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DUANE'S RETRACTION SYNDROME — A REVIEW OF 39 CASES

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Abstract

Thirty nine cases of Duane's Retraction Syndrome who presented to the Eye Clinic at the Children's Hospital over a 12 month period are reviewed. Many of their features are consistent with those of previous authors, although, there was a high incidence of associated abnormalities, which may be explained by the specialist paediatric setting in which they were seen.

Key words: *Paediatrics, associated anomalies, aetiology, lateral rectus.*

Duane's Retraction Syndrome is not a rare condition, and its forms may be as varied as its reported aetiology. It was described by Stilling in 1887 and Turk in 1896. Duane in 1905 provided a more complete definition; and there are currently varied explanations as to its cause.

It was originally described as a musculofascial anomaly, being thought to be due to replacement of normal contractile substance within the lateral rectus with fibrous tissue. Strachan[1] describes increased stiffness in the forced duction test of the lateral rectus of all 15 cases examined. An inability of the lateral rectus to generate a normal force pattern was also reported, lending support to the early theory of fibrotic muscle tissue.

Burian and Von Noorden[2] postulate that it is the result of an innervational disturbance of nuclear or supranuclear origin, with abnormal substitute innervation of the paresed lateral rectus occurring via a branch of the third cranial nerve.

More recent study based on electromyography by Prieto-Diaz suggests the main feature of the syndrome is co-contraction of the horizontal eye muscles.[3] Electromyography distinguishes the normal firing of the medial rectus from the abnormal innervation of the lateral rectus, which show silence on abduction but firing on adduction. Co-contraction of the lateral rectus and, at times co-contraction of the superior or inferior rectus when the patient attempts adduction, produce the retraction of the globe and the palpebral fissure narrowing. Kommerell and Bach[4] believe that the co-contraction occasionally of superior and inferior recti leads to the noted 'A', 'V' and 'X' patterns associated with Duane's syndrome. A case of twitch abduction on attempted upgaze has been reported as a new type of Duane's syndrome with 'V' incomitance; it is thought to be similar to abducting twitches on attempted up/down saccades which can occur following aberrant regeneration after a third nerve palsy. Kommerell and Bach's explanation for this is that the burst impulse in nerve fibres misdirected from a vertical to the medial rectus, pulls the eye away from its direct vertical trajectory.

Retraction is often inconspicuous and may be revealed only by electromyography, as discussed by Huber, Esslen, Kloti and Martenet.[5] Some

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cases described as a sixth cranial nerve palsy are quite possibly Duane's syndrome. In most cases of Duane's, the absence of innervational input to the lateral rectus when the patient attempts abduction, is the cause of that inability to abduct. (It must be noted too, that part of the inability to abduct may be due to restriction which is secondary to a stiff medial rectus.)

Hotchkiss et al describe a clinicopathological study of a case of bilateral Duane's syndrome where the intracranial and orbital pathology show absence of both abducens nuclei and nerves,[6] with the lateral recti being partially innervated by branches of the third cranial nerves. Similarly, Miller describes a unilateral case where the lateral rectus was innervated by the inferior branch of the third cranial nerve; there was an absence of the sixth cranial nerve and no cell bodies of motor neurons in the sixth nerve nucleus.[7]

The diverse anomalies associated with Duane's syndrome (Pfaffenbach, Cross and Kearns,[8] Ramsay and Taylor,[9] O'Malley, Helveston and Ellis[10]) suggest that Duane's syndrome may be of teratogenic origin, with the teratogenic stimulus occurring toward the middle of the first trimester of pregnancy. The congenital anomalies of ear deformity, deafness, facial hypoplasia, facial paresis and thalidomide embryopathy which have appeared concurrently with Duane's syndrome, share a common critical time of embryological development. If Duane's syndrome could be of teratogenic origin then the presence of crocodile tears in some cases could shed light on the site of the lesion.

Thirty nine cases of Duane's syndrome presented to the Eye Clinic at The Children's Hospital over a 12 month period from 1987-1988. Of these, 44% were male, and 56% were female.

Involvement of the left eye only occurred in 61% of cases, the right eye only in 18%, 21% were bilateral. Seventy seven per cent were classified as Type 1, 10% Type 2, and 13% Type 3 (according to Duane's original classification). These findings are consistent with other published studies, as can be seen in Table 1.

In this series a strabismus (most commonly an intermittent esotropia) was the most common reason for presentation, occurring in 41% of cases. Other reasons included 'poor eye movement' (23%), referral for suspected sixth cranial nerve palsy (10%), apparent ptosis (12%), and pain on eye movement, frequent blinking, blocked tear duct and presentation for a general check up due to a family history of unrelated eye problems.

Duane's syndrome usually occurs sporadically but it has been reported that there is some family history in about 2% of cases.[10] In this series there was one case where a male infant's father also had the syndrome. Another female child had one male sibling with the Märcus Gunn Jaw Winking Syndrome and another male sibling with a intermittent exotropia and a significant 'V' pattern. Their parents were first cousins.

The syndrome is a benign entity and treatment is not usually required. Seventy four per cent of this series had good binocularity, usually with a slight compensatory head posture (77% of the whole group had such a head posture). Of the group, 59% were orthophoric or had a well controlled heterophoria, 13% had intermittent esotropia, 3% intermittent hypertropia, 21% constant esotropia and 5% constant exotropia. O'Malley, Helveston and Ellis[10] reported

TABLE
Comparative Incidences (in %) of Features Associated with Duane's Retraction Syndrome

	Right Eye only	Left Eye only	Bilateral	Male	Female	Type		
						I	II	III
This Study	18	61	21	43	56	77	10	13
O'Malley, Helveston and Ellis[1]	16	66	18	48	62	87	11	2
Parks[13]	22	60	18	35	65			
Pfaffenbach,[8] Cross and Kearns	22	60	19	43	57			

'orthotropia' in 31% of their cases, esotropia in 53% and exotropia in 16%.

In this series, 23.1% required correction of some anisometropia and/or astigmatism and in 89% of these cases the greater refractive error was found in the eye affected by Duane's syndrome. Tredici and Von Noorden in a series of 72 cases of Duane's syndrome found 17% had anisometropia of greater than one diopter,[11] and O'Malley et al found a 16% prevalence of the same.[10] Consideration must, however, be given to the approximate 10% of the normal population having anisometropia of greater than one diopter.[12]

One third required some part-time occlusion for amblyopia. Of these, 46% had strabismic amblyopia; 8% refractive amblyopia; 31% a combination of strabismic and refractive amblyopia; and 15% stimulus deprivation amblyopia due to ptosis.

Cosmetic surgery was required in 18% of cases, this was generally to reduce an abnormal head posture. Eight per cent required and responded well to convergence insufficiency treatment.

Duane's syndrome may co-occur with other syndromes. The Klippel Feil Syndrome is said to occur in 3-4% of cases and Congenital Labyrinthine deafness in 7.5-11%.[8,13] When these problems occur together with Duane's syndrome it is called Cervico Oculo Acoustics Syndrome or Wildervanck syndrome. Similarly, Goldenhar's Syndrome may co-occur with Duane's syndrome,[13] and cases of thalidomide embyopathy have also been described by Papst.[14] A rarer co-occurrence is that of the gusto-lacrimal reflex (crocodile tears).[9]

Other ocular conditions such as optic nerve hypoplasia,[15] coloboma of the fundus, iris and lids, pupillary anomalies, persistent hyaloid arteries, cataract[2] and Marcus Gunn Jaw Winking[16] have also been reported in conjunction with Duane's syndrome.

Two of the cases in this series had Goldenhar's syndrome, one with cerebral palsy, deafness and facial hypoplasia, and the other with hydrocephalus, facial hypoplasia, dextrocardia, transverse liver, the left main bronchus on the right side and a malpositioned right ear. One child had middle ear malformation, and another, with bilateral Duane's had developmental delay, curvature of the spine and butterfly vertebrae. One case had neurofibromatosis. There were also one case each of Henoch-Schoelien Syndrome and Congenital Adrenal Hyperplasia.

Ptosis, 'A' and 'V' patterns, convergence weakness, latent nystagmus, chalazion and micropthalmos were also noted.

The high incidence of associated anomalies in this series is no doubt explained by the specialist nature of The Children's Hospital, which deals with the whole range of paediatric medicine. However, it does indicate the need for a thorough examination of any child presenting with this syndrome

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