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CASE REPORT

DUANE RETRACTION SYNDROME (TYPE 1A) IN A MALE CHILD

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ABSTRACT: Duane retraction syndrome is a congenital form of strabismus characterized by horizontal eye movement limitation, globe retraction and palpebral fissure narrowing in attempted abduction. DRS are present in 1% of strabismus patients. It is most commonly unilateral, but can be bilateral. It affects the left eye more frequently than the right eye and females affected more than males. Refractive error commonly associated is hypermetropia. Here we are presenting a case of a 15 year old boy with Type 1A Duane's retraction syndrome in Right eye with myopia.

KEYWORDS: Duane's retraction syndrome, Abduction, Type 1A.

INTRODUCTION: Retraction Syndrome, also known as Stilling-Turk-Duane Syndrome, was originally described by Alexander Duane in 1905. The syndrome was first described by ophthalmologists Jakob Stilling (1887) and Siegmund Türk (1896), and subsequently named after Alexander Duane who discussed the disorder in more detail in 1905.[1,2,3,4,5,6]

DRS is a congenital, incomitant ocular motility disorder characterized by abnormal function of the lateral rectus muscle in the affected eye, together with retraction of the globe and narrowing of the palpebral fissure on attempted abduction.[6,7,8] Generally, the lateral rectus does not abduct the eye, but instead contracts at the same time as the medial rectus on abduction. It is this simultaneous co contraction of the medial and lateral rectus muscles on attempted abduction that causes the retraction of the globe and narrowing of the palpebral fissure when the eye is adducted.[9]

DRS is present in 1% of strabismus patients.[10] It is most commonly unilateral, but can be bilateral.[11] For unknown reasons, it affects the left eye more frequently (approximately 60% incidence),[11-16] and approximately 60% of patients with DRS are females.[8] Although it is usually sporadic, there could be up to 10% of familial cases mostly with autosomal dominant inheritance. Hypermetropia of greater than +1.50 diopter was more frequent in DRS (71%). Myopia and Emmetropia appeared in relatively equal amounts (15% and 14% respectively).[17]

It is usually sporadic, although there could be up to 10% of familial cases mostly with autosomal dominant inheritance. Several autosomal dominant syndromes with dysmorphic features are associated with DRS.[18]

CASE REPORT: A 15 years old school boy attended the outpatient department with complaints of defective vision in both eyes for few years and restricted outward movement of right eye since childhood, according to his father. No history of head ache, nausea or vomiting was present. No history of fever or trauma. No other members in family were affected. No ophthalmic consultations so far.

He was born out of a non-consanguious marriage with full term normal delivery. Antenatal, intranatal and postnatal history was insignificant.

On examination, his unaided visual acuity in both eyes was 6/36, which improved to 6/6 with

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-1.50DS. He had restriction of abduction in RE (Fig. 1) with normal abduction. It was associated with narrowing of palpebral fissure and with retraction of the eye ball on abduction (Fig 3 & 4). No up shoot or down shoot of eye was noted. He had Esotropia of 5 prism diopter in RE (Fig. 2), Very mild degree of head turn towards right side was noted which was cosmetically insignificant. Binocular single vision was maintained. Fundus examination in both eyes showed normal disc, vessels, macula and periphery was normal. The associated ocular features of DRS (Table I) were not seen. Systemic abnormalities known to be associated with DRS were also not present.

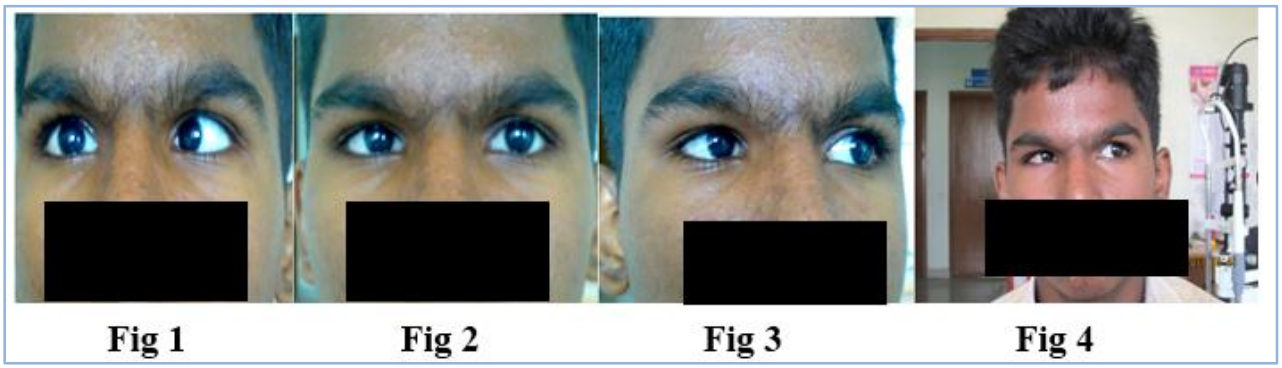

Fig 1 Fig 2 Fig 3 Fig 4

DISCUSSION: Duane's syndrome is a congenital and non-progressive strabismus syndrome. It results from an absent or dysplastic abducens motor neurons with aberrant innervations of the lateral rectus muscle by the oculomotor nerve, from failure of normal development of the pontine abducens nucleus or nerve resulting in failure of the normal innervation of the lateral rectus muscle on the affected side. At the same time, an aberrant branch of the oculomotor nerve innervates the lateral rectus muscle. Thus, globe retraction results from co-contraction of the medial and lateral rectus muscles on attempted abduction.

Huber has classified Duane's syndrome in to three types.[17] (Table 1). Type 1 DRS, Type 2 DRS and Type 3 DRS. Among them Type 1 is the most common with 70-80% prevalence.

Type - 1 (70%-80%)	• Inability to abduct • Normal or minimal defect in abduction • Esotropia with head straight • A or V pattern • Usually up drift or a down drift of affected eye on abduction or attempted abduction. • Globe retraction and palpebral-fissure narrowing on abduction • Usual face turn to affected side • Electromyography shows absence of electrical activity in the lateral rectus muscle on abduction but paradoxical electrical activity on abduction

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Type - 2 (about 7%)	• Inability to abduct • Normal or minimal defect in abduction • Esotropia with head straight • A or V pattern • Usually up drift or a down drift of the affected eye on abduction or attempted abduction • Globe retraction and palpebral-fissure narrowing on abduction • Usual face turn to affected side • Electromyography shows absence of electrical activity in the lateral rectus muscle on abduction but paradoxical electrical activity on abduction
Type -3 (about 15%)	• Inability ability to abduct and adduct • Globe retraction and palpebral-fissure narrowing on attempted abduction • Possible up shoot and down shoot on abduction • Straight or nearly straight head position • The electromyography demonstrates co contraction of the horizontal rectus muscles on both abduction and abduction

Table 1: Huber's classification of DRS

A modification of Huber's classification was proposed by Aluwalis, Gupta, Goel and Khurana based on the deviation in primary position of gaze[17]. This is relevant for only DRS Type 1 because DRS Type 2 is always Exotropia in primary position and DRS Type 3 is always Orthotropia in primary position. The proposed classification is as follows: DRS Type 1A (esotropia in pp), DRS Type 1B (exotropia in pp) and DRS Type 1C (orthotropia in pp).So based on the above classification we diagnosed the patient to have DRS Type 1A in right eye.

As already mentioned it is more common in females and left eye is more commonly involved with hypermetropia being the most common refractive error associated, we are presenting a case of a male child with right eye Duane retraction syndrome type 1A with myopia as the refractive error.

Treatment options include correction of the refractive error, treatment of amblyopia and surgical correction. Surgical correction is required for patients with significant head turn, strabismus in primary gaze, and significant up shoot and down shoot on abduction.[19,20] Surgical treatment has its limitations as it does not assure complete clinical recovery.

As the patient had very mild head turn with no amblyopia with preserved binocular single vision we opted for conservative line of management and prescribed him glasses. Regular follow up was advised and no worsening of symptoms noticed till date.

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