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Review

Surgical treatment of Duane retraction syndrome

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Abstract

Purpose: Surgical treatment in Duane retraction syndrome (DRS) can be very challenging even for the strabismus specialists because of a wide spectrum of diversity in clinical manifestation. The purpose of this article is to review these different surgical treatments.

Methods: A comprehensive search was performed using PubMed database with the different keywords of "Duane retraction syndrome" and "surgery". Articles were selected from original English papers published since 2000. The full text of the selected articles was reviewed, and some articles were added based upon the references of the initial articles. We also provided selected case examples about some of these procedures. *Results*: 125 articles were found in the initial search of which 37 articles were mostly related to the topic of this review. The number finally increased to 59 articles after considering the relative references of the initial articles. Different surgical methods performed on horizontal and vertical rectus muscles (recession, resection, transposition, Y splitting, periosteal fixation and posterior fixation suture) are reviewed. Careful selection of the surgical technique is important to achieve optimal results.

Conclusion: With accurate diagnosis of patients with DRS and proper surgical management, several adverse situations associated with this syndrome (amblyopia, abnormal head posture, upshoot, downshoot, and muscle underaction) can be prevented.

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Keywords: Duane retraction syndrome; Surgery

Introduction

Duane retraction syndrome (DRS) is one of the congenital cranial dysinnervation disorders characterized by limitation of horizontal eye movements, globe retraction, and vertical movements in adduction.^{1,2} The incidence of DRS is 1%-4% of strabismus patients.³ This syndrome is caused by congenital hypoplasia or absence of sixth nerve and nuclei with aberrant innervations by branches from the oculomotor nerve resulting in co-contraction between medial and lateral rectus muscles on adduction.^{4–7} Using electrophysiology tests, Huber classified

Institution at which the study was conducted: Farabi Eye Hospital. Authors obtained consents from the patients for publishing the photos.

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DRS into three types, but because of variability in misinnervation and resultant clinical features, some clinicians have simplified Duane syndrome patients to either esotropic, exotropic, or orthotropic DRS. This simple classification is easier to use for surgical planning.^{8,9}

Various surgical techniques have been described that strabismus surgeons should be aware of the potential benefits and limitations of these procedures. In this article, we review different surgical treatments for DRS with selected case examples for some of them.

Methods

A PubMed search was performed in November 2016 using each of the following key words: "Duane retraction syndrome" and "surgery". Articles were selected from original English papers published since 2000. All article types including original articles, reviews, case reports, and book chapters about

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surgery in DRS were identified. The full text of the selected articles was reviewed, and some articles were added based upon the references of the initial articles. We also provided selected case examples about some of these procedures.

Results

After reviewing English literature using PubMed database, 125 articles were found in the initial search of which 37 articles were mostly related to the topic of this review. The number finally increased to 59 articles after considering the relative references of the initial articles. The results were divided into five main sections of esotropic DRS, exotropic DRS, globe retraction and overshoots, bilateral DRS, and simultaneous abduction. Different surgical methods are categorized and summarized in Table 1.

Surgical approaches

Esotropic Duane retraction syndrome

If strabismus is present, esotropia occurs more frequently than exotropia in patients with DRS. Esotropia is usually less than 30 prism diopters (PD) in primary position.¹⁰ It presents when the limitation in abduction is greater than the limitation in adduction and when the tonus of the lateral rectus muscle in primary position is less than that of the medial rectus muscle. An approach to the surgical treatment of esotropic DRS

Table 1

Summary of different surgical approaches for Duane retraction syndrome (DRS).

depends on the analysis of important anomalies observed in this group of patients such as primary position alignment, abnormal head posture, limitation of ductions, and severity of retraction and overshoots.¹¹ Patients with DRS can have high hyperopia and resulting accommodative esotropia, which is important to correct prior to surgery. Sometimes, spectacles alone can correct abnormal head posture in DRS.¹²

Case 1 (Fig. 1). The patient was a 4-year-old child with left eye esotropic DRS who presented severe left face turn, left eye esotropia, and limitation of abduction. His cycloplegic refraction was +7.00 in both eyes. After prescription of



Fig. 1. *Case 1*. Photograph (upper row) shows motility in a patient with left eye esotropic Duane retraction syndrome (DRS) and hyperopic refractive error. Photograph (lower row) shows head posture before and after prescription of glasses in the same patient.

Туре	Intervention
Esotropic DRS	 Horizontal rectus surgery: Ipsilateral MR recession Symmetric or asymmetric bilateral MR recession Ipsilateral MR recession and contralateral MR posterior fixation suture Ipsilateral MR recession and LR resection Transposition surgery: Full tendon or half tendon VRT with or without fixation suture SRT with and without MR recession
Exotropic DRS	 Ipsilateral LR recession Bilateral lateral rectus recession Periosteal fixation of the LR Full tendon or half tendon nasal VRT with or without fixation suture
Overshoots & Globe retraction	 Mechanical type: Large LR recession Periosteal fixation of the LR Recession of the ipsilateral LR and MR Posterior fixation suture of the ipsilateral LR alone or both the LR & MR Splitting of the LR into a Y configuration Innervational type: Recession of the appropriate vertical rectus muscle
Bilateral DRS	Horizontal muscle recessionVRT
Simultaneous abduction	Large LR recessionPeriosteal fixation of the LRMR resection

DRS: Duane retraction syndrome, MR: Medial rectus, LR: Lateral rectus, VRT: Vertical rectus transposition, SRT: Superior rectus transposition.

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