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ABSTRACT

This paper describes Williams syndrome, a rare genetic disorder that results in mental retardation. An overview of this condition is presented from the perspective of a family who traveled through the many stages of the disability from infancy to young adulthood. The etiology and characteristics of the disability are discussed, including distinctive facial features, delayed motor development, and speech patterns. The paper also addresses the educational implications of the disability. The behavior problems of children with Williams syndrome are identified, including their short attention spans, difficulty in modulating emotions, anxiety around unexpected changes in routine, heightened sensitivity to sounds, perseverating on favorite conversational topics, rocking, and difficulty building friendships. Testing, placement, and academic difficulties are also discussed along with suggestions on how to make appropriate accommodations. The paper closes with a description of the educational career of a child with Williams syndrome. A list of resource organizations is provided and information summary sheets are attached for overhead display. (CR)

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Williams Syndrome

A Family's Journey

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WILLIAMS SYNDROME-A FAMILY'S JOURNEY

Presentation Fifth International Conference on Mental Retardation and Developmental Disabilities
Division on Mental Retardation/Developmental Disabilities of Council for Exceptional Children

October 11, 1996

Austin Marriott at the Capitol

Austin, Texas

Abstract:

This session will present a description of Williams Syndrome, a rare genetic disorder resulting in Mental retardation. An overview of this condition will be presented from the perspective of a family who has traveled through the many stages of this disability from infancy to young adulthood.

Introduction/Background

[Show baby or infant picture of Brian]

This is Brian. Born in 1975. Had rough start. Heart problems. Almost died at birth. Brian began receiving infant training from MHMR. They noted eye-hand coordination and gross motor difficulties among others. At age three a pediatrician made a diagnosis of MR. Released from MHMR at age 3 for early childhood service in a public school district. At this time, the proper diagnosis of Williams Syndrome was made.

Description/Etiology

Williams syndrome is a neurobehavioral congenital disorder (Levine, 1993). It is said to occur sporadically, in about 1 of every 20,000 births. This disorder does not run in families and is not due to any medical, environmental, or psychosocial factors. These children tend to be delayed in development and show a range of learning difficulties which can vary from mild to severe cognitive impairment. There are distinctive patterns of abilities, behavioral, and personality characteristics common to most individuals with this syndrome. These characteristics set them apart from other children with developmental disabilities.

Williams syndrome was characterized and first described by Dr. John C. P. Williams of New Zealand (Williams, 1961). Research has indicated that individuals with Williams are missing an elastin gene on chromosome #7. Elastin is a protein that forms the principal substance of elastic tissue, which is a connective tissue occurring in the walls of arteries and veins. The deletion of this gene is said to account for many of the syndrome's features although it is also believed that there are other genetic material missing from the same region.

Williams syndrome is present at birth and can be diagnosed at that time; however it often goes undiagnosed or is misdiagnosed. Many infants with Williams are colicky for the first several months of life and have difficulty sleeping. Most develop severe feeding difficulties during the first year, including vomiting, constipation and refusal to feed, & they can be irritable and cry excessively. [These symptoms were Brian's parents first clues].

Currently, there is no specific or routine laboratory test used to diagnose Williams syndrome. A diagnosis is usually made when a trained professional in genetic disorders of birth defects obtains information through a case history, physical examination, a developmental assessment, and observing behavior. Testing finds that many children have raised levels of calcium in the blood (Hypercalcemia). Diagnosis can also be made by a cardiologist since many of these kids have cardiac problems. The most common problem is supravalvular aortic stenosis (SAS). This is a narrowing or constriction of the aortic valve. Individuals with SAS have the following signs and symptoms: dyspnea (shortness of breath), profuse sweating, flushing sign, pounding pulsations in the neck arteries, and a heart murmur.

In infancy, children with Williams may show a failure to thrive and some will have hypercalcemia (raised levels of calcium in their blood). They will show motor and mental developmental delays with the motor delays being the most pronounced. Milestones are delayed. Walking occurs at an average of 21 months, talking at 21.6, and toilet training at around 39 months (Morris, 1988).

Characteristics

Williams syndrome is characterized by distinctive facial features, mental retardation (measured IQ 50-70 range), SAS, infantile hypercalcemia, growth deficiency, and acute hearing. Learning problems are common including the inability to remember on a short-term basis but the ability to remember events long after they occur, an unusual friendly personality, small teeth with inadequate enamel covering and spacing, a low pitched voice, and talking as though their speech has been patterned for them. They demonstrate limited independence and are quite uninhibited in approaching strangers.

Children with Williams syndrome generally have characteristic facial features. These include almond shaped eyes, a broad forehead, depressed nasal bridge with an uptilted nose, thick lips with a protuberance of the lower lip, wide mouth, widely spaced teeth, strabismus (a squint), a star shaped pattern of iris in blue and green-eyed children (brown-eyed children usually don't display this pattern), puffiness around the eyes, small head, underdeveloped cheekbones, and elfin ears. Often called 'elfin facies' syndrome or Pixie children. [picture]

Motor development is generally delayed. These children begin walking later than expected. Typically have a very unusual gait. Brian walked on his toes. Often have coordination, balance, and strength problems. Tend to have fine motor difficulties apparent from an early age. These children are often shorter than would be expected when compared to parents. Brian was small for his age. He was wearing clothing for 12 month olds at the age of 3. Also, children with WS have an early puberty, at around 8 or 9 years old. Brian is atypical in that his puberty is late. At 22 years of age, he still acts like a much younger child in terms of his maturity.

People with Williams speak fluently, and yet they often seem to miss the underlying point of what other people are saying. There is a richness to the vocabulary of a child with Williams with a tendency to come up with unusual or unexpected words in spontaneous conversation. This speech pattern is referred to as "cocktail party" speech. Descriptions of cocktail party speech focus on fluent, verbose, adult-like usage, a precocious vocabulary, but output is superficial, inappropriate, and lacking in content (Frank, 1983). Many chatter constantly, but at a superficial level, and often in a formal, adult-like way with frequent use of stereotyped. Interestingly, the onset of language is delayed but, once the child starts talking, language abilities seem to develop at a faster rate.

"Alex Biescar is a good example. At age 3 he hadn't uttered a single word. One day he was at the speech pathologist's office, waiting in the reception area. A floor fan caught his attention, and since Alex shares a common fascination with spinning things, he approached the fan and began staring at it. The concerned receptionist switched it off. Alex switched it back on. She unplugged it. Alex plugged it back in. Next she switched it off and unplugged it. Alex solved this problem as well. Just then, there was a power failure. Alex flipped the switch a couple of times and nothing happened. He wiggled the plug and nothing happened. Frustrated he uttered the first words of his life: "Jesus Christ, this doesn't work!" (Finn, 1991)

Educational Implications/Instructional Strategies

Educators are confused because children with WS test in the MR range, talk like a gifted child, behave at times like an ED child, and function academically like a child with LD. Each of these terms have specific characteristics and meanings for educators, but do not seem to fit the characteristic peaks and valleys of abilities of individuals with WS. Overall result is that these children are not well served by schools (Finn, 1991).

Behavior

There are behavioral characteristics that can pose challenges in classrooms. Most WS children are even-tempered and cooperative but can have quite severe tantrums when they are frustrated, when they want attention, or cannot get their own way. Some possible factors:

- These kids tend to have short attention spans and are very distractible.
- Difficulty modulating emotions. Extreme excitement when happy; tearfulness in response to mild distress, or terror in response to apparently mildly frightening events.
- Anxiety around unexpected changes in routine/schedule

- Heightened sensitivity to sounds (hyperacusis)
- Perseverating on certain "favorite" conversational topics
- Rocking, nail biting, of skin picking
- Difficulty building friendships (peers)

These children may benefit from a behavioral management program. Positive reinforcement or rewards motivate these children. In the classroom, to modify unacceptable behavior, give an alternative appropriate behavior. Social skills may need to be included in the IEP.

Testing

Children with Williams generally do not test well, often scoring lower than they are expected to. Gardner's multiple intelligence theory has been applied to this syndrome, as these children tend to have significant areas of strength and weaknesses (Armstrong, 1987). Correct interpretation of testing data is very important.

Problems with sustained attention is a difficulty in testing situations. Hyperacusis makes it extremely difficult for them to concentrate when sounds are in the environment. Sounds which may seem insignificant, such as water running through pipes or the sound of a lawnmower from outside can cause distraction. Are also aware of fluctuations in sound. Most children with Williams attend better when there is a social component to the test, they are rewarded with a lot of praise, and they can have success at the task. Testing should be done over several sessions.

Placement

The distinctive behavior of children with Williams makes it hard for them to fit easily into a general classroom. They can be hyperactive, us inappropriate social skills, and are easily distracted. An open classroom setting is too distracting. Shifting from one teacher to another does not work well for these children (Cooper, 1988).

These kids tend to do better in small classes of 6-10. There is, however, a great deal of variation in classroom situations. Some do well in regular classroom settings, while getting help outside of class. Others do well in regular classes with an aid. Children with more significant learning and or behavior problems often need a more specialized classroom placement. It is recommended that these children not be placed in classes for students with behavioral disorders as their behavioral issues and needs for support are very different from children with ED.

Academics:

Writing is an area of difficulty due to the inability to control a pencil and maintain correct pressure. Very tedious process. Success has been attained by utilizing markers or felt-tipped pens. Letter formation is usually poor. Typewriters and computers are useful. Cursive is easier to use than manuscript.

Reading. A phonetic and linguistic approach has proven to be best, as opposed to a whole word sight reading approach. Tend to have good blending abilities. Important to remember that kids with WS can be easily distracted by extraneous visual stimuli. Books with many detailed pictures and colors on same page as printed words are not ideal. Books with simple pictures or line drawings are less likely to distract the child.

Math. Arithmetic has been shown to be an area of difficulty. Perceptual, visual-spatial and motor problems make manipulating numbers, carrying out operations, and comprehending principles behind the operations problematic. Also have less abstract reasoning ability. Manipulatives are helpful. Writing numbers and the teaching of concepts should be separated. Has been pointed out that even older children have great difficulty writing down numbers correctly and aligning numbers on the page when adding or subtracting.

Brian's Educational Career

Brian's educational career is a good illustration of what usually happens to these children. At age 3 he entered early childhood educational services in the public school. He left at age four because he was said to be disrupting the class. At age 4 Brian went to a private school for Learning Disabilities. Mother has stated that this was a very beneficial experience, perhaps the most beneficial educational setting. At age five his teacher noted memory problems. Used music to teach concepts. Typical trait-musical abilities. Math a particular weakness-another common trait. Has to use a number line. Can't use a worksheet. Due to heavy phonics instruction, Brian reads at an eight grade level. He has very poor comprehension skills, which is also typical. Very good at puzzles. Concrete, hands-on instruction works best. For instance, he could count money when real coins were used but unable to with play money. Difficulty with handwriting but a felt-tipped pen seemed to be beneficial.

At age 13, his academics peaked. He was placed in a Life-Skills program. The focus was on Functional academics: shopping, cooking, laundry, housekeeping. This is in keeping with information that shows that adults with Williams can be semi-independent. Most require on the site job coaches and most live in group homes. The majority of adults have jobs ranging from certified nurse's aides to janitorial duties. Some Williams adults marry and have families, but with support (Biescar, 1991).

For these reasons, transitional issues have been a major issue in Brian's life. Mother reported that there was no mention of transition on the ARDs at 16 and 17 years of age. He was placed in resource reading but had one-half day of work and half-day of functional academics. At 19 he was supposed to be placed at a paying job. The ITP stated that he would have one-half day of school and a half-day of work. Unfortunately, this didn't come about. The parents had to file a complaint against the district. Brian was placed on a job at a mechanics shop. This was in keeping with his love of machines-another common characteristic of Williams. Unfortunately this arrangement did not last. The school personnel had not properly notified the job site and the owner did not want a child with a disability as an employee. Next, Brian went to work at a local Furrows stocking the shelves. Lost that job in a budget cut. Currently back in Life Skills. The Texas Rehabilitation Commission and MHMR have attended the last two ARDs at mom's request. The TRC will provide job training in auto mechanics starting in July.

Organizations/Information Sources

Williams Syndrome Association Inc.
P.O. Box 297
Clawson, MI 48017-0297
(810) 541-3630 [voice]
(810) 541-3631 [fax]

Canadian Association for Williams Syndrome
P.O. Box 2115
Vancouver, B.C. V6B 3T5

Williams Syndrome Association
Gulf Coast Chapter
Kay Biescar-Regional Director
16211 N. Greenfield Dr.
Klein, TX 77379
(713) 376-1626



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PHYSICAL CHARACTERISTICS

FACIAL FEATURES:

- Small head with broad forehead
- Puffiness around the eyes
- Stellate eye pattern (star shaped)
- Upturned nose and depressed nasal bridge
- Full lips and widely spaced teeth
- Small chin
- 'Elfin' appearance. "Pixie children"

BODY STRUCTURE

- Sloping shoulders
- Elongated neck
- Protruding abdomen
- Distinguishable gait

HEART DISORDERS

- Supravalvular aortic stenosis (narrowing of the aortic valve)
- Pulmonary stenosis

OTHER

- Kidney, bladder, and renal involvement/complications
- Low/poor muscle tone, joint contractures become more common with age

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AGE-RELATED CHARACTERISTICS

EARLY CHILDHOOD

- Premature puberty
- Low birth weight (failure to thrive)
- Hypercalcemia (elevated serum calcium levels) in first 2 years
- Abnormal sleep patterns
- Hyperacusis (sensitive hearing)

SCHOOL AGE

- Mild to severe learning difficulties often associated with visuo-perceptual-spatial deficits
- Math and time concepts may be especially difficult
- Language in older children may show higher levels of production than comprehension
- May continue to exhibit difficulties/delays with gross and fine motor skills

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- May exhibit anxious personalities, impulsive tendencies and poor concentration
- Difficulty forming relationships with peers, preferring company of younger children or adults

ADOLESCENT & ADULT

- Adult stature may be smaller than average
- Physical and medical problems may be progressive
- Potential for hypertension exists

PSYCHOLOGICAL

DEVELOPMENTAL

- Delays in gross and fine motor skills (sitting, walking) and early language development

COGNITIVE AND ADAPTIVE BEHAVIOR

- Exhibits deficits in intellectual functioning, reasoning, social behavior, and self-help skills

PERSONALITY

- Social, outgoing, overly friendly to adults, unafraid of strangers, talkative, great enthusiasm for certain topics
- Hyperactive, social isolation, and anxiety (behavioral temperaments)

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Learning Strengths:

EXPRESSIVE VOCABULARY

LONG TERM MEMORY FOR INFORMATION

INFORMATION FROM PICTURES

ABILITY TO LEARN THROUGH "HANDS ON"

MUSICAL ABILITY

**SHORT AND LONG TERM AUDITORY
MEMORY**

**INTEREST IN & AWARENESS OF EMOTIONS
OF OTHERS**

18

ABILITY TO INITIATE SOCIAL INTERACTION

19

Learning Difficulties

**FINE MOTOR OR VISUAL MOTOR
INTEGRATION**

SPATIAL ANALYSIS

WORD FINDING

MATH SKILLS (Money, time, columns of numbers)

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