Australian Orthoptic Journal 1994; **30: 71-74**

LETTERS TO THE EDITOR

CONTINUING INTRIGUE WITH BROWN'S SYNDROME AND IT'S ASSOCIATION WITH ESOTROPIA

In 1992, we reported on a patient who had congenital Brown's syndrome in association with accommodative esotropia.[1] Although Brown's syndrome is recognised to be mostly associated with normal binocularity and absence of heterotropia in the primary position,[2] accompanying horizontal strabismus, particularly esotropia, has been described by numerous authors over the past 40 years. We apologise for omitting Clarke and Noel[3] and Bourne[4] from our 1992 literature review. Clarke and Noel reported horizontal strabismus and amblyopia in 8 out of 28 Brown's syndrome patients. Bourne, in a series of 20 patients, found 2 had constant esotropia with amblyopia, and 4 had intermittent esotropia in forced primary gaze. Further, Gregerson and Rindziunski[5] have since reported on 10 Brown's syndrome patients who were followed for between 7 and 19 years, 3 of whom had co-existing constant or intermittent esotropia and 1 who had exotropia. In this communication, we wish to update the 'case-law' and report on a further three patients who have congenital Brown's syndrome associated with either accommodative or non-accommodative estropia. Are these coincidental or does the ocular motility deficit seen in Brown's syndrome potentiate an *esotropic force*, either sensory and/or motor?

Case 1. Master R.M., aged 8 years, was diagnosed with bilateral Brown's syndrome of moderate severity (grading of severity based on features described by Eustis, O'Reilly and Crawford, 1987[6]) and an accommodative esotropia that was reduced to microtropia with +4.00/+1.00 correction OU. There was good fusion and stereopsis to 100" (Titmus®), despite the presence of a moderate degree of amblyopia, 6/6 and 6/15.

Case 2. Miss K.K., aged 5 years, presented with severe bilateral Brown's syndrome and large-angle esotropia. She required anisometropic correction of +0.50DS OD and +3.00DS OS. There was no binocular vision and a high degree of amblyopia, 6/10 and 6/30. The ocular rotations were restricted such that on cover testing there was esotropia and hypotropia of each eye under cover. Following some cycles of right occlusion, surgery was undertaken to correct the Brown's sydrome (superior oblique tenotomies) and the esotropia (bimedial rectus recessions). A severe Brown's syndrome on the right side persisted post-operatively, however.

Case 3. Master C.C., aged 8 years, presented with moderate bilateral Brown's syndrome leading to a chin-up compensatory head posture. There was a small-angle intermittent esotropia, tenuously controlled in the primary position and breaking to esotropia with diplopia on slightest chin depression. Correction of the minimal refractive error, +0.25/+0.25 OD and +0.25/+0.50 OS, made no difference to the control. Random-dot stereopsis could be demonstrated during periods of fusion. In view of the problematic head posture, bilateral superior oblique tendon spacer surgery was performed which successfully cured the Brown's syndrome, but not surprisingly caused a troublesome V-pattern and increased esodeviation that will require further operation.

The three patients described all had bilateral Brown's syndrome associated with acquired esotropia. R.M. had a microtropia with fully accommodative esotropia characteristics, K.K. had a non-accommodative constant esotropia, and C.C. had a non-accommodative intermittent esotropia. Forceps duction testing confirmed mechanical restriction in K.K. and C.C. at the time of operation.

We continue to be intrigued by the mechanism that caused esotropia to develop in these patients. It is recognised that mechanical and structural anomalies of the extraocular muscles like Brown's syndrome may be ætiological factors in strabismus.[7] But is this motility deficit alone enough to compel a convergent ocular posture? Since Parks' 1977 report that the superior oblique tendon does not have an anterior sheath, it is generally accepted that the cause of congenital Brown's syndrome in at least the majority of cases lies in the superior oblique tendon itself and/or the trochlea.[8] The mechanical restriction of elevation in adduction could result from a tight or inelastic superior oblique muscle or tendon, or an intrinsic anomaly of the trochlea complex whereby the telescopic movement of the tendon through the pulley during relaxation of the muscle is interfered with. Whatever the exact mechanism, the following occurs in Brown's syndrome. From the primary position, adduction of the globe along the midline plane of movement is not allowed because of the increased linear distance from the trochlea to the insertion of the superior oblique.[9] Rather, adduction is accompanied by a gradual infraduction, thus the characteristic downshoot that is usually seen in Brown's syndrome. Abduction, on the other hand, is not disturbed. It is also unlikely that the inferior oblique's secondary action of abduction could be inhibited when the eye is in the primary position. Therefore, there is no obvious direct mechanical or anatomical esotropic force in Brown's syndrome. Moreover, co-existing exotropia has also been reported in the literature[3,5,6].

The interruption of adduction may have significant sensory consequences however, especially if the condition is bilateral. [3] Vertical incomitant disparity would have adverse effects on the retinal correspondence. Nasal hemiretinal suppression is the most likely antidiplopic mechanism to prevail, certainly in congenital Brown's syndrome. This alone could potentiate an esotropia given the abundance of convergence tonus in early childhood. Its combined presence with hypermetropia of moderate degree, as in Case 3, would have similar effects in producing esotropia.

Having presented these three further cases of congenital Brown's syndrome with acquired esotropia, we wish to reiterate that it is not Brown's syndrome *per se* that induces esotropia, but the consequences of the motor anomaly on the binocular visual system that lead to its suppression and the arrest of fusion, and ultimately, strabismus. Stressing the significance of sensory anomalies in the ætiology of strabismus.

ZORAN GEORGIEVSKI, BAppSc(Orth)Hons
ALISON PITT, MEd DBO(T)
Division of Orthoptics, La Trobe University, Melbourne.

References

1. Pitt A, Georgievski Z. A case study - Brown's syndrome associated with accommodative esotropia. Aust Orthopt J 1992; 28: 47-49.
2. Pratt-Johnson JA, Tillson G. Management of Strabismus and Amblyopia. A Practical Guide. New York: Thieme, 1994: 186.
3. Clarke WN, Noel LP. Brown's syndrome: fusion status and amblyopia. Can J Ophthalmol 1983; 18(3): 118-123.
4. Bourne K. Brown's syndrome, current concepts and a clinical review of twenty cases. Aust Orthopt J 1990; 26: 24-27.
5. Gregerson E, Rindziunski E. Brown's syndrome. A longitudinal long-term study of spontaneous course. Acta Ophthalmol 1993; 71(3): 371-376.
6. Eustis HS, O'Reilly C, Crawford JS. Management of superior oblique palsy after surgery for true Brown's syndrome. J Pediatr Ophthalmol Strabismus 1987; 24(1): 10-16.
7. Von Noorden GK. Binocular Vision and Ocular Motility. Theory and Management of Strabismus. 4th ed. St Louis: CV Mosby, 1990: 139.
8. Wilson ME, Eustis HS, Parks MM. Brown's syndrome. Surv Ophthalmol 1989; 34(3): 153-172.
9. Brown HW. Congenital structural anomalies of the muscles. In: Allen JH (ed.) Strabismus: Ophthalmic Symposium II. St Louis: CV Mosby, 1958: 391-427.