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Surgical treatment of a patient with congenital nystagmus with compensatory head posture: A case report

Kirurško zdravljenje bolnice s prirojenim nistagmusom s kompenzatorno držo glave – prikaz primera

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Abstract

The correction of compensatory head posture in a congenital nystagmus involves surgical treatment that includes recession and resection of extraocular muscles to move the eccentric null zone to a primary position.

A 39-year-old patient presented with an impaired visual acuity and nystagmus that was present since childhood. She had a left head turn with permanent neck pain. At the examination, best corrected visual acuity was 0.4 in both eyes with her glasses. Correction in the right eye was -6.50-1.50/180° and in the left eye -5.50-2.50/180°. Measured objective and subjective angles of squint were +4°, fusion from -3° to +29°, with the presence of stereo vision. During the cover test the nystagmus was present and it enhanced while covering the eye. Ocular motility was not limited. Fundus examination revealed myopic changes in both eyes. Prisms were prescribed, which were well tolerated by the patient. Also, no apparent head turn was noticed while wearing the prisms.

Nine months later, the patient underwent a Kestenbaum procedure. Retroposition of the lateral rectus muscle with resection of the medial rectus muscle in the right eye and retroposition of the medial rectus muscle with resection of the lateral rectus muscle in the left eye were performed. After the procedure nystagmus dampened the most in the minimal left position, the head was in a straight position. Two years after the procedure nystagmus dampened the most in the primary position, the head was in a straight position. Nine years after surgery and refractive correction with contact lenses, the visual acuity was 0.8-0.9p in both eyes.

The presented case showed that adequate functional and surgical treatment led to a good morphological outcome with improved visual acuity in a patient with congenital nystagmus and a compensatory head posture even in adulthood.

Izvleček

Korekcija kompenzatorne drže glave pri prirojenem nistagmusu zahteva kirurško zdravljenje, ki vključuje recesijo in resekcijo zunajočesnih mišic zaradi premika ničte točke v primarni položaj.

39-letna bolnica je bila obravnavana zaradi slabovidnosti in nistagmusa, ki je bil prisoten od otroštva. Glavo je od otroštva obračala v levo in imela stalne bolečine v vratu. Ob prvem pregledu je bila najboljša korigirana vidna ostrina 0,4 obojestransko z očali. Korekcija na desnem očesu je bila –6,50–1,50/180° in na levem očesu –5,50–2,50/180°. Izmerjena objektivni in subjektivni škilni kot sta bila +4°, fuzija od –3° do +29° s prisotnim stereo vidom. Ob izvajanju testa cover je bil prisoten nistagmus, ki se je med pokrivanjem okrepil. Gibljivost zrkel ni bila omejena. Na očesnem

ozadju so bile ugotovljene degenerativne spremembe zaradi kratkovidnosti. Ob prvem pregledu so ji bile predpisane prizme, ki jih je dobro prenašala. Tudi položaj glave je bil ob nošnji prizem poravnan.

Devet mesecev po prvem pregledu je bila pri bolnici opravljena Kestenbaumova operacija. Napravljena je bila retropozicija zunanje preme mišice in resekcija notranje preme mišice na desnem očesu in retropozicija notranje preme mišice in resekcija zunanje preme mišice na levem očesu. Po posegu je bil nistagmus najbolj umirjen v rahlo levi poziciji, položaj glave je bil poravnan. Dve leti po posegu pa je bil nistagmus najbolj umirjen v primarni poziciji, glava pa je ostala izravnana. Pri bolnici je bila 9 let po operaciji in s korekcijo s kontaktnimi lečami vidna ostrina obojestransko 0,8–0,9p.

Primer je prikazal, da sta ustrezna funkcionalna in kirurška obravnava omogočili dober morfološki rezultat z izboljšanjem vidne ostrine pri prirojenem nistagmusu in kompenzatorni drži glave tudi v odrasli dobi.

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1 Introduction

Congenital nystagmus is an eye movement disorder characterized by involuntary oscillations. Patients with congenital nystagmus often turn the head to place the eyes in a direction of gaze with the least nystagmus (null zone). Children with congenital nystagmus tend to prefer gaze in the direction of null zone to gain optimal visual acuity. When the null zone is located eccentric to the primary gaze, a compensatory head posture is frequently adopted (1-5). The surgical correction of compensatory head posture involves extraocular muscle surgery in each eye to move the eccentric null zone to the primary or straight ahead position (2,6-9), but such surgery is probably carried out by a minority of ophthalmologists (10).

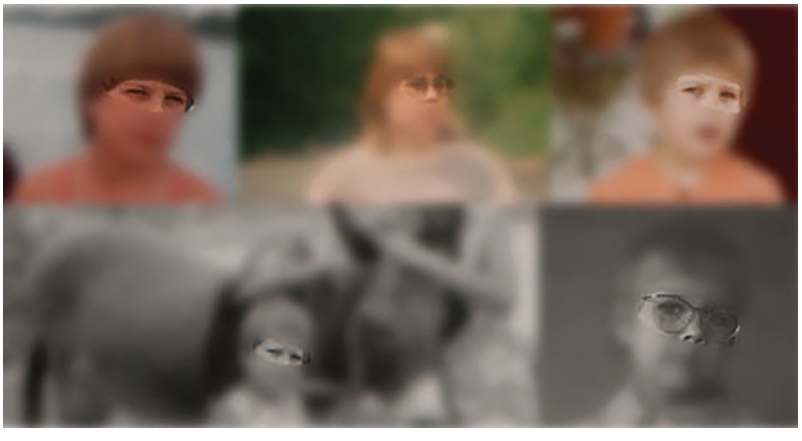

Figure 1: A left head turn and some left tilt adopted by the patient since her childhood.

The purpose of the case report is to present surgical and functional management in an adult patient with congenital nystagmus and a compensatory head posture which led to a good and stable morphological result with improved visual acuity.

2 Case presentation

A 39-year-old patient presented to the Eye Hospital, University Medical Centre Ljubljana with an impaired visual acuity and a horizontal nystagmus that was present since childhood. Since her

childhood, her head was turned to the left and she had permanent neck pain. She did not report any other health problems. At the first examination, best corrected visual acuity was 0.4 in both eyes with her glasses with a left head turn and some left tilt. Correction in the right eye was -6.50-1.50/180° and in the left eye -5.50-2.50/180°. Measured objective and subjective angles of squint were +4°, fusion from -3° to +29°, with the presence of stereo vision. The patient adopted a left head turn and some left tilt (Figure 1), which allowed her to use her null zone in the right gaze in which nystagmus dampened the most and visual acuity was the best. During the cover test, the nystagmus was present and it enhanced while covering the eye. Ocular motility was not limited. Fundus examination revealed myopic changes in both eyes. At the first examination, prisms were prescribed 15Δ base nasally in front of the right eye and 15Δ base temporally in front of the left eye. Prisms were well tolerated by the patient. Also, no apparent head turn was noticed while wearing the prisms.

Nine months after the first examination, the patient underwent surgery - Kestenbaum procedure. Retroposition of the lateral rectus muscle (6.5 mm) with resection of the medial rectus muscle (5.5 mm) in the right eye and retroposition of the medial rectus muscle (4.5 mm) with resection of the lateral rectus muscle (7.5 mm) in the left eye were performed. One week after the surgery, the nystagmus dampened the most in the slight left gaze, the head position was normal. Two years after the procedure the nystagmus dampened the most in the primary position. Contact lenses which she did not wear before, were prescribed to the patient one year after the procedure. Her visual acuity improved slowly during following years. Nine years after

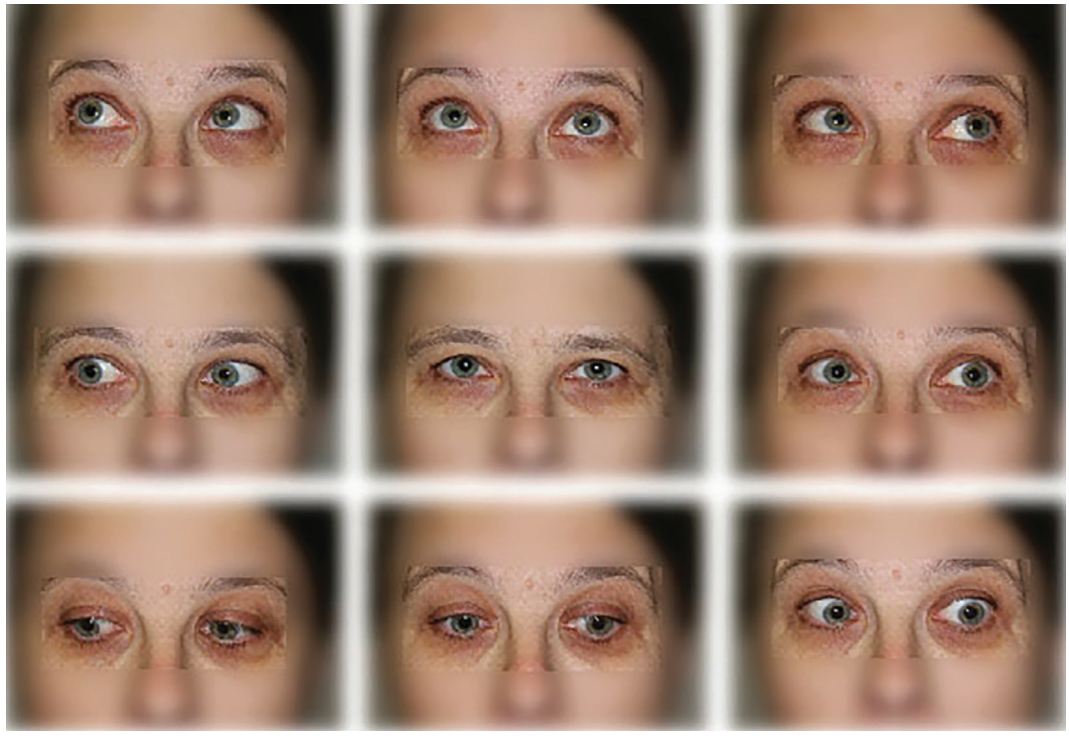

Figure 2: The nine cardinal positions of gaze in a patient 9 years after surgery. In the primary position (image in the middle) the patient looks straight and nystagmus is almost absent. In other cardinal positions of gaze nystagmus is still present.

surgery and refractive correction with contact lenses, the visual acuity was 0.8-0.9p in the right and left eye. Null zone was in the primary position (Figure 2).

3 Discussion

In the presented adult case, head position corresponded to the null zone, similarly as in most patients with infantile nystagmus (4). Attempts to eliminate compensatory head posture in infantile nystagmus with an eccentric null zone with various extraocular muscle surgeries have been tried, but the Kestenbaum procedure as modified by other authors, is probably now the most commonly performed (11).

Surgical procedures for the correction of compensatory head positions in patients with congenital nystagmus began with the independent reports of Anderson, Goto and Kestenbaum in the early 1950s (7,12-15). Since its initial description, the procedure has been modified by Parks, Calhoun and Harley, and Taylor and Jesse (7,15). Anderson proposed to weaken the horizontal rectus muscles that are activated during the slow phase of the nystagmus because they were thought to have a greater tone than their antagonists. Therefore, in a patient with a concordant left head turn, an Anderson-like procedure would consist of a supralarge recession of the right lateral rectus and left medial rectus muscles (4,13,16). A Kestenbaum (4,12) procedure combined 5 mm recessions with resections of the antagonist muscles; however, the initial procedure led to a high rate of hypocorrections and Pratt-Johnson increased the amount of muscle surgery. The former 5 mm recessions and resections were increased to 10 mm (4). Parks modified the Kestenbaum technique to the 5-6-7-8 or the "Classic Maximum" procedure in which a recession of 7 mm was performed to the lateral rectus, a resection of 6 mm to the medial rectus of the abducted eye, a recession of 5 mm to the medial rectus, and finally a resection of 8 mm to the lateral rectus of the adducted eye (4,17). Similar surgery was performed in the presented case.

It has been demonstrated that surgery on all four horizontal muscles was a safe and effective method for altering the compensatory head position in congenital nystagmus (7,8,18,19). It was indicated that varying degrees of improvement may be expected in the studies that included mostly paediatric patients as well as some adults (6-9,18), but complete elimination of the head turn occurred in only 6 of 19 patients (32%) in one older study (18). Newer studies report 26 of 36 patients (72%) with a residual head turn not exceeding 10° after surgery (6) and 34 of 44 (77%) cases with a normal head posture or head turn of less than 5° after surgery (9). The most common anomalous head position following Kestenbaum surgery was usually in the same direction as was present prior to surgery (15). Similarly, in the presented case nystagmus dampened the most in the slight left gaze after surgery and the patient reported to be confused about the head position. Probably the confusion was due to the discrepancy between neck proprioception and new eyeball position, but later this discrepancy has been resolved. It is possible for new or recurrent compensatory head postures to reappear years after the initial treatment, which was reported in the study of four children after Anderson-Kestenbaum surgery (15). But since the head position was still stable 9 years after surgery and the visual acuity improved, new null zone

development in the presented case is probably unlikely.

Compensatory head position was more often reported as horizontal in the literature, but it may also be vertical or take the form of a tilt even though the nystagmus itself is horizontal (10). In the presented case, there was a combination of predominant left head turn and some left tilt, which both improved after surgery on the horizontal rectus muscles. Head tilt and vertical head turns may improve with only weakening surgery of the horizontal rectus muscles in those cases in which the horizontal head turn predominates, at least when other components are not severe. This approach is thought to work by moving the blockage point to the primary position where the cyclovertical muscles' actions are weaker (4,20). Arroyo-Yllanes et al. treated 21 patients with horizontal nystagmus and compensatory head position in all three axes with a predominant head turn. In all cases a modified Anderson procedure was performed - that is, 2 mm retroequatorial recessions of the horizontal yoke rectus muscles responsible for the blockage position, plus corrective surgery for strabismus when needed. The three components of the compensatory head position were improved by surgery of the horizontal yoke rectus muscles only (20). In addition to shifting the nystagmus null zone, the surgery may also broaden the null zone according to the literature (21).

In congenital nystagmus with a neutral zone in dextro- or levoversion binocular functions were often preserved (22). Similarly, the presented case had preserved stereo vision and had good visual acuity with contact lenses which were prescribed due to a higher degree of astigmatism.

In conclusion, the presented case confirms the benefits of Kestenbaum procedure to move the eccentric null zone to the primary position in the management of the patient with congenital nystagmus and a compensatory head posture. The important message of the case is that Kestenbaum procedure was successful even in adulthood. Consequently, it reduced neck pain, eliminated the possibility of additional problems arising from long-term abnormal contracture of the neck muscles and improved cosmesis. Additionally, refractive correction with contact lenses led to improved visual acuity.

Consent was obtained from the patient for the publication of this case presentation.

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