Anterior and nasal transposition of the inferior oblique muscles in patients with missing superior oblique tendons

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INTRODUCTION Patients with missing superior oblique (SO) tendons present with overelevation/underdepression in adduction. Unilateral cases often exhibit abnormal head postures, whereas in bilateral cases, there may be a marked V-pattern with upgaze exotropia. These patients may have craniosynostosis.

METHODS Nine children with unilateral (n = 2) or bilateral (n = 7) absent SO tendons underwent anterior and nasal transposition of the inferior oblique (IO) muscles, some in combination with horizontal rectus recession for horizontal strabismus. They were evaluated 6 to 46 months postoperatively for alignment and oculomotor examination. Cyclodeviations were not evaluated in most children.

RESULTS Postoperatively, all patients improved. Both unilateral cases were orthotropic with no abnormal head posture. In the bilateral cases, vertical deviation in adduction and exotropia in upgaze had largely cleared, although some symptoms remained, most notably vertical deviation in side gaze (3 patients) and V-pattern esotropia in downgaze (2 patients). A patient missing both SO tendons as well as the left superior rectus muscle, who had the anterior and nasal transposition on the right side only, remained with 25$^{\Delta}$ left hypotropia.

CONCLUSIONS Anterior and nasal transposition of the IO muscle reduces overelevation in adduction and helps eliminate or reduce divergence of the eyes in upgaze, but esodeviation may persist in downgaze. This procedure was most effective in unilateral absence of the SO tendon. It is likely to benefit patients with severe congenital fourth nerve palsy in which standard IO muscle weakening procedures have been ineffective. (J AAPOS 2007;11:29-33)

The absence of superior oblique (SO) muscle tendons has been reported in patients showing signs of severe congenital fourth nerve palsy[1] as well as in patients with craniosynostosis.[2-4] In patients with severe congenital fourth nerve palsy, Helveston and colleagues[1,5] described abnormalities in the SO muscle anatomy ranging from redundancy of the tendon to complete absence of the muscle. Craniosynostosis syndromes are known to be associated with extraocular motility problems secondary to abnormalities of the orbit and/or abnormalities of the extraocular muscles themselves.[4,6,7] Abnormalities in all the extraocular muscles have been reported and have included absent or tenuous tendons, abnormal insertion, or abnormality in the muscular microanatomy.[3,4,6,8] Absent or abnormal insertion of the SO tendon is the most common anatomical abnormality of the extraocular muscles in these patients.[2,9] Patients with absent SO tendons are expected to present with overelevation in adduction (traditionally termed "IO overaction"), underdepression in adduction ("SO underaction"), excyclodeviation, head tilt in unilateral cases, and V-pattern strabismus in bilateral cases.

The management of motility problems caused by absent SO tendons is difficult, particularly in patients with craniosynostosis syndromes. In these patients, most known IO-weakening procedures are not effective in dealing with the overelevation in adduction and/or eliminating the V-pattern strabismus.[10] Transpositions of the vertical and horizontal rectus muscles may improve horizontal alignment but typically make the cyclodeviation worse. Anterior and nasal transposition of the IO muscle has been an effective procedure in the management of refractory overelevation in adduction and exodeviation in up gaze.[11,12] Here we report on the results of this procedure in the management of 9 patients with absent SO tendons.

Materials and Methods

A retrospective chart review was performed to identify patients with one or both absent SO muscle tendons who had undergone

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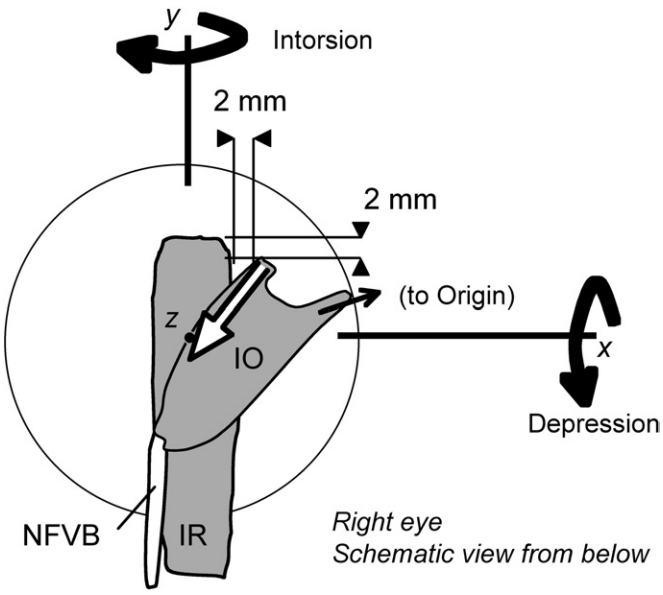

FIG 1. Schematic representation of the IO muscle (IO) in its new position after anterior and nasal transposition, 2 mm nasal and 2 mm posterior to the insertion of the IR. The neurofibrovascular bundle (NFVB) acts as functional origin of the anteriorized IO, rendering it an antielevator and intorter. The white arrow indicates the direction of force on contraction. The letters x, y, and z refer to the axes of Fick.

the anterior and nasal transposition procedure and who had at least 6 months of follow-up. Nine patients who met these criteria were found, ranging in age from 13 months to 10 years (median age, 2.1 years) at the time of the procedure. Data collected from the medical records included age, diagnosis, preoperative and postoperative oculomotor and alignment examination, surgery report, and length of follow-up. This retrospective research was performed in compliance with HIPAA regulations and approved by the Institutional Review Board of the University of Texas Southwestern Medical Center, Dallas.

At the time of the surgery, the absent tendon attachment to the globe was confirmed with the exaggerated traction test[13] and thorough exploration both superotemporally and superonasally. The SO tendon could not be found in one or both eyes in all 9 patients, leading us to conclude that at least the tendon insertion of the SO muscle was absent in these cases. The technique of the anterior and nasal transposition was described elsewhere.[11,12] Similar to the procedure for anterior transposition of the IO muscle, the lateral rectus muscle was isolated with a muscle hook and the eye rotated nasally and superiorly. After isolating the IO muscle with a muscle hook, it was disinserted and reattached to the sclera with the posterior-temporal fibers attached typically 2 mm nasal and 2 mm posterior to the nasal extent of the insertion of the inferior rectus muscle (Figure 1). We currently use permanent sutures made from 6-0 polyester (Mersilene; Ethicon, Somerville, NJ) or nylon (Supramid; S. Jackson, Alexandria, VA) to secure the IO in place. In earlier cases of this series absorbable sutures made from polyglactin 910 (Vicryl; Ethicon, Somerville, NJ) were used. In patients with a significant horizontal deviation in the primary position, recession of one or both medial rectus muscles (for esodeviation) or lateral rectus muscles (for exodeviation) was performed concurrently with the anterior and nasal transposition.

Results

Preoperatively, 2 patients had unilateral missing SO tendons and showed markedly abnormal head posture. The affected eye in both patients showed hypertropia in contralateral gaze (measuring 25^{Δ} and 6^{Δ}) with overelevation in adduction (one eye) and underdepression in adduction (both eyes), but no significant deviation of the eyes in the primary position. The remaining 7 patients had been diagnosed with forms of craniosynostosis (ie, Apert syndrome, Crouzon syndrome, or Pfeiffer syndrome) and had bilateral missing SO tendons. These patients all showed marked V-pattern horizontal deviation with exotropia in upgaze. In primary gaze, 4 patients were exotropic, 1 esotropic, and 2 approximately straight; the patients with horizontal deviation in primary gaze received lateral rectus or medial rectus recession concurrently with the anterior and nasal transposition. All 7 showed marked overelevation in adduction, whereas most also showed underdepression in adduction.

For 3 of the 9 total patients, missing SO tendons were established at previous extraocular muscle surgery, whereas for 6 patients, the missing SO tendons were confirmed during the anterior and nasal transposition procedure. The anterior and nasal transposition procedure was performed unilaterally in the 2 patients with unilateral missing tendons and in 1 patient in whom both SO tendons as well as the left superior rectus (SR) muscle were found missing. The other 6 patients underwent the anterior and nasal transposition procedure in both eyes.

Median follow-up was 8.2 months (range, 6-46 months). At follow-up, both patients with unilateral missing SO tendons were orthotropic in all gaze positions; one showed a very small (5 degrees) residual head tilt, whereas the other showed 6^{Δ} vertical deviation when tilting the head in the ipsilateral direction; this patient also showed a minimal (grade −1) underelevation in adduction (see Figure 2; indicated as "overcorrected"). Figure 2 shows the distribution of overelevation in adduction before and after the procedure for all eyes involved, as well as a classification of change. Of the patients with bilateral missing SO tendons, overelevation in adduction was eliminated in most eyes. In one eye, the problem appeared worse at follow-up; this patient was recommended to undergo exploration of the IO muscle to examine whether it had slipped. The 2 eyes that remained unchanged (Figure 2) were of a single patient who showed great improvements in deviation and currently is undergoing follow-up. The effects on underdepression in adduction are summarized in Figure 3. These symptoms were eliminated or significantly improved in 11 eyes. One patient showed a slight overdepression in adduction in both eyes (indicated as "overcorrected" in Figure 3). This patient underwent additional surgery (recession of the lateral rectus muscle with infraplacement) and seemed to be doing better shortly after

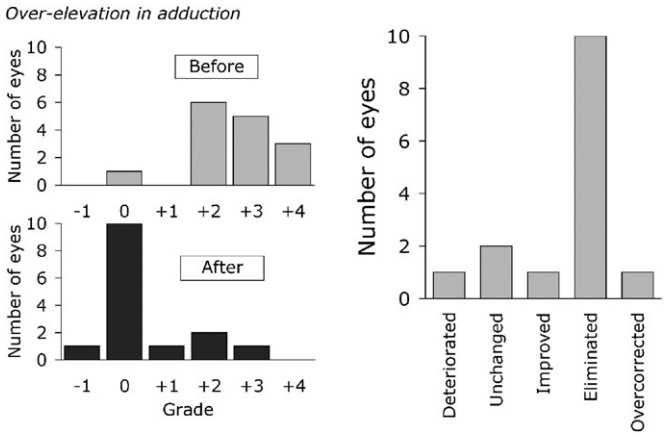

FIG 2. The effects of anterior and nasal transposition on overelevation in adduction using the standard grading scale before (top left panel) and after (bottom left panel). The right panel classifies the change in each eye, where a 1 or greater grade change is considered significant. Eyes with no problems preoperatively and postoperatively are not represented in this graph.

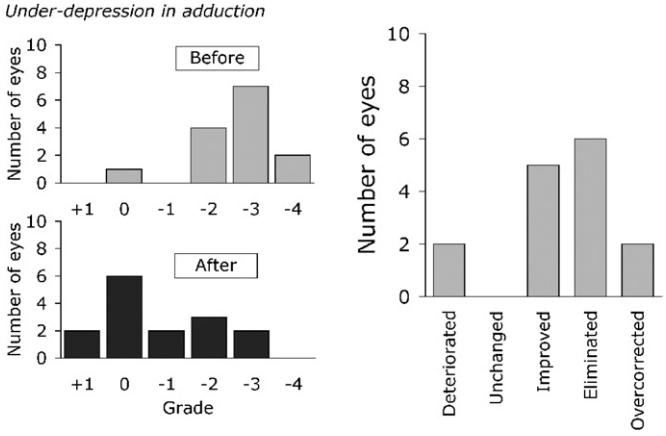

FIG 3. The effects of anterior and nasal transposition on underdepression in adduction using the standard grading scale before (top left panel) and after (bottom left panel). The right panel classifies the change in each eye, where grade change ≥1 was considered significant. Eyes with no problems preoperatively and postoperatively are not represented in this graph.

that, showing full versions and being straight in all gaze directions. In 2 eyes (belonging to 2 different patients) the underdepression in adduction appeared worse than before the anterior and nasal transposition, and both patients will undergo additional surgery. (One of these is the same patient who showed deterioration of overelevation in adduction.)

Improvements in alignment were found in most patients. The deviation in the primary position (Figure 4, top panel) was eliminated or substantially improved in 4 patients, whereas 2 patients were overcorrected and showed 16^{Δ} and 20^{Δ} esotropia. Both these patients originally were exotropic and had undergone recession of the lateral rectus muscles concurrent with the anterior and nasal transposition. One of these patients straightened in the primary

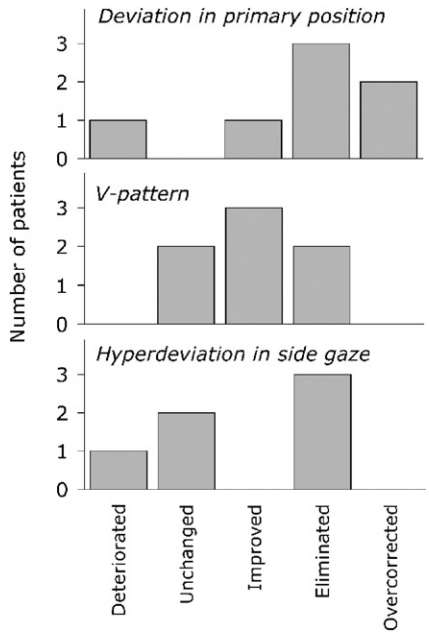

FIG 4. The effects of anterior and nasal transposition on alignment. Shown is the classification of change in deviation in the primary position (top), V pattern (middle), and hyperdeviation in side gaze (bottom), where a change of $\geq 5^{\Delta}$ was considered significant. Note that patients with a significant preoperative deviation in primary gaze underwent rectus muscle recession in combination with the anterior and nasal transposition procedure; the results in the top panel must not be attributed to the transposition. Patients who had no problems preoperatively and postoperatively are not represented in this graph.

position 2 months after additional surgery (advancement of the lateral rectus). A similar additional procedure is planned for the other patient. Another patient showed approximately 10^{Δ} of intermittent exotropia, whereas there had been no significant deviation preoperatively, and it subsequently straightened after lateral rectus recession. The V pattern was eliminated or improved in 5 of the 7 patients who initially presented with V-pattern strabismus (Figure 4, middle panel). Of the 6 patients with hypertropia in side gaze, the problem was elimiated in 3 but became worse in 1 patient (Figure 4, bottom panel). This patients is one who showed worsening of the underdepression in adduction and was recommended to undergo additional surgery to explore whether the IO muscle has slipped. The patient with missing left SR muscle who underwent the anterior and nasal transposition in the right eye only, remained with unchanged hyperdeviation (25^{Δ}) in side gaze.

Overall, 2 patients underwent additional extraocular muscle surgery during follow-up, and 2 others are scheduled or recommended to have additional surgery. All 4 of these patients were in the group of bilateral missing SO tendons. Of the 2 patients with additional procedures, one was asymptomatic at 7 months after lateral rectus recession with infraplacement, whereas the other appeared to have residual hypertropia in side gaze (although orthotropic in

primary) 2 months after the advancement of one lateral rectus and recession of both inferior rectus (IR) muscles.

Discussion

In this report, we describe our findings in a series of patients who, based on surgical findings, were believed to have absence of one or both SO tendons. Imaging techniques, such as high-resolution magnetic resonance imaging,[14] would be needed to further describe the anatomy and pathology of these patients. The exaggerated traction test showed that at least the tendon insertions were absent. We demonstrated that the anterior and nasal transposition procedure can result in significant improvements in overelevation in adduction, underdepression in adduction and upgaze V pattern strabismus (in bilateral cases) and in head posture (unilateral cases) in these patients. Several patients remained with various amounts of V pattern, mainly because of downgaze esotropia. This finding is not an unexpected one, given the mechanics of this procedure that makes it more apt to correct upgaze exotropia than downgaze esotropia. After nasally transposing the IO and anteriorizing, it is converted into an antielevator and intorter in adduction. The adduction in upgaze is caused by the transposed muscle being nasal to the z-axis of Fick,[12] thus eliminating the exotropia in upgaze. The IO muscle, even although an antielevator now, is neurologically inhibited in attempted downgaze. The missing SO tendons do not abduct the eye in downgaze. Hence, esodeviation in downgaze may persist, although a bit of improvement can be expected because the altered torsional position of the globe effectively displaces the IR muscle medially so that its abducting action on depression is somewhat reduced.

The outcome in the 2 cases with congenital unilateral SO tendon absence (without craniosynostosis) was good, with only a very minor residual head tilt in one patient and a small vertical deviation only when tilting the head in the other patient. In contrast, patients with craniosynostosis syndromes frequently have complicating factors in addition to the SO tendon absence that make it more difficult to diagnose and manage. These factors include abnormalities in the shape of the orbit and missing (or attenuated) rectus muscles.

Regardless of the cause of overelevation in adduction in these patients, the treatment has always been a challenge. Most of the attention in the management of such cases was directed to weakening of the IO muscles by means of recession, anterior transposition, myectomy, and denervation and extirpation.[10] The results of these surgeries were not encouraging because they were not very effective in reducing the overelevation in adduction and/or in eliminating the V pattern.[10]

Absence of the SO muscle should be suspected when the standard weakening procedures of the IO do not cause substantial improvements in the clinical picture, when the involved eye is amblyopic, when there is an associated large horizontal deviation, and when the hypertropia is exceptionally large on the involved side.[15] Wallace and von Noorden[15] reviewed the results of the management of patients with isolated absence of the SO muscle. Only one patient in their group had bilateral congenital absence of the SO with a significant V-pattern strabismus and none had craniosynostosis syndromes, where orbital abnormalities and other extraocular anatomy abnormalities are expected to complicate the picture; typically, these patients required more than 2 procedures before achieving acceptable results. In general, all patients with severe congenital fourth nerve palsy should have the exaggerated traction test as described by Guyton[13] at the time of surgery with exploration of the SO muscle tendons if absence is suspected.

Anterior and nasal transposition is a relatively new procedure that has been devised to weaken the IO muscle.[11,12] The procedure places the insertion of the IO muscle on the nasal side of the IR muscle and, hence, nasal to the y-axis and anterior to the x-axis of Fick. This should convert the muscle into an antielevator and an intorter in adduction instead of being an elevator and extorter in adduction.[11,12] This procedure has been used safely and successfully in a variety of eyes with different causes of over elevation in adduction.[12] The patients' age combined with the retrospective nature of the present study prohibited the evaluation of the effects of the procedure on torsion.

In conclusion, anterior and nasal transposition of the IO muscle reduces overelevation in adduction and helps eliminate or reduce divergence of the eyes in upgaze. Horizontal deviations in primary gaze can be treated effectively by concurrent recession of one or more horizontal rectus muscles. Anterior and nasal transposition was most effective for patients with unilateral absence of the SO tendon but also was helpful for patients with craniosynostosis syndromes associated with bilateral absence of the SO tendon. It is likely to benefit patients with severe congenital fourth nerve palsy where standard IO muscle-weakening procedures have been ineffective. Careful, prospective, measurements would be needed to evaluate the effect of this procedure on excyclodeviation in these children.

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An Eye on the Arts – The Arts on the Eye

You had to do your watching quickly, draw conclusions, if there were any to be drawn, set him up for whatever it was you found. Such as, for example, a habit—clearly never pointed out to him—of turning his head to the left before he slashed on the backhand, to let the good right eye follow his blade. And he liked to slash low, sea-raider's attack: a man with a wounded leg was out of a fight, you could move right past him.

— Guy Gavriel Kay (from *The Last Light of the Sun*, Penguin, 2004)