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Case Report

Nystagmus surgery: Our experience

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Introduction

Nystagmus has been an enigma in ophthalmology from the very beginning. It has varying presentations and compensation mechanisms which further complicate this enigma. The authors here present a series of cases of nystagmus which were largely compensated by the face turn. A meticulous approach to investigation and management was followed and it gave remarkable results in all the cases. This article intends to create awareness amongst medical professionals about this compensated nystagmus where effective treatment gives favorable outcome.

Case 1

An 8 years old male child patient presented to the OPD with bilateral involuntary eye movements since birth, and a right face turn apparent since 7–8 months of age. It was not associated with oscillopsia, hearing loss, vertigo or dizziness. There was no history of birth trauma, swelling, mass or injury in the neck region. He grew up with normal developmental milestones. The parent's anxiety was regarding cosmetic disfigurement because of the head turn.

On examination, we discovered a bilateral, conjugate, involuntary, horizontal, right beating jerky nystagmus, in primary gaze. The intensity increased in right gaze, with dampening in left gaze. His Snellen's visual acuity was 20/40 (binocular single vision) with the right head turn, which reduced to 20/80 binocularly in primary gaze because of nystagmus. On occlusion, his visual acuity was 20/200 with right eye and 20/120 with left eye. Cycloplegic refraction showed no significant refractive error. Anterior and posterior segment examination showed no signs suggestive of any sensory deprivation. The head turn was measured using a Goniometer, usually used to measure joint angles by orthopedic surgeons, and found a right head turn of 60° on distance fixation, corresponding with a null point in 60° left gaze [Fig. 1]. Stereoacuity by TNO test was 120 s of arc with a right head turn. Electronystagmography (ENG) confirmed the clinical findings [Fig. 2] suggestive of Idiopathic Manifest Latent Childhood Nystagmus with a compensatory right head turn.

A face turning surgery, to shift the eyes from primary gaze to the direction of the anomalous head posture was planned and a Modified Anderson Operation in which we recessed the

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Fig. 1 – Child with nystagmus (case 1) with right head turn.

left (abducted) eye lateral rectus by 12 mm, and right (adducted) eye medial rectus by 9 mm, using conventional horizontal muscle recession surgery. Post-operatively the child was orthotropic, with a significant improvement of 45° of head turn. The visual acuity also returned back to pre-operative values in primary gaze on 2nd week follow up [Fig. 3]. The intensity of nystagmus in primary gaze was reduced significantly post-operatively and corroborated on ENG.

Case 2

A 6 year old male child patient with myotonic dystrophy presented with abnormal movement of eyeballs, abnormal face turn to left. On examination he had a binocular visual acuity of 20/200 and less than counting finger at 4 feet on monocular testing. His refractive status revealed +0.25 D cyl ax 90° and +0.5 D Sph and +0.5 D Cyl ax 90°. He had bilateral, conjugate, involuntary, horizontal, jerky nystagmus with a left face turn. There was null in 30° of dextroversion. He also had left exotropia of 40 Prism diopter [Figs. 4 and 5]. The child underwent only right lateral rectus recession of 9 mm thus shifting the null to primary gaze and also correcting the exotropia to less than 10 prism diopter [Fig. 6].

Case 3

A 5 year old male child patient presented with abnormal movement of eyeballs with an abnormal face turn to left with a chin depression. The child also had oculocutaneous albinism. On ophthalmic examination his binocular acuity was 20/120. His refractive status was normal. He had bilateral, conjugate, involuntary, horizontal and vertical, jerky nystagmus with a left face turn and chin depression. There was null in 30° of dextroversion and chin depression of 10–20°. The child underwent right lateral rectus recession of 9 mm and left medial rectus recession of 6 mm thus shifting

Fig. 3 – Corrected head turn (case 1).

Fig. 4 – Child with nystagmus (case 2) with left exotropia in primary position.

Fig. 5 – Child with nystagmus (case 2) with left head turn.

Fig. 6 – Case 2 with corrected head turn and orthotropia.

the null to primary gaze. He also underwent recession of both superior rectus by 6 mm. The child was completely orthotropic for primary gaze with full correction of head turn.

The follow up in all three cases was documented for a minimum of 6 months to 1 year and 3 months with consistent results.

Fig. 2 – ENG recording showing a right beating nystagmus in primary gaze, which dampens on left gaze in case 1.

Discussion

Nystagmus is defined as the involuntary rhythmic oscillations of the eyes, with a reported prevalence of approximately 2.4 in 1000.¹ There is significant negative social stigma and relatively poor visual function scores reported with this condition.

The ocular etiologies of anomalous head posture include strabismus, nystagmus, refractive errors, and dissociated ocular deviations. Children with nystagmus will have anomalous head posture due to a “gaze null” (position with respect to the orbit of one or both eyes where the nystagmus intensity is the least and vision is the best) associated with Congenital Nystagmus. An anomalous head posture can also be due to an “adduction null,” where the nystagmus intensity is the least and vision is the best, that is, in the direction of the slow phase.

The surgery to dampen nystagmus is based on 2 principles, immobilization and relocation of the eyes. Immobilization procedures all aim to lessen muscle efficiency and to reduce nystagmus intensity. Their efficiency is unpredictable, and indicated only in the absence of compensatory mechanisms. Relocation procedures were first advocated by Kestenbaum¹ and Anderson² in patients with compensatory head turns. These procedures aim to shift both eyes from an eccentric position of gaze where nystagmus is dampened (null zone) into the primary position.

A compensatory head posture can stabilize nystagmus by placing the eyes at the null point. Head posturing associated with nystagmus can take the form of a face turn, chin elevation, depression, or a head tilt. This can be treated using eye muscle surgery to move the eye position from the null point to primary position.

With a compensatory face turn to the right, the eyes will be shifted to the left, which will be done by moving the right eye out (right medial rectus recession–right lateral rectus resection) and moving the left eye in (left lateral rectus recession and a left medial rectus resection). Parks 5-6-7-8 ‘rule’ (medial recession 5 mm, medial resection 6 mm, lateral recession 7 mm, and lateral resection 8 mm) proposes the amount of surgery for a small face turn. In most cases, however, larger amounts of surgery are needed.³ Eye movements recordings performed on a few patients before and after this surgery have shown that it decreases nystagmus intensity in practically all directions of gaze.⁴ Undercorrection are common for head turns of more than 30°. ^{5–7} and may exceed 10° in some cases.⁷

A 40% increase in surgical dosage has been suggested for patients with larger head turns (Augmented Anderson procedure).⁵ Head postures are improved but patients may be left with a duction deficit.^{5–10}

Children with nystagmus and compensatory head tilt or face turn are often managed conservatively thus depriving them of the advantage following a successful face turning surgery for nystagmus. However proper parent counseling is required to ensure realistic expectations of the outcome.

Conflicts of interest

All authors have none to declare.

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