

ANALYTICAL STUDY OF ESSENTIAL INFANTILE ESOTROPIA AND ITS MANAGEMENT*Kandasamy Sivakumar¹, Periasamy Geetha²*

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ABSTRACT**BACKGROUND**

Essential Infantile Esotropia (EIE) is the most common type of strabismus. About 0.1% of the newborn are found to have esotropia.¹ Though present since birth, it becomes manifest and remains constant around six months of age. The features are large angle constant strabismus, defective Binocular Single Vision (BSV), cross fixation, DVD and latent nystagmus. Most of the patients have mild-to-moderate hyperopia; the amount of deviation is unrelated to the amount and type of refractive error.

MATERIALS AND METHODS

Fifty cases with EIE were included in this prospective study. A thorough ophthalmic and orthoptic evaluation was done in all the patients. For patients more than three years of age, the angle of deviation was measured with prism bar cover test, and for patients less than three years of age, angle of deviation was measured with Hirschberg's test. Associated features like cross fixation, abduction limitation, Dissociated Vertical Deviation (DVD), nystagmus, amblyopia and Inferior Oblique Overaction (IOOA) were documented. Occlusion therapy was given to amblyopic patients prior to surgery. All these patients underwent surgery and were followed up for a period of six months.

RESULTS

The prevalence of EIE in our centre was 0.33%. Of the fifty patients, 28 were males and 22 were female patients. 39 patients (78%) had deviation of 30-50 Prism Dioptres (PD). Incidence of DVD, inferior oblique overaction and nystagmus was found to be lower when compared to western population. Amblyopia should be diagnosed early and treated adequately before surgery. Standard surgical option is bimedial recession. Monocular recession-resection surgery in one eye can be opted for in cases of irreversible amblyopia. Three or four muscle surgery can be done if deviation is very large. If marked inferior oblique overaction is present, the same should be weakened in addition to the horizontal muscle surgery.

CONCLUSION

EIE is the most common type of strabismus. These patients have large and constant angle of deviation. Most of them have mild-to-moderate hyperopia. Amblyopia should be diagnosed early and treated promptly before surgery. Patients show limitation of abduction, cross fixation, dissociated vertical deviation, inferior oblique overaction and nystagmus. Early surgery within two years gives best visual outcome in the form of binocular vision and stereopsis.

KEYWORDS

Essential Infantile Esotropia, Nystagmus, Amblyopia, DVD, IOOA.

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BACKGROUND

Most children exhibit apparent defective abduction and excessive adduction or both. Cross fixation being a characteristic features, the right eye is used to fix the object in the left visual field and the left eye is used to fix the objects in the right visual fields. Amblyopia is a common associated feature in Essential Infantile Esotropia (EIE).

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Overaction of inferior oblique with V-phenomenon and Dissociated Vertical Deviation (DVD) are common components of EIE. In some cases, anomalous head posture associated with latent or manifest nystagmus is seen. The patient turns his head towards fixating eye to gain maximum visual acuity. The treatment of choice for Essential Infantile Esotropia (EIE) is surgical alignment of the eye. Strabismus surgery success depends upon restoration of binocular single vision, normal visual acuity, normal retinal correspondence and stable sensory and motor alignment. Early ocular alignment gives the best chance of the development of some degree of binocular function. Ideally, the eyes should be surgically aligned by the age of 12 months and at the very latest by the age of 2 years, but only after amblyopia and any significant refractory error have been corrected. The initial procedure can be either recession



of both medial recti or unilateral medial rectus recession with lateral rectus resection. Associated significant inferior oblique overaction should also be addressed.

The aim of the study was to find the prevalence, the presence of associated clinical features and to assess the functional and cosmetic results following the appropriate surgical procedures in EIE.

MATERIALS AND METHODS

In this prospective study, fifty cases of Essential Infantile Esotropia (EIE) were analysed in our strabismus and Paediatric Ophthalmology Clinic of Regional Institute of Ophthalmology and Government Ophthalmic Hospital, Chennai, over a period of two years.

Children with esotropia in the age group of six months to ten years, preoperative deviation more than 30 PD, and availability of six months follow up after surgery were included in our study. Patients with previous history of ocular surgery, systemic abnormalities and other congenital abnormalities were excluded from the study. Thorough ophthalmic and orthoptic evaluation were done in all patients. Visual acuity was determined in all patients. In infants and children less than three years of age, spontaneous fixation and following of light was used as an assessment of good vision. In preschool children, Allen cards were used to assess vision and for school going children, Snellen’s chart was used to assess visual acuity. All the children were subjected to cycloplegic refraction with atropine to assess the refractive status.

Angle of deviation was measured with prism bar cover test for patients older than three years of age. For patients less than three years, Hirschberg’s test was done to measure the angle of deviation. Associated features like cross fixation, abduction limitation, Dissociated Vertical Deviation (DVD), nystagmus, amblyopia and inferior oblique overaction were documented. Forced duction test was done to rule out restrictive pathology. Occlusion therapy was given for patients with amblyopia for three months prior to surgery.²

RESULTS

50 children in the age group of six month to ten years were operated upon. 12 of these patients were less than two years, 23 were between two to six years and 15 patients were between seven to ten years of age. Of the fifty patients, 28 were male patients and 22 were females. A positive family history of squint was present in four patients. History of second and third-degree consanguinity was present in 15 patients. 26 patients were emmetropic or with mild hyperopia of less than two dioptres. 20 patients had moderate hyperopia (+2 to +5D) and two patients had high hyperopia (>+5D). Two patients were found to be myopic.

Amblyopia is defined as the difference in visual acuity of two lines between the two eyes of an individual. In our study, amblyopia was present in 17 children. Nine children who were amblyopic and less than 10 years of age underwent occlusion therapy for three months prior to surgery. Of these, five of them showed improvement of vision in the eye with amblyopia. Associated features like

DVD was seen in two patients, nystagmus was seen in six patients, restricted abduction in 40 patients, cross fixation in 11 patients, inferior oblique overaction in six patients and AV phenomenon in six patients.

39 patients had 30-50 PD deviation and 11 patients had more than 50 PD deviation. Alternate convergence squint with free alteration was seen in 36 patients and remaining 14 had dominance of one eye. Standard surgical procedure was bimedial recession. Monocular recession-resection was opted in cases of irreversible amblyopia in one eye. Three or four muscle surgery was done in deviation of more than 50 PD. 22 patients underwent bimedial recession, 13 underwent monocular recession-resection surgery, three underwent three muscle surgery, 10 underwent four muscle surgery and two underwent inferior oblique recession along with horizontal muscle surgery.

Patients were assessed on the first postoperative day, fortnightly for a month and at the end of three and sixth month. 34 patients were found to be orthophoric, 15 patients had residual esotropia of more than 10 PD and one patient had consecutive exotropia. Six patients who had large residual angle of deviation underwent second surgery. Subsequently, good cosmetic alignment and functional improvement were obtained.

Visual improvement was tested with Allen visual acuity cards. 28 patients showed improvement after the first month of surgery and 12 children showed improvement with three months of surgery. Six patients required second surgery.

Binocular single vision was tested in all children who cooperated for the test. 12 patients showed improvement in BSV of which eight children showed Grade 1 BSV, three showed Grade 1 and Grade 2 BSV and one child developed stereopsis.

Types of Refractive Errors	No. of Patients	%
Emmetropic or with mild hyperopia (<+2D)	26	52
Moderate hyperopia (+2 to+ 5D)	20	40
Severe hyperopia (>+ 5D)	2	4
Myopic	2	4

Table 1. Types of Refractive Errors

Extent of Deviation	No. of Patients	%
30-50 PD	39	78
>50 PD	11	22

Table 2. Extent of Deviation

Types of Surgery	No. of Patients	%
Bimedial recession	22	44
Monocular recession and resection	13	26
Three muscle surgery	3	6
Four muscle surgery	10	20
Inferior oblique recession along with horizontal muscle surgery	2	4

Table 3. Types of Surgery

Associated Features	No. of Patients	%
Amblyopia	17	34
DVD	2	4
Nystagmus	6	12
Restricted abduction	40	80
Cross fixation	11	22
Inferior oblique overaction	6	12
AV phenomenon	6	12

Table 4. Associated Features

Surgical Outcome	No. of Patients	%
Orthophoric	34	68
Residual esotropia	15	30
Consecutive exotropia	1	2

Table 5. Treatment Results

Binocular Single Vision (BSV)	No. of Patients	%
Grade 1 BSV	8	16
Grade 1-2 BSV	3	6
Stereopsis	1	2

Table 6. Binocular Single Vision

DISCUSSION

Essential infantile esotropia is defined as a manifest deviation with an onset from birth to six months of age. Prevalence was once estimated to be 1% of the population; Helveston and Costenbader have estimated this to be closer to 0.1%.¹ EIE continues to be the most common form of strabismus.

Little is known about the origin of EIE. Current concepts find their roots in two principal theories. The first theory proposed by Worth states that squint is a defect of fusion faculty putting the eye into an unstable equilibrium to deviate on a slight provocation. Chavasse proposed that peripheral and central factors interfere with the development of binocular reflexes and leading to strabismus.³ The two opposite views have therapeutic implications. If esotropia is due to defect in fusional faculty, no treatment is needed. Normalcy will be restored during development. If on the other hand, no such defects exist, a cure should be accomplished by aligning the eyes as early turned towards the fixating eye is termed as Ciancia syndrome. Association of EIE with latent nystagmus and Dissociated Vertical Deviation (DVD) is termed as Lange syndrome.⁸

The condition that mimics EIE are bilateral sixth nerve palsy, accommodative esotropia, nystagmus blockade syndrome, sensory esotropia and the esotropia associated with Duane’s, cerebral palsy and mental retardation.

The treatment of choice for Essential Infantile Esotropia (EIE) is surgical alignment of the eye. Nonsurgical treatment is directed towards correction of significant refractive error, elimination of amblyopia during preoperative phase and treatment of residual esotropia. Success of strabismus surgery is defined as restoration of binocular single vision, normal visual acuity, normal retinal correspondence and stable sensory and motor alignment. It has been universally accepted that ample restoration of normal binocular single vision with normal restoration of stereopsis is difficult. The

as possible. It is Chavasse’s thinking that triggered the era of early surgery that began with Costenbader.⁴

Essential Infantile Esotropia (EIE) is usually a large angle constant esotropia that measures 30-70 PD. Most of the patients have mild-to-moderate hyperopia; the amount of deviation is unrelated to the amount and type of refractive error. Most children exhibit apparent defective abduction and excessive adduction or both. If amblyopia is present, defective abduction is more prominent in the amblyopic eye. Cross fixation being a characteristic feature, the right eye is used to fix the object in the left visual field and the left eye is used to fix objects in the right visual fields.

Amblyopia is a common associated feature in EIE. It was found in 49% of the 500 patients examined by Costenbader. Amblyopia has to be cured at an early stage because it is an obstacle for the development of binocular single vision.² Overaction of inferior oblique with V-phenomenon and dissociated vertical deviation are common components of EIE. Von Noorden and co-workers reported inferior oblique overaction in 68% of 408 patients. Inferior oblique overaction in infants is elicited with difficulty. DVD is infrequently present before the age of two years.⁵ Incidence of DVD as reported by Von Noorden was 51%. Ciancia observed that latent nystagmus increased during abduction and decreased during adduction in 33% of patients with EIE.⁶ Von Noorden and co-workers reported latent nystagmus in 25% of their patients and they proposed a disturbance in coordination between the vestibular and the optic centre of the oculomotor system.⁷

Asymmetry of optokinetic nystagmus is seen in normal infants upto 3-4 months of age and symmetry develops between 4-6 months of age. Hence, Denver and Von Noorden stated that, if asymmetry is present, there is a high likelihood that strabismus has developed during in the first six months of age. In some cases, anomalous head posture associated with latent or manifest nystagmus is seen. The patient turns his head towards the fixating eye to gain maximum visual acuity. Association of Essential Infantile Esotropia (EIE) with horizontal jerky nystagmus and face

best that can be achieved is normal fixation with peripheral fusion when operated early. The advantage of early surgery before two years of age have greater potential for BSV.^{9,10} The disadvantages are problems with assessment and obtaining reliable measurements, changing anatomical landmarks and difficulty in amblyopia management after surgery.

The advantage of doing surgery after two years of age are reliable measurement and assessment of oblique overaction and Dissociated Vertical Deviation (DVD), improved co-operation for amblyopic treatment and anatomical relationship approaching adult dimensions. The disadvantages are reduced potential for binocular single vision and increased mechanical component of muscle and connective tissue contracture.

The initial procedure can be either recession of both medial recti or unilateral medial rectus recession with lateral rectus resection along with correction of inferior oblique

overaction. Under correction, may require further recession of the medial recti, resection of one or both lateral recti or surgery to the other eye depending on the initial procedure. Inferior oblique overaction may develop subsequently, most commonly at age 2 years. The parents should therefore be warned that further surgery may be necessary despite an initially good result. Initially unilateral, it frequently becomes bilateral within 6 months. Inferior oblique weakening procedures include disinsertion, recession and myectomy. DVD is characterised by updrift with excyclorotation of the eye when undercover or spontaneously during periods of visual inattention. When the cover is removed, the affected eye will move down without a corresponding downdrift of the other eye. It is usually bilateral. Surgical treatment maybe indicated for cosmesis; options include superior rectus recession with or without posterior fixation sutures and inferior oblique anterior transposition. Amblyopia subsequently develops in about 50% of cases as unilateral fixation preference commonly develops postoperatively. An accommodative element should be suspected if the eyes are initially straight or almost straight after surgery and then start to reconverge. Regular refraction is therefore important.

CONCLUSION

Children with EIE form the majority of esotropia. Children upto eight years of age, respond well to treatment of amblyopia than older children. Early diagnosis and appropriate surgical management will render good binocular single vision and stereopsis.

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