

Acquired anomalous head posture following loss of vision in one eye

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ABSTRACT.

Background: We studied the anomalous head postures (AHPs) of five monocular viewing patients and investigated the possible causes and the appropriate surgical strategies to correct each condition.

Methods: Five patients with acquired visual loss in one eye and associated head tilt and/or turn were examined and treated for correcting the head posture according to the etiology of their respective AHPs.

Results: Three types of anomalous head position have been detected: head tilt related to cyclotropia, face turn associated with adduction blocked monocular nystagmus, and face turn to centre the visual field. Surgical plans were prepared according to the mechanism of the AHP in question. After surgery, all patients showed a marked reduction of the head tilt, except one who had a recurrence of the face turn 1 week postoperatively. Mean follow-up time was 19 months.

Horizontal transposition of the vertical muscles for correcting cyclotropia offered stable normalization of the AHP in three monocular viewing patients with head tilt, and represents a safe, viable and easy alternative to the Harada Ito procedure. Horizontal recession of the medial rectus of the fixing eye minimized the abduction nystagmus and relieved the need to adduct the fixing eye and subsequently rotate the head toward the fixing eye in one patient. Recurrence of the AHP occurred in one patient.

Conclusion: Different mechanisms may account for AHP in monocular viewing patients. Different surgical procedures may be used to correct the anomalous position. Careful patient selection and etiological diagnosis of AHP is required prior to developing a surgical strategy.

Key words: amblyopia – strabismus surgery – anomalous head posture.

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Anomalous head posture (AHP) may be associated with a variety of ophthalmologic conditions including strabismus, nystagmus, astigmatism, and visual field defects. In these situations, the AHP is an adaptive or compensatory mechanism to improve visual acuity and/or binocularity. It may be either congenital or acquired and is almost always seen in patients with functional vision in both eyes.

Rarely, abnormal head posture has been reported following visual loss in one eye (Kushner 1979; Helveston et al. 1985; Kushner 1985; and Morad & Nemet 1998). Helveston et al. reported five patients who developed esotropia in the remaining eye after unilateral enucleation in the first few weeks to months of life (Helveston et al. 1985). Esotropia was associated with a face turn toward the opposite side and abduction nystag-

mus with a null point in extreme adduction. An intact globe-ocular muscle relationship, even in a blind eye, may have a stabilizing effect on the fellow eye in the first few weeks to months of life, and this should be considered before enucleation is carried out. Kushner proposed that enucleation was not the relevant element, suggesting the adduction fixation preference as a possible cause of AHP (Kushner 1985). In a case report, Morad and Nemet proposed the hypothesis that torticollis following loss of vision in one eye was probably the result of a subtle abduction nystagmus that occurred only in extreme positions of gaze (Morad & Nemet 1998). More recently, Goltz et al. presented 52 unilaterally enucleated children and adults without nystagmus who had been enucleated at an early age due to retinoblastoma and who went on to exhibit a head turn unrelated to the presence of nystagmus (Goltz et al. 1997). They suggested that the direction of the head turn is 'adaptive' because occlusion by the nose in the lower contralateral field is eliminated by the face turn.

We examined five subjects with significant AHP following visual loss in one eye. Three had a marked head tilt toward the shoulder of the non-seeing eye, possibly related to excyclotropia of the fixing eye. Two presented face turn toward the fixing eye, presumably due to adduction blocking nystagmus.

We report the results of reducing AHP utilizing one of two different surgical strategies on the sound eye: horizontal nasal transposition of the inferior rectus muscle for head tilt, or medial rectus recession of the fixing eye for face turn.

Table 1. Summary table of Patients 1–5

Case	Age	AHP (years)	Degree Preop	Associated findings	VA RE VA LE	Surgery	Follow-up (months)	Degree Postop
1	28	left tilt	12	Re excyclotorsion Le Hypertropia (DVD)	20/20 CF	horizontal nasal transposition inferior Rectus RE	24	0
2	left tilt 15		9	RE Excyclotorsion LE Excyclotorsion (DVD)	20/20 0/400	Horizontal nasal transposition feior Rectus	11	
3	58	left tilt	10	LE DVD	20/20 DF	Horizontal nasal transposition Inferior Rectus RE	16	0
4	12	right face turn	25	Nystagmus RE	20/20 enucl.	6 mm recession MR RE	0	5
5	42	left face turn	20	Nystagmus RE	20/20 CF MR LE	6 mm recession	24	0–4

AHP: anomalous head posture

CF: count fingers

enucl.: enucleated

Material and methods

Five consecutive patients who had lost vision in one eye and then developed an AHP were included in this study. They were seen by one of us (P.N.) during the period 1995–99. All underwent a complete ophthalmological evaluation, including angle measurement using the Krinsky test (placing a prism in front of the sound eye) and motility examination using version and duction observation in the nine positions of gaze. Unilateral Maddox rod testing was performed in all patients to assess subjective cyclotorsion. The Maddox rod lens was placed before the fixing eye in a horizontal direction. The patient's head was maintained in the forced primary position. The rod was rotated until the patient was able to perceive the image in a true vertical direction. The patient's head tilt was measured by photographing the tilt and then superimposing a goniometric scale on the photograph.

Face turn was assessed using a professional magnetic compass mounted on a helmet. While wearing the helmet, the patient was asked to fixate on a target placed in the north position. The examiner recorded the degrees of face turn indicated by the compass arrow pointing constantly to the north. Neurologic evaluation was performed in all patients and included computer tonography (CT) and magnetic resonance imaging (MRI)

of the brain. Neurologic and orthopedic examinations were performed in all patients. No muscular or spine anomalies were diagnosed that would account for the AHP.

Case Reports

Case 1

A 28-year-old man developed a 12 degree left head tilt following traumatic retinal detachment of the left eye many years prior to being seen in our clinic. The visual acuity of the right eye was 20/20 and in the left eye was counting fingers. Motility examination revealed a low frequency small pendular excyclotorsional movement with a variable hypertropia of the left eye measuring 10/15 degrees. No oblique muscle dysfunction was seen. The right eye did not demonstrate nystagmus or any unusual movement except for some difficulty in maintaining fixation for more than 5 seconds. Fundus examination of the right eye revealed excyclotorsional displacement of the macula. Review of pictures from a family album did not show any anomalous head posture prior to the retinal detachment. No other ophthalmologic problems were noted.

Horizontal nasal transposition of the inferior rectus of the right eye was performed through a nasal fornix incision. The tendon was isolated with a double-armed 6.0 vicryl suture, dissected without

cleaning the capsule palpebral attachments and transposed nasally with the temporal end just nasal to the previous insertion and the medial end 1 mm closer to the limbus. At the first follow-up visit, a marked improvement of the head tilt was seen. Posture remained stable after a follow-up period of 2 years.

Case 2

A nine-year-old boy developed a 15 degree left head tilt following a traumatic corneal perforation at age 6 years. A secondary cataract and glaucoma occurred in that eye. Visual acuity was 20/20 in the right eye and 20/400 in the left eye. Examination revealed a low frequency small pendular excyclotorsional movement of the left eye with 8 degrees of left exotropia and a variable vertical deviation of 10–15 degrees. No nystagmus or oblique muscle dysfunction was detected in the right eye. Fundus examination in the right eye disclosed excyclotorsion of the macula. There was no previous history of ocular problems. Ophthalmologic evaluation at age 5 years revealed a visual acuity of 20/20 in each eye with a normal binocular response to the Lang stereo test.

Horizontal nasal transposition of the inferior rectus of the right eye was performed as described in Case 1. The resulting marked improvement of the head tilt had remained stable by the end of a follow-up period of 11 months.

Case 3

A 58-year-old man developed a 10 degree left head tilt following a central retinal artery occlusion of the left eye 7 years earlier. Visual acuity was 20/20 in the right eye and finger counting in the left eye. Motility examination revealed a movement of the left eye with a variable vertical deviation of 10–15 degrees, similar to that associated with dissociated vertical divergence (DVD). No torsional displacement of the macula was seen by fundus examination. Past medical history showed no previous ocular problems. A review of pictures from a family album did not show any APH prior to the retinal vascular insult. We assumed that despite the absence of obvious torsion of the fundus, the head tilt was related to excyclotorsion of the right eye.

Horizontal nasal transposition of the inferior rectus of the right eye was performed as described in Case 1. The first follow-up examination showed the AHP to have normalized. This had remained stable by the end of the 16 month follow-up period.

Case 4

A 12-year-old girl developed a 25 degree right face turn in early childhood. Her left eye was diagnosed with a congenital Peter's anomaly and became phthisical following several surgical procedures for associated glaucoma. Her right eye was normal. There was no history of problems during pregnancy or delivery and she was not premature.

The patient wore an ocular prosthesis covering the left phthisical eye. Vision in the right eye was 20/20. The right eye demonstrated a high frequency unsustained horizontal nystagmus in all gaze directions except when the patient was rotating the head toward the fixing eye. We assumed the patient acquired her head position to eliminate this nystagmus. The remainder of the ophthalmologic examination in the right eye was unremarkable and no cycloplegic refractive error was discovered. Her monocular visual field in the right eye was full.

In an attempt to move the fixing eye laterally and minimize the nystagmus, we performed a 6 mm recession of the medial rectus of the right eye through a fornix incision. A week later, no improvement of the AHP was noted. Surgery did not appear to have created any horizontal imbalance after 20 months of follow up.

Case 5

A 42-year-old woman developed a 20 degree left face turn following a traumatic macular hole 15 years earlier. Visual acuity was 20/20 in the left eye and finger counting in the right eye. The patient demonstrated a DVD-like movement in the right eye. An intermittent high frequency unsustained horizontal nystagmus in the left eye was noted in all gaze fields except when the left eye was adducted. There was no previous history of ophthalmologic problems.

To minimize the nystagmus, a 6 mm recession of the medial rectus of the left eye through a fornix incision was performed. During the first two days following the procedure the AHP was significantly improved. This improvement had remained stable when the patient was last seen 2 years postoperatively. Surgery did not create any horizontal imbalance.

Results

No differences in AHP were detected between Case 4, where the patient suffered visual loss in infancy, and the other cases, where loss of vision was acquired later in life.

In our series, two surgical procedures were performed on the fixing eye: horizontal nasal transposition of the inferior rectus in Cases 1, 2 and 3, and recession of the medial rectus in Cases 4 and 5.

One week postoperatively, all cases reported a normalization of the head position, except Case 4 who showed no improvement at the first follow-up visit. The mean follow-up was 19 months (± 5.57 months). None of patients experienced any worsening of the head position during the follow-up period. The operation did not affect the amount of DVD of the non-fixing eye in Cases 1, 2 and 3. None of the patients or their parents requested or accepted a second procedure for correcting the DVD of the non-fixing eye. Surgery did not create any horizontal deviation in Cases 4 and 5.

Discussion

Head tilt and face turn are uncommon findings following visual loss in one eye. In our series, acquired anomalous head posture was noted immediately after the loss of vision in one eye in patients who had no previous history of strabismus,

nystagmus or anomalous head posture. Two of three patients (Cases 1 and 2) demonstrated head tilt which seemed to be correlated with objective excyclotorsion of the fundus of the normal seeing eye. Patients 1, 2 and 3 showed a vertical deviation of the non-seeing eye similar to DVD. In 1995, Kutluk et al showed that this phenomenon could occur in association with sensory heterotropia and was more common if vision was lost before age 7 years, but could also occur in adults (Kutluk et al. 1995).

DVD is often associated with an anomalous head tilt that may be directed toward or away from the side of the hyperdeviated eye (Betchel et al. 1996; Santiago & Rosenbaum 1998). In some patients, head tilting may represent a compensatory means of controlling or reducing the extent of DVD. Jampolsky recognized the head tilt pattern in DVD in which a hyperdeviation of either eye increases or becomes more evident when the head is tilted to the opposite side (Jampolsky 1986).

Other patients with DVD have a head tilt pattern characterized by a hyperdeviation that increases or becomes manifest when the head is tilted to the same side (Crone 1954). According to Jampolsky, when fixation with one eye predominates in DVD, tonic fixational innervation to the contralateral superior rectus muscle can eventually lead to a contracture of that muscle (Jampolsky 1995). In this setting, surgical recession of a tight superior rectus muscle can reduce or eliminate the compensatory head tilt.

Head tilt associated with DVD of the non-seeing eye has, in our opinion, a peculiar mechanism. When the poorer seeing eye develops a dissociated sursum-extorsional movement, it sends a consensual sursum-intorsional stimulus to the fixing eye. To correct this incyclo-rotation, the head is tilted toward the non-fixing eye, thereby evoking compensatory excyclotorsion of the fixing eye.

Rectus muscle transposition was effective in correcting the acquired head tilt in all three patients in which it was attempted. Medial rectus recession was effective in correcting the AHP when it was an adaptive mechanism to minimize horizontal nystagmus. If the AHP had developed as an adaptive mechanism to avoid seeing the nose and centre the visual field, surgery was unsuccessful. Thus, careful determination of the etiology of the AHP will result in an appropriate treatment strategy.

This is the first reported series addressing the treatment of AHP following loss of vision in one eye. However, the efficacy of selected surgery is hard to evaluate in a case report series and larger studies are needed to forward hypothesis on appropriate treatment strategies.

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