

## Bilateral Duane's Retraction Syndrome: A Rare Case Report with Review of Literature.

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**Abstract:** This report presents case of 6th cranial nerve anomaly typical DRS. We present a rare case of bilateral 6<sup>th</sup> cranial hypoplasia/aplasia.

### INTRODUCTION

Duane's retraction syndrome (DRS) is a congenital eye movement disorder characterized by abduction deficiency, adduction limitation, globe retraction, and palpebral fissure narrowing on attempted adduction. Neurogenic cause (miswiring of third & sixth nerve) for this disorder is well documented.

### CASE REPORT

Twelve year old boy of P3G4 of nonconsanguineous marriage was referred to our dept for CT brain with the h/o squint & blurring of vision. There was no history of birth trauma too. No other family member is having such discomfort. Eye examination [Figure - 1] There was no ocular abnormality, except restriction of ocular movements. Both eyes could not be abducted beyond midline. The other ocular movements were normal. There was narrowing of palpebral fissure with retraction of the globe of right eye on levoversion and likewise left eye was also observed on dextroversion. P/E: Normal hearing, palate, no crocodile tears, normal cervical spine MRI (fiesta-seq) revealed absence of bilateral 6<sup>th</sup> nerve, normal carotid cavernous sinus and ocular muscles. Normal midbrain study [figure-2]



Figure 1



Figure 2: MRI fiesta showing absence of sixth nerve at clivus

### DISCUSSION

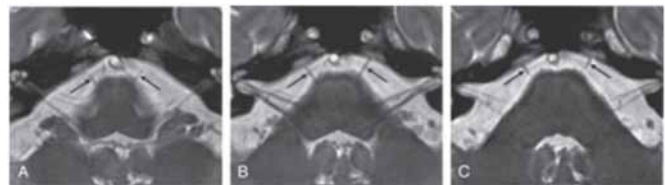
The syndrome was first described by ophthalmologists Jakob Stilling (1887) and Siegmund Türk (1896) and subsequently named after Alexander Duane who discussed the disorder in more detail in 1905<sup>1</sup>. Miswiring of third and sixth nerve was the aetio-pathogenesis of DRS, causing some eye muscles to contract when they shouldn't and other eye muscles not to contract when they should<sup>2</sup> abducent nerve nucleus-nerve was absent or hypoplastic and lateral rectus muscle is innervated by a branch of 3<sup>rd</sup> nerve (EMG study). Most of the patients are diagnosed by the age of 10 years and Duane's is more common unilaterally, in girls (60% of the cases) than boys (40% of the cases) and more on left eye than right eye<sup>4</sup>. The gene sal-like 4 has been implicated as a cause of this condition. Common DRS associated anomalies are Klippel feil anomaly, congenital labyrinthine deafness, Marcus Gun phenomonon,

and paradoxical gustatory lacrimal reflex and Okihiro syndrome, Wildervanck syndrome, Holt-Oram syndrome, morning-glory syndrome, Goldenhar syndrome and congenital labyrinthine deafness

The characteristic features of the syndrome are include limitation of abduction (type 1 Duane's syndrome) or adduction (type 2 Duane's syndrome) or both (type 3 Duane's syndrome) of the affected eye palpebral fissure on attempted abduction. A head turn to the side of the affected eye to compensate for the movement limitations of the eye (s) and to maintain binocular vision, In the primary position, eyes may be straight, esotropia or exotropia.

Majority of the DRS patients remain symptom free are able to maintain binocularity with only a slight face turn. Routine MRI sequences will demonstrate the absence of a tumor but may not be able to demonstrate abducent nerve aplasia or hypoplasia in such a case as they may not visualize the nerve even in normal subjects.

FIESTA sequences reveals visualization of sixth nerve (accuracy 100%)<sup>5</sup>



FIG; Normal study; Three axial MR images show the entire path of the right and left abducens nerves (arrows) as linear dark structures emerging from the pontomedullary sulcus (A), coursing in the superior oblique direction (B), and finally entering into the clivus (C).

### CONCLUSION

MR imaging (3D FIESTA) is a robust tool for diagnosis it is useful for the differential diagnosis of abduction deficit of the eyeball. The **absence of the bilateral/unilateral abducent nerve** strongly suggests DRS in children with bil/unilateral abduction deficit.

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