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## Assessment of torsion after superior rectus transposition with or without medial rectus recession for Duane syndrome and abducens nerve palsy

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### Abstract

**Background**—Superior rectus transposition with or without medial rectus recession has been advocated for the treatment of abducens nerve palsy and esotropic Duane syndrome. Early reports have focused mainly on postoperative ocular alignment, but there is concern that superior rectus transposition may induce torsional misalignment. The purpose of this study was to evaluate torsional outcomes after superior rectus transposition surgery using prospective preoperative and postoperative torsional assessments.

**Methods**—Prospective measurements were performed on all patients undergoing superior rectus transposition. Preverbal infants were assessed using fundus torsion evaluating the position of the fovea relative to the optic nerve; older children/adults underwent double Maddox rod (DMR) assessment of torsion.

**Results**—A total of 11 subjects met the study inclusion criteria. The etiology of strabismus was an abducens nerve palsy ( $n = 7$ ) or Duane syndrome ( $n = 4$ ). For the subjects evaluated by fundus torsion ( $n = 4$ ), there was no significant change in torsion for 3 (75%). For those subjects undergoing DMR ( $n = 7$ ), there was a significant change in subjective torsion ( $4.7 \pm 3.8^\circ$  excyclotorsion vs  $0.0^\circ \pm 5.0^\circ$  excyclotorsion;  $P = 0.004$ ). Esotropic deviation improved significantly for all subjects ( $39 \pm 23$  vs  $6.5 \pm 13$   $P = 0.001$ ) and no significant mean vertical deviation postoperatively, although 1 patient had a clinically significant postoperative hypertropia measuring 14 . Abduction also improved significantly ( $-4.2 \pm 0.9$  vs  $-2.8 \pm 1$ ,  $P = 0.0001$ ).

**Conclusions**—In this patient series, superior rectus transposition with medial rectus recession did not have clinically significant induction of torsional diplopia as a result of the procedure.

Complete or near-complete abduction deficits in Duane syndrome and abducens nerve palsy is has been managed with various surgical techniques. Superior rectus transposition, with or without medial rectus recession, has recently been advocated as an alternative to full-tendon vertical rectus transposition (Johnston SC, et al. IOVS 2006;47:ARVO e-abstract 2475).<sup>1,2</sup>

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Using this technique, investigators have reported improvement in abduction, torticollis, and esotropic deviation, with a low incidence of induced vertical deviation. Significant induction of vertical deviations in the patients included in these studies has not been reported; however, hypothetically, superior rectus transposition can cause torsional misalignment. The largest reported study of patients undergoing superior rectus transposition did not include pre- and postoperative comparisons of torsion.<sup>1</sup> This information is crucial to surgeons who may plan to use this technique, because the induction of torsion may result in postoperative diplopia, difficulties with fusion, and anomalous head posture. The purpose of this study was to evaluate torsional outcomes after superior rectus transposition surgery with prospective pre- and postoperative torsional assessments.

## Methods

This study was approved by the University of California, Los Angeles, Institutional Review Board and conformed to the requirements of the US Health Insurance Portability and Accountability Act of 1996. The clinical records of all patients with esotropic Duane syndrome or complete abducens nerve palsy were reviewed after they prospectively underwent assessments of preoperative and postoperative torsion. Those subjects who underwent full-tendon superior rectus transposition to the lateral rectus muscle insertion with a posterior fixation myopexy between the lateral and superior rectus with or without medial rectus recession at the Jules Stein Eye Institute between 2012 and 2014, and had at least 8 weeks of postoperative follow-up were included. Patients were excluded if there were missing data related to deviation measurements in adduction or abduction, ocular rotations, or torsion. The technique used for this procedure has been published elsewhere.<sup>1</sup> When adjustable sutures were used, the goal after adjustment was orthotropia in primary position.

The following pre- and postoperative characteristics were recorded from patients' medical record: age at onset, age at surgery, preoperative motor alignment at distance and near and in the cardinal positions of gaze, degrees of torticollis, an assessment of ocular ductions, and torsion (either by fundus torsion evaluating for the presence of incyclotorsion or excyclotorsion by evaluating the position of the fovea relative to the optic nerve or double Maddox rods, depending on patient age and ability). For those subjects who underwent additional surgical procedures after the vertical rectus transposition, these characteristics were also recorded at the preoperative visit before the subsequent surgeries.

In general, visual acuity was assessed using best refractive correction with projected age-appropriate optotypes. Ocular alignment was assessed using cover and uncover and prism and alternate cover testing at distance (20 feet) in the seven cardinal gaze positions. Motor alignment at near was assessed at 14 inches. All motor evaluations were done using spectacle correction. Torticollis was estimated by the experienced strabismologist (SJI, FGV, SLP) or measured with a goniometer, in the patient's habitual head position in degrees. Torticollis was assessed with the subject viewing a distant target. Ocular ductions were measured using a standard 4-point scale.<sup>3</sup>

## Statistical analysis

Statistical analyses were performed using statistical software, JMP version 10.0 (SAS Corporation, Cary, NC) and Excel (Microsoft, Redmond, WA). Preoperative and postoperative values were compared using a paired t-test. A *P* value of <0.05 was considered statistically significant.

## Results

A total of 11 subjects were included; no subjects were excluded based on the above-described exclusion criteria. The etiology of strabismus was an abducens nerve palsy in 7 subjects and Duane syndrome in 4 subjects. Ocular torsion was measured using fundus torsion in 4 subjects and double Maddox rods in 7 subjects.

For those subjects undergoing double Maddox rod testing, there was a significant change in subjective torsion ( $4.7^\circ \pm 3.8^\circ$  vs  $0.0^\circ \pm 5.0^\circ$  excyclotorsion; *P* = 0.004). The range of postoperative torsional shift varied from  $1^\circ$  to  $8^\circ$  of incyclotorsional shift. The largest torsional shift was seen in a 50-year-old woman with Duane syndrome (patient 6), diagnosed at 5 years of age, who had subjective torsional diplopia. She had mild myopia ( $-1.75$  D in the right eye and  $-1.00$  in the left eye) but otherwise no other neurologic or ophthalmic diseases. Preoperatively she had 25 of esotropia in primary position, with a  $-4$  abduction and mild ( $-1/2$ ) limitation to adduction. In adduction, her esotropia decreased to 10, and in abduction it measured 50. She underwent superior rectus transposition with suture myopexy to the lateral rectus muscle and medial rectus recession of 5 mm on adjustable suture. Postoperatively, she had a small residual esotropia but developed a symptomatic torsional deviation along with a small hypertropia.

For the subjects evaluated by fundus torsion, there was no significant change in torsion for 3 subjects (75%). However, 1 subject (patient 10), a 10-month-old girl diagnosed with bilateral Duane syndrome at 6 months of age, had new and marked incyclotorsion in the operative eye postoperatively. Patient 10 also had a large overcorrection, with severe torticollis that required reoperation, consisting of reposition of the superior rectus to its original insertion. Preoperatively her cycloplegic refraction was  $+3.50$  D in the right eye and  $+5.00$  D in the left eye. She was wearing her full cycloplegic refraction at the time of her measurements, which revealed an esotropia of 30 in forced primary position. She had a  $-4$  abduction bilaterally and full adduction bilaterally. She underwent a superior rectus transposition with suture myopexy to the lateral rectus and medial rectus recession of 4.5 mm in the right eye. Immediately after surgery she was found to have a large exotropia and fundus torsion.

There was a significant improvement in esotropic deviation for all subjects ( $39 \pm 23$  vs  $6.5 \pm 13$ , *P* = 0.001) and no significant vertical deviation postoperatively overall ( $1 \pm 1.6$  hypotropia vs  $1.9 \pm 6$  hypertropia, *P* = 0.3), although 1 patient had an induced vertical deviation  $>10$  (patient 2, 14). Abduction also improved significantly ( $-4.2 \pm 0.9$  vs.  $-2.8 \pm 1$ , *P* = 0.0001).

## Discussion

In our study, superior rectus transposition was successful in improving ocular alignment and horizontal head posture in patients with Duane syndrome and abducens nerve palsy. There was no significant induction of postoperative vertical deviation in most of our patients although 5 had some degree of vertical deviation and 1 had a deviation  $>10^\circ$ . These findings agree with previous reports by Mehendale and colleagues<sup>1</sup> and Johnston and colleagues (IOVS 2006;47:e-abstract 2475). Our study is unique in that we evaluated pre- and postoperative torsion measurements and found a statistically significant incyclotorsional shift in our patients. Although most of our patients did not have a symptomatic torsional misalignment postoperatively or subjective complaints (range,  $0^\circ$ - $8^\circ$  of incyclotorsion, for those with double Maddox rod testing), the amount of induced incyclotorsion (range,  $1^\circ$ - $8^\circ$  of incyclotorsion shift in 8 of 11 patients) could be significant for patients who have no or a small amount of preoperative incyclotorsion.

The superior rectus transposition procedure was first introduced for the treatment of abduction deficits by Johnston and colleagues (IOVS 2006;47:e-abstract 2475). It was further popularized by Mehendale and colleagues,<sup>1</sup> who described 17 patients who underwent the procedure. This group found that esotropia and abduction deficits improved significantly in the majority of their patients, similar to our findings. However, torsion was only assessed postoperatively in 6 of their patients, and no preoperative torsion measurements were reported. Although the majority of their patients had a small amount of incyclotorsion (mean,  $4^\circ$ ), it was deemed clinically insignificant in most of the patients. The authors mention 1 other patient who had a posterior fixation suture removed in the postoperative care unit due to symptomatic incyclotorsion. The total amount of induced incyclotorsion is unclear from these reported data.

Given the function of the superior rectus muscle, one would expect that transposing it laterally would create both a vertical deviation and incyclotorsion. Surprisingly, a new vertical deviation is not frequently seen after this surgery. Although the reason for this is not clear, it could be due to the minimal change in the vertical vector of the muscle after transposition. Alternatively, there may be a larger vertical deviation induced in ipsilateral gaze that is not appreciated due to the lack of abduction present in most of these patients.

Interestingly, 1 of our 2 patients with severe overcorrection and torsional misalignment was a child under 1 year of age. Our case is quite similar to one described by Mehendale and colleagues,<sup>1</sup> who describe a 1-year-old child with Duane syndrome who developed a consecutive exotropia and adduction limitation postoperatively. Our case and the case described above required reoperation and near-complete reversal of the procedure. These 2 cases may point out a possibility that patients with Duane syndrome who are very young may be at higher risk for overcorrections. This findings has also been demonstrated by our group in younger patients undergoing full vertical rectus transposition (transposition of both the superior and inferior rectus).<sup>4</sup> Torsional misalignment may have been avoided with intraoperative assessment of torsion as described by Holmes and colleagues.<sup>5</sup>

A recent study by Yang and colleagues<sup>6</sup> compares the superior rectus transposition procedure to medial rectus recession for the treatment of Duane syndrome. The authors comment on torsional changes after surgery, stating that 8 of the 9 patients with torsional measurements had no change postoperatively, and 1 patient had asymptomatic intorsion with a “+1” shift in fundus torsion. This study cannot be directly compared to the present study because more than half of the patients had scleral posterior fixation sutures instead of myopexy sutures, and the patient ages and preoperative torsion measurements are not stated.

The results of the present study should be understood within the context of its limitations. First, this study is limited by the small number of subjects included in each subgroup. In addition, multiple surgeons participated in the study, and there may have been small differences in surgical technique. Also, we did not measure diplopia-free fields, as was done by Johnston and colleagues (IOVS 2006;47:ARVO e-abstract 2475). This information would have been helpful in understanding the functional binocular improvements after surgery. Finally, the measurement of ocular torsion was not standardized over the entire study population due to the need for cooperation and understanding of the double Maddox rod test, which was not possible in some of our youngest patients.

Preoperative torsion should be assessed in patients undergoing superior rectus transposition. Although few of our patients had torsional misalignment, the risk should still be considered in patients who preoperatively either have no torsion or incyclotorsion. In these patients intraoperative torsion monitoring should also be considered because of the possibility that the procedure may induce clinically significant post-operative incyclotorsion.<sup>5</sup> However, patients with any amount of excyclotorsion preoperatively are much less likely to have a resultant torsional misalignment that is clinically significant.

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**Table 1**  
**Characteristics of patients undergoing superior rectus transposition**

ID	Age, years	Diagnosis	Surgery <sup>d</sup>	Deviation, PD <sup>b</sup>		Torsion, degrees (method)	
				Preoperative	Postoperative	Preoperative <sup>c</sup>	Postoperative
1	52	6th NP	MRc 6.5 mm	45 4 HoT	2 ET 3 HT	6 (DMR)	5 (DMR)
2	12	6th NP	MRc 6 mm	30 4 HoT	7 ET 14 HT	8 (DMR)	4 (DMR)
3	70	6th NP	MRc 3 mm	30	0 2 HT	5 (DMR)	0 (DMR)
4	49	6th NP	LR plication 7 mm	37 2 HoT	12 2 HT	10 (DMR)	3 (DMR)
5	78	6th NP	MRc 4 mm	25	10 3 HT	4 (DMR)	-2 (DMR)
6	50	Duane	MRc 5 mm	25	10 7 HT	0 (DMR)	-8 (DMR)
7	50	6th NP	MRc 5 mm	85	20	0 (DMR)	-5 (DMR)
8	16	Duane	None	12	12 4 HoT	0 (fundus)	0 (fundus)
9	0.5	Duane	None	25	0 2 HoT	0 (fundus)	0 (fundus)
10	0.8	Duane	MRc 4.5 mm	30	-25 (XT) 4 HoT	0 (fundus)	"large incyclotorsion" ~10 (fundus)
11	0.6	6th NP	MRc 4 mm	80	10	0 (fundus)	0 (fundus)

DMR, double Maddox rods; ET, esotropia; HoT, hypotropia; HT, hypertropia; LR, lateral rectus; MRc, medial rectus recession; PD, prism diopter; 6th NP, abducens nerve palsy; SR, superior rectus muscle; XT, exotropia.

<sup>a</sup>In addition to superior rectus transposition.

<sup>b</sup>Vertical deviations are made with reference to the operated eye. If the operated eye was hypertropic, then the vertical deviation is notated as hypertropia (HT); if the operated eye was hypotropic, then the vertical deviation is notated as hypotropia (HoT).

<sup>c</sup>Plus (+) values represent excyclotorsion; minus (-) values, incyclotorsion.

**Table 2**  
**Pre- and postoperative characteristics of patients undergoing superior rectus transposition**

Clinical measurements	Preoperative <sup>a</sup>	Final postoperative	P value <sup>b</sup>
Torsion measured by DMR, degrees (n = 7)	4.7 excyclo ± 3.8	0 torsion ± 5.0	0.004
Fundus torsion, degrees (n = 4)	0 ± 0	2.5 incyclo ± 5	0.2
Primary position horizontal deviation, PD	39 ET ± 23	6.5 ET ± 13	0.001
Primary position vertical deviation, PD	1 hypo ± 1.6	1.9 hyper ± 7	0.3
Abduction	-4.2 ± 0.9	-2.8 ± 1	0.0001
Torticollis	17.6° ± 9.3°	8.3° ± 11.7°	0.03

DMR, double Maddox rods; ET, esotropia; Excyclo, excyclotorsion; Hyper, hypertropia; Hypo, hypotropia; Incyclo, incyclotorsion; PD, prism diopters.

<sup>a</sup>Plus or minus standard deviation.

<sup>b</sup>Paired *t* test.