Superior Rectus Transposition and Medial Rectus Recession for Duane Syndrome and Sixth Nerve Palsy

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Objective: To describe our results using augmented temporal superior rectus transposition (SRT) with adjustable medial rectus muscle recession (MRc) for treatment of Duane syndrome and sixth nerve palsy.

Methods: Retrospective surgical case review of patients undergoing SRT. Preoperative and postoperative orthoptic measurements were recorded. Minimum follow-up was 6 weeks. Main outcome measures included the angle of esotropia in the primary position and the angle of head turn. Secondary outcomes included duction limitation, stereopsis, and new vertical deviations.

Results: The review identified 17 patients: 10 with Duane syndrome and 7 with sixth nerve palsy. Combining SRT with MRc improved esotropia from 44 to 10 prism diopters (P < .001), reduced abduction limitation from −4.3 to −2.7 (P < .001), and improved compensatory head posture from 28° to 4° (P < .001). Stereopsis was recovered in 8 patients (P = .03). Three patients required a reoperation: 1 for overcorrection and 2 for undercorrection. A new primary position vertical deviation was observed in 2 patients with complex sixth nerve palsy and none with Duane syndrome. No patient described torsional diplopia.

Conclusions: Superior rectus transposition allows for the option of simultaneous MRc in patients with severe abduction limitation who require transposition surgery. Combining SRT and MRc improved esotropia, head position, abduction limitation, and stereopsis without inducing torsional diplopia.


PATIENTS WITH DUANE SYNDROME and sixth nerve palsy have moderate to severe limitation of abduction, often leading to primary position esotropia that is associated with a compensatory head posture, reduced stereopsis, and diplopia. Various approaches to improving eye position have been proposed, including partial and full vertical rectus muscle transposition (VRT) to increase the abduction of the eye. Full VRT procedures are among the most effective for treatment of the abduction deficit; however, new vertical deviations have been described in 6% to 30% of patients.1-3 There is also the concern of anterior segment ischemia with this procedure, especially when a medial rectus muscle recession (MRc) is also required.4,5

Johnston et al6 introduced a modification of the VRT in which only the superior rectus muscle is transposed. Since then, we have adopted this superior rectus transposition (SRT) technique for appropriate patients, usually combining an augmented SRT with an adjustable MRc (SRT + MRc).7 The purpose of this study was to report and evaluate the results of SRT + MRc to correct the esotropia and head turn in patients with Duane syndrome or sixth nerve palsy.

METHODS

This retrospective study was approved by the institutional review board of Children's Hospital Boston. We reviewed the medical records of patients treated in the Department of Ophthalmology at Children's Hospital Boston from January 1, 2006, through October 31, 2010. Patients treated with SRT, with or without augmentation suture, were included. Exclusion criteria consisted of being younger than 1 year, having had prior transposition surgery, or having follow-up of less than 4 weeks. Information obtained included preoperative and postoperative head turn, esotropia and hypertropia in all gaze positions, ductions, and stereopsis. Postoperative complications were also monitored, including the development of a new vertical misalignment or torsion, impairment of abduction, and the need for reoperation or a revision procedure to improve ocular alignment (excluding postoperative adjustment of
adjustable sutures). Alignment was recorded from the orthoptic evaluation results. Ductions were graded on a scale from 0 (indicating full ductions) through −4 (indicating an eye that was able to move to the midline) and −5 (indicating an eye that approached but was unable to reach the midline) to a maximum of −8 (indicating a rare case where the eye was fixed in an extreme adducted position). Head turn measurements (recorded at distance fixation) were taken from patient records or, for 2 patients, measured by estimation using preoperative and postoperative photographs.

SURGICAL TECHNIQUE

After a “short tag noose” adjustable MRc (described by Nihalani et al7 and Hunter et al9) was performed (Figure 1A), a superotemporal incision was made and the superior rectus muscle was isolated. The muscle was then cleared of surrounding attachments, with extra care taken to separate the superior rectus muscle from the levator muscle (best achieved by removing the lid speculum and using a handheld retractor for exposure) and the superior oblique tendon. The muscle was then secured with a double-armed, 6-0 polyglactin 910 suture and detached from the globe. The posterior surface of the muscle was inspected for remaining attachments to the superior oblique tendon or the frenulum. The temporal pole of the muscle was reattached adjacent to the superior pole of the lateral rectus muscle, and the nasal pole was reattached adjacent to the temporal pole of the superior rectus muscle insertion following the spiral of Tillaux (Figure 1B). In most cases, a double-armed 6-0 polyester augmentation suture was then placed by passing one needle through the lateral one-quarter of the superior rectus muscle and the other needle through the superior one-quarter of the lateral rectus muscle, positioning this suture 8 to 12 mm posterior to the insertion of the 2 muscles.2,10,11 The suture was then tied to pull the 2 muscles together as a loop myopexy,12 with no scleral pass (specific exceptions are noted in paragraph 3 of the “Results” section) (Figure 1B).

Patients underwent assessment in the recovery room 1 to 3 hours after the procedure to check alignment and perform suture adjustment if required. The short tag noose approach allowed for a second suture adjustment to be performed (if required) within 10 days of the procedure. The postoperative follow-up visit was scheduled for 6 to 12 weeks after surgery.

STATISTICAL ANALYSIS

The paired t test was used to analyze the preoperative and postoperative results on head turn, esotropia in primary position, and limitation of abduction. Stereopsis data were compared using the McNemar test. A P value of less than .05 was considered statistically significant.

RESULTS

The medical record review identified 17 patients treated with SRT during the study period, all of whom had simultaneous MRc. Patient characteristics and severity grading of all patients are summarized in Table 1. The average postoperative follow-up was 8 (range, 2-32) months, with 9 patients followed up for 6 months or longer.

Patients with Duane syndrome were younger than those with sixth nerve palsy. The average abduction deficit was greater in the sixth nerve palsy group. All patients had a compensatory head turn of more than 20°.

The mean MRc was 5 mm. Postoperative adjustment was performed in 7 of 17 patients. The final amount of recession ranged from 0 to 8 mm after adjustment, but...
only 1 patient had a recession of more than 6 mm. In 6 cases, there were minor variations from the surgical technique noted in the “Methods” section. One patient, the first in the series, did not have an augmentation suture placed. In 3 patients, the polyester augmentation suture was secured to the sclera; 1 of these patients also underwent a simultaneous 9-mm recession of the lateral rectus muscle to correct globe retraction. Three patients, all with a primary position esotropia of at least 50 prism diopters (Δ), had a simultaneous or a subsequent recession of the contralateral medial rectus muscle.

Mean (SD) esotropia in the primary position improved from 44Δ (16Δ) preoperatively to 10Δ (9Δ) postoperatively (Figure 2A; P < .001). Five patients became orthophoric at distance in the primary position without spectacle correction. There was also a mean 21° improvement in head turn (from 28° to 4°, Figure 2B; P < .001) and a 1.6-unit improvement in abduction (from 4.3° to 2.7°, Figure 3A; P < .001), with a 0.6-U reduction in adduction (Figure 3B; P = .009). The improvement in head turn and abduction is documented in a representative pediatric patient (Figure 4). Stereopsis in the primary position was measurable in 5 of 17 patients (29%) before surgery and improved to 13 of 17 patients (76%) after surgery (Table 1; P = .03).

New-onset vertical deviations in the primary position (mean [SD], 10Δ [2.8Δ]) were observed in 2 patients (12%). Both patients had chronic sixth nerve palsy; one of these patients was a 16-year-old boy described in the paragraph about second procedures, whereas the other patient had multiple orbital fractures in addition to a sixth nerve palsy. No patient with Duane syndrome had an induced vertical deviation. There was no new-onset, symptomatic hypotropia in gaze toward the affected side. In several cases, a hypotropia was already evident in the affected eye before surgery owing to aberrant innervation (eg, Figure 4) or multiple cranial nerve palsies.

Torsion was assessed postoperatively in 6 patients using the double Maddox rod test (3 patients) or anatomic torsion (3 patients). Of these, 2 patients did not have torsion and 3 had incyclotorsion (mean, 4°; values, 2°, 2°, and 5°). None of these patients were symptomatic. In another case, the augmentation suture (placed through the sclera) was removed in the postanesthesia care unit to reverse incyclotorsion and diplopia that were noted in the immediate postoperative period; however, at the final postoperative visit, that patient had 2° to 3° excyclotorsion.

A second procedure was required in 3 cases. Two patients had undercorrections; one was a 4-year-old boy with a residual esotropia of 20Δ who responded to full hypercorrection and a 7.5-mm recession of the contralateral medial rectus muscle. The second was a 16-year-old boy with a sixth nerve palsy secondary to pilocytic astrocytoma (after irradiation) who developed a recurrence (with a 14Δ hypotropia) during the first 2 months after surgery. He was subsequently treated with an augmented inferior rectus transposition, which partially reduced the head turn and improved the esotropia from 25Δ to 10Δ but with a persistent hypotropia. The overcorrection occurred in a 1-year-old girl with Duane syndrome who developed a consecutive esotropia and abduction limitation after surgery; this patient was treated with reversal of the transposition, recession of the lateral rectus muscle, and advancement of the medial rectus muscle so that the final net procedure was simply a 2-mm MRc and lateral rectus recession.

**Table 1. Patient Demographics and Distribution**

<table>
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<tr>
<th>Patient Groups</th>
<th>Duane Syndrome (n = 10)</th>
<th>Sixth Nerve Palsy (n = 7)</th>
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<tbody>
<tr>
<td>Age range, mo</td>
<td>1 to 33</td>
<td>6 to 60</td>
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<td>Sex, No. of patients</td>
<td>Male: 6</td>
<td>1</td>
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<tr>
<td></td>
<td>Female: 4</td>
<td>6</td>
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<td>Follow-up, mo</td>
<td>2 to 32</td>
<td>2 to 16</td>
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<td>Abduction deficit, U</td>
<td>−3 to −5</td>
<td>−4 to −5</td>
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<td>Head turn &gt;20°, No. (%)</td>
<td>10/10 (100)</td>
<td>7/7 (100)</td>
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<tr>
<td>Stereopsis, No. (%)</td>
<td>4/9 (44)</td>
<td>1/6 (17)</td>
</tr>
<tr>
<td>Postoperative</td>
<td>8/9 (89)</td>
<td>5/6 (83)</td>
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**COMMENT**

During the past century, a variety of partial and full transposition procedures of the vertical rectus muscles have been proposed in an effort to improve eye alignment in patients with abduction limitation. In 2004, Rosenbaum reviewed the results of VRT with posterior fixation, orbital fixation, and partial VRT in patients with sixth nerve palsy and Duane syndrome. His study showed a marked improvement in the range of binocular single vision of patients who had undergone a VRT with posterior fixation. Phamnvaechavan et al have described a vessel-sparing, “crossed adjustable” VRT that allows for adjustment of the superior and inferior rectus muscles; this technique does not allow for an augmentation suture.

New vertical strabismus is a concern with VRT; in one report, 6% to 30% of patients treated with VRT had a clinically significant new vertical strabismus. Vertical rectus transposition also carries a theoretical increase in the risk of anterior segment ischemia, especially if MRc is required. Johnston et al were the first to propose that it might be possible to gain the benefits of transposition surgery by transposing only the superior rectus muscle (with or without MRc), thus reducing the amount of surgery required and the theoretical risk of anterior segment ischemia. We adopted this technique, modifying it with the addition of a short tag noose adjustable suture on the medial rectus muscle to allow for optional suture adjustment up to 14 days after surgery, along with routine use of an augmentation suture.

Many patients with abduction limitation will develop tightness or contracture of the medial rectus muscle over time. This condition can limit the effectiveness of a transposition procedure. Because we transposed only...
the superior rectus muscle in our procedure, we were comfortable performing routine MRc to reduce any potential abduction limitation. By making this recession adjustable, we were able to fine-tune the horizontal alignment in the postoperative period, an added benefit considering that a transposition procedure might have a less predictable result on horizontal alignment than a recession or resection. The addition of an abducting force to the eye has a theoretical advantage of preventing recurrence of esotropia over time, but the duration of follow-up in the present study is not sufficient to address this question. Because only 1 muscle is transposed, even patients who have had a prior recession or resection may be considered candidates for the SRT (or the SRT + MRc) procedure.

The results of SRT + MRc in our hands appear to be comparable to the results of VRT (Table 2 and Table 3). However, in one patient with undercorrection (with a sixth nerve palsy secondary to pilocytic astrocytoma), the subsequent transposition of the inferior rectus muscle improved the ocular alignment. In our experience, SRT + MRc and VRT are superior to ipsilateral MRc alone for patients with severe abduction limitations because the amount of MRc required alone tends to cause a new adduction limitation and MRc contributes no chronic abducting force to prevent recurrence. One patient experienced overcorrection, but we do not believe it was a result of the transposition because, after fully reversing the transposition, the overcorrection persisted until the previously recessed medial rectus muscle was advanced (and the lateral rectus muscle was recessed). None of the patients with Duane syndrome developed a hypotropia after surgery, but 2 patients with sixth nerve palsy did; we believe these two were anomalous cases because one had orbital fractures requiring surgery and the other had the augmentation suture attached to the sclera. Torsional diplopia was not a problem, except in one unusual case in which the patient (treated...
with scleral fixation of the augmentation suture) described such distressing torsional diplopia in the recovery room that the suture was removed before discharge, yet that patient ended up with excyclotorsion after surgery. Although it is not possible to make general recommendations about the augmentation suture based on the

<table>
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<th></th>
<th>Preoperative</th>
<th>Postoperative</th>
<th>Preoperative</th>
<th>Postoperative</th>
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<tr>
<td>Head turn, degrees</td>
<td>20</td>
<td>5</td>
<td>38</td>
<td>12</td>
<td>24</td>
<td>3</td>
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<td>Esotropia, ∆</td>
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<td>6</td>
<td>32</td>
<td>9</td>
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<td>10</td>
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<td>Abduction, U (change, %)</td>
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<td>-2.7 (29)</td>
<td>-4</td>
<td>-2</td>
<td>-4</td>
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<td>18</td>
<td>5/64</td>
<td>3/38</td>
<td>8</td>
<td>8/10</td>
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<tr>
<td>Induced vertical deviation, No. of patients</td>
<td>20</td>
<td>5</td>
<td>38</td>
<td>12</td>
<td>24</td>
<td>3</td>
</tr>
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</table>

Abbreviations: ∆, prism diopters; MRC, medial rectus muscle recession; SRT, superior rectus muscle transposition; VRT, vertical rectus muscle transposition.

Figure 4. Head position and motility in a boy aged 5½ years. A and C, Preoperative views. B and D, Two-month postoperative views. The patient underwent augmented superior rectus transposition and a 4.5-mm left medial rectus muscle recession.
latter 2 cases, we advise surgeons adopting the SRT procedure to use a simple loop myopexy of superior rectus muscle to lateral rectus muscle for augmentation until more experience is gained and published.

This study is limited by its retrospective nature and the follow-up of less than 6 months in 8 patients (47%). No control group was treated with MRc alone. Some patients were too young to assess stereopsis adequately at the initial examination; therefore, some of the observed improvement in stereopsis may have been the result of patient maturation. Many patients were also too young to permit objective assessment of torsion. Field of binocular vision was not tested in this retrospective study; however, the 1.6-U improvement in abduction combined with a 0.6-U limitation of adduction is consistent with an increase in the range of motion of the eye.

The study addresses only temporal SRT; it does not address the question of nasal SRT.24 We performed nasal SRT in one patient but had to reverse the procedure 3 days later owing to intolerable torsional diplopia. This patient was not included in the present study.

We can only speculate how a vertical deviation and torsional diplopia did not result from an unbalanced transposition of the superior rectus muscle. We believe that because the transposition also involves a slight advancement of the superior rectus muscle (to follow the spiral of Tillaux), the procedure increases the effective strength of the superior rectus muscle to counterbalance the weakening of the vertical effect that results from the transposition. However, such an alteration of the superior rectus muscle insertion, extending the temporal edge to the lateral rectus tendon insertion and then following the muscle to the point of the augmentation suture, should increase torsion that much further. Enhanced modeling of the forces operating within the orbit may be required to begin to explain this apparent inconsistency. The clinically insignificant hypotropia noted in abduction in several cases after surgery appeared to be a consequence of aberrant innervation because it was present before surgery in all patients.

In conclusion, in patients undergoing SRT + MRc, there was a markedly reduced esotropia in the primary position, increased abduction, and improved head position with minimal effect on adduction and few cases of vertical or torsional diplopia. Considering these results, we recommend SRT + MRc for patients with profoundduction limitation in which there is no reasonable chance that a horizontal rectus muscle procedure alone will be satisfactory. The procedure is especially helpful in cases in which there may be simultaneous contracture of the medial rectus muscle, because it allows an MRc to be combined with a transposition procedure without greatly increasing the risk of anterior segment ischemia.

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REFERENCES