The effectiveness of surgery for congenital nystagmus

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The paper by Atilla, Erkam and Isikcelik in this issue investigates the effect of recessions of all four horizontal recti on 12 patients with congenital nystagmus (CN). Since this procedure was suggested by Bietti and Bagolini[1] rather limited improvements in visual function have been reported, despite some reduction of the CN.[2,3] The results of this latest paper are more encouraging as the authors find an improvement in visual acuity (VA) along with a reduction of CN.

The gaps in our knowledge of CN present several interesting dilemmas; before deciding on surgery the patient's nystagmus needs to be carefully diagnosed.

Nystagmus characteristics

Accurate classification of nystagmus is important. If the patient has strabismus (stereoacuity is a useful indicator here, as small squints are difficult to observe in the presence of nystagmus) they should be carefully examined for the presence of manifest latent nystagmus (MLN). In MLN, the nystagmus fast phase beats towards the fixing eye and can be reversed by cover testing. Some cases of CN exhibit similar behaviour, and diagnosis will then rely upon eye movement recordings. The slow phase of CN has an increasing velocity whereas MLN shows decreasing, or constant velocity slow phases. Two further features of MLN are dissociated vertical deviation (DVD) and the absence of a true null zone – instead, nystagmus amplitude tends to damp when the fixing eye is adducted. Many patients with MLN have good vision, and surgical management is usually directed at correcting any strabismus.

Individuals with CN occasionally exhibit more than one null zone; this may be noticeable on testing acuity, but a more stimulating, dynamic stimulus (watching a video for example) will tend to bring out multiple null zones. The patient should also be observed for several minutes to check for periodic alternating nystagmus (PAN) where the null zone shifts with time. PAN is common in albinism[4] and is easy to miss in CN.[5] The presence of multiple null zones or PAN may rule out null zone surgery.

Two salient features of CN complicate measuring the effects of treatment: the nystagmus increases when visual acuity (VA) is measured (thereby reducing VA), and VA tends to improve on repeat visits. Unfortunately, this may still catch some researchers unawares. Studies of CN treatments are strengthened by the inclusion of an untreated control group, or by having measures of visual performance over several visits before treatment,[6] and by an analysis of eye movements. High-quality eye movement recordings not only aid the diagnosis of nystagmus but also allow potential visual acuity to be predicted[7] and objective comparisons to be made.

The CN waveform contains periods when the eyes are moving relatively slowly. Individuals with CN maximise their acuity by placing their fovea at these 'foveation periods' where eye velocity is least.[8] Subjects with albinism (who have no distinct fovea) behave in a similar fashion by consistently using a specific retinal locus[9] corresponding to an area of retina that gives maximum vision. Such strategies suggest that ocular motor control can be precise despite the presence of CN.

Non-surgical options

Any refractive correction that would be normally prescribed should be tried; VA may not improve in the clinic but patients benefit from their refractive correction in a more normal, relaxed environment if a clearer image reduces the 'effort to see'. Base-out prisms (with low minus lenses in pre-presbyopes) may help in cases with a strong reduction of CN on near viewing (it is the convergence, rather than the accommodation that is important[10]). Other therapies such as auditory biofeedback[11] and cutaneous stimulation[12] can reduce CN in some cases. BT therapy, which can be helpful in reducing oscillopsia in acquired nystagmus, has also been suggested for CN.

Surgery

The goals of surgery for CN are to reduce the need for head postures by re-aligning the null zone, or to improve vision by reducing the nystagmus, or both.

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The Kestenbaum–Anderson procedure is an approach for the patient with an established null zone in eccentric gaze. The aim is to shift the null zone to primary gaze. Since the first reports,[13–15] the suggested amounts of surgery have gradually increased in an attempt to reduce the chance of the head posture returning.

Bilateral recessions of both medial recti combined with a Faden suture (or Kestenbaum–Anderson procedure) have been reported to take advantage of a convergence effect.[16] Caution should be exercised where there is potential to induce a divergent strabismus, particularly for near viewing.

Questions that have previously surrounded the type of surgery reported by Atilla *et al.* in this journal are that it may reduce conjugacy in eccentric gaze, which may limit benefits for those with an eccentric null zone. Subjects with a central null zone may be happy to trade reduced binocular vision in eccentric gaze if their CN waveform is improved following surgery. Any technique that reduces the effectiveness of extraocular muscles could affect fine ocular motor control. A simple measure such as saccadic peak velocity and accuracy could provide useful insights here. An interesting feature of Kestenbaum–Anderson surgery is a general dampening of the CN at all gaze angles, as well as a re-location of the null zone.[17,18] Recent work on achiasmatic dogs with CN and see-saw nystagmus has demonstrated that the damping effect can be obtained by performing tenotomies without any resection or recession.[19] This promising finding may deliver useful insights into the effects of surgery.

Summary

Many individuals with CN cope remarkably well. With several different surgical approaches available, there needs to be reasonable certainty that a chosen treatment is the best for an individual patient. It is hoped that continued research into the mechanisms underlying CN will deliver a deeper understanding of exactly how extraocular surgery brings about an improvement in some cases while still being less than perfectly reliable in others.

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