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This article expands on the March/April 2005 article (on "CdLS and The Eye" written by Drs. Berger, Schloff and Schoedel, describing the implications of many structures of the eyes and various conditions common to persons who have CdLS) in regard to the educational, communication, and social aspects related to visual functioning. "CVI" or "cortical/cerebral visual impairment" is an often overlooked condition that affects many children with CdLS, impinging on not only their vision, but also their ability to learn and interact with their environment.

Vision is complex. It requires (1) the ability of the eyes to receive visual information, (2) the brain's ability to process the visual information in combination with and at the same time as other sensory information and (3) motivation to use the visual system.

All people with CdLS should have examinations by an ophthalmologist or optometrist to determine, as much as possible, the health of the eyes, how clearly they are able to see visual information and/or if they have conditions that may be relieved by surgery, glasses and/or other means. It is the eye that takes in the visual information.

However, it is the brain that processes the information and allows us to understand the visual messages the eyes receive. Understanding all the information the eyes receive may be difficult for some persons because it takes many areas of the brain, working as a team, to process such information. Thus, a major challenge for many people is in understanding the visual information.

Finally, effective use of the visual system requires motivation. It is a lot of work to (1) determine what to look at from the vast array of visual images that bombard us almost every waking hour and (2) coordinate the movements of the ocular muscles in the right direction, track and scan. The most critical aspect, as related to motivation, is the desire to learn from and socially interact with the environment.

Understanding Visual Information

There are some persons with CdLS who have visual behaviors similar to those individuals who have been diagnosed with cortical/cerebral visual impairment – otherwise referred to as CVI. CVI is a disability resulting from either an insult to the brain or how the brain was configured during prenatal development. CVI affects how the brain processes the visual information received by the eyes. Many persons with CVI display wide variations in the functional use of their vision, not just day-to-day but also minute-to-minute.





People with CVI also tend to display some/many of the following behaviors (very few persons have all these behaviors):

- Great variability in how they use their eyes from being visually attentive at one moment and visually inattentive at another moment. May appear to stare into space.
- A tendency to use peripheral vision more than central vision. The midline is rarely the preferred gazing orientation.
- Limited visual attention and a seeming lack of visual curiosity about their environment/surroundings.
- Great difficulty in looking at crowded or complex visual displays for informational purposes.
- A tendency to want to get close to the object(s) they are viewing either to magnify the image or to reduce the complexity of what they are looking at.
- A tendency to be light sensitive and/or a tendency to gaze at lights. Some individuals gaze at light even though they are light sensitive.
- A tendency to look away when reaching.
- A tendency to avoid looking at the human face OR a tendency to stare at the human face OR a
- tendency to stare at just one part of the face.
- Difficulty in managing multi-sensory demands and planning/implementing motor responses; relying mostly on other senses for help/information.
- Possible depth perception difficulties which may affect accurate reaching for objects and/or mobility.
- Many people with CVI have difficulty in making the quantum leap of learning from three-dimensional objects to two-dimensional line drawings or photographs, except when trained picture-by-picture. Even then, it is difficult for many to generalize the understanding of these pictures to different environments and situations.
- For many who are print readers, there may be inconsistent difficulties interpreting print, a need for larger and bolder size print even though visual acuity is near-normal, and a short attention span when using print materials. The latter frequently is due to visual fatigue as a result of the effort to see and interpret the crowded visual



symbols. The major difference between readers with CVI and readers with learning disabilities is that the former is inconsistent. At one moment the person may be able to read well and in the next moment, not be able to read at all. With learning disabilities, there is more consistency in the type of problems related to reading.

- Difficulty in using vision when moving OR difficulty when the object is moving. Some people may have difficulty when there is no movement. This may make moving around in the environment difficult.
- Difficulty in discriminating between and identifying people based solely on vision, relying on the other senses for information. This is called *prosopagnosia* or, in its more extreme form, *facial agnosia*.

Numerous other possible behaviors may be associated with CVI including difficulty in understanding the full range of language used by others as well as the language they, themselves, use. Such communication difficulties may range from people who understand only the emotional and melodic aspect of language to other people who may talk a lot but have significant difficulty in understanding the social aspect of the verbal exchange.

The Implications of Cortical/Cerebral Visual Impairment (CVI) for Students Who Have CdLS:

A primary implication of CVI is the confusion that is created for families and professionals unfamiliar with this condition when visual interest is not obvious or is demonstrated only occasionally. Such confusion is even greater when the medical eye examination is relatively normal. Such variability in visual responsiveness may be highly dependent on the person's level of fatigue, stress, medication, motivation for the task, competing sensory demands, position, and motor requirements of the task. Visual behaviors also may be highly influenced by physical challenges that do not allow the person easy visual access to the environment and by the nature of a seizure condition, should that exist.

Another primary implication of CVI, especially in relating with students who have CdLS and do not talk, is the monumental task in determining what type(s) of communication systems would be the most appropriate. It must be understood that the use of line drawings (e.g., Mayer-Johnson), photographs and/or print assumes the ability to interpret abstract visual symbols. As part of this consideration, it is critically important to remember that when a person looks at something, the visual gaze does not automatically translate into understanding (although it does show interest). The difficulty in understanding may be due to a lack of appropriate visual experiences, a cognitive status that does not support understanding two-dimensional abstract representation or a form of CVI. Thus, when planning what type of communication system might be introduced to a student who does not talk, the following must be considered:





- Does the student demonstrate many of the behaviors described previously that might suggest there is a possibility of some degree of cortical/cerebral visual impairment?
- Does the student consistently demonstrate knowledge of object function and anticipation of the subsequent activity based solely on looking at an object that is an integral component of the activity? If the student cannot visually interpret the three-dimensional world (e.g., objects), how can the student be expected to interpret the two-dimensional world (e.g., pictures, line drawings)?
- Does the student prefer to use objects to communicate while rejecting the use of pictures, line drawings and/or communication devices (for more than cause & effect purposes)?

Finally, the diagnosis of CVI is not restricted to persons who have severe and obvious physical and/or cognitive challenges. Nor is CVI restricted to persons who do not talk or those who have acuity measurements in the visual impairment range. Recent research has demonstrated that CVI also may be a "hidden disability" for many walking, talking academic students who also have near-normal visual acuity. Diagnosing CVI is difficult when there is no focal brain insult. The best diagnostic methods to determine CVI include the medical eye care examination and systematic observations of functional visual behaviors by families and involved professionals. It is especially helpful to have a certified teacher of the visually impaired, who has training and experience with cortical/cerebral visual impairment (CVI), as part of the diagnostic team. Many students, with and without CdLS, have been given visual communication systems without appropriate and thorough observations of their understanding of visual information. Such decisions may have a life-long impact on the educational, communication, and social aspect of these students' lives.

