This document presents the agenda and materials distributed at a 1-day introductory workshop on deaf-blindness. Introductory material explains the workshop's purpose and rules. A short test contrasts facts and myths about deaf-blindness. A handout presents information on the dynamics of deaf-blindness, etiologies in the adult deaf-blind population, and categories of deaf-blindness. Information on the anatomy and function of the eye and visual system is provided. Visual acuities and visual fields are explained, followed by information on five types of vision loss, an explanation of how a child's vision is affected by a specific eye disorder, and possible developmental delays of a child with severe vision impairment. An activity in which participants attempt to identify objects by touch alone is suggested. The workshop then addresses how hearing works, the importance of sound, speech perception, otological care, anatomy of the ear, causes and types of hearing losses, measuring hearing loss, and degrees of hearing loss. A hearing loss simulation activity is also provided. Remaining informational handouts cover communication modes, language levels, and a functional approach to language intervention and assessment. A workshop summary notes that deaf-blind individuals may have problems in the areas of anticipation, motivation, communication, and learning. (DB)
Introduction to Deaf-Blindness Workshop

18 December 1995
Central Missouri Deaf-Blind Task Force

Presented by:
Larry Rhodes - Missouri 301.77 Project

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Agenda
Introduction to Deaf-Blindness Workshop

18 December 1995
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Agenda

9:00 am - 9:10 am Introductions
9:10 am - 9:15 am Purpose of the workshop
9:15 am - 9:30 am Dynamics of Deaf-Blindness and Etiologies in the Deaf-Blind Population
9:30 am - 9:45 am The Eye and How It Works (The Eyeball Made Easy)
9:45 am - 10:00 am What are Visual Acuities and Visual Fields?
10:00 am - 10:15 am The Five Types of Vision Loss
10:15 am - 10:30 am Break
10:30 am - 11:00 am The Five Types of Vision Loss (Con't)
11:00 am - 11:30 am Activity: "Whatzit?"
11:30 am - 11:45 am Possible Developmental Delays of a Child with a Severe Vision Impairment
11:45 am - 12:00 am Set-Up for Activity: Lunch
12:00 am - 12:30 pm Activity: Lunch
12:30 pm - 12:45 pm  Process Lunch Activity
12:45 pm - 1:00 pm  Break
1:00 pm - 1:15 pm  How We Hear
1:15 pm - 1:30 pm  Types and Degree of Hearing Loss
1:30 pm - 2:00 pm  Activity: Hearing Loss Simulations
2:00 pm - 2:15 pm  How Hearing Loss Affects Communication and Concept Development
2:15 pm - 2:30 pm  Break
2:30 pm - 3:00 pm  Communication Modes
3:00 pm - 3:45 pm  Activity: Non-Verbal Deaf-Blind Communication
3:45 pm - 4:00 pm  Tying It All Together
4:00 pm - 4:15 pm  Wrap-up and Evaluation
Purpose and Rules
Purpose of the Workshop

To provide:

- a basic awareness of vision and hearing losses and how these losses affect an individual's development and functioning in her/his environment;

- a basic awareness of how the combination of deafness and blindness creates a whole new set of dynamics affecting how the Deaf-Blind consumer relates to other individuals and to the environment;

- an opportunity to experience the dynamics of deaf-blindness; and,

- a foundation for future, expanded training in specific topic areas relating to deaf-blindness: Communication, Social Skills, Orientation and Mobility, Independent Living Skills, Recreation, Job Development and Placement, etc...

Rules for the Workshop

1. Participant must plan to stay the entire day.

2. The presenter will *not* use "People First Language" when referring to Deaf-Blindness. The presenter will adhere to the preference of the adult Deaf, Blind, and Deaf-Blind Communities and refer to these individuals as Deaf Individuals/Consumers, Blind Individuals/Consumers, and/or Deaf-Blind Individuals/Consumers.

3. The presenter will start and stop on time.

4. There are no "bad" questions.

5. Relax and enjoy yourself during this workshop.
FACTS AND MYTHS ABOUT DEAF-BLINDNESS

(A short test)
FACTS AND MYTHS ABOUT DEAF-BLINDNESS

Many people hold a number of misconceptions and misunderstandings about the dual sensory impairments described by the label of deaf-blindness. Read each of the following statements. Which ones do you think are facts? Which ones do you think are myths? Circle the one best response for each statement.

1. People who have been assigned the label deaf-blindness are both totally deaf AND totally blind
   FACT  MYTH

2. As a result of the rubella vaccine developed in 1969, children are seldom labelled as deaf-blind and professionals need not worry about developing the skills to address this low-incidence disability group.
   FACT  MYTH

3. Usher Syndrome, the combination of congenital deafness and progressive blindness due to Retinitis Pigmentosa, is a leading cause of deaf-blindness among adolescence and young adults.
   FACT  MYTH

4. There are minimal differences in the characteristics and support needs of individuals born deaf-blind (congenitally deaf-blind) and those of individuals who become deaf-blind later in life (adventitiously deaf-blind).
   FACT  MYTH

5. Regardless of the age of onset or severity of the dual disabilities, communication and orientation and mobility are common support needs of individuals who have been labeled deaf-blind.
   FACT  MYTH

6. Typically, individuals who have been labeled deaf-blind are unable to work or live in integrated community settings.
   FACT  MYTH  (developed by HKNC-TAC)
Dynamics of Deaf-Blindness

and

Etiologies in the Deaf-Blind Population
Dynamics of Deaf-Blindness and Etiologies in the Adult Deaf-Blind Population

What is Deaf-Blindness?

"Deaf-Blind" refers to two sensory impairments, visual and hearing, occurring in combination with each other. The combination of these sensory losses causes significant education and rehabilitation problems.

For the purpose of this workshop, we will use the Federal definition of Deaf-Blindness used to identify deaf-blind children:

The Federal Definition of Deaf-Blindness

The term "Children with deaf-blindness," means children and youth having auditory and visual impairments, through combination of which creates such severe communication and other developmental and learning needs that they cannot be appropriately educated without special education and related services, beyond those that would be provided solely for children with hearing impairments, visual impairments, or severe disabilities, to address their educational needs due to these concurrent disabilities.

- The Federal Register, October 11, 1991, page 51585

Why do we want to use the Federal definition?

Because it is functional.

When we are working with a Deaf-Blind adult, we need to ask ourselves:

-Does this individual have enough vision to compensate for her/his lack of hearing?
- Does this individual have enough hearing to compensate for her/his lack of vision?

If the answer is "NO" to both of the above, you are working with an individual who is deaf-blind.

The term "Deaf-Blind" does not always mean totally blind and totally deaf. Most deaf-blind people actually have some residual hearing and/or vision.

What makes the combination of deafness and blindness so unique?

Vision and Hearing are our two distance senses; they bring the world to us. Loss of these two senses deprives us of the two primary senses through which we learn about the environment and the world at large.

Because of these two losses - vision and hearing - the person who is deaf-blind, particularly if deaf-blind since birth, may experience cognitive problems that may resemble those experienced by individuals with developmental disabilities. Be aware of an individual who has been deaf-blind since birth may resemble those of an individual with autism. This is because the deaf-blind individual's incoming information about the world is distorted.

On the next page is a chart that provides a simple way of looking at sensory integration and how a combined vision and hearing loss can create cognitive delays. The diagram is a modification one developed by Jean Ayers.
NORMAL

Sensory input → Brain organizes and prepares a response → Output provides feedback which becomes new input

good feedback reinforces good responses

DEAF-BLIND

Sensory input is not quite right → Brain does not organize it not quite right → Output is faulty; as time goes by this results in poor feedback which further creates problems

poor feedback reinforces poor responses

If, in addition, the person also has developmental disabilities or cerebral palsy, the poor feedback cycle is further reinforced.
Four Categories of Deaf-Blindness

1. Congenitally Deaf, Adventitiously Blind (born Deaf/Hearing Impaired and later lost vision)

Some causes of vision loss:
- Retinitis Pigmentosa (RP)/Ushers/Other Related
- Macular Degeneration
- Diabetic Retinopathy
- Cataracts
- Retinal Detachment

2. Congenitally Blind, Adventitiously Deaf (born Blind/Visually Impaired and later lost hearing)

Some causes of hearing loss:
- Unknown
- High Fevers
- Diabetes
- Kidney Failure
- Pollution/Toxins
- Trauma
- Genetic Factors
- Presbycusis

3. Congenitally Deaf-Blind (born Deaf-Blind)

Some Causes of Congenital Deaf-Blindness:
- Prenatal Dysfunction:
  - AIDS
  - Rubella
  - Toxoplasmosis
- Syndromes
  - Down's
  - Trisomy 13
- Multiple Congenital Anomalies
  - CHARGE Association
  - Fetal Alcohol Syndrome
  - Maternal Drug Abuse
Prematurity
Pre- and Post Natal Trauma
Meningitis
Encephalitis

4. Adventitiously Deaf-Blind (born with normal vision and hearing and later lost both)

Some causes of deaf-blindness:
Illness
Trauma
Genetic Factors
Age related vision and hearing losses
How Can I Begin to Understand the Dynamics of Deaf-Blindness?

To help you better understand the dynamics of deaf-blindness, let's first explore vision and hearing losses separately, examining how each alone affects an individual's development and relationship with the world.

Not only will you learn about the effects of vision and hearing loss - individually and together - "academically" through lecture, but also experientially through planned activities.

At the end of this workshop, we'll tie together all of your experiences by taking a closer look at deaf-blindness.
The Eye and How It Works
(The Eyeball Made Easy)
INSITE RESOURCES FOR FAMILY CENTERED INTERVENTION
FOR INFANTS, TODDLERS, AND PRESCHOOLERS
WHO ARE VISUALLY IMPAIRED

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TOPIC 1: ANATOMY OF THE VISUAL SYSTEM

GOAL

Parents and other caregivers will know the parts of the visual system and be able to explain what is wrong with the child’s visual system.

MATERIALS

- Diagram V-1, "The External Structures Surrounding the Eye"
- Diagram V-2, "The Human Eye"
- Diagram V-3, "The Natural Blind Spot"
- Diagram V-4, "Optic Nerve Pathway In the Brain"
- An eye model or colored eye anatomy chart (A model and chart can be purchased from the American Optometric Association, 243 North Lindberg Blvd, St. Louis, MO 63141. A simple eye anatomy illustration entitled "How We See" is available free from the National Society for the Prevention of Blindness, 79 Madison Ave, New York, NY 10016)
- A penlight
- Mirror

INSTRUCTIONS TO EARLY-childhood SPECIALIST

For many parents, it is advisable to shorten this discussion to cover only the key anatomical parts of the visual system. Another option is to share the information over two or more visits. Specific names of some ocular disorders will be listed in brackets as a note about what disorders relate to damage or abnormalities in the part of the visual system being discussed. When talking about the parts of the visual system which relate to the child’s ocular disorder, take time to cover what has been damaged in that part of the child’s visual system.

This means that the early childhood specialist will need to know what the child’s ocular disorder is ahead of time and should have done some reading on it or have discussed it with a vision consultant. Ask the parents (or caregivers what information they have obtained about the child’s ocular disorder from the child’s eye doctor(s). This, and the following topic, should help them make sense of the information if they do not, as of yet, understand it. Stop occasionally to see if they have questions or comments. A reference guide to ocular disorders in young children is found in Appendix C on page 638. Look up the specific ocular disorder in that guide. Remember that information on ocular disorders and treatment is constantly changing with medical advances, so consult the child’s eye doctor(s) for the most up-to-date information.

SAMPLE DISCUSSION

Today, we will look at how the human visual system is put together, some of the things that may be damaged, and what, specifically, has happened to your child’s visual system. We use the words "visual system" because vision and seeing involves more than just the eyes; they also involve special nerves called the optic nerves that connect the eye to the brain as well as a certain part of the brain called the visual cortex that actually does the "seeing". All of this together is called the visual system. When something happens to any of these parts, vision loss
can occur.

Let's start at the very beginning. The majority of the eye's development takes place during the first three months of pregnancy, the embryonic period. The eyeballs appear as two little buds of brain material extending from the rest of the brain and connected to it by what will later become the optic nerves. Therefore, the eyes are really an extension of the brain exposed to the outside world!

It is during this first three months of pregnancy that things can differ from what is expected. For example, viruses (e.g., rubella, cytomegalovirus or CMV), infections and certain drugs taken by the mother can alter how the eye is formed. If the genes that control the development of the eyes are abnormal, this is when malformations take place. Often, these are things and events over which the mother has no control and which the best eye doctor in the world could not have predicted or prevented. They just happen. [If the child's eye report indicates an etiology such as albinism, retinoblastoma, Down or CHARGE syndrome; or if the child's mother contracted a virus or other infection during her pregnancy that resulted in vision problems such a cataracts caused by rubella; or if the child has an ocular deformity such as anophthalmia or coloboma, take time to discuss it with the parents if they are comfortable in doing so.]

Slowly, the structures of the visual system--the eyeballs, the optic nerves, and the visual cortex--develop and become refined. A full term baby with no visual problems is born with an almost completely developed visual system. The newborn's visual acuity (his ability to see things sharply or clearly) is poor, but the baby's acuity approximates 20/20 by the age of 12 months due to maturation of the nervous system. Because a young baby sees objects much better when the objects are within one to two feet from his eyes, human faces attract a good deal of his visual attention. This attraction is reinforced because a baby is generally held cradled in someone's arms when he is being nursed or fed. This also helps in the bonding process between the infant and his parents. Bright, colorful objects and toys within arm's reach also attract his attention.

During the first few months of life, the baby's eyes are not always in alignment when he is looking at something or following it with his eyes. One eye may turn in or out. However, by the time the baby is 6 months old, both eyes work well together as a team.

Just as there are things that can cause damage to the visual system when it is developing during the first three months of pregnancy, a number of things can cause damage to the visual system during delivery and in the weeks and months after a baby is born. They can include a difficult birth (e.g., the baby does not get enough oxygen), premature birth, infection and disease, prolonged hydrocephalus and accidents resulting in head injuries. From birth until about 18 - 24 months, a baby's brain is still considered to be immature because the nerve pathways in the brain are still forming. This makes the baby more susceptible to brain injury. [If the child's eye report indicates an etiology that occurred during birth or the months thereafter, take time to discuss it with the parents.]

Let's take a closer look now at this visual system, starting with the eyeball and working our way back to the visual cortex. [To aid in describing the information contained in the next three paragraphs, point out the structures being discussed using Diagram V-1, "The External Structures Surrounding the Eye"].

The eyeball lies within an indentation of the skull called the orbit. The orbit is made up of bones in the skull. The upper bone also forms the eyebrow and the lower bones form the cheekbone. These bones, along with soft, cushiony tissue around the eye, help protect it from direct impact. The front of the eye is also protected by the eyelids and lashes. Glands found in the lids and above the eye (not indicated in the diagram), make the tears that bathe the front of the eye, lubricate it and help to prevent infections. Every time you blink, this happens.
Try to keep your eyes open, and do not blink. What do you feel? Burning or stinging? This is because the front of your eye, or cornea, is drying out.

Each eyeball has six muscles connected to it on the outside called extraocular muscles. These muscles hold the eyeballs in place as well as move the eyeballs. Two or more of these muscles, working together, move each eyeball up, down, diagonally and to the side. These eye movements are important for visually searching for things, looking at these things, and visually following these things as they move. The muscles are hooked up to nerves that are controlled by the front part of the brain, the frontal lobes. When everything is working right with these muscles, they allow both eyeballs to move together in the same direction. When both eyeballs are aligned and move together, we see only one image and have depth perception.

[If the child has an extraocular muscle imbalance such as strabismus, exotropia, esotropia, or crossed eyes, take time now to discuss it. Is the child’s problem due to damage to the frontal lobes, which control the coordinated movements of the extraocular muscles? Is one muscle shorter or longer than normal, upsetting the muscular balance? Is the imbalance the result of another disorder? For example, reduced vision in the right eye may cause it to wander and not move in conjunction with the left eye. Abnormal muscle tone in children with cerebral palsy can also affect the extraocular muscles, thereby affecting eye movements. If appropriate, discuss the treatment approach recommended by the doctor such as patching, eye drops, optical aids or surgery.]

[To aid in describing the information contained in the next part of this discussion, point out the structures being discussed using Diagram V-2, "The Human Eye."

There is a very thin clear membrane, with tiny, very thin blood vessels, that starts at the inside edge of the eye lids, runs along the inside surface of the eye lids, then fold back over and covers the outside front of the eye ball. This is called the conjunctiva. This helps protect the eye from infections and keeps anything from slipping behind the eyeball. It also makes fluids that lubricate the eye. If the conjunctiva becomes infected, it may become pink. This is what we call "pinkeye".

The human eye is oval in shape. This means it is shaped more like an egg and not a ping-pong ball. In an adult, the eye is about 1 inch across. A child’s eye is a little smaller.

The eyeball has three layers to it. Most of the outside layer of the eye is called the sclera. It is also known as the "whites of the eyes". The sclera is made up of fibers that criss-cross each other, making the sclera very tough. Since the sclera is tough, it makes a protective coat for the eye protecting everything else inside it. It also keeps light from entering the eye except at the front of the eye.

At the front of the eye the sclera becomes clear so that light can enter the eye. This clear area of the outermost layer of the eye is called the cornea. If you look at someone’s eyes from the side, you can see a clear bulge, or bubble, at the front of the eyes. This is cornea.

Because the surface of the cornea is curved, it helps to bend and focus the light entering the eye onto the retina at the back of the eye. This bending of light is called refraction and the cornea refracts 80% of the light being refracted on its way to the back of the eye. If the curvature of the cornea is too steep, not steep enough, or irregular, it does not allow the light to focus, forming a clear image on the retina at the back of the eye. We will discuss the retina a bit later. These may cause or help cause the "refractive errors" we call near sightedness, far sightedness, and astigmatism. In most cases, glasses and contact lenses are all that is needed to correct these errors.

The cornea is also the only part of the eye that can be transplanted, an operation that is less successful in young children.

[If the child has a refractive error or a problem with the cornea, discuss it. High refractive errors are associated with some syndromes such as Down syndrome. Disorders
related to the cornea are corneal dystrophy, keratoconus and herpes.

Behind the cornea is a space called the **anterior chamber**. It is filled with a liquid called the **aqueous humor**. We will discuss more about the anterior chamber and the aqueous humor a little later.

Behind the cornea and the anterior chamber is a round ring shaped part of the eye called the **iris**. This is the part of the eye we refer to when we say that a person has blue, green, hazel, brown - but not black! - eyes. The iris is part of the middle layer of the eyeball.

Just as our skin has pigmentation which gives it color, the middle layer of the eye, which includes the iris, also has pigmentation which gives the eye its color. The amount of pigmentation determines the color of the eye. An iris with a lot of pigment is brown. An iris with a small amount is blue. People with certain types of albinism have little or no pigmentation, that is why most people with albinism have very blue (and almost pink) irides. (Irides is the plural of iris.)

As we learned earlier, the iris is shaped like a ring with a hole in the middle. This hole is called the **pupil**. The pupil is actually an opening that lets light pass on through to the retina at the back of the eye.

The purpose of the iris is to control the amount of light entering the eye through the pupil. The iris does this by getting wider, making the pupil smaller, or by getting thinner, making the pupil larger. In dim light, the iris gets thinner, or dilates, making the pupil larger allowing more light to enter the eye. In bright light, the iris get wider, or constricts, making the pupil smaller, keeping too much light from entering the eye. Just as there are six extraocular muscles outside the eye that moves it, there are two muscles inside the iris that allows it to dilate or constrict. [Use your hand with fingers and thumb forming a circle to demonstrate constriction and dilation or have the parents watch pupils in a mirror as you shine a penlight in their eyes, then remove it. Remind them of what it is like when they walk from a dark movie theater into the sunny outdoors and vice versa.]

Disorders related to the iris are aniridia, coloboma and albinism. Also, the pupillary response in some neurologically impaired children may be slow or unequal because of damage to the brainstem which controls this involuntary reflex. These children have difficulty adapting to changes in lighting. For example, if the pupil is fixed large and open, the child may be very light sensitive. Or if the pupil is fixed small and nearly closed, the child may have trouble at night and in dark rooms. Some children may have "hippus" where the pupil dilates and constricts continually, meaning they have little control over the amount of light entering the eye. An abnormal pupillary response may also be caused by medication.]

The iris is connected to another part of the middle layer of the eye called the **ciliary body**. The ciliary body is made up mostly of muscle and blood vessels. The ciliary body is connected to the lens by thin strands of fiber called suspensory ligaments or the Zonules of Zinn. The muscles of the ciliary body pull on the suspensory ligaments, which in turn pull on the lens which allows the lens to change shape. We will discuss more about this a little later today as well as in the future during another lesson.

The ciliary body also has another function. It also produces a watery fluid, the aqueous humor. We will discuss what this fluid does in a little bit.

The rest of the middle layer of the eye is the **choroid**. It starts at the ciliary body, going all the way around the inside of the eye and back to the ciliary body again. Along with the iris and ciliary body, the choroid is part of the vascular, or blood carrying, layer of the eye. The choroid provides oxygen and nutrients to the retina, the inner most layer of the eye, keeping it alive.

Before we talk about the innermost, and last layer of the eye, we need to go back and talk about the eye's **lens**. The normal lens in a child has no color and is almost transparent.
It is made up of layers, like an onion, and is very flexible. It is encased in a clear "skin", that helps the lens keep its shape.

As we discussed above, the lens is connected to the ciliary body by the suspensory ligaments. As the muscles of the ciliary body change shape, they tug or let loose of the suspensory ligaments, which, in turn, changes the shape of the lens so the eye can focus on objects both near and far. Along with the cornea, the lens is also responsible for a lot of the refractive power of the eye. We discussed this term earlier today when we talked about the cornea. Unlike the cornea, however, the lens can change shape, fine focusing the light onto the retina depending on the distance of the object at which we are looking. If it needs to focus the image of an object far away onto the retina, the lens will become somewhat "fatter and shorter". If it needs to focus the image of an object close up onto the retina, the lens will become somewhat "thinner and longer". A young child's lens is much more flexible than an adult's. When a low vision child brings objects very close to his face, it is a natural way for him to magnify the images, making it easier for him to see them.

[If the child has cataracts or is aphakic, discuss this and go over the treatment recommended by the eye doctor.]

At the back of the eye is the innermost layer of the eye, the retina. It is a very thin tissue made up mostly of nerves and special cells, somewhat like nerve cells, called photoreceptor. There are two types of photoreceptor cells: rods and cones.

The cones are connected almost one-to-one to the nerves in the retina. The cones give us the ability to see fine detail and color. They work best in daylight or in high lighting. Although cones can be found throughout the retina, most are concentrated tightly in the middle back part of the retina called the macula or central retina. Since most of the cones are concentrated in the macula, we use the macula part of the retina to see fine detail and color. That is why the macula is in the middle back part of the eye. In the very middle of the macula is an area called the fovea centralis. The eye sees the sharpest detail here. [Disorders related to the macula area of the retina include Stargardt's Disease and other types of juvenile macular degeneration; albinism; macular hypoplasia and a few types of retinitis pigmentosa. Discuss these disorders if appropriate.]

The rods are connected 20 or more per nerve. They work best in minimum lighting. Although rods can be found throughout the retina, they are concentrated on the rest of the retina away from the macula, the peripheral retina. The rods provide us with the ability to see gross forms and movement. [Disorders related to the peripheral area of the retina include glaucoma and retinitis pigmentosa. Discuss these disorders if appropriate.]

The rods and cones work as a team in the retina. They answer the questions "Where?" and "What?" when we see. The movement of something or someone is best picked up by the rods located in the peripheral part of the retina, our peripheral or "side" vision. This causes a reflex that turns our eyes towards the source of the movement (Where?) so that the image of that something or someone falls on the cones located in the macular, the central part of the retina, in order to identify that thing or that person (What!).

There is a very thin layer of cells with pigment of the retina that touches against the choroid layer of the eye. This thin layer is called the pigment epithelium. (This structure is not labeled on Diagram V-2, "The Human Eye"). The "feet" of the rods and cones come in contact with the pigment epithelium. Light entering the eye and striking the pigment epithelium bounces back against the rods and cones. Light sensitive chemicals found here changes the light into neural, or "electrical", signals which are then sent back to the brain through the optic nerve.

The pigment epithelium and rods/cones layers of the retina lay next to the choroid and get their oxygen and nutrients from the choroid. If the retina should separate from the choroid, the rods, cones and pigment epithelium would eventually die and vision will be lost in that area.
[There are many other disorders related to the retina such as retinal detachment, retinopathy of prematurity, and retinal coloboma. Discuss these disorders as appropriate at this time.]

All the nerve endings from the rods and cones come together at the back and slightly to one side of the eye to form one big bundle called the **optic nerve**. At this place there is a slight depression in the retina called the **optic disc**. Only nerve fibers and blood vessels entering and leaving the eye are present at this spot. There are no rods or cones in this disc, therefore, there is one part of the back of each eye that is "blind", that has no vision. This is called the "natural blind spot" or "physiological blind spot". Sometimes the Latin word *scotoma* is used for the words blind spot. Most of us have never noticed the blind spot of each eye because each eye covers for the other's blind spot. [Do the exercise described on Diagram V-3, "The Natural Blind Spot," which lets the parents experience their own natural blind spot in each eye.]

When the eye doctor looks into a child's eyes with certain instruments, he will become concerned if the disc is a pale color rather than a healthy red. The color and shape of the disc can indicate the health of the optic nerve tract (or even alert the doctor to the possibility of a brain tumor).

Before we continue along the optic nerve and leave the eye itself, we still need to talk about two other things found inside the eye: the aqueous humor and vitreous humor.

Earlier, we discussed that along with changing the shape of the lens, the ciliary body also produces a watery fluid called the aqueous humor or aqueous fluid. Humor is a Latin word that means "water". The aqueous humor provides oxygen and nutrients to the lens and the cornea which do not have blood vessels within them. The aqueous fluid also bathes these parts of the eye and keep them clean and free of infection.

The ciliary body constantly makes aqueous fluid. The aqueous leaves the ciliary body and enters a small area between the front side of the lens and the back side of the iris called the **posterior chamber**. It continues to flow across the front of the lens and out through the pupil, which is the opening in the iris, into the large space between the front side of the iris and the back side of the cornea called the **anterior chamber**. Since aqueous is constantly being made, it has to leave the eye somehow. It does so through a little sieve-like opening called the **trabecular meshwork**. The trabecular meshwork is found where the inside edge of the cornea and the outside edge of the iris come together. The aqueous drains through this trabecular meshwork into a "tube" called the **Canal of Schlemm**. This carries the aqueous to veins which then carry the aqueous out and away from the eye. [Glaucoma is a disorder that relates to these structures and the flow of the aqueous.]

The entire area between the back of the lens and the retina is filled with the **vitreous humor** or **vitreous fluid**. The vitreous is clear and jell-like. Like air does to a basket ball, the vitreous keeps the eye round. Without the vitreous, the eyeball would collapse.

[Refer to Diagram V-4, "Optic Nerve Pathway In the Brain," for the remainder of this topic.] The optic nerves leaving the back of both eyes come together at a point, just in front of the brain itself, called the **optic chiasm**. At this point, some fibers in the optic nerve from the right eye "cross over" and join some fibers from the optic nerve of the left eye and, together, go on to the left side of the brain. Likewise, some fibers from the left optic nerve "cross over", joining some fibers from the right optic nerve and, together, they go on to the right side of the brain. We will discuss this again later when we get to Topic 2: How the Visual System Works.

Each bundle of nerves continue, one along the right side and the other along the left side of the brain, to points on each side of the brain called **lateral geniculate bodies**. These can be thought of as control towers which direct traffic (messages) from the eyes to where they need to go.
Eighty percent of the information goes to an area in the back of the brain called the visual cortex. The visual cortex is the part of the brain that interprets the signals traveling along the optic nerves. The visual cortex is divided in half between the left and right occipital lobes.

The remaining twenty percent of the information goes to the brainstem which controls involuntary visual functions such as eye blinks and the changes in the size of the pupils.

Disorders related to the optic nerve tract are optic nerve atrophy, optic nerve hypoplasia, hydrocephalus, and tumors putting pressure on the nerves and thereby damaging them.

A doctor once said, "The eye doesn't know a horse from a barn. It is the brain that sees." The eye is connected via the optic nerve to the visual cortex of the brain. It is this special part of the brain which forms a picture from information collected by the retina. The visual cortex in turn sends information to other areas of the brain which process and respond to it. For example, when you read your name, the frontal lobe causes the extraocular muscles to move, which causes the eyes to move during reading; the visual centers in the brain process information about the line configurations and shapes; the language centers of your brain help you identify your name; motor, language, and auditory centers help you say it and so on. Hundreds of pieces of information are being shared and processed in seconds. There are more interconnections between the visual cortex and other areas of the brain than between any other sense area and other areas of the brain. The visual system provides the brain with 75% of its information. [The eye disorder related to this part of the visual system is cortical visual impairment.]

In our next topic, we will discuss how the visual system works. We will refer back to this topic any time we need to do so.

SAMPLE ACTIVITIES

1. Use the activities suggested in the Sample Discussion (pupillary response, natural blind spot activities).

2. Show and discuss a simple book on the visual system (e.g., Your Eyes, Chapter 1, see Reference and Reading List below) or an article or chapter on the visual system which could be obtained from the early intervention program or a local library.

SAMPLE CHALLENGES

1. Read through this article (chapter or book) on the eye.

2. Go to the library and find additional information to read about the eye and the visual system.

REFERENCE AND READING LIST


Diagram V.1

THE EXTERNAL STRUCTURE SURROUNDING THE EYE

RIGHT EYE

FRONT VIEW
Behind the Eyelids

SIDE VIEW

Extraocular
Muscles

Upper Eyelid

Lower Eyelid

Lashes
**THE HUMAN EYE**

(Cut in half and looking from the top downward)

**RIGHT EYE**
Hold this page at arm's length, and close your left eye. Stare hard, with your right eye at the dot. Move the page slowly towards you, still staring at the dot. What happens to the cross?

Try the experiment again, but with your right eye closed. Stare with your left eye at the cross - then start moving the page slowly toward your face. What happens to the dot?
OPTIC NERVE PATHWAY IN THE BRAIN

Left Eye
Optic Nerve
Optic Tract
Lateral Geniculate Body of Thalamus
Visual Cortex
Right Eye
Optic Nerve
Optic Tract
Lateral Geniculate Body of Thalamus
Visual Cortex

Farside of Retina
Noseside of Retina
Midbrain
Superior Colliculus and Pretectal Area
Left Occipital Lobe
Right Occipital Lobe

Diagram V.4
TOPIC 2: FUNCTION OF THE VISUAL SYSTEM

GOAL
Parents and other caregivers will understand how the visual system works and relate this to the child's vision problem.

MATERIALS

- Diagram V-5, "Light Rays And How They Are Bent In The Eye"
- Diagram V-6, "Normal Visual Field"
- Diagram V-7, "Accommodation of The Lens"
- Diagram V-8, "Ambiguous Figures"
- An eight to 10 foot length of thin rope
- Prism
- Clear glass of water and a pencil
- Piece of paper with a lot of print on it

INSTRUCTIONS TO EARLY CHILDHOOD SPECIALIST

Adapt and simplify this topic for parents and caregivers who are not as interested in how the visual system works. Relate the information to the child's own vision problem as much as possible. For example, when talking about how light is bent and focused on the retina, talk about how the child's refractive error is being corrected if the child has this problem. Or, when talking about how the eye moves, point out how the child's muscle imbalance affects those normal movements.

If needed, go back over any points in the Sample Discussion section of the previous topic, Anatomy of the Visual System. If the parents should forget names, locations, or functions of any part of the visual system that is related to their child's vision problem, encourage them to review the needed information in the previous topic.

SAMPLE DISCUSSION

In order to see, we need light. The eye is the sense organ that receives rays of light that reflect off of objects that we see. The eye then changes those light rays into electric, or neural, signals and sends them on to the brain for interpretation. From the patterns of light rays striking against the retina and the storehouse of past experiences recorded in our brains, we perceive a world of colorful objects and living things in three dimensional space.

Light rays travel in waves. Scientists have found that the rays of light for each of the colors we see—red, orange, yellow, green, blue and purple—travel at different frequencies. A frequency is how many cycles, or "waves", per second a ray of light has. [Use the length of rope to demonstrate light frequency and what is meant by a low frequency verses a high frequency. Have a parent or someone else hold one end of the rope, or tie it to a chair or door handle. You take the other end of the rope. For the purpose of this demonstration, only shake the rope up and down.] This length of rope will show us how light travels and what I mean by frequency. As I shake this rope, you can see the "waves" traveling from my end of the rope to the other. Light travels in much the same way. You can see that the waves go up and down,
like peaks and valleys. From the top of the peak of one wave to the top of the peak of the next wave, is one cycle. [Shaking the rope slowly, continue by saying:] Right now I'm shaking the rope slowly. The distance from the peak of one wave to the peak of the next is long. The waves are traveling slowly. Not many waves or cycles are traveling from my end of the rope to the other. This is a low frequency. [Increasing the shaking of the rope...] You can see that as I shake the rope faster and faster, the distance from the top of one peak to the top of the next is getting shorter and shorter. The waves or cycles are traveling more quickly. This is a higher frequency.

The light rays of each color travels at a different frequency. Violet and blue light have the highest frequency. They have the quickest, shortest cycles or waves. Yellow and red light rays have the slowest, longest cycles or waves.

Sunlight is all the frequencies of light combined together. We can see this with the help of a prism. [Show the parents how light which passes through a prism is broken into the various colors.] When sunlight hits a prism, it breaks up into each of the different colors of the rainbow. You can see that violet and blue are at one end and yellow and red are at the other. Red light has the lowest frequency and violet has the highest. There is a form of light that travels slower than red light and faster than violet, which our eyes cannot see.

During our discussion for our last topic, Anatomy of the Visual System, we discussed that the part of the retina called the macula had a high concentration of cells called cones. Cones are the cells in the retina that can best tell the difference between the different frequencies of light. The cones are the cells responsible for seeing color. Have you ever wondered why babies start out preferring bright colors such as red and blue instead of soft pastel colors? It is thought that the cone cells responsible for seeing brighter colors are the first to fully develop.

As billions of light rays travel through the air, they bump into objects such as the furniture in your house, my book--just about everything. Like all objects, my book absorbs some of the waves of certain frequencies of light and reflect other waves of certain other frequencies. As you look at my book, the light rays reflected back in different patterns enter our eyes and strike and stimulate the retina. The patterns of light reflected by my book are different from those reflected by the different furniture in this room. This allows the brain to tell the difference between my book and, let's say, the chair where I am sitting. Different objects--people, plants, animals and buildings--look different or similar depending on the patterns of light reflected from them as well as our past experiences with these objects.

It is also very important to know that light rays bounce off objects in all directions. Remember this because we will discuss this more in just a little bit.

As light enters the eye, it is refracted, or bent, as it passes through the cornea, the lens and the fluids in the eye. This happens because as light rays pass from one clear substance, in this case, the air, into a more dense substance, such a water, the light rays slow down a little bit and change direction. [Demonstrate this by putting a pencil in a glass of water.] Look what happens to the image of this pencil as the rays of light from it travel through the water. When you look at the pencil from the side, the pencil appears to bend. The light rays bouncing off the pencil have been refracted, or bent.

Let's follow the path light takes as it enters the eye. (Show and refer to Diagram V-5, "Light Rays And How They Are Bent In The Eye"). As light rays enter the curved surface of the cornea, they are bent towards the pupil and the back of the eye. They are bent even more by the aqueous fluid and then by the lens of the eye. The light rays are also bent by the vitreous humor as well. All of these--the cornea, the lens and the fluids in the eyes--work together to focus the light rays into a clear image on the macular area of the retina on the back of the eye. Notice in the picture here that the image is upside down on the retina. This happens when the light rays go through the lens. The image is put back right-side-up by our brain.
Both of our eyes working together can see things in a large area above, below and to the sides of our eyes. The area we can see is called a visual field. From our far left to our far right, this area or visual field, ranges from about 160 - 180 degrees. (Show Diagram V-6, "Normal Visual Field").

Let's do a little demonstration now. (Give the parents the piece of paper with writing on it and have them hold it out to the sides of their heads as they look straight ahead at you.) What are you visually aware of about the paper you are holding? Can you read anything on it? No? That is because the light rays bouncing off it and into your eyes are landing on the periphery or outside edges of your retina. Can you remember what kind of receptor cells are in this part of the retina and the information they give you? These are the rods. They give gross form, awareness, and movement information. This peripheral vision is what enables you to avoid all the toys on the floor as you run to grab the stool your baby is about to pull over on himself.

Now, slowly move the piece of paper around to the front of your eyes until you are able to read it. The light rays reflecting off the print on the paper are now entering the eye in such a manner that they are focused on the macula area of the retina. Can you remember what kind of receptors cells are found there and the role they play? These are the cones. They allow us to see fine detail and color. Actually, your eyes are only able to read a small area of print. The surrounding print is blurry. This is because, as we discussed earlier, the area on the retina that gives us our finest vision is very small, like a pinpoint.

[For this next paragraph, show Diagram V-7, "Accommodation Of The Lens"].

Not only does the position of an object in relation to the eyes, to the sides or straight ahead, determines how our eyes have to adjust in order to see areas clearly, but also its distance from the eyes. Pretend you are looking at a tree 20 feet or more away. Remember a few minutes ago I said that light rays bounce off objects in all directions? These rays are traveling away from each other. We call this diverging rays. When an object is 20 or more feet away, the only light rays that are coming towards our eyes from that object, in this case, the tree, are all traveling in the same direction--they are parallel. All the others have traveled in different directions quite a distance from the eye. Since the light rays from the tree are already traveling in a parallel fashion, they do not have to be bent much to be brought into focus on the retina. Therefore the lenses in your eyes are relaxed and are flatter in their shape.

Now pretend that you are looking at one leaf on the tree as you are walking closer to it. The leaf appears to be getting bigger. Try this with some real object in this room. [Pause and let the parents do this.] This is because the image of that object is taking up more and more of your visual field and stimulating a larger and larger area on your retinas. The rays now entering your eyes from that object are no longer parallel. They are diverging, or moving out in all directions. The lenses in your eyes now must bend those diverging rays of light so that they travel in the same direction towards the back of your eyes and the retinas. The lenses now have to become "fatter" to bend or refract those light rays.

This process of the lens changing shape to keep objects in focus at both near and far distances is called accommodation. Accommodation happens so quickly we are not aware of it. Try glancing around the room at different objects at different distances, including some very close and some very far away. Think about how quickly your lenses are accommodating to keep things in focus. Here is another exercise to try: Hold two fingers of one hand at arm's length in front of you. Focus on those two fingers and notice how blurry the distant background is. Now, focus on an object across the room as you look past your fingers. How do your fingers appear now? (Hopefully, blurry. The parents may say it looks like they are holding up three fingers instead of two.)
When following an object as it moves closer to you, both of your eyes must begin to turn in equally towards each other and the nose. We call this convergence. As you follow the object back out as it moves away from your face, your eyes move apart or diverge. Watch my eyes as I demonstrate this. (Do this so that the parents can see the movements of convergence and divergence.)

Your eyes are never completely still. When they are searching for something they move together in small rapid jerks called saccades. (Let the parents observe this as you visually search and scan the environment.) When I am watching something move, or tracking, notice how my eyes move more slowly. When my eyes are focused on something like that picture on your wall, my vision will start to fade after a few seconds. My eyes have to repeatedly sweep the image of that picture over the receptor cells on my retina so that those cells do not start to adapt and cease sending a signal of the image of that picture to the back of my brain. What is so amazing is that in spite of all the little eye movements, the saccades, the picture does not appear to be moving! Now you try this.

The eyes are slightly separated so that each one sees the world from a slightly different angle. Point to a small object on the wall or on a table, looking at it with both eyes. Keep your finger in the same position as you close your right eye, then quickly open it while shutting your left eye. Continue switching back and forth between your eyes. Did you notice anything? (The object and finger shifted position back and forth.) The brain uses this slight difference between the images of each eye along with other clues to give us depth perception which is also called binocular vision or stereo vision. Depth perception is the ability to judge distances so that we can do such things as reach for and grasp an object, react in time to keep from getting hit by a flying object or to parallel park a car. Close one eye and reach for a pencil on the floor as you use your open eye to do the reaching. What did you experience?

It has often been said that children with monocular vision, which is usable vision in only one eye, do not have depth perception. This is not quite true. These children may have difficulty with some tasks requiring depth perception, such as reaching, climbing steps or catching a ball, but many of these children have found ways to get around this. Their brains, like ours, learn to use experience and other clues for judging distances. Some of these clues are:

1. Relative size: objects look smaller the further away they are and bigger the closer they are;
2. Shadows which are cast by the objects;
3. Brightness;
4. Clarity: close objects are in sharper focus; and
5. Motion of the objects.

Up until now, we just talked about how the eyes move, how large of an area they can see, and so on; but now let's discuss what happens when light rays strike and stimulate certain areas on the retina.

Remember that the receptor cells, the rods and cones, turn the light energy into electrical, or neural, signals which then quickly travels along the optic nerve path. (Use Diagram V-4, "Optic Nerve Pathway In the Brain" from Topic 1 as you discuss the optic nerve pathway.) At the chiasm, where the two optic nerves meet, some of these signals, or messages, cross over to the opposite optic nerve while others stay on the same side. On their way from the chiasm to the brain, the nerves along the optic tract pass through a relay station on each side of the brain called the lateral geniculate bodies. Here, the signals are divided up and sent to different areas of the brain.
Eighty percent of the signals and information goes to an area in the back of the brain called the visual cortex. The visual cortex is the part of the brain that interprets the signals traveling along the optic nerves. The visual cortex processes the information about the form, brightness, color, size, distance, depth, lines and edges of the object being seen. All of this is put together into an image that we see. The visual cortex then sends this image to other areas of our brain which in turn help us to recognize, name, interpret and use the information provided by the visual system.

Seeing, or visual perception, involves many sources of information beyond those which meet the eye. It involves knowledge of an object or person that we get from previous experience which include touching, smelling and hearing the object or person. For example, your baby comes to know you over time because of all the different things you have done with him. He has come to know your touch, your smell and your voice.

During visual perception, or seeing, your brain is actively looking for the best way to make sense out of what it sees when looking at an object. Along with the image being formed in the visual cortex, your brain searches its memory of experiences to locate any other information associated with the object. For example, if you turned the TV on and people and animals started coming out of it into your living room, you would think that your TV was not a TV after all. Why? Because based on your past experiences, the thing that looked like a TV would not fit your previous experience about how a TV works; namely, animals and people just do not come out of it!

To show you how active, or dynamic, seeing really is, I have some pictures for you to look at. (Use Diagram V-8, "Ambiguous Figures" and ask the parents what they see in each picture. Then let them read the comments below each picture.) Did you notice how your mind kept trying to figure out the drawings as you looked at them? It tried to compare them to things you know about, searching for the best interpretation or understanding about them.

Does this give you a bit of a feel for how complex the visual system is? In a sense, we learn to see. This becomes crucial when dealing with a child whose visual system is not working quite as it should. To a certain degree, he, too, can learn to put meaning to the blurs and blobs in his world through repeated multisensory experiences with people, things and events he comes in contact with on a daily basis.

We will discuss this later in other topics.

**SAMPLE ACTIVITIES**

1. Do all the activities suggested in the Sample Discussion.

2. Share a simple book, article or chapter on visual perception checked out from the program or local library (e.g., *Your Eyes*, Chapters 2 and 9. See Reference and Reading List below).

**SAMPLE CHALLENGES**

1. Look through this article (book or chapter) about how the visual system works. You may even wish to see if your local library has any useful information on this topic. We can discuss some of the key things you learned from your reading next week.

2. This week think about how your child’s visual impairment has affected the normal visual process and what this might tell you about how he sees. Be ready to discuss this during our next home visit.
REFERENCE AND READING LIST


Diagram V.5

LIGHT RAYS AND HOW THEY ARE BENT IN THE EYE

RIGHT EYE
(Cut in half and looking from the top downward)
NORMAL VISUAL FIELD

When the two eyes function together, the fields of view fuse in the brain as a single, three-dimensional image.

Central Field

About 25°

Peripheral Field

160° - 180°

Unclear Image in the Periphery
ACCOMMODATION OF THE LENS

FOR CLOSE OBJECTS:
Light rays are divergent and need to be bent more in order to focus them on the retina. Notice that the image on the retina is larger when looking at near objects.

FOR DISTANT OBJECTS:
Light rays do not need to be bent as much to focus them on the retina.
What are Visual Acuities and Visual Fields?
What are Visual Acuities and Visual Fields?

Visual Acuities:

*Visual Acuity:* Is the term used to state how clearly the consumer is able to see the fine detail of an object and at what distance. In short, it states how clearly the consumer sees.

There are two types of visual acuities that are measured:

- *Far Distance Visual Acuity* (often just called "Distance Visual Acuity")
- *Near Distance Visual Acuity* (often just called "Near Visual Acuity")

Doctors in the United States usually record far distance visual acuities in what is called Snellen Equivalents. The numbers - 20/20, 20/200, etc. - all provide usable information. The top number, the numerator, is the distance from the consumer's eye(s) to the target, usually an eye chart. The bottom number, the denominator, is the size of the print or target read/identified.

These numbers can provide a meaningful reference point to help get an functional idea of just what the consumer can see. A consumer with the far distance visual acuity of 20/20, which is "normal" distance vision, sees an object clearly at 20 feet what *should* be seen clearly at 20 feet. If a consumer has the distance visual acuity of 20/70, which is the visual acuity sometimes used to determine "visual impairment", the consumer can see an object clearly at 20 feet that another person with "normal" distance vision (20/20) sees at 70 feet. A consumer who has the distance visual acuity of 20/200, which is the best corrected distance visual acuity used to determine "legal blindness", can see an object clearly at 20 that another person with "normal" distance vision (20/20) sees clearly at 200 feet!

Near acuities indicate how well a consumer can see to perform near, detailed tasks such as reading, sewing, and drawing. Unlike distance acuities which are written in the United states almost solely using the Snellen Equivalent, near visual acuities are recorded in several different ways.

To show the relative size of letters/numbers/targets these various systems indicate, a *Table of Approximate Equivalents of Near Notations* is provided.
<table>
<thead>
<tr>
<th>Metric Equivalent</th>
<th>Snellen Equivalent</th>
<th>Jaeger Equivalent</th>
<th>Point/N Equivalent</th>
<th>Approx. Letter Size</th>
<th>Examples of Test Size</th>
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</thead>
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<tr>
<td>13M</td>
<td>20/640</td>
<td>---</td>
<td>---</td>
<td>18mm</td>
<td>headlines</td>
</tr>
<tr>
<td>12M</td>
<td>20/600</td>
<td>---</td>
<td>---</td>
<td>16mm</td>
<td>headlines</td>
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<tr>
<td>10M</td>
<td>20/500</td>
<td>---</td>
<td>---</td>
<td>14mm</td>
<td>headlines, can labels</td>
</tr>
<tr>
<td>9M</td>
<td>20/450</td>
<td>J #20</td>
<td>48 Pt</td>
<td>12mm</td>
<td>headlines, can labels</td>
</tr>
<tr>
<td>8M</td>
<td>20/400</td>
<td>J #19</td>
<td>30 Pt</td>
<td>11mm</td>
<td>subheadlines, can labels</td>
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<tr>
<td>6M</td>
<td>20/300</td>
<td>J #16</td>
<td>27 Pt</td>
<td>9mm</td>
<td>subheadlines, can labels</td>
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<td>5M</td>
<td>20/250</td>
<td>J #15</td>
<td>24 Pt</td>
<td>7mm</td>
<td>subheadlines, can labels</td>
</tr>
<tr>
<td>4M</td>
<td>20/200</td>
<td>J #18</td>
<td>20 Pt</td>
<td>5.5mm</td>
<td>&quot;jumbo&quot; large print, can labels</td>
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<tr>
<td>3M</td>
<td>20/150</td>
<td>J #16</td>
<td>20 Pt</td>
<td>4.5mm</td>
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<td>2.5M</td>
<td>20/125</td>
<td>J #13</td>
<td>18mm</td>
<td>3mm</td>
<td>some large print, labels, children's books</td>
</tr>
<tr>
<td>2M</td>
<td>20/100</td>
<td>J #11</td>
<td>18 Pt</td>
<td>2.5mm</td>
<td>children's books (grades 1 - 3)</td>
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<tr>
<td>1.5M</td>
<td>20/80</td>
<td>J #10</td>
<td>14 Pt</td>
<td>2mm</td>
<td>children's books (grades 4 - 7), some magazine print</td>
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<tr>
<td>1.2M</td>
<td>20/60</td>
<td>J #7</td>
<td>10 Pt</td>
<td>1.5mm</td>
<td>some hard bound books</td>
</tr>
<tr>
<td>1M</td>
<td>20/50</td>
<td>J #5</td>
<td>9 Pt</td>
<td>1mm</td>
<td>hardbound books, some magazines, and typing</td>
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<tr>
<td>.80M</td>
<td>20/45</td>
<td>J #4</td>
<td>7 Pt</td>
<td>.80mm</td>
<td>(a.k.a., <em>standard print</em>)</td>
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<tr>
<td>.75M</td>
<td>20/40</td>
<td>J #3</td>
<td>6 Pt</td>
<td>.75mm</td>
<td>most paperbacks, some magazines, typing</td>
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<tr>
<td>.50M</td>
<td>20/25</td>
<td>J #1</td>
<td>4 Pt</td>
<td>.50mm</td>
<td>(a.k.a., <em>standard print</em>)</td>
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<tr>
<td>.37M</td>
<td>20/20</td>
<td>J #1+</td>
<td>3 Pt</td>
<td>.30mm</td>
<td>newspapers, phonebooks, paper backs</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
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<td>this is extremely fine print</td>
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Samples of *Approximate* Point and Metric Print Sizes

This text is in 6 Pt/7.5M print

This text is in 8 Pt/1M print

This text is in 10 Pt/1.2M print

This text is in 12 Pt/1.25M print

This text is in 14 Pt/1.5M print

This text is in 18 Pt/2M print *

This text is in 24 Pt/3M print

This text is in 36 Pt/7M print

This text is in 48 Pt/9M print

* Most books produced by the American Printing House for the Blind (APH) are printed in 18 Pt/2M print.
Visual Fields:

*Visual Fields:* A functional definition of visual fields is the total area a person can see - above, below, to the sides, and straight ahead - while looking straight ahead and keeping the head and eyes still.

Any area in the visual field where there is missing or distorted vision is called a blind spot or *scotoma* (pl. *scotomas* or *scotomata*).
The Five Types of Vision Loss
The Five Types of Vision Loss

Visual problems are the result of one or more of the following areas of visual functioning:

*Visual acuity:* The consumer does not see as clearly as she/he should; images are not sharp.

*Visual field:* The area the consumer can see - above, below, to the sides, and straight ahead - is limited when holding her/his head and eyes still.

*Contrast sensitivity:* The consumer is unable to see the relative difference between the lightness and darkness of objects (something like looking at a faded photograph).

*Ocular motor control:* The consumer has difficulty moving her/his eyes when visually fixating, following, and/or scanning objects.

*Visual processing:* The consumer's brain is having difficulty making sense out of what she/he is seeing.
INSITE RESOURCES FOR FAMILY CENTERED INTERVENTION FOR INFANTS, TODDLERS, AND PRESCHOOLERS WHO ARE VISUALLY IMPAIRED

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TOPIC 3: HOW VISION IS AFFECTED

GOAL

Parents and other caregivers will know the implications of the child's eye disorder(s), including how the child sees and the treatment recommended for the disorder.

MATERIALS

- Diagram V-9, "The Eye Compared To a Video System"
- Diagram V-10, "How Visual Losses Appear"
- Diagram V-11, "Different Problems That Can be Found in the Optic Nerve Pathway With the Resulting Visual Field Loss"
- Diagram V-12, "Normal Gazes"
- Diagram V-13, "Heterotropia or Strabismus"
- Diagram V-14, "Vision is More Than 20/20 Eyesight" (copies of this, in groups of 500 cards, can be purchased from Optometric Extension Program, 2912 South Daimler Street, Santa Anna, CA 92705)
- Low vision simulators, one of which is similar to the child's visual loss. (If the child has both hearing and vision problems, be sure to add earplugs and headphones.)
- An acuity chart, preferable a Snellen with the big "E"

INSTRUCTIONS TO EARLY CHILDHOOD SPECIALIST

Because of the importance of this topic, it may be helpful to spend at least a couple of weeks on it. Know ahead of time if the child's ocular disorder has resulted in one or more of the following functional visual problems:

1. Loss of visual acuity - distance and/or near;
2. Loss of visual field;
3. Contrast sensitivity;
4. Oculomotor problems; and/or
5. Visual processing difficulties.

Share the introductory information on the first page of the Sample Discussion. Then, go over the information which relates specifically to the child's ocular disorder and apply it to his specific situation. For example, if the child has cataracts only, the information on acuity and contrast sensitivity loss may be all that applies to him. If the child has brain damage with cerebral palsy and a diagnosis of cortical visual impairment with strabismus, the information on processing and oculomotor disorders will be the most applicable.

When doing activities with the vision simulators for the visual acuity and visual field loss, make sure that you have the parents experience several activities. For example, have them:

1. Walk around or find something upon request;
2. Find an item to order out of a catalog, read a book, do a puzzle, or do some handwork; or
3. Pour a cup of milk or brush their teeth.
Instructions for making low vision simulators are found in Appendix B on page 635. If the child is also physically impaired, try to simulate a physical handicap in some of the activities (e.g., parents can use only one hand, or can have no head control). If the child is also hearing impaired, give a simulated auditory impairment (cotton balls or earplugs may be placed in ears with earphones on top of all of that, or speak very softly or not at all, or use only gestures). When finished with an activity, ask the parents (or caregiver) how it felt and what kinds of things helped or hindered their efforts. Relate their experiences to what it might be like for their child to function with his visual impairment.

SAMPLE DISCUSSION

The visual system - the eyes, the optic nerves, and the brain - can be compared to a video system: the video camera, the cable, and the television set [show parents Diagram V-9, "The Eye Compared To a Video System"]. Of course this analogy is simplistic since both the visual system and the video system are much more complex than shown in this figure.

Many things can go wrong in either system. For example, if the lens on the video camera is scratched, the image on the TV will not come through clearly. If the camera is out of focus, the picture will be blurred. If the cable connecting the video camera and the TV is defective, the signal to the TV will be weak or absent. Problems within the TV itself will also affect the picture on the TV. All the parts have to be working well together for a clear image to appear on the television screen. In the same way, the eye (which collects information), the optic nerve (which carries it), and the brain (which interprets it) must all work together for good vision to occur.

There are numerous things that can go wrong with parts of the visual system, resulting in hundreds of eye problems such as cataracts, retinopathy of prematurity, or strabismus. Most children with vision impairments have more than one eye problem at the same time which then affects how they see in many ways.

Most eye problems fall into one or more of five categories used to describe visual loss. These are:

1. **Loss of Visual Acuity:** The child does not see as clearly as he should, images are not sharp.
2. **Loss of Visual Field:** The area the child can see - above, below, and to the sides of his eyes - is limited when holding his head and eyes still; or he may have islands of vision.
3. **Oculomotor Problems:** The child has difficulty with moving his eyes when fixating, following, and/or scanning objects with his eyes.
4. **Reduced Contrast Sensitivity:** The child is unable to see the relative difference between the lightness and darkness of objects, something like looking at a faded photograph.
5. **Problems with Visual Processing:** The child’s brain is having difficulty making sense out of what he is seeing.

Remember, most children with vision impairments have problems in more than one of the above areas. For example, a child with retinopathy of prematurity may have visual acuity, visual field and contrast sensitivity losses.

We have had a chance to talk a little about your child’s eye problems. Today we want to look more closely at how it affects his vision.
For discussing each of the five categories used to describe visual loss, refer to Figure 10, "How Visual Losses Appears", as appropriate.

LOSS OF VISUAL ACUITY

A child who has a loss of visual acuity does not see his world as clearly as he should. Let me show you what I mean. [Conduct an activity with several acuity loss simulators, such as 20/200, Light Perception, and No Light Perception. See Appendix B on page 635 for instructions on how to make or purchase low vision simulators.] As you can see, there are different amounts of loss of visual acuity, and no two children see the world in quite the same way.

There are two types of visual acuity losses—distance and near. If a child has a distance visual acuity loss, distant objects appear blurry. If a child has a near visual acuity loss, near objects appear blurry. A very large number of children who have vision impairments have both losses.

Doctors in the United States usually record visual acuities in what is called Snellen equivalents. Have you heard of the term "20/20 vision"? Well, this is a Snellen equivalent.

[If the child's eye exam resulted in a Snellen acuity measure for the child, use the following explanation. If it did not, do not spend much time on the explanation unless the parents are interested.]

- What do these Snellen visual acuity numbers mean? The top number - the first number said, the numerator - tells us how far your child stood from the eye chart when he was tested. The bottom number - the second number said, the denominator - tells us what was the smallest line your child was able to read or identify.

These numbers also mean something else. The big "E" at the top of the standard eye chart is a "200 foot" sized letter. This means that a person with normal, or 20/20, vision can read the "E" at 200 feet - 2/3 of the length of a football field. A legally blind child with visual acuities of 20/200 could read the "E" at only 20 feet away while the person with normal vision could read the "E" at 200 feet away. Some children with vision impairments have visual acuities of 20/400 or 20/800. This means that they could not stand any further than ten or five feet away to read the big "E". A child's visual acuities may be so poor that the acuities cannot be written in numbers. Words like "sees shadows and gross forms" or "sees light" are used.

There are some methods that can be used to get an acuity measurement on babies and children who cannot identify letters or pictures on a chart. These include such testing methods as preferential looking using the Teller Acuity cards and the STYCARS Test (see information on second page of Diagram V-16). These tools and methods are still not commonly used by eye physicians. [Note: If parent wants to know more about these, go into the information on them in greater detail.]

It is difficult to measure exact acuities for babies and children who have multiple impairments. That is why you rarely see numbers like 20/100 written on their eye reports. Often there is just a description of what the doctor thinks the child can see.

By careful observations over a period of time, we can get a good idea of what your child sees. For example, does your child seem to recognize you visually? Can he choose his bottle from other bottles of similar shape or color? Does he recognize a familiar toy using only his eyes?

The causes of loss of visual acuity can vary. The child may be very near or far sighted and even glasses may not be enough to correct his vision to normal. For example, the child may have albinism resulting in a macula that never fully developed, so he does not see clearly enough to read to the 20/20 line. Glasses will not fix the problem. However, he may also have a
refractive error which causes him to be near sighted as well. If you remember from our previous topic discussions, a refractive error means that his eyes are not able to bend light rays appropriately to have the rays focus clearly on the retina at the back of the eye. Glasses can fix that part of the vision loss. Without glasses, he may have 20/200 vision, but with the glasses, his vision may improve to 20/100.

Another cause of acuity loss may be something within the eye that blocks the light rays from reaching the back of the eye. This could be something like cataracts and corneal opacifications - a cloudy lens or cornea. Or else it could be that the retina, particularly the macula, and/or the optic nerve may be damaged or not fully developed. The bottom line is that the brain does not get a clear picture signal.

Surgery does not always fix visual acuity loss either. Sometimes it helps with cataracts and corneal opacifications. Usually, not much can be done about problems in the retina or optic nerve, except surgery for some kinds of retinal detachments or to relieve pressure caused by a tumor or fluid on the optic nerve. We cannot transplant an eye or optic nerve because of the billions of fibers that would have to be correctly reconnected.

In later topics, we will have a chance to explore more closely what your child sees and how well he sees. We will work on ways to help him understand and make sense out of the blurs and blobs in his world. We will find ways to help make it easier for him to see, such as making things larger, getting your child closer to objects, adding more color or contrast, and using appropriate lighting.

Common disorders which have associated visual acuity loss are: high refractive errors such as found with children with Down syndrome; cataracts; retinal detachment; ROP; glaucoma; Leber’s amaurosis; underdeveloped eye as in microphthalmia; retina, macula, and/or optic nerve hypoplasia; optic nerve atrophy; albinism; and, aniridia. (See Appendix C on page 638 for descriptions of common ocular disorders with associate functional implications found in young children.)

LOSS OF VISUAL FIELD

A child with a visual field loss is limited in how large of an area he can see. Let me show you what I mean. [Have the parents look through a couple of field loss simulators, such as those representing tunnel vision or central field loss. Make sure one of the simulators is a mixed simulator which simulates both the child’s field and acuity loss.] As you can see, there are varying degrees of field loss. Most of us can see an area of about 160° - 180° in front, above, below, and to the sides of the eyes while holding both our head and eyes still and looking forward. A child with a visual field of 20° or less in his best eye, which is like looking down a tunnel or the tube of a roll of toilet paper, is considered legally blind. The acuity in that eye may or may not be normal. More often than not, it is poor. As you experienced, even with good visual acuity, a severe visual field loss can make reading and just getting around quite difficult. Some children have "islands" of vision which might be like looking through swiss cheese. Other children have just the opposite with little "blind spots" scattered around their visual field. If a child has lost his central visual fields, or central vision, he may have problems with seeing color and fine detail. He will have to use his peripheral visual fields, or side vision, for getting around and seeing things.

It is not uncommon to see the word scotoma which means a blind spot or area in which a child does not see. The child has to learn to look around it. When a total loss of half of the visual field occurs, it is called a hemianopsia. For example, a child with a left hemianopsia may only see the right half of his high chair tray at one time and may not see the food on the left side of the tray.

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It is common to see a child with a central field loss holding his head in a different position when he is looking at people or things. This allows the image of the person or thing to fall on the area of his retina that is giving him the best vision. This is one way in which the child has learned to compensate for not having a full visual field. Some children with nystagmus (jerky eye movements) may also use a different head posture when looking. This is because of where their "null" point is. This is usually a point in the periphery where they are better able to hold their eyes to decrease the jerky movement, thus giving them a slightly clearer image of what they are looking at.

It is very difficult for doctors to measure a young child's visual fields accurately. Sometimes, the doctor may only get a general idea as to where any field problems may be and where your child sees the best and the worst. By carefully observing over time your child's behavior when he is looking, we can get a good idea as to where your child sees the best. For example, when he looks at you or a toy, does he have a tendency to look from one side? Does he bump into things on one side? Does he look at things quite often? Does he look at you by dropping his chin and tilting his eyes? If the child utilizes an unusual head and/or eye position when looking or using his vision, you and the parents can try to analyze what that means. Begin to talk about the behaviors that indicate where the child might see best. Remember, when the child also has a motor impairment, it may be more difficult to sort out how much of the head and/or eye positioning is due to his visual loss and how much is due to the motor impairment. Talk to the child's physical or occupational therapist to make sure that the head position is not a head or neck problem in need of strengthening.

Visual field losses can be caused by any eye problems that block the light rays from reaching the back of the eyes. These could be things like cataracts and corneal opacifications - a cloudy lens or cornea. More often, a field loss is the result of damage or abnormalities of the retina or optic nerve such as a detached retina, retinitis pigmentosa, or a brain tumor putting pressure on the optic nerve. A field loss does not always mean that the child sees nothing in a particular area. It can mean that vision may be more blurred in that area than in other areas. If there is damage somewhere along the optic nerve tract, doctors can sometimes estimate where it is by looking at how a child's visual fields are affected. (Use Diagram V-11, "Different Problems That Can be Found in the Optic Nerve Pathway With the Resulting Visual Field Loss", if it would help the parents better understand the child's field loss when damage to the visual pathway or visual cortex is involved. You may need to help orient the parents to this somewhat complex figure to aid in their understanding it.)

Usually, there is very little that can be done medically for a field loss unless the disorder happens to be operable, as in the case when a cataract is removed or part of a retina is reattached. Has your child's doctor discussed any type of treatment that he feels would help reduce the severity of your child's visual field loss?

In later topics, we will have a chance to find out more about your child's visual field loss. We will teach him some ways to compensate for his loss, such as using search patterns and scanning to piece his fragmented views of things into wholes.

[Common disorders that result or could result in field loss are Stargardt's disease (or any other type of macular degeneration), retinitis pigmentosa, retinopathy of prematurity, glaucoma, retina detachments, and tumors putting pressure on portions of the optic nerve tract. See Appendix C on page 638 for descriptions of specific ocular disorders and their associated functional problems.]
REDUCED CONTRAST SENSITIVITY

A child with reduced contrast sensitivity has difficulty telling the relative difference between the lightness and darkness of whatever he is looking at. Everything can appear washed out, like looking at a faded photograph. This is especially a problem for a child trying to tell things apart that are almost the same color.

Reduced contrast sensitivity can affect how a child perceives his world around him visually. It can affect his recognizing faces, his ability for locate things with his eyes, and even his posture.

Reduced contrast sensitivity can be the result of reduced visual acuities and visual fields, glare, or even problems in the functioning of the visual cortex at the back of the brain.

Some things can be done to enhance contrast sensitivity such as by using sunglasses/filters, changing the amount and/or type of lighting, or other modifications in the environment. (Such modifications will be covered later in Sections 2.)

[Virtually all disorders affect contrast sensitivity to one degree or another. Some primary eye care practices and low vision services can assess a child’s contrast sensitivity and make specific recommendations on how to utilize or enhance the child’s current contrast sensitivity.]

OCULOMOTOR PROBLEMS

A child with oculomotor problems - problems with the muscles of the eyes - has difficulty with coordinated movements of his eyes. He may not use his eyes together when looking at or following a moving person or thing. He may look with one eye and then the other. As he watches people or things come in close to him, one eye may turn in or out, or both eyes may turn in (cross) or out. The child may have trouble following fast moving things such as balls. He may lose track of them completely. He may have difficulty in reaching for things accurately.

In the case of oculomotor problems, the coordinated way the six muscles that move the eye together stops happening. There are several possible causes for this. The frontal lobe of the brain, which helps control the movements of the muscles that move the eyes, may be damaged. A nerve to one of the muscles may be damaged, thereby paralyzing the muscle. One muscle may be longer or shorter than normal so it may not pull far enough or too far. An eye problem may have decreased the visual acuity in only one eye so much that that eye is no longer able to work equally with the better eye.

Problems caused by the muscles that move the eye not working properly can be very complex and difficult to treat. Some problems are corrected or at least improved with glasses and/or medication, or even surgery. Others might be helped by patching the better eye in order to make the poorer or weaker eye learn to work and become stronger. The younger the child when treatment is provided to correct the imbalance, the better is his prognosis for developing improved vision. So, it is critical that muscle problems are identified during the infant and toddler years.

In some cases, such as a child with cerebral palsy who has had damage to the frontal lobe of the brain, there may be nothing that can be done. The problem is the brain itself. With children who have other motor impairments, it is often hard to determine how much of the problem is caused by the child’s vision problems and how much by the child’s motor problems. Again, it may be helpful to consult with the child’s OT or PT on this problem.

Often a problem with the eyes not moving together causes the child to see double. The more severely affected eye starts to be used less and less. The information sent to the brain by that eye may start to be ignored by the brain (suppression) in order avoid the double vision. If
if this goes uncorrected, the child can become amblyopic - or functionally blind - in that eye.

Another oculomotor problem that is present in many children with vision problems is nystagmus. This means the child’s eyes move in rhythmic jerks. This movement may be in a side to side pattern (horizontal), in a circular pattern (rotary), etc. This is not because the muscles that move the eyes are not working together. It is believed that the normal saccades (movements) of the eye as it fixates become exaggerated in low vision children to the degree that these movements can be observed. Nystagmus sometimes decreases as the child gets older. (If the parents want to know more about this, or it is present in the child, take more time to discuss it.)

[Common terms related to oculomotor problems are strabismus, exotropia, esotropia, alternating exotropia, hypotropia, hypertropia, phorias, and amblyopia. See Diagram V-12, "Normal Gazes" and Diagram V-13, "Heterotropia or Strabismus". See Appendix C on page 647 for descriptions of specific ocular disorders.]

PROCESSING PROBLEMS

A child who has had damage to his brain and also has a cortical visual impairment has a visual cortex that is having difficulty processing and making sense of the information sent to it by the eyes. Very often, the child has healthy, normal eyes and optic nerves.

Let me give you an idea of what this is like. (Show the parents Diagram V-14, "Vision is More Than 20/20 Eyesight". Ask them if they know what the picture is. Let them hold it in any position they would like. If they cannot figure it out after a few minutes, then point out the features and tell them what it is: a cow. Remind them that they have normal eyes and nerves, but their brain is not making sense of the picture.)

Very early in life, children with visual processing problems function as if they are blind. However, with time, stimulation, and neurological maturation, they often progress. This even happens after four or five years of age. In other words, a baby who appears to be blind, one year later may be looking, reaching, and following toys and faces with his eyes. The amount of progress varies from child to child. It will be less if the child has additional problems in the eye or optic nerve. In later topics, we will learn about ways to help children who have cortical vision problems make sense out of their world. However, there is very little that can be done for these children medically.

If an eye doctor suspects cortical visual loss and is unable to get any type of visual response from a child with low vision and multiple handicaps, he may recommend that the child be given a special electronic test called a VER (Visual Evoked Response). This test may let the doctor know for sure if a breakdown has happened in the child's visual cortex by showing whether or not the visual signals are reaching the visual cortex at the back of the brain. The MRI (magnetic resonance imagery) is another test that may be used to diagnose cortical visual impairment.

Although many children with multiple and severe impairments have cortical vision impairments, the term is often overused for something we know little about. The visual cortex can be damaged by severe head injuries, infection like meningitis, prolonged hydrocephalus, and/or difficult birth with associated oxygen deprivation. In addition, some children are born with underdeveloped or even absent brains. See Appendix C, page 642, for more information on CVI.

SAMPLE ACTIVITIES

1. Follow through with the activities in the Sample Discussion (e.g., using low vision
simulators, looking at and discussing the confusing picture of the cow, etc.).

2. Together do some brief observations related to determining the child’s visual acuity and visual field (if and how he positions his head and eyes, if he has an oculomotor problem, etc.).

SAMPLE CHALLENGES

1. Read an article (in a pamphlet or section of a book) about your child’s vision problem. See if you can find out more information about his vision problem at the library or from your child’s eye doctor. You might also want to write to an organization, such as the one listed below, for more information about your child’s vision problem.

   The National Association for Parents of Visually Impaired
   2180 Linway Dr.
   Beloit, WI 53511
   (608) 362-1380

2. I will leave you with the simulator most like your child’s vision loss. Use it off and on throughout the week as you do routine tasks about your home or work - but be careful! You may even want to spend a whole day wearing it. Again, be careful, and make sure someone else is around in case you need help. We will talk about your experiences next week.

3. Make some informal observations this week of how your child functions with his eye problem.

REFERENCE AND READING LIST

6. Hanson, Mary. Beyond tracking: Enhancing vision development from birth to one year of age.
THE EYE COMPARED TO A VIDEO SYSTEM

Diagram V-9

TV video camera

cord

brain

eye

nerve

TV set
HOW VISUAL LOSSES APPEAR

LOSS OF VISUAL ACUITY

Normal visual acuity lets us see sharp details of this house both close up and far away.

The loss of visual acuity causes "blurry" vision. The child may be able to see the shape of something, but not the details.

A person has low vision if the acuity in his best eye with the best correction is between 20/50 and 20/200 (AFB).

A person is legally blind if visual acuity is 20/200 or worse in the better eye with the best correction possible.

In the U.S., one in 500 people are legally blind.
"ISLANDS" OF VISION: a child with this type of field loss only sees isolated parts of people or things when he is looking at them. Like with scattered blind spots, the child's functioning will be determined by how big, how many, and where the "islands of vision" are.

HEMIANOPSIA: A child with this type of field loss only sees half of the people or things when he is looking at them. This half of visual field could be left or right or top or bottom. The child may not notice or bump into people or things 'hidden' in the half of the visual field that has no vision.

A person is legally blind if their visual field is 20° or less in the better eye with the best correction possible.
LOSS OF VISUAL FIELDS

LOSS OF PERIPHERAL (CENTRAL) VISION: A child with this type of field loss will be able to only see straight ahead and will not be able to see people or things at the sides without turning his eyes or head. This is sometimes called tunnel vision. It is like looking through an empty tube of toilet paper.

LOSS OF CENTRAL VISION: A child with this type of field loss will only be able to see a blind or grey spot straight ahead no matter which way he turns his eye. It is like looking at a picture with a spot in the middle.

SCATTERED BLIND SPOTS OR SCOTOMAS: A child with this type of field loss may loose ‘parts’ of people or things when he is looking at them. How big the blind spots are, where they are located, and how many there are, will affect how severe the child’s visual functioning will be.
CONTRAST SENSITIVITY is our ability to tell the difference between the lightness and darkness of things we see. It can also be explained as being able to tell things apart that are almost the same color.

To a child with the loss of contrast sensitivity, everything can appear 'washed out'. It's like looking at a faded photograph.
DIFFERENT PROBLEMS THAT CAN BE FOUND IN THE OPTIC NERVE PATHWAY WITH THE RESULTING VISUAL FIELD LOSS

1. Lesion in left superior temporal retina causes a corresponding field defect in the left inferior nasal visual field.

2. Total blindness, right eye. Complete lesion of right optic nerve.

3. Chiasmal lesion causes bilateral hemianopsia.

4. Left nasal hemianopsia due to a lesion involving the outer aspect of one side of the chiasm (pressure on the optic nerve from aneurysm of left internal carotid artery).

5. Right homonymous hemianopsia due to a lesion of the left optic tract (least common site of hemianopsia).

6. Right homonymous inferior quadrantanopsia due to involvement of optic radiations (upper left optic radiation in this case).

7. Right homonymous hemianopsia due to a lesion of the left occipital lobe. The pupillary light reflex is not impaired.
When you move your eyes straight up, each eye is being pulled by 2 muscles. One moves it up and in; the other moves it up and out. These are the superior rectus and inferior oblique muscles.

When you move your eyes straight down, the two muscles being used on each eye are the inferior rectus and superior oblique.

When you move your eyes up and to the left, the superior rectus and inferior oblique work together. On each eye, one muscle must exert a stronger pull than the other to achieve the turn to the left. In the left eye it is the inferior oblique; in the right eye, it is the superior rectus that pulls harder. The opposite would take place when looking up and to the right.

When you move your eyes to the side, just one muscle is exerting a pull on each eye. When looking to the left, the muscles exerting the pull are the external rectus in the left eye and internal rectus in the right eye.

When looking down and left, the inferior rectus and superior oblique exert the pull in the left eye; the inferior rectus and superior oblique exert the pull in the right eye (underlined muscle exerts greater pull).

When cross-eyed, the internal rectus in both eyes exerts the pull on each eye.
## Phorias

<table>
<thead>
<tr>
<th>Phoria</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Exophoria</strong></td>
<td>A tendency for one or both eyes to turn out when fusion is interrupted.</td>
</tr>
<tr>
<td><strong>Esophoria</strong></td>
<td>A tendency for one or both eyes to turn in towards nose when fusion is interrupted.</td>
</tr>
<tr>
<td><strong>Right hyperphoria</strong></td>
<td>Right eye tends to turn upwards; left turns downwards.</td>
</tr>
<tr>
<td><strong>Left hyperphoria</strong></td>
<td>Left eye tends to turn upwards; right turns downwards.</td>
</tr>
</tbody>
</table>

*These can be overcome by natural desire for fusion. They can cause eye strain and headaches or blurred vision. Are not as serious as conditions in the right column nor are they as obvious. Correct with glasses, medication, patching, and/or surgery.*

## Tropias

<table>
<thead>
<tr>
<th>Tropia</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Exotropia</strong></td>
<td>Turning outwards of one or both eyes, causing person to see double or suppress vision in one eye and use the other to see.</td>
</tr>
<tr>
<td><strong>Esotropia</strong></td>
<td>Turning inwards of one or both eyes causing the above.</td>
</tr>
<tr>
<td><strong>Right Hypertropia</strong></td>
<td>Right eye turns upwards and left turns downwards causing the above.</td>
</tr>
<tr>
<td><strong>Left Hypertropia</strong></td>
<td>Left eye turns upwards and right eye turns downwards causing the above.</td>
</tr>
</tbody>
</table>

*These conditions are worse in degree than the ones on the left and are obvious when looking at child's eyes. May be corrected with patching, glasses, medication, and/or surgery.*
You may not immediately see the subject of the photograph on the reverse side because the seemingly meaningless blobs of light and dark require VISUAL interpretation. When you “see” the subject, you have VISION; before that you had only eyesight. Vision is the ability to get MEANING from eyesight.

Many people have clear “eyesight,” but are slow readers, or have difficulty to get MEANING through the visual pathway. Such persons are handicapped in business, social and school life. Many also suffer visual discomfort or fatigue while reading or doing other close work.

The correction of these problems and the care of this complex physical and mental process called VISION require a highly specialized professional service - a service provided by the modern optometrist concerned with seeing as applied to clarity of eyesight AND the ability to get MEANING through vision.

Published in the interest of better vision by the Optometric Extension Program Foundation, Inc. (Nonprofit), Duncan, Oklahoma, an international organization dedicated to postgraduate education and research in vision.
ACTIVITY:
"WHATZIT?"
Activity:
"Whatzit?"

There are 20 numbered paper bags on the side table. Each bag contains a familiar object.

Without looking inside, reach into a paper bag and feel the object. Are you able to identify it by touch alone? Look at the number on the outside of the bag and write what you think that object is on the appropriate line below. Try to do this with all 20 bags - or as many as time allows.

1. __________________________
2. __________________________
3. __________________________
4. __________________________
5. __________________________
6. __________________________
7. __________________________
8. __________________________
9. __________________________
10. _________________________
11. __________________________
12. __________________________
13. __________________________
14. __________________________
15. __________________________
16. __________________________
17. __________________________
18. __________________________
19. __________________________
20. __________________________
Possible Developmental Delays of a Child with Severe Vision Impairment
Possible Developmental Delays of a Child with a Severe Vision Impairment

(The following information is adapted from the INSITE TRAINER'S MANUAL - SKI*HI Institute)

Social:

The child's bonding with parent(s) is harder to establish and more easily interrupted. There are several reasons for this. The smile response is established late and may not be as frequent; it is harder to evoke. Since the child's vision is affected, the child does not respond to a parent's approach as would a child with normal vision: thrashing, making noises, etc. Rather the child becomes still in order to hear the parent better. The parent may view this as rejection by the child. There may be minimal or no eye contact by the child. Parents must learn to be observant and watch the baby's hands for signals that the child wants to interact with the parents. Since visual input is missing, the baby's main interest is focused on her own body; her parents may interpret this to mean that the baby wants to be left alone. The baby needs a lot more touching, however if the parent feels that the child is not bonding with them, they may feel reluctant to touch or hold the child.

Parents are often overprotective; combining this with the child's lack of vision for incidental learning by observation, social norms often need to be taught. This child may have difficulty in developing a concept of self as separate from other. Because of delays in concept development (and, often, from overprotection by the parents) the child is often much less independent. Due to the lack of vision input and incidental learning, the child may lack awareness of social expectations.

Because the child does not have the benefit of seeing the many ways we show our feelings through body language (facial and body), the child does not appear expressive.

Play:

Interaction skills with other children in play may be a problem. The child may demonstrate less variety in play. Make believe play is delayed (may appear as late as four to five years of age) and once the child has entered this stage of play, the child may remain in fantasy play longer. The child is often less
interested in miniatures (such as doll houses, toy cars, etc). As a toddler, the child may sit and stay rather than explore her environment; the child may need to be motivated to find out all the interesting things in her environment with which she can play.

Cognition:

Object permanence is slower in appearing. The child may need help in learning how to problem solve. The child has a difficult time transforming experiences of touch and sound into mental images. The child may experience delays in whole-to-part concept development. The child may be less flexible, more rigid in thinking. The child may have trouble with problem solving and predicting outcomes. The child continues an egocentric view of the world longer than normal. Concept of time and space is not normal. Accurate and meaningful experience are critical for the child to develop good cognition. The child may need more help with organizing concepts.

Communication and Language:

The baby may babble a lot the first year, but in a subdued way. The baby may quiet quickly when people are present in order to hear what's happening around her. In some cases, the child has problems with the function and purpose of language; the child may exhibit verbalism and echolalia (this is a form of stimulation). Many words can be meaningless to the child because she can't see clearly the picture of object connected with the sounds. The child may have problems with pronouns (the child may not use the word "I" but may refer to herself by her name) and abstract words. The child's development of speech and basic syntax may be normal by the age of four to five years, however, the child may be overly verbal.

Self Care:

The child may need longer time to learn specific skills; the child may need more hand-over-hand/co-active help form parents and others in the early learning stage of an activity. Parent over protection may get in the way.

Child may be messy self-feeders - parents may be tempted to prolong feeding the child delaying independence. Child may initially be resistant to various food textures. Child may not chew properly and may only want to eat pureed food (this may be the result of not seeing chewing modeled). The child may delay
desire to self spoon feed since she does not see others do it and, therefore, doesn't understand that this is the social, "grown up" thing to do.

For dressing, the child may not understand colors and patterns. The child may not see front/back or inside-out so these concepts may be delayed in being established.

May mix up day and night sleeping patterns.

Gross Motor:

Child may have delayed head control and balance due to lack of a visual reference, therefore, other mobility skills such as getting into sitting, raising upper torso, crawling, and walking are quite delayed. A child with normal vision is motivated to raise her head off the floor in order to see her world, this motivation may not be there for the child with a vision impairment; furthermore keeping her chin on the floor may provide that child with additional sensory input. Sitting - first supported and unsupported - matches that of a child with no vision loss since the child with the vision impairment does not need to move - getting into and out of the sitting position, may be a problem. Child may "cruise" along objects for a longer period of time before starting to walk. Child may keep a wide base in her stance. Walking tends to be delayed until ages 18 - 19 months. Jumping, skipping, and running are hard for the child to learn and perform.

Fine Motor:

Child has to be taught to use her hands. While a 3 - 4 month old child with full vision is fascinated with watching her hands engage in the midline, the child with a severe vision loss brings her hands together only accidentally and not in a purposeful way. Reaching for sound is delayed 9 - 12 months. Child may be tactually defensive. The child may lack the vision to adequately monitor reach, grasps, and manipulation, therefore, tasks take longer until learned. Child may need hand-over-hand/co-active assistance and sound to help in the learning process. Many visually orientated tasks are not exciting to the child so many of the fine motor skills generally used in these tasks are therefore not re-enforced.
Other Behaviors:

The child may develop other behaviors ("blindisms"): eye poking; head shaking; rocking; reciting jingles and echolalia, etc.
How We Hear
How We Hear

Sound enters the external ear canal and makes the tympanic membrane (eardrum) vibrate. The vibrations are transmitted to the cochlea through a series of three small bones, the ossicles. The vibrations set up in the fluid contained in the cochlea result in stimulation of the nerve endings in the cochlea and transmission of impulses to the brain through the auditory nerve.

The eustachian tube connects the middle ear and the throat, allowing equalization of air pressure on both sides of the eardrum. The semicircular canals play no role in hearing, but help a person maintain his balance.
THE INSITE MODEL

Home Intervention for Infant, Toddler, and Preschool Aged Multihandicapped Sensory Impaired Children

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TOPIC 1: SOUND; CONTACT WITH THE WORLD

PARENT GOALS

1. Parents will describe four reasons why sound is so important:
   
   A. communication, language;
   B. physical sensory development;
   C. psychological sensory development; and
   D. warning.

2. Parents will define:
   
   A. frequency: pitch of a sound;
   B. Hertz (Hz): frequency or pitch; and
   C. decibel (dB): loudness or intensity of a sound.

MATERIALS

- Figure H-1, "Amounts Of Hearing Loss"
- rubber band
- pencil
- pitch pipe, piano, or other musical instrument (can use voice to show different pitches)

INSTRUCTIONS TO PARENT ADVISOR

Discuss with parents the need for them to understand why sound is important. This information and the understanding of several professional terms will make it easier for them to deal with their child's hearing aids as well as other professionals they will come in contact with.

SAMPLE DISCUSSION

Importance Of Sound

Suppose you were very young like your child and did not know any words and suppose there was no sound to hear these words. Let's pretend there is no sound. I will not use my voice. See how much you can understand. [Drop voice and say a few words or sentences in a foreign language or use nonsense words.]

What did you understand? [Response: Nothing.] You see, when a child does not know a language (the words), it is very hard to understand what is being said, but it is even more difficult if there is no sound. So sound is important to help your child learn to communicate. We all learn language by hearing it during critical periods of readiness. It appears that the development of speech and language is based upon innate biologically programmed factors (Lenneberg, 1967). Thus, the earlier the child's hearing is helped by amplification, the better for the child's speech and language development.
Sound is important to learn to communicate, but it is also very important for other reasons:

1. Sound stimulates the development of your child’s sense of hearing. Without sound, sensory deprivation (in this case, sound deprivation) results. The hearing system may deteriorate by prolonged lack of sound (sound deprivation) in early life.

2. The absence of sound can also affect psychological aspects of development. Sound allows your infant contact with the world so he can begin to trust the people and things in his environment. Sounds make your child feel part of the world. If the hearing aids only help your child come into contact with his world, it will be enough help. The expense and effort of putting on the hearing aids will be worth it.

3. Sound provides warning and helps your child to know how to act. If something falls behind him, he will know where to look. If a car honks, he can get out of the way. If mother shouts, he can come to her (or go the other way!). The hearing aid will help bring sound to your child.

What Is Sound?

Sound is something vibrating; i.e., something moving back and forth. As it moves forward, it pushes on air molecules and the molecules squeeze together. As it moves back, the molecules spread out.

When a plucked rubber band (or string on a musical instrument) moves back and forth one time, [demonstrate by moving finger back and forth], that is one cycle. If it moves back and forth ten times it would be ten cycles. If it moves back and forth 250 times in one second [try to demonstrate], that would be 250 cycles per second (refer to Figure H-1 and show 250 on audiogram). This would be middle C [play on piano or use pitch pipe or other instrument]. Another name for cycles per second is Hertz (Hz). If the source vibrates 250 times in one second that would be 250 Hz (middle C). How fast the sound source vibrates determines how high or low the sound will be. If it moves very slowly, 125 times in one second, a low sound (octave below middle C) would be heard. If it moves very rapidly (500 times in one second), a higher sound (one octave above middle C) would be heard. How high or low a sound is, is called pitch or frequency. A particular pitch or frequency is called a pure tone. A sound can be high or low and it can also be loud or soft.

Decibel (dB) is the term utilized for how loud a sound is. If a sound is very soft, it would be about 10 dB. Normal voice would be about 60 dB. If a truck passed by, that would be about 110 dB. Decibel is simply a measurement of loudness (like an inch is a measurement of length and a pound is a measurement of weight). This example audiogram [point to Figure H-1] shows what dB range is usually referred to as normal hearing, mild hearing loss, etc. Different audiologists may divide up the amount of hearing loss slightly differently; however, this is usually how it is done.

SAMPLE ACTIVITIES

none

SAMPLE CHALLENGES

none

REFERENCE AND READING LIST

Figure H-1

Amounts of Hearing Loss

Hertz (Hz)
(Cycles per second, pitch, frequency)

AUDIGRAM

Decibels (dB) (loudness or intensity)

Hearing Threshold Level in Decibels Re: 1964 Standard

0-20 dB = Range of Normal Hearing
21-40 dB = Mild Hearing Loss
41-55 dB = Moderate Hearing Loss
56-70 dB = Moderately Severe Loss
71-90 dB = Severe Hearing Loss
91+ dB = Profound Hearing Loss

BEST COPY AVAILABLE
TOPIC 2: PERCEPTION OF SPEECH; MINIMIZING BACKGROUND NOISE

PARENT GOALS

1. Parents will be aware how the perception of speech is affected by:
   A. the pitch of the speaker's voice;
   B. the loudness of the speaker's voice;
   C. the distance of the speaker's voice from the listener; and
   D. the presence of background noise.

2. Parents will establish four conditions for the best home listening environment:
   A. use a normal, conversational tone of voice;
   B. speak normally loud;
   C. be as close to the child as possible; and
   D. minimize background noise while speaking to the child.

MATERIALS

- Figure H-2, "Comparison Of The Frequency And Intensity Of Various Environmental And Speech Sounds"
- "A Sound Approach" (slide/tape presentation available through HOPE Inc., 1780 North Research Park Way, Suite 110, Logan, UT 84321)
- tape recorder
- slide projector

INSTRUCTIONS TO PARENT ADVISOR

For parents of less severely handicapped children with mild-moderate hearing losses, a discussion is available in the Middle Ear Program (SKI*HI Outreach Monograph Series) on the perception of speech relative to word meanings, functional relations of words (grammar), and prosody. This monograph can be obtained from HOPE Inc., 1780 North Research Park Way, Suite 110, Logan, UT 84321.

If available, show the slide/tape presentation, "A Sound Approach," and discuss its implications.

SAMPLE DISCUSSION

The perception of speech is the ability to be aware of, or notice, speech. People with normal hearing can hear the frequency range between 20 and 20,000 cycles per second (Hz) [point out range from far below to way above the 125 to 8,000 Hz range shown on Figure H-2]. It is known that normal hearing infants can hear the differences among many speech sounds almost from birth.

Infants are cued to their parents' speech by its distinctive pitch (the "musical" sound of speech). Men's vocal pitches tend to be in lower frequency regions (100-150 Hz) than women's (200-225 Hz). A child's speech is even higher in pitch than that of a woman's. The normal way you vary your pitch patterns while talking to your child helps him to perceive (be aware of) your voice.
The loudness of your speech varies very quickly within the range of about 30-60 dB [point out 'on Figure H-2]. For example, if you say the sound [sh], it may be approximately 20 dB louder than if you said the sound [th] as in the word "thing." Therefore, when you speak softly or when you are more than about two feet away from your child, he may not be able to hear the softer sounds ([l], [th], [s], [p]) of your speech.

Also, your child's listening environment often includes noise from television, other people talking, fans, air conditioners, appliances, etc. [show and discuss Figure H-2]. Often this background noise, which is almost as loud or even louder than your speech, masks out a significant amount of what you are saying to your child.

Distracting and competing noises make it difficult to hear conversations clearly. All surrounding noises, as well as speech sounds, are equally amplified with a hearing aid; thus, important communication and speech signals may be lost in background noise. When it is impossible to eliminate background noises completely, speaking close to the child makes the speech signal stronger and clearer.

Thus, when you are talking to your child, you should try to do the following:

1. Use a normal conversational tone of voice (pitch patterns).
2. Be sure to speak normally loud (not too softly).
3. Be as close as possible to your child when you are speaking to him, the closer the better.
4. Keep background noises at a minimum when communicating with your child.

SAMPLE ACTIVITIES

none

SAMPLE CHALLENGES

1. Make a list of background noises occurring during the home visit, during a normal day, in the car, at the store, etc.
2. Work to reduce distracting noises; discuss examples at next home visit.
3. Listen to competing background noises with your child's hearing aid on to determine the distraction effects of such noises.
4. Blindfold yourself and listen to distracting sounds. Discuss how to eliminate them.

REFERENCE AND READING LIST

Figure H-2

Comparison of the Frequency and Intensity of Various Environmental and Speech Sounds

Frequencies in Cycles Per Second (Hertz)

Hearing Loss in Decibels

Softest Speech

Average Speech

Loudest Speech
TOPIC 3: OTOLOGICAL CARE; ANATOMY OF THE EAR; CAUSES AND TYPES OF HEARING LOSSES

PARENT GOALS

1. Parents will be aware of the various medical personnel available to their child and be able to state the need for continued medical care.
   A. Otolaryngologists or otologists give medical clearance for wearable amplification.
   B. Continued, periodic medical follow-up is necessary:
      1. to detect and treat middle ear infections;
      2. to detect progressive hearing loss; and
      3. to evaluate a balance problem.

2. Parents will be aware of the five parts of the hearing system and what is in each part.
   A. Outer Ear
      1. Auricle/pinna
      2. Ear canal
   B. Middle Ear
      1. Eardrum
      2. Bones: malleus, incus and stapes
      3. Oval window
      4. Eustachian tube
   C. Inner Ear
      1. Semicircular canals
      2. Cochlea
      3. Auditory nerve
   D. Brainstem
      1. Auditory nerve
   E. Brain
      1. Auditory cortex

3. Parents will be aware how sound travels from the sound source, through the ear and to the brain.
   A. Sound pushes eardrum.
   B. Eardrum pushes three bones.
   C. Last bone pushes on oval window.
   D. Oval window moves fluid in cochlea.
E. Moving fluid in cochlea stimulates the nerves in patterns.
F. Patterns travel up the brainstem.

4. Parents will be aware what can go wrong in each of the five parts of the hearing system and state what has gone wrong with their child's hearing system.

A. Outer Ear
   1. Wax plug in canal
   2. Canal and/or pinna not formed correctly or missing

B. Middle Ear
   1. Three bones broken
   2. Middle ear infections
   3. Hole (perforation) in eardrum

C. Inner Ear
   1. Nerve cells in cochlea damaged or missing
   2. Possible damage to balance mechanism

D. Brainstem
   1. Auditory nerves in brainstem damaged or failed to develop
   2. Auditory nerve tumors

E. Brain
   1. Brain cells damaged or missing
   2. Brain failed to develop

F. Area(s) damaged for this particular child

5. Parents will explain cause of their child's loss, if known.

MATERIALS

- Figure H-3, "Professionals Involved With The Hearing Impaired"
- Figure H-4, "Anatomy Of The Hearing Mechanism"
- Figure H-5, "The Cochlea"
- Figure H-6, "Anatomy Of The Brain"
- "Anatomy Of Hearing Loss" (slide/tape show available from HOPE Inc., 1780 North Research Park Way, Suite 110, Logan, UT 84321; use as review after the discussion)
- tape recorder
- slide projector
INSTRUCTIONS TO PARENT ADVISOR

Remember to utilize only the goal(s) from this lesson that the family has requested or that you select based on your awareness of the child's specific cause and type of hearing loss.

SAMPLE DISCUSSION

Medical Personnel Available And Periodic Follow-Up

The first step in obtaining hearing aids is medical clearance. [Show the parents Figure H-3, "Professionals Involved With The Hearing Impaired" and leave a copy with them.] This clearance can be provided by an otologist (ear specialist) or an otolaryngologist (ear, nose, and throat specialist) or a family doctor if a specialist is not available. Medical clearance is needed for hearing aid use because hearing aids may not be recommended if the hearing loss can either be medically or surgically improved in the immediate future.

Equally as important as initial medical clearance is the need for continued medical follow-up, especially of infants and young children who have frequent colds, allergies, influenza, other viruses, and bacterial infections of the throat that can cause associated middle ear problems. Many infants do not let their parents know when they have a middle ear infection; for example, by rubbing or tugging at their ears. Therefore, periodic checking is essential to determine if a middle ear infection is present when an upper respiratory infection (cold) is present. Even children with permanent hearing loss have middle ear infections which can temporarily cause a significantly greater loss of hearing.

Also, in a few infants, sensorineural hearing loss is progressive and this needs to be checked by periodic audiological testing. It is essential for hearing impaired infants and young children who experience dizziness or other balance problems to receive otologic care as soon as possible.

Anatomy And What Can Go Wrong
With The Five Parts Of The Hearing System

There are five parts of the hearing system [show parents Figure H-4]: (1) outer ear; (2) middle ear; (3) inner ear; (4) brainstem; and (5) brain.

The first area, the outer ear, has the ear flap (auricle or pinna) and the ear canal. Wax in the ear canal can build up and become compacted behind an earmold. Every child should be checked regularly for ear wax because it can cause temporary hearing loss and increase feedback (sound coming back out the ear) during hearing aid use.

The second area, the middle ear, contains the eardrum. The eardrum is connected to three tiny bones (malleus, incus, and stapes). The last bone (the stapes) is connected to the oval window. The eustachian tube is a small tube which goes from the top of the throat to the middle ear. It has two purposes. The first function of the eustachian tube is to let air into the middle ear and thus make the air pressure in the middle ear the same as the pressure outside the ear. For example, while driving up a mountain or flying in a plane, the ear may begin to feel plugged. The air outside is very "thin" but the air inside the ear is not. This heavy air inside the ear pushes out on the eardrums and causes a "plugged" feeling. Yawning will cause the "thin" air from the outside to rush into the mouth, up the eustachian tube and into the middle ear. This feels like a "pop." Now the air pressure is the same on the inside and outside of the middle ear or eardrum and the plugged feeling is gone. Letting air into the middle ear is also very important because having air in the middle ear helps to keep it from collecting too much fluid.

The second purpose of the eustachian tube is to drain fluid from the middle ear space. If there is fluid in the middle ear, it will run out of the ear and down the eustachian tube to the throat. Again, since young children have more colds, infections, and allergies, it is important to make sure there is no infection in the middle ear.
The third part of the hearing system is the inner ear. It has in it: (1) the semicircular canals which help with balance; (2) the cochlea which looks like a snail; and (3) the auditory nerve which goes to the fourth area, the brainstem. The brainstem carries sound to the fifth area, the brain.

**How Sound Travels From The Sound Source Through The Ear And To The Brain**

Something must vibrate for sound to occur. When something vibrates, air molecules are pushed together. The first molecule pushes the second molecule which pushes the third and so on until the molecules push in on the eardrum. The eardrum vibrates or moves back and forth at the same speed (or cycles per second) as the source. When the eardrum moves back and forth, it moves the three little bones, the malleus, incus, and stapes, back and forth [refer to Figure H-4]. The cochlea is filled with a fluid and has thousands of tiny nerve cells in it. As the last tiny bone (stapes) moves back and forth, it pushes on the oval window, which pushes the fluid in the cochlea back and forth [refer to Figure H-5]. As the fluid moves back and forth, the nerves are activated (stimulated) in a certain pattern. The nerves join together into one large nerve in the brainstem that carries the pattern to the brain.

As this pattern is traveling through the brainstem to the brain, it passes through several centers, apparently designed to perform a particular function. Some reflexes (for example, startle responses to loud sounds) and hypersensitive responses to sound are established at the brainstem level. The brainstem likely has other functions that at present are unknown or poorly understood.

The area of the brain where sound is analyzed is called the auditory cortex [show Figure H-6]. Although a great deal about how the brain functions is understood, scientists are still uncertain about the specific functions of the brain that underlie many of the problems observed in children with hearing loss.

**What Can Go Wrong In Each Of The Five Parts Of The Hearing System?**

When someone is hearing impaired, there is a problem with sound going through the ear to the brain. If there is a problem in the outer ear (wax block in the canal or the pinna and/or the canal fails to develop), sound cannot travel successfully through it.

Sound cannot travel through the middle ear if there are problems such as:

1. one or more of the three tiny bones are broken;
2. there is fluid or infection in the middle ear;
3. there is a hole (perforation) in the eardrum;
4. the middle ear is congenitally malformed (Pert's syndrome, Treacher Collins, Pierre Robin, osteogenesis imperfecta, etc.); associated disorders are often present.

If there is a problem in the outer or middle ear, an otolaryngologist or otologist can usually fix it (surgery).

In the inner ear (cochlea) when the nerve cells are damaged, the child will have a permanent hearing loss. Some of the things that can cause damage to the nerve cells are:

1. Spinal meningitis: Bacterial, viral, or fungal infection or inflammation of the meningeal membrane surrounding the brain and spinal cord. Although a relatively uncommon occurrence, spinal meningitis may set in as a complication of untreated otitis media (Connelly, 1988).
2. Drugs: There are several drugs that adversely affect hearing. Parents need to ask their ear specialist which ones should be avoided in case their pediatrician or another doctor inadvertently prescribes one of these drugs.
3. Extremely loud noises: Sounds of sufficient intensity and duration can cause injury to the inner ear, producing a temporary or permanent hearing loss. Firecrackers, model airplanes tested indoors, toy firearms (caps), farm machinery, and extremely loud music are some of the sounds capable of producing injury. Consequently, it is important for the child to avoid exposure to these sounds.

4. Viral infections: Hearing loss may result from the mother having measles (rubella) during her pregnancy and/or other viral infections (such as cytomegalovirus) which can damage the baby during pregnancy or during the birth process.

5. Heredity: In most cases of hereditary deafness, the cochlea is damaged.

6. Rh factor: Due to blood incompatibility of mother and infant, damage in the cochlea may ensue. Often the brainstem is also damaged.

7. Syndromes (childhood deafness associated with defects): Various syndromes are associated with malformations of the inner ear (Waardenburg, Klippel-Feil, etc.). Some syndromes are characterized by both malformed low-set ears and damage to the middle ear and/or inner ear (CHARGE, Pierre Robin, etc.). Many syndromes include eye and kidney disorders. Progressive hearing loss is also possible.

The causes of about 40% of inner ear hearing losses are not known. Basically, problems in the inner ear cannot be corrected with surgery. A great deal of research is going on in the area of cochlear implants (sending electrical stimulation directly to the auditory nerve). Several implantation schemes are under investigation (inside the cochlea, at the oval window, etc.). Investigators have reported that electrical stimulation does produce auditory sensations and discrimination of loudness changes within normal limits. However, discrimination among frequencies (pitch) is poor. Word recognition skills of implanted patients (mostly adults) vary. There seems to be agreement that the use of implanted devices helps adult patients, who learned language before they lost their hearing, to control their own speech output and to engage in speech reading.

At the present time, cochlear implants for children are classified by the FDA as investigational devices. A lower age limit of two years is generally appropriate for being considered a candidate for a cochlear implant. There are not studies that adequately separate the effect of the implant from improvement due to maturation and training. The cochlear implant is an important step in the long-range goal of understanding, preventing and treating hearing impairment. The foremost limitation is that implantation does not restore normal hearing. It is likely that the future will bring significant improvements in this field of research.

Whatever causes damage to the cochlea or nerves to the brain may also cause damage to the balance mechanism, the semicircular canals. When this occurs, gross motor skills will be delayed and the infant may try to use sight and touch to compensate for damage to the balance mechanism.

Many multisensory impaired children have damage in the brainstem resulting in a problem with the way sound information is sent to the brain (auditory cortex). This could happen because of the following:

1. anoxia (lack of oxygen at birth);
2. maternal rubella;
3. birth trauma;
4. prematurity;
5. drugs;
6. Rh problem (incompatibility); and
7. encephalopathic diseases (brain diseases).
These same conditions can also damage the auditory cortex itself (refer to Figure H-6). When the auditory cortex is damaged, the child may have difficulty analyzing sounds, inhibiting inappropriate motor responses to sound, understanding sounds, remembering sounds, forming an auditory "space" (finding where a sound comes from and reasoning its location), and choosing which sound to listen to.

Some multisensory impaired children may also have suffered damage to other parts of the brain by the same mechanism that caused hearing loss. If the frontal lobe is injured, a child may have expressive aphasia which is the inability to produce language (understanding of language may not be affected). If the parietal area of the brain is damaged, a child may experience difficulty thinking about his experiences and thus be unable to make use of language. These children sometimes are diagnosed autistic. Whenever the temporal lobe of the brain is damaged, a child will be unable to attach meaning to sound and/or attend to sound.

It is often very difficult to diagnose specific brain disorders. The important issue at this point is not to waste time trying to label but to take advantage of these early years and begin stimulating the hearing system.

Cause Of The Child's Loss, If Known

Here is the location of your child's hearing problem (point to the outer, middle, inner ear, brainstem, and/or brain on Figure H-4). The cause of your child's hearing loss is (discuss the cause of the child's loss if known, such as damaged nerves, fluid in the middle ear, brain damage, or combinations of these).

SAMPLE ACTIVITIES

none

SAMPLE CHALLENGES

1. Explain the location and cause of your child's hearing loss to your spouse (if not in attendance) or other family member.

REFERENCE AND READING LIST


**FIGURE H-3**

PROFESSIONALS INVOLVED WITH THE HEARING IMPAIRED

<table>
<thead>
<tr>
<th>1. Otolaryngologist (ENT) or Otologist:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A physician (M.D., D.O.) knowledgeable in diseases of the ear, whose goal is to establish the medical characteristics of an individual’s hearing loss and offer appropriate treatment recommendations. FTC (Federal Trade Commission) regulations require physician approval prior to the purchase of a hearing aid; either the otolaryngologist or otologist is the best qualified physician to do this. The intent of the medical examination is to assure that your child’s best medical interests are protected prior to the purchase of a hearing aid. An otologist is a trained otolaryngologist (ear, nose and throat specialist) who has specialized in the ear.</td>
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<tr>
<th>2. Audiologist:</th>
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</thead>
<tbody>
<tr>
<td>A professionally trained individual with a masters (M.A., M.S.) or doctorate (Ph.D., Ed.D.) degree in audiology. The audiologist has the basic responsibilities to assess hearing, determine the person’s ability to use hearing, and to increase the ability of the hearing handicapped individual to cope with the situations of everyday life (through hearing aids, assistive listening devices, etc.).</td>
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<tr>
<th>3. Dispensing Audiologist:</th>
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</thead>
<tbody>
<tr>
<td>An audiologist who, in addition to selecting a hearing aid and providing subsequent follow-up care, orders the hearing aid and sells it to the patient.</td>
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</table>

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<thead>
<tr>
<th>4. Hearing Aid Dispenser:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A person with no special training who obtains hearing aids directly from the manufacturer and sells them to patients upon receipt of a prescription from a physician or audiologist. This person does not have direct contact (except written) with the patient. Many audiologists have begun offering this service to patients. Some physicians offer this as well.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>5. Hearing Aid Dealer:</th>
</tr>
</thead>
<tbody>
<tr>
<td>A hearing aid salesperson who provides a retail outlet for hearing aids. A dealer is not an audiologist and is not trained to provide audiological services.</td>
</tr>
</tbody>
</table>
ANATOMY OF THE HEARING SYSTEM

EXTERNAL EAR
- Ear Flap or Pinna
- Ear Canal
- Eardrum

MIDDLE EAR
- Middle Ear Bones
- Semicircular Canals
- Eustachian Tube

INNER EAR
- Cochlea
- Auditory Nerve

INNER EAR
- Auditory Nerve
- Auditory Cortex
- Brainstem
- Brain

Figure H-4
Figure H-5

The Cochlea

ANATOMY OF THE COCHLEA

ILLUSTRATION OF INTERIOR OF THE COCHLEA SHOWING THE NERVE CELLS
TOPIC 4: MEASURING HEARING LOSS

PARENT GOAL

Parents will explain their child's hearing loss in terms of:

A. amount (mild, moderate, etc., or in dB);
B. configuration or shape (sloping, flat, rising); and
C. discomfort or tolerance level (does/does not have a tolerance problem).

MATERIALS

* Figure H-1 (in Topic 1)
* Figure H-7, "Sample Audiogram" (parent advisor may want to copy so she can write on it)
* Figure H-8, "Two Illustrations Of Hearing Loss With A Flat Audiogram"
* Figure H-9, "Two Illustrations Of Hearing Loss With A Sloping Audiogram"
* Figure H-10, "Impedance Testing/What Do Different Tympanograms Mean?"
* Figure H-11, "Auditory Brainstem Responses"
* Figure H-12, "Air Conduction Testing"
* Figure H-13, "Bone Conduction Testing"
* Figure H-14, "Illustration Of Conductive Hearing Loss"
* Figure H-15, "Illustration Of Sensorineural Hearing Loss"
* audiometer (if possible)
* child's own audiogram and other audiological test results
* patterns for vests, carrier pockets, and/or toupee tape

INSTRUCTIONS TO PARENT ADVISOR

If appropriate, describe how the audiologist determines if the child has a conductive loss (problem in the outer or middle ear) versus a nerve loss (problem in the inner ear). For most parents, the explanation will be difficult. Parent advisors may want to give it to interested parents only. A sample discussion is given in Supplemental Information For Parents at the end of this discussion.

If the child will be fitted with hearing aids on a trial or diagnostic basis, utilize the sample discussion also given in Supplemental Information For Parents. The guidelines for the introduction of hearing aids (Topic 7) and the auditory stimulation activities in the Auditory Development Section (pages 393 to 397) should be utilized prior to any decision being made that wearable amplification is not appropriate.

Utilize the child's own audiological evaluation, the sample discussion below, and Figures H-7 through H-14 to help the parents meet the goals for this topic. It is a good idea to review this information with the parents each time the child returns for an audiological evaluation. Remember to select and present only the portions of this topic which are applicable to the particular child and family.

SAMPLE DISCUSSION

Hearing Testing

You will remember when your child had his hearing tested, the audiologist wrote down what your child could hear on a paper that looked like this. This is called an audiogram [show sample audiogram in Figure H-7]. Let's talk about how the audiologist tests hearing and what an audiogram is. When your child has his hearing tested, the audiologist first wants to know what kind of response your child makes to
sound and second how loud a sound must be for him to hear it. First, the audiologist will observe your child's general level of awareness to the environment. The audiologist will be on the lookout for responses to touch (tactile), to light (visual), and to sound (auditory). While the audiologist is observing, she will want you to describe the changes in your child's behavior, observed by you at home, that seem to signal responses to sound; for example:

1. startle responses;
2. eye blink;
3. change in ongoing behavior (either increase or decrease of activity);
4. tensing of entire body;
5. orienting or searching for sound;
6. smiling;
7. head turn;
8. vocalization response (grunt, change in sucking, babbling);
9. negative or withdrawal response; and
10. inconsistent response.

The audiologist will also want you to describe the circumstances under which responses to sound are noted (for example, only when your child is in his support chair or only when it is completely quiet in the room), what specific sounds your child responds to (noises, voices, etc.), and how consistently he responds to these sounds (e.g. once only, over and over, only when not watching something).

Second, the audiologist will use what is called behavioral observation audiometry in the sound field room (sound comes through speakers) and under earphones to determine how loud sounds need to be for your child to hear them. The audiologist will utilize a variety of sounds (e.g. speech, environmental sounds, music, and sound toys). These sounds are recorded on a tape and then played through an audiometer where the loudness (intensity) can be regulated. The audiologist may also use tape recorded speech calibrated or filtered at a certain pitch.

The audiologist will pick one of these sounds with one frequency (pitch) like 1000 Hz which is here on the audiogram [show Figure H-7]; lower pitches are here, and higher pitches are here. She will send the 1000 Hz sound to your child very softly. This side of the audiogram tells us how loud the sounds are. Very soft sounds are here, very loud sounds are here [point out]. So the audiologist gives your child a 1000 Hz sound very softly (maybe 10-15 dB). Your child cannot hear the sound so he does nothing. Then the audiologist makes the sound louder and louder. When your child hears the sound, he will blink or turn his head or decrease his activity or in some way let the audiologist know he hears the sound.

If your child hears the sound when it is 80 dB loud, the audiologist will make a mark at 80 dB [point out]. If he hears the sound at 95 dB, the audiologist will make a mark at 95 dB [point out]. When the audiologist gives your child a sound in only one of his ears, she will send the sound into just one earphone. If your child hears the 1000 Hz sound in his right ear at 80 dB, the audiologist will make a circle like this at 80 dB [demonstrate]. If your child hears this sound in his left ear at 90 dB, the audiologist will make an "X" like this at 90 dB [demonstrate]. The audiologist will then select another pitch, maybe an octave above 1000 Hz or 2000 Hz [point out], and make that sound loud enough for your child to hear it. When the speakers are being utilized, the audiologist will mark an "S" [demonstrate] and you will know that at least one ear is hearing the sound at this level. The only way to find out about each ear is to use earphones.

It is not likely that a complete test (responses at every frequency) will be obtained during one evaluation period. Usually several visits to the audiologist are required. Initially, a multihandicapped sensory impaired infant may have only primitive or reflexive responses such as startle or eye-blink. As your child develops, it may be possible to "condition" a response to sound. This means he can be taught to look at an animated toy or blinking light each time he hears a sound. He is rewarded for looking each time he hears by getting to see the toy or light. Thus, he continues to look each time he hears. This procedure is appropriate for
most visually impaired (not totally blind) children. When a conditioned response to sound can be established, pure tones (sounds with only one frequency or pitch) can be used to obtain a complete audiogram. Once responses to all the pitches important for hearing speech can be obtained, the audiologist will have a picture that looks like this [complete a sample audiogram]. This audiogram is now complete. If a child does not hear the sound even when it is presented at the limit of the audiometer (which is 110 dB on some audiometers and 120 dB on others), the audiologist may not make a mark or may use the audiometric symbols $\rho$ and $\chi$, indicating no response.

Some people have the same amount of hearing at each pitch. Their audiogram may look like this [show Figure H-8]. Some people have different amounts of hearing at each pitch. Their audiogram may look like this [show Figure H-9].

**Additional Audiological Tests**

The audiologist may also use additional tests such as impedance audiometry ( tympanometry) to help determine what kind of loss (conductive or sensorineural) your child has. The impedance testing may also help determine:

1. existing middle ear pressure (air pressure in middle ear versus outer ear);
2. eardrum mobility or movement (can eardrum still move when sound strikes it?);
3. eustachian tube function (is tube open so air can get into the middle ear?);
4. continuity and mobility of the middle ear bones (are bones properly connected to each other and moving when sound strikes them?); and
5. acoustic reflex thresholds (does middle ear muscle contract when a loud sound is presented?).

Tympanometry is the technique for measuring the movement (mobility) of the eardrum which in turn gives information about almost any problem in the eardrum or middle ear. For example, if there is negative pressure in the middle ear (less air in the middle ear than the outer ear), this may cause the eardrum to be pulled into the middle ear cavity (retracted) and cause a mild conductive type hearing loss. This information can be detected by tympanometry (impedance testing) even though there may be no observable fluid in the child’s middle ear [use Figure H-10 to discuss child’s particular impedance results].

The acoustic reflex threshold is the loudness level (dB) at which the stapedial muscle contracts. The stapedial muscle is the little muscle in the middle ear going from the stapes bone out to the wall of the middle ear. It is known at what level the muscle contracts in normal hearing ears. The level at which reflexes are present, or their absence, gives the audiologist additional information about the kind (cochlea versus middle ear) and amount of hearing loss. This type of testing is very valuable for infants and young children who cannot cooperate fully in other diagnostic tests.

Remember, the pure tone test results may miss a middle ear problem when severe and profound sensorineural hearing losses exist. Therefore, it is important to “watch” for these with tympanometry and medical checkups of the ears.

In addition to these test methods, many multisensory impaired infants are evaluated with brainstem evoked response (BSER) audiometry, also called auditory brainstem response (ABR) audiometry. This procedure is helpful with many infants, particularly those who do not appear to respond to standard procedures. BSER is a recording of brain activity (electrical) for sound stimulation recorded via electrodes from the top of the head (vertex) to either mastoid (or earlobe) [show and discuss Figure H-11]. It is most easily evoked (produced, observed) with clicks repeatedly presented and then summed (or averaged) by
computer analysis. These sound clicks are not frequency (pitch) specific and thus the evoked potential (electrical response) is not as specific as an audiogram. The latency of the potential (how long before it occurs after the click) corresponds to specific causes of hearing loss (middle ear versus cochlea versus auditory nerve problems). Despite the fact that children up through the age of three or four must be sedated to perform the test, it does give a reliable indication of the amount of hearing loss in each ear (up to the limits of the equipment) for frequencies above approximately 1500 Hz.

The Child's Own Audiogram

Let's look at your child's audiogram and talk about what he hears. At this very low pitch, your child hears at ___ dB. At this higher pitch, your child hears at ___ dB.

Of course, different people have different amounts of hearing. A person who has normal hearing can hear sounds that are 0-20 dB loud [refer to Figure H-1 in Topic 1, page 307]. A person who has a very small (mild) hearing loss can hear sounds when they are about 21-40 dB loud. If the sounds must be 41-70 dB, the person has a moderate loss. If the sounds must be 71-90 dB before they can be heard, that person has a severe hearing loss. A person who has a profound loss hears sounds only when they are 91 dB or louder. Your child's hearing loss is in this area [show on child's audiogram].

Clarity And Tolerance Testing

In addition to how loud sounds have to be for your child to hear, the audiologist is also interested in how clearly your child hears. For infants, judging how clearly one hears has to be accomplished by long-term observation of responses to sound. The most basic level is whether or not a child can perceive the difference between the presence or the absence of sound. Beyond this very basic skill, the audiologist looks for the ability to: (a) grossly discriminate between two very different sounds, (b) use sound in some systematic way, and (c) produce consistent responses to consistent sounds.

The last area the audiologist looks at is tolerance, or in other words, how loud the sound can be before your child gives signs of discomfort. It is important to get this information for each ear separately. If the audiologist cannot obtain a level, she will assume one within the range of safety for your child. If observations in the home indicate your child is not tolerating this level (startle responses to many sounds), your child may need a special adjustment in his hearing aid. The audiologist should be contacted and the aid adjusted or replaced as soon as possible.

SAMPLE ACTIVITIES

none

SAMPLE CHALLENGES

none

SUPPLEMENTAL INFORMATION FOR PARENTS

Conductive Loss Versus Nerve Loss

Pure tone testing is usually accomplished using two different methods of getting sound to the ear. Pure tone air conduction testing uses earphones. In air conduction testing, sound must travel through the entire auditory system as shown in Figure H-12. In general, if damage to the auditory system exists anywhere, the result will be some degree of hearing loss.
Pure tone bone conduction testing uses a bone vibrator which usually fits behind the ear. Vibration from the bone vibrator bypasses the external ear and middle ear and is transferred directly to the cochlea as shown in Figure H-13. Thus, if the inner ear can hear better than the entire auditory system (due to middle ear infection, external ear malformation, etc.), the bone conduction responses will be better than the air conduction responses. For example, if a 40 dB hearing loss exists as revealed by the air conduction responses shown in Figure H-14, but the bone conduction responses are normal (0-15 dB), the problem must necessarily lie in the external and/or middle ear. This type of hearing loss is referred to as a conductive hearing loss. In this example, the inner ear (cochlea) is functioning normally. On the other hand, if the pure tone air conduction and bone conduction responses are equivalent as shown in Figure H-15, the problem must necessarily lie in the cochlea. Such a hearing loss is referred to as a sensorineural loss. [Note: In case 2 on Figure H-15, bone conduction thresholds remain at 65 dB whereas the air conduction thresholds continue to drop since 65 dB (approximately) is the limit for bone conduction testing.] It is, of course, possible for a person to have hearing loss due to both conductive and sensorineural factors in the same ear or to have a conductive loss in one ear and a sensorineural loss in the other ear.

**Diagnostic Trial Fitting of Hearing Aids**

The purpose of a diagnostic trial hearing aid fitting is to observe the reaction of your child to amplified sound. This can help the audiologist determine whether or not your child has a significant hearing loss that warrants hearing aid use. Hearing aids that are set very softly are used initially. We will carefully observe your infant's responses, as will the audiologist, to his hearing aids. If your child's responses to sound improve with amplification, that is a good indication the aids are helping.

**REFERENCE AND READING LIST**


325
Sample Audiogram

KEY

O = RIGHT EAR
X = LEFT EAR
[] = BONE CONDUCTION
S = SPEAKER
A = AIDED

Figure H-7
Two Illustrations of Hearing Loss with a Flat Audiogram
Two Illustrations of Hearing Loss
with a Sloping Audiogram
**Impedance Testing**

What Do Different Tympanograms Mean?

<table>
<thead>
<tr>
<th>TYMPANOGRAM</th>
<th>PRESSURE</th>
<th>MOBILITY</th>
<th>MEANING</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image" alt="Normal" /></td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td><img src="image" alt="Normal" /></td>
<td>Normal</td>
<td>Low</td>
<td>Draining from the middle ear and/or thickened ear drum and/or immobile ossicle bones and/or fluid in the middle ear</td>
</tr>
<tr>
<td><img src="image" alt="Normal" /></td>
<td>Normal</td>
<td>High</td>
<td>Floppy ear drum and/or broken ossicle bones</td>
</tr>
<tr>
<td><img src="image" alt="Negative" /></td>
<td>Normal</td>
<td>Normal</td>
<td>High negative pressure (ear drum pulled into middle ear) with or without draining from the middle ear</td>
</tr>
<tr>
<td><img src="image" alt="Negative" /></td>
<td>Low</td>
<td>Low</td>
<td>Draining from the middle ear and/or thickened ear drum and/or immobile ossicle bones and/or fluid in the middle ear</td>
</tr>
<tr>
<td><img src="image" alt="Negative" /></td>
<td>High</td>
<td>High</td>
<td>Floppy ear drum and high negative pressure (ear drum pulled into middle ear) or broken ossicles and high negative pressure</td>
</tr>
<tr>
<td><img src="image" alt="Positive" /></td>
<td>Normal</td>
<td>Normal</td>
<td>High positive pressure (ear drum pushed out from middle ear) with or without draining from the middle ear</td>
</tr>
</tbody>
</table>
Auditory Brainstem Responses (A.B.R.)

A.B.R. Testing

Normal A.B.R. Recording

65 dB S.L.

A.B.R. Recording Showing No Response

65 dB S.L.

Figure H-11
Figure H-12

Air Conduction Testing

AIR CONDUCTION PURE TONE TESTING
— ACCOMPLISHED WITH EARPHONES —
Figure H-13

Bone Conduction Testing
Figure H-14

Illustration of Conductive Hearing Loss

\[ X = \text{LEFT EAR} \\
O = \text{RIGHT EAR} \\
[] = \text{Bone Conduction} \]
Illustration of Sensorineural Hearing Loss

Figure H-15

Hearing Threshold Level in Decibels Re: ISO 1964 Standard

Case 1:  
Case 2:
TYPES AND DEGREES OF HEARING LOSS
Types and Degrees of Hearing Loss

There are three types of hearing loss: Conductive, Sensorineural, and Cortical/Perceptual. For conductive and sensorineural hearing loss, there are four degrees (mild, moderate, severe, and profound). There are no degrees for cortical/perceptual hearing loss.

<table>
<thead>
<tr>
<th>Types of Hearing Loss</th>
<th>Degrees of Hearing Loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>Conductive</td>
<td>Mild</td>
</tr>
<tr>
<td></td>
<td>Moderate</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
</tr>
<tr>
<td></td>
<td>Profound</td>
</tr>
<tr>
<td>Sensorineural</td>
<td>Mild</td>
</tr>
<tr>
<td></td>
<td>Moderate</td>
</tr>
<tr>
<td></td>
<td>Severe</td>
</tr>
<tr>
<td></td>
<td>Profound</td>
</tr>
<tr>
<td>Cortical/Perceptual</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Conductive Hearing Loss:

This hearing impairment is due to a disruption of the transmission of sound through the outer and middle ear.

What are some of the causes of this type of hearing loss?

This type of hearing loss can be caused by:
- blockage of the ear canal (by wax, some foreign object, etc.);
- blockage of the eustachian tube opening (by infection, allergy, etc);
- damage to the eardrum (by infection, blow to the heard, puncture by foreign object, etc.);
- damage to the three tiny bones in the middle ear (infection, etc.);
- congenital malformation of the middle ear (Pert's syndrome, Treacher Collins, Pierre Robin, etc.);
- congenital malformation of the outer ear; and,
- any other situations that affect the outer and middle ear's ability to transmit sound waves to the inner ear.

How does this type of hearing loss affect hearing?

1. Sounds seem dull with little volume, there is a reduction of sound of all frequencies.
2. Sounds that are heard are not distorted.
3. Most speech sounds can be heard but are softer.
4. Words and sentences can be heard and understood with amplification.

What can be done?

1. Excess wax can be removed - by a physician or nurse.
2. The foreign object can be removed - by a physician or nurse.
3. There is some treatment that can be done such as providing medication for allergies and infections; lancing the eardrum to drain fluid from the middle ear; inserting tubes to equalize pressure between the outer and middle ears; and surgically repairing a damaged eardrum.
4. There are also some treatments for some genetic disorders such as providing prosthetic ossicles (the tiny bones of the middle ear) and other surgery.
5. A hearing aid is also often used.

Sensorineural Hearing Loss:

This hearing impairment is due to sensory or nerve damage in the inner ear, the auditory nerve, and/or the brain stem.

What are some of the causes of this type of hearing loss?

This type of hearing loss can be caused by:
- illness (such spinal meningitis - bacterial, viral, and/or fungal, rubella, and other viral/bacterial/fungal infections);
- hereditary (in most cases it is damage to the cochlea);
- Rh factor;
- trauma;
- extremely loud noises;
- syndromes (CHARGE Association, Waardenburg, Pierre Robin, etc.);
- drugs (pre and/or post);
- prematurity; and,
- anoxia (lack of oxygen at birth).

How does this type of hearing loss affect hearing?

1. There is a hearing loss starting with high frequencies.
2. Perceived volume of sound can be severely reduced.
3. Sounds tend to be distorted - even with hearing aids.
4. Not all speech sounds can be heard.
5. Words and sentences can be difficult or impossible to understand through listening alone.

What can be done?

1. Hearing aids can be used - but they only amplify the sound, not improve the quality of the sound.
2. Cochlear implant - may or may not really work for the person. This only improves detection of sound; it does not guarantee recognition of sound without speechreading.
Cortical/Perceptual Hearing Loss:

This hearing impairment is due to damage to the auditory cortex and/or other parts of the brain used to process sound (such as the frontal lobe, parietal lobe, etc.).

What are some of the causes of this type of hearing loss?

This type of hearing loss can be caused by:

- anoxia (lack of oxygen at birth);
- prenatal infections (such as rubella, etc.);
- birth trauma;
- post natal trauma;
- prematurity;
- drugs (both pre- and post natal);
- Rh factor; and,
- brain diseases

How does this type of hearing loss affect hearing?

This depends on which part of the brain is affected.

If the auditory cortex is affected, the person may have difficulty analyzing sounds, remembering sounds, and choosing which sound to attend.

If the frontal lobe is affected, the person may have expressive aphasia which means that person can understand language but not be able to produce it.

If the parietal lobe is affected the person may be unable to process experiences and therefore not be able to use language.

If the temporal lobe is affected, the person may not be able to attach meaning to sound and/or attend to sound.
ANATOMY OF THE HEARING SYSTEM

Figure H-4

EXTERNAL EAR

INNER EAR

MIDDLE EAR

BRAINSTEM

Ear Flap or Pinna

Middle Ear Bones

Semicircular Canals

Cochlea

Eustachian Tube

Auditory Nerve

Auditory Cortex

Eardrum

Ear Canal
Figure H-5
The Cochlea

ANATOMY OF THE COCHLEA

ILLUSTRATION OF INTERIOR OF THE COCHLEA
SHOWING THE NERVE CELLS

Auditory Nerve

Oval Window

319
ANATOMY OF THE BRAIN

Figure H-6

CEREBELLUM

FRONTAL LOBE

SENSORY MOTOR AREA

PARietal LOBE

AUDITORY CORTEX

TEMPORAL LOBE

OCcipital LOBE

BRAINSTEM

320
Sample Audiogram

KEY
O = RIGHT EAR
X = LEFT EAR
[] = BONE CONDUCTION
S = SPEAKER
A = AIDED

Figure H-7

Hearing Threshold Level in Decibels Re: ISO 1964 Standard
Figure H-1

Amounts of Hearing Loss

Hertz (Hz)
(Cycles per second, pitch, frequency)

0-20 dB = Range of Normal Hearing
21-40 dB = Mild Hearing Loss
41-55 dB = Moderate Hearing Loss
56-70 dB = Moderately Severe Loss
71-90 dB = Severe Hearing Loss
91+ dB = Profound Hearing Loss
<table>
<thead>
<tr>
<th>dB Threshold</th>
<th>Degree of Hearing Loss</th>
<th>Ability to Detect Speech</th>
<th>Familiar Sounds</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td>Can hear speech with no difficulty</td>
<td>Rustling leaves</td>
</tr>
<tr>
<td>10</td>
<td>Normal</td>
<td>Can hear speech with no difficulty</td>
<td>Whisper</td>
</tr>
<tr>
<td>20</td>
<td>Mild</td>
<td>Can hear speech unless distant or faint</td>
<td>Watch tick</td>
</tr>
<tr>
<td>30</td>
<td>Moderate</td>
<td>Can hear conversational speech at 3 to 5 feet - needs hearing aid</td>
<td>Air conditioner</td>
</tr>
<tr>
<td>40</td>
<td>Severe</td>
<td>Can hear loud conversations, has great difficulty in group situations</td>
<td></td>
</tr>
<tr>
<td>50</td>
<td>Severe</td>
<td>Max hear loud voice about one foot away, may identify noises in environment, may distinguish vowels</td>
<td>Piano</td>
</tr>
<tr>
<td>60</td>
<td>Severe</td>
<td>Max hear loud voice about one foot away, may identify noises in environment, may distinguish vowels</td>
<td>Food blender</td>
</tr>
<tr>
<td>70</td>
<td>Severe</td>
<td>Max hear loud voice about one foot away, may identify noises in environment, may distinguish vowels</td>
<td></td>
</tr>
<tr>
<td>80</td>
<td>Severe</td>
<td>Max hear loud voice about one foot away, may identify noises in environment, may distinguish vowels</td>
<td>Piano</td>
</tr>
<tr>
<td>90</td>
<td>Severe</td>
<td>Max hear loud voice about one foot away, may identify noises in environment, may distinguish vowels</td>
<td>Food blender</td>
</tr>
<tr>
<td>100</td>
<td>Severe</td>
<td>Max hear loud voice about one foot away, may identify noises in environment, may distinguish vowels</td>
<td>Lawn mower</td>
</tr>
<tr>
<td>110</td>
<td>Severe</td>
<td>Max hear some loud sounds</td>
<td>Jet at 10,000 feet</td>
</tr>
<tr>
<td>120</td>
<td>Severe</td>
<td>Max hear some loud sounds</td>
<td>Motorcycle</td>
</tr>
<tr>
<td>125</td>
<td>Severe</td>
<td>Max hear some loud sounds</td>
<td>Rock band</td>
</tr>
<tr>
<td>130</td>
<td>Severe</td>
<td>Max hear some loud sounds</td>
<td>Jackhammer</td>
</tr>
<tr>
<td>140</td>
<td>Threshold of Pain</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Degrees of Hearing Loss**
Figure H-2

Comparison of the Frequency and Intensity of Various Environmental and Speech Sounds

Frequencies in Cycles Per Second (Hertz)
Two Illustrations of Hearing Loss with a Flat Audiogram
Figure H-9

Two Illustrations of Hearing Loss with a Sloping Audiogram
Figure H-14

Illustration of Conductive Hearing Loss

X = LEFT EAR
O = RIGHT EAR
[] = Bone
[-] = Conduction
Illustration of Sensorineural Hearing Loss

Case 1:

Case 2:
Activity:

Hearing Loss Simulations
Moderate Hearing Loss Simulation
Sensorineural/High Frequency
(Sentences)

1.

2.

3.

4.

5.
## Sensorineural/High Frequency Hearing Loss (Spelling Test)

<table>
<thead>
<tr>
<th>Severe Hearing Loss</th>
<th>Moderate Hearing Loss</th>
<th>Mild Hearing Loss</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>1.</td>
<td>1.</td>
</tr>
<tr>
<td>2.</td>
<td>2.</td>
<td>2.</td>
</tr>
<tr>
<td>3.</td>
<td>3.</td>
<td>3.</td>
</tr>
<tr>
<td>4.</td>
<td>4.</td>
<td>4.</td>
</tr>
<tr>
<td>5.</td>
<td>5.</td>
<td>5.</td>
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<tr>
<td>6.</td>
<td>6.</td>
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</tr>
<tr>
<td>7.</td>
<td>7.</td>
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</tr>
<tr>
<td>8.</td>
<td>8.</td>
<td>8.</td>
</tr>
<tr>
<td>9.</td>
<td>9.</td>
<td>9.</td>
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Transcriptions for the Hearing Loss Simulations

Moderate Hearing Loss Simulation
Sensorineural/High Frequency
(Sentences)

1. It is tough to pass a calm pool on a hot day.
2. Please sell me a nice book with white pages
3. Walk back and knock on the ranch house door.
4. Your wife bought a fine chair after a long search.

Sensorineural/High Frequency Hearing Loss
(Story)

Normal Hearing:

During the early 1900's, the bustling (sic) city of Mobile, Alabama, was experiencing its first automobiles. Auto drivers were terrible. They demanded to drive as they pleased. Their only concern was for their destination and getting there as quickly and as precisely as possible.

It was not uncommon to see an auto drive through someone's backyard, or through a vacant lot just to get somewhere faster. Drivers commanded the roads as they pleased and at the speeds they pleased. Collisions were frequent and damage to property was tremendous.
Mild Hearing Loss:

The City Fathers of Mobile became very concerned. They tried countless safety precautions, all without the slightest success.

If it hadn't been for Random Persons, a citizen of Mobile, the automobile problems would quite likely never have ended. And it all happened as the result of an unlikely string of events.

Random Persons was a salesman. He had spent a long, hot, and tiring day making sales calls far to the south. As he drove home, his mind constantly wandered from his driving. As he neared the edge of the city, he recalled later, he hit a series of bumps that startled him.

Moderate Hearing Loss:

The next morning, Random called the County Road Crew to report those road bumps of the night before. When the Road Crew arrived at the section of road reported by Random, they beheld a curious sight and smell. There in the exact center of the road and exactly five feet apart were a dozen white stripes.

It didn't take the Crew Boss long to see that the stripes were skunks which had been flattened and merged into the freshly oiled road.

Severe Hearing Loss:

It looked as though a family of skunks had been partaking in a familiar game of Follow-the-Leader, strolling down the highway only to be flattened into the road surface by Random's unattentive (sic) driving.

The scent from the flattened skunks easily persuaded the Road Crew to leave them be. As a consequence, motorists driving that stretch of road were forced to stay to the extreme sides of the road to avoid the strong odor.

Mild Hearing Loss:

The City Fathers were quick to see, and smell, the safety implications of the entire mishap. They were quick to observe that the white stripes divided the road and kept opposing traffic apart. The odor also kept everyone awake and moving along.
The City Fathers knew that they couldn't divide every road with skunks. Soon skunks would become extinct. So they simply painted white stripes in the middle of all Mobile roads. And it worked.

Soon an entire nation followed the Mobile example, dividing roads with painted white lines.

**High Frequency Sensorineural Hearing Loss**
*(Spelling Test)*

1. Bath
2. Pearl
3. Sour
4. Mouse
5. Learn
6. Wheat
7. Vine
8. Tape
9. Hedge
10. Mood
How Hearing Loss Affects Communication and Concept Development
How Hearing Loss Affects Communication and Concept Development

After having had two experiences involving hearing loss, how do you think hearing loss affects communication and concept development?
Communication Modes
HELEN KELLER NATIONAL CENTER
COMMUNICATIONS LEARNING CENTER

LANGUAGE LEVEL 1

CHARACTERISTICS

a. Minimal expressive and receptive communication skills
b. No formalized linguistic symbol system
c. May display behaviors of tactual defensiveness
d. May have no previous exposure to sign language
e. Methods of communication may include natural body gestures, object cues, environmental cues and establish daily routines
f. Limited past experience
g. Difficulty generalizing information across a variety of domains

INSTRUCTIONAL PROGRAMS

Language training (concept and skill development) must occur in the natural environment during "hands on" meaningful activities. Instruction most successful outside the domain of the classroom throughout the course of the day.

Personal Grooming Program
Meal Time Program
Leisure Time Activities
Work Training Program
Community Activities Program

OBJECTIVE EXAMPLES

a. Manipulates objects and persons to make needs known.
b. Accept molding of hand to form sign for functional objects in the environment during daily routine with a familiar person. Also attends to "speaker's" gestures and signs.
c. Responds appropriately to signed directions paired with concrete object during daily routine.
d. Identifies self using name sign or tangible symbol.
e. Pairs object with picture.
f. Indicates basic needs (toilet, drink, food), using sign, pointing to picture.
g. Indicates preference given a variety of objects representing choice of activity.
LANGUAGE LEVEL 2

CHARACTERISTICS

a. Adequate basic communication skills to make a variety of needs known.
b. Responds appropriately to concrete messages presented during daily routine.
c. May minimally initiate communication but can interact with others possessing common mode.
d. Language may be minimally descriptive.
e. Topics of communication are generally concrete and dependent upon past experience. Abstract ideas are difficult to comprehend or express.
f. Limitations or gaps in linguistic competence influences independence in other areas.
g. Supplemental communication aids or alternative methods are necessary.

INSTRUCTIONAL PROGRAM

1. Intensive one-to-one instruction in classroom domain
   a. Communication books
   b. Vocabulary development - sign, written, sight word
   c. Basic written language skills (language experience approach)

2. Opportunities to acquire skills in the functional domain (Community Neighborhood Experience) Communication with non-signing public

3. Conversation groups

OBJECTIVE EXAMPLES

1. Creates simple sentences using NOUN plus VERB structure while relating events of personal experience.

2. Responds appropriately to WH? (What, who, where, when)
3. Increases ability to interact with peers in a group situation - asks appropriate questions to gain additional information from others, elaborates on topics of conversation etc...

4. Utilizes communication book in a restaurant to order and pay for food

5. Increases functional sign vocabulary (foods, activities, feeling, cooking term, etc...)

6. Increase sight word identification for functional items, survival phrases, etc...
CHARACTERISTICS

a. Possesses a strong preferred modality of communication (ASL or English)
b. Highly skilled in at least one method of communication
c. Must learn new modality of communication due to recent sensory loss (ex. braille vs. print, tactual sign language vs. visual sign language)
d. Onset and/or degree of sensory loss influences type of training pursued
e. Requires alternate techniques due to recent sensory loss so as to increase independence in the public domain

INSTRUCTIONAL PROGRAMS

a. Vocabulary development - sign, written, braille
b. Reading comprehension
c. Remedial English Grammar
d. Advanced written language skills
e. Spelling
f. Conversational sign language skills

OBJECTIVE EXAMPLES

a. Writes descriptive essay
b. Uses written language as an alternate method of communication in the public domain
c. Writes a business letter
d. Demonstrates adequate language skills to utilize aids and devices for communication purposes
e. Accommodates and adapts to other individuals with communication differences
f. Demonstrates ability to read and comprehend various publications, i.e.; newspaper, magazine articles for information and enjoyment purposes

LANGUAGE EVALUATION

A variety of formal and informal testing tools may be used to collect and evaluate language samples.

WRITTEN LANGUAGE

1. Cued language samples using pictures
2. Informal (natural) samples (i.e., notes, letters, autobiographies, journal entries)
3. Stanford Achievement Test
4. Gallaudet Diagnostic Test S:EP
5. Basic "who","what", "when", "where", questions
   Test of Syntactic Abilities

ORAL/SIGN LANGUAGE

1. Conversation
2. Teacher directed commands
3. Vocabulary lists
4. Observation of sign language style: ASL, PSE, SEE, Gesture
5. Pictures/objects to elicit descriptive language
6. Pictures/objects to elicit question responses

READING COMPREHENSION

1. Basic "who", "what", "when", "where", questions
2. Selected paragraphs followed by discussion (i.e., re-tell the story)
3. Selected paragraphs followed by questions
4. Stanford Achievement Test
FUNCTIONAL APPROACH TO LANGUAGE INTERVENTION AND ASSESSMENT

Language is a Social Tool - the conversational approach to teaching language skills uses the environment.

You must manipulate variables in the environment to create opportunities for language learning. Training must occur in the functional environment inside eyes or during hands on meaningful activities.

The communication behavior taught must have a purpose. Preferable it is a high frequency behavior. This will enhance the generalization of this behavior.

You have communication before you learn language. A communication system motivates a person to learn language. A person must have a reason to learn symbols.

So, the first step is to establish a conventional communication modality (visual, auditory, tactile, mixed).

Pre-symbolic skills include
1. Physical imitation
2. Object permanence
3. Turn taking
4. Functional use of objects
5. Gestures
6. Receptive language

Non-symbolic Communication
1. Body movements
2. Facial expressions
3. Tactile cues (ie: on the body touch cues)
4. Vocal sounds
5. Eye contact
6. Gestures
7. Imitation

Emergency of symbol system
1. More formalized gestures
2. Object cues
3. Basic signs iconic ----- arbitrary
4. Pictures - line drawings, photos
5. Signed phrases
6. Communication books
Tying It All Together
Tying It All Together

Through this workshop you have found that there are four major areas of concern when working with Deaf-Blind individuals:

1. Anticipation
2. Motivation
3. Communication
4. Learning

Deaf-Blind individuals may:

* lack the ability to communicate with their environment in a meaningful way
* have a distorted perception of their world
* lack the ability to anticipate future events or the results of their actions
* be deprived of many of the most basic extrinsic motivations
* be unable to benefit from opportunities for secondary or tertiary learning
* have had medical problems that led to serious developmental lags
* have been mislabelled as mentally retarded/developmentally delayed or emotionally disturbed
* have been forced to develop unique learning styles to compensate for their multiple disabilities
* have extreme difficulty in establishing and maintaining interpersonal relationships with peers
* have been placed in inappropriate educational and adult programs designed to serve other developmental disabilities - such placements resulted in further developmental delays
What are some strategies when working with Deaf-Blind Adults?

1. Use one-on-one interaction initially
2. Provide constant, non-distorted information to the consumer
3. Use the consumer's communication system (signals and cues, coactive signing, interactive signing, pictures, Braille, etc...)
4. Make the task meaningful and relevant - provide motivation
5. Support
6. Adapted learning strategies.