

ABSTRACT

Title of Document: Reliability of The CVI Range – A Functional Vision Assessment for Children with Cortical Visual Impairment

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Children identified as visually impaired under the Individuals with Disabilities Education Act (IDEA) need to have a functional vision assessment to determine how the visual impairment affects educational performance. Most current functional vision assessments have been based on the needs of children with ocular visual impairments (children with damage to the eye structures). Children with visual impairment due to brain damage, or cortical visual impairment (CVI), have unique vision characteristics that are often different from children with ocular visual impairments. Given this situation, Roman-Lantzy (2007) developed The CVI Range for conducting a functional vision assessment of children with CVI. The purpose of this study was to examine the reliability of The CVI Range.

In this study, 104 children were assessed with The CVI Range. Twenty-seven children were tested by two examiners to determine inter-rater reliability; 20 children were tested on two occasions to determine the test-retest reliability; and 57 children were tested one time by a single examiner. The CVI Range had an internal consistency measure or alpha of .96. The inter-rater reliability coefficient was .98 and the test-retest

reliability coefficient was .99. In addition, the CVI Range has two sections that are scored differently and the scores from the two sections were compared to determine if they provided similar scores and therefore similar implications for intervention. Kappa, or the index of agreement, for the two parts of the assessment was .88. Results of this study indicate that The CVI Range is a reliable instrument. Future research needs to focus on training requirements related to administration of The CVI Range as well as training of the many professionals that serve children with CVI. Research is also needed to determine appropriate and effective interventions for children with CVI. The CVI Range can be used to document progress and therefore determine the effectiveness of interventions and further knowledge in the field of evidence-based practices that are appropriate for children with CVI.

THE RELIABILITY OF THE CVI RANGE -
A FUNCTIONAL VISION ASSESSMENT FOR CHILDREN WITH
CORTICAL VISUAL IMPAIRMENT

By

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Dedication

“You do the best you can with what you know at the time.

When you know better, you do better.”

Dr. Maya Angelou

This dissertation is dedicated to Gregory James (Jamie) Stepanek, one of the first children

I knew with cortical visual impairment or CVI. At the time I did not know it was CVI, but now I know better. This dissertation is about doing better. This work is for you Jamie, and for all of the other Jamie’s out there.

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CHAPTER I

INTRODUCTION

Visual impairment in young children can be divided into two major categories: ocular visual impairment and cortical visual impairment. Ocular visual impairment involves conditions in which the eye structures are underdeveloped or damaged due to insult, disease, or infection (Teplin, 1995). When ocular structures of the visual system are damaged, the child does not get visual information at all or at best gets unclear or incomplete visual information. With cortical visual impairment (CVI) the eye structures are healthy but the child's brain is damaged or malformed; as a result, the child is unable to interpret the information received from the eyes (Good, et al., 1994). Children can have either type of visual impairment, or they can have co-existing ocular and cortical visual impairment (Hoyt, 2003).

Historically, ocular conditions were the leading cause of visual impairment in young children (Robinson, Jan, & Kinnis, 1987). According to Teplin (1995), examples of these impairments include: retinopathy of prematurity (i.e., disorganized growth of retinal blood vessels due to prematurity which may result in scarring and retinal detachment); cataracts (i.e., clouding of the eye lens which obscures vision); and, optic nerve hypoplasia (i.e., underdeveloped optic nerve which limits transmission of visual information to the brain). With improvement in medical treatments for many of these visual conditions, fewer children today have visual impairment due to ocular conditions (Jan, Good, & Hoyt, 2006; Khan, O'Keefe, Kenny, & Nolan, 2007; Rosenberg, et al., 1996).

As the number of children with visual impairment due to ocular conditions has declined, the number of children with cortical visual impairment has increased (Jan, Good, & Hoyt, 2006). Concurrent with improvements in the treatment of ocular conditions, there have been advances in medical technology such that many children who previously died due to significant brain damage now survive (Groenveld, 2003; Rosenberg, et al., 1996). As a result, CVI is now the leading cause of visual impairment in developed countries (Blind Babies Foundation, 1995; Flanagan, Jackson, & Hill, 2003; Goggin & O'Keefe, 1991; Hatton, 2001; Hatton, Schwietz, Boyer, & Rychwalski, 2007; Khan, O'Keefe, Kenny, & Nolan, 2007; Rahi & Dezateux, 1998; Rogers, 1996; Rosenberg, et al., 1996). This trend is expected to continue (American Printing House for the Blind, 2007; Morse, 1990).

Another recent development in the field of visual impairment is an increase in the number of young children with multiple disabilities in addition to visual impairment (Flanagan, Jackson, & Hill, 2003). Regardless of whether visual impairment is due to ocular or cortical impairment, the percentage of children with additional neurological impairments has increased. Overall estimates are as high as 70% of children with visual impairment have additional disabilities (Teplin, 1995). For children with CVI, the estimates are as high as 88% of children with acquired CVI (i.e. children who experienced brain damage after the perinatal period) to 100% of children with congenital CVI have associated neurological impairments (Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985; Wong, 1991).

Under the Individuals with Disabilities Education Act (IDEA) children who are identified with a visual impairment are required to have an assessment to determine how

the disability affects the educational performance of the child (IDEA, 2004). Medical reports and visual acuity numbers determine eligibility for educational services, but they do not provide useful information about the impact of a particular child's vision loss on that child's education (Teplin, 1995). Instead, educational impact is determined through a functional vision assessment. "Functional vision represents vision-mediated performance on tasks required for daily life" (Mayer & Fulton, 2006, p.66). A functional vision assessment determines how a child uses vision to accomplish activities in the classroom or activities of daily living (Langley, 1998). Based on this information, appropriate developmental and educational modifications and accommodations can be determined that will give the child access to the educational curriculum.

For children with ocular visual impairment, there are a variety of assessment tools and a well-known framework to accomplish this task (Appleby, 2002; Hyvärinen & Appleby, n.d.; Roman-Lantzy, 2006; Teplin, 1995). Assessment of ocular visual functioning typically includes response to light, awareness of visual input or fixation, ocular motor functioning, near and distance acuity measures, color perception, contrast sensitivity, visual field assessments, and visual perceptual skills (Appleby, 2002; Hyvärinen & Appleby, n.d.; Teplin, 1995). Accurate assessment of visual skills and needs allows professionals to provide appropriate accommodations and modifications that address the child's vision needs.

Functional vision assessments can be complicated by the fact that most of the children with visual impairments have multiple disabilities (Morse, 1992; Teplin, 1995). The assessment of vision in children with multiple disabilities is therefore often a challenge (Hall, Orel-Bixler, & Haegerstrom-Portnoy, 1991; Morse, 1991, 1992, 1999).

Children with multiple disabilities may not be able to complete typical vision assessments such as identifying letters or shapes on an acuity chart, or matching colors. Many children with multiple disabilities are non-verbal and not able to follow instructions needed to complete typical vision assessments.

Functional Vision Assessment of Children with Multiple Disabilities and Visual Impairment

In response to the need for assessment guidelines for a functional vision assessment of children with multiple disabilities, the Individualized Systematic Assessment of Visual Efficiency (ISAVE) was developed (Langley, 1998). ISAVE was specifically developed for students with significant cognitive, neurological, and sensory impairments who cannot respond to standard measures of visual functioning (Langley, 1998). In ISAVE, Langley describes specific assessment procedures in the following areas: structural integrity, minimal responsiveness (when a child has light perception only), alignment and ocular mobility, oculomotor skills, acuity (near and distant), visual fields, visual perceptual skills, and social attentional gaze behaviors. In addition to assessment of specific vision skills, ISAVE includes a vision screening test, a baby screening test, and developmental vision inventory. There is also a section on cortical visual impairment that includes diagnostic criteria for CVI. The CVI assessment protocol is used to determine the presence or absence of CVI (Langley, 1998).

Individual school systems sometimes develop guidelines for functional vision assessments for children with multiple disabilities who cannot complete standard vision assessments. One example of a locally developed functional vision assessment is the South Carolina Functional Vision Assessment (South Carolina Department of Education,

1992). The purpose of this instrument is to provide educators with a framework for assessing the functional vision of students across age and ability levels. The South Carolina Functional Vision Assessment includes assessment of the following visual skills: blink reflex, pupillary response, awareness and localization, fixation, muscle balance, eye preference, tracking, fields, shift gaze, convergence, depth perception, visual acuity, scanning, and color preference.

In Maryland, an informal survey was conducted of six local jurisdictions (i.e., Montgomery, Howard, Prince George's, Baltimore, Baltimore City, and Anne Arundel Counties) to determine what instruments were used to complete a functional vision assessment for children with multiple disabilities (Newcomb, unpublished). Each county used a different, locally developed, checklist of vision skills. Several counties also used ISAVE or the South Carolina Functional Vision Assessment. Typical skills to be assessed included: pupillary response, fixation, light perception, near and distance acuity, ocular motor functioning, tracking, shift gaze, contrast sensitivity, muscle balance, visual fields, and visual perceptual skills.

Clearly many professionals have responded to the need for an appropriate functional vision assessment for children with multiple disabilities. However, the actual vision skills assessed are derived from an ocular visual impairment model (e.g., Langley, 1998). In addition, none of the functional vision assessments for children with multiple disabilities had any reliability or validity data available. Consistently, researchers who have studied the characteristics of children with CVI have found that the visual characteristics of children with CVI are different from children with ocular visual impairment (Jan & Groenveld, 1993; Nielsen, 1993; Teplin, 1995). Therefore, ocular

models of functional vision assessment may not be appropriate for children with cortical visual impairment.

Differences in Ocular and Cortical Visual Impairment

Jan and Groenfeld of the Visually Impaired Program of Children's Hospital, Vancouver, British Columbia, Canada were among the first researchers to systematically document the characteristics of children with CVI (Jan, Groenfeld, & Anderson, 1993; Jan, Groenfeld, Sykanda, & Hoyt, 1987; Jan, Groenfeld, & Sykanda, 1990). From their research and observations across two decades, they summarized the primary differences between ocular and cortical visual impairment in children (Jan & Groenfeld, 1993).

According to these researchers, some of the primary differences include:

- Children with CVI often have a normal medical eye exam while children with ocular impairments typically have an abnormal medical eye exam.
- Children with ocular impairments demonstrate consistent visual functioning with a normal visual attention span. Children with CVI demonstrate variable visual functioning with a markedly short visual attention span.
- Coordinated eye movements are usually present in CVI and not in an ocular impairment.
- Children with CVI often demonstrate compulsive light gazing but rarely eye pressing. Children with ocular visual impairment seldom light gaze and often demonstrate eye pressing.
- Sensory nystagmus is often present when an ocular loss is congenital or early and not usually present in CVI.

- Color perception is usually preserved in CVI and may be absent in ocular visual impairment, depending on the disorder.
- Children with CVI almost always have a peripheral field loss. Children with ocular visual impairment usually do not have a field loss.
- Close viewing is present in both types of visual impairment, but for different reasons. Children with ocular visual impairment use close viewing for magnification while children with CVI use close viewing for a reduction in crowding (inability to perceive objects spaced closely together) as well as for magnification.
- Children with CVI usually do not “look” visually impaired (p.101), while children with ocular visual impairment appear visually impaired (e.g., eyes look abnormal).
- Children with ocular visual impairment may have additional neurological disabilities, while children with CVI nearly always have additional disabilities (Jan & Groenveld, 1993).

In further research, Groenveld identified additional differences in ocular and cortical visual impairment (Groenveld, 2003). Children with CVI often show a head turn when they look at or reach for an object of interest; they seem to look and then look away as they reach for the object. Children with ocular visual impairment look and reach at the same time. In addition to the crowding phenomenon mentioned previously, children with CVI often have difficulties with foreground/background perception. They have difficulties when they look for objects against a patterned background as well as objects spaced closely together. Distance viewing is related to this difficulty with background

information. Complexity of visual input often affects the performance of a child with CVI. Gaze aversion, or looking away, increases with more complex visual information (Baker-Nobles & Rutherford, 1995). For children with ocular visual impairment, improved functioning is often seen with enhanced visual input, but for children with CVI, enhanced input often is detrimental to the use of their vision (Geruschat, 2005; Groenveld, Jan, & Leader, 1990).

Despite these documented differences in the visual skills of children with ocular versus cortical visual impairment, the functional vision assessments currently used in the field rely heavily on the needs of children with ocular visual impairment and do not take into account the unique characteristics of children with CVI. In response to this need, Roman-Lantzy developed a functional vision assessment for children with CVI, The CVI Range (Roman-Lantzy, 2007).

Functional Vision Assessment of Children with CVI

Based primarily on the findings of researchers in British Columbia (Good & Hoyt, 1989; Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985) Roman-Lantzy developed an observational instrument, The CVI Range, that specifically addresses the unique visual characteristics of children with CVI (Roman-Lantzy, 2007). The CVI Range builds on her earlier work to validate an interview instrument that could differentiate children with CVI from children with ocular visual impairment (Roman, 1996). In her original study, characteristics of children with CVI that had been identified in the literature were used to design an interview and to develop a behavioral observation protocol. Experts in the field of CVI helped to establish the

content validity of the interview questions. Results of the study indicated that the interview could successfully differentiate children with CVI from children with ocular visual impairment and there was concurrent validity with the visual behavioral observations of the infants in the study. Her study is described in detail in Chapter 2.

In 1998, she conducted further examination of the content validity of her assessment when she met with Jan and his team in Vancouver to discuss her tool and to verify the characteristics delineated on the assessment (Roman-Lantzy, personal communication, September 17, 2007). Content validity is the extent to which a measurement reflects a specific domain of content (Carmines & Zeller, 1979) and Jan and his team felt that The CVI Range addressed the critical characteristics of children with CVI. This assessment tool is used to determine the child's level of visual functioning and the effects of the various characteristics of CVI on the child's visual functioning. The CVI characteristics assessed with this tool are: color, movement, visual latency, field preferences, complexity, light-gazing/non-purposeful gaze, distance viewing, visual reflexes, novelty, and visual motor skills. (See Appendix A for a copy of The CVI Range.)

The CVI Range can be used to assess the visual skills of children with CVI and to determine the educational impact of the child's visual performance. After a thorough review of the literature (see Chapter 2), this instrument has emerged as the only functional vision assessment that is designed to specifically address the unique visual behaviors of children with CVI. Assessment results from The CVI Range can be used to develop appropriate interventions, individualized to each child's specific vision needs and to document change in vision skills across time. In addition, The CVI Range, as a

measure of progress, can document effective evidenced-based interventions for children with CVI.

Significance of Visual Interventions and Modifications for Children with CVI

Hubel and Wiesel (1970) conducted some of the first research that addressed brain development and visual skills. They demonstrated that kittens deprived of visual input to one eye for six weeks following birth remained permanently blind in that eye even when visual input was restored to that eye. No amount of visual input restored normal function to the deprived eye. From their work came the idea of critical periods in visual development (Bruer, 2001). Critical periods were defined as fixed times during which specific input was required to facilitate development in that area.

Research subsequent to their initial work demonstrated that critical period effects were not necessarily permanent or irreversible (Bruer, 2001). Chow and Stewart (1972) demonstrated that visually deprived kittens, when forced to use the deprived eye, could learn to use that eye. Harweth, Smith, Crawford, and van Noorden (1989) found that monkeys who had one eye closed during the critical period could recover near normal functioning if forced to use the deprived eye. Due to the ability of these animals to learn to use vision, even after what was considered the fixed critical period, the term sensitive period is now preferred. The core idea of sensitive periods that is accepted by most researchers is that having a certain experience at a designated point in development has a profound impact on future development in that area (Hoyt, 2003).

In addition to the idea of sensitive periods, is the idea of increased plasticity of the brain in infants and young children (Hoyt, 2003). While the exact time that is considered a sensitive period for visual development in children is not clearly defined, most

researchers agree that the early years are the time of the most plasticity (Hoyt, 2003). The fact that most children with CVI demonstrate some improvement in vision (Khetpal & Donahue, 2007) is usually attributed to plasticity and the brain's ability to develop visual functions despite damage to visual pathways and areas of the brain responsible for vision.

The potential for change in visual functioning in children with CVI makes it critical to determine the child's current visual functioning and identify the factors that could facilitate visual improvement. The CVI Range (Roman-Lantzy, 2007) is an assessment that addresses the unique visual characteristics of children with CVI and determines how each characteristic is having an impact on visual functioning. Using information gained through this assessment, professionals can provide visual experiences and modifications that capitalize on the brain's plasticity. In addition, documentation of progress could help build a foundation for interventions that are evidenced-based. For The CVI Range to provide this valuable information it needs to be a consistent or reliable instrument; however, there are currently no formal reliability data available on this test.

Importance of Reliability

Reliability is a major consideration in any assessment procedure (Nunnally, 1978; Salvia & Ysseldyke, 2007). Reliability involves the ability to measure consistently the behavior of interest. The extent to which a test or measuring procedure yields the same results on repeated trials is the extent to which we consider a test to be reliable (Carmines & Zeller, 1979). Consistent results are necessary to use assessment information in decision making for intervention and tracking child progress. Several types of reliability are critical for assessment information to be useful including: internal consistency, test-retest, and, for observational tests, inter-rater reliability (Nunnally, 1978).

Internal consistency is a measure of how well the items in a test correlate with each other. High correlations are suggestive of consistent measurement of a single construct (Carmines & Zeller, 1979). In The CVI Range, the ten characteristics of CVI are each rated (0-1) and the ratings are summed to provide a measure of the severity of CVI. Reliability as demonstrated by how well the individual items correlate with each other, will provide a measure of internal scale consistency.

A second type of reliability is test-retest. Test-retest reliability is an index of stability across time (Salvia & Ysseldyke, 2007). A behavior that is seen today should also be observed next week. If the behavior of interest involves a developmental process or learning, scores need to be consistent across a short period of time. Generally, the shorter the time between tests, the higher the test-retest reliability will be (Traub, 1994). Many researchers have described children with CVI as having variable visual functioning (Dutton, et al., 1996; Good & Hoyt, 1989; Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985). Roman-Lantzy (2007) has argued that children with CVI do not have visual functioning that varies; instead, changes in the environment may be responsible for a child's change in visual behavior. If child scores change significantly from one test situation to the next, and the testing situations are similar, then the test-retest reliability will be low. If environmental factors are controlled and child scores are similar from one test to the next, then test-retest reliability will be high. High test-retest reliability will support Roman-Lantzy's argument that variability in visual functioning is related more to environmental factors than to actual changes in the child's vision. In

Chapter 3, issues related to methodology and test-retest reliability are examined in more detail.

Another type of reliability that is important for any observational assessment is inter-rater reliability (Nunnally, 1978). For inter-rater reliability, two similarly qualified examiners must get the same or similar results on a given assessment protocol (Salvia & Ysseldyke, 2007). The CVI Range is an assessment that is completed by observation of the child's visual functioning. The scale and scoring guideline for observations must be clear enough that two people can obtain similar results when using the tool. Inter-rater reliability is a function of how well the examiners understand and consistently score visual responses. Establishing consistency across people is critical for an observational instrument such as The CVI Range.

Purpose of Study

Children identified as visually impaired under IDEA need to have a functional vision evaluation to determine how the visual impairment affects educational performance (IDEA, 2004). Once this is determined, an appropriate and individualized education plan can be developed and implemented. This plan should address the child's vision needs and how these needs relate to accessing the educational curriculum. Children with CVI have unique vision needs that are often different from children with ocular visual impairments. In response to this situation, Roman-Lantzy developed The CVI Range for conducting a functional vision assessment for children with CVI. For The CVI Range, there is preliminary content validity data (Roman, 1996; Roman-Lantzy, personal communication, September 17, 2007). At the time of this study, there was no formal reliability data available for the assessment. For this assessment to provide useful

information for teachers and parents, the assessment needs to be reliable. The purpose of this study was to examine the reliability of The CVI Range. To address reliability, the following research questions were asked:

1. To what degree does The CVI Range have good internal consistency as measured by coefficient alpha?
2. To what degree does the CVI Range have good test-retest reliability as measured by stable scores across time?
3. To what degree does The CVI Range have good inter-rater reliability as measured by having two qualified examiners obtain the same or similar score?
4. To what degree does The CVI Range have consistency across the two sections of the assessment as measured by comparing the scores on the two sections?

CHAPTER II

REVIEW OF LITERATURE

This chapter presents a review of the literature relative to cortical visual impairment. First, the definition of CVI and origination of the term will be examined, followed by a discussion of the prevalence of CVI in developed countries. Research concerning the etiology and the behavioral characteristics of children with CVI will be reviewed. Next, the current status of measurement of vision in children with CVI, including a medical model of assessment as well as functional vision assessment will be examined. Finally, outcome studies and intervention research will be reviewed.

Literature Review Search Strategies

Studies included in this literature review were found using a number of search strategies. First, a computer search was conducted of ERIC, PsycLIT, PsycINFO, Medline, PubMed, OVID, and EBSCOhost databases. In addition, a search was conducted with *The Journal of Visual Impairment and Blindness* and *Developmental Medicine and Child Neurology*, two professional peer-reviewed journals that deal with children with visual impairment and children with brain damage. The database and journal searches were conducted using various combinations of the following keywords: *blindness, cortical, visual impairment, vision disorders, cortical visual impairment, cerebral visual impairment, child, infant, and preschool.*

Another search strategy was to conduct an ancestral search of the reference lists of literature reviews and research articles written by preeminent researchers in the area of cortical visual impairment. The final strategy was to review the American Printing House

for the Blind (APH) CVI website. APH created CVI Synergy, a group of researchers, educators, and physicians who work with children diagnosed with CVI. The CVI Synergy group identifies articles and research related to CVI and the APH website is a compilation of the meetings of CVI Synergy.

Definition of Cortical Visual Impairment

Vision as a sense is associated primarily with the eye (Roman-Lantzy, 2007), however as early as the 17th century scientists became interested in the path of the optic nerve fibers (Hoyt, 2003). By the early 19th century many scientists began to suggest that vision was served by specific parts of the cerebral cortex. The term cortical blindness began to be used in the 20th century for adults who were blind due to bilateral damage to the occipital cortex (Whiting, et al., 1985). Cortical blindness was diagnosed when an adult had complete loss of vision, including light perception, but had normal pupillary responses, normal eye movements, and normal retinal exams.

Early in the history of the diagnosis of pediatric visual impairment due to cortical issues, the term cortical blindness was used. In 1985, Whiting et al. argued that acquired cortical blindness in adults may be quite different from congenital or early-onset cortical visual loss in children. Due to the nature of the immature brain and early plasticity, children with cortical visual loss may initially present and subsequently develop very differently than adults with cortical visual loss (Hoyt, 2003; Whiting, et al., 1985). Total absence of vision in children due to brain damage is extremely rare. Children with cortical visual impairment usually have some visual responses and often experience significant visual recovery; therefore, the term cortical blindness was not deemed appropriate for children (Good, et al., 1994; Hoyt, 2003; Roland, Jan, Hill, & Wong,

1986). Since that time, the term cortical visual impairment has been the preferred term for use with children (Edmond & Foroozan, 2006; Good et al., 1994; Hoyt, 2003; Jan, Groenvelt, Sykanda, & Hoyt, 1987). Although the term cortical visual impairment is preferred, it is not universally used for children with vision loss due to brain damage (Jan, Good, & Hoyt, 2006; Roman-Lantzy, 2007). Some researchers prefer the term cerebral visual impairment because damage to the brain that can cause vision loss may not always originate in the visual cortex (Dutton, 2006; Hyvärinen, 2006). In this paper, the term cortical visual impairment will be used to refer to any loss of vision due to brain damage or malformation. In the case of children with co-existing ocular conditions, cortical visual impairment is diagnosed when the severity of loss cannot be explained solely by the ocular condition (Roman-Lantzy, 2007).

Prevalence of Cortical Visual Impairment

The concept of visual impairment in young children has changed across the past 30 years (Flanagan, Jackson, & Hill, 2003). There has been a decrease in the number of children with isolated visual impairment and an increase in the number of children with coexisting neurological disability. Medical technology has improved perinatal care and resulted in increased survival rates of very premature and very sick term infants (Khetpal & Donahue, 2007). Concurrent with this increased survival rate is the emergence of cortical visual impairment (CVI) as a major cause of visual impairment in young children from developed countries.

The Blind Babies Foundation of Northern California was one of the first agencies in the United States to establish a database of clients they served (Blind Babies Foundation, 1995). In 1994, a data base was established and data from over 1,200 client

files were entered. The files included children with birth dates between 1980 and 1995 and contained information on cause of visual impairment. Cortical visual impairment was the most frequent cause of visual impairment with over 30% of all of the children they served diagnosed with CVI (Blind Babies Foundation, 1995).

In 1995, the Model Registry of Early Childhood Visual Impairment Consortium was established with support of the Hilton-Perkins Program (Hatton, 2001). The purpose of the Consortium was to develop and implement a model registry of children from birth to 3 with visual impairments. Nine states (Alaska, Arizona, California, Colorado, Iowa, Massachusetts, New Mexico, North Carolina, and Utah) participated and collected data from 1998 to 1999. The most prevalent visual condition reported was CVI (26%).

Based on the Model Registry developed by the Consortium, a national registry, Babies Count, was developed by American Printing House for the Blind (Hatton, Schwietz, Boyer, & Rychwalski, 2007). To date, 26 states have submitted data on children birth to 36 months with visual impairment. Again, cortical visual impairment is the most prevalent visual condition reported (24%).

In addition to data from the United States, a number of other countries have collected and analyzed data about children with visual impairment. In the Republic of Ireland a national survey of visually impaired children under 16 years of age was conducted. Between July 1989 and June 1990, 172 children were examined by an ophthalmologist. Of these children, 27 (16%) were diagnosed with cortical visual impairment (Goggin & O'Keefe, 1991). Flanagan, Jackson, and Hill (2003) used multiple hospital and community sources to examine the characteristics of children with visual impairment in South and East Belfast in the year 2000. Seventy six children were

identified and 34 (45%) were diagnosed with CVI. Khan, O'Keefe, Kenny, and Nolan (2007) reviewed data from 1990-2004 from ophthalmology departments and the National Council of Blind and Visually Impaired in Ireland. They divided the data into categories (i.e., genetic, prenatal, perinatal, and childhood) based on the age of the child at the time of vision loss. In the perinatal group, CVI was the largest single cause of visual impairment. They noted that the most significant trend across the 14 years was the decrease in blindness due to ROP and the increase in visual impairment due to cortical issues.

In Great Britain, Fleck and Dangata (1993) examined all 93 children attending the Royal Blind School during the 1991-92 academic school year. Children with cortical visual impairment represented 26% of the school population. Rogers (1996) examined the database of the Liverpool vision assessment team. The data base included children ages birth to 16 who were identified as visually impaired in 1995. Of the 199 children identified 130 (65%) had additional disabilities. Of those, 64 (49%) had cortical visual impairment. This represents approximately one-third of the total population with CVI. Rahi and Dezateux (1998) reviewed data in Great Britain relative to the prevalence and causes of visual impairment in Britain. They indicated that the principle causes of serious vision loss were congenital cataract, cortical visual impairment and optic atrophy. Exact percentages were not reported.

Rosenberg, et al. (1996) completed a study of the national registers for the blind during 1993 from 5 Nordic countries (Denmark, Iceland, Finland, Norway, and Sweden). National registers represent the population of children ages birth to 17 who are registered

as visually impaired. Brain disorders accounted for approximately 45% of the cases of visual impairment (Rosenberg, et al., 1996).

In summary, during the last 20 years vision loss due to brain damage has been recognized as a significant cause of vision impairment in children. This condition is now known as cortical visual impairment. Prevalence data that are available from developed countries indicate that CVI is now the single leading cause of vision impairment in children. This is due in part to improvements in medical technology that have increased the survival rates of children who experience brain damage and in part due to improvements in medical technology that have improved outcomes for children with ocular problems. In the next section, research about the causes of CVI will be reviewed. Appendix B contains a summary of the studies reviewed that relate to the causes of CVI. Appendix C contains a summary of the studies reviewed relative to the relationship of specific brain injuries and CVI in preterm and full term children.

Causes of Cortical Visual Impairment

Some of the earliest work in CVI came out of the Visually Impaired Program at the Children's Hospital, British Columbia. Whiting, et al. (1985) examined the records and testing data of children seen at the clinic from 1970 – 1984. From those records, 50 children with permanent CVI were identified. Etiology was divided according to time of damage: prenatal, perinatal, and acquired. Perinatal was defined as the period within the first 28 days of life. These researchers identified the primary causes of CVI during the prenatal period as toxemia, intra-uterine infection, and cerebral dysgenesis. During the perinatal period the primary cause was asphyxia, followed by hemorrhage and

meningitis/encephalitis. Acquired CVI was caused by shunt malfunctions, trauma, meningitis, cortical vein thrombosis, and cardiac arrest (Whiting, et al., 1985).

Jan, Groenvelde, Sykanda, and Hoyt (1987), also of the Visually Impaired Program at the Children's Hospital, British Columbia studied 50 children seen in the program between 1983 and 1985. They divided the etiology into two categories based on age of the child at time of insult: prenatal/perinatal and acquired. The primary cause of CVI during the pre- and perinatal time period was asphyxia, followed by cerebral dysgenesis, cerebral hemorrhage, and infection. For CVI acquired after the perinatal period the causes were, in decreasing order of numbers of children: shunt failure, asphyxia, injury, and dehydration. In a study of light gazing in children with CVI, Jan, Groenvelde, and Sykanda (1990) examined the records and assessment data of 69 patients with CVI seen in their clinic from January 1987 – May 1989. The major causes of CVI were asphyxia, followed by central nervous system anomalies, injury, and infection.

Huo, Burden, Hoyt, and Good (1999) reviewed the records from 1979-1994 in a large pediatric ophthalmology practice in California. From these records, 170 cases of children diagnosed with CVI were identified. The most common causes of CVI were, in order of prevalence: perinatal hypoxia, cerebral vascular accident, meningitis and acquired hypoxia (Hoyt, 2003; Huo, Burden, Hoyt, & Good, 1999).

Most recently, Khetpal and Donahue (2007) examined the records from patients visiting the Vanderbilt University Pediatric Ophthalmology Center, Tennessee, from 2002 to 2005. Ninety eight children were identified as having CVI. The most common etiologies were perinatal hypoxia, prematurity, hydrocephalus, structural central nervous system abnormalities, and seizures.

Specific Brain Injury and CVI

Premature infants are especially vulnerable to brain injury and possibly subsequent vision issues. Some of the common complications of prematurity that may result in vision impairment include: intraventricular hemorrhages, cerebral infarcts, and periventricular leucomalacia (Pike, et al., 1994). Intraventricular hemorrhage (IVH) involves bleeding into ventricles of the brain thought to be due to changes in blood flow or perfusion in the brain cells. The lack of blood flow results in cell death and subsequent breakdown of the blood vessel walls, leading to bleeding (Hoyt, 2003). IVH is graded I-IV, according to severity. Excessive bleeding can cause further brain damage. Cerebral infarcts are any focal area of bleeding and cell death usually also caused by hypoperfusion. In premature infants, dehydration or swings in blood pressure can cause focal infarcts (Hoyt, 2003). Periventricular leucomalacia (PVL) results from hypoxic ischemic damage of the white matter surrounding the ventricles in the brain (Jacobson & Dutton, 2000). Pike, et al. (1994) studied 42 preterm children followed at Hammersmith Hospital, London, England. Children were divided into three groups: children with PVL; children with a large IVH, Grade II-III; and, children with cerebral infarction. They found that visual impairments were more common with ischemic lesions (PVL and infarcts) than with hemorrhagic ones. However, 38 of the 42 children showed some impairment in one or more aspects of visual functioning, so they cautioned that each child needs individual assessment.

Jacobson, Ek, Fernell, Flodmark, and Broberger (1996) also studied children with PVL. The 13 children in their study were born between 1980 and 1989, were between 4 and 14 years of age during the study, and were all diagnosed with PVL. All children

presented with visual impairment with or without visual perceptual difficulties. The children often demonstrated greater visual difficulties than would have been predicted by visual acuity scores alone (Jacobson, et al., 1996). They concluded that children with PVL should be monitored closely for signs of visual difficulties.

Lanzi, et al. (1998) conducted a study of children with cerebral palsy (CP) due to PVL to determine the presence of CVI in children with CP due to PVL. Their subjects were 38 children, ages 20-66 months, who were born prematurely, and who had diagnoses of CP and PVL. The children were born between 1992 and 1996. Severity of PVL was determined by neuroimaging (i.e., brain MRI). The results of the study were that CVI occurs frequently in children with CP due to PVL (66%). In addition, the severity of PVL was correlated with severity of vision loss (Lanzi, et al., 1998).

Cioni et al. (1997) also conducted a study of preterm infants with PVL to determine the frequency of CVI and to correlate the severity of visual deficit with the severity of PVL. Their subjects were 14 children with severe PVL and 34 with moderate PVL born between 1990 and 1994. The results of the study were consistent with Lanzi et al. (1998). Children with severe PVL were more likely to have CVI; however, there were 3 of the 14 children with severe PVL who had normal vision at one year. Children with moderate PVL were less likely to demonstrate visual impairment at one year. Because vision outcome could not be predicted in individual cases, they concluded that all children with PVL should be monitored for visual impairment (Cioni, et al., 1997).

Mercuri et al. (1997) studied the effects of hypoxic-ischemic events in full term infants. Thirty one infants diagnosed with hypoxic-ischemic encephalopathy (HIE), born between 1991 and 1996, were included in the study. Infants were included if they were

full term and demonstrated neurological abnormalities within the first 48 hours of life. The purpose of the study was to determine if the degree or site and size of HIE could predict visual outcome. Twenty of the 31 infants demonstrated abnormal visual results. Visual outcome was worse with more severe HIE, however, visual outcome could not be predicted for individual children based only on HIE (Mercuri, et al., 1997).

Brodsky, Fray, and Glasier (2002) examined the records of 100 children seen at Arkansas Children's Hospital Eye Clinic between 1989 and 1999. In this study they compared the visual impairment of 50 children with cortical loss (predominately cortical gray matter) and 50 children with subcortical loss (predominately subcortical white matter or PVL). They found differing profiles of vision function between the two groups. They concluded that the developing brain's level of maturity at the time of injury affects the part of the brain that is injured and the subsequent visual outcome. For preterm infants, the injury tends to affect white matter (i.e., PVL). For term infants the injury tends to affect the cortical gray matter. The actual mechanism of injury is thought to be related to the differing watershed zones in the two groups of children. The watershed zone is the area at the end of the vascular system that is most susceptible to injury from lack of oxygen. Lack of oxygen precipitates a decrease in systemic blood flow which leads to a loss of autoregulation of cerebral blood flow and decrease in perfusion of the brain (Hoyt, 2003). In the preterm brain, the watershed zone is in the subcortical white matter and in the full term child the watershed zone is in the cerebral cortex; hence the differing patterns of brain injury due to lack of oxygen.

Hoyt (2003) also conducted a similar study comparing the records of 96 children with CVI, 41 with damage in the visual cortex and 26 with damage in the periventricular

white matter (i.e., PVL). The children included in his study were seen in a pediatric ophthalmology practice in San Francisco between 1979 and 1994. The purpose of his study was to compare the visual outcomes and recovery of the two groups of children. The average length of time that the children were followed was 5.9 years. In general, the children with PVL had less improvement in vision than children with damage in the visual cortex.

Summary and Methodological Issues

From the early research done in British Columbia in the 1980s to the most recent work at Vanderbilt in 2000-2004, asphyxia/hypoxia clearly emerged as the most frequent cause of CVI in the pre- and perinatal period for both preterm and full term children. While lack of oxygen can cause vision problems in any child, a number of researchers have documented that lack of oxygen affects the newborn brain differently depending on the child's gestational age (Brodsky, Fray, & Glasier, 2002; Hoyt, 2003). Other frequent causes of CVI include: shunt malfunctions, trauma, meningitis, infections, and central nervous system abnormalities.

All of the studies that addressed causes of CVI were accomplished by retrospective review of medical records in large ophthalmology clinics (e.g., Visually Impaired Program of Children's Hospital, British Columbia; Vanderbilt University Pediatric Ophthalmology Center, TN). Researchers reviewed eye medical records for all children seen during a specified time span of several years and identified those children diagnosed with CVI. From those records they reviewed other medical information, such as brain imaging results, to determine the causes of CVI. Several researchers examined the relationships between specific brain injuries and CVI. Two of those studies were also

retrospective studies accomplished through review of medical records (Brodsky, Fray, & Glasier, 2002; Hoyt, 2003). The remaining studies were prospective studies conducted in medical facilities. Subjects were recruited from children who were seen in a follow up clinic across several years. Researchers set inclusion criteria (e.g., child with PVL or HIE) and subsequent children were included who met their descriptive criteria. In the next section, the visual characteristics of children with CVI will be described. A summary of the results of studies that describe the characteristics of children with CVI can be found in Appendix D.

Characteristics of Children with Cortical Visual Impairment

Jan and his colleagues in British Columbia were among the first researchers to document the characteristics of children with CVI. In a series of studies (Good & Hoyt, 1989; Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985) across a 20 year span, they systematically documented characteristics of children with CVI. The children ranged in age from 6 months to 19 years with a variety of causes of CVI including: perinatal hypoxia, injury, infection, central nervous system abnormalities, and trauma. In summary, the characteristics of children with CVI that they identified included:

- Children looked towards moving objects; and, children who were mobile did not bump into objects.
- Children did not look blind.
- Children were visually inattentive and lacked visual curiosity.
- Children did not have nystagmus.

- There was an absence of visual self-stimulation (e.g., eye-pressing, flickering fingers in front of eyes).
- Some children stared into lights and a few children were photophobic.
- Children exhibited variable visual functioning.
- Children often supplemented vision with touch, but looked away when they touched or reached for an object.
- Some children could identify color, but not objects; or color but not shape.
- Many children had visual field loss.
- Children often recognized familiar objects in one environment but not in another, and recognized familiar objects more than novel ones.
- Children had difficulty with distance vision and usually viewed objects close.
- All children had other neurological problems (e.g., developmental delay or mental retardation, CP, seizures, hearing loss).
- Many children had co-existing ocular impairment, usually optic nerve atrophy (Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985).

Jan, Groenveld, and Sykanda (1990) did further research into light gazing by children with CVI. Light gazing was defined as compulsive staring into lights for longer than 15 seconds. They studied children with CVI evaluated by the Visually Impaired Program of British Columbia, Canada, between the years of 1987-1989. Of the 69 children with CVI, approximately 60% were light-gazers and the most multiply disabled

children were the most likely to gaze at lights. Jan, Groenveld, and Anderson (1993) studied photophobia in children with CVI. Photophobia was defined as visual discomfort in normal lighting conditions. They studied children during the years between 1987 and 1991 and found that approximately one-third of the children demonstrated photophobia. In most cases it was mild and diminished with time. Some children who were photophobic were also light gazers.

Dutton (2003, 2004, 2006; Dutton et al., 1996) has also done extensive research on the characteristics of children with CVI. At the Vision Assessment Clinic in Glasgow, Scotland, Dutton, et al. (1996) examined 90 children with CVI between the years 1992-1994. Visual functioning ranged from no evidence of vision in 16 children to children with only mild acuity loss. In children with some evidence of vision but significant cognitive disabilities, he noted variable visual functioning that was often affected by fatigue and other distractions, as well as better visual functioning in familiar environments.

In 20 children where there was only a mild acuity loss, Dutton et al. (1996) documented complex disorders of cognitive vision. Acuity is the ability to distinguish details and cognitive vision involves visual tasks that require analysis and interpretation of the visual world (Dutton, et al., 1996). Cognitive vision disorders included impairment in: recognition, orientation, depth perception, perception of movement, and simultaneous perception. Difficulties in recognition included recognition of objects and people. Difficulties in orientation involved problems getting lost, especially in new environments or difficulties finding where objects were left. Problems with depth perception included skills such as distinguishing a line on the floor from a step. Impaired movement

perception involved difficulties seeing moving objects. Impaired simultaneous perception involved not being able to see more than one part of a whole, e.g., not seeing more than one object or picture when multiple objects or pictures were presented. He concluded that children with CVI have a range of visual functioning and even when visual acuity numbers would not indicate poor vision, a child with CVI may have complex visual difficulties that cannot be explained by acuity alone (Dutton, et al., 1996).

Based on these early observations, Dutton developed a model of how the visual system works and the effects of damage to the different visual pathways (Dutton, 2003, 2006; Dutton & Jacobson, 2001). He used the term “cognitive visual disorder” (Dutton, 2003, p. 290) to refer to problems with misinterpretation of the visual world either with respect to where things are or what things are. He described two primary visual pathways that affect different areas of visual functioning. Visual information is primarily processed in the occipital cortex where it is passed to two principal locations, the dorsal and ventral streams. The two streams are closely interlinked.

The dorsal stream links the visual cortex with the posterior parietal lobes. The posterior parietal lobes process an entire visual scene and help to focus attention to details of interest and aid in planning motor actions. Dysfunction in the dorsal stream (most often due to PVL) can lead to difficulties handling complex visual scenes such as finding a toy among many toys or finding an object at a distance (which includes more information to sort out). This inability to sort out part of a whole can also lead to problems with crowding of text when a child is learning to read. Dorsal stream dysfunction can also lead to difficulties moving through space. Stairs, curbs, and other environmental features are difficult for a child with dorsal stream dysfunction (Dutton,

2003, 2006; McKillop & Dutton, 2008). Lower field loss is common, leading to further difficulties with moving through space.

The ventral stream links the visual cortex with the temporal lobes. The temporal lobes are responsible for recognition of visual information. Ventral stream dysfunction is associated with difficulty in recognizing faces, as well as difficulties in recognizing shape or form. A child who is learning to read may have difficulties recognizing letters and words (Dutton, 2003, 2006; McKillop & Dutton, 2008). Lack of recognition of objects in the environment makes getting around without getting lost a challenge for children with ventral stream damage.

The dorsal stream and ventral stream work closely together as a child attempts many visual tasks throughout the day. The child sees and recognizes using ventral stream functions and reaches out and picks up objects using dorsal stream functions. However, when brain damage occurs, specific parts of the task may be more difficult and it is often hard to understand why a child with CVI seems to sometimes see and respond and other times not (Dutton, 2003, 2006; McKillop & Dutton, 2008). He concluded that comprehensive vision assessment of multiple visual functions in addition to traditional acuity measures may help to sort this out.

Jacobson et al. (1996) examined a group of children ages 4-14 who were diagnosed with PVL and had subsequent CVI. They documented the following characteristics:

- Acuity was worse with symbols in a line instead of in isolation (crowding effect).
- Visual fields were restricted in all children.

- The children had normal color vision.
- Most children had normal contrast sensitivity.
- Many children had difficulties identifying forms and often guessed from parts of an object or used color cues.
- Children tired easily with visual tasks.
- Many children also had optic nerve atrophy.

Pike, et al. (1994) described the vision in 42 children born preterm with either PVL, cerebral infarct, or IVH. They noted the following characteristics of children with CVI: color vision is preserved in children with CVI, children have more difficulties with complex pictures than with single symbols, and crowding is an issue for most children. Groenendaal and van Hof-van Duin (1992) tested a group of 38 children with CVI and noted absence of visual threat response in approximately one-third of the children, field deficits in children that could be tested, no fixations in half of the children, and reduced acuity in children that could be tested. Cohen-Maitre and Haerich (2005) conducted a study to evaluate the effects of color and movement on visual attention in children with CVI. Both color and movement were salient features in getting and maintaining visual attention in children with CVI. Movement was preferred over color and movement plus color was the most powerful stimulus.

Summary and Methodological Issues

With one exception, all of the studies that described characteristics of children with CVI were done in large medical ophthalmology clinics. The studies were retrospective reviews of medical records. Cohen-Maitre and Haerich (2005) conducted an experimental study to determine the differential effects of movement and color on visual

performance. They did not describe the severity of CVI in their sample of children. Despite prior research that describes children with CVI as functioning with mild impairment to near blindness, no information was given as to the level of visual functioning of the children who participated. While movement and color emerged as salient features in visual attention, without some knowledge of the level of visual impairment, it is difficult to determine to whom the results would generalize.

Consistently in the research across the past 20 years several characteristics of children with CVI have emerged. Children with CVI have difficulties with complex visual input, often seen in crowding or difficulties with complex environments or pictures. Sometimes this issue with complexity is seen in difficulties with distance viewing. Often visual functioning seems to be variable. Children with CVI respond to color and often to movement although some have difficulties seeing a moving object. Children respond better in familiar settings or with familiar objects. Often visual inattention or light gazing is seen. Almost all children have field deficits. Some children cannot look and touch at the same time, while for others touch helps them to identify an object. Many children cannot recognize objects. These characteristics have emerged consistently in children with CVI from a variety of causes (e.g., PVL, HIE, IVH, seizures, injury, and infections), in children with varying levels of additional disabilities (e.g., mild to severe cognitive and motor disabilities), and across a wide age span (ages 6 months to 19 years). In the next section research about assessment of children with CVI will be examined.

Measurement of Vision in Children with Cortical Visual Impairment

Measurement of vision in children with CVI is often a challenge (Morse, 1992). There are two major types of information that are of interest to professionals working with children with visual impairments. First, testing is needed that confirms a diagnosis of CVI and determines the level of visual impairment. Appropriate medical documentation of a visual disability is usually needed to access appropriate vision services. Second, beyond a diagnosis, professionals are interested in functional measures of vision. How does the child use vision in daily tasks? Functional assessment of vision should guide intervention and provide a measure to assess the effectiveness of that intervention (Roman-Lantzy, 2007).

Medical testing to confirm diagnosis. Most researchers and ophthalmologists use several types of neuroimaging to document brain damage or central nervous system malformations. The most often used are computed tomography (CT scan) (Dutton, et al., 1996; Jacobson, Ek, Fernell, Flodmark, & Broberger, 1996; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting et al., 1985); magnetic resonance imaging (MRI) (Brodsky, Fray, & Glasier, 2002; Khetpal & Donahue, 2007; Lanzi, et al., 1998); and, ultrasound (Cioni, et al., 1997; Mercuri, et al., 1997; Pike, et al., 1994).

One of the most common electrophysiologic tests of vision is the visual evoked potential (VEP). In a VEP, the child is shown a series of stripes or checkerboard patterns on a lighted screen and brain activity over the occipital cortex is measured. Sometimes a bright flash of light is used as a visual stimulus (flash VEP). If there is no brain activity during the presentation of the visual stimulus, the assumption is that the child cannot see the stimulus. Whiting et al. (1985) used visual evoked potential mapping (VEPM) to

measure vision. While VEP measures response to flashes of light in the visual cortex, VEPM measures response to light over large portions of the brain and is displayed as a multicolored moving picture depending on the degree of electrical activity in that part of the brain. The VEPM was used to diagnose CVI and results of the VEPM were more accurate in identifying children with CVI than VEP alone (Whiting, et al., 1985).

In addition to documentation of brain damage or brain responses (e.g., VEP), documentation of vision loss is critical to the diagnosis of CVI. In documentation of vision loss, Colenbrander (2006) suggested a distinction between vision functions and functional vision. Vision functions describe how the eye functions and describes skills such as fixation, tracking, visual fields, and various acuity measures. Visual functioning describes how the child functions in vision-related tasks and includes measures such as using vision to orient in the environment and using vision to recognize and manipulate objects. The following sections will look at how various researchers have attempted to measure vision functions as well as functional vision. A summary of testing of vision functions in children with CVI can be found in Appendix E.

Assessment of vision functions. The most common vision function measured is acuity or the ability to discriminate and recognize detail (Teplin, 1995). The standard method of measuring acuity in a literate child is to have the child read letters from a Snellen chart with letters of standardized sizes from a set distance (Teplin, 1995). This type of acuity is called an otype acuity. Normal otype visual acuity for an older child or adult is 20/20. The denominator is the distance at which a person with normal vision can read the target letters. The numerator is the distance that the tested individual can

read the letters. For example, an acuity of 20/200 means that the tested individual can read the letters at 20 feet that a person with normal vision can read at 200 feet.

For very young children, or children with significant disabilities who cannot identify letters or shapes, other methods of determining acuity must be used. One of the most common tests is the visual evoked potential (VEP). Acuity is determined by the smallest stripes or checkerboard pattern to which the child has a response (Odom, et al., 2004). The acuity measure from response to black and white stripes or checkerboards is called grating or resolution acuity (Cavallini, et al., 2002). Grating acuity is the child's ability to perceive separate elements of a stimulus.

VEP testing is often used with children with CVI. Granet, Hertle, Quinn, and Breton (1993) studied the use of flash VEP to predict vision improvement in children with CVI. For 10 children ages 5-48 months, all with initial abnormal flash VEP results, the initial testing did not predict which children would show improvements at follow up (2 to 30 months after initial testing). Clarke, Mitchell, and Gibson (1997) also examined the use of flash VEP in assessment of children with CVI. They studied 44 children and performed a flash VEP at initial and follow up visits. There was no information about the ages of the children, or the length of time of follow up. They found a marginal relationship between initial VEP and improvement at follow up. A normal VEP was predictive of a more positive outcome; however, an initial abnormal VEP was no better than chance in predicting outcome.

Good (2001) used a sweep VEP procedure to measure the vision of children with CVI. Sweep VEP is similar to VEP in that it involves the measurement of brain activity, but the stimulus is moving black and white lines. He tested 41 children with CVI ages 6

months to 16 years with a sweep VEP procedure, and retested 23 for test-retest reliability. He conducted the testing under low and normal background luminance conditions. He compared the results to normative data as well as a clinical rating of vision for each child. He concluded that sweep VEP is a reliable and valid measure of vision in children with CVI. He also noted that some children with CVI performed better under low luminance conditions.

Good and Huo (2006) repeated the study of sweep VEP under two luminance conditions with 20 children (ages 7 months to 4 years) with CVI and 17 age-matched control subjects. They found that children with CVI had improved grating acuity under low luminance conditions. In the control group, luminance had no effect on the children's responses. They concluded that this finding had implications for optimal viewing and learning conditions for children with CVI.

Skoczenski and Good (2004) examined yet another type of acuity with a sweep VEP procedure. They examined vernier acuity in a group of children with CVI. Vernier acuity is the ability to localize pattern elements or to detect a discontinuity in a line or a misalignment in a segment of a line (Skoczenski & Norcia, 1999). They tested 35 children ages 4 months to 16 years (mean: 3.5 years) diagnosed with CVI using two different stimuli for the sweep VEP. The first stimulus was the standard black and white stripes used in determining a grating acuity. The second stimulus was lines with vernier offsets that appeared and disappeared. The offsets were of smaller and smaller magnitude. They found that vernier acuity was lower than grating acuity in children with CVI. They concluded that vernier acuity is cortically mediated and may be a more sensitive measure to quantify vision deficits in children with CVI.

Watson, Orel-Bixler, and Haegerstrom-Portnoy (2007) used sweep VEP as a quantitative measure of visual acuity to document progress in children with CVI. They had assessment and follow up data on 34 children with CVI. The age of the children ranged from 1 to 16 years (mean: 5 years), and the time between measures was 0.6 to 13.7 years (mean: 6.5 years). They documented significant improvement in vision in approximately half of the children and concluded that the sweep VEP provided a quantitative measure of vision that could be used to document progress in children with CVI.

In summary, VEP is often one of the tests administered to children with CVI or suspected CVI; however, VEP results often do not predict future visual functioning (Clarke, Mitchell, & Gibson, 1997; Granet, Hertle, Quinn, & Breton, 1993). Good (2001) found sweep VEP to be a useful quantitative measure of vision in children with CVI, and one study indicated that sweep VEP could be used to document progress in children with CVI (Watson, Orel-Bixler, & Haegerstrom-Portnoy, 2007). Despite the frequency of use, there are some drawbacks to the use of electrophysiologic testing. The procedures are expensive, time consuming, require highly specific medical equipment, and can only be administered in a medical facility (Jan & Groenveld, 1993).

Another acuity measure often used with young children or children with developmental disabilities is the Teller Acuity Card procedure (Cavallini, et al., 2002; Mash & Dobson, 1998; Teller, McDonald, Preston, Sebris, & Dobson, 1986). Teller Acuity Cards (TAC) are based on the observations that typical newborns and infants, when given visual stimuli (black and white stripes) and a non-patterned stimuli of equal luminance, will prefer to look at the patterned stimuli. The preference is noted by the

child's gaze and/or head turn towards the patterned stimuli. The patterned cards consist of black and white stripes of smaller and smaller widths. The widths are measured in cycles per degree or the number of stripes per degree of visual angle (Teller, et al., 1986). Normative data on full term infants (Cavallini, et al., 2002) have been established and are available for comparison of children with suspected visual impairments. Teller Acuity Cards are a measure of resolution acuity, or the ability to perceive the separate elements of a stimulus (Cavallini, et al., 2002) and results are reported as a grating acuity. A number of researchers have used Teller Acuity Cards or some similar preferential looking technique to attempt to measure acuity in children with CVI (Brodsky, Fray, & Glasier, 2002; Dutton et al., 1996; Jan, Groenveld, & Anderson, 1993).

Some of the first researchers to examine the use of preferential looking techniques in children with CVI were Birch and Bane (1991). They examined 132 children, ages birth to 12 years, diagnosed with CVI. They had follow up data for 62 of the original sample. The purpose of their study was to evaluate the concurrent and predictive validity of preferential looking acuity estimates for children with CVI. Each child's score was compared to the mean acuity of age-matched peers. In addition, they performed an ophthalmology exam and noted the child's ability to fix and follow. Their results were that preferential looking acuity co-varied with the child's ability to fix and follow; and, that there was a positive correlation between initial and follow up preferential looking acuities. One limitation that they noted was that some children did not show consistent responses to the stimuli.

Van Hof-van Duin et al. (1998) conducted a longitudinal study of children at risk of CVI. They tested 39 children, (7 born at term with a history of HIE and 32 born

prematurely with a history of PVL or IVH) and used TAC procedures at 1-2 years of age to predict visual impairment at 5 years of age. They also examined ultrasound and MRI tests. The results were that 21 of the 39 children had some degree of visual impairment at 5 years of age. TAC scores at 1 to 2 years of age were accurate in predicting outcome for 27 of the 39 children, however not for the remaining 12. They cautioned that, on an individual basis, TAC scores may not predict future outcome and each child at risk of CVI needs ongoing assessment.

Westall, Ainsworth, and Buncic (2000) compared the results of VEP and Teller Acuity Card (TAC) procedures in children with cortical and ocular visual impairments. They reviewed the charts of 175 children ages 3 months – 13 years (mean: 13.8 months) who were referred for visual acuity testing to the Visual Electrophysiology Unit of a Children's Hospital in Ontario, Canada. Children included in the study were required to have both TAC and VEP testing as well as a complete eye exam. Their data showed that in 48% of the children there were discrepancies between the two scores. Children with developmental delay usually had poorer TAC acuity scores than VEP acuity scores and the more severe the disability, the more likely there was to be inconsistency between the two scores. They concluded that visual motor and attentional factors may contribute to a poorer TAC score for children with significant disabilities. For longitudinal studies and studies of progress, they suggested that measures of acuity need to be consistent across time or a comparison is not a valid measure of progress. This is especially true when measuring progress for individual children with significant disabilities. These researchers did not analyze the results by CVI versus ocular impairments, nor did they look at

children who had a combination of ocular and cortical impairments such as CVI with optic nerve atrophy.

Weiss, Kelly, and Phillips (2001) examined 31 children born full term who were visually unresponsive despite normal medical eye exams. They used TAC and VEP with brain neuroimaging with children who were developmentally normal (14 infants less than 1 year of age) and developmentally delayed (17 infants less than 1 year of age). The developmentally normal infants all had normal VEPs and TAC scores. They were diagnosed with visual inattention. The developmentally delayed infants all had abnormal VEPs and abnormal neuroimaging. Acuity scores with TAC ranged from normal to no response to TAC. These children were subsequently diagnosed with CVI.

Lim, et al. (2005) studied the development of acuity in children born at term with a history of HIE. They examined 19 children ages 6 months to 6 years who had both TAC and VEP measurements at the same session on at least one date, and who had acuity measured (by either method) at more than one session. In almost all children both TAC and VEP acuities were below normal for their age with TAC acuity scores below VEP acuity scores. Lower TAC scores are also typical in children with normal vision. At follow up, all demonstrated improvement in acuity but with a rate of improvement lower than normal. In many children there were substantial discrepancies in TAC and VEP scores. They concluded that acuity increases for many children with CVI, but at a slower rate than for children with normal vision.

Stiers, Vanderkelen, and Vandenbussche (2004) studied the difference in grating and otype acuities in children with ocular and cortical visual impairment. The subjects were 81 children ages 5-24 years attending a special school for children with visual

impairment in Belgium. For grating acuity they used a preferential looking technique, similar to TAC. For ototype acuity they used the Landolt-C instead of Snellen letters. The Landolt-C is presented with the gap in the "C" in one of four directions (i.e., up, down, left, or right) and the child indicates the direction of the gap by pointing or verbal response. Overall, for all of the children grating acuity was better than ototype acuity. The lower the ototype acuity, the bigger the discrepancy in the two scores. The greatest discrepancies were seen in children with brain abnormalities and a diagnosis of CVI. They concluded that the complexity of the response required for ototype acuity could have an effect on the scores. Otype acuity requires recognition and response to a stimulus. Grating acuity only requires awareness of a stimulus.

There are two other measures of acuity that are occasionally used in testing children with CVI. Optokinetic nystagmus (OKN) is a gross measure of the child's awareness of a moving black line. OKN is based on the fact that a child's eyes tend to follow or track the motion of one line at a time in a steadily moving display. As the tracked line moves out of sight, the eyes will "snap back" to fixate and follow another one (Salamanca & Kline, n.d.). The examiner spins a large cylinder with black and white stripes in front of a child. They look for the movement of the eyes and the shift back to follow subsequent lines (nystagmus) to determine if the child is aware of the visual stimulus. OKN is usually part of a battery of assessments of vision functions.

Groenendaal and van Hof-van Duin (1992); Cioni, et al. (1997); and, Brodsky, Fray, and Glasier (2002) all used OKN as one measure of visual awareness in children with CVI. Results are reported as normal or abnormal OKN responses in children with CVI.

The Stygar ball test involves presenting a series of balls of various sizes and noting the smallest ball the child responds to and at what distance (Gould & Sonksen, 1991). Again, this test is often used as a gross measure of acuity for children who do not respond to two dimensional materials; however, the test relies on the child's ability to follow a moving target. Groenendaal and van Hof-van Duin (1992) used the Stygar ball test as part of a battery of tests with children who had experienced perinatal hypoxia. Results were not reported for that test. Pike et al. (1994) also used the Stygar ball test for children who could not respond to standard acuity measures.

Visual field deficits are common in children with CVI (Jacobson, Ek, Fernell, Flodmark, & Broberger, 1996; Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985). There are two methods of testing visual fields described in the research on children with CVI. The first method is an arc perimetry as used by Cioni et al. (1997). The arc perimetry is a device that the child sits behind. It has two bands shaped in an arc going left/right and up/down. The child maintains fixation on a target centrally and lights are presented along the arc to determine the child's field of vision to the left, right and in upper and lower fields. Jan, Groenveld, and Anderson (1993) also used this method to determine field losses.

The second method of testing for visual field deficits is called confrontational testing. When the child is fixated on a target, other targets are presented in upper, lower, left and right fields. The examiner notes if the child notices the object presented in the periphery and in which fields. Dutton et al. (1996); and, Jacobson et al. (1996) used the confrontational method to determine possible field loss.

Finally, in addition to more formal assessment of vision functions, most researchers use informal observations of various other vision functions. Dutton et al., (1996) made note of each child's ability to fix and follow, reach near objects, and show awareness of faces. In addition to acuity by preferential looking tests, Jan, Groenveld, and Anderson (1993) made note of each child's response to bright objects. Cioni et al. (1997), rated the following: eye contact, presence of nystagmus, child's ability to fix and follow, convergence (eyes turning in to view close objects), eye alignment, and blink to threat reflex. Brodsky, Fray, & Glasier (2002) used behavioral responses to light and moving objects when children could not respond to TAC acuity procedures.

Issues in testing vision functions. Many of the tests of vision functions, especially electrophysiological testing such as VEP involves the use of very expensive and highly specialized equipment. The tests can only be administered in a medical setting by specially trained personnel. In addition, the measures only provide information about grating acuity or the ability to distinguish lines. The Teller Acuity Card procedure provides a measure of grating acuity that is less expensive and easier to administer. When children with CVI are tested with a grating acuity measure, the results can be compared to age-norms that are available for both TAC and VEP. Grating acuity, when compared to age norms, may be useful for quantifying vision loss to document eligibility for educational services, but does not provide any information about how the child uses vision in the context of daily activities. A final limitation of grating acuity scores are that they cannot be converted to ototype equivalents (e.g. 20/200) because grating acuity only involves awareness, and not recognition. Grating acuities are usually higher than ototype acuity measures for the same child (Hyvärinen, 2006).

Other visual awareness testing, such as OKN or Stygar ball testing, can also provide comparisons to normative data, but only provides a gross estimate of awareness of contrast and movement. These data do not provide any information about how the child uses vision in functional activities. Visual field testing provides a useful measure of where the child is aware of objects or people, and can be used to document field losses. This information has educational implications and applications in terms of placement of materials, or seating of the child relative to instruction. However field testing is just one aspect of how a child uses vision and additional educationally relevant testing is needed to adequately provide for all of the child's vision needs.

In summary, assessment of vision functions in children with CVI includes various measures of acuity including electrophysiologic testing (e.g., VEP), preferential looking (e.g., Teller Acuity Cards), optokinetic nystagmus (OKN), and the Stygar ball test. Testing of visual fields is done by arc perimetry or confrontational fields. And finally, most researchers make note of each child's ability to fix and follow and to respond to objects and faces. Another important measure of vision in children with CVI is the assessment of visual functioning or how the child uses vision in various tasks of daily living and in various environmental settings. In the next section, assessment of visual functioning will be examined.

Assessment of visual functioning. Whiting et al. (1985) and Jan, Groenveld, Sykanda, and Hoyt (1987), in their early work in British Columbia used a five point classification of vision to measure functional vision: No apparent vision, light perception, vision within 3 feet, vision within 10 feet, and vision beyond 10 feet. They rated each child at each visit to get an estimate of functional vision. Although, this rating was

created to be an informal means to document functional vision, the scale did not address the child's vision in the context of functional tasks of everyday life. There were no reliability or validity data presented on their measure of functional vision.

In 1999, Huo, Burden, Hoyt, and Good developed a six level measure of functional vision. The six levels were:

- Level 1 – light perception only.
- Level 2 – occasional fixation on large objects, faces or movement.
- Level 3 – Occasional fixation on small objects (i.e., pennies or stickers) or reliable fixation on faces.
- Level 4 – Reliable fixation on small objects; visual acuity 20/400 to 20/200.
- Level 5 – Reliable visual acuity not better than 20/50 (both eyes open).
- Level 6 – Completely normal vision.

They implemented this system as a method of quantifying functional vision. They rated each child at the initial visit and at the last visit. They calculated visual improvement in terms of changes in levels. A number of other researchers (Hoyt, 2003; Khetpal & Donahue, 2007) have used the Huo scale (as it is called) as a clinical measure of functional vision to supplement or compare to other measures of vision (e.g., TAC or VEP) and to document progress in children with CVI. Even though the Huo scale is used as a method to quantify functional vision, it does not include any information about how the child actually uses vision in tasks of everyday life or in an environment other than the medical clinic. There are no data about the reliability or validity of the scale.

Assessment of functional vision is often a challenge for children with CVI (Langley, 1998; Morse, 1992, 1999). To address this problem, a number of professionals have offered suggestions, but none have provided systematic research on these assessment methods. Hyvärinen (2003, 2006) suggested that assessment of functional vision in children with CVI should begin with assessments of vision skills such as fixation, tracking, accommodation, and contrast sensitivity. Next, assessment should involve ongoing observations that include information such as: recognition and reading, perception of pictures, perception of space, eye hand coordination, integration of visual information with information from other senses; and, use of other compensatory functions such as hearing. She has developed a number of commercially available products to assess vision skills such as acuity, grating acuity, contrast sensitivity, color vision in young children (Lea-Tests Ltd, 2006). Her assessment materials were developed for and have been widely used with young children. Many of the materials require skills such as recognition of shapes and verbal abilities (e.g., the ability to name shapes or follow verbal directions). She suggests the materials are to be used with children with ocular or cortical visual impairment; however, there is no data available on the use of Lea materials with children with CVI.

Dutton (2004) describes assessment of vision for children with CVI. The goals of a functional vision assessment are to determine the vision available for communication, education, and movement in the environment. He suggests observations of typical vision skills (e.g., acuity, contrast sensitivity, visual fields) as well as observations of higher visual processing skills. These skills include: simultaneous visual processing problems, recognition, problems with reading, problems with orientation, perception of movement,

visual memory, visual imagination, lack of visual attention, and prolongation of visual tasks. For each of these skills he provides a brief description of the skill, but no directions for specific observations or direct assessment of these skills. There is no information for the use of findings from observations for educational interventions. Finally, he offers no way to quantify these skills or measure progress.

Roman-Lantzy (2007) has developed The CVI Range, a functional vision assessment specifically designed to address the vision skills of children with CVI. She based her assessment on the work of Jan, Groenvelde and their team in British Columbia as well as her own work (Roman, 1996). Her assessment involves a combination of parent interview, child observation and direct assessment. Specific directions for assessment and scoring of each item on The CVI Range are provided in her book: *Cortical Visual Impairment: An Approach to Assessment and Intervention* (Roman-Lantzy, 2007). In addition, she details how assessment results are directly linked to interventions specific to each child's vision needs. The CVI Range is the first functional vision assessment designed specifically for children with CVI that makes a connection between the child's specific CVI vision characteristics and educational interventions. As such, it fills a gap in the field relative to functional vision assessments for children with CVI. The CVI Range will be described in more detail later in this chapter.

Issues in functional vision assessment for children with CVI. In summary, functional vision assessment for children with CVI is often accomplished with informal checklists that estimate overall visual functioning. Professionals recognize the importance of understanding how a child uses vision for communication and daily activities, but rarely do they provide specific guidance for conducting functional vision assessments.

They suggest observations of various vision skills, but again suggestions are lacking guidance and any means to objectively quantify observation results. Some materials, i.e. Lea-tests, require the ability to follow verbal directions, label shapes, and to use complex cognitive responses such as matching. Roman-Lantzy (2007) was the first professional in the field to develop a specific functional vision assessment for children with CVI; to provide directions for observations; and, to quantify observations so progress can be tracked in a systematic way across time. In the next section, outcomes for children with CVI will be examined. Appendix F provides a summary of outcome studies.

Outcomes for Children with Cortical Visual Impairment

Early researchers in the field of cortical vision loss in children noted that, possibly due to the nature of the immature brain and early plasticity, children with cortical visual loss often develop vision skills across time (Good, Jan, Burden, Skoczinski, & Candy, 2001; Groenvelde, Jan, & Leader, 1990; Hoyt, 2003; Whiting, et al., 1985). After the initial insult to the brain, vision tends to improve in a majority of the children (Afshari, Afshari, & Fulton, 2001; Groenvelde, Jan, & Leader, 1990). Chen, Weinberg, Catalano, Simon and Wagle (1992) examined the development of object vision in children with CVI. Object vision is defined as the ability to recognize faces or hand-held toys. They examined 30 infants who were diagnosed with CVI in the first year of life. Each child was followed for a minimum of 12 months (mean: 43 months). Object vision developed in 15 of the 30 children. There was no correlation between neuroimaging results and development of object vision.

Groenendaal and van Hof-van Duin (1992) studied a group of 38 children with CVI due to perinatal hypoxia. Of the 38 children (ages 7 weeks to 17 years), 22 were

seen more than once. The researchers tested various visual skills (e.g., acuity, visual threat, visual fields). At follow-up all children demonstrated improvement in visual skills, even children up to 16 years of age.

Castano, Lyons, Jan, and Connolly (2000) followed 10 children with CVI due to infantile spasms. At follow up visits (range 14 months to 6 years later) 5 children (50%) showed no improvement and 50% showed small amounts of improvement. All children continued to exhibit significant impairment in visual functions such as acuity and visual attention.

Huo, Burden, Hoyt, and Good (1999) reviewed the records of children seen in a pediatric ophthalmology clinic across 15 years. A total of 170 children with CVI were rated on a six level clinical evaluation of vision (i.e., the Huo scale described in the previous section). The ratings range from light perception only to normal vision. Children were rated at initial visit and at most recent visit. Average length of time for follow up was 5.9 years. They found that 60.42% of the children demonstrated improvement in vision as measured by levels of improvement.

Hoyt (2003) further analyzed the outcomes for these children by dividing them into two groups: children with damage to the visual cortex (41 children), and children with damage in the periventricular white matter (26 children with PVL). For the two groups the initial ratings on the 6 point scale were similar. At follow up (mean: 5.9 years), the children with visual cortex damage showed more improvement as measured by levels of change than the children with PVL. Both studies were retrospective studies completed by review of records. The age at entry and number of follow up visits was not reported for either group.

Matsuba and Jan (2006) reviewed the records of children seen in their clinic in British Columbia from 1985 to 2004 to evaluate long-term outcome for children with congenital CVI. Using medical records they identified 259 children with initial and follow up data. They included children with follow up assessments occurring after the child was at least 3 years old and with follow up at least 2 years from initial assessment. They divided the children into two groups by age of initial assessment (greater than and less than 3 years). Most of the children (46%) demonstrated improvement in visual acuity as measured by the Teller Acuity Card procedure. More improvement was seen in children whose initial assessment was prior to age three.

Khetpal and Donahue (2007) reviewed the records of children with CVI seen at Vanderbilt Eye Institute between the years 2002 and 2005. They had initial and follow up data on 52 children (ages 2 month to 19 years). Average length of follow up was 2.33 years. They used a modified 6 point scale based on the one developed by Huo (Huo, Burden, Hoyt, & Good, 1999). They did not describe the scale in the article. Forty percent of the children showed no improvement in visual functioning. Thirty four percent had minimal improvement; 17% had mild improvement; and, 6% had significant improvement. They concluded that the overall outcome for children with CVI has not changed across the past 20 years. They discussed the limitations of using a retrospective study. Many children did not have follow up data. In addition, there was no way to determine how many children had a delay from initial brain insult and their initial examination. That delay could prevent researchers from detecting any early improvements made by the children.

Summary and Methodological Issues

In summary, many researchers have followed children with CVI to document changes in vision across time. All of the large studies are descriptive and involve a retrospective review of medical records. Most researchers have not made any attempt to examine factors, other than location or extent of brain damage, which could affect visual outcome. Matsuba and Jan (2006) did divide children by ages and noted improved outcomes for children seen prior to age three. Within studies, the children cover a wide age range and length of time of follow up varies greatly, sometimes by years. There is no information in any study about any intervention or special education services that the child may or may not have received or what factors may have had a positive influence on the development of vision skills.

Measures of improvement vary from study to study which makes it difficult to compare results across studies. Some studies measure improvement with the Huo scale (described earlier), some with grating acuity, and some with specific skills such as object recognition. Acuity as a single measure does not provide information about how a child uses vision in the context of daily tasks (Good, et al., 1994; Teplin, 1995). A few studies have been conducted that begin to examine outcomes after specific interventions. These will be described in the next section.

Intervention Studies of Children with Cortical Visual Impairment

Baker-Nobles and Rutherford (1995) presented a case study of one child born who experienced seizures and apnea at two weeks of age and was subsequently diagnosed with left-sided hemiparesis and CVI. The child was initially seen at 5 months of age and her initial visual skills consisted of brief peripheral glance at colored gel on a

lightbox (i.e., a large box with florescent lighting covered with a white opaque plastic). Intervention strategies included: use of lightbox, use of lighted toys, and simplification of the child's visual environment (e.g., single objects on plain background). At 18 months of age, the child demonstrated central fixation on simple pictures, shifted gaze between objects, moved towards a toy she saw in her environment, and attempted imitation of an action with a toy that she had observed. Intervention strategies were described, but there was no information regarding frequency or duration of intervention activities. Treatment fidelity was not examined.

Farrenkopf, McGregor, Nes, and Koenig (1997) conducted a single subject study with a 17 year old girl with cortical visual impairment. They wanted to improve her skills in looking at, reaching for, and drinking from a cup. The cup was modified with a preferred color. They compared two treatment strategies across two settings. The first treatment involved verbal prompting and the second treatment involved physical prompting (i.e., placing a hand on her head to assist her in keeping her head still so she could fixate on her cup). The settings were home and school. The physical prompting resulted in more consistent attempts to look at, reach for and drink from her cup. They concluded that modifications that met her visual needs resulted in improved use of vision in a functional task. They described the intervention procedures, materials, and data collection in detail. The intervenors were trained and inter-observed agreement was checked on 25% of the data. Internal, external, and social validity were discussed.

Lueck, Dornbusch, and Hart (1999) studied the effects of training to improve visual functioning in a young child with CVI. They used a combination of visual environmental management, visual skill training, and visually dependent task training.

They used an A-B-A single subject design. The child showed significant improvement in his visual skills, however, concurrent with his treatment he began a new seizure medication that decreased seizure activity and increased alertness. Results of this study are confounded by factors outside of the treatment. While the child improved in visual functioning (i.e., visually guided reach, visual attention to toy), given outside events, the effects of treatment cannot be determined.

Ek, Fellenius, and Jacobson (2003) conducted a longitudinal study of 4 children with CVI who were learning to read. All of the children had a history of periventricular white matter damage. The researchers followed the children from first grade, age 6 until eighth grade, age 13. All children improved in visual acuity and learned to read (three print and one Braille). Full scale IQ scores declined. The children continued to have difficulties with visual cognitive organization and abstract logical reasoning. None of them liked to read and did not choose it as a leisure activity. All children continued to have visual problems such as low acuity, crowding, field deficits, and difficulty with fixations. This study did not examine a specific intervention strategy. It was a descriptive case study of 4 children learning to read.

McKillop and her colleagues (2006) conducted a discussion group of parents and caregivers. From their ophthalmology clinic they invited parents, grandparents, and caregivers of children with CVI. Over 40 people who were involved with 17 children participated in the group. The families were asked to discuss some of the problems of their children and some of the strategies that they had found helpful to their child. Families reported a variety of strategies which included: enlarging print to reduce complexity; using bright or favorite colors; teaching children with field losses

compensatory strategies (e.g., teaching a child with a lower field loss to regularly look down); using verbal cues to help the child; presenting materials on plain backgrounds; reducing clutter in environments; allowing for rest from visual fatigue; previewing new environments; teaching key routes; limiting the number of toys out at one time; and, use of visualization and social stories for social problems. No information was provided about the success of these interventions other than being described as “helpful.” There was no information about ages of the children, severity of CVI, other disabilities, or educational settings.

In summary, several single subject and case review studies have been completed with children with CVI. In individual cases, modifications of the visual environment and/or materials seem to result in improved use of vision for children with CVI. Parents also report a variety of strategies that are helpful for children with CVI. While these initial results are promising, many more studies are needed to document effective strategies that will lead to solid evidence-based interventions that are appropriate for children with CVI.

The CVI Range and the Proposed Study

Children with CVI as their primary visual diagnosis constitute a growing number of children with visual impairment (Blind Babies Foundation, 1995; Hatton, 2001; Hatton, Schwietz, Boyer, & Rychwalski, 2007). In addition, research has demonstrated that children with CVI can demonstrate improvements in vision (Khetpal & Donahue, 2007). Visual improvement is usually attributed to plasticity and the brain’s ability to develop visual skills despite damage to visual pathways and areas of the brain responsible for vision. The potential for change in such an important area of development makes it

critical for educators to understand how to best educate children with CVI by providing appropriate visual input and environmental modifications that meet their unique visual needs.

The first step in developing a foundation of evidenced based interventions is having an appropriate, valid, and reliable measure to systematically document strategies that are effective. In response to this need in the field, Roman-Lantzy (2007) developed The CVI Range. Prior to development of The CVI Range, Roman (1996) conducted a study to determine the efficacy of a parent/caregiver interview to identify infants with CVI. Answers to interview questions were compared to infant visual behaviors to establish concurrent validity of the interview. This infant observation protocol was the beginning of The CVI Range.

The interview questions for Roman's study (1996) were derived from visual characteristics of children with CVI described in the literature. The final interview contained 25 interview questions that covered the following categories: light gazing, use of movement in visual attention, color preference, visual field preference, visually guided reach, nonpurposeful gaze/visual attention, visual novelty, appearance of eyes/normal eye exam, and visual array/visual complexity. Several steps were taken to insure the content validity of the interview questions. Individuals with expertise in CVI were identified and asked to rate a set of 55 possible interview questions. The 55 questions covered the nine characteristics listed above. Based on the rankings of nine experts, the 55 questions were condensed to 25. Once content validity of the interview questions was established, several interviewers were trained to reliably score parental answers to the questions.

As mentioned previously, a functional vision assessment of each infant was conducted to establish concurrent validity of the interview. Concurrent validity was established by correlating the interview results with results of a functional vision assessment. The functional vision assessment was completed by observing each infant using behavioral indicators that represented vision behaviors from the CVI categories covered in the interview. For example, three interview questions asked parents about their child's color preferences. When the child was observed by an independent examiner, the color/s that the child looked at was noted. The results of the study indicated that the responses to the interview questions differentiated children with CVI from children with other types of visual impairment. In addition, the correlation between interview scores and observed visual behaviors was .936. The CVI characteristics identified and included in the interview, as well as included in the behavioral observations laid the foundations for The CVI Range.

The CVI Range is the first instrument specifically designed to be used for conducting a functional vision assessment of children with CVI. The assessment is based on the visual characteristics of children with CVI that have been documented by Jan and his team (Good & Hoyt, 1989; Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985) as well as other researchers in the area of CVI (Dutton 2003, 2004, 2006; Dutton et al., 1996; Jacobson, Ek, Fernell, Flodmark, and Broberger, 1996). These are also the characteristics that differentiated children with CVI from children with ocular visual impairment (Roman, 1996). The CVI characteristics assessed with this tool are:

- Color – does the child have a preference for a particular color, or respond better to certain colors?
- Movement – does the child need movement to initiate visual attention?
- Visual latency – is there a lag between presentation of materials and the child’s visual attention, is latency affected by familiarity of toy, time of day, or fatigue?
- Field preferences – does the child respond better when materials are in a specific visual field (e.g. only to the right or left)?
- Complexity – does the child have trouble looking when an object is complex, when there are too many objects, or when there is competing sensory information?
- Light-gazing/nonpurposeful gaze – does the child gaze at lights or spend time not looking at anything at all (i.e., nonpurposeful gaze)?
- Distance viewing – does the child have trouble looking at things at a distance?
- Visual reflexes – are the following reflexes present: blink to touch between eye brows and blink to threat?
- Novelty – does the child show a visual preference for familiar objects?
- Visual motor skills – does the child have a visually guided reach, or are look and touch performed separately? (See Appendix A for The CVI Range.)

The CVI Range is completed using information gathered from three sources: parent or teacher interview, child observation, and direct assessment (Roman-Lantzy,

2007). First, a parent, caregiver, or teacher is interviewed about the child's medical background, eye medical information, and visual behaviors. The second source of information is observation of the child in living and/or learning environments. Finally, information is gathered by systematic presentation of visual stimuli or direct assessment.

From all of the gathered information, The CVI Range is scored in two ways. The first section of The CVI Range is the Across-CVI Characteristics method. This method provides a "snapshot" of the child's visual abilities at each of the different levels of visual functioning (Roman-Lantzy, 2007, p.54). The CVI Range is in Appendix A. A complete scoring guide can be found in Roman-Lantzy (2007). The assessment protocol lists the specific skills that are included in each level of functioning listed below. In this section, five levels of visual functioning are possible:

- CVI Range 1-2: Student functions with minimal visual response;
- CVI Range 3-4: Student functions with more consistent visual response;
- CVI Range 5-6: Student uses vision for functional tasks;
- CVI Range 7-8: Student demonstrates visual curiosity; and,
- CVI Range 9-10: Student spontaneously uses vision for most functional activities.

The scoring guide describes how to score each individual item. The items are scored until a ceiling effect is reached. A ceiling effect is reached when the pluses that describe the child's current visual functioning end and a series of minuses occur for four or more items. The student's score on this section of The CVI Range is determined by where the plus scores stop. If the pluses stop in the middle of one of the levels, assign the lower number. If the pluses include all of the items in that level, assign the higher number. For

example, if the child passed half of the items in the “CVI Range 3-4”, the child would score “3”. If the child passed all of the items in the “CVI Range 7-8”, the child would score an 8. The child is assigned an overall score between 1-10 in the Across-CVI Characteristics section of the test.

The second section of the CVI Range is the Within-CVI characteristics method. In the second method, each characteristic is scored separately to describe the degree to which each is interfering with the child’s visual functioning. Each characteristic is scored from 0-1 as follows:

- 0 = not resolved; usually or always a factor affecting visual functioning;
- .25 = resolving;
- .5 = resolving; sometimes a factor affecting visual functioning;
- .75 = resolving; and,
- 1 = resolved; not a factor affecting visual functioning.

A characteristic is considered resolved if it no longer has an effect on the child’s visual functioning. For example, the score of “0” in the area of color would indicate that an object had to be a certain color for the child to look at it. Color is considered resolved and scored “1” when the child can look at objects of any color. See Appendix A for a copy of the The Resolution Chart that describes the progression of resolution for each characteristic. The total of all 10 characteristics yields a score of 0 – 10.

Finally, the two scores from the Across- and Within-CVI Characteristics Methods are compared to give a range (i.e., lowest total number to highest total number scored). For example, if a child scored a 4 on the Across-CVI Characteristics Method and scored a 3 on the Within-CVI Characteristics Method, the child’s functioning range would be 3-4.

The overall scores on The CVI Range are then divided into three broad intervention categories or Phases I, II, and III. Phases have implications for intervention in that at each Phase, there are broad overarching visual goals. Phase I encompasses CVI Range scores from 1-3 and the major goal of Phase I is to build consistent visual behaviors. Phase II encompasses CVI Range scores from 4-7 and the major goal of Phase II is to integrate vision into all functional routines. And finally, Phase III encompasses CVI Range scores 8-10 and the major goal of Phase III is to demonstrate visual curiosity and to consistently and spontaneously use vision in all tasks. Phases guide intervention strategies and environmental considerations. In Phase I, the child needs a high level of environmental modifications to use vision at all. Typically a child in Phase I cannot look and do anything else at the same time (e.g., touch or listen). Looking is infrequent and the child spends most of the time not visually attending to anything at all. By Phase II, the child can use vision in the context of daily activities. The child can look and also act on the environment at the same time. Vision use is not spontaneous and there is often prolonged times of nonpurposeful gaze. In Phase III, the child begins to attend to two dimensional input (i.e., pictures) and demonstrates visual curiosity. A child in Phase III spontaneously uses his vision most of the time (Roman-Lantzy, 2007).

For The CVI Range, there is preliminary content validity data (Roman, 1996; Roman-Lantzy, personal communication, September 17, 2007). At this time there is no formal reliability data available for the assessment. In the next section the concepts of reliability as they relate to measurement theory and specifically to The CVI Range will be examined.

Reliability

Measurement is concerned with quantifying the amount of a characteristic possessed by a person (Traub, 1994). To some extent, chance error is involved in all measurements (Carmines & Zeller, 1979). Chance or random error comes from all of the factors that confound the measurement of any phenomenon. A person's true score on any measurement would be the expected value of the variable that is being measured, however on any given instrument the person's true score is unknown. What is available is the person's actual score on the test or the observed score. The true score is considered to be the sum of the observed score and the random error associated with that measurement. A test is considered reliable if the observed score is close to the true score and random error is at a minimum (Traub, 1994).

Reliability theory is based on the notion that limited variance in observed scores, or consistency of observed scores means less measurement error. To the extent that measurement error is slight, an instrument is said to be reliable (Nunnally, 1978). Researchers can never know exactly the size of an error component of a measurement, but can estimate that size by looking at the variance of error in obtained scores. When the differences in observed scores on repeated measures are large, the random error is considered to be large. An examination of the score variance in repeated measures over many subjects yields the average amount that observed scores vary, or the standard error of measure (SEM). Therefore, two important qualities of any measurement are the strength of the relationship between repeated measures and the standard error of measurement. The strength of the relationship is equal to the correlation between the two measures. This correlation is known as the reliability coefficient. In general, the higher

the reliability coefficient and the smaller the standard error of measurement, the higher the quality of the instrument (Nunnally, 1978).

In order to obtain a reliability coefficient for an instrument, there must be two measurements for each student (Traub, 1994). The same test can be administered two times to the same group of students. This is the test-retest method of establishing reliability. Parallel or equal tests can be given to students to compare results on the two measures. In the case of The CVI Range, a parallel instrument does not exist, so this method of establishing reliability is not appropriate for the proposed study. And finally, a single test can be divided into half and the halves compared to each other. A test can be halved in any number of ways or statistical tests (e.g. Chronbach's alpha) can be performed that consider all possible combinations of items split into two halves (Traub, 1994). This method of determining reliability is called internal consistency.

For instruments that are scored by subjective judgements such as observational instruments, an additional way of examining reliability is the inter-rater reliability (Nunnally, 1978). Inter-rater reliability is determined by comparing the ratings of two examiners on the same students. When a score on an instrument is dependent on the judgement of a rater, it is important that the instrument and criterion for scoring are clear enough that two raters would get the same or a similar score. A measurement must be able to provide the same results for a child, no matter who observes the child, for the score to be meaningful (Nunnally, 1978). Inter-rater reliability is critical for an instrument such as The CVI Range because it is an instrument based on observations and subsequent judgments in scoring by the examiner.

In summary, one of the critical aspects of determining the quality of an assessment instrument is the reliability of that instrument. For The CVI Range to be used as a quality functional vision assessment for children with CVI, the reliability of the instrument must be examined. Several types of reliability are important to examine for The CVI Range including internal consistency, test-retest, and inter-rater reliability. The current study was designed to determine the reliability of The CVI Range by asking the following research questions:

1. To what degree does The CVI Range have good internal consistency as measured by coefficient alpha?
2. To what degree does the CVI Range have good test-retest reliability as measured by stable scores across time?
3. To what degree does The CVI Range have good inter-rater reliability as measured by having two qualified examiners obtain the same or similar score?
4. To what degree does The CVI Range have consistency across the two sections of the assessment as measured by comparing the scores on the two sections?

CHAPTER III

RESEARCH METHODOLOGY

The CVI Range (Roman-Lantzy, 2007) is a functional vision assessment for children with cortical visual impairment (CVI). Functional vision assessments describe how a child uses vision in vision-related tasks such as recognizing and manipulating objects. The purpose of this study was to examine the reliability of The CVI Range. To address reliability, the following research questions were asked:

1. To what degree does The CVI Range have good internal consistency as measured by coefficient alpha?
2. To what degree does the CVI Range have good test-retest reliability as measured by stable scores across time?
3. To what degree does The CVI Range have good inter-rater reliability as measured by having two qualified examiners obtain the same or similar score?
4. To what degree does The CVI Range have consistency across the two sections of the assessment as measured by comparing the scores on the two sections?

Description of Study

Reliability involves being able to consistently measure a behavior of interest (Nunnally, 1978). In the case of this study, reliability meant being able to consistently measure the functional vision of children with CVI. Children with CVI were assessed by professionals who had been trained through the Multi-state CVI Mentorship Project. The CVI Range was used by these trained mentors. These assessments were analyzed to determine the reliability of the CVI Range. Prior to initiating data collection, Institutional

Review Board (IRB) approval was obtained from the University of Maryland. The following sections describe data management, the Multi-state CVI Mentorship Project, training of the mentors, the professionals who conducted assessments for the study, study participants, assessment procedures, methodological considerations, and data analysis.

Data Management

Examiners who tested children sent all of the assessment protocols to the student investigator of this study along with the signed Informed Consent forms. Each single assessment and each pair of assessments were assigned a number to protect the confidentiality of the child. Raw data was stored in a locked cabinet.

CVI Mentorship Project and Training of the Mentors

The CVI Multi-state Mentorship Project was a regional collaborative project among four state deaf-blind programs. This multi-state project was included as one component of the 2003-2008 Office of Special Education Program (OSEP) deaf-blind grants for each state.

During year one, each state chose CVI mentors. Mentors were professionals from the fields of vision, physical therapy, occupational therapy and early intervention. These professionals agreed to 5 years of extensive training and field work to become CVI mentors. For training, the mentors participated yearly in a 3-day conference led by Dr. Christine Roman-Lantzy. In addition to the annual conference, mentors participated 3 times a year in video conferences and/or webinars. Training content included: relevant literature in causes and treatment of CVI; assessment of children with CVI; intervention for children with CVI; and, mentorship skills such as planning and delivering

presentations, adult learning styles, and peer coaching. Assessment training involved use of The CVI Range with children of varying ages and levels of CVI.

Within each state, mentors met monthly to review training and assignments. Field work included conducting assessments of children with CVI as well as designing educational interventions to be implemented by the child's educational team.

Assessments were videotaped periodically for supervision and training purposes.

Supervision of fieldwork was provided by each state's planning team members. Results of assessments and interventions were also periodically reviewed by Dr. Roman-Lantzy and individual feedback was provided by her to each mentor. These highly trained mentors, including the author of this study, conducted the assessments for this study. In addition, Dr. Roman-Lantzy also conducted assessments included in these analyses.

Evaluators

Twelve professionals, including this author, and Dr. Roman-Lantzy conducted the assessments that were included in this study. The evaluators included three special education teachers/early interventionist, six teachers of the visually impaired, two occupational therapists, and one neonatologist. The teachers and occupational therapists were all members of the CVI Multi-State Mentorship project. The neonatologist has worked with Dr. Roman-Lantzy at West Penn Hospital in Pittsburgh for more than 10 years and was a member of her dissertation committee. He participated in nine inter-rater assessments with Dr. Roman-Lantzy. The assessments were conducted in four states representing a variety of rural, urban, and suburban settings.

As part of the final evaluation of the CVI Multi-State Mentorship project, the mentors completed evaluation surveys that included a question about the number of CVI

assessments each mentor had conducted as part of this 5-year project. Number of assessments completed per mentor ranged from 30-80, with an average of 44 assessments; thus, each mentor who completed assessments for this study had extensive experience in conducting CVI assessments using The CVI Range.

Study Participants

When a child was referred to the CVI Mentorship Project, the mentor explained the reliability study and asked the parent or guardian's permission to use the assessment as part of this study. If the family chose to participate, they signed Informed Consent that had been approved by the University of Maryland Institutional Review Board (IRB) (See Appendix G). If families chose not to participate in the study, their child was still eligible for the services of the CVI Mentorship Project (i.e., assessment and intervention).

Children were referred by local school systems and early intervention programs. Advertising was done through general deaf-blind program descriptions including brochures and websites for each state (e.g., www.cbss.umd.edu), through statewide CVI training provided by CVI mentors, and through informal contacts of mentors with special education professionals in their state. There were no specific recruitment brochures or flyers advertising the CVI Mentorship project or this study. Any child assessed by a CVI Mentor was eligible for participation in the study. Subjects were recruited from early intervention programs (birth to 3) or special education programs (3-21 years old) if they were diagnosed with CVI or suspected of having CVI. Subjects could also have other disabilities such as developmental delay, mental retardation, cerebral palsy, seizure disorder, or co-existing ocular conditions. For each child, the mentors reported results of assessment using The CVI Range, the child's date of birth, current age at time of testing,

other disabilities, and a brief description of services the child received. Sixty-three children were tested by mentors. In addition to children tested as part of the CVI Multi-state Mentorship Project, nine assessments were conducted by Dr. Roman-Lantzy. For these children, Dr. Roman-Lantzy and her neonatologist colleague conducted inter-rater reliability assessments.

The total sample for data analysis in this study included 72 children assessed as part of the study and 32 children from an existing data set, a total of 104 children. All of the 104 assessments were used for internal consistency analyses. Of the 72 children assessed specifically for this study, 27 children were assessed by 2 examiners for inter-rater reliability; and, 20 children were assessed twice for test-retest reliability. The average age of the sample was 46.5 months with a range of 6-144 months. See Table 1 for details.

Dr. Roman-Lantzy regularly conducts many CVI assessments using The CVI Range at West Penn Hospital in Pittsburgh, PA and has a large existing data set of assessments. IRB approval was given to use assessments from this existing data set for analyses in this study. An additional 32 assessments from this data set were included in analyses in this study. The average age of this sample was 43 months with a range of 5-173 months. See Table 1 for more details.

In addition to age, additional disabilities and a brief description of services was reported for each child. All of the children had additional disabilities. Additional disabilities included developmental delay, mental retardation, cerebral palsy, health impairment, hearing impairment, and/or other ocular conditions. For children with ocular conditions, optic nerve atrophy was the most frequently occurring condition (43%). Table

2 provides details about additional disabilities divided by IDEA program category, i.e. early intervention (EI) which includes children birth to 36 months and special education (SPED) which includes children 36 months and older. Most children received early intervention or special education services. Additional services included PT, OT, speech therapy, vision services, and hearing services. Table 3 shows the different services received by the children, again divided by IDEA program category. For children eligible for early intervention services (i.e., birth to 36 months), home was the most frequent placement. For children eligible for special education services (i.e., 36 months and older) placements represented a continuum from home-based to special centers. See Table 4 for a summary of placements for children in early intervention and special education.

Table 1: Age of subjects

Group	N	Mean age in months (range)	SD
Single assessment	25	47.2 (7-144)	36.9
Inter-rater	27	50.4 (6-119)	32.5
Test-retest	20	40.7 (9-116)	26.3
Roman-Lantzy	31*	43 (5-173)	42.9
Total sample	103*	45.3 (5-173)	35.7

* missing age data on 1 student

Table 2: Additional Disabilities by IDEA Program Category

Group (N)	Cognitive - DD/MR	CP/motor	Hearing Impaired	Health impaired	Ocular
EI * (55)	55 - 100%	24 - 44%	11 - 20%	26 - 47%	12 - 22%
SPED* (49)	37 - 76%	35 - 71%	11 - 22%	26 - 53%	18 - 37%
Total (104)	92 - 88%	59 - 57%	22 - 21%	52 - 50%	30 - 29%

* EI = Early intervention; SPED = Special Education

Table 3: Services by IDEA Program Category

Group (N)	Special instr'n N - %	Special Educ. N - %	PT N - %	OT N - %	Speech N - %	Vision N - %	Hearing N - %
EI * (55)	40-73%		55-91%	37-67%	14-25%	42-76%	4-7%
SPED * (49)		46-94%	39-80%	37-76%	34-69%	39-80%	4-8%
Total (104)	86-83%		94-90%	74-71%	48-46%	81-78%	8-8%

* EI = Early intervention; SPED = Special Education

Table 4: Placements by IDEA Program Category

Group (N)	Home-based N - %	Special center N - %	Special class N - %	Combo N - %	Reg. Ed. N - %
EI* ** (54)	46 – 85%	5 – 9%	3 – 6%		
SPED* (49)	6 – 12%	9 – 18.5%	19 – 39%	9 – 18.5%	6 – 12%
Total** (103)	52 – 50%	14 – 14%	22 – 21%	9 – 9%	6 – 6%

* EI = Early intervention; SPED = Special Education

** Placement data missing on 1 student

Assessment Procedures for The CVI Range

The CVI Range was completed by gathering information from three primary sources: interview, observation, and direct assessment (Roman-Lantzy, 2007). Interviews were conducted with family members or service providers who knew the child well. Observations were made as the evaluator interacted with the child in living and learning environments. Direct assessment involved presenting specific visual stimuli and noting the child's responses. The specific assessment procedures for each of these sources of data will be described in the next sections.

Interview procedures. At the beginning of the assessment, the evaluator talked to the family or service provider about the child, the child's history, and their observations of the child's vision skills. The evaluator gathered information about the child's medical history. Families were asked a variety of questions about how the child uses vision. The answers to the interview questions indicated behaviors that were positive or negative for CVI. Table 5 provides sample interview questions. Table 6 provides sample responses

that would be positive for CVI. A complete set of interview questions can be found in Roman-Lantzy (2007).

Table 5: Sample Interview Questions (Roman-Lantzy, 2007 p.34)

-
1. Tell me what you do to get your child interested in a toy.
 2. When you show your child a toy, how do you know he sees it?
 3. What has the doctor told you about your child's eyes?
 4. Do you think your child has a favorite color?
 5. How does your child react when you give him new things to look at?
 6. Are you usually able to identify (be certain of) what your child is looking at?
 7. Does your child have a favorite side or favorite head position?
-

Observation procedures. Observations began as soon as the evaluator met the child. Visual behaviors were noted as the child participated in routine activities and interacted with significant people. Whether in the home or a school environment, the evaluator noted what the child looked at, if the child looked at the evaluator, or if the child seemed to be looking at nothing. The evaluators noted if the child displayed visual curiosity. Did the child stare at lights? Visual behaviors in the context of the child's real world situation gave the evaluators valuable information about the child's functional vision (Roman-Lantzy, 2007).

Table 6. Sample Answer Guide: Parent Interview Questions (Roman-Lantzy, 2007, p. 41)

Interview question	Characteristics of CVI or other features	Positive for CVI: Sample responses	Negative for CVI: Sample responses
Tell me what you do to get your child interested in a toy.	Movement Visual fields	I move it, or shake it back and forth, or activate it. I present it to my child's right or left side and move it or try to get it to make a motion.	I set it up in the center of where my child is positioned. I put it in his or her hand. I put it in front of him or her. He or she notices the toy san then I bring it to him or her.
When you show your child something, how do you know he or she sees it?	Visual attention/ nonpurposeful gaze	I'm not always sure he or she sees what I show him or her. When I show him or her favorite or familiar objects, he or she stops doing other things. He or she smile or moves toward the object. I don't think there are many things he or she likes to look at.	He or she likes to see most things as long as I move them close enough. He or she seems to like the same things other babies like. He or she looks right at the toy and gets "excited."

Direct assessment procedures. In addition to gathering information through interview and observations, evaluators conducted direct assessment of specific visual skills. Evaluators typically began the assessments by interviewing the child's parent or providers. As the interview was in progress, the evaluator made informal observations about the child's use of vision. Based on the answers to the interview questions and informal observations, the evaluator then chose materials and began direct assessment procedures. Direct assessment involved presentation of specific visual stimuli that would

elicit typical of a child with CVI. The evaluator then noted the child's responses.

Materials were presented that addressed the ten specific characteristics of CVI including: color, movement, visual latency, visual field preference, complexity, light-gazing and nonpurposeful gaze, distance viewing, visual reflexes, novelty, and visual motor responses. The specific materials used depended on the preliminary information gathered from interview and observations. For example, if the parent reported that the child's favorite color was yellow, then yellow materials were used for direct assessment.

Because children with CVI often look at familiar objects better than novel ones, the child's own materials were often used in the assessment. The environment was controlled during direct assessment procedures (e.g., quiet, plain background) to construct an optimal situation for the child to use vision (Roman-Lantzy, 2007). Table 7 provides sample suggestions for direct assessment.

Table 7. Sample Suggestions for Direct Assessment (Roman-Lantzy, 2007, p. 55)

-
- **Color preference:** Present materials that are made of the student's reported preferred color and then compare the response to behavior when objects of a nonpreferred color are presented.
 - **Visual latency:** When presenting both known and novel objects, note the amount of time it takes for the student to notice the presence of the object. Also note when the latency, or slowness to respond, occurs in the session and what conditions or materials are associated with latency.
 - **Atypical visual reflexes:** Attempt to elicit the visual blink and visual threat reflexes several times in a row—perhaps two or three times—and several times during the assessment session. Be aware of the possibility of habituation; that is, the student's blink responses may lessen if the evaluator repeats the touch or threat too many times in a row.
-

Scoring Procedures for The CVI Range

The CVI Range was scored using two different methods. In the Across-CVI Characteristics Assessment Method the child was assigned an overall score along a 10-

point range that provided an overview or “snapshot” of the child’s visual functioning (Roman-Lantzy, 2007, p. 54). In this method, five levels of functioning were used:

- CVI Range 1-2: Student functions with minimal visual response;
- CVI Range 3-4: Student functions with more consistent visual response;
- CVI Range 5-6: Student uses vision for functional tasks;
- CVI Range 7-8: Student demonstrates visual curiosity; and,
- CVI Range 9-10: Student spontaneously uses vision for most functional activities.

In each range, there are lists of behaviors that are scored in the following manner:

- R Statement represents resolved visual behavior
- + Describes current functioning of the child
- +/- Partially describes child
- - Does not apply to the child

In addition, for each behavior the evaluator indicated whether the score was obtained from interview, observation, or direct assessment. In this method of scoring, not every characteristic is represented at each range. A complete scoring guide can be found in Roman-Lantzy (2007). The CVI Range can also be found in Appendix A.

The second method of scoring The CVI Range was the Within-CVI Characteristics Assessment Method. In this method each of the ten characteristics was scored on a 0 to 1 scale to indicate the degree to which this characteristic was interfering with visual functioning. Each characteristic was scored from 0-1 as follows:

- 0 = not resolved; usually or always a factor affecting visual functioning;
- .25 = resolving;

- .5 = resolving; sometimes a factor affecting visual functioning;
- .75 = resolving; and,
- 1 = resolved; not a factor affecting visual functioning.

The total score yielded a number between 0-10. The CVI Resolution Chart provided a guideline for scoring each characteristic. See Table 8 for sample CVI Resolution Chart scoring guide. See Appendix A for complete CVI Resolution Chart.

When the scoring was complete, there were two scores, each a number between 0 and 10. The scores were then placed into one of three broad phases that guided the development of intervention strategies. Phase I included CVI Range scores 0 – 3; Phase II included CVI Range scores 3.25 – 7; and, Phase III scores included CVI Range scores 7.25 – 10.

Table 8. Sample CVI Resolution Chart Scoring Guides

Score = 0	Score = .25	Score = .5	Score = .75	Score = 1
Objects viewed are generally a single consistent color	Student has a “favorite” color	Objects viewed may have two to three colors	More colors and familiar patterns are regarded	Student has no color or pattern preferences
Prolonged periods of visual latency	Latency slightly decreases after periods of consistent viewing	Latency present only when student is tired, stressed, or over stimulated	Latency rarely present	Latency resolved

In general, children in Phase I need maximum environmental modifications and the major goal of Phase I is to build consistent visual behaviors. In Phase II children are able to integrate vision with functioning. Less environmental controls are needed and the major goal of Phase II is for the child to use vision in the context of functional activities. By Phase III children are demonstrating visual curiosity and are spontaneously using their

vision most of the time. In Phase III children can look at pictures and other two-dimensional materials. Only the most complex environments affect visual functioning. In the total sample, the mean score on The CVI Range was 4.72 (SD = 2.5) with a range of scores from 1.00 to 9.25. Of the 104 children in this study, 34 (33%) scored in Phase I, 45 (43%) scored in Phase II, and 25 (24%) scored in Phase III. Table 9 provides a further break down of Phase data relative to group, (e.g., inter-rater, test-retest, etc).

Table 9: Phase by Group

Group (N)	Phase I N - %	Phase II N - %	Phase III N - %
Inter-rater (27)	6 – 22%	13 – 48%	8 – 30%
Test-retest (20)	9 – 45%	7 – 35%	4 – 20%
Single test (25)	11 – 44%	7 – 28%	7 – 28%
Data set from Roman-Lantzy (32)	8 – 25%	18 – 56%	6 – 19%
Total sample (104)	34 – 33%	45 – 43%	25 – 24%

Methodological Considerations

Traub (1994) described four factors to consider when designing and implementing a reliability study:

The sample should be representative of the population of examinees that is of interest, the measurements should be experimentally independent, the procedure for collecting data in the reliability experiment should duplicate that used in practical applications of the measuring instrument, and the experiment should produce at least two measurements on every participant. (p. 67)

Each of these will be discussed relative to this study.

Representative sample. For this study, subjects were recruited from early intervention and special education programs that referred children for a CVI assessment to a CVI mentor. The children who were referred during this study ranged in age from 6 months to 12 years old, with a variety of other disabilities (e.g., cerebral palsy, mental retardation, seizure disorder). They all had a medical history that was suggestive of CVI (e.g., PVL – periventricular leucomalacia, HIE – hypoxic ischemic encephalopathy, central nervous system anomalies) and represented a wide range of visual functioning from almost no vision responses to subtle visual difficulties. As such, they were representative of the children with CVI described by other researchers.

Independence of measures. Independent measures are ones that have not been influenced by or will not influence the measurement of another student. Scores on the assessment of one student should not affect the scores of another student. For example, if a paper-pencil test were administered to a group of students no student should be able to copy another student's answers. When students are tested more than once, every measurement should be independent. The score on the first assessment should in no way affect or generate the score on the second assessment.

In this study, students were tested one at a time in home or educational settings. Student responses did not have an influence on other student's responses. Separate student scores were independent. For inter-rater reliability assessments, the rating of one examiner did not influence the rating of the second examiner, making the two scores independent of each other. In this study, two mentors participated in the same assessment, but scored the assessment independently. Participation in the assessment usually involved one mentor interviewing the family or provider and interacting with the child while the

other mentor observed, but occasionally the second mentor would talk to the family or interact with the child also. Mentors were not given instructions about how to participate in the assessment together, but were given strict instructions not to discuss scoring of the assessment when they participated in an inter-rater reliability assessment. The special considerations of the test-retest situation will be discussed further in a subsequent section.

Identical administration procedures. The procedures for conducting assessments in a reliability study should be identical to the procedures used in practical application of the assessment. The CVI Range is an assessment that is completed in a child's home or educational setting using a combination of interview, observation, and direct assessment strategies. For this study, the assessments were conducted in the same manner. Students were assessed in natural settings (home or school). The procedures described in Roman-Lantzy (2007) and described in a previous section were used to collect assessment data.

Two measures for each student. A reliability study has to yield more than one score on each student. For inter-rater reliability, two examiners scored the same assessment. For internal consistency, statistical analysis (described later) was used that allowed a single administration to be divided into parts to yield the required multiple scores for each student. For test-retest reliability the same student was tested on two occasions to yield the required two scores. In test-retest reliability studies the researcher must consider several factors to insure that the measures are independent including: learning between test administrations, variations in the subjects (health, fatigue), or variations in the testing conditions.

First, when a test is administered two times the length of time between administrations should be short to avoid the problems associated with development or

learning (Traub, 1994). The usual convention is two weeks between test administrations. For this study, each test-retest assessment was completed in a time frame of two weeks or less. The range in terms of number of days between test one and test two was 1 – 14 days, and the average length of time between tests was 6.7 days (SD = 3.6 days).

Another consideration for test-retest situations is the child's recall of previous answers. The CVI Range is an observational assessment and the children were observed in settings and activities that were familiar to the child. Vision responses were observed to be present or not, unlike a test with correct and incorrect answers. The child either had the vision skill or did not, therefore, recall in the test-retest reliability assessments was not a factor, and did not affect the test-retest reliability coefficient.

Finally, for a test-retest reliability study, the testing procedures should be identical in the two situations. For this study, children were tested two times in the same setting, with the same materials, by the same examiner. CVI mentors reported the following: date of original assessment, date of retest, setting, positions of the child (including equipment used to position the child), materials used, and any other factors that might affect the child's performance on either occasion (including hunger, fatigue, seizures, illness, etc.). There were not situations where the second testing was completed in a different setting. In all of the 20 test-retest assessments, there were no reported interfering events such as an illness or seizures that could have affected the results. There were no other major discrepancies in materials, positions, and time of day between test one and test two for the 20 children used for the test-retest reliability analysis.

Data Analysis

An investigation of the reliability of The CVI Range was conducted by analyzing assessment data gathered using The CVI Range. Reliability coefficients were determined by examining internal consistency, test-retest reliability and inter-rater reliability. In addition, a comparison of the scores on Part I and Part II of the CVI Range was made.

For internal consistency, test-retest, and inter-rater reliability analyses, Part II, the Within-CVI Characteristics Method section of The CVI Range, was used. In Part II every characteristic was represented at numeric values from 0 to 1 and the rating of all 10 characteristics was totaled, yielding an overall CVI Range score between 0 and 10. Scores from 0 to 10 represented severity of CVI with lower scores representing more severe CVI than higher scores.

A total of 104 children were assessed using The CVI Range. Twenty-seven children were tested by two examiners for inter-rater reliability, 20 children were tested two times by the same examiner for test-retest reliability, and 57 children were tested one time by a single examiner. For internal consistency and for comparison of scores from Part I and Part II of The CVI Range, all 104 children were included. When a child had more than one assessment (i.e., inter-rater reliability or test-retest reliability), the researcher randomly chose one of the assessments for inclusion in the analyses.

Internal consistency. One of the assumptions of reliability is that all of the items on an assessment are measuring one underlying construct. Internal consistency is a measure of how well the items in a test correlate with each other. High correlations are suggestive of consistent measurement of a single construct (Carmines & Zeller, 1979). Chronbach's alpha is often used to determine internal consistency. It was important to

look at internal consistency of The CVI Range because the total score is used to describe severity of CVI and is used in other reliability analyses. If alpha was high, then the scale was likely measuring a single underlying construct. In addition, the total score could then be used for determining test-retest and inter-rater reliability. For this study 104 assessments were analyzed using Chronbach's alpha as a measure of internal consistency. Results are presented in Chapter 4.

Inter-rater reliability. For inter-rater reliability, two similarly qualified examiners must get the same or similar results on a given assessment protocol (Nunnally, 1978). For this study, 27 children were assessed by two trained examiners who then independently scored The CVI Range. Total scores on Part II of The CVI Range were used to determine inter-rater reliability. The two sets of scores were correlated using Pearson's correlation coefficient. Pearson's coefficient assumes a normal distribution of scores and interval level data, however, Pearson's r has been shown to be robust to violations of those assumptions (Harris, 2001; Havlicek & Peterson, 1977) and is therefore preferred to a non-parametric measure of correlation.

In addition to correlation, inter-rater reliability was computed using Cohen's kappa. Kappa is appropriate for inter-rater reliability calculations with categorical data (Fleiss, Levin, & Paik, 2003). The scores on The CVI Range place the child in one of three categories, i.e. Phase I, II, or III and phase placement has practical intervention implications; therefore, kappa was used to determine how well two examiners placed the child in the same phase. Results of correlational and kappa analyses are presented in Chapter 4.

Test-retest reliability. Test-retest reliability is an index of stability across time (Nunnally, 1978). For this study, 20 children were tested on two different occasions, not more than 14 days apart by the same examiner. The same examiner was used to avoid confounding the results with inter-rater issues. The time between test one and test two ranged from 1-14 days with an average of 6.7 days apart. Total scores on Part II of The CVI Range were used to determine test-retest reliability. The two sets of scores were correlated using Pearson's correlation coefficient. As with inter-rater reliability, Pearson's was chosen due to its robustness.

In addition to correlation, test-retest reliability was also computed using Cohen's kappa. Kappa is appropriate for agreement calculations with categorical data (Fleiss, Levin, & Paik, 2003). Because phase placement is categorical and has intervention implications, kappa was used to determine if test-retest scores on The CVI Range place the child in the same Phase. Results of test-retest analyses are presented in Chapter 4.

Comparison of across-CVI and within-CVI characteristics methods. The CVI Range is made up of two sections that are scored differently, but both provide a total score between 0 – 10. To this point, the Within-CVI Characteristics Method section of the test was used for reliability calculations. The two sections are scored differently and the scoring guidelines are different. Appendix A has a copy of The Resolution Chart guide for scoring the Within-CVI Characteristics Method. A complete scoring guide for scoring the Across-CVI Characteristics Method section can be found in Roman-Lantzy (2007). The important clinical question was do the two parts of the test place the child in the same phase and therefore have similar intervention implications. For this final analysis, kappa was again used to determine the level of agreement in group placement.

One assessment from each child, a total of 104 children, was used for comparison. For children who were tested more than once or by more than one person, the researcher randomly picked one of the assessments to be used in the analysis. Results are presented in Chapter 4.

CHAPTER IV

RESULTS

The purpose of this study was to examine the reliability of The CVI Range. To address reliability, the following research questions were asked:

1. To what degree does The CVI Range have good internal consistency as measured by coefficient alpha?
2. To what degree does the CVI Range have good test-retest reliability as measured by stable scores across time?
3. To what degree does The CVI Range have good inter-rater reliability as measured by having two qualified examiners obtain the same or similar score?
4. To what degree does The CVI Range have consistency across the two sections of the assessment as measured by comparing the scores on the two sections?

Results will be presented for each question.

Internal Consistency

A total of 104 children were assessed using The CVI Range. Twenty-seven children were tested by two examiners for inter-rater reliability, 20 children were tested two times by the same examiner for test-retest reliability, and 57 children were tested one time by a single examiner. When a child had more than one assessment, i.e. inter-rater and test-retest, the researcher randomly chose one of the assessments to include in the analysis. Chronbach's alpha was used to determine the reliability of The CVI Range. Alpha was examined for the total sample and for each of the following subgroups: inter-rater, test-retest, single assessments, and data set from Roman-Lantzy. Table 10 shows

the results of these analyses. As a general rule, reliabilities are acceptable at .80 for most scales. At that level the measurement represents mostly true score and little random error (Carmines & Zeller, 1979). The CVI Range had an internal consistency measure of .96 which represents a very high reliability. The higher the alpha level, the more likely that the scale is measuring a single underlying construct, in this case, severity of CVI.

Table 10: Chronbach's Alpha for The CVI Range

Group	N	Alpha
Inter-rater	27	.959
Test-retest	20	.968
Single assessments	25	.969
Roman-Lantzy data set	32	.956
Total sample	104	.962

Alpha is based on the average correlation among items on a scale. The inter-item correlation table for the 10 test items is presented in Table 11.

Test-retest Reliability

Twenty children were tested two times by the same examiner to determine test-retest reliability of The CVI Range. The children were tested by the same examiner, in the same setting, the same time of day, with the same materials. The average time between tests was 6.7 days (range 1-14 days). The average age of the children in this group was 40.7 months (range 9-116 months). Using the scores from Part II, the Within-CVI Characteristics Method section of The CVI Range, the two sets of scores were correlated. Pearson's correlation coefficient for test-retest reliability was .99.

Table 11: Inter-Item Correlation Matrix for The CVI Range

	Color	Movem't	Latency	Fields	Complex	Lt gaze	Distance	Reflexes	Novelty	Vis motor
Color	1.00									
Movem't	.819	1.00								
Latency	.774	.788	1.00							
Fields	.610	.674	.646	1.00						
Complexity	.785	.872	.770	.663	1.00					
Lt gaze	.747	.774	.789	.630	.753	1.00				
Distance	.787	.791	.821	.658	.756	.737	1.00			
Reflexes	.614	.620	.649	.561	.655	.526	.631	1.00		
Novelty	.781	.814	.792	.616	.819	.757	.793	.595	1.00	
Vis motor	.804	.833	.799	.660	.800	.745	.761	.653	.787	1.00

In addition to correlation, test-retest reliability was examined using Cohen's kappa. Children were placed into one of three phases based on their scores on The CVI Range. Phase I included children whose scores were 0-3; Phase II included children whose scores were 3.25 – 7.0; and Phase III included children whose scores were 7.25 – 10. In Roman-Lantzy (2007), Phase I is considered scores from 0-3; Phase II is considered scores from 4-7; and Phase III is considered scores from 8-10. For the purposes of this study, it was necessary to decide what to do with scores between 3 and 4 (i.e., 3.25, 3.5, 3.75) as well as between 7 and 8 (i.e., 7.25, 7.5, 7.75). In a discussion with Roman-Lantzy (personal communication, January 29, 2009), a decision was made to make the cutoff points for Phase II any score above 3 and for Phase III, any score above 7. Nine children (45%) scored in Phase I; 7 children (35%) scored in Phase II; and 4 children (20%) scored in Phase III. The measure of agreement for group placement, or kappa, was 1.0. There was a perfect agreement in phase placement from the first test to the second for all 20 children. Based on the correlation coefficient (.99) and kappa (1.0) The CVI Range has very high test-retest reliability.

Inter-rater Reliability

Twenty-seven children were tested by two examiners to determine inter-rater reliability. The average age of the children in this group was 50.4 months (range 6-119 months). Part II, the Within-CVI Characteristics Method section of The CVI Range, was used for this analysis. Pearson's correlation coefficient for inter-rater reliability was .98. The CVI Range had very high inter-rater reliability.

In addition to correlation, group placement or kappa was also examined as a measure of inter-rater reliability. Children were placed into one of three phases based on

their scores on The CVI Range. In this group 6 children (22%) scored in Phase I; 13 children (48%) scored in Phase II; and 8 children (30%) scored in Phase III. Kappa greater than .75 represents excellent agreement beyond chance (Fleiss, Levin, & Paik, 2003) and kappa was .83 for inter-rater reliability of The CVI Range. Further analysis of the absolute difference in the scores of the two examiners revealed a mean difference in scores of .31 points (range 0 – 2 points). In other words, on average the total scores of the two raters differed by less than half a point. Differences in group placement was affected by cut-off points (e.g., 3.0 is considered Phase I and 3.25 is considered Phase II) more than by large differences in total scores.

Comparison of Across-CVI and Within-CVI Characteristics Method

As described earlier, The CVI Range is scored by two different methods. The final question examined was whether the two parts of The CVI Range placed the child in the same phase. All 104 assessments were used for this analysis. When a child had more than one assessment, the researcher randomly chose only one of the assessments to be used in the analysis. In the total sample 34 children (33%) scored in Phase I; 45 children (43%) scored in Phase II; and 25 children (24%) scored in Phase III. Kappa was .88 for The CVI Range. The average score difference in Part I and Part II was .55 (range 0 – 1.5 points). Differences in phase placement were the result of cut off scores more than large differences in the two scores. There is a very high agreement in the scoring of Part I and Part II of The CVI Range.

Post Hoc Analysis

To further examine internal consistency, Chronbach's alpha was examined by phase. Does The CVI Range have good internal consistency at differing levels of severity of CVI? Alpha was computed for the 34 children who scored in the Phase I range (0-3 points). Alpha was .55 for Phase I. Forty-five children scored in Phase II (scores from 3.25 – 7) and alpha was .78. For Phase III (scores 7.25 – 10) there were 25 children, and alpha was .55.

Alpha can be affected by several factors. First, a small number of items can affect alpha (Traub, 1994). The CVI Range has only 10 items, however, that did not affect alpha for the total sample or for the smaller groups such as test-retest or inter-rater. Even with a 10-item assessment, alpha was very high (.95 and above) in the previous analyses.

Alpha is also greatly affected by score variability. Alpha is the highest when there is minimal within-subject variability and greater variability between subjects (Garson, 2002). Restricting the scores to a small part of the assessment, which is what happened when scores were analyzed by phase, had a negative effect on alpha. The between subject variability was too low when the data was analyzed by phase. Alpha was lower at the two extremes (Phase I and III) and higher in the midranges (Phase II). There was even less variability between subjects at extremes. On The CVI Range, scores were rarely at the far extremes (e.g., 0, .25, 9.75, 10). In this sample, the lowest score was 0.75 (n = 1) and the highest score was 9.25 (n = 1). In Phase II, there were scores representing each possible point value that was included in that group (i.e., 3.25 – 7); therefore, alpha was higher in Phase II. In each group analyzed previously (e.g., inter-rater, test-retest) as well as in the total sample, there was a sufficient variability between students. Each group had children

from all phases representing a range of CVI severity. The lower alpha scores when data was analyzed by phase was a result of the nature of alpha and the effects of variability.

Summary

The reliability of The CVI Range was examined in a number of ways to answer the research questions posed by this study. According to the results of this study, The CVI Range had excellent internal consistency, inter-rater, and test-retest reliability. In addition, the two parts of The CVI Range consistently agreed on placement of the child into phases. Implications of these findings and directions for future research will be discussed in Chapter 5.

CHAPTER V

DISCUSSION

Children identified as visually impaired under the Individuals with Disabilities Education Act (IDEA) need to have a functional vision evaluation to determine how the visual impairment affects educational performance. Based on an extensive review of the literature, most current functional vision assessments have been based on the needs of children with ocular visual impairments (children with damage to the eye structures). Researchers have consistently documented that children with visual impairment due to brain damage, or cortical visual impairment (CVI), have unique vision characteristics that are often different from children with ocular visual impairments. Given this situation, Roman-Lantzy (2007) developed The CVI Range for conducting a functional vision assessment of children with CVI. The purpose of this study was to determine the reliability of The CVI Range and results of this study indicated that this assessment has good inter-rater, test-retest, and internal consistency reliability. Implication of these findings will be discussed in the following sections. Discussion will include: validity of The CVI Range; implications for assessment and intervention of children with CVI; and, future training and research needs in the field.

Validity of The CVI Range

For any measure to be useful in research and practice, it must provide results that are consistent as well as results that are representative of the construct of interest. In other words, assessments must be reliable and valid. The reliability of The CVI Range has been established in this study. The validity of The CVI Range has previously been addressed

in several ways. First, results of this study indicated that Cronbach's alpha was very high, which is often an indicator that a scale is measuring a single construct. The CVI Range is designed to measure severity of CVI and the obtained alpha score indicates that there is a single underlying construct that is addressed by these characteristics.

Roman-Lantzy has examined the validity of The CVI Range in two ways. First, she met with Dr. Jan and his colleagues to discuss the assessment and get expert opinion about the content of the scale (Roman-Lantzy, personal communication, September 17, 2007). In addition, her dissertation involved interviewing parents about visual characteristics of their children. Through the interview she identified a number of characteristics that could differentiate children with CVI from children with ocular visual impairment (Roman, 1996). The actual interview questions that represented these characteristics were chosen by a cohort of experts, thus the interview had content validity. As part of the study she completed an observation of children with CVI using the same characteristics as guidelines (e.g., color, movement, distance). She demonstrated that the characteristics could be described by parents in interviews and observed in children by professionals trained to observe those same characteristics. The behavioral observations had concurrent validity with the interview. The CVI Range was based on the characteristics identified in the interview and behavioral observation checklist. Having a functional vision assessment that is reliable and valid has implications for research and practice.

Use of The CVI Range

Cortical visual impairment is the most frequent cause of visual impairment in young children. The CVI Range is an appropriate functional vision assessment for

children with CVI and this study demonstrates that it is a reliable instrument. The CVI Range needs to be consistently used as one of the assessments to determine the child's vision needs when the child has cortical visual impairment. In the studies reviewed for this paper, no one who assessed or designed interventions for children with CVI used this assessment as a measure of functional vision. This could be in part due to the fact that this is a recent assessment. However, no one described a functional vision assessment that addressed any of the known characteristics of children with CVI. This research demonstrates that The CVI Range has good psychometric properties and is therefore a good instrument to use as a functional vision assessment for children with CVI.

Results of The CVI Range indicate which of the characteristics of CVI are interfering with the child's use of vision. The CVI Range thus gives valuable information about how materials and the environment need to be adapted for the child to use his or her vision. For example, if the examiner discovers that the child responds best to certain colors, or that the child needs to be in a supported position to use vision, that information is critical in designing appropriate interventions. Each characteristic that is affecting functional vision needs to be addressed in designing interventions.

Training and The CVI Range

The inter-rater reliability in this study was very high ($r = .98$), however, the assessments were conducted by highly trained CVI Mentors. Each mentor had participated in 5 years of training with Roman-Lantzy and conducted numerous CVI assessments. While this is one of the strengths of the study, it can also be seen as a limitation. When considering to whom these results can generalize, there are few individuals who may use this instrument who have such a high level of training and

supervision. With confidence the conclusion can be made that well trained evaluators can score The CVI Range reliably. The question then becomes, how much training is enough? Does the training have to be conducted by Roman-Lantzy? The CVI Range is in Roman-Lantzy's book, *Cortical Visual Impairment: An Approach to Assessment and Intervention* (2007), however the results of an assessment given by someone who only reads the book may be very different from the results of someone who has trained extensively with Dr. Roman-Lantzy. This does not make the instrument unreliable; it only means that the question of training must be addressed in future research. There are several ways of addressing the training issues raised by this study.

One way to address training is to check the reliability of evaluators who have varying levels of training. How much training, with how much supervision, and how much practice is needed to score The CVI Range reliably with Roman-Lantzy or with a well trained evaluator, such as one of the CVI mentors? This is a question that can be addressed through systematic research with future trainees. The Multi-state CVI Mentorship Project is continuing training of a new cohort of professionals, but for a shorter period of time, i.e., two years. These new evaluators can and should be checked periodically for reliability with Dr. Roman-Lantzy and/or with one of the CVI mentors. This strategy will help to document training needs around The CVI Range, however, it does not address training for professionals outside of these four states.

Another strategy to insure properly trained professionals would be to develop commercially available training materials including videos of children with CVI. Training materials should include training relative to the 10 characteristics addressed on The CVI Range, as well as interview, observation, and direct assessment strategies.

Especially in respect to direct assessment strategies, there needs to be video clips with the opportunity for the trainee to score the behavior they are seeing and then check their observations with a standard, such as Dr. Roman-Lantzy's score. There also needs to be videos of complete assessments and a chance for the trainee to check for reliability. Because children with CVI vary in terms of severity, training materials must cover all levels or phases of CVI. Production of training materials is a very involved project, however it is necessary to find some way to ensure that professionals who use The CVI Range meet the criteria for a conducting a quality assessment. Intervention and documentation of progress are dependent on quality assessments.

A final training need that emerged from this study was not related to administration of The CVI Range, but instead to demographic data collected about all of the children. All of the children in this study had other disabilities in addition to CVI. They all had multiple service providers and received many different services including early intervention, special education, PT, OT, speech therapy, vision services, and hearing services. Children with CVI are served by multiple professionals from multiple disciplines. This fact highlights the need for training about CVI across disciplines. While not everyone may need to be qualified to administer The CVI Range, everyone on the child's team needs to understand the results of the CVI assessment. All professionals as well as family members need to understand the characteristics of CVI that affect a child's vision, and how interventions are designed to meet vision needs. A child needs to use vision across all activities of the day. The speech therapist needs to understand how the child's vision affects the development of a communication system. Can the child look at pictures? Do object symbols need to be presented on a plain background? The teacher

needs to know how to present materials in a group setting. Does the wall behind her need to be plain? Does the child's cubby need to be outlined in a specific color so that the child can find it and be successful in finding his or her coat at the end of the day? The PT needs to know that the child cannot use vision when in a challenging position, so while working on therapy activities music may be a more appropriate motivator.

Many of the children in this study had medical needs and as a result were also involved with many medical professionals. Medical professionals were often the first people who offered information to families. As part of a final evaluation of the past CVI Multi-state Mentorship Project, several families were interviewed about how the project services had helped their child and what improvements need to be made for the next CVI project. Families often commented that medical professionals had not provided them with any information about CVI. If information was provided, they did not provide information about interventions that might work or professionals in special education or early intervention who might be able to help. Again, medical professionals may not need to know how to administer The CVI Range, but they do need to understand the information covered by the assessment and that there are resources available to families. They also need to understand that intervention can make a difference for children with CVI. Families report being told their child has CVI and there is nothing that can be done, or CVI means to treat your child as blind and focus on other senses. Training around The CVI Range needs to include professionals from all disciplines, developmental and medical, that serve children with CVI and their families.

The Stability of Visual Functioning in Children with CVI

One of the most significant findings of this research study was the test-retest reliability ($r = .99$). For a long time, the conventional wisdom from the field has been that children with CVI have vision that fluctuates from day to day or even hour to hour (Dutton, 2003, 2004, 2006; Good & Hoyt, 1989; Jan, Good, & Hoyt, 2006; Jan, Groenveld, & Anderson, 1993; Jan, Groenveld, & Sykanda, 1990; Jan, Groenveld, Sykanda, & Hoyt, 1987; Whiting, et al., 1985). The test-retest reliability finding in this study is in direct opposition to previous findings. Given stability in examiners, settings, materials, and time of day, children with CVI in this study did not have functional vision that fluctuated. This was true for children across all three phases of CVI.

The test-retest reliability finding has several implications. First, the finding needs to be replicated. Another study of test-retest reliability needs to be conducted with more children at each level of CVI. The current study had children at all levels, but only included 20 children. The results need to be replicated with a larger sample of children.

Another implication of this finding is that there may be another plausible explanation for previous findings of variable visual functioning. Looking at the results of an assessment using The CVI Range, one can see that any number of factors can affect a child's visual functioning. Especially at the more severe levels of CVI, there are multiple factors which have an influence on the child's use of vision. In earlier observations researchers did not take into account any reasons for the child's variable responses. In addition, they did not use a systematic measure of functional vision such as The CVI Range. They used informal observations without a measure to quantify their findings. The child may have looked at a toy at home, but in the doctor's office with a different

toy, novelty was affecting visual functioning. The child may have looked at a cup when the mother was wearing a plain shirt and did not look when presented with the same cup when the mother was wearing a flower print shirt, thus increasing the complexity of the background. In both cases, the conclusion could be drawn that the child looks sometimes, but not other times, while in fact, it was something in the environment more than the child's vision that changed. When more of the characteristics are interfering with visual functioning, there is a greater chance that there are outside factors influencing the child's vision, not that the vision itself is variable.

The test-retest finding also has implications for assessment and intervention. When assessment is used for pre- and post-test measures, conditions need to be similar in both situations. If assessment results are used to make decisions about the effectiveness of an intervention, there needs to be stability in assessment conditions. Understanding that the child's vision is stable also has an effect on how families, teachers, and therapists view the child. If significant adults hold the belief that the child has stable vision, they are more likely to take responsibility for engineering the environment for the child's success. This author has personally observed that when a child is not looking at something, adults who believe the child's vision is variable are likely to conclude, "Well, that's what kids with CVI are like. Sometimes they look, sometimes they don't." If they do not believe that vision is variable, adults are more likely to examine other factors in the environment and modify them so the child can be successful. Understanding that vision is stable, but can be influenced by many things in the environment, puts the responsibility for success on the adult, not the child.

Directions for Future Research

Several areas of possible research have been identified in this chapter including research related to training evaluators in administration of The CVI Range and the stability of visual functioning in children with CVI. Another area of research that has not been adequately addressed in previous studies is which interventions are effective for children with CVI. Future research needs to focus on building a body of evidenced-based literature about effective interventions for children with CVI.

Early research with brain development and visual skills came from the work of Hubel and Wiesel (1970) and newborn kittens deprived of visual input. From their work came the idea of critical periods in visual development. Subsequent research with cats and monkeys indicated that the effects of vision deprivation were not permanent or irreversible (Bruer, 2001; Chow & Stewart, 1972; Harweth, Smith, Crawford, & vanNoorden, 1989). Due to the ability of animals to learn to use vision, the idea emerged that certain experiences at a designated point in time can have a profound effect on development in that area. It is generally accepted, due to brain plasticity, that for children, the earlier years are the most important for change in vision (Hoyt, 2003). Previous research has indicated that children with CVI often demonstrate improved vision (Afshari, Afshari, & Fulton, 2001; Good, Jan, Burden, Skoczenski, & Candy, 2001; Groenveld, Jan, & Leader, 1990; Hoyt, 2003; Huo, Burden, Hoyt, & Good, 1999; Khetpal & Donahue, 2007; Matsuba & Jan, 2006). This change is usually attributed to plasticity and the brain's ability to develop visual functions despite damage to the visual pathways. The potential for change in children with CVI makes it critical to determine which factors and what interventions can facilitate visual improvement. Most of the

research on child change was completed through record reviews in large medical clinics. None of the research that described change in vision documented what intervention strategies facilitated or inhibited improvement.

Effective interventions were examined in very few studies (Baker-Nobles & Rutherford, 1995; Farrenkopf, McGregor, Nes, & Koenig, 1997; Lueck, Dornbusch, & Hart, 1999). While these early results were promising, future studies need to look at a wider variety of children at varying levels of severity of CVI. There has to be a recognized measure that can document improvement such as The CVI Range. Interventions should be designed based on the results of a quality assessment. Systematic studies that match child characteristics to interventions and documents progress across time will lead to a body of literature about practices that are effective. Only with good assessment to design intervention and a way to monitor child change, can any intervention be called evidenced-based.

Conclusion

CVI is the leading cause of visual impairment in young children. Early research focused on understanding who the children were – causes of CVI and characteristics of children with CVI. Early assessment studies focused on medical diagnosis and typical measures of vision functions such as acuity. As more of these children have been identified in early intervention and special education, the need to understand functional vision and develop appropriate interventions for children with CVI has increased. This study represents one step forward in identifying the needs of children with CVI. The CVI Range has good inter-rater, test-retest, and internal consistency reliability and as such can be used with confidence as a quality functional vision assessment for children with CVI.

A good functional vision assessment is the bridge to the next research focus in the field which is the development of evidenced-based interventions. Children with CVI can and often do make progress in vision. Professionals owe it to them to know what works and to implement strategies that help children improve vision functioning.

Appendix A

CVI Forms

The CVI Range

The CVI Resolution Chart

Essential Forms

THE CVI RANGE

Student/child's name: _____ Age: _____

Evaluator(s): _____ Evaluation Date: _____

This assessment protocol is intended for multiple evaluations over a period of time.
Suggested scoring (no less than three times per school year):

- a. Initial assessment (red)
- b. Second assessment (blue)
- c. Third assessment (green)

Further assessments will require a new form.

Totals:	Evaluation #1 (red)	Evaluation #2 (blue)	Evaluation #3 (green)
1. Range for Rating 1			
2. Total for Rating 2			
3. Combine both ratings to get overall CVI Range			

0 1 2 3 4 5 6 7 8 9 10

No functional Vision Typical or near-typical Visual functioning (continued)

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The CVI Range: Across-CVI Characteristics Assessment Method

Rating 1

Rate the following statements as related to the student/child's visual behaviors by marking the appropriate column to indicate the methods used to support the scores:

O = information obtained through observation of the child/student

I = information obtained through interview regarding the child/student

D = information obtained through direct contact with the child/student

In the remaining columns, indicate the assessed degree of the CVI characteristic:

- R The statement represents a resolved visual behavior
- + Describes current functioning of student/child
- +/- Partially describes student/child
- - Does not apply to student/child

CVI Range 1-2: Student functions with minimal visual responses

O	I	D	R	+	+/-	-	
							May localize, but no appropriate fixations on objects or faces
							Consistently attentive to lights or perhaps ceiling fans
							Prolonged periods of latency in visual tasks
							Responds only in strictly controlled environments
							Objects viewed are a single color
							Objects viewed have movement and/or shiny or reflective properties
							Visually attends in near space only
							No blink in response to touch or visual threat
							No regard of the human face

(continued)

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CVI Range 3-4: Student functions with more consistent visual responses

O	I	D	R	+	+/-	-	
							Visually fixates when the environment is controlled
							Less attracted to lights: can be redirected
							Latency slightly decreases after periods of consistent viewing
							May look at novel objects if they share characteristics of familiar objects
							Blinks in response to touch and/or visual threat, but the responses may be latent and/or inconsistent
							Has "favorite" color
							Shows strong visual field preferences
							May notice moving objects at 2 to 3 feet
							Look and touch completed as separate events

CVI Range 5-6: Student uses vision for functional tasks

O	I	D	R	+	+/-	-	
							Objects viewed may have two to three colors
							Light is no longer a distractor
							Latency present only when the student is tired, stressed, or overstimulated
							Movement continues to be an important factor for visual attention
							Student tolerates low levels of background noise
							Blink response to touch is consistently present
							Blink response to visual threat is intermittently present
							Visual attention now extends beyond near space, up to 4 to 6 feet
							May regard familiar faces when voice does not compete

(continued)

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CVI Range 7-8: Student demonstrates visual curiosity

O	I	D	R	+	+/-	-	
							Selection of toys or objects is less restricted; requires one to two sessions of “warm up”
							Competing auditory stimuli tolerated during periods of viewing; the student may now maintain visual attention on objects that produce music
							Blink response to visual threat consistently present
							Latency rarely present
							Visual attention extends to 10 feet with targets that produce movement
							Movement not required for attention at near distance
							Smiles at/regards familiar and new faces
							May enjoy regarding self in mirror
							Most high-contrast colors and/or familiar patterns regarded
							Simple books, picture cards, or symbols regarded

(continued)

CVI Range 9-10: Student spontaneously uses vision for most functional activities

O	I	D	R	+	+/-	-	
							Selection of toys or objects not restricted
							Only the most complex environments affect visual response
							Latency resolved
							No color or pattern preference
							Visual attention extends beyond 20 feet
							Views books or other two-dimensional materials, simple images
							Uses vision to imitate actions
							Demonstrates memory of visual events
							Displays typical visual-social responses
							Visual fields unrestricted
							Look and reach completed as a single action
							Attends to two-dimensional images against complex backgrounds

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The CVI Range: Within-CVI Characteristics Assessment Method

Rating II

Determine the level of CVI present or resolved in the 10 categories below and add to obtain total score. Rate the following CVI categories as related to the student/child's visual behaviors by circling the appropriate number (the CVI Resolution Chart may be useful as a scoring guide):

- 0 Not resolved; usually or always a factor affecting visual functioning
- .25 Resolving
- .5 Resolving; sometimes a factor affecting visual functioning
- .75 Resolving
- 1 Resolved; not a factor affecting visual functioning

	Not resolved		Resolving		Resolved
1. Color preference	0	.25	.5	.75	1
Comments:					
2. Need for movement	0	.25	.5	.75	1
Comments:					
3. Visual latency	0	.25	.5	.75	1
Comments:					
4. Visual field preference	0	.25	.5	.75	1
Comments:					
5. Difficulties with visual complexities	0	.25	.5	.75	1
Comments:					
6. Light-gazing and nonpurposeful gaze	0	.25	.5	.75	1
Comments:					
7. Difficulty with distance viewing	0	.25	.5	.75	1
Comments:					

(continued)

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	Not resolved		Resolving		Resolved
8. Atypical visual reflexes	0	.25	.5	.75	1
Comments:					
9. Difficulty with visual novelty	0	.25	.5	.75	1
Comments:					
10. Absence of visually guided reach	0	.25	.5	.75	1
Comments:					

CVI Resolution Chart

Date _____ Student's Name _____ Evaluator _____

Use the following chart to help develop areas of needs for development of IEP goals and objectives.

	Phase I Building Visual Behavior Level I Environmental Considerations		Phase II Integrating Vision with Function Level II Environmental Considerations		Phase III Resolution of CVI Characteristics Level III Environmental Considerations
CVI Characteristics	Range 1-2 (0)	Range 3-4 (.25)	Range 5-6 (.50)	Range 7-8 (.75)	Range 9-10 (1)
Color preference	Objects viewed are generally single color	Has "favorite" color	Objects may have 2-3 colors	More colors, familiar patterns regarded	No color or pattern preference
Need for movement	Objects viewed generally have movement/reflective properties	More consistent localization, brief fixations on movement & reflective materials	Movement continues to be an important factor to initiate visual attention	Movement not required for attention at near	Typical responses to moving targets
Visual latency	Prolonged periods of visual latency	Latency slightly decreases after periods of consistent viewing	Latency present only when student is tired, stressed, or over stimulated	Latency rarely present	Latency resolved
Visual field preferences	Distinct field dependency	Shows visual field preferences	Field preferences decreasing with familiar inputs	May alternate use of right and left fields	Visual fields unrestricted

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	Phase I Building Visual Behavior Level I Environmental Considerations	Phase II Integrating Vision with Function Level II Environmental Considerations	Phase III Resolution of CVI Characteristics Level III Environmental Considerations		
CVI Characteristics	Range 1-2 (0)	Range 3-4 (.25)	Range 5-6 (.50)	Range 7-8 (.75)	Range 9-10 (1)
Difficulties with visual complexity	Responds only in strictly controlled environments	Visually fixates when environment is controlled	Student tolerates low levels of familiar background noise Regards familiar faces when voice does not compete	Competing auditory stimuli tolerated during periods of viewing - student may now maintain visual attention on music toys Views simple books/symbols Smiles at/regards familiar and new faces	Only the most complex visual environments affect visual response Views books or other 2-dimensional materials Typical visual-social responses
Light-gazing and nonpurposeful gaze	May localize briefly but no prolonged fixations on objects or faces Overly attentive to lights or perhaps ceiling fans	Less attracted to lights - can be redirected to other targets	Light is no longer a distractor		

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Difficulty with distance viewing	Visually attends in near space only	Occasional visual attention on familiar, moving or large targets at 2-3 feet	Visual attention extends beyond near space, up to 4-6 feet	Visual attention extends to 10 feet with targets that produce movement	Visual attention extends beyond 20 feet Demonstrates memory of visual events
Atypical visual reflexes	No blink in response to touch and/or visual threat	Blinks in response to touch but response may be latent	Blink response to touch consistently present Visual threat response intermittently present	Visual threat response consistently present (both near 90% resolved)	Visual reflexes always present, resolved
Difficulty with visual novelty	Only favorite or known objects solicit visual attention	May tolerate novel objects if the novel objects share characteristics of familiar objects	Use of "known" objects to initiate looking sequence	Selection of objects less restricted, requires 1-2 sessions of "warm up" time	Selection of objects not restricted
Absence of visually guided reach	Look & touch occur as separate functions Large &/or moving targets	Look & touch on smaller objects that are familiar, lighted, or reflective Look and touch are still separate	Visually guided reach with familiar objects or "favorite" color	Look and touch occur in rapid sequence but not always together	Look and touch consistently

Key:

- Draw an "X" through boxes that represent resolved visual behaviors
- Use highlighter to outline boxes describing current visual functioning
- Draw an "O" in boxes describing visual skills that may never resolve because of co-existing ocular conditions

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Appendix B
Causes of CVI

Appendix B: Causes of CVI

Authors	Subjects	Location and dates	Causes of CVI (arranged in order of frequency)
Whiting, Jan, Wong, Flodmark, Farrell, & McCormick (1985)	50 children, 6 months to 19 years old	Children's Hospital, British Columbia Seen 1970-1984	Prenatal - toxemia - intrauterine infection - cerebral dysgenesis Perinatal - asphyxia - hemorrhage - meningitis Acquired - shunt - trauma - meningitis - thromobosis - cardiac arrest
Jan, Groenveld, Sykanda, & Hoyt (1987)	50 children, 6 months to 17 years old	Children's Hospital, British Columbia Seen 1983-1985	Pre- and perinatal - asphyxia - cerebral dysgenesis - cerebral hemorrhage - infection Acquired - shunt failure - asphyxia - injury - dehydration
Jan, Groenveld, & Sykanda (1990)	69 children, 4 months to 11 years old	Children's Hospital, British Columbia Seen 1987-1989	Asphyxia CNS anomalies Injury Infection Other
Huo, Burden, Hoyt, & Good (1999)	170 children, 3 months to 15 years old	Pediatric ophthalmology practice in CA Seen 1979-1994	Perinatal hypoxia Cerebral vascular accident Meningitis Acquired hypoxia
Khetpal & Donahue (2007)	98 children, 2 months to 19 years old	Children's Center, Vanderbilt Hospital, TN Seen 2002-2005	Hypoxia Prematurity Hydrocephalus CNS anomalies Seizures

Appendix C
Specific Brain Injury and CVI

Appendix C: Specific brain injury and CVI

Author	Subjects	Brain injury	Results
Pike, et al. (1994)	42 premature children, at least 2 years old at time of study	PVL IVH Cerebral infarcts	Children with PVL and cerebral infarcts were more likely to have CVI; Overall, 38 of 42 children had some level of CVI
Jacobson, Ek, Fernell, Flodmark, & Broberger (1996)	13 children born premature, ages 4-14 years old	PVL	All children had CVI
Lanzi, et al. (1998)	38 children born premature, ages 20-66 months old, diagnosed with cerebral palsy	PVL	66% of children had CVI; Severity of PVL was correlated with severity of CVI
Cioni, Fazzi, Coluccini, Bartalena, Boldrini, & van Hof-van Duin, (1997)	48 premature children, 24 months old	14 with severe PVL 34 with moderate PVL	11 of 14 children with severe PVL had CVI; 16 of 34 children with moderate PVL had CVI
Mercuri, et al. (1997)	31 full term children, ages 5-31 months old	HIE	20 of 31 children had CVI; Children with more severe HIE had worse CVI
Brodsky, Fray, & Glasier (2002)	100 children with CVI (record review), ages not reported	Cortical damage (gray matter) Subcortical damage (white matter)	Different gestational ages of child at time of insult results in different pattern of brain damage; Preterm infants have white matter damage or PVL; Term infants have gray matter or cortical damage
Hoyt (2003)	96 children with CVI (record review); ages	PVL Damage to visual cortex	Children with PVL demonstrate less improvement in vision than children with damage to visual cortex

PVL – periventricular leucomalacia;
 IVH – intraventricular hemorrhage;
 HIE – hypoxic-ischemic encephalopathy

Appendix D

Characteristics of Children with CVI

Appendix D: Characteristics of children with CVI

Research studies by location and authors	Subjects	Visual characteristics
Visually Impaired Program, British Columbia, Canada Whiting, Jan, Wong, Flodmark, Farrell, & McCormick (1985) Jan, Groenveld, Sykanda, & Hoyt, (1987) Good & Hoyt (1989) Jan, Groenveld, & Sykanda (1990) Jan, Groenveld, & Anderson (1993) Jan, Good, & Hoyt (2004)	169 children, ages 6 months to 19 years old	<ul style="list-style-type: none"> - attention to movement - mobile children do not bump into objects - do not look blind, e.g. malformed eyes - visually inattentive - lack of visual curiosity - no nystagmus - no visual self-stimulation - light gazing in 60% - photophobia in 33% - variable visual functioning - look away when touching - retains color vision - better vision in familiar environments - better vision with familiar objects - field loss - close viewing - presence of other neurological problems - can have co-existing ocular conditions
Vision Assessment Clinic Glasgow, Scotland Dutton, et al. (1996) Dutton (2003, 2004, 2006)	90 children, ages less than 1 year to 16 years old	<ul style="list-style-type: none"> - range of visual functioning from mild to total blindness - visual functioning affected by fatigue and distractions - problems with recognition of objects and people - problems with orientation or getting lost - poor depth perception - difficulty seeing moving objects - impaired simultaneous perception or inability to see more than one part of a whole - field losses

Appendix D: Characteristics of children with CVI (continued)

Tomtebodas Resource Center Stockholm, Sweden Jacobson, Ek, Fernell, Flodmark, & Broberger (1996)	13 children, ages 4-14 years old	<ul style="list-style-type: none"> - crowding effect - field restrictions - normal color vision - normal contrast sensitivity - impaired form identification - easily fatigued with visual tasks - many children also have optic nerve atrophy
Hammersmith Hospital UK Pike, et al. (1994)	42 children, age 24 months old	<ul style="list-style-type: none"> - color vision preserved - difficulty with complex pictures - single symbols easier to identify (crowding)
Erasmus University The Netherlands Groenendaal & van Hof-van Duin (1992)	38 children, ages 1.5 months to 19 years old	<ul style="list-style-type: none"> - visual threat reflex absent in 33% - field loss common - no fixation in 50% - reduced acuity in children who could perform acuity tasks
California Cohen-Maitre & Haerich, 2005	11 children, ages 18-72 months old	<ul style="list-style-type: none"> - color and movement are salient features of getting and maintaining visual attention

Appendix E

Testing of Vision Functions in Children with CVI

Appendix E: Testing of vision functions in children with CVI

Vision function	Author	Subjects	Results
Grating acuity measured with visual evoked potential (VEP)	Granet, Hertle, Quinn, & Breton (1993)	10 children, ages 5-48 months old	VEP results did not predict improvements at follow up
	Clarke, Mitchell, & Gibson (1997)	44 children, ages not given	VEP results did not predict improvements at follow up
Grating acuity measured with Sweep VEP	Good (2001)	41 children, ages 6 months to 16 years old	Sweep VEP is a reliable and valid measure of vision in children with CVI
	Good & Huo (2006)	20 children, ages 7 months to 4 years old	Luminance had effect on acuity for children with CVI
	Watson, Orel-Bixler, & Haegerstrom-Portnoy (2007)	34 children, ages 1 to 16 years old	Sweep VEP provides a quantitative measure to document progress in children with CVI
Vernier acuity measured with sweep VEP	Skoczenski & Good (2004)	35 children, ages 4 months to 16 years old	Vernier acuity is lower than grating acuity in children with CVI
Grating acuity measured with preferential looking (PL)/Teller Acuity Cards (TAC)	Birch & Bane (1991)	132 children, ages birth to 12 years old for initial evaluation; 62 of those children had follow up data	PL acuity correlated with ability to fix and follow; Positive correlation between initial and follow up TAC acuity
	van Hof-van Duin, Bertuccelli, Fazzi, Romano, & Boldrini (1998)	39 children, ages 1-2 years old at initial and 5 years old at follow up	TAC predictive of outcome for only 27 of 39 children

Appendix E: Testing of Vision Functions in Children with CVI (continued)

Comparison of VEP and TAC grating acuity	Westall, Ainsworth, & Buncic (2000)	175 children with ocular or cortical visual impairment, ages 3-13 years old	48% of children had discrepancies in TAC and VEP; Children with severe disabilities more likely to have a discrepancy
	Weiss, Kelly, & Phillips (2001)	31 children, age less than 1 year old - 14 dev. normal - 17 dev. delayed	Dev. normal infants had normal VEP and TAC; All delayed infants had abnormal VEPs; some had abnormal TAC
	Lim, Soul, Hansen, Mayer, Moskowitz, & Fulton (2005)	19 children, ages 6 months to 6 years old	TAC and VEP below age norms; TAC lower acuity than VEP; many children had large discrepancy
Comparison of grating (TAC) and ototype acuity (Landolt-C)	Stiers, Vanderkelen, & Vandebussche (2004)	81 children, ages 5-24 years old - 14 with CVI - 48 with ocular VI - 19 with both	Grating acuity is better than ototype acuity; children with CVI have greatest discrepancy
OKN as measure of visual awareness	Groenendaal & van Hof-van Duin (1992)	38 children, ages 7 weeks to 19 years old	32 of 38 children had abnormal OKN
	Cioni, Fazzi, Coluccini, Bartalena, Boldrini, & van Hof-van Duin (1997)	48 children, age 24 months old	17 of 48 children with abnormal OKN
	Brodsky, Fray, & Glasier (2002)	100 children, ages not reported	OKN results not reported
Stygar ball test of acuity	Groenendaal & van Hof-van Duin (1992)	38 children, ages 7 weeks to 19 years old	Stygar test given to children who could not respond to other acuity measures, results not given

 Appendix E: Testing of vision functions in children with CVI (continued)

Stygar ball test of acuity	Pike, et al. (1994)	42 children, age 24 months old	Stygar test given to children who could not respond to other acuity measures; results not given
Visual fields tested with arc perimetry	Cioni, Fazzi, Coluccini, Bartalena, Boldrini, & van Hof-van Duin (1997)	48 children with PVL, age 24 months old	9 of 14 children with severe PVL had field loss; 4 of 34 children with moderate PVL had field loss
	Jan, Groenveld, & Anderson (1993)	35 children, ages not given	Results of arc perimetry not reported
Visual fields tested with confrontational testing	Dutton, et al. (1996)	90 children, ages less than 1 year to 16 years old	52% of the children had a field loss
	Jacobson, Ek, Fernell, Flodmark, & Broberger (1996)	13 children with PVL, ages 4-14 years old	13 of 13 children had field loss

Appendix F
Outcomes for Children with CVI

Appendix F: Outcomes for children with CVI

Authors	Subjects	Outcomes
Chen, Weinberg, Catalano, Simon, & Wagle (1992)	30 children – initial visit in first year; Follow up at least 12 months later, (mean length of follow up - 43 months)	15 of 30 children developed object vision (ability to recognize faces or toys)
Groenendaal & van Hof-van Duin (1998)	22 children seen more than one time, ages 7 weeks to 17 years Length of follow up not reported	All children improved in vision skills (acuity, visual threat, visual fields)
Castano, Lyons, Jan, & Connolly (2000)	10 children – initial visit at 2 to 8 months of age, length of follow up ranged from 14 months to 6 years	50% showed improvement in vision skills (e.g., acuity, visual attention)
Huo, Burden, Hoyt, & Good (1999)	Record review of 170 children seen more than once, average length of follow up 5.9 years	60% of children showed improvement on Huo scale of functional vision
Hoyt (2003)	Further analyzed data to compare children with PVL vs. children with visual cortex damage	Children with visual cortex damage made more improvements than children with PVL
Matsuba & Jan (2006)	Record review of 259 children Follow up was at least two years from initial visit; Children divided by initial visit prior to age 3 or older than 3 years	46% of children improved in visual acuity as measured by TAC; Children seen before 3 showed more improvement
Khetpal & Donahue (2007)	Record review of 52 children – initial visit at ages 2 months to 19 years old Average length of follow up: 2.33 years	- 40% had no improvement; - 34% had minimal improvement; - 17% had mild improvement; - 6% had significant improvement; Improvement measured with modified Huo scale

Appendix G
Informed Consent

CONSENT FORM

Project Title	<i>Reliability of the CVI Range, by Dr. Chris Roman</i>
WHY IS THIS RESEARCH BEING DONE?	<i>This is a research project being conducted by Dr. Diane Kelly and Ms. Sandra Newcomb at the University of Maryland, College Park. We are inviting you to participate in this research project because you have a child who is being assessed with the CVI Range. The purpose of this research project is to examine the CVI Range to determine if it is an assessment that consistently measures the characteristics of CVI and to determine if multiple raters are consistent in rating characteristics of CVI.</i>
What will I be asked to do?	<i>If you agree to participate, your child's CVI assessment will be videotaped. More than one observer may be present during the assessment. Your child may be assessed on two different days, approximately 1-2 weeks apart. The CVI assessment involves observations of your child with various visual stimuli (toys or familiar household objects) to determine what he looks at, and what environmental supports are needed to help him use his vision. CVI assessments take from 1-2 hours.</i>
What about confidentiality?	<p><i>We will do our best to keep your personal information confidential. To help protect your confidentiality, assessment data will be coded with a number for identification. All forms will be locked and stored in a secure place, and only project personnel will have access to the data.</i></p> <p><i>This research project involves making videotapes of your child's assessment. The tapes will be used to determine if multiple raters are consistent in rating characteristics of CVI. The videotapes will be locked in a secure place and only project personnel will have access to them.</i></p> <p><input type="checkbox"/> <i>I agree to have my child videotaped during participation in this study.</i></p> <p><input type="checkbox"/> <i>I do not agree to have my child videotaped during participation in this study.</i></p> <p><i>If we write a report or article about this research project, your identity will be protected to the maximum extent possible. Your information may be shared with representatives of the University of Maryland, College Park or governmental authorities if you or someone else is in danger or if we are required to do so by law.</i></p>

Project Title	<i>Reliability of the CVI Range, by Dr. Chris Roman</i>
What are the risks of this research?	<i>There are no known risks associated with participating in this research project.</i>
What are the benefits of this research?	<i>This research is not designed to help you personally, but the results may help the investigator learn more about assessment of children with CVI. We hope that, in the future, other people might benefit from this study through improved understanding of assessment of children with CVI.</i>
Do I have to be in this research? May I stop participating at any time?	<i>Your participation in this research is completely voluntary. You may choose not to take part at all. If you decide to participate in this research, you may stop participating at any time. If you decide not to participate in this study or if you stop participating at any time, it will not affect your child's assessment or intervention programming by the CVI Mentorship Project.</i>
What if I have questions?	<p><i>This research is being conducted by Dr. Diane Kelly and Sandra Newcomb, Department of Special Education at the University of Maryland, College Park. If you have any questions about the research study itself, please contact Dr. Kelly at 301-405-7915 or Ms. Newcomb at 301-405-6476.</i></p> <p><i>If you have questions about your rights as a research subject or wish to report a research-related injury, please contact: Institutional Review Board Office, University of Maryland, College Park, Maryland, 20742; (e-mail) irb@deans.umd.edu; (telephone) 301-405-0678</i></p> <p><i>This research has been reviewed according to the University of Maryland, College Park IRB procedures for research involving human subjects.</i></p>
Consent	<i>Your signature indicates that: the research has been explained to you; your questions have been fully answered; and you freely and voluntarily choose to participate in this research project.</i>
Signature and Date	Name of Child:
	Name of Parent/Guardian:
	SIGNATURE OF Parent/Guardian:
	Date:

Appendix H
Glossary of Terms

Glossary of terms

Acuity – the ability to discriminate and recognize detail

Otype acuity – ability to distinguish letters or shapes, e.g., Snellen chart

Grating acuity – ability to perceive separate elements of a stimulus, e.g. black and white stripes; also known as resolution acuity

Resolution acuity – ability to perceive separate elements of a stimulus, e.g. black and white stripes; also known as grating acuity

Vernier acuity – ability to localize pattern elements or detect discontinuity in a line or misalignment in segment of a line, e.g., Landolt-C

Asphyxia – lack of oxygen or excess of carbon dioxide usually caused by not breathing or inadequate oxygen supply

Cataracts – clouding of the eye lens which obscures vision

Cerebral vascular accident/infarct – focal area of bleeding and cell death in the brain

Cerebral dysgenesis – lack of normal development of the brain

Cortical visual impairment – loss of vision due to damage or malformation in the brain

Crowding – inability to perceive objects, letters, or words spaced close together

Hypoxia – lack of oxygen

Hypoxic-ischemic damage – brain damage due to lack of oxygen and lack of blood flow to the brain

Hypoxic-ischemic encephalopathy – profuse brain damage due to the combination of lack of oxygen and lack of blood flow

Intraventricular hemorrhage – bleeding into the ventricles of the brain

Ischemia – lack of or inadequate blood flow

Nystagmus – involuntary movement of eyes, usually an indicator of poor vision

Optic nerve atrophy – damage to the optic nerve

Optic nerve hypoplasia – underdeveloped optic nerve

Optokinetic nystagmus (OKN) – test of child's awareness of moving black lines

Periventricular leucomalacia – damage to the white matter surrounding the ventricles in the brain

Perfusion – blood flow which delivers oxygen and nutrients

Retinopathy of prematurity – disorganized growth of retinal blood vessels due to prematurity which may result in scarring and retinal detachment

Visual evoked potential – electrophysiologic test of visual acuity

Visual evoked potential mapping – electrophysiologic test of brain's response to light over large portions of the brain.

Sweep VEP – electrophysiologic test of visual acuity using moving black and white lines

Watershed zone – area at the end of vascular system that is most susceptible to injury from lack of oxygen

Commonly used abbreviations

CP – cerebral palsy

CVI – cortical visual impairment

HIE – hypoxic ischemic encephalopathy

IVH – intraventricular hemorrhage

OKN – optokinetic nystagmus

ONA – optic nerve atrophy

ONH – optic nerve hypoplasia

PVL – periventricular leucomalacia

ROP – retinopathy of prematurity

TAC – Teller acuity cards

VEP – visual evoked potential

VEPM – visual evoked potential mapping

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