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Exploring Cortical Visual Impairment: Disease Classification, Parental Concerns, and Functional Vision Assessment

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Abstract

Background

Cortical visual impairment (CVI) is an increasingly recognized cause of childhood visual impairment resulting from damage to the retrochiasmal visual pathways. Children with CVI experience difficulties in visual processing that can impact perception, mobility, learning, and interaction with their surroundings. Early diagnosis is often difficult because the clinical presentation is variable and conventional visual acuity assessments may not adequately reflect functional visual limitations. Functional assessment tools such as the CVI Range, together with caregiver-reported observations, provide important insights into everyday visual performance. However, data exploring the relationship between parental concerns and functional vision in children with CVI remain limited in low-resource settings such as Nepal.

Methods

This prospective cross-sectional study included children aged 7 months to 18 years diagnosed with cortical visual impairment (CVI) at a tertiary pediatric ophthalmology clinic. Caregiver-reported visual concerns were recorded, and functional vision was assessed using the CVI Range to classify children into three functional phases. Visual acuity and contrast sensitivity were measured using age-appropriate methods, and their associations with CVI phase and caregiver-reported concerns were analyzed.

Results

A total of 70 children with CVI were included, with a mean age of 4.66 ± 4.65 years; 67.1% were male. Nearly half (48.6%) were classified as Phase 1, 18.6% as Phase 2, and 32.9% as Phase 3, with mean age increasing across phases. The most common parental concern overall was difficulty maintaining eye contact (37.1%), followed by academic concerns (17.1%) and failure to look directly at objects (17.1%). Parental concerns varied by CVI phase, shifting from deficits in visual attention in Phase 1 to academic, mobility, and distance vision difficulties in later phases. Visual assessment methods also varied by phase, with fixation and following predominating in Phase 1 (79.4%), grating acuity in Phase 2 (76.9%), and Snellen visual acuity in Phase 3 (60.9%). Mean grating acuity improved across phases, while contrast sensitivity was poorer in Phase 1 compared with later phases. However, differences in grating acuity, contrast sensitivity, and LogMAR visual acuity across phases were not statistically significant ($p > 0.05$).

Conclusion

Parental concerns in children with CVI evolve with age and functional vision status, reflecting the changing nature of visual difficulties. Caregiver-reported concerns, combined with functional vision

assessment using the CVI Range, may improve early identification of CVI and support individualized rehabilitation planning.

Background

Cortical visual impairment (CVI) is a neurovisual disorder caused by dysfunction of the retrochiasmal visual pathways, resulting in impaired visual processing despite structurally normal eyes [1]. It has become one of the most common causes of childhood visual impairment worldwide, particularly in developing countries, where improved neonatal survival has increased the number of children at risk of neurological injury. [2–3] [3]Early identification of CVI remains challenging because its manifestations are often subtle, heterogeneous, and may overlap with developmental or behavioral disorders. [4]. In addition, conventional visual acuity testing alone may not fully capture the functional visual difficulties experienced by these children, as many may demonstrate relatively preserved acuity despite significant challenges in perception, mobility, learning, and environmental interaction. This reliance on conventional testing may result in missed or delayed diagnosis of cortical visual impairment (CVI), emphasizing the need for functional vision assessments to identify the broader impact of CVI on daily life.

To address this, functional vision assessment tools such as the CVI Range have been developed.[5] The CVI Range is a structured and validated tool designed to evaluate the unique visual and behavioral characteristics associated with CVI and to classify children according to the severity of functional visual impairment. [6]. Unlike standard visual acuity tests, it provides insight into how vision is used in everyday activities, helping guide rehabilitation planning and monitor changes over time.

Caregiver observations have become increasingly recognized as an important component in the assessment of children with CVI, particularly in those who are too young or unable to participate in formal vision testing.[4]. Parents are often the first to notice abnormal visual behaviors such as poor eye contact, difficulty recognizing familiar faces, or reduced visual attention. Recent studies have highlighted the value of these concerns in identifying CVI and understanding functional vision deficits. A study from India by Sumalini et al. demonstrated that parent-reported concerns varied according to CVI severity, with children in earlier phases showing deficits in visual attention and those in later phases exhibiting more complex visual-perceptual difficulties.[7]

Despite increasing global recognition of CVI, there remains limited evidence from low- and middle-income countries, including Nepal, regarding the relationship between parental concerns and functional vision assessment. Understanding these patterns may improve early diagnosis, support family-centered care, and guide targeted rehabilitation strategies. Therefore, this study aimed to explore parental concerns in children with CVI and evaluate their relationship with functional vision assessment findings at a tertiary eye care center in Nepal.

Methods

Study design and participants

This prospective cross-sectional study was conducted in a pediatric ophthalmology clinic at a tertiary eye care center to classify children with cortical visual impairment (CVI) into three functional phases and identify the most common caregiver-reported visual concerns.

The study aimed to characterize children with CVI by examining the relationship between CVI phase, caregiver-reported visual concerns, age, visual acuity, and contrast sensitivity. Secondary objectives included evaluating the association between parent-reported concerns and functional vision assessment using the CVI Range, as well as exploring the relationship between functional vision scores and measured visual acuity.

Children aged 7 months to 18 years diagnosed with CVI based on magnetic resonance imaging findings, medical history, and clinical observation of visual behavior were included. Caregivers were asked an open-ended question regarding their child's vision-related difficulties: *"What vision-related problems have you noticed in your child?"* No prompts were provided, and all reported concerns were documented.

Functional vision assessment

Functional vision was assessed using the CVI Range, a validated tool designed to evaluate the unique visual and behavioral characteristics of children with CVI. The assessment included ten characteristics, each graded individually through examiner observation, caregiver interview, and direct assessment. Total scores ranged from 0 to 10, with higher scores indicating better functional vision. Based on these scores, children were classified into three phases: Phase 1 (0–3), Phase 2 (3–7), and Phase 3 (7–10). Only the within-the-characteristics scale was used in this study.

Visual acuity and contrast assessment

Binocular grating acuity was assessed using Lea Grating Paddles, presenting spatial frequencies of 1, 2, 4, and 8 cycles/cm. The child's visual response was determined by observing eye movements toward the grating stimulus compared to a gray control stimulus. Testing progressed from coarse to finer gratings, and the highest spatial frequency with two positive responses was recorded as the threshold. Grating acuity was adjusted for testing distance and recorded in cycles per degree.

Binocular contrast sensitivity was measured using the Hiding Heidi Low Contrast Face Test. Cards with progressively lower contrast levels (100%, 25%, 10%, 5%, 2.5%, and 1.25%) were presented sequentially. Infants were assessed through behavioral responses, while older children identified the face verbally or by pointing. The lowest contrast level at which the child responded consistently was recorded as the contrast threshold.

Statistical analysis:

Data were entered into Microsoft Excel and cleaned prior to analysis. The cleaned dataset was exported to IBM SPSS Statistics version 20 for statistical analysis. Categorical variables were summarized using frequencies and percentages. Fisher's exact test was used to compare categorical variables when the

expected cell count was less than five. For continuous variables, an independent-samples *t*-test was used to compare two groups, while one-way analysis of variance (ANOVA) was used to compare more than two groups. All statistical tests were two-tailed, and a *p*-value of less than 0.05 was considered statistically significant.

Results

A total of 70 children with cortical visual impairment (CVI) were included in the study. The mean age of the participants was 4.66 ± 4.65 years. The majority of children were aged 1–5 years (51.4%), followed by 6–10 years (21.4%), < 1 year (14.3%), and 11–19 years (12.9%). There was a male predominance, with 47 males (67.1%) and 23 females (32.9%). Of the 70 children enrolled, 48.6% (n = 34) were classified as Phase 1, 18.6% (n = 13) as Phase 2, and 32.9% (n = 23) as Phase 3. The distribution of participants across the three CVI phases was statistically significant (p = 0.009). Mean age increased progressively across CVI phases, from 2.0 ± 2.65 years in Phase 1 to 5.94 ± 4.56 years in Phase 2 and 7.85 ± 4.82 years in Phase 3.(Table 1)

Table 1
Frequency distribution and mean age of children across CVI phases

CVI Phase	Frequency (n)	Mean Age in years	Standar Deviation (SD)
1	34	2.00	2.65
2	13	5.94	4.55
3	23	7.85	4.82
Total	70	4.65	4.64

Parental Concerns

Difficulty maintaining eye contact was the most frequently reported parental concern (37.1%), followed by academic concerns and failure to look directly at objects (17.1% each), difficulty with distance viewing (15.7%), eye deviation (11.4%), and bumping into objects (10.0%). Other concerns, including difficulty walking, fluctuating vision, and frequent falls, were reported less commonly.(Table 2)A total of 70 caregivers reported visual concerns regarding their children with CVI. Most caregivers (55.7%, n = 39) reported a single concern, while 28.6% (n = 20) reported two concerns, 14.3% (n = 10) reported three concerns, and 1.4% (n = 1) reported four concerns. Multiple concerns were therefore reported by 31 caregivers (44.3%), indicating that a substantial proportion of children presented with more than one functional visual difficulty.

Table 2
Frequency and distribution of parental concerns according to CVI phase

Findings	CVI Phase							
	1		2		3		Total	
	Count	%	Count	%	Count	%	Count	%
difficulty in recognising faces	6	17.6%	1	7.7%	0	0.0%	7	10.0%
unable to look at light	3	8.8%	0	0.0%	1	4.3%	4	5.7%
difficulty in distance viewing	4	11.8%	1	7.7%	6	26.1%	11	15.7%
missing object on the lower / side field	2	5.9%	0	0.0%	1	4.3%	3	4.3%
eye deviation	2	5.9%	1	7.7%	5	21.7%	8	11.4%
academic concerns	1	2.9%	6	46.2%	5	21.7%	12	17.1%
bumping into objects	1	2.9%	4	30.8%	2	8.7%	7	10.0%
no visual concerns	1	2.9%	0	0.0%	3	13.0%	4	5.7%
stares at light	1	2.9%	0	0.0%	0	0.0%	1	1.4%
difficulty in walking in stairs	0	0.0%	0	0.0%	2	8.7%	2	2.9%
difficulty in walking in narrow roads	0	0.0%	0	0.0%	1	4.3%	1	1.4%
does not look at objects directly	11	32.4%	1	7.7%	0	0.0%	12	17.1%
cannot walk confidently	1	2.9%	3	23.1%	3	13.0%	7	10.0%
difficulty in walking in uphill and downhill	1	2.9%	0	0.0%	2	8.7%	3	4.3%
sees very up close	0	0.0%	1	7.7%	1	4.3%	2	2.9%
frequent falling	0	0.0%	1	7.7%	0	0.0%	1	1.4%
difficulty in maintaining eye contact	19	55.9%	4	30.8%	3	13.0%	26	37.1%
fluctuating vision	1	2.9%	0	0.0%	1	4.3%	2	2.9%

Findings	CVI Phase							
	1		2		3		Total	
	Count	%	Count	%	Count	%	Count	%
Total Person	34	100.0%	13	100.0%	23	100.0%	70	100.0%

Parental concerns varied across CVI phases. In Phase 1, difficulty maintaining eye contact (55.9%) and failure to look directly at objects (32.4%) were the most common concerns. In Phase 2, academic concerns (46.2%) and bumping into objects (30.8%) predominated, while in Phase 3, difficulty with distance viewing (26.1%), academic concerns (21.7%), and eye deviation (21.7%) were most frequently reported. Overall, concerns shifted from visual attention deficits in the early phases to functional and spatial vision difficulties in the later phases.(Table 2)

Parental concerns also varied across age groups. In infants (< 1 year), difficulty maintaining eye contact (60%) and failure to look directly at objects (40%) were the most common concerns. Among children aged 1–5 years, difficulty maintaining eye contact remained predominant (47.2%), with increasing reports of distance viewing difficulties. In children aged 6–10 years, academic concerns (40%) and difficulty walking confidently (26.7%) were most frequently reported. Among adolescents aged 11–19 years, distance viewing difficulties and academic concerns (33.3% each) were the leading concerns. Overall, parental concerns shifted from visual attention deficits in younger children to functional, academic, and distance vision challenges in older age groups.

Visual Assessment Methods

Visual assessment methods varied across CVI phases. Overall, Fixation and Following (FNF) was the most commonly used assessment method (40.0%, n = 28), followed by grating acuity (37.1%, n = 26) and Snellen visual acuity testing (22.9%, n = 16). FNF was predominantly used in Phase 1 (79.4%), reflecting limited cooperation and visual responsiveness in younger children. In Phase 2, grating acuity was the most frequently employed method (76.9%), whereas Snellen visual acuity testing predominated in Phase 3 (60.9%).

Among the 28 children assessed using FNF, more than half of those in Phase 1 (53.6%, n = 15) did not demonstrate fixation and following, while 46.4% (n = 13) showed a positive response.(Table 3) All children assessed in Phase 2 demonstrated fixation and following, and no children in Phase 3 required FNF assessment.

Grating acuity was assessed in 26 children, with a mean value of 2.94 cycles and a median of 2 cycles. Lower grating acuity values were more common in Phase 1, whereas higher values were observed in later phases. In Phase 2, 4 cycles was the most common value (30%), while in Phase 3, 4 cycles (33.3%) and 8 cycles (22.2%) were most frequently recorded.(Table 4)

Snellen visual acuity was assessed in 16 children, none of whom were in Phase 1. Among children in Phase 3, visual acuity ranged from 2/60 to 6/6, with 6/12 being the most common value (21.4%, n = 3), indicating considerable variability in visual function even among children with higher CVI phases. (Table 4)

Table 3
Association between fixation and following (FNF) responses and CVI phase

FNF Values	CVI Phase				p value
	1		2		
	Count	%	Count	%	
Fix and follow light	12	44.4%	1	100.0%	0.464
Not fixing and following	15	55.6%	0	0.0%	
Total	27	100.0%	1	100.0%	

Table 4
Comparison of grating acuity, contrast sensitivity, and LogMAR visual acuity across CVI phases

Variables	**Phase1**		**Phase 2**		**Phase3**		**p value**
	n	**mean ± SD**	**n**	**mean ± SD**	**n**	**mean ± SD**	
Grating Values	7	2 ± 2.69	10	3.33 ± 2.87	9	3.25 ± 3.19	0.617
Contrast acuity	16	69.53 ± 41.03	12	37.08 ± 46.94	23	51.63 ± 47.75	0.178
Logmar Vision	-	-	2	0.48 ± 0.42	14	0.61 ± 0.47	0.723

Of the 28 children assessed with the Fixation and Following (FNF) test, 55.6% of Phase 1 children did not demonstrate fixation and following, whereas all children assessed in Phase 2 showed a positive response. The association between FNF response and CVI phase was not statistically significant (p = 0.464).(Table 3)

Mean grating acuity increased from 2.00 ± 2.69 cycles in Phase 1 to 3.33 ± 2.87 cycles in Phase 2 and 3.25 ± 3.19 cycles in Phase 3. Although children in the higher CVI phases demonstrated better grating acuity, the difference was not statistically significant (p = 0.617).

Contrast acuity values were highest in Phase 1 (69.53 ± 41.03), followed by Phase 3 (51.63 ± 47.75), and lowest in Phase 2 (37.08 ± 46.94). As higher values indicate poorer contrast sensitivity, children in Phase 1 demonstrated reduced contrast sensitivity compared with later phases, although this difference was not statistically significant (p = 0.178).

LogMAR visual acuity could only be assessed in children from Phases 2 and 3. Mean LogMAR visual acuity was 0.48 ± 0.42 in Phase 2 and 0.61 ± 0.47 in Phase 3, with no significant difference between the

groups (p = 0.723).

Overall, while children in higher CVI phases tended to demonstrate better grating acuity and were more likely to complete formal visual acuity testing, no statistically significant differences were observed in grating acuity, contrast sensitivity, or LogMAR visual acuity across CVI phases.

Discussion

In this study, parental concerns regarding children with cortical visual impairment (CVI) were assessed alongside functional vision evaluation using the CVI Range. The mean age of the participants was 4.66 years, which is similar to that reported in previous studies, including the work of Rice et al. [8]The study cohort demonstrated a predominance of male participants, a trend that has also been reported in the existing literature on CVI.

A total of 113 concerns were identified from 70 parents, with 31 parents (44.3%)reporting multiple concerns .To facilitate analysis, the reported concerns were consolidated into 19 distinct categories of visual concerns.

Functional vision was evaluated using the CVI Range, a standardized assessment tool designed for children with cortical visual impairment (CVI). While conventional visual assessments typically measure visual acuity, visual fields, and visual awareness, the CVI Range focuses on functional vision—that is, how children use their vision to interact with their environment and perform daily activities.[6]

The pattern of parental concerns observed across CVI phases is consistent with the developmental trajectory described by Sumalini et al. [7]While early-phase children primarily demonstrated difficulties in visual attention and eye contact, later phases were associated with concerns related to academic performance, mobility, and distance viewing. These findings suggest that as functional vision improves, caregivers become less focused on fundamental visual behaviors and more aware of higher-order visual and functional challenges. This aligns with the observations of Sumalini et al., who reported that functional vision outcomes varied according to CVI severity and were influenced by factors such as visual acuity, contrast sensitivity, and age. Together, these findings highlight the importance of assessing functional vision alongside traditional measures of visual function in children with CVI.

Parental concerns varied across age groups, with younger children predominantly presenting with difficulties in visual attention and fixation, while older children demonstrated concerns related to learning, mobility, and distance vision. Infants and preschool-aged children were most frequently reported to have difficulties maintaining eye contact and looking directly at objects, reflecting the early visual attention deficits that are characteristic of CVI. Similar findings have been reported by Roman-Lantzy and other investigators, who described impaired visual attention, fixation, and social visual engagement as common early manifestations of CVI.

In contrast, school-aged children in the present study were more likely to present with academic difficulties, mobility challenges, and distance viewing problems. This finding is consistent with previous studies reporting that higher-order visual processing deficits in CVI often become more apparent as children enter formal education and are required to navigate increasingly complex visual environments. Difficulties with reading, recognizing faces, spatial orientation, and environmental navigation have been widely documented in older children with CVI. The increasing prevalence of academic concerns observed in this study is also supported by the work of Dutton and colleagues, who emphasized the significant impact of CVI on educational performance and everyday functioning.[9]

Overall, the findings suggest that the clinical presentation of CVI evolves with age, progressing from deficits in basic visual attention and fixation during early childhood to more complex functional and perceptual visual difficulties in later childhood and adolescence. This highlights the importance of age-specific assessment strategies and functional vision evaluations when managing children with CVI.

The pattern of visual acuity assessment in the present study was similar to that reported in previous CVI literature. Children in Phase 1 were primarily assessed using fixation and following (FNF), while grating acuity was most commonly used in Phase 2 and Snellen visual acuity testing predominated in Phase 3. Similarly, the study of 194 children with CVI found that observational assessments were more commonly used in children with severe impairment, whereas Teller Acuity Cards and optotype-based tests were increasingly feasible in children with less severe CVI. These findings suggest that the choice of visual acuity assessment is closely related to the child's functional vision level and CVI severity.

Contrast sensitivity demonstrated a trend toward improvement across advancing CVI phases. Although the difference was not statistically significant, this finding may reflect better functional visual performance in children classified in the higher CVI phases.

This study has several limitations. First, the distribution of participants across the three CVI phases was unequal, with a larger proportion of children in Phase 1, which may have influenced intergroup comparisons. Second, visual assessment tools were not uniformly applied across all phases, as fixation and following was primarily used in Phase 1, grating acuity in Phase 2, and recognition acuity in Phase 3. This limited direct comparison of visual function across phases. Additionally, the relatively small sample size in some subgroups, particularly for LogMAR visual acuity assessment, may have reduced the statistical power to detect significant differences.

Conclusions

Children with cortical visual impairment present with distinct parental concerns that vary according to age and CVI phase. Early phases are characterized by deficits in visual attention and fixation, while later phases show increasing concerns related to academic performance, mobility, and distance vision. Functional vision assessment using the CVI Range provides valuable insights beyond conventional visual acuity testing and may support earlier identification and targeted rehabilitation strategies. Understanding

caregiver-reported concerns alongside structured functional vision assessment may improve clinical evaluation and management of children with CVI.

Declarations

Ethics Approval and Consent to Participate:

Ethical approval for this study was obtained from the Institutional Review Committee (IRC) of the Tilganga Institute of Ophthalmology, Kathmandu, Nepal (Approval No:19/2024). The study was conducted in accordance with the ethical principles of the Declaration of Helsinki and its subsequent amendments. Written informed consent was obtained from the parents or legal guardians of all participating children prior to enrollment.

Funding Declaration:

This study did not receive any external funding. The research was conducted using the institutional resources and facilities of Tilganga Institute of Ophthalmology, Kathmandu, Nepal.

Consent for publication:

Not applicable

Availability of data and materials:

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Competing interest :

The authors declare that they have no competing interests

Authors' contributions:

AJL conceived and designed the study, developed the research methodology, coordinated data collection, performed data analysis and interpretation, and drafted the manuscript. AJL served as the Principal Investigator.

PT contributed to data collection and assisted with study implementation as Co-Principal Investigator.

SA contributed to the study concept and design, provided scientific mentorship and supervision throughout the study, and critically reviewed the manuscript as Co-Principal Investigator.

MP performed the statistical analysis, contributed to data interpretation, and critically reviewed the manuscript as Co-Principal Investigator.

All authors read and approved the final manuscript.

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Not applicable

Data Availability

Data Availability Statement The datasets generated and analyzed during the current study are not publicly available because they contain potentially identifiable participant information. De-identified data are available from the corresponding author on reasonable request and subject to approval by the Institutional Review Committee of Tilganga Institute of Ophthalmology.

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