

NASET Lesser Known Disorders in Special Education Series

Issue # 2 - February 2010

Each issue of this series contains at least three lesser known disorders. Some of these disorders may contain subtypes which will also be presented. You will also notice that each disorder has a code. These codes represent the coding system for all disabilities and disorders listed in the [Educator's Diagnostic Manual \(EDM\)](#) Wiley Publications.

Disorders in this issue:

- [Gerstmann's Syndromes](#)
 - [Hyperlexia](#)
 - [Cornelia de Lange Syndrome](#)
-

LD 6.00-Gerstmann's Syndromes

Disability Category - Learning Disability

Definition

Gerstmann's syndrome is a neurological disorder. The disorder should not be confused with Gerstmann-Sträussler-Scheinker disease, a type of transmissible spongiform encephalopathy.

In adults, the syndrome may occur after a stroke or in association with damage to the parietal lobe. In addition to exhibiting the above symptoms, many adults also experience aphasia, (difficulty in expressing oneself when speaking, in understanding speech, or in reading and writing).

Diagnostic Symptoms

This disorder characterized by four primary symptoms:

- 1.) a writing disability (agraphia or dysgraphia)
- 2.) a lack of understanding of the rules for calculation or arithmetic (acalculia or dyscalculia)
- 3.) an inability to distinguish right from left, and
- 4.) an inability to identify fingers (finger agnosia)

Further Key Points

There are few reports of the syndrome, sometimes called developmental Gerstmann's syndrome, in children. The cause is not known. Most cases are identified when children reach school age, a time when they are challenged with writing and math exercises. Generally, children with the disorder exhibit poor handwriting and spelling skills, and difficulty with math functions, including adding, subtracting, multiplying, and dividing. An inability to differentiate right from left and to discriminate among individual fingers may also be apparent. In addition to the four primary symptoms, many children also suffer from constructional apraxia, an inability to copy simple drawings. Frequently, there is also an impairment in reading. Children with a high level of intellectual functioning as well as those with brain damage may be affected with the disorder (National Institute of Neurological Disorders and Stroke, 2004).

AU 5.00-Hyperlexia

Disability Category - Autism

Definition

A type of syndrome often associated with autistic features characterized by above normal ability to read coupled with a below normal ability to understand spoken language (American Hyperlexia Association, 2005).

Diagnostic Symptoms

Oral Hyperlexia is a syndrome with the following three main characteristics (Autism Support Network, 2002):

1. Early precocious and/or intense fascination with letters or numbers
2. Delays in verbal language
3. Social skills deficits

Examples of the above include (American Hyperlexia Association, 2005):

- Learn expressive language in a peculiar way, echo or memorize the sentence structure without understanding the meaning (echolalia), reverse pronouns
- Listen selectively, appear to be deaf
- Normal development until 18–24 months, then regression
- Rarely initiates conversations
- Self-stimulatory behavior
- Significant difficulty in understanding verbal language
- Specific, unusual fears
- Strong auditory and visual memory
- Think in concrete and literal terms, difficulty with abstract concepts

Further Key Points

Hyperlexia has characteristics similar to autism, behavior disorder, language disorder, emotional disorder, Attention Deficit Disorder, hearing impairment, giftedness or, paradoxically, mental retardation.

To develop effective teaching strategies and more typical childhood development, it is important to differentiate Hyperlexia from other disorders. Thorough psychological evaluation by a psychologist who is familiar with the syndrome of Hyperlexia is a crucial first step. Hearing, neurological, psychiatric, blood chemistry, speech and language and genetic evaluations can be performed to rule out other disorders but are not needed to identify Hyperlexia (Kay, 2004).

According to the American Hyperlexia Association (2005), “Hyperlexia's place on or outside of the autistic spectrum is a matter of much debate. Be that as it may, hyperlexia is a trait commonly seen in autistic spectrum disorders. Autistics with hyperlexia have a unique learning style and a better prognosis than those without this reading skill. Hyperlexia is often written off as a "meaningless splinter skill" but it is much more than that even if comprehension lags because reading can be a very useful tool for learning other skills and can be the doorway to language in general” (p.1).

SL 1.00 Cornelia de Lange Syndrome (CdLS)

Disability Category - Mental Retardation

Definition

Cornelia de Lange syndrome (CdLS) is a rare genetic disorder that is apparent at birth (congenital). Associated symptoms and findings typically include delays in physical development before and after birth (prenatal and postnatal growth retardation); characteristic abnormalities of the head and facial (craniofacial) area, resulting in a distinctive facial appearance; malformations of the hands and arms (upper limbs); and mild to severe mental retardation. Many infants and children with the disorder have an unusually small, short head (microbrachycephaly); an abnormally long vertical groove between the upper lip and nose (philtrum); a depressed nasal bridge; upturned nostrils (anteverted nares); and a protruding upper jaw (maxillary prognathism). Additional, characteristic facial abnormalities may include thin, downturned lips; low-set ears; arched, well-defined eyebrows that grow together across the base of the nose (synophrys); an unusually low hairline on the forehead and the back of the neck; and abnormally curly, long eyelashes. Affected individuals may also have distinctive malformations of the limbs, such as unusually small hands and feet, inward deviation (clinodactyly) of the fifth fingers, or webbing (syndactyly) of certain toes. Less commonly, there may be absence of the forearms, hands, and fingers. Infants with Cornelia de Lange syndrome may also have feeding and breathing difficulties; an increased susceptibility to respiratory infections; a low-pitched "growling" cry; heart defects; delayed skeletal maturation; hearing loss; or other physical abnormalities. The range and severity of associated symptoms and findings may be extremely variable from case to case.

In most individuals with the disorder, Cornelia de Lange syndrome appears to occur randomly for unknown reasons (sporadic). However, there have been some familial cases, suggesting autosomal dominant inheritance (National Organization of Rare Disorders, 2004).
