Research Article

Prevalence of Strabismus in Patients with Cerebral Palsy

Elizabeth Mathew, Jasmine Mary Jacob and Sheldon Goudinho*

Dr. Somervell memorial CSI medical college, Karakonam, India
*corresponding author: Email: sheldonjasmine@gmail.com

[Received-15/02/2015, Accepted-25/02/2015]

ABSTRACT

Background: Cerebral palsy is a condition usually associated with treatable ocular pathologies such as strabismus. Objectives: 1. To observe and estimate the prevalence of various types of strabismus in children with cerebral palsy. To estimate the strength of association between the presence of squint and birth weight, and squint and gestational age of the child. Materials and Methods: This cross sectional, hospital based study was conducted in Dr SM CSI Medical College, Karakonam, Trivandrum. Eighty children diagnosed with cerebral palsy were included in the study. A detailed ophthalmic evaluation was done. Presence of and type of strabismus was noted. Results: Strabismus was found in 32 of 80 (40%) cases enrolled in the study. Exotropia was found to be 3.4 times more than esotropia. Alternating exotropia was the commonest type accounting for 17 cases (21.3%). No statistical association was found between squint and birth weight or gestational age.

Keywords: cerebral palsy, strabismus, birth weight, gestational age,

INTRODUCTION

Cerebral palsy is a diagnostic term used to describe a group of motor syndromes resulting from disorders of early brain development [2]. It is the most common and costly form of chronic motor disability that begins in childhood with a prevalence of 2/1000[2]. It is caused by a broad group of developmental, genetic, metabolic, ischemic, infectious and other acquired etiologies that produce a common group of neurological phenotypes. Neurological features can change or progress over time [2]. While it is usually associated with a wide spectrum of developmental disorders including mental retardation, visual, hearing, speech, cognitive and behavioural handicap, the motor handicap may be the least [2]. The association of certain eye defects with cerebral palsy has been known since Little’s early papers on the disease [1]. Strabismus, nystagmus, optic atrophy & refractive errors are common in children with cerebral palsy. A multidisciplinary approach is most helpful in the treatment of such children. Thus an ophthalmologist should be included in the initial assessment. This article aims to highlight the occurrence of various types of squint in children with cerebral palsy.

Aim of the study

1. To observe and estimate the prevalence of various types of strabismus in children with cerebral palsy.
2. To estimate the strength of association between the presence of squint and birth weight, and squint and gestational age of the child.
MATERIALS AND METHODS
This cross sectional, hospital based study was conducted in Dr SM CSI Medical College, Karakonam, Trivandrum. Eighty children diagnosed with cerebral palsy, who attended the Paediatrics outpatient department between 2012 and 2014 were included in the study.

INCLUSION CRITERIA
1. Children aged between one and fourteen years
2. Children already diagnosed with cerebral palsy

EXCLUSION CRITERIA
Children diagnosed to have other disorders/syndromes apart from cerebral palsy which are associated with ocular pathology.

Informed consent was granted by their parents prior to participation in the study.

A detailed history regarding gestational age, antenatal period, birth weight, postnatal period, parity of mother and the child’s treatment history was taken.

A detailed ophthalmic evaluation was done. Presenting visual acuity was assessed monocularly. Depending on the age and cooperation of the patient, a variety of methods were used to assess visual acuity: Snellen’s chart, Lea Symbol chart (colour plate 1) and Central Steady Maintained technique (CSM). CSM involves covering one eye while the child fixates on an object. The non covered eye should maintain central, steady fixation which is maintained through a blink. The presence of nystagmus and resistance to occlusion was recorded in patients whose visual acuity could not be measured.

This was followed by a complete ocular examination. Head posture, facial symmetry, extraocular movements, abnormal movements, Orthoptics assessment including cover-uncover test to detect presence of deviation and to measure angle of deviation for near and distance, Hirschberg test and prism bar cover test were done.

Refraction under cycloplegia was performed in all children. Cycloplegia was accomplished with one drop of 1% cyclopentolate instilled every five minutes for a total of three drops. Retinoscopy was performed after 30 minutes.

Detailed evaluation of the anterior segment was carried out with a standard slit lamp biomicroscope (Zeiss). If biomicroscopic evaluation was not possible due to poor level of cooperation then torch light examination of the anterior segment was done. The posterior segment evaluation in every patient was carried out in a fully dilated state of the eye with direct and indirect ophthalmoscope, whichever was possible. Post mydriatic test was done three days later and glasses prescribed for those with refractive error.

The data was stored on a computerised database and analysed using SPSS Computer software (version15.0). Chi square test, Fisher extract test & Mann Whitney U test were used in the statistical analysis and a p value of below 0.05 was considered as significant.

RESULTS
A total of 80 children were evaluated in our study, majority of whom (35%) were between 10-14 years. Mean age of patients was 8.8 years, ranging from 1 year to 14 years.

65% of the children examined were boys.

Strabismus was found in 32 of 80 (40%) cases enrolled in the study. Exotropia was found to be 3.4 times more than esotropia. Alternating Exotropia was the commonest type accounting for 17 cases (21.3%).

Table 1. Percentage distribution of the sample according to age

<table>
<thead>
<tr>
<th>Age (years)</th>
<th>Count</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;=5</td>
<td>19</td>
<td>23.8</td>
</tr>
<tr>
<td>5 – 10</td>
<td>26</td>
<td>32.5</td>
</tr>
<tr>
<td>&gt;10</td>
<td>35</td>
<td>43.8</td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>8.8 ± 4</td>
<td></td>
</tr>
</tbody>
</table>

Fig. 1. Percentage distribution according to age.
Table 2. Percentage distribution of the sample according to sex

<table>
<thead>
<tr>
<th>Sex</th>
<th>Count</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male</td>
<td>52</td>
<td>65.0</td>
</tr>
<tr>
<td>Female</td>
<td>28</td>
<td>35.0</td>
</tr>
</tbody>
</table>

There were 52 (65%) males and 28 (35%) females.

Table 4. Percentage distribution of the sample according to strabismus

<table>
<thead>
<tr>
<th>Strabismus</th>
<th>Count</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>48</td>
<td>60.0</td>
</tr>
<tr>
<td>Alternating Exotropia</td>
<td>17</td>
<td>21.3</td>
</tr>
<tr>
<td>Alternating Esotropia</td>
<td>3</td>
<td>3.8</td>
</tr>
<tr>
<td>Unilateral Exotropia</td>
<td>5</td>
<td>6.3</td>
</tr>
<tr>
<td>Unilateral Esotropia</td>
<td>4</td>
<td>5.0</td>
</tr>
<tr>
<td>Intermittent Exotropia</td>
<td>2</td>
<td>2.5</td>
</tr>
<tr>
<td>Hypertropia</td>
<td>1</td>
<td>1.3</td>
</tr>
</tbody>
</table>

BILATERAL EXOTROPIA

Table 5. Association between strabismus and birth weight

<table>
<thead>
<tr>
<th>Strabismus</th>
<th>Very low birth weight</th>
<th>Low birth weight</th>
<th>Normal</th>
<th>Z#</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Count</td>
<td>Percent</td>
<td>Count</td>
<td>Percent</td>
<td>Count</td>
</tr>
<tr>
<td></td>
<td>4.0</td>
<td>57.1</td>
<td>11</td>
<td>39.3</td>
<td>17</td>
</tr>
</tbody>
</table>

We attempted to find out the strength of association between presence of strabismus and the birth weight of the child.

Large p values from Mann Whitney U test show that there was no statistical association for birth weight with development of strabismus, although majority of the very low birth weight children did have some type of squint.

We also attempted to determine the association between strabismus and the gestational age of the child at birth.

However, no statistical association for gestational age with strabismus (p=0.407) was found.

DISCUSSION

Cerebral palsy describes a group of permanent disorders of the development of posture,
movement and tone causing activity limitation due to static encephalopathy acquired during brain growth in fetal life, infancy or early childhood. Though the brain disorders are unchanging the effects are dynamic as the brain matures and the child’s developmental capabilities extend [5].

The motor disorders are often accompanied by disturbances of sensation, perception, cognition, communication, behaviour, epilepsy and secondary musculoskeletal problems. Cerebral palsy refers to the first 2-5 years when there is active growth of the brain.

With an incidence of 2-2.5% per thousand live births, it is considered the most common cause of physical disability in childhood [6-11]. The most recent consensus definition emphasized the non motor features of cerebral palsy, listing “disturbance in sensation, cognition, communication, perception and or seizure disorder” that often accompanies the required motor impairment.[12]

Children with cerebral palsy are at risk of disturbance in multiple components of the visual system, including the primary visual pathway (eye, optic nerves, thalami, optic radiations, and primary visual cortices), visual association areas, and the oculomotor system [13, 14].

Prevalence of visual disturbances in cerebral palsy varies depending on the specific diagnoses included, but it has been estimated that between 5% and 9% of children with cerebral palsy have severe visual impairment [15-17].

At present 75-80% of causes of cerebral palsy point to antenatal factors responsible for abnormal development of the brain.

Perinatal and neonatal causes such as sepsis, neonatal seizures, cerebral ischemia and low Apgar scores are present in substantial number of children with cerebral palsy.

Main etiological factors in 10-20% of children with cerebral palsy are intra partum asphyxia and exposure to maternal infections such as chorioamnionitis, sepsis, urinary tract infections and fever with elevated levels of cytokines.

About 5-15% of new born infants with birth weight less than 1500gm manifest severe neurological abnormalities, mainly cerebral palsy, and 25-50% , more subtle cognitive, perception and behavioural defects [24].

Periventricular leucomalacia (PVL) is a brain lesion caused by episodes of hypoxia or ischemia at a gestational age of 24-34 weeks [25]. The lesion affects the corticospinal and or the geniculo-striate tracts, giving rise to spastic diplegia and or visual impairment [26-29].

The association of certain eye defects with cerebral palsy has been known since Little’s early papers on the disease in 1834 [1]. Both squint and refractive errors are usually common in these children [2]. They should be screened for these defects as early as possible. In practice the detailed examination and treatment given to a neurologically normal child is often denied to children with cerebral palsy [4].

Ophthalmological problems are reported with a range from 50% to 90% in cases with diagnosis of CP (37,46,47,50-52). Relation between frequency and severity of visual sensory problems and motor- mental deficits in children with CP are also reported [31, 37, 53].

In our study ocular abnormalities were found in 77.5% of the cases and this rate was found to be consistent with literature.

Ophthalmological problems were reported to be frequent in the spastic diplegic subtype of disease which is indicated as the most common type of CP in literature (37, 52-55). This was consistent with our study. On the other hand ocular problems were rarely reported in cases with dyskinetic type of CP [50].

Previous studies demonstrated CP related ophthalmological problems including strabismus and refractive errors with rates between 28% and 86%. The frequency of strabismus may be directly linked to the severity of visual motor system involvement [31, 37-39, 50-55]. The high incidence of strabismus is probably related to lesions in the subcortical oculomotor centres or cerebellar lesions which disrupt binocular vision [9]. Research demonstrates association between severity of physical and intellectual impairment in cerebral palsy and accommodative dysfunction [60]. The reduction of accommodation in such patients due to high prevalence of significant refractive error was
also found to be linked with the high frequency of squint [61].

**Table 6.** Association between strabismus and the gestational age of the child at birth.

<table>
<thead>
<tr>
<th>GA &lt;= 30 weeks</th>
<th>GA &gt;30 weeks</th>
<th>Z#</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Count</td>
<td>Percent</td>
<td>Count</td>
<td>Percent</td>
</tr>
<tr>
<td>Strabismus</td>
<td>15</td>
<td>46.9</td>
<td>17</td>
</tr>
</tbody>
</table>

In our study the overall incidence of strabismus was 40%, and the ratio of Exotropia to esotropia was found as 3.4:1. Strabismus was most frequently seen in diplegic and tetraplegic subtypes of CP.

Pigassou-Albouy had reported a 50% incidence of strabismus in a group of CP patients while incidence in general population is only 3% [56]. A study conducted by David Dufresne, Lynn Dagenais, Michael I. Shevell, on a total of 301 cases of cerebral palsy concluded that 49.8% had visual impairment. The majority of these individuals had strabismus (55.7%) and a slightly lesser fraction had refractive error (20.7%).

In a study done by Peter Black in 1982 involving 120 children with cerebral palsy the authors concluded that 20% had normal eyes; squint was found in 52.5% children and significant refractive errors in 50%, nystagmus (15.8%), ptosis (2.5%) and it was shown that spastic diplegia carries a high risk of refractive error and athetoid the least risk. There was a high incidence of strabismic and anisometric amblyopia (15%) [37].

A study conducted by P.D. Black in 1980 involving 117 children with cerebral palsy showed that only 20.5% of these children had normal eyes. Refractive errors accounted for nearly 50% of these abnormalities. Squint was found in more than 50% of the children. Sixteen out of the fifty nine squints were paralytic. Sixteen children had an amblyopic eye with a vision of 6/36 or less, associated with squint or anisometropia. Nystagmus was present in nineteen children (16%), half of which had ataxic cerebral palsy; cataract was present in 3 children [5].

Sabita Katoch, Anjana Devi, Prajanka Kulkarni evaluated a group of 200 cerebral palsy patient, it was found that 68% had significant visual morbidity. Seventy eight patients had strabismus. A study conducted by Jitendra Jethani at Aravind eye hospital, Madurai on children with PVL showed that thirty one (81.6%) had strabismus, predominant group being esotropia (57.9%), fourteen (36.8%) had nystagmus [39].

Research performed by A. Taylan Ozturk, A. Tulin Berk and Aylin Yaman involving a group of 194 patients diagnosed with cerebral palsy concluded that strabismus was found in one hundred and seven of cases (55.2%). Esotropia was found to be 2.5 times more than exotropia. Vertical deviation was seen in five cases, three of which had the diagnosis of spastic tetraplegia.

Both lower gestational age and low birth weight were found to be risk factors for the development of strabismus [40].

A study conducted by Sanjay Marasini, Nabin Paudel, Prakash Adhikari, Jyoti Baba Shrestha, Merrill D. Bowan involving thirty six children with cerebral palsy concluded that 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. Strabismus was the second most common finding and was present in 36% of the children. Fourteen percent of the children had a variant of nystagmus. Central cortical visual impairment was suspected in 11% [42].

Sasmal NK, Mandal R, Das D, Sarkar P, Biswas MC, Dey AK, Chatterjee S who studied one hundred and forty patients with cerebral palsy concluded that overall incidence of ocular abnormalities were 42.1%. The major ocular abnormalities included strabismus (36.4%), myopia (12.9%), hypermetropia (8.6%), astigmatism (3.6%), non glaucomatous optic atrophy (10.7%) and nystagmus (9.3%). Cortical visual impairment was seen in 20.7% children [44].

In a study conducted by I.A. Langunju, T.S. Oluleye involving one hundred and forty nine patients with cerebral palsy concluded that forty two had ocular abnormalities. More than
half (61.9%) were completely blind. The major ocular abnormalities identified in the affected cases were strabismus (50%), optic atrophy (50%), cortical visual impairment (47.7%), nystagmus (9.5%), and refractive errors (4.8%). Presence of spastic quadriplegia was associated with an increased risk of ocular abnormalities [45].

ASSOCIATION BETWEEN DIFFERENT VARIABLES

There was no statistical association for birth weight with strabismus and refractive errors in our study. Large p values from Mann Whitney U test indicate insignificant association (p = 0.522, 0.810 respectively).

According to the Mann Whitney U test there was no statistical association for gestational age with strabismus and refractive errors in our study (p = 0.83, 0.25 respectively). Although in other studies significant association was found for gestational age and birth weight with strabismus and refractive errors.

Study conducted by Taylan Ozturk et al reported that lower gestational age and birth weight were found as an aetiological factor for increasing risk of strabismus (p= 0.001, p = 0.003 respectively) , especially esotropia compared to Exotropia (p= 0.009, and p= 0.024) which is already mentioned in literature [31, 39, 58]. Jitendra Jethani reported that 81.6% of children in their study had strabismus, the predominant group being esotropia 22(57.9%) .The average birth weight was 2.17 kg. The mean gestational age was 31.56 months [39].

One of the limitations of our study is that it failed to demonstrate statistical association between variables due to a smaller sample size. Aetiological studies in order to find out the major predisposing factor for CP development may have a role in lowering the prevalence of the disease

Prematurity was found to be the leading aetiological factor for CP in 41.4% of our cases. The rate of prematurity as an aetiological factor for CP was published between 26% and 71% in the previous studies conducted in Turkey [46, 47]. From the history of our patients asphyxia was shown as second most common aetiological factor with a rate of 32.5%.

In developed countries spastic diplegic subtype of CP was found to be more frequent, in parallel with the increase in the survival of premature babies whereas in developing countries spastic tetraplegic type of CP was seen more frequently [48, 49]. In our study spastic diplegia was the most common subtype of CP with a rate of 67.5%. Spastic quadriplegia accounted for 25% of the cases.

Thus the literature tells us how important it is for children with cerebral palsy to have an ophthalmic evaluation at the earliest. It confirms that children with CP are at more risk of developing ocular abnormalities. Parents and 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 the child’s physical, social, academic and visual development. A complete eye examination should be sought as soon as diagnosis of CP is made and yearly thereafter.

CONCLUSION

1. Ocular abnormalities are common in children with cerebral palsy. In the present study ocular abnormalities were found in 77.5% of cases. We found a prevalence of 40% of for strabismus, which correlates with other published studies.

2. a) There was no statistical association for birth weight with refractive errors and strabismus in our study (p = 0.810, p = 0.522 respectively).

b) There was no statistical association for gestational age with refractive errors and strabismus in our study (p = 0.800, p = 0.407 respectively)

3. The most common aetiological factor associated with cerebral palsy was prematurity (37.5%) followed by birth asphyxia (32.5%) in our study.
The diagnosis of cerebral palsy is usually been made by the time a child reaches twelve months of age and rarely after eighteen months. It is at this time that the child should have his or her initial ophthalmological assessment. Thorough ocular assessment of cerebral palsied patients may be difficult, however familiar surroundings and caring environment, adequate clinical attention can facilitate assessment of all ocular disorders of these patients. Early referral of children diagnosed with CP for ocular examination is of utmost importance for better visual prognosis as many of the ocular pathologies are amenable to treatment.

REFERENCES
1. Little WJ. Treatment of flat foot or spurious valgus. Lancet 1834;44:679-84.


