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DUANE'S RETRACTION SYNDROME TYPE 1 ASSOCIATED WITH DISSOCIATED VERTICAL DEVIATION: A CASE REPORT

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Abstract

A case is reported of an eight year old girl with a left Duane's retraction syndrome type 1, bilateral dissociated vertical deviation (DVD) and an infantile esotropia. The patient also had congenital hydrocephalus which was treated with a ventriculoperitoneal shunt. This is the third case report of a Duane's syndrome and DVD presenting together. The clinical characteristics are discussed.

Key words: *Hydrocephalus, infantile esotropia.*

INTRODUCTION

The retraction syndrome as described by Stilling (1887), Turk (1896) and Duane (1905) has the following typical and most frequently observed clinical characteristics: marked restriction or complete absence of abduction, normal or a slight restriction of adduction, narrowing of the palpebral fissure and retraction frequently with an upshoot or a downshoot of the affected eye on attempted adduction, widening of the palpebral fissure on attempted abduction.[1] The various manifestations of Duane's retraction syndrome have been categorised by Huber[2] into the 3 types:

1. palsy of abduction
2. palsy of adduction
3. palsy of abduction and adduction

Other reported associated signs include horizontal strabismus (mainly esotropia), 'A', 'V' or 'X' patterns, defective convergence and a compensatory head posture, upshoot of the affected eye on attempted adduction, elevation or depression on attempted abduction and bulging of the orbital fat through the septum orbitale.[2-4]

Duane's retraction syndrome is most commonly of congenital origin and may also be acquired.[3,5,6] The syndrome is more common in females and on the left side. Bilateral cases have been reported in about 20% of cases,[5] though it has been argued by Rowe et al. that bilateral Duane's may be more common.[4] There have also been reports of associated abnormalities including congenital deafness, seizures, congenital facial palsies, microcephaly, generalised hypotonia and crocodile tears and oculocutaneous albinism.[4,7-9]

The aetiology of this syndrome proposed by

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Huber was 'paradoxical anomalous innervation of the lateral rectus'.[2] More recent electrophysiological studies have proved this hypothesis and have shown an abnormality of the brainstem in which the lateral rectus is innervated by the third nerve.[10]

Dissociated vertical deviation is characterised by an upward movement of the eye when occluded. On removal of the occluder, the eye "drifts" downwards. Other associated signs are: extorsion, latent nystagmus, horizontal deviations, asymmetry of Optokinetic nystagmus (OKN), a positive Bielschowsky phenomenon where the affected eye elevates behind the occluder, but as the density of the red filter is increased, the eye behind the occluder returns to the primary position, inferior oblique overaction, 'A' pattern and weak (at best) binocular functions.[3,11]

DVDs are commonly associated with infantile esotropia, though it may become manifest after surgery for the correction of the horizontal deviation. Since Stevens was reportedly the first to describe DVD in 1895[7] a plethora of descriptions have been used to describe this condition. In 1990, Wilson and McClathey[12] argued that DVD implied a purely vertical deviation, and suggested the term 'dissociated strabismus complex', to be divided into vertical, horizontal and torsional components.

Various hypotheses have been put forward to explain this phenomenon.[13-15] To date the aetiology is unknown though it is apparent that there is a disturbance of both the sensory and motor systems.

This paper presents a case of a patient with a Duane's retraction syndrome type 1 and an infantile esotropia with a DVD. This is only the third case reported of a Duane's retraction syndrome type I occurring with DVD. It is the first case report of these two conditions occurring with an infantile esotropia.

CASE REPORT

E.O. was born at term after a normal pregnancy. The labour was of fifteen hour duration and there was a forceps delivery. The circumference of E.O.'s head at birth was noted to be 36.5 cms. This increased to 37.5 cms five days later. Neonatal jaundice was also evident and was treated with phototherapy. One week after birth E.O. presented with a low occipital encephalocele, which was repaired. Hydrocephalus then developed and required a ventriculo-peritoneal shunt.

At 7 months of age, E.O.'s shunt became occluded. A cerebral ultrasound revealed grossly dilated lateral ventricles, a moderately dilated third ventricle and a slightly dilated fourth ventricle. The shunt was replaced. At this time a left esotropia was noted by the neuro-surgeon, though E.O.'s mother has reported subsequently the presence of the esotropia since birth.

Six months later E.O. was referred to an ophthalmologist. The presence of a right esotropia of approximately twenty degrees and rotary nystagmus was confirmed. The right visual acuity was thought to be reduced. There was bilateral failure of abduction and cross fixation was queried.

The neuro-surgeon's report at this time diagnosed a midline syndrome. The term 'midline syndrome' encompasses developmental, neuroplastic, vascular and/or traumatic damage to any of the following: corpus callosum, cingulate gyrus, septum pellucidum, septal area or fornix. The features may include ocular abnormalities, especially optic atrophy, strabismus, nystagmus and chorio-retinitis. Epilepsy and mental retardation are often present, but were not in this case.[16]

At three years of age E.O. underwent a right medial rectus recession and a maximal right lateral resection for correction of the esotropia. Cosmesis was good post-operatively in the presence of a left hypertropia. As time progressed a left esotropia became manifest when E.O. was tired.

Later that year E.O. was admitted to hospital with symptoms of right sided headaches, drowsiness and vomiting. The CT scan was normal, but a right 6th nerve palsy was queried by the neuro-surgeon. At this time, an ophthalmological investigation revealed the presence of a left esotropia and a left Duane's syndrome type 1 with lid retraction in abduction. No papilloedema was

evident. One month later, E.O.'s shunt blocked once again and a new catheter was inserted.

E.O. was investigated at the Orthoptic Clinic at La Trobe University in 1991 at eight years of age. Poor cosmesis was the presenting complaint. The hydrocephalus was under control and E.O. was not on any medication. There was an insignificant refractive error of hypermetropic astigmatism. The fundi and media were normal. There was a family history of strabismus. E.O.'s father (now deceased) and her two paternal aunts had a history of strabismus, but the types of deviations are unknown.

The orthoptic examination revealed a face turn to the left; visual acuity of 6/9, N5 in both the right and left eyes. On cover testing at both ⅓ metre and 6 metres there was a moderate left esotropia with left hypertropia, bilateral DVD (left greater than right) and extorsion. Measurements were made by prism cover test. At near with either eye fixing the esotropia was measured at 18△; the right hypertropia was 6△ and the left hypertropia was 30△. At 6 metres with either eye fixing the esotropia was measured at 12△. The right hypertropia was 7△ and the left hypertropia was 20△. The measurement of the vertical deviations is considered to be an estimate only due to the variability of the deviations. OKN was intact with both eyes open when the drum was rotated from left to right and from right to left. When each eye was tested separately, OKN was present when the drum was rotated from temporal to nasal, but was absent when the drum was rotated from nasal to temporal. The Sbiza bar was used to examine the Bielschowsky phenomenon. However, it could not be demonstrated. Testing of ocular pursuit movements showed no movement of the left eye beyond the midline with an upshoot of the left eye and widening of the palpebral fissure on attempted abduction, retraction of the left eye and narrowing of the palpebral fissure on adduction. The left eye diverged on elevation. Binocular functions were tested with Worth's lights, Bagolini glasses, the Lang two pen test and the synoptophore. Simultaneous perception could be demonstrated only on the synoptophore; fusion and stereopsis could not be demonstrated. A visuscope examination revealed latent micro-nystagmus.

DISCUSSION

Based on the well documented history and the most recent orthoptic investigation, the diagnosis of a Duane's retraction syndrome, type 1 associated with an infantile esotropia and bilateral DVD was made.

This is only the third case to be documented. The first (case 1) was reported by Clarke et al. in 1988[15] and the second (case 2) by Rimmer and Katz in 1990.[7]

Both previously reported cases were of Duane's syndrome type 1 with bilateral DVD. There was no detailed past history available for either Case 1 or Case 2. Case 1 had an associated alternating esotropia and Case 2 had a small hypertropia. Case 2 had an upshoot of the affected eye on attempted abduction. Both cases had latent nystagmus. No fusion or stereopsis could be demonstrated in Case 1. There was no report of assessment of binocular function in Case 2. Neither had undergone any treatment.

E.O.'s findings demonstrate some similarities and differences between these two cases and frequently reported characteristics of Duane's syndrome and DVD. E.O. had a type 1 Duane's syndrome with an upshoot of the affected eye on attempted abduction. This is in contradiction to most reports which are of an upshoot on attempted adduction. Micro latent nystagmus was evident, which is frequently reported in association with DVD. Only simultaneous perception could be demonstrated therefore the head posture of a face turn to the left was not in the interest of binocularity. In this case a detailed past history was available, which confirmed the presence of an infantile esotropia.

The most significant difference between this case and the other two cases was that E.O. had hydrocephalus. The sudden appearance of a squint in patients with hydrocephalus is diagnostic of sudden raised intra-cranial pressure. While the first documented appearance of the squint at 7 months of age was prior to an episode of raised pressure, the presence of the squint was reportedly noted at birth. This cannot be

confirmed. The DVD was first documented when E.O. was eight years of age, so the exact time of onset is impossible to determine.

Duane's syndrome is generally considered to be a congenital condition. However, there are reported cases of acquired retraction syndrome[6] and Raab[5] has quoted findings of a patient in which a Duane's syndrome apparently evolved. Was E.O.'s Duane's syndrome a congenital condition or was it an acquired condition secondary to complications caused by the hydrocephalus? While the diagnosis of Duane's in this case was first documented when E.O. was 5 years, limitation of abduction had been noted at 13 months, but this was considered to be associated with the infantile esotropia. It is possible that the presence of the infantile esotropia associated with cross fixation masked the Duane's syndrome until E.O. was old enough to be more accurately examined. Conversely, when the Duane's syndrome had been diagnosed the upshoot of the left and the divergence of that eye on elevation may have masked the presence of the DVD.

Neither Clarke et al.[15] nor Rimmer and Katz[7] could offer an explanation for the concurrence of these conditions. Clarke et al. commented that it was "interesting to speculate that our patient may have a single abnormality accounting both for the presence of a Duane's syndrome and DVD" and Rimmer and Katz stated that "the concurrence of both syndromes implies that a clinically common strabismic entity can occur with differing underlying neuroanatomy". The authors are in agreement with Clarke et al. that if both conditions are caused by a brain stem anomaly the co-existence of these conditions would be more commonly found. The most likely explanation is one of coincidence, though it is interesting that within four years there have been three case reports.

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