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Bilateral Type-I Duane Syndrome with Multiple Anamolies: A Case Report

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Abstract: *The Duane syndrome is a strabismus syndrome which is characterized by congenital non-progressive horizontal ophthalmoplegia which primarily affects the abducens nerve. Approximately 70% of the individuals with the Duane syndrome have an isolated disease. We have described here, a case of bilateral Duane syndrome with associated anamolies.*
Keywords: *Duane syndrome , Bilateral Congenital Zonular cataract , Hypertelorism , Lash Ptosis.*

I. Case Report

A 10-year old boy who was born out of a non consanguineous marriage with an uneventful perinatal period was brought with a complaint of decreased vision in both eyes. On examination the child has Hypertelorism and Telecanthus (3.4cm) . Vision in both eyes – 6/60. Has lash ptosis in both eyes. Ocular movements – Bilateral restriction of Abduction , with retraction of globe and narrowing of palpebral fissure with down shoots and nystagmus on attempted adduction. Eyelids , Conjunctiva and Cornea are normal. Pupil reactions normal. Zonular lenticular opacity present in both eyes. Fundus is normal.

II. Discussion

Duane's retraction syndrome (DRS), also known as Stilling-Turk-Duane syndrome, is defined as a congenital miswiring of the lateral and medial recti muscles, resulting in an impaired ocular motility syndrome that includes palpebral fissure narrowing. The incidence of DRS is approximately 1% of the total cases of strabismus. Eighty percent of cases are unilateral and characterized by either limited abduction, limited adduction, or both. It was first described by Sinclair, Turk and Stilling in 1895. In 1905, Duane summarized the clinical presentations and postulated the theory for its pathogenesis and treatment. It is caused by an absence or hypoplasia of both the abducens nucleus and the nerve with an anomalous innervation of its target, the lateral rectus muscle, by a branch of the oculomotor nerve. Duane's syndrome has three variants:

- Type I: Limited abduction with or without esotropia
- Type II: Limited adduction with or without exotropia
- Type III: Limitation of both abduction and adduction and any form of horizontal strabismus

The first type is more common and accounts for 85% of the cases.[1][10]
Esotropia is the most common type of strabismus which is encountered and characteristic up shoots and down shoots occur in adduction. Most patients are diagnosed by the age of 10 years and Duane's is more common in girls (60 percent of the cases) than boys (40 percent of the cases). The other associated ocular anomalies which can occur in subjects with the Duane syndrome can include nystagmus, anisocoria, ptosis, optic nerve colobomas, epibulbar dermoids, crocodile tears and aniridia. This syndrome is also associated with skeletal (limb hypoplasia, polydactyly and hypoplastic or absent radius and/or thumb), vertebral (scoliosis, spina bifida, "butterfly" vertebrae and Klippel-Feil anomaly), genitourinary (renal agenesis and vesicoureteral reflux), and cardiac (patent ductus arteriosus and auricular septal defect) defects [5,6]. The treatment of the DS may involve correction of the refractory error and squint and surgical procedures like muscle recession procedures, vertical transposition of the rectus muscle, or a combination of the two, for improving or eliminating the head turns and misalignment of the eyes.

III. Conclusion

The DS is a rare syndrome and bilateral DS is all the more so. Any child with bilateral DS should be completely evaluated to rule out other syndromic associations. Immediate and early ophthalmological interventions with refractory error treatment, use of prisms and surgical corrections, will go a long way in helping such children to lead a normal life. Our child presented with Type-1, bilateral DS and "Congenital cataract"- a rare combination which has been scarcely reported in the Indian medical literature .

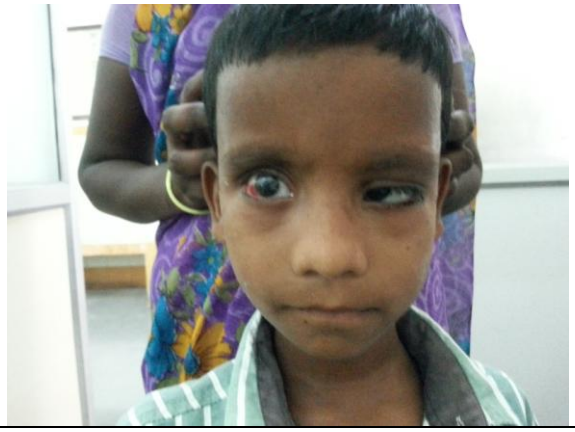

Fig 1 : Right eye abduction limited and Retraction of globe Left eye.

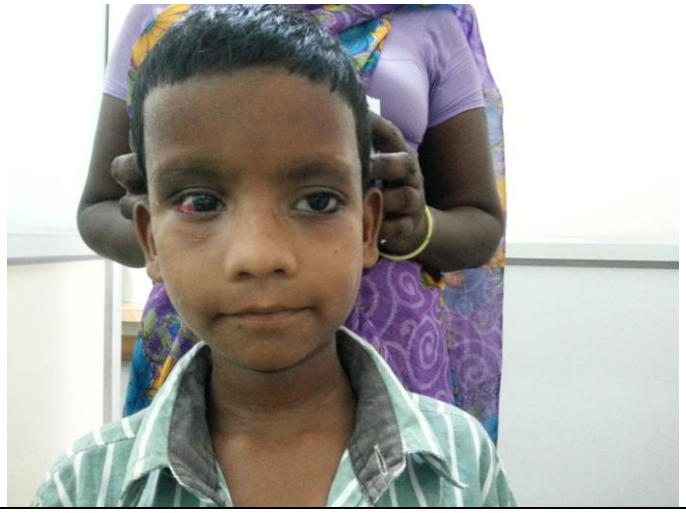

Fig 2 : Left eye Abduction limited

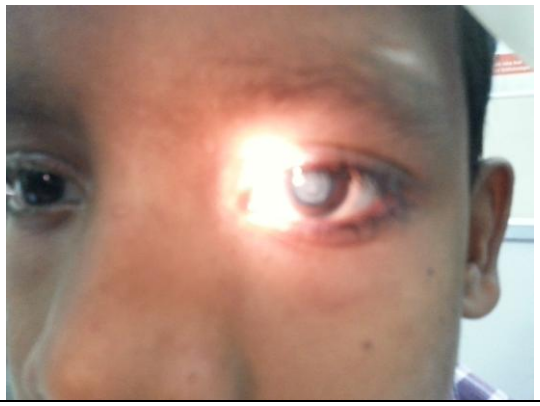

Fig 4 : Right eye Pseudophakia and Left eye Zonular Lenticular opacity

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