A DEFINITIVE SURGICAL CORRECTION TO RARE UNCORRECTED INFANTILE ESOTROPIA WITH ANOMALOUS MEDIAL RECTUS INSERTION
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ABSTRACT

INTRODUCTION
Infantile esotropia is an idiopathic esotropia previously called as congenital esotropia where there is an inward movement of one or both eyes. Up to 4 months of age, infrequent episodes of convergence are normal but thereafter ocular misalignment is abnormal. Congenital esotropia commonly referred as crossed eyes is not present since birth but usually develops within first six months of life in an otherwise normal infant with no significant refractive error and no limitation of ocular motility. Angle of deviation is usually constant and fairly large >30°. Infantile esotropia is usually associated with inferior oblique over action usually developing after one year of age, dissociated vertical deviation (DVD) in about 70-90% cases and latent horizontal nystagmus.

Patients with infantile esotropia usually do not develop binocular vision. There is alternate fixation in primary gaze and in lateral gaze there is cross fixation. Amblyopia develops in 25-40% when patient fixes more with one eye. Thus surgical correction should be done early in life to avoid amblyopia. Here we encounter a rare case of infantile esotropia with medial rectus insertion anomaly who presented in her adolescence and underwent a definite cosmetic surgical correction by medial rectus recession and ipsilateral lateral rectus resection at same time.

KEYWORDS
Infantile esotropia, Amblyopia, Medial Rectus Recession, Anomalous Medial Rectus Insertion, Lateral Rectus Recession.

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INTRODUCTION: Infantile esotropia develops in the early months of infancy and is not present since birth. Parents often give history of transient episodes of misalignment at age of 2 to 4 months. Aetiology of infantile esotropia is still unknown. Many theories were proposed, among them Worth1 had significant assertion that there is irreparable congenital defect in infant’s visual system and surgery is only for cosmetic reason later date. Chavasse supported by Costenbader and Parks suggested that the neural components necessary for normal binocular vision are present in strabismic individuals at birth, but the development of fusion is eventually impeded by abnormalities of optical input. So early surgery gives good outcome.

Hereditary component plays a role in etiology. Others being prematurity, perinatal or gestational complications, supplemental oxygen use at birth, use of systemic medications, and male sex.2 Instrumental delivery is also a contributory factor in it. Awareness of these risk factors can lead to early detection and management of esotropia. Amblyopia3 develops in structurally normal eye due to lack of fixation. If the brain is not stimulated binocularly the esotropic uncorrected eye goes for amblyopia hampering both the vision and cosmesis of the child.

CASE REPORT: A 14 years old girl attended to the outpatient department of AIMS, Bellur with history of crossed eyes and diminution of vision in left eye. Her grandmother who accompanied her said girl’s eyes have been crossed since two years of age, and the left eye seems to cross more than the right. They have not consulted any doctor these many years, now the girl complains of diminution of vision in left eye since 2 years. She was a full term infant with spontaneous normal delivery without perinatal complications, and no adjoining medical conditions. Her aunt suffers from the same problem in the family.

Her general physical examination was normal. On Ocular Examination she had a head tilt towards left side and left eye was clearly crossed inward (esotropic). There is no facial hemiparesis and any neurological deficit adjoining it. The best corrected visual acuity was 6/6 in right eye but left eye had 6/36 with no improvement with pin hole. With the left eye covered, she fixes and follows easily. However, with the right eye covered and patient faces more trouble following with her left eye as the binocular vision is hampered. Left side on extraocular movement lateral rectus movement was restricted not moving beyond the midline in left eye. The AV phenomenon was not associated with the above eye. There was no nystagmus. Pupils were equally round and reactive to light; no afferent pupil defect with no leukocoria. Corneal reflection test (Hirschberg test) showed 35° to 40° esotropia when penlight was directed towards the cornea, and the reflected image was located temporal to the center.
of left pupil. Cover-uncover test: On covering the right eye, there is an outward shift of the left eye. When the eye is uncovered, the left eye shifts back inward. Alternate-cover test: On switching the cover to the left eye, there is an outward shift of the right eye. When the cover is alternated from one eye to the other, there is always an outward shift of the opposite eye. Though patient presented late, surgery was planned in view of cosmetic reason and to prevent further deterioration of vision in her left eye due to amblyopia. The patient was diagnosed with infantile esotropia with amblyopia left eye and was planned for medial rectus recession with lateral rectus resection under GA. Intraoperatively after conjunctival and Tenon’s dissection anomalous medial rectus insertion was observed about 3mm from limbus for which 8 mm medial recession and mm lateral rectus resection was done. Postoperatively, her eyes were orthophoric in primary position.

Fig. 1: Pre-Op Photos of Infantile Esotropia Case

Fig. 2: Intra-Operative Photos
DISCUSSION: Strabismus is misalignment of visual axes of two eyes. Broadly it can be classified as 1) Apparent squint or pseudo strabismus. 2) latent squint 3) Manifest squint (heterophoria) - a) concomitant squint b) incomitant squint. Infantile esotropia is one of the concomitant squint and also the most common type of infantile strabismus. Congenital esotropia is a misnomer as esotropia is not present at birth and presents within six months of age. Genetic component plays a major role. Other factors associated are prematurity, instrumental delivery, use of supplemental oxygen, hydrocephalus, developmental delay, seizure disorders, intraventricular haemorrhage.

Strabismic amblyopia is the other major problem of infantile esotropia. In most cases, one eye the fixing one remains dominant and the other eye does not focus, and hence it fails to develop the normal visual pathway in childhood as the brain ignores the signals from that eye to avoid diplopia. Thus amblyopia develops and the esotropic eye is known as lazy eye. If the eyes are alternatingly fixing it’s a good sign, as the vision can actually be equal in both eyes, but up to 40% will have associated amblyopia. Angle of deviation is usually constant and large. An estimate of the amount of esodeviation can be made with corneal reflection testing. The Hirschberg corneal reflex test is a rough but handy method to estimate the angle of manifest squint involving shining a light onto the cornea. If an eye is deviated inward, the light reflex will be temporal to the pupil center. The definitive method of testing for strabismus is the cover-uncover test. An occluder is placed over the fixing eye and the opposite eye is observed. In case of esotropia there will be inward deviation.

Pseudoesotropia (pseudo strabismus) is the most common differential diagnosis, where a wide, flat nasal bridge with prominent epicanthal folds gives a crossed appearance. Accomodative esotropia is also frequent, which is treated with glasses as the patient is far-sighted (hyperopic) and the strain to focus causes the eyes to turn inward. Other differential diagnosis of infantile esotropia includes bilateral congenital sixth nerve palsy, secondary due to organic diseases, mechanical limitations of eye movements such as Duane syndrome (agenesis of the sixth nerve nucleus, accompanied by globe retraction on adduction) and Mobius syndrome (palsy of sixth, seventh, and twelfth cranial nerves). Nystagmus blockage syndrome in which convergence dampens a horizontal nystagmus.

Treatment of infantile esotropia is mainly focused to correct amblyopia if present. The weaker eye is made to focus by patching the fixing eye to treat the amblyopia. Once vision improves surgical alignment is planned. Most commonly done surgery is medial rectus recession and lateral rectus resection. This procedure involves detaching the medial rectus muscles from their scleral insertion sites, then suturing them to the sclera several millimeters behind the original insertion sites. This effectively weakens the muscles, diminishing their adducting effect. Earlier the surgery better the level of stereoscopic depth perception (stereopsis). So surgery should be planned within two years of age more recent studies suggest by one year of age.

A close follow up should be kept after surgery for any postoperative misalignment and amblyopia. Parents should be counselled for requirement of repeated surgeries for misalignment and other motility disorder in future.
Alignment to near-orthophoria is the typical goal. This results in a stable alignment with an excellent appearance with good visual outcome.

CONCLUSION: Very early surgery can result in excellent motor alignment and high-grade stereo acuity in some patients with congenital esotropia but in our case patient presented at 14 years of age so a medial rectus recession with lateral rectus resection was planned giving the patient excellent cosmetic correction with little correction to amblyopia.

REFERENCES: