Original Article

Setting up of a cerebral visual impairment clinic for children: Challenges and future developments

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Aim: The aim of this study is to describe the setting up of a cerebral visual impairment (CVI) clinic in a tertiary care hospital in South India and to describe the spectrum of cases seen. **Materials and Methods:** The CVI clinic, set up in February 2011, receives interdisciplinary input from a core team involving a pediatrician, neurologist, psychiatrist, occupational therapist, pediatric ophthalmologist, and an optometrist. All children, <18 years of age, with cerebral palsy (CP), learning disability, autism, neurodegenerative diseases, and brain trauma are referred to the clinic for functional vision assessment and opinion for further management. **Results:** One thousand four hundred and seventy-eight patients were seen in the CVI clinic from February 2011 to September 2015. Eighty-five percent of the patients were from different parts of India. In the clinic, 61% had CP, 28% had seizure disorders, autism was seen in 9.5%, and learning disability, neurodegenerative conditions, and brain injury together constituted 1.5%. Most of the children (45%) had moderate CP. Forty percent of CVI was due to birth asphyxia, but about 20% did not have any known cause for CVI. Seventy percent of patients, who came back for follow-up, were carrying out the habilitation strategies suggested. **Conclusions:** Average attendance of over 300 new patients a year suggests a definite need for CVI clinics in the country. These children need specialized care to handle their complex needs. Although difficult to coordinate, an interdisciplinary team including the support groups and voluntary organizations is needed to facilitate the successful implementation of such specialized service.

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Vision is not just visual acuity or "sight;" it also entails how the person recognizes and understands the world around him. It comprises collection, processing, and coding of information.[1] The process of seeing involves conversion of the analog signal of the visual image on the retina to digital signals that are then transmitted to the brain to accord the experience of vision. The brain is thus the seeing organ of the body because vision in its broadest sense is accorded by multiple brain components[2] including white matter, occipital cortex, thalamus, brain stem,[3] cerebellum,[4] and the vestibular system.[5] Vision has two separate but interacting functions: (1) to perceive objects and their relations, which is the basis for the person's conscious experience of the world and visual cognitive function and (2) to visually guide movement and actions brought about by the motor system.[6,7] Visual functions served by the occipital lobes include visual acuity, contrast sensitivity, color vision, visual fields, and in the adjacent middle temporal lobes, perception of movement.[8] The posterior parietal lobes help with overviewing and creating an internal emulation of the visual scene, identifying an element within it, moving the gaze to that element, and guiding our movement through the surrounding environment.[7,9] The temporal lobes facilitate recognition of faces, facial expressions, objects, shapes, and route finding.[10] Damage to these retrogeniculate structures and higher visual areas in any combination or degree results in cerebral visual impairment (CVI). This comprises a complex primary visual and often cognitive cum perceptual dysfunction of the brain.[11,12] The two principal higher pathways affected by CVI are the occipitoparietal pathway (dorsal stream) and the occipitotemporal pathway (ventral stream).[13] The causes of CVI are many, but the most frequent cause is perinatal hypoxic-ischemic damage.[14,15] This causes the development of neuromotor disorders such as cerebral palsy (CP) and can affect geniculate/extrageniculate visual pathways and the visual association areas,[3] thus explaining the relationship between CP and CVI. Children with CP not only have motor dysfunction but also have problems with vision that is less evident and thus may go unidentified.[16,17] Early recognition and salient intervention founded upon facilitating communication, ensuring access, and enhancing mobility, clearly has the potential to avert significant developmental impairment.

Visual function assessment in these children needs to go beyond the standard visual acuity and visual field testing taught and practiced in eye departments across the country. Therefore, a pediatric ophthalmologist or optometrist without special training is ill-equipped to manage these patients. The purpose of a CVI clinic is to assess all aspects of functional vision enabling early recognition of visual difficulties due to

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CVI in children with CP and other similar disorders and using this knowledge to implement appropriate parenting and educational strategies. It is important, not only to understand the wide spectrum of visual disability but also to identify the child's skills that can be employed for habilitation.[2] The aim of this paper is to describe the setting up of a CVI clinic in a tertiary care hospital in South India and to describe the spectrum of cases seen.

Materials and Methods

This clinic has been set up in a multispecialty tertiary care hospital in South India that already has a specialized unit for Developmental Pediatrics, Pediatric Neurology, and Pediatric Psychiatry. The author underwent specialized CVI vision assessment training abroad. The training involved vision assessment and rehabilitation of children with CVI in hospital and community setup. Our CVI clinic was set up in the eye department of the parent institution in India in February 2011. Assessment to identify and characterize CVI requires dedicated space, time, and an interdisciplinary team approach.[18] Our existing examination rooms have been modified to reduce surrounding noise and to make it more appealing to these children. The clinic waiting area has walls painted with cartoon characters, space to run around, play with toys, and drawing materials to keep the child entertained. The examination rooms have been modified to be more child-friendly and are clutter and noise free. The capital and human resources needed for such modification are presented in Table 1.

Selection and description of the participants

All children presenting to our hospital, <18 years of age, with CP, learning disability, autism, neurodegenerative diseases, and brain trauma are referred to the clinic. Each child is referred to the CVI clinic by his/her pediatrician, neurologist, psychiatrist, occupational therapist, or ophthalmologist. The child's referring clinician provides the necessary information regarding the neurological and developmental/cognitive assessment and radiological input. The pediatric ophthalmologist gathers this information from the hospital's electronic medical records (EMR).

Running of the clinic

In the CVI clinic, the trained pediatric ophthalmologist performs the neuro-ophthalmological assessment. This begins when the child comes into the waiting room of the clinic accompanied by the parents/caregivers and includes observing the children's activities in the waiting area. Spontaneous visual behaviors are carefully observed. Throughout the CVI examination and assessment, the child is observed for what he/she is looking at, and the size and nature of the object that gains and maintains the child's attention, as well as the distance the child can appreciate the object of regard. Initial identification of visual-perceptual disorders is carried out, based initially on in-depth history taking, first eliciting spontaneously volunteered information about visual behaviors, then using a Structured Clinical Question Inventory as a guide to ensure that key issues are not omitted.[2] Visual acuity is evaluated using the best-corrected dioptric correction by the pediatric orthoptist. Binocular functional vision is usually assessed as uniocular assessment is difficult in many children. Depending on the age and severity of physical disability, different tests are used. For children with severe CP, in whom visual acuity cannot be quantified, we look for responses to light and high contrast pictures/toys, such as movement of limbs or heading toward the visual target, postural reactions, avoiding reactions, and changes in facial expression. Eliciting optokinetic nystagmus also provides information on the visual potential in the child. Children with mild-to-moderate CP are assessed with age and ability appropriate visual acuity tests. These include Lea grating paddles, Cardiff visual acuity cards, Kay pictures, and Snellen visual acuity chart. Oculomotor functions including fixation, saccadic eye movements, accommodation, visual axis alignment, and stereopsis are also assessed. The visual field in children is assessed based on the behavioral responses, reflecting the child's ability to locate targets presented in different areas of the visual field. Neuropsychological tests were not employed in our subset of patients due to their multiple neuropsychological disabilities, rendering their administration impossible.

Results

From the inception of the clinic in February 2011 to September 2015, 1478 patients have been referred. Most of these patients (85%) come from different parts of India. The international patients have come from Bangladesh, Nepal, Sri Lanka, UAE, and Kuwait. Of the patients referred into the clinic, 61% had CP, 28% had seizure disorders, autism was seen in 9.5%, and learning disabilities, neurodegenerative conditions, and brain injury together constituted 1.5%. Of the 902 children with CP, 39% had severe CP (GMFCS level 5, MACS level 5, and CFCS level 5), 45% had moderate CP, and 16% had mild CP. The most common perinatal cause of CVI in our clinic was birth asphyxia (40%), but there were about 20% of cases where a cause of the CVI could not be established. Visual acuity assessment is shown in Fig. 1. There was a mixed picture of

Table 1: Resources required for setting up cerebral visual impairment clinic

Infrastructure required	Equipment required	Personnel
Spacious enough for child to move and play freely	Toys, coloring books, low set tables, and chairs	Interdisciplinary team approach - pediatricians both at community and hospital level, pediatric neurologist, developmental pediatricians, child psychiatrist, occupational therapist
Clutter-free environment, with minimal decoration	Vision assessment tools such as OKN strips, Cardiff visual acuity cards, Lea grating paddle, Kay pictures, Snellen's visual acuity charts	Ophthalmologist and pediatric orthoptist trained in CVI assessment
Limited background auditory noise	Visual field assessment kits	

OKN: Optokinetic nystagmus, CVI: Cerebral visual impairment

higher visual dysfunction similar to that previously reported in the literature.[1]

Of the 1478 patients seen in the clinic, 295 (20%) of the patients have returned for review. Of these, 70% (206 patients) were utilizing strategies recommended in the CVI clinic, and all reported significant progress in their child's visual abilities. This was corroborated on clinical examination where the children were found to have gained improved eye contact, greater interest in toys and puppets, and enhanced interest in their surroundings. Of the remainder, 5% had carried out the visual strategies suggested for a few months but had discontinued the training as they did not find any improvement in their child's condition. The rest was more concerned about motor improvement than visual habilitation. A description of one of the cases presented to our clinic is described in Box 1.

Liaison between team members was conducted through telephonic conversations and hospital EMR. Interdisciplinary clinics were precluded owing to the constraints of opportunity and time. As the clinical service has expanded, another pediatric ophthalmologist and pediatric orthoptist have gained in-house training in CVI assessment.

Discussion

The visual brain reconstructs a dual internal emulation of the visual scene, with the ventral stream: temporal lobes creating the imagery and the dorsal stream; posterior parietal lobes mapping it in space and time. Disorders of this process are unique to each child and involve incomplete mapping of the scene, which necessitates a dedicated quiet environment for optimum assessment. Vision has an input and processing system.[19] Vision fundamentally facilitates learning throughout a child's development, resulting in knowledge, skills, and traits that will shape the child's personality and cognition. CVI is the preferred term as it is the result of damage to both gray and white matter of the brain.[2] Despite its absence from ICD 10, CVI is one of the leading causes for visual impairment in children both in the developed and developing world.[20] Visual dysfunctions are one of the main features of CP and are on a par with the motor disorders.[2] Up to 60%–70% of children with CP may have CVI.[21]

In many Western countries, such as the United Kingdom, Italy, the Netherlands, Germany, and Sweden, this specialized service is routinely available for affected children. There is also involvement of governmental and nongovernmental organizations in their rehabilitation and follow-up. Such services can also help maintain a database of visual impairment in CP children for the nation. In India, such specialized services or databases have yet to be developed. According to the published literature from India, the estimated incidence of CP is 3 per 1000 live births[22] and CVI in CP is reported at 28%,[23] this is just the tip of the iceberg. It is likely to be higher if visual perceptual dysfunction is included in the study. Training in CVI is not yet part of postgraduate (PG) ophthalmology training program in India, neither is there any fellowship offering this facility. Development of this specialized care will improve PG training and care for these neglected children.

Of late, there has been a growing awareness of CVI among pediatric ophthalmologists in the country. In spite of the availability of all the personnel in our hospital, getting them all together remains a challenge. The combined travel and discussion time currently renders setting up a clinic where all the multidisciplinary team (MDT) specialists are physically present is a challenge. Currently, the pediatric ophthalmologist assessing CVI has access through hospital's EMR to the child's systemic condition, intelligent quotient scoring, medication, physiotherapy and occupational therapy, and also neuroimaging. The primary clinician and the therapist also have access to the CVI assessment reports for further management of the child. We are developing a regular fortnightly meeting of the MDT to discuss and manage difficult cases.

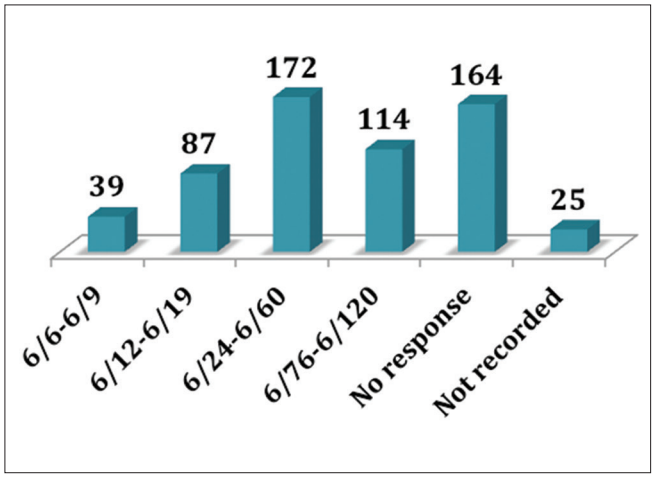

Figure 1: Visual acuity assessment (number of patients)

Box 1: Clinical presentation of a case seen in cerebral visual impairment clinic

An 11-year-old, Mast X (Grade 1 student) was brought to this multispecialty hospital by his father with concerns of learning difficulties. He is first born to nonconsanguineous parents, born at 36 weeks of gestation by cesarean section with a birth weight of 2.25 kg. There is a history suggestive of birth asphyxia necessitating specialized neonatal care for 10 days. There was a delay in attaining his developmental sequences. At 7 years of age, he developed right-sided tonic-clonic seizures of about 2 mins duration every 3-4 days, partially controlled with oral medication. He was largely independent in self-care skills, and his parents did not have any concern about vision. His developmental pediatrician's assessment scores were as follows: On SBIT his mental age was 4 years and 7.5 months with an IQ score of 44. On VSMS, he had a social age of 9 years and 8 months. On the GLADS, he could read and write at Grade 1 level in his native language but had poor numerical and mathematical skills for this grade. The electroencephalography and VEP reports were normal. The MRI of the brain showed bilateral symmetrical parieto-occipital gliosis with gyral thinning and white matter hyperintensities [Fig. 2]. Ophthalmological examination showed that he had a uniocular BCVA with uncrowded Kay pictures of 6/9, N6, but 6/24 with crowded Kay pictures. His oculomotor examination was normal and did not show any refractive error. Using the SCQI and neuro-ophthalmological examination, findings of CVI are summarized in the form of a report [Table 2] with strategies suggested for the parents/caregivers and clinicians

SBIT: Stanford-Binet intelligence test, IQ: Intelligent quotient, VSMS: Vineland Social maturity Scale-Indian adaptation, GLADS: Grade Level Assessment Device for children with learning problems in school, VEP: Visual evoked potential, MRI: Magnetic resonance imaging, BCVA: Best-corrected visual acuity, SCQI: Structured Clinical Question Inventory, CVI: Cerebral visual impairment

Examination of a child with CVI is time consuming, and for any clinical examination to be fruitful, it is important that the child is comfortable throughout. Children with CVI are at ease in a quiet environment with fewer distractions, which is not possible in a regular busy pediatric ophthalmology clinic. The examination rooms are accordingly designed to be spacious, uncluttered, and child-friendly with minimum background auditory noise. Making the required modifications to our examination rooms was a challenge as the existing concrete structure needed to be refurbished with limited funds. We therefore involved volunteers.

It is important to remember that the needs and well-being of children with CVI and their families are central to establishing such a service. For appropriate service delivery to provide accurate and useful vision assessment of the child, it is important to have both professional and parental input. CVI assessment necessitates giving an assessment report to the referring clinician and therapists/teachers [Table 2] that includes the information needed by teachers to ensure that their teaching is accessible to the child. Advice regarding salient habilitative strategies is also suggested in the report. Key links to the CVI clinic include community health-care workers, special educators, voluntary organizations, and parents/caregivers support groups. Our limitation is a poor rate of return to the clinic, principally owing to low socioeconomic background and distant locations from where these children come, rendering regular review impracticable, and necessitating the development of alternative approaches to follow-up. Assessment in CVI clinics is fruitless if recommended strategies are not understood nor implemented at home. The support groups have an important role to address the following issues:

- Spread education and awareness about CVI
- It is a daunting task for parents not only to accept the physical condition of their wards but also to carry out

Figure 2: T2 axial and T1 sagittal images showing gliosis in bilateral parietal lobe, thinning of the gray matter and volume loss in the white matter (arrowhead), marked thinning involving the body (posterior portion), splenium of the corpus callosum (long arrow)

Table 2: Report of cerebral visual impairment clinic assessments and strategies for habilitation for Mast X

Based on SCQI	Based on examination	Probable reason	Strategies suggested
Dorsal stream dysfunction			
Difficulty in copying from blackboard		Impairment in visual search of source and target documents, and remembering the information	Reduce distraction. To use a friend's notes to copy at home and use upper limbs to mask the surrounding text
Dislikes crowded places such as family functions, fairs	Unsettled in the waiting area with so many children and noise	Crowded places and noise causes confusion	To go these places at a time when crowd is less
Difficulty in finding objects against a patterned background	Difficulty in locating the pen kept on patterned tablecloth	Pattern causes distraction and confusion	To use plain tabletops and tablecloth
	Placement of objects inaccurately on the edge of the table and wide-open mid-flight fanning of the fingers while gathering the pen	Damage to the posterior parietal cortices leads to optic ataxia	Learn to extend the little finger or part of the body as a tactile guide to the location of the target surface to avoid misplacement of the object
Goes close to the television to watch it and watches only traditional music		Visual search impairment limits how much can be seen	Can go close to TV
	Getting irritated when he was doing a task and simultaneously instructions for doing the task were being given	Difficulty dividing attention between doing a task and listening	To instruct before the task is being asked to carry out
Needed help to navigate steps and stairs and to wear foot wear	Lower field defect on visual field assessment	Cannot see the feet well due to lower field defect	To use umbrellas/walking sticks to walk. While wearing footwear, bring foot to the eye level by placing the foot on a reasonably high chair/stool
Ventral stream dysfunction	Difficulty in recognizing objects	Prosopagnosia resulting due to damage to the temporal lobe	The child could feel the objects (tactile recognition)

SCQI: Structured Clinical Question Inventory

the strategies suggested. They need constant motivation, with regular encouragement and support. Engaging with support groups and voluntary organizations proves beneficial
- There is a constant need for economically viable tools and equipment to be developed because affected families are commonly under financial strain
- An important role of these organizations is to give feedback on services provided by the CVI clinic to assist in service improvements.

Conclusions

CVI is an important cause of vision impairment in children with special needs. There is a need to set up specialized clinics in all parts of the country for patient care and training. For a holistic and successful management of CVI, a multidisciplinary approach is ideal.

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Conflicts of interest

There are no conflicts of interest.

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