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Ocular abnormalities in children with cerebral palsy

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Summary

This prospective study was undertaken to evaluate the prevalence of ocular abnormalities, as well as describe the various eye defects seen among cases of cerebral palsy presenting at the Paediatric Neurology Clinic, University College Hospital, Ibadan, Nigeria. All cases of cerebral palsy seen at the Paediatric Neurology Clinic, University College Hospital, Ibadan, over a period of 18months were carefully evaluated by the Paediatric Neurologist for signs of ocular abnormalities. Those in whom such abnormalities were found were referred to the Consultant Ophthalmologist for detailed eye examination and accurate description of the ocular abnormalities. One hundred and forty nine children with cerebral palsy were seen during the period of study, forty two had associated ocular abnormalities, giving a prevalence rate of 28.2%. More than half (61.9%) of the cases were completely blind. The major ocular abnormalities identified in the affected cases were strabismus (50%), optic atrophy (50%) and cortical visual impairment (47.7%). Other eye defects less frequently seen were nystagmus (9.5%) and refractive errors (4.8%). Presence of spastic quadriplegia was associated with an increased risk of ocular abnormalities. Ocular abnormalities are a frequent problem in children with cerebral palsy. Evaluation of all children with cerebral palsy must include amongst other things, a full ophthalmologic evaluation, even when no gross eye anomalies are visible to the attending physician. Early identification of these defects in children with CP is crucial in order to institute prompt therapy in cases with defects that are amenable to treatment.

Keywords: *Ocular, cerebral palsy*

Résumé

Cette étude prospective était effectuée pour évaluer la prévalence des signes d'anomalies oculaires et à décrire les différents défaut des yeux parmi les cas de palsie cérébrale évalué dans la clinique pédiatrie neurologique du Centre Universitaire Hospitalier, Ibadan, Nigeria pendant 18 mois ; ceux ayant des anomalies étaient transparents aux consultations ophtalmologiques pour diagnostiquer les yeux et plus précisément décrire les anomalies oculaires. Cent quarante neuf enfants ayant le cérébrale étaient vu durant cette période, quarante deux avaient des anomalies oculaire d'un taux de 28.2%, plus de la moitié(61.9%) des cas étaient complètement aveugle. l'anomalie majeur identifiée était le strabisme(50%) l'atrophie optique (50%) et l'impair de la cortique visuel (47.7%). Autres défauts oculaires moins fréquents étaient le nystagmus (9.5%) et les erreurs réfractaires (48%). La présence de la quadriplégie spastique était associée à une augmentation du risque d'anomalie oculaire. Nous recommandons que l'évaluation de cette condition chez les enfants doit inclure une complète évaluation oculaire/ophtalmologique même sans anomalie grandiose visible par le médecin. Une détection précoce chez les enfants est importante afin d'initier le traitement précis selon les types de défauts.

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Introduction

Cerebral palsy (CP) remains a major cause of disability in childhood [1]. Cerebral palsy (CP) refers to a group of non-progressive, but often changing motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of development [2]. The association of certain eye defects with cerebral palsy has been known since Little's early papers on the disease in 1834[3] and various forms of eye defects have been described in children with cerebral palsy. These include nystagmus, strabismus, refractive errors, optic atrophy, cataracts, field defects, corneal abnormalities and cortical visual impairment [4,5]. Various ocular abnormalities have been reported in all the recognized forms of CP.

With increasing survival of preterm infants, periventricular leucomalacia (PVL), a major problem of preterm infants, particularly those delivered between gestational age 24-34 weeks has become a principal cause of visual impairment in children in

the developed world [6,7]. Cerebral visual dysfunction caused by PVL is characterised by delayed visual maturation, sub normal visual acuity, crowding, visual field defects and visual perceptual cognitive problems [6]. Examination of the affected children often reveal optic disc abnormalities, strabismus, nystagmus, and deficient visually guided movements [6,7]. Other ocular abnormalities described in children with CP and a background history of prematurity include cortical visual impairment and retinopathy of prematurity [8].

A high incidence of visual defects have also been reported in children with hemiplegic CP [9]. Guzzetta *et al* in a study of children with early-onset hemiplegia and hemiplegic CP, reported impaired visual acuity, visual field defects and optokinetic nystagmus as the major ocular abnormalities in these children [7]. Major visual abnormalities in spastic quadriplegic CP include refractive errors, strabismus, nystagmus and optic atrophy [9].

Vision is one of the most important senses required for normal physical and cognitive development in childhood. Presence of severe visual defects might pose a significant problem to the rehabilitation of children with cerebral palsy. Visual dysfunction has been shown to further compromise postural control in children with cerebral palsy [10]. Early detection of ocular abnormalities in children with cerebral palsy will improve the overall prognosis in affected children as the treatable forms of these defects are given early and prompt intervention. Although CP is one of the leading causes of disability in the developing world [11,12] the ocular defects associated with this common childhood neurological disorder in African children is not well documented in literature. This study was undertaken to evaluate the prevalence and nature of ocular abnormalities in children with cerebral palsy. The findings in the study will help to improve the care of children with cerebral palsy.

Materials and methods

The study was prospective and was carried out at the paediatric neurology clinic and the ophthalmology clinic of the University College Hospital, Ibadan, Nigeria. The University College Hospital, Ibadan provides tertiary health care services, serves as the major referral centre for the South Western part of the country and also receives cases from other parts of the country.

The subjects studied comprised children with a diagnosis of cerebral palsy seen at the paediatric neurology clinic of the hospital over a period of 18 months, May 2004 – October 2005. Informed consents were obtained from the caregivers of all the children studied. The diagnosis of cerebral palsy was clinical, based upon a history of abnormal motor development that was not progressive, coupled with the presence of abnormal neurological signs that localised the lesion to the brain [13]. All the cases were evaluated by and the diagnoses of cerebral palsy made by the paediatric neurologist. A detailed history of the illness, development, pregnancy, birth and neonatal periods were taken. A thorough neurologic examination was carried out on each patient at presentation and this entailed assessment of consciousness, speech, cranial nerves, cerebellar, motor and sensory functions.

Classification: Spasticity involving one upper limb and the ipsilateral lower limb was classified as hemiplegia, spasticity of all four limbs, with the upper limb involvement more marked than or equal to that of the lower limbs, spastic quadriplegia, spasticity of the lower extremities, with a variable but lesser involvement of the upper limbs, spastic diplegia. Cases with extrapyramidal symptoms were classified as dyskinetic, those with hypotonia classified as hypotonic, while the mixed class had a combination of features of the spastic and extrapyramidal types [14].

A preliminary eye examination was carried out by the paediatric neurologist in the paediatric neurology clinic. This involved gross ocular examination for ocular alignment, extraocular movements and presence of strabismus or nystagmus. Visual acuity was assessed by light fixation and response to visual threat. The pupils were examined using a pen torch to assess pupillary size and response to light reflex. All the children with ocular abnormalities detected on preliminary examination by the paediatric neurologist were referred to the ophthalmologist for a detailed ocular examination at the eye clinic. At presentation in the eye clinic, each child's ocular examination began with the assessment of visual acuity. Children under 4 years were tested for light fixation, ability to identify and match pictures and letters. Those above 4 years were tested with the Snellen charts. The anterior segment examination was done with a pen torch, while dilated fundoscopy and mydriatic retinoscopy was done under light sedation. Topical Tropicamide 0.5% was used as dilating agent, while Paraldehyde at a dose of 1ml per year of life, up to a maximum of 5ml, was given intramuscularly as a sedative. The findings were recorded in a datasheet prepared for the study.

Data were entered into a microcomputer and analysis done with the SPSS 11 for windows software. Categorical variables were compared using the Chi square. A p-value of less than 0.05 was regarded as significant.

Table 1: Pattern of ocular abnormalities in 42 children with cerebral palsy

Ocular abnormality	Number of cases**	% Total**
Optic atrophy	21	50.0
Strabismus	21	50.0
Cortical visual impairment	20	47.7
Nystagmus	4	9.5
Refractive errors	2	4.8

***Some children had more than one form of ocular abnormality*

Results

One hundred and forty nine children with cerebral palsy were recruited into the study, their ages ranged from 6 months to 13 years, median 18.0 months. There were 95 males and 54 females, giving a male to female ratio of 1.8:1. The major underlying causes of CP were severe birth asphyxia (49.7%), severe neonatal jaundice (26.8%), post infectious brain damage (10.7%) and prematurity (3.4%). Other less common causes of CP found were craniosynostosis (1.3%), intrauterine infections (2.0%), and metabolic disorders (1.3%). The cause could not be ascertained in 6 (4.0%) cases. The spastic form of CP was the most predominant, found in 123 (82.6%) cases. Other forms of CP seen in the study were dyskinetic 8.1%, mixed 6.7% and hypotonic in 2.7%.

Ocular abnormalities were found in 42 (28.2%) of the 149 children with cerebral palsy. Of these, 26 (61.9%) had total blindness, 13 (31.0%) were partially blind and only 3 (7.1%) had normal vision. The most frequent abnormalities were optic atrophy (50.0%), strabismus (50.0%) and cortical visual impairment (47.7%). Five (25%) of the 20 children with cortical visual impairment had total blindness, while the remaining 15 (75%) had varying degrees of residual vision. Table 1 shows the frequencies of the various eye defects in the study population. A larger proportion of children with spastic quadriplegia (39.5%) had associated visual impairment when compared with other topographic groups (17.8%). This observation was statistically significant ($\div^2$= 6.18, p==0.01). Table 2 shows the ocular abnormalities in the various types of CP

Discussion

The study identified ocular defects as a common associated deficit in children with cerebral palsy and this was found in 28.2% of the 149 children with CP studied. The reported prevalence of ocular abnormalities in children with CP varies widely, ranges between 17-90% and has been shown to vary with the severity of CP [4,14-17]. The prevalence of 28.2% found in this study is consistent with previous reports. Sankar and Mundkur [17] found ocular abnormalities in 28% of Indian children with CP and Sharma *et al* [15] reported ocular defects in 35% of the children

Table 2: Prevalence of ocular abnormalities in the principal types of CP

Type of CP	Blindness Total/ Partial *n (%)*	Optic atrophy *n (%)*	Cortical blindness *n (%)*	Strabismus *n (%)*	Nystagmus *n (%)*	Refractive error *n (%)*
Spastic Quadriplegia (n= 78)	31 (39.7)	17 (21.8)	16 (20.5)	14 (17.9)	4(5.1)	1 (1.3)
Spastic Hemiplegia (n= 32)	4 (12.5)	2 (6.25)	2 (6.25)	2 (6.25)	-	-
Spastic Diplegia (n=9)	-	-	-	-	-	-
Spastic Triplegia (n=3)	1 (33.3)	1 (33.3)	-	1 (33.3)	-	-
Spastic Monoplegia (n=1)	-	-	-	-	-	-
Dyskinetic (n=12)	-	-	-	1 (8.3)	-	-
Mixed (n=10)	2 (20.0)	-	1 (10.0)	2 (20.0)	-	1 (10.0)
Hypotonic(n=4)	1 (25.0)	1 (25.0)	1 (25.0)	1 (25.0)	-	-

with CP. Black [16] reported a higher prevalence of ocular defects among the 117 children with CP studied as 78% of them had eye defects. The higher prevalence of eye defects found in his study has been attributed to the marked severity of CP in the cases studied.

The major ocular abnormalities found in this study were optic atrophy, strabismus and cortical visual impairment. Half of the children with cerebral palsy in this study had strabismus and this is consistent with findings from previous studies [15,16]. Strabismus is one of the treatable ocular defects in cerebral palsy and early identification of this abnormality will optimize visual potential, as well as improve the management and overall prognosis of the child with cerebral palsy. Optic atrophy was found in 50% of the cases studied. In all the cases, the degree of optic atrophy was severe and all the affected patients were completely blind. Perinatal asphyxia is a well recognized cause of optic atrophy [18] and this condition was found to be the leading underlying cause of CP in this study. The relatively high prevalence of optic atrophy in this study may therefore be related to the aetiology of CP in the cases studied. Cortical visual impairment was found in nearly half of the cases in this study. Like optic atrophy, perinatal hypoxia has been reported as one of the leading causes of cortical visual impairment in childhood [19]. Five (25%) of the 20 children with cortical visual impairment had total blindness, while the remaining had residual vision in the eyes.

Nearly two-thirds of the CP cases with ocular abnormalities in this study had total blindness. This represents an additional limitation to the functional independence of the affected children and adversely impacts prognosis. Early recognition of these defects in children with CP is important in order to provide the much-needed support to the affected child and the family.

The prevalence of refractive errors found in this study was rather low (4.8%), when compared with previous studies [15,16]. Objective retinoscopy is difficult in children with cerebral palsy in the presence of nystagmus and strabismus. Automated refraction would have aided the identification of refractive errors but this could not be used in this study as this technique requires the cooperation of the subject which could not be obtained in most of the cases studied. The photoscreener is an effective means of screening children and young adults with severe learning disabilities for refractive errors and strabismus so that the children with these abnormalities may be targeted for a more detailed evaluation.[20] This equipment is however not available in our centre. These factors may have contributed to the low prevalence of refractive errors among children with cerebral palsy in the study. Multi-centred studies in other parts of the country will be useful in corroborating this finding. Eye anomalies associated with prematurity, which have been on the increase in the developed world, were an infrequent finding in this study. This may be attributed to the relatively high perinatal mortality associated with prematurity and very low birth weight in a developing country like Nigeria.

Ocular abnormalities are a frequent problem in children with cerebral palsy. They pose considerable challenge to the management of these children as their education and rehabilitation rely heavily on visual stimulation. Early identification of these defects in children with CP is crucial in order to institute prompt therapy in cases with defects that are amenable to treatment. Evaluation of all children with cerebral palsy must include amongst other things, a full ophthalmologic evaluation, even when no gross eye anomalies are visible to the attending physician.

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