Oblique muscle surgery for treatment of nystagmus with head tilt

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BACKGROUND Patients with nystagmus may adopt an abnormal head posture if they have a null zone in eccentric gaze. These patients uncommonly present with torticollis due to a null zone when the head is tilted. We describe the results of surgery on the oblique muscles to improve the abnormal head posture in this condition.

METHODS This was a retrospective review of patients who had head tilts due to null zones of nystagmus. Surgery consisted of an anterior 50% tenectomy of the superior oblique tendon on one side and recession of the inferior oblique muscle to a position 6 mm posterior to the insertion of the inferior rectus muscle on the contralateral side. The patients' clinical histories and outcomes were reviewed.

RESULTS Six patients underwent the procedure. Of these, four had infantile nystagmus syndrome and two were born prematurely and had histories of intraventricular hemorrhages. Five of the patients had previous Kestenbaum surgery that corrected the horizontal component of their abnormal head postures. Age at time of surgery for the head tilt ranged from 3 to 13 years. Postoperative follow-up ranged from 1.5 to 3 years. The preoperative head tilts ranged from 25° to 45° (mean, 39°). The postoperative improvement ranged from 20° to 40° (mean, 28°). One of the patients with a history of intraventricular hemorrhage required additional surgery for strabismus unrelated to nystagmus.

CONCLUSIONS Anterior tenectomy of the superior oblique tendon combined with contralateral recession of the inferior oblique muscle improved head tilts related to a null zone of nystagmus. (J AAPOS 2012;16:322-326)

Patients with nystagmus frequently experience a decrease in their abnormal eye movements in eccentric gaze. The area of minimal movement is called a null zone. Placing the eyes in the null zone provides optimal visual acuity. Because of this, patients often develop a compensatory head posture to maximize their vision by keeping their eyes in the null zone. Null zones are most commonly horizontal, less commonly vertical, and least commonly torsional.

Surgery to correct abnormal head postures has been well described for horizontal and vertical head positions.[1-5] The procedure consists of a combination of recession, resection, or transposition of the extraocular muscles to shift the zone of minimal nystagmus from eccentric to primary gaze. This allows patients to function with their maximal acuity without the need to turn their heads. Few reports have been published concerning surgery for head tilts due to nystagmus. Surgical procedures employed in these reports have included slanting or transpositions of the rectus muscles, or procedures involving the oblique muscles.[1,6-10] We describe the results of surgery on the oblique muscles to treat such patients.

Methods

This retrospective study was approved by the Washington University School of Medicine Institutional Review Board and was compliant with provisions of the Health Insurance Portability and Accountability Act. The records of six patients evaluated from 2005 through 2009 who were diagnosed with head tilt due to null zones of nystagmus and treated with oblique muscle surgery were reviewed. The patients underwent complete ophthalmologic examinations pre- and postoperatively, including clinical assessments of visual acuity, pupillary response, extraocular movements and nystagmus, cycloplegic refraction, and examination of the anterior segment and posterior pole. Best-corrected visual acuity was measured with the patients' heads in the preferred postures. The abnormal head posture was recorded as the maximal angle measured with a goniometer when the patient was fixating on the smallest identifiable target on a distant eye chart. Age at surgery, other systemic and ocular diagnoses, and duration of follow-up were recorded.

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Submitted July 31, 2011.
Revision accepted May 6, 2012.

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1091-8531/$36.00
http://dx.doi.org/10.1016/j.jaapos.2012.05.005

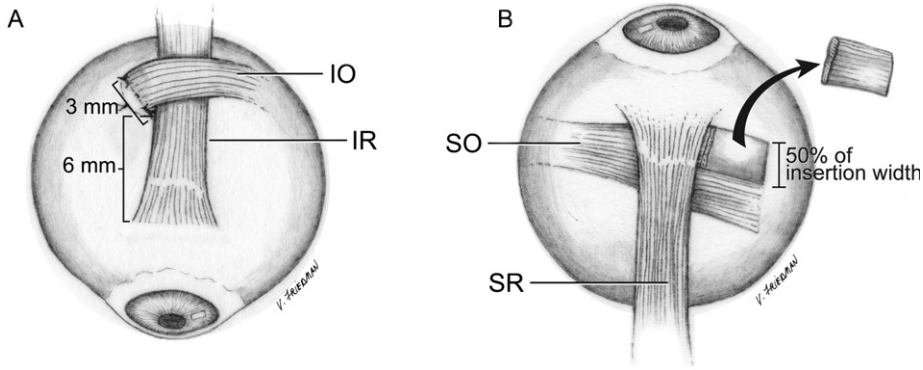

FIG 1. A, Inferior oblique muscle recession, left eye, surgeon's view. The inferior muscle has been cut near its insertion. Its anterior edge is reattached 6 mm posterior to the lateral border of the inferior rectus muscle insertion. The posterior fibers are reattached at a 45° angle posteriorly and laterally from the anterior insertion. B, Superior oblique tenectomy, right eye, surgeon's view. The fibers of the superior oblique tendon are disinserted from the anterior border of the insertion to 50% of the width of the insertion. The anterior 50% of the tendon is then removed between the insertion and the lateral border of the superior rectus muscle. *IO*, inferior oblique muscle; *IR*, inferior rectus muscle; *SO*, superior oblique tendon; *SR*, superior rectus muscle.

The surgical procedure consisted of a superior oblique tendon anterior 50% tenectomy and a contralateral left inferior oblique muscle recession. For patients with right head tilts, surgery was performed on the right superior oblique tendon and the left inferior oblique muscle, and oppositely for patients with left head tilts. The inferior oblique recession was performed through an inferotemporal fornix incision. The inferior oblique muscle was hooked with a small hook and brought forward. The connective tissue was dissected and the inferotemporal quadrant was inspected to confirm that the entire muscle was hooked. The muscle was cut near the insertion and a double-armed 6-0 polyglactin 910 absorbable suture was placed on the cut end. The anterior suture was reattached to the globe 6 mm posterior to the lateral border of the inferior rectus muscle, and the posterior suture was placed at a 45° angle 3 mm posteriorly and laterally away from this point (Figure 1A). The superior oblique tenectomy was performed through a superotemporal fornix incision. The superior rectus muscle was hooked and the insertion and lateral border were exposed. The tendon insertion was identified with a small hook, and the width of the insertion was visualized. An anterior 50% tenectomy was performed by disinserting the anterior 50% of the tendon and then resecting the anterior one half of the tendon between the insertion and the lateral border of the superior rectus muscle (Figure 1B).

Results

Six patients aged 2 to 13 years underwent the procedure (Table 1). Of these, four had infantile nystagmus syndrome and two were premature infants who had intraventricular hemorrhages during the neonatal period. One patient had cerebral palsy and had additional strabismus surgery that was unrelated to the nystagmus. The patients with infantile nystagmus syndrome had uniplanar horizontal nystagmus that presented within the first few months of life, normal or near-normal visual acuity, normal visual-evoked potential responses, and no structural abnormalities of the optic nerves or retinas. The patients with histories of intraventricular hemorrhages had horizontal pendular nystagmus, with no retinal abnormalities associated with premature birth. None of the patients had vertical strabismus or fundus torsion preoperatively.

Of the six patients, five initially had Kestenbaum procedures (bilateral recessions and resections of the horizontal rectus muscles) that improved the horizontal component of their abnormal head postures. The procedure described herein was used to treat head tilts that either persisted or developed after correction of the horizontal null zone in these five patients. The nystagmus in all of the patients was minimal with the preferred head tilt and increased with the head held straight.

Three of the patients were treated for amblyopia and three wore spectacles to correct refractive errors.

Preoperative head tilts ranged from 25° to 45° (mean, 39°). Mean follow-up time after surgery was 2.2 years (range, 1.5-3 years). The postoperative improvement ranged from 20° to 40° (mean, 28°). All of the patients demonstrated decreased nystagmus in the primary position with the head straight postoperatively, and increased nystagmus with forced head tilts. Patient 4 had a small hyperphoria that was not symptomatic and that did not change in side gazes. None of the other patients developed a vertical deviation in primary gaze or in eccentric gaze positions (including head tilts). None of the patients had horizontal deviations or fundus torsion at the last follow-up examination. The visual acuity improved postoperatively in three patients and did not decrease in any patient. Four of the patients demonstrated fusion (three by prism vergence testing and one by stereopsis).

Discussion

The most common abnormal head posture in patients who have nystagmus with a null zone is a horizontal head turn.