrhea**, which presents far less frequently among the adolescent age group, is usually associated with some pathologic process in the pelvis. The pain is unusually severe and it (1) begins at menarche (obstructive form of secondary dysmenorrhea); (2) begins more than 3 years after menarche (e.g., endometriosis); or (3) is acute and related to one particular menstrual period (e.g., a complication of sexual activity such as a sexually transmitted infection or pregnancy).

<table>
<thead>
<tr>
<th>Conditions Associated with Secondary Dysmenorrhea</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Genital tract infections, specifically sexually transmitted endometritis or salpingitis</td>
</tr>
<tr>
<td>2. Complications of pregnancy, e.g., threatened or ectopic pregnancy</td>
</tr>
<tr>
<td>3. Endometriosis</td>
</tr>
<tr>
<td>4. Congenital malformations of the genital tract (with or without a component of blood flow obstruction)</td>
</tr>
<tr>
<td>5. Genital tract cysts and neoplasms</td>
</tr>
<tr>
<td>6. Intrauterine devices</td>
</tr>
</tbody>
</table>


DYSMORPHISM

Diagnosing Dysmorphism

The dysmorphic infant or child presents the pediatrician and the parent with several unsettling requirements: a correct diagnosis and, where possible, an etiology; a comprehensive management guide; and, a careful assessment of recurrence risks and rates.
The pediatrician's best tools in the diagnosis of the dysmorphic child are his or her eyes, the child's parents and other family members, and several guiding principles. The task involves the determination of the pathogenesis and close attention to the appearance of marked or subtle patterns (Table 1).

**Table 1. Pathogenic Mechanisms of Dysmorphism**

<table>
<thead>
<tr>
<th>TYPE OF DYSMORPHISM</th>
<th>DEFINITION</th>
<th>ASSOCIATED FACTORS</th>
<th>EXAMPLES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malformation</td>
<td>A rudimentary abnormality involving differentiation or organization of an organ part, an organ, or body part representing an embryologic field. Occurs during embryogenesis.</td>
<td>Genetic or chromosomal abnormality; teratogenic effect</td>
<td>Cleft lip/palate</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Spina bifida</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Congenital heart defects</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Down syndrome</td>
</tr>
<tr>
<td>Deformation</td>
<td>Represents a response of normal tissue to abnormal external forces. Tends to occur late in pregnancy.</td>
<td></td>
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<tr>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Disruption</td>
<td>Abnormality involving a destructive process in a normally formed organ.</td>
<td>Disruptive agent</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Radiation</td>
<td>Craniostenosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Multiple fetuses</td>
<td>Plagiocephaly-torticollis sequence</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malformed fetus</td>
<td>Micrognathia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Oligohydramnios</td>
<td>Ear deformities</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Unusual placental site</td>
<td>Pectus carinatum</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Maternal: Primigravida</td>
<td>Scoliosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Small mother</td>
<td>Dorsiflexion of foot</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Small uterus</td>
<td>Clubfoot</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Malformed uterus</td>
<td>Facial nerve palsies</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Uterine fibroids</td>
<td>Erb's palsy</td>
</tr>
<tr>
<td>Dysplasia</td>
<td>Abnormality in the development (organization or differentiation) of cells and tissues as opposed to whole organs.</td>
<td>Genetic or unknown</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Looking for patterns:

The anomalies described in the above table can be characterized as "major" (those with functional, surgical, or cosmetic consequences) or "minor" (those without consequences). Bear in mind that normal variants may constitute minor anomalies in the context of a syndrome.

A pattern of anomalies will often reveal the diagnosis and can present as syndromes, sequences, or associations.

Sequence:

A sequence refers to an isolated developmental abnormality and its subsequent structural consequences. An example is the initial mandibular hypoplasia of the Pierre Robin sequence that results in small chin, cleft palate, obstructive airway, and anoxia. The cause of a sequence is not necessarily defined.

Syndrome:

A syndrome consists of a pattern of malformations that are recognized to result from a specified cause, such as trisomies.

Association:

Associations are recognized as nonrandom and significant groupings of malformations without known etiologies. Two of the common associations are VACTERL and CHARGE. VACTERL, previously known as VATER, describes vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula with atresia, radial and renal defects, and limb defects. CHARGE consists of coloboma of the eye, heart defects, atresia of the choanae, growth and mental retardation, genital anomalies in the male, and ear anomalies.

In addition to the examination directed at recognition of patterns, there are several measurements that can be made to facilitate a diagnosis. The size of the hands and feet as well as the ratio of upper body length (crown to pubis symphysis) to lower body length (symphysis to soles) can signal dwarling syndromes. The measurement of inner canthal distance, interpupillary distance, corneal diameter, internipple distance, and penile length may lead to diagnostic clues.

Be aware of the risk factors for structural anomalies (Tables 2 and 3).

**Table 2. Children at Risk for Structural Anomalies**

| 1. The child with a family history of structural anomalies | 4. The infant of a mother with diabetes mellitus, phenylketonuria, epilepsy, or alcoholism |
| 2. The child with one known structural anomaly | 5. The mentally retarded child |
| 3. The small-for-dates infant | 6. The “strange-looking” child |

**Table 3. Anomalies and Associated Conditions**

<table>
<thead>
<tr>
<th>LOCATION</th>
<th>ANOMALY</th>
<th>CONDITION*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Back</td>
<td>Scoliosis</td>
<td>Stickler syndrome, neurofibromatosis</td>
</tr>
<tr>
<td>Chest</td>
<td>Pectus excavatum</td>
<td>Marfan’s syndrome</td>
</tr>
<tr>
<td>Digits</td>
<td>Curved fifth fingers (clinodactyly)</td>
<td>Russell-Silver syndrome</td>
</tr>
<tr>
<td></td>
<td>Finger-like thumbs</td>
<td>Holt-Oram syndrome, Aase syndrome</td>
</tr>
</tbody>
</table>

*These are illustrative conditions; often other conditions will also have the anomaly.

Table continued on next page.
<table>
<thead>
<tr>
<th>LOCATION</th>
<th>ANOMALY</th>
<th>CONDITION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Digits (Cont.)</td>
<td>Polydactyly</td>
<td>Carpenter's syndrome, trisomy 13 syndrome</td>
</tr>
<tr>
<td></td>
<td>Broad thumbs/great toes</td>
<td>Rubinstein-Taybi syndrome</td>
</tr>
<tr>
<td></td>
<td>Syndactyly</td>
<td>Apert's syndrome</td>
</tr>
<tr>
<td></td>
<td>Nail hypoplasia</td>
<td>Fetal hydantoin syndrome, Coffin-Siris syndrome</td>
</tr>
<tr>
<td></td>
<td>Second finger overlaps third finger, fifth finger overlaps fourth finger</td>
<td>Trisomy 18, trisomy 13</td>
</tr>
<tr>
<td>Ears</td>
<td>Low-set</td>
<td>Trisomy 18, Treacher Collins' syndrome</td>
</tr>
<tr>
<td></td>
<td>Crumpled</td>
<td>Beal syndrome</td>
</tr>
<tr>
<td></td>
<td>Small</td>
<td>Down syndrome</td>
</tr>
<tr>
<td></td>
<td>Large</td>
<td>Sotos' syndrome</td>
</tr>
<tr>
<td></td>
<td>Preauricular tags</td>
<td>Goldenhar's syndrome</td>
</tr>
<tr>
<td></td>
<td>Preauricular pits</td>
<td>BOR syndrome</td>
</tr>
<tr>
<td>Eyes</td>
<td>Microphthalmos</td>
<td>Oculodentodigital dysplasia</td>
</tr>
<tr>
<td></td>
<td>Coloboma of iris</td>
<td>CHARGE association</td>
</tr>
<tr>
<td></td>
<td>Stellate pattern of iris</td>
<td>Williams syndrome</td>
</tr>
<tr>
<td></td>
<td>Absent iris</td>
<td>Aniridia-Wilms' tumor</td>
</tr>
<tr>
<td></td>
<td>Cataracts</td>
<td>Hallermann-Streiff syndrome</td>
</tr>
<tr>
<td>Face</td>
<td>Unique facial gestalt</td>
<td>Down syndrome, Williams syndrome, Prader-Willi syndrome, Cornelia de Lange syndrome, whistling face syndrome</td>
</tr>
<tr>
<td>Genitalia</td>
<td>Hypospadias</td>
<td>Smith-Lemli-Opitz syndrome</td>
</tr>
<tr>
<td></td>
<td>Micropenis</td>
<td>Robinow's syndrome</td>
</tr>
<tr>
<td></td>
<td>Scrotal shawl</td>
<td>Aarskog's syndrome</td>
</tr>
<tr>
<td></td>
<td>Hypoplasia of labia majora</td>
<td>Escobar syndrome</td>
</tr>
<tr>
<td>Hair</td>
<td>Widow's peak</td>
<td>Fronto-ocular dysplasia</td>
</tr>
<tr>
<td></td>
<td>White forehead</td>
<td>Waardenburg syndrome</td>
</tr>
<tr>
<td></td>
<td>Low posterior hairline</td>
<td>Turner's syndrome, Noonan's syndrome</td>
</tr>
<tr>
<td></td>
<td>Scalp defect</td>
<td>Trisomy 13</td>
</tr>
<tr>
<td></td>
<td>Sparse to absent</td>
<td>Hypohidrotic ectodermal dysplasia</td>
</tr>
<tr>
<td></td>
<td>Kinky</td>
<td>Menkes' syndrome</td>
</tr>
<tr>
<td>Hands/feet</td>
<td>Short</td>
<td>Prader-Willi syndrome</td>
</tr>
<tr>
<td></td>
<td>Long</td>
<td>Marfan's syndrome</td>
</tr>
<tr>
<td></td>
<td>Abnormal palm/sole creases</td>
<td>Sotos' syndrome, Down syndrome</td>
</tr>
<tr>
<td>Head</td>
<td>Macrosephaly</td>
<td>Hydrocephalus, Sotos' syndrome</td>
</tr>
<tr>
<td></td>
<td>Microcephaly</td>
<td>Many syndromes</td>
</tr>
<tr>
<td>Joints</td>
<td>Dislocations</td>
<td>Larsen's syndrome</td>
</tr>
<tr>
<td></td>
<td>Elbow abnormalities</td>
<td>Turner's syndrome, XXXXY syndrome</td>
</tr>
<tr>
<td></td>
<td>Absent patella</td>
<td>Trisomy 8</td>
</tr>
<tr>
<td></td>
<td>Contractures</td>
<td>Beal syndrome, arthrogrypotic conditions</td>
</tr>
<tr>
<td>Limbs</td>
<td>Long</td>
<td>Beal syndrome, Stickler syndrome, XXY syndrome</td>
</tr>
<tr>
<td></td>
<td>Short</td>
<td>Many short-limbed dwarfing syndromes</td>
</tr>
<tr>
<td></td>
<td>Radial hypoplasia</td>
<td>VATER association</td>
</tr>
<tr>
<td>Mandible</td>
<td>Hypoplasia</td>
<td>Pierre Robin sequence</td>
</tr>
<tr>
<td>Maxilla</td>
<td>Hypoplasia</td>
<td>Nager syndrome</td>
</tr>
<tr>
<td>Mouth</td>
<td>Large</td>
<td>Goldenhar's syndrome</td>
</tr>
<tr>
<td></td>
<td>Multiple frenula</td>
<td>Orofaciodigital syndrome</td>
</tr>
<tr>
<td></td>
<td>Large tongue</td>
<td>Beckwith-Wiedemann syndrome</td>
</tr>
</tbody>
</table>

Table continued on next page.
Table 3. Anomalies and Associated Conditions (Cont.)

<table>
<thead>
<tr>
<th>LOCATION</th>
<th>ANOMALY</th>
<th>CONDITION*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neck</td>
<td>Webbed</td>
<td>Noonan's syndrome</td>
</tr>
<tr>
<td></td>
<td>Short</td>
<td>Klippel-Feil syndrome</td>
</tr>
<tr>
<td>Nose</td>
<td>Small</td>
<td>Fetal warfarin effect</td>
</tr>
<tr>
<td></td>
<td>Large</td>
<td>Seeker's syndrome</td>
</tr>
<tr>
<td></td>
<td>Choanal atresia</td>
<td>CHARGE association</td>
</tr>
<tr>
<td>Ocular region</td>
<td>Hypertelorism</td>
<td>Opitz syndrome, Aarskog’s syndrome</td>
</tr>
<tr>
<td></td>
<td>Hypotelorism</td>
<td>Holoprosencephaly</td>
</tr>
<tr>
<td></td>
<td>Short palpebral fissures</td>
<td>Fetal alcohol syndrome, blepharophimosis</td>
</tr>
<tr>
<td></td>
<td>Epicanthis fold</td>
<td>Fetal trimethadione syndrome</td>
</tr>
<tr>
<td></td>
<td>Upward slant of palpebral fissures</td>
<td>Down syndrome</td>
</tr>
<tr>
<td></td>
<td>Downward slant of palpebral fissures</td>
<td>Treacher Collins' syndrome</td>
</tr>
<tr>
<td></td>
<td>Synophrys (fusion of eyebrows in midline)</td>
<td>Cornelia de Lange syndrome</td>
</tr>
<tr>
<td>Philtrum</td>
<td>Prominent</td>
<td>Trichorhinophalangeal syndrome</td>
</tr>
<tr>
<td></td>
<td>Long</td>
<td>Williams syndrome</td>
</tr>
<tr>
<td></td>
<td>Short</td>
<td>Cohen syndrome</td>
</tr>
<tr>
<td></td>
<td>Smooth</td>
<td>Fetal alcohol syndrome</td>
</tr>
<tr>
<td>Skin</td>
<td>Cafe au lait spots</td>
<td>Neurofibromatosis, Russell-Silver syndrome, Bloom's syndrome</td>
</tr>
<tr>
<td></td>
<td>Pigmented nevi</td>
<td>Turner's syndrome</td>
</tr>
<tr>
<td></td>
<td>Multiple lentigines</td>
<td>Leopard syndrome</td>
</tr>
<tr>
<td></td>
<td>Telangiectases</td>
<td>Ataxia-telangiectasia syndrome, Bloom's syndrome, Rothmund-Thomson syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hemangiomaata</td>
<td>Sturge-Weber syndrome</td>
</tr>
<tr>
<td>Teeth</td>
<td>Hypodontia</td>
<td>Hyphidrotic ectodermal dysplasia</td>
</tr>
<tr>
<td></td>
<td>Caries</td>
<td>Dentinogenesis imperfecta</td>
</tr>
<tr>
<td></td>
<td>Neonatal</td>
<td>Ellis-van Creveld syndrome</td>
</tr>
<tr>
<td>Thorax</td>
<td>Small</td>
<td>Jeune's syndrome</td>
</tr>
</tbody>
</table>

Lastly, the pediatrician must be armed with the knowledge of recurrence risks and be able to appropriately counsel the concerned parent(s) (Table 4).

Table 4. Recurrence Risk in Families of Dysmorphic Children

<table>
<thead>
<tr>
<th>RISK IN SIBLINGS</th>
<th>PERCENTAGE</th>
<th>CONDITION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low</td>
<td>1 2</td>
<td>Trisomy 21 syndrome, trisomy 13, trisomy 18</td>
</tr>
<tr>
<td></td>
<td>3 7</td>
<td>Spina bifida, cleft palate/tip, hypospadias</td>
</tr>
<tr>
<td>Moderate</td>
<td>25</td>
<td>Autosomal recessive disease: Smith-Lemli-Opitz syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td>X-linked recessive disease: X-linked hydrocephalus</td>
</tr>
<tr>
<td>High</td>
<td>50</td>
<td>Autosomal dominant disease: neurofibromatosis, tuberous sclerosis</td>
</tr>
<tr>
<td>Total</td>
<td>100</td>
<td>Chromosomal disorder: 21/21 translocation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Down's syndrome with carrier parent</td>
</tr>
</tbody>
</table>

DYSPHAGIA

Common Causes

Chemical mucositis
Caustic ingestion
Gastroesophageal reflux with esophagitis
Radiation/chemotherapy
Immature sucking/swallowing mechanism

Oropharyngeal infections
Cervical adenitis
Epiglottitis
Gingivitis
Herpetic stomatitis
Peritonsillar abscess
Pharyngitis
Retropharyngeal abscess
Tooth abscess
Physiologic expulsion reflux

Uncommon Causes

Cerebral palsy
Cleft palate
Esophageal spasm
Esophageal stricture
External compression of the esophagus
Esophageal diverticuli
Esophageal duplication
Mediastinal masses/tumors
Vascular anomalies

Foreign body
Infectious esophagitis
Candida, herpes
Macroglossia (any cause)
Megagnathia
Pharyngeal diverticuli
Physiologic (globus hystericus)
Submucosal cleft
Tracheoesophageal fistula

Rare Causes

Choanal atresia
Collagen vascular disease
Dermatomyositis
Scleroderma
Diphtheria
Esophageal atresia, web, cyst
Laryngeal cyst, cleft
Muscular hypertrophy of the esophagus
Neuromuscular causes
Botulism
Bulbar and suprabulbar palsy
Mobius syndrome
Chalasia/achalasia of the esophagus
Congenital laryngeal stridor
Cranial nerve palsy

Neuromuscular causes (Cont.)
Demyelinating disease
Guillain-Barré syndrome
Hypotonias
Muscular dystrophy
Myasthenia gravis
Myotonic dystrophy
Pharyngeal or cricopharyngeal incoordination
Tetanus
Pharyngeal cyst, cleft
Rumination
Temporomandibular ankylosis/hypoplasia
Tumors (oropharynx, esophagus)
DYSRHYTHMIA

Common Causes

Acidemia
Congenital heart disease
Drugs
  - Antiarrhythmics
  - Beta blockers
  - Caffeine
  - Cocaine

Drugs (Cont.)
  - Psychotropics
  - Sympathomimetics
  - Hypoxemia
  - Idiopathic
  - Postoperative (cardiac procedures)

Uncommon Causes

Cardiomyopathy (dilated, hypertrophic, infiltrative)
Electrolyte disturbances (especially K, Ca, Mg)
Myocarditis

Sickle-cell disease
Sick-sinus syndrome
Wolff-Parkinson-White syndrome
  (and/or other necessary bypass tracts)

Rare Causes

Anomalous coronary artery
Central nervous system
  - Hemorrhage
  - Infection
  - Trauma
Collagen vascular disease
Complete congenital heart block
Endocrine (thyrotoxicosis, secondary electrolyte disturbance)

Kawasaki disease
Myocardial ischemia
Myocardial trauma
Myocardial tumors
Neonatal lupus
Prolonged QT syndrome
Rheumatic fever

DYSURIA

Common Causes

Candidal dermatitis/vaginitis
Chemical urethritis (bubble bath)
Contact dermatitis/vulvitis

Urethritis
Urinary tract infection
Viral cystitis

Uncommon Causes

Foreign body
Herpes simplex
Meatitis

Pinworms
Urethral trauma
102—Dysuria

Rare Causes

Appendicitis
Bladder diverticulum
Bladder outlet obstruction
  Posterior urethral valves
Bladder stones
Constipation
Drugs
  Amitriptyline
  Cytoxan
Hematospermia
Interstitial cystitis

Meatal stenosis
Posthitis
Prostatitis
Reiter’s syndrome
Schistosomiasis
Stevens-Johnson syndrome
Tuberculosis
Urethral prolapse
Urethral stricture
Varicella

I'm sorry you are wiser,
I'm sorry you are taller;
I liked you better foolish,
  And I liked you better smaller.

Aline (Mrs. Joyce) Kilmer
For the Birthday of a Middle-
Aged Child, Stanza 1
EARS

Low-set Ears

Because of their association with various syndromes, low-set ears can be a finding of extreme importance. There is still a controversy over what constitutes low-set ears. We prefer a method of evaluation suggested by Dr. Murray Feingold. In the figure, the face has a sheet of x-ray film held over it. On the margins of the film are measurement scales, and the center of the film is bisected by a horizontal line that is aligned with the medial canthi of the eyes. The amount of the ear lying above the line is measured as well as the overall length of the ear. If the ear is below the center line, the ear is low set. If 10% or less of the overall length of the ear is above the line, the ears are said to be low set. If low-set ears are found, look carefully for other physical abnormalities.

\[
\text{If } \frac{a}{b} \times 100 \leq 10, \\
\text{the ears are low set.}
\]

Some Syndromes Associated with Low-set Ears

<table>
<thead>
<tr>
<th>Apert's syndrome</th>
<th>Down's syndrome</th>
<th>Seckel's syndrome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Camptomelic syndrome</td>
<td>Fetal hydantoin syndrome</td>
<td>Trisomy 13 syndrome</td>
</tr>
<tr>
<td>Carpenter's syndrome</td>
<td>Hallermann-Streiff syndrome</td>
<td>Trisomy 18 syndrome</td>
</tr>
<tr>
<td>Cri-du-chat syndrome</td>
<td>Noonan's syndrome</td>
<td>Treacher-Collins syndrome</td>
</tr>
<tr>
<td>Deletion of the short arm of chromosome 13</td>
<td>Saethre-Chotzen syndrome</td>
<td>Turner's syndrome</td>
</tr>
</tbody>
</table>

ENCOPRESIS

Common Causes
- Chronic constipation
- Diarrheal disorders
- Emotional disturbance

Uncommon Causes
- Hirschsprung’s disease

Rare Causes
- Diastematomyelia
- Epidural abscess
- Poliomyelitis
- Postanorectal surgery
- Sacral agenesis
- Spinal cord tumor
- Syringomyelia
- Transverse myelitis

ENDOTRACHEAL INTUBATION

Complications of Endotracheal Intubation

Endotracheal intubation in the setting of an intensive care unit, emergency room, or delivery suite is rarely as controlled a procedure as the same performed in an operating room. Unrecognized esophageal placement of an endotracheal tube is the most common complication of emergency intubations and can lead rapidly to brain damage or death. Below are the phases of progressive hypoxemia, their clinical pictures, and recommended resuscitative maneuvers.

Precritical Phase: Arterial oxygen content decreases from normal 19 vol% to 12 vol% (± 70% saturation).
- Sympathetic tone increases: HR increases by approximately 10 bpm.
- Systolic BP increases 10–15 mmHg.
- Diastolic BP unchanged.
- Pulse pressure widens.

Critical Phase: Arterial oxygen content ranges from 12 vol% to 9 vol% (50% saturation).
- Vagal tone increases: systolic BP decreases, HR decreases.
- Cyanosis evident in nonanemic patients.

Terminal Phase: Arterial oxygen content falls below 9 vol% (<50% saturation).
- HR slow to 40 or fewer bpm.
- BP readings cannot be obtained.
- ECG readings may show marked sinus bradycardia in the absence of a palpable pulse.

Resuscitation
In the absence of a known and immediately remediable problem, the tube should be removed quickly and an oral or nasal airway should be placed. The patient should be administered 100% oxygen with a bag and mask until an unhurried reintubation can be performed. The use of atropine or vasopressors will not correct the hypoxia.

Recognition and correction of the hypoxemia during the precritical and critical phases will generally avert any catastrophic event. Terminal phase may require initiation of BLS and ALS.*

* BLS = basic life support; ALS = advanced life support.
Other Complications

Foreign bodies: Due to excessive lymphoid tissue in childhood, semirigid naso-tracheal tubes may cause dislodging of adenoid tissue and subsequent obstruction.

Bronchial intubation: The right bronchus is more susceptible to catheterization due to the obtuse angle at its junction with the trachea. Partial catheterization of the right bronchus may result in occlusion and collapse of the right upper lobe.

Tube distortion: Tubes constructed from soft rubber or metal reinforced plastic are less likely to kink, compress, or twist, but even they may distort, leading to occlusion and hypoxemia.

Effect of tube size: Due to the diameter of the pediatric airway, increased airway resistance is a significant problem with thick-walled tubes. Refer to the chart below for selection of the appropriate thin-walled endotracheal tube.

### Selection of Endotracheal Tube

<table>
<thead>
<tr>
<th>Infant Weight (g)</th>
<th>Endotracheal Tube Internal Dia. (Mm)</th>
<th>Suction Catheter Size (French Gauge)</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;1250</td>
<td>2.5</td>
<td>5 Fr</td>
</tr>
<tr>
<td>1250-3000</td>
<td>3.0</td>
<td>6 Fr</td>
</tr>
<tr>
<td>&gt;3000</td>
<td>3.5</td>
<td>8 Fr</td>
</tr>
</tbody>
</table>

By Age

<table>
<thead>
<tr>
<th></th>
<th>Internal Dia. (Mm)</th>
<th>Suction Catheter Size (French Gauge)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Full term 9 mo</td>
<td>3.5</td>
<td>5 Fr</td>
</tr>
<tr>
<td>12-20 mo</td>
<td>4.0</td>
<td>6 Fr</td>
</tr>
<tr>
<td>2 yr</td>
<td>4.5</td>
<td>8 Fr</td>
</tr>
<tr>
<td>&gt;2 yr</td>
<td>4.5 + age (yr) / 4</td>
<td>10 Fr</td>
</tr>
</tbody>
</table>


Verification Techniques for Proper Placement

Although no verification technique for the placement of endotracheal tubes is completely infallible, there are several methods that are quite reliable and ought to be routinely performed after intubation.

1. The most reliable method of verification, detection of end-expired CO₂, requires instruments that may not be available in all emergency situations. As carbon dioxide analyzers become less cumbersome and more available, this will be the method of choice.

2. Auscultation of breath sounds is historically accepted as a reliable method for verification of tracheal intubation. Recent studies have found that it may be dependable only two-thirds of the time. Confounding factors in the pediatric age group include short necks and obesity. An important point: auscultation is most useful when performed both before and after intubation. Nonidentical sounds or unequal chest expansion should alert the pediatrician to a possible problem.
3. **Pulse oximetry**, now widely available, is a good adjunct to auscultation but in general should not be the sole method of verification. Oximetry is an excellent monitoring device for the intubated child at risk for mucous plugging or compression. Regardless of the patient's risk status, pulse oximetry is useful because pediatric patients tend to desaturate more quickly than their adult counterparts.

4. **Mouth-to-tube-insufflation** remains a reliable, simple, and universally available technique for verifying tube placement. A quick, but not too forceful, breath is expired into the tube connector by the physician. A properly placed tube will allow gradual insufflation of the lungs. Expiration, against minimal resistance, should be felt against the physician's turned cheek. If esophageal placement has occurred, the expired air will not equal the insufflated amount, the expulsion will be rapid, and the air will not be felt on the cheek.


**ENURESIS**

Common Causes

Developmental delay of bladder function and capacity
Psychological

Uncommon Causes

Diabetes
Food allergy

Obstructive abnormalities of the urinary tract
Urinary tract infections

Rare Causes

Compulsive water drinking
Diabetes insipidus, central or nephrogenic

Lumbosacral anomalies
Sickle-cell anemia
Spinal cord tumors

Natural History of Nocturnal Enuresis

Most children stop wetting their beds between 2 and 4 years of age. Does that mean that persistent bedwetting after 4 indicates an abnormality? Almost certainly not. An organic disorder is rarely found in enuretic children, that is, children who are bedwetting after the age of 5.

Bedwetting after age 5 years is more common in boys, in children from large families, and in children from lower socioeconomic groups, and it occurs more commonly in families where one of the parents may have been a bedwetter. Nocturnal enuresis occurs once a month or more in 8% of school-age children.
When bedwetting recurs after a period of dryness (termed regressive or secondary enuresis), an explanation should be sought. The polyuria of diabetes mellitus may frequently present in this way. Also rule out urinary tract disease and diabetes insipidus.

<table>
<thead>
<tr>
<th>Age by Which Bedwetting Stopped</th>
<th>BOYS</th>
<th>GIRLS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>White (%)</td>
<td>Black (%)</td>
</tr>
<tr>
<td>&lt;2</td>
<td>28</td>
<td>37</td>
</tr>
<tr>
<td>2 3</td>
<td>70</td>
<td>73</td>
</tr>
<tr>
<td>4 5</td>
<td>85</td>
<td>83</td>
</tr>
<tr>
<td>6 7</td>
<td>91</td>
<td>89</td>
</tr>
</tbody>
</table>

What becomes of the children, about 10 to 15%, who continue to wet after 4 to 5 years of age?

There is a relative constancy to the percentage of children who spontaneously remit from their enuresis each year. The figure to remember is that any one child during the course of 1 year has about a 15% chance of this problem going away by itself. Thus, a child who wets his bed at age 5 will have about a 3% chance of bedwetting by age 20. These figures apply only to the natural history of bedwetting and not to those children whose bedwetting is related to an environmental factor.

<table>
<thead>
<tr>
<th>Percentage Spontaneously Ceasing Bedwetting per Year</th>
</tr>
</thead>
<tbody>
<tr>
<td>AGES: 5 TO 9</td>
</tr>
<tr>
<td>Percentage</td>
</tr>
</tbody>
</table>

Diurnal or daytime enuresis occurs much less frequently than nocturnal, with an incidence of less than 1% in a 7- to 12-year-old group.


**EOSINOPHILS**

**Cerebrospinal Fluid Eosinophilia**

Many laboratories now perform cytocentrifugation and differential cytologic staining of the WBCs found in cerebrospinal fluid. The finding of CSF eosinophilia using this technique has led to the development of a list of the various agents—infected and otherwise—that can cause eosinophilic pleocytosis in the central nervous system. The following list includes the reported causes of "eosinophilic meningitis." For most of the causes listed, a careful history and physical examination will provide the diagnosis.
Fungal infection
- Coccidiomycosis meningitis

Viral infection
- Coxsackie virus meningitis
- Chronic lymphocytic choriomeningitis virus
- Subacute sclerosing panencephalitis

Bacterial infection
- Tuberculous meningitis
- Neurosyphilis

CSF inflammation
- Radiologic dye used in myelography
- Rubber CSF shunt tubing
- CNS malignancy
- Multiple sclerosis
- CNS hemorrhage

Parasitic infection
- Neurocysticercosis (caused by the tapeworm *Taenia solium* and found in Asia and Africa)
- *Toxocara canis* and *T. cati*
- Trichinella
- Nematode meningoencephalitis, including that due to *Angiostrongylus cantonensis* and *Gnathostoma spinigerum*, found in Taiwan, Thailand, and the Pacific islands
- Amebic meningitis caused by the free-living amebae *Naegleria fowleri* and *Acanthamoeba* found in warm fresh or brackish water all over the world.

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**EPIDIDYMITIS**

**Epididymitis in Children and Adolescents**

Epididymitis, though considered rare in prepubertal males, is also a common cause of acute scrotum in childhood. Like torsion of the spermatic cord, the common clinical presentation includes pain, swelling, and erythema. Unlike torsion, the onset tends to be less acute.

Although definitive differentiation between epididymitis and testicular torsion requires radionuclide scanning or surgery, the history and physical exam should point the pediatrician in the correct direction. Once established, the diagnosis of epididymitis often suggests further urologic investigation.

**Epididymitis Compared with Testicular Torsion**

<table>
<thead>
<tr>
<th>FEATURE</th>
<th>EPIDIDYMITIS</th>
<th>TESTICULAR TORSION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset:</td>
<td>Insidious onset often accompanied by signs and symptoms of urethritis or systemic bacterial and/or viral infection. History may be significant for recent trauma.</td>
<td>Acute onset of pain</td>
</tr>
<tr>
<td>Physical signs:</td>
<td>Elevation of testis decreases pain.</td>
<td>Elevation of testis increases pain.</td>
</tr>
<tr>
<td></td>
<td>Gastrointestinal symptoms rare.</td>
<td>Abdominal pain and gastrointestinal symptoms common.</td>
</tr>
<tr>
<td></td>
<td>Cremasteric reflex present.</td>
<td>Cremasteric reflex present.</td>
</tr>
<tr>
<td>Causes:</td>
<td>Bacterial: Coliform organisms, gonorrhea, <em>Staphylococcus</em>, <em>M. Tuberculosis</em>, <em>C. trachomatis</em>; viral; traumatic; chemical (i.e., reflux); systemic diseases such as sarcoid, Kawasaki's, Henoch-Schönlein purpura; idiopathic</td>
<td>Anatomical defect</td>
</tr>
</tbody>
</table>
In the infant with suspected epididymitis, a thorough work-up for sepsis is indicated, because epididymitis in infancy is often a signpost of systemic illness. In the child less than 2 years of age and in older patients with recurrent episodes, a urologic work-up with an IVP and VCUG is recommended to rule out any associated genitourinary abnormality.


**EPISTAXIS**

**Common Causes**
- Allergic rhinitis
- Repeated sneezing
- Secondary to dryness and crusting over anterior portion of nasal septum
- Trauma
  - External
  - Self-inflicted (nose picking)
- Upper respiratory infection

**Uncommon Causes**
- Factor XI deficiency
- Hypertension
- Platelet dysfunction syndrome
- Sickle cell anemia
- Thrombocytopenia from any cause
- von Willebrand's disease

**Rare Causes**
- Angiofibroma
- Ataxia-telangiectasia
- Congenital syphilis
- Ehlers-Danlos syndrome
- Foreign body
- Malaria
- Measles
- Nasal angiomas
- Nasal diphtheria
- Nasal polyp
- Oral contraceptives
- Osler-Weber-Rendu disease
  (hereditary hemorrhagic telangiectasia)
- Pertussis
- Rheumatic fever
- Scarlet fever
- Scurvy
- Typhoid fever
- Varicella
- Wegener's granulomatosis

**EPSTEIN-BARR VIRUS**

**Epstein-Barr Viral Antibody Titers**

Although the heterophile antibody test is sometimes positive in young children with infection caused by the Epstein-Barr virus, heterophile positivity is certainly not as reliable in pediatric patients as it is in adults. But don't despair. More specific antibody assays are available. They allow you to determine on a single serum sample whether a patient is currently infected or has had the infection at some time in the past. The most helpful tests are the following:
**Interpretation of EBV Serum Antibody Patterns**

<table>
<thead>
<tr>
<th>INTERPRETATION</th>
<th>IgM CAPSID ANTIGEN</th>
<th>IgG CAPSID ANTIGEN</th>
<th>IgG EARLY ANTIGEN</th>
<th>ANTINUCLEAR ANTIGEN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Susceptible</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute primary infection</td>
<td>+</td>
<td>-</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>(IM presentation)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute primary infection</td>
<td>+</td>
<td>+</td>
<td>-</td>
<td>+ or -</td>
</tr>
<tr>
<td>(non-IM presentation or asymptomatic)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Old, quiescent infection</td>
<td>-</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Reactivated infection</td>
<td>±</td>
<td>+</td>
<td>+</td>
<td>+§</td>
</tr>
</tbody>
</table>

* A few (<10%) adults and an even greater number (10% to 20%) of children with acute IM develop an antibody response directed to R instead of D component.
* A low antibody titer (≤1:5 in our laboratory) may also be detected in acute infection.
* Occasionally a weak, probably nonspecific, antibody response to R component is present.
* Moderate, stable titers of antibody should be present.
* Stable levels of antibody, although in low or absent levels in immunosuppressed and immunodeficient patients, are present.

From Sumaya CV, Epstein-Barr virus serologic testing: Diagnostic indication and interpretations. Pediatr Infect Dis 5:337, 1986. Reproduced by permission of Williams & Wilkins Co.

The timing of these antibody rises is depicted in the accompanying figure.

Duration of serum IgM and IgG antibody responses to EBV early antigen and nuclear antigen. The antibody response to EBV early antigen components may persist for years after an episode of EBV infectious mononucleosis. Antibodies to EBV nuclear antigen may be absent during acute infection but, once present, remain for life. (From Sumaya CV, Ench Y. Pediatrics 75:1011, 1985. Reproduced by permission of the American Academy of Pediatrics.)

Heterophile antibodies are nonspecific and not directed against the virus. They cause agglutination of sheep, horse, beef, and goat red blood cells. Since other antibodies may cause such agglutination, care should be taken to avoid
false-positive reporting. Testing sera with horse RBCs is the most sensitive method, whereas the beef cell hemolysin test gives the most specific results. The most reliable method for single slide tests is to absorb the sera with guinea pig kidney cells before adding horse RBCs. When the antibody is due to EBV infection, the test should remain positive after such absorption. Many of the rapid test kits include guinea pig kidney cells for this purpose. Rapid test kits may be accurate and helpful, but experienced personnel and fresh testing material are essential for best results.

The list below provides a summary of the contents of some of the commercially available kits.

<table>
<thead>
<tr>
<th>TRADE NAME</th>
<th>RED BLOOD CELL USED</th>
<th>USE OF GUINEA PIG KIDNEY ABSORPTION</th>
<th>MANUFACTURER</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mono-test</td>
<td>Horse</td>
<td>No</td>
<td>Wampole Labs.</td>
</tr>
<tr>
<td>Mono-Diff</td>
<td>Horse</td>
<td>Yes</td>
<td>Wampole Labs.</td>
</tr>
<tr>
<td>Monospot</td>
<td>Horse</td>
<td>Yes</td>
<td>Ortho Diagnostics</td>
</tr>
<tr>
<td>Diagluto</td>
<td>Horse</td>
<td>Yes</td>
<td>Beckman Instruments, Inc.</td>
</tr>
<tr>
<td>Monosticon</td>
<td>Sheep</td>
<td>Yes</td>
<td>Organon Diagnostic Products</td>
</tr>
<tr>
<td>Mono-Stat</td>
<td>Native and papain-treated sheep</td>
<td>Not needed</td>
<td>Colab Labs., Inc.</td>
</tr>
<tr>
<td>Confirmikit</td>
<td>Native and enzyme-treated horse</td>
<td>Not needed</td>
<td>BBL-BioQuest, Div. of Becton, Dickinson &amp; Co.</td>
</tr>
<tr>
<td>Heterol</td>
<td>Native and enzyme-treated horse</td>
<td>Not needed</td>
<td>Difco</td>
</tr>
</tbody>
</table>

Reference: Andiman WA: J Pediatr 95:171, 1980. Figure courtesy of Dr. John Sullivan, X-Linked Lymphoproliferative Syndrome Registry. Department of Pathology, University of Massachusetts Medical Center, Worcester, MA.


**ERYTHROCYTE SEDIMENTATION RATE**

**The Slow Sedimentation Rate**

All of the factors responsible for determining the rate at which erythrocytes sediment have not been identified. Factors that are known to influence the sedimentation rate include the quantity of fibrinogen, alpha1-globulin, the gamma-M globulin, and the serum cholesterol, with the quantity of fibrinogen perhaps playing the most important role. In addition, alterations in the morphologic characteristics of the red cell or in cell surface charge that hinder rouleau formation will affect the erythrocyte sedimentation rate.

Everyone is familiar with the long and nondescript list of diseases that produce an increase in the erythrocyte sedimentation rate. It is generally not appreciated that certain disorders or drugs characteristically produce a slow sedimentation rate or a rate that is slower than would be anticipated. Disorders that produce a slow sedimentation rate include:
Anorexia nervosa
Hypofibrinogenemia, congenital or acquired
Abetalipoproteinemia (acanthocytosis)
Sickle cell anemia (if many sickled forms are present)
Pyruvate kinase deficiency (usually postsplenectomy if associated with marked morphologic alterations of the erythrocytes)

In patients with the nephrotic syndrome in whom an infection is suspected, the measurement of the C-reactive protein provides a useful alternate screening test.


EYE

Causes of Severe Eye Injuries in Children

Approximately 160,000 school age children in the U.S. suffer from traumatic eye injuries of varying severity each year. Indeed, the only more common pediatric ophthalmologic entity requiring hospital admission is strabismus. Although trauma is the usual descriptive word for these eye injuries among children, a variety of offending etiologies is responsible, including balls, sticks, fists, fingers, falls, glass, animal bites, and metallic foreign bodies. Listed below are the most frequent diagnoses made among children with eye trauma severe enough to warrant hospital admission, subdivided into age groups.

Primary Diagnoses of Eye Trauma

<table>
<thead>
<tr>
<th>DIAGNOSIS</th>
<th>AGF GROUP, NO. OF CASES</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>TOTAL (%)</td>
</tr>
<tr>
<td>Hyphema</td>
<td>81 (31.4)</td>
</tr>
<tr>
<td>Globe lacerations</td>
<td>48 (18.6)</td>
</tr>
<tr>
<td>Traumatic cataract</td>
<td>31 (12)</td>
</tr>
<tr>
<td>Eyelid laceration</td>
<td>28 (10.9)</td>
</tr>
<tr>
<td>Retinal detachment</td>
<td>19 (7.4)</td>
</tr>
<tr>
<td>Foreign body in eye</td>
<td>18 (7)</td>
</tr>
<tr>
<td>Orbital fractures</td>
<td>10 (3.9)</td>
</tr>
<tr>
<td>Injury to orbital tissue</td>
<td>7 (2.7)</td>
</tr>
<tr>
<td>Burns</td>
<td>4 (1.6)</td>
</tr>
<tr>
<td>Vitreous hemorrhage</td>
<td>4 (1.6)</td>
</tr>
<tr>
<td>Traumatic glaucoma</td>
<td>3 (1.2)</td>
</tr>
<tr>
<td>Corneal abrasion</td>
<td>2 (&lt;1)</td>
</tr>
<tr>
<td>Lens subluxation</td>
<td>2 (&lt;1)</td>
</tr>
<tr>
<td>Conjunctival laceration</td>
<td>1 (&lt;1)</td>
</tr>
<tr>
<td>Total (%)</td>
<td>258 (100)</td>
</tr>
</tbody>
</table>
Primary admitting diagnoses of pediatric patients with ocular trauma (ages 0 to 20 years). Others include those less-frequent diagnoses listed in the table. RD indicates retinal detachments.


The Eyelash Syndrome

Adults can appreciate the fact that a foreign body in the eye is both annoying and painful. Indoors the most common foreign body to produce such discomfort is the eyelash. We frequently forget that eyelashes may get into the eyes of infants as well. The next time you are confronted with the problem of an irritable, crying baby for which you cannot find a suitable explanation, be sure to check the eyes for a foreign body. If you find it, you can produce an instant cure.

If an older child complains of the sensation of a foreign body, there usually is one, particularly indoors. Oblique illumination with a flashlight can aid detection. If nothing is seen, check the upper tarsal conjunctiva by eversion of the lid. This may call for instillation of a local anesthetic.

Most foreign bodies on or in the cornea or conjunctiva can be removed by irrigation or gentle wiping with a wet cotton applicator (also see next entry). In the event of an abrasion, instill an antibacterial ointment and follow up within 24 hours to check on healing and for the presence of infection.

Removal of a Foreign Body

You may be called upon to remove a simple nonpenetrating foreign body from the eye—objects such as eyelashes, dust, or dirt. All you need is a clean Band-Aid. Simply touch the foreign body with the adhesive portion of the Band-Aid. You can even do this to yourself with the aid of a mirror when necessary.

Remember that as a teenager you are at the last stage in your life when you will be happy to hear that the phone is for you.

Fran Lebowitz
FAILURE TO THRIVE

Common Causes

Neglect
  Inadequate ingestion/metabolism of calories
  Depression with anorexia
  Manipulative behavior
  Rumination as self-stimulation
  Secondary malabsorption
  Self-induced (vomiting, laxative abuse)
  Specific deficiency (e.g., zinc, biotin)
  Starvation
  Secondary neuroendocrine abnormalities
  Abnormal cycling of growth hormone
  Cortisol deficiency
  Physical neglect/abuse
  Psychosocial deprivation
  Withholding of food as neglect/abuse
  Intentional withholding of food
  “Unintentional” withholding of food
  “Overwhelmed” caretaker
  Lack of support systems (financial/social)
  Primary personal needs (e.g., drug/alcohol abuse)
  Time constraints (e.g., unsupervised eating, bottle propping)
  Psychotic or depressed caretaker

Nonorganic failure to thrive
  Inadequate volume of feeds
  Two feeds per day
  Too little per feed
  Colic
  “Difficult” feeder
  Financial factors
  Ignorance
  Inexperienced/impatient ± compounding child factors
  Inappropriate foods for age
  Cultural factors
  Fad diets
  Financial factors
  Ignorance
  Incorrect preparation of formula
  Chronic dilution
  Financial factors
  Ignorance
  Prolonged use after gastroenteritis
  Inappropriate additives

Normal variants
  Delayed growth spurt
  Early onset growth retardation
  Genetic “slightness”

Organic failure to thrive
  CNS etiologies
    Mental retardation/cerebral palsy
    Neurodevelopmental retardation
  Gastrointestinal etiologies
    Chronic gastroenteritis
    Gastroesophageal reflux
    Pyloric stenosis
  Prematurity
  Small for gestational age
**Uncommon Causes**

- Defective utilization of calories
  - Chronic hypoxemia
  - Diabetes mellitus
- Defects in absorption
  - Cystic fibrosis
  - Enzymatic deficiencies
  - Food sensitivity/intolerance
  - Hepatitis
  - Inflammatory bowel disease
  - Milk allergy
  - Starvation
- Inadequacy of food intake
  - Cleft lip/palate
  - Dyspnea of any cause
    - Congenital heart disease
    - Respiratory disease/insufficiency
- Increased metabolism
  - Chronic anemias
  - Chronic/recurrent infections
  - Otitis, sinusitis, pneumonia
  - Parasites
  - Tuberculosis
  - Urinary tract infection
  - Chronic respiratory insufficiency
  - Congenital heart disease
  - Malignancies

**Rare Causes**

- Defective utilization of calories
  - Adrenal insufficiency
  - Chromosomal syndromes
  - Diabetes insipidus
  - Diencephalic syndrome
  - Drugs/toxins
  - Dysmorphogenic syndromes
  - Fetal exposure syndromes
  - Hypopituitarism
  - Hypothyroidism
- Metabolic disorders
  - Aminoacidopathies
  - Galactosemia
  - Organic acidurias
  - Storage diseases
- Parathyroid disorders
- Renal tubular acidosis
- Defects in absorption
  - Acrodermatitis enteropathica
  - Biliary atresia/cirrhosis
  - Celiac disease
  - Hirschsprung's disease
  - Immunologic deficiency
  - Necrotizing enterocolitis
  - Pancreatic insufficiency
  - Short gut syndrome
- Inadequacy of food intake (Cont.)
  - Immature suck/swallow
  - Pharyngeal incoordination
- Increased metabolism
  - Acquired heart disease
  - Adrenocortical excess
  - Chronic inflammation
    (e.g., JRA, SLE)
  - Chronic seizure disorder
  - Drugs/toxins
  - Hyperaldosteronism
  - Hyperthyroidism
FATIGUE

Common Causes

Acute recovery from surgery, trauma, most illnesses
Anemia
Chronic atopy
Eating disorders
   Excessive dieting (± anorexia nervosa, bulimia)
Excessive physical exertion
Mononucleosis (and most viral infections)
Obesity

Pregnancy
Psychosocial
   Chronic boredom
   Chronic depression/anxiety
   Grief
   Stress (prolonged and severe)
Sedentary lifestyle
Sleep disorders
   Insomnia
   Sleep pattern disruption
      (lack of REM sleep)

Uncommon Causes

Acute bacterial infections
   Bacteremia
   Meningitis
Chronic hypoxemia
   Asthma
   Cardiomyopathy
   Chronic pulmonary disease
   Congenital heart disease
   Congestive heart failure
   Cystic fibrosis
   Heart disease
   Pericarditis
   Pulmonary hypertension

Chronic infections (Cont.)
   Brucellosis
   Cytomegalic inclusion disease

Chronic infections
   Histoplasmosis
   Osteomyelitis
   Parasitic infestations
   Pyelonephritis
   Sinusitis
   Subacute bacterial endocarditis
   Toxoplasmosis
   Tuberculosis
   Urinary tract infection
   Dehydration
   Hepatitis
   Upper airway obstruction
      (sleep apnea)
   Pickwickian syndrome
   Tonsillar-adenoidal hypertrophy

Rare Causes

Acquired immunodeficiency syndrome (AIDS)
Allergic tension fatigue syndrome
Connective tissue diseases
   Dermatomyositis
   Juvenile rheumatoid arthritis
   Mixed connective tissue disease
   Scleroderma
   Systemic lupus erythematosus
Endocrine disorders
   Diabetes insipidus
   Diabetes mellitus

Endocrine disorders (Cont.)
   Hyper/hypoadrenalism
   Hyper/hypopituitarism
   Hyper/hypothyroidism
   Hyperparathyroidism
   Hepatic insufficiency
   Hypoglycemia
   Inborn errors of metabolism
   Inflammatory bowel disease
   Intussusception
   Malignancy
   Leukemia
118—Fever

Malignancy (Cont.)
  Lymphoma
  Solid tumors
Metabolic disturbances
  Hypermagnesemia
  Hypokalemia
  Hypomagnesemia
  Hyponatremia
Neurologic
  Intracranial hematomas
  Myasthenia gravis
  Narcolepsy
Renal tubular acidosis
Toxins and drugs
  Alcohol
  Analgesics and salicylates
  Anticonvulsants

Toxins and drugs (Cont.)
  Antihistamines
  Barbiturates
  Carbon monoxide
  Corticosteroids
  Digitalis
  Heavy metals
  Insulin
  Nicotine
  Pesticides
  Progesterones
  Sedatives
  Tetracycline
  Vitamin A
  Vitamin D
  Uremia

FEVER

Fever of Unknown Origin

Fever is defined here as a temperature, higher than 38.5°C for more than 2 weeks.

Common Causes

Collagen vascular disease
  Juvenile rheumatoid arthritis
  Lupus erythematosus
  Periarthritis nodosa
Factitious
Infections
  Atypical mycobacterial infections
  Epstein-Barr virus infections
  Osteomyelitis
  Sinusitis, mastoiditis
  Urinary tract infections
  “Viral syndromes”

Inflammatory bowel diseases
  Regional enteritis
  Ulcerative colitis
Malignancy
  Acute lymphoblastic leukemia
  Neuroblastoma
  Hodgkin’s disease
  Non-Hodgkin’s lymphoma

Uncommon Causes

Drug-induced
Infections
  Cat-scratch disease
  Cytomegalic inclusion disease
  Lung abscess
  Hepatitis

Infections (Cont.)
  Histoplasmosis
  Pelvic inflammatory disease
  Salmonellosis
  Kawasaki disease
  Lyme disease
Rare Causes

Infection
- Behcet's syndrome
- Diabetes insipidus
- Central
- Nephrogenic
- Diencephalic syndrome
- Ectodermal dysplasia
- Familial dysautonomia
- Hepatoma
- Infection
  - Blastomycosis
  - Brucellosis
  - Human immunodeficiency virus infection
  - Leptospirosis
  - Liver abscess
  - Lymphogranuloma venereum
  - Malaria
  - Perinephric abscess

Infection (Cont.)
- Psittacosis
- Q fever
- Rocky Mountain spotted fever
- Streptococcosis
- Subdiaphragmatic abscess
- Toxoplasmosis
- Tuberculosis
- Tularemia
- Viral encephalitis
- Visceral larva migrans

Fever of Unknown Origin—Continued

Prolonged episodes of fever without an apparent explanation are an uncommon diagnostic problem in pediatrics. Because of their rarity, they represent an exacting challenge and provide the clinician with an unequaled opportunity to demonstrate his skills in both careful history taking and physical examination. At least 50% of “fevers of unknown origin” can be diagnosed by thoughtful attention to details and very simple laboratory studies. Unfortunately, the designation “fever of unknown origin” often prompts a myriad of tests and radiographic procedures in a nonsystematic fashion.

What are the usual causes of obscure, prolonged fevers in children and how do they differ in etiology from those observed in adults? The accompanying table summarizes the findings in two studies involving infants and children and contrasts them with a representative study of adult patients. Fever was defined as the presence of a rectal temperature of 38.5°C (99.8°F) on at least four occasions over a minimum period of 2 weeks.

<table>
<thead>
<tr>
<th>Causes of Fever of Unknown Origin</th>
</tr>
</thead>
<tbody>
<tr>
<td>INFANTS AND CHILDREN</td>
</tr>
<tr>
<td>Pizzo and Associates</td>
</tr>
<tr>
<td>Infections (52%)</td>
</tr>
<tr>
<td>Viral syndromes</td>
</tr>
<tr>
<td>Respiratory</td>
</tr>
<tr>
<td>Central nervous system</td>
</tr>
<tr>
<td>Urinary tract</td>
</tr>
<tr>
<td>Osteomyelitis</td>
</tr>
</tbody>
</table>

Table continued on next page.
Fever

Causes of Fever of Unknown Origin (Cont.)

<table>
<thead>
<tr>
<th>Infants and Children</th>
<th>Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pizzo and Associates</td>
<td>McClung</td>
</tr>
</tbody>
</table>

**Infections (Cont.)** (52%)
- Endocarditis
- Tuberculosis
- Herpes simplex, generalized
- Sinusitis
- Salmonellosis

**Collagen-Vascular (20%)**
- Rheumatoid arthritis
- Vasculitis
- Anaphylactoid purpura
- Lupus erythematosus

**Neoplastic (6%)**
- Leukemia
- Lymphoma

**Miscellaneous (10%)**
- Agranulocytosis
- Lamellar ichthyosis
- Milk allergy
- Agammaglobulinemia
- Behçet's syndrome
- Anicteric hepatitis
- Ruptured appendix
- Central nervous system fever
- Aspiration pneumonia

**Physically well children (9%)**

**Undiagnosed (12%)**

**The Diagnostic Evaluation**

1. *Initial studies* should be determined by clues provided by the history and physical examination. One must particularly search for a history of recent immunizations, transfusions, travel, risk factors for HIV infection, and exposure to animals or other sick individuals.
2. *Initial diagnostic procedures* should include a complete blood count, urinalysis, erythrocyte sedimentation rate, chest film, and serum protein electrophoresis in addition to more specific studies indicated from the history and physical examination.

3. If sedimentation rate is elevated, if serum electrophoresis reveals a reversed albumin-globulin ratio or increase in the alpha globulin fraction, or if leukocytosis exists, these should all be considered evidence of an active disease process.

4. If initial studies fail to provide a diagnosis, other useful studies might include:
   - Blood cultures, urine cultures, stool cultures
   - Liver function tests
   - Bone marrow biopsy and culture
   - Antinuclear antibodies
   - Latex fixation test
   - Lupus erythematosus preparations
   - Upper gastrointestinal films
   - Barium enema
   - Intravenous pyelogram
   - Bone scan
   - Sinus films

5. Ultimately, the diagnosis may require a biopsy of skin, muscle, and/or liver.

6. It is useful to establish an orderly timetable for the pursuit of the diagnosis. All too often the investigation proceeds in an aimless fashion without a logical schedule.


The Symptomatic Treatment of Fever

Would you throw cold water on the walls of an overheated room or would you turn down the thermostat? The answer seems obvious, yet the same logic is often not present when it comes to the management of fever in children. The rational treatment of fever requires an understanding of its pathophysiologic basis. Most febrile states in infants and children result from an abnormal elevation in the hypothalamic setpoint triggered by the release of interleukins. When this occurs, heat production is increased and heat loss is minimized. Much less commonly, fever is a result of excessive heat production alone, or when heat production is normal but heat loss is faulty.

The accompanying table examines the pathophysiologic basis for fever and describes the corresponding appropriate treatment.

### Pathophysiologic Basis for Symptomatic Treatment of Fever

<table>
<thead>
<tr>
<th>Disease Process Causing Fever</th>
<th>Pathophysiology of Fever</th>
<th>Clinical Findings</th>
<th>Appropriate Non-specific Treatment</th>
<th>Inappropriate Non-specific Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infection, malignancy, allergy, steroid fever, collagen disease</td>
<td>Endogenous pyrogen causes rise in hypothalamic setpoint</td>
<td>Patient complains of feeling cold; piloerection; cold extremities; absent of minimal sweating; body positioned to minimize surface area, shivering</td>
<td>Drug-induced lowering of hypothalamic setpoint (e.g., with aspirin, acetaminophen); supply sufficient clothing and covers for maximal comfort; avoid shivering</td>
<td>Physical removal of heat, e.g., sponging, ice blanket, ice water enemas; without change in setpoint, these measures will cause discomfort, increase metabolic rate and will only lower body temperature for brief period</td>
</tr>
<tr>
<td>CNS lesion, DDT poisoning, scorpion venom, radiation, epinephrine and norepinephrine overdose</td>
<td>Agent or illness acts directly on hypothalamus to raise setpoint</td>
<td>Same as above</td>
<td>Drug-induced lowering of hypothalamic setpoint theoretically indicated as above; it is not clearly established, however, as possible with presently available drugs</td>
<td>Same as above</td>
</tr>
<tr>
<td>Malignant hyperthermia, hyperthyroidism, hypernatremia, primary defect in energy metabolism, aspirin overdose</td>
<td>Heat production exceeds heat loss mechanisms</td>
<td>Patient complains of feeling hot; no piloerection; hot extremities; active sweating; body positioned to maximize surface area</td>
<td>Undress patient; physical removal of heat, e.g., ice blanket, sponging</td>
<td>Attempt to lower setpoint (which is already set normally) with drugs, e.g., aspirin - possible toxicity of drug without potential benefit</td>
</tr>
<tr>
<td>Overuse of sauna, exposure to industrial heat, overdressing</td>
<td>Environmental heat load exceeds normal heat loss mechanisms</td>
<td>Same as above</td>
<td>Eliminate heat source; undress patient; physical removal of heat is effective but is not usually necessary</td>
<td>Same as above</td>
</tr>
<tr>
<td>Ectodermal dysplasia, burns, phenothiazine, anticholinergic overdose, heat stroke</td>
<td>Defective heat loss mechanisms cannot cope with normal heat load</td>
<td>Patient complains of feeling hot; sweating decreased (secondary to disease process); hot extremities; body positioned to maximize surface area</td>
<td>Provide cool environment; undress patient; physical removal of heat may be necessary</td>
<td>Same as above</td>
</tr>
</tbody>
</table>

Decline in Fever Following Acetaminophen—What Does It Mean?

Many physicians assume that a febrile child who exhibits a reduction in fever along with an improvement in general appearance following acetaminophen administration is not likely to have a bacterial infection. However, the degree of fever reduction does not distinguish between viral and bacterial infection, nor does it help in selecting those children who are bacteremic. Comparison of the degree of fever reduction in children with their eventual etiologic diagnosis has yielded the following conclusions:

1. The improvement in clinical appearance following fever reduction makes the identification of patients with potentially life-threatening infection more difficult.
2. Patients with occult bacteremia and bacterial deep tissue infections experience at least as great a reduction in fever 1 and 2 hours following acetaminophen administration as do patients with self-limited viral infections.
3. Even patients with bacterial meningitis experience a mean temperature reduction of 1.1°C following acetaminophen administration.

These studies demonstrate that neither the observation of clinical improvement nor the history of defervescence following antipyretic administration should comfort the physician when the patient's condition prior to antipyretic therapy gave cause for concern.


Hospitalization of Febrile Infants—What Is the Risk?

When we hospitalize young infants and treat them with intravenous antibiotics for presumed sepsis, we believe we are decreasing their risk of serious disease and complications. In fact, hospitalization of young febrile infants is not only costly, it is risky. Of 190 febrile infants under 2 months of age evaluated in the outpatient clinics of The Johns Hopkins University Hospital and hospitalized for observation and treatment, 37 patients (19.5%) had 48 separate complications. Twenty-four (50%) of the complications resulted from intravenous administration of fluids and/or antibiotics.

<table>
<thead>
<tr>
<th>Complications During In-hospital Treatment of Febrile Infants</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>TYPE OF COMPLICATION</strong></td>
</tr>
<tr>
<td>Preventable</td>
</tr>
<tr>
<td>IV infiltrates requiring compresses</td>
</tr>
<tr>
<td>Sloughing of skin with IV therapy</td>
</tr>
<tr>
<td>Fluid overload</td>
</tr>
<tr>
<td>Gentamicin overdose</td>
</tr>
<tr>
<td>Fever secondary to high isostate temperature</td>
</tr>
<tr>
<td>Untreated urinary tract infection</td>
</tr>
<tr>
<td>Distraught mother secondary to multiple lumbar punctures</td>
</tr>
<tr>
<td>Stolen Infant</td>
</tr>
<tr>
<td><strong>Total</strong></td>
</tr>
</tbody>
</table>

Table continued on next page.
Complications During In-hospital Treatment of Febrile Infants (Cont.)

<table>
<thead>
<tr>
<th>TYPE OF COMPLICATION</th>
<th>NO. OF COMPLICATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Other</td>
<td></td>
</tr>
<tr>
<td>Diarrhea with onset &gt; 72 hr after admission</td>
<td>12</td>
</tr>
<tr>
<td>Thrush/candidiasis</td>
<td>6</td>
</tr>
<tr>
<td>Chloramphenicol sodium succinate-induced bone marrow</td>
<td>1</td>
</tr>
<tr>
<td>suppression</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>19</td>
</tr>
</tbody>
</table>

In addition to the complications listed above, diagnostic misadventures during hospitalization can lead to unnecessary costs and patient trauma. The table below lists the diagnostic misadventures encountered during the hospitalization of the same 190 infants mentioned above.

Misadventures During In-hospital Treatment of Febrile Infants

<table>
<thead>
<tr>
<th>TYPE OF MISADVENTURE</th>
<th>NO.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Contaminated CSF cultures</td>
<td>12</td>
</tr>
<tr>
<td>Contaminated blood cultures</td>
<td>4</td>
</tr>
<tr>
<td>Abnormal urinalysis findings with no follow-up</td>
<td>4</td>
</tr>
<tr>
<td>3 normal chest roentgenograms in 1 patient</td>
<td>1</td>
</tr>
<tr>
<td>Suprapubic examination done to check contaminated,</td>
<td>1</td>
</tr>
<tr>
<td>improperly labeled urine culture</td>
<td></td>
</tr>
<tr>
<td>2 repeated lumbar punctures, both negative after positive</td>
<td>1</td>
</tr>
<tr>
<td>counterimmunoelectrophoresis</td>
<td></td>
</tr>
<tr>
<td>Traumatized infant 2° to multiple lumbar punctures</td>
<td>2</td>
</tr>
<tr>
<td>Kept 48 hr for neurologic consultation that was not done</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>26</td>
</tr>
</tbody>
</table>

The next time you consider hospitalizing an infant “just for observation,” remember these potential complications and try to assure more good than harm comes from your decision.


FONTANELS

How Many Fontanels Are Present at Birth in the Infant's Skull?

There are actually six fontanels, but only two, the anterior and posterior, are normally palpable.

The anterior fontanel is at the meeting place of the coronal, sagittal, and frontal sutures. It is a diamond-shaped, fibrous tissue membrane covering a transient defect in ossification. It is the largest fontanel and measures about 4 cm in the A-P direction, and 2.5 cm transversely. The membrane pulses with the
infant's pulse and can be observed to be slightly depressed when the baby is upright and quiet. Molding of the skull from pressures during labor and delivery can cause temporary overriding of the sutures and the impression of a smaller fontanel. Other less benign conditions causing smaller fontanel size are discussed in the following section.

The posterior fontanel is at the meeting place of the sagittal and lambdoid sutures. It is triangular and usually less than 1 cm at the widest point.

Two pairs of fontanels, the sphenoidal and mastoid, appear on each side of the skull, are small and irregular, and are difficult to palpate.

**Abnormal Fontanel Size**

An abnormality in size of the anterior fontanel may be a tip-off to abnormality in the infant. The figure displays the fontanel size, defined as $\frac{\text{length} \times \text{width}}{2}$, as measured with a steel tape in 201 normal infants.

The following tables list conditions associated with an unusually small (or prematurely closed) fontanel or with an unusually large fontanel.

<table>
<thead>
<tr>
<th>Disorders in Which Premature Closure or Small Fontanel for Age May Be a Feature</th>
</tr>
</thead>
<tbody>
<tr>
<td>Microcephaly</td>
</tr>
<tr>
<td>High Ca++/vitamin D ratio in pregnancy</td>
</tr>
<tr>
<td>Craniosynostosis</td>
</tr>
<tr>
<td>Hyperthyroidism</td>
</tr>
<tr>
<td>Normal variant</td>
</tr>
</tbody>
</table>

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Disorders in Which Large Fontanel for Age May Be a Feature

<table>
<thead>
<tr>
<th>Skeletal Disorders</th>
<th>Chromosomal Abnormalities</th>
<th>Other Conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Achondroplasia</td>
<td>Down's syndrome</td>
<td>A thyric hypothyroidism</td>
</tr>
<tr>
<td>Aminopterin-induced syndrome</td>
<td>13 Trisomy syndrome</td>
<td>Hallermann-Streiff syndrome</td>
</tr>
<tr>
<td>Apert's syndrome</td>
<td>18 Trisomy syndrome</td>
<td>Malnutrition</td>
</tr>
<tr>
<td>Clidocranial dysostosis</td>
<td></td>
<td>Progeria</td>
</tr>
<tr>
<td>Hypophosphatasia</td>
<td></td>
<td>Rubella syndrome</td>
</tr>
<tr>
<td>Kenny's syndrome</td>
<td></td>
<td>Russell-Silver syndrome</td>
</tr>
<tr>
<td>Osteogenesis imperfecta</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pyknodysostosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vitamin D deficiency rickets</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


The Bulging Fontanel

A bulging fontanel in an infant is generally regarded as a sign of serious CNS disease, such as:

- Meningitis
- Encephalitis
- Hydrocephalus
- Cerebral hemorrhage
- Intracranial abscess
- Subdural hematoma
- Lead poisoning
- Sinus thrombosis
- Tumor

The history, however, may suggest a benign cause. A congenital subgaleal cyst over the anterior fontanel may simulate a bulging fontanel. *Benign intracranial hypertension*, a syndrome of increased intracranial pressure, normal ventricular system and CSF composition, and absence of focal neurologic signs can also produce a bulging fontanel.

The causes of benign intracranial hypertension in infancy:

- Impaired CSF absorption
- Obstructed inferior vena cava secondary to intrathoracic mass or obstructive lung disease
- Obstruction of sagittal sinus secondary to skull fracture or other cause
- Drugs (Cont.)
  - Nalidixic acid
  - Infections
  - Roseola infantum (herpes virus 6)
  - Guillain-Barré syndrome
  - Nutritional
  - Hypovitaminosis A
  - Rapid brain growth following starvation
  - Miscellaneous
  - Polycythemia vera
  - Heart disease
  - Allergic diseases
  - Anemia (severe)
  - Wiskott-Aldrich syndrome

FOREIGN BODY

Where Is the Coin—in the Esophagus or Trachea?

The list of foreign bodies encountered in various openings of children’s bodies is almost endless. A frequent problem is the swallowing or aspiration of objects. Coins are favored for this purpose (but note that hot dogs are one of the most common causes of fatal aspiration). Opaque objects such as coins are accurately localized with radiography. However, a frequent question that comes up in removing a coin is whether it is in the esophagus or trachea.

In the esophagus, foreign bodies are usually found at one of three areas of physiologic narrowing: (1) below the cricopharyngeal muscle; (2) at the level of the aortic arch; or (3) just above the diaphragm. Foreign bodies lie in the plane of least resistance, and if a coin enters the esophagus, it will lie in a frontal plane and thus appear head-on in the anterior-posterior film of the chest and on-edge in a lateral film. In contrast, a coin in the trachea will come to rest in the sagittal plane and will appear on-edge in the A-P view and head-on in the lateral view.

Of course foreign bodies can lodge in the larynx and in bronchi, as well as in the trachea. Objects in the upper airway can cause dysphagia from swelling, and objects in the esophagus can cause airway problems from compression or overflow of food or other secretions.

Other common locations for foreign bodies in children are the eye (see entry under “EYE”), ear, nose, stomach and intestine.


Foreign Bodies in the Air and Food Passages

A wise pediatrician once said, “When deliberating over a difficult diagnosis in a child, even if it’s as clear-cut a case as otitis media, always consider the ingestion of a foreign body!” The following should facilitate that thought process.

1. Underlying factors leading to foreign bodies of the esophagus
   Neuromuscular disorders (uncoordinated swallowing)
   Vascular compression (double aortic arch, etc.)
   Stricture or stenosis secondary to:
      Congenital deformity
      Repaired tracheoesophageal fistula
      Reflux esophagitis
      Caustic ingestion
   Altered mental status (e.g., poor judgment secondary to age, underlying medical condition)
   Alteration of sensation

2. Signs and symptoms of esophageal foreign bodies
   Refusal to take oral feedings
   Gagging
   Increased salivation
   “Foreign body” sensation
   Vomiting
   Pain radiating to the sternal or back area
   Pain or discomfort with swallowing
   Drooling
3. Signs and symptoms of foreign bodies of the airway

**Foreign Bodies of the Airway**

<table>
<thead>
<tr>
<th>SIGNS, SYMPTOMS</th>
<th>LOCATION OF FOREIGN BODY</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>LARYNX</td>
</tr>
<tr>
<td>Hoarseness</td>
<td>+</td>
</tr>
<tr>
<td>Aphonia</td>
<td>+</td>
</tr>
<tr>
<td>Odynophagia</td>
<td>+</td>
</tr>
<tr>
<td>Drooling</td>
<td>+</td>
</tr>
<tr>
<td>Audible slap</td>
<td>+</td>
</tr>
<tr>
<td>Cough</td>
<td>+</td>
</tr>
<tr>
<td>Hemoptysis</td>
<td>+</td>
</tr>
<tr>
<td>Stridor</td>
<td>+</td>
</tr>
<tr>
<td>Wheeze</td>
<td>+</td>
</tr>
<tr>
<td>Dyspnea</td>
<td>+</td>
</tr>
<tr>
<td>Airway obstruction</td>
<td>+</td>
</tr>
<tr>
<td>Sudden death</td>
<td>+</td>
</tr>
</tbody>
</table>

4. Progression of radiographs: Imaging that may be required for diagnosis
   - Chest x-ray (anteroposterior and lateral)
   - Lateral neck (if indicated)
   - Inspiratory/expiratory films (if patient is cooperative)
   - Chest fluoroscopy
   - Ultrasound; overpenetrated films of unresolved areas of density
   - Possible computed tomography; possible contrast studies

5. An ounce of prevention
   - Certain foods are more easily aspirated than others (e.g., peanuts—the most commonly aspirated food, chunks of carrots and apples, hot dogs, etc.). These foods should be withheld from children 0–4 years until they can chew them properly (i.e., after their molars have erupted). Small objects, such as tiny plastic toys, etc., should always be avoided in this age group.


**FRAGILE X SYNDROME**

Recognition of the Fragile X Syndrome in Young Children

Fragile X syndrome is the most commonly inherited form of mental retardation. Although it is thought to be an X-linked recessive trait with variable expression and incomplete penetrance, 30% of all carrier women are also affected. The syndrome is named “fragile X” because there exists a fragile site or gap at the end of the long arm of the X-chromosome in lymphocytes of affected patients when grown in a folate-deficient medium. Carrier females typically have a 30 to 40% chance of giving birth to a retarded male and a 15 to 20% chance of having a retarded female. Further, there frequently exists a maternal family history for a relative with mental retardation or developmental and learning disabilities. Most studies have dealt with recognition of this syndrome in older children and young
adults, but many of the physical features, behavioral characteristics, and family history features are apparent far earlier.

Prominent parental concerns that might bring such a child to a pediatrician's attention include:

- Developmental delay
- Speech delay
- Short attention span or hyperactivity
- Mouthing of objects persisting at an age beyond when it would be expected
- Difficulty in disciplining the child
- Frequent temper tantrums
- Autistic-like behaviors such as rocking, talking to oneself, spinning, unusual hand movements, difficulty with transitions, preference for being alone, echolalia, and poor eye contact
- Poor gross motor coordination
- History of vomiting, spitting up, or colic during infancy
- History of frequent otitis media
- Self-abusive behaviors
- Hand flapping
- Drooling persisting at an age beyond when it would be expected
- Hypotonia
- Fighting with others
- Pica
- Hand/thumb sucking

While older children (8 to 12 years of age) are more likely to display the classic physical features of fragile X syndrome (long face with a prominent jaw, large prominent ears, and post-pubertal macroorchidism), patients as young as 2 or 3 years have been noted to exhibit the following physical findings:

- Long and/or wide and/or protruding ears
- Prominent jaw or long face
- High arched palate
- Flattened nasal bridge
- Microcephaly or relative macrocephaly
- Apparent hypertelorism
- Epicanticth folds

- Simian creases of palms; vertical creases of soles
- Long philtrum
- Hemangioma
- Hyperextensible joints
- Antimongoloid slant to eyes
- Clinical impression of macroorchidism
- Prominent forehead

It is not feasible or sound to recommend chromosomal studies on all children with developmental, learning, and behavioral disabilities. But these problems (particularly speech delay, unusual behaviors, and developmental delay) taken in context with a maternal family history of mental retardation or developmental disabilities and the physical findings of long, wide, or protruding ears, a long face, flat nasal bridge, and a high arched palate probably warrant a search for the fragile X chromosome. A new method of identifying carriers of these mutations by direct DNA analysis has recently been described.

To make a prairie it takes a clover
and one bee, —
One clover, and a bee,
And revery.
The revery alone will do
If bees are few.

Emily Dickinson
GASTROENTERITIS

Rotavirus Versus Astrovirus

Over the past 20 years, pediatricians and virologists have gained great insights into the causes and nature of gastroenteritis. The enteric adenoviruses, such as rotavirus, have been of particular focus as a cause of diarrhea in infants and young children. Recently, reports of astrovirus-caused gastroenteritis has come to the attention of clinicians, although their medical importance remains poorly defined. Offered below is a comparison of the clinical findings associated with astrovirus and rotavirus gastroenteritis:

Clinical Findings Associated with Astrovirus and Rotavirus Gastroenteritis*

<table>
<thead>
<tr>
<th>FINDINGS</th>
<th>ASTROVIRUS INFECTION (N = 44) (percent)</th>
<th>ROTAVIRUS INFECTION (N = 175) (percent)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Watery stools</td>
<td>61</td>
<td>67</td>
</tr>
<tr>
<td>Loose stools</td>
<td>41</td>
<td>35</td>
</tr>
<tr>
<td>Mucoid stools</td>
<td>55</td>
<td>51</td>
</tr>
<tr>
<td>Bloody stools</td>
<td>7</td>
<td>6</td>
</tr>
<tr>
<td>Nausea^</td>
<td>71</td>
<td>88</td>
</tr>
<tr>
<td>Abdominal pain^</td>
<td>58</td>
<td>63</td>
</tr>
<tr>
<td>Vomiting</td>
<td>61</td>
<td>67</td>
</tr>
<tr>
<td>Fever</td>
<td>80</td>
<td>83</td>
</tr>
<tr>
<td>Dehydration ≥5%</td>
<td>5</td>
<td>15</td>
</tr>
</tbody>
</table>

* Only stool samples in which no bacterial, parasitic, or other viral pathogens were detected are included.
^ Includes data from Study 1 only (33 children with astroviruses and 116 with rotaviruses).


Chitterlings: Another *Yersinia* Food Group

*Yersinia enterocolitica* was first recognized as a cause of human infection in 1933. In young children, infection generally causes acute gastroenteritis. In older children a picture of mesenteric adenitis that can easily be confused with appendicitis predominates. A 1988–89 outbreak of *Yersinia enterocolitica* in Atlanta has added chitterlings (also *chitlins*), the small intestines of pigs, to the list of agents of transmission. Although the epidemiology remains poorly understood, previously cited agents include contaminated milk products, contact with sick pets, transfusion of contaminated blood products, and ingestion of raw pork.
The Atlanta outbreak was traced to households where chitterlings were prepared for holiday meals. Preparation of chitterlings involves boiling the raw small intestines of pigs after fat and fecal matter have been removed. In nearly all of the Atlanta cases, the affected infants and children had contact with the chitterlings’ preparers, although not the raw intestines themselves.

The take-home message is that when an infant or child presents with a picture of acute gastroenteritis, a thorough investigation into intrahousehold contacts as well as ingestion of possible contaminants is called for.

**Clinical Manifestations of Yersiniosis**

- Diarrhea
- Mucoid stools
- Bloody diarrhea or pus (10–20%)
- Fever
- Vomiting
- Abdominal pain—colicky, diffuse or localized to RLQ

**Differential Diagnosis**

- *Shigella*
- Enteroinvasive *E. coli*
- *Salmonella*
- *Campylobacter*
- *Yersinia pseudotuberculosis* can cause identical picture to the mesenteric adenitis of older children.


**GASTROINTESTINAL BLEEDING**

**In the Neonate**

**Common Causes**

- Esophagitis
- Gastritis
- Ingested maternal blood
- Necrotizing enterocolitis
- Stress ulcer (gastric)

**Uncommon Causes**

- Acquired coagulopathy
- Gastroenteritis (*Campylobacter* infections)
- Hemophilia
- Rectal trauma or gastrointestinal trauma
- Thrombocytopenia
- Vitamin K deficiency
- Volvulus

**Rare Causes**

- Acute ulcerative colitis
- Gastric polyp
- Gastrointestinal duplication cyst
- Intussusception
- Leiomyoma
- Milk allergy
- Nasal or pharyngeal bleeding
- Severe cyanotic congenital heart disease
- Vascular malformation of the gut (hemangiomia, telangiectasia, arteriovenous malformation)
In Infancy

Common Causes

- Anal fissures
- Esophagitis
- Gastritis (possibly due to drug ingestion)
- Gastroenteritis
- Polyps

Uncommon Causes

- Acquired coagulation disturbance
- Hemophilia
- Henoch-Schönlein purpura
- Inflammatory bowel disease
- Meckel's diverticulum
- Parasitism
- Peptic ulcer
- Thrombocytopenia

Rare Causes

- Chronic granulomatous disease
- Diverticulitis
- Esophageal varices
- Hemangiomas and telangiectasia
- Hemolytic-uremic syndrome
- Hemorrhoids
- Intestinal foreign body
- Lymphosarcoma
- Peutz-Jeghers syndrome
- Pseudoxanthoma elasticum
- Scurvy

Using the BUN/Creatinine Ratio in Localizing the Source of Gastrointestinal Bleeding in Children

Although most children with gastrointestinal bleeding make the diagnostician's job somewhat easier by presenting with a chief complaint of either hematemesis or bright red blood per rectum, there exists a gray zone in localizing the source of bleeding when a child presents with melena or altered blood in the stool. Frequently, this child undergoes a wide variety of costly and invasive diagnostic procedures in order to determine the site of blood loss. A useful and inexpensive means of identifying upper gastrointestinal bleeding, however, lies in an evaluation of the blood urea nitrogen to creatinine ratio (BUN/Cr).

In a retrospective study of 40 children hospitalized for evaluation and treatment of gastrointestinal bleeding at the Children's Hospital of Los Angeles, a BUN/Cr ratio > 30 indicated an upper gastrointestinal bleeding source with a specificity of 100%. Documentation of the source of bleeding was confirmed by endoscopy, surgery, or presence of hematemesis or bright red blood per rectum. The rise in BUN after an upper gastrointestinal hemorrhage is probably a result of increased hepatic catabolism of the absorbed amino acid load from the intraluminal blood. The bleeding, therefore, must occur proximal to the small intestine's absorptive surface (e.g., proximal to the ligament of Treitz) in order to cause a significant rise in the BUN/Cr ratio. The sensitivity of the BUN/Cr ratio for upper gastrointestinal bleeding, however, was 39%, because there existed a wide range of BUN/Cr ratios among the upper GI bleeders (range = 10–140; mean = 34 ± 29 SD) in comparison to the lower GI bleeders (range = 3.3–30; mean = 16 ± 8.5 SD). A BUN/Cr ratio less than or equal to 30, therefore, can be consistent with either an upper or a lower gastrointestinal bleeding site. The wide range of BUN/Cr ratios seen in children with upper gastrointestinal bleeding may
have been due to smaller volumes of blood loss, vomiting, or more rapid gastrointestinal transit times in children when compared to adults.


**Kool-Aid Colitis**

A mother brings her child to your Emergency Room frightened by the new onset of bright red stools in the child's diaper. The child appears hemodynamically stable and is without abdominal tenderness. What do you do?

If the stool guaiac is negative, ask the mother whether her child has had any cherry- or strawberry-containing beverages. Don't stop there. Ask about beets, tomato-containing products, and cherry or strawberry candies. Any and all of these substances can be the culprit when an alarmingly scarlet, nonclotting effluent accompanies a stool.


**GENETICS**

What is a Human Being's "Unique Genetic Make-up"?

The uniqueness of each individual stems from the fact that over one-fifth of his or her genes (i.e., proteins) are in a form that differs from that present in the majority of the population. All human diseases are a result of the interaction of the person's unique genetic make-up and the environment, and therefore genetics can be considered the basis of all medicine.


**Ethnicity As a Risk Factor**

Disorders transmitted by the inheritance of a single mutant gene, termed mendelian or "simply inherited disorders," occur with increased frequency in specific ethnic groups. Some examples of these relationships are listed in the table below.

**Examples of Simply Inherited Disorders that Occur with Increased Frequency in Specific Ethnic Groups**

<table>
<thead>
<tr>
<th>ETHNIC GROUP</th>
<th>SIMPLY INHERITED DISORDER</th>
</tr>
</thead>
<tbody>
<tr>
<td>African blacks</td>
<td>Hemoglobinopathies, especially Hb S, Hb C, persistent Hb F, α and β thalassemias</td>
</tr>
<tr>
<td>Armenians</td>
<td>Glucose-6-phosphate dehydrogenase deficiency</td>
</tr>
<tr>
<td>Armenians</td>
<td>Familial Mediterranean fever</td>
</tr>
</tbody>
</table>

Table continued on next page.
# Examples of Simply Inherited Disorders that Occur with Increased Frequency in Specific Ethnic Groups (Cont.)

<table>
<thead>
<tr>
<th>ETHNIC GROUP</th>
<th>SIMPLY INHERITED DISORDER</th>
</tr>
</thead>
</table>
| Ashkenazi Jews | Abetalipoproteinemia  
|               | Bloom's syndrome  
|               | Dystonia musculorum deformans (recessive form)  
|               | Factor XI (PTA) deficiency  
|               | Familial dysautonomia (Riley-Day syndrome)  
|               | Gaucher's disease (adult form)  
|               | Neimann-Pick disease  
|               | Pentosuria  
|               | Tay-Sachs disease |
| Chinese | $\alpha$-Thalassemia  
|         | Glucose-6-phosphate dehydrogenase deficiency  
|         | Adult lactase deficiency |
| Eskimos | Pseudochoolinesterase deficiency  
|         | Adrenogenital syndrome |
| Finns | Congenital nephrosis  
|         | Mulibrey nanism |
| French Canadians | Tyrosinemia |
| Japanese | Acatalasemia |
| Lebanese | Homozygous familial hypercholesterolemia |
| Mediterranean peoples | $\beta$-Thalassemia  
| (Italians, Greeks, Sephardic Jews) | Glucose-6-phosphate dehydrogenase deficiency  
|               | Familial Mediterranean fever  
|               | Glycogen storage disease, type III |
| Northern Europeans | Cystic fibrosis |
| Scandinavians | $\alpha_1$-antitrypsin deficiency  
|               | LCAT (lecithin:cholesterol acyltransferase) deficiency |
| South African whites | Porphyria variegata  
|                     | Homozygous familial hypercholesterolemia |


## GROWTH

### Predicted Weight and Height from Age

If you know the age of a child and want a rough estimate of the weight and height:

- **For 3–12 mo:** \[ \text{Weight (lb)} = \text{age (mo)} + 11 \]
- **For 1–6 yr:** \[ \text{Weight (lb)} = \text{age (yr)} \times 5 + 17 \]
- **For 7–12 yr:** \[ \text{Weight (lb)} = \text{age (yr)} \times 7 + 5 \]
- **For 2–14 yr:** \[ \text{Height (in)} = (2\frac{1}{2} \times \text{age}) + 30 \]

More on Predicted Heights

The formula of Tanner et al. demonstrates that height at age 3 years correlates better with height at maturity than it does at any other age:

Adult height (cm) = 1.27 \times \text{height (at 3 yr)} + 54.9 \text{ cm (males)}

Adult height (cm) = 1.29 \times \text{height (at 3 yr)} + 42.3 \text{ cm (females)}

If you cannot remember these formulas or your programmable calculator has been stolen, the commonly accepted statement that the child at age 2 has achieved one half his or her final height is quite satisfactory. For girls, however, 10 to 12 cm (2.54 to 4.00 in) must be subtracted from this predicted height. If the height at 3 years is known, an alternative to the Tanner equation to predict final adult height is to multiply the age 3 height by 1.87 for boys and 1.73 for girls.


Growth During Infancy: Weight

When assessing the growth of infants in a well-child care setting, a handy rule of thumb is offered: the newborn infant typically loses between 5 and 10% of his or her birth weight during the first few days of life (due to water loss). After that, the infant should gain 1 ounce or 30 grams, on average, per day. This weight gain, approximately 1 to 2 pounds per month, results in a doubling of the birth weight by six months of age. By the age of 2 years, weight gain should slow down to about one-half pound per month. Please note that although this tip is quite useful in the growing infant, it is not sensitive in the older child.

Growth During Infancy: Length

The greatest rise in linear growth occurs during infancy (the first 2 years of life). The average length of a newborn American infant is 50.4 cm (2.0 SD) or 19.8 inches (0.8 SD) in males and 49.7 cm (1.9 SD) or 19.6 inches (0.75 SD) in females. The growth increment during this period should be 25 to 30 cm (10 to 12 in) during the first year of life and 12 cm (5 in) during the second year of life. Male infants are typically heavier (by about 0.5 kg) and longer (by about 0.5 cm) than their female counterparts.

Growth During Infancy: Cranial and Brain Growth

The human brain begins its peak growth rate at birth and during the postnatal period. It should be noted that at birth, the infant's brain is only one-sixth of its final weight. Consequently, the growth of the cranial vault parallels this rapid development in order to accommodate the increasing brain size. In fact, careful evaluation of the skull size (head circumference) and shape gives the pediatrician a great deal of insight into the infant's neurologic development.

Mean head circumference at birth is approximately 35.3 cm (1.2 SD) or 13.9 inches (0.5 SD). There should be a 5 cm increase in head circumference during
the first 3 months of life and an additional 6 cm increase by the end of the first year of life. Whereas the head circumference of a newborn infant is greater than his or her chest, this ratio should approximate 1:1 by age 1 year. (N.B.: Closure of the fontanel is covered under “Fontanels.”)

GYNECOMASTIA

The Differential Diagnosis of Gynecomastia

Gynecomastia is defined as the visible or palpable development of breast tissue in boys or men. It has been divided into four types:

Type I gynecomastia (pubertal gynecomastia or benign adolescent breast hypertrophy) refers to the common entity seen in pubertal males. In fact, many cite an incidence of 60 to 70% in this population. It is typically a firm, tender subareolar mass anywhere from 1 to 5 cm in diameter. The pubertal adolescent frequently complains of pain in the breasts, particularly when wearing binding clothing. It usually spontaneously resolves within 2 years.

Type II gynecomastia (physiologic gynecomastia without evidence of underlying disease, or with evidence of organic disease including the effects of specific drugs) refers to a generalized, nonpainful breast enlargement. It is essential to differentiate between physiologic gynecomastia and breast enlargement due either to a pathologic process or to the use of a specific drug. The physician should, therefore, obtain a careful history regarding the time of onset, family history, duration of the enlargement, history of systemic illness, weight change, and drug or medication use. Physical examination should include height, weight, blood pressure, breast size, and Tanner staging of both breasts and genitals, in addition to a neurologic assessment. The most frequent causes of Type II gynecomastia are listed in tabular form below.

Type III gynecomastia is general obesity simulating gynecomastia, and Type IV gynecomastia is pectoral muscle hypertrophy.

Common Causes of Type II Gynecomastia

I. Idiopathic
II. Familial causes
   a. Associated with anosmia and testicular atrophy
   b. Reifenstein's syndrome (a type of familial male pseudohermaphroditism secondary to partial androgen insensitivity)
   c. Associated with hypogonadism and small penis
   d. Others
III. Specific illnesses or syndromes
   a. Kleinfelter's syndrome
   b. Male pseudohermaphroditism
   c. Testicular feminization syndrome
   d. Tumors (e.g., seminoma, Leydig cell tumor, teratoma, feminizing adrenal tumor, hepatoma, bronchogenic carcinoma)
   e. Leukemia
   f. Hemophilia
   g. Leprosy
   h. Thyroid dysfunction (hyper- and hypothyroidism)
   i. Cirrhosis of the liver
   j. Traumatic paraplegia
   k. Chronic glomerulonephritis
   l. Starvation (on refeeding)
IV. Miscellaneous Drugs
   a. Amphetamines
   b. Anabolic steroids
   c. Birth control pills
   d. Busulfan (and other chemotherapeutic agents)
   e. Cimetidine
   f. Clomiphene
   g. Diazepam
   h. Corticosteroids
   i. Digitalis
   j. Estrogens
   k. Human chorionic gonadotropin
   l. Insulin
   m. Isoniazid (and other antituberculosis drugs)
   n. Ketoconazole
   o. Marijuana
   p. Methadone (and other narcotics)
   q. Methyldopa
   r. Reserpine
   s. Spironolactone
   t. Testosterone
   u. Tricyclic antidepressants

Head Circumference in Term Infants

When you are stuck without your growth grid, there is still a simple means of determining if a head is growing normally. The little table below lists the expected rate of increases in head circumference for term infants during the first year of life.

<table>
<thead>
<tr>
<th>PERIOD</th>
<th>HEAD CIRCUMFERENCE INCREMENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>First 3 months</td>
<td>2 cm/month = 6 cm</td>
</tr>
<tr>
<td>4-6 months</td>
<td>1 cm/month = 3 cm</td>
</tr>
<tr>
<td>6-12 months</td>
<td>0.5 cm/month = 3 cm</td>
</tr>
<tr>
<td>First year</td>
<td>12 cm</td>
</tr>
</tbody>
</table>

HEADACHE

Common Causes

- Extracranial infection
  - Otitis/mastoiditis
  - Pharyngitis
  - Sinusitis
  - Tooth abscess
- Febrile illness
- Migraine
- Tension
- Anxiety
- Environmental stress

Uncommon Causes

- Depression
- Eye strain
- Meningitis/encephalitis
- Temporomandibular joint disease

- Trauma
- Concussion
- Occipital neuralgia

Rare Causes

- Allergy
- Arnold-Chiari malformation
- Cervical osteoarthritis
- Chronic renal disease
- Congenital erythropoietic porphyria
- Cranial bone disease
- Decreased intracranial pressure
- Post-lumbar puncture
HEARING

Which Infant Is at Risk for Hearing Loss?

Long before a delay in language development secondary to a hearing loss is noted by a parent or physician, the astute pediatrician can single out infants at risk. Screening for hearing loss is relatively inexpensive and easy to perform, and the benefits of early detection are immeasurable. Be aware of the risk factors and look for the absence of the normal newborn’s response to sound: startling, blinking, crying, quieting, or other forms of alertness are the normal newborn’s reactions to sound.

Factors that Mandate Screening for Hearing Loss

A blood relative with childhood hearing impairment.
Anatomic malformations involving the head and neck.
Bacterial meningitis—especially *H. influenzae*.
Birthweight less than 1,500 g.
Severe asphyxia as evidenced by low Apgar scores, arterial pH ≤ 7.25, coma, seizures, or the need for continuous assisted ventilation.
Unconjugated bilirubin > 17 mg/100 ml of serum.
Viral or other nonbacterial intrauterine fetal infections.
Any neonate with known congenital defect: Recessive syndromes constitute 4% to 40% of childhood deafness; dominant syndromes constitute about 15%; x-linked syndromes about 2%; and rubella, 9 to 20% of cases of childhood deafness.

Screening Tests

These screening tests allow evaluation of the infant at risk as early as 2 weeks of age:

1. Crib-o-gram: Crib movement is recorded in response to sound stimulus application.
2. Brainstem auditory evoked response (BAER): More sensitive than a crib-o-gram, BAERs record electrical potential at the skin overlying the nuclei and tracts of the auditory pathway.


Exogenous Causes of Hearing Loss

In addition to heredity, gestational age, dysmorphisms, and asphyxia, there are several exogenous causes of hearing loss to consider in the evaluation of communicative delay in infants and children.

Preconception and Prenatal

Cytomegalovirus
Rubella
Hypoxia
Syphilis
Maternal drug abuse and alcoholism
Toxemia, diabetes, or other severe maternal systemic disease
Ototoxic drugs (quinine)
Toxoplasmosis
Irradiation of parent

Perinatal

Hypoxia
Prematurity
Maternal infection
Traumatic delivery
Ototoxic drugs (aminoglycosides)

Neonatal and Postnatal

Erythroblastosis fetalis
Meningitis, encephalitis
Hypoxia
Ototoxic drugs (aminoglycosides)
Infantile measles or mumps

HEMATOLOGY

Laboratory Test Results in Disorders Producing Hypochromia and Microcytosis

<table>
<thead>
<tr>
<th>DISORDER</th>
<th>SERUM IRON</th>
<th>IRON-BINDING CAPACITY</th>
<th>FERRITIN</th>
<th>FEP*</th>
<th>HEMOGLOBIN ELECTROPHORESIS</th>
<th>MARROW IRON STORES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Iron deficiency</td>
<td>Decreased</td>
<td>Increased</td>
<td>Decreased</td>
<td>Increased</td>
<td>Normal</td>
<td>Decreased</td>
</tr>
<tr>
<td>Chronic disease anaemia</td>
<td>Decreased</td>
<td>Decreased</td>
<td>Increased</td>
<td>Increased</td>
<td>Normal</td>
<td>Increased</td>
</tr>
<tr>
<td>Sideroblastic anaemia</td>
<td>Increased</td>
<td>Normal</td>
<td>Increased</td>
<td>Decreased</td>
<td>Normal</td>
<td>Ring sideroblasts</td>
</tr>
<tr>
<td>β-thalassemia trait</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Hemoglobin A₃ increased</td>
<td>Normal</td>
</tr>
<tr>
<td>α-thalassemia trait</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
</table>

*FEP indicates free erythrocyte protoporphyrin.
Adapted from Steinberg MH, Dreiling BJ: Microcytosis. JAMA 249:85, 1983, with permission.

HEMATURIA

Common Causes

Benign causes
- Benign recurrent hematuria
- Familial hematuria
- Idiopathic recurrent gross hematuria
- Postural hematuria

Contamination
- Menstrual
- Munchausen’s syndrome
- Munchausen’s syndrome by proxy
- Pregnancy-related bleeding

Hemoglobinopathies
- Hgb C
- Hgb SC
- Sickle-cell disease/trait (Hgb SS/SA)
- Sickle-thalassemia trait

Hypercalciuria
- Distal renal tubular acidosis
- Diuretic therapy
- Endocrine disorders
  - Diabetes mellitus

Hypercalciuria (Cont.)
- Endocrine disorders (Cont.)
  - Hyperadrenocorticism
  - Hyperparathyroidism
  - Hypothyroidism

Hypocalcemia

Hyperphosphatemia

Hypertension

Immobilization

Juvenile rheumatoid arthritis

Medullary sponge kidney

Metabolic acidosis

Neoplasm

Renal tubular dysfunction

Sarcoidosis

Vitamin D excess

Hypoxia, asphyxia, and circulatory compromise

Acute tubular necrosis

Cortical and medullary necrosis

155
Infections
- Cystitis (viral, bacterial)
- Pyelonephritis
- Urethritis
- Meatal stenosis
- Noninfectious cystitis
  - Cytoxan
  - Radiation
- Perineal irritation
- Phimosis

Post-infectious glomerulonephritis

Trauma
- Fractured pelvis
- Postcatheterization
- Postcircumcision
- Postsurgery
- Renal contusion
- Renal fracture
- Urethral trauma
- Urethral ulceration

Uncommon Causes

Bladder diverticuli/polyps

Coagulopathies

Drug-induced
  - Analgesic nephropathy
  - Cephalosporins
  - Cytoxan
  - Penicillin
  - Sulfonamides

Exercise

Glomerular disorders
  - Mesangioproliferative
  - Minimal change disease

Hydronephrosis

Infections
  - Epididymitis
  - Prostatitis

Masturbation

Periureteritis (appendicitis, ileitis)

Polycystic disease

Reflux nephropathy

Renal calculi

Renal vein thrombosis

Thrombocytopenia

Ureteropelvic junction obstruction

Urethral foreign body

Wilms' tumor

Rare Causes

Allergy
  - "Apparent"
    - "Beeturia"
  - Betadine
  - Biliuria
  - Desferoxamine
  - Dyes
    - Analine
    - Congo red
  - Hemoglobinuria
  - Myoglobinuria
  - Phenothiazines
  - Porphyria

Diabetic nephropathy

Glomerular disorders
  - Amyloidosis
  - Crescentic glomerulonephritis (GN)
  - Familial nephritis (Alport's)
  - Focal segmental proliferative GN
  - Focal segmental sclerosis

Goodpasture's syndrome

IgA nephropathy

Membranous GN

Mesangiocapillary GN

Subacute bacterial endocarditis

Systemic lupus erythematosus (SLE)

Wegener's granulomatosis

Hemangioma

Hematospermia

Immunologic
  - Hemolytic-uremic syndrome
  - Henoch-Schönlein purpura
  - Polyarteritis nodosa

SLE

Infections
  - Leptospirosis
  - Malaria
  - Schistosomiasis
  - Toxoplasmosis
Evaluation of Hematuria in Children and Adolescents

After urinary tract infections, hematuria is the most frequently occurring abnormality in the genitourinary tract. One must carefully assess the complaint of hematuria in order to rule out or diagnose from a large list of potentially serious disorders. It is important, first, to assess whether or not actual blood or simply a red substance is being excreted into the urine. A reagent strip (impregnated with orthotolodine-peroxide and enhanced with 6-methoxyquinolone) makes this quite simple. False-negative results with the dipstick method are rare but can result from the presence of high urinary concentrations of ascorbic acid. False-positive results, which are also rare, can occur in the presence of a raging urinary tract infection where bacterial peroxidase is released in high quantities. Following a positive dipstick microscopic evaluation is required; if no red blood cells (RBCs) are noted in a fresh urine sample, one should entertain the diagnosis of hemoglobinuria or myoglobinuria.

Although gross hematuria is easily recognized upon visual inspection, the urine should always be examined with dipstick and microscopic analysis to rule out other causes of red urine. Microscopic hematuria is usually defined as (a) three or more consecutive urine samples with positive reagent strip test results and either two or more RBCs per cubic millimeter in a fresh, uncentrifuged urine sample or (b) six or more RBCs per high-power field in a fresh urine sediment specimen.

Common Causes of Red Urine

1. **Negative Dipstick Test** (e.g., dyes, drugs, pigments)
   a. Pink, red, Coca-Cola, burgundy-colored urine (drug and food ingestion):
      Aminopyrine, anthocyanin, azo dyes, beets,* blackberries, chloroquine, desferoxamine, mesylate, ibuprofen, methyldopa, nitrofurantoin, phenazopyridine, phenolphthalein, rifampin, rhodamine B, sulfasalazine, urates.
   b. Dark brown, black:
      i. Disease-associated: alkaptonuria, homogentisic aciduria, melanin, methemoglobinemia, tyrosinosis
      ii. Drug and food ingestion: alanine, resorcinol, thymol.

2. **Positive Dipstick Test but No RBCs on Microscopic Examination**
   a. Hemoglobinuria
      i. Drugs and chemicals: aspidium, betanaphthyl, carbolic acid, carbon monoxide, chloroform, fava beans, mushrooms, naphthalene, pamaquine, phenylhydrazine, quinine, snake venom, sulfonamides.

*The excretion of red-purple or beet-colored urine (beeturia) after ingesting beets should prompt the clinician to work up the patient for iron-deficiency anemia.
ii. Disease-associated and other causes: all types of hemolytic anemias, hemolytic-uremic syndrome, septicemia, paroxysmal nocturnal hemoglobinuria, freshwater drowning, mismatched blood transfusions, cardiopulmonary bypass.

3. Positive Dipstick Tests with RBCs on Microscopic Examination
   a. No RBC casts present: tumors, cysts, stones, obstruction
   b. RBC casts present:
      i. Without proteinuria: IgA nephropathy, familial nephritis, benign or familial hematuria (in association with hypercalciuria)
      ii. With proteinuria: Acute glomerulonephritis, Henoch-Schönlein purpura, systemic lupus erythematosus, chronic glomerulonephritis
     iii. Heavy proteinuria: Nephrotic syndrome

The basic work-up and evaluation for the patient with hematuria should proceed as follows:

1. History (associated symptoms, precipitating events, pattern of hematuria, familial occurrence).
2. Physical examination (presence of edema, elevated blood pressure, skin lesions, joint involvement).
3. Laboratory tests
   a. Urinalysis (with confirmation of microscopic hematuria on two or more occasions).
   b. Urine culture (gross hematuria occurs in 5% to 10% of children with symptomatic urinary tract infection; the frequency of those children with symptomatic urinary tract infection and microscopic hematuria is not known).
   c. Complete blood count with examination of peripheral smear
   d. Serum creatinine
   e. ASO titer, streptozyme, C3 complement, anti-DNA antibody (e.g., poststreptococcal glomerulonephritis, SLE, proliferative GN)
   f. Quantitative urine protein, calcium, creatinine excretion
   g. Imaging studies (e.g., ultrasonography of the urine tract, intravenous pyelography, voiding cystourethrography).


Hematuria After Blunt Trauma

Hematuria is a common finding in the aftermath of blunt trauma to the abdomen. Traditional thinking suggests that hematuria is a significant consequence of genitourinary (GU) injury, and its presence typically elicits consideration of radiologic imaging of the GU tract. Which patient, in fact, requires a study and which study should you choose?
Hematuria

Asymptomatic hematuria is not an indication for computed tomography (CT) of the abdomen. In a study of 378 consecutive children evaluated by CT of the abdomen, those who were asymptomatic had no evidence of organ injury. A child with no signs or symptoms of abdominal injury (e.g., tenderness, ecchymoses, distention) despite the presence of hematuria does not warrant a CT exam. Children with asymptomatic hematuria may be evaluated on a nonemergent basis with Doppler ultrasonography, excretory urography, or a radioactive renal scan. In contrast, hematuria with abdominal symptoms is a significant marker of injury to both urinary and nonurinary organs in the setting of blunt abdominal trauma.


Drug-induced Causes of Hematuria

<table>
<thead>
<tr>
<th>THE DRUG</th>
<th>THE DISORDER</th>
</tr>
</thead>
<tbody>
<tr>
<td>Penicillin and cephalosporin analogues, phenytoin</td>
<td>Allergic interstitial nephritis</td>
</tr>
<tr>
<td>Phenacetin, nonsteroidal anti-inflammatory agents</td>
<td>Papillary necrosis</td>
</tr>
<tr>
<td>Cyclophosphamide, mitotane</td>
<td>Chemical cystitis</td>
</tr>
<tr>
<td>Cyclophosphamide, phenacetin</td>
<td>Malignant neoplasm of uroepithelium</td>
</tr>
<tr>
<td>Anticoagulants</td>
<td>Spontaneous bleeding or induction of bleeding from an occult lesion</td>
</tr>
</tbody>
</table>


Neonatal Hematuria

Gross hematuria is a rare presentation during the first month of life. The findings in one series of 35 patients are demonstrated below.

Findings in Series of 35 Patients

<table>
<thead>
<tr>
<th>CAUSE</th>
<th>NO. OF PATIENTS</th>
<th>AGE AT ONSET</th>
<th>REMARKS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Unknown</td>
<td>11</td>
<td></td>
<td>1 died of hyaline membrane disease and pneumothorax; normal BUN level in 9, elevated in 2</td>
</tr>
<tr>
<td>Renal vein thrombosis</td>
<td>7</td>
<td>3</td>
<td>3 had diabetic mothers; Bun level &gt; 40 mg/100 ml in all; 4 had thrombocytopenia</td>
</tr>
</tbody>
</table>

Table continued on next page.
Findings in Series of 35 Patients (Cont.)

<table>
<thead>
<tr>
<th>CAUSE</th>
<th>NO. OF PATIENTS</th>
<th>AGE AT ONSET</th>
<th>REMARKS</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>1ST WEEK</td>
<td>2ND WEEK</td>
</tr>
<tr>
<td>Polycystic disease of kidney</td>
<td>6</td>
<td>6</td>
<td></td>
</tr>
<tr>
<td>Obstructive uropathy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hydronephrosis</td>
<td>3</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Ureteral valve</td>
<td>3</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td>Bladder neck</td>
<td>1</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td>Sponge kidney</td>
<td>3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wilms' tumor</td>
<td>2</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Abdominal masses were palpated in all patients later found to have renal vein thrombosis or polycystic kidneys. Intravenous pyelograms were normal in all patients in whom no cause was found for the hematuria, in the patients with posterior urethral valves, and in the one with bladder neck obstruction. IVP was abnormal in all other patients. Voiding cystourethrogram demonstrated the abnormality in the patients with obstruction.

**Conclusion:** Abdominal palpation, IVP, and blood urea nitrogen levels are warranted in all newborns presenting with gross hematuria. If the diagnosis is still unavailable, voiding cystourethrogram is in order. A significant number of these patients, however, will have no evident cause for their hematuria, and will recover spontaneously.


**HEMOGLOBIN**

**Classification of Red Cell Hemolytic Disorders by Predominant Morphology**

In the following lists, nonhemolytic disorders of similar morphology are enclosed in parentheses for reference.
148—Hemoglobin

**Spherocytes**
Hereditary spherocytosis
ABO incompatibility in neonates
Immunohemolytic anemias with IgG- or C3-coated red cells
Acute oxidant injury (hexose monophosphate shunt defects during hemolytic crisis, oxidant drugs and chemicals)
Hemolytic transfusion reactions
*Clostridium welchii* septicemia
Severe burns, other red cell thermal injury
Spider, bee, and snake venoms
Severe hypophosphatemia
Hypersplenism*

**Bizarre Polkilocytes**
Red cell fragmentation syndromes
(\(\text{\textdollar}\)) micro- and macroangiopathic hemolytic anemias)
Acute oxidant injury*
Hereditary elliptocytosis in neonates
Hereditary pyropoikilocytosis

**Elliptocytes**
Hereditary elliptocytosis
Thalassemias
(Other hypochromic-microcytic anemias)
(Megaloblastic anemias)

**Stomatocytes**
Hereditary stomatocytosis
Rh\(_{null}\) blood group
Stomatocytosis with cold hemolysis
(Liver disease, especially acute alcoholism)
(Mediterranean stomatocytosis)

**Irreversibly Sickled Cells**
Sickle cell anemia
Symptomatic sickle syndromes

**Intraerythrocytic Parasites**
Malaria
Babesiosis
Bartonellosis

**Spiculated or Crenated Red Cells**
Acute hepatic necrosis (spur cell anemia)
Uremia
Red cell fragmentation syndromes*
Infantile pyknocytosis
Embden-Meyerhof pathway defects*
Vitamin E deficiency*
Abetalipoproteinemia
Heat stroke*
McLeod blood group
(Postsplenectomy)
(Transiently after massive transfusion of stored blood)
(Anorexia nervosa)*

**Target Cells**
Hemoglobins S, C, D, and E
Hereditary xerocytosis
Thalassemias
(Other hypochromic-microcytic anemias)
(Obstructive liver disease)
(Postsplenectomy)
(Lecithin:cholesterol acyltransferase deficiency)

**Prominent Basophilic Stippling**
Thalassemias
Unstable hemoglobins
Lead poisoning*
Pyridine 5'-nucleotidase deficiency

**Nonspecific or Normal Morphology**
Embden-Meyerhof pathway defects
Hexose monophosphate pathway defects
Unstable hemoglobins
Paroxysmal nocturnal hemoglobinuria
Dyserythropoietic anemias
Copper toxicity (Wilson's disease)
Cation permeability defects
Erythropoietic porphyria
Vitamin E deficiency
Hemolysis with infections*
Rh hemolytic disease in neonates
Paroxysmal cold hemoglobinuria*

* Disease sometimes associated with this morphology.
The Rise in Hemoglobin with Iron Therapy

How fast should the hemoglobin rise when you start treating your iron deficient patient with oral iron?

Two important factors must be considered when judging the adequacy of the hematologic response. They are (1) the initial hemoglobin value and (2) the duration of the period of observation. The lower the initial hemoglobin, the greater is the hemoglobin rise per day. The shorter the observation period, the greater is the calculated hemoglobin rise per day. As one gets closer to a normal hemoglobin value, the daily rise in hemoglobin is much less. In our own experience in treating patients with hemoglobin values of less than 8.0 g/100 ml, one may anticipate a hemoglobin rise of 0.2 to 0.3 g per day and during the first 7 to 10 days of therapy. During the period of 10 to 24 days, the hemoglobin rises at a rate of 0.15 g per day and slows after that point to a rate of 0.10 per day, until a normal level is achieved. Normally, the reticulocyte count begins to increase in 48 to 72 hours and reaches a peak 7 to 10 days after the initiation of therapy.

Listed below is another guide to the expected response as a function of the initial hemoglobin level.

<table>
<thead>
<tr>
<th>INITIAL HEMOGLOBIN (g/dl)</th>
<th>HEMOGLOBIN RISE IN ONE WEEK (g/dl)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.0 - 5.0</td>
<td>1.61</td>
</tr>
<tr>
<td>5.1 - 6.0</td>
<td>1.53</td>
</tr>
<tr>
<td>6.1 - 7.0</td>
<td>1.17</td>
</tr>
<tr>
<td>7.1 - 8.0</td>
<td>1.11</td>
</tr>
<tr>
<td>8.1 - 9.0</td>
<td>0.98</td>
</tr>
<tr>
<td>9.1 - 10.0</td>
<td>0.57</td>
</tr>
<tr>
<td>10.1 - 11.0</td>
<td>0.72</td>
</tr>
<tr>
<td>11.1 - 12.0</td>
<td>0.40</td>
</tr>
</tbody>
</table>

Optimal responses to oral iron therapy are achieved by treating the patient with ferrous sulfate. A patient should receive 2 to 3 mg of elemental iron per kg three times per day. The iron should be given between meals and never administered with milk.


HEMOPTYSIS

The Child with Hemoptysis

Hemoptysis is defined as the spitting up of blood that originates from the lungs or bronchial tree. Although this is a rare sign in children, it can be potentially life-threatening. Rapid and thorough evaluation is vital in order to identify and control the source of bleeding.
Hemoptysis

In the work-up of the child with hemoptysis, it is necessary to begin by anatomically delineating where the bleeding is actually occurring. Most children who spit up blood have an identifiable source of bleeding outside of the lower respiratory tract, such as epistaxis or trauma to the oropharynx. Differentiating hemoptysis from hematemesis, on the other hand, can be difficult. Distinguishing features of these two signs are summarized below:

Hemoptysis

- Blood is usually bright red, frothy, and mixed with sputum (pus, organisms, and macrophages may be present).
- Anemia is not a common finding.
- Bleeding is often preceded by a gargling noise and is associated with coughing.
- pH of blood is alkaline.

Hematemesis

- Blood is usually dark red to brownish in color (e.g., coffee ground emesis).
- Often associated with anemia.
- Bleeding and emesis are often preceded or accompanied by nausea and retching.
- pH is acidic.

The different causes of hemoptysis are wide and varied, as detailed below:

1. Infectious Causes
   a. Bacterial
      i. Bronchiectasis (most commonly associated with cystic fibrosis, immunodeficiency disorders, dyskinetic cilia syndrome, and pertussis).
      ii. Necrotizing pneumonias (e.g., Pseudomonas, Staphylococcus, Klebsiella)
      iii. Pulmonary tuberculosis
      iv. Lung abscess
      v. Bronchitis
      vi. Tracheitis
      vii. Pertussis

   b. Fungal
      i. Aspergillosis
      ii. Coccidioidomycosis
      iii. Actinomycosis
      iv. Mucormycosis
      v. Candidiasis

   c. Parasitic
      i. Paragonimiasis
      ii. Echinococcosis
      iii. Strongyloidiasis
      iv. Ancylostomiasis

   d. Viral
      i. Laryngitis
      ii. Laryngotracheobronchitis
      iii. Pneumonitis

2. Foreign Bodies

3. Intrathoracic Defects and Lesions (Congenital or Acquired)
   a. Pulmonary arteriovenous fistula
   b. Hemangiomatous malformation
   c. Neurenteric cyst
   d. Bronchogenic cyst
   e. Mitral stenosis
   f. Other cardiac anomalies
   g. Pulmonary embolism and infarction
   h. Aortic aneurysm
   i. Pulmonary sequestration
   j. Arteriovenous fistula
   k. Venous obstructive condition
   l. Anomalous vessel
   m. Congenital telangiectasia
Henoch-Schönlein Purpura

4. Autoimmune Conditions
   a. Wegener's granulomatosis
   b. Pulmonary hemosiderosis
   c. Milk allergy
   d. Goodpasture's syndrome
   e. Collagen vascular disease

5. Trauma
   a. Compression, crush injury, or penetrating injury
   b. Iatrogenic (e.g., postsurgical, postdiagnostic lung puncture, posttransbronchial biopsy, barotrauma)

6. Neoplastic conditions
   a. Endobronchial metastasis (e.g., metastatic osteogenic sarcoma)
   b. Bronchial adenoma
   c. Mediastinal teratoma
   d. Choriocarcinoma
   e. Endometriosis
   f. Bronchiogenic carcinoma

7. Hemoglobinopathy with pulmonary infarct (e.g., the “chest syndrome” of sickle cell disease)

8. Factitious (as a manifestation of Munchausen syndrome)


HENOCHE-SCHÖNLEIN PURPURA

Neurologic Manifestations of Henoch-Schönlein Purpura (HSP)

Although the classic triad of purpuric rash, arthritis, and crampy abdominal pain remains pathognomonic for Henoch-Schönlein purpura, nervous system involvement may be more common than we believe. Osler first described neurologic manifestations of HSP in 1914. It has not been until recently, however, that the protean neurologic symptoms have been recognized. Histopathology has demonstrated the characteristic fibrinoid necrosis in meningeal and cerebral parenchymal arterioles and small arteries that is found in the cutaneous lesions, Gl blood vessel walls, and kidney mesangium (nephritis is also common in HSP).

The neurologic symptoms of HSP can be nonspecific or dramatic, transient or permanent, primary or late. Regardless of the case, the manifestations may be indicative of significant CNS disease requiring treatment and/or neurologic assessment and follow-up.

CNS Involvement in HSP

<table>
<thead>
<tr>
<th>Headache</th>
<th>Focal neurologic deficits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mental status changes</td>
<td>Aphasia</td>
</tr>
<tr>
<td>Behavior</td>
<td>Hemiparesis</td>
</tr>
<tr>
<td>Depressed state of consciousness</td>
<td>Paraparesis</td>
</tr>
<tr>
<td>Seizures</td>
<td>Quadriplegia</td>
</tr>
<tr>
<td>Partial, partial complex, generalized</td>
<td>Cortical blindness</td>
</tr>
<tr>
<td>Status epilepticus</td>
<td>Chorea</td>
</tr>
<tr>
<td></td>
<td>Ataxia</td>
</tr>
</tbody>
</table>
Peripheral NS Involvement in HSP

Mononeuropathy
- Facial nerve
- Ulnar nerve
- Femoral nerve
- Sciatic nerve
- Peroneal nerve

Polyneuropathy
- Guillain-Barré
- Polyradiculoneuropathy
- Brachial plexopathy


HEPATOMEGALY

Common Causes

Benign cystic disease
Benign transient hepatomegaly
(usually with GI viral illness)
Biliary tract obstruction
- Alagille's disease
- Ascending cholangitis
- Biliary atresia
- Choledochal cyst
Congestive heart failure
Cystic fibrosis
Diabetes mellitus
Hyperalimentation
Iron-deficiency anemia
Leukemia, lymphoma
Malnutrition
Maternal diabetes
Neonatal hepatitis

Pulmonary hyperinflation
("apparent" hepatomegaly)
Septicemia
Sickle-cell anemia
Toxin/drug reactions (hepatitis, cholestasis, fatty infiltration)
Acetaminophen
Birth-control pills
Corticosteroids
Hydantoins
Phenobarbital
Sulfonamides
Tetracycline
Viral hepatitis
CMV, EBV, coxsackievirus
Hepatitis A, B, non-A, non-B

Uncommon Causes

Chronic active hepatitis
Chronic anemias
Erythroblastosis fetalis
Hartoma
Hemangioma
Hemolytic anemias
Hepatic abscess (pyogenic)
Hepatoblastoma
Inflammatory bowel disease
Liver hemorrhage
Metastatic tumors
Pericarditis
Reye's syndrome
Rocky Mountain spotted fever
Systemic inflammatory disease (e.g., JRA, SLE)
Visceral larva migrans

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### Rare Causes

<table>
<thead>
<tr>
<th>α₁-Antitrypsin deficiency</th>
<th>Infantile pyknocytosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amyloidosis</td>
<td>Infantile sialidosis</td>
</tr>
<tr>
<td>Beckwith-Wiedemann syndrome</td>
<td>Klippel-Trenaunay-Weber syndrome</td>
</tr>
<tr>
<td>Brucellosis</td>
<td>Lipodystrophy</td>
</tr>
<tr>
<td>Budd-Chiari syndrome</td>
<td>Malaria</td>
</tr>
<tr>
<td>Carnitine deficiency</td>
<td>Mannosidosis</td>
</tr>
<tr>
<td>Chediak-Higashi syndrome</td>
<td>Methylmalonic acidemia</td>
</tr>
<tr>
<td>Crigler-Najjar syndrome</td>
<td>Moore-Federmann syndrome</td>
</tr>
<tr>
<td>Farber's disease</td>
<td>Mucolipidosis</td>
</tr>
<tr>
<td>Galectosemia</td>
<td>Mucopolysaccharidoses</td>
</tr>
<tr>
<td>Gaucher's disease</td>
<td>Niemann-Pick disease</td>
</tr>
<tr>
<td>Glycogen storage disease</td>
<td>Parasitic infections</td>
</tr>
<tr>
<td>Granulomatous hepatitis</td>
<td>Amebiasis</td>
</tr>
<tr>
<td></td>
<td>Flukes</td>
</tr>
<tr>
<td></td>
<td>Schistosomiasis</td>
</tr>
<tr>
<td>Chronic granulomatous disease</td>
<td>Rendu-Osler-Weber syndrome</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>Rickets</td>
</tr>
<tr>
<td>Tuberculosis</td>
<td>Tangier's disease</td>
</tr>
<tr>
<td>Hemochromatosis</td>
<td>Tyrosinemia</td>
</tr>
<tr>
<td>Hemophagocytic syndrome</td>
<td>Urea cycle defects</td>
</tr>
<tr>
<td>Hepatic porphyrias</td>
<td>Veno-occlusive disease</td>
</tr>
<tr>
<td>Hepatocellular carcinoma</td>
<td>Wilson's disease</td>
</tr>
<tr>
<td>Hereditary fructose intolerance</td>
<td>Wolman disease</td>
</tr>
<tr>
<td>Histiocytic syndromes</td>
<td>Zellweger syndrome</td>
</tr>
<tr>
<td>Histoplasmosis</td>
<td></td>
</tr>
<tr>
<td>Homocystinuria</td>
<td></td>
</tr>
<tr>
<td>Hyperlipoproteinemia 1</td>
<td></td>
</tr>
<tr>
<td>Hypervitaminosis A</td>
<td></td>
</tr>
</tbody>
</table>

---

**HERPES**

### Neonatal Herpes Simplex Virus Infections

Herpes simplex virus (HSV) infection of the newborn is a feared and potentially devastating illness. As herpes virus infections continue to rise among women in the child bearing years, so has the incidence of neonatal cases of HSV. To further complicate matters, at least 60% of neonates who develop HSV infections are born to women who have never recognized symptoms of genitourinary HSV infection. Common means of transmitting HSV to the neonate include: (1) vaginal delivery through an infected birth canal; (2) ascending infection after rupture of membranes in a woman shedding HSV from the cervix; (3) postnatal contact with individuals exhibiting active skin lesions; and (4) transplacental infections.

Serious neonatal HSV infection is less likely when the infant is born to a mother with recurrent lesions, apparently because maternal antibody provides some measure of protection. Eighty percent of neonatal HSV infections are caused by HSV-2 and 20% are due to HSV-1. Although serious neonatal disease
due to HSV-1 has been described, neonates infected with HSV-1 are more likely to survive without serious morbidity than those infected with HSV-2. Listed below are the common clinical manifestations of neonates with HSV infections:

**Disseminated HSV Disease** (mean age of presentation = 7 days)

- Fever
- Lethargy
- Convulsions or other CNS findings
- Poor feeding
- Respiratory distress
- Pneumonitis
- Shock
- Bleeding
- Hepatosplenomegaly
- Jaundice
- Skin lesions

**Localized HSV Disease** (mean age of presentation = 11 days)

- Skin and mucosal lesions (vesicular or ulcerated lesions on an erythematous base)
- Keratoconjunctivitis
- CNS findings

*Although 50% of all infants with disseminated HSV develop skin lesions, they may not be present with the onset of symptoms.*

*Up to 60% of infants with local involvement of the skin, eyes, or mouth will progress to disseminated disease.*


**HIP**

**Congenital Dislocation of the Hip: A New Diagnostic Method**

Congenital dislocation of the hip (CDH) is known to result from several etiologies (see below), including mechanical, hormonal, and hereditary factors. It occurs six times more frequently in otherwise normal females than in males. The Catch-22 of CDH is that it is difficult to diagnose in infants, but the success of treatment initiated after the child begins walking is poor.

In the past, pediatricians have relied on the Barlow and Ortolani tests to diagnose CDH in the neonate. These tests are only reliable in the newborn period but remain an effective screening test. Since approximately 60% of unstable hips in the newborn period resolve as laxity disappears, we need a reliable screening test for the infant from 2-4 months.

A recent study from Asahikawa, Japan, demonstrates the usefulness of abnormal or asymmetrical inguinal folds in the frog-leg position for diagnosing CDH in the 3-4 month old child. Although the number of false positives was high, the coincidence rates between radiographic diagnosis and abnormal inguinal folds for dislocation, subluxation, and acetabular dysplasia were 100%, 100%, and 47%, respectively. Comparative coincidence rates for the limited abduction test (Aliss' or Galeazzi's sign) were 0%, 60%, and 0%. Thus, abnormal inguinal folds are a more sensitive indicator of CDH than limited abduction and should reduce the number of radiologic exams performed on infants.
**Factors Involved in CDH**

**Mechanical:** Breech deliveries
First born child
Oligohydramnios

**Hormonal:** Generalized ligamentous laxity; results from increased circulating estrogens and relaxin at the time of birth (6:1).

**Hereditary:** Positive family history in 20% of cases of CDH.

**Physical Signs of CDH in 3-4 Month Old**
- Shortened leg
- Limited passive abduction
- Asymmetric folds of femoral skin
- Abnormal inguinal folds

**Tests for CDH**
- Barlow’s and Ortolani’s tests are performed with thumb and forefinger on the lesser and greater trochanters. Only one hip should be examined at a time.
  - **Barlow:** Adduction and posterior pressure may produce a “clunk” of subluxation or dislocation.
  - **Ortolani:** Abducting and “lifting” hip back into place relocates the dislocation caused by Barlow’s test.

**Abnormal inguinal folds:** See figure.

---

**Hirschprung’s Disease**

**Ruling Out Hirschprung’s Disease**

Hirschprung’s disease, or congenital megacolon, is the most common cause of obstruction of the colon in the neonatal period (about 33% of all neonatal obstructions).

Clinically, it is difficult to distinguish which infants have Hirschprung’s disease in the neonatal period. Meconium plug syndrome, cystic fibrosis, hypothyroidism, and many other abnormalities may present with constipation.
obstruction, abdominal distention, or emesis. In contrast, infants who do not develop gastrointestinal signs or symptoms during the first 30 days of life do not have Hirschsprung's disease and do not require biopsy to rule out the disease.


**HIRSUTISM**

**Common Causes**
- Familial or racial factors
- Idiopathic hirsutism
- Physiologic hirsutism
- Pregnancy
- Puberty

**Uncommon Causes**
- Central nervous system injury
- Drugs
  - Anabolic steroids
  - Birth-control pills
  - Cyclosporine
  - Diazoxide
  - Dilantin
  - Drugs (Cont.)
    - Minoxidil
    - Progestrone
    - Testosterone
    - Emotional stress (?)
    - Polycystic ovarian disease
    - Severe malnutrition

**Rare Causes**
- Achard-Thiers syndrome
- Acromegaly
- Adrenal disorders
  - Adrenal carcinoma
  - Congenital adrenal hyperplasia
  - Cushing's syndrome
  - Virilizing adrenal adenoma
  - Congenital erythropoietic porphyria
- Dysmorphogenic syndromes (many)
- Hyothyroidism
- Male pseudohermaphroditism
- Ovarian disorders
  - Pure gonadal dysgenesis
  - Virilizing ovarian tumors
    - Arrhenoblastoma
    - Granulosa-theca cell tumors

**HOARSENESS**

**Common Causes**
- Caustic ingestion
- Excessive use of the voice
- Foreign body
- Infectious mononucleosis
- Instrumentation (naso/orogastric tube)
- Laryngitis
- Laryngotracheitis
- Laryngotracheobronchitis
- Postintubation hoarseness
- Postnasal drip
- Vocal cord nodules
- Vocal cord paralysis (postsurgical trauma)
Uncommon Causes

Congenital vocal cord paralysis
Epiglottitis
Hypocalcemia (e.g., hyperparathyroidism)
Hypothyroidism
Laryngeal trauma

Rare Causes

Amyloidosis
Angioneurotic edema
Chromosomal abnormalities
Achondroplasia
Bloom's syndrome
Cockayne's syndrome
Cri du chat syndrome
DeLange's syndrome
Diastrophic dwarfism
Dubowitz's syndrome
Dysautonomia
Williams' syndrome
Congenital abnormalities
Arytenoid cartilage displacement
Clefts
Cysts
Webs
Criccarytenoid arthritis (JRA)
Diphtheria

Recurrent laryngeal nerve dysfunction (Cont.)
Central nervous system disease (Cont.)
Polyneuritis
Pseudobulbar palsy
Ramsay Hunt syndrome
Storage disease
Syphilis
Syringobulbia
Toxin
Trauma
Tumor
Wilson's disease
Motor unit dysfunction
Botulism
Muscular dystrophy
Myasthenia gravis
Toxins
Werdnig-Hoffmann disease
Sarcoidosis
Storage disease (e.g., lysosomal)
Tetany
Tuberculosis
Tumors of the larynx
Adenoma
Carcinoma
Chondroma
Ectopic thyroid
Fibroangioma
Fibroma
Fibrosarcoma
Hamartoma
Hemangioma
Hygroma
Vocal cord hemorrhage
(Wegener's granulomatosis)
HUMAN BITES

A Differential Diagnosis for Lesions That Mimic Human Bites

Human bites are a common occurrence, particularly in conjunction with fighting, abuse (both sexual and physical), patients who have been institutionalized, homicides, self-inflicted injuries, and in association with systemic illnesses (e.g., Lesch-Nyhan syndrome, bulimia). Although most human bites are superficial abrasions, deeper lesions can be quite difficult to treat because of polymicrobial infection and cellulitis. A thorough history and physical examination are important to differentiate human bite lesions from other dermatologic lesions that resemble them. Frequently, however, the history is inaccurate or absent in cases of human bites.

The lesion of a human bite is usually annular or ovoid in shape; teeth marks may be present but these usually become confluent with the passage of time. A key finding in distinguishing human bites from other dermatologic disorders is the presence or absence of scaly lesions; human bites are characteristically nonscaly. Listed below is a table of dermatologic disorders that can mimic human bites; identifying such lesions is of particular importance in suspected cases of child abuse.

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>DESCRIPTION OF LESION</th>
<th>HISTOPATHOLOGY</th>
<th>PREDILECTION SITES</th>
<th>SPECIAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fixed drug eruptions</td>
<td>One or several sharply demarcated erythematous lesions</td>
<td>Epidermis: hydropic degeneration of basal layer; dyskeratotic keratinocytes</td>
<td>Often involve face or genitalia</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dermis: pigment incontinence</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subacute cutaneous lupus erythema (SLE)</td>
<td>Scaly erythematous papules that enlarge, become confluent to form annular and polycyclic lesions</td>
<td>Epidermis: hyperkeratosis, follicular plugging, liquefaction degeneration of basal cells</td>
<td>Shoulders, extensor surfaces of arms, dorsum of hands, upper back, chest</td>
<td>Mild systemic illness; SS-A (Ro) and SS-B (La) antibodies</td>
</tr>
<tr>
<td>Pityriasis rosea</td>
<td>Herald patch: oval or round lesion with central salmon color and darker peripheral zone separated by collarette of scale Symmetric secondary eruption in &quot;Christmas tree&quot; distribution (smaller than herald patch)</td>
<td>Epidermis: hyperkeratosis, hypogranulosis, acanthosis, spongiosis Dermis: mixed superficial perivascular infiltrate with eosinophils</td>
<td>Usually on trunk</td>
<td>Variable prodromal symptoms</td>
</tr>
</tbody>
</table>

Table continued on next page.
Dermatologic Disorders That May Mimic Human Bites (Cont.)

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>DESCRIPTION OF LESION</th>
<th>HISTOPATHOLOGY</th>
<th>PREDILECTION SITES</th>
<th>SPECIAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dermatophytosis: tinea corporis</td>
<td>Most common shows annular lesion with active, erythematous border and central clearing; scales</td>
<td>Fungal organisms in stratum corneum</td>
<td>Glabrous skin</td>
<td>KOH positive</td>
</tr>
<tr>
<td>Granuloma annulare</td>
<td>Skin colored, erythematous, or violaceous papules that assume an annular configuration</td>
<td>Dermis: foci of collagen degeneration (necrobiosis)</td>
<td>Hands and feet: trunk</td>
<td></td>
</tr>
</tbody>
</table>


HUMAN IMMUNODEFICIENCY VIRUS

Indicator Diseases for HIV Infection in Children

Acquired immunodeficiency syndrome (AIDS) is increasing among children as a result of perinatal transmission from infected mothers. By 1987 AIDS was the leading cause of death in the U.S. among children 1-4 years of age. Though many of these children are born to women known to be infected with the human immunodeficiency virus (HIV), mothers are often asymptomatic and undiagnosed at the time their children become ill. Recognizing the diseases with which AIDS in children is likely to present initially may allow for more rapid diagnosis among the children of undiagnosed mothers.

AIDS Indicator Diseases Among 1026 Patients with Perinatally Acquired HIV Infection

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>NO.</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pneumocystis carinii pneumonia</td>
<td>345</td>
<td>34</td>
</tr>
<tr>
<td>Lymphoid interstitial pneumonitis</td>
<td>283</td>
<td>28</td>
</tr>
<tr>
<td>Recurrent bacterial infections</td>
<td>246</td>
<td>24</td>
</tr>
<tr>
<td>HIV wasting syndrome</td>
<td>165</td>
<td>16</td>
</tr>
<tr>
<td>Candida esophagitis</td>
<td>132</td>
<td>13</td>
</tr>
<tr>
<td>HIV encephalopathy</td>
<td>116</td>
<td>11</td>
</tr>
<tr>
<td>Cytomegalovirus disease</td>
<td>77</td>
<td>7</td>
</tr>
<tr>
<td>Pulmonary candidiasis</td>
<td>51</td>
<td>5</td>
</tr>
<tr>
<td>Cryptosporidiosis</td>
<td>31</td>
<td>3</td>
</tr>
<tr>
<td>Herpes simplex disease</td>
<td>30</td>
<td>3</td>
</tr>
<tr>
<td>Mycobacterium avium infection</td>
<td>29</td>
<td>3</td>
</tr>
</tbody>
</table>

*Some children had more than one reported disease.


Recognition and Management of the Infant at Risk

As the incidence of pediatric AIDS continues to rise in the U.S., it becomes incumbent upon the pediatrician to be both facile and competent in its diagnosis and management. Listed below are some helpful hints:

### Table 1. When to Suspect HIV Infection

<table>
<thead>
<tr>
<th>Evidence of maternal HIV infection</th>
<th>Evidence of infant HIV infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Clinical evidence of HIV infection or immunodeficiency</td>
<td>Generalized lymphadenopathy</td>
</tr>
<tr>
<td>Intravenous drug user and/or sexually promiscuous</td>
<td>Hepatosplenomegaly</td>
</tr>
<tr>
<td></td>
<td>Salivary gland enlargement</td>
</tr>
<tr>
<td></td>
<td>Unexplained developmental delay or encephalopathy</td>
</tr>
<tr>
<td></td>
<td>Recurrent or persistent infections</td>
</tr>
</tbody>
</table>

- From a geographic area where HIV infection is prevalent
- Sexual partner of IV drug abuser or HIV-infected man
- Unexplained wasting/failure to thrive
- Chronic pneumonitis
- Immune thrombocytopenic purpura
- Lymphoid malignancy
- Unexplained hepatitis, nephropathy, or cardiomyopathy

### Table 2. Initial Laboratory Evaluation for Neonatal HIV Infection

1. Perform an ELISA test for HIV antibody. If positive, repeat. If repeat ELISA is positive, perform Western blot. A positive Western blot confirms presence of HIV antibody. If possible, obtain a DNA PCR test, p24 antigen test, or HIV culture.
2. If HIV ELISA test is negative and the patient is clinically well: Schedule regular (every other month) visits Consider repeat HIV ELISA test if child's condition changes
3. If HIV ELISA test is negative and patient is ill, obtain DNA PCR, p24 antigen test, or HIV culture

### Table 3. Immunodeficiency Syndromes That May Present in Infancy

| Fetal alcohol syndrome | Disorders of neutrophil function (chronic granulomatous disease) |
| DiGeorge syndrome | Wiskott-Aldrich syndrome |
| Adenosine deaminase deficiency | Severe combined immunodeficiency |
| Nucleoside phosphorylase deficiency | Malignancy |
| Quantitative immunoglobulin disorders | |

### Table 4. Assessment of Immune Function in Infants at Risk for HIV Infection

| CBC with differential | Functional tests of cell-mediated immunity: skin testing for candida, trichophyton, mumps (if previously immunized for mumps) |
| Quantitative assay of IgG, IgA, and IgM | Assay of isohemagglutinins or antibodies to previously administered vaccines |
| Assay of IgG subclasses | Lymphocyte stimulation response to antigens and mitogens |
| CD4/CD8 lymphocyte counts and ratio ("helper/suppressor" ratio) | |

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Table 5. If the Infant Is HIV-positive but without Signs of Immune Deficiency

<table>
<thead>
<tr>
<th>Mother should avoid breast-feeding</th>
<th>Recheck ELISA every 3 mo in healthy child</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physician should examine child every other month</td>
<td>Child should become HIV-negative (by ELISA) by 15 mo of age</td>
</tr>
<tr>
<td>Provide education for the mother/household re: mechanisms of transfer of HIV, caring for baby, HIV transmission routes</td>
<td>Continue follow-up through at least 3 yr of age</td>
</tr>
<tr>
<td></td>
<td>Recheck HIV ELISA later if change in clinical condition is suspicious</td>
</tr>
</tbody>
</table>

Table 6. Immunizing the Infant with HIV Infection

| Table 6. Immunizing the Infant with HIV Infection |
| Do give: |
| DPT (regular schedule) | MMR (at 15 mo) |
| Hib (regular schedule) | Pneumococcal vaccine (at 2 yr) |
| Inactivated polio vaccine (same schedule as for OPV)* | Influenza vaccine (at 6 mo or later) |
| Do not give: |
| Live polio vaccine | BCG vaccine |

* Inactivated polio vaccine should also be given to infants who are not infected but live in a household with an HIV-infected person.

Table 7. When to Start Prophylaxis for PCP*

<table>
<thead>
<tr>
<th>AGE OF PATIENT</th>
<th>CD4+ COUNT (CELLS/μL)**</th>
<th>ACTION RECOMMENDED</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-11 mo</td>
<td>&lt;1,500</td>
<td>Start PCP prophylaxis</td>
</tr>
<tr>
<td></td>
<td>1,500-2,000</td>
<td>No prophylaxis; recheck CD4+ count in 1 mo</td>
</tr>
<tr>
<td></td>
<td>&gt;2,000</td>
<td>No prophylaxis; recheck CD4+ count every 3-4 mo</td>
</tr>
<tr>
<td>12-23 mo</td>
<td>&lt;750</td>
<td>Start PCP prophylaxis</td>
</tr>
<tr>
<td></td>
<td>750-1,000</td>
<td>No prophylaxis; recheck CD4+ count in 1 mo</td>
</tr>
<tr>
<td></td>
<td>&gt;1,000</td>
<td>No prophylaxis; recheck CD4+ count every 3-4 mo</td>
</tr>
<tr>
<td>24-72 mo</td>
<td>&lt;500</td>
<td>Start PCP Prophylaxis</td>
</tr>
<tr>
<td></td>
<td>500-750</td>
<td>No prophylaxis; recheck CD4+ count in 1 mo</td>
</tr>
<tr>
<td></td>
<td>750-1,500</td>
<td>No prophylaxis; recheck CD4+ count every 3-4 mo</td>
</tr>
<tr>
<td></td>
<td>&gt;1,500</td>
<td>No prophylaxis; recheck CD4+ count every 6 mo</td>
</tr>
<tr>
<td>&gt;72 mo</td>
<td>&lt;200</td>
<td>Start PCP prophylaxis</td>
</tr>
<tr>
<td></td>
<td>200-300</td>
<td>No prophylaxis; recheck CD4+ count in 1 mo</td>
</tr>
<tr>
<td></td>
<td>&gt;300-600</td>
<td>No prophylaxis; recheck CD4+ count every 3-4 mo</td>
</tr>
<tr>
<td></td>
<td>&gt;600</td>
<td>No prophylaxis; recheck CD4+ count every 6 mo</td>
</tr>
</tbody>
</table>

* Applies to children who are HIV-infected, HIV-seropositive, or less than 12 mo of age and born to an HIV-infected mother.
** Regardless of CD4+ count, start PCP prophylaxis if the CD4+ % is less than 20%. Prophylaxis is also indicated, regardless of age and CD4+ count, for any child who had an episode of PCP.

Adapted from Centers for Disease Control.

Nonimmune Hydrops Fetalis

The original definition of nonimmune hydrops fetalis is attributed to E.L. Potter who described the condition in 1943 as "universal edema of the fetus unassociated with erythroblastosis." Currently the term hydrops is used to describe the accumulation of fluid in specific interstitial tissues or body cavities, whereas hydrops fetalis refers to the generalized, pathologic accumulation of fluid in serous cavities in the fetus with edema of the soft tissue.

Ultrasound is the most common means of diagnosing this condition prenatally. The findings of such an examination typically reveal fetal skin edema, effusions in the body cavities (e.g., ascites), hydramnios, and placental edema. Upon delivery, the newborn infant displays gross edema and may be extremely difficult to resuscitate due to ascites, pleural effusions, and an associated lung hypoplasia.

Etiology

In general, hydrops fetalis is separated into two categories: immune and nonimmune. Immune hydrops is most commonly secondary to Rh isoimmunization. Since the advent of anti-D globulin in the early 1960s, the majority of cases of fetal hydrops are nonimmune in nature. The incidence of nonimmune hydrops fetalis is between 1/2500 to 1/3500 newborns.

The pathologic mechanisms leading to this condition can be categorized as follows:

1. Increased intracapillary hydrostatic pressure
2. Decreased intracapillary osmotic pressure
3. Damage to the peripheral capillary/vascular integrity.

Various combinations of these mechanisms in different conditions are seen (see accompanying table, "Causes of Nonimmune Hydrops").

The Investigation of Hydrops Fetalis

When hydrops fetalis is discovered, either by ultrasound during a pregnancy or at birth, the following approach is recommended in order to extract the maximum amount of information:

1. Maternal work-up
   - Complete blood count and indices
   - Hemoglobin electrophoresis
   - Kleihauer-Betke stain of peripheral blood
   - VDRL and TORCH titers
   - Anti-Ro, systemic lupus erythematosus preparation, and sedimentation rate
   - Oral glucose tolerance test

2. Fetal assessment
   - Continued ultrasound-cardiac work-up
   - Limb-length, fetal movement

3. Amniocentesis
   - Karotype (α-fetoprotein)
   - Virus cultures
   - Establish culture for appropriate metabolic or DNA testing

4. Fetal blood sampling
   - Kerotype
   - Hemoglobin analysis
   - IgM; specific cultures
   - Albumin and total protein
5. At delivery
   Karotyping (as appropriate)  Detailed autopsy
   Photography                (as appropriate)
   X-ray films                Urinalysis
   Fluid from effusion, ascites Complete blood count
   (chyle, protein, culture)    Liver function studies
   Placental examination       Viral titers

**Causes of Nonimmune Hydrops**

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>CONDITIONS</th>
<th>APPROXIMATE % OF CASES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hematologic</td>
<td>Homozygous α-thalassemia; chronic fetomaternal transfusion; twin-to-twin transfusion; acardius; atrioventricular shunts; hemorrhage or thrombosis; maternal drugs (chloramphenicol)</td>
<td>10</td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Severe congenital heart disease (e.g., complex congenital heart defects, atrioventricular septal defects, premature closure of the foramen ovale, hypoplastic left and right heart); arrhythmias or congenital heart block; myocardial and endocardial disease; cardiac tumors (e.g., rhabdomyomas)</td>
<td>20</td>
</tr>
<tr>
<td>Respiratory</td>
<td>Cystic adenomatoid malformation of lung; diaphragmatic hernia; pulmonary lymphangiectasia; pulmonary sequestration; intrathoracic mass</td>
<td>5</td>
</tr>
<tr>
<td>Gastrointestinal</td>
<td>Bowel atresias; volvulus; duplications of the gut; peritonitis</td>
<td>5</td>
</tr>
<tr>
<td>Urinary/renal</td>
<td>Urethral and ureteral atresia</td>
<td>5</td>
</tr>
<tr>
<td>disorders</td>
<td>Bladder neck obstruction</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Posterior urethral valves</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Cloacal malformation</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Congenital nephrosis</td>
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</tr>
<tr>
<td>Chromosomal</td>
<td>Turner syndrome; trisomies 13, 18, 21; triploidy; miscellaneous aneuploidy</td>
<td>16</td>
</tr>
<tr>
<td>Placenta</td>
<td>Umbilical vein thrombosis; torsion of cord; chorioangioma</td>
<td></td>
</tr>
<tr>
<td>Intrauterine</td>
<td>Cytomegalovirus; toxoplasmosis; syphilis; parvovirus; parasitic diseases</td>
<td>8</td>
</tr>
<tr>
<td>infection with or</td>
<td></td>
<td></td>
</tr>
<tr>
<td>without hemolysis</td>
<td></td>
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<tr>
<td>Recognized</td>
<td>Dwarfing syndromes (e.g., thanatophoric, Jeune, hypophosphatasia, achondrogenesis); arthrogryposis; Neu-Laxova syndrome; Pena-Shokeir syndrome; Noonan syndrome; multiple pterygium syndromes; Meckel syndrome</td>
<td>11</td>
</tr>
<tr>
<td>Metabolic disorders</td>
<td>Lysosomal storage disorders (including mucopolysaccharidoses); Gaucher disease; gangliosidosis; sialidosis</td>
<td>5·10</td>
</tr>
<tr>
<td>Miscellaneous</td>
<td>Amniotic band syndrome; fetal tumors (e.g., teratoma, neuroblastomas, Wilms, angiomas)</td>
<td></td>
</tr>
</tbody>
</table>


164—Hyperhidrosis/Hyperleukocytosis

HYPERHIDROSIS

Common Causes

Emotional stimuli
Exercise
Fever, recovery from fever

Increased environmental temperature
Ingestion of spicy foods

Uncommon Causes

Atopic predisposition
Chronic illness
Brucellosis
Pulmonary tuberculosis
Cluster headaches

Congestive heart failure
Drug withdrawal
Hypoglycemia
Respiratory failure
Salicylate intoxication

Rare Causes

Acrodynia
Acromegaly
Auriculotemporal syndrome
Carbon monoxide poisoning
Carcinoid syndrome
Citrullinemia
Diencephalic syndrome
Familial dysautonomia
Familial periodic paralysis
Hyperthyroidism
Insulin overdose

Ipecac ingestion
Myocardial infarction
Organophosphate poisoning
Phenylketonuria
Pheochromocytoma
Pyridoxine deficiency
Spinal cord injury
Thrombocytopenia-absent radius syndrome (TAR)
Vasoactive intestinal peptide-secreting tumor

HYPERLEUKOCYTOSIS

Blood Gas Determinations with Extreme Leukocytosis

Patients with hyperleukocytosis—white cell counts in excess of 200,000 per mm³, as seen with leukemia—may have respiratory distress and/or hypoxia secondary to leukocyte-thrombocyte aggregation in the lungs, to hyperviscosity, or to pneumonia. Because of these complications, arterial blood gas determinations are often essential to management of these patients. Because leukocytes consume oxygen, accurate determinations of arterial oxygen tension can only be made if blood samples are immersed immediately in crushed ice and injected into the gas analyzer within 1 minute. Blood gas measurements in patients with hyperleukocytosis should be considered unsuitable if there is a delay of more than 1 minute.

HYPERLIPIDEMIA

Causes of Secondary Hyperlipidemia in Childhood

Drug use—steroids, thiazides, \( \beta \)-blockers, isotretinoin (Accutane), anticonvulsants, oral contraceptives, alcohol

Obesity
Diabetes mellitus
Hypothyroidism
Lipodystrophy
Pregnancy

Storage disease—Tay-Sachs, glycogen storage diseases, etc.
Renal failure
Nephrotic syndrome
Systemic lupus erythematosus and collagen diseases
Cholestasis
Anorexia nervosa
Idiopathic hypercalcemia

HYPERSENSITIVITY

Clinical Examples of Hypersensitivity Reactions

The immune system generally plays a protective role in maintaining a host’s response to potentially dangerous immunologic and infectious stimuli, yet there are circumstances where the immune system’s machinations, once set in motion, can produce tissue injury. These hypersensitivity reactions are divided into four types: (1) the immediate or anaphylactic type, (2) the cytotoxic type, (3) the immune complex or Arthus type, and (4) the delayed hypersensitivity reaction. Listed below are clinical examples of each reaction.

1. **Type 1 hypersensitivity (allergic reactions mediated by IgE)**
   a. Urticaria  
   b. Hay fever  
   c. Allergic rhinitis  
   d. Allergic conjunctivitis  
   e. Allergic asthma  
   f. Systemic anaphylaxis (e.g., reactions to antibiotics, vaccines, foreign sera, hormones, medications, contrast agents, hymenoptera stings, snake venom, blood products, and foods)

2. **Type 2 hypersensitivity (antibody = dependent cytotoxicity)**
   a. Erythrobastosis fetalis  
   b. Acquired hemolytic anemia  
   c. Thrombocytopenia  
   d. Pemphigus  
   e. Goodpasture’s syndrome (glomerulonephritis associated with hemoptysis)  
   f. Graft rejection  
   g. Neutropenia  
   h. Chronic keratitis

3. **Type 3 hypersensitivity (immune complex or Arthus type)**
   a. Microbial infection  
   i. Bacterial (e.g., streptococcal, glomerulonephritis, and lepromatous leprosy)  
   ii. Viral (e.g., cytomegaloviral choriomeningitis)  
   iii. Parasitic (e.g., toxoplasma retinochoroiditis)  
   b. Malignancy (e.g., solid tumor metastases, lymphoma, and leukemia)  
   c. Autoimmune disorders (e.g., systemic lupus erythematosus, sympathetic ophthalmia, rheumatoid arthritis, Sjogren’s syndrome, lens-induced uveitis)
Hypertension

d. Vasculitis  
   i. Secondary to immune complexes involving any infection or tissue antigens  
   ii. Erythema multiforme (Stevens Johnson)  
   iii. Serum sickness

4. Type 4 hypersensitivity (delayed cell-mediated immunity)  
   a. Mantoux tuberculint test (which only becomes positive after 48 hours)  
   b. Reactions to poison ivy, poison sumac, and poison oak contact  
   c. Hashimoto’s thyroiditis  
   d. Transplantation reaction and graft rejection


HYPERTENSION

Common Causes

Agitation  
Anxiety  
Coarctation of the aorta  
Essential hypertension  
Immobilization  
Obesity  
Pain  
Renal causes  
   Acute tubular necrosis  
   Congenital anomalies  
      Hydronephrosis  
      Nephrophthisis  
      Polycystic kidneys  
      Renal aplasia/hypoplasia/dysplasia  
      Segmental hypoplasia  
   Glomerulonephritis (acute and chronic)  
      Membranoproliferative, etc.  
      Postinfectious  
      Liddle’s syndrome  
   Miscellaneous nephropathy  
      Amyloidosis  
      Diabetes mellitus  
      Gout  
   Nephrolithiasis  
   Nephrotic syndrome  
      Idiopathic  
      Minimal change disease  

Renal causes (Cont.)  
   Obstructive uropathy  
   Other nephritides  
      Familial nephritis  
      Hemolytic-uremic syndrome  
      Henoch-Schönlein purpura  
      Hypersensitivity/transfusion reaction  
      Periarteritis nodosa  
      Radiation  
      Systemic lupus erythematosus  
   Pyelonephritis  
   Renal failure (acute and chronic)  
   Renal transplantation  
   Renal vascular disease  
      Renal artery  
      Aneurysm  
      Arteritis  
      Embolic disease  
      External compression  
      Fibromuscular dysplasia  
      Fistula  
      Stenosis  
      Thrombosis  
      Trauma  
   Renal vein thrombosis  
   Retroperitoneal fibrosis  
   Trauma
Renal causes (Cont.)

Tumors
Extrinsic tumors
Adrenal carcinoma
Neuroblastoma
Small pressure-cuff size

Renin-secreting tumors
(J-G cell)
Wilms' tumor

Uncommon Causes

Cardiovascular etiologies
Anemia
Aortic aneurysm/thrombosis
Arteriovenous fistula
Aortic insufficiency
Aortcopulmonary window
Patent ductus arteriosus
Bacterial endocarditis
Iatrogenic hypervolemia
Polycythemia
Pseudoxanthoma elasticum
Radiation aortitis
Takayasu's arteritis

Drugs and chemicals
Glucocorticoids
Glycyrrhizic acid (licorice)
Heavy metals (lead, cadmium, mercury)
Methysergide
Mineralocorticoids
Monoamine-oxidase inhibitors
Oral contraceptives
Phencyclidine
Sodium salts
Sympathomimetics (decongestants)
Tricyclic antidepressants

Rare Causes

Burns
Central nervous system
Dysautonomia (Riley-Day syndrome)
Encephalitis
Guillain-Barré syndrome
Increased intracranial pressure
Poliomyelitis
Neurofibromatosis
Collagen vascular
Dermatomyositis
Scleroderma
Cystinosis
Endocrine
Congenital adrenal hyperplasia
11-β-hydroxylase deficiency
17-hydroxylase deficiency
Cushing's syndrome

Endocrine (Cont.)
Hyperaldosteronism
Primary
Conn's syndrome
Dexamethasone-suppressible
Idiopathic nodular hyperplasia
Secondary
Hyperthyroidism
Pheochromocytoma
Fabry's disease
Hypoxia
Malignant hyperthermia
Metabolic
Hypercalcemia
Hypernatremia
RTA with nephrocalcinosis
Sickle-cell anemia
Stevens-Johnson syndrome

Malignant Hypertension in Children

The crisis of hypertension, or a hypertensive emergency, is heralded by a blood pressure high enough to cause damage to such target organs as the brain (hypertensive encephalopathy), eye (retinopathy, infarction of anterior visual pathways), kidneys (renal failure), and the heart (left ventricular hypertrophy and
subsequent failure). Longstanding hypertension can also yield these effects but over a much longer time period. Severe hypertension in the pediatric age range has an incidence of 1 out of 1000 and is usually secondary to renal disease (e.g., renal scarring from chronic pyelonephritis or obstructive uropathy, glomerulonephritis, and renovascular disease). Nonrenal etiologies include coarctation of the aorta and catecholamine-excess states. In tertiary medical centers, some of the most frequent and severe cases of hypertension are a result of complications from end-stage renal disease and postrenal transplantation.

Given the child or infant with an exceedingly high blood pressure, whether asymptomatic or not, the following data collection may be useful in elucidating the etiology and duration of the hypertension. Intravenous hypertensive therapy, in the case of a hypertensive crisis, should be initiated quickly to prevent end-organ damage. If no hypertensive crisis exists and hypertension has been documented on at least three different occasions, maintenance therapy with oral hypertensive agents should be initiated.

1. **Key Historical Data**

<table>
<thead>
<tr>
<th>History</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. Neonatal umbilical artery catheterization</td>
<td>Renal artery stenosis</td>
</tr>
<tr>
<td>b. History of unexplained fever, urinary tract infection, failure to thrive</td>
<td>Reflux nephropathy</td>
</tr>
<tr>
<td>c. Nocturia, peripheral edema, hematuria, failure to thrive</td>
<td>Renal disease</td>
</tr>
<tr>
<td>d. Joint pain and swelling, rash</td>
<td>Connective tissue disease</td>
</tr>
<tr>
<td>e. Palpitations, flushing, sweating, fever, weight loss</td>
<td>Pheochromocytoma</td>
</tr>
<tr>
<td>f. Weakness, muscle cramps</td>
<td>Hyperaldosteronism</td>
</tr>
<tr>
<td>g. Ingestion of drug</td>
<td>Drug-induced hypertension</td>
</tr>
<tr>
<td>h. Family history of hypertension, renal disease</td>
<td>Inherited renal disease</td>
</tr>
</tbody>
</table>

2. **Physical Findings**

   |                                                                                   | Renal disease                    |
   | a. Short stature, peripheral edema, pallor                                      | Neurofibromatosis                 |
   | b. Cafe-au-lait spots                                                            | Pheochromocytoma                 |
   | c. Tachycardia, increased sweating at rest, flushing                           | Cushing syndrome; steroid abuse   |
   | d. Moon facies, truncal obesity, striae                                         | Coarctation of the aorta         |
   | e. Absent or delayed femoral pulses, leg pressure significantly lower than arm pressure | Renovascular disease             |
   | f. Abdominal bruit                                                              | Congestive heart failure secondary to severe or longstanding hypertension |
   | g. Tachycardia, tachypnea, hepatomegaly, rales                                  |                                  |
2. Physical Findings (Cont.)

h. Hypertensive fundoscopic changes
i. Bell's palsy
j. Neurologic deficit (e.g., absent pupillary reflex, hemiparesis)

C. Chronic severe hypertension
D. Side-effect of chronic or severe acute hypertension

3. Initial Laboratory Examination

<table>
<thead>
<tr>
<th>Test</th>
<th>Possible Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. Complete blood count</td>
<td>Low hemoglobin—chronic renal disease</td>
</tr>
<tr>
<td>b. Urinalysis</td>
<td>Low platelets and white cell count—connective tissue disease</td>
</tr>
<tr>
<td>c. Urea, creatinine</td>
<td>Renal disease</td>
</tr>
<tr>
<td>d. Sodium, potassium, blood gases</td>
<td>Renal disease; mineralocorticoid excess</td>
</tr>
<tr>
<td>e. Renin</td>
<td>Renovascular disease</td>
</tr>
<tr>
<td>f. Chest x-ray</td>
<td>Evidence of cardiac failure</td>
</tr>
<tr>
<td>g. ECG</td>
<td>Hypertensive cardiomyopathy</td>
</tr>
</tbody>
</table>


HYPOGONADISM

Hypogonadism and Obesity

The association between obesity and gonadal insufficiency or dysfunction has long been noted, yet the exact relationship of these two problems remains unclear. Endocrinologists have subdivided the myriad of syndromes and disease entities into four major categories:

1. Abnormalities of the peripheral metabolism of sex hormones
   a. Obese adult men have been noted to have low serum testosterone levels and poorly developed secondary sexual characteristics.
   b. Eunuchoid, hypogonadic males are frequently obese and often lose weight with the exogenous administration of testosterone.
   c. Obesity in adult women is frequently associated with dysfunctional uterine bleeding, amenorrhea, and increased conversion of circulating androgens to estrogens.

2. Acquired hypothalamic conditions or Frohlich's syndrome (specifically lesions to the ventromedial nucleus of the hypothalamus)
   a. Craniopharyngioma
   b. Trauma
Hypotonia

3. Extragonadal endocrine disorders
   a. Hypothyroidism
   b. Cushing's syndrome
   c. Pseudohypoparathyroidism

4. Genetic syndromes of hypogonadism and obesity
   a. Hypogonadotropic hypogonadism
      i. Kallmann's syndrome (anosmia or hyposmia, midline defects including cleft lip and palate, color blindness, neurosensory defects, renal and bone abnormalities)
      ii. Prader-Willi syndrome (hypotonia in the newborn period, short stature, mental retardation, diabetes mellitus and insulin resistance, distinctive facial features such as almond-shaped eyes)
      iii. Laurence-Moon-Bardet-Biedl syndrome (retinitis pigmentosa, polydactyly, syndactyly, brachydactyly, mental retardation, spastic paraplegia, genitourinary tract anomalies)
      iv. Biemond syndrome (iris coloboma, mental retardation, polydactyly)
      v. Börjeson-Forsman-Lehman syndrome (severe mental retardation, short stature, coarse facies, microcephaly, seizures, nystagmus, ptosis)
      vi. Carpenter syndrome (acrocephalosyndactyly, mental retardation)
   b. Hypergonadotropic hypogonadism
      i. Klinefelter syndrome (XXY chromosome, small testes, gynecomastia)
      ii. Alström syndrome (retinal degeneration, nerve deafness, diabetes mellitus, acanthosis nigricans, nephropathy)


HYPOTONIA—NEONATAL

Common Causes

Asphyxia
Benign, congenital

Sepsis
Trauma

Uncommon Causes

Congenital joint laxity
Down syndrome
"Hypermobility syndrome"
Hypothyroidism

Neonatal myasthenia
Spinal cord injury
Werdnig-Hoffmann disease

Rare Causes

Achondroplasia
Cerebro-hepato-renal syndrome
Congenital lactic acidosis

Congenital myopathies
Central core disease
Myotubular myopathy
The Differential Diagnosis of Hypotonia

Hypotonia, or decreased muscle tone, can be a presenting sign in a number of pathologic conditions affecting the central or peripheral nervous system. It is important to distinguish hypotonia from muscle weakness, which is a diminution of muscle power or strength but normal tone. Typically, when decreased muscle strength and tone appear together, diseases of the peripheral nervous system should be suspected; when hypotonia occurs without alterations in muscle strength, a disorder of the central nervous system should be investigated.

Anatomic localization of the process producing hypotonia is useful in generating a differential diagnosis. This can be done easily at the bedside.

1. **Diseases of the central nervous system or upper motor neuron disease** (processes involving the brain and spinal cord but not the anterior horn cells):
   a. Muscle strength is usually preserved and often normal.
   b. Deep tendon reflexes of the affected limbs are always preserved. (They may be exaggerated if the disease in question affects the pyramidal tracts.)
   c. Infantile reflexes either persist or, if lost, return (e.g., the Babinski reflex, palmar and plantar grasp, sucking, rooting, and snouting reflexes).
   d. Sensation is generally preserved and normal.
   e. The sign of hypotonia may be associated with dysequilibrium, such as gait imbalance, wide stance, ataxia, or dysmetria with or without a tremor.

2. **Diseases affecting the peripheral nervous system or lower motor neuron disease** (processes involving the anterior horn cell, myoneural junction, and innervation of muscle):
   a. Patients exhibit prominent weakness in concert with hypotonia.
   b. Deep tendon reflexes may be normal or decreased, but they are never hyperreflexive; in diseases involving the anterior horn cells and peripheral motor neurons, deep tendon reflexes are frequently absent.
   c. Primitive or infantile reflexes are absent (with the exception of newborns).
   d. Sensation abnormalities are not present (with the exception of specific disorders that affect both the peripheral motor and sensory nerves).
   e. Equilibrium is compromised secondary to impaired muscle strength; the dysequilibrium, therefore, should be in proportion to the associated muscle weakness.

The following two tables list acute and subacute causes and chronic causes of hypotonia.
### Acute and Subacute Diseases Producing Hypotonia

<table>
<thead>
<tr>
<th>BRAIN AND SPINAL CORD</th>
<th>ANTERIOR HORN CELL</th>
<th>PERIPHERAL NERVE</th>
<th>MYONEURAL JUNCTION</th>
<th>MUSCLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Metabolic</td>
<td></td>
<td>Poliomyelitis</td>
<td>Botulism</td>
<td>Infectious Myositis</td>
</tr>
<tr>
<td>Encephalopathy</td>
<td></td>
<td>Guilian-Barré</td>
<td>Myasthenic Syndromes</td>
<td>Viral</td>
</tr>
<tr>
<td>Hypoxia/ischemia</td>
<td></td>
<td>Syndrome</td>
<td>Antibiotics</td>
<td>Parasitic</td>
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<tr>
<td>Hypoglycemia</td>
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<td>Trauma</td>
<td>Metabolic poisons</td>
<td>(trichinosis)</td>
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<tr>
<td>Bilirubin</td>
<td></td>
<td>Peripheral</td>
<td>(organophosphates)</td>
<td>Endocrine</td>
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<tr>
<td>Ammonia</td>
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<td>Neuropathy</td>
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<td>Hypothyroidism</td>
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<td>Acidosis</td>
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<td>Vitamin deficiency</td>
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<td>Addison's disease</td>
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<tr>
<td>Toxic</td>
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<td>(B1, B6, B12, folate)</td>
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<td>Collagen vascular</td>
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<tr>
<td>Encephalopathy</td>
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<td>Drug induced</td>
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<td>disease</td>
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<td>Drugs</td>
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<td>Heavy metal (Pb)</td>
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<td>Poisons</td>
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<td>Diabetes</td>
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<tr>
<td>Animal bites</td>
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<td>Uremia</td>
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<td>(reptile, insect)</td>
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<td>Porphyrin</td>
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<td>Vaccinal</td>
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<td>Diphtheria</td>
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<td>Vaccinal</td>
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<td>Concussion</td>
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<td>Collagen vascular</td>
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<td>Hemorrhage</td>
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<td>Encephalitis</td>
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<td>Meningitis</td>
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<td>Myelitis (transverse)</td>
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<td>Para-Infectious Acute</td>
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<tr>
<td>Cerebellar Ataxia</td>
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<tr>
<td>Hydrocephalus</td>
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<tr>
<td>Neoplasia</td>
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<tr>
<td>Posterior fossa tumors</td>
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<tr>
<td>Collagen Vascular</td>
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<tr>
<td>Disease</td>
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</table>


### Chronic Causes of Hypotonia

<table>
<thead>
<tr>
<th>BRAIN AND SPINAL CORD</th>
<th>ANTERIOR HORN CELL</th>
<th>PERIPHERAL NERVE</th>
<th>MYONEURAL JUNCTION</th>
<th>MUSCLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
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<tr>
<td>Anencephaly</td>
<td></td>
<td>(e.g., Charcot-Marie Tooth, Dejerine-Sottas syndromes)</td>
<td></td>
<td>1. The muscular dystrophy syndromes</td>
</tr>
<tr>
<td>Encephalocele</td>
<td></td>
<td></td>
<td>3. Inherited recurrent focal neuropathies</td>
<td>2. Congenital myopathies (e.g., central core disease, minicore disease, nemaline myopathy, severe x-linked myotubular myopathy)</td>
</tr>
<tr>
<td>Myelomeningocele</td>
<td></td>
<td></td>
<td>4. Familial dysautonomia</td>
<td>3. Mitochondrial disorders</td>
</tr>
<tr>
<td>b. Disorders of diverticulation</td>
<td></td>
<td></td>
<td></td>
<td>4. Periodic paralyses</td>
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<tr>
<td>Holoprosencephaly</td>
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<td>5. Inflammatory myopathies</td>
</tr>
<tr>
<td>Arnold-Chiari malformation</td>
<td></td>
<td></td>
<td></td>
<td>6. Myotonic syndromes</td>
</tr>
<tr>
<td>Dandy-Walker syndrome</td>
<td></td>
<td></td>
<td></td>
<td>7. Endocrine myopathies</td>
</tr>
<tr>
<td>c. Disorders of commissuration</td>
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<td></td>
<td>8. Metabolic myopathies (e.g., Glycogen storage disease of heart and muscle or Pompe's disease)</td>
</tr>
<tr>
<td>Agenesis of the corpus callosum</td>
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</tr>
<tr>
<td>Agenesis or hypoplasia of the septum pellucidum</td>
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</tr>
</tbody>
</table>

Table continued on next page.
Hypotonia—173

Chronic Causes of Hypotonia (Cont.)

<table>
<thead>
<tr>
<th>BRAIN AND SPINAL CORD</th>
<th>ANTERIOR HORN CELL</th>
<th>PERIPHERAL NERVE</th>
<th>MYONEURAL JUNCTION</th>
<th>MUSCLE</th>
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<tbody>
<tr>
<td>d. Disorders of histogenesis</td>
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<tr>
<td>Microcephaly vera</td>
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<tr>
<td>Cerebral gigantism</td>
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</tr>
<tr>
<td>Cerebellar aplasia</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neuronal heterotopias</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lissencephaly</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2. Inborn Errors of Metabolism</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(e.g., carbohydrate, amino acid, fatty acid)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3. Storage Disorders</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>a. Glycogen</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>b. Gangliosidosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>c. Mucopolysaccharidosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>d. Mucolipidosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>e. Peroxisomal</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4. Toxic encephalopathies</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>5. Infectious encephalitis</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>(e.g., TORCH)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>6. Hypothyroidism (congenital)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>7. Hydrocephalus</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

I but use you a minute, then I resign you, stallion,
Why do I need your paces when I myself out-gallop them?
Even as I stand or sit passing faster than you.

Walt Whitman
From *Song of Myself*
IDIOPATHIC THROMBOCYTOPENIC PURPURA

Who Needs a Bone Marrow Examination?

Is a bone marrow examination necessary when you are faced with a child with the clinical findings of acute idiopathic thrombocytopenic purpura whose examination results are otherwise normal and whose blood cell count and blood smear reveal only thrombocytopenia?

Naturally the clinician, and the parent, worry that the thrombocytopenia may be a manifestation of leukemia. A study in which the records of 2239 patients with acute lymphoblastic leukemia were reviewed showed that none of these children had significant thrombocytopenia with no other hematologic or physical manifestations of the leukemia.

A bone marrow examination is unnecessary in your patient if:

1. No blasts are present in the peripheral blood film;
2. The platelet count is less than 50,000/mm³;
3. The hemoglobin concentration is more than 11.00 g/dl;
4. The absolute neutrophil count is more than 1500/mm³; and
5. There is no organomegaly.


IMMUNODEFICIENCY

The Humoral Immunodeficiency Syndromes

Humoral immunodeficiency syndromes are characterized by an impairment in the host's capacity to manufacture antibodies. Although the particular syndrome may be congenital or acquired, these patients exhibit little or no immunoglobulin upon serum testing. The typical child with humoral immunodeficiency presents with frequent, recurrent, and persistent bacterial infections in association with low immunoglobulin levels or a particular impairment in the production of a specific antibody.

Children with humoral immunodeficiency are particularly susceptible to infections by encapsulated organisms (e.g., *Streptococcus pneumoniae*, group A streptococci, and *Hemophilus influenzae*) because of an inability to produce opsonizing antibodies. Other pathogens that frequently infect these patients include mycoplasma, *Giardia lamblia*, *Clostridium difficile*, and *Staphylococcus aureus*. The types of frequently occurring infections include pneumonia, upper respiratory tract infections, otitis media, sinusitis, conjunctivitis, diarrhea, and furunculosis. Children
with humoral immunodeficiency, on the other hand, have an intact ability to form T cells, natural killer cells, phagocytic cells, and complement, so that frequent or recurrent viral, fungal, and parasitic infections are not a prominent feature.

The evaluation of a child who presents with frequent infections of the type described above begins with a thorough history, specifically addressing which infections the child has had, the etiologic agents identified, and their natural history in that host. A careful physical examination with attention to particular findings consistent with the humoral immunodeficiency syndromes is also warranted. The laboratory examination should begin with determination of quantitative serum IgG, IgA, and IgM; a functional antibody response to immunization with diphtheria and tetanus; and complete blood count with attention to white cell morphology on the peripheral smear. Further work-up might include quantitative serum isohemagglutinins; a bone marrow biopsy to detect plasma cells; a lymph node biopsy for histologic analysis of primary follicles, germinal centers, and plasma cells; B cell phenotypic markers; IgG subclasses; and salivary IgA. The essential features of the primarily humoral immunodeficiency syndromes are summarized in tabular form, below:

### Humoral Immunodeficiency Syndromes

<table>
<thead>
<tr>
<th>SYNDROME</th>
<th>IMMUNE DEFECT</th>
<th>CHARACTERISTIC FEATURES</th>
<th>CLINICAL ASSOCIATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>X-linked agammaglobulinemia</td>
<td>Block at level of pre-B → B cell</td>
<td>Very low Ig levels</td>
<td>Echovirus infection</td>
</tr>
<tr>
<td></td>
<td></td>
<td>No B cells in PB</td>
<td>Dermatomyositis</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Lymphoreticular malignancy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>IVGG indicated</td>
</tr>
<tr>
<td>Autosomal recessive agammaglobulinemia</td>
<td>Very low B cells in PB</td>
<td>Females also affected</td>
<td>Similar to X-linked (Bruton's)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>IVGG indicated</td>
</tr>
<tr>
<td>Transient hypogammaglobulinemia</td>
<td>Unknown</td>
<td>B cell population normal</td>
<td>IgG and infections</td>
</tr>
<tr>
<td>of infancy</td>
<td></td>
<td></td>
<td>↓ by age 2</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>IVGG not indicated</td>
</tr>
<tr>
<td>Common variable immunodeficiency</td>
<td>Intrinsic B cell defects</td>
<td>B cells present in PB</td>
<td>Autoimmune disease</td>
</tr>
<tr>
<td></td>
<td>T suppressor activity</td>
<td></td>
<td>lymphadenopathy</td>
</tr>
<tr>
<td></td>
<td>T or B auto-antibodies</td>
<td></td>
<td>splenomegaly</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Sprue-like syndrome</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Lymphoreticular malignancy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>IVGG not indicated</td>
</tr>
<tr>
<td>IgA deficiency</td>
<td>Intrinsic B cell defect</td>
<td>Common</td>
<td>Autoimmune disease</td>
</tr>
<tr>
<td></td>
<td>T suppressor activity</td>
<td>May be drug-induced</td>
<td>Risk of anti-IgA anaphylactoid reaction</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Anti-IgA antibodies common</td>
<td>IVGG not indicated</td>
</tr>
<tr>
<td>Hyper-IgM syndrome</td>
<td>Defective isotype switch</td>
<td>↓↓ IgM with low IgG, IgA</td>
<td>Autoimmune disease</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Lymphadenopathy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>IVGG indicated</td>
</tr>
<tr>
<td>Selective or qualitative</td>
<td>Lack of T cell “help”</td>
<td>IgG2, IgG4 most common</td>
<td>IgA deficiency</td>
</tr>
<tr>
<td>deficiency</td>
<td></td>
<td>Ig levels normal or high</td>
<td>IVGG used for broadly</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>response to antigen challenge</td>
</tr>
</tbody>
</table>

Nonimmunologic Defense Mechanisms

The human body is blessed with a number of nonimmunologic defense mechanisms that serve as the “first line of defense” in preventing microbial invasion. These include the skin, mucous membranes, and their secretory components. It should also be noted that normal vascular perfusion of tissues, adequate flow of urine, bile and respiratory secretions, and the presence of normal commensal bacterial flora are necessary in the daily prevention of microbial invasion. Indeed, many pediatric patients who suffer from chronic recurrent infections have defects in these anatomic and physical barriers rather than true immunodeficiencies. The workup of such a patient, therefore, should always establish the integrity of such anatomic, nonimmunologic barriers to infection.

Defects in Nonimmunologic Defense Mechanisms Contributing to Recurrent Infections

<table>
<thead>
<tr>
<th>Abnormal barriers</th>
<th>Microbiologic flora</th>
<th>Foreign bodies</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eczema</td>
<td>Alteration by antibiotic therapy</td>
<td>Pulmonary</td>
</tr>
<tr>
<td>Burns</td>
<td>Abnormal drainage</td>
<td>Heart valves</td>
</tr>
<tr>
<td>Skull fractures</td>
<td>Ureteral stenosis</td>
<td>Vascular catheters</td>
</tr>
<tr>
<td>Sinus tracts</td>
<td>Vesicoureteral reflux</td>
<td>Urinary catheters</td>
</tr>
<tr>
<td>Abnormal vascular perfusion</td>
<td>Dysfunction of eustachian tubes</td>
<td></td>
</tr>
<tr>
<td>Angiopathy (e.g., diabetes)</td>
<td>Cystic fibrosis</td>
<td></td>
</tr>
<tr>
<td>Edema (e.g., nephrotic syndrome,</td>
<td>Ciliary dysfunction</td>
<td></td>
</tr>
<tr>
<td>congestive heart failure)</td>
<td>Tracheoesophageal fistula</td>
<td></td>
</tr>
<tr>
<td>Infarction (e.g., sickle cell disease)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>


Inborn Errors of Metabolism Presenting in the Neonatal Period

Inborn errors of metabolism are not as rare as we might believe. Although incidence rates are hard to come by due to undiagnosed cases, the possibility that a healthy full-term neonate who becomes suddenly ill has a treatable metabolic error is nearly as likely as that infant having an acquired infection. The tragedy of inborn errors of metabolism in the neonatal period is that a missed diagnosis can lead to rapidly progressive neurologic deterioration, coma, and death.

The characteristic symptoms of inborn errors are, like those of sepsis, largely nonspecific and variable: lethargy, failure to thrive, vomiting, seizures, and respiratory distress in the immediate neonatal period. Taken as a group, conservative estimates suggest that 20% of disease among full-term neonates without risk factors can be accounted for by metabolic errors. While by no means inclusive, the tables below indicate the more common inborn errors of metabolism that present within the first days of life and the results of appropriate tests.

Early diagnosis and treatment are the keystones to the prevention of neurologic sequelae or death. When presented with a sick, full-term infant, pursue the usual sepsis work-up but do not neglect the evaluation for metabolic disease.
## Inborn Errors of Metabolism with Enzyme Deficiencies and Diagnostic Signs (Part I)

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Deficient Enzyme</th>
<th>Acid Base</th>
<th>Anion Gap</th>
<th>Plasma Glucose</th>
<th>Plasma Lactate</th>
<th>Plasma Pyruvate</th>
<th>Plasma Amino Acids</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Urea Cycle Disorders</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Carbamylphosphate synthetase (CPS I)</td>
<td>Respiratory alkalosis</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Absent citrulline, glutamine, alanine, and arginine</td>
<td></td>
</tr>
<tr>
<td>OTC deficiency</td>
<td>Respiratory alkalosis</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Same as CPS def.</td>
<td></td>
</tr>
<tr>
<td>Citrullinemia</td>
<td>Respiratory alkalosis</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Citrulline, glutamine, alanine, arginine</td>
<td></td>
</tr>
<tr>
<td>Argininosuccinase</td>
<td>Respiratory alkalosis</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>N</td>
<td>Argininosuccinate and its anhydrides, citrulline, glutamine, and arginine</td>
<td></td>
</tr>
</tbody>
</table>

**Disorders of Branched-chain Amino Acid Metabolism and Leucine Degradation**

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Acid Metabolism and Leucine Degradation</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Maple syrup urine</td>
<td>Branched-chain ketoacid dehydrogenase</td>
<td>Metabolic acidosis</td>
<td>N = 1</td>
<td>N</td>
</tr>
<tr>
<td>Isovaleric acidemia</td>
<td>Isovaleryl-CoA dehydrogenase</td>
<td>Metabolic acidosis</td>
<td>N = 1</td>
<td>N</td>
</tr>
<tr>
<td>Propionic acidemia</td>
<td>Propionyl-CoA dehydrogenase</td>
<td>Metabolic acidosis</td>
<td>N = 1</td>
<td>N</td>
</tr>
<tr>
<td>Methylmalonic acidemia</td>
<td>Methylmalonyl-CoA mutase, adenosylhomocysteinase</td>
<td>Metabolic acidosis</td>
<td>N = 1</td>
<td>N</td>
</tr>
</tbody>
</table>

**Disorders of Carbohydrate Metabolism**

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Metabolism</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Galactosemia</td>
<td>L-G-6-phosphatase; Lb.G-6-phosphatase translocase</td>
<td>Metabolic acidosis</td>
<td>1</td>
</tr>
<tr>
<td>Glycogen storage disease</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Others**

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Metabolism</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Nonketotic hyperglycinenia</td>
<td>Glycine cleavage complex</td>
<td>N to respiratory acidosis</td>
<td>N</td>
</tr>
<tr>
<td>Multiple carboxylase deficiency</td>
<td>Biotin, holocarboxylase synthetase</td>
<td>Metabolic acidosis</td>
<td>N = 1</td>
</tr>
<tr>
<td>Type II glutaric aciduria</td>
<td>Multiple acyl-CoA dehydrogenase, electron transfer flavoprotein (ETF)</td>
<td>Metabolic acidosis</td>
<td>N</td>
</tr>
<tr>
<td>Congenital lactic acidosis</td>
<td>Pyruvate dehydrogenase (PDH) complex, pyruvate carboxylase, mitochondrial electron transport defect</td>
<td>Metabolic acidosis</td>
<td>1</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
### Inborn Errors of Metabolism with Enzyme Deficiencies and Diagnostic Signs (Part II)

<table>
<thead>
<tr>
<th>Disorder</th>
<th>Plasma NH4</th>
<th>Urine DNPH*</th>
<th>Urine Ketones</th>
<th>Urine RS*</th>
<th>Urine Organic Acids</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Urea Cycle Disorders</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Carbamylphosphate synthetase (CPS)</td>
<td>4+</td>
<td></td>
<td></td>
<td></td>
<td>2-oxoisovaleric, 2-hydroxyisovaleric, 2-oxoisocaproic, 2-hydroxyisocaproic, 2-oxo-3-methylkalic, N-isovalerylglucose, 3-hydroxyisovaleric, free isovaleric acid</td>
<td>Odor of maple syrup</td>
</tr>
<tr>
<td>OTC deficiency</td>
<td>4+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>X-linked inheritance</td>
</tr>
<tr>
<td>Citrullinemia</td>
<td>4+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Argininosuccinic acidemia</strong></td>
<td>4+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Disorders of Branched-chain Amino Acid Metabolism and Leucine Degradation</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Maple syrup urine</td>
<td>N</td>
<td>+</td>
<td></td>
<td></td>
<td>3-hydroxypropionic acid, methylleuate</td>
<td>Neutropenia, thrombocytopenia</td>
</tr>
<tr>
<td>Isovaleric acidemia</td>
<td>N → 2+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Neutropenia, thrombocytopenia</td>
</tr>
<tr>
<td>Propionic acidemia</td>
<td>N → 4+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Methylmalonic acidemia</td>
<td>N → 4+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Disorders of Carbohydrate Metabolism</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Galactosemia</td>
<td>N</td>
<td></td>
<td>+</td>
<td></td>
<td>May have tyrosine metabolites with liver dysfunction</td>
<td>May present with gram-negative sepsis, cataracts, and hyperbilirubinemia</td>
</tr>
<tr>
<td>Glycogen storage disease</td>
<td>N</td>
<td></td>
<td>+</td>
<td></td>
<td></td>
<td>Cholesterol, triglycerides, and uric acid, may be masked in newborns by frequent feedings</td>
</tr>
<tr>
<td><strong>Others</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nonketotic hyperglycinemia</td>
<td>N</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Seizures usually prominent</td>
</tr>
<tr>
<td>Multiple carboxylase deficiency</td>
<td>N → 2+</td>
<td></td>
<td></td>
<td></td>
<td>3-methylcrotonyl, 3-methylglucose, 3-methylcapropropionic, 3-hydroxyisovaleric, methylleuate</td>
<td></td>
</tr>
<tr>
<td>Type II glutaric aciduria</td>
<td>N → 2+</td>
<td></td>
<td></td>
<td></td>
<td>Glutaric acid, 2-hydroxyglutaric acid, and ethylmalonic and 2-hydroxyisovaleric dicarboxylic acids</td>
<td>Odor of sweaty feet; dysmorphic features</td>
</tr>
<tr>
<td>Congenital lactic acidosis</td>
<td>N → 2+</td>
<td></td>
<td></td>
<td></td>
<td>Lactic acid</td>
<td>PBDH-facial dysmorphology may be present</td>
</tr>
</tbody>
</table>

*DNPH: 2-dinitrophenylhydrazine; RS: reducing substances*
### Other Disorders of Inborn Errors of Metabolism Reported in the Neonatal Period

- 3-Methylcrotonylglycinuria and 3-hydroxyisovaleric aciduria
- 2-Methylacetoacetyl-CoA thiolase deficiency
- Succinyl-CoA: 3-ketoacid-CoA transferase deficiency
- D-Glyceric acidemia
- 5-Oxoprolinuria
- Hyperornithinemia/hyperammonemia-homocitrullinuria syndrome
- 5,10-Methylene tetrahydrofolate reductase deficiency
- Molybdenum cofactor deficiency
- Short-chain acyl-CoA dehydrogenase deficiency
- Long-chain acyl-CoA dehydrogenase deficiency
- 2-Ketoadipic aciduria
- 3-Hydroxy-3-methylglutaryl-CoA lyase deficiency
- Fructose 1,6-diphosphatase deficiency
- Peroxisomal disorders: Zellweger syndrome, neonatal adrenoleukodystrophy, infantile Refsum’s disease, pseudo-Zellweger syndrome
- Hepatorenal tyrosinemia

#### Symptoms
- (vomiting, lethargy, coma) after 24 hours of age

- **Hyperammonemia**
  - Absent
  - Present
    - Galactosemia, MSUD, others
    - Acidosis and/or ketosis
      - Present
      - Absent
        - Disorder of organic acid metabolism—PAA, MMA, others
        - Urea cycle defect
          - Plasma citrulline
            - Absent-trace
            - 100-300 μM
            - >1000 μM
              - Urine orotate
                - Low
                - High
                - CPSD
                - OTCD
                - ALD
                - ASD

#### Key:
- MSUD = maple syrup urine disease
- PAA = propionic acidemia
- MMA = methylmalonic acidemia
- CPSD = carbamylphosphate synthetase deficiency
- OTC = ornithine transcarbamylase deficiency
- ASA = arginosuccinic acid
- ALD = arginosuccinate dehydrolase deficiency
- ASD = arginosuccinate synthetase deficiency

Making the diagnosis when initial tests show hyperammonemia.
General Protocol for Diet-responsive Disorders

1. Discontinue intake of offending compounds and precursors.
2. Correct fluid and electrolyte abnormalities.
3. Institute hemodialysis in cases of progressive hyperammonemia or coma.
4. Provide a minimum of 120 cal/kg/d utilizing intravenous and oral nutrition. Mead Johnson product MJ80056 is a convenient source of nonprotein calories.
5. Institute pharmacologic trial of specific vitamin cofactor.
6. Add minimal amounts of the essential offending compounds, as indicated by careful and frequent monitoring of plasma levels.
7. Adjust calories, fluids, and amounts of offending compounds individually according to growth and plasma concentrations.


Clinical Symptomatology of Inborn Errors of Metabolism in the Neonate or Infant

The newborn or older infant with an acute onset of nonspecific symptoms is an all too frequent dilemma for pediatricians. Along with infections, cardiac defects, gastrointestinal diseases, and insults to the central nervous system, however, an inborn error of metabolism (IEM) must be considered.

Symptoms indicating a possibility of an IEM (one or all):
1. Infant becomes acutely ill after period of normal behavior and feeding; this may occur within hours or weeks.
2. Neonate or infant has seizures and/or hypotonia, especially if seizures are not retraceable.
3. Neonate or infant has an unusual odor.

Symptoms indicating strong probability of an IEM, particularly when coupled with the above symptoms:
1. Persistent or recurrent vomiting
2. Failure to thrive (failure to gain weight or weight loss)
3. Apnea or respiratory distress (tachypnea)
4. Jaundice or hepatomegaly
5. Lethargy
6. Coma (particularly intermittent)
7. Unexplained hemorrhage
8. Family history of neonatal deaths, or of similar illness, especially in siblings
9. Parental consanguinity
10. Sepsis (particularly Escherichia coli, a common form of sepsis in patients with galactosemia)

INCIDENCE VS. PREVALENCE

The incorrect use of these two terms is prevalent among clinicians. This is unfortunate both for interpretation of clinical information and statistics, and for the accuracy of personal expression. The correct definitions are:

**Incidence**: The expression of the rate at which a certain event occurs. In particular, the incidence rate is a rate in which the numerator is the number of new cases of a disease in a population during a specified time and the denominator is the number of the population at risk.

\[
\text{Incidence rate} = \frac{\text{Number of new cases of a disease}}{\text{Total population at risk}} \quad \text{(Per unit of time)}
\]

**Prevalence**: The total cases in existence at a certain time in a designated area expressed as a rate.

\[
\text{Prevalence rate} = \frac{\text{Number of existing cases}}{\text{Total population}} \quad \text{(At a certain time in a designated area)}
\]

If the incidence is stable over time,

\[
\text{Prevalence} = \text{Incidence} \times \text{Average duration of disease}
\]

INJURIES

Is It Possible to Make a Difference?

Sweden has the lowest child injury death rate of any country in the world, 88 deaths in 1988 in a population of 8.5 million. We admit up front the list of caveats needed in comparing the homogeneous society of Sweden with the complex population of the United States. However, notice in the figure the progress in Sweden in lowering childhood injury fatalities since 1957-1959. In 1988 10 Swedish children drowned, compared to 100 in 1954. What happened?

![Bar chart showing injury fatalities, Sweden and the United States, 1957 through 1959 and 1986. Data from World Health Organization.](graphic)

For the past 25 years the task of lowering childhood injuries and deaths in Sweden was the responsibility of the Joint Committee for the Prevention of Childhood Accidents, which embarked on a three-pronged approach that has achieved remarkable results. The three parts of the process were:

1. A system for injury surveillance and prevention research to identify the important problems, to test countermeasures, and to implement better trauma care.
2. The provision of a safer environment for children through legislation and regulation. This included, for example, separating children from hazards such as traffic and making the child's environment safer (windows, stairs, stoves, etc.).
3. Educating the public about injury prevention.

It has made a difference.


**INTOEING**

"Doctor, My Kid Walks Funny," or What to Do About Intoeing

Parents of infants and toddlers just learning how to walk frequently notice the symptom of "toeing-in." Despite frantic expectations of special exercises, braces, or corrective shoes, most conditions that cause an intoed posture are rarely due to a pathologic or orthopedic anomaly and are not much affected by interventions. Indeed, the overwhelming majority of such cases are within the norms of physical development.

**A General Game Plan for the Child Who Presents With Intoeing**

1. Reassure most parents that the condition of intoeing will resolve on its own as the child progresses in his or her growth and development.
2. Identify those patients within the limits of normal development, explain the condition carefully to the parents, and monitor the child over the course of time to ensure resolution.
3. Avoid unnecessary treatments.
4. Distinguish the rare orthopedic entities from the normal variants in order to provide interventional therapy (see table).

<table>
<thead>
<tr>
<th>Causes of Intoeing in Childhood</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>INITIAL PRESENTATION</strong> (NONAMBULATORY CHILD)</td>
</tr>
<tr>
<td>Hips</td>
</tr>
<tr>
<td>b. Minimal or no internal rotation</td>
</tr>
</tbody>
</table>

Table continued on next page.
Causes of Intoeing in Childhood (Cont.)

<table>
<thead>
<tr>
<th>INITIAL PRESENTATION (NONAMBULATORY CHILD)</th>
<th>NORMAL RESOLUTION (AMBULATORY CHILD)</th>
<th>PROBLEMS REQUIRING CORRECTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Femur</td>
<td>High angle of anteversion (40-60° at birth) apparent only after external rotational contractures have been stretched out by ambulation. This angle is a cause of actual intoeing (see figure).</td>
<td>Gradual remodeling of angle in childhood reaching adult configuration of 10-20° between ages 5 and 10 years.</td>
</tr>
</tbody>
</table>

The angle of femoral anteversion is created by the long axis of the neck of the femur and the transcondylar axis of the knee joint and is normally 10° to 20° in adults (top). The angle is higher in newborns (40° to 60°) and young children (bottom), producing intoeing, but usually remodels to the adult angle by age 5 to 8.

Table continued on next page.
### Causes of Intoeing in Childhood (Cont.)

<table>
<thead>
<tr>
<th>Lower Leg</th>
<th>Initial Presentation (Nonambulatory Child)</th>
<th>Normal Resolution (Ambulatory Child)</th>
<th>Problems Requiring Correction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Internal tibial torsion is normal in a nonwalking child and a cause of actual intoeing (see figure).</td>
<td>Tibial torsion gradually unwinds, disappearing between ages 18 months and 4 years.</td>
<td>Unresolved tibial torsion that persists beyond age 6 or 7 is rare. It requires corrective osteotomy.</td>
<td></td>
</tr>
</tbody>
</table>

**Internal tibial torsion** is one cause of intoeing. When the patella is straight and facing forward, the normal position of the lateral malleolus is 10° to 15° posterior to the medial malleolus (top). If the lateral malleolus is on the same level as the medial malleolus, or anterior to it, internal tibial torsion is present (bottom).

*Table continued on next page.*
Causes of Intoeing in Childhood (Cont.)

<table>
<thead>
<tr>
<th>INITIAL PRESENTATION (NONAMBULATORY CHILD)</th>
<th>NORMAL RESOLUTION (AMBULATORY CHILD)</th>
<th>PROBLEMS REQUIRING CORRECTION</th>
</tr>
</thead>
<tbody>
<tr>
<td>Foot</td>
<td></td>
<td></td>
</tr>
<tr>
<td>a. Positional varus (the borders of the foot are straight and there is no actual foot deformity)</td>
<td>a. Positional varus disappears with standing, usually between 9 and 14 months. Intoeing caused by internal tibial torsion and increased femoral anteversion ceases as these conditions begin to resolve.</td>
<td></td>
</tr>
<tr>
<td>b. Metatarsus adductus (a fixed deformity of the foot in which the forepart of the foot is angled away from the foot’s main longitudinal axis toward the midline).</td>
<td>b. Rigid metatarsus adductus requires serial casting followed by the use of a passive holding device.</td>
<td></td>
</tr>
<tr>
<td>c. Equinovarus (or clubfoot)</td>
<td>c. Clubfoot requires correction with casting and surgery.</td>
<td></td>
</tr>
</tbody>
</table>

The physical examination should also include an assessment of the child’s spine. A search for spinal curvatures and dermatologic evidence of possible neurologic disease should be made. Minor neural tube defects, for example, may not be obvious in a preadolescent patient, but after the pubertal growth spurt they can present as asymmetrical muscle weakness and rotational weakness of the leg.


INTRAOSSEOUS INFUSION

The child presenting emergently with shock secondary to overwhelming sepsis, dehydration, trauma, and life-threatening status epilepticus demands immediate vascular access. This noble goal, unfortunately, is not always easily achieved. The intraosseous infusion of fluids and drugs directly into the bone marrow, however, is an especially useful skill for the pediatrician to acquire for such situations. It should be reserved for the emergencies noted above and employed only when other methods of intravenous access have failed.

Anatomy

The marrow sinusoids of long bones drain into medullary venous channels; nutrient and emissary veins drain into the systemic venous system. Marrow cavities are particularly appealing in patients with severe hypovolemia and peripheral circulatory shock, because they act as rigid and uncollapsible veins. Further, medications injected via intraosseous infusion are absorbed almost immediately into the general circulation. One caveat that needs mentioning is to be aware of the child’s age; the soft, vascular red marrow in long bones seen in...
infants and young children is physiologically replaced by the less vascular yellow marrow at approximately 5 years of age.

**Technique (see figure)**

1. The patient's leg should be restrained with a small sandbag placed behind the knee for support. The skin should be cleansed with povidone-iodine or alcohol using an aseptic technique. Local anesthetic is optional and may not be necessary in patients with depressed mental status.

   ![Diagram of bone marrow aspiration](image)

   Placement of the Illinois sternal or iliac bone marrow-aspiration needle in the proximal tibial location. The disposable needle has a flange at the top to make it easier to grip, a locking stylet to prevent the needle from being plugged with bone during insertion, and a screw mechanism to adjust the length of the exposed shaft. Depending on the gauge and manufacturer, the length of the shaft can be adjusted from 0.16 to 4.76 cm (1/16 to 1 7/8 in).

2. Disposable sternal or iliac bone-marrow aspiration needles (15 to 18 gauge) are preferable. The shaft should be short with a protective sheath in order to prevent the needle's tip from being forced too deeply into or through the bone.

3. The proximal tibia is the optimal site of insertion, because it precludes interference from ventilation or chest compressions during placement. The site of insertion should be on the midline of the anterior tibia, 1 to 3 cm or two fingers' width below the tibial tuberosity at an angle of 60° to 90° away from the growth plate. Advance the needle using a screwing or boring type of motion.

   [N.B.: alternative insertion sites include the distal tibia and the femur, 2 to 3 cm above the external condyles; the sternum and ilium are less suitable sites and should be avoided.]

4. Entry into the marrow space is confirmed by noting a lack of resistance after the needle has passed through the cortex. Marrow should be easily aspirated into a syringe and fluids should infuse freely. Before injection, hypertonic and alkaline solutions should be diluted.
5. The needle should stand upright without support but must be stabilized and secured by taping the flanges of the needle in order to prevent loss of access.
6. Flush the needle with a heparin-containing saline to prevent clotting before administering a conventional solution.
7. The insertion site must be observed for evidence of extravasation.
8. Once conventional vascular access has been obtained (normally within 1 to 2 hours after establishing the intraosseous infusion), the bone-marrow needle should be removed. Longer intraosseous infusions increase the risk of infectious complications.
9. A sterile dressing should be placed over the dressing site and pressure applied to the dressing for 5 minutes.

Possible Complications

1. Subcutaneous and occasional subperiosteal infiltration of fluid or leakage at the puncture site.
2. Clotting of marrow in the needle, which can impede access.
3. Localized cellulitis and subcutaneous abscesses have been reported in about 0.7% of all cases.
4. Osteomyelitis has been reported in approximately 0.6% of cases.
5. Theoretically, damage to the growth plates and marrow elements can occur, but this is rarely observed. Fat and bone emboli are also of concern but rarely occur.

Contraindications

The procedure is contraindicated in children with osteogenesis imperfecta, osteopetrosis, and an ipsilateral fractured extremity due to risk of subcutaneous extravasation. The risk of infectious complication increases when the needle is introduced through an area affected by cellulitis or burn injuries. The procedure should not be performed in children over the age of 5 years.


INTUSSUSCEPTION

The Need for Prompt Recognition

The pediatrician is usually the first physician to see a child with an intussusception. Prompt recognition of this acute disorder will reduce morbidity and mortality. Remembering the following facts will facilitate early diagnosis and improve management.

<table>
<thead>
<tr>
<th>Age of Patients</th>
<th>% Presenting at Given Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Under 12 months</td>
<td>52%</td>
</tr>
<tr>
<td>1-2 years</td>
<td>24%</td>
</tr>
<tr>
<td>2-3 years</td>
<td>10%</td>
</tr>
<tr>
<td>3-7 years</td>
<td>11%</td>
</tr>
<tr>
<td>Over 7 years</td>
<td>3%</td>
</tr>
</tbody>
</table>
**Iron Deficiency Anemia**

### Signs and Symptoms

<table>
<thead>
<tr>
<th>Sign or Symptom</th>
<th>% Presenting</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain</td>
<td>94%</td>
</tr>
<tr>
<td>Vomiting (at least once)</td>
<td>91%</td>
</tr>
<tr>
<td>Gross blood with stool</td>
<td>66%</td>
</tr>
<tr>
<td>Abdominal mass</td>
<td>59%</td>
</tr>
</tbody>
</table>

Patients typically are healthy infants and children with no previous history of gastrointestinal disease. Nearly all infants present with recent onset of abdominal pain and at least one episode of vomiting. The pain is characterized by the child's crying and drawing his legs into his abdomen. Males are affected about twice as often as females. The mass is usually sausage-shaped and is palpable along the course of the colon. On occasion one may elicit Dance's sign—an emptiness in the right lower quadrant that reflects the fact that the intussuscepting bowel has moved out of this portion of the abdomen.

### Etiology of Intussusception

In less than 10% of patients will an etiologic factor be determined. Specific causes include Meckel's diverticulum (most common), ileal polyp, ileal granuloma, inspissated meconium in patients with cystic fibrosis, Henoch-Schönlein purpura, and lymphosarcoma.

Although the barium reduction will successfully reduce approximately 75% of all intussusception, *it is advisable for all patients over 6 years of age to have elective exploratory laparotomy because of the high probability that intussusception at this age has a specific cause; it is frequently produced by an intestinal lymphosarcoma.*


---

**Iron Deficiency Anemia—189**

### A Progression of Findings

Throughout the world anemia is the most common manifestation of nutritional deficiency. In the U.S. iron deficiency is associated with the majority of nutritional anemias.

The onset of iron deficiency anemia is preceded by a sequence of abnormalities that may exist a considerable time before the anemia. Given that iron deficiency is a multisystem problem and that symptoms may occur well in advance of the onset of anemia, screening tests have been devised to detect iron deficiency before anemia is present. The sequential changes are indicated in the table.
Sequential Changes in the Development of Iron Deficiency

<table>
<thead>
<tr>
<th>STAGE I</th>
<th>STAGE II</th>
<th>STAGE III</th>
</tr>
</thead>
<tbody>
<tr>
<td>IRON DEPLETION</td>
<td>IRON-DEFICIENT ERYTHROPOIESIS</td>
<td>IRON DEFICIENCY</td>
</tr>
<tr>
<td>Serum ferritin EAR</td>
<td>Decreased levels</td>
<td>Decreased levels</td>
</tr>
<tr>
<td>Bone marrow iron</td>
<td>Decreased staining</td>
<td>Decreased staining</td>
</tr>
<tr>
<td>Total serum iron binding capacity (TIBC)</td>
<td>Normal</td>
<td>Elevated</td>
</tr>
<tr>
<td>Serum iron</td>
<td>Normal</td>
<td>Decreased levels</td>
</tr>
<tr>
<td>Erythrocyte protoporphyrins</td>
<td>Normal</td>
<td>Increased levels</td>
</tr>
<tr>
<td>Hemoglobin</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Mean corpuscular volume (MCV)</td>
<td>Normal</td>
<td>Normal or low</td>
</tr>
<tr>
<td>Red cell morphology</td>
<td>Normal</td>
<td>Normal</td>
</tr>
</tbody>
</table>


Blue Sclerae as a Sign of Iron Deficiency Anemia

In his classic textbook, *The Principles and Practice of Medicine*, Sir William Osler described a common but rarely noted finding among iron-deficient, undernourished teenage girls: the presence of blue sclerae. A group of British gastroenterologists recently studied the presence of blue sclerae in association with iron deficiency anemia and found the sign to be far more sensitive and equally specific for iron deficiency anemia than the presence of mucosal pallor. Further, the search for blue sclerae is easier to confirm, because it is not affected by skin thickness, blood transfusions, pigmentation, or perfusion, which can be confusing in an accurate assessment of mucosal pallor.

The blue sclerae sign in iron deficiency anemia most likely results from impaired collagen synthesis; iron is a vital cofactor in the hydroxylation of proline and lysine residues. The result is a thin sclera that allows greater visibility of the choroid, and a bluish color is observed. Blue sclerae are associated with other diseases, as listed below, but these disorders are extremely rare and
Iron Overdose—191

certainly less common than iron deficiency anemia in the general population. The presence of blue sclerae, therefore, should prompt the physician to consider a possible underlying iron deficiency.

**Disease Entities Associated with Blue Sclerae**

<table>
<thead>
<tr>
<th>Iron deficiency anemia</th>
<th>Rheumatologic disorders</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anemia secondary to chronic blood loss (e.g., duodenal ulcers, inflammatory bowel disease, gluten enteropathy)</td>
<td>(e.g., rheumatoid arthritis, systemic lupus erythematositis)</td>
</tr>
<tr>
<td>Osteogenesis imperfecta</td>
<td>Malignancies of the gastrointestinal tract</td>
</tr>
<tr>
<td>Inherited connective tissue disorders (e.g., Ehlers-Danlos syndrome, pseudoxanthoma elasticum)</td>
<td>Corticosteroid use</td>
</tr>
<tr>
<td></td>
<td>Anemia secondary to hookworm infection (ankylostomiasis)</td>
</tr>
<tr>
<td></td>
<td>Myasthenia gravis</td>
</tr>
</tbody>
</table>


**Koilonychia—A Sign of Iron Deficiency**

Koilonychia, or spooning of the nails, refers to the loss of the longitudinal and lateral convexity of the nail associated with thinning and fraying of its distal portion. The terminal and lateral borders are flared dorsally as the normal convexity is replaced by flattening and concavity of the nail plate.

Although spooning of the nail can be seen as a result of local fungal infections or as a hereditary abnormality, it is also an early sign of iron deficiency in infants and children as well as adults.

The nail of the index finger is more frequently and more severely deformed. The third finger and the thumb are also commonly involved. The spooning occurs symmetrically, which should provide a clue to diagnosis. If still in doubt, then look at the feet. Most infants with spoon nails will also display the same involvement on the nail of the big toe.

This provides a simple way of making a presumptive diagnosis of iron deficiency.


**IRON OVERDOSE**

**Iron Poisoning—An Emergency Assessment**

Because most patients with serum iron values in excess of 300 μg/dl are likely to develop signs and symptoms of iron overdose, which clinical signs and laboratory determinations do, in fact, predict a serum iron concentration of greater than 300 μg/dl and serve as a guide for admission and therapy?
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Five clinical and laboratory findings have been found to be significantly different in patients with serum iron concentrations greater than or less than 300 μg/dl. These five findings are:

1. White cell count > 15,000/mm³
2. Blood sugar > 150 mg/dl
3. Presence of vomiting
4. Presence of diarrhea
5. Radiopaque material visible on flat plate of abdomen.

Vomiting has the highest sensitivity value, as well as the highest negative predictive value. The absence of vomiting makes it highly unlikely that the patient has a serum iron in excess of 300 μg/dl.

Importantly, patients do not develop signs or symptoms of acute iron toxicity more than 6 hours after ingestion. Any patient who remains asymptomatic for 6 hours following ingestion may be discharged with minimal risk of having a dangerously elevated serum iron concentration.

In summary, initial management of iron overdose should consist of induced emesis or lavage with a large bore tube. If any of the five screening tests described above is positive, a serum iron concentration of greater than 300 μg/dl is likely, and a serum iron concentration should be obtained. If a delay in obtaining a serum iron is anticipated, the patient should be given an intramuscular dose of deferoxamine (50 mg/kg up to a maximum of 1 g). If the drug produces a “vin rose” color to the urine, indicating that the serum iron concentration exceeds the total iron-binding capacity, the patient requires treatment. Patients who have negative results for all five tests should be observed for at least 6 hours for symptoms. Those remaining asymptomatic may be discharged.


Never lend your car to anyone to whom you have given birth.

Erma Bombeck
Jaundice in Infancy

Visible jaundice occurs at serum bilirubin levels greater than 5 mg/dl. Most infants, full-term and premature, exhibit signs of a transient, unconjugated hyperbilirubinemia during the first week of life. This "physiologic jaundice" is the result of a complex interplay among several factors such as:

1. an elevated bilirubin load secondary to increased red blood cell (RBC) volume, decreased RBC survival, and an increased enterohepatic circulation.
2. defective hepatic uptake of bilirubin from the serum due to diminished levels of ligandin and competition for binding to intracellular proteins.
3. defective bilirubin conjugation caused by decreased UDP-glucuronyl-transferase activity.
4. defective bilirubin excretion.

The National Collaborative Perinatal Project has determined that the majority of infants with physiologic jaundice will not have a serum bilirubin level > 12.9 mg/dl if full term, or 16 mg/dl if premature. Risk factors for developing hyperbilirubinemia during the first week of life include breastfeeding, maternal diabetes, induction of labor with oxytocin, male sex, and Oriental race.

The essential questions to answer, when evaluating a jaundiced newborn, are the severity of the hyperbilirubinemia and the type (conjugated vs. unconjugated). A rapid rise in serum bilirubin, such as an infant who develops hyperbilirubinemia in the first 24 hours of life or a rise in serum bilirubin greater than 5 mg/dl/day, warrants immediate investigation. A jaundiced infant without risk factors or prolonged jaundice (>1 week in a full term; >2 weeks in a premature infant) also needs to be evaluated, as does the infant with conjugated hyperbilirubinemia.

Unconjugated Hyperbilirubinemia

If the fractionation of the serum bilirubin documents as unconjugated hyperbilirubinemia, then the search for its cause should progress as follows (please note that the history, physical examination and clinical course will help guide the extent of the evaluation):

1. Fractionate serum bilirubin
2. Blood type and Rh (mother and infant)
3. Hemoglobin, hematocrit, and reticulocyte count
4. Coombs test (direct and indirect)
5. Peripheral blood smear
6. Prothrombin time/partial thromboplastin time
7. Platelet count
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8. Alkaline denaturation of hemoglobin test (of emesis)—adult vs. fetal hemoglobin
9. Sepsis work-up (blood, urine, cerebrospinal fluid culture)
10. Thyroid screen thyroxine, triiodothyronine, thyroid-stimulating hormone
11. Phenobarbital trial
12. Interruption of breastfeeding

Differential Diagnosis for Unconjugated Hyperbilirubinemia

1. Physiologic jaundice
2. Hemolysis (e.g., ABO incompatibility; erythroblastosis fetalis; and red blood cell defects such as spherocytosis, elliptocytosis, G-6-PD deficiency, and pyruvate kinase deficiency)
3. Hemorrhage (e.g., birth trauma, cephalohematoma)
4. Breast milk jaundice
5. Swallowed maternal blood
6. Placental dysfunction
7. Sepsis
8. Clotting disorders
9. Infant of a diabetic mother
10. Hypothyroidism
11. Intestinal obstruction (e.g., pyloric stenosis, duodenal stenosis, or atresia)
12. Crigler-Najjar syndrome (an hereditary disorder of glucuronyl transferase resulting in an elevated unconjugated bilirubin level; Type II Crigler-Najjar is distinguished from Type I by a rapid decline in serum bilirubin level with phenobarbital therapy [5 mg/kg/day])
13. Lucey-Driscoll syndrome (a syndrome of retention jaundice due to defective bilirubin conjugation in infants resulting from an unidentified factor transmitted by the mother to her infant).

Conjugated Hyperbilirubinemia

Conjugated hyperbilirubinemia is always pathologic in the neonate and its presence demands a diagnostic evaluation. The primary emphasis of this work-up is to identify those infants with treatable infectious and metabolic diseases, recognizable congenital or genetic disorders, or extrahepatic obstruction who would benefit from surgical intervention. This evaluation should begin with a complete history and physical examination, and the laboratory evaluation should proceed as follows:

1. Fractionate serum bilirubin
2. Serum transaminases, alkaline phosphatase (or 5'-nucleotidase), albumin, cholesterol
3. Prothrombin time
4. Stool color
5. Cultures (blood, urine, CSF, etc.)
6. Hepatitis B surface antigen, TORCH titers, VDRL
7. Serum α1-antitrypsin level
8. Metabolic screen (urine/serum amino acids; urine for reducing substances)
9. Thyroid screen
10. Ophthalmologic examination
11. Sweat chloride
12. Skull, long bones, abdominal, and chest x-ray films
13. Abdominal ultrasound
14. Duodenal intubation (string test for duodenal fluid color, bilirubin, bile acids)
15. Hepatobiliary scintigraphy
16. Percutaneous liver biopsy
Differential Diagnosis of Conjugated Hyperbilirubinemia*

1. Extrahepatic obstruction
   a. Infantile obstructive cholangiopathy
      - Biliary atresia
      - Neonatal hepatitis
      - Choledocholithiasis
   b. Other causes
      - Bile plug syndrome
      - Choledocholithiasis
      - Spontaneous bile duct perforation
      - Extrinsic bile duct compression

2. Genetic and metabolic disorders
   a. Disorders of carbohydrate metabolism
      - Galactosemia
      - Fructosemia
      - Glycogen storage disease type IV
   b. Disorders of amino acid metabolism
      - Tyrosinemia
   c. Disorders of lipid metabolism
      - Niemann-Pick disease
      - Gaucher disease
      - Wolman disease
      - Cholesterol ester storage disease
   d. Chromosomal disorders
      - Trisomy 18
      - Down syndrome
   e. Miscellaneous genetic & metabolic disorders
      - α1-Antitrypsin deficiency
      - Neonatal hypopituitarism
      - Cystic fibrosis
      - Zellweger cerebrohepatorenal syndrome
      - Familial hepatosteatosis

3. Persistent intrahepatic cholestasis
   a. Paucity of intrahepatic bile ducts
   b. Arteriohepatic dysplasia
   c. Benign recurrent intrahepatic cholestasis
   d. Byler disease
   e. Hereditary cholestasis with lymphedema
   f. Trihydroxycoprostanic acidemia

4. Acquired intrahepatic cholestasis
   a. Infections
      - Hepatitis B (non-A, non-B?)
      - Syphilis
      - Toxoplasmosis
      - Rubella
      - Cytomegalovirus
      - Herpes
      - Varicella
      - Echovirus
      - Coxsackievirus
      - Leptospirosis
      - Tuberculosis
      - Bacterial sepsis
   b. Drug-induced cholestasis
   c. Cholestasis associated with parenteral nutrition


Progression of Dermal Icterus in the Newborn

The rate of rise of serum bilirubin in the newborn infant who is jaundiced should always be monitored by laboratory determinations. The pediatrician, however, through simple examination of the infant, may make some estimation as to the rate of rise of serum bilirubin.
Dermal icterus has been shown to progress in a cephalopedal fashion; that is, as the infant's bilirubin rises, more of the skin becomes icteric. The icterus begins at the head and neck and progresses caudally to the palms and soles. The following table correlates the level of indirect bilirubin with the area of skin that is icteric in full-term infants whose jaundice is not due to Rh incompatibility.

<table>
<thead>
<tr>
<th>Area of the Body</th>
<th>Range of Indirect Bilirubin (mg/100 ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Head and neck</td>
<td>4-8</td>
</tr>
<tr>
<td>Upper trunk</td>
<td>5-12</td>
</tr>
<tr>
<td>Lower trunk and thighs</td>
<td>8-16</td>
</tr>
<tr>
<td>Arms and lower legs</td>
<td>11-18</td>
</tr>
<tr>
<td>Palms and soles</td>
<td>&gt;15</td>
</tr>
</tbody>
</table>

As icterus progresses, the area that had been jaundiced remains jaundiced, so that the entire body is icteric when the bilirubin rises above 15 mg/100 ml. The fading of the icterus as the bilirubin level falls affects all body areas at the same time, so that the intensity rather than the extent of the staining fades. The staining may progress more rapidly in the low birth weight infant, whereas the infant with Rh disease may demonstrate a relative lag in dermal staining.

Correct estimation of the extent of icterus involves the examination of the completely undressed infant. Blue-white fluorescent light. Icterus may be detected by blanching the skin with pressure of the thumb and noting the color of the underlying skin. This is a more difficult determination to make in deeply pigmented black infants, but the palms and soles, at least, may be easily examined even in these patients.


JOINT PAIN

Common Causes

Chondromalacia patellae
Growing pains
Osteomyelitis
Overuse
Septic arthritis
Sickle-cell disease
Sympathetic effusion
Tietze's syndrome
Transient synovitis
Trauma
Contusion

Trauma (Cont.)
Fracture
Hemarthrosis
Sprain/strain
Viral arthritis
Adenovirus
Epstein-Barr virus
Hepatitis
Mumps
Rubella
Varicella
Uncommon Causes

Attention-seeking behavior
Child abuse
Foreign body
Legg-Calve-Perthes disease
Mycoplasma
Osgood-Schlatter disease
Osteochondritis dissecans
Popliteal cyst
Psoriatic arthritis

Reactive arthritis
Brucella
Campylobacter
Salmonella
Shigella
Yersinia

Refered pain (retroperitoneal/intra-peritoneal inflammation)
Slipped capital femoral epiphysis
Subluxation of the patella

Rare Causes

Bone tumors
Carpal-tarsal osteolysis
Congenital joint laxity
  Ehlers-Danlos syndrome
  Marfan syndrome
  Stickler’s syndrome
Cystic fibrosis
Fabry’s disease
Gaucher’s disease
Giardia
Gout
Hyperlipoproteinemia
Hyperparathyroidism
Idiopathic chondrolysis
Immunodeficiency
  Complement deficiency
  Hypogammaglobulinemia
Immunologic (Cont.)
  Kawasaki’s disease
  Mixed connective tissue disease
  Polyaarteritis nodosa
  Reiter’s syndrome
  Scleroderma
  Serum sickness
  Sjögren’s syndrome
  Systemic lupus erythematosus
  Leukemia
  Lipogranulomatosis
  Lyme disease
  Mucopolysaccharidosis
  Mycobacterial disease
  Psychogenic rheumatism
  Reflex sympathetic dystrophy
  Rickets
  Sarcoidosis
  Stevens-Johnson syndrome
  Subacute bacterial endocarditis
  Syphilis
    Charcot joint
    Infection
    Thyroid disease
    Villonodular synovitis
    Whipple’s disease

Juvenile Rheumatoid Arthritis—197

Features of Juvenile Rheumatoid Arthritis

Juvenile rheumatoid arthritis (JRA) is an entity with protean manifestations. There is a remarkable heterogeneity of JRA, which has been divided into three
Juvenile Rheumatoid Arthritis

major categories: (1) systemic, (2) pauciarticular-onset type (in which four or fewer joints are affected), and (3) polyarticular-onset type (the type most similar to typical rheumatoid arthritis). There has been some acceptance among rheumatologists and orthopedists to further subdivide the pauciarticular group. Early-onset pauciarticular JRA primarily affects young girls; chronic iridocyclitis is a common feature, as is the tendency to have positive antinuclear antibody tests. Late-onset pauciarticular JRA, on the other hand, affects mostly older boys; sacroiliitis is a common feature and many patients go on to develop ankylosing spondylitis in adult life.

<table>
<thead>
<tr>
<th>Features of the Three Major Types of JRA</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
</tr>
<tr>
<td><strong>SYSTEMIC ONSET</strong></td>
</tr>
<tr>
<td>High fever</td>
</tr>
<tr>
<td>Rheumatoid rash</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
</tr>
<tr>
<td>Splenomegaly</td>
</tr>
<tr>
<td>Hepatomegaly</td>
</tr>
<tr>
<td>Pericarditis</td>
</tr>
<tr>
<td>Myocarditis</td>
</tr>
<tr>
<td>Pneumonitis, pleuritis</td>
</tr>
<tr>
<td>Chronic iridocyclitis</td>
</tr>
<tr>
<td>Subcutaneous nodules</td>
</tr>
<tr>
<td>Leukocytosis</td>
</tr>
<tr>
<td>Rheumatoid factor</td>
</tr>
</tbody>
</table>

++++ = most cases; +++ = many cases; ++ = some cases; + = occasional cases; 0 = rare or no cases.


The greatest poem ever known
Is one all poets have outgrown:
The poetry, innate, untold,
Of being only four years old.

Christopher Morley
From To A Child
KAWASAKI’S DISEASE

Is It Really Kawasaki’s Disease?

Kawasaki’s disease, also known as mucocutaneous lymph node syndrome, occurs worldwide. It is an acute, febrile, multisystemic disease of children that is usually benign and self-limited. In the United States the annual incidence is 4.5–8.5 cases per 100,000 children below 5 years of age. In an outbreak in a community, the incidence can rise to 150 per 100,000 children.

Every house officer seems to make this diagnosis five times more frequently than that of scarlet fever and other more common red rashes. Is it really Kawasaki’s disease? The figure shows us what to expect if and when we are to make this diagnosis, and the list below helps us to distinguish this disorder from its imitators.

**Table:**

<table>
<thead>
<tr>
<th>Symptom/Sign</th>
<th>First Week</th>
<th>Second Week</th>
<th>Third Week</th>
<th>Fourth Week</th>
<th>Second Month</th>
</tr>
</thead>
<tbody>
<tr>
<td>Temperature</td>
<td>May respond</td>
<td>100%</td>
<td></td>
<td>94%</td>
<td>7%</td>
</tr>
<tr>
<td>Cervical adenitis</td>
<td>88%</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Conjunctival hyperemia</td>
<td>90%</td>
<td>50%</td>
<td></td>
<td>0%</td>
<td></td>
</tr>
<tr>
<td>Erythematous oral mucosa</td>
<td>90%</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Strawberry tongue</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Polymorphous exanthem</td>
<td>92%</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Edema, hands &amp; feet</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peeling nails</td>
<td>75%</td>
<td></td>
<td>94%</td>
<td>7%</td>
<td></td>
</tr>
<tr>
<td>Peeling palms &amp; soles</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Kawasaki’s Disease

Familiarity with its features will help you to make the diagnosis.

Major Manifestations

- Fever in excess of 38.5°C for 5 days
- Redness and induration of palms and soles
- Desquamation of skin over fingers during convalescence
- Polymorphous exanthem over trunk; no vesicles
- Conjunctivitis
- Redness and fissuring of the lips
- Strawberry tongue
- Diffuse redness of oropharynx
- Acute, nonpurulent swelling of cervical lymph nodes

Other features of the disease may include tachycardia, gallop rhythm, distant heart sounds, heart murmurs, EKG changes, diarrhea, proteinuria, pyuria, leukocytosis, mild anemia, elevated platelet count, increased erythrocyte sedimentation rate, and increasing level of IgE during period of illness.

Less frequent manifestations include arthralgia, arthritis, aseptic meningitis, and mild jaundice.

Mortality is approximately 0.3 to 2%. The most serious complication is vasculitis of the coronary arteries, although early deaths may result from severe myocarditis. Deaths are due primarily to thrombosis of large coronary-artery aneurysms and resultant myocardial infarction, which occurs late in the course of the disease (between the third and fourth week). Coronary angiography during the illness may reveal abnormalities in as many as 60% of patients. These include aneurysms, dilatation, stenosis, tortuosity, and irregularity of arterial vessel walls. These appear to regress with recovery, but at present, the long-term prognosis is unknown.

Age incidence: The incidence is highest in children of 1 year of age and approximately 80% of all patients are under 4 years of age.

Etiology: Unknown. No evidence of point source or person-to-person transmission has ever been documented.

Treatment: Unresponsive to antibiotics. When administered early, high-dose intravenous gamma globulin together with aspirin has been shown to be effective in reducing coronary artery abnormalities.

Recurrences: Rare.

May mimic some of the features of scarlet fever, measles, atypical measles, rubella, Stevens-Johnson syndrome, juvenile rheumatoid arthritis, staphylococcal scalded-skin syndrome, and acrodynia (mercury poisoning).

Clinical Features Associated with Kawasaki’s Disease

The clinical features that substantiate a diagnosis of Kawasaki’s disease are listed below.

Diagnostic Criteria for Kawasaki’s Disease

1. Fever lasting for at least 5 days
2. Presence of four of the following five conditions:
   a. Bilateral conjunctival injection
   b. Changes of the mucosa of the oropharynx, including injected pharynx, injected and/or dry fissured lips, strawberry tongue
   c. Changes of the peripheral extremities, such as edema and/or erythema of hands and/or feet, desquamation usually beginning periungually
   d. Rash, primarily truncal; polymorphous but nonvesicular
   e. Cervical lymphadenopathy
3. Illness not explained by other known disease process.

Other features of Kawasaki’s disease may help establish the diagnosis, may explain complications as they arise, or may allow for anticipatory management of the patient. These associated findings include the following:

Clinical Manifestations Associated with Kawasaki’s Diseases

<table>
<thead>
<tr>
<th>NONCARDIAC MANIFESTATIONS</th>
<th>CARDIAC MANIFESTATIONS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior uveitis</td>
<td>Aortic aneurism</td>
</tr>
<tr>
<td>Arthritis/arthralgia</td>
<td>Coronary aneurism</td>
</tr>
<tr>
<td>Aseptic meningitis</td>
<td>Late coronary artery occlusion</td>
</tr>
<tr>
<td>Hepatic dysfunction</td>
<td>Myocarditis</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>Pericardial effusion</td>
</tr>
<tr>
<td>Hydrops of the gallbladder</td>
<td></td>
</tr>
<tr>
<td>Pneumonitis</td>
<td></td>
</tr>
<tr>
<td>Sterile pyuria</td>
<td></td>
</tr>
<tr>
<td>Tympanitis</td>
<td></td>
</tr>
</tbody>
</table>

*Many experts believe that, in the presence of classic features, the diagnosis of Kawasaki’s disease can be made (and limited treatment instituted) before the fifth day of fever by experienced individuals.


Anterior Uveitis and Kawasaki’s Disease

Anterior uveitis involves inflammation of the vessels of the anterior uveal tract, including the iris and the ciliary body. Anterior uveitis, also referred to as iridocyclitis, is a common manifestation of Kawasaki’s disease, and its detection during the first week of illness may help establish the diagnosis. Slit-lamp examination is required in order to establish the presence of anterior uveitis, and, because the usual symptoms of eye pain and photophobia may be minimal, it has been suggested that ophthalmologic examination should be a routine part of the evaluation of patients suspected of having Kawasaki’s disease.
Other illnesses that may be associated with conjunctivitis in the pediatric patient are listed below. Anterior uveitis may be found in some of these conditions.

- Streptococcal and staphylococcal toxin-mediated diseases
- Adenovirus and other viral infections
  - (enterovirus, measles*)
- Stevens-Johnson syndrome*
- Leptospirosis*
- Yersinia pseudotuberculosis infection
- Rickettsial infection
- Reiter's syndrome*

Inflammatory bowel disease*
Post-infectious immune complex disease* (e.g., post-meningococcal)
Sarcoidosis*
Systemic lupus erythematosus*
Behçet's syndrome*
Juvenile rheumatoid arthritis
  - (esp. early-onset pauciarticular-type JRA)

*May have evidence of anterior uveitis on slit lamp examination.


KIDNEYS

A Technique for the Palpation of the Kidneys of Neonates

Congenital malformations of the urogenital tract occur in approximately 12% of all newborns. In 0.5% of all newborns, significant renal anomalies are present. These should be detected early in life in order to avoid subsequent complications. Almost all significant anomalies can be detected by careful abdominal palpation. A simple technique that will enable you to palpate the kidneys of 95% of all neonates is as follows:

1. Support the infant in a semireclining position facing you by placing your left hand behind the infant's shoulders, neck, and occiput.
2. Place the fingers of your right hand in the infant's left costovertebral angle posteriorly.
3. Use the thumb of your right hand to search the infant's abdomen systematically, at first superficially and then deeply.
4. Deep palpation is performed by applying gentle, steadily increasing pressure subcostally in a posterior and cephalad direction. The thumb can then be slipped downward without reducing the posteriorly directed pressure. Usually, the upper pole of the kidney can be felt trapped between the descending thumb and the posteriorly placed fingers.
5. Next, change hands and examine the opposite side of the abdomen.

After practice on some two dozen infants, this technique can be mastered and subsequently performed in 30 seconds. Because of its high yield, it deserves your optimal skill and attention.


**Pattern of Normal Language Development—A Summary**

<table>
<thead>
<tr>
<th>AGE</th>
<th>VOCALIZATION AND SPEECH</th>
<th>RESPONSE COMPREHENSION</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 mo</td>
<td>Much crying and whimpering; produces some vowel and few consonant sounds.</td>
<td>Smiles: decreases activity; startles at loud sounds.</td>
</tr>
<tr>
<td>3 mo</td>
<td>Different cries for pain, hunger, and discomfort; decreased crying time; some repetitive sounds (&quot;ga, ga, ga&quot;); coos and sighs.</td>
<td>Vocal gurgle in response to soothing voice; some imitative response to speech.</td>
</tr>
<tr>
<td>5 mo</td>
<td>Babbles; vocal play; many repetitive sounds, all vowels, m, k, g, b and p; laughs out loud.</td>
<td>Imitative response to speech decreased; turns and looks to sound; recognizes familiar voice; vocalizes displeasure.</td>
</tr>
<tr>
<td>7 mo</td>
<td>Considerable variety in babbling, loudness and rhythm of all vocalizations; adds d, t, n and w to repertory of sounds; talks to toys.</td>
<td>Gestures increase as part of vocal responses to stimuli; response to sound is increasingly influenced by visual factors.</td>
</tr>
<tr>
<td>9 mo</td>
<td>Cries to get attention; increasing variations in pitch: &quot;mama,&quot; &quot;dada&quot; and &quot;baba&quot; part of vocal play but not associated with a person or object.</td>
<td>Retreats from strangers, often accompanied by crying; may imitate hand clapping.</td>
</tr>
<tr>
<td>11 mo</td>
<td>May use one word correctly; imitates sounds and correct number of syllables, little crying.</td>
<td>Comprehends &quot;no no,&quot; responds to &quot;bye-bye&quot; or &quot;patty-cake&quot; with appropriate gestures.</td>
</tr>
<tr>
<td>1-2 yr</td>
<td>Much unintelligible jargon; all vowels present; improves articulation so that 25% of words intelligible; names many objects by 24 mo; much echolalia.</td>
<td>Recognizes 150-300 words by 24 mo; responds correctly to several commands, e.g., &quot;sit down,&quot; &quot;give me that,&quot; &quot;stand up,&quot; &quot;come here,&quot; etc.</td>
</tr>
<tr>
<td>2-3 yr</td>
<td>Tries new sounds but articulation lags behind vocabulary; 50-75% of words intelligible; often omits final consonants; jargon nearly absent.</td>
<td>Comprehends 800-1,000 words by 3 yr; responds to many commands using &quot;on,&quot; &quot;under,&quot; &quot;up,&quot; etc.</td>
</tr>
<tr>
<td>3-4 yr</td>
<td>Speech nears 100% intelligibility; faulty articulations of /l and /r/ frequent; uses 3-4 words in sentences; uses a few plurals by 4 yr.</td>
<td>Recognizes plurals, sex differences, adjectives and adverbs; comprehends complex sentences.</td>
</tr>
<tr>
<td>4-6 yr</td>
<td>Syntax correct by 6 yr, forms 5- or 6-word sentences that are compound or complex with some dependent clauses; fluent; articulation good except for /sh, z, ch/ and /j/; can express temporal relations; voice well modulated in conversation.</td>
<td>Understands 2,500-3,000 words; carries out commands involving 3-4 actions; comprehends &quot;if,&quot; &quot;because&quot; and &quot;why.&quot;</td>
</tr>
</tbody>
</table>

LEUKOKORIA

Leukokoria, or the white pupil, is generally viewed by the examining physician as a sign of ocular disease. It occurs when a lesion interferes with the path of the ophthalmoscope’s white light shined into a patient’s eyes. Under normal circumstances, the ingoing beam passes through a clear cornea, aqueous humor, pupil, lens, vitreous, and retina to reflect off the vascular choroid. Given the vascularity of the choroid, the light reflected out of the eye appears red in color and is termed the “normal red reflex.” While the red reflex is certainly important to document when performing an eye exam, it should be noted that even when it occurs intraocular lesions may be present. Indeed, any lesion not directly in the path of the ingoing beam would not interfere with the red reflex and would subsequently be undiagnosed. Listed below, in order of frequency, are some of the more common causes of leukokoria.

1. Cataracts
2. Persistent hyperplastic primary vitreous
3. Retrolental fibroplasia
4. Retinal dysplasia
5. Retinoblastoma
6. Chorioretinal coloboma
7. Retinal detachment
8. Retinoschisis
9. Congenital retinal folds
10. Persistent pupillary membrane
11. Hyaloid cysts
12. Uveitis
13. Nematode endophthalmitis (toxocara)
14. Panophthalmitis
15. Coats disease (unilateral exudative retinitis)
16. Norrie’s disease (sex-linked recessive disorder characterized by bilateral blindness following severe retinal detachments, deafness, and mental retardation)
17. Juvenile xanthogranuloma
18. Ocular tumors
19. Ocular trauma
20. Vitreous hemorrhages
21. Medullated nerve fibers
22. Chorioretinal degeneration
23. Incontinentia pigminti
24. Phakomatoses
25. High myopia
26. Intraocular foreign body


LIMB PAIN

Common Causes

Growing pains
Infection
Cellulitis
Osteitis
Osteomyelitis
Post-rubella vaccination
Infection (Cont.)
Septic arthritis
Soft-tissue abscess
Toxic synovitis
Viral myositis
Sickle-cell disease-vaso-occlusive crisis
Trauma

Chondromalacia patellae
Compartment syndromes
Dislocation and subluxation
Fracture
Hypermobility syndrome
Joint strain, sprain, internal damage
Myositis ossificans

Trauma (Cont.)

Pathologic fracture
Postimmunization
Shin splints
Soft-tissue contusion or hemorrhage
Stress fracture
Tendonitis, fasciitis, bursitis
Traumatic periostitis

Uncommon Causes

Accessory tarsal ossicle
Collagen vascular disease
(e.g., dermatomyositis, lupus)
Conversion reactions
Henoch-Schönlein purpura
Juvenile rheumatoid arthritis

Legg-Calvé-Perthes disease
Osgood-Schlatter disease
Osteochondritis dissecans
Rheumatic fever
Tarsal coalition

Rare Causes

Bone tumors (osteogenic sarcoma, Ewing's sarcoma, chondrosarcoma)
Cushing's syndrome
Familial Mediterranean fever
Hemophilia
Histiocytosis X
Hyperparathyroidism
Hypervitaminosis A
Inflammatory bowel disease
Leukemia
Mucopolysaccharidosis

Myopathies
Neuroblastoma
Osteoporosis
Popliteal cyst
Rickets
Scurvy
Slipped-capital femoral epiphysis
Soft-tissue tumors (rhabdomyosarcoma, fibrosarcoma)
Sympathetic reflex dystrophy

Common Causes

Attention-seeking behavior (usually after minor trauma)
Calluses/corn/ingrown toenails
Chondromalacia patellae
Contusion
Foreign body (especially plantar surface)
Fracture (may be occult)
Growing pains
Hemophilia (hemarthrosis, soft-tissue bleed)
Immunization (local reaction)
Leg length discrepancy

Mimicry
Myositis (acute viral)
Poorly fitting shoes (tight or loose)
Shin splints
Sickle-cell disease (painful crisis/infarction)
Soft-tissue/cutaneous infection
Sprain/strain
Tendonitis
Torsion deformities
Transient synovitis
## Uncommon Causes

<table>
<thead>
<tr>
<th>Arthritis (septic)</th>
<th>Neuromuscular disease (Cont.)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baker's cyst</td>
<td>Peripheral neuropathy (Cont.)</td>
</tr>
<tr>
<td>Blount's disease</td>
<td>Periodic paralysis</td>
</tr>
<tr>
<td>Bone tumor (benign and malignant)</td>
<td>Poliomyelitis</td>
</tr>
<tr>
<td>Calcaneal spurs</td>
<td>Tick paralysis</td>
</tr>
<tr>
<td>Child abuse</td>
<td>Radiculopathy</td>
</tr>
<tr>
<td>Congenital contractures</td>
<td>Spastic paralysis</td>
</tr>
<tr>
<td>Coxa vara</td>
<td>Osgood-Schlatter disease</td>
</tr>
<tr>
<td>Erythema nodosum</td>
<td>Osteochondritis dissecans</td>
</tr>
<tr>
<td>Legg-Calvé-Perthes disease</td>
<td>Osteomyelitis</td>
</tr>
<tr>
<td>Leukemia</td>
<td>Phlebitis</td>
</tr>
<tr>
<td>Neuromuscular disease</td>
<td>Plantar wart</td>
</tr>
<tr>
<td>Ataxia</td>
<td>Referred pain</td>
</tr>
<tr>
<td>CNS bleed</td>
<td>Discitis</td>
</tr>
<tr>
<td>CNS infection</td>
<td>Epidural/paraspinal abscess</td>
</tr>
<tr>
<td>Flaccid paralysis</td>
<td>Iliac adenitis</td>
</tr>
<tr>
<td>Migraine</td>
<td>Infratereitoneal infection/inflammation</td>
</tr>
<tr>
<td>Muscular dystrophy</td>
<td>Pelvic inflammatory disease</td>
</tr>
<tr>
<td>Peripheral neuropathy</td>
<td>Retroperitoneal mass</td>
</tr>
<tr>
<td>Causalgia</td>
<td>Slipped capital femoral epiphysis</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>Subluxation of the patella</td>
</tr>
<tr>
<td>Guillain-Barré syndrome</td>
<td></td>
</tr>
<tr>
<td>Heavy metal intoxication</td>
<td></td>
</tr>
</tbody>
</table>

## Rare Causes

<table>
<thead>
<tr>
<th>Arthritis/arthralgia</th>
<th>Hepatitis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Acute rheumatic fever</td>
<td>Hypervitaminosis A</td>
</tr>
<tr>
<td>Dermatomyositis</td>
<td>Hysteria</td>
</tr>
<tr>
<td>Henoch-Schönlein purpura</td>
<td>Intervertebral disc herniation</td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
<td>Köhler's disease</td>
</tr>
<tr>
<td>Juvenile rheumatoid arthritis</td>
<td>Larsen-Johanson disease</td>
</tr>
<tr>
<td>Kawasaki's disease</td>
<td>Neuroblastoma</td>
</tr>
<tr>
<td>Polyarteritis nodosa</td>
<td>Pott's disease</td>
</tr>
<tr>
<td>Serum sickness</td>
<td>Pyomyositis</td>
</tr>
<tr>
<td>Systemic lupus erythematosus</td>
<td>Rickets</td>
</tr>
<tr>
<td>Brucellosis</td>
<td>Scurvy</td>
</tr>
<tr>
<td>Caffey's disease</td>
<td>Sever's disease</td>
</tr>
<tr>
<td>Congenital joint laxity (Ehlers-Danlos)</td>
<td>Sinding-Larsen disease</td>
</tr>
<tr>
<td>Erythromelalgia</td>
<td>Trichinosis</td>
</tr>
<tr>
<td>Freiberg's disease</td>
<td></td>
</tr>
</tbody>
</table>

### LUMBAR PUNCTURE

#### Estimating Lumbar-Puncture Depth in Children

The lumbar puncture (LP) is a frequently used diagnostic procedure particularly among infants and young children presenting with an acute infectious illness. Yet
the procedure can be difficult, particularly for physicians who do not perform LPs with regularity, often resulting in inserting the needle too deeply and disrupting the venous plexus that lies beneath the dura on the anterior wall of the vertebral canal. Such a traumatic lumbar puncture is contaminated with blood, rendering the cerebrospinal fluid white cell count all but useless. Further, a bloody tap may confuse matters if a culture result is positive because the patient is bacteremic but does not have meningitis.

A group of pediatricians at the Medical College of Wisconsin have developed a linear regression analysis of how deep to insert the lumbar puncture needle based upon the child's body surface area in square meters and the depth at where CSF reflux occurred. Each lumbar puncture was performed by positioning the child in the right lateral decubitus position with maximal flexion at the waist and neck. The needle was inserted, perpendicular in relation to the back, at the L3 or L4 vertebral interspace. Using the following equation:

\[
\text{Depth of lumbar puncture} = 0.77 \text{ cm} + 2.56 \text{ (body surface area in m}^2\text{)}
\]

these physicians were able to estimate the depth of lumbar puncture to within approximately 5 mm in most young children without incurring trauma or CSF reflux (see figure).

Relation of the depth of lumbar puncture to body-surface area. Dotted lines represent the 95% confidence limits for the predicted depth of lumbar puncture.

How to Interpret the Bloody Tap

Spinal fluid is supposed to be clear. When it isn’t, the preliminary information received from the laboratory may be difficult to interpret. The effect of blood on CSF results (usually as a result of inserting the spinal needle too deeply and piercing the vascular plexus ventral to the epidural space) has actually been studied, and these studies make it possible to take a logical approach to the interpretation of the bloody tap.

**CSF WBC.** The effect of blood on the WBC is the most difficult parameter to calculate. Though it is intuitive that the WBC count would be altered in proportion to the WBC count in the peripheral blood, many studies have demonstrated that it is not this simple. Certainly when the observed CSF WBC is higher than would be predicted from the peripheral WBC/RBC ratio, or when the percentage of PMNs is higher than that in the peripheral blood count, infection involving the central nervous system should be suspected. However, management decisions should not be made based on the CSF WBC alone when the CSF is contaminated with blood.

**CSF Glucose.** When experiments have been done to determine the effect of mixing blood with CSF, no change in CSF glucose concentration can be demonstrated. Though it has been suggested that hypoglycorrhachia (abnormally low glucose in CSF) may result from RBC contamination, experimental studies have not confirmed this contention, and a low CSF glucose should be regarded as a low CSF glucose.

**CSF Protein.** There is no question that blood in the CSF raises the protein concentration. The increase in protein has been found to be approximately 1 mg/dl for every 1000 RBCs. This is only an approximation, however, and for most purposes the CSF protein concentration is not helpful when the tap is bloody.

**Xanthochromia.** When hemoglobin from lysed RBCs remains in the CSF for an extended period of time, the breakdown products oxyhemoglobin, methemoglobin, and bilirubin create a yellowish discoloration of the CSF after the specimen is centrifuged. CSF contaminated by fresh blood, as in a traumatic lumbar puncture, remains clear and colorless after centrifugation. The pigmentation that results from RBC breakdown persists for about 7 days after the hemorrhage has stopped. Since RBC lysis occurs in CSF after about 4 hours, specimens that are not analyzed within that time period may be xanthochromic even if the blood resulted from fresh contamination.

Traumatic lumbar puncture occurs in one out of every 5 LP attempts in pediatric patients, and the likelihood of a traumatic tap increases inversely with the age of the patient. Since bleeding usually occurs as a result of introducing the spinal needle too far, a method to avoid this problem has been devised. This method involves removing the spinal needle stylet once the needle has been introduced through the epidermis and the dermis. As the needle is very slowly advanced, the flow of spinal fluid can be seen as soon as the needle tip is in the subarachnoid space. The stylet should be replaced prior to withdrawing the needle to minimize the pressure gradient between the subarachnoid space and the atmosphere as the needle is withdrawn. It is important that the stylet remain in the needle until the epidermis and the dermis are traversed. Implants of small islands of skin during lumbar puncture have been reported to lead to the later development of epidermoid tumors along the needle track.

Lumbar Punctures and the Peripheral White Blood Cell Count

As if there weren't enough factors to consider in the performance of a lumbar puncture (LP), a team of Israeli pediatricians have brought up one more: the timing of obtaining the peripheral white blood cell (WBC) in relation to performing the LP. A prospective study of 26 neonates and infants suspected of having meningitis noted a significant increase in the peripheral WBC after the LP was completed (10,960 ± 3,500 cells/μl before the LP; 13,300 ± 3,970 cells/μl after the LP, p < 0.001). The greatest increase in white cells was seen in the neutrophil and lymphocyte fraction, presumably because these cells are so quickly released from the marginal granulocyte pool. There were no significant differences, before and after LP, in serum glucose, urea, hemoglobin, and platelet counts. While the LP does not impair interpretation of the CSF glucose to serum glucose ratio, the data depicted below make a strong case for obtaining the peripheral WBC count prior to performing the LP (see figure and table).
**Mean ± SD of the Blood Tests Studied Before and 10–15 Minutes After the LP Procedure**

<table>
<thead>
<tr>
<th>MEAN BLOOD LEVELS</th>
<th>BEFORE LP</th>
<th>AFTER LP</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC count (cells/µl)</td>
<td>10,960 ± 3,500</td>
<td>13,300 ± 3,970*</td>
</tr>
<tr>
<td>Glucose (mg/dl)</td>
<td>85.3 ± 13.4</td>
<td>84.1 ± 12.6</td>
</tr>
<tr>
<td>Hemoglobin (gr/dl)</td>
<td>11.6 ± 2.2</td>
<td>11.7 ± 2.2</td>
</tr>
<tr>
<td>Thrombocytes (x10³)</td>
<td>269 ± 113</td>
<td>315 ± 93</td>
</tr>
<tr>
<td>Urea (mg/dl)</td>
<td>6.3 ± 2.3</td>
<td>6.5 ± 2.5</td>
</tr>
</tbody>
</table>

* p < 0.001 (T-paired test).


**LYME DISEASE**

**Clinical Manifestations of Lyme Disease**

Lyme disease is an arthropod-borne infection that is heralded by a distinctive skin eruption (erythema chronicum migrans), followed, in stages, by neurologic or cardiac complications and arthritis. Its vector is the deer tick, *Ixodes dammini*, and the etiologic agent is the spirochete *Borrelia burgdorferi*. Because the clinical manifestations of Lyme disease are so protean in their presentation, with considerable overlap between the three major stages and their time of appearance, the accompanying temporally organized flow chart of its natural history ought to be useful, as well as the table.

**Tick Bite** 3 to 4 weeks

**Stage One**
- PRIMARY ERYTHEMA CHRONICUM MIGRANS
  - malaise, fever
  - "flu"-like illness
  - regional lymphadenopathy

**Stage Two**
- NEUROLOGIC COMPLICATIONS
  - VII nerve palsy
  - meningitis
  - radiculoneuritis

  (4 weeks after rash)

50%

**Stage Three**
- CARDITIS
  - heart block
  - myopericarditis

  (4 to 5 weeks after rash)

8%

Clinical manifestations of Lyme disease.
### Manifestations of Lyme Disease by Stage*

<table>
<thead>
<tr>
<th>System</th>
<th>Early Infection</th>
<th>Late Infection</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Localized (Stage 1)</strong></td>
<td><strong>Disseminated (Stage 2)</strong></td>
<td><strong>Persistant (Stage 3)</strong></td>
</tr>
<tr>
<td><strong>Skin</strong></td>
<td>Secondary annular lesions, malar rash, diffuse erythema or urticaria, evanescent lesions, lymphocytoma</td>
<td>Acrodermatitis chronica atrophicans, localized sclerodema-like lesions</td>
</tr>
<tr>
<td><strong>Musculo-skeletal system</strong></td>
<td>Migratory pain in joints, tendons, bursae, muscle, bone; brief arthritis attacks; myositis; osteomyelitis; panniculitis</td>
<td>Prolonged arthritis attacks, chronic arthritis, peripheral enthesopathy, periostitis or joint subluxations below lesions of acrodermatitis</td>
</tr>
<tr>
<td><strong>Neurologic system</strong></td>
<td>Meningitis, cranial neuritis, Bell's palsy, motor or sensory radiculoneuritis, subtle encephalitis, mononeuropathy, multiple, myelitis, chorea, cerebellar ataxia</td>
<td>Chronic encephalomyelitis, spastic parapareses, ataxic gait, subtle mental disorders, chronic axonal polyradiculopathy, dementia</td>
</tr>
<tr>
<td><strong>Lymphatic system</strong></td>
<td>Regional or generalized lymphadenopathy, splenomegaly</td>
<td></td>
</tr>
<tr>
<td><strong>Heart</strong></td>
<td>Atrioventricular nodal block, myopericarditis, pancarditis</td>
<td>Keratitis</td>
</tr>
<tr>
<td><strong>Eyes</strong></td>
<td>Conjunctivitis, iritis, choroiditis, retinal hemorrhage or detachment, panophthalmitis</td>
<td></td>
</tr>
<tr>
<td><strong>Liver</strong></td>
<td>Mild or recurrent hepatitis</td>
<td></td>
</tr>
<tr>
<td><strong>Respiratory system</strong></td>
<td>Nonexudative sore throat, nonproductive cough, adult respiratory distress syndrome</td>
<td></td>
</tr>
<tr>
<td><strong>Kidney</strong></td>
<td>Microscopic hematuria or proteinuria</td>
<td></td>
</tr>
<tr>
<td><strong>Genitourinary system</strong></td>
<td>Orchitis</td>
<td></td>
</tr>
<tr>
<td><strong>Constitutional symptoms</strong></td>
<td>Minor</td>
<td>Severe malaise and fatigue</td>
</tr>
</tbody>
</table>

*The classification by stages provides a guideline for the expected timing of the illness's manifestations, but this may vary from case to case.
* Systems are listed from the most to the least commonly affected.
† The inclusion of this manifestation is based on one or a few cases.


**What Is the Long-term Course of Lyme Disease in Children?**

The natural history of Lyme disease is not yet completely known. In a recent report the authors studied the long-term course of Lyme arthritis in 46 children in whom the onset of the disease occurred between 1976 and 1979 and who received...
no antibiotic therapy for at least the first 4 years of the illness. Of the 46 children (age range, 2 to 15 years), 33 (72%) initially had erythema migrans, 7 (15%) had influenza-like symptoms, and 6 (13%) had migratory joint pain. These manifestations were followed by brief attacks of arthritis, particularly affecting the knee. The percentage of children with recurrent episodes of arthritis declined each year. By year 4, only 10 children still had a mean of two episodes of arthritis per year; the duration of arthritis was generally longer in older children (P < 0.05). During the sixth year of illness, two children had keratitis, and more than 10 years after the onset of disease, a subtle encephalopathy developed in two other children. Of the 39 children whom the authors were able to contact in 1988–1989, 12 (31%) still had occasional brief episodes of joint pain and 1 had marked fatigue. All 46 children had positive IgG antibody responses to *Borrelia burgdorferi* throughout the illness and on long-term follow-up. As compared with those who became asymptomatic, the children with recurrent symptoms more often had IgM responses to the spirochete and had significantly higher IgG titers (P < 0.05).

The long-term course of initially untreated Lyme disease in children may include acute infection followed by attacks of arthritis and then over several years by keratitis, subtle joint pain, or chronic encephalopathy. This combination of symptoms in a patient with a high IgG antibody titer to *B. burgdorferi* is of concern, and the appropriate treatment for these patients is not yet certain.


**LYMPHADENOPATHY (GENERALIZED)**

**Common Causes**
- Infection (viral, fungal, spirochetal)
- Juvenile rheumatoid arthritis
- Serum sickness

**Uncommon Causes**
- Drug reactions
  - Anticonvulsants, antithyroid, isoniazid
  - Hodgkin’s disease
  - Infection, bacterial
  - Leukemia
  - Non-Hodgkin’s disease
  - Systemic lupus erythematosus

**Rare Causes**
- Angioimmunoblastic lymphadenopathy
- Dysgammaglobulinemia
- Gaucher’s disease
- Hemophagocytic syndromes
- Histiocytic medullary reticulosis
- Histiocytosis
- HIV infection
- Hyperthyroidism
- Metastatic neuroblastoma
- Niemann-Pick disease

**A Diagnostic Approach to Lymphadenopathy**

Is the lymphadenopathy generalized or localized? Generalized lymphadenopathy is defined as enlargement of more than two noncontiguous node regions. Generalized lymphadenopathy is caused by generalized disease.
Generalized Lymphadenopathy

What are associated signs and symptoms?
- Rash?
- Hepatosplenomegaly?
- Thyroid enlargement?
- Joint involvement?
- Heart and lung abnormalities?
- Pallor?
- Easy bruising?

Infections
- Exanthems
- Cytomegalovirus
- Infectious mononucleosis
- Infectious hepatitis
- Typhoid fever
- Malaria

Collagen Vascular Disease
- Lupus erythematosus
- Rheumatoid arthritis

Immunologic Reactions
- Serum sickness, drug reactions
- Granulomatous disease (sarcoid)

Storage Disease
- Gaucher’s disease
- Niemann-Pick disease

Malignancies
- Leukemia
- Lymphoma
- Histiocytosis
- Neuroblastoma, metastatic

Hyperthyroidism

Localized Lymphadenopathy

Signs of infection in the involved node?
Evidence of infection in the drainage area of node?
History of recent antigenic introduction in the node’s drainage area?


Axillary — Secondary to infections in the hand, arm, lateral chest wall, or lateral portion of the breast. May be result of recent immunization in the arm.

Epitrochlear — Secondary to infections on ulnar side of hand and forearm. Observed in tularemia when bite occurs on finger. Also seen in secondary syphilis.

inguinal — Infection in lower extremity, scrotum, penis, vulva, vagina, skin of lower abdomen, perineum, gluteal region, or anal canal. May be seen in lymphogranuloma venereum. May represent metastatic disease from testicular tumors or bony tumors of the leg. Immunization in leg.

Cervical — Generally the result of localized infection. See accompanying table for differential diagnosis.

Causes of Cervical Adenitis

<table>
<thead>
<tr>
<th>CAUSE</th>
<th>COMMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Viral upper respiratory infections</td>
<td>Most common cause. Nodes soft, minimally tender, and not associated with evidence of redness and warmth of overlying skin.</td>
</tr>
<tr>
<td>Bacterial infection</td>
<td>Streptococcus and staphylococcus most common etiologic agents. Usually secondary to previous or associated infection in drainage area of node. More frequently unilateral. Signs of infection — tenderness, warmth, and redness generally present. Look for primary focus of infection in scalp, mouth, pharynx, and sinuses.</td>
</tr>
<tr>
<td>Tuberculosis</td>
<td>Mycobacterium tuberculosis infections generally bilateral, involve multiple nodes. Associated with evidence of chest disease and systemic signs. Atypical mycobacteria infections more commonly unilateral initially. Not associated, in general, with other foci of disease. With either agent, evidence of local warmth and redness uncommon.</td>
</tr>
</tbody>
</table>

Table continued on next page.
### CAUSE

<table>
<thead>
<tr>
<th>Condition</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infectious mononucleosis</td>
<td>Fever, malaise, preceding upper respiratory infection often noted. Splenomegaly common. Atypical lymphocytes present. Epstein-Barr virus titers required for diagnosis in younger children.</td>
</tr>
<tr>
<td>Cytomegalovirus/Toxoplasmosis</td>
<td>Indistinguishable clinically from Epstein-Barr virus infections. Requires serologic studies to make the diagnosis.</td>
</tr>
<tr>
<td>Cat-scratch disease</td>
<td>History of contact with young cat. May be preceded by history of fever and malaise. Adenopathy restricted to area drained by initial cat scratch.</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>Disease bilateral. Chest x-ray almost always abnormal. May have keratitis, iritis and evidence of bone disease.</td>
</tr>
<tr>
<td>Hodgkin's disease</td>
<td>Common presenting symptom. Frequently unilateral at time of initial manifestation. Node is rubbery, nontender, and not associated with signs of inflammation. Make certain that supraclavicular involvement is not present. When present, strongly suspect lymphoma.</td>
</tr>
<tr>
<td>Non-Hodgkin's lymphoma</td>
<td>Bilateral at time of initial presentation in approximately 40% of patients. Cervical and submaxillary nodes commonly involved together.</td>
</tr>
</tbody>
</table>

### Algorithm: Generalized Lymphadenopathy

1. Is patient taking phenytoin, hydralazine, allopurinol, antithyroid or antileprosy medications?
   - Yes: Stop drug treatment and reevaluate in 2–3 wk
   - No:
     1. EBV titer or monospot
     2. Toxoplasma titer
     3. CMV serology
        - Positive: Dx specific disease
        - Negative: Blood cultures
          1. PPD
          2. Histoplasmosis, coccidioidomycosis, or brucella serology (if suggested by history)
             1. ANA
             2. CBC
             3. CXR
                - Positive: Dx specific disease
                - Negative: Exceptional biopsy

### Notes
- Chest x-ray almost always abnormal.
- Make certain that supraclavicular involvement is not present.

**BEST COPY AVAILABLE**
CERVICAL LYMPHADENOPATHY

SYSTEMIC ACUTE SYMPTOMS WITH FEVER

SMALL NODE < 3 cm

- NONFLUCTUANT
  - Measure node size
  - Throat cult. for strep.
  - Ant. nose culture for staph.
  - (opt)

- FLUCTUANT
  - Needle asp. with gm stain & culture
  - "A"

LARGE NODE > 3 cm

- SAME AS IN "C"

SINGLE, UNILOBAR NECK MASS

- NEEDLE ASP.

MULTILOBULAR

ASYMPTOMATIC

Careful Hx & PE to exclude systemic cause of cervical mass

CXR

CBC

IF ALL TESTS NONCONCLUSIVE AND NODE SIZE PERSISTS OR ENLARGES WITHIN 4-6 WK

TREAT WITH APPROPRIATE ANTIBIOTICS

REEVALUATE IN 2-3 WK

IF NO CHANGE IN SIZE, OR LARGER, CONT. Rx AND REEVALUATE IN 2-3 WK

IF NO CHANGE IN SIZE OR LARGER

EXCISIONAL BIOPSY
Algorithm: Localized Lymphadenopathy

**SUPRACLAVICULAR AREA**
- Chest x-ray
- Excisional biopsy
- Local infection?

**OCCIPITAL AREA**
- Local infection?

**PREAURICULAR AREA**
- Conjunctivitis
- Hx of exposure to kitten?
- PPD skin test
- VDRL (optional)

**SUBMAXILLARY and SUBMENTAL AREAS**
- Local Infection
- Bilateral
- PPD skin test
- VDRL
- Throat culture
- Consider excisional bx for non-Hodgkin's lymphoma

**AXILLARY AREA**
- Local Infection
- Hx of exposure to kitten?
- Consider PPD skin test, ANA, sed. rate

**EPITROCHLEAR AREA**
- Local infection?
- Consider ANA, VDRL, sed. rate

**INGUINAL and ILIAC AREAS**
- Local infection?
- Consider VDRL, culture for gonorrhea

**POPLITEAL AREA**
- Local Infection?
- Local Infection?

**ABDOMINAL and PELVIC AREAS**
- PPD skin test
- Sed. rate


MAGNESIUM

Magnesium Deficiency: A Common Problem

Magnesium deficiency has a reported incidence of 10% among all patients in tertiary care hospitals. It is frequently associated with hypocalcemia and/or hypokalemia. Despite one's good intentions, dosing the hypomagnesemic, hypokalemic, hypocalcemic patient with large amounts of calcium and potassium salts will do little to correct his or her electrolyte imbalance until the serum magnesium is restored to a normal value. Magnesium deficiency has numerous causes as noted in the table below; nutritional deficiency leads the list. Symptoms of hypomagnesemia are manifested primarily as neuromuscular irritability (e.g., tetany, tremors, and seizures). Changes in personality, anorexia, nausea, abnormal cardiac rhythms and EKG changes can also be seen (see table).

<table>
<thead>
<tr>
<th>Causes of Magnesium Deficiency*</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Nutritional</strong></td>
</tr>
<tr>
<td>Prolonged parenteral fluid</td>
</tr>
<tr>
<td>administration</td>
</tr>
<tr>
<td>Total parenteral nutrition</td>
</tr>
<tr>
<td>without magnesium</td>
</tr>
<tr>
<td>Starvation with metabolic</td>
</tr>
<tr>
<td>acidosis</td>
</tr>
<tr>
<td>Protein-calorie malnutrition</td>
</tr>
<tr>
<td>Kwashiorkor</td>
</tr>
<tr>
<td>Alcoholism</td>
</tr>
<tr>
<td><strong>Intestinal</strong></td>
</tr>
<tr>
<td>Chronic diarrhea from any cause</td>
</tr>
<tr>
<td>(e.g., chronic ulcerative colitis,</td>
</tr>
<tr>
<td>Crohn's disease, laxative abuse,</td>
</tr>
<tr>
<td>villous adenoma, adenocarcinoma</td>
</tr>
<tr>
<td>of rectum)</td>
</tr>
<tr>
<td>Malabsorption</td>
</tr>
<tr>
<td>Short-bowel syndrome</td>
</tr>
<tr>
<td>Gluten enteropathy</td>
</tr>
<tr>
<td>Pancreatic insufficiency with</td>
</tr>
<tr>
<td>steatorrhea</td>
</tr>
<tr>
<td>Tropical sprue</td>
</tr>
<tr>
<td>Familial malabsorption of</td>
</tr>
<tr>
<td>magnesium</td>
</tr>
<tr>
<td><strong>Renal</strong></td>
</tr>
<tr>
<td>Disease related</td>
</tr>
<tr>
<td>Renal tubular acidosis</td>
</tr>
<tr>
<td>Acute tubular necrosis</td>
</tr>
<tr>
<td>(diuretic phase)</td>
</tr>
<tr>
<td>Chronic glomerulonephritis</td>
</tr>
<tr>
<td>Chronic pyelonephritis</td>
</tr>
<tr>
<td>Familial and sporadic renal</td>
</tr>
<tr>
<td>magnesium loss</td>
</tr>
<tr>
<td>Drug related</td>
</tr>
<tr>
<td>Diuretics (furosemide, ethacrynly acid,</td>
</tr>
<tr>
<td>thiazides)</td>
</tr>
<tr>
<td>Antibiotics (gentamicin, tobramycin,</td>
</tr>
<tr>
<td>ticarcillin, carbenicillin,</td>
</tr>
<tr>
<td>amphotericin B)</td>
</tr>
<tr>
<td>Antineoplastic drugs (cisplatin, combinations of</td>
</tr>
<tr>
<td>antibiotics and cytotoxic agents)</td>
</tr>
<tr>
<td>Cyclosporine</td>
</tr>
</tbody>
</table>

*From Hospital Practice, February 15, 1987, with permission.

Table continued on next page.
Causes of Magnesium Deficiency (Cont.)

Endocrine and Metabolic

<table>
<thead>
<tr>
<th>Condition</th>
<th>Associated Malignancy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary and secondary aldosteronism</td>
<td>Primary hyperparathyroidism (due to hypercalcemia; immediately postoperatively in patients with osteitis fibrosa cystica)</td>
</tr>
<tr>
<td>Hyperthyroidism</td>
<td>Hyperparathyroidism (due to hypercalcemia; immediately postoperatively in patients with osteitis fibrosa cystica)</td>
</tr>
<tr>
<td>Excessive lactation</td>
<td>Uncontrolled diabetes with marked glucosuria</td>
</tr>
<tr>
<td>Pregnancy (third trimester)</td>
<td>Acute intermittent porphyria</td>
</tr>
<tr>
<td>Hypercalcemia</td>
<td></td>
</tr>
</tbody>
</table>

Congenital, Neonatal

<table>
<thead>
<tr>
<th>Maternal diabetes</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Maternal hyperparathyroidism or hypoparathyroidism</td>
<td></td>
</tr>
<tr>
<td>Exchange transfusions (citrate effect)</td>
<td></td>
</tr>
</tbody>
</table>


MALIGNANT DISEASE

Clues to Malignant Disease

Certain congenital malformations and acquired diseases are recognized to be associated with an increased incidence of malignancy. The conditions listed below should signal a warning and cause a high index of suspicion, regular observation, and appropriate studies for early detection of the associated malignancies.

In addition there are familial associations connected to certain tumors, such as brain tumors, Hodgkin’s disease, and Ewing sarcoma, which have been reported in siblings more frequently than chance alone would explain. Awareness of these associations may allow for earlier detection of both malignancies and congenital and other syndromes.

<table>
<thead>
<tr>
<th>Congenital and Acquired Conditions Associated with Increased Risk of Malignancy in Childhood</th>
</tr>
</thead>
<tbody>
<tr>
<td>CONDITION</td>
</tr>
<tr>
<td>Agammaglobulinemia</td>
</tr>
<tr>
<td>Albinism</td>
</tr>
<tr>
<td>Aniridia (non-familial)</td>
</tr>
<tr>
<td>Ataxia telangiectasia</td>
</tr>
<tr>
<td>Beckwith’s syndrome</td>
</tr>
<tr>
<td>Bloom’s syndrome</td>
</tr>
<tr>
<td>Chédiak-Higashi syndrome</td>
</tr>
<tr>
<td>Congenital X-linked immunodeficiency</td>
</tr>
<tr>
<td>D-trisomy</td>
</tr>
<tr>
<td>Down syndrome</td>
</tr>
</tbody>
</table>

Table continued on next page.
<table>
<thead>
<tr>
<th>CONDITION</th>
<th>ASSOCIATED MALIGNANCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>11p syndrome</td>
<td>Wilms’ tumor</td>
</tr>
<tr>
<td>Familial pelyposis of colon</td>
<td>Colonic carcinoma</td>
</tr>
<tr>
<td>Family history (first degree) of malignancy</td>
<td>Same or other malignancy</td>
</tr>
<tr>
<td>Fanconi anemia</td>
<td>Leukemia, hepatoma</td>
</tr>
<tr>
<td>Genitourinary anomalies</td>
<td>Wilms’ tumor</td>
</tr>
<tr>
<td>Giant cell hepatitis</td>
<td>Carcinoma of liver</td>
</tr>
<tr>
<td>Gonadal dysgenesis</td>
<td>Gonadal cancer</td>
</tr>
<tr>
<td>Hemihypertrophy</td>
<td>Wilms’ tumor, adrenal cortical carcinoma, liver carcinoma, hepatoblastoma</td>
</tr>
<tr>
<td>Hippel-Lindau disease</td>
<td>Pheochromocytoma</td>
</tr>
<tr>
<td>Horner syndrome</td>
<td>Neuroblastoma</td>
</tr>
<tr>
<td>IgM deficiency</td>
<td>Lymphoma</td>
</tr>
<tr>
<td>Irradiation:</td>
<td></td>
</tr>
<tr>
<td>in utero</td>
<td>Leukemia</td>
</tr>
<tr>
<td>of head and neck in early life</td>
<td>Thyroid carcinoma, brain and parotid tumors</td>
</tr>
<tr>
<td>for retinoblastoma</td>
<td>Osteosarcoma</td>
</tr>
<tr>
<td>for Wilms’ tumor</td>
<td>Osteosarcoma, ostcochondroma</td>
</tr>
<tr>
<td>for neuroblastoma</td>
<td>Osteosarcoma, ostcochondroma</td>
</tr>
<tr>
<td>Klinefelter’s syndrome</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Multiple endocrine adenomatosis I (Wermer syndrome)</td>
<td>Schwannoma</td>
</tr>
<tr>
<td>Multiple endocrine adenomatosis II (Sipple syndrome)</td>
<td>Thyroid carcinoma, pheochromocytoma</td>
</tr>
<tr>
<td>Multiple mucosal neuromas</td>
<td>Medullary thyroid carcinoma</td>
</tr>
<tr>
<td>Maternal stilbestrol during pregnancy</td>
<td>Vaginal adenocarcinoma</td>
</tr>
<tr>
<td>Neurofibromatosis</td>
<td>Pheochromocytoma, sarcoma, schwannoma, leukemia</td>
</tr>
<tr>
<td>Nevus sebaceous</td>
<td>Basal cell carcinoma</td>
</tr>
<tr>
<td>Poland’s syndrome</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Renal dysplasia</td>
<td>Wilms’ tumor</td>
</tr>
<tr>
<td>Severe combined immunodeficiency</td>
<td>Lymphoma, leukemia</td>
</tr>
<tr>
<td>13q syndrome</td>
<td>Retinoblastoma</td>
</tr>
<tr>
<td>Thyroid cancer (medullary)</td>
<td>Pheochromocytoma</td>
</tr>
<tr>
<td>Ulcerative colitis/regional ileitis</td>
<td>Colonic carcinoma</td>
</tr>
<tr>
<td>Wiskott-Aldrich syndrome</td>
<td>Lymphoma, lymphosarcoma</td>
</tr>
<tr>
<td>Xeroderma pigmentosum</td>
<td>Basal cell or squamous cell carcinoma</td>
</tr>
</tbody>
</table>

**MAPLE SYRUP URINE DISEASE**

**What Is the Intellectual Outcome?**

Maple syrup urine disease (MSUD) is the most common inborn error of amino acid metabolism and presents acutely in the neonatal period. Classic MSUD is characterized by lethargy, poor feeding, vomiting, and alternating periods of hypertonicity and flaccidity. In untreated disease, progressive neurologic deterioration, seizures, cerebral edema, coma, and death will usually occur within the first month of life.

A recent report of a controlled study of the intellectual outcome in 16 children with MSUD compared the outcome of MSUD diagnosed after symptoms became apparent with that of MSUD diagnosed prospectively and treated presymptomatically. Affected children treated presymptomatically had higher IQ scores than their affected siblings treated after their disease became symptomatic. The authors concluded that early and meticulous treatment of MSUD can result in intellectually normal children.

Therapy for MSUD consists of a diet low in branched-chain amino acids. However, little is known about the long-term clinical course of these patients or their lifespan.


**MARFAN'S SYNDROME**

**Diagnostic Criteria for Marfan's Syndrome**

**Diagnostic Manifestations**

**Skeletal**
- Anterior chest deformity, especially asymmetric pectus excavatum or carinatum
- Dolicostenomelia not due to scoliosis
- Arachnodactyly
- Vertebral column deformity
- Tall stature, especially compared with unaffected first-degree relatives
- High, narrowly arched palate and dental crowding
- Protrusio acetabulae
- Abnormal appendicular joint mobility
  - Congenital flexion contractures
  - Hypermobility

**Ocular**
- Ectopia lentis*
- Flat cornea
- Elongated globe
- Retinal detachment
- Myopia

**Cardiovascular**
- Dilation of the ascending aorta*
- Aortic dissection*
- Aortic regurgitation
- Mitral regurgitation due to mitral valve prolapse
- Calcification of mitral annulus
- Mitral valve prolapse
- Abdominal aortic aneurysm
- Dysrhythmia
- Endocarditis

* A major manifestation.
Pulmonary
Spontaneous pneumothorax
Apical bleb

Skin and integument
Striae atrophicae
Inguinal hernia
Other hernia

Central nervous system
Dural ectasia*
  Lumbosacral meningocele
Dilated cisterna magna
Learning disability (verbal performance discrepancy)
Hyperactivity with or without attention deficit disorder

Requirements for Diagnosis
In the absence of an unequivocally affected first-degree relative:
  Involvement of the skeleton and at least two other systems; at least one major manifestation.

In the presence of at least one unequivocally affected first-degree relative:
  Involvement of at least two systems: at least one major manifestation preferred, but this will depend on family’s phenotype.

Urine amino acid analysis in the absence of pyridoxine supplementation confirms absence of homocystinuria.

Conditions Most Often Considered in Differential Diagnosis
Homocystinuria
Familial or isolated mitral valve prolapse
Familial or isolated annuloaortic ectasia (Erdheim disease)
Congenital contractural arachnodactyly
Stickler syndrome


MEAN CORPUSCULAR VOLUME

Causes of Elevated MCV
Normal newborn
Reticulocytosis
Spurious elevations (cold agglutinins)
Hypothyroidism
Liver dysfunction
Down syndrome
Hereditary orotic aciduria
B12/folate deficiency
Aplastic anemia
Preleukemia
Leukemia
Diamond-Blackfan syndrome

Iron Deficiency or Thalassemia Trait?

Children with mild microcytic anemias are commonly encountered in the practice of pediatrics. Most of these patients have either iron deficiency or
thassemia trait. The use of red cell indices can provide a simple means of making a presumptive diagnosis without requiring serum iron determinations or hemoglobin electrophoresis.

Two formulas employing these indices have been proposed. They are as follows:

1. The Mentzer formula = \( \frac{MCV}{\text{Red cell count}} \)

   Interpretation: Values in excess of 13.5 strongly suggest that the patient has iron deficiency anemia, whereas values below 11.5 indicate that thalassemia trait is the most likely diagnosis.

2. The discriminant function = \( MCV - RBC - (5 \times \text{Hb}) - 3.4 \)

   Interpretation: Positive values suggest a diagnosis of iron deficiency, while negative values indicate that thalassemia trait is the cause of the microcytic anemia.

Caution: These formulas are useful only in uncomplicated situations. Confusing answers may be obtained in patients with associated hemolytic anemias or in patients with thalassemia minor who have hemorrhage or are pregnant, or in patients who are polycythemic secondary to chronic hypoxemia.

These formulas are useful in initial evaluation of patients. If iron deficiency is suggested by the formula and the patient does not respond to iron therapy, then further evaluation is indicated. A diagnosis of thalassemia trait should be confirmed in at least one family member.


MENINGITIS

Meningeal Signs

Inflamed meninges of any etiology (i.e., meningitis, intracranial bleeding, exposure to chemical agents, and CNS tumors) will produce the signs of Kernig and Brudzinski. These signs are frequently looked for and mentioned in the physical evaluation of a toddler or child suspected of having meningitis, yet the eponyms are often mixed up or interchanged in the excitement of describing such a patient. This confusion over the meningeal signs would, undoubtedly, inflame Drs. Kernig and Brudzinski, each of whom thought his sign was superior to the other's in diagnosing meningitis. In actuality, as bedside signs of meningeal irritation, both signs are of equal value.
**Kernig's sign.** This sign is named for the Russian physician Vladimir Michailovich Kernig (1840–1917), who described it in 1884. The examiner should place the patient in a supine position and passively flex the hip to 90° while the knee is also passively flexed to about 90° (see figure below). In a positive Kernig's sign, the patient's knee will resist passive extension and he may complain of intense pain, presumably induced by stretching inflamed sciatic nerve roots. The key here is that Kernig's sign begins with the knee.


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**Brudzinski's sign.** Several signs are named for the Polish pediatrician Josef von Brudzinski (1874–1917) who described the “neck sign” in 1909. With passive flexion of the patient's neck, the examiner should note a flexion at the knee and hips (see figure below). This sign, like Kernig's sign, is a reflection of the patient's protective response to preventing the eager examining physician from stretching his or her inflamed sciatic and intradural nerve roots.


---

**Brudzinski's leg signs.** Even before describing the neck sign, Brudzinski described contralateral reflex signs (the identical contralateral sign and the reciprocal contralateral sign). They are elicited less often than the neck sign.
As described by Verghese and Gallemore, the identical contralateral reflex sign is elicited in the supine position. When the hip and knee on one side are passively flexed by the examiner, the contralateral leg begins to flex.

The reciprocal contralateral reflex occurs when the leg that has flexed in response to passive flexion of the other leg begins to extend spontaneously. The reciprocal contralateral reflex then follows the identical contralateral reflex and looks like a little kick (see figure below). The contralateral reflex was present in 66% of the cases of meningitis observed by Brudzinski.

Brudzinski's leg signs. (A), Examiner passively flexes patient's leg (large arrow). (B) The identical contralateral sign: contralateral leg begins to flex (small arrows). (C) The reciprocal contralateral sign: the same leg that exhibited the active flexion begins to extend spontaneously, a reflex resembling a little kick (double arrows). (From Verghese A, Gallemore G: Rev Infect Dis 9:1191, 1987, with permission.)

References:

Persistent Pleocytosis

What can be learned from a repeat lumbar puncture in a child with bacterial meningitis? Not very much.

The following table lists the CSF findings in 30 patients with bacterial meningitis who had sequential lumbar punctures. None of the 30 suffered a relapse of meningitis.
### Sequential Spinal Fluid Changes in Bacterial Meningitis

#### H. INFLUENZAE (21 PATIENTS)

<table>
<thead>
<tr>
<th>Day of Therapy</th>
<th>Cells (mm³)</th>
<th>Glucose (mg/dl)</th>
<th>Protein (mg/dl)</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>3162 ± 905*</td>
<td>36 ± 7</td>
<td>126 ± 22</td>
<td>21</td>
</tr>
<tr>
<td>(0-15, 250)</td>
<td>(0-104)</td>
<td>(20-330)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>3925 ± 1477</td>
<td>52 ± 8</td>
<td>88 ± 29</td>
<td>8</td>
</tr>
<tr>
<td>(135-9300)</td>
<td>(27-76)</td>
<td>(40-260)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>1948 ± 732</td>
<td>45 ± 6</td>
<td>97 ± 16</td>
<td>9</td>
</tr>
<tr>
<td>(162-6100)</td>
<td>(16-58)</td>
<td>(70-140)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>544 ± 252</td>
<td>50 ± 7</td>
<td>108 ± 37</td>
<td>5</td>
</tr>
<tr>
<td>(51-1368)</td>
<td>(23-61)</td>
<td>(34-218)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4-7</td>
<td>305 ± 164</td>
<td>42 ± 4</td>
<td>107 ± 22</td>
<td>11</td>
</tr>
<tr>
<td>(48-1617)</td>
<td>(22-64)</td>
<td>(42-240)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8-10</td>
<td>44 ± 8</td>
<td>47 ± 3</td>
<td>38 ± 3</td>
<td>11</td>
</tr>
<tr>
<td>(11-77)</td>
<td>(32-55)</td>
<td>(29-54)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11-15</td>
<td>76 ± 10</td>
<td>51 ± 7</td>
<td>51 ± 7</td>
<td>18</td>
</tr>
<tr>
<td>(3-160)</td>
<td>(32-63)</td>
<td>(23-122)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;15</td>
<td>94 ± 20</td>
<td>48 ± 3</td>
<td>40 ± 3</td>
<td>11</td>
</tr>
<tr>
<td>(4-176)</td>
<td>(38-61)</td>
<td>(22-54)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

#### S. PNEUMONIAE (9 PATIENTS)

<table>
<thead>
<tr>
<th>Day of Therapy</th>
<th>Cells (mm³)</th>
<th>Glucose (mg/dl)</th>
<th>Protein (mg/dl)</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>3496 ± 934</td>
<td>28 ± 10</td>
<td>261 ± 57</td>
<td>9</td>
</tr>
<tr>
<td>(7-7535)</td>
<td>(0-100)</td>
<td>(13-530)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>6940 ± 6629</td>
<td>56 ± 16</td>
<td>--</td>
<td>2</td>
</tr>
<tr>
<td>(330-13,500)</td>
<td>(40-71)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>2006 ± 715</td>
<td>52 ± 6</td>
<td>142 ± 44</td>
<td>3</td>
</tr>
<tr>
<td>(495-3580)</td>
<td>(42-70)</td>
<td>(56-196)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>544 ± 252</td>
<td>50 ± 7</td>
<td>108 ± 37</td>
<td>5</td>
</tr>
<tr>
<td>(51-1368)</td>
<td>(23-61)</td>
<td>(34-218)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Numbers in parentheses represent the range.
*Mean ± standard error.
* = data insufficient.


### The Risk for Epilepsy Following Bacterial Meningitis

Most neurologic abnormalities following acute episodes of bacterial meningitis are transient and resolve without permanent loss or subsequent seizures. However, children with persistent neurologic deficits from cerebral injuries sustained during bacterial meningitis are at great risk for seizures, particularly if they had seizures during the acute episode. In most cases the epilepsy that followed occurred within 5 years of the acute illness and the seizures were focal or had a focal onset and therefore were difficult to control.

Children with normal neurologic examinations after the acute episode have an excellent chance of escaping serious neurologic sequelae, including seizures.


### Meningococcal Infection

#### Skin Lesions and Prognosis in Meningococcal Infections

The presence, type, and location of skin lesions in meningococcal infections can serve as a useful, immediate indicator of prognosis. The skin manifestations may be of three types:
Microcytosis

1. No lesions or other abnormalities
2. Erythematous, macular, and/or petechial lesions in a generalized distribution over the trunk and extremities.
3. Large purpuric or ecchymotic lesions, usually on the extremities, in association with petechiae.

The clinical manifestations of the disease vary little in groups with no lesions or in those with the generalized macular or petechial eruption, although the incidence of meningitis tends to be increased in those with no skin manifestations.

In contrast, patients with ecchymotic and purpuric lesions have a greater incidence of hyperpyrexia, coagulation abnormalities, shock, and death. The table below illustrates these differences.

<table>
<thead>
<tr>
<th>Type of Skin Lesions Related to Various Clinical and Laboratory Factors and Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>CLINICAL AND LABORATORY FACTORS</strong></td>
</tr>
<tr>
<td>------------------------------------</td>
</tr>
<tr>
<td>Meningitis</td>
</tr>
<tr>
<td>Leukocytosis</td>
</tr>
<tr>
<td>Hyperpyrexia</td>
</tr>
<tr>
<td>Shock</td>
</tr>
<tr>
<td>Bleeding diathesis</td>
</tr>
<tr>
<td>Mortality</td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>


Microcytosis

**Screening Methods in Evaluating Microcytosis**

<table>
<thead>
<tr>
<th>METHOD</th>
<th>FORMULA</th>
<th>THALASSEMA</th>
<th>IRON DEFICIENCY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Discriminant function</td>
<td>MCV ((5 \times Hb)) RBC 8.4</td>
<td>&lt;1</td>
<td>&gt;1</td>
</tr>
<tr>
<td></td>
<td>MCH (\div) RBC</td>
<td>&lt;3.8</td>
<td>&gt;3.8</td>
</tr>
<tr>
<td></td>
<td>MCV (\div) RBC</td>
<td>&lt;13</td>
<td>&gt;13</td>
</tr>
<tr>
<td></td>
<td>0.01 (\times) MCH (\times) MCV</td>
<td>&lt;1,530</td>
<td>&gt;1,530</td>
</tr>
<tr>
<td>RBC count</td>
<td>(&gt;5.0 \times 10^{12}/L)</td>
<td>(&lt;5.0 \times 10^{12}/L)</td>
<td></td>
</tr>
<tr>
<td>Osmotic fragility</td>
<td>Percent hemolysis</td>
<td>&lt;95%</td>
<td>&gt;95%</td>
</tr>
<tr>
<td>Coefficient of variation*</td>
<td>(\sigma/\mu \times 100)</td>
<td>&lt;14%</td>
<td>&gt;14%</td>
</tr>
<tr>
<td>Volume distribution curve</td>
<td>EVR(_{50})**</td>
<td>&lt;26 fl</td>
<td>&gt;27 fl</td>
</tr>
</tbody>
</table>

* Where \(\mu\) = median cell volume and \(\sigma\) = standard deviation.
** Estimated volume range for 50% of cells.

MILESTONES

Milestones of Development—A Summary

Most important milestones in italics

Newborn  Prone—pelvis high, knees under abdomen.
2-4 weeks  Watches mother intently as she speaks to him.
1 month  Ventral suspension (held prone, hand under abdomen)—head up momentarily: elbows flexed: hips partly extended, knees flexed.
4-6 weeks  Smiles at mother in response to overtures.
6 weeks  Ventral suspension—head held up momentarily in same plane as rest of body. Some extension of hips and flexion of knees and elbows.
Prone—pelvis largely flat, hips mostly extended. (But when sleeping the baby lies with pelvis high, knees under abdomen, like newborn baby.)
Pull to sit from the supine—much head lag, but not complete: hands often open.
Supine—follows object 90 cm away over angle of 90°.
2 months  Ventral suspension—maintains head in same plane as rest of body. Hands largely open.
Prone—chin off couch. Plane of face 45° to couch.
Smiles and vocalizes when talked to.
Eyes—follow moving person.
3 months  Ventral suspension—holds head up long time beyond plane of rest of body.
Prone—plane of face 45°-90° from couch.
Pulled to sit—only slight head lag.
Hands loosely open.
Holds rattle placed in hand.
Vocalizes a great deal when talked to.
Follows object for 180° (lying supine).
Turns head to sound (3 to 4 months) on a level with the ear.
4 months  Prone—plane of face at 90° to couch.
Hands come together.
Pulls dress over face.
Laughs aloud.
5 months  Prone—weight on forearms.
Pulled to sit—no head lag.
Supine—feet to mouth. Plays with toes.
Able to go for object and get it.
6 months  Prone—weight on hands, extended arms.
Pulled to sit—no head lag.
Supine—lifts head spontaneously.
Sits on floor, hands forward for support.
6 months
(Cont.) Held in standing position—full weight on legs.
Rolls, prone to supine.
Begins to imitate (e.g., a cough).
Chews.
Transfers cube from one hand to another.

7 months Sits on floor seconds, no support.
Roll, supine to prone.
Held standing—bounces.
Feeds self with biscuit.
Attracts attention by cough or other methods.
Turns head to sound below level of ear.

8 months Sits unsupported. Leans forward to reach objects.
Turns head to sound above level of ear.

9 months Stands, holding on. Pulls to stand or sitting position.
Crawls on abdomen.

9–10 months Index finger approach.
Finger thumb apposition—picks pellet between tip of thumb and tip of forefinger.

10 months Creeps, hands and knees, abdomen off couch.
Can change from sitting to prone and back.
Pulls self to sitting position.
Waves bye.
Plays patacake.
Helps to dress—holding arm out for coat, foot for shoe, or transferring object from one hand to another for sleeve.

11 months Offers object to mother, but will not release it.
One word with meaning.
Sitting—pivots round without over-balancing.
Walks, holding on to furniture: walks 2 hands held.

One year 2–3 words with meaning.
Prone—walks on hands and feet like bear.
Walks, one hand held.
Casting objects, one after another, begins.
Gives brick to mother.

13 months Walks, no support.
Mouthing of objects stopped.
Slobbering largely stopped.

15 months Creeps up stairs. Kneels.
Cubes—tower of two.
Takes off shoes.
Feeds self, picking up an ordinary cup, drinking, putting it down.
Imitation of mother in domestic work ('Domestic mimicry').
Jargon.

18 months No more casting.
Gets up and down stairs, holding rail.
Jumps, both feet.
<table>
<thead>
<tr>
<th>Age</th>
<th>Milestones</th>
</tr>
</thead>
<tbody>
<tr>
<td>18 months</td>
<td>Seats self in chair.</td>
</tr>
<tr>
<td>(Cont.)</td>
<td>Cubes—tower of 3–4.</td>
</tr>
<tr>
<td></td>
<td>Throws ball without falling.</td>
</tr>
<tr>
<td></td>
<td>Takes off gloves, socks, unzips.</td>
</tr>
<tr>
<td></td>
<td>Manages spoon well.</td>
</tr>
<tr>
<td></td>
<td>Points to 3 parts of body on request.</td>
</tr>
<tr>
<td></td>
<td>Books—turns pages, 2 or 3 at a time.</td>
</tr>
<tr>
<td></td>
<td>Points to some objects, on request.</td>
</tr>
<tr>
<td></td>
<td>Toilet control—tells mother that he wants potty. Largely dry by day.</td>
</tr>
<tr>
<td>21–24 months</td>
<td>Spontaneously joins 2 or 3 words together to make sentence.</td>
</tr>
<tr>
<td>2 years</td>
<td>Picks up object from floor without falling.</td>
</tr>
<tr>
<td></td>
<td>Runs.</td>
</tr>
<tr>
<td></td>
<td>Kicks ball without overbalancing.</td>
</tr>
<tr>
<td></td>
<td>Turns door knob, unscrews end.</td>
</tr>
<tr>
<td></td>
<td>Cubes—tower of 6 or 7.</td>
</tr>
<tr>
<td></td>
<td>Puts on shoes, socks, pants: takes off shoes, socks.</td>
</tr>
<tr>
<td></td>
<td>Points to 4 parts of body on request.</td>
</tr>
<tr>
<td></td>
<td>Pencil—imitates vertical and circular strokes.</td>
</tr>
<tr>
<td></td>
<td>Book—turns pages singly.</td>
</tr>
<tr>
<td></td>
<td>Mainly dry at night.</td>
</tr>
<tr>
<td></td>
<td>Climbs stairs, two feet per step.</td>
</tr>
<tr>
<td>24 months</td>
<td>Motor: Gross Runs well, no falling.</td>
</tr>
<tr>
<td></td>
<td>Walks up and down stairs alone.</td>
</tr>
<tr>
<td></td>
<td>Kicks large ball on request.</td>
</tr>
<tr>
<td></td>
<td>Fine Turn pages of book singly.</td>
</tr>
<tr>
<td></td>
<td>Adaptive Builds tower of 6–7 cubes.</td>
</tr>
<tr>
<td></td>
<td>Aligns cubes for train.</td>
</tr>
<tr>
<td></td>
<td>Imitates vertical and circular strokes.</td>
</tr>
<tr>
<td></td>
<td>Language Uses pronouns.</td>
</tr>
<tr>
<td></td>
<td>Three-word sentences; jargon discarded.</td>
</tr>
<tr>
<td></td>
<td>Carries out 4 directions with ball (“on the table,” “to mother,” “to me,” “on the chair”).</td>
</tr>
<tr>
<td></td>
<td>Personal-social Verbalizes toilet needs consistently.</td>
</tr>
<tr>
<td></td>
<td>Pulls on simple garment.</td>
</tr>
<tr>
<td></td>
<td>Inhibits turning of spoon in feeding.</td>
</tr>
<tr>
<td></td>
<td>Plays with domestic mimicry.</td>
</tr>
<tr>
<td>30 months</td>
<td>Motor: Gross Jumps up and down.</td>
</tr>
<tr>
<td></td>
<td>Walks backward.</td>
</tr>
<tr>
<td></td>
<td>Fine Holds crayon in fist.</td>
</tr>
<tr>
<td></td>
<td>Adaptive Copies crude circle, closed figure.</td>
</tr>
<tr>
<td></td>
<td>Names some drawings: house, shoe, ball, dog.</td>
</tr>
<tr>
<td></td>
<td>Language Refers to self as “I”.</td>
</tr>
<tr>
<td></td>
<td>Knows full name.</td>
</tr>
<tr>
<td></td>
<td>Personal-social Helps put things away.</td>
</tr>
<tr>
<td></td>
<td>Unbuttons large buttons.</td>
</tr>
</tbody>
</table>

212
3 years  
*Motor: Gross* Alternates feet going upstairs.  
Jumps from bottom step.  
Rides tricycle, using pedals.  

*Fine* Holds crayon with fingers.  

*Adaptive* Builds tower of 9–10 cubes.  
Imitates 3-cube bridge.  
Names own drawing.  
Copies circle and imitates cross.  

*Language* Uses plurals.  
Gives action in picture book.  
Gives sex and full name.  
Obey 2 prepositional commands ("on," "under").  

*Personal-social* Feeds self well.  
Puts on shoes.  

4 years  
*Motor* Walks downstairs alternating feet.  
Does broad jump.  
Threws ball overhand.  
Hops on one foot.  

*Adaptive* Draws man with 2 parts.  
Copies cross.  
Counts 3 objects with correct pointing.  
Imitates 5-cube gate.  
Picks longer of two lines.  

*Language* Names 1 or more colors correctly.  
Obey 5 prepositional commands ("on," "under,"  
"in back," "in front," "beside").  

*Personal-social* Washes and dries face and hands; brushes teeth.  
Distinguishes front from back of clothes.  
Laces shoes.  
Goes on errands outside of home.  

5 years  
*Motor* Skips, alternating feet.  
Stands on 1 foot more than 8 seconds.  
Catches bounched ball.  

*Adaptive* Builds 2 steps with cubes.  
Draws unmistakable man with body, head, etc.  
Copies triangle.  
Counts 10 objects correctly.  

*Language* Knows 4 colors.  
Names penny, nickel, dime.  
Descriptive comment on pictures.  
Carries out 3 commissions.  

*Personal-social* Dresses and undresses without assistance.  
Asks meaning of words.  
Prints few letters.  

6 years  
*Motor* Advanced throwing.  
Stands on each foot alternately, eyes closed.  
Walks line backward, heel-toe.  

*Adaptive* Builds 3 steps with blocks.  
Draws man with neck, hands, and clothes.  

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6 years  Adaptive  Adds and subtracts within 5.  (Cont.)  (Cont.)  Copies diamond.  Language  Uses Stanford-Binet items (vocabulary).  Defines words by function or composition, e.g.,  “house is to live in.”  Personal-  social  Ties shoelaces.  Differentiates A.M. and P.M.  Knows right from left.  Counts to 30.


MONONUCLEOSIS

“Alice in Wonderland” Syndrome and Infectious Mononucleosis

Central nervous system involvement is estimated to occur in anywhere from 0.7 to 20% of patients with infectious mononucleosis. The 20% figure includes electroencephalographic abnormalities as a sole manifestation of central nervous system disease. The neurologic abnormalities may range from acute meningoencephalitis to facial diplegia, retinal abnormalities, mononeuritis, and the Guillain-Barré syndrome.

To this list of neurologic complications should be added the presence of metamorphopsia or the “Alice in Wonderland” syndrome. Metamorphopsia refers to the complaints of distortions in the apparent sizes, shapes, and spatial relations of objects seen. This symptom has previously been recognized in some patients with migraine, epilepsy, or drug-induced hallucinations.

When it occurs in infectious mononucleosis, as a manifestation of central nervous system involvement, it may last from three weeks to three months.

When the patient begins to see things peculiarly, be sure you see correctly the peripheral blood smear, the Mono Spot Test, and, if necessary, the Epstein-Barr virus titers.


Complications of Infectious Mononucleosis

Most children with infectious mononucleosis experience a typical episode without complications. However, complications, when they do occur, may be so dramatic that they become the principal manifestation of the disease. Among the most severe complications, and one perhaps most feared by clinicians, is splenic
rupture, which can occur with minor trauma. Also, swelling of the upper airway may be very severe and cause occlusion. It is obviously important to recognize the many complications of this common disease. They include the following:

<table>
<thead>
<tr>
<th>Neurologic</th>
<th>Cardiac</th>
<th>Hematologic</th>
</tr>
</thead>
<tbody>
<tr>
<td>Encephalitis</td>
<td>Pericarditis</td>
<td>Hemolytic anemia</td>
</tr>
<tr>
<td>Guillain-Barré syndrome</td>
<td>Myocarditis</td>
<td>Thrombocytopenia</td>
</tr>
<tr>
<td>Facial nerve palsy</td>
<td></td>
<td>Granulocytopenia</td>
</tr>
<tr>
<td>Meningoencephalitis</td>
<td></td>
<td>Aplastic anemia</td>
</tr>
<tr>
<td>Aseptic meningitis</td>
<td></td>
<td>Hemolytic-uremic syndrome</td>
</tr>
<tr>
<td>Transverse myelitis</td>
<td></td>
<td>Disseminated intravascular coagulation</td>
</tr>
<tr>
<td>Seizures</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peripheral neuritis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mononeuritis multiplex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Optic neuritis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute psychosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diplopia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Reye's syndrome</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subacute sclerosing panencephalitis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Perceptual distortions (Alice in Wonderland syndrome)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Cardiac</th>
<th>Hematologic</th>
<th>Pulmonary</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pericarditis</td>
<td>Hemolytic anemia</td>
<td>Airway obstruction</td>
<td>Splenic rupture</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>Thrombocytopenia</td>
<td>Interstitial pneumonitis</td>
<td>Glomerulonephritis</td>
</tr>
<tr>
<td></td>
<td>Granulocytopenia</td>
<td>Pulmonary infiltration</td>
<td></td>
</tr>
</tbody>
</table>


**MOVEMENT DISORDERS**

**Disorders of Movement**

The patient is observed to be making unusual involuntary movements. Is it a tic, a tremor, chorea, athetosis, or some other involuntary movement? The recognition and classification of the movement disorder is essential for the establishment of a correct diagnosis.

*Athetosis* refers to a writhing, irregular movement associated with increased tone in the distal extremities. These movements are primarily around the long axis of the limb. Hyperextension of the digits is common. The movements are often continuous, with the amplitude increased by volition or excitement. It is usually the result of birth injury or kernicterus.

*Ballismus* refers to rapid movements occurring usually at the shoulder, but they may also be observed at the hip. They are irregular and consist of violent hurling, flinging, and throwing in the upper extremity and kicking or circumduction in the lower extremity. It is usually unilateral (hemiballismus). In the adult the lesion in the contralateral subthalamic nucleus is of vascular origin, while in children it represents a severe form of chorea.
Chorea, Greek for dance, may seem an incongruous term for these rapid, involuntary, nonrhythmic jerks of various parts of the body. They involve both proximal and distal portions of the limbs but may involve the face and trunk as well.

Dystonia refers to a movement disorder characterized by simultaneous contraction of agonist and antagonist muscles. The muscular contraction occurs prior to the onset of movement, leading to a tightening and stiffening of the affected parts of the anatomy. The end position, following a movement, is maintained for a prolonged period.

Myoclonus is an involuntary, repetitive, instantaneous, irregular contraction of a group of muscles, or more rarely, a single muscle.

Tremor is a rhythmic, oscillatory movement of a body part. It may be distinguished from myoclonus and tics by the regularity and the equal force and speed of the movement in both directions.

Tic, the most common movement disorder, consists of rapid stereotyped movements in areas about the face, neck, and shoulder that are usually directed away from the midline. They occur irregularly and last less than a second or may occur repetitively over several minutes. They are most obvious during excitement or emotional stress.

The table below summarizes the characteristic features of these movement disorders:

<table>
<thead>
<tr>
<th>Characteristics of Abnormal Movements</th>
</tr>
</thead>
<tbody>
<tr>
<td>MOVEMENT</td>
</tr>
<tr>
<td>Athetosis</td>
</tr>
<tr>
<td>Ballismus</td>
</tr>
<tr>
<td>Chorea</td>
</tr>
<tr>
<td>Dystonia</td>
</tr>
</tbody>
</table>

Table continued on next page.
Murphy’s Law

Characteristics of Abnormal Movements (Cont.)

<table>
<thead>
<tr>
<th>MOVEMENT</th>
<th>SPEED</th>
<th>LOCATION</th>
<th>DIRECTION</th>
<th>STEREOTYPE</th>
<th>RHYTHMICITY</th>
<th>INTERVAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myoclonus</td>
<td>Very rapid</td>
<td>Localized or generalized</td>
<td>Any</td>
<td>Stereotyped</td>
<td>Irregular</td>
<td>0.5 to 5 seconds</td>
</tr>
<tr>
<td>Tic</td>
<td>Rapid</td>
<td>Usually in area supplied by motor cranial nerve (face, shoulder, neck)</td>
<td>Rotational; away</td>
<td>Stereotyped</td>
<td>Irregular</td>
<td>1 second to minutes</td>
</tr>
<tr>
<td>Tremor</td>
<td>Variable</td>
<td>Usually localized, often in hand</td>
<td>Complex or simple</td>
<td>Extreme stereotype</td>
<td>Very rhythmic; may be irregular</td>
<td>0.1 to 1 second</td>
</tr>
</tbody>
</table>


MURPHY’S LAW (MEDICAL MURPHOLOGY)

Spitzer’s Laws of Neonatology (Abridged)

1. The more stable a baby appears to be, the more likely he will “crump” that day.
2. The distance that you have to go to a transport is directly proportional to the degree of illness of the baby.
3. The nicer the parents, the sicker the baby.
4. The incidence of neonatal problems increases dramatically if either parent is a physician or a nurse.
5. Endotracheal tubes are designed to fall out (become plugged, etc.) at the most critical moment.
6. The milder the RDS, the sooner the infant will find himself in 100% oxygen and maximal ventilatory support.
7. The longer a patient is discussed on rounds, the more certain it is that no one has the faintest idea what’s going on or what to do.
8. The sickest infant in the nursery can always be discerned by the fact that he is being cared for by the newest, most inexperienced nursing orientee.
9. The surest way to have an infant linger interminably is to inform the parents that death is imminent.
10. The probability of infection is directly proportional to the number of antibiotics that an infant is already receiving.
11. Lasix® (vitamin L) will squeeze urine out of bricks. Unfortunately, it doesn't always work as well in babies.
12. Antibiotics should always be continued for ____ days. (Fill in the blank with any number from 1 to 21.)
13. If you can’t figure out what’s going on with a baby, call the surgeons. They won’t figure it out either, but they’ll sure as hell do something about it.

Six Variations for Patients

1. Just because your doctor has a name for your condition doesn't mean he knows what it is.
2. The more boring and out-of-date the magazines in the waiting room, the longer you will have to wait for your scheduled appointment.
3. Only adults have difficulty with child-proof bottles.
4. You never have the right number of pills left on the last day of a prescription.
5. The pills to be taken with meals will be the least appetizing ones.

Corollary
Even water tastes bad when taken on doctor's orders.

6. If your condition seems to be getting better, it's probably your doctor getting sick.

Matz's Warning
Beware of the physician who is great at getting out of trouble.

Erma Bombeck's Rule
Never go to a doctor whose office plants have died.

Cochrane's Aphorism
Before ordering a test, decide what you will do if it is (1) positive or (2) negative. If both answers are the same, don't do the test.

Bernstein's Precept
The radiologist's national flower is the hedge.

Lord Cohen's Comment
The feasibility of an operation is not the best indication for its performance.

Telesco's Laws of Nursing

1. All the IVs are at the other end of the hall.
2. A physician's ability is inversely proportional to his availability.
3. There are two kinds of adhesive tape, that which won't stay on and that which won't come off.
4. Everybody wants a pain shot at the same time.
5. Everybody who didn't want a pain shot when you were passing out pain shots wants one when you are passing out sleeping pills.

"Why, yes... we do have two children who won't eat their vegetables."

THE FAR SIDE cartoon by Gary Larson is reprinted by permission of Chronicle Features, San Francisco, CA.
NEUROFIBROMATOSIS

Diagnosing Neurofibromatosis in Children Under 6

One of the most common single gene disorders is neurofibromatosis, which occurs in 1 of 4000 live births. There is virtually a complete dominant penetrance of the gene for von Recklinghausen's neurofibromatosis (neurofibromatosis-1) localized at the centromeric region of chromosome 17. Yet the diagnosis, particularly in young children, is difficult because there exists so much variation in gene expression with age. Indeed, without a positive family history—which only occurs in 50% of all the cases—the diagnosis is based solely upon clinical signs. The need to commit to memory these signs often occurs to a pediatric intern the morning after admitting a child with café-au-lait spots. Fortunately a recent National Institutes of Health Consensus Conference has delineated the diagnostic guidelines for neurofibromatosis-1 and -2.

Criteria for Diagnosis of Neurofibromatosis-1
(von Recklinghausen's Neurofibromatosis)

Two or more of the following criteria are required for diagnosis:

1. Six or more café-au-lait macules larger than 5 mm in greatest diameter in prepubertal individuals and larger than 15 mm in postpubertal individuals.
2. Two or more neurofibromas of any type, or one plexiform neurofibroma.
3. Freckling in the axillary or inguinal region.
4. Optic glioma.
5. Two or more Lisch nodules (pigmented hamartomas of the iris).
6. A distinctive osseous lesion, e.g., sphenoid dysplasia or thinning of the long bone cortex with or without pseudarthrosis.
7. A first-degree relative (parent, sibling, or offspring) with neurofibromatosis-1 according to the above criteria.

Criteria for Diagnosis of Neurofibromatosis-2
(Bilateral Acoustic or Central Neurofibromatosis)

1. Having a bilateral eighth nerve mass that can be seen with appropriate imaging techniques (e.g., computed tomography, magnetic resonance imaging).
2. Having a first degree relative with neurofibromatosis-2 and either a. an eighth nerve mass or b. two of the following:
   i. neurofibroma
   ii. meningioma
   iii. glioma
   iv. schwannoma
   v. juvenile posterior subcapsular lenticular opacity

Neurologic Signs of Infancy

A good pediatrician should know the time of appearance and the time of disappearance of the normal reflexes observed during infancy. If your patient displays alterations from the sequence described in the accompanying table, it should alert you to the possibility of neurologic dysfunction.

### Normal Reflexes Appearing in Infancy

<table>
<thead>
<tr>
<th>RESPONSE</th>
<th>AGE AT TIME OF APPEARANCE</th>
<th>AGE AT TIME OF DISAPPEARANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Reflexes of position and movement</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Moro reflex</td>
<td>Birth</td>
<td>1-3 months</td>
</tr>
<tr>
<td>Tonic neck reflex (unsustained)</td>
<td>Birth</td>
<td>5-6 months (partial up to 2-4 years)</td>
</tr>
<tr>
<td>Neck righting reflex</td>
<td>4-6 months</td>
<td>1-2 years</td>
</tr>
<tr>
<td>Landau response</td>
<td>3 months</td>
<td>1 2 years</td>
</tr>
<tr>
<td>Palmar grasp reflex</td>
<td>Birth</td>
<td>4 months</td>
</tr>
<tr>
<td>Adductor spread of knee jerk</td>
<td>Birth</td>
<td>7 months</td>
</tr>
<tr>
<td>Plantar grasp reflex</td>
<td>Birth</td>
<td>8-15 months</td>
</tr>
<tr>
<td>Babinski response</td>
<td>Birth</td>
<td>Variable</td>
</tr>
<tr>
<td>Parachute reaction</td>
<td>8-9 months</td>
<td>Variable</td>
</tr>
<tr>
<td><strong>Reflexes to sound</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Blinking response</td>
<td>Birth</td>
<td></td>
</tr>
<tr>
<td>Turning response</td>
<td>Birth</td>
<td></td>
</tr>
<tr>
<td><strong>Reflexes of vision</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Blinking to threat</td>
<td>6-7 months</td>
<td></td>
</tr>
<tr>
<td>Horizontal following</td>
<td>4-6 weeks</td>
<td></td>
</tr>
<tr>
<td>Vertical following</td>
<td>2 3 months</td>
<td></td>
</tr>
<tr>
<td>Optokinetic nystagmus</td>
<td>Birth</td>
<td></td>
</tr>
<tr>
<td>Postrotational nystagmus</td>
<td>Birth</td>
<td></td>
</tr>
<tr>
<td>Lid closure to light</td>
<td>Birth</td>
<td></td>
</tr>
<tr>
<td>Macular light reflex</td>
<td>4-8 months</td>
<td></td>
</tr>
<tr>
<td><strong>Food reflexes</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rooting response awake</td>
<td>Birth</td>
<td>3 4 months</td>
</tr>
<tr>
<td>Rooting response - asleep</td>
<td>Birth</td>
<td>7 8 months</td>
</tr>
<tr>
<td>Sucking response</td>
<td>Birth</td>
<td>12 months</td>
</tr>
<tr>
<td><strong>Handedness</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>2 3 years</td>
<td></td>
</tr>
<tr>
<td><strong>Spontaneous stepping</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Birth</td>
<td></td>
</tr>
<tr>
<td><strong>Straight line walking</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>5 6 years</td>
<td></td>
</tr>
</tbody>
</table>

NEUTROPENIA

What to Look for When the Pregnancy Is Complicated by Hypertension

The association between maternal hypertension and neutropenia of the newborn had been recognized for some time. What remained a mystery was the etiology, the mechanism of neutropenia, and whether any clinical consequences existed. A 1989 study from the University of Utah removed the shroud from some of the questions and advanced hypotheses regarding the etiology (Table 1).

Table 1. Apparent Risk Factors for Neutropenia in the Neonatal Period in Association with Maternal Hypertension

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>NEUTROPENIA</th>
<th>NO NEUTROPENIA</th>
<th>P VALUE*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intrauterine growth retardation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Premature birth</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Severe pregnancy-induced hypertension (BP &gt; 160/110, proteinuria &gt; 5 g/24 h)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Maternal HELLP syndrome (hemolysis, elevated liver enzymes, low platelets)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

The Utah study documented neutropenia (duration of 1 h to 30 d) in nearly 50% of the infants of mothers with maternal hypertension. Nosocomial infections occurred in 23% of those infants as opposed to 3% of healthy, non-neutropenic controls. The prevalence of neutropenia differed with respect to the type of hypertension (Table 2).

Table 2. Characteristics of Infants with Neonatal Neutropenia and Their Mothers

<table>
<thead>
<tr>
<th></th>
<th>NEUTROPENIA (N = 35)</th>
<th>NO NEUTROPENIA (N = 37)</th>
<th>P VALUE*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Infants</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Birth weight (g)*</td>
<td>1550 ± 770</td>
<td>2530 ± 880</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Gestational age (wk)*</td>
<td>31.5 ± 3.5</td>
<td>36.0 ± 3.7</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Intrauterine growth retardation (n = 12)</td>
<td>10</td>
<td>2</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Mothers</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age (yr)*</td>
<td>25.8 ± 4.7</td>
<td>24.4 ± 5.7</td>
<td>NS</td>
</tr>
<tr>
<td>Race</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nonwhite</td>
<td>4</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>White</td>
<td>29</td>
<td>28</td>
<td>NS vs. nonwhite</td>
</tr>
<tr>
<td>Hypertension</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pregnancy-induced</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mild</td>
<td>5</td>
<td>18</td>
<td></td>
</tr>
<tr>
<td>Severe</td>
<td>13</td>
<td>5</td>
<td>&lt;0.002 vs. mild</td>
</tr>
<tr>
<td>HELLP syndrome</td>
<td>8</td>
<td>3</td>
<td>&lt;0.01 vs. mild</td>
</tr>
</tbody>
</table>

*The comparisons of birth weight, gestational age, maternal age, and the interval from membrane rupture to delivery were made according to Student's t-test; the other comparisons were made with Fisher's exact test. NS denotes not significant.

† Mean ± SD.

Table continued on next page.
Kinetic studies performed on cord blood of the neutropenic infants revealed diminished neutrophil production as opposed to accelerated destruction or excessive margination. The authors proposed two hypotheses to explain the diminished production: (1) deficiency of neutrophil-specific growth factors or (2) inhibition of neutrophil differentiation. The molecular mechanism remains unknown. Until the mechanism is elucidated, therapy rests in recognition of the phenomenon and prophylactic antibiotic use as indicated.


**NORMOBLASTEMIA**

The Cause of Nucleated Red Blood Cells in the Peripheral Blood in Children (Normoblastemia)

*Childhood Diseases Associated with Normoblastemia*

<table>
<thead>
<tr>
<th>1. Hematologic/Oncologic</th>
<th>Myelofibrosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Severe anemia of any cause</td>
<td>Preleukemia</td>
</tr>
<tr>
<td>Hemolytic anemias</td>
<td>Leukemia</td>
</tr>
<tr>
<td>Iron deficiency</td>
<td>Lymphoma</td>
</tr>
<tr>
<td>Blood loss</td>
<td>Myeloproliferative disorders</td>
</tr>
<tr>
<td>Megaloblastic anemia</td>
<td>Solid tumor invasion of bone marrow</td>
</tr>
<tr>
<td>Histiocytosis</td>
<td></td>
</tr>
</tbody>
</table>

*Table continued on next page.*
Childhood Diseases Associated with Normoblastemia (Cont.)

2. Infections
   - Bacterial infection (especially sepsis)
   - Tuberculosis
   - Osteomyelitis
   - Fungal

3. Hypoxia
   - Congestive heart failure
   - Cyanotic heart disease
   - Asthma and other respiratory disease

4. Other
   - Collagen vascular diseases
   - Sarcoïdosis
   - Inflammatory bowel disease
   - Osteopetrosis
   - Gaucher's and other storage diseases
   - Diabetic ketoacidosis
   - Thermal injury
   - Vinca alkaloids
   - Asplenia
   - Newborn (physiologic)
   - Uremia
   - Normal finding

From this long list the most common disorders include cardiac disease, hemolytic disorders, pulmonary disease, and bone marrow replacement.


NURSEMAID'S ELBOW

Reducing Nursemaid's Elbow to Simple Terms

Subluxation or partial dislocation of the head of the radius is affectionately termed "nursemaid's elbow," because it typically arises subsequent to a sudden jerk or pull of a toddler's arm. Such a maneuver can be seen at any park, playground, or shopping mall on an hourly basis! More specifically, 90% of all cases of nursemaid's elbow are due to the sudden longitudinal pull or traction at the wrist when the elbow is fully extended and the forearm is pronated. It is typically seen in toddlers between the ages of 1 and 5 years, with a peak incidence among children aged 15 to 30 months.

The child with nursemaid's elbow also tends to hold the arm slightly flexed at the elbow and slightly pronated in order to avoid pain. Typically, the child who has incurred such an injury will refuse to move the affected arm and complains vociferously and painfully at any such attempt to manipulate the elbow, particularly in supination and pronation.

The Anatomy of Nursemaid's Elbow

Dislocation of the head of the radius is best understood by reviewing its anatomy (see figure). The radial head is wrapped by a cuff-like annular ligament. The annular ligament attaches the radius to the ulna but also allows rotary motion of the radial head. These ligamentous fibers combine with other ligaments of the elbow at the radiohumoral joint. Sudden longitudinal traction, when applied to a toddler's pronated forearm, stretches and tears the annular ligament at its distal attachment on the radial neck. With continued traction, the annular ligament slips over the radial head and, once the traction is released, these fibers can become caught between the articular surface of the radial head and the capitellum. The result is pain.
The annular ligament covers the radial head and attaches the radius to the ulna. With sudden longitudinal traction, the ligament stretches and tears. Fibers of the ligament are then caught between the radial head and the capitellum.

**A Prescription for Reducing Nursemaid's Elbow to a Mere Memory**

X-rays are rarely indicated in this type of injury. Instead, following the maneuvers illustrated in the figure below should correct the problem quickly and simply.

To reduce the injury, hold the elbow slightly flexed. Apply pressure over the radial head with the thumb, then hold the child's wrist with the other hand and quickly move the forearm to either a supine or pronated position.

NUTRITION

Infant Foods—Calories and Their Distribution

When the infant is ready for strained or junior foods, it is important to be aware of the number of calories being provided and their source. The accompanying table lists estimated calories derived from analysis of a variety of products in each category.

### Strained Foods

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>KCAL, 100 GM</th>
<th>PERCENTAGE OF CALORIES</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Protein</td>
</tr>
<tr>
<td>Juices</td>
<td>65 (45-98)</td>
<td>2</td>
</tr>
<tr>
<td>Fruits</td>
<td>85 (79-125)</td>
<td>2</td>
</tr>
<tr>
<td>Vegetables</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Plain</td>
<td>45 (27-28)</td>
<td>14</td>
</tr>
<tr>
<td>Creamed</td>
<td>63 (42-94)</td>
<td>13</td>
</tr>
<tr>
<td>Meats</td>
<td>106 (86-194)</td>
<td>53</td>
</tr>
<tr>
<td>Egg yolks</td>
<td>192 (184-199)</td>
<td>21</td>
</tr>
<tr>
<td>High meat dinner</td>
<td>84 (63-106)</td>
<td>29</td>
</tr>
<tr>
<td>Desserts</td>
<td>96 (71-136)</td>
<td>4</td>
</tr>
<tr>
<td>Cereal</td>
<td>360 (349-393)</td>
<td>39</td>
</tr>
<tr>
<td>Cereal-fruit</td>
<td>85 (76-98)</td>
<td>18</td>
</tr>
</tbody>
</table>

### Junior Foods

<table>
<thead>
<tr>
<th>CATEGORY</th>
<th>KCAL, 100 GM</th>
<th>PERCENTAGE OF CALORIES</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Protein</td>
</tr>
<tr>
<td>Fruits</td>
<td>85 (69-116)</td>
<td>2</td>
</tr>
<tr>
<td>Vegetables</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Plain</td>
<td>46 (27-71)</td>
<td>12</td>
</tr>
<tr>
<td>Creamed</td>
<td>64 (45-72)</td>
<td>13</td>
</tr>
<tr>
<td>Meats</td>
<td>103 (88-135)</td>
<td>56</td>
</tr>
<tr>
<td>Soup dinner</td>
<td>61 (39-100)</td>
<td>15</td>
</tr>
</tbody>
</table>


Are We Eating the “Wrong” Fruits & Vegetables? (Or What Could Be More Nutritious Than a Fresh Orange?)

Eighteen common fruits and vegetables are listed here, first in order of their nutrient density and a second in order of their total nutrient contribution to the U.S. diet (density times tonnage). Our diets would improve considerably if we ate more from the top of the lefthand list than from the bottom. There is nothing “wrong” with lettuce and oranges, of course, but notice how far down they are in nutritional value. Note too how much more nutritious vegetables are than fruits.
### NUTRIENT CONTRIBUTION OF CONCENTRATION NUTRIENTS TO DIET

<table>
<thead>
<tr>
<th>CROP</th>
<th>RANK</th>
<th>CROP</th>
<th>RANK</th>
</tr>
</thead>
<tbody>
<tr>
<td>BROCCOLI</td>
<td>1</td>
<td>TOMATOES</td>
<td>1</td>
</tr>
<tr>
<td>SPINACH</td>
<td>2</td>
<td>ORANGES</td>
<td>2</td>
</tr>
<tr>
<td>BRUSSELS SPROUTS</td>
<td>3</td>
<td>POTATOES</td>
<td>3</td>
</tr>
<tr>
<td>LIMA BEANS</td>
<td>4</td>
<td>LETTUCE</td>
<td>4</td>
</tr>
<tr>
<td>PEAS</td>
<td>5</td>
<td>SWEET CORN</td>
<td>5</td>
</tr>
<tr>
<td>ASPARAGUS</td>
<td>6</td>
<td>BANANAS</td>
<td>6</td>
</tr>
<tr>
<td>ARTICHOokes</td>
<td>7</td>
<td>CARROTS</td>
<td>7</td>
</tr>
<tr>
<td>CAULIFLOWER</td>
<td>8</td>
<td>CABBAGE</td>
<td>8</td>
</tr>
<tr>
<td>SWEET POTATOES</td>
<td>9</td>
<td>ONIONS</td>
<td>9</td>
</tr>
<tr>
<td>CARROTS</td>
<td>10</td>
<td>SWEET POTATOES</td>
<td>10</td>
</tr>
<tr>
<td>SWEET CORN</td>
<td>12</td>
<td>PEAS</td>
<td>15</td>
</tr>
<tr>
<td>POTATOES</td>
<td>14</td>
<td>SPINACH</td>
<td>18</td>
</tr>
<tr>
<td>CABBAGE</td>
<td>15</td>
<td>BROCCOLI</td>
<td>21</td>
</tr>
<tr>
<td>TOMATOES</td>
<td>16</td>
<td>LIMA BEANS</td>
<td>23</td>
</tr>
<tr>
<td>BANANAS</td>
<td>18</td>
<td>ASPARAGUS</td>
<td>25</td>
</tr>
<tr>
<td>LETTUCE</td>
<td>26</td>
<td>CAULIFLOWER</td>
<td>30</td>
</tr>
<tr>
<td>ONIONS</td>
<td>31</td>
<td>BRUSSELS SPROUTS</td>
<td>34</td>
</tr>
<tr>
<td>ORANGES</td>
<td>33</td>
<td>ARTICHOokes</td>
<td>36</td>
</tr>
</tbody>
</table>

### The "Skinniest" Cuts of Beef

For those patients who have been placed on a low fat, low cholesterol diet but still insist "real people eat beef," here are some of the "skinniest" cuts of beef you can recommend:

#### Lean Beef

<table>
<thead>
<tr>
<th>CUT OF BEEF</th>
<th>FYI OF ROUND</th>
<th>TOP LOIN</th>
<th>ROUND TIP</th>
<th>ROUND TIP</th>
<th>FENDERLOIN</th>
<th>TOP SIRLOIN</th>
</tr>
</thead>
<tbody>
<tr>
<td>Calories</td>
<td>143 cal</td>
<td>176 cal</td>
<td>157 cal</td>
<td>153 cal</td>
<td>179 cal</td>
<td>165 cal</td>
</tr>
<tr>
<td>Total Fat</td>
<td>4.2 g</td>
<td>8.0 g</td>
<td>5.9 g</td>
<td>4.2 g</td>
<td>8.5 g</td>
<td>6.1 g</td>
</tr>
<tr>
<td>Saturated Fat</td>
<td>1.5 g</td>
<td>3.1 g</td>
<td>2.1 g</td>
<td>1.4 g</td>
<td>3.2 g</td>
<td>2.4 g</td>
</tr>
<tr>
<td>Cholesterol</td>
<td>59 mg</td>
<td>65 mg</td>
<td>69 mg</td>
<td>72 mg</td>
<td>72 mg</td>
<td>76 mg</td>
</tr>
</tbody>
</table>

*Figures for a cooked and trimmed 3 oz serving; 4 oz uncooked beef yields a 3 oz cooked portion.*


OBSESSIVE-COMPULSIVE DISORDER

Step on a Crack and You’ll Break Your Mother’s Back

Obsessive-compulsive disorder is a significant disturbance of childhood that has not been well studied until recent years. The disorder appears to occur with greater frequency than previously thought, usually in adolescence, and it is more common in boys than girls by at least 2 to 1. It has a presentation very similar to adult OCD. Common obsessional thoughts concern contamination (e.g., feces, dirt, disease) and fears of wrongdoing; common compulsions are hand-washing rituals, grooming, and checking rituals.

Major Presenting Symptoms in 70 Consecutive Children and Adolescents with Severe Primary Obsessive-Compulsive Disorder

<table>
<thead>
<tr>
<th>REPORTED SYMPTOM AT INITIAL INTERVIEW</th>
<th>NO. (%) OF PATIENTS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excessive or ritualized hand washing, showering, bathing, tooth brushing, or grooming</td>
<td>60 (85)</td>
</tr>
<tr>
<td>Repeating rituals (e.g., going in/out door, up/down from chair)</td>
<td>35 (51)</td>
</tr>
<tr>
<td>Checking (doors, locks, stove, appliances, emergency brake on car, paper route, homework, etc.)</td>
<td>32 (46)</td>
</tr>
<tr>
<td>Rituals to remove contact with contaminants</td>
<td>16 (23)</td>
</tr>
<tr>
<td>Touching</td>
<td>14 (20)</td>
</tr>
<tr>
<td>Measures to prevent harm to self or others</td>
<td>11 (16)</td>
</tr>
<tr>
<td>Ordering/arranging</td>
<td>12 (17)</td>
</tr>
<tr>
<td>Counting</td>
<td>13 (18)</td>
</tr>
<tr>
<td>Hoarding/collecting rituals</td>
<td>8 (11)</td>
</tr>
<tr>
<td>Rituals of cleaning household or inanimate objects</td>
<td>4 (6)</td>
</tr>
<tr>
<td>Miscellaneous rituals (e.g., writing, moving, speaking)</td>
<td>18 (26)</td>
</tr>
<tr>
<td>Concern with dirt, germs, or environmental toxins</td>
<td>28 (40)</td>
</tr>
<tr>
<td>Something terrible happening (fire/death/illness of self or loved one, etc.)</td>
<td>17 (24)</td>
</tr>
<tr>
<td>Symmetry, order, or exactness</td>
<td>12 (17)</td>
</tr>
<tr>
<td>Scrupulosity (religious obsessions)</td>
<td>9 (13)</td>
</tr>
<tr>
<td>Concern or disgust with bodily wastes or secretions (urine, stool, saliva)</td>
<td>6 (8)</td>
</tr>
<tr>
<td>Lucky/unlucky numbers</td>
<td>6 (8)</td>
</tr>
</tbody>
</table>

*Obsessions or compulsions are totaled, so the total exceeds 70.

Table continued on next page.
Occam’s Razor/Odors of Disease

Major Presenting Symptoms in 70 Consecutive Children and Adolescents with Severe Primary Obsessive-Compulsive Disorder (Cont.)

<table>
<thead>
<tr>
<th>COMPULSIONS</th>
<th>REPORTED SYMPTOM AT INITIAL INTERVIEW NO. (%) OF PATIENTS*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Forbidden, aggressive, or perverse sexual thoughts, images, or impulses</td>
<td>3 (4)</td>
</tr>
<tr>
<td>Fear might harm others/self</td>
<td>3 (4)</td>
</tr>
<tr>
<td>Concern with household items</td>
<td>2 (3)</td>
</tr>
<tr>
<td>Intrusive nonsense sounds, words, or music</td>
<td>1 (1)</td>
</tr>
</tbody>
</table>


OCCAM’S RAZOR

A Diagnostic Principle—Occam’s Razor

Without knowing it many clinicians apply Occam’s razor to their diagnostic thinking. Occam’s razor is a logical principle attributed to William of Occam, although it was used by some scholastic philosophers prior to him. The principle states that a person should not increase, beyond what is necessary, the number of entities required to explain anything, or that the person should not make more assumptions than the minimum needed. This principle is often called the Law of Parsimony. Since the Middle Ages it has played an important role in eliminating unnecessary elements from explanations. Remember William of Occam when you attempt to explain multiple symptoms in your patient with a single diagnosis.

ODORS OF DISEASE

Unusual Odor as a Clue to Diagnosis

Can you smell a rat or sniff out a diagnosis? The sense of smell is not used enough as part of the physical examination. Listed below are diseases associated with unusual odors.

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>ENZYME DEFECT</th>
<th>ODOR</th>
<th>CLINICAL FEATURES</th>
<th>TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diabetes mellitus</td>
<td>Lack of insulin or insulin activity</td>
<td>Acetone on breath, fruity</td>
<td>Polyuria, polyphagia, polydipsia, weight loss, acidoses, coma</td>
<td>Insulin administration</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Diseases Associated with Unusual Odors (Cont.)

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>ENZYME DEFECT</th>
<th>ODOR</th>
<th>CLINICAL FEATURES</th>
<th>TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Phenylketonuria</td>
<td>Phenylalanine hydroxylase</td>
<td>Musty, &quot;mousy,&quot; &quot;horsey&quot;</td>
<td>Progressive mental retardation, eczema, decreased pigmentation, seizures, spasticity</td>
<td>Diet low in phenylalanine</td>
</tr>
<tr>
<td>Maple syrup urine disease</td>
<td>Branched chain dehydroxylase</td>
<td>Maple syrup</td>
<td>Marked acidosis, seizures, coma leading to death in first year or two of life or mental subnormality without acidosis or intermittent acidosis without mental retardation</td>
<td>Diet low in branched chain amino acids; protein restriction and/or thiamine in large doses</td>
</tr>
<tr>
<td>Oasthouse urine disease</td>
<td>Defective transport of methionine, branched chain amino acids, tyrosine, and phenylalanine</td>
<td>Yeast-like; dried-celery-like</td>
<td>Mental retardation, spasticity, hyperpnea, fever, edema</td>
<td>Restrict methionine in diet</td>
</tr>
<tr>
<td>Odor of sweaty feet, Syndrome I</td>
<td>Isovaleryl CoA dehydrogenase</td>
<td>Sweaty feet</td>
<td>Recurrent bouts of acidosis, vomiting, dehydration, coma, aversion to protein foods</td>
<td>Restrict leucine in diet</td>
</tr>
<tr>
<td>Odor of sweaty feet, Syndrome II</td>
<td>Green acyldehydrogenase</td>
<td>Sweaty feet</td>
<td>Onset of symptoms in first week of life with acidosis, dehydration, seizures, and death</td>
<td>High CHO diet (?) Low fat diet (?)</td>
</tr>
<tr>
<td>Odor of cats syndrome</td>
<td>Beta-methylcrotonyl-CoA carboxylase</td>
<td>Cat's urine</td>
<td>Neurologic disorder resembling Werdnig-Hoffmann disease, ketoacidosis, failure to thrive</td>
<td>Leucine restriction (?) Biotin administration</td>
</tr>
<tr>
<td>Fish odor syndrome</td>
<td>Unknown</td>
<td>Like dead fish</td>
<td>Stigmata of Turner's syndrome, neutropenia, recurrent infections, anemia, splenomegaly</td>
<td>Unknown</td>
</tr>
<tr>
<td>Fish odor syndrome</td>
<td>Trimethylamine oxidase</td>
<td>Like dead fish</td>
<td>Unusual odor of sweat, skin and urine. Normal development</td>
<td>Elimination of fish from the diet</td>
</tr>
<tr>
<td>Odor of rancid butter syndrome</td>
<td>Unknown</td>
<td>Rancid butter</td>
<td>Poor feeding, irritability, progressive neurologic deterioration with seizures and death; hepatic dysfunction; possibly same as acute tyrosinosis</td>
<td>Response to decreased phenylalanine and tyrosine intake (?)</td>
</tr>
</tbody>
</table>

Serum Osmolality

It is often important to estimate serum osmolality before the laboratory measurement becomes available. The following formula will make that estimation more accurate.

The short cut approach is

\[
\text{Serum osmolality} = [\text{Na(mEq/L)} = K \text{ (mEq/L)}] \times 2 + \frac{\text{Glucose}}{18} + \frac{\text{BUN}}{3}
\]

The normal value is 280.

I know a baby
Who smells like fresh muffins
Wrapped in warm linen
Just dried by the breezes
Blown over the lilacs
Brought out by the spring sun
And back from the oceans
With Orient spices
The Precordial Catch Syndrome

In 1955 Miller and Texidor first described an entity in young adults they termed "precordial catch." It has proven to be a common entity. Perhaps as many as 50% of older adolescents and young adults will experience this sensation of a sudden, brief, nonradiating, periapical pain that is unrelated to exercise or exertion. Both patients and their parents are naturally concerned about heart disease, but you can reassure them that the pain is of no cardiac significance. The precise cause of this painful sensation of "something being caught" and being forced to "freeze" in place is still unknown. The characteristics are described in the list below.

The Pain Itself

Onset: Sudden, unexpected, unprovoked.
Location: Left lower anterior aspect of chest; typically infra-apical at the sternal border.
Duration: Brief (<3 minutes, usually 1 minute or less).
Description: Variable but superficial, knife- or needle-like, burning, stabbing, shooting, sharp, something catches.
Localization: Site often localized by patient using one or more fingers.
Radiation: Nonradiating.

Related Factors

Respiration: Taking a deep breath accentuates pain and makes patient "freeze." Forced inspiration, if possible, relieves pain.
Exertion: Unrelated to strenuous activity; usually occurs at rest.
Posture: Pain sometimes occurs when patient bends over or is slouched. Pain is relieved by stretching and straightening if possible.


PALSY

Neonatal Phrenic Nerve Palsy—The "Belly Dancer's Sign"

Unilateral diaphragmatic paralysis with or without brachial plexus injury may present in neonates as "respiratory distress." The chest roentgenogram may be
Pancreatitis

misleading unless obtained in deep inspiration. Fluoroscopy is required to demonstrate paradoxical motion of the diaphragm on the involved side.

It should be remembered that the existence of diaphragmatic paralysis can be recognized by merely observing the movement of the umbilicus during the respiratory cycle. To perform this maneuver, note the position of the umbilicus at full expiration. Mark this position by placing your pen at the spot. During inspiration, the umbilicus can be seen to shift upward and toward the side of the paralyzed diaphragm. Other suggestive physical findings include unexplained tachypnea without dyspnea, slightly decreased breath sounds on the paralyzed side, fine inspiratory rales on the paralyzed side if atelectasis is present, widening of the subcostal angle on the affected side during inspiration, and flattening of the epigastrum on the side of the paralyzed diaphragm during inspiration. The movement of the umbilicus is the sign most easily identified.


PANCREATITIS

Acute Pancreatitis in Children

Pancreatitis is an acknowledged but infrequently recognized cause of abdominal pain in children. The diagnosis is sometimes difficult. The following clinical description may help.

Etiology

<table>
<thead>
<tr>
<th>Drugs/toxins</th>
<th>Infection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thiazides</td>
<td>Mumps (even in the absence of parotitis)</td>
</tr>
<tr>
<td>Steroids</td>
<td>Hepatitis B virus</td>
</tr>
<tr>
<td>Azathioprine</td>
<td>Coxsackie B5</td>
</tr>
<tr>
<td>Alcohol</td>
<td>Epstein-Barr virus</td>
</tr>
<tr>
<td>Tetracycline</td>
<td>Mycoplasma</td>
</tr>
<tr>
<td>Salicylazosulfapyridine</td>
<td>Influenza B</td>
</tr>
<tr>
<td>Chlorthalidone</td>
<td>Diabetes mellitus (ketoacidosis)</td>
</tr>
<tr>
<td>Furosemide</td>
<td>Parotitis</td>
</tr>
<tr>
<td>L-asparaginase</td>
<td>Hyperparathyroidism</td>
</tr>
<tr>
<td>Oral contraceptives</td>
<td>Septic shock</td>
</tr>
<tr>
<td>Trauma/surgery/child abuse</td>
<td>Cystic fibrosis</td>
</tr>
<tr>
<td>Biliary tract disease</td>
<td>Pregnancy</td>
</tr>
<tr>
<td>Choledochal cyst</td>
<td>Acute porphyria</td>
</tr>
<tr>
<td>Stricture of the common bile duct</td>
<td>Kwashiorkor</td>
</tr>
<tr>
<td>Congenital stenosis of the ampulla</td>
<td>Hyperlipoproteinemia I and V</td>
</tr>
<tr>
<td>of Vater</td>
<td>Scorpion bites</td>
</tr>
<tr>
<td>Anomalous insertion of the common</td>
<td>Idiopathic</td>
</tr>
<tr>
<td>bile duct</td>
<td></td>
</tr>
<tr>
<td>Cholelithiasis/cholecystitis</td>
<td></td>
</tr>
</tbody>
</table>
Pancreatitis—251

Signs and Symptoms

1. Abdominal pain. Children may not localize the pain very well. It is usually noted to be in the upper quadrants or the periumbilical area. The pain is usually constant, but it may be intermittent, and it may be made worse by eating. The knee-chest position will usually relieve the pain.

2. Vomiting. Vomiting is aggravated by eating or drinking. It does not relieve the pain.

3. Abdominal tenderness. Tenderness may be accompanied by guarding and rebound. Maximal tenderness is usually in the midepigastric region. Bowel sounds may be normal, hypoactive, or absent.

4. Fever.

5. Upper gastrointestinal hemorrhage. The hemorrhage is thought to result from stress and may originate in the stomach, duodenum, or be caused by penetration of an ulcer into the head of the pancreas.

Laboratory Evaluation

1. Elevated bilirubin. This may be due to a stone in the common duct or to edema in the head of the pancreas.

2. X-ray changes. X-rays may document pleural effusion (most commonly on the left side) and/or ascites. There may also be a dilated segment of small bowel adjacent to the inflamed pancreas (sentinel loop). Isolated gaseous distention of the ascending colon and hepatic flexure may be present (colon cutoff sign). A CT of the pancreas often reveals the presence of a boggy, swollen organ.

3. Hyperglycemia. Diabetes mellitus may or may not follow pancreatitis.

4. Hypocalcemia.

5. Elevated serum amylase. The serum amylase usually begins to rise within hours of the onset of symptoms. It usually peaks within the first 24 hours of illness and returns to normal within 48 to 72 hours. Daily amylase determinations are helpful in following patients. If the amylase remains elevated for over two weeks, a pseudocyst should be suspected. Amylase values may be normal in patients with acute hemorrhagic pancreatitis.

6. Elevated serum lipase. These values tend to follow those of the serum amylase.

7. Elevated urinary diastase. Timed urine collections are necessary for this determination.

8. Amylase clearance test. Amylase clearance may be elevated in patients with severe burns or diabetic ketoacidosis, as well as in those with pancreatitis. It is calculated from the following formula:

\[
\frac{\text{Cam}}{\text{Ccr}} = \frac{\text{Amylase (urine)}}{\text{Amylase (serum)}} \times \frac{\text{Creatinine (serum)}}{\text{Creatinine (urine)}} \times 100
\]

Treatment

1. Relief of pain. This is best accomplished with meperidine given every three hours. Its effect may be potentiated by promethazine.

2. Reduction of exocrine pancreatic secretion. The patient should fast, and intravenous fluids should be supplied. If intravenous fluids are required for more than five days, parenteral alimentation should be initiated. A nasogastric tube should be placed if the patient is nauseated or vomiting, or has an ileus.
When oral feedings are initiated, they should consist of carbohydrates alone initially, because they cause the least stimulation to the pancreas. Feedings should be restarted when abdominal tenderness has disappeared, any ileus has resolved, and urinary diastase or amylase clearance has become normal.

3. Treatment of shock and electrolyte abnormalities.

Although anticholinergic drugs and antibiotics have been used in the treatment of pancreatitis, their use has not improved the prognosis. Mortality may range from about 20% with acute interstitial pancreatitis to about 80% with hemorrhagic pancreatitis.


**PANNICULITIS**

What Do Popsicles and Horses Have in Common?

Both are associated with forms of cold-exposure panniculitis, characterized by single or multiple crops of tender nodules in the subcutaneous fat. Blood vessels also are usually affected, resulting in a histologic picture of fat-cell necrosis. The nodules can be of a size less than 1 cm to over 10 cm across. The clinical picture is one of reddish-purple discoloration and erythematous, enlarging nodules that are often painful to palpation. The lesions are most obvious 24 to 48 hours after the cold injury. They are commonly confused with a cellulitis. The patients are afebrile and feel well, and the lesions subside without treatment in 2 to 3 weeks, leaving no permanent injury.

The popsicle form of panniculitis is produced by sucking on cold objects, such as popsicles and ice cubes, or the lengthy application of the popsicle to any area of the skin.

In equestrian cold panniculitis, the lesions appear on the outer thighs as a result of prolonged horseback riding in freezing weather.

**PARASITES**

Sushi Eaters Beware!

H. L. Mencken used his father's method of separating the world's population into two groups: those who pay their bills and those who do not. An equally all-encompassing method might be those who eat sushi (the Japanese delicacy of raw fish) and those who don't. Recently, however, a report in the New England Journal of Medicine appeared that might diminish the legions of raw fish eaters. The report noted a patient who presented with mild abdominal distension, direct and rebound tenderness in the right lower quadrant, and an elevated white blood cell count. After 6 hours observation, the patient's right lower quadrant tenderness worsened, and she was taken to the operating room for emergency appendectomy. At operation the appendix appeared grossly normal and a pinkish-red, sinuous worm was found moving onto the surgical drapes just prior
to surgical closure of the wound. The worm was identified as an early 4th stage larva of the genus *Eustrongylides*, a nematode parasite of fish-eating birds that is frequently found in raw fish. The patient's medical history was remarkable for eating sushi, prepared at her friend's house, the day before.

This case only adds to the growing list of parasitic diseases that can be acquired by ingesting infected fish that is raw or insufficiently cooked, smoked, salted, or marinated. The clinical presentation varies depending upon where the parasite has localized (e.g., the stomach or the intestines). Dominating features of these parasitic infections include acute abdominal symptoms such as discomfort, guarding, nausea, severe epigastric pain, and rebound tenderness. An elevated eosinophil count (30 to 40%) is also suspicious for a parasitic infection.

There are a number of parasites that have been reported to be indigenous to many marine fish typically caught off the coasts of the U.S., Japan, and Europe (e.g., salmon, cod, whiting, herring, and haddock). They include:

- *Anisakis simplex*, the most common parasitic disease of sushi eaters.
- *Pseudoterranova* (formerly *Phocanema*) *decipiens*
- *Contracecum* species
- *Heterophyes heterophyes*
- *Diphyllobothrium latum* (broad or fish tapeworm)
- *Nanophyetus salmincola*
- *Eustrongylides*
- *Gambierdiscus toxicus* (ciguatera fish poisoning)

In order to avoid these parasitic infections, the CDC in Atlanta suggests that you cook seafood before consuming it; heating fish to 65°C for 10 minutes appears to kill most worms. Freezing fish for a minimum of 5 days at 20°C (4°F) also kills most parasitic species. Other methods of preparing raw fish, such as brining or marinating, cannot be relied upon to destroy helminths. Visual inspection and candling (holding the fish up to light) seem to be most reliable in the hands of an experienced sushi chef.

Other fish-related diseases include scombroid (histamine fish poisoning) and a number of viral and bacterial infections from raw shellfish ingestion.


**PARVOVIRUS**

**Beyond Fifth Disease: The Clinical Spectrum of Parvovirus B19**

Once the viral etiology of erythema infectiosum, or fifth disease, was determined to be human parvovirus B19, the true clinical spectrum of B19 infection could be investigated. We now know that B19 infection in healthy children can occur without the usual facial (“slapped cheek”) rash or subsequent reticular rash on the extremities and trunk. We also know that B19, when it
Pelvic Inflammatory Disease

Pelvic inflammatory disease (PID) is a serious problem among sexually active adolescent women. Over 1,000,000 women of reproductive age contract PID each year; teenagers make up 20% of these cases. The incidence of PID among adolescents rises each year as more and more teenagers become sexually active and engage in unprotected sexual intercourse. The short-term complications of PID include perihepatitis (Fitz-Hugh–Curtis syndrome) and tubo-ovarian abscesses, while long-term sequelae include an increased incidence of infertility and ectopic pregnancies. For example, women with a history of one or more episodes of PID were found to have an involuntary infertility rate of 21% (compared to 3% in controls) and a sixfold increase in ectopic pregnancies even with antimicrobial intervention. In light of these complications the successful PID intervention that minimizes the patient’s risk must include (1) early recognition, (2) the use of broad spectrum antibiotics to treat the polymicrobial nature of the disease, (3) an emphasis on careful clinical reevaluation of the suspected PID patient within 24 hours to detect antibiotic failure or a misdiagnosis, and (4) evaluation and treatment of the patient’s sexual partners.
Unfortunately, the most frequently occurring symptoms of PID lack specificity in their delineation from other disease processes involving the reproductive, urinary, and gastrointestinal tracts, as depicted in the table below:

### Common Clinical Symptoms of PID by Organ System

<table>
<thead>
<tr>
<th>SYMPTOM</th>
<th>REPRODUCTIVE</th>
<th>URINARY</th>
<th>GASTROINTESTINAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower abdominal pain</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Urinary symptoms</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Intermenstrual bleeding</td>
<td>+</td>
<td>±</td>
<td>±</td>
</tr>
<tr>
<td>Dysmenorrhea</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Secondary amenorrhea</td>
<td>+</td>
<td>±</td>
<td>±</td>
</tr>
<tr>
<td>Nausea/vomiting</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Fever</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Malaise</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Dyspareunia</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
</tbody>
</table>

The history alone will lead the physician potentially to misdiagnose many women with acute PID, and this can put them at great risk for the short-term and long-term sequelae. Although laparoscopy is the definitive means of determining the presence of acute PID, it is not feasible to perform the procedure on every adolescent with lower abdominal pain because of its risks, costs, and the availability of manpower. The performance of a careful pelvic examination, however, in context with the patient's history and chief complaints, can be useful in at least separating reproductive tract disease from urinary and gastrointestinal complaints.

### Common Clinical Signs in Acute Lower Abdominal Pain Among Adolescent Females by Organ System

<table>
<thead>
<tr>
<th>SYMPTOM</th>
<th>REPRODUCTIVE</th>
<th>URINARY</th>
<th>GASTROINTESTINAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lower abdominal tenderness</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Perineal rash</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Vaginal discharge</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Cervical mucopus</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Uterine bleeding</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Cervical motion tenderness</td>
<td>+</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>Uterine tenderness or mass</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Adnexal tenderness or mass</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>


### The Criteria for the Diagnosis of Acute PID

Given the limitations of history-taking, the pelvic examination, and laparoscopy for PID, the following clinical diagnostic criteria are offered:

All three should be present:

1. Lower abdominal tenderness
2. Cervical motion tenderness
3. Adnexal tenderness (may be unilateral), plus (see following list)
One of the following should be present:

1. Temperature $\geq 38^\circ C$
2. White blood cell count $\geq 10,500/mm^3$
3. Purulent material obtained by culdocentesis
4. Inflammatory mass present on bimanual exam ± sonogram
5. ESR $> 15$ mm/hr
6. Evidence of *N. gonorrhoeae* and/or *C. trachomatis* in the endocervix.
   a. Gram stain with gram-negative diplococci
   b. Monoclonal antibody for *C. trachomatis*
7. $> 5$ white blood cells per oil-immersion field on Gram's stain of endocervical discharge.


**Whooping Cough and the White Blood Cell Count**

The white blood cell count can often be helpful in diagnosing pertussis. A marked leukocytosis (WBC $> 25,000/mm^3$) with a differential demonstrating the presence of 50 to 90% lymphocytes is usually considered presumptive evidence of pertussis in infants and children with a cough. (Definitive diagnosis is made by recovery of *B. pertussis*, *B. parapertussis*, or *B. bronchiseptica* from a Dacron or calcium alginate nasopharyngeal swab.)

Despite the availability of vaccines, pertussis is unfortunately still very much with us. It is not generally appreciated that infants under 6 months of age often do not display the aforementioned degree of leukocytosis and that the white cell count may be normal during the prodromal phase of the illness.

The table below illustrates the range of white cell counts by age in patients with pertussis.

<table>
<thead>
<tr>
<th>Total WBC</th>
<th>0 to 6 Months</th>
<th>6 Months to 2 Years</th>
<th>2 to 5 Years</th>
<th>5 Years</th>
<th>Total Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>5,000 to 15,000</td>
<td>38%</td>
<td>6%</td>
<td>14%</td>
<td>31%</td>
<td>23%</td>
</tr>
<tr>
<td>15,000 to 25,000</td>
<td>31%</td>
<td>49%</td>
<td>32%</td>
<td>31%</td>
<td>36%</td>
</tr>
<tr>
<td>25,000 to 50,000</td>
<td>29%</td>
<td>33%</td>
<td>45%</td>
<td>31%</td>
<td>34%</td>
</tr>
<tr>
<td>&gt;50,000</td>
<td>2%</td>
<td>12%</td>
<td>9%</td>
<td>7%</td>
<td>7%</td>
</tr>
</tbody>
</table>

The percentage of lymphocytes in this group of patients varied from 27 to 99%, with a mean of 70.4%.

Patients with a leukemoid reaction (WBC $> 50,000$) are more likely to have pulmonary complications such as atelectasis and pneumonia.

The relation of the total white cell count to the stage of the illness is illustrated in the following table.
Phagocytes

Phagocytes—257

Relation of the Total White Cell Count to the State of the Illness

<table>
<thead>
<tr>
<th>WHITE CELL COUNT</th>
<th>CATARRHAL STAGE (WEEKS 1-2) (%)</th>
<th>PAROXYSMAL STAGE (WEEKS 3-5) (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5,000 to 15,000</td>
<td>28</td>
<td>12</td>
</tr>
<tr>
<td>15,000 to 25,000</td>
<td>42</td>
<td>34</td>
</tr>
<tr>
<td>&gt;25,000</td>
<td>30</td>
<td>54</td>
</tr>
</tbody>
</table>


PHAGOCYTES

Congenital and Acquired Phagocytic Defects

Phagocytic cells are required to destroy microorganisms that would otherwise lead to disease in the host. The functions of these polymorphonuclear cells may be divided as follows: (1) adherence to the vascular endothelium; (2) chemotaxis or the recognition of and migration to a specific chemical stimulus; (3) phagocytosis; and (4) the killing of the ingested microorganism. Deficiency in serum opsonins (antibody or complement) or a deficiency in the number of phagocytes (neutropenia or asplenia) can also yield phagocytic dysfunction. Defects in one or more of these roles can lead to recurrent and severe infections. The phagocytic defects can either be congenital or acquired (in association with another disease). The congenital causes, which are extremely rare in frequency, include defects of cell movement (e.g., hyperimmunoglobulinemia E syndrome, actin dysfunction, and glycoprotein deficiency) and defects of microbial activity (e.g., chronic granulomatous disease, glucose 6-phosphate dehydrogenase deficiency, myeloperoxidase deficiency, and Chédiak-Higashi syndrome). Acquired or secondary phagocytic defects are far more common as detailed in the table below:

Acquired or Secondary Phagocytic Defects

<table>
<thead>
<tr>
<th>PRIMARY DISEASE OR CONDITIONS</th>
<th>ASSOCIATED CELLULAR DEFECT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Malnutrition</td>
<td>Chemotaxis, phagocytosis, killing</td>
</tr>
<tr>
<td>Hypophosphatemia</td>
<td>Phagocytosis, killing</td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td>Chemotaxis, adherence</td>
</tr>
<tr>
<td>Leukemia</td>
<td>Adherence, chemotaxis, phagocytosis, killing</td>
</tr>
<tr>
<td>Acute alcohol intoxication</td>
<td>Chemotaxis</td>
</tr>
<tr>
<td>Thermal injury</td>
<td>Chemotaxis, killing</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>Chemotaxis, phagocytosis, opsonins</td>
</tr>
<tr>
<td>Systemic lupus erythematosis</td>
<td>Chemotaxis</td>
</tr>
<tr>
<td>Inflammatory bowel disease</td>
<td>Chemotaxis, phagocytosis, opsonins</td>
</tr>
<tr>
<td>Systemic infections</td>
<td>Chemotaxis</td>
</tr>
<tr>
<td>Iron deficiency</td>
<td>Chemotaxis, killing</td>
</tr>
<tr>
<td>Pregnancy</td>
<td>Chemotaxis, killing</td>
</tr>
<tr>
<td>Steroids</td>
<td>Chemotaxi, opsonization</td>
</tr>
<tr>
<td>Viral infection</td>
<td>Chemotaxis, killing</td>
</tr>
</tbody>
</table>

Needle Phobia

A recent case report of a physician who displayed an involuntary fear of receiving injections and having his blood drawn reminds us of the distinct entity psychiatrists call “needle phobia.” (The 5-dollar word is belonephobia, from the Greek belone, meaning needle.)

Typically, phobias manifest themselves with the physical sign of transient tachycardia. Patients with needle or blood injury phobias, on the other hand, experience a diphasic cardiovascular response: tachycardia followed by bradycardia, hypotension, nausea, diaphoresis, vertigo, and syncope. On rare occasions, shock and death have been reported.

It is difficult to ascertain just how many children and adults have bona-fide needle phobia as opposed to simply not liking needles inserted into their bodies. Some experts estimate it at 5% of the population. Whether or not patients with true needle phobia will even present to a medical clinic or simply avoid all forms of health care is a different question.

Psychiatrists and evolutionary biologists have hypothesized that needle phobia may have a selective value over more traditional “fight or flight” responses to bodily harm. This is to say, there may be protective benefits in a fainting response or adverse cardiovascular reflex when confronted with an aggressive intern armed with a needle. On the other hand, a fainting toddler might be preferable to a screaming one, especially when one is compelled to obtain blood!


Photophobia

Photophobia is an abnormal intolerance for light, usually a result of inflammation of the iris and ciliary body. It is not to be confused with photosensitivity, which is also associated with a long list of diseases, usually with skin signs. A better term for photophobia is photodysphoria, but the latter is seldom used.

Although the diseases associated with photophobia are often obvious and relatively easily diagnosed (e.g., viral conjunctivitis, measles, or bacterial meningitis), there are other, more subtle conditions that must be considered when the primary diagnosis is not so obvious. Some of these associations are listed below.

More Common Associated Infections
Measles
Coxsackie B infection
Lymphocytic choriomeningitis
Viral conjunctivitis
Arbovirus infection
Bacterial meningitis

Less Common Associated Infections
Phlyctenular conjunctivitis
Yellow fever
Psittacosis infections
Rickettsial infections
(Rocky Mountain spotted fever, murine typhus)
Noninfectious Associations

Infantile glaucoma
Albinism
Vitamin A deficiency
Keratitis (e.g., Reiter's syndrome)
Erythropoietic porphyria
Acute cerebellar ataxia
Chédiak-Higashi syndrome
Aniridia
Cystinosis

Migraine
Corneal ulcer
Hysteria (in older child)
Arsenic poisoning
Mercury poisoning
Drug toxicity
Trimethadione
Ethosuccimide
PAS


School Phobia

Vague physical complaints are frequently heard in the pediatrician's office. When combined with normal physical and laboratory findings and poor school attendance because of the complaints, the child is often found to have "school phobia," a descriptive term for anxiety over leaving home in the 6 to 10 year old group.

Once a significant child-teacher conflict or fear of harassment by other children has been ruled out, the physician's immediate goal should be the return of the child to full school attendance. Steps in this direction to be discussed during the visit are listed below.

1. Do a thorough physical examination and pertinent laboratory studies as soon as possible. The child should then be given an unequivocal "clean bill of health." The findings should be conveyed to the parents along with a brief but sympathetic explanation about the reality of symptoms caused by anxiety or depression.

2. The parents should be gently but firmly convinced that immediate return of the child to school is essential. The parents must insist on the child's return to school for this step to be effective. Delay in return to school makes it increasingly difficult for the child to go back.

3. What to expect and what to do on school mornings should be reviewed with the mother. She should not ask the child how he feels. If he is up he should go to school, even if he is late or has missed the school bus. If he comes home at lunch he should be returned. If the child says he is ill, the mother should do one of two things. If questionably or mildly ill, he can be sent to school. If the mother feels the child is truly ill, he should be seen by the physician early that same morning. The child is not to stay at home without seeing a physician.

4. The person to be in charge of taking the child to school if he refuses to go should be clarified. This may be one of the parents, another relative, school social worker, or other responsible adult.

5. The school principal should be contacted by the physician, who can ask the school's cooperation in helping the child return to school. This is especially important if the child has a real or imaginary fear regarding some condition at school. The school nurse should also be contacted if she has been sending the child home for minor illness. She should be asked to have the child rest in her office for a time rather than send him home.
6. Weekly visits for several weeks are important for follow-up. A final visit several months later will allow long-term assessment.

Failure of this program, if conscientiously carried out by parents, physician and school personnel, suggests the necessity of psychiatric referral to explore the severe dependency problems that are often present.

Prevention of school phobia and related dependency problems can be aided by the encouragement of independence at appropriate times during infancy and preschool problems. The following milestones of independence may be useful guidelines.

<table>
<thead>
<tr>
<th>WHEN</th>
<th>CHILD SHOULD</th>
</tr>
</thead>
<tbody>
<tr>
<td>By 6 months</td>
<td>be left with baby sitter while parents have evenings out.</td>
</tr>
<tr>
<td>By 2 years</td>
<td>be left home, while awake, with baby sitter.</td>
</tr>
<tr>
<td>By 3 years</td>
<td>experience being left somewhere other than his home.</td>
</tr>
<tr>
<td>As soon as ready</td>
<td>be allowed to feed, dress, and wash himself.</td>
</tr>
<tr>
<td>By 3-4 years</td>
<td>be allowed to play in yard by himself.</td>
</tr>
<tr>
<td>By 4-5 years</td>
<td>be allowed to play in neighborhood by himself.</td>
</tr>
</tbody>
</table>


**PIGMENTURIA**

**Myoglobinuria, Hemoglobinuria, or Porphyria?**

The passage of large quantities of pigment in the urine often produces diagnostic confusion. Many substances may color the urine, but few mimic the appearance of hemoglobin. Hemoglobinuria must be distinguished from myoglobin or porphyrin compounds. Both myoglobin and hemoglobin will give positive results on the commonly employed dipstick (Labstix) for heme. The following table should provide a guide in the initial differential diagnosis of the three major causes of pigmenturia.

**Physical and Biochemical Features of the Pigmenturias**

<table>
<thead>
<tr>
<th>PHYSICAL EXAMINATION</th>
<th>MYOglobINURIA</th>
<th>HEMOGLOBINURIA</th>
<th>PORPHYRIA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Muscles</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Weakness</td>
<td>+</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>Pain</td>
<td>±</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>Edema</td>
<td>+</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Neuropathy (peripheral and autonomic)</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>CNS dysfunction</td>
<td>-</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>Skin lesion</td>
<td>-</td>
<td>-</td>
<td>±</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>Rare</td>
<td>-</td>
<td>+</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
### Physical and Biochemical Features of the Pigmenturias (Cont.)

<table>
<thead>
<tr>
<th>PHYSICAL EXAMINATION</th>
<th>MYOglobinuria</th>
<th>HEMOglobinuria</th>
<th>Porphyrria</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Laboratory Tests</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Urine</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Color</strong></td>
<td>Brown</td>
<td>Red-brown</td>
<td>Burgundy</td>
</tr>
<tr>
<td>Benzidine</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>Hematase-orthotoluidine</td>
<td>+</td>
<td>+</td>
<td>-</td>
</tr>
<tr>
<td>80% (NH₄)₂SO₄PPT</td>
<td>-</td>
<td>+</td>
<td>?</td>
</tr>
<tr>
<td>80% (NH₄)₂SO₄SUPER</td>
<td>+</td>
<td>-</td>
<td>?</td>
</tr>
<tr>
<td>Porphobilinogen</td>
<td></td>
<td></td>
<td>+</td>
</tr>
<tr>
<td>Spectrophotometry (α band)</td>
<td>582 (oxymyo)</td>
<td>577 (oxyhemo)</td>
<td>594 to 624*</td>
</tr>
<tr>
<td>Taurine</td>
<td>Increased</td>
<td>Normal</td>
<td>?</td>
</tr>
<tr>
<td>Immunodiffusion</td>
<td>Specific</td>
<td>Specific</td>
<td>-</td>
</tr>
<tr>
<td><strong>Serum</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Appearance</strong></td>
<td>Clear</td>
<td>Pink</td>
<td>Clear</td>
</tr>
<tr>
<td>Haptoglobin</td>
<td>Normal</td>
<td>Low</td>
<td>Normal</td>
</tr>
<tr>
<td>Creatine phosphokinases</td>
<td>Marked increase</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Carnitine</td>
<td>Increased</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Immunodiffusion</td>
<td>Specific</td>
<td>Specific</td>
<td>-</td>
</tr>
<tr>
<td>Triglycerides</td>
<td>↑ in specific defects</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

*Varies with type.

The clinical circumstances may provide the most help in defining the cause of pigment in the urine. The following conditions are associated with myoglobinuria.

### Causes of Myoglobinemia and Myoglobinuria

- **Trauma and ischemic disease**
  - “Crush” syndrome
  - Arterial ischemia of extremities, myocardial infarction
  - Pressure necrosis (comatose states)
  - Surgical procedures (orthopedic, vascular, cardiac)

- **Exertional states**
  - Exertion in otherwise normal individuals (military recruits)
  - Convulsive disorders

- **Metabolic disorders**
  - Alcoholic myopathy
  - Anesthetic associated syndromes (malignant hyperthermia)
  - Defects in carbohydrate metabolism (McArdle’s disease, phosphofructokinase deficiency, syndrome of abnormal glycolysis)
  - Defect in lipid metabolism (deficiency of carnitine, palmitoyl transferase)
  - Hypokalemia
  - Toxins (heroin user’s rhabdomyolysis, quail eater’s disease, Haff disease, snake and hornet venoms)

Most hospital laboratories will make a definitive differentiation between myoglobin and hemoglobin by performing a cellulose acetate electrophoresis. Unfortunately, if it is not between 8:00 A.M. and 5:00 P.M., you may be out of
Pleural Effusion

luck getting the laboratory to perform this test. An alternative test, which is presumptive of the presence of myoglobin, is based on differential solubility in ammonium sulfate. This is based on the principle that myoglobin is soluble in 80% saturated ammonium sulfate solution, whereas hemoglobin is not.

The test is performed as follows:

1. Clear the urine specimen by centrifugation or filtration.
2. Add 2.8 gm of (NH₄)₂SO₄ to 5 ml of urine, making an 80% saturated solution of (NH₄)₂SO₄. Allow the solution to stand for 5 minutes, then filter.
3. If myoglobin is in the urine, it will remain in solution. If hemoglobin is in the urine, it will precipitate and will be detected on the filter paper.

A presumptive positive test should be followed by an electrophoresis when available. Whatever test is used, the urine must be absolutely fresh.


PLEURAL EFFUSION

Pleural Effusions: Exudates or Transudates?

It seems the question always comes up. Is it a transudate or an exudate? Only an analysis of the fluid obtained by a thoracentesis can answer the question and even then you can't always be certain.

A transudate occurs when the mechanical factors influencing the formation or reabsorption of pleural fluid are altered. Decreased plasma oncotic pressure, and elevated systemic or pulmonary hydrostatic pressure are alterations that commonly produce transudates. In contrast, an exudate results from inflammation or other diseases of the pleural surface. Common conditions producing an exudate include: pneumonia, tuberculosis, pancreatitis, pulmonary infarction, and systemic lupus erythematosus.

Reliance on a single test to distinguish an exudate from a transudate will frequently be misleading. In the past, the measurement of pleural fluid protein, or specific gravity, or cell count has been employed as a diagnostic aid. Any single test will give unacceptably high “false positive” or “false negative” results.

The use of the following three tests will enable you to correctly classify virtually all pleural effusions:

1. Pleural fluid protein
   Serum protein
   ≥ 0.5 (suggests exudate)

2. Pleural fluid LDH
   ≥ 200 IU (suggests exudate)

3. Pleural fluid LDH
   Serum LDH
   ≥ 0.6 (suggests exudate)
The presence of two of these criteria strongly suggests a diagnosis of exudate—the presence of all three virtually assures it.

Some other helpful facts include:

About 80% of transudates will have a white cell count of less than 100/mm³, while 80% of exudates will have white cell counts above 1000/mm³.

Pancreatitis often produces a left-sided pleural effusion.

If congestive heart failure is associated with a unilateral effusion, it is usually right-sided.


**POISONING**

**An Unknown Poison**

It is axiomatic that the most useful information in an accidental poisoning is the label on the container. In over 90% of poisonings, the poison is known from the label or other source. However, situations sometimes arise where a possible poisoning has occurred, but the amount and nature of the ingested substance are unknown. Features that should suggest poisoning in an ill child include:

1. Abrupt onset of illness
2. Child’s age 1 to 4 years
3. History of previous ingestion
4. Multiple organ system involvement that does not fit single disease

A combination of symptoms will sometimes suggest the drug or poison involved:

<table>
<thead>
<tr>
<th>Symptoms and Signs</th>
<th>Possible Poison</th>
</tr>
</thead>
<tbody>
<tr>
<td>Agitation, hallucinations, dilated pupils, bright red color to the skin, dry skin, and fever</td>
<td>Atropine-like agents</td>
</tr>
<tr>
<td>Marked activity, tremors, headache, diarrhea, dry mouth with foul odor, sweating, tachycardia, arrhythmia, dilated pupils</td>
<td>Amphetamines</td>
</tr>
<tr>
<td>Slow respirations, pinpoint pupils, euphoria, or coma</td>
<td>Opiates</td>
</tr>
<tr>
<td>Salivation, lacrimation, urination, defecation, miosis, and pulmonary congestion</td>
<td>Organic phosphates or poison mushrooms</td>
</tr>
<tr>
<td>Sleepiness, slurred speech, nystagmus, ataxia</td>
<td>Barbiturates or tranquilizers</td>
</tr>
<tr>
<td>Hypernea, fever, and vomiting</td>
<td>Salicylates</td>
</tr>
<tr>
<td>Oculogyric crises, ataxia, and unusual posturing of head and neck</td>
<td>Phenothiazines</td>
</tr>
<tr>
<td>Nausea, vomiting, sweatiness, and pallor are early manifestations; late manifestations include stupor and signs of liver failure</td>
<td>Acetaminophen</td>
</tr>
</tbody>
</table>

The following list expands on signs and symptoms and the toxins with which they may be associated.
264—Poisoning

Ataxia
- Alcohol
- Barbiturates
- Bromides
- Carbon monoxide
- Diphenylhydantoin

Convulsions and muscle twitching
- Alcohol
- Amphetamines
- Antihistamines
- Boric acid
- Camphor
- Chlorinated hydrocarbon insecticides (DDT)
- Cocaine
- Cyanide

Coma and drowsiness
- Alcohol—ethyl
- Antihistamines
- Barbiturates and other hypnotics
- Carbon monoxide

Paralysis
- Botulism
- Heavy metals

Pupils
- Pinpoint
  - Mushrooms (muscarine type)
  - Narcotic depressants (opiates)
- Dilated
  - Amphetamines
  - Antihistamines
  - Atropine
  - Barbiturates (coma)
  - Cocaine

Nystagmus on lateral gaze
- Barbiturates
- Minor tranquilizers (meprobamate, benzodiazepine)

Pulse rate
- Slow: Digitalis, Lily of the valley, Narcotic depressants
- Rapid: Alcohol, Amphetamines, Atropine, Ephedrine
Respiratory alterations

**Rapid**
- Amphetamines
- Barbiturates (early)
- Carbon monoxide
- Methanol
- Petroleum distillates
- Salicylates

**Slow or depressed**
- Alcohol
- Barbiturates (late)
- Narcotic depressants (opiates)
- Tranquilizers

*Wheezing and pulmonary edema*
- Mushrooms (muscarine type)
- Narcotic depressants (opiates)
- Organic phosphate insecticides
- Petroleum distillates

*Paralysis*
- Organic phosphate insecticides
- Botulism

**Mouth**

*Salivation*
- Arsenic
- Corrosive
- Mercury
- Mushrooms
- Organic phosphate insecticides
- Thallium

*Dryness*
- Atropine
- Amphetamines
- Antihistamines
- Narcotic depressants

**Breath odor**
- Acetone: acetone, alcohol (methyl, isopropyl), phenol, salicylates
- Alcohol: alcohol (ethyl)
- Bitter almonds: cyanide
- Coal gas: carbon monoxide
- Garlic: arsenic, phosphorus, organic phosphate insecticides, thallium
- Oil of wintergreen: methyl salicylate
- Petroleum: petroleum distillates
- Violets: turpentine

**Skin color**

*Jaundice (hepatic or hemolytic)*
- Aniline
- Arsenic
- Carbon tetrachloride
- Castor bean
- Fava bean
- Mushroom
- Naphthalene
- Yellow phosphorus

*Cyanosis*
- Aniline dyes
- Carbon monoxide
- Cyanide

*Red flush*
- Alcohol
- Antihistamines
- Atropine
- Boric acid
- Carbon monoxide
- Nitrites
- Strychnine
- Nitrites
- Rifampin

**Violent emesis often with hematemesis**
- Acetaminophen
- Aminophylline
- Bacterial food poisoning
- Boric acid
- Corrosives
- Fluoride
- Heavy metals
- Phenol
- Salicylates
Poisoning

Abdominal colic
- Black widow spider bite
- Heavy metals
- Narcotic depressant withdrawal

Oliguria-anuria
- Carbon tetrachloride
- Ethylene glycol
- Heavy metals
- Hemolytic poisons (naphthalene, plants)
- Methanol
- Mushrooms
- Oxalates
- Petroleum distillates
- Solvents


Carbamate and Organophosphate Poisoning

Despite many parents’ valiant attempts, “Mr. Yuk” does not always deter curious children. Carbamates and organophosphates remain ubiquitous components of insecticides, and their toxic effects can reach the nervous system through inhalation, absorption, and ingestion.

The carbamates and organophosphates are anticholinesterases that lead to the accumulation of unhydrolyzed acetylcholine at the receptors. The result is continued stimulation and, ultimately, paralysis of cholinergic transmission. The organophosphates penetrate the central nervous system and will show central effects that the carbamates do not.

Clinical Features of Carbamate and Organophosphate Insecticide Poisoning

<table>
<thead>
<tr>
<th>Muscarinic</th>
<th>Nicotinic</th>
<th>Central nervous system (organophosphates)</th>
</tr>
</thead>
<tbody>
<tr>
<td>“Sludge”</td>
<td>Muscle symptoms</td>
<td>Severe headache</td>
</tr>
<tr>
<td>Salivation</td>
<td>Fasciculations</td>
<td>Tremor</td>
</tr>
<tr>
<td>Lacrimation</td>
<td>Cramps and fatigue</td>
<td>Ataxia</td>
</tr>
<tr>
<td>Urination</td>
<td>Loss of deep tendon reflexes</td>
<td>Restlessness</td>
</tr>
<tr>
<td>Defecation</td>
<td>Paralysis</td>
<td>Slurred speech</td>
</tr>
<tr>
<td>Gastrointestinal pain &amp; cramping</td>
<td>Tachycardia</td>
<td>General weakness</td>
</tr>
<tr>
<td>Emesis</td>
<td>Hypertension</td>
<td>Seizures</td>
</tr>
<tr>
<td>Miosis: if present, look for hyperactive bowel sounds</td>
<td></td>
<td>Coma</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cardiorespiratory depression</td>
</tr>
</tbody>
</table>

Recognition and Treatment

The muscarine effects generally precede the nicotinic effects. If you have a case of suspected carbamate or organophosphate poisoning, the patient will be atropine-refractory: atropinization should occur in the nonpoisoned patient within 5-20 minutes after a dose of 0.05 mg/kg for a child, or 1-2 mg for an adult.

Prior to the atropine test, the ABCs of Basic Life Support are mandatory. The patient should be thoroughly disrobed and cleansed due to possible continued absorption through dermal contact. If ingestion occurred, initiate ipecac/lavage and charcoal.
Atropine is the cornerstone of therapy. Give the child 0.05 mg/kg as needed; give the adult 0.4 to 2.0 mg IV every 15–30 minutes until the patient cannot spit. In general, a 6–12 h course of atropine is necessary. With carbamate poisoning, the cholinesterase complex is reversible. This is not the case with organophosphates, so additional therapy with pralidoxime, a cholinesterase regenerator, is necessary.


POLYPOSIS

Familial Polyposis

Familial polyposis is an autosomal dominant disorder notable for development of multiple adenomatous polyps in the colon and rectum. The polyps usually do not become apparent until after puberty. The incidence of familial polyposis ranges from 1 in 7,000 to 1 in 10,000 births. The risk of developing colon cancer in affected individuals approaches 100% by age 55. Patients with familial polyposis tend to seek medical attention because of either a family history of polyposis or symptoms of abdominal discomfort, rectal bleeding, and diarrhea. Frequently, the abdominal symptoms do not appear until 10 years after the development of polyps.

Clinical Features

1. Autosomal dominant inheritance (rare spontaneous mutations have been reported).
2. Onset in adolescence
3. Onset of symptoms begin about 10 years after appearance of polyps.
4. Multiple colonic adenomatous polyps
5. Associated extracolonic lesions
   a. Epidermoid cysts (usually on head, neck, and trunk)
   b. Subcutaneous fibromas (usually on the scalp, shoulders, arms and back)
   c. Desmoid tumors of the abdominal wall, mesentary, and retroperitoneum
   d. Osteoma (symptoms a–d in association with polyposis is also known as Gardner syndrome).
   e. Sebaceous cysts
   f. Gastric and duodenal polyps
   g. Congenital hypertrophy of the retinal pigment epithelium (multiple pigmented patches on one or both fundi)
   h. Abnormal dentition (including odontomas, dentigenous cysts, and unerupted, missing, or supernumerary teeth)
6. High risk of colon cancer
   a. 100% risk in untreated patients
   b. Early age of onset (median age is 39 years)
   c. Cancers arise from adenomatous polyps
   d. Synchronous colon carcinomas are common.
   e. Metachronous colon carcinomas are common.
**Polyuria**

**Protocol for Screening Patients at Risk of Familial Polyposis**

<table>
<thead>
<tr>
<th>AGE</th>
<th>ASYMPTOMATIC</th>
<th>SYMPTOMATIC</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 13</td>
<td>None</td>
<td>Flexible sigmoidoscopy</td>
</tr>
<tr>
<td>14-10</td>
<td>Annual flexible sigmoidoscopy</td>
<td>Colonoscopy or double-contrast barium enema</td>
</tr>
<tr>
<td>20-45</td>
<td>Annual flexible sigmoidoscopy, baseline colonoscopy, or double-contrast barium enema at age 20, repeated every 3 years</td>
<td>Colonoscopy or double-contrast barium enema each year</td>
</tr>
</tbody>
</table>

**Surgical Management of Familial Polyposis Patients**

<table>
<thead>
<tr>
<th>COLONIC POLYPS</th>
<th>PROCEDURE</th>
<th>POSTOPERATIVE FOLLOW-UP</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 10</td>
<td>Endoscopic removal</td>
<td>Every 6 months</td>
</tr>
<tr>
<td>Multiple polyposis with &lt;20 rectal polyps</td>
<td>Colectomy with ileorectal anastomosis, or colectomy with mucosal proctectomy and reservoir ileal anastomosis</td>
<td>Every 6 months</td>
</tr>
<tr>
<td>Multiple polyposis with &gt;20 rectal polyps</td>
<td>Total proctocolectomy with ileostomy</td>
<td>Annual</td>
</tr>
</tbody>
</table>


**POLYURIA**

**Common Causes**

- Diabetes mellitus
- Diuretic abuse
  - Alcohol
  - Caffeine
  - Medications
- Iatrogenic
  - Aggressive parenteral hydration
  - Diuretic use
- Psychogenic polydipsia
- Renal failure
- Sickle-cell anemia
- Urinary tract infection

**Uncommon Causes**

- Diabetes insipidus (central)
- Interstitial nephritis
  - Analgesic abuse
  - Diphenylhydantoin
- Mercury poisoning
- Methicillin reaction
- Sulfonamides
- Renal calculi/hypercalcemia
- Renal tubular acidosis

**Rare Causes**

- Bartter's syndrome
- Cystinosis
- Medullary cystic disease of the kidney
- Nephrogenic diabetes insipidus
- Neuroblastoma/ganglioneuroblastoma
- Pheochromocytoma
POTASSIUM

Hyperkalemia is defined here as a serum potassium level higher than 5.5 mEq/L.

**Common Causes**
- Acidosis
- Renal failure
- Severe dehydration

**Uncommon Causes**
- Excessive potassium infusion
- Shock

**Rare Causes**
- Addison's disease (adrenal insufficiency)
- Cell lysis syndromes

Hypokalemia is defined here as a serum potassium level lower than 3.5 mEq/L.

**Common Causes**
- Chronic diarrhea
- Diuretics
- Malnutrition
- Metabolic alkalosis

**Uncommon Causes**
- Excessive corticoids
- Renal tubular disorders

**Rare Causes**
- Amphotericin B therapy
- Bartter's syndrome
- Cushing's syndrome
- Familial periodic paralysis

---

PORPHYRIAS

Which Type Is Present?

It is important to distinguish porphyrias from simple porphyrinuria, which is associated with a number of common conditions. It is also possible to have some notion of the various forms of porphyrias by clinical signs and routine laboratory results.

In general, of the porphyrias that are associated with excretion of excessive amounts of porphyrin precursors, only acute intermittent porphyria is associated with abdominal pain. Those porphyrias in which the latter part of the heme synthesis pathway is affected are associated with excretion and accumulation of porphyrins. These forms of the disease include congenital erythropoietic porphyria, erythropoietic protoporphyria, and porphyria cutanea tarda. Dermatologic manifestations predominate in these forms.

The forms of the disease in which both porphyrias and their precursors are excreted are associated with both abdominal pain and dermatologic manifestations. These forms include porphyria variegata and hepatic coproporphyria.

The two most common forms of porphyria encountered in clinical practice in the U.S. are porphyria cutanea tarda, with cutaneous signs, and acute intermittent porphyria, with neurologic symptoms, usually occurring in acute episodic attacks.


Fist Clenching Pseudohyperkalemia

There are some lessons in medicine that must be learned over and over. The causes of pseudohyperkalemia appear to be one of those lessons.

When a non-hemolyzed specimen results in a laboratory report of hyperkalemia in the absence of excessive intake or decreased renal excretion, question yourself or question the phlebotomist. Did the adolescent clench his or her fist or use an isometric handgrip? Did the infant or child struggle or resist during blood drawing? Muscular contractions cause local release of potassium and can cause false elevations in serum values (1 to 2 mmol per liter). Do not get caught in the grip of pseudohyperkalemia!

Causes of Hyperkatemia (Serum (K⁺) > 4.9 mmol/L)

1. Pseudohyperkalemia
   a. Local release due to muscular contraction
   b. Hemolyzed specimen
   c. Severe thrombocytosis (pH > 10⁶ ml)
   d. Severe leukocytosis (WBC > 10⁵ ml)

2. Excessive intake
   a. Potassium replacement therapy
   b. Potassium salts of antibiotics
   c. Salt substitutes
   d. High-potassium diet: bananas, orange juice, carrots, celery, broccoli

3. Decreased renal excretion
   a. Potassium-sparing diuretics (e.g., triamterene, spironolactone, amiloride)
   b. Renal insufficiency
   c. Mineralocorticoid deficiency
   d. Hyporeninemic hypoaldosteronism (diabetes mellitus)
   e. Tubular unresponsiveness to aldosterone (e.g., sickle cell disease, SLE)
   f. Heparin administration

4. Redistribution (excessive cellular release)
   a. Acidemia (each 0.1 decrease in pH, 0.4-0.6 mmol/L increase in K⁺)
   b. Insulin deficiency
   c. Hypertonicity
   d. Hemolysis
   e. Tissue necrosis, rhabdomyolysis, burns
   f. Hyperkalemic periodic paralysis


PROcedures

Site and Depth of Heel Skin Punctures in the Newborn

Every day, including Sundays, literally thousands of newborn infants have heel punctures performed in order to obtain blood samples.
Many of these punctures are badly performed. Little attention is paid to normal anatomy. Serious complications of heel punctures in newborns include calcaneal osteomyelitis and necrotizing chondritis.

The skin's primary arterial blood supply comes from an arterial network at the junction of the lower dermis and upper subcutaneous tissue. Branches from one side of this network supply blood to the subcutaneous tissue, and those from the other side supply the dermis. A large network of veins is also present at the dermal subcutaneous junction. Because of the anatomy, most of the blood obtained from a skin puncture flows from vessels at the dermal subcutaneous junction, and for this reason it is not necessary to extend the puncture any deeper to obtain adequate blood flow.

How deep is this junction? The accompanying figure (below right) illustrates the distance from the skin to the subcutaneous junction (S-S) and the distance from the skin to the periosteum of the calcaneus (S-P) as a function of body weight. A lancet puncture of 2.4 mm will extend below the dermal subcutaneous junction but will not penetrate the perichondrium in even the smallest infants. Do not go deeper than 2.4 mm.

The side-to-side limits of the calcaneus are illustrated in the drawing (left). A line extending posteriorly from a point between the fourth and fifth toes and running parallel to the lateral aspect of the heel, and another line extending posteriorly from the middle of the big toe and running parallel to the medial aspect of the heel, serve as useful guidelines. Heel punctures should be performed on the plantar surface of the heel and beyond the lateral and medial limits of the calcaneus. These safe areas are marked by the hatched lines in the illustration. Don't be responsible for bone spurs.


PROTEINURIA

Common Causes

Chronic pyelonephritis
Isolated transient/intermittent proteinuria
Cold exposure
Congestive heart failure
Exercise
Febrile illness
Idiopathic proteinuria
Orthostatic proteinuria
Pregnancy
Trauma
Urinary tract infection

Uncommon Causes

Nephritis sediment
Membranoproliferative glomerulonephritis
Postinfectious glomerulonephritis
Nephrotic sediment
Minimal change disease
Preeclampsia
Tubular proteinuria
Acute tubular necrosis
Obstructive uropathy
Polycystic kidney disease

Rare Causes

Drugs
Captopril
Fenoprofen
Gold
Penicillamine
Probenecid
Nephritic sediment
Hereditary nephritis
IGA nephropathy
Mixed cryoglobulinemia
Rapidly progressive glomerulonephritis
Subacute bacterial endocarditis
Systemic lupus erythematosus
Nephrotic sediment
Amyloidosis
Diabetes mellitus

Uncommon Causes (Cont.)
Focal glomerulonephritis
Membranous nephropathy
Miscellaneous infections
Hepatitis B
Malaria
Syphilis
Overflow proteinuria
Bence Jones proteinuria
Lysozymuria (in leukemia)
Tubular proteinuria
Analgesic abuse
Chronic hypertension
Hypercalcuiuria
Hyperuricemia
Radiation nephritis

PRURITUS

Common Causes

Atopic dermatitis
Cholestasis of pregnancy
Contact allergens (plants, cosmetics, dyes, medications)
Contact irritants (soaps, chemicals, excrement, wool)
Dermatitis herpetiformis

Drugs
Aminophylline
Aspirin
Barbiturates
Erythromycin
Gold
Griseofulvin
Drugs (Cont.)

- Isoniazid
- Opiates
- Phenothiazines
- Vitamin A

Dry skin

- Advanced age
- Excess bathing/strong detergents
- Low humidity

Foreign body

Hepatitis

Herpes gestationis

High humidity

Insect bites/infestations

- Fleas, mosquitos, scabies miters, lice mitis, chiggers
- Iron-deficiency anemia
- Parasitic infection
  - Pinworms
  - *Toxocara canis*

Pityriasis rosea

Psoriasis

Seborrheic dermatitis

Skin infections (bacterial/viral/fungal)

Urticaria

Water contact (aquagenic)

Uncommon Causes

- Biliary obstruction
  - Drug induced
  - Extrahepatic biliary obstruction
  - Primary biliary cirrhosis
- Chronic renal failure
- Hematopoietic malignancies
  - Hodgkin's disease
  - Leukemia

Hematopoietic malignancies (Cont.)

- Lymphoma
- Neurodermatitis
- Parasitic infection
  - Cercaria
  - Hookworms
  - Trichinosis

Rare Causes

- Autoimmune (SLE, JRA)
- Congenital ectodermal disorders
- Endocrine disorders
  - Carcinoid syndrome
  - Diabetes mellitus
  - Hyper/hypothyroidism
  - Hypoparathyroidism
- Erythropoietic protoporphyria

Hematopoietic malignancies

- Mastocytosis
- Multiple myeloma
- Polycythemia vera
- Malignant solid tumors
- Neurologic syndromes
- Psychosis

Relieving the Sting or the Itch

Although a number of drugs and lotions are available for the treatment of bites and rashes, home remedies are often as good or even better in producing relief. Here are some tried and true home remedies:

**Home Remedies for the Treatment of Bites and Rashes**

<table>
<thead>
<tr>
<th>FOR</th>
<th>TRY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bee stings, wasp stings, and jellyfish bites</td>
<td>Adolph's meat tenderizer. Add a little water to the powder and rub into the bite. Expect relief in minutes.</td>
</tr>
<tr>
<td>Poison ivy</td>
<td>Ban roll-on deodorant. Just rub on the rash and rub away the itch.</td>
</tr>
</tbody>
</table>

Table continued on next page.
Delayed Puberty and the Adolescent Female

Surely none of us can forget the emotional and physiologic complexities of puberty. The adolescent female who believes her development is delayed can be caught in a maelstrom of anxiety. For the majority of girls with “delayed” puberty, the cause is none other than normal variance. In these cases, the pediatrician is the perfect person to provide the needed reassurance that development will occur. For the minority of females with a pathologic cause of pubertal delay, diagnosis and, in some cases, treatment, are within reach. The tables and evaluation plan that follow will help you and your patient in this delicate matter.

Late, Delayed or Arrested?

“Late” defines the onset of puberty at an age older than the average but within 2 standard deviations of the mean.

“Delayed”—when no signs of sexual development have begun by the age of 13. “Arrested”—when more than 5 years have passed between adrenarche or thelarche and menarche.

---

Tanner Stages and Their Mean Age of Appearance

<table>
<thead>
<tr>
<th>STAGE</th>
<th>BREAST DEVELOPMENT</th>
<th>PUBIC DEVELOPMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>MEAN AGE (YR)</td>
<td>RANGE (YR)</td>
</tr>
<tr>
<td>Tanner I</td>
<td>11.2</td>
<td>8.0-13.0</td>
</tr>
<tr>
<td>(prepubertal)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tanner II</td>
<td>12.2</td>
<td>10.0-14.3</td>
</tr>
<tr>
<td>Tanner III</td>
<td>13.1</td>
<td>10.8-15.3</td>
</tr>
<tr>
<td>Tanner IV</td>
<td>15.3</td>
<td>11.9-18.8</td>
</tr>
<tr>
<td>Tanner V</td>
<td>12.6</td>
<td>10.0-16.0</td>
</tr>
<tr>
<td>Menarche</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
## Causes of Delayed Puberty

### Constitutional Delay in Growth and Development (CD GD): Family History

### Hypogonadotropic hypogonadism

**Hypothalamic-pituitary disorders**
- Isolated deficiency of GnRH
- Isolated deficiency of LH, FSH, or both
- Panhypopituitarism

**Associated abnormalities**
- Kallman syndrome (anosmia)
- Prader-Willi syndrome
- Bardet-Biedl syndrome

**Postinflammatory**
- Autoimmune (hypophysitis)
- Infectious (meningitis, encephalitis)

**Trauma**

**Infiltration**
- Histiocytosis X
- Hemochromatosis

**Irradiation**

**Tumor**
- Craniopharyngioma
- Optic glioma
- Adenoma

### Functional gonadotropin deficiency

**Chronic systemic or endocrinologic disease**
- Cardiovascular (congenital or acquired)
- Pulmonary (asthma, cystic fibrosis)
- Hematologic (sickle cell disease)
- Gastrointestinal (celiac disease, chronic inflammatory bowel disease, other causes of malabsorption)
- Renal (renal tubular acidosis, renal failure)
- Immunologic (chronic/persistent infection, immunocompromise)
- Collagen-vascular (SLE, JRA)
- Endocrine (hypothyroidism, glucocorticoid excess, hyposomatotropism, IDDM)
- Psychiatric (emotional stress)

### Hypogonadotropic hypogonadism Cont.

**Weight loss**
- Anorexia nervosa
- Malabsorption
- Exercise

**Hyperprolactinemia**
- Prolactinoma

### Hypergonadotrophic hypogonadism

**Primary gonadal abnormalities**
- Gonadal dysgenesis and its variants
- Insensitivity to gonadotropins
- Defects in steroidogenesis

**Acquired gonadal failures**
- Postinflammatory
- Autoimmune
- Infectious
- Posttraumatic
- Vascular
- Surgical
- Infiltration
- Galactosemia
- Myotonic dystrophy
- Ataxia-telangiectasia

**Toxic**
- Irradiation
- Chemotherapy

**Tumor**

### Hyperandrogenism

**Polycystic ovary syndrome (PCOS)**
- Nonclassical congenital adrenal hyperplasia (CAH)

### Anatomic genital abnormalities

**Rokitansky syndrome (congenital absence of uterus and vagina)**
- Transverse vaginal septum and/or imperforate hymen
- End-organ insensitivity to androgens (testicular feminization)

Evaluating Delayed Puberty in Girls

History → Family delay in growth and sexual development

Physical examination → Abnormal

Bone age

LH, FSH

Extremely elevated LH, FSH

Chromosome analysis

Moderately elevated LH

Pelvic sonography

Polycystic ovaries

GnRH

Elevated prolactin

Abnormal

Normal

Gonadal dysgenesis

Ovarian antibodies

Galactosemia

Gonadotropin insensitivity

Defect in steroidogenesis

Other causes of ovarian failure (see previous listing)

Absent, Prepubertal

Possible hypogonadotropism

Possible CDGD

Intracranial/parasellar lesion

Precocious Puberty: The Other Side of the Coin

Like delayed puberty, precocious puberty can cause significant emotional trauma in the preadolescent female. Unlike delayed puberty, precocious puberty is more often a sign of underlying disease with possible long-term consequences. Since precocious puberty is 4 to 8 times more common in girls, the following differential and algorithm are directed to the evaluation of the female patient.

By definition, isosexual precocious puberty in girls is the appearance of secondary sexual characteristics before the child's eighth birthday. An oft-confused phenomenon is that of "early adolescence" where signs of puberty appear between the ages of 8 and 10. The subsets of girls that are prone to "early adolescence" include those with simple obesity, advanced bone age, or increased body mass index. Below is a differential for each of the three categories of precocious puberty and a recommended work-up.
Categories of Precocious Puberty (True and Complete)

1. Central Isosexual precocity
   - Idiopathic, often familial
   - CNS anatomic defects
     - Septo-optic dysplasia
     - Hydrocephalus
     - Cysts
   - Postinflammatory
     - Meningitis
     - Encephalitis
     - Brain abscess
   - Trauma
   - Irradiation
   - Tumor
     - Hamartoma
     - Neurofibroma
     - Optic glioma
     - Astrocytoma
     - Ependymoma
     - Dysgerminoma
     - Craniopharyngioma
     - Postpseudosexual precocity

2. Pseudosexual precocity
   - Gonadotropin-dependent
     - HcG secreting tumors (very rare)
     - LH&FSH secreting tumors (very rare)
   - Gonadotropin-independent
     - Ovarian estrogen
       - Granulosa cell tumor
       - Follicular cysts
     - McCune-Albright syndrome
       - (irregularly contoured café au lait spots, polyostotic fibrous dysplasia, and precocious puberty)
     - Adrenal estrogen
       - Feminizing tumors
     - Exogenous estrogen or estrogen-like substances
       - Oral contraceptives
       - Topical estrogen
       - Cimetidine
       - Cannabis
       - Spironolactone
       - Digitalis

3. Primary hypothyroidism

Incomplete Isosexual Precocity

Premature thelarche, with or without galactorrhea: peak incidence 6 m–2 y; persists from birth in 23% of girls.

Premature menarche: any vaginal bleeding in prepubertal females should alert the pediatrician to possible sexual abuse or trauma, vaginal infection, tumors, foreign body, or urethral prolapse.


Algorithm (see top of next page.)

Treatments exist to “turn off” the hypothalamic-pituitary ovarian axis through pharmacologic or surgical therapies. The long-term consequences of precocious sexual development include compromise of final adult stature, possibly increased risk of cervical and breast cancers due to extended exposure to unopposed estrogen, and psychological trauma. When precocious puberty is suspected, a referral to a pediatric endocrinologist is suggested.

Timing of Puberty and Its Biopsychosocial Correlates

Research on the timing of maturation, precocious puberty, and environmental change has reported fairly consistent findings. The influence of puberty and its timing appear to have the greatest effects in the areas of self-conceptions (body image and self-esteem), developmental needs (heterosexual relationships, peer affiliations, family independence), school performance (academic performance and problem behaviors), and environmental responses (peer, parental, and teacher expectations). These effects vary as a function of:

1. gender,
2. the relationship of the individual’s pubertal status to that of his or her peers,
3. definitions of early and late timing, and
4. the behavior under investigation.

In general, the most negative effects have been reported for early-maturing females. Some recent work has shown that the effects of early maturation may be detrimental for both sexes, with early maturation in males being associated with the early initiation of sexual activity and other risk behaviors.

## Abnormal Pupils

<table>
<thead>
<tr>
<th>DESCRIPTION</th>
<th>NAME</th>
<th>DIFFERENTIAL DIAGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Shape</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absent iris</td>
<td>Aniridia</td>
<td>Wilms' tumor</td>
</tr>
<tr>
<td>Scalloped or asymmetric retraction</td>
<td>Irregular iris</td>
<td>Adhesions, old iritis, persistent pupillary membrane, trauma</td>
</tr>
<tr>
<td>Tearing the root of the iris from ciliary attachment</td>
<td>Iridodialysis</td>
<td>Trauma</td>
</tr>
<tr>
<td>Loss of circular shape</td>
<td>Coloboma</td>
<td>Congenital or operative</td>
</tr>
<tr>
<td><strong>Movement and Size</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Loss of light reflex, preservation of accommodation, miosis</td>
<td>Argyll Robertson pupil</td>
<td>Syphilis, also seen occasionally in encephalitis, multiple sclerosis, CNS tumor</td>
</tr>
<tr>
<td>Very slow light reflex, preservation of accommodation, mydriasis</td>
<td>Adie's pupil</td>
<td>Benign</td>
</tr>
<tr>
<td>Preservation of light reflex, loss of accommodation</td>
<td>Reverse Argyll Robertson pupil</td>
<td>Bilateral: Diabetes mellitus, syphilis, basilar meningitis, tumor of the corpora quadrigemina</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Unilateral: Diphtheria, intoxication (alcohol), syphilis</td>
</tr>
<tr>
<td>Loss of all reflex movements of the pupil</td>
<td>Ophthalmoplegia interna</td>
<td>Third nerve nucleus damage, diabetes mellitus, syphilis, diphtheria, tumor, trauma</td>
</tr>
<tr>
<td>Loss of ipsilateral light reflex, loss of contralateral consensual reflex</td>
<td>Optic nerve lesion</td>
<td>Lesion between chiasma and globe</td>
</tr>
<tr>
<td>Loss of psychic or sensory mydriasis (may be associated with Horner's syndrome)</td>
<td>Sympathetic pupil</td>
<td>Syringomyelia, paralysis of cervical sympathetic nerve</td>
</tr>
<tr>
<td>Miosis, preservation of light and accommodation reflexes</td>
<td>Miotic, reactive pupil</td>
<td>Neonates, the elderly, stimulation of pupillary sphincter, paralysis of dilator pupillae (encephalitis, syringomyelia, CNS abscess), tumor or hemorrhage irritating the center for constriction. Opiates, organic phosphates, pilocarpine.</td>
</tr>
<tr>
<td>Mydriasis, preservation of light and accommodation reflexes</td>
<td>Mydriatic, reactive pupil</td>
<td>Mania, schizophrenia, irritation without destruction of cervical sympathetics (i.e., aneurysm, tumor, blood infection), LSD</td>
</tr>
</tbody>
</table>

*The atropines cause cycloplegia: dilatation and paralysis of the iris.*

Table continued on next page.
### Abnormal Pupils

<table>
<thead>
<tr>
<th>DESCRIPTION</th>
<th>NAME</th>
<th>DIFFERENTIAL DIAGNOSIS</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Movement and Size (Cont.)</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pupils alternately dilate and contract rapidly (&quot;tremor of the iris&quot;)</td>
<td>Hippus</td>
<td>Multiple sclerosis, drug/alcohol overdose, homocystinuria, central scotoma with macular damage or disease or injury to axial fibers of optic nerve</td>
</tr>
<tr>
<td>More than one pupil in an eye</td>
<td>Polycoria</td>
<td>Congenital, traumatic, surgical</td>
</tr>
<tr>
<td>Inequality of size of pupils</td>
<td>Anisocoria</td>
<td>Variation of normal, iritis, diabetes mellitus, cervical sympathetic lesion, eye drops, glaucoma, unilateral damage to third nerve fibers, syphilis, trigeminal neuralgia, carotid or aortic aneurysm, cranial lesion, cerebral herniation, artificial eye</td>
</tr>
<tr>
<td>Pupils dilate under light stimulus</td>
<td>Paradoxical pupil (rare)</td>
<td>Syphilis</td>
</tr>
<tr>
<td>With strong deviation of the eyes, the pupil of the abducted eye is larger than that of the adducted eye</td>
<td>Tournay's sign</td>
<td>Normal</td>
</tr>
</tbody>
</table>


---

### PURPURA (PETECHIAL AND ECCHYMOSES)

#### Common Causes
- Thrombocytopenia
- Trauma
- Viral infections

#### Uncommon Causes
- Abnormal platelet function
- Child abuse
- Cupping and coin rubbing
- Drug ingestion (aspirin)
- Factitious
- Henoch-Schönlein purpura
- Hereditary coagulation disturbance
- Infection
- Sentic emboli
- Ulceria
- Vasculitis
- Violent coughing

#### Rare Causes
- Autoerythrocyte sensitization
- Bernard-Soulier (giant platelet) syndrome
- Cushing's syndrome
- Dysproteinemias
- Glanzmann's thrombasthenia
- Hereditary hemorrhagic telangiectasia
- Macular cerulae
- Marfan's syndrome
- May-Hegglin anomaly
- Osteogenesis imperfecta
- Osteopetrosis
- Platelet storage pool disease
- Protein C deficiency
- Protein S deficiency
- Purpura fulminans
- Schamberg's disease
- Scurvy
- Vitamin K deficiency
Purpura and Petechiae—Interpreting the Sign

Every little bruise can have a meaning all its own. It is important to look carefully at a hemorrhagic lesion for the clue to its underlying disease. Listed below are some guides to the interpretation of this sign.

<table>
<thead>
<tr>
<th>Condition</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Thrombocytopenic purpura</td>
<td>Petechiae are nonpalpable. Platelet count &lt; 20,000/ml.</td>
</tr>
<tr>
<td>Thrombocytopathic purpura</td>
<td>Easily bruised. Petechiae are rare.</td>
</tr>
<tr>
<td>Vasculitic purpura (+/- thrombocytopenia)</td>
<td>Petechiae are palpable.</td>
</tr>
<tr>
<td>Drug purpura (+/- thrombocytopenia)</td>
<td>Often associated with hemorrhagic bullae in the mouth.</td>
</tr>
<tr>
<td>Allergic purpura (Henoch-Schönlein)</td>
<td>Pruritic crops of symmetrical purpura on proximal extremities (4+ lower) associated with urticarial and erythematous lesions.</td>
</tr>
<tr>
<td>Purpura fulminans (skin manifestations of DIC)</td>
<td>Large symmetrical ecchymoses, particularly on distal extremities, complicated by acral gangrene. Petechiae are rare.</td>
</tr>
<tr>
<td>&quot;Devil’s pinches&quot; (autoerythosensitization vs. factitious)</td>
<td>Females. “Spontaneous” painful ecchymoses (+/- erythematous base) on anterior-lateral aspect of thigh and abdomen in a stepladder distribution.</td>
</tr>
<tr>
<td>Hyperglobulinemic purpura</td>
<td>Lower extremities (after exercise or prolonged standing). Tendency for skin to develop brownish pigmentation. Identical to idiopathic nonhyperglobulinemic syndrome, called Schamberg’s disease, except that the latter has normal serum globulin levels.</td>
</tr>
<tr>
<td>Cryoglobulinemic purpura</td>
<td>Purpura (+/- gangrene) on exposed acral areas (fingers, nose, ears, face).</td>
</tr>
<tr>
<td>Amyloid purpura</td>
<td>Spontaneous periorbital purpura (usually post Valsalva maneuver). “Touch purpura.”</td>
</tr>
<tr>
<td>Scorbutic purpura</td>
<td>Purpura around hair follicles (perifollicular petechiae). Characteristically associated with corkscrew hairs. Saddle distribution.</td>
</tr>
<tr>
<td>Senile purpura</td>
<td>Purple flat ecchymotic spots on extensor surface of forearms, dorsum of hands, and neck in the elderly. Identical lesions are found in cachetic states and chronic hypercortisonism.</td>
</tr>
</tbody>
</table>
| Embolic purpura                  | A. Septic embolic
  White centered petechial lesion often located on mucous membrane and conjunctivae (e.g., bacterial endocarditis).
  B. Fat emboli
  Petechiae limited to upper one-half of the body, particularly to anterior chest. Never seen on the face or back (skimming effect). |
| Palatine petechiae               | Infectious mononucleosis.
  Sepsis.
  Trauma (e.g., dentures). |
Pyuria and Bacteriuria

Petechiae (lesion ≤ 3 mm) Almost always indicates a disturbance of platelets or a vasculopathy. Rarely do they indicate an abnormality of coagulation.


PYURIA AND BACTERIURIJA

Does pyuria always signify the presence of bacteriuria? Is bacteriuria always associated with the presence of pyuria? The answer to both questions is “no.” Pyuria in the absence of bacteriuria can be seen:

1. In dehydration
2. In trauma
3. In the presence of an irritating agent in the renal pelvis, bladder, or ureter.
4. In renal tuberculosis
5. In acute and chronic glomerulonephritis
6. After administration of oral polio vaccine
7. After administration of intramuscular iron
8. In renal tubular acidosis
9. In association with a variety of viral infections
10. In Kawasaki disease

Listed below is a comparison between urine bacterial counts and leukocyte counts. Urines were collected by a clean voiding technique, and 5 ml was centrifuged for 3 minutes at 3000 RPM.

<table>
<thead>
<tr>
<th>COLONY COUNT (BACTERIA/ml)</th>
<th>UNCENTRIFUGED</th>
<th>CENTRIFUGED</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>10 or more WBC/mm³</td>
<td>100 or more WBC/mm³</td>
</tr>
<tr>
<td>10⁵</td>
<td>61%</td>
<td>23%</td>
</tr>
<tr>
<td>10⁴-10⁵</td>
<td>28%</td>
<td>0%</td>
</tr>
<tr>
<td>10³-10⁴</td>
<td>23%</td>
<td>0%</td>
</tr>
<tr>
<td>10²-10³</td>
<td>21%</td>
<td>1%</td>
</tr>
<tr>
<td>Sterile</td>
<td>10%</td>
<td>1%</td>
</tr>
</tbody>
</table>

WHAT SEASON OF "PROFILES ON PARADE" WOULD BE COMPLETE WITHOUT A VISIT FROM OUR OLD FRIEND, DR. DAN ASHER, TASTEMAKER TO THE MELLOW?

DAN'S LATEST HANDBOOK IS FOR ALL YOU BABY-BOOMERS WHO'VE FINALLY GOTTEN INTO CHILD-REARING, RIGHT, DOCTOR?

THAT'S RIGHT, MARK! I CALL IT, "THE MELLOW PARENT: SHARING YOUR SPACE WITH DEPENDENTS."

WHOA! SOUNDS LIKE ANOTHER TIMELY TITLE, DAN!

FOR SURE, MARK! WITH SO MANY UPSCALE FOLKS GETTING INTO PARENTING THESE DAYS, THERE'S JUST A BIG NEED FOR YOUR BASIC HOW-TO!
IN "THE MELLOW PARENT," I DISCUSS SUCH PROBLEMS AS LOOKING FOR A GOOD HOUSE-KEEPER, PREVENTING YOUR BABY FROM EATING YOUR FERNS, AND THE IMPORTANCE OF BUDGETING QUALITY TIME.

THAT'S A SMALL PART OF THE DAY YOU SET ASIDE TO SPEND WITH YOUR KID. IT DOESN'T HAVE TO BE MUCH, TEN MINUTES IS USUALLY ENOUGH, BUT IT HAS TO BE QUALITY! YOU CAN'T JUST WATCH T.V. — YOU HAVE TO FOCUS ON THE CHILD'S NEEDS.

WHAT'S QUALITY TIME, DAN?

WHAT IF ONE OF ITS NEEDS IS MORE TIME?

THAT'S QUANTITY TIME. I'M NOT REALLY TALKING ABOUT THE PROBLEM CHILD HERE, MARK.
QUESTIONS

Thirty Questions That Parents Ask and Pediatricians
Should Be Able to Answer

Listed below are questions frequently asked by parents. No one can have all the answers, but you should begin to learn some of them in your pediatric training. We have stratified the questions, with their answers, as a function of probable experience. If you have any questions about our answers, drop us a line.

Questions that a PIA should be able to answer:

1. If I chop up foods, such as hot dogs, very fine, is it safe to feed these to my 2-year-old?
   A. If they are chopped into very small pieces, they are safe; but it is important to remember that hot dog chunks are a common cause of aspiration in the 2-year-old. Another very dangerous food for any child under 3 years of age is peanuts. "Only a nut would feed a nut to a child." Instruct parents in the Heimlich maneuver.

2. Can shaking a baby or tossing him or her into the air playfully be harmful?
   A. Yes, both can be extremely harmful. Such play can injure the brain, neck, or spinal cord and has been known to produce retinal hemorrhages.

3. Is it true that it is harmful to swing or lift a child by holding his hands and pulling up on his arms?
   A. Yes, this can produce a "nursemaid's elbow"—which is a dislocation of the elbow.

4. My husband smokes in the house. Is this really a health danger to my young children?
   A. Absolutely. Smoking at home is known to aggravate asthma. There is a higher incidence of otitis media and other forms of respiratory illness among children living in a home with smokers. By the way, side-stream smoke also increases the non-smoking adult's risk for cancer. Get your husband to stop smoking or get rid of him.

5. I have a healthy 4-year-old. Should she receive the influenza vaccine?
   A. It is not necessary. The vaccine is advised for children with a variety of chronic illnesses.

6. Is it true that penicillin will cure the symptoms of a sore throat within 24 hours?
   A. Yes, but only if the symptoms are a result of an infection with the streptococcus microorganism. Without treatment, it will take 48 to 72 hours before symptoms subside.

7. Is it really possible for my son to have a streptococcal infection in his anal area?
   A. Definitely. It is characterized by pain and redness, usually without fever. Penicillin is the cure.
Questions

8. What is “croup”?
   A. Croup is the term applied to a condition that is characterized by a inspiratory noise called “stridor.” It is often accompanied by a cough that sounds like the bark of a seal. Croup can be caused by infections or allergic swelling in the larynx or “voice-box.”

9. Is it safe to give my 10-month-old daughter swimming lessons?
   A. Not a good idea. She can swallow a great deal of water while learning and that may cause convulsions. She will not really learn to swim at this age and does not have sufficient judgment to avoid water dangers when left alone.

10. Can you recommend a good book for me, a new parent, on child health?
    A. Yes. The best book on this subject is entitled Your Child’s Health, by Dr. Barton D. Schmitt and is published by Bantam Books and is available in paperback.

Questions that a PL-2 should be able to answer:

1. My 16-year-old daughter often eats several glasses of ice cubes each day. Is there something wrong with her?
   A. Your daughter suffers from a condition termed “pagophagia” or ice-eating. “Pagos” is the Greek word for “ice” and “phagia” in Greek means “to eat.” Compulsive ice-eating is usually a sign of iron deficiency and can often be promptly cured with iron therapy.

2. Can the rash from poison ivy be spread by exposure to the fluid from popped blisters?
   A. No. Poison ivy results from a hypersensitivity reaction to the plant’s resins. Transmission only occurs by direct contact with the plant or with the hands, clothing, or pets that have resin on them. The blister fluid does not contain the resin.

3. My daughter is almost 3 and still not toilet trained. What am I doing wrong?
   A. Nothing. Toilet training should not begin until your child is ready. Readiness means that your child has the neurologic capacity to control bodily functions, understands the concepts of toileting, knows the language of the toilet, and, most importantly, wants to be clean and dry. This process may start as early as 1 year of age. I suspect that your child will train herself very soon.

4. We have a kitten at home. Can a kitten or cat cause any health problems for my 2-year-old son?
   A. Yes, definitely yes. Diseases and parasites such as cat scratch fever, toxoplasmosis, and Toxocara cati can be spread from a cat or kitten. Cat dander is often responsible for asthma. Now that you have a child you don’t need any other pet. Get rid of the cat.

5. Is it safe to allow my son to play football with his high school team?
   A. It depends on the status of his physical maturity relative to others his own age, the protective equipment available, and the concern of the coach for the emotional and physical welfare of the members of the team.
6. My 5-year-old periodically becomes hoarse. What should I do?
   A. Get some ear plugs for yourself. The episodes of hoarseness are probably a consequence of excessive screaming on your child's part. "Screecher's nodules"—actual thickening of the vocal cords—can result from overuse of the voice. If hoarseness persists, your child should be seen by an otolaryngologist.

7. My child has pink urine after eating beets. What does this mean?
   A. About 7% of the population will demonstrate "beeturia" after eating the equivalent of one beet. This is a genetic trait. Beeturia is also seen in a very high percentage of children and adults with iron deficiency anemia. This form of beeturia disappears with iron therapy. To be certain of the cause of your child's problem, a blood count should be obtained.

8. Is it appropriate for a young child to attend the funeral of his grandparent?
   A. If he wants to go, then you should let him. Never force a child, of any age, to attend a viewing or a funeral.

9. Does an exclusively breast-fed infant require supplemental iron?
   A. Not during the first 6 months of life. Studies have shown that the iron status of an exclusively breast-fed infant is as good as an infant receiving an iron-fortified formula at age 6 months. If breast milk remains the sole source of nutrition beyond 6 months of age, iron supplements are indicated, because studies have shown that about 15% of such infants will be iron deficient by 9 months of age.

10. What kind of shoes should I buy for my baby when she starts walking?
    A. The least expensive sneakers you can find. Shoes are only necessary for the protection of the feet from the dirt, glass, and manure of the streets and are not required to enhance the act of locomotion.

**Questions that a PL-3 should be able to answer:**

1. My 2-year-old is a finnicky eater and I'm not sure she's eating all the foods she needs. Should I supplement her diet with vitamins?
   A. No, they are unnecessary. Put a balanced meal before the child and she will take what she needs. Vitamins are only "fish-food" for a person eating a balanced diet. The excess vitamins you provide will appear in the child's urine, and when they are flushed down the toilet they will eventually reach the fish in the sea.

2. I'm a working mother with three young children. Not surprisingly, I often come home tired. Can you give me some advice to make returning home after a tough day better for all of us?
   A. There are no easy answers for a task that stumps even "superwoman." But these suggestions may help:

   - When you pick up your child from day care, bring a snack for the trip home.
   - Take turns with your spouse leaving work early, so one of you can pick up your child before you are both exhausted.
• Establish a homecoming routine that gives you recovery time. Tell your children that they will get your full attention after you have a chance to catch your breath and get dinner started.
• Reward your children for good behavior. Try a chart with gold stars or a special picture to color.
• Expect your school-age children to begin their homework after school and before you arrive.
• Reduce meal preparation during the week by cooking on weekends and freezing the servings until you need them.
• If you can afford it, hire a high school student as a mother's helper for that first hour at home.
• Involve the children with dinner chores, such as setting the table, clearing the table, and the like.
• Share the evening tasks with your spouse.
• If possible, take the family out to dinner occasionally to give everyone a break.

3. Should I give my child aspirin?
   A. No, except under a physician's direction for some very special circumstances, such as Kawasaki's disease or acute rheumatic fever.

4. Does it help to put butter on burns?
   A. The best thing to put on a burn immediately is ice. The ice will relieve the pain and reduce the damage done by the burn. Topical vitamin E is much better than butter in reducing the scarring and damage done by a small burn.

5. Can iron deficiency anemia produce any harm in a 1-year-old?
   A. Yes. Recent studies demonstrate that iron deficiency anemia results in cognitive delays in 1- and 2-year-olds that may not be correctable with iron therapy.

6. When can you begin giving a child an allowance?
   A. When you can afford it and when the child has some appreciation of the value of money and when you have agreed upon the fact that the allowance is dependent on the satisfactory performance of some responsibilities. This usually begins at age 8-9.

7. Can fruit juice cause stomach aches or diarrhea in my 6-year-old?
   A. Yes, if they consume large quantities of juice and if the juice contains non-absorbable sugars such as sorbitol. Apple juice, for example, contains large quantities of sorbitol.

8. Can a child have an ear infection without a fever?
   A. Yes. About one-half of episodes of otitis media are not associated with temperature elevation beyond 100.4°

9. Our 12-year-old son wants an All-Terrain vehicle. Are they safe?
   A. No. Don't buy it either for him or for you.

10. My teenage daughter says that kissing is good exercise. Is this true?
    A. Not if her goal is to lose weight. It is estimated that a single kiss burns up 9 calories and that by kissing three times a day for 1 year, she could lose 2.8 pounds.
RED CELL

Red Cell Distribution Width

Age-appropriate Values for RBC Distribution Width

<table>
<thead>
<tr>
<th>AGE</th>
<th>NO. OF PATIENTS</th>
<th>RBC DISTRIBUTION WIDTH (MEAN ± SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-6 mo</td>
<td>68</td>
<td>13.0 ± 1.5</td>
</tr>
<tr>
<td>7-12 mo</td>
<td>84</td>
<td>13.7 ± 0.9</td>
</tr>
<tr>
<td>13-24 mo</td>
<td>108</td>
<td>13.4 ± 1.0</td>
</tr>
<tr>
<td>2-3 yr</td>
<td>119</td>
<td>13.2 ± 0.8</td>
</tr>
<tr>
<td>4-5 yr</td>
<td>151</td>
<td>12.7 ± 0.9</td>
</tr>
<tr>
<td>6-8 yr</td>
<td>106</td>
<td>12.6 ± 0.8</td>
</tr>
<tr>
<td>9-11 yr</td>
<td>98</td>
<td>12.8 ± 1.0</td>
</tr>
</tbody>
</table>


RENAL FAILURE

The FENa Test: Use in the Differential Diagnosis of Acute Renal Failure

The physician is frequently faced with the problem of distinguishing prerenal azotemia from acute tubular necrosis in patients with acute renal failure.

In the oliguric phase of these two conditions, the renal tubule handles sodium in distinctly different fashions. In prerenal azotemia, the renal tubule avidly reabsorbs the filtered sodium; in acute tubular necrosis, the reabsorption of sodium is restricted.

These observations provide the basis for a simple test for differentiating these two conditions—the "FENa test" (FENa is the excreted fraction of the filtered sodium).

The test is performed by measuring both sodium and creatinine in simultaneously collected samples of plasma and urine.

The FENa is calculated as follows:

\[
\frac{[\text{Sodium}]_U}{[\text{Sodium}]_P} \times \frac{[\text{Creatinine}]_U}{[\text{Creatinine}]_P} \times 100
\]

U and P represent concentrations in urine and plasma, respectively.
In general, an FE\textsubscript{Na} of less than 1 indicates prerenal azotemia, and an FE\textsubscript{Na} of more than 3 indicates acute tubular necrosis.


**RESPIRATORY DISTRESS**

The Diagnosis of Newborns with Acute Respiratory Distress

The newborn with clinical signs of acute respiratory distress—central cyanosis, tachypnea (>60 breaths/min), tachycardia (>160 beats/min), retractions, grunting, and nasal flaring—demands immediate attention. A review of the maternal history, the age of onset of respiratory distress, a physical examination, and laboratory tests (particularly a chest x-ray) will greatly aid the clinician in assessing this problem further. The following differential diagnosis should be of use:

1. **Upper airway obstruction**
   - Choanal atresia
   - Masses (encephalocele, tumor)
   - Macroglossia
   - Nasal stiffness
   - Cleft palate
   - Laryngeal obstruction (paralysis, web, tumor, stenosis, atresia, malacia)
   - Tracheal obstruction (mass, web, stenosis, atresia, malacia, cleft, vascular ring, goiter)

2. **Pulmonary**
   - Hyaline membrane disease
   - Transient tachypnea of the newborn
   - Aspiration of meconium, gastric, or amniotic fluid
   - Pneumonia
   - Pneumothorax, pneumomediastinum
   - Persistent pulmonary hypertension
   - Tracheoesophageal fistula
   - Pulmonary hemorrhage
   - Hypoplasia or agenesis of the lungs
   - Cystic disease (emphysema, cysts)
   - Pleural effusions (e.g., chylothorax)
   - Pulmonary sequestrations

3. **Cardiac**
   - Cyanotic congenital heart disease
   - A cyanotic congenital heart disease
   - Arrhythmia (paroxysmal supraventricular tachycardia, block)
   - Increased intravascular volume (iatrogenic fluid overload)
   - High output failure (hyperthyroidism, arterial-venous malformation)
   - Pneumopericardium
   - Cardiomyopathy (infection, endocardial fibroelastosis, hypertrophic cardiomyopathy)

4. **Thoracic**
   - Chest wall deformities (chondrodystrophies, rib deformities)
   - Masses (tumors, cysts)

5. **Metabolic**
   - Hypoglycemia
   - Infant of a diabetic mother
   - Inborn errors of metabolism

6. **Diaphragmatic**
   - Hernia (foramen of Bochdalek)
   - Paralysis (phrenic nerve)
   - Eventration
7. Neuromuscular
- CNS damage (trauma, hemorrhage)
- Medication (maternal sedation, narcotic withdrawal)
- Muscular weakness (e.g., myasthenia gravis)
- Congenital defects

8. Infections
- Sepsis
- Pneumonia (viral, bacterial)

9. Hematologic/vascular
- Hyperviscosity, hypervolemia
- Anemia
- Hemoglobinopathy

10. Other
- Asphyxia
- Acidosis
- Hypothermia
- Hyperthermia


RESPIRATORY VIRUSES

Clinical Syndromes Produced by Respiratory Viruses

Upper respiratory tract infections and pneumonia are among the most frequently made diagnoses in pediatric clinics and emergency rooms, particularly during the winter months. All too frequently, these infections are lumped together as upper respiratory infections (URI), viral syndrome, or "influenza-like" syndrome. Many viruses, aside from influenza, are capable of producing respiratory symptoms.

Clinical Syndromes Produced by Respiratory Viruses

<table>
<thead>
<tr>
<th>VIRUSES</th>
<th>CORYZA</th>
<th>PHARYNGITIS</th>
<th>CROUP</th>
<th>FLU-LIKE ILLNESS</th>
<th>FEVER</th>
<th>PNEUMONIA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Influenza A</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
</tr>
<tr>
<td>Influenza B</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Influenza C</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Parainfluenza (13)</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Respiratory syncytial</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
</tr>
<tr>
<td>Coxsackie A</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Coxsackie B</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Echo</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Adenoviruses (17, 14, 21)</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Rhinoviruses (&gt;100)</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
</tr>
<tr>
<td>Coronavirus</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Herpesviruses (1, 2)</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
<tr>
<td>Epstein-Barr</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td>⬤</td>
<td></td>
<td>⬤</td>
</tr>
</tbody>
</table>

* In children only
292—Respiratory Virus

The term influenza, incidentally, originated in Italy during the 15th century. A particularly severe epidemic of a respiratory viral syndrome at that time was attributed to the influenza (influence) of the stars and evil forces.


The Clinical Manifestations of Respiratory Syncytial Virus

Respiratory syncytial virus (RSV) is an all too common cause of epidemic winter respiratory disease in infants, children, and adults. Its severity spans a wide spectrum ranging from a common cold to bronchiolitis (the most common presentation) to respiratory distress, apnea, cyanosis, and pneumonia. RSV is potentially most life-threatening in infants with other underlying conditions such as congenital heart disease (especially conditions with high pulmonary flow), bronchopulmonary dysplasia, other chronic irreversible pulmonary disorders (e.g., cystic fibrosis, pulmonary hemosiderosis, bronchiolitis obliterans, and idiopathic pulmonary hypertension), and all children on immunosuppressive regimens (e.g., transplantation patients, cancer patients treated with chemotherapy or radiation therapy, and patients on high-dose steroid regimens) or children with immunodeficiency syndromes (e.g., severe combined immune deficiency).

Although bronchiolitis is the most frequent presentation of an RSV infection, pneumonia is the most common admitting diagnosis for infants infected with RSV who require hospitalization. Factors that contribute to the severity of illness, in addition to those described above, include age, size of inoculum, and characteristics of an RSV infection:

Common characteristics (> 75% of all cases)
- Rhinorrhea
- Cough
- Airway hyperactivity
- Hypoxemia
- Air trapping (hyperinflated lungs on x-ray examination)

Uncommon Characteristics (< 20% of all cases)
- Prolonged fever (T > 102°F)
- Otitis media
- Hoarseness
- Pleural effusion
- Hepatosplenomegaly
- Enlarged cardiac silhouette
- Hilar adenopathy

The differential diagnosis for RSV pneumonia should include:
1. Chlamydial pneumonia as it occurs in the same age group and presents with air trapping and wheezing. The distinguishing point between these two diseases is that chlamydial disease is more prolonged and insidious than RSV.
2. Congenital anomalies of the respiratory tract
3. Foreign bodies
4. Reactive airway disease
5. Infection with other respiratory agents (e.g., influenza virus, parainfluenza viruses, adenovirus, pertussis (*Bordetella*), or mycoplasma).


**RETROPHARYNX**

**Retropharyngeal Abscess**

The retropharyngeal space extends from the base of the skull to about the level of the second thoracic vertebra. Abscess in this space results from suppuration of the lymph nodes, which run in two parallel chains and drain the nasopharynx, adenoids, and posterior paranasal sinuses. During childhood these nodes are prominent, but they atrophy during adolescence. Though trauma and adjacent vertebral osteomyelitis can predispose to the development of retropharyngeal abscess, local respiratory tract infection is usually felt to be the initiating event. Extension of the infection may result in mediastinitis or asphyxia due to increasing pressure or rupture of the abscess.

Both aerobic and anaerobic bacteria may be isolated from retropharyngeal abscesses, and more than one organism is often found. The frequent isolation of *Staphylococcus aureus* and beta-lactamase-producing anaerobes warrants the use of antibiotics effective against penicillin-resistant oropharyngeal flora. Surgical drainage of the abscess is critical.

The 63 bacteria isolated from 17 children with retropharyngeal abscess are listed below:

<table>
<thead>
<tr>
<th>Bacteria Isolated from Children with Retropharyngeal Abscess</th>
</tr>
</thead>
<tbody>
<tr>
<td>ISOLATES</td>
</tr>
<tr>
<td>----------</td>
</tr>
<tr>
<td>Aerobic and facultative Gram-positive cocci</td>
</tr>
<tr>
<td>Viridans streptococci</td>
</tr>
<tr>
<td><em>Staphylococcus aureus</em></td>
</tr>
<tr>
<td>Beta-hemolytic <em>Streptococcus</em> Group A</td>
</tr>
<tr>
<td><em>Streptococcus pneumoniae</em></td>
</tr>
<tr>
<td><em>Streptococcus constellatus</em></td>
</tr>
<tr>
<td><em>Streptococcus morbillorum</em></td>
</tr>
<tr>
<td>Micrococcus</td>
</tr>
<tr>
<td>Gram-negative cocci</td>
</tr>
<tr>
<td><em>Neisseria</em> sp.</td>
</tr>
<tr>
<td>Gram-negative bacilli</td>
</tr>
<tr>
<td><em>Eikenella corrodens</em></td>
</tr>
<tr>
<td><em>Haemophilus influenzae</em> (nontypable)</td>
</tr>
<tr>
<td><em>Haemophilus parainfluenzae</em></td>
</tr>
<tr>
<td>Total no. of aerobes</td>
</tr>
</tbody>
</table>

Table continued on next page.
Bacteria Isolated from Children with Retropharyngeal Abscess (Cont.)

<table>
<thead>
<tr>
<th>ISOLATES</th>
<th>NO. OF ISOLATES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anaerobic Anaerobic cocci</td>
<td></td>
</tr>
<tr>
<td><em>Peptostreptococcus</em> species</td>
<td>3</td>
</tr>
<tr>
<td><em>Veillonella parvula</em></td>
<td>1</td>
</tr>
<tr>
<td>Microaerophilic streptoccus</td>
<td>1</td>
</tr>
<tr>
<td>Gram-positive bacilli</td>
<td></td>
</tr>
<tr>
<td><em>Eubacterium lentum</em></td>
<td>2</td>
</tr>
<tr>
<td>Gram-negative bacilli</td>
<td></td>
</tr>
<tr>
<td><em>Bacteroides melaninogenicus</em></td>
<td>7</td>
</tr>
<tr>
<td><em>Bacteroides capillosus</em></td>
<td>2</td>
</tr>
<tr>
<td><em>Bacteroides</em> species</td>
<td>1</td>
</tr>
<tr>
<td><em>Fusobacterium</em> species</td>
<td>1</td>
</tr>
<tr>
<td>Total no. of anaerobes</td>
<td>18</td>
</tr>
</tbody>
</table>


RETT SYNDROME

The Diagnosis of Rett Syndrome

The diagnosis of Rett syndrome, a severe developmental disorder occurring in young girls, usually between 6 and 18 months of age, can be difficult in view of the fact that it is dependent on history and physical findings alone. There is no laboratory test to confirm the diagnosis. These children experience rapid decline in motor and cognitive function after a period of apparently normal development. In those children who have begun to speak, all meaningful communication is lost, including eye contact. The patients often experience interrupted sleep and periods of uncontrollable screaming. Prevalence is between 1/10,000 and 1/15,000.

The implications of this diagnosis are tragic for both the patient and family. The criteria that can be used to support or exclude the diagnosis are listed below:

1. **Necessary criteria**
   - Normal prenatal and perinatal period
   - Apparently normal development first 6 mo
   - Normal head circumference at birth
   - Deceleration of head growth between 5 mo and 4 yr
   - Loss of purposeful hand skills between 6 mo and 30 mo; communication dysfunction; social withdrawal
   - Stereotypic hand movements
   - Gait apraxia and truncal ataxia between 1 to 4 yr
   - Diagnosis tentative until 2 to 5 yr of age
2. Supportive Criteria

Breathing dysfunction  
EEG abnormalities  
Seizures  
Spasticity  
Peripheral vasomotor disturbances  
Scoliosis  
Growth retardation  
Hypotrophic small feet

3. Exclusion Criteria

Intrauterine growth retardation  
Organomegaly—signs of storage disease  
Retinopathy/optic atrophy  
Microcephaly at birth  
Evidence of perinatally acquired brain damage  
Identifiable metabolic disorder  
Evidence of serious CNS infection or trauma

RHINITIS

Diagnosis and Natural History of Allergic Rhinitis

Allergic rhinitis, which is ranked by the National Center for Health Statistics as the sixth most prevalent chronic condition in the U.S., has its peak incidence in childhood and adolescence. It is an atopic hypersensitivity response to foreign allergens mediated by IgE antibodies, but not all persons with IgE antibody have clinical disease. The most common allergens are the following:

Grass pollens (late spring/early summer)  
Tree pollens (early spring)  
Weed pollens (late summer/autumn)  
Animal danders  
House-dust mites  
Insects  
Mold spores  
Foods (uncommonly associated)

Symptoms include paroxysmal sneezing; watery, profuse rhinorrhea; nasal congestion (stuffy nose); itching of the nose and eyes; and lacrimation and ocular redness. Other symptoms that can occur are noisy breathing, snoring, hyposmia or anosmia, itchy palate or pharynx, throat clearing, and cough. Children may have “allergic shiners,” a dark discoloration in the infraorbital regions secondary to obstruction of venous drainage. The key to diagnosis is the temporal correlation of symptoms with allergic exposure.

As with most allergies, avoidance of the offending allergens is the most effective treatment, which makes identification of the allergens an important component of the therapeutic strategy. However, many of the methods commonly used to diagnose allergies are only minimally helpful in managing allergic rhinitis, including early and late skin-test responses, measurement of IgE levels, and calculation of histamine release by basophils. The natural history of allergic rhinitis is presented in the schematic of sensitization (Phase I) and clinical disease (Phase 2) shown on the following page.
Simplified schematic representation of the natural history of allergic rhinitis. During phase 1 persons become sensitized to an allergen, and during phase 2 clinical disease develops. The overwhelming majority of patients have an early response on reexposure to allergen. The early response is dominated by activation of mast cells and release of mediators. After the early response, most patients have cellular infiltration of the nasal mucosa that causes late inflammatory events. These include the spontaneous recurrence of release of mediators (late-phase reaction), hyperresponsiveness to irritants, and increased responsiveness to allergen (priming). The circles indicate the heterogeneity of these late inflammatory events. The inflammation can resolve spontaneously, cause a complication, or potentially lead to an irreversible form of chronic rhinitis. (From Naclerio RM, N Engl J Med, 325: 861, 1991, with permission.)

Persistent Rhinitis in the Newborn

Because neonates are often obligate nose breathers, nasal congestion and rhinorrhea may be a difficult problem. The causes of persistent rhinitis in the newborn are listed below along with the treatment of each type.

**Rhinitis in the Newborn**

<table>
<thead>
<tr>
<th>ENTITY</th>
<th>CAUSE</th>
<th>TREATMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transient idiopathic stuffy nose of the newborn</td>
<td>Unknown</td>
<td>Normal saline nosedrops may be instilled and then removed after a few minutes with cotton-tipped applicators or gentle suction on a rubber bulb syringe. If the congestion interferes with feeding, 2 drops of 0.125% phenylephrine (Neo-Synephrine) may be instilled in the nose just before meals for several days.</td>
</tr>
<tr>
<td>Chemical rhinitis</td>
<td>Due to overtrement of idiopathic stuffy nose with topical nasoconstrictors</td>
<td>Discontinue nosedrops. Use oral decongestants 1-2 days.</td>
</tr>
<tr>
<td>Pyogenic rhinitis</td>
<td>These infants have bacterial infection despite absence of purulent discharge. Diagnose via cultures of discharge</td>
<td>Same as for idiopathic stuffy nose.</td>
</tr>
<tr>
<td>Congenital syphilis</td>
<td>Maternal syphilis</td>
<td>Penicillin.</td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td>Congenital hypothyroidism</td>
<td>Thyroid hormone replacement.</td>
</tr>
<tr>
<td>Choanal atresia</td>
<td>Congenital defect</td>
<td>Place oral airway immediately. Definitive surgery by otolaryngologist.</td>
</tr>
<tr>
<td>Nasal fracture</td>
<td>Birth trauma</td>
<td>Diagnose by examination for subluxation of the nasal septum causing occlusion of the nasal passages. Refer to otolaryngologist.</td>
</tr>
</tbody>
</table>


**RHYTHMIC BEHAVIORS**

**Rocking and Rolling—And Head Banging**

Body rocking, head banging, and head rolling are three rhythmic behaviors that may show up in normal infants between 6 and 10 months of age and may last up to 18 months. Head banging is the most upsetting to parents, who often consult pediatricians because of concern about self-injury, as well as the disruption to the household, often in the middle of the night. Neighbors also have been known to urge intervention.
The average banger is a male (about 3 to 1), usually awake and in bed, usually banging against the headboard, and usually not crying or showing any evidence of temper. Some seem exceptionally relaxed and even blissful during the activity.

The most common positions for head banging were described by de Lissevoy and further discussed by Hoder and Cohen:

1. **The hands and knees position**, in which the child stands on hands and knees and rocks back and forth; on the forward motion the forehead or cranial cap is struck against the crib.
2. **The sitting position**, in which the child is braced or sitting against the side of the crib or the head board. The knees are drawn up or the legs may be straight out; the arms and hands serve to brace the body in motion. The motion is mainly a trunk movement, or it is limited to throwing the head repeatedly to the rear, striking the crib.
3. **The prone position**, in which the child is lying prone; the head is raised and then dropped on the pillow or mattress or brought down with considerable force.
4. **Multiple positions**, in which the child kneels, stands, or sits as he holds onto the bars or the railing of the crib while striking his forehead.
5. **The supine position**, in which, while supine, the child rolls either his head or his whole body from side to side with the head striking the sides of the crib.

In most cases these patterns of motor behavior are transient and resolve spontaneously. Parents should be reassured that no brain damage will result.


**RICKETS**

**Origin of the Name**

Most scholars believe that the term is derived from the Greek work "rachitis" (a disease of the spine), and hence the use of the medical synonym rachitis for "rickets." Professor H. A. Skinner felt the term originated from the Anglo-Saxon term "wricken" (to twist). A 17th century writer named John Aubrey added still another "twist" to this story when he wrote:

I will whilst 'tis in my mind insert this Remarque, viz., about 1620 one Ricketts of Newberye, a Practitioner in Physick, was excellent at the Curing of Children with swoln heads, and small legges; and the Disease being new, and without a name, He being so famous for the cure of it, they called the Disease the Ricketts ... and now 'tis good sport to see how they vex their lexicons, and fetch it from the Greek.

At any rate, when trying to look erudite on the wards, be careful not to confuse the 17th century "Dr. Ricketts" with Howard Taylor Ricketts (1871-1910), the American pathologist who in 1906 discovered the etiology of Rocky Mountain spotted fever and other typhus-like diseases. These microorganisms are designated the genus *Rickettsia* and the family Rickettsiaceae in his honor.

The Three-Stage Chemical Evolution of Rickets

Rickets—or avitaminosis D—is a defect in the mineralization of the growing skeleton, including bone and the cartilage of the growth plate. The three stages of the chemical evolution of the disease are as follows:

**Stage 1 (Intestinal Calcium Transport Decreased)**

- Serum calcium decreased
- Serum phosphorus normal
- Serum alkaline phosphatase normal

**Stage 2 (Compensatory Hyperparathyroidism)**

- Serum calcium normal
- Serum phosphorus decreased
- Serum alkaline phosphatase increased
- Serum bicarbonate decreased
- Serum chloride increased
- Aminoaciduria
- X-ray—active rickets

**Stage 3 (Parathyroid Response No Longer Sustains Normal Serum Calcium)**

- Serum calcium decreased
- Serum phosphorus decreased
- Serum alkaline phosphatase increased
- Serum bicarbonate decreased
- Serum chloride increased
- Aminoaciduria
- X-ray—florid rickets
- Tetany may occur


**Causes of Rickets**

<table>
<thead>
<tr>
<th>Cause</th>
<th>Source of Deficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Diet</strong></td>
<td></td>
</tr>
<tr>
<td>Calcium deficiency</td>
<td>Low intake</td>
</tr>
<tr>
<td>High phytin content (e.g., soy formula)</td>
<td>Malabsorption</td>
</tr>
<tr>
<td>Inadequate sunlight and vitamin D supplementation</td>
<td>Malabsorption</td>
</tr>
<tr>
<td><strong>Medications</strong></td>
<td></td>
</tr>
<tr>
<td>Antacids</td>
<td>Malabsorption</td>
</tr>
<tr>
<td>Furosemide</td>
<td>Excretion</td>
</tr>
<tr>
<td>Anticonvulsants (phenytoin or phenobarbital)</td>
<td>Malabsorption</td>
</tr>
<tr>
<td><strong>Prematurity</strong></td>
<td></td>
</tr>
<tr>
<td>Inadequate calcium intake</td>
<td>Low intake</td>
</tr>
<tr>
<td>Inadequate phosphate intake</td>
<td>Low intake</td>
</tr>
<tr>
<td>Vitamin D deficiency</td>
<td>Malabsorption</td>
</tr>
<tr>
<td>Increased requirement (suspected but not proved)</td>
<td>Malabsorption</td>
</tr>
</tbody>
</table>
300—Rickets

### Cause of Calcitriol Deficiency*

<table>
<thead>
<tr>
<th>Cause</th>
<th>Source of Deficiency</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insufficient UV exposure and inadequate vitamin D supplementation</td>
<td>Malabsorption of supplemental vitamin D in steatorrhea (acholic or celiac)</td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>

---

*Calcitriol deficiency leads to inadequate calcium and phosphate reabsorption.


---

Children at High Risk for Rickets

- Small premature infants
- Urban breast-fed infants who do not receive supplemental vitamin D
- Children with chronic renal insufficiency
- Children with biliary atresia or liver disease

Radiographic Findings in Rickets

The radiographic signs of rickets are the same regardless of the disorder responsible for undermineralization.

**Knees and wrists**
- Epiphyseal centers are indistinct or visible
- Metaphyseal zones of provisional calcification have faint, irregular outlines
- Increased distance from the visible mineralized portion of the shafts to the epiphyseal centers is apparent
- Ends of ulna and fibia are concave
- Ends of bones are widened
- In severe rickets, density of the bone shafts is reduced

**Chest**
- Ends of ribs are expanded, cupped, indistinct, and appear farther than usual from the sternum
- Proximal humeri show changes listed for knees and wrists but lesser in degree because linear growth is slower

**When rickets heals**
- Supernormal amounts of mineral, visible as dense transverse bands, appear in the formerly deficient zones of provisional calcification
- Dense lines may also appear in subperiosteal osteoid parallel to the bone shafts and can be misinterpreted as evidence of trauma


### Diagnosis and Management of Rickets

<table>
<thead>
<tr>
<th>CAUSE</th>
<th>DIAGNOSTIC TOOLS</th>
<th>MANAGEMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Calcium deficiency</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Low intake</td>
<td>History</td>
<td>Modify diet to include at least 500 mg/d of Ca</td>
</tr>
<tr>
<td>Marginal intake + excess phytin</td>
<td>History</td>
<td>Adjust intake to 200 mg/kg/d</td>
</tr>
<tr>
<td>Extreme prematurity (birth weight &lt; 1,500 g)</td>
<td>Stool fat, Serum 25-OH-D, low</td>
<td>25-OH-D, (5-7 µg/kg/d), if serum level is low, supplement dietary Ca</td>
</tr>
<tr>
<td>Steatorrhea</td>
<td></td>
<td>Vitamin D 1,000 2,000 IU/d</td>
</tr>
<tr>
<td>Anticonvulsants (phenobarbital or phenytoin)</td>
<td>History</td>
<td></td>
</tr>
<tr>
<td>Furosemide</td>
<td>Hypercalcemia (urine Ca/Cr &gt; 0.2)</td>
<td>Base supplement: 3 10 mM/kg/d as NaHCO₃ or citrate</td>
</tr>
<tr>
<td>Renal tubular acidosis</td>
<td>Serum CO₂ low, Urine pH 6.0 or above, Hypercalcemia (urine Ca/Cr &gt; 0.2)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Vitamin D deficiency</th>
<th>History</th>
<th>400 IU/d of vitamin D</th>
</tr>
</thead>
<tbody>
<tr>
<td>Insufficient UV light</td>
<td>Low 25-OH-D</td>
<td></td>
</tr>
<tr>
<td>No vitamin D supplement</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Table continued on next page.*
<table>
<thead>
<tr>
<th>CAUSE</th>
<th>DIAGNOSTIC TOOLS</th>
<th>MANAGEMENT</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitamin D deficiency (Cont.)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Liver disease</td>
<td>Low 25-OH-D$_3$</td>
<td>25-OH-D 5-7 μg/kg/d$^{12}$</td>
</tr>
<tr>
<td>Renal disorders may reduce calcitriol formation: Hypoplasia or parenchymal damage</td>
<td>BUN or Cr high</td>
<td>Calcitriol 0.25-1.0 μg/d</td>
</tr>
<tr>
<td>Liver disease</td>
<td>Serum P usually high</td>
<td>CaCO$_3$ to restrict P absorption and supplement dietary Ca$^{13}$</td>
</tr>
<tr>
<td>Renal disorders may reduce calcitriol formation: Hypoplasia or parenchymal damage</td>
<td>Serum Ca low (may be normal in secondary hyperparathyroidism) Alkaline phosphatase high</td>
<td>Restrict milk and protein sources to lower P load</td>
</tr>
<tr>
<td>Specific hydroxylase deficiency</td>
<td>Chemical results of Ca deficiency</td>
<td>Calcitriol 0.5-1.0 μg/d</td>
</tr>
<tr>
<td>Vitamin D present but ineffective</td>
<td>25-OH-D$_3$ normal Calcitriol low</td>
<td></td>
</tr>
<tr>
<td>Receptor defect</td>
<td>Phenotype (alopecia)</td>
<td>Calcitriol 10-30 μg/d</td>
</tr>
<tr>
<td>Nuclear translation defect</td>
<td>Chemical results of Ca deficiency High calcitriol</td>
<td>Parenteral Ca 1 g/d</td>
</tr>
<tr>
<td>Skin fibroblast cultures to differentiate receptor from nuclear translation defects</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Phosphorus deficiency</td>
<td>Low serum P</td>
<td>Adjust formula or parenteral source to give 100 mg/kg/d</td>
</tr>
<tr>
<td>Diet (limited to very premature infants)</td>
<td>Radiographic signs of rickets</td>
<td>Alternative gastric HCl control (e.g., cimetidine)</td>
</tr>
<tr>
<td>Antacid excess (aluminum hydroxide)</td>
<td>History Low serum P</td>
<td>Supplement P and calcitriol$^{14}$ if low</td>
</tr>
<tr>
<td>Excessive phosphaturia from tubular dysfunction (calcitriol formation may also be deficient)</td>
<td>History</td>
<td></td>
</tr>
<tr>
<td>Isolated, X-linked normocalciuric (common)</td>
<td>Urine Ca/Cr normal Calcitriol normal</td>
<td>Supplement P and calcitriol</td>
</tr>
<tr>
<td>Isolated, recessive hypercalciuric (very rare?)</td>
<td>History</td>
<td>Supplement P</td>
</tr>
<tr>
<td>With acidosis, glucosuria, and aminoaciduria alone (Fanconi syndrome) or the result of metal poisoning, fructose intolerance, tyrosinemia, galactosemia, cystinosis, or Wilson's disease</td>
<td>Urine Ca/Cr high Calcitriol high</td>
<td>Supplement alkali, P and calcitriol as indicated by serum analysis</td>
</tr>
<tr>
<td>Fanconi syndrome plus cerebral and eye defects (Lowe syndrome)</td>
<td>History</td>
<td>Same as for Fanconi syndrome</td>
</tr>
<tr>
<td>Physical findings</td>
<td>Serum and urine analysis (same as for Fanconi syndrome)</td>
<td></td>
</tr>
<tr>
<td>Tumors</td>
<td>Phosphaturia</td>
<td>Excision; if not feasible, calcitriol and P supplement</td>
</tr>
<tr>
<td>Mesenchymal$^9$</td>
<td>Calcitriol low (mesenchymal tumors may be small and cryptic)</td>
<td></td>
</tr>
<tr>
<td>Sebaceous nevi$^{10}$</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurofibromatosis</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Key: BUN = blood urea nitrogen; CA = calcium; CaCO$_3$ = calcium carbonate; Cr = Creatinine; NaHCO$_3$ = sodium bicarbonate; P = phosphorus; 25-OH-D$_3$ = 25 hydroxyvitamin D; UV = ultraviolet.

The Infant and Child with Acute Scrotum

The presentation of an infant or child with an enlarged, tender, and discolored scrotum is generally a call for alarm, both on the part of the patient and the physician. It is essential to diagnose quickly and correctly the patient with an acute scrotum to avoid irreversible testicular injury. This is particularly the case with the most common cause of acute scrotum in children, torsion of the testicle.

Many urologists recommend testicular radionucleotide scans and Doppler and scrotal ultrasound examinations to confirm the diagnosis of the acute scrotum. A simpler means to differentiate torsion from less threatening problems such as epididymitis and orchitis, however, is to test for the cremasteric reflex—the reflex is almost always absent in a patient with torsion (see also p. 304).

Causes of the Acute Scrotum

1. Testicular torsion
   a. Extravaginal torsion is found exclusively in neonates. It is caused by a twisting of the spermatic cord above the tunica vaginalis, thus cutting off the blood supply. Transillumination can be useful in distinguishing extravaginal torsion from hydrocele.
   b. Intravaginal torsion is a twisting of the testes within the tunica vaginalis. It can occur anytime in life but is most common in the prepubertal and postpubertal male.
2. Epididymitis
3. Torsion of testicular appendages yields localized tenderness at the upper pole of the testis or epididymis. A “blue-dot” sign is pathognomonic of this entity, representing the necrotic appendage beneath the skin.
4. Orchitis
5. Strangulated inguinal hernia
6. Idiopathic scrotal edema
7. Henoch-Schönlein purpura
8. Tumor
9. Trauma
10. Extrascrotal disease (e.g., intraabdominal sepsis and formation of a pyocele).

The Importance of the Cremasteric Reflex in Acute Scrotal Swelling in Children

The importance of accurate and rapid diagnosis of the acute scrotum cannot be overemphasized. There exists a wide variety of diagnostic modalities that have been reported to improve assessment and dictate which patient should undergo surgical exploration. The most valuable aid in differentiating testicular torsion (which requires rapid correction to avoid testicular damage) from other causes of acute scrotal swelling remains the presence or absence of the cremasteric reflex. (The testicles are suspended by the cremaster muscle [from the Greek kremaster, to hang].)

In a prospective study of 245 boys, from the newborn period to age 18; who presented with acute scrotal swelling, the presence of the cremasteric reflex (stroking the inner thigh to cause an elevation of the ipsilateral testis by contraction of the cremaster muscle) was the most reliable clinical finding in ruling out testicular torsion. The correlation between the presence of ipsilateral cremasteric reflex and the absence of testicular torsion was 100%. Absence of this reflex, therefore, should strongly increase your suspicion of torsion.

Cremasteric Reflex in Acute Scrotal Swelling

<table>
<thead>
<tr>
<th>DIAGNOSIS</th>
<th>NO OF TS</th>
<th>CREMASTERIC REFLEX</th>
</tr>
</thead>
<tbody>
<tr>
<td>Testis torsion</td>
<td>56</td>
<td>0</td>
</tr>
<tr>
<td>Hydatid torsion</td>
<td>77</td>
<td>58</td>
</tr>
<tr>
<td>Epididymitis</td>
<td>47</td>
<td>31</td>
</tr>
<tr>
<td>Hernia/hydrocele</td>
<td>22</td>
<td>8</td>
</tr>
<tr>
<td>Trauma</td>
<td>22</td>
<td>19</td>
</tr>
<tr>
<td>Leukemia</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Varicocele</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Orchialgia</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>Idiopathic scrotal edema</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Henoch-Schönlein purpura</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>Testis tumor</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Insect bite</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td>245</td>
<td>125</td>
</tr>
</tbody>
</table>

Rare Causes

CNS infection
  Congenital infection
Parasitic infection
Syphilis
Tetanus
Tuberculosis

Congenital CNS malformation
  Agenesis/dysgenesis
  Holoprosencephaly
  Porencephaly
  Hydrocephalus

Drugs/toxins
  Aminophylline
  Amphetamines
  Antihistamines
  Atropine
  Camphor
  Carbon monoxide
  Drug withdrawal
  Heavy metals
  Hexachlorophene
  Hydrocarbons
  Local anesthetics
  Narcotics
  Organophosphates
  Penicillin
  Pertussis toxoid
  Phencyclidine
  Scabicides
  Steroids
  Tricyclic antidepressants

Inborn errors of metabolism
  Aminoacidopathy
  Galactosemia
  Organic aciduria
  Storage disease

Metabolic
  Hypernatremia
  Hypocalcemia
  Hypomagnesemia
  Hyponatremia

Miscellaneous
  Arrhythmia
  Dysmorphogenic syndromes (many)
  Kernicterus
  Metachromatic leukodystrophy
  Pyridoxine deficiency

Miscellaneous (Cont.)
  Rett syndrome
  Reye's syndrome
  Subacute sclerosing panencephalitis

Neurocutaneous syndrome
  Incontinentia pigmenti
  Linear sebaceous nevus
  Neurofibromatosis
  Sturge-Weber disease
  Tuberous sclerosis

Seizure mimics
  Breathholding spells
  Hyperventilation
  Malingering
  Masturbation
  Migraine
  Myoclonus
  Narcolepsy
  Orthostatic hypotension
  Pallid infantile syncope
  Panic disorder
  Paroxysmal torticollis of infancy
  Pseudoseizures
  Sandifer’s syndrome (gastro-esophageal reflux)
  Shivering on urination
  Shuddering attacks
  Sleep disorders
  Syncope
  Tics
  Vertigo

Systemic infection
  Roseola
  Shigella

Tumors

Vascular
  A-V malformation
  Embolic phenomenon
  Hemorrhage
  Hypertension
  Sickle-cell disease
  Thrombosis
  Vasculitis

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Classification of Seizures and Epilepsy

Seizures and epilepsy are classified according to a scheme developed by the International League Against Epilepsy (ILAE). An abbreviated version of the classification is shown below:

<table>
<thead>
<tr>
<th>Classification of Seizures and Epileptic Syndromes</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Partial seizures</strong></td>
</tr>
<tr>
<td>Simple partial seizures (consciousness preserved)</td>
</tr>
<tr>
<td>With motor signs (jacksonian, adverseive)</td>
</tr>
<tr>
<td>With somatosensory or special sensory symptoms</td>
</tr>
<tr>
<td>With autonomic symptoms or signs</td>
</tr>
<tr>
<td>With psychic symptoms</td>
</tr>
<tr>
<td>Complex partial seizures (consciousness impaired)</td>
</tr>
<tr>
<td>Simple partial onset followed by impaired consciousness</td>
</tr>
<tr>
<td>Impaired consciousness at onset</td>
</tr>
<tr>
<td>Secondarily generalized seizures</td>
</tr>
<tr>
<td>Simple partial seizures evolving to generalized tonic-clonic seizures</td>
</tr>
<tr>
<td>Complex partial seizures evolving to generalized tonic-clonic seizures</td>
</tr>
<tr>
<td>Simple partial seizures evolving to complex partial seizures, then to generalized tonic-clonic seizures</td>
</tr>
</tbody>
</table>

| Generalized-onset seizures                         |
| Tonic-clonic seizures                               |
| Absence seizures                                    |
| Atypical absence seizures                           |
| Myoclonic seizures                                  |
| Tonic seizures                                      |
| Atonic seizures                                     |

| Localization-related (focal) epilepsies             |
| Idiopathic                                         |
| Benign focal epilepsy of childhood                  |
| Symptomatic                                        |
| Chronic progressive epilepsy                        |
| partialis continua                                 |
| Temporal-lobe epilepsy                              |
| Extratemporal epilepsy                              |

| Generalized epilepsy                                |
| Idiopathic                                         |
| Benign neonatal convulsions                         |
| Childhood absence epilepsy                          |
| Juvenile myoclonic epilepsy                         |
| Other generalized idiopathic epilepsy               |
| Cryptogenic or symptomatic                         |
| West syndrome (infantile spasms)                   |
| Early myoclonic encephalopathy                     |
| Lennox-Gastaut syndrome                             |
| Progressive myoclonic epilepsy                      |

| Special syndromes                                  |
| Febrile seizures                                   |


What Are the Criteria for Simple Febrile Seizure?

Age 6 months to 6 years  Normal results from neurologic and developmental examination
Generalized seizure (indicating involvement of both cerebral hemispheres) of less than 20 minutes duration  Negative family history of afebrile seizures
Occurs within 24 hours of fever onset

To Treat or Not to Treat After a First Seizure?

Should the child receive antiepileptic drug therapy after a nonfebrile first seizure? This is a central and still controversial issue in the management of epilepsy. It begs the follow-up question, why treat seizures?

Seizures are treated mainly because of their psychosocial consequences. Children with epilepsy often have difficulty with interpersonal relationships and self-esteem, as well as vocational problems later in life. There are reports of slightly increased mortality in association with seizure disorders, but there is yet no proof that patients on medication have less mortality than untreated individuals. Because 25–41% of patients taking antiepileptic drugs have recurrent seizures, the effectiveness of antiepileptic drugs in preventing recurrence of seizures has also been questioned. Also, there is no evidence at this time that seizures beget further seizures.

So which children should be treated?

The decision to treat should be based on several factors, including:

- Age
- Type of seizure
- Frequency of seizures and time between
- Timing and circumstance of occurrence of seizures
- Risk of further occurrence
- Precipitating factors
- Risk of drug treatment (30% of patients have side-effects requiring modification of therapy)
- Probable consequences of further seizures
- Probability of treatment success

The chance of having an additional seizure after the first is about 30% (range 16–62%), and the second seizure tends to occur within 12 months. In children with absence seizures, the occurrence is at the high end of the range. The chance of having an additional seizure after the second is 50–75%. Studies of predictors of seizure recurrence have reported that it is often possible to identify patients with a relatively low risk of seizure recurrence.

Although the final decision to treat or not to treat must be made individually for each patient, the following guidelines may be offered:

1. Treat a child who has experienced two or more tonic-clonic seizures.
2. Treat children who experience seizures that impair consciousness, such as absence and partial complex seizures, which tend to occur more often— sometimes daily—than the rare generalized tonic-clonic seizures and can impair function because of their frequency.


Questions frequently arise in the clinic surrounding the topics of sexuality and sexual behavior in children. More often than not, these questions reveal a great deal about the comfort level and value system parents attach to this developmental issue. To aid in this dialogue, listed below are the frequencies of a large variety of sexual behaviors noted among 880 preadolescent boys and girls (ages 2-12).

<table>
<thead>
<tr>
<th>No.</th>
<th>Item (Abbreviated)</th>
<th>Overall</th>
<th>2-6 Boys</th>
<th>7-12 Boys</th>
<th>11-12 Girls</th>
</tr>
</thead>
<tbody>
<tr>
<td>10</td>
<td>Puts mouth on sex parts</td>
<td>0.1</td>
<td>0.4</td>
<td>0.0</td>
<td>0.0</td>
</tr>
<tr>
<td>15</td>
<td>Asks to engage in sex acts</td>
<td>0.4</td>
<td>7.2</td>
<td>0.0</td>
<td>0.0</td>
</tr>
<tr>
<td>7</td>
<td>Masturbates with object</td>
<td>0.8</td>
<td>0.8</td>
<td>0.8</td>
<td>0.0</td>
</tr>
<tr>
<td>17</td>
<td>Inserts objects in vagina/anus</td>
<td>0.9</td>
<td>0.0</td>
<td>2.8</td>
<td>0.0</td>
</tr>
<tr>
<td>9</td>
<td>Imitates intercourse</td>
<td>1.1</td>
<td>0.8</td>
<td>0.4</td>
<td>2.4</td>
</tr>
<tr>
<td>14</td>
<td>Sexual sounds</td>
<td>1.4</td>
<td>0.4</td>
<td>0.8</td>
<td>3.9</td>
</tr>
<tr>
<td>30</td>
<td>French kisses</td>
<td>2.5</td>
<td>1.6</td>
<td>4.0</td>
<td>2.4</td>
</tr>
<tr>
<td>28</td>
<td>Undresses other people</td>
<td>2.6</td>
<td>4.4</td>
<td>4.4</td>
<td>0.5</td>
</tr>
<tr>
<td>29</td>
<td>Asks to watch explicit television</td>
<td>2.7</td>
<td>0.0</td>
<td>1.6</td>
<td>6.8</td>
</tr>
<tr>
<td>19</td>
<td>Imitates sexual behavior with dolls</td>
<td>3.2</td>
<td>0.8</td>
<td>4.0</td>
<td>1.5</td>
</tr>
<tr>
<td>2</td>
<td>Wants to be opposite sex</td>
<td>4.9</td>
<td>7.3</td>
<td>7.5</td>
<td>1.9</td>
</tr>
<tr>
<td>22</td>
<td>Talks about sexual acts</td>
<td>5.7</td>
<td>2.4</td>
<td>2.8</td>
<td>9.2</td>
</tr>
<tr>
<td>21</td>
<td>Dresses like opposite sex</td>
<td>5.8</td>
<td>6.0</td>
<td>9.5</td>
<td>3.4</td>
</tr>
<tr>
<td>8</td>
<td>Touches others' sex parts</td>
<td>6.0</td>
<td>8.9</td>
<td>5.6</td>
<td>4.9</td>
</tr>
<tr>
<td>16</td>
<td>Rubs body against people</td>
<td>6.7</td>
<td>8.5</td>
<td>8.3</td>
<td>4.4</td>
</tr>
<tr>
<td>31</td>
<td>Hugs strange adults</td>
<td>7.3</td>
<td>6.5</td>
<td>14.3</td>
<td>2.4</td>
</tr>
<tr>
<td>32</td>
<td>Shows sex parts to children</td>
<td>8.1</td>
<td>15.7</td>
<td>7.5</td>
<td>4.4</td>
</tr>
<tr>
<td>62</td>
<td>Uses sexual words</td>
<td>8.8</td>
<td>4.8</td>
<td>1.2</td>
<td>19.9</td>
</tr>
<tr>
<td>33</td>
<td>Overly aggressive, overly passive</td>
<td>10.4</td>
<td>8.1</td>
<td>17.5</td>
<td>6.3</td>
</tr>
<tr>
<td>27</td>
<td>Talks flirtatiously</td>
<td>10.6</td>
<td>8.5</td>
<td>15.9</td>
<td>2.9</td>
</tr>
<tr>
<td>13</td>
<td>Pretends to be opposite sex</td>
<td>13.0</td>
<td>16.9</td>
<td>20.6</td>
<td>2.9</td>
</tr>
<tr>
<td>4</td>
<td>Masturbates with hand</td>
<td>15.3</td>
<td>22.6</td>
<td>16.3</td>
<td>11.2</td>
</tr>
<tr>
<td>21</td>
<td>Looks at nude pictures</td>
<td>15.5</td>
<td>11.3</td>
<td>7.9</td>
<td>27.2</td>
</tr>
<tr>
<td>20</td>
<td>Shows sex parts to adults</td>
<td>16.0</td>
<td>25.8</td>
<td>17.9</td>
<td>9.7</td>
</tr>
<tr>
<td>3</td>
<td>Touches sex parts in public</td>
<td>19.7</td>
<td>35.5</td>
<td>19.0</td>
<td>15.5</td>
</tr>
<tr>
<td>34</td>
<td>Interested in opposite sex</td>
<td>23.0</td>
<td>21.0</td>
<td>20.6</td>
<td>19.9</td>
</tr>
<tr>
<td>18</td>
<td>Tries to look at people undressing</td>
<td>28.5</td>
<td>33.9</td>
<td>33.3</td>
<td>27.7</td>
</tr>
<tr>
<td>6</td>
<td>Touches breasts</td>
<td>30.7</td>
<td>43.5</td>
<td>48.4</td>
<td>11.7</td>
</tr>
<tr>
<td>26</td>
<td>Kisses nonfamily children</td>
<td>33.9</td>
<td>41.1</td>
<td>55.2</td>
<td>9.7</td>
</tr>
<tr>
<td>23</td>
<td>Kisses nonfamily adults</td>
<td>36.2</td>
<td>41.1</td>
<td>52.4</td>
<td>18.9</td>
</tr>
<tr>
<td>25</td>
<td>Sits with crotch exposed</td>
<td>36.4</td>
<td>35.1</td>
<td>59.1</td>
<td>15.5</td>
</tr>
<tr>
<td>24</td>
<td>Undresses in front of others</td>
<td>41.2</td>
<td>49.6</td>
<td>61.9</td>
<td>21.4</td>
</tr>
<tr>
<td>11</td>
<td>Touches sex parts at home</td>
<td>45.8</td>
<td>64.1</td>
<td>54.4</td>
<td>36.4</td>
</tr>
<tr>
<td>5</td>
<td>Scratches crotch</td>
<td>52.2</td>
<td>58.1</td>
<td>67.9</td>
<td>40.8</td>
</tr>
<tr>
<td>35</td>
<td>Boy-girl toys</td>
<td>53.9</td>
<td>63.3</td>
<td>71.4</td>
<td>30.6</td>
</tr>
</tbody>
</table>

Additional items (Dec-Jan)

<table>
<thead>
<tr>
<th>No.</th>
<th>Item (Abbreviated)</th>
<th>Overall</th>
<th>2-6 Boys</th>
<th>7-12 Boys</th>
<th>11-12 Girls</th>
</tr>
</thead>
<tbody>
<tr>
<td>42</td>
<td>Touches animal sex parts</td>
<td>1.3</td>
<td>4.5</td>
<td>0.0</td>
<td>0.0</td>
</tr>
<tr>
<td>37</td>
<td>Mouth on mother's breast</td>
<td>2.6</td>
<td>0.0</td>
<td>7.7</td>
<td>0.0</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Frequency of Sexual Behaviors (Percent Endorsement) (Cont.)

<table>
<thead>
<tr>
<th>NO.</th>
<th>ITEM (ABBREVIATED)</th>
<th>OVERALL</th>
<th>2-6, BOYS</th>
<th>2-6, GIRLS</th>
<th>7-12, BOYS</th>
<th>7-12, GIRLS</th>
</tr>
</thead>
<tbody>
<tr>
<td>40.</td>
<td>Overly friendly with strange men</td>
<td>7.1</td>
<td>4.5</td>
<td>11.5</td>
<td>2.9</td>
<td>8.0</td>
</tr>
<tr>
<td>36.</td>
<td>Stands too close</td>
<td>11.6</td>
<td>6.8</td>
<td>15.4</td>
<td>14.7</td>
<td>8.0</td>
</tr>
<tr>
<td>41.</td>
<td>Shy about undressing</td>
<td>38.7</td>
<td>29.5</td>
<td>32.7</td>
<td>50.0</td>
<td>52.0</td>
</tr>
<tr>
<td>43.</td>
<td>Walks around nude</td>
<td>41.9</td>
<td>47.7</td>
<td>65.4</td>
<td>20.6</td>
<td>12.0</td>
</tr>
<tr>
<td>38.</td>
<td>Walks around in underwear</td>
<td>52.9</td>
<td>54.5</td>
<td>75.0</td>
<td>44.1</td>
<td>16.0</td>
</tr>
<tr>
<td>39.</td>
<td>Shy with strange men</td>
<td>64.5</td>
<td>63.6</td>
<td>80.8</td>
<td>47.1</td>
<td>56.0</td>
</tr>
</tbody>
</table>


## SHORT STATURE

### Use of Bone Age Determination in the Diagnosis of Short Stature

The cause for short stature may often be determined by careful history and physical examination. Nutritional or emotional deprivation, chronic disease, or a history of short stature in other family members may provide an explanation for decreased height. Facial appearance may suggest a genetic or chromosomal abnormality. Organ enlargement may lead to a diagnosis of a storage disease.

Often, however, the diagnosis is not readily apparent. In these cases, it is helpful to begin with a comparison of skeletal maturation (bone age) to height age and chronologic age. The table lists the diagnoses that should be suggested by such a comparison, and the clinical features accompanying each diagnosis.

### Comparison of Bone Age to Height Age and Chronologic Age

<table>
<thead>
<tr>
<th>MEASUREMENT</th>
<th>DIAGNOSIS SUGGESTED</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone age equal to or slightly behind chronologic age</td>
<td>Primordial short stature</td>
<td>Birth weight and length below normal for gestational age.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Subsequent growth parallel to, but below, 3rd percentile.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Normal onset and progression of puberty.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Minor skeletal abnormalities.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Includes genetic and chromosomal aberrations, e.g.,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Down’s syndrome and Turners syndrome.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Short stature as adult.</td>
</tr>
<tr>
<td>Familial short stature</td>
<td>Normal length and weight for first 1 to 2 years of life.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Height falls below 3rd percentile at 5 to 10 years of age.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Puberty not delayed.</td>
<td></td>
</tr>
<tr>
<td></td>
<td>&quot;Normal&quot; adult height not attained.</td>
<td></td>
</tr>
</tbody>
</table>

Table continued on next page.
### Comparison of Bone Age to Height Age and Chronologic Age (Cont.)

<table>
<thead>
<tr>
<th>MEASUREMENT</th>
<th>DIAGNOSIS SUGGESTED</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone age retarded in relation to chronologic age, but less retarded than height age</td>
<td>Constitutional short stature</td>
<td>Appropriate weight and length for gestational age at birth. Slow growth during childhood. Delayed onset of puberty. Other family members may remember similar growth pattern. Important to differentiate from hypothyroidism and growth hormone deficiency. Ultimately reach “normal” adult height.</td>
</tr>
<tr>
<td>Metabolic disorders, e.g.: Hypophosphatemic rickets, Hypophosphatasia, Mucopolysaccharidoses, Glycogen storage diseases, Renal tubular acidosis, Bartter’s syndrome, Vasopressin-resistant diabetes insipidus</td>
<td>Clinical and laboratory findings consistent with these disorders.</td>
<td></td>
</tr>
<tr>
<td>Organic acidemias and acidurias, Hemolytic anemias, Disorders of mineral metabolism, Immunoglobulin or white blood cell abnormality, Others</td>
<td>Clinical and laboratory findings consistent with these disorders.</td>
<td></td>
</tr>
<tr>
<td>Chronic disease, e.g.: Chronic infection, Hepatic disease, Pulmonary disease, Renal disease, Malabsorption, Malignancy, Collagen vascular disease, Others</td>
<td>Clinical and laboratory findings consistent with the disease; initial clue may be increased erythrocyte sedimentation rate. May exhibit variable growth rate over several years.</td>
<td></td>
</tr>
<tr>
<td>Bone age equal to or advanced in comparison with height age</td>
<td>Familial short stature</td>
<td>See above.</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Comparison of Bone Age to Height Age and Chronologic Age (Cont.)

<table>
<thead>
<tr>
<th>MEASUREMENT</th>
<th>DIAGNOSIS SUGGESTED</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bone age equal to or advanced in comparison with height age (Cont.)</td>
<td>Sexual precocity with androgen excess</td>
<td>Increased linear growth early in life with early closure of epiphyses. Clinical signs of androgen excess (facial, axillary, and pubic hair, penile or clitoral enlargement).</td>
</tr>
<tr>
<td></td>
<td>Sexual precocity with estrogen excess</td>
<td>Early closure of epiphyses without prior augmentation of linear growth. Clinical signs of estrogen excess (breast enlargement, galactorrhea in females, and so on).</td>
</tr>
<tr>
<td>Bone age greatly decreased and less than or equal to height age</td>
<td>Hypothyroidism</td>
<td>Degree of growth retardation depends upon age of onset. Congenital hypothyroidism is associated with severe growth failure. In juvenile hypothyroidism, the growth retardation is more insidious. Delayed dental age.</td>
</tr>
<tr>
<td></td>
<td>Cushing’s syndrome (most often iatrogenic)</td>
<td>Truncal obesity, moon facies, violaceous striae, hirsutism, muscle weakness, hypertension.</td>
</tr>
<tr>
<td></td>
<td>Hypopituitarism and growth hormone deficiency. Causes include: Congenital absence of pituitary Infection Reticuloendothelioses Vascular infarcts and anomalies Trauma Irradiation Surgical resection Malnutrition</td>
<td>Delayed dental age. Puberty often delayed. May have neurologic abnormalities.</td>
</tr>
<tr>
<td></td>
<td>Maternal deprivation</td>
<td>May have impaired motor and intellectual development. May or may not be associated with malnutrition. May have growth hormone deficiency.</td>
</tr>
</tbody>
</table>

SHWACHMAN'S SYNDROME

Pancreatic Insufficiency and Neutropenia

When Shwachman's syndrome was first described in 1964, the hallmarks of this rare entity were exocrine pancreatic insufficiency, bone marrow hypoplasia and associated neutropenia, metaphyseal chondroplasia, growth retardation, and recurrent soft tissue infections. Since that initial case report, many more manifestations of Shwachman's syndrome have been elaborated and described. These protean features of the disorder are listed in the table below.

The exact pathogenic basis for the hematologic and other features of this multisystem illness has yet to be determined, although some have hypothesized that the basic defect of the Shwachman syndrome may lie in the function of the microtubular and microfilament elements of many different cell types in the body. The relative contributions of impaired cellular motility, instead of neutropenia, to these patients' increased susceptibility toward infections is also unclear.

Features Associated with Shwachman's Syndrome

| Exocrine pancreatic insufficiency | Neonatal problems |
| Steatorrhea                      | Poor feeding, respiratory distress |
| Growth retardation               | Psychomotor retardation |
| Skeletal abnormalities           | Hypotonia |
| Metaphyseal dyschondroplasia, delayed maturation, rib abnormalities, long bone tubulation, clinodactyly | Hepatomegaly |
| Narrow thorax                    | Raised SGOT and SGPT |
| Hematologic abnormalities        | Renal tubular dysfunction |
| Bone marrow hypoplasia, neutropenia, thrombocytopenia, raised HbF, lymphoproliferative and myeloproliferative neoplasia | Ichthyosis |
| Recurrent infections             | Dental abnormalities |
| Defective neutrophil mobility     | Delayed puberty |
|                                 | Diabetes mellitus |
|                                 | Dysmorphic features |
|                                 | Endocardial fibrosis |
|                                 | Hirschsprung's disease |


SINUSES

The Paranasal Sinuses and the Mastoid Sinus

At what age does sinusitis become a diagnostic possibility?
It is useful to remember the ages at which the sinuses are pneumatized. Once a true sinus is present, the possibility of infection exists.

Sinuses present at birth Anterior and posterior ethmoid.
Maxillary antra.
Skin Signs/Sleeping Patterns—313

Two to four years

Pneumatization of frontal sinuses begins—complete by 5 to 9 years of age.

Sphenoid sinus becomes visible by age 3.

The mastoid antrum is present at birth, and pneumatization of the temporal bone starts in early infancy. The mastoid process is not present at birth, but begins to grow during the first year. Pneumatization is a slow, irregular process, but is generally complete prior to adolescence.

Sinusitis is seen with increased frequency in patients with cyanotic heart disease, in leukemia and aplastic anemia while patients are neutropenic, in cystic fibrosis, and in patients with a history of nasal allergies.


SKIN SIGNS

Tache Cérébrale and Dermatographia

Diagnostic information can be obtained from stroking the skin. When the skin over the abdomen, back, or chest is gently stroked with the fingernail or a blunted point, two major responses may be elicited: (1) tache cérébrale and (2) dermatographia.

In tache cérébrale (cerebral spot), the stroking produces a red streak that is flanked by thin, pale margins. This sign develops within 30 seconds of stroking and persists for several minutes. It has been noted to be present in patients with scarlet fever, hydrocephalus, a variety of febrile illnesses, and, most particularly, in meningitis. It can be used as an early clue to the presence of meningitis, particularly in the neonatal period. The French name derives from the presence of the sign as a concomitant of several nervous (or “cerebral”) diseases.

Dermatographia, meaning literally “writing on the skin,” is the marking of the skin by rubbing with a blunted point at sufficient pressure. The stroking produces a white or pale line with red margins. This wheal is seen in patients with fair skin, in those with vasomotor instability, or in extreme form in patients with urticaria pigmentosa (Darier’s sign). Dermatographism, the tendency to show dermatographia, is present in 2-5% of the population, but only a subgroup has symptomatic dermatographism, one of the physical urticarias.


SLEEPING PATTERNS

Crying, Feeding, and Sleeping Patterns in Infants 1 to 12 Months of Age

What is normal crying time, feeding time, or sleeping time for infants? Mothers often are concerned or complain that their baby is abnormal. With the following guidelines you can either reassure them or be alerted to a possible problem.
Mothers who feel a need for additional help generally have babies that cry for more than 6 hours per day, take more than 6.0 hours to feed, and spend less than 7 hours sleeping.

### Mean Times for Infant Activities

<table>
<thead>
<tr>
<th>ACTIVITY</th>
<th>&lt; 3 Mo</th>
<th>3-5 Mo</th>
<th>6-8 Mo</th>
<th>&gt; 9 Mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>Crying</td>
<td>1.6</td>
<td>1.3</td>
<td>1.4</td>
<td>1.1</td>
</tr>
<tr>
<td></td>
<td>(0.5-0)</td>
<td>(0.9-5)</td>
<td>(0.3-0)</td>
<td>(0.3-5)</td>
</tr>
<tr>
<td>Feeding</td>
<td>3.1</td>
<td>2.4</td>
<td>2.0</td>
<td>2.1</td>
</tr>
<tr>
<td></td>
<td>(1.0-6.8)</td>
<td>(1.3-5.3)</td>
<td>(1.0-3.8)</td>
<td>(0.8-4.5)</td>
</tr>
<tr>
<td>Sleeping</td>
<td>15.2</td>
<td>14.3</td>
<td>13.5</td>
<td>13.4</td>
</tr>
<tr>
<td></td>
<td>(11.8-20.5)</td>
<td>(10.0-18.5)</td>
<td>(10.3-17.8)</td>
<td>(10.3-16.0)</td>
</tr>
</tbody>
</table>


---

### Snake Bite

#### Is This a Poisonous Snake Bite?

This question is posed to physicians in offices and emergency rooms several thousand times a year in this country. A correct and prompt answer is essential for proper treatment. Failure to use antivenom early when indicated can be fatal; its inappropriate employment for a bite by a harmless snake may be hazardous due to severe reactions.

The problem has two parts. First, was the bite due to a harmless or a poisonous snake? Second, if the snake was venomous, is envenomation present or likely? The question may be resolved by examination of the snake so that it may be put in the category of either a venomous or a nonvenomous variety and by examination of the patient to determine if venomation has occurred. Basic dependable guidelines for attaining both these goals will be outlined. They may be used by the amateur who has no knowledge of serpents at all. This discussion applies only to those snakes that are native to the continental United States and does not relate to foreign species introduced into this country as pets or exhibits.

#### Examination of the Snake

One should not attempt to identify the exact species, since this is often a challenge for even the genuine expert due to pitfalls involving confusing color variate (albinism and malanism) and deceptive patterns (atypical or absent). Undue delay may result by waiting to locate an available herpetologist in a nearby zoo, museum, or zoology department. One should instead inspect the snake and, from the guidelines provided, assign it to the harmless or harmful group. The problem is somewhat simplified since in the mainland United States there are only two families of indigenous poisonous serpents.

- **Crotalidae.** These are the pit vipers, which include all rattlesnakes, cottonmouths (water moccasin), and copperheads (highland moccasin). One or more of this family has been found in all states, with the exception of Maine, Alaska, and Hawaii. The head is large and triangular. The neck is relatively slender, so that it is readily distinguishable from the thick, heavy body. The pupil is vertically
elliptical. Pit organs ( loreal pits) are pat'ognomorphic of all members of this group. A pit is present on each side of the head and it resembles an extra nostril. The pits are deep, readily visible between the eye and the nostril, and located just below a line connecting these two structures. One or two fangs are found on the upper jaw of all pit vipers. They are specialized hollow or grooved teeth, which are recurved and longer than the other teeth. It is through these that the venom is injected. In this family the fangs are movable and when not in use are folded up against the palate. A white membrane may cover the fang down to the tip. Normally there are two fangs in the upper jaw one on each side of the maxilla so the classic bite pattern shows two fang punctures. However, one or both may be broken off or shed, in which instance there may be only a single fang mark present or none at all. No envenomation is possible if fangs are absent. Reserve fangs are always present, so the missing fang is replaced soon. Rarely, one or two reserve fangs may be functioning along with the customary complement of one or two fangs. In such a circumstance the bite pattern will be atypical, demonstrating three or four fang punctures.

Herpetologists identify species by meticulous scale counts of the head, neck, body, and tail. For practical purposes one may observe the scales (scutes, shields, plates) on the ventral surface of the body just posterior to the anus (subcaudal scales). In this family the subcaudal scales are usually arranged in a single row, but exceptions occur in which the rows are double. In the majority of harmless snakes the subcaudal scales are double, but this is not infallible either since exceptions are found. Rattles, of course, are specific for rattlesnakes and are not present in the copperhead, cottonmouth, other venomous species, or harmless snakes. They break off because of wear and tear, or during ecdysis (molting), so the number is inconstant regardless of the age of the reptile. If all rattles have been lost or if the specimen is a baby that has not yet developed rattles, there will be a slight enlargement at the tip of the tail known as the button. Other poisonous snakes do not show a button, nor do nonvenomous species. Also, the end of the tail of a rattlesnake that has lost its rattles is short and blunt, whereas the tip of the tail of a harmless snake is usually gradually tapered.

Elapidae. This family is represented in this country only by the coral snake. Unlike the pit vipers the coral snake is restricted to the southern states and is generally not found north of Arizona, Arkansas, or the Carolinas. Compared to the rattlesnakes and moccasins, the head is narrow and the neck and body are slender, giving a cylindrical configuration which is quite different from the shape of the pit vipers. The pupil is circular, thus resembling that of our indigenous harmless varieties. Pit organs are not present. Two fangs are present one on each side of the maxilla (unless one or both have been shed). They are erect, fixed, and smaller than those of the pit vipers. Subcaudal scales tend to be in a double row similar to those of nonvenomous snakes, but rarely may be in a single row in the coral snake. Both rattles and tail button are absent. The coral snake is an exception to the general rule of not trying species identification, since there are confusing imitators that are harmless. Fortunately the nonvenomous mimics are easy to differentiate from the potent coral snake. The coral snake has a black snout and broad body rings of red and black that are separated by a narrower band of yellow. The mnemonic “red next to yellow kills a fellow” is helpful to keep in mind. The harmless look-alikes (scarlet snake, scarlet king snake) show a grey or red snout and red and yellow rings that are separated by a black band. Here the mnemonic to remember is “red against black is venom lack.”
Summary of Family Characteristics—Native Harmless Versus Native Venomous Snakes in the Continental United States

### Ways to Differentiate Poisonous From Harmless Snakes

<table>
<thead>
<tr>
<th>Poisonous (Pit vipers)</th>
<th>Harmless</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oval head</td>
<td>Large and triangular (pit vipers); small and narrow (coral snake)</td>
</tr>
<tr>
<td>Vertical elliptical pupil</td>
<td>Vertically elliptical (pit vipers); round (coral snake)</td>
</tr>
<tr>
<td>Absent pit organs</td>
<td>Present in all pit vipers (copperhead, cottonmouth, rattlesnakes); absent in coral snake</td>
</tr>
<tr>
<td>Absent fangs</td>
<td>Present in all venomous species. Large, long, recurved teeth. Long and movable in pit vipers. Short, erect, and fixed in coral snake. Usually 2 (1 on each side upper jaw) unless shed or reserve fangs also in use</td>
</tr>
<tr>
<td>Double row subcaudal scales, usually, but exceptions</td>
<td>Single row in pit vipers, but with exceptions; double row in coral snake, but exceptions</td>
</tr>
<tr>
<td>Absent rattles</td>
<td>Present in all rattlesnakes unless lost or undeveloped in baby. If missing a button, present at tip of tail. No rattles in other venomous species or in harmless snakes</td>
</tr>
</tbody>
</table>

Table continued on next page.
Nonvenomous Versus Venomous Characteristics of Continental U.S. Snakes (Cont.)

<table>
<thead>
<tr>
<th>NONVENOMOUS</th>
<th>VENOMOUS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Usually ends in gradual taper</td>
<td>Tip of tail Short and blunt in rattlesnake if rattles lost</td>
</tr>
<tr>
<td>Bite pattern (seldom perfect)</td>
<td>Total of 6 with no fang marks; four rows maxillary teeth and 2 rows mandibular teeth</td>
</tr>
<tr>
<td>Total of 6 with no fang marks; four rows maxillary teeth and 2 rows mandibular teeth</td>
<td>Number of rows Total of 4 with 1-2 fang marks (rarely 3-4 if reserve fangs functional). Two rows with fang marks from maxillary teeth. Two rows without fang marks from mandibular teeth</td>
</tr>
<tr>
<td>Series scratches or tiny punctures (1-2 mm deep); pattern of mandibular teeth often imperfect</td>
<td>Appearance Series scratches or tiny punctures (1-2 mm deep) plus fang marks. Fang marks recognizable as larger and deeper punctures than those from nonfang teeth. Mandibular teeth often indistinct</td>
</tr>
</tbody>
</table>

Examination of the Patient

This is to determine if envenomation has taken place. If it has occurred, immediate vigorous therapy is required. If the snake was not captured, presence or absence of envenomation will be the sole criterion available for deciding if the snake was harmless or poisonous. Verbal descriptions of escaped snakes are generally unreliable. The bite pattern is helpful and may be diagnostic as indicated earlier, but it does not indicate if envenomation has in fact occurred. Even though the victim has been struck by a venomous serpent, envenomation may not ensue. This is the result of various circumstances that influence the flow of venom, the amount of venom injected, and the toxicity of the venom. Evaluation of envenomation depends on the development of local and systemic symptoms and signs. The following are the usual clinical effects which may appear after injection of a sufficient amount of a potent venom.

Local Symptoms and Signs. The two P’s (puncture and pain) and the two E’s (edema and erythema) constitute the classic local reaction to deposition of a potent venom in the tissues. At least two should be present to substantiate the diagnosis.

I. Puncture: One or two fang marks are present (rarely three or four if reserve fangs in use). These punctures are larger and deeper than those from the other teeth. A wheal or vesicle may develop at the site. If at least one fang mark is not present, then envenomation could not have taken place. Bleeding is usually brisk.

II. Pain: Usually develops within 5 to 10 minutes of the strike. It may be delayed up to an hour under certain conditions and may be lacking with a coral snake bite. In a classic case of moderate to severe envenomation involving a pit viper, the pain appears promptly and is severe and unremitting.

III. Edema: Typically obvious within 5 to 10 minutes. It also may be delayed up to an hour or absent with a coral snake bite. The swelling may progress up the limb during the next 36 hours and eventually reach the trunk. The overlying skin becomes tense and shiny. The extent of the edema is one of the criteria used in the clinical grading of the severity of envenomation for assessing the amount of antivenom required and for monitoring the progress of the case.
IV. **Erythema:** Redness is ordinarily visible within 5 to 10 minutes. It may not appear for an hour and may be absent after a bite by a coral snake. Later, other types of discoloration develop with pit viper bites as hemorrhages occur in the tissues. Eventually, some blueness usually follows unlike the typical reaction to a severe insect bite.

**NOTE:** Exceptions in the time of appearance and number of these four cardinal signs occur in some pit viper bites owing to variability of potency and amount of venom injected. Also in the event of a fortuitous strike directly into a vessel, there may be absence of local manifestations along with the rapid appearance of systemic signs. Local signs may be missing with coral snake bites owing to the predominance of neurotoxin over hemotoxin.

V. **Hemorrhage:** Petechiae and ecchymoses commonly occur, particularly with pit viper bites. Oozing from fang marks often continues for several hours. This is in contradistinction to the wounds from nonfang teeth, which cease bleeding promptly with both poisonous and nonpoisonous species.

VI. **Paresthesias:** Numbness and/or tingling frequently are noted at the bite site and around the mouth.

VII. **Late local signs:** Tissue necrosis and thrombosis may develop, with sloughing of tissues and gangrene of the extremities. This type of response is common with pit vipers but less so with coral snakes due to the difference in the venoms. Localized lymphadenopathy is a feature in some.

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**Systemic Symptoms and Signs.** These are produced by the hematogenous or lymphogenous dissemination of the venom. Some of the toxins have enzymatic activity. Owing to the multiplicity of the effects of these diverse protein molecules, the clinical manifestations are numerous and protean. A consistent clinical picture may not be present.

I. **General:** Lassitude, weakness, fatigue, nonwhirling dizziness, diaphoresis, sialorrhea, and the sensation of a “full” or “thick” tongue.

II. **Pulmonic:** Edema, respiratory failure, and death.

III. **Cardiac:** Hypotension, congestive failure, cardiac arrest, and death.

IV. **Renal:** Hematuria, proteinuria, azotemia, and renal failure with death.

V. **Gastrointestinal:** Nausea, emesis, hematemesis, and melena.

VI. **Hematologic:** Alterations of the coagulation system, with petechiae, ecchymoses, bleeding into subcutaneous and muscle tissues, hemorrhages into viscera, and bloody effusions into serous cavities. Laboratory determinations may demonstrate prolonged prothrombin time, thrombocytopenia, fibrinolysis, and prolonged bleeding and clotting times. Epistaxis, hematuria, hematemesis, and melena are common with severe envenomation.

VII. **Central nervous system:** Headache, blurred vision, paresthesias, slurred speech, bulbar palsies, generalized convulsions, and paralyses of the extremities. Deep tendon reflexes are variable. The sensorium typically remains intact, with a lucid and oriented patient. Sometimes somnolence may be a feature, and occasionally euphoria is present if the individual has been dosed with the traditional snake bite remedy, whiskey. Disorientation and states bordering on delirium and mania may occur in some victims owing to the hysteria and snake phobia seen in some adults following exposure to a serpent.

VIII. **Death:** Fatalities are due to respiratory failure, cardiac decompensation, renal shutdown, hemorrhage, or irreversible shock.
The physician who has no knowledge of snakes can render an intelligent decision about a snake bite by following the guidelines given regarding inspection of the snake, observation of the bite pattern, and examination of the patient. Irrefutable proof of the poisonous nature of a snake native to the continental United States includes the presence of fangs, pit organs, rattles, and a vertically elliptical pupil. All of these are present in the pit vipers (rattlesnakes, copperheads, and cottonmouth moccasins). The coral snake lacks the pit organs and rattles and has a circular pupil. Otherwise, all indigenous snakes with a round pupil are harmless. Fang marks are diagnostic of a venomous species and are larger and deeper than the scratches or superficial punctures produced by the nonfang teeth. If the patient shows pain, puncture, edema, and erythema, envenomation has taken place. These local signs appear within an hour of the bite, and systemic signs develop later. Local manifestations may be absent with coral snake bite, in which case generalized signs and symptoms develop rapidly.


References:

Other readings:
SODIUM: HYPERNATREMIA

Hypernatremia is defined here as a serum sodium level higher than 145 mEq/L.

Common Causes
- Diarrhea
- High environmental temperatures

Uncommon Causes
- Nephrogenic diabetes insipidus
- Postobstructive diuresis
- Salt poisoning
- Sickle-cell nephropathy

Rare Causes
- Cushing's disease
- Hypercalcemia nephropathy

SODIUM: HYPONATREMIA

Hyponatremia is defined here as a sodium level lower than 130 mEq/L.

Common Causes
- Diarrhea
- Excessive salt-free infusions
- Syndrome of inappropriate ADH secretion (SIADH)
- Water intoxication

Uncommon Causes
- Acute renal failure
- Chronic renal failure
- Congestive heart failure
- High environmental temperatures

Rare Causes
- Adrenal insufficiency
- Cirrhosis
- Cystic fibrosis and excessive sweating

SPLENOMEGALY

Common Causes
- Acute infections (bacterial, viral, rickettsial, protozoal, spirochetal, myobacterial)
- Congenital hemolytic anemias
  - Hemoglobinopathies
  - Hereditary spherocytosis
  - Thalassemia major; thalassemia intermedia


Spoiled Child Syndrome—321

Uncommon Causes

Congestive splenomegaly
Cyanotic congenital heart disease
Hodgkin's disease
Juvenile rheumatoid arthritis

Leukemia
Lupus erythematosus
Non-Hodgkin's disease
Severe iron-deficiency

Rare Causes

Acquired autoimmune hemolytic anemia
Amyloidosis
Beckwith-Wiedemann syndrome
Brucellosis
Chronic granulomatous disease
Congenital erythropoietic prophyria
Dysgammaglobulinemia
Hemophagocytic syndromes
Histiocytosis
Hurler's syndrome and other mucopolysaccharide disorders

Malaria (in the United States)
Metastatic neuroblastoma
Myelofibrosis
Osteopetrosis
Sarcoidosis
Serum sickness
Splenetic cyst or hemangioma
Storage disease (e.g. Gaucher's, Neimann-Pick)
Wolman's disease

THE SPOILED CHILD SYNDROME

The pediatrician is called upon to wear many hats. One of the more difficult roles is counselor to the parent concerned about a “spoiled” child. The concept demands differentiation between normal behavior patterns and the excessive self-centeredness that marks the spoiled child. The cause of the spoiled child syndrome is often not a lack of discipline by the parents, but a lack of consistent limit-setting.

Age-Related Normal Behavior Pattern

The Crying Infant. Brazelton's time-honored study of crying in infancy (Pediatrics 29:579-588, 1962) indicated that the average infant cried 2 1/2 hours per day for the first 7 weeks of life. Whether due to hunger, colic, or want of attention, the infant's cry represents a genuine need to which the parent ought to be encouraged to respond. After 3 to 4 months of age, a cry may become a manipulation demanding modification techniques on the part of the parent.

The Exploring Toddler. As the infant discovers his or her mobility, curiosity becomes infinite. The “search and destroy” or “baby taste test” activities can frighten or aggravate the most equanimitous parent. An understanding of this stage as a normal part in development will help the parent to “child proof” the home and to begin the process of setting limits for the child.

The Terrible Two's. As the child approaches 2 years of age, his sense of autonomy is beginning to emerge. This is often the time that conflicts between the parent and the newly assertive child begin. The characteristic independence and resistance to parental authority can be damped by a variety of techniques. One way to avoid confrontation is to offer the child choices within the parent's
limits. Thus, both the parent and the emerging individual maintain a sense of control.

As with most human behaviors, those of the normal child are on a continuum with those labelled disruptive or spoiled. The family environment, the presence of stressors, and the individual's inherent coping abilities can shift the balance. In the absence of childhood handicaps or family stresses, such as separation and divorce, parental alcoholism, or parental mental illness, there are certain behavior patterns that do not fit any but the "spoiled" category. In these cases, the parent needs both guidance and assurance that the setting of consistent limits and appropriate punishments for infractions can be effective.

Behavior Patterns Suggestive of True Spoiling

**Trained Night Feeding.** Beal's 1969 study on night feeding concluded that by 4 months of age, 95% of infants should sleep through the night without a feeding. The older infant who continues to cry for a 2 A.M. meal is often the child of caring parents who have attended every cry with a breast or bottle. Failure to break the snacking cycle and use cuddling or a pacifier can lead to the development of spoiling.

**Trained Night Crying.** As with trained night feeding, trained night crying represents the infant's training of the parent. The new parent in particular is loathe to leave a crying infant in bed. But here, as with other infant behaviors, the "need" has to be distinguished from the "want" if the parent desires rest. A helpful hint might be placing the infant in his or her crib while still awake with an assurance that 10 to 15 minutes of crying is part of the infant's own settling mechanism. Trained responsiveness from the parent can diminish the baby's own ability to achieve sleep.

**Recurrent Temper Tantrums.** A tantrum is a "fit of bad temper" representing both anger and frustration. They generally surface in the toddler who is attempting to assert independence and can be a frightening spectacle for the parent. Tantrums, like limitations, are occasionally inevitable. If the tantrum is rewarded by the loosening of restrictions, it is apt to recur. Reassure the parents that though the desire to please their child may be strong, ignoring the tantrum will likely bring this behavior to an end without harming the child. With tantrums, as with other disruptive behaviors, a little anticipatory guidance can go a long way.

**The Toddler Who Is Out of Control.** This is often a child whose parents have "tried everything" to modify the kicking, biting, refusals to eat, sleep, or toilet train. If the modifications have been inconsistent, so will be the responses. Instruction in the use of "time out" and the importance of its regular use should help the troubled parents. Remind the parents that the un-training of disruptive behaviors will not be miraculous and will require patience, but that with continued enforcement, they will be successful.

In summary, the parents of the unruly child need guidance and support. It is probably best not to label a child "spoiled" but to stress the normal behavior patterns of children at various ages and to offer solutions for avoiding or correcting disruptive behaviors. By emphasizing consistency and de-emphasizing worry you can help the parents learn the effectiveness of limits.

What Are the Guidelines for Disqualifying Conditions for Sports Participation?

In 1988 the American Academy of Pediatrics published new guidelines for participation in competitive sports, which are summarized in the tables below:

Classification of Athletic Events According to Probability for Contact and Degree of Strenuousness

<table>
<thead>
<tr>
<th>CONTACT COLLISION</th>
<th>LIMITED CONTACT IMPACT</th>
<th>NONCONTACT</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Strenuous</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Boxing</td>
<td>Baseball</td>
<td>Aero dancing</td>
</tr>
<tr>
<td>Field hockey</td>
<td>Basketball</td>
<td>Crew</td>
</tr>
<tr>
<td>Football</td>
<td>Bicycling</td>
<td>Fencing</td>
</tr>
<tr>
<td>Ice hockey</td>
<td>Diving</td>
<td>Field</td>
</tr>
<tr>
<td>Lacrosse</td>
<td>Field</td>
<td>Disleus</td>
</tr>
<tr>
<td>Martial arts</td>
<td>High jump</td>
<td>Javelin</td>
</tr>
<tr>
<td>Rodeo</td>
<td>Pole vault</td>
<td>Shot put</td>
</tr>
<tr>
<td>Soccer</td>
<td>Gymnastics</td>
<td>Running</td>
</tr>
<tr>
<td>Wrestling</td>
<td>Horseshack rising</td>
<td>Swimming</td>
</tr>
<tr>
<td></td>
<td>Ice</td>
<td>Tennis</td>
</tr>
<tr>
<td></td>
<td>Roller</td>
<td>Track</td>
</tr>
<tr>
<td></td>
<td>Skiing</td>
<td>Weight lifting</td>
</tr>
<tr>
<td></td>
<td>Cross-country</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Downhill</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Water</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Softball</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Squash, handball</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Volleyball</td>
<td></td>
</tr>
</tbody>
</table>


Recommendations for Participation in Competitive Sports

<table>
<thead>
<tr>
<th>CONTACT COLLISION</th>
<th>LIMITED CONTACT IMPACT</th>
<th>NONCONTACT</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Strenuous</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Atlantoaxial instability</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>*Swimming: no butterfly, breast stroke, or diving starts.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Acute illness</td>
<td>* Needs individual assessment, e.g., contagiousness to others, risk of worsening illness.</td>
<td></td>
</tr>
<tr>
<td>Cardiovascular</td>
<td>Moderate</td>
<td></td>
</tr>
<tr>
<td>Carditis</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Hypertension</td>
<td>Mild</td>
<td></td>
</tr>
<tr>
<td>* Needs individual assessment.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Table continued on next page.
### Recommendations for Participation in Competitive Sports (Cont.)

<table>
<thead>
<tr>
<th></th>
<th>CONTACT COLLISION</th>
<th>LIMITED IMPACT</th>
<th>NONCONTACT MODERATELY STRENUEOUS</th>
<th>NONCONTACT NONSTRENUEOUS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiovascular (Cont.)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congenital heart disease</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Patients with mild forms can be allowed a full range of physical activities; patients with moderate or severe forms, or who are postoperative, should be evaluated by a cardiologist before athletic participation.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Eyes</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Absence or loss of function of one eye</td>
<td>*</td>
<td>+</td>
<td>*</td>
<td>+</td>
</tr>
<tr>
<td>Detached retina</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>* Availability of American Society for Testing and Materials (ASTM)-approved eye guards may allow competitor to participate in most sports, but this must be judged on an individual basis.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>* Consult ophthalmologist.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Inguinal hernia</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Kidney: Absence of one</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Liver: Enlarged</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Musculoskeletal disorders</td>
<td>*</td>
<td>*</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>* Needs individual assessment.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Neurologic</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>History of serious head or spine trauma, repeated concussions, or craniotomy</td>
<td>*</td>
<td>*</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Convulsive disorder</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Well controlled</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Poorly controlled</td>
<td>No</td>
<td>No</td>
<td>Yes*</td>
<td>Yes*</td>
</tr>
<tr>
<td>* Needs individual assessment.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>* No swimming or weight lifting.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>* No archery or riflery.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ovary: Absence of one</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Respiratory</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pulmonary insufficiency</td>
<td>*</td>
<td>*</td>
<td>*</td>
<td>*</td>
</tr>
<tr>
<td>Asthma</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>* May be allowed to compete if oxygenation remains satisfactory during a graded stress test.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sickle cell trait</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Skin: Boils, herpes, impetigo, scabies</td>
<td>*</td>
<td>*</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>* No gymnastics with mats, martial arts, wrestling, or contact sports until not contagious.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spleen: Enlarged</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Testicle: Absence or undescended</td>
<td>Yes*</td>
<td>Yes*</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>* Certain sports may require protective cup.</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

---


Women and Menstruation in the Athlete

The Committee on Sports Medicine of the AAP has recommended that any medical evaluation of a female athlete should include a focus on menstrual history. Pubertal development appears to be delayed in thin athletes, especially ballet dancers and runners.

![Graph showing age of menarche in sedentary controls, high school athletes, college athletes, Olympic athletes, and ballet dancers.](image)

Exercise and age of menarche. Pubertal development appears to be delayed in thin athletes.

![Graph showing age of menarche in athletes trained premenarcheally and postmenarcheally.](image)

Athletes who began their training premenarcheally experienced a delay in menarche.

**PREMENARCHAL ATHLETIC TRAINING**

\[ \times 1 \text{ year} \quad \Rightarrow \quad \text{DELAY IN MENARCHE BY 5 MONTHS} \]

For each year of training before menarch, menarche is delayed by 5 months.
When should initiation of pubertal changes in an athlete be considered abnormal? The following guidelines are helpful:

1. If no pubertal changes occur by the chronologic age of 13 years (two standard deviations outside the normal variation), examination should be done to rule out thyroid abnormalities, prolactin-secreting adenomas, ovarian dysgenesis, and chromosomal abnormality.
2. If there is no period by the age of 16 with some pubertal growth, then a definite search must be made for anatomic causes of amenorrhea and mullerian agenesis.
3. The AAP Committee on Sports Medicine has recommended that a work-up be instituted if menarche is delayed by 1 year beyond the age of onset of menses of other female family members.
4. If the patient, her parents, or coach are anxious about late menarche or delayed puberty, a limited individualized work-up should be offered.

Some common causes of delayed menarche are listed below:

**Some Common Causes of Delayed Menarche**

<table>
<thead>
<tr>
<th>Hypothalamic</th>
<th>Ovarian</th>
</tr>
</thead>
<tbody>
<tr>
<td>Space-occupying lesions (e.g., cranio-pharyngioma, glioma)</td>
<td>Gonadal dysgenesis - chromosomal abnormalities</td>
</tr>
<tr>
<td>Functional disturbances of hypothalamic-pituitary axis (e.g., anorexia nervosa, emotional stress, athletics, eating disorders, drugs)</td>
<td>Tumors</td>
</tr>
<tr>
<td>Hypopituitarism - idiopathic</td>
<td>Polycystic ovaries</td>
</tr>
<tr>
<td>Prolactin-secreting adenomas</td>
<td>Resistant ovary syndrome</td>
</tr>
<tr>
<td>Pituitary</td>
<td>Uterine or vaginal</td>
</tr>
<tr>
<td>Hypopituitarism - idiopathic</td>
<td>Absence of uterus (e.g., mullerian agenesis)</td>
</tr>
<tr>
<td>Prolactin-secreting adenomas</td>
<td>Complete or partial absence of vagina</td>
</tr>
<tr>
<td>Other</td>
<td>Imperforate hymen resulting in hematocolpos</td>
</tr>
<tr>
<td>Congenital adrenal hyperplasia</td>
<td></td>
</tr>
<tr>
<td>Hypothyroidism or hyperthyroidism</td>
<td></td>
</tr>
<tr>
<td>Debilitating chronic disease (e.g., congenital heart disease, Crohn's disease, collagen disorders, renal failure)</td>
<td></td>
</tr>
</tbody>
</table>


**STOOL**

**Stool Frequency in Healthy Infants and Children**

Knowledge of the normal range of bowel movements can help physicians and parents deal with concerns regarding both constipation and diarrhea. Although both of these entities are more a function of the state of stool hydration, the issue of frequency frequently enters into the thinking. Listed in the following table are some norms based on age:
**Number of Stools Per Day**

<table>
<thead>
<tr>
<th>AGE</th>
<th>PERCENTILES</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>3</td>
</tr>
<tr>
<td>5 days 1 mo</td>
<td>0.9</td>
</tr>
<tr>
<td>1-5 mo</td>
<td>0.6</td>
</tr>
<tr>
<td>5-12 mo</td>
<td>0.8</td>
</tr>
<tr>
<td>1-3 yr</td>
<td>0.6</td>
</tr>
<tr>
<td>3-6 yr</td>
<td>0.4</td>
</tr>
<tr>
<td>Over 6 yr</td>
<td>0.4</td>
</tr>
</tbody>
</table>


**Stool Frequency in Infants as a Function of Feeding Style**

Do breastfed infants have more stools than bottle-fed babies? On average, the answer is yes, but great individual variation exists according to different feedings, as is revealed by the numbers below from a study of 185 infants under 3 months of age.

<table>
<thead>
<tr>
<th>Stool Frequency</th>
</tr>
</thead>
<tbody>
<tr>
<td>TYPE OF FEEDING</td>
</tr>
<tr>
<td>Human milk</td>
</tr>
<tr>
<td>Formula</td>
</tr>
<tr>
<td>Human milk + formula</td>
</tr>
</tbody>
</table>


**The Floating Stool**

There is a persistent myth that it is the fat content that buoys the floating stool. We thought this myth had been deflated in the early 1970s in a series of poetic testaments in *The New England Journal of Medicine*. The following sample gives ample argument to the fact that it's the air in stool that keeps it afloat, not the fat.

**Floaters and Sinkers**

*To the Editor:* The recent article “Floating Stools—Flatus versus Fat.” inspired me to embrace the Muse as follows:

While safe's the stool that comes a sinker,
The floater's apt to be a stinker.

So it's not fat but, rather, flatus
Imparts the elevated status.

Freehold, NJ

STRABISMUS

Strabismus, or squint, is a result of one of the three major pathologic processes:

1. An imbalance in the ocular muscles of the two eyes as a result of maldevelopment or innervation.
2. A difference in the refraction of the two eyes.
3. A visual defect in one eye.

Strabismus may be either paralytic or nonparalytic. Nonparalytic strabismus is seen frequently in infants during the first 6 months of life. After this age strabismus requires an explanation and treatment in order to avoid amblyopia. A paralytic squint is abnormal at any age.

When the squint is of the nonparalytic type (concomitant), all muscles move the eye normally, but they do not work in conjunction with each other. The two eyes are in the same position relative to each other, whatever the direction of gaze. The nonparalytic squint is not associated with diplopia. In young infants the presence of strabismus can easily be confirmed by shining a light at the eyes from directly in front of the patient. The reflection of the light should normally be in the center of the pupil or at a corresponding point on both corneas.

When the squint is of the paralytic type (nonconcomitant) owing to muscle paralysis, the eyes are straight except when moved in the direction of the paralyzed muscle. If full ocular movements are elicited in one eye when the other is covered, then a paralytic strabismus can be excluded.

Nonparalytic squint is seen in children with hydrocephalus, cerebral palsy, retinoblastoma, corneal opacities, and refractive errors.

Paralytic squint should suggest the presence of a brain stem lesion and increased intracranial pressure.


STRIDOR

Common Causes

<table>
<thead>
<tr>
<th>Allergic reaction</th>
<th>Retropharyngeal abscess</th>
</tr>
</thead>
<tbody>
<tr>
<td>Croup</td>
<td>Secretions</td>
</tr>
<tr>
<td>Foreign body aspiration</td>
<td>Spasmotic croup</td>
</tr>
<tr>
<td>Hypertrophied tonsils/adenoids</td>
<td>Subglottic stenosis (congenital, postintubation)</td>
</tr>
<tr>
<td>Peritonsillar abscess</td>
<td>Vocal cord nodules</td>
</tr>
<tr>
<td>Postinstrumentation edema</td>
<td></td>
</tr>
</tbody>
</table>

Uncommon Causes

<table>
<thead>
<tr>
<th>Corrosive ingestion</th>
<th>Tracheitis (bacterial)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epiglottitis</td>
<td>Vocal cord paralysis (congenital postsurgical)</td>
</tr>
<tr>
<td>Granuloma (postintubation/tracheostomy)</td>
<td>Vocal cord polyps</td>
</tr>
<tr>
<td>Laryngeal trauma</td>
<td></td>
</tr>
</tbody>
</table>
Stridor—329

Rare Causes

- Angioneurotic edema
- Congenital goiter
- Cricoarytenoid arthritis (JRA)
- Diphtheria
- Ectopic thyroid
- Esophageal foreign body
- External tracheal compression
  - Hemorrhage
  - Infection
  - Tumor
- Farber’s disease
- Glossoptosis
- Hemangioma
- Hypoplastic larynx
- Internal laryngocele
- Laryngeal papilloma
- Larangeal tumors
- Laryngismus stridulus (rickets)
- Marcoglossia
- Opitz-Frias syndrome
- Pierre Robin syndrome
- Post-tracheostomy stricture
- Psychogenic stridor
- Tetany
- Thyroglossal duct cyst
- Tracheoesophageal fistula
- Tracheo-laryngo-esophageal cleft
- Vascular ring

Stymied by Stridor?

Stridor is a harsh, high pitched sound made during breathing, especially inspiration. It is always indicative of a pathologic problem. Think in anatomic terms and you will usually find the cause.

Stridor at the Epiglottis

Congenital anomalies:
- Aryepiglottic cyst
- Dermoid cyst
- Thyroglossal duct cyst
- Lingual thyroid
- Flabby epiglottis

Inflammatory disease:
- Epiglottitis: bacterial origin, allergic origin

Stridor at the Larynx and Subglottic Region

Congenital anomalies:
- Hemangioma or lymphangioma
- Unilateral or bilateral vocal cord paralysis
- Laryngeal and/or subglottic stenosis
- Laryngomalacia
- Laryngeal cyst
- Papilloma

Trauma:
- Birth injury
- Postlaryngoscopy
- Postlaryngeal catheterization

Inflammatory disease:
- Laryngitis
- Laryngeal abscess
- Subglottic edema (of allergic origin)

Foreign body:
- Radiopaque or radiolucent

Metabolic disorders:
- Laryngismus stridulus (rickets)
330—Stroke

**Stridor from the Trachea**

Congenital anomalies: Hemangioma or lymphangioma
Tracheomalacia
Cartilage ring abnormalities ("segmental malacia")

Foreign body: Radiopaque or radiolucent

Postoperative: After tracheal intubation
Stricture after tracheostomy
Narrowing at the level of tracheoesophageal fistula

**Stridor from Causes Originating Outside the Respiratory Tract**

Congenital anomalies: Vascular ring or anomalous innominate artery
Esophageal atresia
Tracheoesophageal fistula
Aberrant or ectopic thyroid tissue
Congenital goiter
Carcinoma of thyroid

Inflammatory origin: Retropharyngeal abscess
Retroesophageal abscess

Foreign body: Within the esophagus

Postoperative: After tracheoesophageal fistula closure
After mid-mediastinal surgery


---

**STROKE**

**Stroke in Children and Teenagers**

The incidence of stroke in children and adolescents is extremely low; this is largely due to the rare occurrence of significant atherosclerosis in these age groups. When strokes do occur in children and teenagers, they are often severe and frequently associated with seizure disorders, motor deficits, and death. Persistent aphasia, on the other hand, which is a common feature of stroke in adults, rarely accompanies stroke in children. When confronted with a child presenting with signs and symptoms of a cerebrovascular accident, the clinician needs to consider three important factors before applying the differential diagnosis that appears below: (1) the patient's age, (2) the presence of other or underlying medical conditions, and (3) the clinical presentation of the stroke.

1. Strokes not associated with underlying systemic disease

   a. **Acute hemiplegia of childhood** (acute infantile hemiplegia) refers to the sudden onset of hemiparesis that is not associated with intracranial hemorrhage. Of these cases 60% present with severe, generalized seizures and coma. The neurologic examination is remarkable for weakness. Although
there exists a large number of pathologic entities that can cause acute hemiplegia of childhood, we shall divide them into five major processes:

i. Occlusive vascular disease at the base of the brain associated with telangiectasia of the basal ganglia (Moyamoya syndrome)

ii. Occlusive vascular disease at the base of the brain without telangiectasia.

iii. Narrowing of the origin of the internal carotid artery

iv. Distal branch occlusion of the intracranial arteries

v. Corkscrew pattern in small terminal arteries

b. **Intracranial hemorrhage** is strongly suggested by the sudden onset of a neurologic deficit in association with headache, somnolence, and nuchal rigidity. The CT scan of the head is usually diagnostic.
   i. **Arteriovenous malformations** are the most common cause of subarachnoid hemorrhage in children. They may or may not be associated with a neurologic deficit; a history of seizures may exist.
   ii. **Aneurysms** are rare in infants but are more common as age increases. The initial episode of aneurysmal hemorrhage may not be associated with focal neurologic signs, but subsequent episodes often yield significant deficits. Aneurysmal hemorrhages may rarely present as frequent headaches or cranial nerve palsies. Polycystic kidney disease and coarctation of the aorta are predisposing factors to aneurysms.

2. Strokes associated with underlying systemic diseases

   a. **Congenital heart disease** (particularly cyanotic heart disease)
      i. Stroke may occur to a *right to left shunt* that allows emboli to bypass the lungs and enter the arterial circulation of the brain.
      ii. During surgical procedures requiring *cardiac bypass*, air emboli, foreign material, and thrombi can yield neurologic deficits.
      iii. **Venous thrombi**
      iv. **Polycythemia**

   b. **Purulent venous thrombosis** (secondary to pyogenic infections of the mastoids, paranasal sinuses, scalp, or face)
      i. **Lateral sinus thrombosis** can present with increased intracranial pressure and abducens (cranial nerve VI) paralysis
      ii. **Sagittal sinus thrombosis** can present with increased intracranial pressure and evolving neurologic signs.
      iii. **Cavernous sinus thrombosis** is classically associated with proptosis, vascular engorgement of the bulbar conjunctivae, retinal hemorrhages, and extraocular muscle palsies.

   c. **Trauma**
      i. *Direct injury to the head*
      ii. *Injury to the neck* with intraoral damage (e.g., a traumatic injury to the posterior pharyngeal wall) can yield a dissecting aneurysm of the carotid vessels. With traumatic injuries to the head and neck, there is typically a latent period of 2 to 24 hours, followed by the onset of hemiparesis in association with somnolence and increased intracranial pressure; this latency period is probably because the trauma causes an intimal tear in a major artery, followed by dissecting aneurysm formation and thrombosis of the vessel.
d. **Sickle cell anemia:** Acute hemiparesis is a complication of older children and adolescents with sickle cell disease, most likely secondary to thrombosis in the capillaries and venules of the white matter. Vasoocclusive crises of the cerebral vessels have also been theorized to contribute to the incidence of stroke in these children. Arterial thrombi in major vessels and intracranial hemorrhage occur rarely.

e. **Homocystinuria:** This is an autosomal recessive defect in methionine metabolism that manifests itself as mental retardation, dislocation of the lenses, and tall stature. The defect also affects platelet function, which can yield an arterial or venous occlusion. Further, homocystine deficiency causes endothelial damage and leads to increased platelet consumption.

f. **Rare causes of stroke in children**
   i. **Hematologic**
      (a) Thrombotic thrombocytopenic purpura and other consumption coagulopathies
      (b) Thrombocytosis
      (c) Polycythemia
   ii. **Cardiac disorders**
      (a) Arrhythmias
      (b) Bacterial endocarditis
      (c) Atrial myxoma
   iii. **Rheumatologic disorder**
      (a) Vasculitis (e.g., periarteritis nodosa, giant cell arteritis, Takayasu's arteritis)
      (b) Systemic lupus erythematosus
   iv. **Migraine**
      (a) Hemiplegic migraine
      (b) Basilar artery migraine
      (c) Alternating hemiplegia of childhood
   v. **Viral infections** (e.g., coxsackie A-9 encephalitis)
   vi. **Neurocutaneous disorders**
      (a) Neurofibromatosis
      (b) Sturge-Weber syndrome
      (c) Tuberous sclerosis
   vii. **Metastatic neoplasms**
      (a) Rhabdomyosarcoma
      (b) Neuroblastoma
      (c) Primary brain tumors
   viii. **Atherosclerotic disease**
      (a) Progeria
      (b) Hypercholesterolemias
      (c) Hyperlipidemias
   ix. **High-dose radiation to head and neck yielding occlusion or stenosis of the internal carotid arteries** (usually occurs 2 to 22 years after the course of radiotherapy)
   x. **Necrotizing angiitis associated with intravenous methaamphetamine abuse.**

Sudden Death Among Young People

Although an uncommon phenomenon among children and young adults, sudden death is a startling and complex problem that presents itself with a frequency of 1.3 to 8.5 per 100,000 patient years. One-third to one-half of these deaths are secondary to cardiac disease. Congenital cardiac disease is a more common cause of sudden death during infancy and early childhood. Hypertrophic cardiomyopathy and precocious atherosclerosis are more frequent causes of sudden death among adolescents and young adults.

Common Causes of Sudden Death in Young Persons

<table>
<thead>
<tr>
<th>Noncardiac causes</th>
<th>Occult “unexpected cardiac causes”</th>
</tr>
</thead>
<tbody>
<tr>
<td>Toxic substance abuse</td>
<td>Conduction-system abnormality</td>
</tr>
<tr>
<td>Abdominal hemorrhage</td>
<td>Heart block (primary or secondary)</td>
</tr>
<tr>
<td>Cerebral hemorrhage</td>
<td>Sinus-node dysfunction (primary or secondary)</td>
</tr>
<tr>
<td>Pulmonary disease or abnormality</td>
<td>Ventricular tachyarrhythmia</td>
</tr>
<tr>
<td></td>
<td>Myocardial tumor</td>
</tr>
<tr>
<td>Pre-existing “known and clinical diagnosable cardiac causes”</td>
<td>Right ventricular dysplasia</td>
</tr>
<tr>
<td>Myocarditis</td>
<td>QT prolongation syndrome</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>Primary arrhythmia</td>
</tr>
<tr>
<td>Mitral-valve prolapse</td>
<td>Wolff-Parkinson-White syndrome</td>
</tr>
<tr>
<td>Major congenital heart lesions</td>
<td>Coronary arteritis or precocious atherosclerosis</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>Intramural coronary artery</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Anomalous origin of left coronary artery from pulmouary trunk</td>
</tr>
<tr>
<td>Ebstein’s anomaly</td>
<td>Aberrant origin from “wrong” sinus of Valsalva</td>
</tr>
<tr>
<td>Pulmonary vascular obstruction (primary or secondary)</td>
<td>Dissecting aortic aneurysm as a result of Marfan’s disease</td>
</tr>
<tr>
<td>Cystic medial necrosis</td>
<td></td>
</tr>
</tbody>
</table>


Sun Exposure: To Have or Have Not

We have entered a new era. Fewer and fewer people covet the once sought after “healthy glow” of a summer tan. As reports surface about the risks of a blistering burn before the third decade, more parents want advice about sun exposure. As a pediatrician, be prepared to answer questions about summer sun intensity and keep a mental list of the diseases and drugs that predispose young (and old) skin to photosensitivity.
Advise parents that the sun is at its maximum intensity between the hours of 11 A.M. and 3 P.M. and offer this handy rule of thumb: “When your shadow is shorter than you are tall, the sun is more likely to burn you than at other times, so seek protection with proper clothing, shade, sun screens or other means” (Lancet 1:44, 1990).

**Diseases Exacerbated or Precipitated by Sunlight**

**Viral**
- Herpes simplex
- Certain viral exanthems

**Genetic and metabolic**
- Xeroderma pigmentosum
- Albinism
- Vitiligo
- Darier’s disease (pseudoxanthoma elasticum)
- Bloom’s syndrome
- Rothmund-Thompson Syndrome
- Certain porphyrias
- Hartnup’s disease
- Phenylketonuria

**Collagen vascular**
- Systemic lupus erythematosus
- Discoid lupus erythematosus
- Dermatomyositis

**Miscellaneous**
- Solar urticaria
- Hydroa aestivale (Hutchinson’s summer prurigo) and vacciniforme
- Photosensitive eczema
- Polymorphous light eruption

**Drugs Predisposing to Sunburn or Photoreaction**

**Antibiotics**
- Sulfonamides
- Tetracyclines
- Nalidixic acid
- Griseofulvin

**Acne preparations**
- Retinoids (topical and systemic)

**Antiepileptics**
- Hydantoins
- Trimethadione
- Barbiturates

**Other**
- Certain chemotherapeutic agents
- Antimalarials
- Phenothiazines
- Coal tars
- Psoralens
- Chlorothiazide diuretics


**SWEAT TEST**

**The False Positive Sweat Test**

A sweat chloride value in excess of 60 mEq/L is generally considered diagnostic of cystic fibrosis. In adults, the normal range may be somewhat higher, i.e., 60–80 mEq/L. But even when the sweat test has been carefully performed (and this is not always the case), other causes of an elevated sweat chloride must be carefully considered before making a diagnosis. These other causes include:
Adrenal insufficiency, untreated
Ectodermal dysplasia
Hereditary nephrogenic diabetes insipidus
Glucose-6-phosphatase deficiency
Pupillatonia, hyporeflexia, and segmental hypohydrosis with autonomic dysfunction

False-negative sweat chloride results may be caused by edema. A diagnosis of cystic fibrosis should not be made on the basis of a positive sweat test alone. At least one of the following four criteria must also be present:

1. Documented family history of cystic fibrosis
2. Chronic pulmonary disease
3. Pancreatic insufficiency
4. A genotype consistent with the diagnosis.

SYNCOPE

The Work-up of Syncopal Episodes

The symptom of syncope has been defined as the reversible, atraumatic loss of consciousness and is usually associated with an inability to stand upright. This reaction can be due to some underlying impairment in cardiac output resulting in diminished cerebral perfusion. Careful evaluation of the patient presenting with syncopal episodes is clearly warranted, because that person may be at risk for injury, toward himself or others, especially if engaged in some activity such as driving, playing sports, crossing the street, and so on. Furthermore, patients who are having syncopal attacks secondary to some form of cardiac impairment are at risk for serious arrhythmias and sudden death.

1. The Etiology of Syncope
   a. Vascular/reflex
      Vasodepressor, orthostatic hypotension, cough, micturition, swallow, migraine, Takayasu disease, hyperventilation, carotid sinus, pregnancy, anemia, volume loss.
   b. Psychologic
      Hysteria, hyperventilation, fearful or threatening stimuli
   c. Cardiac
      Obstruction, arrhythmia, heart block, myocarditis, cardiomyopathy, mitral valve prolapse, pericardial effusion, prolonged QT syndrome, coronary anomaly, pulmonary artery hypertension, right ventricular dysplasia
   d. Neurologic
      Epilepsy, vertigo, central autonomic insufficiency (e.g., Riley-Day syndrome and Shy-Drager syndrome)
e. **Metabolic**

Low values for glucose, calcium, magnesium, or pO$_2$; abnormal values for sodium, potassium, or chloride

f. **Drugs**

Tricyclic antidepressants, antihypertensives, diuretics, barbiturates, phenothiazines, nitrates, cocaine, and other drugs of abuse.

2. **The Prodrome in Vasovagal Syncope**

(Vascular/reflex/psychologic categories of diagnosis account for the vast majority of syncopal episodes in both children and adults)

a. **Physiologic factors**
- Hunger
- Fatigue
- Illness
- Hot, crowded rooms
- Pain
- Anxiety
- Perceived threat
- Sight of blood

b. **Symptoms**
- Pallor/clammy skin
- Sweating
- Dilated pupils
- Blurred vision
- Nausea/epigastric distress
- Lightheadedness
- Dizziness
- Weakness

3. **The Reflex of Physiology in Syncopeal Episodes**

a. Recall the following important equations in cardiac physiology:
   i. \[ \text{Heart rate} \times \text{stroke volume} = \text{cardiac output} \]
   ii. \[ \text{Cardiac output} \times \text{total peripheral resistance} = \text{blood pressure} \]
   iii. \[ \text{Heart rate} \times \text{stroke volume} \times \text{total peripheral resistance} = \text{blood pressure} \]

b. Factors that determine heart rate:
   - Vagal tone (inhibitory)
   - Catecholamines (stimulant)
   - Sympathetic tone (stimulant)

c. Factors that determine stroke volume:
   - Circulating blood volume
   - Venous return (e.g., muscle tone, respiratory motion, tissue pressure, pregnancy)
   - Sympathetic tone

d. Factors that determine total peripheral resistance:
   - Baroreceptor tone (e.g., carotid sinus, aortic arch)
   - Arteriolar tone (as determined by electrolyte balance, catecholamines, and autonomic tone)
   - Drugs

4. **Primary Workup for Patient with Syncope**

Although the etiology of syncope may frequently be revealed from an accurate history, a number of laboratory tests are available when the history alone is not sufficient. The following table (Table 1) summarizes the primary workup of these patients:
Table 1. Primary Work-up for Patient with Syncope*

<table>
<thead>
<tr>
<th>ATHLETE WITH SYNCOPE</th>
<th>PATIENT WITH CARDIAC SYNCOPE</th>
<th>PATIENT ON DRUGS</th>
<th>PATIENT WITH NEUROLOGIC PSYCHIATRIC PROBLEMS</th>
<th>PATIENT WITH RECURRENT SYNCOPE OR SYNCOPE OF UNDETERMINED ORIGIN</th>
<th>PATIENT WITH REFLEX SYNCOPE</th>
</tr>
</thead>
<tbody>
<tr>
<td>History</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Physical examination</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>ECG</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Laboratory: CBC, electrolytes, glucose, Ca²⁺, Mg²⁺</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Holter monitor</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
</tr>
<tr>
<td>Hyperventilation test</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td></td>
</tr>
<tr>
<td>Serum bicarbonate</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td></td>
<td></td>
</tr>
<tr>
<td>EEG</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Carotid sinus massage</td>
<td></td>
<td>XX</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Telemetry</td>
<td>XX</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Echocardiogram</td>
<td>XX</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stress test</td>
<td>XX</td>
<td>XX</td>
<td>XX</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intracardiac electrophysiology study</td>
<td>XX</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Specific laboratory serum for drugs</td>
<td>XX</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*X indicates test, technique, or procedure that should be used in all patients with syncope. This is the initial screen to help sort the patients into one of the six general categories (column heads). XX indicates additional testing that may be required based on the results of the initial screen.

As described by Branch, “fainting spells share the common mechanism of transient inadequacy of cerebral perfusion due to inappropriate vasodilatation with pooling of blood in the extremities. They share common characteristics of being brief, usually without adverse consequences, and usually occurring while the patient is standing, sometimes while sitting, and rarely if ever while recumbent. Diagnostic clues are provided by the setting, the onset, the patient’s appearance, and the recovery (Table 2).”

Table 2. Differentiating Vasovagal Syncope from Seizure from Cardiac Syncope

<table>
<thead>
<tr>
<th>VASOVAGAL SYNCOPE</th>
<th>SEIZURE</th>
<th>CARDIAC SYNCOPE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Prodromal weakness, nausea, diaphoresis, lasting seconds to minutes</td>
<td>Sudden onset, or brief aura: deja vu, olfactory, gustatory, visual, etc.</td>
</tr>
</tbody>
</table>

Table continued on next page.
Table 2. Differentiating Vasovagal Syncope from Seizure from Cardiac Syncope (Cont.)

<table>
<thead>
<tr>
<th>Typical settings</th>
<th>VASOVAGAL SYNCOPE</th>
<th>SEIZURE</th>
<th>CARDIAC SYNCOPE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Emotionally upset,</td>
<td>Emotional upset,</td>
<td>Any setting, including</td>
<td>Any setting, often</td>
</tr>
<tr>
<td>prolonged standing,</td>
<td>prolonged standing,</td>
<td>sleep, sometimes blinking lights, monot-</td>
<td>without warning</td>
</tr>
<tr>
<td>uncomfortable sur-</td>
<td>uncomfortable sur-</td>
<td>onous music</td>
<td></td>
</tr>
<tr>
<td>roundings, or on first</td>
<td>roundings, or on first</td>
<td></td>
<td></td>
</tr>
<tr>
<td>arising with full</td>
<td>arising with full</td>
<td></td>
<td></td>
</tr>
<tr>
<td>bladder</td>
<td>bladder</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Occurrence</td>
<td>Only when upright</td>
<td>Any position</td>
<td>Any position</td>
</tr>
<tr>
<td>Appearance</td>
<td>Pallor, weak pulse</td>
<td>Cyanosis, stertorous breathing</td>
<td>Pallor, variable pulse</td>
</tr>
<tr>
<td>Residuum</td>
<td>Rapid recovery but</td>
<td>Prolonged recovery with postictal state, Todd's paresis</td>
<td>Recovery may be rapid</td>
</tr>
<tr>
<td></td>
<td>may recur on standing, occasional brief clonic movements, or urinary incontinence</td>
<td></td>
<td>or prolonged; if cardiac arrest: seizure-like activity, signs of cerebral hypoxia</td>
</tr>
</tbody>
</table>


SYNDROMES AND EPONYMS

Despite mounting opposition in some quarters, the use of eponymic names for syndromes, diseases, signs, et al. is likely to continue during all our lifetimes. The date and source of the originally published or reported description connected to the eponym (not always the first published description) are interesting, both for old (e.g., Pott's, 1779) and new (e.g., Kawasaki, 1974) syndromes and diseases. Listed below is a sampling:

Addison's disease.

Budd-Chiari syndrome.

Calvé-Legg-Perthes syndrome.

**Cockayne’s syndrome.**


**Down’s syndrome.**


**Ebstein’s anomaly.**


**Ehlers-Danlos syndrome.**

Ehlers E: Cutis laxa, Neigung zu Haemorrhagien in der Haut, Lockerung mehrerer Artikulationen. (Case for diagnosis.) Derm Zschr 8:173-174, 1901.


**Tetralogy of Fallot.**


**Fitz-Hugh and Curtis syndrome.**


**Goodpasture’s syndrome.**


**Guillain-Barré syndrome.**


**Hodgkin’s disease.**


**Kawasaki disease.**

Klinefelter's syndrome.

de Lange syndrome.

Marfan's syndrome.

Meckel's diverticulum.

Ménière's syndrome.

Mibelli's disease.

Münchausen's syndrome.

Niemann-Pick disease.

Pott's disease.
Pott P: Remarks on that kind of palsy of the lower limbs which is frequently found to accompany a curvature of the spine and is supposed to be caused by it, together with its method of cure. London, Johnson, 1779.

Reye's syndrome.

Riley-Day syndrome.

Pierre Robin syndrome.
Schönlein-Henoch purpura.

Stevens-Johnson syndrome.

Whipple’s disease.

Wilson’s disease.

Wolff-Parkinson-White syndrome.


Who Was Down?

John Langdon Down was born near Plymouth, England on November 18, 1828. He enrolled as a medical student at the London Hospital in 1853 and obtained a doctorate in 1859, after receiving the university gold medal for physiology. In the same year—1859—he was appointed medical superintendent of the Eastwood Asylum for idiots, at Redhill, Surrey, England, a post he held for 10 years and where he wrote a paper titled “Observations on an ethnic classification of idiots.” He noted that many of his patients had similar clinical features and described them as follows:

The face is flat and broad and destitute of prominence. Cheeks are roundish and extended laterally. The eyes are obliquely placed and the internal canthi more than normally distant from one another. The palpebral fissure is very narrow. The lips are large and thick with transverse fissures. The tongue is long, thick and much roughened. The nose is small.

He stated that “Their resemblance to each other was such that, when placed side by side, it is difficult to believe that they are not the children of the same parents.

Down’s work at Eastwood brought him much recognition, and in 1869 he was able to establish an institution at Redhill, Surrey for mentally retarded children of the wealthy. He named it Normansfield after his friend, Norman Wilkinson. At Normansfield, Down wrote his monograph titled Mental Affections of Childhood and Youth, published in 1887, which contained the classic description of Down’s syndrome. He also mentioned adrenogenital dystrophy, which
Syphilis

subsequently gained recognition as Fröhlich's syndrome. Down worked at Normansfield until his death in 1896.

For about 100 years the term "mongolism" was used as the primary descriptive name for Down's syndrome, with the eponyms "Down's" and "Langdon-Down's" used as alternatives, the hyphenated form having been preferred by Down in his later life. However, controversy eventually arose because some regarded the reference to the Mongol ethnic group as insulting, and in 1965 representatives of the Mongolian People's Republic in the World Health Organization approached the Director General and petitioned him to abandon the term "mongolism." Their request was accepted, and the eponym Down's syndrome was adopted.


SYPHILIS

Approaching Congenital Syphilis

The diagnosis of congenital syphilis, like the diagnosis of many congenital infections, is often confounded by the absence of symptoms or signs in the newborn, as well as by the difficulty in interpreting neonatal serologic responses to infection. The abrupt rise in the number of reported cases of congenital syphilis in the late 1980s, however, has increased the need for guidelines that will insure the detection and appropriate management of newborns with this treatable disease. The tables and figures below represent the recommended approach for surveillance, diagnosis, evaluation, and treatment of congenital syphilis.

Congenital syphilis may be missed if serologic tests are not performed for both the mother and her infant at the time of delivery. Even when these tests are performed, some infants are not identified as having syphilis probably because the infection is very recent and there has been insufficient time for an antibody response to develop. Some infants with congenital syphilis of later onset do not present with a typical rash; therefore, at least in areas where the disease is prevalent, serologic tests for syphilis should be included in the evaluation of all febrile infants, even those with negative results on serologic testing at birth.

Table 1. Surveillance Case Definition for Congenital Syphilis

For reporting purposes, congenital syphilis includes cases of congenitally acquired syphilis in infants and children, as well as syphilitic stillbirths.

1. **Confirmed.** A confirmed case of congenital syphilis is a case in which *Treponema pallidum* is identified by darkfield microscopy, fluorescent antibody, or other specific stains in specimens from lesions, placenta, umbilical cord, or autopsy material.

Table continued on next page.
Table 1. Surveillance Case Definition for Congenital Syphilis (Cont.)

2. Presumptive. A presumptive case of congenital syphilis is either of the following:
   a. Any case in which the infant’s mother had untreated or inadequately treated* syphilis at delivery, regardless of findings in the infant
   b. Any case in which the infant or child is reactive to a treponemal test for syphilis and in which any one of the following is present:
      i. Any evidence of congenital syphilis on physical examination (Table II)
      ii. Any evidence of congenital syphilis on a long bone radiograph
      iii. Reactivity to a CSF VDRL test*
      iv. Elevated CSF cell count or protein (without other cause)*
      v. Quantitative nontreponemal serologic titer that is fourfold higher than the mother’s (both specimens drawn at birth)
      vi. Reactive test for FTA-ABS 19S-IgM antibody*

3. Syphilitic stillbirth. A syphilitic stillbirth is defined as a fetal death in which the mother had untreated or inadequately treated syphilis at delivery of a fetus after a 20-week gestation or of a fetus weighing more than 500 gm.


*Inadequate treatment consists of any nonpenicillin therapy or penicillin given less than 30 days before delivery.

*It may be difficult to distinguish between congenital and acquired syphilis after infancy. Signs may not be obvious and stigmata may not yet have developed. Abnormal values of CSF VDRL test cell count, and protein, as well as IgM antibodies, may be found in either congenital or acquired syphilis. Findings on long bone radiographs may help to indicate congenital syphilis. The diagnosis may ultimately be based on maternal history and clinical judgment; the possibility of sexual abuse also needs to be considered.

Table 2. Evaluation for Early Congenital Syphilis

| 1. Maternal history, including results of serologic testing and treatment |
| 2. Thorough physical examination |
| 3. Long-bone radiographs |
|   a. Diaphyseal periostitis |
|   b. Osteochondritis |
|   c. Wimberger sign |
| 4. Nontreponemal antibody titer |
|   a. VDRL test (simultaneous quantitative serum titer for mother and neonate) |
| 5. Treponemal antibody titer |
|   FTA-ABS test |
|   FTA-ABS on 19S-IgM fraction of serum (CDC) |
| 6. CSF analysis |
|   a. Cell count |
|   b. Protein level determination |
|   c. VDRL test |
| 7. Other tests as clinically indicated |
|   a. Chest radiography |
|   b. Complete blood cell count |
|     i. Leukemoid reaction with or without monocytosis or lymphocytosis |
|     ii. Coombs negative hemolytic anemia |
|   c. Platelet count |
|     i. Thrombocytopenia |
|   d. Liver function tests |
|   e. Urinalysis |
| 8. HIV antibody test |
Figure 1. Algorithm for management of newborn infant born to mother with positive nontreponemal (VDRL or rapid plasma reagin) test result.
Table 3. Recommended Antimicrobial Treatment Regimens for Infants Born to Mothers with Positive VDRL Test Result

1. For confirmed or presumptive congenital syphilis (either item A or item B)
   a. Crystalline penicillin G, 100,000 to 150,000 units/kg/day administered intravenously in divided doses every 8-12 hours for 10-14 days
   b. Procaine penicillin G, 50,000 units/kg/day administered once daily intramuscularly for 10-14 days

2. Recommended only for infants at low risk for congenital syphilis who were born to HIV-seronegative mothers adequately treated for syphilis and in whom close follow-up cannot be ensured.
   a. Benzathine penicillin G, 50,000 units/kg (administered intramuscularly as one-time dose)

FOLLOW-UP FOR UNTREATED INFANTS:
VDRL AND FTA-ABS TITER AT 1, 2, 4, 6, and 12 MONTHS
- VDRL TITER IS DECREASING BY 3-4 MONTHS OF AGE, and
- VDRL IS NEGATIVE BY 6 MONTHS OF AGE, and
- FTA-ABS IS NEGATIVE BY 12 MONTHS OF AGE

YES ▼ NO
UNINFECTED REEVALUATE AND RE-TREAT WITH A REGIMEN RECOMMENDED FOR SYPHILIS OF MORE THAN 1 YEAR’S DURATION

FOLLOW-UP FOR TREATED INFANTS:
VDRL TITER AT 1, 2, 4, 6 and 12 MONTHS, and
CSF ANALYSIS EVERY SIX MONTHS
- VDRL TITER IS DECREASING BY 3-4 MONTHS OF AGE, and
- VDRL IS NEGATIVE BY 6 MONTHS OF AGE, and
- CSF VDRL IS NEGATIVE BY 6 MONTHS OF AGE, and
- CSF CELL COUNT IS DECREASING, and
- CSF CELL COUNT IS NORMAL BY 2 YEARS OF AGE

YES ▼ NO
YES ▼ NO
ADEQUATELY TREATED

Figure 2. Follow-up management for an infant examined or treated for congenital syphilis.

MY HEART LEAPS UP

My heart leaps up when I behold
A rainbow in the sky:
So was it when my life began;
So is it now I am a man;
So be it when I shall grow old,
Or let me die!
The Child is father of the Man;
And I could wish my days to be
Bound each to each by natural piety.

William Wordsworth
Eruption of Deciduous Teeth

The eruption of the first tooth in an infant is accompanied by parental pride in the fact that yet another milestone is reached. The figure below indicates the age and the order in which deciduous teeth erupt. The dot represents the mean age, whereas the wavy line demonstrates normal variation. Exceptions to the sequence of eruption are uncommon. Late eruption is unlikely to be of significance; however, it has been associated with both hypothyroidism and rickets.


TENNIS ELBOW

Tennis Elbow in Breastfeeding Mothers

Lateral epicondylitis or tennis elbow has been described in a variety of patients. It is classically seen when a person engages in repetitive, similar movements of the forearm extensor muscles, e.g., the continuous and monotonous swinging of tennis racket. The pain of lateral epicondylitis is particularly exacerbated by putting tension on the origin of the forearm extensor muscle, such as the active dorsiflexion of the wrist while grasping an object. Tennis elbow has also been
reported to be secondary to infections, trauma, arthritis, and peripheral neuropathy. Recently, however, a new etiology of lateral epicondylitis has been noted—among breastfeeding mothers who used hand-operated breast pumps improperly. The figure below demonstrates improper and proper body mechanics while using a piston style breast pump in order to avoid this new entity: breast pump-induced tennis elbow.

A. Subject demonstrating improper body mechanics while using a piston style breast pump. Note the flexion and abduction at the shoulder, pronation of the forearm, and dorsiflexion at the wrist resulting in the prominent bulge of the contralateral sternocleidomastoid muscle (arrow) compensating to maintain proper body alignment. B. Subject demonstrating proper body mechanics with shoulder adducted and lying comfortably against the body, forearm in supination, and wrist slightly flexed.

TERMINOLOGY

What Is Meant By “Usually”?  

We all too often use words that lack precision. We, ourselves, may not have a clear idea of what we mean let alone our listener.

A group of 51 individuals, highly skilled or professional workers, were asked to quantitate a number of inherently imprecise terms. Listed below are the words and what the readers believed to be the occurrence rate signified by the terms. The mean value as well as two standard deviations from the mean are provided so that you can appreciate how diverse are the interpretations of these commonly used words.

<table>
<thead>
<tr>
<th>Term</th>
<th>Occurrence Rate</th>
<th>± 2 S.D. (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Always</td>
<td>100%</td>
<td>--</td>
</tr>
<tr>
<td>Almost always</td>
<td>89%</td>
<td>75-100</td>
</tr>
<tr>
<td>Usually</td>
<td>71%</td>
<td>35-100</td>
</tr>
<tr>
<td>Frequently</td>
<td>68%</td>
<td>42-93</td>
</tr>
<tr>
<td>Often</td>
<td>59%</td>
<td>28-92</td>
</tr>
<tr>
<td>Occasionally</td>
<td>20%</td>
<td>0-42</td>
</tr>
<tr>
<td>Infrequently</td>
<td>12%</td>
<td>0-28</td>
</tr>
<tr>
<td>Rarely</td>
<td>55%</td>
<td>0-17</td>
</tr>
<tr>
<td>Never</td>
<td>0%</td>
<td>--</td>
</tr>
</tbody>
</table>

Since nothing is ever “never” or “always” you can see the problems you usually create in interpretation every time you use these imprecise words.


THALASSEMIAS

Classification of the Thalassemias

The thalassemias are a group of inherited blood disorders in which production of one or more of the hemoglobin polypeptide chains is diminished. The resultant erythrocytes produced in these disorders have a low intracellular hemoglobin content, or hypochromia, and are smaller in size than normal red blood cells (i.e., microcytosis). Further, the polypeptide globin chains that are produced in the patient with thalassemia are unstable and aggregate within the red blood cell, yielding membrane damage and early destruction both in the bone marrow and the peripheral circulation.

Classification of the thalassemias is based upon the type of globin chain, which is either absent or produced in diminished amounts.
Clinical Features of Thalassemias

<table>
<thead>
<tr>
<th>TYPE</th>
<th>CLINICAL FEATURES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alpha-thalassemia Type</td>
<td>No clinical stigmata</td>
</tr>
<tr>
<td>Silent carrier</td>
<td>Mild anemia; hypochromic and microcytic red cells</td>
</tr>
<tr>
<td>Thalassemia trait (heterozygous)</td>
<td></td>
</tr>
<tr>
<td>Hemoglobin H disease</td>
<td>Splenomegaly; moderate-to-severe hemolytic anemia; mild jaundice</td>
</tr>
<tr>
<td>Hydrops fetalis (homozygous)</td>
<td>Death in utero</td>
</tr>
<tr>
<td>Beta-thalassemia Type</td>
<td>No clinical stigmata</td>
</tr>
<tr>
<td>Silent carrier</td>
<td>Mild anemia; hypochromic and microcytic red cells</td>
</tr>
<tr>
<td>/3-thalassemia trait or minor (heterozygous)</td>
<td></td>
</tr>
<tr>
<td>Thalassemia intermedia</td>
<td>Splenomegaly and severe anemia. Skeletal deformities, frequent fractures and arthritis are complications.</td>
</tr>
<tr>
<td>/3-thalassemia major</td>
<td>Severe anemia incompatible with life unless regular blood transfusions are given.</td>
</tr>
</tbody>
</table>


Complications of /3-Thalassemia Major

/3-thalassemia major or Cooley’s anemia is one of the most serious of the thalassemias. Patients with this disease can only survive with frequent blood transfusions, careful attention to iron balance, and supportive therapy. The complications of /3-thalassemia major result mainly from (1) excessive hematopoesis; (2) chronic hemolysis; and (3) iron overload with resultant organ damage.

1. Complications due to excessive hematopoesis
   a. Marked bone marrow hypertrophy and cortical thinning
   b. Bony changes, particularly in the craniofacial area (“rodent facies”) secondary to maxillary overgrowth, protrusion of teeth, separation of the orbits, flattening of the nasal bridge, and malar prominence. These bony changes may yield:
      i. Chronic sinusitis
      ii. Impaired hearing
   c. Pathologic fractures (particularly in weight-bearing bones)
   d. Lymphadenopathy
   e. Hepatosplenomegaly

2. Complications due to chronic hemolysis
   a. Gallstones
   b. Leg ulcers (usually seen in late adolescence and early adulthood)

3. Complications due to iron overload as a result of chronic transfusion therapy (all tissues are affected by this iron overload, but the liver, spleen, and pancreas retain iron in the highest concentrations)
   a. Cardiac disease
      i. Pericarditis
      ii. Atrial and ventricular arrhythmias
      iii. Congestive heart failure
   b. Hepatic disease (hepatic fibrosis)
4. Growth and endocrine dysfunction
   a. Growth failure: Although children with thalassemia major on a chronic transfusion program grow normally until the age of 12, their growth velocity diminishes thereafter, and they fail to exhibit a pubescent growth spurt. Growth hormone levels are usually normal or elevated. The growth failure is probably secondary to chronic disease and iron overload.
   b. Delayed or incomplete sexual maturation
   c. Acquired hypothyroidism, hypoparathyroidism, and diabetes mellitus due to hemochromatosis.

Incidentally, Thomas Cooley, the Detroit pediatrician who first described thalassemia major in 1927, required only a microscope and his patients' peripheral smears to describe a disease that influenced the fields of hematology, human heredity, and population genetics.


THEOPHYLLINE

Factors That Alter Theophylline Clearance in Children

Theophylline remains the most commonly used drug for treating children with asthma in the U.S. Despite improved formulations of theophylline that provide a sustained release of the xanthine derivative, and the ability to measure serum theophylline levels, many children are extremely variable in their absorption, metabolism, and clearance of the drug.

Theophylline has a theoretical bioavailability of 100%, and its metabolism and clearance are controlled principally by the liver, where 90% of the drug is metabolized and then excreted in the urine. Any disease or condition affecting the metabolic machinery of the liver, therefore, will play a significant role in altering a patient's metabolism and absorption of theophylline.

Factors Affecting Theophylline Metabolism

<table>
<thead>
<tr>
<th>Factors that reduce clearance:</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Liver disease</td>
</tr>
<tr>
<td>2. Congestive heart failure: cor pulmonale</td>
</tr>
<tr>
<td>3. Prolonged fever, particularly from viral infection</td>
</tr>
<tr>
<td>4. Macrolide antibiotics such as erythromycin and troleandomycin</td>
</tr>
<tr>
<td>5. Cimetidine</td>
</tr>
<tr>
<td>6. Age less than 1 year</td>
</tr>
<tr>
<td>7. Influenza vaccine</td>
</tr>
<tr>
<td>8. Acute hypoxemia</td>
</tr>
<tr>
<td>9. High carbohydrate, low protein diet</td>
</tr>
<tr>
<td>10. Propranolol</td>
</tr>
<tr>
<td>11. Furosemide</td>
</tr>
</tbody>
</table>

Table continued on next page.
Factors Affecting Theophylline Metabolism (Cont.)

Factors that increase clearance
1. Tobacco or marijuana smoking
2. Charcoal broiled foods (consumed in large quantities over a long period of time)
3. Phenobarbital
4. Phenytoin
5. Isoproterenol
6. High protein/low carbohydrate diet
7. Pregnancy
8. Rifampin
9. Tegretol


THROMBOCYTOSIS

Significance of Thrombocytosis

In these days of automated cell counters, platelet counts are often determined whether we ask for them or not. As is often the case with unrequested and unnecessary laboratory studies, “abnormal” results are frequently reported. It turns out that platelet counts that previously would have been considered to be abnormally elevated are not particularly unusual among healthy pediatric patients. The figure below represents the distribution of platelet counts obtained from 805 ambulatory pediatric patients. Although the largest number of patients had platelet counts between 200,000 and 400,000/mm³, 12.9% of the children had counts of greater than 500,000/mm³, and 2% were greater than 700,000/mm³. Children with elevated platelet counts were most often completely healthy, but some had evidence of viral or bacterial infection, and they tended to be younger than the children with “normal” counts.

Distribution of platelet counts in 805 ambulatory pediatric patients.
Although "elevated" platelet counts among healthy children may be normal, extreme thrombocytosis is usually associated with a recognizable disease state. Among 94 children with platelet counts greater than 900,000/mm³, only one child was completely healthy. Recognized conditions associated with extreme thrombocytosis include the following:

- Infection of any kind
- Recovery from chemotherapy
- Iron deficiency
- Splenectomy
- Malignancies
- Respiratory distress
- Inflammatory diseases such as Kawasaki disease, juvenile rheumatoid arthritis, and anaphylactoid purpura
- Recent surgery
- Metabolic diseases
- Nephrotic syndrome


Thrombocytosis in Childhood

With the increased availability of electronic cell counting, increased platelet counts are being noted more frequently. Listed below are the major causes of platelet counts in excess of 900,000/mm³.

Infection
- Both viral and bacterial infections, particularly of the central nervous system.

Hematologic
- Conditions in this group include consequences of chemotherapy, iron deficiency anemia, chronic myelogenous leukemia, and the early postsplenectomy state.

Respiratory
- Respiratory distress syndrome, with or without bronchopulmonary dysplasia; severe respiratory obstruction.

Tissue damage or response to surgery
- Trauma, postoperative response

Collagen vascular disease
- Juvenile rheumatoid arthritis
- Wegener granulomatosis
- Anaphylactoid purpura

Metabolic diseases
- When complicated with acidosis and dehydration

Nephrotic syndrome
- An increase in platelet count can be viewed in many circumstances as an acute phase reactant.
- Thrombocytosis, infants and children, is rarely associated with thrombotic consequences and requires no therapy.

Tick-Related Infection

Ticks are small, but they are large enough to carry with them many even smaller microorganisms that they inject into unsuspecting human hosts. Most of us are familiar with the clinical characteristics of the tick-borne infections in our own locale, but we may not appreciate the importance of these diseases in patients who have traveled prior to their illness. It is important, first, to remember to think of tick-related infection, and then to consider tick activity in the area, the geographic distribution of ticks known to carry specific pathogens, sites of tick exposure, and signs and symptoms at presentation related to the time of exposure. The incubation period for most of the infections carried by ticks is 3 to 5 days to 2 weeks.

If you have trouble remembering the regional and clinical characteristics of tick-borne infection, the following table may help.

**Tick-borne Diseases in Children**

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>ORGANISM</th>
<th>VECTOR</th>
<th>RESERVOIR</th>
<th>GEOGRAPHIC DISTRIBUTION</th>
<th>TYPE OF ILLNESS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Babesiosis</td>
<td>Babesia microti</td>
<td><em>Ixodes dammini</em></td>
<td>Rodents</td>
<td>Coastal area, islands of Massachusetts, Rhode Island, New York</td>
<td>Malaria-like, fever, anemia, renal failure</td>
</tr>
<tr>
<td>Lyme disease</td>
<td>Borrelia burgdorferi</td>
<td><em>Ixodes dammini</em>, <em>I. pacificus</em>, <em>Amblyomma americanum</em></td>
<td>Migratory birds</td>
<td>Northeast, Midwest and Western United States*</td>
<td>Fever, rash (ECM), headache, myalgias, multiple stages</td>
</tr>
<tr>
<td>Tularemia</td>
<td>Francisella tularensis</td>
<td><em>Dermacentor andersoni</em>, <em>D. variabilis</em>, <em>Amblyomma americanum</em></td>
<td></td>
<td>Southern, Southwestern and Midwest United States</td>
<td>Fever, lymphadenopathy, pneumonia</td>
</tr>
<tr>
<td>Rocky Mountain spotted fever</td>
<td>Rickettsia rickettsii</td>
<td><em>Dermacentor andersoni</em>, <em>D. variabilis</em>, <em>Amblyomma americanum</em>, <em>Haemaphysalis leporis-palustris</em></td>
<td>Dogs, cats, rodents, rabbits</td>
<td>Western hemisphere, especially So. eastern United States</td>
<td>Fever, headache, myalgias, rash, toxicity</td>
</tr>
<tr>
<td>Erlichiosis</td>
<td><em>Ehrlichia canis</em></td>
<td><em>Rhipicephalus sanguineus</em></td>
<td>Dogs</td>
<td>Southern, Southwestern and Midwest United States, Japan</td>
<td>Fever, chills, myalgias, hematologic abnormalities, similar to RMSF</td>
</tr>
<tr>
<td>Relapsing fever</td>
<td><em>Ehrlichia nenensis</em></td>
<td><em>Rhipicephalus sanguineus</em>, <em>Ornithodoros moubata</em></td>
<td>Dogs, rodents, opossums, squirrels, armadillos</td>
<td>Western Mountains, Southern Plains, United States</td>
<td>Fever, chills, headache, myalgia, relapsing course</td>
</tr>
<tr>
<td>Queensland tick typhus</td>
<td><em>Rickettsia australis</em></td>
<td><em>Ixodid ticks</em></td>
<td>Rodents, dogs marsupials</td>
<td>Eastern Australia</td>
<td>Similar to RMSF, usually milder</td>
</tr>
<tr>
<td>Fievre boutonneuse</td>
<td><em>Rickettsia conorii</em></td>
<td><em>Ixodid ticks</em></td>
<td>Dogs, rodents</td>
<td>Worldwide</td>
<td>Similar to RMSF, usually milder</td>
</tr>
</tbody>
</table>

*a Recent evidence for widespread disease in United States.

*b Animals become ill with infection.

Table continued on next page.
Tick-borne Diseases in Children (Cont.)

<table>
<thead>
<tr>
<th>DISEASE</th>
<th>ORGANISM</th>
<th>VECTOR</th>
<th>RESERVOIR</th>
<th>GEOGRAPHIC DISTRIBUTION</th>
<th>TYPE OF ILLNESS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Asian tick typhus</td>
<td><em>Rickettsia siberica</em></td>
<td>Ixodid ticks</td>
<td>Dogs, rodents</td>
<td>Central Asia, Russia</td>
<td>Similar to RMSF, usually milder; regional lymphadenopathy</td>
</tr>
<tr>
<td>Q fever</td>
<td><em>Coxiella burnetii</em></td>
<td>All endemic species</td>
<td>Cattle, sheep, goats</td>
<td>Worldwide</td>
<td>Fever, headache, pneumonia</td>
</tr>
<tr>
<td>Colorado tick fever</td>
<td>Orbivirus</td>
<td><em>Dermacentor andersoni</em></td>
<td>Rodents, deer</td>
<td>Rocky Mountain states, Western Canada and Northern Sierras</td>
<td>Fever, headache, malaise, myalgias, leukopenia</td>
</tr>
<tr>
<td>Tick-borne encephalitis</td>
<td>Flavivirus</td>
<td><em>Ixodes persulcatus, Ixodes ricinus</em></td>
<td>Cattle, sheep, goats, rodents</td>
<td>Central Asia, Eastern Europe, Russia</td>
<td>Fever, headache, encephalitis, photophobia, hyperesthesias</td>
</tr>
<tr>
<td>Tick-bite granuloma</td>
<td></td>
<td>All species</td>
<td></td>
<td></td>
<td>Local reaction, granuloma, complement-mediated</td>
</tr>
<tr>
<td>Tick paralysis</td>
<td><em>Dermacentor andersoni</em>, <em>Dermacentor variabilis</em></td>
<td></td>
<td></td>
<td></td>
<td>Toxin-mediated, neurologic syndrome, ataxia, areflexia, ascending flaccid paralysis, mild fever</td>
</tr>
</tbody>
</table>

*Infection usually acquired by inhalation from animals; ticks important in animal transmission. ECM, erythema chronicum migrans.

Because the diagnosis of most of these infections depends upon serologic testing, which often must be performed in a reference laboratory, empiric antibiotic therapy is often required. Most of the tick-related infections can be treated with tetracycline or chloramphenicol; however, amoxacillin or penicillin are the appropriate alternatives to tetracycline in the child younger than 8 or 9 years with suspected Lyme disease.


**TORTICOLLIS**

**Common Causes**

Congenital, muscular, or vertebral anomalies

**Uncommon Causes**

Cervical adenopathy
Congenital nystagmus
Drug-induced (e.g., phenothiazines, haloperidol, metoclopramide, trimethobenzamide)
Paroxysmal

Pharyngitis
Retropharyngeal abscess
Secondary to reflux esophagitis
(Sandifer's syndrome)
Superior oblique muscle weakness

355 BEST COPY AVAILABLE
356—Tourette's Syndrome

Rare Causes

Calcification of intervertebral disks
Dystonia musculorum deformans
Eosinophilic granuloma of cervical vertebrae
Fibromyositis
Hepatolenticular degeneration
Juvenile rheumatoid arthritis
Kernicterus

Osteomyelitis of the cervical vertebrae
Pneumonia of an upper lobe
Posterior fossa tumor
Spasms nutans
Spinal tumor
Subluxation or dislocation of cervical vertebrae

TOURETTE'S SYNDROME

Tics can occur commonly during childhood. Several studies estimate that about 3% of all children exhibit tics at some time. These symptoms are most commonly transient, however, and present as excessive blinking or grimacing. Less common are transient nonspecific vocalizations (e.g., clearing of the throat, sniffing, or frequent coughing).

There are occasions when the symptoms may become more chronic and persistent. When limited to motor symptoms, the condition is called multiple chronic motor tic disorder. In the child between ages 2 and 14 presenting with both vocal and motor symptoms that wax and wane in severity over time and have been present for more than a year, the term Gilles de la Tourette's or Tourette syndrome (TS) is used. Its etiology, unfortunately, remains poorly understood. The types of tics seen in this fascinating syndrome are listed below:

The Symptoms

1. Motor tics
   Blinking
   Grimacing
   Shrugging
   Mouth-opening
   Head-jerking
   Tongue movements

2. Vocal tics
   Sniffing
   Coughing
   Clearing of the throat
   Hissing
   Barking
   Honking
   Snorting
   Squeaking

   Extending or flexing neck
   Jerking of trunk
   Jerking of extremities
   Tensing of abdominal muscles
   Kicking
   Lip-licking
   Burping
   Repetition of letters
   Repetition of words or phrases
   Involuntary cursing (coprolalia)*
   Clicking
   Whistling
   Spitting
   Shrieking

*Although Georges Gilles de la Tourette's original 1885 description (Arch Neur, Paris, 9:19 42, 158-200, 1885) stressed coprolalia as a cardinal feature, subsequent studies have noted a 20 to 35% incidence rate of this symptom among patients with Tourette syndrome. Its absence, therefore, does not contradict the diagnosis.
3. **Complex symptoms**

- Squatting
- Jumping
- Twirling
- Repetitive touching of objects and people
  
  Repetitive sniffing
  
  Obscene gestures (copropraxia)
  
  Head-banging
  
  Self-injury (biting, scratching)

**Diagnosis and Treatment**

A careful history and physical are warranted, because between 21% and 54% of children with TS have symptoms of attention deficit disorder (ADD). Further, there exists a strong relationship between ADD patients treated with stimulant medications such as methylphenidate (Ritalin) and the subsequent development or exacerbation of tics.

It should be stressed that all of the medications suggested for Tourette syndrome provide only symptomatic relief and are not curative. Further, no evidence exists to suggest that early therapy with these medications has any effect on the long-term prognosis of tic disorders. Many Tourette syndrome patients have symptoms mild enough not to require pharmacologic intervention. In light of the side-effects from the medications listed, many physicians prefer not to use them.

1. **Agents available**

   - Haloperidol
   - Pimozide
   - Fluphenazine
   - Clonidine
   - Clonazepam

2. **Potential side-effects of neuroleptic drugs**

   - Acute dystonic reactions
   - Parkinsonian symptoms
   - Anticholinergic symptoms
   - Sedation
     
     Tardive dyskinesia
     
     Increased appetite
     
     Depression
     
     Cognitive blunting
     
     School phobias


**TRACHEOESOPHAGEAL FISTULA**

Tracheoesophageal fistula (TEF) and esophageal atresia are the two types of esophageal malformations causing upper intestinal obstruction. TEF is the failure of the trachea and esophagus to divide linearly during embryogenesis. Esophageal atresia is the developmental occlusion of the esophagus in a localized segment of lumen. Both may present at birth with aspiration and both may occur as an isolated finding.

The diagnosis of tracheoesophageal fistula may be suggested by a variety of clinical observations. The five types of fistula are depicted below, along with the symptoms and signs that typically accompany them.
Tracheoesophageal Fistula

Type A
Symptoms and Signs:
Excessive mucus, aspiration of saliva.
Scaphoid abdomen.
No gas in bowel on x-ray.
Cannot pass catheter into stomach.
Gradually increasing respiratory distress.
Polyhydramnios.

Type B
Symptoms and Signs:
Polyhydramnios.
Coughing, choking and pneumonia from birth.
Scaphoid abdomen.
No gas in bowel on x-ray.

Type C
Symptoms and Signs:
Most common (80% of cases).
Excessive mucus.
Gradually increasing respiratory distress.
Polyhydramnios frequent but not severe.
Gas in bowel on x-ray.

Type D
Symptoms and Signs:
Coughing, choking, and pneumonia from birth.
Gas in bowel on x-ray.

Type E
Symptoms and Signs:
Difficult to diagnose.
Coughing or cyanosis with feeding.
Chronic aspiration pneumonia.

The differential diagnosis includes pharyngeal muscle weakness, vascular rings, and esophageal diverticula.

Discovery of a tracheoesophageal fistula should alert the physician to the possibility that other congenital anomalies may be present. Anomalies that have been found to be associated include:

Vertebral  Anal  Cardiac  Renal  Limb

UMBILICUS

Abnormalities of the Pediatric Umbilicus

During both the prenatal and neonatal periods, the umbilicus is a site of many embryologic and structural changes (Fig. 1). Consequently, there exists a wide variety of umbilical abnormalities that require accurate diagnosis and, potentially, subsequent treatment. These abnormalities can be divided in terms of congenital anomalies, infections, signs of remote or underlying disorders, and rare causes of malignancy. We use an anatomic approach to delineate these disorders.

**FIGURE 1.** Relationship of umbilical structures in the fetus to those in the infant. Left. The left umbilical vein and both umbilical arteries persist; Above. Cross-section of the cord. Structures may be compared between the fetus and the child.

1. Anatomical Disorders of the Cord and Umbilicus

   a. **Single umbilical artery**

      i. Found in 0.5 to 0.9% of all births, the single umbilical artery is more common in whites than blacks; it is also more common in females and has been associated with infants of diabetic mothers, certain trisomy anomalies, and with thalidomide use during pregnancy.
ii. There is a reported incidence of congenital anomalies associated with the single umbilical artery, including renal, cardiovascular, pulmonary, genitourinary, cerebrospinal, musculoskeletal, facial, and ocular abnormalities. The work-up of a single umbilical artery, therefore, should include a thorough physical examination, especially for dysmorphism, parental counseling, good follow-up for identification of problems that may not be obvious at birth, and special screening studies (e.g., for cardiac or renal anomalies) as indicated.

b. **Umbilical dysmorphism**
   i. Aarskog syndrome: short stature with facial, digital, and genital malformations and a prominent, protruding central portion of the umbilicus surrounded by a deep ovoid depression.
   ii. Reiger syndrome: goniodygenesis, hypodontia, and a broad prominent umbilicus with redundant umbilical skin.
   iii. Robinow syndrome: fetal face, short forearms with brachydactyly, genital hypoplasia, moderate dwarfing, and an abnormally high positioned, broad, and poorly epithelialized umbilicus
   iv. Beckwith-Wiedemann syndrome: macroglossia, gigantism, hyperinsulinemia and frequently associated with omphalocele.

c. **Disorders of the umbilical stump**
   i. Omphalitis is an infection of the stump, which is frequently followed by sepsis.
   ii. Persistent omphalomesenteric remnants or ectopic viscera in a retained stalk (usually the stump atrophies and separates 12–14 days after birth; a stalk still present at 3 to 4 weeks should be surgically explored and excised).
   iii. Umbilical or pyogenic granuloma (these should be ablated with silver nitrate)

d. **Incomplete obliteration of the omphalomesenteric duct** (Fig. 2)
   All of these entities require surgical evaluation and repair.
   i. Umbilical polyp
   ii. Umbilical sinus
   iii. Omphalomesenteric cyst (cysts are usually not diagnosed unless they undergo torsion, become infected, or enlarge when distended by secretions)
   iv. Fibrotic bands between the umbilicus and intestine
   v. Meckel’s diverticulum
   vi. Omphalomesenteric fistula (persistent vitelline duct)

e. **Incomplete obliteration of the allantois** (Fig. 3)
   These entities require surgical repair.
   i. Urachal sinus
   ii. Urachal cyst
   iii. Urachal diverticulum
   iv. Patent urachus

f. **Abnormalities of the umbilical ring**
   Embryologically, the umbilical ring must constrict and close after the intestine has migrated into the abdominal cavity.
   i. Umbilical hernia (over 85% will close without surgical repair)
FIGURE 2. Omphalomesenteric anomalies: A, an umbilical polyp; B, umbilical sinus; C, an omphalomesenteric cyst, D, fibrotic bands between the umbilicus; E, Meckel's diverticulum; F, an omphalomesenteric fistula.

ii. Gastrochisis is a defect in the abdominal wall of an extraumbilical location. It may represent a rupture, in utero, of an umbilical hernia at a weak point. Gastrochisis requires surgical treatment and closure to return the abdominal contents to their proper space.

iii. Omphalocele results from a failure of the intestine to return from the vitelline duct to the abdomen. The omphalocele is covered by a thin, membranous sac, which encloses the intestine and often allows the liver with the cord arising from it. Risks include infection, dehydration, and hypothermia, and surgical closure is indicated. Both omphalocele and gastrochisis have a high association with other congenital anomalies (e.g., gastrointestinal, cardiac, renal, and chromosomal).
2. Other Disorders of the Umbilicus

a. Umbilical tumors
   i. Primary sarcoma
   ii. Primary hemangiomas
   iii. Arteriovenous malformations

b. Intraabdominal hemorrhage can present with a bluish discoloration of an umbilical hernia (Hoffstater’s sign)

c. Acute pancreatitis can be heralded by periumbilical bruising (Cullen’s sign).

References: Adapted from Black CT: Disorders of the pediatric umbilicus. Resident and Staff Physician 35:64-84, 1989, with permission.
Upper Airway Obstruction—363

UPPER AIRWAY OBSTRUCTION

Infectious Causes of Upper Airway Obstruction:
Distinguishing the Features of Viral Croup,
Epiglottitis, and Bacterial Tracheitis

The child presenting to the clinic or emergency room with upper airway obstruction demands immediate attention. There exist many causes of acute upper airway obstruction, including foreign bodies, diphtheria, infectious mononucleosis, and measles. More chronic or recurrent causes of stridor and upper airway obstruction include vascular rings, congenital heart disease, tracheal stenosis from previous intubation, severe allergic reactions leading to laryngospasm, and recurrent angioneurotic edema. The most common causes of potentially life-threatening upper airway obstruction, however, are infectious in origin: (1) viral laryngotracheobronchitis or croup; (2) epiglottitis; and (3) bacterial tracheitis. A prompt diagnosis, obviously, requires a sound knowledge of their distinguishing features in order to assure proper medical management.

Clinical Features of Viral Croup, Epiglottitis, and Bacterial Tracheitis

<table>
<thead>
<tr>
<th>CLINICAL FEATURE</th>
<th>VIRAL CROUP</th>
<th>EPIGLOTTITIS</th>
<th>BACTERIAL TRACHEITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Site of airway obstruction</td>
<td>Infraglottis</td>
<td>Supraglottis</td>
<td>Infraglottis</td>
</tr>
<tr>
<td>Patient age Peak</td>
<td>2 yrs</td>
<td>3-6 yrs</td>
<td>2 yrs</td>
</tr>
<tr>
<td>Range</td>
<td>8 mo 5 yr</td>
<td>17 mo adult</td>
<td>1 mo 9 yr</td>
</tr>
<tr>
<td>Sex</td>
<td>M &gt; F (2:1)</td>
<td>M = F</td>
<td>M = F</td>
</tr>
<tr>
<td>Duration of illness prior to admission (hours)</td>
<td>12-78 hrs</td>
<td>2-48 hrs</td>
<td>24-96 hrs</td>
</tr>
<tr>
<td>Prodrome</td>
<td>Viral URI</td>
<td>Uncommon</td>
<td>Viral URI</td>
</tr>
<tr>
<td>Clinical features on presentation</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stridor</td>
<td>Common</td>
<td>Uncommon</td>
<td>Common</td>
</tr>
<tr>
<td>Barking cough</td>
<td>&gt;60%</td>
<td>Uncommon</td>
<td>&gt;50%</td>
</tr>
<tr>
<td>Temperature (°C)</td>
<td>37.8</td>
<td>38.6</td>
<td>39.2</td>
</tr>
<tr>
<td>Hoarseness</td>
<td>20%</td>
<td>Uncommon</td>
<td>None</td>
</tr>
<tr>
<td>Retractions</td>
<td>Common</td>
<td>Uncommon</td>
<td>Varied</td>
</tr>
<tr>
<td>Wheezing</td>
<td>5%</td>
<td>None</td>
<td>10%</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>10%</td>
<td>20%</td>
<td>10%</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>None</td>
<td>10%</td>
<td>None</td>
</tr>
<tr>
<td>Drooling</td>
<td>None</td>
<td>10%</td>
<td>None</td>
</tr>
<tr>
<td>Appearance</td>
<td>Lying down, non-toxic</td>
<td>Sitting up, toxic</td>
<td>Varied</td>
</tr>
<tr>
<td>Season</td>
<td>Late spring; late fall</td>
<td>Year round</td>
<td>Year round</td>
</tr>
<tr>
<td>Progression</td>
<td>Slow</td>
<td>Rapid</td>
<td>Varied: slow rapid</td>
</tr>
<tr>
<td>WBC range</td>
<td>5 11,000</td>
<td>16 22,000</td>
<td>8 20,000</td>
</tr>
<tr>
<td>PMN range (%)</td>
<td>40 80</td>
<td>60 95</td>
<td>40 80</td>
</tr>
<tr>
<td>Mean bands (%)</td>
<td>7</td>
<td>30</td>
<td>30</td>
</tr>
</tbody>
</table>

Table continued on next page.
Clinical Features of Viral Croup, Epiglottitis, and Bacterial Tracheitis (Cont.)

<table>
<thead>
<tr>
<th>CLINICAL FEATURE</th>
<th>VIRAL CROUP</th>
<th>EPIGLOTTITIS</th>
<th>BACTERIAL TRACHEITIS</th>
</tr>
</thead>
<tbody>
<tr>
<td>X-ray findings</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Subglottic narrowing (%)</td>
<td>90–100</td>
<td>None</td>
<td>60–100</td>
</tr>
<tr>
<td>Enlarged epiglottis (%)</td>
<td>None</td>
<td>100</td>
<td>Rare</td>
</tr>
<tr>
<td>Infiltrate on admission (%)</td>
<td>10</td>
<td>&lt;30</td>
<td>&gt;50</td>
</tr>
<tr>
<td>Infiltrate during admission (%)</td>
<td></td>
<td>&lt;30</td>
<td>100</td>
</tr>
<tr>
<td>Response to racemic epinephrine (%)</td>
<td>95</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Positive tracheal cultures (%)</td>
<td>0</td>
<td>70 (epiglottis)</td>
<td>100</td>
</tr>
<tr>
<td>Bacteria</td>
<td>100</td>
<td>0</td>
<td>50</td>
</tr>
<tr>
<td>Viral</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Positive blood cultures (bacteria) (%)</td>
<td>0</td>
<td>90</td>
<td>0</td>
</tr>
<tr>
<td>Hospitalization (%)</td>
<td>10</td>
<td>100</td>
<td>100</td>
</tr>
<tr>
<td>Respiratory arrest</td>
<td>Rare</td>
<td>Increased risk</td>
<td>25%</td>
</tr>
<tr>
<td>Treatment (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intubation</td>
<td>3</td>
<td>99</td>
<td>65</td>
</tr>
<tr>
<td>Tracheostomy</td>
<td>&lt;1</td>
<td>1</td>
<td>30</td>
</tr>
<tr>
<td>Mean duration of hospitalization (days)</td>
<td>7</td>
<td>5</td>
<td>21</td>
</tr>
<tr>
<td>Recurrence</td>
<td>5%</td>
<td>Rare</td>
<td>Rare</td>
</tr>
<tr>
<td>Etiologic agents</td>
<td>Parainfluenza type 1, 2, 3; respiratory syncytial virus; rhinovirus</td>
<td><em>Haemophilus influenzae</em> type b, <em>β</em>-hemolytic streptococci</td>
<td><em>S. aureus; Haemophilus influenzae</em> type b; *streptococcus, group A; β- and hemolytic streptococci; Neisseria; E. coli; others</td>
</tr>
</tbody>
</table>


URINALYSIS

Dangers of the Dipstick

The urine dipstick is an extremely quick and convenient method of screening for pH, protein, glucose, hemoglobin, ketones, bilirubin, and urobilinogen. The dipstick is a screening test, however, and its limitations must be kept in mind. The situations in which false positive and false negative results may be obtained are listed below:

**pH.** This determination is not altered except when the pH of the urine is acutally altered by drugs.

**Protein.** A highly alkaline urine may cause a false positive dipstick for protein.
The alternative test used for detecting protein other than albumin involves denaturing the protein with heat or sulfosalicylic acid to produce turbidity. A false positive result in this alternate test may be produced by:

- Buniodyl
- Chlorpromazine
- Promazine
- Carinamide
- Cephaloridine
- Cephalothin
- Radiographic agents such as iopanoic acid, iodopyracet, and iophenoxic acid
- Sulfamethoxazole
- Thymol
- Tolbutamide

**Glucose.** The dipstick technique for glucose may be falsely positive in the presence of bleach in the collecting vessel and vaginal powders containing glucose. Ascorbic acid may produce a false negative result by retarding color development.

The Clinitest method may be used to quantitate the urinary glucose; however, other reducing agents may produce a false positive result. These include:

- Sugars: galactose, lactose, levulose, maltose, or pentose
- Homogentisic acid
- Glucuronic acid
- Bleach

Drugs that produce a false positive Clinitest result include:

- Acetanalide
- p-aminosalicylic acid
- Antipyrine
- Cephaloridine
- Cephalithin
- Chloramphenicol
- Chlortetracycline
- Cinchophen
- Diatrizoate
- Isoniazid
- Levodopa
- Nalidixic acid
- Oxytetracycline
- Tetracycline

**Hemoglobin.** A false positive test may be produced by myoglobin or the presence of oxidizing agents such as ascorbic acid.

**Ketones.** Aspirin may cause ketonemia in children, but the presence of aspirin in a ketonuric urine may produce a false negative dipstick.

False positive tests for ketones may be seen in the presence of:

- Levodopa
- Paraldehyde in the presence of ethanol
- Phenformin
- Porphobilinigen
- Skatole
- Indole
- Large quantities of bilirubin
- p-aminosalicylic acid
- Antipyrene
- Apronalide
- Bromsulfophthalein
- Chlorpromazine
- Phenazopyridine
- Phenothiazines
- Sulfadiazine
- Sulfamethoxazole
- Sulfanilamide
- Sulfonamides

URINE OUTPUT

Urine Output Measurement in Premature Infants

Lest we forget, urine, like water, does evaporate. This is especially true in the setting of the neonatal intensive care unit. A recent study of the rate and degree of fluid evaporation from disposable diapers under radiant warmers or in infant isolettes showed that evaporation was a function of time and was inversely related to the volume of fluid added to the diaper. Considering the importance of accurate determination of urine output for assessment of hydration status, renal function and nutrient retention in the premature and term neonate, the moral of this tale is clear: adhesive urine bags and frequent diaper inspection are imperative in the care of the low birth weight infant.


URINARY TRACT

Delayed Urination in the Newborn

One of the kindest acts a neonate can perform for his pediatrician is to urinate early in life. Ninety-nine to 100% of all normal infants urinate at least once by 48 hours of age. Approximately 23% will void first in the delivery room, and the act may not be reported to the nursery.

Failure to urinate by the first 1 to 2 days of life may be due to obstruction of urine flow or to inability to form urine. Causes of obstruction include

- Imperforate prepuce
- Urethral strictures
- Urethral diverticulum
- Hypertrophy of the verumontanum
- Neurogenic bladder
- "Megacystic syndrome"
- Ureterocele
- Renal tumors
- Cystic kidneys

Inability to form urine may result from

- Postnatal intravascular hypovolemia
- Restriction of oral fluids
- Bilateral renal agenesis
- Cortical necrosis
- Tubular necrosis
- Bilateral renal vein thrombosis
- Congenital nephrotic syndrome
- Congenital pyelonephritis
- Congenital nephritis

Nonspecific symptoms or signs such as excessive crying, irritability, poor feeding, pallor, emesis, mottled skin, or weak pulse may suggest the development of uremia.
The physical examination may be more useful in establishing a specific diagnosis.

**Physical Examination Following Delayed Urination**

<table>
<thead>
<tr>
<th>PALPATION OR PERCUSSION OF DISTENDED BLADDER</th>
<th>NO KIDNEYS PALPABLE</th>
<th>PALPABLE RENAL MASS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Obstruction of urine flow</td>
<td>Bilateral renal agenesis</td>
<td>Renal vein thrombosis</td>
</tr>
<tr>
<td>Examine meatus for patency and look for epispadias or hypospadias</td>
<td>These infants are usually males and tend to have lowset ears, epicanthal folds, and a flattened nose</td>
<td>Infantile polycystic kidneys</td>
</tr>
<tr>
<td>A urethral diverticulum may give rise to a bulge along the dorsum of the penis</td>
<td></td>
<td>Hydronephrosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Cystic dysplasia</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neoplasm</td>
</tr>
</tbody>
</table>


**Urinary Tract**

It is tempting to omit a urine culture in young infants being evaluated for a source of fever, although it is well recognized that urinary tract infections in such infants are associated with nonspecific signs and symptoms. In their evaluation of 100 infants 5 days to 8 months of age, all of whom had been discharged from the nursery in good health, Ginsburg and McCracken found the following characteristics:

- 75% of all infants with UTIs were less than 90 days of age.
- 75% of those infants with UTIs who were younger than 3 months were male.
- 95% of the male infants with UTI were uncircumcised.
- Fever, irritability, vomiting, and diarrhea were the only symptoms in over 90% of infants.
- Although bacteria could be visualized in 81% of stained urine samples, over 50% of the urine samples had less than 10 WBCs per high power field when examined microscopically.
- 25% of the infants had positive blood cultures, but all but one of the bacteremic infants were less than 3 months of age.
- 45% of girls and only 7% of boys with UTI were found to have radiologically detected abnormalities.

Every night and every morn
Some to misery are born.
Every morn and every night
Some are born to sweet delight.
Some are born to sweet delight,
Some are born to endless night.

William Blake
From *Auguries of Innocence*
VAGINAL BLEEDING

Abnormal Vaginal Bleeding in Adolescents

Pediatricians caring for adolescent females frequently see patients with the presenting complaint of vaginal bleeding. Skill in defining the cause of this source of bleeding is vital in order to differentiate benign processes from those that are potentially deleterious. By convention, abnormal vaginal bleeding is excessive in duration and quantity, occurs more frequently than once every 20 days, or is associated with anemia. While the overwhelming majority of such abnormal vaginal bleeding during the teenage years is caused by dysfunctional uterine bleeding, a condition most likely secondary to an immature hypothalamic-pituitary-gonadal axis, careful analysis and evaluation are indicated. Listed below are some entities that should be considered before applying the label of dysfunctional uterine bleeding to such a patient.

1. Vagina
   a. Foreign bodies (usually heralded by a foul-smelling, bloody discharge)
   b. Lacerations
   c. Adolescents whose mothers were prescribed diethylstilbestrol (DES) during their pregnancy in order to suppress spontaneous abortions

2. Cervix (over 1 million teenagers a year become pregnant)
   a. Spontaneous abortion
   b. Incomplete abortion
   c. Threatened abortion
   d. Ectopic pregnancy
   e. Molar pregnancy
   f. Submucosal myomas

3. Ovaries
   a. Functional ovarian cysts (follicular or corpus luteal)
   b. Tumors
   c. Polycystic ovary disease

4. Hypothalamic-pituitary (e.g., prolactinomas)

5. Adrenals
   a. Addison's disease (adrenal insufficiency)
   b. Congenital adrenal hyperplasia

6. Thyroid
   a. Hypothyroidism
   b. Hyperthyroidism

7. Sexually transmitted diseases
   a. Vaginitis (e.g., Trichomonas vaginalis infection)
   b. Cervicitis (e.g., Chlamydia trachomatis or Neisseria gonorrhoeae infections)
   c. Uterus and salpinx (e.g., pelvic inflammatory disease)

8. Endometriosis
9. Medications
   a. Complications of contraceptives
      i. Oral contraceptive pill
      ii. Intrauterine devices
   b. Anticoagulants
   c. Gonadal and adrenal steroids

   a. Chronic illness
   b. Emotional stress, eating disorders, crash diets, obesity, exercise (all of the
      aforementioned are more commonly associated with amenorrhea than
      excessive bleeding).

11. Bleeding disorders
   a. Hereditary or acquired thrombocytopenia
   b. Hereditary or acquired platelet function defect
   c. Von Willebrand’s disease
   d. Factor XIII or IX deficiency

12. Dysfunctional uterine bleeding (which is defined as abnormal vaginal
    bleeding that occurs in the absence of pregnancy, infection, neoplasms, or
    any other pathologic entity or disease).

References: Anderson MM, Irwin CE, Snyder DI: Abnormal vaginal bleeding in
Cowan BD, Morrison JC: Management of abnormal genital bleeding in girls and

VAS DEFERENS

Unilateral Absence of the Vas Deferens: A Useful Clinical Sign

The vas deferens of an adolescent male is easily palpated during a routine
physical examination. The unilateral absence of the vas deferens, on the other
hand, is associated with a 79% likelihood of finding a missing ipsilateral func-
tioning renal unit. Such an association makes the absence of the vas deferens a
significant anomaly, and examination for the presence of the ipsilateral renal unit
(e.g., intravenous pyelogram) is mandatory.

Reference: Donohue RE, Fauver HE: Unilateral absence of the vas deferens. A useful

VESICLES

Vesicular and Vesiculopustular Eruptions in the Newborn

There exists a large differential diagnosis for the newborn infant presenting
with vesicular or vesiculopustular lesions. The first distinguishing factor the
physician must ascertain is whether the lesions are caused by an infectious or
noninfectious etiology. This can typically be done rather quickly and inexpensively with the following diagnostic tests:

1. Gram stain
2. Potassium hydroxide (KOH) preparation
3. Tzanck smear
4. Bacterial culture
5. VDRL of the mother and infant
6. Viral culture
7. Fungal culture
8. If necessary, a skin biopsy

Given the potential for sepsis, one should assume an infectious etiology in the newborn presenting with vesicular or vesiculopustular lesions and treat appropriately before the culture results are known. A differential diagnosis is presented below, separated in terms of infectious and noninfectious etiologies.

1. **Infectious vesicular and vesiculopustular lesions**
   a. Herpes simplex (vesicles are on an erythematous base and are usually 1 to 3 mm in diameter, arranged either in clusters or singly; bullae, macular exanthems, purpura, and zosteriform eruptions have also been reported).
   b. Congenital varicella
   c. Varicella-zoster (grouped vesicles on an erythematous base arranged in a dermatomal or segmental pattern).
   d. Congenital cutaneous candidiasis (lesions are typically erythematous macules that progress over the course of 1 to 3 days through papular, vesicular, and pustular stages, followed by superficial desquamation. Yellow vesicles have also been reported).
   e. *Staphylococcal aureus* infection can cause bullae, erosions, or diffuse superficial desquamation (i.e., the staphylococcal scalded skin syndrome).
   f. Congenital syphilis can yield vesicles and bullae (typically on the palms and soles but any location is possible).

2. **Noninfectious vesicular and vesiculopustular lesions**
   a. Erythema toxicum neonatorum (erythematous macules with a central vesicle or pustule, primarily on the trunk). A Wright's stain of the vesicle or pustular contents revealing sheets of eosinophils is diagnostic.
   b. Transient neonatal pustular melanosis (vesicopustules, collarettes of scale, and hyperpigmented macules may be noted on the neck and trunk. The vesicles and pustules resolve 48 hours after birth, leaving only scaly lesions or hyperpigmented macules.
   c. Heat rash or miliaria (usually a papular, vesicular or pustular rash). Frequently seen in the neonate who has been excessively warmed.
   d. Letterer-Siwe disease (infantile form of histiocytosis X). Infants characteristically present with scalp eruptions similar to that of seborrheic dermatitis. Purpura, ulcers, vesicles, and pustules have also been reported. Biopsy is usually diagnostic. Multiorgan involvement is common.
   e. Congenital self-healing reticulohistiocytosis (possibly a variant of Letterer-Siwe disease). Infants may present with vesicles but more typically display erythematous or blue papules and nodules.
   f. Urticaria pigmentosa (lesions are classically tan or reddish-brown papules, macules or nodules that appear urticaric with rubbing. [Darier's sign]). The skin biopsy reveals a perivascular and epidermal mast cell proliferation. These lesions can yield vesicles and bullae during the newborn period.
   g. Bullous mastocytosis (lesions are classically large bullae or erosions; vesicles are seen rarely).
Vertigo and Syncope

h. Epidermolysis bullosa (a heterogeneous group of inherited skin disorders yielding easy blistering, with bullae and erosions, of the skin). Gentle rubbing, or Nicolsky's sign, can produce such lesions.
i. Dermatitis herpetiformis (vesicles and hemorrhagic crusts, particularly on the extremities).
j. Pemphigus vulgaris (blisters)
k. Herpes gestationis (blisters)
l. Incontinentia pigmenti (also known as Bloch-Sulzberger syndrome, melanoblastosis cutis linearis sive systematisata, melanosis corii degenerativa, and Absoe-Hansen's disease). It typically presents with erythema or vesicles or both at birth and often looks like erythema toxicum neonatorum. Seen almost exclusively in females, it is believed to be x-linked dominant and lethal in males. The lesions may be scattered but are more frequently linear in pattern. Neurologic, ocular, and dental anomalies are frequently present.
m. Incontinentia pigmenti achromians or hypomelanosis of Ito.


VERTIGO AND SYNCOPE

Vertigo (dizziness) and syncope (lightheadedness, fainting) may be difficult symptoms for a child to distinguish between with certainty. Many entities that are traditionally thought to cause syncope may also cause vertigo. Syncope will therefore be discussed as a subheading of causes of vertigo.

Common Causes

Benign paroxysmal vertigo
Drugs
- Alcohol
- Anticonvulsants
- Antihypertensives
- Aspirin
- Dilantin
- Gentamicin
- Narcotics
- Sedatives
- Streptomycin
Ear disease
- External canal impaction
  - Cerumen
  - Foreign body
- Inner ear disease
  - Cholesteotoma (with extension)
  - Fistula
  - Mastoiditis (with extension)
  - Suppurative labyrinthitis

Ear disease (Cont.)
- Inner ear disease (Cont.)
  - Vestibular neuronitis
  - Viral (acute) labyrinthitis
- Middle ear disease
  - Chronic suppurative otitis (with extension)
  - Hemotympanum (basilar skull fracture)
  - Otitis media (rare as isolated finding)
  - Serous otitis media
  - Tympanic membrane perforation
Headache
- Basilar artery migraine complex
- Migraine
- Hyperventilation syndrome
Seizure
- Aura/recovery phase
- Reflex seizure
- Visual impairment
Uncommon Causes

Central nervous system infection
Abscess
Encephalitis
Meningitis
Hypotension

Trauma
Basilar skull fracture
Cerebellar lesion/hemorrhage
Labyrinthine trauma
Postconcussion syndrome

Rare Causes

Adrenal insufficiency
Anemia
Arnold-Chiari malformation
Benign positional vertigo
Brain stem ischemia
Breath-holding spells
Central nervous system tumors
Acoustic neuroma
Brain stem glioma
Cerebellar glioma
Ependymoma
Medulloblastoma
Demyelinating disease
Multiple sclerosis
Endocrine disorders
Adrenal insufficiency
Diabetes mellitus
Thyrotoxicosis
Hypertension
Hypoglycemia
Increased intracranial pressure
Ménière’s syndrome
Pellagra
Psychosomatic illness
Ramsay Hunt syndrome

Syncope (many causes previously discussed)
Cardiovascular etiologies
Arrhythmia
Atrioventricular block
Cardioauditory syndrome
Emery-Dreifuss muscular dystrophy
Mitral valve prolapse
Paroxysmal atrial tachycardia
Paroxysmal ventricular tachycardia
Prolonged QT syndrome
Sick-sinus syndrome
Cardiac anomalies
Aortic stenosis
Pulmonary stenosis
Tetralogy of Fallot
Transposition
Truncus arteriosus
Carotid sinus syncope
Dysautonomia (Riley-Day syndrome)
Idiopathic hypertrophic subaortic stenosis
Left atrial myxoma
Myocardial infarction
Orthostatic hypotension
Pulmonary hypertension
Vasovagal stimulation
Vestibulocerebellar ataxia

VISION

The Visual Acuity of Normal Children

A friend of yours tells you that her 1 year old child has been examined by her pediatrician and is said to have "perfect 20/20" vision. Your child, who is also 1, has 20/200 vision. Who should get a new physician?

Visual acuity at birth is poorer than at any other time of life and only gradually improves to the 20/20 range at the time of entrance to kindergarten. The accompanying table indicates the expected average acuity of preschool children. An acuity of 5/200 should not be misinterpreted to mean that the
newborn is practically blind. Just stick your tongue out at a newborn and see what he does back to you!

### Visual Acuity

<table>
<thead>
<tr>
<th>AGE</th>
<th>AVERAGE UNCORRECTED ACUITY</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth</td>
<td>5/200</td>
</tr>
<tr>
<td>1 year</td>
<td>20/200</td>
</tr>
<tr>
<td>2 years</td>
<td>20/40</td>
</tr>
<tr>
<td>3 yrs</td>
<td>20/30</td>
</tr>
<tr>
<td>4 yrs</td>
<td>20/25</td>
</tr>
<tr>
<td>5 yrs</td>
<td>20/20</td>
</tr>
</tbody>
</table>


### VULNERABLE CHILD SYNDROME

Parents of a child who was expected to die, or parents of an only child, or parents who have experienced the death of a child often react in a manner that produces a disturbance in the psychosexual development of their offspring. Learn to recognize the circumstances that produce “the vulnerable child” syndrome and its manifestations. The psychosexual disturbance manifests itself most commonly in the following ways:

1. **Difficulty with separation.** Child may be briefly entrusted to the care of grandparents, but babysitters are rarely used. In extreme instances, mother and child never separate. Sleep problems are common. The child frequently sleeps with parents or in parents' room. Mother or father wakes frequently during the night to check on the status of the child.

2. **Infantilization.** Parents are unable to set disciplinary limits. Parent is overprotective, overindulgent, and oversolicitous. Child is overly dependent, disobedient, irritable, argumentative, and uncooperative. Children may be physically abusive to parents. Feeding problems are common.

3. **Bodily overconcerns.** Hypochondriacal complaints, recurrent abdominal pain, headaches, and infantile fears are prominent. School absence is common. Mothers express concern about minor respiratory infections, stool habits, “poor color,” circles under the eyes, and blueness when crying.

4. **School underachievement.** Unspoken agreement that the child is only safe with mother may produce separation anxiety that results in poor school performance.

### Predisposing Factors in the Production of the Vulnerable Child

1. Child is first-born to older parents who had resigned themselves to being childless.

2. Parents cannot have additional children as a result of a hysterectomy or other sterilization procedure.
3. The patient was born with congenital anomaly.
4. The patient was born prematurely.
5. The patient has an acquired handicap, e.g., epilepsy.
6. The child has had a truly life-threatening illness, such as erythroblastosis, nephrosis, or severe asthma.
7. During pregnancy the mother was told that the fetus might die.
8. Mother had a postpartum depression.
9. Mother has ambivalent feelings about child, such as instances where child was born out of wedlock.
10. Parents have unresolved grief reaction as a result of loss of another child.
11. A hereditary disorder is present in the family, such as cystic fibrosis or muscular dystrophy.
12. There is a psychological need on the part of the parents to find something physically wrong with the child in order to displace unacceptable feelings about the patient. Child is frequently brought to physicians because of parents' suspicion of leukemia, brain tumor, rheumatic fever, or other serious illness.
13. Separation of infant from his or her mother for phototherapy.

Treatment

1. Recognize the circumstances that may produce a vulnerable child and try to reassure parents about the health of the infant or child before symptoms appear.
2. Make authoritative statements about the child's well-being based on a thoughtful, cumulative history, physical examination, and pertinent measurements and laboratory findings.
3. Point out to the parents and get them to accept the reasons for their unnecessary concern, the child's responsive behavior, and the mutual reinforcement that is present.

Do not produce the syndrome yourself with comments such as "I thought for sure he was going to die," or "If she hadn't gotten here when she did we wouldn't have been able to save her," or "You are very lucky parents that we saved your child."

WALKERS

The AMA has summarized the epidemiology of walker injuries as follows:

1. Between 70% and 80% of infants will use a walker, mostly between ages 5 and 12 months; twice as many boys use walkers as girls.

2. Of infants who use walkers, 30% to 40% will have an accident.

3. Most walker accidents are minor and relatively few result in contact with a physician.

4. The most common types of accident involve falling down stairs, tipping over, and finger entrapment. Other injuries result from infants pulling objects down onto themselves.

5. Of infants seen in emergency departments for a walker injury, almost all serious trauma results from falling down stairs. Over 90% of all stairwell injuries among infants less than 12 months of age are related to use of walkers. Closed head injury is the most common serious walker injury, followed by fractures (skull, arm, clavicle) and other trauma, such as burns, dental injuries, and lacerations.

6. Of infants with serious injury, about one third stop walker use immediately, one third stop use within 2 months (usually because infants begin walking on their own), and one third are still using a walker 2 months after the injury.

7. Most walker injuries occur in the home with one or both parents present. Of injuries involving stairs, about half occur in houses with stairwell gates.

8. Although the occurrence of trauma is unrelated to the age at first use, a number of siblings, and parents' occupations, it is related to the amount of time spent in the walker. Fewer than 30% who spend less than 2 hours a day in a walker suffer a nonserious fall, compared with approximately 55% of infants who spend more than 2 hours per day in a walker.

9. The types of walkers involved in serious injury are fairly evenly divided between the X-frame, in which the steel support bars form an X, and the circular frame, in which the support bars go up in a straight vertical pattern to reach the upper tray.

Approximately one million walkers are sold in the U.S. each year. Although relatively rare, serious trauma does occasionally occur and physicians should counsel parents about the use of walkers, especially near stairwells.

There is no evidence that walkers promote bipedal ambulation.

WHEEZING

Common Causes

Aspiration
  Direct (e.g., defective swallow, neuromuscular disease)
  Indirect (gastroesophageal reflux, emesis)
Asthma
Atopic disease
Bronchiectasis
Bronchiolitis
Bronchitis
Foreign-body aspiration
Pneumonitis

Arterial Causes

Bronchopulmonary dysplasia
Congestive heart failure
Cystic fibrosis
Hypersensitivity pneumonitis
  Allergic bronchopulmonary aspergillosis
Mediastinal mass/adenopathy
Pulmonary edema
Tracheobronchomalacia

Rare Causes

α1-Antitrypsin deficiency
Angioneurotic edema
Carcinoid syndrome
Factitious wheezing
Lobar emphysema
Neoplasm/tumor
Psychogenic airway obstruction
Pulmonary hemosiderosis

WOUND CARE

In their zest to achieve antisepsis and ensure good wound healing, pediatricians and nurses may be creating the cosmetic surgery cases for tomorrow. Routine wound care in the acute setting usually consists of a 1:1 dilution of hydrogen peroxide and stock povidone iodine. Both of these solutions are extremely toxic to exposed fibroblasts and are, therefore, counterproductive agents as often used. Recommendations regarding the use of these antiseptics include:

- A 1:100 dilution of hydrogen peroxide (greater strength may be used on already mature granulation tissue).
- A 1:1000 (1 ml/L) dilution of povidone iodine. This is problematic, because although fibroblast toxicity is minimized, so is bactericidal effect. The answer is to paint the perimeter of the wound but not the wound itself.
- Avoid antibiotic solutions in wounds in the acute stages due to recognized cytotoxicity.

Put Sugar, Not Salt, in Their Wounds

It may seem counterintuitive, but granulated sugar has proven to be one of the safest, least expensive, and most “universal” antimicrobial treatments for infected wounds and superficial lesions. The success of sucrose solution as an antimicrobial depends on its water activity (aw), which defines the water requirements for growth of a given microorganism.

Studies demonstrated in The Lancet showed that 195 g of sugar in 100 g of water (aw = 0.858) completely inhibited growth of Staphylococcus aureus. S. aureus (aw = 0.86) happens to have the lowest aw of common bacterial pathogens, including streptococci, Klebsiella, E. coli, Corynebacterium, C. perfringens and other clostridia, and Pseudomonas.

The limiting factor in this extraordinarily available and effective method of wound care is that application of solution to a wound causes an osmotic change in surrounding tissue. The osmotic pressure change results in dilution of the sucrose solution, necessitating the addition of more granulated sugar. This seems a small inconvenience given the ubiquity of sucrose. When preparing your next First Aid Kit, do not forget the sugar!

It is doubtful that any resident or physician can imagine a single work day in the hospital without ordering an x-ray of some kind. One of the most frequent concerns parents have when consenting to a radiologic procedure has to do with the amount of radiation exposure a particular x-ray yields. Listed below are the range of radiation doses generated in a variety of medical procedures and "nonmedical" activities:

### Range of Radiation Doses Received in Various Medical and Nonmedical Activities

<table>
<thead>
<tr>
<th>TYPE OF RADIATION</th>
<th>DOSE (RAD, REM, VERY APPROXIMATE)*</th>
<th>LENGTH OF EXPOSURE</th>
<th>WHERE RECEIVED</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chest film, newborn</td>
<td>0.004</td>
<td>Msec</td>
<td>Skin entrance dose; exit dose lower</td>
</tr>
<tr>
<td>CT, contiguous slices, child</td>
<td>2.5</td>
<td>Sec</td>
<td>Scanned volume</td>
</tr>
<tr>
<td>Lateral of lumbosacral spine, adult</td>
<td>0.5</td>
<td>Sec</td>
<td>Skin entrance dose; exit dose much lower</td>
</tr>
<tr>
<td>Cardiac catheterization</td>
<td>10-100</td>
<td>Hr</td>
<td>Skin entrance dose; exit dose much lower</td>
</tr>
<tr>
<td>Curative radiotherapy</td>
<td>7,000</td>
<td>Wk</td>
<td>Tumor and adjacent structures</td>
</tr>
<tr>
<td>Natural background at sea level</td>
<td>0.08</td>
<td>Yr</td>
<td>Whole body</td>
</tr>
<tr>
<td>Some professional jet pilots and flight crews, from cosmic rays</td>
<td>1</td>
<td>Yr</td>
<td>Whole body</td>
</tr>
<tr>
<td>Residents of certain areas of India with radioactive soil</td>
<td>3</td>
<td>Yr</td>
<td>Whole body</td>
</tr>
<tr>
<td>Radiation workers, current permitted dose</td>
<td>5</td>
<td>Permitted/yr</td>
<td>Radiation badge</td>
</tr>
<tr>
<td>Dose at which half of population dies, nuclear warfare</td>
<td>450</td>
<td>Min</td>
<td>Whole body</td>
</tr>
</tbody>
</table>

*To convert to grays, divide by 100.

Disorders of nail pigmentation have been associated with numerous conditions. The term *chromonychia* refers to an abnormality in color of the substance or surface of the nail plate and/or subungual tissues. Practically the entire color spectrum is represented in abnormalities of the nails, but this section will focus only on yellow nails. White nails (*leukonychia*) are the most common variant.

The nail should be studied with the fingers relaxed and not pressed against a surface, which can alter the hemodynamics and give a false appearance. The fingertip should then be blanched to try to differentiate between discoloration of the nail plate and the vascular bed. If a tropical agent is suspected as the cause, try removing it by scraping or by rubbing with a solvent.

The causes of staining include contact with exogenous agents, cosmetics, other topical applications, tobacco, trauma, physical agents, and fungal and bacterial infections.

Nails can provide an extended historical record of abnormalities of skin pigment that might otherwise go unnoticed.

### Causes of Yellow Nails

<table>
<thead>
<tr>
<th>TOPICAL/CONTACT</th>
<th>EXTERNAL AGENTS (CONT.)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Cosmetics</strong></td>
<td></td>
</tr>
<tr>
<td>Chloroxine (+ aluminum)</td>
<td>Diquat</td>
</tr>
<tr>
<td>Formaldehyde in hardeners</td>
<td>Epoxy systems</td>
</tr>
<tr>
<td>Formaldehyde-phenol resins</td>
<td>Hydrofluoric acid</td>
</tr>
<tr>
<td>Hair dyes</td>
<td>Nitric acid and derivatives</td>
</tr>
<tr>
<td>Nail lacquers</td>
<td>Picric acid</td>
</tr>
<tr>
<td>Resorcinol in nail varnish</td>
<td>Tetryl</td>
</tr>
<tr>
<td>Tobacco</td>
<td></td>
</tr>
<tr>
<td><strong>Dermatoses</strong></td>
<td>Weed and insect poisons</td>
</tr>
<tr>
<td>Fogo selvagem (wildfire pemphigus)</td>
<td>Yellow dyes, paints, polishes, and stains</td>
</tr>
<tr>
<td>Psoriasis</td>
<td>Therapeutic agents</td>
</tr>
<tr>
<td>Onychomycosis</td>
<td>Amphotericin B</td>
</tr>
<tr>
<td>Yeast infection (dermatophytes and nondermatophytes)</td>
<td>Dinitrochlorobenzene</td>
</tr>
<tr>
<td></td>
<td>Fluorescein</td>
</tr>
<tr>
<td><strong>External agents</strong></td>
<td></td>
</tr>
<tr>
<td>Chromium salts (yellow ochre color)</td>
<td>Trauma</td>
</tr>
<tr>
<td>Coal tar derivatives</td>
<td>Caustic soda</td>
</tr>
<tr>
<td>Dichromates</td>
<td>Hematoma in resolution</td>
</tr>
<tr>
<td>Dinitrophenol</td>
<td>Thermal injury</td>
</tr>
</tbody>
</table>

*Table continued on next page.*
## Causes of Yellow Nails (Cont.)

<table>
<thead>
<tr>
<th>HEREDITARY/SYSTEMIC</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Aplasia cutis with dystrophic nails</td>
<td></td>
</tr>
<tr>
<td>Beta carotene</td>
<td></td>
</tr>
<tr>
<td>Chronic pulmonary disease</td>
<td></td>
</tr>
<tr>
<td>Diabetes mellitus</td>
<td></td>
</tr>
<tr>
<td>D-penicillamine</td>
<td></td>
</tr>
<tr>
<td>Familial amyloidosis with poly-neuropathy</td>
<td></td>
</tr>
<tr>
<td>Hyperbilirubinemia</td>
<td></td>
</tr>
<tr>
<td>Incontinentia pigmenti (slightly yellow)</td>
<td></td>
</tr>
<tr>
<td>Macular amyloidosis with familial nail dystrophy</td>
<td></td>
</tr>
<tr>
<td>Pachyonychia congenita</td>
<td></td>
</tr>
<tr>
<td>Peripheral vascular disease</td>
<td></td>
</tr>
<tr>
<td>Porphyria cutanea tarda</td>
<td></td>
</tr>
<tr>
<td>Progeria</td>
<td></td>
</tr>
<tr>
<td>Rifampin ingestion</td>
<td></td>
</tr>
<tr>
<td>Tetracycline ingestion</td>
<td></td>
</tr>
<tr>
<td>Yellow nail syndrome and lymphedema</td>
<td></td>
</tr>
</tbody>
</table>

Z

ZOO NOSES

Diseases You Can Acquire from Pets and Animals

Everyone knows that dogs are "man’s best friend"—that is, unless you own a cat. Indeed, pets are a valuable addition to the family and can enrich a child’s life with affection, companionship, and a sense of responsibility. But as wonderful as pets are, they present a potential risk to the health of humans, particularly children. Listed below are the more common zoonoses and the animals with which they are associated:

Potential Host Distribution of Selected Zoonoses

<table>
<thead>
<tr>
<th>Viral Diseases</th>
<th>Domestic Animals</th>
<th>Wild Animals</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Horses</td>
<td>Cattle</td>
</tr>
<tr>
<td>Arbovirus encephalitis</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Cat-scratch disease (virus suspected)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lymphocytic choriomeningitis</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Newcastle</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Rabies</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Vesicular stomatitis</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Yellow Fever</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Rickettsial Diseases</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Q fever</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Rocky Mountain spotted fever</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Spirochetal Diseases</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Leptospirosis</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Rat-bite fever</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bacterial Disease</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anthrax</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Brucellosis</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>Erysipelas</td>
<td>X</td>
<td>X</td>
</tr>
</tbody>
</table>

Table continued on next page.
### Potential Host Distribution of Selected Zoonoses (Cont.)

<table>
<thead>
<tr>
<th>Bacterial Disease (Cont.)</th>
<th>Domestic Animals</th>
<th>Wild Animals</th>
<th>Mammals</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Horses</td>
<td>Cattle</td>
<td>Sheep</td>
</tr>
<tr>
<td>Hemorrhagic septicemia</td>
<td>X X X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Listeriosis</td>
<td>X X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Melioidosis</td>
<td>X X X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Plague</td>
<td>X X X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Pseudotuberculosis</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Psittacosis</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Salmonellosis</td>
<td>X X X X X X X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Scarlet fever</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Septic sore throat</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Staphylococcosis</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Tetanus</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Tuberculosis</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Tularaemia</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
<tr>
<td>Vibriosis</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
<td>X X X X X X</td>
</tr>
</tbody>
</table>

### Fungal Diseases

| Actinomycosis             | X X X X X X X X X X X X | X X X X X X |
| Aspergilosis              | X X X X X X | X X X X X X | X X X X X X |
| Coccidioidomycosis        | X X X X X X | X X X X X X | X X X X X X |
| Cryptococcosis            | X X X X X X | X X X X X X | X X X X X X |
| Epizootic lymphangitis    | X X X X X X | X X X X X X | X X X X X X |
| Histoplasmosis            | X X X X X X | X X X X X X | X X X X X X |
| Nocardiosis               | X X X X X X | X X X X X X | X X X X X X |
| North American blastomycosis | X X X X X X | X X X X X X | X X X X X X |
| Rhinosporidiosis          | X X X X X X | X X X X X X | X X X X X X |
| Ringworm                  | X X X X X X | X X X X X X | X X X X X X |
| Sporotrichosis            | X X X X X X | X X X X X X | X X X X X X |
| Streptothricosis          | X X X X X X | X X X X X X | X X X X X X |

### Protozoan

| Amebiasis                 | X X X X X X | X X X X X X | X X X X X X |
| Balantidiasis             | X X X X X X | X X X X X X | X X X X X X |
| Leishmaniasis             | X X X X X X | X X X X X X | X X X X X X |
| Plasmodium (malaria)      | X X X X X X | X X X X X X | X X X X X X |
| Sarcocystis               | X X X X X X | X X X X X X | X X X X X X |
| Toxoplasmosis             | X X X X X X | X X X X X X | X X X X X X |
| Trypanosomiasis           | X X X X X X | X X X X X X | X X X X X X |

From Fowler ME: Curr Probl Pediatr 4:3, 1974, with permission.

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