ABSTRACT

Purpose: To evaluate the surgical results of asymmetric bilateral lateral rectus recession in exotropic Duane retraction syndrome with abnormal face turn toward the opposite side according to secondary deviation measurements.

Methods: Retrospective chart review.

Results: Seven cases of unilateral exotropic Duane retraction syndrome were reviewed. All cases had globe retraction on adduction and exotropia with limited adduction, five of which also had mild limitation of abduction. Four cases had upshoot/downshoot on adduction and all patients had face turn. Exotropia was measured in forced primary position. The average lateral rectus recession was 6.36 mm (range: 5.5 to 7.5 mm) in the affected eye and 7.36 mm (range: 6.5 to 8.5 mm) in the healthy eye. The mean follow-up period was 282 days. Mean exotropia in the forced primary position improved from 27.9 ± 5.7 prism diopters (PD) preoperatively to 7.9 ± 16.8 PD postoperatively (P = .025). Head position resolved completely in all but one case (P = .031). There were no significant changes in ductions.

Conclusions: The results suggest that asymmetric bilateral lateral rectus recession in exotropic Duane retraction syndrome with abnormal head turn posture successfully eliminates abnormal head turn posture and exotropia in most cases.

INTRODUCTION

Exotropic Duane retraction syndrome with abnormal head turn posture toward the opposite side (usually referred to as type II Duane retraction syndrome) is less common than esotropic or orthotropic Duane retraction syndrome and there are limited data regarding the outcome of different surgical approaches.

Exotropic Duane retraction syndrome is believed to occur when the force generated by the lateral rectus muscle through dysinnervation is greater than the corresponding force of the medial rectus muscle. All patients with exotropic Duane retraction syndrome have an adduction deficit and many also have an abduction deficit. In the classification by Huber, exotropic Duane retraction syndrome cases with adduction deficit are considered type II, whereas those with both adduction and abduction deficits are considered type III. Distinguishing between the two former types of Duane retraction syndrome is challenging because the clinical presentation is a result of the severity of one abnormality: the larger the number of oculomotor nerve fibers that move from innervating the medial rectus muscle to innervating the lateral rectus muscle, the more force the medial rectus muscles lose and the lateral rectus muscles gain. Therefore, we preferred to define all cases as exotropic Duane retraction syndrome and to specifically describe individual characteristics of each case (alignment, with/without upshoot/downshoot, limited adduction, abduction) and not according to Huber’s classification.

According to the ophthalmic literature, the most common surgical approach in cases of exotropic...
Duane retraction syndrome is unilateral lateral rectus recession (of either the affected eye or, less commonly, the contralateral eye). The purpose of this study was to report for the first time a series of patients undergoing a bilateral lateral rectus recession procedure for unilateral exotropic Duane retraction syndrome with abnormal head turn posture according to the secondary deviation, as measured by the alternate cover test in forced primary position.

**PATIENTS AND METHODS**

This retrospective study was approved by the institutional review boards of the Tel Aviv Medical Center and Assuta Hospital, Tel Aviv, Israel, and followed the tenets of the Declaration of Helsinki.

We retrospectively collected data from the records of all patients operated on by a single surgeon (CS) between 1990 and 2014 for unilateral exotropic Duane retraction syndrome. Included in this analysis were only patients who had bilateral lateral rectus recession as their first surgical procedure and at least 1 month of postoperative follow-up. The medical records were identified through a manual search of the surgical logbooks in Tel Aviv Medical Center and a computerized search of the surgical logbooks in Assuta Hospital, Tel Aviv, Israel.

For each patient, the following data were reviewed: age at the time of surgery, gender, laterality of the ocular motility defect, presence of head turn, ductions, presence of upshoot or downshoot, globe retraction, type of surgery performed, maximal motor misalignment at near and distance fixation (measured by the alternate cover test at forced primary position), and need for further intervention. Postoperative outcomes were recorded from the last postoperative visit. In a case in which a second procedure was needed, we documented the last follow-up visit prior to the second surgery.

Ductions were graded on a scale from 0 (full ductions) to -4 (inability to move the eye past the midline). The exotropia was always measured at the primary position using the alternate cover test while the prism bar was held in front of the healthy eye to detect the maximal secondary deviation. When there was no movement of the healthy eye when moving the cover from that eye to the eye affected by Duane retraction syndrome, measurement of secondary deviation was achieved.

In all patients, an asymmetric bilateral lateral rectus recession was performed using the fixed recession technique. The amount of recession of the lateral rectus muscle of the affected eye was determined according to the maximal motor exotropia measured at distance fixation in the forced primary position. The lateral rectus muscle of the unaffected eye was recessed by 1 mm more than that of the affected eye.

Y-splitting of the lateral rectus muscle was performed in all cases with upshoot or downshoot according to the technique previously described by Rao et al. Statistical analyses were performed using Statistical Package for the Social Sciences software (version 21.0; IBM Corporation, Armonk, NY). The paired samples Wilcoxon signed-rank test and McNemar test with Yates continuity correction were used to assess the difference between preoperative and postoperative continuous and categorical characteristics, respectively. A two-sided $P$ value of .05 was considered statistically significant.

**RESULTS**

Seven patients (2 males and 5 females) with unilateral exotropic Duane retraction syndrome who had bilateral rectus muscle surgery and a follow-up period of at least 1 month were identified during the 24-year period that was searched. All patients had globe retraction on adduction and exotropia with limited adduction. In 5 of the 7 cases, mild limitation of abduction was documented. All patients had face turn toward the opposite side. Four patients had upshoot/downshoot on adduction.

The mean age at the time of surgery was 14.9 years (range: 3 to 43 years). The left eye was involved in 4 (57%) patients. The mean follow-up period was 282 days (median: 312 days; range: 30 to 678 days). The average lateral rectus recession was 6.36 mm (range: 5.5 to 7.5 mm) in the affected eye, with most patients receiving more than 6 mm of recession.

Evaluations of exotropia and head turn after surgery are depicted in Table 1. Mean exotropia in the forced primary position improved from 27.86 ± 5.7 prism diopters (PD) preoperatively to 7.86 ± 16.8 PD postoperatively ($P = .025$). Head position resolved completely in all but one case ($P = .031$). Figure 1 demonstrates an improved head position after surgery for patient 5. The upshoot/downshoot resolved in all four patients ($P = .031$). There were no significant changes in ductions ($P = .655$ and .180 for
adduction and abduction deficit, respectively. Figure 2 demonstrates both the improvement in head position and exotropia after surgery and the absence of a significant change in ductions for patient 7. Only one patient (patient number 1) needed a second procedure due to persistent exotropia with abnormal head turn. The patient originally had a large exotropia (40 PD). Two months after the first surgery, the patient had a transposition procedure of the superior and inferior rectus muscles nasally with foster augmentation, after which the exotropia improved to 25 PD. Interestingly, on the first postoperative day, there was only one case of overcorrection (patient 3), which resolved within 1 week.

**DISCUSSION**

Duane retraction syndrome is caused by the absence or partial development of the sixth cranial nerve nucleus. The lateral rectus muscle is innervated anomalously by branches from the third cranial

| Patient | Sex | Age (y) | Eye | Alignment (PD) | Extent of Lateral Rectus Recession (mm) | ADD | ABD | US/DS | Ipsilateral | Contralateral | Y-splitting | ABH Resolved | AHP Resolved | Alignment | US/DS | AHP Resolved |
|---------|-----|---------|-----|----------------|----------------------------------------|-----|-----|-------|-------------|-------------|-------------|--------------|-------------|-----------|-------------|-----------|---------|-------------|
| 1       | F   | 31      | Right | 40 XT         | 0                                      | -4  | 0   | No    | Yes          | Yes         | No          | No           | No          | Yes       | Yes         | Yes       |
| 2       | F   | 43      | Left  | 30 XT         | -3                                     | -4  | -4  | No    | Yes          | Yes         | No          | Yes          | No          | Yes       | Yes         | Yes       |
| 3       | M   | 3       | Left  | 25 XT         | -2                                     | -3  | -3  | No    | Yes          | Yes         | No          | Yes          | No          | Yes       | Yes         | Yes       |
| 4       | F   | 3       | Right | 25 XT         | -2                                     | -2  | -2  | No    | Yes          | Yes         | No          | Yes          | No          | Yes       | Yes         | Yes       |
| 5       | M   | 7       | Left  | 25 XT         | 0                                      | -2  | -1  | No    | Yes          | Yes         | No          | Yes          | No          | Yes       | Yes         | Yes       |
| 6       | F   | 5       | Right | 25 XT         | 0                                      | -2  | -1  | No    | Yes          | Yes         | No          | Yes          | No          | Yes       | Yes         | Yes       |
| 7       | M   | 7       | Left  | 25 XT         | 0                                      | -2  | -1  | No    | Yes          | Yes         | No          | Yes          | No          | Yes       | Yes         | Yes       |

PD = prism diopters; ADD = adduction; ABD = abduction; US/DS = upshoot/downshoot; ABH = abnormal head position; XT = exotropia.
nerve with or without subnormal innervation originated from the abducens cranial nerve itself. This causes the pathognomonic co-contraction of the medial and lateral rectus muscles with globe retraction on adduction,\(^9,10\) variable degrees of adduction and abduction limitation, and possible strabismus.

The exotropic type of Duane retraction syndrome (usually referred to as type II Duane retraction syndrome) is a relatively uncommon form. Several surgical methods have been proposed in the ophthalmic literature to treat exotropic Duane retraction syndrome with abnormal head turn posture, most of which are supported by case reports or small case series only.

Merino et al.\(^4\) described the surgical results of simple unilateral rectus recession in two cases of exotropic Duane retraction syndrome with abnormal head turn posture. Resolution of the exotropia was reported in only one of the cases.

Snir et al.\(^5\) recently presented the surgical results of 11 cases of exotropic Duane retraction syndrome. Three patients had ipsilateral lateral rectus recessions with unsatisfactory results and 8 patients had contralateral lateral rectus recession with ipsilateral Y-splitting of the lateral rectus muscle as needed, 7 of which had satisfactory surgical results. Velez et al.\(^6\) described the largest series of unilateral rectus recession together with Y-splitting. In their series of 10 cases, 3 patients required a second procedure, 2 of whom had contralateral lateral rectus recession as a second procedure due to persistent exotropia.

Barbe et al.\(^7\) described a large series of horizontal muscle recession for Duane retraction syndrome. Eight cases of unilateral exotropic Duane retraction syndrome (type II: 5 patients; type III: 3 patients) in this series had an ipsilateral lateral rectus recession procedure, 2 (25%) of which needed a second procedure.

Another surgical approach that was described in three case reports is to weaken the lateral rectus muscle with periosteal fixation or attachment to the lateral canthal tendon.\(^11\,13\)
The current study evaluated the outcome of bilateral asymmetric lateral rectus recession in exotropic Duane retraction syndrome instead of the traditional unilateral lateral rectus recession. In 4 (57%) patients, Y-splitting was performed due to upshoot/downshoot; however, our study was too small to compare the results with and without Y-splitting. Postoperative alignment was good, with resolution of abnormal head turn posture in all but one case in which the original exotropia was high (40 PD).

The mean preoperative exotropia in primary position measured in our study was significantly higher than in both previous larger series by Snir et al.5 and Velez et al.6 (27.9 ± 5.7 PD with the first case included and 25.8 ± 1.9 PD without the first case included compared to 17.3 ± 3.5 PD and 18.4 ± 7.3 PD, respectively; P < .05). We do not know whether this difference is incidental or can be attributed to the different measurement techniques used. In our series, alignment was always measured at forced primary position using the alternate cover test to reveal maximal motor deviation (secondary deviation).

By dividing the recession between the two lateral rectus muscles rather than only one, whether it was either the affected muscle as described by Velez et al.8 or the unaffected muscle as described by Snir et al.,5 we avoided increased abduction deficit.

Jampolsky’s “fixation duress” theory applies to the more common Duane retraction syndrome type I. According to his theory, based on Sherrington’s law of reciprocal innervation and Hering’s law of equal innervation, a weakening procedure for the medial rectus muscle in the unaffected eye obligates stronger innervational impulses for any desired adduction, leading to equal innervation of the yoke lateral rectus muscle in the affected eye and pulling the eye more toward the center.14,15 Although motility laws in Duane retraction syndrome are abnormal, this was found to be successful.16 The senior author of this article (CS) has followed Jampolsky’s technique for both esotropic and exotropic Duane retraction syndrome with head turn for many years.16

An optional explanation for the advantage of asymmetrical surgery is that, by recessing the lateral rectus muscle in the healthy fixating eye more than the affected eye, we increase the stimulation for the yoke medial rectus muscle in the affected eye, thereby pulling the affected eye more toward the center. Of course, this is a theoretical explanation only.

Interestingly, on review of our cases, although our patients had larger angles of exotropia compared to previous series, we had a better success rate. There are three main differences between our case series and previous series: a relatively large exotropia as measured according to secondary deviation, bilaterality of the surgery, and an asymmetric approach. We believe that operating according to secondary deviation measurement leads to sufficient surgery with less undercorrection. Additionally, the bilateral nature of the surgery allows one to operate less on each side and, therefore, cause less impairment in ductions postoperatively. Finally, the difference in the recession between the affected and healthy eyes is relatively small (1 mm); however, in surgical tables of lateral rectus recession, each millimeter in surgery is equivalent to 5 PD of misalignment. Unfortunately, due to the retrospective nature of this study, we cannot compare symmetric to asymmetric bilateral lateral rectus recession for exotropic Duane retraction syndrome.

The results of this study should be interpreted within the context of its limitations. This is a retrospective review and subject to the selection and follow-up bias inherent to all retrospective reviews. Because of the nature of this study and the young age of most patients at the time of surgery, we failed to secure documentation of stereopsis, ocular alignment on side gaze, and precise measurements of abnormal head turn posture before surgery. Because exotropic Duane retraction syndrome is relatively rare and the described surgical approach is the preferred approach of the senior author (CS), we were unable to select an appropriate control group with which to compare the postoperative results to determine whether a different surgical procedure would have worked equally as well or better.

Despite its limitations, this study represents one of the largest series of exotropic Duane retraction syndrome and is the first to describe the results of bilateral lateral rectus recession with an asymmetric approach. We attribute our good success rates to operation according to secondary deviation measurement and the bilateral approach. As previously stated, we do not know whether a symmetric approach would have given us similar results.

REFERENCES


