Combined surgical strategy for management of unilateral exotropic Duane retraction syndrome associated with limitation of abduction

Mohamed F. Farid, MD, FRCS

PURPOSE To report the effect of asymmetrical bilateral lateral rectus recession combined with augmented partial vertical rectus transposition (VRT) in the management of exotropia, head turn, limited abduction, and anomalous vertical movements associated with unilateral exotropic Duane retraction syndrome (XT-DRS).

METHODS The medical records of all patients with unilateral XT-DRS associated with limitation of abduction who underwent surgery during a 5-year period from 2013 to 2018 with at least 6 months' follow-up were reviewed retrospectively. Outcome measures were changes in head turn, primary position distance and near exodeviation, degree of limited abduction, and anomalous vertical movements on adduction.

RESULTS A total of 11 patients (6 males) were included. Mean patient age was 16.3 years (range, 6-29). Exodeviation at distance and near fixation were corrected by means of 26.4$^{\Delta}$ and 24.8$^{\Delta}$, respectively. Head turn was improved by a mean of 17.3°. Limited abduction and anomalous vertical movements were corrected by means of 1.6 and 1.5 units, respectively. No patients developed symptomatic induced vertical deviation or anterior segment ischemia.

CONCLUSIONS In our study cohort, the combined strategy of asymmetrical bilateral lateral rectus recession with unilateral augmented partial VRT yielded satisfactory results in the management of unilateral XT-DRS associated with limited abduction with no recorded intra- or postoperative complications. (J AAPOS 2019;23:323.e1-5)

Unilateral exotropic Duane retraction syndrome (XT-DRS) associated with limitation of abduction, which accounts for approximately 17%-26% of all cases of Duane syndrome,[1-3] is characterized by primary position exotropia with abnormal head posture (AHP), anomalous vertical movements, and globe retraction on adduction as well as by variable degrees of limited abduction.[4,5] Although its clinical features are well recognized, management of XT-DRS remains challenging. Recession of the lateral rectus muscle, either unilaterally or bilaterally, is the most common surgical approach; however, in cases with preexisting limitation of abduction, lateral rectus recession also accentuates the limitation as it improves the degree of exotropia.[6] Few publications in the literature address management of the associated clinical problems of exotropia and limitation of abduction in XT-DRS. The current study evaluated the results of a combined surgical strategy tailored to correct the primary position exotropia by asymmetrically recessing both lateral rectus muscles as well as improving limitation of abduction and vertical shooting by partial vertical rectus transposition augmented by dual scleral fixation and loop myopexy sutures in unilaterally affected patients.

Subjects and Methods

This study was approved by the Institutional Review Board of Benha University Hospital. The medical records of patients with unilateral XT-DRS associated with limited abduction who underwent asymmetrical lateral rectus recession combined with unilateral dually augmented partial vertical rectus transposition (VRT) at Benha University Hospital from 2013 to 2018 were retrospectively reviewed. Only unilaterally affected patients with limited abduction who underwent the combined procedure were included. Patients with XT-DRS, either unilateral or bilateral, who were associated with isolated limitation of adduction or combined limitation of abduction and adduction were excluded, as were those who had undergone previous strabismus surgery or had <6 months' follow-up.

Patients received complete ophthalmic and orthoptic assessment preoperatively, including best-corrected visual acuity

Author affiliations: Department of Ophthalmology, Benha Faculty of Medicine, Benha University, and Benha University Hospital, Benha, Egypt
Submitted February 25, 2019.
Revision accepted September 6, 2019.
Published online November 2, 2019.
Correspondence: Mohamed F. Farid, MD, FRCS, PO Box 13511, 1 Fareed Nada Street, Benha, Egypt (email: mohamed_fathy_10@yahoo.com).

1091-8531/$36.00
https://doi.org/10.1016/j.jaapos.2019.09.013

measurement, cycloplegic refraction, stereopsis (Titmus fly test), and slit-lamp and fundus examination. Head turn, if present, was assessed by observation and photographic documentation and quantified using an orthoptic goniometer with a 5° scale while the patient fixated on a target at 6 m. Angle of deviation in the forced primary position was estimated by alternate prism and cover test while the patient fixated on targets at 6 m and 0.33 m, respectively, and the largest angle measured at distance and near was used to guide the amount of lateral rectus recession required. Assessment of defective abduction was performed using a four-point scale (−3 to 0), which was modified to match the fact that affected eyes were already exotropic in forced primary position (meaning they already passed the midline). In this modified scale, −3 referred to patients who could not abduct their eyes from their presenting exotropic position, whereas 0 referred to full abduction in which the temporal cornea reached the lateral canthus. Degrees of limited abduction were given intermediate values of −2 and −1.

Anomalous vertical movements were measured on a four-point scale similar to the one used for quantification of inferior oblique overaction, with +1 referring to minimal anomalous vertical movements and +4 referring to up- or downward globe rotation to the limit that the cornea was merely visible.[7] Globe retraction was assessed preoperatively and was documented as present or absent without quantitative measurements. Postoperatively, changes in globe retraction were assessed by comparing current photographs with preoperative clinical photographs and were documented as improved, stationary, or worsened.

All procedures were performed under general anesthesia by the same consultant (MFF). After intraoperative forced duction testing, a large (270°) limbal conjunctival periotomy with generous posterior release was performed in the affected eye to gain access to the lateral rectus, superior rectus, and inferior rectus muscles. Lateral rectus recession was performed using the conventional scleral fixation technique, followed by partial superior rectus and inferior rectus transpositions, as described by Britt and colleagues,[8] and each transposed muscle was dually augmented by loop myopexy suture combined with posterior scleral fixation suture 10 mm posterior to the lateral rectus muscle insertion, as described by Farid.[9] The superior rectus and inferior rectus muscles were isolated and cleaned from their fascial connections and septae. Each muscle was then split posteriorly into temporal and nasal halves from its insertion site, with preservation of nasal anterior ciliary vessels in the attached nasal portion of the muscle. The temporal portion of each muscle was then sutured using single-double armed 6-0 polyglactin 910 suture before it was disinserted, laterally transposed, and reattached to the corresponding upper and lower poles of lateral rectus muscle insertion along the spiral of Tillaux. Partial VRT was dually augmented by muscle-to-muscle union sutures that approximate each transposed segment of superior rectus and inferior rectus muscles to the corresponding upper and lower halves of the lateral rectus using a nonabsorbable 5-0 polyester suture, including a scleral fixation close to the edge of the lateral rectus muscle and at 10 mm posterior to the original lateral rectus muscle insertion. At the end of the surgery, forced duction was performed to ensure that there was no restriction on adduction. The lateral rectus muscle of the contralateral free eye was recessed using a fixed scleral suture technique. The amount of lateral rectus recession was based on the largest angle of deviation measured preoperatively and was asymmetrically split between both eyes, with the recession of the lateral rectus muscle in the noninvolved eye recessed by 2 mm more than the recession in the involved eye.

At each postoperative visit, all patients were subjected to the same examinations performed preoperatively. Careful slit-lamp examination of the anterior segment was performed at each follow-up visit to detect any signs of anterior segment ischemia. Surgical results were obtained from postoperative patients' notes. Postoperatively, outcomes included correction of primary position exotropia, head turn, degree of limitation of abduction, and change of anomalous vertical movements. Statistical analysis was performed using SPSS software (SPSS for Windows V.17.0; SPSS Inc, Chicago, IL). A paired t test was used to compare pre- and postoperative values. A P value of <0.05 considered significant.

Results

A total of 11 patients (6 males) with unilateral XT-DRS were included. Mean patient age at surgery was 16.3 years (range, 6-29 years). In 7 patients the left eye was involved. Mean follow-up was 12.2 months (range, 6-21 months). Preoperatively, no amblyopia was observed in any patient. Mean spherical refractive error was -0.3 ± 0.6 in the right eye and -0.4 ± 0.7 in the left eye. Mean best-corrected visual acuity was 0.8 ± 0.1 in the right eye and 0.9 ± 0.08 in the left eye. Patient characteristics before and after surgery are provided in eSupplement 1 (available at jaapos.org).

On intraoperative forced duction testing, positive restriction on adduction was found in 10 cases: 6 severe, 3 moderate, and 1 mild. The mean amount of lateral rectus recession was 6.2 mm (range, 4–7 mm) in the involved eye and 8.2 mm (range, 6–9 mm) in the contralateral eye. Following the combination procedure, there was significant improvement of distance exotropia, near exotropia, AHP, limitation of abduction ($P < 0.00001$), and anomalous vertical movements ($P = 0.0013$). See Table 1. Preoperatively, 9 of 11 patients (82%) showed anomalous vertical movements on adduction (5 patients upshoot and 4 patients downshoot). Of those patients, anomalous vertical movements improved in all but 1 patient with mean values improved from 1.8 (range, 0-4) to 0.27 (range, 0-1). See Figure 1. Globe retraction on adduction was observed in 9 patients preoperatively (81%). Postoperatively, 7 patients had their globe retraction improved as evident by comparison with preoperative photographs (Figure 1), whereas in 2 patients, globe retraction remained unchanged, and no patient showed worsening of the preoperative globe retraction.

Stereopsis data was available for 10 patients, 2 of whom had no stereoacuity pre- or postoperatively. Of the remaining 8 patients, 5 had stereoacuity of 100–3000 arcsec, and 3 had stereoacuity of >100 arcsec preoperatively. After surgery, 5 patients had stereoacuity of >100 arcsec, and 3

Table 1. Mean and ranges of pre- and postoperative values for main study parameters

Item	Preoperative (range)	Postoperative (range)	Mean change (range)	*P* value
Near XT, PD	28 (12-40)	3.2 (0-6)	24.8 (8-34)	<0.00001
Distance XT, PD	33.6 (20-45)	7.2 (0-12)	26.4 (10-40)	<0.00001
AHP, degrees	21.3 (10-40)	4 (0-10)	17.3 (10-30)	<0.00001
Limited abduction	−2.4 (−2 to −3)	−0.8 (0 to −2)	1.6 (1-2)	<0.00001
Vertical shooting	1.8 (0-4)	0.27 (0-1)	1.5 (0-4)	0.0013

AHP, abnormal head posture; *PD*, prism diopter; *XT*, exotropia.

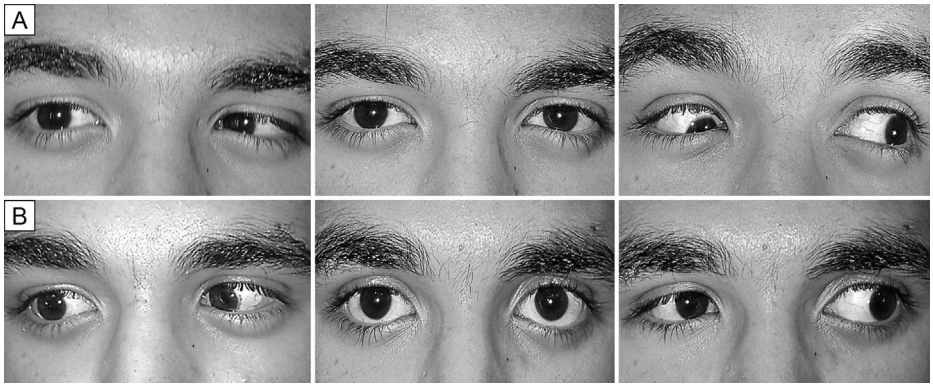

FIG 1. Pre- (A) and postoperative (B) clinical photographs of patient 1, with right XT-DRS associated with −2 abduction limitation and +3 downshoot on adduction. Limited abduction improved to −1 with complete elimination of downshoot on adduction after asymmetrical lateral rectus recession (6 mm in the right eye and 8 mm in the left eye) combined with dually augmented vertical rectus transposition in the right eye.

had stereoacuity of 100–3000 arcsec. Mean stereoacuity improved from 682.5 (range, 80–3000 arcsec) to 297.5 (range, 60–800 arcsec) postoperatively.

No intraoperative complications were reported in any patient. Postoperatively, no limitation of adduction was noted in any patient. There were no recorded cases of induced vertical deviations or limitation of abduction in the contralateral eye. No patient reported postoperative subjective torsional diplopia nor showed signs of anterior segment ischemia.

Discussion

In two large studies that reviewed the demographics and subtypes of large groups of DRS patients in two different geographical locations,[1,2] unilateral XT-DRS associated with limitation of abduction constituted a considerable percentage of Duane retraction syndrome patients. In those trials, 118 of 441 patients (26.7%) and 67 of 404 (16.5%) had unilateral XT-DRS associated with limitation of abduction, respectively. However, to our knowledge, there has been no published specific surgical strategy to manage unilateral XT-DRS with limited abduction, with its combined challenging features of exotropia (in which the lateral rectus recession seems a reasonable option) and limitation of abduction (which is accentuated by lateral rectus recession). The current study reviews the results of a combined surgical strategy tailored to alleviate comorbidities associated with unilateral XT-DRS with limitation of abduction. In general, we have found that the combined technique is effective in the correction of primary position exotropia and AHP and has the added advantage of improving the limitation of abduction and anomalous vertical movements in patients with unilateral XT-DRS associated with limitation of abduction.

Unilateral or bilateral recession of the lateral rectus muscle has been described as the procedure of choice to correct primary position exotropia and AHP associated with XT-DRS.[10-12] Theodorou and Burke[12] reported that in 82% of their bilateral XT-DRS patients exotropia resolved postoperatively to within 10^{Δ} of orthotropia/phoria, and 86% of those with AHP experienced resolution or improvement after lateral rectus recession. However, 6 of 11 patients showed an increase in limitation of abduction postopertively.[12] Barbe and colleagues[10] retrospectively reviewed 59 DRS patients, of whom 11 (8 unilateral) had XT-DRS, with variable degrees of limitation of abduction. Following lateral rectus recession, exotropia improved significantly in 8 patients (73%); AHP, in 7 (64%). However, limitation of abduction worsened postoperatively in 5 cases, remained stationary in 5 cases, and improved in only 1 case. Farid and colleagues[11] performed large (8.5–10 mm) lateral rectus recession in unilateral XT-DRS and reported that although primary position exotropia and AHP were statistically improved, the mean limitation of abduction increased from −3 to −3.6 postoperatively. Compared with the previously published reports,

restriction of the lateral rectus muscle in the current series improved statistically from −2.4 preoperatively to −0.8 postoperatively.

Kekunnaya and colleagues[6] have stated that although lateral rectus recession was the recommended treatment for XT-DRS, it worsened limitation of abduction, if present. Therefore, they advised that asymmetrical lateral rectus recession be performed in cases with unilateral XT-DRS, with the greater amount of recession to be performed in the contralateral free eye. However, no data was available to support this suggestion. Recently, Mezad-Koursh and colleagues[13] evaluated the results of asymmetrical bilateral lateral rectus recession, with recession of the lateral rectus in the unaffected eye by 1 mm more than in the affected eye, in 7 patients with unilateral XT-DRS associated with isolated limitation of adduction or combined limitation of abduction and adduction. They reported improvement of exotropia from 28^{Δ} to 8^{Δ}, with complete resolution of AHP in all but 1 case. However, preoperative limitation of abduction and adduction remained unchanged after surgery. In the current series, the recession of the lateral rectus muscle was asymmetrically divided between both eyes, with the recession of the lateral rectus in the contralateral free eye 2 mm more than the lateral rectus recession in the affected eye. Decreasing the amount of lateral rectus recession in the affected eye would minimize the negative effect of muscle recession on the abducting force of an already compromised lateral rectus muscle, and this would yield the maximum chance for the success of the transposition procedure, which, in the current study, has resulted in significant improvement of abduction.

To overcome the problem of the limitation of abduction in Duane retraction syndrome, a variety of vertical rectus lateral transposition procedures have been proposed. They include lateral transposition of full vertical rectus muscles,[14] parti,al vertical rectus muscles,[8] or even the superior rectus muscle (Johnston SC, et al. Invest Ophthalmol Vis Sci 2006;47: ARVO e-Abstract 2475), with the option of augmenting each procedure of transposition by incorporating part of the transposed muscle to the sclera,[15] the lateral rectus muscle,[16] or both.[9] VRT combined with other horizontal rectus muscle's procedures has been advocated by some authors to solve ocular deviation in the primary position associated with limited ocular duction in Duane retraction syndrome.[14,17] Sharma and colleagues[18] performed partial VRT combined with periosteal fixation of the lateral rectus muscle in 7 XT-DRS patients and found that abduction was improved from −3.6 to −2.8, and exotropia was corrected from 21.3^{Δ} to 8^{Δ}. Britt and colleagues[8] reported the results of partial VRT in 5 esotropic Duane retraction syndrome patients with limited abduction. Improved abduction was noted in 3 patients; the transposition was combined with ipsilateral weakening of the lateral rectus muscle to reduce co-contraction and pseudoptosis. Potential complications associated with VRT include anterior segment ischemia and induced vertical deviations.[8,14]

Another advantage of the combined strategy presented here is its efficacy in treating anomalous vertical movements on adduction associated with XT-DRS, with no need for additional procedures. Fixation of the lateral rectus muscle to both vertical rectus muscles and to the sclera prevents its slippage above or below the globe during adduction, which is responsible for the phenomenon of anomalous vertical movements noticed on adduction. Stabilization of the lateral rectus muscle to the sclera by any means, including scleral fixation, has proved effective in the management of anomalous vertical movements associated with DRS (Eisenbaum AM, Parks MM. A study of various surgical approaches to the leash effect in Duane's syndrome. AAPOS and AAO, Chicago, IL, November 5, 1980).[19] In some previous reports, significant reduction of anomalous vertical movements was recorded after isolated lateral rectus recession in XT-DRS.[7] In the same way, lateral rectus recession in the Duane syndrome eye could contribute to the improvement of anomalous vertical movements recorded in the current study. With dual augmentation of VRT performed in the current series, XT-DRS patients with anomalous vertical movements on adduction could have been spared procedures that may cause more weakening of an already compromised muscle, such as Y splitting of the lateral rectus muscle.

References

1. Kekunnaya R, Gupta A, Sachdeva V, et al. Duane retraction syndrome: series of 441 cases. J Pediatr Ophthalmol Strabismus 2012;49:164-9.
2. Khan AO, Oystreck DT, Wilken K, Akbar F. Duane retraction syndrome on the Arabian Peninsula. Strabismus 2007;15:205-8.
3. Park WH, Son DH, Yoon SW, Baek SH, Kong SM. The clinical features of Korean patients with Duane's retraction syndrome. Korean J Ophthalmol 2005;19:132-5.
4. Duane A. Congenital deficiency of abduction associated with impairment of adduction, retraction movements, contraction of the palpebral fissure and oblique movements of the eye. Arch Ophthalmol 1905;34:133-59.
5. Huber A. Electrophysiology of the retraction syndromes. Br J Ophthalmol 1974;58:293-300.
6. Kekunnaya R, Kraft S, Rao VB, Velez FG, Sachdeva V, Hunter DG. Surgical management of strabismus in Duane retraction syndrome. J AAPOS 2015;19:63-9.
7. Farid MF. Y-split recession vs isolated recession of the lateral rectus muscle in the treatment of vertical shooting in exotropic Duane retraction syndrome. Eur J Ophthalmol 2016;26:523-8.
8. Britt MT, Velez FG, Thacker N, Alcorn D, Foster RS, Rosenbaum AL. Partial rectus muscle-augmented transpositions in abduction deficiency. J AAPOS 2003;7:325-32.
9. Farid MF. Dual-augmented transposition of vertical recti in chronic abducens palsy. Am J Ophthalmol 2019;197:59-64.
10. Barbe ME, Scott WE, Kutschke PJ. A simplified approach to the treatment of Duane's syndrome. Br J Ophthalmol 2004;88:131-8.
11. Farid M, Saeed A, Elbarki A. Unilateral large lateral rectus recession in treatment of unilateral exotropic Duane retraction syndrome. J Egypt Ophthalmol Soc 2016;109:5-9.

12. Theodorou N, Burke J. Surgical and functional outcomes in bilateral exotropic Duane's retraction syndrome. Br J Ophthalmol 2013;97:1134-7.
13. Mezad-Koursh D, Leshno A, Klein A, Stolovich C. The efficacy of bilateral lateral rectus recession according to secondary deviation measurements in unilateral exotropic Duane retraction syndrome. J Pediatr Ophthalmol Strabismus 2018;55:47-52.
14. Rosenbaum AL. Costenbader Lecture. The efficacy of rectus muscle transposition surgery in esotropic Duane syndrome and VI nerve palsy. J AAPOS 2004;8:409-19.
15. Foster RS. Vertical muscle transposition augmented with lateral fixation. J AAPOS 1997;1:20-30.
16. Wright KW. Color Atlas of Strabismus Surgery: Strategies and Techniques. 2nd ed. New York: Springer; 2007.
17. Jampolsky A. Duane syndrome. In: Rosenbaum AL, Santiago AP, eds. Clinical Strabismus Management. Philadelphia: WB Saunders Co; 1999.
18. Sharma P, Tomer R, Menon V, Saxena R, Sharma A. Evaluation of periosteal fixation of lateral rectus and partial VRT for cases of exotropic Duane retraction syndrome. Indian J Ophthalmol 2014;62:204-8.
19. Berndt K, Berndt E. Operative treatment of Duane's syndrome associated with extreme vertical divergence [in German]. Klin Monbl Augenheilkd 1979;174:62-7.