Ocular Manifestations in Children with Cerebral Palsy
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ABSTRACT

Background. Cerebral palsy (CP) is a group of non-progressive disorders of movement or posture. In patients with CP, a visual handicap can have a direct impact on psychosocial status. This study was conducted to determine the nature and frequency of ocular abnormalities in children with CP in Nepal.

Methods. A total of 36 children with cerebral palsy residing in Kathmandu Valley underwent a detailed optometric examination. The examination consisted of assessment of visual acuity, extraocular motility, cover test, anterior segment and posterior segment evaluation. Cycloplegic refraction was performed on all children.

Results. The mean age of the children was 5.48 ± 4.75 years. Ocular abnormalities were present in 86% of the children. Refractive error was the most common ocular abnormality, and was present in 78% of the children. Refractive error was the sole finding in 19% of the children tested. Strabismus was the second-most common ocular abnormality, and was present in 36 percent of the children. Fourteen percent of the children had a variant of nystagmus. Central cortical visual impairment was suspected in 11%.

Conclusion. Refractive error and strabismus are common ocular manifestations in children with CP in Nepal. Increased awareness regarding these abnormalities to the parents and health professionals may contribute to improving these children's academic abilities as well as their overall developmental process.

Keywords: cerebral palsy, Nepal, nystagmus, refractive error, strabismus

Introduction

Cerebral palsy (CP) describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behavior, and/or by a seizure disorder.1 The diagnosis of CP is usually made on the basis of uncoordinated muscle movements and delays in reaching developmental milestones. In addition to a physical examination, computerized tomography and/or magnetic resonance imaging of the child’s brain to look for the brain insults and abnormalities may help to diagnose the condition.2 The assessment and management of visual disorders in physically or intellectually impaired children presents a complex challenge for the clinician.3 In different CP motor disorder syndromes, a number of visual disorders have been previously described, including: strabismus (33%-50%),4-10 refractive anomalies (28.5% - 54%),4-6,9,10,11 cortical visual impairment (28%-48%),4,7 optic atrophy (2%-50%),5,7,8 nystagmus (1% - 18%),4-6,8-10 ptosis (1% - 2.5%),5-6,8 and accommodative dysfunctions.12

Refractive errors, nystagmus and strabismus often greatly affect the quality of vision of CP patients. The earlier the diagnosis can be made and appropriate
management instituted, the more the children will be helped in their learning and academic success.

This study explores and documents the variety of ocular problems in Nepalese children with Cerebral Palsy and educates the parents of these children concerning the importance of eye and vision examinations. Several similar studies have been conducted in different parts of the world with varying findings. This study is the first of its kind to look for ocular abnormalities in persons with CP in Nepal. It is believed this study will help to promote earlier examination of CP children so that quality of life (QOL) in these children may be improved.

Methods

Our study was cross-sectional, descriptive, and community/hospital-based. It was conducted at two centers: the Self Help Group for Cerebral Palsy (SGCP) at Dhapakhel, Lalitpur (a rehabilitation center for CP children in Nepal) and BP Koirala Lions Center for Ophthalmic Studies (BPKLCOS) (a tertiary eye hospital in Kathmandu, Nepal). All the patients were diagnosed with CP by a pediatrician. Of the 36 children with CP, 23 children were from SGCP and the remaining 13 children were referred to BPKLCOS from Kanti Children’s Hospital (general pediatric hospital) Kathmandu, Nepal. The study took a full calendar year to conduct.

Informed consent was granted by the guardians and a responsible administrator from SGCP. A brief history of the visual behavior of the child, along with histories of antenatal, postnatal and other maternal factors were taken.

Every child was seated either on their parent’s lap or in adaptive wheel chairs throughout the examination. Presenting visual acuity (aided or unaided) was assessed monocularly. Depending upon the age and cooperation of the patients, a variety of methods were used for assessing visual acuity: Snellen chart; Kay Picture test; Preferential Looking Chart, and Central Steady Maintained technique (CSM). CSM involves covering one eye while the child is fixating on an object (small toy). The non-covered eye should maintain central, steady fixation, which is maintained through a blink. LogMAR notations of the visual acuity recorded were converted into Snellen equivalent fractions. For children where other visual acuity tests could not be applied, the Central Steady Maintained (CSM) technique was used.

Ocular motility was evaluated by moving a highly interesting colorful object or torchlight in six different cardinal gazes. In uncooperative patients, the Doll’s Head Tilt test was used to assess the extra ocular motility. A cover test, Hirschberg test or Brückner’s test, was completed to detect the presence of strabismus. This was done for both near and distance. The laterality and constancy of any deviation were noted and nystagmus was assessed as well when possible.

Refraction under cycloplegia was performed in all of the children. Cycloplegia was obtained with one drop of 1% cyclopentolate (regardless of age) instilled every 5 minutes for a total of 3 drops. Cycloplegia was followed by retinoscopy 30 minutes after the instillation of the last drop. The standards for quantifying refractive error were as follows: myopia was considered to be a mean spherical equivalent of ≥0.50D; hyperopia as ≥ +1.00D; astigmatism as ≥ -1.00D in any meridian; and anisometropia (mean sphere) as ≥ 1.00D.

Detailed examination of the anterior segment was carried out with either a hand held slit lamp or a standard slit lamp biomicroscope (Haag-Streit). If biomicroscopic evaluation was not possible due to a poor level of cooperation, a torchlight (flashlight) was used. The posterior segment evaluation in every patient was carried out in a fully dilated state of the eyes with direct ophthalmoscope or indirect ophthalmoscope, whichever was possible. Findings of the anterior and posterior segments were recorded in a preset proforma especially designed for the study. If the patient was found to have reduced visual acuity despite normal ocular health, cortical visual impairment (CVI) was suspected. The data were evaluated using SPSS Computer software (version 14.0) and the results were analyzed accordingly.

Results

A total of 36 children with cerebral palsy were included in the study. Among them 75% were males.
The mean age of the children was 5.48 ± 4.75 years (Range: 6 months to 21 years). Sixty-six percent (24) of the children had not had any previous eye examination. Ocular abnormalities were detected in 86% (31) of the children. Multiple ocular abnormalities were present in 66% of the children. The overall co-morbidity findings of 36 children with CP are summarized in Figure 1.

**Visual Acuity.** Presenting visual acuity was assessed in all the children. The most frequently used method to assess visual acuity was the Kay Picture test, which was used for 41% (15) of the children and was followed by the CSM technique which was used for 36% (13) of the patients. The percentage of children and the tests that were used for them is depicted in Table 1. All the children had equal visual acuity in both eyes, with only one exception.

A presenting visual acuity of 6/6 was measured in 8% (3) of the children, 6/9 to 6/18 in 22% (8) of the children and 6/18 to 6/60 in 28% (10) of the children. Three percent of the children had measured visual acuity less than 6/60. A positive CSM was recorded in 22% (8) of the children and negative in 17% (11) of the children.

**Extraocular Motility.** Extraocular motility examination was performed in 83% (30) of the children. Among them, 93% (28) had full ocular motility in all positions of gazes. Only 7% (2) of the children had a restriction of ocular motility. One patient had restriction of ocular motility in all gazes; one patient demonstrated unilateral limitation of abduction.

**Refraction.** Refractive error as defined by the parameters of this study was present in 78% of the children. In 19% of the children, refractive error was the sole cause of visual deficit. Thirty percent of the children had myopia, 24% hyperopia, and 30% (11) had astigmatism, either in isolation or compounded with myopia or hyperopia. Eight percent of the children had mild to moderate anisometropia. Mean spherical equivalent refractive error of right eye was +0.25 ±2.16DS (range -4.75 to +5.25) and + 0.27 ± 2.21DS for left eye. Figure 2 shows the pattern of mean equivalent sphere refractive error of the left eye in relation to age.

**Binocular function.** Binocularity was assessed using various tests according to the cooperation of children and feasibility for the examiner. The cover test could be performed on only 30% (11) of the children. All other cases needed multiple tests (Bruckner test, Hirschberg reflex test or Krimsky test) for diagnosis of strabismus and measurement of the deviation. Strabismus was present in 36% (13) of the patients. The most common type of strabismus was alternating exotropia (61%), followed by three cases of unilateral exotropia and two cases of alternating esotropia. Two cases had strabismic angle magnitude of ≤15° and all other 11 strabismic cases had an angle of ≥ 25°. Nystagmus was present in 14% (5) of the children.

**Pupil and optic nerve status.** Normal pupillary responses were found in 75% of our subjects. Relative afferent pupillary defect (RAPD) was seen in 19% of the children. One child had congenital unilateral mydriasis and one had a unilateral mid-dilated pupil unresponsive to light. The optic disc appeared normal in 78% of the children. Complete optic atrophy was seen in 8% (3) of the children and temporal disc pallor was seen in 14% (5) of the children. Of all the cases, 11% of the children were suspected to have central cortical visual impairment (CVI).

**Other findings.** Other ocular diagnoses were ptosis (3%), Peter’s anomaly (3%), and Retinopathy of Prematurity (3%).

### Discussion

Only one-third of the children studied had a previous history of ocular examination, which strongly suggests that eyecare practitioners and the parents of children in Nepal are still unaware of the high

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**Table 1: Methods used for assessing presenting Visual acuity in children with Cerebral Palsy**

<table>
<thead>
<tr>
<th>Test used</th>
<th>Percentage of children</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kay Picture test</td>
<td>41%</td>
</tr>
<tr>
<td>Central Steady and Maintained</td>
<td>36%</td>
</tr>
<tr>
<td>Sheridan Gardiner</td>
<td>11%</td>
</tr>
<tr>
<td>Preferential looking test</td>
<td>11%</td>
</tr>
</tbody>
</table>

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*Figure 2. Mean equivalent sphere of refractive error of left eye against age*
incidence of ocular problems in those with cerebral palsy. Most of the children (81%) were undergoing rehabilitation at their schools for their disabilities, so the majority of our subjects were examined at their usual rehabilitation center. This gave the clinician an important advantage in that the subjects cooperated more fully in the examination because of the familiar environment. Moreover, their teachers were immediately available to be educated about the children’s difficulties and were offered interventional suggestions.

A Snellen equivalent visual acuity of 6/6 was recorded in three children (8%). Seventy percent of the children had visual acuity poorer than 6/18. One factor for this might have been some children’s relative non-motivation, fatigue and prolonged inattention to the acuity tests during the testing duration. Moreover, it cannot be said whether the presenting acuity recorded for each child was actually their best acuity or merely the best effort the child could offer. Even the children in whom CSM was positive might have had better visual acuity than estimated.

The accurate measurement of visual acuity in CP patients is a difficult task. A very large range of visual acuity is noted in the literature on CP. More reliable tests like Kay Picture Chart, Snellen /Sheridan Gardiner Chart and Preferential Looking Chart were used in only about two thirds of the patients. The less reliable and qualitative vision assessment method, CSM technique, was used in the remainder. A VEP acuity measurement technique is an alternative option for assessing visual functions in uncooperative and non-verbal children, however the test couldn’t be used in this study due to lack of such facility in Nepal.

A spectrum of visual disorders is prevalent in CP and has been described extensively in the literature, ranging from a frequency of 28% to 86%. Our findings are in agreement with the higher figure, showing a prevalence of 86%. In this study, (66%) children had more than one visual deficit.

The prevalence of ocular abnormalities in children with CP in Nepal is significantly higher (p<0.001) in comparison to the general population of school children in Kathmandu (age range 5-16 years). This emphasizes the need for a proper ocular examination of all persons diagnosed with cerebral palsy.

Refractive error, as defined in this study, was the most common type of abnormality documented (78%). This is in contrast to the study by Govinda et al. (N=70) who found strabismus (35.7%) to be the most frequent abnormality. However, different prevalence of abnormal refractive errors in patients with CP, from 28.5% to 54% have been reported in other parts of the world in CP. In Nepal, the prevalence of refractive error in normally developing children of school age is 3%-18% which is far less than what we found in our CP patients (78%). Moreover, only 25% of our CP patients were wearing spectacles when first examined. This emphasizes the need for appropriate referral and management of refractive problems, and counseling of parents of the need for vision care in CP children.

Much of the literature quotes a higher prevalence of hyperopia in CP but our study did not agree with the prior studies. Instead, we found myopia occurred to a greater extent (39%) than hyperopia (29%). We found the incidence of myopia and hyperopia to be considerably larger than that reported in normal children. Fant and Perlstein report a higher prevalence of myopia in those with spastic CP and found that hypermetropia was more prevalent in CP with dyskinesia. We could not analyze our refractive findings according to the CP type due to lack of sufficient medical records.

The prevalence of astigmatism in our study population (39%) is similar to some previous studies. Kozeis and Anogeianaki et al. reported the incidence of astigmatism to be 40.9% and Govinda and Lamba reported an even higher incidence (50%). The higher prevalence of refractive error even in the lower age group suggests the emmetropization process may have been hampered. Our findings on the prevalence of refractive error were similar to the findings in children with Down syndrome of Nepal, but hyperopia occurred in greater amounts. As the present study shows a significant prevalence of refractive errors, additional studies should be conducted to understand the development of refractive errors in CP in our context.

The prevalence of strabismus observed in the present study (36%) matches well with other parts of the world (India, 35.7%, 39%, Japan, 33.1%, Africa, 50%) Alternating exotropia was seen to occur more frequently in CP patients than in normally developing children (χ2=51.25, df=1, p<0.001). Various ranges for the incidence of nystagmus have been reported in CP (1.02% - 18%) and our study agrees with this range (14%).

The prevalence of ptosis (3%) found in our study agrees with that reported in other studies (1.43%,
1.02%, 2.5%). Another finding of our study, comparable to similar studies, is the prevalence of cataracts (2 cases, 5.5%). We also found agreement with the literature on the prevalence of retinopathy of prematurity (3%) and Peter’s anomaly (3%). Katoch et al. found normal pupillary reactions in 94% of the CP children they studied. This was observed in 75% of our cases. Complete optic atrophy was present in three (8%) children. Similar findings are quoted by Black (10%). Four children (11%) had very poor visual acuity not attributable to any ocular abnormalities or severe disorders. These children were suspected of having either retrochiasmal or central cortical lesions. This data on CVI was less than the study done by Katoch et al. (28%) and Lagunja and Oluluyev (48%).

We agree with many authors writing on the topic of CP that timely referral and appropriate management, including refractive correction, surgery, and intervention with rehabilitative counseling may help to improve the QOL children with CP.

**Conclusion**

Ocular abnormalities are very common in children with CP. Our findings are similar to previous studies in the literature and confirm that children with CP are at more risk of developing ocular abnormalities. Parents and the health practitioners who are responsible for the health and overall development of CP children should be aware of the ocular defects that may be present in these children. Early intervention will help for the child's physical, social, academic and visual development. A full eye examination should be sought as soon as a diagnosis of CP is made and yearly thereafter.

Thorough ocular assessment of cerebral palsied patients can be difficult. However, familiar surroundings and caring environment, adequate clinical attention can facilitate assessment of all the visual disorders of these patients. Early referral of the children diagnosed with CP for ocular examination is of utmost necessity for better visual prognosis.

**Acknowledgement**

We would like to acknowledge the institute B P Koirala Lions Center for Ophthalmic Studies for technical support with the instruments and vehicle. We are thankful to Mr. Bimal Lal Shrestha; the director and Aaradhana RajBhandari, the staff of Self Help Group for Cerebral Palsy for supporting us to conduct the study. Finally, we are most sincerely grateful to all the parents of CP children who allowed us to conduct the study. We thank all the children for their involvement. We also wish to thank Drs. Mark Taub and Dominick Maino for their editorial assistance in the preparation of this manuscript.

**References**