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Restoration of binocular single vision following multiple treatments for longstanding bilateral internuclear ophthalmoplegia and unilateral third nerve paresis in Chiari II malformation

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Abstract

Aim: **To present a patient with longstanding bilateral internuclear ophthalmoplegia and unilateral third nerve paresis in Chiari II malformation who achieved binocular single vision with the help of prisms following two surgical and two botulinum toxin procedures.**
Method: **The case is discussed of a 34-year-old man who presented with a longstanding exotropia and a gradually increasing abnormal head posture to achieve fixation. Investigation, treatment and follow-up to discharge are described.**
Results: **The patient initially underwent surgery with the aim of improving cosmesis of the exotropia and the abnormal head posture, but functionally benefical stereopsis and field of binocular single vision were achieved with multiple treatments including Fresnel prisms.**
Conclusion: **Functional results are achievable, even in cases of longstanding strabismus associated with complex ocular motility and neurological defects.**

Key words: Bilateral internuclear ophthalmoplegia, Chiari II malformation, Longstanding, Restoration of binocular single vision, Third nerve palsy

Introduction

Internuclear ophthalmoplegia (INO) is characterised by a decreased ability to adduct one or both eyes that is caused by a lesion in the medial longitudinal fasciculus (MLF). A dissociated jerk nystagmus is present in the abducting eye, and normal convergence and a reduction in the velocity of the adducting eye on horizontal saccades may also be evident. Patients with a unilateral or bilateral INO may demonstrate binocular single vision (BSV) in primary position, often in the presence of an exophoria. Bilateral cases that exhibit a large exotropia in primary position are sometimes referred to as 'wall-eyed bilateral INO' (WEBINO).

The most common aetiologies of INO are demyelination and vascular. Other aetiologies include trauma, infection, hydrocephalus, brainstem and fourth ventricle tumours, and Chiari malformation. In Chiari II malformation part of the cerebellum, the lower part of the brainstem and part of the fourth ventricle herniate down through the foramen magnum into the neck area; and it is associated with spina bifida.[1] Patients with spina bifida are known to suffer from a range of ophthalmic problems, mostly related to hydrocephalus, which is caused by the coexisting Chiari malformation.

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Case report

A 34-year-old man presented complaining of increasingly poor cosmesis of a right exotropia and the need to gradually adopt a right face-turn to be able to see. He reported the onset of exotropia at age 8 years, although it is possible the onset was prior to this. He had received no previous ocular treatment and he reported the angle of the exotropia had continued to increase over time. The patient had spina bifida with hydrocephalus and Chiari II malformation, was a wheelchair user, but could walk short distances with the aid of crutches and held a disabled driving licence. He had undergone neurosurgery soon after birth to repair an open spinal canal defect, at which time a ventriculoperitoneal (VP) shunt had been inserted. This VP shunt was revised after 2 months and a ventriculoatrial (VA) shunt inserted. At the age of 2 years the VA shunt had become blocked, but no further intervention was planned as he had 'compensated for this himself'. A neurology review was requested following his presentation to the ophthalmology department, and this concluded the shunt system was non-functional and had not been functioning for a long time. No further neurological intervention was planned unless the patient became symptomatic from raised intracranial pressure in the future. A CT examination of the head was performed instead of an MRI scan due to the presence of an older type intraventricular shunt. The CT scan was reported to show the intracranial manifestations of a Chiari II malformation associated with spina bifida, including the intraventricular shunt from the right parietal approach, with massive ventriculomegaly, which appeared to be longstanding. No mass lesions or other structural abnormalities were evident on imaging. Full Goldmann visual fields were documented and there were no medical contraindications to strabismus surgery.

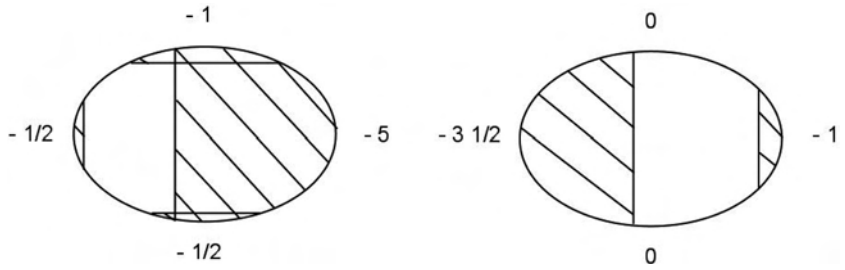

Fig. 1. Pre-operative ocular movement restrictions.

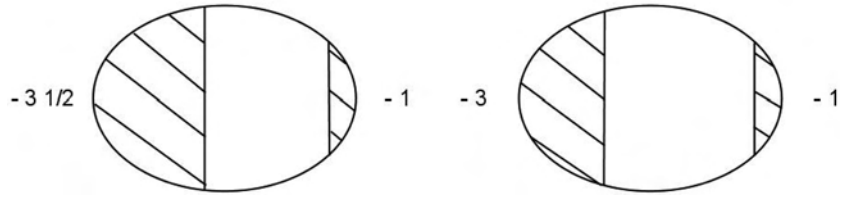

Fig. 2. Ocular movement restrictions following two surgical procedures and two BT injections.

Visual acuity with myopic and astigmatic correction was 6/12+2 right eye, with a moderate face-turn left to achieve fixation and the upper lid lifted; 6/6+2 left eye with a slight face-turn right to achieve fixation. Cover testing revealed a marked right exotropia and a significant face-turn left was necessary to take up right fixation. The patient was unaware of any diplopia or panoramic vision. Ocular motility testing revealed a 4 mm ptosis, −5 limitation of adduction, −1/2 limitation of abduction, −1 limitation of elevation and −1/2 limitation of depression of the right eye; and a −3½ limitation of adduction and a −1 limitation of abduction of the left eye (Fig. 1). Abducting nystagmus movements were evident in each eye on abduction. Both fourth cranial nerves were thought to be intact. Pupils were of equal size. The exotropia measured greater than 70 dioptres (D) by prism reflection test at near and 32° by reflections on the synoptophore. A forced duction test (FDT) performed under local anaesthesia confirmed a mild limitation of adduction of the right eye. At this stage the patient was diagnosed as having right third nerve paresis and bilateral INO with contracture of the antagonist lateral rectus muscles.

Follow-up measurements were stable and surgery was performed with the aim of improving the cosmesis of the exotropia. Under general anaesthesia both lateral recti were found to be slightly tight on FDT. A 3.0 mm resection of the right inferior rectus and superior rectus muscles and a nasal transposition of the muscle insertions to the borders of the right medial rectus muscle were made and augmented with Foster sutures. One day post-operatively the right exotropia had reduced to 60–70D by prism reflection test at near. However, at 2 weeks post-operatively the deviation had increased to 85–90D right exotropia and 5D right hypertropia by prism cover test at near and 75D right exotropia and 5D right hypertropia by prism cover test at distance. Ocular movement testing revealed the right adduction was still a −5 limitation, abduction was still a −1/2 limitation; however, elevation was now a −3 limitation and depression was now a −2 limitation. Despite little change in his adduction and the size of the exotropia, the patient felt he had gained significant functional improvement post-operatively. In an attempt to improve the adduction of the right eye, 5 international units (IU) of botulinum toxin (BT) A were injected into the right lateral rectus muscle at 4 weeks post-operatively.

After the right lateral rectus BT A injection the patient once again felt there was a functional improvement, despite prism cover test measurements and the right adduction improving minimally. A further right lateral rectus BT A injection of 5 IU was performed, after which the right adduction improved to −4 limitation. The exotropia at this time measured 60D at near and 70D at distance with a slight right hypotropia by the prism reflection test. Further surgery was performed with the aim of improving the adduction of the right eye. The right lateral rectus was found to be slightly tight at 7 mm from the limbus and it was recessed to 20 mm from the limbus.

At 1 day post-operatively following this second surgical procedure the limitation of right adduction improved to 1 and limitation of right abduction increased to −3½ (Fig. 2), culminating in a more balanced limitation on right gaze. Prism cover testing measured a 14D exotropia and 4D right hypotropia at near, and a 14D exotropia and 7D right hypotropia at distance. The patient was aware of vertical diplopia and a 4D base-up Fresnel prism was fitted to the right lens of his glasses. At follow-up the patient demonstrated binocular single vision (BSV) with Fresnel prisms and these were subsequently altered according to the patient's symptoms. The patient reported single vision 'most of the time' and BSV was documented as a small horizontal prism fusion range (near: 12D base-out, 4D base-in;

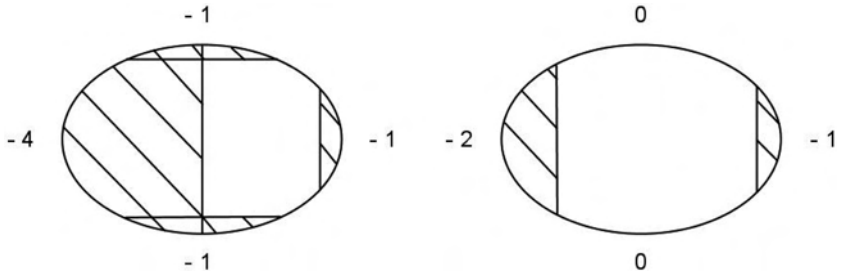

Fig. 3. Ocular movement restrictions at discharge.

distance: 14D base-out, 2D base-in), binocular convergence to 10 cm and 80″ arc stereopsis (TNO) at best, although this varied depending on the patient's level of tiredness.

At 4 years follow-up the patient had a −4 limitation of abduction, −1 limitation of adduction, −1 limitation of elevation and a −1/2 limitation of depression of the right eye, with slight right ptosis; and a −2 limitation of adduction and a −1 limitation of abduction of the left eye (Fig. 3). No abnormal head posture was present and useful field of BSV was demonstrated with incorporated prisms (a total of 5D base-out and 5D base-up right lens); however, the patient continued to be aware of diplopia when very tired. The patient was happy with this functional result and was discharged.

Discussion

This patient presented with an increasing angle right exotropia, which had been present for at least 26 years. Due to a unilateral right third nerve paresis and bilateral INO the patient also had complex limitations of his ocular movements and was progressively adopting an abnormal head posture (face-turn right) to achieve fixation with his left eye and function visually. Two surgical and two BT injections to his right eye only allowed him to regain a useful and central field of BSV with the aid of prisms, and the need for an abnormal head posture was eliminated.

Conservative management options in cases of unilateral and bilateral INO include occlusion and prismatic correction of any primary position deviation; of which occlusion is often the most successful due to the incomitant nature of the strabismus and the variability of symptoms in different positions of gaze.

Murthy *et al*.[2] reported their experiences using BT in managing 16 patients with unilateral and bilateral INO. The injections were successful in reducing the angle of deviation in 94% of cases; however, only 3 patients (19%) achieved any form of BSV. In the long term 31% of patients in their series continued with maintenance injections, 19% were discharged with a stable angle of deviation without troublesome diplopia and 31% were discharged with occlusion. Two patients (13%) went on to have further surgery and 1 patient (6%) was discharged symptom-free with prisms, although no BSV was documented with the prisms. Interestingly in this series 14 of the 16 patients also commented that subjectively they felt better and had functional improvements in everyday aspects of living despite continued diplopia symptoms following the injections. This is similar to our case, in that despite initially achieving only minimal objective improvements in adduction and the angle of the deviation in primary position, the patient commented that he felt significant functional improvements.

Cosmetic and functional benefits may also be gained from surgery in patients with INO. Adams *et al*.[3] reported 3 cases of multiple sclerosis causing bilateral INO with large angle exotropia in primary position that achieved improvements in primary position alignment and function following surgery. One patient had two surgical procedures, 7 years apart, which enabled her to be diplopia free, although in the interim period she had been managed with prisms. The second patient was asymptomatic with a small exophoria in primary position after one surgical procedure. The final patient was symptom free after one surgical procedure; however, they had suppression in the presence of a small manifest strabismus in primary position.

Roper-Hall *et al*.[4] reported on 8 cases of WEBINO that underwent one or two surgical procedures in an attempt to either improve alignment, eliminate diplopia, restore BSV or reduce an abnormal head posture. Seven of the eight achieved some level of BSV following surgery and in 4 of these 7 it was with the aid of prisms. BSV was most commonly sensory fusion as demonstrated by the Worth Four Dot test and motor fusion, where the testing method used is unclear. Only 2 patients achieved stereopsis post-operatively and again the testing method used is unclear.

Conclusion

This case highlights the fact that cosmetic as well as substantial subjective and objective functional improvements can be achieved in longstanding strabismus, even in cases of Chiari II malformation causing bilateral INO and unilateral third nerve palsy. The patient reported significant functional improvements in daily activities even when minimal objective improvements were achieved in adduction and primary position alignment with the initial surgery and BT injections. Useful BSV was regained with the help of prisms following the second surgery. Prisms may not be as useful in the initial stages of treatment of bilateral INO and WEBINO cases, but they should remain an invaluable treatment option when managing residual deviations following surgery.

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