

Research Article

# Profiling children with cerebral visual impairment in a tertiary eye care center

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## Abstract

**Background:** Cerebral visual impairment (CVI) is a bilateral visual impairment that affects children in all industrialized countries. It has become more common in low-income countries as a result of the increased survival rates of children who suffer from severe neurological conditions during the perinatal period. The purpose of this study was to determine the characteristics of children with CVI in a tertiary children's eye care center population.

**Methods:** From October 2020 to September 2021, a cross-sectional study was conducted to select all consecutive patients with a diagnosis of cerebral visual impairment aged 6 months to 16 years. On the neurological deficit, information was gathered from the patient's referral: parental interviews, observations, and direct assessment were used for functional vision characteristics, and an ophthalmic examination was performed for eye findings. The interviewees' responses were matched to the ten specific behavioral characteristics shared by children with CVI. Cortical visual impairment was diagnosed using three criteria: [the vision loss is not explained by abnormalities found on the eye examination, a neurological medical diagnosis, and the child exhibits one of the unique visual and behavioral characteristics described by Roman Lantz]. A descriptive statistical analysis (frequency, mean, and range) was calculated.

**Results:** Forty children with CVI (1.96% of total children) were seen. The mean age was 2.56 ( $\pm$  1.98) years. There were 24 (60%) males. On a referral paper of 28, hypoxic-ischemic encephalopathy was the commonest cause mentioned (70.0%). Seizures were the most frequent neurological deficit at presentation. Ophthalmic and neurologic impairments were found in 42.5% of children with CVI. Based on Roman-Lantz's three phases of the CVI Range, 90% of children with CVI at the test time had Phase I or Phase II vision.

**Conclusion:** According to the findings of this study, visual impairment is critical in the diagnosis of CVI. The prevalence of CVI as a cause of childhood vision impairment is significant. Hypoxic-ischemic encephalopathy is the most common cause of CVI. All children with CVI have serious neurological issues, and the majority have associated ophthalmic abnormalities.

## Introduction

Cerebral visual impairment (CVI) refers to a broad range of bilateral visual and perceptual impairments caused by dysfunction, anomaly, or injury to the brain's retinogeniculate visual pathways and centers, specifically the optic radiations, occipital cortices, and visual associative areas, in any combination or degree, and oculomotor control, which is more profound if the thalamus is affected [1,2]. Cerebral visual impairment, which is already the leading cause of low vision in children in developed countries, is becoming more common in low-income countries as medical technology improves the survival of children who suffer from severe neurological conditions during the perinatal period [3,4]. CVI has the potential to affect at least 2.4% of children [5]. Because

it is not a consciously symptomatic condition, many affected children go unidentified [5]. Indeed, the most common causes and associations with CVI are cerebral anoxia (or hypoxia) and periventricular leukomalacia [6,7]. It can also occur as a result of meningitis [8], encephalitis [9], traumatic brain injury [10,11], hydrocephalus [12], or metabolic abnormalities [13].

A child with CVI may have a normal to near-normal ability to detect details in what he or she sees, but the child may be completely unaware of what he or she is seeing [14]. Early brain damage is frequently diffuse and affects multiple brain functions, resulting in concomitant neurological disorders such as seizures, intellectual disability, and cerebral palsy, which may exacerbate the detrimental effects of CVI on cognitive, motor, and social development [9,15].

### More Information

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**Submitted:** February 20, 2022

**Approved:** April 06, 2022

**Published:** April 07, 2022

**How to cite this article:** Teshome AW, Worku MD. Profiling children with cerebral visual impairment in a tertiary eye care center. Int J Clin Exp Ophthalmol. 2022; 6: 007-012.

**DOI:** 10.29328/journal.ijceo.1001043

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**Keywords:** Profile; Children; Cerebral visual impairment





Cerebral visual impairment is a heterogeneous group of disorders with distinct neuro-visual pathologies that necessitate a distinct adaptation to the underlying neurological deficits. The assessment of visual function in these children must go beyond the standard methods of visual acuity and visual field testing used in routines.

Although studies have been conducted in various parts of the world, there is a scarcity of data on the epidemiology of the “new” visual disability CVI in Ethiopia. The purpose of this study was to ascertain the characteristics of children with CVI in the population of a tertiary children’s eye care center, Menelik II hospital. The knowledge gained from this study will aid in effective planning to deal with childhood blindness in the country and to improve patient care with CVI by implementing a screening procedure to detect children who demonstrate the behaviors associated with CVI.

## Materials and methods

A cross-sectional study included all consecutive patients diagnosed with cerebral visual impairment for the first time, as well as those children with settled diagnoses returning for follow-up visits. From October 2020 to September 2021, researchers at Menelik II Hospital’s Children’s Eye Care Center in Addis Ababa, Ethiopia, conducted the study. An ethics committee of Addis Ababa University’s College of Health Science, Department of Ophthalmology research, and publication board reviewed and approved the study protocol, and the research was carried out under the 1975 Helsinki Declaration, as revised in 1983. By assigning a code to each patient, confidentiality and anonymity are preserved.

### Criteria for inclusion

1. The baby must be at least 6 months old [16].
2. The child must have reduced visual acuity or difficulty seeing when compared to other children his or her age.
3. The abnormalities discovered during the eye exam do not account for vision loss.
4. The child has a medical diagnosis that affects his or her brain (neurological insults).

A child with an ocular problem, as well as CVI, was excluded, making it difficult to determine whether the behavior was caused by the ocular or cerebral condition.

The study subjects were chosen based on a referral to a pediatric ophthalmology clinic for an ophthalmic evaluation of the child. The invitation to participate in the study has been extended and written consent was obtained.

The patient’s sociodemographic data (age at test, age at CVI onset, sex, place of delivery, gestational age), presenting compliant, causes, systemic findings, and additional handicapping conditions from referral paper or interdisciplinary

consultation, parent interview for the child’s functional vision evaluation, and ophthalmic examination were entered into the structured questioner.

The referring clinician for the child provided the necessary information regarding the neurological, developmental assessment, and radiological input, all of which are known to cause damage to the visual pathways and visual processing areas of the brain. The information was gathered from the patient’s referral or chart consultation notes, interviews, observations, and direct assessment [17].

A parent interview was conducted for the child’s functional vision evaluation in order to identify visual perceptual disorders. (Interview questionnaire) The interviewees’ responses were matched to the ten specific behavioral characteristics shared by children with CVI.

The examination in the pediatric ophthalmology clinic began with a quiet observation of the child’s spontaneous visual behavior and motor functions. The level of visual interest displayed by a child when entering a new environment provides a general indication of their overall functioning (Novelty).

Children were examined after regular clinic hours in a quiet room, while seated in the lap of a caregiver who could provide truncal or neck support as needed. Supine examinations of children were also performed. The examiner approached the child from a safe distance (about two-thirds of a meter), addressed him/her by name, and smiled. Throughout the functional vision assessment, the child was observed for what he or she was looking at, the size and nature of the object that captured and held the child’s attention, as well as the distance the child was able to appreciate the object of regard.

During the direct assessment, the tester spent several minutes depending on the age of the child, completing the tasks in a long time while talking and playing with the child to evaluate the presence and degree of individual characteristics. No sudden jerky movements or using loud toys to attract attention, which may be alarming to the child, were used.

The binocular functional vision was usually assessed. Depending on the age and severity of the physical disability, different tests were used. The objects used for visual stimulation were red or yellow toys and white torch lights for pupillary reflex.

The following visual behavioral examinations were primarily performed for CVI study subjects to assess the nature of the visual insult and attempt to establish the three phases of CVI severity [18].

Moving target toys that light up to attract the child’s attention were used to do visual behavioral assessments using a behavioral assessment method that had been drawn from Roman- Lantzy characteristics:



- 1) Color preference
- 2) Need for movement
- 3) Visual latency
- 4) Visual field preferences
- 5) Light-gazing
- 6) Decreased distance viewing
- 7) Atypical visual reflexes
- 8) Novelty
- 9) Complexity
- 10) Visual-motor

Expected response: the examiner was looking for responses to light and high-contrast (red and yellow) toys, such as movement of limbs, or heading towards the visual target, postural reactions, avoiding reactions, and changes in facial expression.

The visual field test in CVI children (Visual field preferences) was assessed based on the behavioral responses, reflecting the child's ability to locate targets presented in different areas of the visual field.

The classification of visual behavior as mild, moderate, and severe was based on the CVI range: a functional vision assessment tool designed specifically for use with individuals with CVI that was developed by Dr. Christine Roman-Lantzy [17]. Visual functioning in CVI is measured across the CVI Range [1-10].

- 0-3 Phase I Most severe impact on visual functioning
- 3-7 Phase II Moderate impact on visual functioning
- 7-10 Phase III Closest to typical visual functioning

Completing an examination of the eyes:

- Intraocular pressure of both eyes was measured using I care when the child gets calm or sleep and results were recorded.
- Oculomotor function assessment including fixation, ocular alignment, and motility was checked by corneal light reflex tests and a near cover test, when possible by using toys that light up to attract the child's attention at 2/3 of a meter distance.
- Portable slit lamp biomicroscopy was used to examine the adnexa, conjunctiva, cornea, anterior chamber, iris, and lens and positive pertinent findings were recorded.
- The pupillary examination was performed with a moderately bright torchlight with the room lights

dimmed. Direct and consensual pupillary reflexes were noted.

- Fundus examination using a direct ophthalmoscope was performed after dilating the pupil using 1% Tropicamide and pertinent positive findings were recorded.
- Cycloplegic refraction was done by an optometrist.

### Operational definition

Cerebral visual impairment was diagnosed: if the patient meets the following three criteria:

1. The vision loss is not explainable by abnormalities found on the eye examination. A normal pupillary reflex, normal eye/near normal abnormal eye exam that cannot explain the lack of functional vision.
2. When a child exhibits one or more visual or behavioral characteristics: Impairment in his /her visual and/or visual perceptual abilities based on parent/caregiver interview and direct assessment of the visual behaviors [17].
3. A neurological medical diagnosis; (From history or referral paper the child's birth or medical history to determine the presence or probability of brain damage, which might have caused CVI + Neuro-imaging result) [18].

Refractive error: Hyperopia and myopia are defined as the spherical equivalent of +/- 3.00 D Sph [19].

### Data analysis procedure

The questionnaires were checked for completeness, cleaned manually, and entered into an Excel spreadsheet daily. SPSS version 20 was used to analyze the data. The results were presented in tables and graphs. Categorical variables were presented in percentage while continuous variables were presented in mean (SD) and ranges.

### Results

Forty children met the criteria of inclusion in this study, representing 1.96% of all children seen at the children's eye care center in the study period. From these 40 children, the study was able to construct a profile for the CVI children. The mean ( $\pm$  SD) age of children was 2.56 ( $\pm$  1.98) years. Their ages ranged from 6 months to 9 years. As shown in Table 1, there were 24 (60%) males. The highest frequency of consultation for children with CVI was recorded among pediatric patients between 1 and 5 years of age, constituting 75.0% of the patients. Thirty-three (82.5%) of children with CVI were residents of urban areas.

Table 2 summarizes the causes and patterns of neurologic abnormalities found in the sample. As shown in the Table,



the most common cause of CVI among the study population as reported in the referral papers was hypoxic-ischemic encephalopathy, which was mentioned in 28 (70.0%) of the referral papers. All subjects in this study had a neurological deficit, and the most frequent deficit was seizures, with a rate of 47.7%.

The brain magnetic resonance imaging [MRI] findings were abnormal in 60% and were not done in 27.5% of children with brain damage or disruption of brain development referred for assessment of functional vision. Fourteen out of 40 children (35%) with CVI had ischemic encephalopathy on their neuroimaging report. Other abnormal MRI findings included central nervous system [CNS] malformation in 6 (15%), global cortical atrophy in 3 (7.5%), and periventricular leukomalacia in 1 (2.5%).

Thirty-one of the 40 children (77.5%) with CVI were referred to the hospital's children's eye care center by pediatricians or pediatric neurologists, and 9 (22.5%) by ophthalmologists from the same hospital and other secondary eye care units.

During the parental/caregiver interview, 47.5% of them stated that they had no vision concerns when they went to the treating physician, and the child was referred for vision and eye evaluation by their treating physician. Those parents

with concerns about their children's vision (66.7%) reported an inability to see and follow, followed by squinting (28.6%) and 4.8% of the concerns were related to rapid eye movement.

Table 3 shows compiled behavioral characteristics and associated ocular findings in children with CVI. A child had many of the CVI functional vision characteristics. All the study participants had difficulties with visual complexity. In phase I, 47.5% of CVI patients in the study population had a functional vision; 42.5% had it in phase II, and 10% had a functional vision in phase III.

## Discussion

Prior research has shown that cerebral visual impairment (CVI) is becoming more common in low-income countries as a result of advances in perinatal care; Huo, et al. [19], for example, reported that CVI occurred in 2.4% of all participants examined and is frequently accompanied by additional ophthalmological problems and is almost always associated with other, serious neurological abnormalities.

However, these studies have either been retrospective or have not focused on patients whose disorder was not appropriately and timely recognized using the relevant symptomology of CVI. In this study, we determined the profile of CVI prospectively in a group of children with referrals suggestive of brain damage or disruption of brain development at a tertiary hospital children's eye care center.

We found that CVI accounted for 1.96% of children visiting the children's eye care center in the study period: as a district subgroup of patients having functional visual deficits with associated ophthalmic and neurologic impairments.

In our study, prematurity was found in 3 (7.5%) of the study participants, which is comparable with the study done in New Zealand where it was 7% [12].

**Table 1:** Characteristics of children with CVI as seen at the children's eye care center of Menelik II hospital, Addis Ababa, Ethiopia.

	Categories	Number of patients	% of study population (n = 40)
Age at examination (years)	< 1	7	17.5
	1-5	30	75.0
	> 5	3	7.5
Age at CVI onset	At Birth	39	97.5
	After birth	1	2.5
Sex	Male	24	60.0
	Female	16	40.0
Address	Urban	33	82.5%
	Rural	7	17.5%
Place of birth	Home	2	5.0
	Health unit	38	95.0
Gestational age	Premature	3	7.5
	Term	37	92.5

**Table 2:** As reported in the referral paper, the causes and neurological deficits in children with CVI.

	Number of patients	% of study population (n = 40)
Causes		
Hypoxic-ischemic encephalopathy	28	70.0
Brain structural abnormalities	6	15.0
Cerebral vascular accident	3	7.5
Periventricular leukomalacia	1	2.5
Meningitis/ encephalitis	1	2.5
Idiopathic	1	2.5
Neurological deficits		
Seizures	19	47.7
Cerebral palsy	13	32.5
Mental/developmental delay	4	10.0
Trans-tentorial hearing loss	1	2.5
Microcephaly	1	2.5
Unknown	2	5.0

**Table 3:** Functional visual characteristics and ophthalmic findings in children with CVI.

	Number of patients	% of study population (n = 40)
Functional Visual characteristics		
Color preference	21	52.5
Need for movement	33	82.5
Visual latency	35	87.5
Visual field preferences	32	80.0
Complexity	0	0.00
Light-gazing & non-purposeful gaze	26	2.5
Distance viewing	6	15.0
Atypical visual reflexes	16	40.0
Visual novelty	5	12.5
Visual guided reach	2	5.0
Ophthalmic findings		
Refractive error (Myopia (40%), Hyperopia (15%))	22	55.0
Strabismus (Exotropia (22.5%), Esotropia (7.5%))	12	30.0
Primary motor nystagmus	5	12.5
Temporal optic disc pallor	3	7.5
Corneal opacity	1	2.5





Our findings are in accordance with previous studies which have reported that hypoxic-ischemic encephalopathy is the commonest cause of CVI mentioned in a referral paper [10,12,13]. Meningitis was etiological in our study group, as has been reported elsewhere [8].

CVI is almost always seen in children who have neurological problems [19]. All of the CVI patients in our study had associated neurological deficits. However, 42.5% of children had additional ophthalmic deficits according to the data. This means that ophthalmologists must be aware of the steps that must be taken to improve the visual experience of children with CVI.

Our study revealed seizures as the most frequent occurrence in CVI children. Also, we found cerebral palsy, mental/developmental delay, trans-tentorial hearing loss, and microcephaly comparable to the Huo, et al. study [19].

The present study has shown that ischemic encephalopathy was the most common neuroimaging finding seen in the study population. The other neuroimaging findings are CNS malformation, global cortical atrophy, and periventricular leukomalacia [21].

In our study, refractive error necessitating spectacle correction accounts for the most common ocular problem seen, followed by strabismus, nystagmus, and corneal opacity. This finding was also similar to a study done in India [12].

Detailed observation of the visual behavior added new elements which allowed us to gain a better understanding of visual function indicators for possible cerebral visual disorders at an early stage. There is a wide variety of CVI behaviors. A child may have one or more of these CVI behaviors, and the degree of severity of the presenting functional vision characteristic may vary considerably [17]. Based on Roman-Lantzy's three phases of the CVI Range, we found that 90% of children with CVI at the test time had Phase I or Phase II vision.

Our analysis is based on a referral practice and describes the age of the majority of our study population, which was between 1 and 5 years at presentation and had most of the eye findings. There is a need for children's eye care centers to equip themselves with the necessary skills and facilities to ensure children receive the additional support they require at the earliest possible age. Furthermore, many milder cases of CVI went unidentified since they were not able to get a referral for ophthalmic evaluation. To further support a diagnosis of a mild form of CVI in the child, known causes of CVI in the child's medical history should be the most important consideration for CVI screening.

Most notably, this is the first study to our knowledge to describe the causes, associated ophthalmic and neurological deficits, and visual behavioral characteristics in Ethiopia at

the children's eye care center. However, some limitations are worth noting. The first limitation was that children with mild forms of CVI may not have been included in the study because we used visual perception testing as a diagnostic functional vision test, and may have underestimated the prevalence of the disorder. The other limitation was that incomplete referral papers resulted in difficulty in analysis and drawing conclusions due to missing data.

## Conclusion and recommendations

The findings of this study demonstrated that CVI as a cause of childhood vision impairment is significant. Hypoxic-ischemic encephalopathy is the most common cause of CVI. The majority of children with CVI have serious neurological and ophthalmic abnormalities. The study emphasizes the importance of evaluating the visual behaviors (target needs for movement, visual latency, visual field preference, and color preference) in unexplainable vision loss in normal eyes. Providing children's eye care centers with the necessary skills and facilities to ensure children with CVI receive the necessary support and strengthening team approaches to address challenges.

## Acknowledgment

Professor Abebe Bejiga's expert assistance is gratefully acknowledged. The staff at Menelik II hospital children's eye care center, the ophthalmic residents, and the families who inspired this work deserve our deepest gratitude.

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