

COMMON FEATURES ASSOCIATED WITH CONGENITAL ESOTROPIA

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ABSTRACT

OBJECTIVES: The objectives of this study were to assess the clinical features associated with congenital esotropia and to study the management regime of features associated with congenital esotropia.

METHODS: A descriptive cross-sectional study was carried out analyzing data from 61 patients, examining patients of Congenital Esotropia up to 2 years of age, from August to November 2018 in College of Ophthalmology and Allied Vision Sciences, Lahore. This study mainly included the study of associated features of Congenital Esotropia like dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, cross fixation and refractive errors. Prior approval was sought from ethical review board of College of Ophthalmology and Allied Vision Sciences to conduct this study.

RESULTS: The study included 31 male patients and 30 female patients. 62.3% of patients were using spectacles before the study. 18% participants were having a birth history of the full term with C-section and 82% patients were having birth History of the full term with normal delivery. This study of associated features of Congenital Esotropia showed that 23% of patients were having Dissociated Vertical Deviation (DVD), 27.9% patients were having Manifest Latent Nystagmus (MLN), 65.6% patients were having Inferior Oblique over Action (IOOA), 88.5% patients were having Cross Fixation, and 78.7% patients were having abduction Deficit. The refractive status showed that 8.2% patients were having Myopia, 52.5% were hyperopic, 9.8% were Astigmatic while refractive error was absent in 29.5% (18) patients. Hyperopia was more common in male patients having Esotropia than female patients. Likewise, DVD, MLN, IOOA, cross fixation and abduction deficit were more in patients of Congenital Esotropia having Hyperopia. DVD was maximum among patients having 2 years of age and it occurred in 5(8.20%) patient of CET ($p = 0.000$). Manifest latent nystagmus was maximum among patient having 1 year of age and it occurred in 6(9.84%) of patients ($p = 0.001$). IOOA was maximum among patients having 2 years of age and it occurred in 19(31.15%) of the patients ($p = 0.015$). Cross fixation was maximum among patients having 2 years of age and it occurred in 29(47.54%) of patients ($p = 0.000$). Abduction deficit was maximum among patients having 2 years of age and it occurred in 24(39.34%) of patients ($p = 0.000$). Hyperopia was maximum in 19(31.15%) patients of CET ($p = 0.000$).

CONCLUSION: Clinical features like Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, cross fixation and refractive errors are associated with Infantile Esotropia.

KEYWORDS: Binocular single vision, Dissociated Vertical Deviation, Latent Nystagmus, Congenital esotropia, Inferior Oblique Over action,

INTRODUCTION

Squint is an ocular misalignment that is due to abnormal neuromuscular control of ocular motility during the binocular single vision. Squint is an irregular alignment of the visual axis of the eyes. Squint affects 1-3% of children throughout the world. Strabismus occurs in children with a history of premature birth, systemic diseases, cerebral palsy, genetic syndromes (i.e. Down syndrome) and family history of squint.¹ The classification of Strabismus is based on the direction of the deviation (horizontal, vertical, torsional, frequency,

the age of onset and deviation in different fields of gaze (comitant or incomitant).² Congenital Esotropia is also called "Infantile Esotropia" and is defined as alternating, cross fixational esotropia which develops within first six months of life and the characteristic finding of congenital esotropia (CET) mostly develops within the first three years of childhood.³ CET is a large, constant, and stable angle inward deviation that begins within the first 6 months of infancy. Esotropia is also associated with albinism.⁴

Congenital Esotropia (CE) is one of the most common forms of squint in children, with a prevalence of births of 25 per 10,000 newborns. The characteristics associated with CET includes alternating esotropia, cross-fixation, latent-manifest nystagmus, over-action of the inferior oblique muscles, dissociated vertical deviation (DVD) and reduced binocular vision.⁵ In CET, it is determined that achieving a certain degree of binocular function is related to the early alignment of the eyes. In addition, close follow-ups and the precise treatment of the accompanying vertical deviation, especially the IOOA and DVD can improve in a timely manner heads to the binocular sensory function.⁶

A study was conducted in 2016 where the prevalence of infantile esotropia at birth was thoroughly investigated. It was reported to vary from one in 403 live births to one in 50 of all newborns. The associated conditions like DVD and IOOA tend to develop more frequently after the age of two years and reported to occur as high as 57% and 78% of the time respectively. The study also found the higher frequency of nystagmus with a latent type.

Another study was conducted where 165 patients with congenital esotropia were examined from April 1991 to September 2001. The study found the amblyopia in 48.4% of patients, DVD in 12.7 %, IOOA in 11.3%, LN in 6.0% and the average refractive errors in 1.18% (ranging from spherical -7D to spherical +6D).⁶

Esotropia is the most common form of heterotropia. In a chain of 558 successive cases of esotropia, if about 4 out of 5 patients of heterotropia who access the orthoptist ensure an inward deviation, the treatment of this specific object is particularly significant. The therapy of inward deviation by non-surgical and surgical means is far away from precise.⁷ It is suspected that the damage or deterioration of the posterior fossa is a possible causal factor in the development of infantile esotropia with nystagmus in the abduction.⁸ Infantile esotropia is considered a family disease where relatives who do not meet all the criteria for diagnosis still show some of its characteristics. Optokinetic Nystagmus (OKN) asymmetry, reduction of stereopsis and nystagmus in abduction are subclinical signs of the condition. It is emphasized that the importance of a positive family history of strabismus is important to understand the genetic factors responsible for the transmission of infantile esotropia.⁹

The clinical findings associated with Infantile Esotropia include refractive errors, amblyopia, Dissociated Vertical Deviation (DVD), Inferior Oblique Muscle Over-Action (IOOA), and nystagmus. Due to a similar feature, it needs to be distinguished from the Duane retraction syndrome, Moebius syndrome, nystagmus block syndrome and early onset accommodative esotropia, as well as other causes of esotropia in childhood.¹⁰

DVD is a disorder of ocular motility with unexplained etiology and occurs in 75% of patients with infantile esotropia. A study explained that the DVD can appear in up to 90% of cases of infantile esotropia.⁹ DVD is an upward drifting of one eye when the other is fixating on a target and it is a bilateral condition with a distinct asymmetry.¹¹ It is a common hyper deviation in most of the congenital esotropia and it develops in individuals with normal binocular single vision and exotropia.. Clinical characteristics of DVD include elevation, extorsion, and abduction of non-fixating and with fixating eye incyclotorting incomitance. DVD is also associated with a compensatory head tilt. The occurrence of related amount of abnormal posture of the head in patients with DVD may be enhanced with squint surgery.¹²⁻¹³

The surgery performed prior to two years of age leads to greater potential to achieve Binocular Single Vision (BSV) and it is also associated with the lack of development of DVD, Latent Nystagmus (LN) and Symmetric Optokinetic Nystagmus (OKN).¹⁴ Due to the interruption in binocularity produced by the early onset of strabismus, it inhibits the normal development of cortical inputs to the optokinetic pathway of the brain stem which produces an asymmetry in monocular optokinetic responses.¹⁵ Infantile esotropia with jerk nystagmus in abduction and DVD are characterized by the early onset.⁸

The asymmetry of the pursuit and optokinetic system is evident in the latent nystagmus that reflects a tonic dominance, directed nasally with reference to fixation eye. The defect lies between the cortex and the brainstem. The gaze-evoked component is a purposeful reaction that diminishes the nystagmus in adduction at the expense of an increase in abduction.¹⁶ Children affected by congenital esotropia can cross-fixate to see an object on the contra lateral side with the adducted eye; in doing so, they do not need to abduct the eye ipsilateral to the object and, therefore abduction

defects may appear. Less frequently, an esotropic child is unable to abduct and cross-fixate to allow side gaze.¹⁷

Patients with infantile esotropia and manifest latent nystagmus take a turn of the face towards the fixating eye due to nystagmus dampening mechanism. In this case, the purpose of the surgical plan is to correct both face turn and strabismus.¹⁸

Abduction deficit gets improved after the surgical procedure.¹⁹ According to the results of a study, the abduction deficit can be considered as a risk factor in patients with congenital esotropia who have undergone a strabismus surgery at the age of more than 2 years.¹⁹

MATERIALS AND METHODS

It was a descriptive Cross-sectional study and held in College of Ophthalmology and Allied Vision Sciences, King Edward Medical University/Mayo Hospital Lahore from September to November 2018. Inclusion criteria were Children with congenital esotropia and Children below 2 years of age. Exclusion criteria were acquired esotropia/squint other than esotropia, any ocular pathology, Pseudostrabismus, Duane's retraction Syndrome, Down syndrome, Cerebral palsy and 6th nerve palsy. Prior approval was sought from ethical review board of College of Ophthalmology and Allied Vision Sciences to conduct this study.

Data has been collected through a questionnaire which is constituted of information about congenital esotropia and features associated with it like Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, and Refractive error. Data has been analyzed by SPSS software. Qualitative data were measured by applying the Chi-square test and quantitative data were measured by taking mean & standard deviation (\pm SD). The quantitative variables were presented as frequency as a percentage and for other variable suitable statistical techniques applied.

RESULTS

This study included a total number of 61 patients with Congenital Esotropia. The study was constituted of 31 male patients and 30 female patients. 62.3% of patients were using spectacles before the study. Like 18% participants were having a birth history of the full term with C-section and 82% patients were having birth History of the full term with normal delivery.

This study of associated features (table 1) of Congenital Esotropia showed that 23% of patients were having Dissociated Vertical Deviation (DVD). Likewise, 27.9% patients were having Manifest Latent Nystagmus (MLN). In addition, 65.6% patients were having Inferior Oblique Over action (IOOA). Likewise, 88.5% patients were having Cross Fixation. Furthermore, 78.7% patients were having abduction Deficit. The refractive status was; 8.2% patients were having Myopia, 52.5% were hyperopic, 9.8% were Astigmatic and refractive error was absent in 29.5% (18) patients. Hyperopia was more common in male patients having Esotropia than female patients. Likewise, DVD, MLN, IOOA, cross fixation and abduction deficit are more in patients of Congenital Esotropia having Hyperopia. DVD was maximum among patients having 2 years of age and it occurred in 5(8.20%) patient of CET. Manifest latent nystagmus was maximum among patients having 1 year of age and it occurred in 6(9.84%) of patients. IOOA was maximum among patients having 2 years of age and it occurred in 19(31.15%) of the patients. Cross fixation was maximum among patients having 2 years of age and it occurred in 29(47.54%) of patients. Abduction deficit was maximum among patients having 2 years of age and it occurred in 24(39.34%) of patients. Hyperopia was maximum and it occurred in 19(31.15%) patients of CET. P-value is less than 0.05 of above all features it means these clinical features like Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, cross fixation and refractive errors are associated with Infantile Esotropia.

Table 1:

Dissociated Vertical Deviation		
	Frequency	Percent
YES	14	23.0
NO	47	77.0
Total	61	100.0

Table 2:

Type of Refractive Errors		
	Frequency	Percent
Myopia	5	8.2
Hyperopia	32	52.5
Astigmatism	6	9.8
No refractive error	18	29.5
Total	61	100.0

Table 2 shows that most of the subjects were having hyperopia (n=32, 52.5%). p=0.000

Table 3: Chi-Square test Results

	Dissociated Vertical Deviation	Manifest Latent Nystagmus	Inferior Oblique Over action	Cross Fixation	Abduction Deficit	Type of Refractive Errors
Chi-Square	17.852	11.951	5.918	36.213	20.082	31.393
Df	1	1	1	1	1	3
Asymp. Sig.	.000	.001	.001	.000	.000	.000

P-value is less than 0.05 of above all features it means these clinical features like Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, cross fixation and refractive errors have a statistically significant association with Infantile Esotropia.

Figure 1: Comparison of Age with Manifest Latent Nystagmus

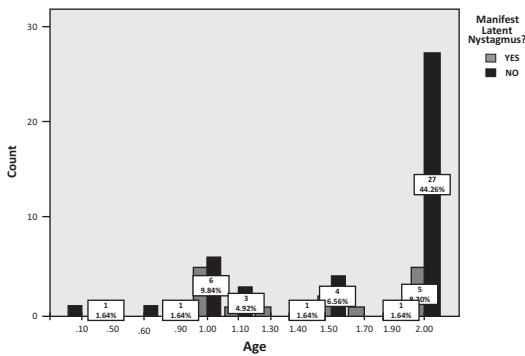


Figure 1 shows the comparison between Age of patient with Manifest Latent Nystagmus in CET. Manifest latent nystagmus was maximum among patient having 1 year of age and it occurred in 6(9.84%) of patients. The p value with chi square test was 0.001

Figure 2: Comparison of Age with Manifest Latent Nystagmus

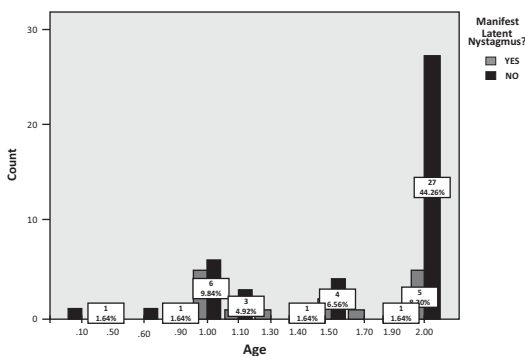


Figure 2 shows comparison between Age of patient with Manifest Latent Nystagmus in CET. Manifest latent nystagmus was maximum among patient having 1 year of age and it occurred in 6(9.84%) of patients. The p value with chi square test was 0.001

DISCUSSION

Squint is a disorder where the eyes do not line up in the same direction while focusing. The condition is more commonly known as crossed eye.

There are many types of esotropia, Congenital esotropia occurs in the first 6 months of infancy. This study has been emphasized on the associated features of congenital esotropia and their prevalence among patients of CET. This study has been done in Orthoptics clinic of Institute of Ophthalmology Mayo Hospital Lahore; Pakistan. This study has helped us to accurately measure the association of different associated factors of CET. If Inferior Oblique over action is treated then Latent Manifest Nystagmus and DVD are avoided.

This study of associated features of congenital Esotropia showed that 23% of patients were having Dissociated vertical deviation. Likewise, 27.9% patients were having Manifest Latent Nystagmus. In addition, 65.6% patients were having Inferior Oblique Overaction. Likewise, 88.5% patients were having Cross Fixation. Furthermore, 78.7% patients were having Abduction Deficit and 8.2% patients were Myopic, 52.5% were hyperopic, 9.8% were Astigmatic and refractive error was absent in 18/61 (29.5%) patients. This study revealed that Cross fixation, Inferior Oblique Overaction and Abduction deficit were most among patients of CET. Among refractive errors, Hyperopia is more in patients with CET. CET start in first 6 months of infancy.

Though in North America there is a tendency to perform surgery for congenital esotropia in general prior to 1 year of age (early on surgery), in Europe it is postponed more commonly until 1 to 3 years (standard surgery). In practice, children are not operated before 1 year owing to the improved frequency of standard reoperation for early on surgery. Accordingly our data, assessment of inferior oblique hyper function is hardly precise in infants. In addition, the treatment of potentially partial amblyopia, the unsteadiness of the angle and the late maturation of the sensor motor system, and the opportunity of at least partly unplanned declaration

justify our option.⁵

From this study, it is concluded that Hyperopia is more in males having Esotropia. Likewise, DVD, MLN, IOOA, cross fixation and Abduction deficit are more in patients of CET having Hyperopia. p-value was less than 0.05 of all features which means that these clinical features like Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, cross fixation and refractive errors are associated with Infantile Esotropia

CONCLUSION

Strabismus not only cosmetically affects the aesthetic perspective of a child but also produces a negative effect on visual acuity. Untreated strabismus causes amblyopia in children. The early diagnosis of patients with CET and the regular monitoring of patients help the pediatric ophthalmologist to perform the surgery as soon as possible to align the visual axis. It is important because these patients are going through their plastic period and their vision develops up to 8 years. The analysis of the data was done through the use of SPSS. The configuration level was set at 5%. The data was summarized on the nature of the data. The descriptive data were analyzed by percentages and frequencies and summarized in tables and pie charts. All the preliminary analysis was done to verify that the data comply with the assumptions. Hyperopia was more in male having Esotropia. Likewise, DVD, MLN, IOOA, cross fixation and Abduction deficit were more in patients of CET having Hyperopia. p-value is less than 0.05 of all features which means these clinical features like Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, cross fixation and refractive errors are associated with Infantile Esotropia.

RECOMMENDATIONS

Demographic factors, such as gender and family strabismus history, are associated with a higher risk of occurrence. On the other hand, some clinical features, including Dissociated vertical deviation, Inferior oblique over action, Latent manifest nystagmus, Abduction deficit, and Refractive errors will be manifest in patients of CET. Surgery must be planned between 12 and 24 months to achieve an optimal alignment. After studying this it, is recommended that, early diagnosis of congenital esotropia is very important. Strictly regular follow-ups and need for surgery must be ruled out early to align

visual axis, so the binocular single vision develops in infants.

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