Superior Rectus Transposition vs Medial Rectus Recession for Treatment of Esotropic Duane Syndrome

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IMPORTANCE Superior rectus transposition (SRT) with or without medial rectus recession (MRC) has been introduced as an alternative to MRC alone for treatment of esotropic Duane syndrome; however, the effectiveness of these procedures has not been compared previously.

OBJECTIVE To compare the safety and efficacy of MRC and SRT in treatment of Duane syndrome.

DESIGN, SETTING, AND PARTICIPANTS Retrospective medical record review of all patients with esotropic Duane syndrome who underwent surgical treatment from January 1, 2006, through December 31, 2012, in a multispecialty, hospital-based pediatric ophthalmology/adult strabismus practice at Boston Children’s Hospital. Patients in the SRT group underwent SRT with or without MRC; those in the non-SRT group underwent unilateral or bilateral MRC.

EXPOSURES Surgical treatment of esotropic Duane syndrome.

MAIN OUTCOMES AND MEASURES Binocular alignment, ocular ductions, head position, stereopsis, and fundus torsion were recorded before surgery and at the 2-month and final postoperative visits. We also evaluated postoperative drift.

RESULTS The medical record review identified 36 patients who underwent 37 procedures, including 19 in the SRT group (13 SRT + MRC and 6 SRT alone) and 18 in the non-SRT group (11 unilateral MRC and 7 bilateral medial rectus recession). Mean MRC was smaller when performed with SRT (3.3 vs 5.3 mm; P = .004). Although the initial deviation was larger in the SRT group, both groups had a similar improvement in esotropia and head turn. Abduction improved by at least 1 unit in 15 of 19 patients in the SRT group (79%) vs 5 of 18 in the non-SRT group (28%). In 24 patients followed up for more than 6 months, mean esotropia decreased from 8.2 to 6.1 prism diopters (Δ) in the SRT group (n = 12) but increased from 7.2 to 10.9Δ in the non-SRT group (n = 12).

CONCLUSIONS AND RELEVANCE The combination of SRT and MRC was more effective than MRC or bilateral medial rectus recession at improving abduction while allowing for a smaller recession to align the eyes and eliminate a compensatory head posture. Although any surgery on the vertical rectus muscles should in theory increase the risk for vertical or torsional complications, to date this theory has not been borne out in our patients. Patients treated with SRT appear to have a reduced likelihood of long-term undercorrection. We therefore recommend SRT with adjustable MRC for treatment of Duane syndrome in patients with larger amounts of esotropia.

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Duane syndrome is a congenital cranial dysinnervation disorder characterized by limitation of horizontal eye movement(s) with aberrant innervation causing globe retraction in adduction. Most patients have esotropia in primary gaze, limited abduction, and a head turn toward the involved side (esotropic type 1 Duane syndrome). Various surgical approaches have been proposed to improve binocular alignment and reduce head turn in Duane syndrome. Unilateral or bilateral medial rectus resections (MRCs or BMRs, respectively) are common procedures and are usually satisfactory for correcting alignment in primary gaze and reducing torticollis; however, MRC does not improve the abduction limitation. Indeed, a large MRC limits abduction, which in turn may cause difficulty with convergence and in contralateral gaze.

In an effort to improve abduction without limiting adduction in patients with Duane syndrome, a variety of transposition procedures have been developed, including augmented transposition of both vertical rectus muscles (VRT). The superior rectus transposition (SRT) was developed subsequently to reduce the number of muscles requiring surgery while decreasing the likelihood of inducing a vertical deviation observed with VRT. A recently published experience combining SRT with an adjustable MRC in patients with esotropic Duane syndrome found results comparable to those of VRT but with a lower incidence of induced hypertropia. However, whether the potential benefits of SRT or similar transposition procedures warrant the added complexity and potential risk for vertical deviation associated with the procedure remains unclear. The purpose of this study is to address this question by comparing the results of SRT with those of MRC in terms of postoperative alignment, head turn, and ocular ductions and long-term stability of alignment.

Methods

This retrospective study was approved by the institutional review board of Boston Children’s Hospital. We used billing records to identify patients treated for Duane syndrome in the Department of Ophthalmology from January 1, 2006, through December 31, 2012. We included patients with esotropic Duane syndrome who were treated with SRT or MRC. Patients were divided into the SRT group, consisting of patients who had SRT alone or in combination with MRC (SRT + MRC), and the non-SRT group, consisting of patients who had unilateral MRC or BMR. Patients were excluded if follow-up was less than 1 month or if VRT surgery had been performed previously. In cases in which patients underwent MRC follow-up more than 2 years later by SRT, the 2 procedures were analyzed separately: the first in the non-SRT group, the second in the SRT group.

Superior rectus transposition was performed as described previously. Briefly, the superior rectus muscle was isolated, with care taken to clear the muscle of attachments to overlying levator and underlying superior oblique tendon. The muscle was secured, detached, and then reattached to the eye along the spiral of Tillaux, spanning the gap between the original lateral and superior rectus muscle insertions. A loop myopexy was then placed between the lateral and superior rectus muscles 8 to 10 mm posterior to the insertion, incorporating approximately one-quarter of the muscle width, in most cases without a scleral pass. Medial rectus recession was performed per the preference and surgical tables of the individual surgeon.

Sensorimotor examination data were extracted from the preoperative evaluation, 2 months after surgical intervention, and at the final follow-up examination. Only patients with follow-up longer than 6 months were included for analysis of long-term stability (postoperative drift). At each point, we recorded the angle of esotropia in near and distance straight-ahead gaze, head turn in degrees, limitation of abduction (including limitation in elevation and depression), limitation of adduction, stereopsis (by Titmus fly and Randot circles), hypertropia in straight-ahead gaze and elevation, and torsion (by the double-Maddox rod test and/or indirect ophthalmoscopy as available in the record).

Alignment was recorded from the orthoptic evaluation of distance deviation. Ductions were graded on a scale from 0 (indicating full duction) 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 in which the eye was nearly fixed in an extremely adducted position). Head turn was measured with a goniometer at distance fixation or using clinical photographs. In SRT cases performed toward the end of the study period, we monitored intraoperative torsional changes as described by Holmes et al (Supplement [eFigure 1]).

Statistical calculations were performed using commercially available software (Prism 5.0; GraphPad Software, Inc [http://www.graphpad.com]). We used 2-way analysis of variance to compare the preoperative and postoperative results of the SRT and non-SRT groups and 1-way analysis of variance to analyze the change in deviation angle and head turn of the groups undergoing SRT + MRC, SRT alone, MRC alone, and BMR. If a patient in the SRT group had undergone previous MRC, the amount of MRC was included in the calculation of MRC dosage. Titmus stereopsis of greater than 2 lines was considered clinically meaningful. We used Bonferroni post hoc tests for multiple comparisons and an unpaired t test for comparisons where appropriate. P < .05 was considered statistically significant.

Results

The medical record review identified 37 patients with esotropic Duane syndrome who underwent 38 procedures. One procedure was excluded for lack of adequate follow-up, leaving 36 patients who underwent 37 procedures by 9 surgeons, with 20 of the 37 performed by 2 of us (L.R.D. and D.G.H.), including 13 of 19 SRT procedures. The SRT group included 13 cases of SRT + MRC and 6 cases of SRT alone. Six of 13 SRT + MRC cases underwent muscle-to-muscle augmentation (superior to lateral rectus muscle) without scleral fixation, and 7 underwent muscle-to-sclera augmentation (when the lateral rectus muscle was recessed or lax). The non-SRT group included 11 cases of MRC alone and 7 cases of BMR.
Among the 6 cases of SRT without MRC, 2 had undergone an MRC procedure more than 2 years previously (for which the MRC amount was included in the calculation of MRC performed); in 2 additional cases, the MRC was reduced to 0 at the time of postoperative suture adjustment. One case of SRT alone was combined with an ipsilateral inferior oblique recession to treat hypertropia in adduction.

Characteristics of patients in the SRT and non-SRT groups are summarized and compared in the Table. The amount of MRC performed (Supplement [eFigure 2]) was significantly larger in the non-SRT group (mean [SD], 5.3 [1.3] mm) than in the SRT group (3.3 [2.3] mm) ($P = .004$). The mean postoperative follow-up was 14.6 months for the SRT group and 32.0 months for the non-SRT group.

Although the initial deviation was greater in the SRT group and both procedures significantly decreased the angle of esotropia, the SRT + MRC procedure appeared to be most effective (Figure 1A-C). Superior rectus transposition alone was comparable in effect to MRC alone and BMR. The SRT and non-SRT groups showed notable improvements in head turn (Figure 1D-F). As with angle of deviation, the SRT + MRC group appeared to show the greatest efficacy, but the difference in head position was not statistically significant.

Stereopsis improved in both groups (Table), with 3 patients in each group improving by 3 or more lines on the Titmus test. No patient had a reduction in stereopsis.

Preoperative and postoperative limitation of adduction and abduction were available for all procedures. Abduction significantly improved in the SRT group but not in the non-SRT group (Figure 2A and B). We found a small but significant increase in adduction limitation in the SRT group.
non-SRT groups (Figure 2C and D). Two patients from each group developed postoperative adduction limitation of more than −0.5.

In 15 patients in the SRT group, abduction was measured not only in the horizontal meridian but also in elevation and depression (Figure 3). In some cases, preoperative abduction was slightly better in elevation (Figure 3B) or in depression (Figure 3C). After surgery, we found a trend toward more improvement in abduction in elevation than in depression (Figure 3D and Supplement eFigure 3).

No patient in either group described torsional diplopia postoperatively. Of 9 patients in whom torsion was measured before and after surgery (all but 1 in the SRT group), 8 showed no change postoperatively. A +1 (asymptomatic) increase in intorsion was observed in 1 SRT patient. Intraoperative monitoring of torsion performed in 3 patients in the SRT group showed no change at the end of the procedure.

Small, asymptomatic vertical deviations in adduction were present in 3 of 18 patients in the SRT group and 3 of 19 in the MRC group. No patient developed a symptomatic vertical strabismus after surgery; however, 2 SRT patients had a new postoperative hypotropia or hypophoria. An 18-year-old girl developed hypotropia of 4 prism diopters (Δ) in straight-ahead gaze without diplopia; at the final postoperative visit, she had 20 seconds of stereopsis with a 5° residual head turn. A 5-year-old girl underwent a 6-mm ipsilateral MRC and developed a hypophoria after a subsequent SRT. Three years later, she had a residual esotropia of 8Δ and a persistent left hypophoria of 10Δ but was asymptomatic with good fusion and 70 arc seconds of stereopsis with a 5° residual left head turn.

Measurements of long-term drift in postoperative alignment were available for evaluation in 24 patients (12 in the SRT group [mean follow-up, 22 months] and 12 in the non-SRT group [47 months]). Although we found a trend toward further increase in surgical effect in the SRT group (mean esotropia decreased from 8.2Δ to 6.1Δ), the trend was toward a loss of effect in the non-SRT group (mean esotropia increased from 7.2Δ to 10.9Δ) (Figure 4A). These trends were not statistically significant (P < .001). A similar trend was observed in the improvement in head position (Figure 4B), with an improvement over time in the SRT group (decrease from 5.0° to 2.5°) compared with a recurrence over time in the non-SRT group (increase from 3.3° to 5.5°). Again, these observed trends were not statistically significant (P < .14). No significant long-term changes in limitation of abduction (Figure 4C) or adduction (Figure 4D) were observed.

Discussion

Debate is ongoing about the best approach for treatment of esotropic Duane syndrome and other causes of abduction limitation. Recession of the ipsilateral medial rectus muscle is an attractive option for its simplicity and predictability, and indeed this approach will improve the deviation and head turn in straight-ahead gaze.1-3,5 Unfortunately, to correct larger deviations and head turns, a large MRC is required, which often induces limitation of abduction without improving abduction.5,7 Bilateral MRC is used for larger angles. Recessions of the contralateral medial rectus muscle are sometimes used, but the approach remains controversial,2,13-15 with a paradoxical increase in head turn described in some cases.16,17

Transposition procedures have been proposed as a means of increasing lateral force to the eye to compensate for the lack of lateral rectus muscle function. At present, full, augmented VRT is a widely used approach.19-21 Rosenbaum22 showed a marked improvement of abduction and in the field of binocular single vision in patients with Duane syndrome who undergo VRT. Concerns about induced hypotropia have limited widespread adoption of VRT.23 In addition, when the medial rectus muscle is tight, concerns about anterior segment ischemia limit the option of MRC.

We adopted the SRT in 2006 after Johnston and colleagues8 first presented results in the treatment of Duane syndrome and sixth nerve palsy. Mehendale and colleagues9 reported successful treatment of patients with complete loss of lateral rectus function. Morad and Kekunnaya24 have also reported good results of SRT + MRC for Duane syndrome.

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In our multiphysician strabismus practice, patients with esotropic Duane syndrome were treated with SRT and non-SRT procedures according to surgeon preference, and all patients were examined by the same orthoptic team according to the same documentation guidelines. More patients undergoing non-SRT procedures had surgery earlier in the study period, which resulted in a longer overall follow-up for the non-SRT group. Although the present study was not a randomized...
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Abduction improved significantly in patients treated with SRT compared with MRC alone. We found a comparable reduction of esotropia in straight-ahead gaze, but the SRT group had a larger preoperative esotropia than the non-SRT group; thus, surgeons might have selected SRT for patients with a larger preoperative angle. The improvement in abduction with SRT (15 of 19 patients [79%]) was similar to the results reported previously in a smaller group of patients. The improvement in abduction with MRC (5 of 18 patients [28%]) was similar to the 29% improvement reported by Merino et al (although they graded abduction limitation only on a scale of –1 through –3).

By combining SRT with MRC, a significantly smaller MRC was required to treat the esotropia. We also found a trend toward better preservation of abduction in the SRT group, which matches our clinical impression. A prospective study collecting more complete documentation of preoperative and postoperative abduction in both groups is necessary to confirm or refute this claim.

The SRT patient in our study who developed an early, severe overcorrection is the same patient described in an earlier study by Mehendale and colleagues. Overcorrection has also been reported by Morad and Kekunnaya, especially when SRT was combined with BMR. We recommend reserving the SRT procedure for patients with a larger angle of deviation and severe abduction limitation. When the deviation is greater than 50Δ, we combine the SRT with BMR. In all cases (SRT and non-SRT), we place the medial rectus muscle(s) on adjustable sutures25,26 to reduce the likelihood of an early overcorrection. In 2 SRT patients in the present series, the medial rectus muscle was adjusted back to a 0-mm recession in the recovery room 1 hour postoperatively, with excellent long-term results in both cases.

We observed a trend toward long-term drift in opposite directions in the 2 groups (2.0Δ decrease in esotropia and 2.5° improvement in head turn in the SRT group vs a 3.7Δ increase in esotropia and 2.1° recurrence of head turn in the non-SRT group). Although these differences were not statistically significant, they align with our clinical impression that the SRT procedure seems to have a more sustained effect over time than the MRC procedure. If valid, a difference in drift may occur because the force of the medial rectus muscle is unbalanced with MRC alone, allowing development of further contracture and inelasticity over time. Perhaps a balancing abducting force provided by SRT prevents this evolution. Although the mean follow-up was not identical in the 2 long-term groups, the shorter follow-up for the SRT group was longer than 2 years, which should be sufficient to allow for valid comparison.

A change in outward vector force provided by SRT would carry with it a possible increased likelihood of long-term overcorrection. Velez and colleagues reported late exotropia in 14 of 51 patients followed up after VRT with or without subsequent MRC. We advise against targeting an overcorrection when performing a transposition procedure in esotropic Duane syndrome.

We observed better abduction in elevation or in depression preoperatively in 7 of 16 patients. The improvement in abduction observed with SRT may have been greater in elevation, which is not unexpected considering the asymmetric transfer of force in the single muscle transposition.

In our clinical practice, the field of binocular single vision is not measured routinely after surgery. However, if the improvement in abduction exceeded the limitation of adduction after surgery, one can infer that the field of single binocular vision might have been enhanced in patients treated with SRT.

Based on our current state of understanding of extraocular muscle function, one would predict that an unbalanced temporal transposition of the superior rectus muscle would create torsional diplopia. This prediction was not supported by our observations intraoperatively or postoperatively. Superior rectus transposition may induce some intorsion; however, our observations suggest that the risk for symptomatic torsional diplopia seems to be very low. We recommend that future studies document torsion with photographic methods before and after surgery.

With the transposition of the superior rectus laterally, a portion of the muscle force vector directed vertically was displaced laterally. This loss of vertical force would be expected to cause a hypotropia, but none was observed. We speculate that the hypotropia did not occur when the muscle was attached along the spiral of Tillaux because it was also advanced slightly, which compensated for the reduced vertical force. If torsion and vertical strabismus were induced by surgery, they could adversely affect fusion or stereopsis, but our data show no negative impact, with improved binocular function after surgery.

Conclusions

Compared with MRC or BMR alone, SRT + MRC appears to be more effective at improving abduction while allowing for a smaller MRC to align the eyes and eliminate a compensatory head posture in patients with esotropic Duane syndrome. Although not confirmed by this study alone, patients treated with SRT probably will have a reduced likelihood of developing a long-term undercorrection. In our experience, SRT + MRC is technically straightforward and allows for recession of a tight medial rectus muscle with transposition in a single procedure. It also allows for suture adjustment to reduce the effect if required. We therefore continue to recommend SRT combined with adjustable MRC for treatment of patients with profound limitation of abduction and large-angle esotropia.
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