Unilateral large lateral rectus recession in treatment of unilateral exotropic Duane retraction syndrome

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Purpose
The purpose of this study was to determine the effect of unilateral large lateral rectus (LR) recession on head turn and horizontal ocular deviation in patients with unilateral exotropic Duane retraction syndrome.

Patients and methods
This was a prospective, interventional study of 10 consecutive patients with unilateral exotropic Duane retraction syndrome associated with contralateral head turn. All patients underwent large (8.5–10 mm) unilateral LR recession on the affected eye. All patients were followed up for at least 6 months. Preoperative and postoperative ocular deviation, angle of head turn, stereoacuity, and severity of limitation in abduction and adduction were recorded and analyzed.

Results
Mean head turn decreased from 30.5±9.56° (range, 15°–40°) to 6.5±4.74° (range, 0°–15°) (P<0.001). Mean exotropia decreased from 33.0±6.32 prism diopter (range, 25–45 prism diopter) to 5.7±4.9 prism diopter (range, 0–15 prism diopter) (P<0.001). Mean limitation in adduction decreased from 2.8±0.79 to 1.6±0.52 (P<0.001). Mean limitation in abduction increased from 3.0±1.16 to 3.6±0.7 (P=0.024). All patients had variable degrees of stereopsis (ranging between 100 and 800 arcsec, which remain unchanged postoperatively).

Conclusion
Unilateral large LR recession abolishes or ameliorates the wide scale of head turn and exotropia in unilateral exotropic Duane retraction syndrome with noticeable improvement of defective adduction.

Keywords:
abnormal head posture, duane syndrome, exotropia, lateral rectus recession

Introduction
Duane retraction syndrome is a well-known form of congenital incomitant ocular motility disorders. Its main features are limited abduction and/or adduction, globe retraction, and vertical shooting on attempted adduction [1]. The underlying pathology of Duane syndrome was thoroughly studied, with most recent researches revealing an underlying agenesis of the sixth cranial nerve nuclear complex. All ocular motility features were found to be secondary to the original neurological developmental defect [2].

Since the electromyography findings of Huber [3], Duane syndrome was classified into three main types: type 1 with mainly a limitation in abduction, type 2 with limited adduction, and type 3, which carries features of both. In types 2 and 3, which are relatively rare, the patient is usually exotropic and adopts face turn to the contralateral side to maintain binocularity. Upshoots and downshoots are relatively more common in these subtypes. Exotropia (EX) occurs in Duane syndrome when lateral rectus (LR) dominates its antagonist either neurologically secondary to misinnervation or mechanically secondary to fibrotic contracture. In cases with normal abduction, EX will follow as a result of strong unopposed action of the LR; however, EX can still occur in cases with diminished abduction secondary to synergistic divergence [4].

In exotropic Duane, surgery is mainly indicated for large primary position EX and significant head turn. Additional surgical goals such as severe vertical shooting and globe retraction are addressed and managed accordingly [5,6]. Many authors pointed out the number of surgical approaches to treat exotropic Duane, mainly targeting weakening of LR muscle by recession either unilaterally or bilaterally [5,7]. This study reviews the surgical results in a selected group of patients with unilateral exotropic Duane treated with unilateral LR muscle recession (8.5–10 mm from original muscle insertion) and

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analyzes the effect of surgery on a wide range of head turn and primary position EX.

Patients and methods
The study protocol was approved by Benha University Research Ethics Committee. The study and data collection conformed to all local laws and complied with the principles of the Declaration of Helsinki. A prospective study was performed on all patients with unilateral exotropic Duane syndrome with significant head turn and primary position EX from May 2011 to April 2014. Inclusion criteria include patients with unilateral limitation of adduction and/or abduction associated with significant head turn (≥15°) and primary position EX (≥10 prism diopter). Cases of bilateral exotropic Duane were excluded from the study. Unilateral exotropic Duane patients with primary position EX and head turn less than 10 prism diopter and 15°, respectively, were also excluded, as well as patients who had previous eye muscle surgery.

All patients underwent complete orthoptic and ophthalmic examination at least twice preoperatively and at each postoperative visit. Ocular examinations included assessment of visual acuity by Snellen or E chart, fixation pattern, cycloplegic refraction, and fundus examination. Significant refractive errors were corrected by appropriate glasses. None of the patients were found to be amblyopic. Cover test was done for near at 1/3 m and at distance 6 m. Head turn was determined by observing the angle suspended between the patient’s primary position during visual acuity measurement and patient’s nose, and it was measured by an orthopedic goniometer with a five-degree scale. Strabismus measurements were taken by alternate prism cover test in forced primary position for near and distance using loose prisms. Assessment of ocular motility was performed, and any limitations were documented on a scale of 0–4. Binocularity was assessed using Titmus fly test while patients were allowed to use their compensatory head turn. Patient’s head turn, horizontal deviation, and ocular versions were recorded preoperatively and at each postoperative visit. No electromyography confirmation was performed. Informed consent for the surgery was obtained from all patients after thorough explanation of all surgical steps and potential complications.

Intraoperative forced duction testing was performed on the Duane eye and revealed combined limitation of abduction and/or adduction of variable intensities in all cases. All patients underwent large (8.5–10 mm) LR recession on the affected eye performed by the same surgeon (M.F.F.). The choice of recession dose depended on the magnitude of the abnormal head posture (AHP) and EX. Higher grades of AHP and EX (≥40) received maximum recession (10 mm), whereas lower grades received 8.5 mm recession. Recession was performed using standard limbal approach, and the LR muscle was anchored to sclera at the selected distance measured from the original muscle insertion. Postoperative evaluations were performed at 1 week, 1, 3, and 6 months. The clinical data obtained at 6 months postoperatively were recorded on a report form and compared with the baseline measurements. Surgical intervention was considered successful when postoperative head turn decreased to less than or equal to 15° and when primary position EX was within 10 prism diopter. These data were tabulated and analyzed using the computer program statistical package for social science, version 16 (SPSS Inc., Chicago, Illinois, USA). In the statistical comparison between the different groups, the significance of difference was tested using paired t-test and Wilcoxon test. A P value less than 0.05 was considered statistically significant, and P value less than 0.01 was considered highly significant in all analyses.

Results
Ten unilateral exotropic Duane patients with contralateral head turn fulfilled the inclusion criteria. Summary of raw preoperative and postoperative data including head turn, EX, stereopsis, and limitation of horizontal ductions are shown in Table 1.

Mean age of patients at the time of surgery was 7.9 ±2.33 years (range, 5–13 years). Female-to-male ratio was 3 : 2. The left eye was involved in eight of the patients (80%). None of the patients were ambylopic. All patients had variable degrees of stereopsis with a mean of 400.0±294.39 arcsec (ranging between 100 and 800 arcsec), which remain unchanged postoperatively.

Mean head turn decreased from 30.5±9.56° (range, 15°–40°) preoperatively to 6.5±4.74° (range, 0°–15°) at the last postoperative visit (P≤0.001). Mean EX decreased from 33.0±6.32 prism diopter (range, 25–45 prism diopter) to 5.7±4.9 prism diopter (range, 0–15 prism diopter) (P≤0.001) (Fig. 1). Orthotropia in primary position was achieved in one patient (10%). In seven patients (70%), EX had decreased to be within the fusional range of 10
In two cases (20%), EX stabilized at 12 and 15 prism diopter postoperatively; one did not need further interventions, as visual acuity and stereopsis were stable, whereas the other underwent contralateral LR recession because of cosmetic requirements.

Preoperatively, all patients had limitation of abduction and adduction of variable severity. Mean limitation of adduction decreased from 2.8±0.79 (range, 2 to 4) to 1.6±0.52 (range, 1 to 2) (P≤0.001). Mean limitation in abduction increased from 3.0±1.16 (range, 1 to 4) to 3.6±0.7 (range, 4 to 2) (P=0.024) (Fig. 2). Four patients had preoperative vertical shooting on adduction (two upshoots and two downshoots). Vertical shooting responded significantly to LR recession alone with no added procedures (Fig. 3). None of the patients experienced postoperative consecutive esotropia or diplopia.

**Discussion**

Many factors stand behind the difficulty in the management of Duane retraction syndrome such as its mysterious pathophysiology together with its wide

### Table 1 Preoperative and postoperative patients' raw data

| Case | Demography | Preoperative AHP (deg.) | Preoperative XT | Preoperative duction | Stereopsis (sec.) | Surgical procedure (mm) | Postoperative AHP (deg.) | Postoperative XT | Postoperative duction |
|------|-------------|-------------------------|----------------|----------------------|------------------|------------------------|--------------------------|----------------|----------------------|
| 1    | OS, 7 years, female | 40 | 40Δ | LR – 4/MR – 2 | 800 | LRR: 10 | 15 | 12Δ | LR – 4/MR – 2 |
| 2    | OS, 6 years, male | 35 | 35Δ | LR – 3/MR – 2 +upshoot | 200 | LRR: 8.5 | 5 | 8Δ | LR – 3/MR – 1 no upshoot |
| 3    | OD, 8 years, female | 40 | 25Δ | LR – 3/MR – 3 | 400 | LRR: 8.5 | 10 | 4Δ | LR – 4/MR – 2 |
| 4    | OS, 6 years, male | 20 | 30Δ | LR – 4/MR – 4 ++ upshoot | 200 | LRR: 8.5 | 0 | 2Δ | LR – 4/MR – 2 no upshoot |
| 5    | OS, 5 years, female | 25 | 35Δ | LR – 1/MR – 3 +downshoot | 100 | LRR: 8.5 | 5 | 0Δ | LR – 2/MR – 2 no downshoot |
| 6    | OS, 7 years, female | 40 | 45Δ | LR – 4/MR – 3 | 800 | LRR: 10 | 10 | 15Δ | LR – 4/MR – 2 |
| 7    | OS, 10 years, male | 35 | 35Δ | LR – 3/MR – 2 | 400 | LRR: 8.5 | 5 | 2Δ | LR – 4/MR – 1 |
| 8    | OS, 9 years, female | 35 | 25Δ | LR – 3/MR – 2 | 100 | LRR: 8.5 | 10 | 4Δ | LR – 4/MR – 1 |
| 9    | OS, 8 years, female | 15 | 30Δ | LR – 4/MR – 4 | 800 | LRR: 8.5 | 0 | 8Δ | LR – 4/MR – 2 |
| 10   | OD, 13 years, male | 20 | 30Δ | LR – 1/MR – 3 +downshoot | 200 | LRR: 8.5 | 5 | 2Δ | LR – 3/MR – 2 no downshoot |

Δ, prism diopter; AHP, abnormal head posture; LR, lateral rectus; LRR, lateral rectus recession; MR, medial rectus; OD, right; OS, left; XT, exotropia.
range of subtypes and diverse spectrum of the presenting signs. The difficulty is experienced in the management of the rare subtype of the syndrome, exotropic Duane, to a greater extent [8,9]. Poor surgical results that were obtained in literature could be explained by noncommitment to strict indications of surgical intervention and – to a great extent – the lack of universally agreed surgical protocol and dosage to manage such cases [10].

Classically, the main indications for surgical intervention in Duane syndrome have been to correct strabismus in the primary position together with elimination of AHP without causing an overcorrection or additional limitation of ocular motility. Management of globe retraction, upshoots, and downshoots where those signs are evident need additional surgical modifications. There are not many published articles about surgical management of unilateral Duane syndrome, especially exotropic subtype [11]. The current study evaluated the procedure of large unilateral LR muscle recession, and assessed its outcome regarding head turn and primary position EX in a consecutive series of patients.

The results of the current work revealed the success of this simple procedure in improving face turn in all patients (postoperative face turn ≤15°). With regard to primary position EX, 80% of the patients are considered successful ‘EX ≤10 prism diopeter’, whereas only one patient required additional contralateral LR recession for cosmetically unacceptable residual EX of 15 prism diopeter. No changes were reported regarding the level of the preoperative visual acuity and stereopsis secondary to this procedure.

In Barbe’s et al. [8] study, eight patients out of 59 were unilateral exotropic Duane. Those patients were treated by unilateral LR recession but with different and diverse surgical recession doses. Four patients (50%) received large recession (range 7–8.5 mm), whereas the others were treated with less amounts of LR recession (range 3.5–6 mm). All patients of the large recession group achieved acceptable postoperative results, whereas two patients of the less recession group needed additional recessions for improving the significant postoperative AHP. The results of Barbe’s large recession group considerably match current series’ results.

Unilateral LR recession combined with Y-splitting of LR to correct upshoots in unilateral exotropic Duane had been reported in the past. Sukhija et al. [12] performed super-maximum LR recession (10–12 mm) with an average of 10.3±0.8 mm, combined with Y-splitting for the treatment of unilateral exotropic Duane with severe upshoot and downshoot on adduction. According to their results, head turn improved from 27.9±6.4° to 2.6±4.4° and EX from 26.4±5.6 to 2.28±4.07 prism diopter. Upshoot decreased from 2.8±0.44 to 1.0±0.70 and downshoots from 2.5±0.70 to 0. In another study by Velez et al. [13] with a mean LR recession of 8.7±2.9 mm, head turn decreased from 12.7±4.4° to 4.8±5.3°. EX improved from 18.4±7.3 to 6.2±5.9 prism diopter postoperatively. Added Y-shaped LR splitting decreased upshooting from 2.2±0.7 to 1±1.6 and downshooting from 1.9±1.2 to 0.2±1.6.

Natan and Traboulsi [11] aimed at evaluating the results of single muscle recession (medial or LR muscle) in the treatment of all subtypes of Duane syndrome (esotropic and exotropic). Only three cases out of 27 were exotropic. They underwent average unilateral LR recession of 6.3 mm (range 5–7 mm). Unfortunately, there were no specific preoperative or postoperative data for those particular patients.

Barbe et al. [8] recessed single horizontal muscle in 59 patients with esotropic and exotropic Duane syndrome. Postoperatively, 85% of all their patients achieved ≤ 10° head turn. Kubota et al. [14] retrospectively analyzed 124 patients of all subtypes of Duane syndrome who underwent the same approach; 89% of cases showed improvement in primary horizontal deviation and AHP. Kaban et al. [15] performed unilateral medial rectus recession for their 28 unilateral Duane type 1 patients. They reported that 71% of their patients showed reduction of head turn from 19° preoperatively to 5° postoperatively.
According to Barbe et al. [8], most of their patients who had preoperative version deficit in the affected gaze of −3 or worse were improved, but the deficits were never eliminated and normal motility was not restored. Furthermore, they concluded that motility of opposite gaze may become more restricted. On the other hand, the abduction deficits of all patients of the current study were improved with variable amplitudes. Half of our patients showed stabilization of their abduction deficits, whereas the other half experienced an expected increase in limitation of abduction, as a result of the large LR recession. More importantly, such increased limitation had no effect on postoperative AHP and ocular primary position. In agreement with Barbe et al.[8], there was no relation between version deficit and the AHP in both preoperative and postoperative measurements. Despite persistent postoperative limited version, strabismus and head turn nearly totally disappeared and did not recur. Barbe et al. [8] concluded that the AHP was not the result of the deficit versions, but of the need to obtain binocularity. However, their opinion could be argued, as AHP could be found in cases that have binocularity and also in amblyopic cases.

Recession of normal LR in contralateral dominant eye is discouraged by some surgeons. They claim that on doing so more innervation is needed to keep the eye in primary position, which will flow to the affected eye, thus increasing the anomalous vertical movements and retraction [13].

Many authors have shared their experience regarding single muscle recession for the treatment of Duane syndrome [11,14,15]. However, their studies were lacking common protocol and surgical dose for such weakening. In this study, we review our results after large recession of LR to treat a wide range of primary position EX and head turn in unilateral exotropic cases. The results are encouraging, and this simple procedure seems to work fairly well on a wide scale of AHP and EX in presented cases with no reported clinically significant undercorrections or overcorrections.

The results of the current study should be interpreted in the context of its limitations. First, it includes a limited number of patients. However, this could be compensated by a narrow spectrum of inclusion criteria that involve only unilateral cases of the rare exotropic form of Duane syndrome. Second, there is a lack of an appropriate control group to compare with whether a different surgical option would be equally effective or even better. Finally, given the small age group of selected patients, it was difficult to assess the result of surgery on patients’ binocular single field of vision.

Despite its limitations, this study reveals that unilateral large LR recession for patients with unilateral exotropic Duane syndrome and significant head turn is almost equally effective over a broad range of signs. Overall, ocular deviation, AHP, and adduction improved significantly without imposing undercorrections or overcorrections.

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Conflicts of interest
There are no conflicts of interest.

References
1 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; 34:133–159.
2 Miller NR, Kiel SM, Green WL. Clark AW. Unilateral Duane’s retraction syndrome (Type 1). Arch Ophthalmol 1982; 100:1468–1472.
3 Huber A. Electrophysiology of the retraction syndromes. Br J Ophthalmol 1974; 58:293–300.
4 Souza-Dias C. New considerations about Duane’s syndrome. Rev Bras Oftalmol 2009; 68:107.
5 DeRespinis PA, Caputo AR, Wagner RS, Guo S. Duane’s retraction syndrome. Surv Ophthalmol 1993; 38:257–288.
6 Jampolsky A. Duane’s syndrome. In: Rosenbaum AL, Santiago AP, editors. Clinical strabismus management. Philadelphia, PA: WB Saunders; 1999. 325–346.
7 Kraft SP. A surgical approach for Duane syndrome. J Pediatr Ophthalmol Strabismus 1968; 25:119–130.
8 Barbe ME, Scott WE, Kutschke PJ. A simplified approach to the treatment of Duane’s syndrome. Br J Ophthalmol 2004; 88:131–138.
9 Saunders RA, Wilson ME, Bluestein EC, Sinatra RB. Surgery on the normal eye in Duane retraction syndrome. J Pediatr Ophthalmol Strabismus 1994; 31:162–169.
10 Greenberg MF, Pollard ZF. Poor results after recession of both medial rectus muscles in unilateral small-angle Duane’s syndrome, type I. J AAPOS 2003; 7:142–145.
11 Natan K, Traboulsi EI. Unilateral rectus muscle recession in the treatment of Duane syndrome. J AAPOS 2012; 16:145–149.
12 Sukhjia J, Kaur S, Singh U. Isolated lateral rectus recession with Y splitting versus anchoring of the lateral rectus muscle in patients with exotropic Duane syndrome. J AAPOS 2014; 18:147–150.
13 Velez FG, Velez G, Hendler K, Pineles SL. Isolated Y-splitting and recession of the lateral rectus muscle in patients with exo-duane syndrome. Strabismus 2012; 20:109–114.
14 Kubota N, Takahashi H, Hayashi T, Sakaue T, Maruo T. Outcome of surgery in 124 cases of Duane’s Retraction Syndrome (DRS) treated by intraoperatively graduated recession of the medial rectus for esotropic DRS, and of the lateral rectus for exotropic DRS. Binocul Vis Strabismus Q 2001; 16:15–22.
15 Kaban TJ, Smith K, Day C, Orton R, Kraft S, Cadera W. Single medial rectus recession in unilateral Duane syndrome type I. Am Orthoptic J 1995; 45:108–114.