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Duannes Retraction Syndrome

KEYWORDS

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INTRODUCTION

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] "Congenital Deficiency of Abduction associated with impairment of adduction, retraction movements, contraction of the palpebral fissure and oblique movements of the eye". Archives of ophthalmology

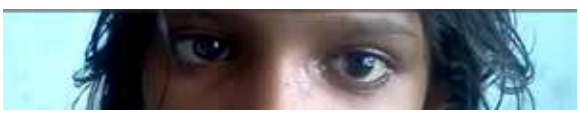
Other names for this condition include: Duane's Retraction Syndrome, Eye Retraction Syndrome, Retraction Syndrome, Congenital retraction syndrome and Stilling-Turk-Duane Syndrome.[2]There are three types of DRS .Type-1-limitation of adduction,Type -2-Limitation of abduction,Type -3-Limitation of both adduction & abduction.

Mutations in the ***CHN1*** gene are associated with familial isolated Duane syndrome. Direct sequencing of the ***CHN1*** gene is available as a clinical test, and has to date detected missense mutations in seven patients and affected family members. The ***CHN1*** mutations have not been found to be a common cause of simplex Duane retraction syndrome.

CASE VIGNETTE:

12 YEAR old female admitted with c/o diplopia on looking for distant objects & to the left &with diminished visual acuity ,had on examination- limitation of adduction in the right eye &limitation of abduction in the left eye with narrowing of the palpebral fissures on adduction &widening of the palpebral fissures on abduction ,retraction of globe,horizontal nystagmus on all positions of gaze

She belonged to the type -3 DRS .



DISCUSSION

The basics in DRS are congenital absence of abducent nucleus ,aberrant innervations of 6th nerve by inferior division of 3rd cranial nerve nucleus.This aberrant innervations causes retraction of the globe &narrowing of the palpebral fissure on adduction.In 70 percent of DS cases, this is the only disorder the individual has. However, other conditions and syndromes have been found in association with DS. These include malformation of the skeleton, ears, eyes, kidneys and nervous system, as well as:

Okihiro's syndrome, an association of DS with forearm malformation and hearing loss, Wildervanck syndrome, fusion of neck vertebrae and hearing loss, Holt-Oram syndrome, abnormalities of the upper limbs and heart, Morning Glory syndrome, abnormalities of the optic disc or "blind spot", and Goldenhar syndrome, malformation of the jaw, cheek and ear, usually on one side of the face.

REFERENCE ADAMS TEXT BOOK OF NEUROLOGY-page no263a LOCALISATION IN CLINICAL NEUROLOGY-PAUL BRAZIS De jongs-193,211,214,215 • Duane A (1905 ". Archives of ophthalmology (Chicago) 34: 133–50. ". Archives of ophthalmology"Learning About Duane Syndrome". Retrieved 2007-06-06.