

**OCULAR DISORDERS IN CHILDREN WITH DEVELOPMENTAL DELAY***Meera Suresh Joshi<sup>1</sup>, Omkar Jagdish Telang<sup>2</sup>, Vrunda Gangadhar Morepatil<sup>3</sup>*<sup>1</sup>Resident, Department of Ophthalmology, K. J. Somaiya Medical College and Hospital, Sion, Mumbai.<sup>2</sup>Associate Professor, Department of Ophthalmology, K. J. Somaiya Medical College and Hospital, Sion, Mumbai.<sup>3</sup>Lecturer, Department of Ophthalmology, K. J. Somaiya Medical College and Hospital, Sion, Mumbai.**ABSTRACT****BACKGROUND**

In India, an estimated 1.5-2.5% children below 2 years of age are developmentally delayed. A higher incidence of ocular disability is seen in these children, refractive errors and strabismus being most common. These can add to the overall burden of health as most of them have developmental comorbidities.

The aim of the study is to study the ocular disorders in children with developmental delay.

**MATERIALS AND METHODS**

We studied 112 children between the 2-12 years of age diagnosed to have developmental delay. All the subjects underwent a detailed ophthalmic evaluation including visual acuity testing using Snellen's charts (3m and 6m) and Log MAR charts (recorded as per Snellen's vision testing to maintain uniformity), cycloplegic refraction, torchlight and slit-lamp evaluation and dilated fundus examination. The data was tabulated and represented using bar diagrams, Pie charts and graphs. The results were expressed as percentages.

Design-Cross-sectional, observational study.

**RESULTS**

66 boys and 46 girls (total 112) were evaluated. The mean age of the study population was 7.8 years  $\pm$  2.4 SD. The aetiology of developmental delay was cerebral palsy (64%), Down syndrome (22%), autism (7%), intellectual disability (4.5%) and 1 case each of congenital hypothyroidism and ataxia telangiectasia. The prevalence of ocular disorders was found to be 84.8%, which was slightly higher in girls (87%) as compared to boys (83%). Refractive error (79.5%) was the commonest ocular disorder followed by strabismus (46.4%). Astigmatism (44.6%) was the commonest refractive error, which was divided into myopic astigmatism (19.6%), hyperopic astigmatism (13.8%) and mixed astigmatism (11.2%). Simple hyperopia was seen in 21.9% subjects and simple myopia in 12.1%. Exotropia (52%) was commoner than esotropia (48%). Other ocular abnormalities included optic atrophy, nystagmus, epicanthal folds, cataract, mongoloid slant, ptosis, telecanthus, conjunctival telangiectasia and blepharitis. Almost, 10% children with cerebral palsy had optic atrophy and 25% of those with Down syndrome had cataracts.

**CONCLUSION**

Ocular disorders are commonly seen in children with developmental delay. Refractive errors and strabismus are commonest and can easily be treated. Early diagnosis, prompt intervention and a close follow up are essential in order to prevent amblyopia.

**KEYWORDS**

Ocular Disorders, Developmental Delay, Refractive Errors, Strabismus, Cerebral Palsy, Down Syndrome.

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**BACKGROUND**

Vision plays a pivotal role in the overall development of a child. Uncorrected visual disturbances can hamper the acquisition of skills such as language interpreting facial

expressions and skills requiring hand-eye coordination. This in turn can affect the performance of the child at various levels, besides causing a socioeconomic burden upon the family.

Developmental delay is the condition in which a child is failing to meet expected developmental milestones in one or more of the following areas, viz. physical, social, emotional, intellectual, speech and language and/or adaptive development (sometimes called self-help skills, which include dressing, toileting, feeding, etc.). These delays are diagnosed when a child performs approximately 20-30% below age-related norms in one or more of these

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areas (with adjustment for prematurity in affected children).<sup>1</sup>

WHO estimated that 5% of the world's children 14 years and below have some type of moderate-to-severe disability.<sup>2,3</sup> In India, the prevalence of Developmental Delay (DD) is estimated to be 1.5-2.5% in children less than 2 years of age.<sup>2</sup>

An increased risk of having developmental delay is associated with having genetic syndromes, post-infection syndromes, cerebral palsy, seizures, CNS malformations, low birth weight and abnormal movements and tone.<sup>4</sup>

The incidence of ocular disorders is considerably high among children with developmental delay. This may be accounted for by several underlying causes of their disability with prenatal and perinatal factors and acquired injury, all being of relevance. Visual disorders are thus particularly higher in children born preterm having low birth weight, genetic/congenital anomalies and those who have suffered brain injury as seen in cerebral palsy.<sup>5</sup>

Refractive errors are the most commonly encountered ocular finding.<sup>6,7</sup> These tend to go unnoticed until an older age, primarily due to the lack of the child's ability to recognise poor vision. The more obvious disorders such as strabismus maybe picked up by a vigilante parent. However, reluctance to seek medical help leads to delayed and inadequate treatment. Prolonged deprivation of good vision causes incomplete development of the visual pathway. The plasticity of the visual system is lost with limited chances of recovery of sufficient vision.<sup>6</sup>

The following study aims to investigate the types of ocular disorders in children with developmental delay in a tertiary care hospital in a metropolitan city.

## MATERIALS AND METHODS

This was an observational study conducted over a period of one year at the Ophthalmology Department at K. J. Somaiya Hospital, Mumbai. Children between 2-12 years of age with developmental delay were included in this study. Children diagnosed to have neuroregression, previous history of ocular surgery and with family history of hereditary ocular disorders were excluded. The guardian of the child was explained the details of the study. A written informed consent was obtained from the guardian. A detailed antenatal, perinatal and postnatal history was obtained from the guardian. Any additional information regarding the child's exact systemic diagnosis was obtained from the treating paediatrician. IQ was obtained for 5 children who were diagnosed to have intellectual disability.

The sample size has been estimated using open Epi software based on prevalence of ocular disorders among children with learning disabilities in special education schools in Pune is 45.3% and absolute precision is 10% at 90% confidence level and 80% power of study, the estimated sample size is 65. In the present study, we have included 112 children.

All the above children underwent a detailed ophthalmic evaluation in the ophthalmology OPD-

1. Visual acuity was tested when possible by (a) Snellen's visual acuity charts and Lea symbol (LogMAR) charts (recorded as per Snellen's vision testing to maintain uniformity), (b) Ability of the child to fixate and follow light, (c) Whether the fixation was central, steady and maintained, (d) Visual axis, (e) Whether the child allowed occlusion of either eye.
2. Cycloplegic refraction was done under 1% atropine ointment (whenever not contraindicated) in children less than 7 years of age and in those with esotropia. In these children, the parents/guardians were instructed to instil atropine 1% ointment in the eyes of the children twice a day for 3 days and asked to follow up on the 4<sup>th</sup> day for retinoscopy. In older children, 1% cyclopentolate drops (whenever not contraindicated) were used for cycloplegia. Best corrected visual acuity was noted on their follow up visits whenever possible. In others, the results were noted based on objective retinoscopy, which was carried out at a distance of 1 metre. From the retinoscopic findings, 1 was deducted for working distance and an additional 1 or 0.75 for atropine or cyclopentolate respectively as per standard norms. Children who were diagnosed to have refractive errors were prescribed glasses, while in those who were previously wearing glasses, the spectacle prescription was confirmed or altered wherever required.
3. Ocular motility was evaluated in all 9 directions of gaze when possible. Ocular deviation was assessed by cover/uncover test and graded using Prism Bar Cover test/Krimsky test.
4. Ptosis evaluation was done if present and graded into mild, moderate and severe.
5. An external torch light evaluation was done followed by a slit-lamp evaluation before as well as after dilatation whenever possible.
6. Fundus examination was done by direct ophthalmoscopy and indirect ophthalmoscopy using 20 dioptre condensing lens under mydriasis.

## RESULTS

Of the 112 children, 66 were boys (58.9%) and 46 (41.1%) were girls and majority of children were in the age group of 6 to 9 (59.8%). The aetiology of developmental delay was cerebral palsy (64.3%), Down syndrome (22.3%), autism (7.1%), intellectual disability (4.5%) and one case each of congenital hypothyroidism and ataxia telangiectasia (0.9%). Prevalence of all ocular disorders in these children was 84.8% (95/112). 87% of girls and 83.3% of boys had ocular disorders. Various ocular findings were mainly refractive error (79.5%), strabismus (46.4%), followed by epicanthal folds (14.3%), nystagmus (12.5%), cataract (7.1%) and optic atrophy (6.3%). Also, seen were mongoloid slant (4.5%), ptosis (2.7%), telecanthus (2.7%) and on each of conjunctival telangiectasia and blepharitis.

178/224 (78.6%) eyes had refractive error. Simple hyperopia (21.9%) was more common than simple myopia (12.1%). Astigmatism was seen in 100 (44.6%) eyes. Myopic astigmatism (19.6%) was more common than

hyperopic astigmatism (13.8%) and mixed astigmatism (11.2%).

52 (46.4%) children had strabismus. Out of those, 27 (52%) had exotropia and 25 (48%) had esotropia. Incidence of exotropia (24.1%) was found to be higher than that of esotropia (22.3%).

51 of the 52 children with strabismus had some or the other refractive error. Astigmatism followed by hyperopia was more commonly associated with strabismus compared to myopia. In our study, 14 (12.5%) of the children had horizontal nystagmus.

Aetiology	Refractive Error	Strabismus	Epicanthal Fold	Nystagmus	Cataract	Optic Atrophy	Ptosis	Blepharitis
CP (72)	58	46	1	11	-	7	3	-
DS (25)	20	-	15	1	6	-	-	1
Autism (8)	5	3	-	-	2	-	-	-
ID (5)	4	2	-	1	-	-	-	-
AT (1)	1	-	-	1	-	-	-	-
CH (1)	1	-	-	-	-	-	-	-
<b>Total</b>	<b>89</b>	<b>51</b>	<b>16</b>	<b>14</b>	<b>8</b>	<b>7</b>	<b>3</b>	<b>1</b>

***Aetiology of Developmental Delay with Ocular Disorder***

CP= cerebral palsy, DS= Down syndrome, ID = intellectual disability, AT= ataxia telangiectasia, CH = congenital hypothyroidism.

61 out of 72 (84.7%) children with CP showed ocular findings (51 males, 21 females), 23 out of 25 (92%) children with DS showed ocular findings (6 males, 19 females), 5 out of 8 children (5 males, 3 females) with autism showed ocular findings, 4 out of 5 children (3 males, 2 females) with ID showed ocular findings-the IQ of all 5 children was in the range of 35-50 (moderate ID).

## DISCUSSION

Global developmental delay can be defined as significant delay in two or more developmental domains- gross and fine motor; speech and language; cognition; personal and social development; or activities of daily living.<sup>8</sup> Significant delay maybe defined as performance 2 or more standard deviations below the mean on developmental screening or assessment tests.<sup>9</sup>

The causes of developmental delay include genetic syndromes, post-infectious syndromes, cerebral palsy, seizures, CNS malformations, low birth weight, extreme prematurity, etc.<sup>4</sup>

More than 200 million children below 5 years of age in developing countries do not reach their developmental potential and thus are developmentally delayed.<sup>10</sup> As per the 2011 Census of India, visual disabilities (48.5%) have emerged as the top category among five types of disabilities on which data has been collected. The others in sequence are movement, mental, speech and hearing disabilities.<sup>11</sup> This shows that the magnitude of visual disturbances among developmentally delayed population is significantly high. Unfortunately, in many developing countries, this issue remains unaddressed. The present study is aimed at observing the various ocular disorders found in children with developmental delay.

We found the gender ratio of children with ocular disorders to be 1.22:1 (males:females). In a study by Wu H.J.<sup>12</sup> et al on 41 children with developmental delay, 68% were males and 32% females. Nielsen LS<sup>13</sup> et al found the gender ratio of males to females to be 1.66:1 in their study

conducted on developmentally delayed children in Denmark with 1.55:1 being the ratio of visually impaired children.

When we divided our subjects into 3 age groups namely those of preschool age (2-5 years), primary school age (6-9 years) and middle school age (10-12 years), we found majority of them to be in primary school age group (59.8%). The mean age was found to be 7.8 years with a standard deviation of  $\pm 2.4$ . In the study by Nielsen LS<sup>13</sup> et al, the mean age was  $10.1 \pm 3.19$  SD. In our study, the prevalence of ocular disorders was similar in all 3 age groups.

The aetiology of developmental delay in our study was cerebral palsy (64%), Down syndrome (22%), autism (7%), intellectual disability (mental retardation) (4.5%) and one case each of congenital hypothyroidism and ataxia telangiectasia.

In our study, 84.8% children showed presence of one or the other ocular disorder. The prevalence of ocular disturbances in our study is much higher than other studies. Gogate P<sup>6</sup> et al showed this incidence to be 45.3% in children with learning disabilities. Joshi R and Somani A<sup>7</sup> found ocular disorders in 51.45% of children with mental retardation. A study conducted by Reena A<sup>14</sup> et al in children with delayed milestones, 64.6% had ocular manifestations.

Refractive error, in our study was on the top of the list (79.5%) followed by strabismus (46.4%). Other significant findings in our study, besides refractive error were nystagmus, cataract, optic atrophy and ptosis.

In the 224 eyes that we studied, astigmatism was seen in 100 (44.6%) eyes. Simple hyperopia (21.9%) was more common than simple myopia (12.1%). Myopic astigmatism (19.6%) was more common than hyperopic astigmatism (13.8%) and mixed astigmatism (11.2%). The study conducted by Reena A<sup>14</sup> et al showed that refractive errors (41.3%) were the most common ophthalmic manifestation in children with delayed milestones of which 18% had significant hyperopia ( $>+3D$ ), 12% had myopia and 11% astigmatism. In a similar study by Nielson LS<sup>15</sup> et al, 15.3% had hyperopia  $>+3D$ , 10.8% were myopic and 20.6% had astigmatism. Joshi R and Somani A<sup>7</sup> found myopia (60%)

to be more common than hyperopia (20%) and astigmatism (20%) in children with mental retardation. Vora U<sup>16</sup> et al showed the prevalence of refractive errors to be 58.5% in children with special needs as compared to 2.9% in age-matched healthy subjects in Oman. They found that astigmatism (27.1%) was more common than myopia (24.3%) followed by hyperopia (18.6%). Gogate P<sup>6</sup> et al found the prevalence of myopia (10.5%) and hyperopia (10.6%) to be similar with astigmatism (5.7%) being much lower.

In our study, 52 (46.4%) children had strabismus. Out of those, 27 (52%) had exotropia and 25 (48%) had esotropia. Incidence of exotropia (24.1%) was found to be higher than that of esotropia (22.3%). 51 of the 52 children with strabismus had some or the other refractive error. Astigmatism followed by hyperopia was more commonly associated with strabismus compared to myopia. Nielsen LS<sup>15</sup> et al found strabismus in 26.8% of their subjects with esotropia in 14.9% and exotropia in 10.3%. Reena A<sup>14</sup> et al showed that the prevalence of strabismus was 40%. Joshi R and Somani A<sup>7</sup> found the presence of squint in 10.37% of their subjects. Gogate P<sup>6</sup> et al found the prevalence of strabismus to be 15.7% of which 54.2% had exotropia and 45.7% has esotropia, which is similar to our study. They found only 51.8% of those with strabismus to have an associated refractive error, out of which, myopia and hyperopia were more commonly associated with strabismus than astigmatism. In our study, however, we found almost 100% (51 of 52) of the children with strabismus had an associated refractive error with astigmatism being the most prevalent one.

Ocular abnormalities are common in children with Cerebral Palsy (CP) with an incidence of about 50-70%.<sup>17</sup> A study by Katoch S and Kulkarni P<sup>18</sup> reported 68% ocular morbidity in 200 children with CP. Black PD<sup>19</sup> reported in his study on 117 CP children, 79% with ocular findings and Douglas<sup>20</sup> found 58.4% CP children with abnormal ocular findings. In our study, 84% CP children had ocular abnormalities and although refractive errors (80.1%) and squint (63.9%) were the most common ocular findings in CP, 15% of these children had nystagmus and almost 10% of CP children had optic atrophy.

The pathology of spastic CP is extensive and diffuse cerebral cortex involvement with periventricular haemorrhage, subcortical haemorrhage and cortical atrophy, while in athetoid CP, lesions are mainly located in basal ganglia. This extensive CNS involvement explains the higher incidence of ocular defects in children with CP.<sup>18</sup>

Down syndrome is a risk factor of having visual impairment such as refractive errors, reduced accommodation, strabismus and early cataracts. The cause is believed to be poor maturation of the higher visual pathways.<sup>21</sup> Vora U<sup>16</sup> et al found that 80% with Down syndrome had refractive error. Akinci<sup>22</sup> et al noted that 97.4% of children with Down syndrome had ocular findings. This was in contrast to 42.4% without Down syndrome with ocular findings.

Da Cunha and Moreira<sup>23</sup> reported the occurrence of early cataract among children aged up to 17 years with Down syndrome to be from 5% up to 50%. In previous studies of congenital or infantile cataract, 3-5% of cases were associated with Down syndrome. Any insult (e.g., infectious, traumatic, metabolic) to the nuclear or lenticular fibres may result in an opacity of the clear lenticular media.

In our study, male:female ratio in Down syndrome was almost 1:3. Refractive error and epicanthal folds were the most common ocular findings in our study in children with Down syndrome. 80% of our children had refractive errors, which is similar to the study conducted by Vora<sup>16</sup> et al. Almost, 25% of them had cataracts. However, they were not visually significant and did not get in the way of clinical examination of posterior segment.

Ophthalmic findings reported in children with autism include abnormal electroretinograms, deficient evoked visual potentials, atypical optokinetic nystagmus, abnormal saccades and a higher incidence of strabismus than normal.<sup>24</sup> In our study, 8 children with developmental delay were diagnosed to have autism, 5 were boys and 3 were girls. Refractive error, squint and cataract were the chief ophthalmic findings noted.

Several studies have explored ocular and visual disorders in intellectually challenged individuals. Visual assessment is quite challenging in this group of children. The various disorders described in this setting include simple refractive errors, strabismus, reduced visual acuity, oculomotor abnormalities and field alterations. Optic atrophy, retinal dystrophies and structural eye anomalies too maybe associated.<sup>5</sup>

Banks<sup>25</sup> found in his study in the Netherlands, 72.3-92% with intellectual disability had ocular problems and the prevalence increased with age and severity of disability. 49% mentally handicapped children had some form of refractive error. Warburg found prevalence of myopia to be at 43% and of hyperopia at 21% in severe/profoundly intellectually impaired individuals.<sup>26</sup> Van den Broek<sup>27</sup> found refractive errors in 22% of people with severe and profound multiple disabilities.

In our study, 5 children were intellectually challenged, having moderate intellectual disability and 4 of them had ophthalmic findings such as refractive error, strabismus and nystagmus.

The higher prevalence of visual disorders in children with developmental disabilities is accounted for by the many underlying causes of their disability with prenatal and perinatal factors and acquired injury all of relevance. Hence, visual disorders are particularly increased in children born preterm, children who have suffered brain damage with subsequent learning difficulty and/or CP (cerebral palsy) and children with congenital cerebral anomalies or other genetic syndromes that may predispose to ocular anomalies.<sup>28</sup>

Gogate<sup>6</sup> et al found that 46.5% of children with learning disabilities having known perinatal insult had some ocular disorder. The distribution of ocular disorder varied with the type of perinatal insult suffered.

We had one patient of ataxia telangiectasia who was 7 years boy who showed telangiectatic blood vessels on conjunctiva besides nystagmus, exotropia, compound myopic astigmatism.

Specific ocular abnormalities are not described in congenital hypothyroidism in literature. However, our patient was an 11-year-old girl who had congenital hypothyroidism with global developmental delay and was found to have mixed astigmatism.

Despite the magnitude of the problem, affected children are underserved due to lack of awareness about their problems, not only among their parents and guardians, but also among the healthcare providers. The presence of more than one disability can have a multiplicative rather than additive effect on their life experience.

The paediatrician or general practitioner who is caring for an adolescent with an eye disorder during the transition to adulthood should be aware of social, psychological and professional impact of ocular disorders on the person. Ongoing ophthalmologic care is needed for these people and should be recommended in most cases. In addition, impact of the vision disorder on the choices that young adult can make should be considered. Vision screening programmes should be undertaken for early detection and care of eye problems in these children, especially because most of these children do not complain on their own and these disorders can be completely unrecognised and neglected.

## CONCLUSION

Children with special needs have other disabilities that are prioritised and the need for eye care often remains unnoticed or neglected. Refractive errors, if left untreated, results in visual disability adding to compromised quality of life, which is quite avoidable. Refractive error correction at an early age prevents amblyopia and thus prevents the child from spending productive years of life with avoidable blindness. Eye healthcare providers should educate parents and school teachers about the need for early eye screening. It is important that in developing countries national programs should at least focus on the high-risk groups for visual impairment and disabilities.

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