Surgical management of nystagmus

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All nystagmus degrades visual acuity by reducing the stability of fixation and lessening foveation time. In addition, adults with acquired nystagmus may complain of *oscillopsia*, a subjective impression of movement of the perceived world. Surgery may be performed for congenital nystagmus (CN), for latent or manifest-latent nystagmus (LN, MLN) or for acquired nystagmus. Nystagmus surgery aims to reduce the amplitude of nystagmus with either improvement of visual acuity or a reduction of a bothersome head posture. The commonest reason for patients' seeking surgical treatment is to allow them to acquire a driving licence. Regrettably, this is frequently not possible.

SURGERY FOR CONGENITAL NYSTAGMUS

Congenital nystagmus can occur in patients with no other ocular abnormality (*idiopathic motor nystagmus*), or may be associated with other ocular conditions such as oculocutaneous or ocular albinism, early visual deprivation or retinal dystrophies. All patients require a careful ophthalmological examination to exclude other ocular abnormalities, a cycloplegic refraction and, if a refractive error is detected, a trial of optical correction with spectacles or contact lenses. *Recession* of a rectus muscle is the commonest operation to reduce its action. The muscle insertion is moved to a more posterior point on the eye, usually 5–7 mm from its original insertion, where it reattaches. *Resection* is the commonest strengthening operation. A segment of muscle (5–8 mm) is removed and the shortened muscle reattached at its original insertion.

Four main surgical strategies have been advocated in management of congenital nystagmus—Kestenbaum surgery for compensatory head posture with null zone; artificial divergence surgery; maximum recession of horizontal rectus muscles; and rectus muscle anterior tenotomy.

Kestenbaum surgery

It has never been adequately explained why certain patients with congenital motor nystagmus (CN) have a position of gaze where the intensity of their nystagmus is minimal. Such patients discover that their vision is at its best when their eyes are placed in the position of least ocular instability, and commonly demonstrate a compensatory head malposition to bring the zone of best vision into the straight-ahead position. In contrast, patients with manifest-latent nystagmus (MLN), who typically fix with only one eye, may find that their latent nystagmus, which invariably involves a nasalward drift of the fixing eye, is minimized by adopting a face turn towards the fixing eye.

In 1953, Anderson[1] and Kestenbaum[2] independently suggested that an abnormal head posture related to nystagmus could be alleviated by surgery. In the following year Goto[3] made similar suggestions. Anderson's proposal was for recession of the pair of rectus muscles whose action was in the direction of the face-turn. Goto suggested resection of the antagonist muscles, and Kestenbaum favoured surgery on all four muscles, although he also suggested the two eyes should have sequential surgery. It is the Kestenbaum strategy, with modifications, that is normally performed today, and his name tends to be attached to this surgical approach to nystagmus.

Types of head posture

Face-turns to one side or the other are the commonest type of posture, although minor degrees of head tilt or chin elevation or depression are seen. Large degrees of chin up/down or head tilt are unusual. The typical head posture is in moderately but not extremely eccentric gaze, and nystagmus intensity will often increase again in gaze beyond this 'null zone'.

Care should be taken to identify, by history or examination, patients with *variable head-turns*, as these do not benefit from active treatment. Some patients find that pushing their eyes into either position of extreme horizontal gaze helps to stabilize the eyes. Others have the rare condition of periodic alternating nystagmus, or the even rarer periodic alternating null zone.

Measurement of compensatory head positions is notoriously difficult. The most extreme head positions are often adopted only when the patient is making a maximum effort to read a distant test-type. Frequently there is no head posture for near, and when vision is not critical the head tends to be kept in the most comfortable position, usually

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straight ahead. Various methods have been adopted. Many authorities make a subjective estimate. Sradj [4] has devised a 'torticollometer'. Others[5] have used eccentric fixation on an array of light emitting diodes. Mitchell *et al.*[6] utilized an orthopaedic goniometer.

Assessment before surgery

All cases require careful *refraction*, with cycloplegia if indicated, since astigmatism and anisometropia are not rare. Even apparently trivial refractive errors should be corrected, as such correction may occasionally produce spectacular improvement in ocular stability. *Contact lenses* move with the eye and therefore offer an improvement in potential best visual acuity.

Prisms will allow the eyes to be placed in the eccentric null position while the head is straight (i.e. change the direction of gaze). They should be placed with the base towards the head-turn. Prisms may be very useful as a guide to likely benefit from surgical treatment, but have little to offer as a definitive therapy, since most head-turns are of 30° or more, requiring around 60Δ of prism. If they are ordered as Fresnel membrane prisms, acuity will be degraded. Solid glass prisms are unacceptably heavy and obvious; they will also cause chromatic aberration.

Surgical treatment

The principle underlying surgical treatment is to rotate the eyes in the direction of the head-turn—in other words, to produce a relative gaze palsy towards the side to which the eyes are normally directed. The technique is essentially that advocated by Kestenbaum. However, he advocated recessions and resections of only 5 mm, and this is not adequate to improve the head position in the typical case. Parks[7] in 1973 suggested his popular (and memorable) '5, 6, 7, 8' procedure. This dosage is based on the amounts felt to represent maximal surgery without sacrificing rotations—i.e. recession of one medial rectus of 5 mm, and one lateral rectus 7 mm, with appropriate resections of the other medial rectus 6 mm and lateral rectus 8 mm. Calhoun and Harley[8] and later Nelson *et al.*[9] suggested 'augmented Parks' surgery. Where the head-turn is up to 30°, the surgical amounts are increased by 40%—i.e. 7, 8.4, 9.8, 11.2 mm. Where the turn equals or exceeds 45°, they advocate 60% augmentation—i.e. 8, 9, 11.2, 12.8 mm. On the other hand, Pratt-Johnson[10] obtained excellent results by making all recessions and resections 10 mm. Other authors[11–13] have suggested various surgical dosages.

Most authorities agree that the early response to surgery wears off in some cases, with recurrence of the head posture. Reoperation—for example, re-resection of the previously resected muscles by 5 mm and a posterior fixation procedure on the previously recessed pair of muscles—is safe and effective.

Vertical postures and head tilts are much less common, although many face-turns are accompanied by minor degrees of vertical head displacement. When chin elevation or depression is the main feature, the principle of rotating the eyes in the direction of the head posture should be followed. Therefore for chin depression, both inferior recti should be resected 4–6 mm and both superior recti recessed a similar amount. For pure head tilt, the intortor on the side of the tilt (e.g. the superior oblique) should be weakened and the extortor on the other side (e.g. the inferior oblique) strengthened. Conrad and de Decker[14] operate on all four oblique muscle insertions, slanting the insertions to either increase or decrease their cyclotorsional effect. An alternative approach has been described by Spielmann[15], who slants the insertions on all the rectus muscles of each eye. Both approaches seem logical.

The benefit of surgery is largely cosmetic, but Dell'Osso and Flynn[5] have shown 'broadening of the null zone' postoperatively. Best corrected visual acuity is usually unchanged.

Artificial divergence surgery

This surgical approach is based on the common observation that many patients with congenital nystagmus show a significant reduction in its amplitude on convergence. Their near visual acuity is therefore frequently disproportionally better than distance acuity. In some patients, base-out prisms suppress nystagmus and improve vision. The aim of surgery is to induce a latent divergent ocular position (exophoria), which the patient will then overcome by exerting fusional convergence, thereby damping the nystagmus. Patients must have adequate fusional reserves to overcome the induced deviation, so preoperative testing with prisms is mandatory. Sedler *et al.*[16] reported 26 patients assessed preoperatively with prisms. 20 responded to prism adaptation and of these 17 did well with artificial divergence surgery; 3 patients required a combination of artificial divergence surgery with the classic Kestenbaum procedure. 6 patients who did not respond to prisms had Kestenbaum surgery as a primary procedure. Zubcov *et al.*[17] reported 18 cases treated with combinations of artificial divergence surgery and Kestenbaum surgery with particular reference to visual acuity. 6 had artificial divergence surgery alone: 2 gained one line of Snellen visual acuity, and one gained four lines. 7 patients had standard Kestenbaum surgery: one gained a single Snellen line of vision. 5 had a combined approach: 4 gained two or more lines of Snellen acuity.

Maximum recession of horizontal rectus muscles

This procedure is designed to symmetrically weaken the horizontal rectus muscles and reduce the amplitude of the

nystagmus, all congenital nystagmus being either purely or largely horizontal in direction. It was first suggested by Bietti and Bagolini in 1960[18], but was then revived by von Noorden[19] and Helveston[20] in 1991.

All four horizontal recti are recessed to around the equator of the globe, the medials being 10 mm from their insertions, and the laterals at 12 mm. The procedure causes remarkably little reduction in horizontal rotations of the eye and distinctly improves vision. The Snellen acuity is only slightly improved, but nearly all patients report a decrease in the time taken to identify an object of regard. This has been studied by Sprunger *et al.*[21].

Anterior tenotomy of rectus muscles

Dell'Osso *et al.*[22] have recently reported some studies in an animal with naturally occurring nystagmus, the achiasmatic Belgian sheepdog. Their thesis is that removal of the tendon organ responsible for proprioception abolishes the nystagmus. Anterior tenotomy of all rectus and oblique muscles certainly reduced nystagmus in two animals, but the results in man are still awaited.

Neurosurgery

In 1988 Funahashi[23] reported a series of 106 patients with congenital nystagmus of whom 10 (aged 18–42, mean 27) underwent stereotactic neurosurgical superior colliculectomy. 7 showed a reduction of nystagmus amplitude by 40%. There has been no replication of this study.

SURGERY FOR MANIFEST-LATENT NYSTAGMUS

Most patients with latent and manifest-latent nystagmus have no important visual symptoms as long as both eyes are open. A small group, however, have a significant face-turn towards the fixating eye in order to damp the nystagmus, which typically increases on abduction and decreases on adduction. If surgery is contemplated it should be directed at the fixing eye to prevent it drifting in a nasalward direction. Typical surgery would involve recession and posterior fixation of the medial rectus of the dominant eye. Many cases of manifest-latent nystagmus have esotropia and a face-turn. Here the amount of surgery for the face-turn and that for the esotropia should be summed. Typically, a standard recession of the medial rectus and resection of the lateral rectus on the dominant eye will be required. If patients are unhappy about surgery on their 'good' eye, it is possible to demonstrate the expected effects of surgery by means of an injection of botulinum toxin to the medial rectus of the dominant eye[24].

NYSTAGMUS SURGERY IN CHILDREN

Although congenital nystagmus is usually present by the age of 3 months, and children may be referred at quite an early age, surgery is normally deferred until the child is around 7–8 years old. There seems no evidence of permanent skeletal change due to an intermittently adopted head posture, and young children do not as a rule complain of the cosmetic defect. In binocular cases, there is a small risk in visually immature children of *inducing* strabismus, especially if the surgery is on vertical recti. In cases with manifest-latent nystagmus, amblyopia treatment should take priority and there is no urgency in surgical treatment since such cases are rarely if ever binocular.

SURGERY FOR ACQUIRED NYSTAGMUS

Regrettably, little can be done surgically for most patients with acquired nystagmus. Sometimes the causal lesion can be removed, as shown by the reports by Senelick[25] who improved down-beat nystagmus secondary to basilar impression and Arnold–Chiari malformation by transoral removal of the odontoid process, and Spooner and Balogh[26], who reported reduction of nystagmus in 4 of 5 cases of Arnold–Chiari malformation following suboccipital decompression. Roberts *et al.*[27] operated on 7 patients with nystagmus (acquired in 3 cases) and an abnormal head posture with extreme chin elevation or depression; the patients with acquired nystagmus improved by one or two Snellen lines.

Patients, especially adult patients, with nystagmus are seeking a therapy for an inherently incurable condition. They are understandably keen to try *any* therapy that might help. The condition can be very variable, with emotion, tiredness and stress all known to have effects on its severity. The placebo effect of any treatment, especially surgery, must be very great, and randomized trials of treatment are difficult to design. One should therefore try not to raise hopes of radical improvement with any therapy.

CONCLUSIONS

Patients with congenital nystagmus and a compensatory head posture should be fully assessed medically and ophthalmologically. All cases require careful refraction and a trial of spectacle or contact lens treatment. Prisms may be of value in assessment and selection of cases for surgery, but are of little value for long-term therapy. The modified Kestenbaum surgical approach to congenital nystagmus with an abnormal head posture has proved its worth over the past 47 years. Additional amounts of surgery may be beneficial in cases with exceptionally large face-turns. Recurrence of head position may be managed by reoperation. Artificial divergence surgery and maximal recession of the horizontal rectus muscles may benefit patients who do not adopt a compensatory head posture. In most cases of acquired nystagmus surgical treatment is unlikely to be of value.

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