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Essential Infantile Esotropia: An Unusual Case Report

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Abstract:- Esotropia is a condition in which one eye is turned in or nasally. These in turning may be alternate or constant. Congenital or essential infantile esotropia syndrome is a common type of strabismus. It is mostly present in the first 6 to 12 month of age. Association may be with cross fixation, other extraocular muscle overaction, amblyopia, large deviation mostly with no refractive error and with or without nystagmus. It should be differentiated from other causes of esotropia. Management include mostly surgical. Here we report a case of 10-year-old male child with left eye congenital esotropia (40 PD BO) with no oblique overaction, no amblyopia with normal fundus. The case was managed by left eye medial rectus recession and lateral rectus resection. So, in conclusion we report uniocular surgery has same result as binocular surgery. However, surgery done in an early age has a good outcome as compared to delayed surgery.

Keywords:- *Amblyopia, Base Out (BO), Cross Fixation, Esotropia, Prism Diopter (PD), Overaction*

I. INTRODUCTION

Congenital or essential infantile esotropia is an inward deviation of eye with an onset earlier than six months of age.In 1903 Worth proposed that the cause of squint was a "congenital defect of the fusion faculty" and that rebuilding of binocularity in affected patients wasdejected. [1]Afterward, much clinical verification has accumulated propose that surgical alignment of the infantile esotropia within 10 PD of orthotropia before two year of age rise the constancy of obtaining binocular vision. [2,3,4]

The history of strabismus has been discussed broadly by Duke-Elder. [5]Because of their clear and ugly looks, cross-eyed figures are mentioned prominently in primitive tradition andmythology.The word strabismus derives from the Greek mythology (strabismos means squinting). [5]

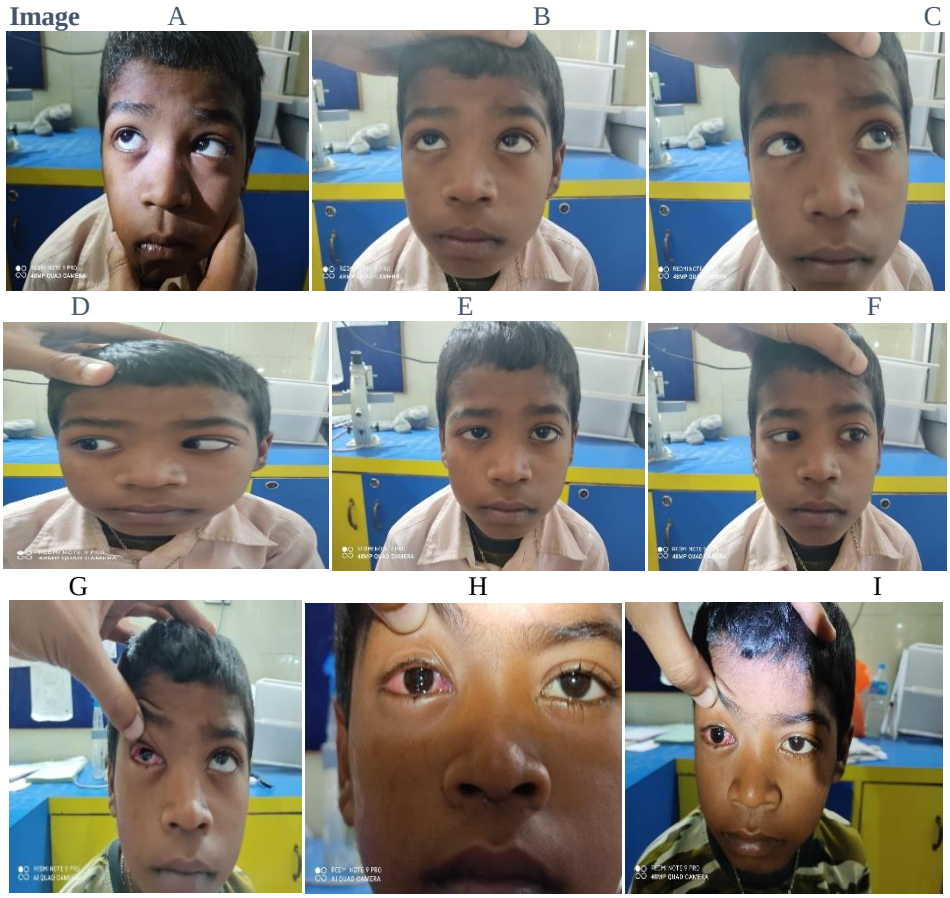
The frequency of congenital or infantile esotropia varies from 0.1 to 1%.[6,7,8] It is seen earlier at 6 months of age, with angle more than 30 (PD base out), with or no refractive error. Associations generally include latent nystagmus, dissociated vertical deviation (DVD), decreased binocular single vision, with or without inferior oblique overaction and cross fixation of eyes. Infantile (congenital) esotropia need detail history including birth history, slit lamp examination, family history, old family photographs if required and orthoptic examinations to identify amblyopia, type of squint, judge the angle of squint, examine binocular single vision which consist of simultaneous macular perception, fusion, stereopsis. Orthoptic evaluation must rule out sensory - motor evaluation of strabismus, anomalous retinal correspondence and suppression.

II. CASE HISTORY AND EXAMINATION

A 10-year-old male child presented to pediatric eye out patient department at RIO IGIMS medical college, Patna, Bihar with a history of left eye inward deviation alternately. His mother had noticed an ocular misalignment 5 years before. He was misdiagnosed as a case of ocular palsy elsewhere and was treated outside since last four years. He had visual acuity of 6/6 in both eyes on Snellen visual acuity chart. There was no history of trauma or fall, no history of diplopia and abnormal head posture. There was no history of fever, vomiting, seizures, rashes or lesion. Birth history and milestones were normal. Immunization history is complete till date. Family history of was not significant. Old photograph of a child showed left eye esotropia.

III. STRABISMUS AND ORTHOPTIC WORK UP

Cover -uncover test showed alternate esotropia (left eye is more preferred then right eye. Extraocular movements both duction – version was full and free in all diagnostic cardinal gaze position. Dolls eye maneuver showed full eye movements. Forced duction test was negative which signify non paralytic (concomitant) strabismus. Wet retinoscopy with tropicamide -phenylephrine (0.8 %- 5 %) express +2.0D in both axis in both eyes. Hirschberg corneal light reflex test manifest 15 ^{o} esotropia. PBCT (prism barcover test) both near and distance appear 40PDBO in primary position, 40 PDBO in elevation and 45 PDBO in depression. AC/ A ratio was absent. Worth four dot test signify alternate suppression. So, we diagnosed it as an alternate esotropia with no oblique overaction. Patient underwent surgery under general anesthesia. Right eye medial rectus recession 5.5 mm and lateral rectus resection 7 mm was done. Picture A and C showing no oblique overaction whereas picture E showing right eye esotropia. Picture H and I denote normal ocular alignment (orthotropia) after surgery.



IV. DISCUSSION

Post-operative satisfactory strabismus surgery was defined as orthotropic (0 PD); small angle residual esotropia (0 to +10 PD); small secondary exotropia (0 to -10 PD); esotropia (> +10 PD) and exotropia (< -10 PD).[9] The phrase infantile esotropia is identical with congenital esotropia but hardly very small number of cases are actually present at birth. Meanwhile the actual date of eye deviation is difficult to determine so that 6 months is take on by eye surgeon to diagnose this condition. Nelson in 1987 chose to employ congenital (infantile) esotropia which is classic postpartum start of eye asymmetry.[10] There are different theories to diagnose congenital esotropia. Worth [11] in 1903 put forward that it is a congenital deficiency of fusion faculty. He also states that it is an irreversible defect in the brain ability to fuse so that binocularity is lost. Worth sensory theory was altered by Chavasse's motor theory in 1939. He projected that the neural components of binocular vision are available in congenital esotropia. He explained that the primary problem is mechanical and curable if the deviation could be eliminated in infancy. Chavasse [12] suggested that deformity in binocular vision may be the result and not the cause of early squint. Up to 1960 studies look to bear Worth's theory of irreversible loss of binocularity, but more than the past fifty years studies stated favorable sensory results in some infants who went surgical correction between 6 months and 2 years. These findings formed for the current practice of early surgery for patients with congenital esotropia. For evaluation, cases of essential infantile esotropia require detail history like birth history, family history and milestone history. Examination consists of vision, ocular movements, squint evaluation by (Hirschberg test, PBCT, Krimsky test) cycloplegic refraction, nystagmus evaluation, cross fixation, forced duction test if possible and Dolls head rotation test. For vision testing in preverbal child we use CSM method to rule corneal light reflex central, steadiness with cover uncover test and maintained fixation with the help of occluding other eye if uncover eye moves less than three second, we called as unmaintained which is gross indication for amblyopia. Differential diagnosis consists of accommodative esotropia, 6th CN palsy, DRS syndrome, sensory esotropia and nystagmus blockage syndrome. Hiles [13] in 1980 explained that latent nystagmus occurs in one third of patients, inferior oblique overaction and DVD found in more than half of patients with essential infantile esotropia. In our case inferior oblique overaction, amblyopia and DVD were absent. Alternate suppression was present. The main aim of management of congenital esotropia is to reduce deviation to orthotropic as much as possible. Post-operative alignment within 10 PD results in monofixation syndrome which is associated with central suppression and peripheral fusion. This small angle squint represents stable, normal cosmetic aspect and is considered a good surgical outcome even if fusion was absent. The common surgery for infantile esotropia is both eye medial rectus recession. Other technique like recess resect in single eye is also done. Two muscle surgery spares the other two horizontal muscles for next surgery if required. Scott et al [14] in 1986 states that some surgeon will perform three or four muscle surgery if angle of squint was more than 50 PD.

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V. CONCLUSION

It is essential for pediatric ophthalmologist to discuss surgical, medical management, prognosis and follow up of any case of infantile esotropia with the child's parents. Parents are worried about their child and ophthalmologist or optometrist have an important role to rule out any doubt of child's parents like risk of amblyopia, post-surgery complication and chances of secondary surgery.

Image J **K**

Picture J shows another a 14 years old girl with right eye infantile esotropia. Picture K was one week post operative with orthotropia.

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