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Contralateral lateral rectus muscle recession in a patient with unilateral exotropic Duane retraction syndrome type II: A case report

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ABSTRACT

A fixation preference for the affected eye is uncommon in patients with unilateral Duane retraction syndrome (DRS), and surgery on the fellow eye is rarely advocated. We are presenting a case report of a 9-year-old boy with unilateral DRS type II in the left eye who received lateral rectus muscle recession in his right amblyopic eye. The patient was orthophoric and his face turn was gone 6 months postoperatively. Surgery on the fellow amblyopic eye is a good choice for unilateral DRS where the affected eye dominates the fixation, and the satisfactory outcome suggests that alignment in the primary position can correct the face turn effectively despite the muscle duction deficit in the affected eye and further extend the binocular single visual field.

KEYWORDS

Duane retraction syndrome; strabismus surgery; fixation preference; case report



Introduction

Duane retraction syndrome (DRS) is a rare congenital eye movement disorder caused by hypoplasia or absence of the sixth cranial nerve and nuclei combined with dysinnervation of one or two branches of the oculomotor nerve.¹ It is characterized by horizontal ocular motility deficit, ocular deviation in the primary position, retraction of the globe, and narrowing of the palpebral fissure during attempted adduction, which are caused by the simultaneous contraction of the medial and lateral rectus muscles.^{2,3} Associated in many cases of DRS is an upshoot or downshoot: a “flipping” up or down movement which occurs when the eye moves above or below the horizontal plane in an adducted position. This abnormality is considered to be related to a sudden slippage of the tight lateral rectus muscle over the globe.⁴ Patients who showed esotropia or exotropia commonly adopt a face turn to maintain binocular single vision and make up for the duction deficit, depending on which duction is maximally affected.^{5,6} Huber categorized DRS into three types according to electrophysiology, among which type II (4–13%) is the least common

presentation and presents with poor eye adduction and exotropia.¹

3–25% of patients with DRS suffer from amblyopia, while strabismus and anisometropia were considered as strong reasons among studies.^{7–9} In a majority of cases, the unaffected eye is the dominant eye for those patients who exhibit fixation preferences with unilateral DRS (80–90% of the cases).^{7,10} Currently, only a few cases with fixation preference for the affected eye have been reported. But these cases were all DRS type I.^{7,11,12} In this case, it is DRS type II which is different from the previous reports.

In the treatment of unilateral DRS, surgery on the involved eye is recommended by most authors. For a patient who is exotropic in the primary position, an ipsilateral lateral rectus recession for deviations <20D and a bilateral lateral rectus recession for deviations >20D are widely adopted. In addition, Y-splitting recession of the ipsilateral rectus muscle is considered a reliable surgical option in some patients with significant overshoot.^{13,14} Surgery restricted on the fellow eye is rarely advocated.¹⁵ Here, we present a case report of

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The study was conducted in Zhongshan Ophthalmic Center, Sun Yat-sen University, Guangzhou, China.

Abbreviations

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a child patient with unilateral DRS who received lateral rectus muscle recession in his fellow amblyopic eye and obtained a satisfactory outcome.

Case presentation

A 9-year-old boy was presented to our hospital due to an outward deviation in his right eye for 3 years. He has no history of ocular trauma or ocular surgery, and his birth and development were not affected.

On examination, the patient showed moderate horizontal torticollis, with a right-sided face turn of 40° while reading the distance vision chart. The best-corrected visual acuity was 20/16 in his left eye and 20/32 in the right eye. Cycloplegic refraction manifested anisometropia, $+1.00\text{DS}/-0.50\text{DC}\times 1$ in the left eye and $+1.75\text{DS}/-3.50\text{DC}\times 5$ in the right eye. Anterior segment and retinal examinations were unremarkable. In the primary position of gaze, the patient presented with 15 prism diopters (PD) exotropia, L/R 5PD in primary position measured with prism alternative cover test, and an angle kappa of zero degree in both eyes were noted (Figure 1a). Globe retraction of grade 2 of the left eye in dextroversion was observed.¹⁶ Hertel exophthalmometer measurements were 12 mm OD and 12 mm OS in primary gaze, 12 mm OD and 9 mm OS in 30° dextroversion, and 12 mm OD and 12 mm OS in 30° levoversion. Besides, an upshoot of the left eye was noted as the globe adducts (grade 4) and moves above the horizontal plane (grade 3) in adduction, respectively, but no

downshoot was present.¹⁶ Adduction was mildly limited in the left eye, while the extraocular movement was full in the right eye (Figure 2a). Force duction test revealed no contracture in both eyes. Synoptophore test showed fusion field arranging from -7° to $+8^\circ$. In diplopia quantification using the cervical range of motion (CROM) method, binocular single vision appeared exclusively in 30° of left gaze and habitual reading gaze, and the score was 84 (Figure 3a).

He was diagnosed with exotropic DRS in the left eye. A 6-mm right lateral rectus recession was performed in his right eye. No extraocular muscle abnormalities were found. Postoperatively, the patient was orthotropic for distance and near in primary gaze, with resolved face turn in 6 months (Figure 1b). There was mild limitation of abduction in the right eye and adduction in the left eye, but no manifest strabismus in the horizontal gaze position except for the full right gaze, where the upshoot remained in the left eye. Globe retraction and upshoot in right gaze in the left eye remained unchanged (Figure 2b). The score of CROM diplopia examination was 6 as diplopia only appeared in 30° of right gaze (Figure 3b).

Discussion

Single horizontal rectus muscle recession of the affected eye in these cases was proposed by Duane in 1905 and has been widely used due to its simplicity, reliable correction of primary deviation as well as abnormal face turn for deviation

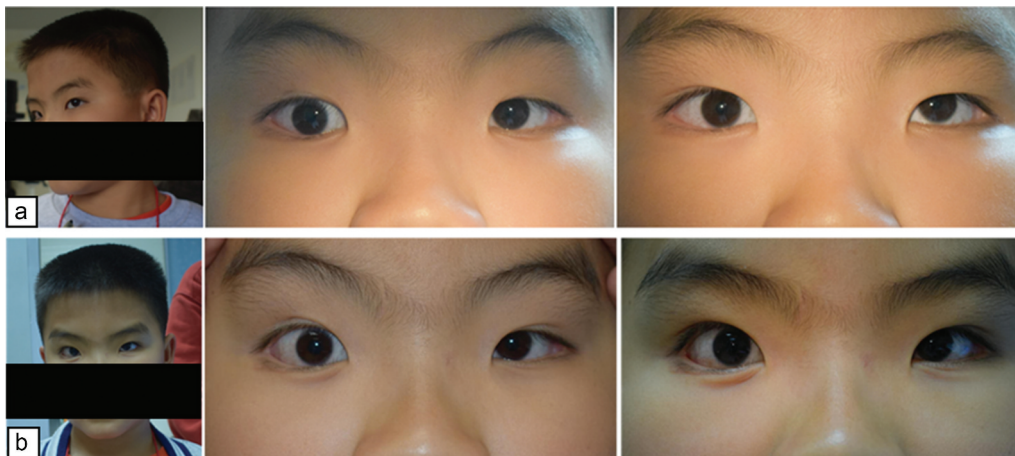


Figure 1. Head posture and eye position of the patient. Preoperative (a) and postoperative (b) head posture and eye position in the patient with DRS. The patient was orthotropic without abnormal head position after 6-mm lateral rectus recession.

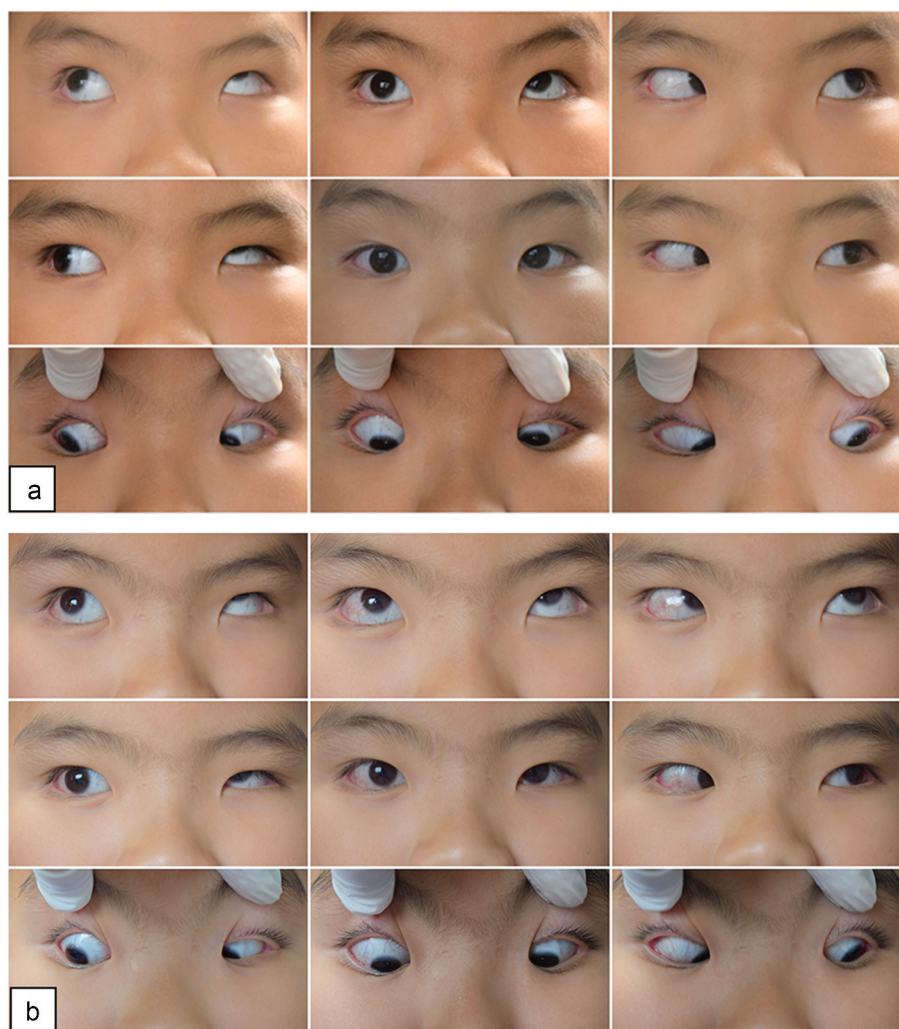


Figure 2. 9-gaze photographs of the patient. (a) 9-gaze preoperative photographs showed mild limitation of adduction with globe retraction, palpebral fissure narrowing, and upshoot on attempted adduction of the left eye. (b) 9-gaze postoperative photographs showed mild-to-moderate limitation of adduction in the right eye with persistent symptoms in the left eye.

less than 20PD.^{3,14} Moreover, recession of lateral rectus in Duane's eye can simultaneously diminish globe retraction, palpebral fissure narrowing, and up/down shoot in adduction.¹¹ However, recession of the ipsilateral lateral rectus muscle may worsen the abduction limitation and increase the esotropia in lateral gaze postoperatively, while the small recession in the fellow eye is adequate to realign the eye in primary position without this complication.¹⁵

Surgery on the fellow eye is rarely advocated. Several authors declared that weakening of the rectus muscles in the normal eye can balance the adduction deficit and thereby expand the binocular single visual (BSV) field.¹⁵ In addition, when the unaffected eye is the dominant eye, this procedure

can reduce the likelihood of any recontracture in the DRS eyes simultaneously, according to Sherrington's and Hering's laws.^{11,17} However, Duane's syndrome is a disorder in which Sherrington's and Hering's laws do not apply. Thus, this theory, in which "fixation duress" plays a significant role, remains doubtful. As far as we are aware, operation restricted in the fellow eye has not been reported and discussed before in the case that the affected eye is the dominant eye.

Amblyopia in the fellow eye could be due to long-term uncorrected anisometropia in this patient. Based on his examination and his parents' operation preference on the "bad" eye, we performed a 6-mm lateral rectus recession on the amblyopic right eye. The patient was orthophoric

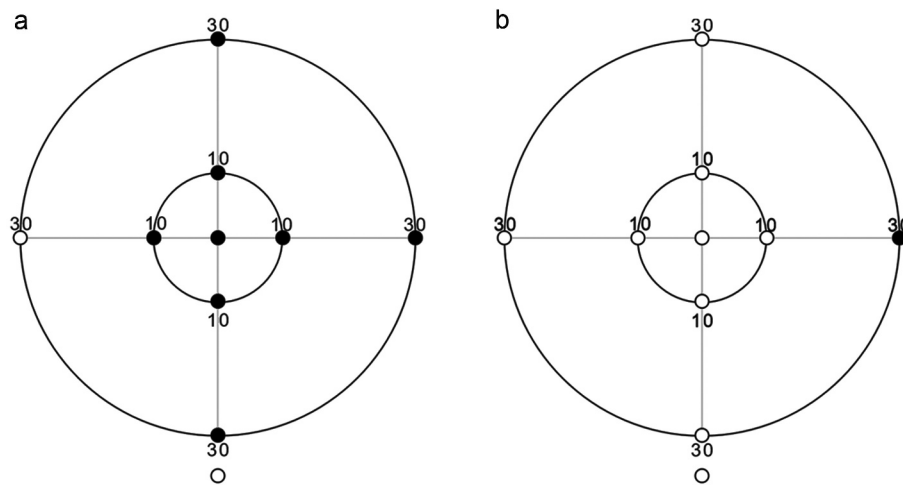


Figure 3. Results of cervical range of motion method. Single vision represented by white point and diplopia by black point. Point below 30° represents habitual reading gaze. (a) Preoperative cervical range of motion diplopia scored 84. (b) Postoperative cervical range of motion diplopia scored 6.

at distance and near, and his face turn was gone 6 months postoperatively. This suggests that alignment in the primary position can correct the face turn effectively despite the muscle duction deficit in the affected eye. Since the primary goal for surgery in DRS is the correction of primary position deviation and torticollis,¹² single contralateral rectus recession may be a proper choice for a patient with poor visual acuity in the fellow eye when the affected eye can reach the midline.

Since a precise measurement of BSV field using the Goldmann perimeter was unavailable due to the noncooperation of the child, we adopted the CROM method instead. This method has been recommended as a simple alternative to the Goldmann perimeter for quantifying diplopia.¹⁸ In this case, the CROM score dipped significantly from 84 preoperation to 6 postoperation, indicating an extended BSV field with balanced horizontal rotations. Besides, we noticed a disappearance of vertical deviation along with correction of horizontal deviation by recession of the lateral rectus muscle. Further studies are needed to explore the underlying mechanisms.

This case suggests that surgery restricted on the fellow eye is a good choice to obtain alignment in the primary position, correct the face turn, and further extend the BSV field for unilateral DRS where the affected eye dominates the fixation.

Ethics approval and consent to participate

This study followed the tenets set forth in the Declaration of Helsinki, and approval was obtained from the Ethics Committee of the Zhongshan Ophthalmic Center of Sun Yat-sen University.

Consent for publication

Written informed consent was obtained from the guardian for publication of this case report and any accompanying image consent.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

KM designed the study and was a major contributor in writing the manuscript. XY and KD analyzed and interpreted the patient data and substantively revised the manuscript. LC collected the data and checked and revised the manuscript and participated in the surgery as the first aider. XL designed the study, performed all the treatments, and agreed to be accountable for all aspects of the work. All authors read and approved the final manuscript.

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