SURGICAL TREATMENT OF DUANE'S SYNDROME TYPE I BY RECESSION OF THE MEDIAL RECTUS OF THE AFFECTED EYE AND FADEN OPERATION OF THE CONTRALATERAL MEDIAL RECTUS

TRATAMIENTO QUIRÚRGICO DEL SÍNDROME DE DUANE, TIPO I MEDIANTE RETROINSERCIÓN DEL RECTO MEDIAL DEL LADO AFECTO Y FADEN DEL RECTO MEDIAL CONTRALATERAL

ABSTRACT

Purpose: Different surgical approaches have been described for the treatment of Duane’s syndrome. The purpose of our study is to report the results of patients undergoing recession of the medial rectus (MR) muscle of the affected eye and placement of contralateral MR faden posterior fixation sutures.

Methods: Retrospective study of 11 patients treated by a 4-7 mm recession of the MR of the affected eye and 13 mm faden posterior fixation suture of the contralateral MR in order to correct abnormal head position and esotropia in primary position.

Results: After surgery, there was no torticolis in 81.8% of patients, with less than 10º of torticolis in the remainder. In all patients, postoperative esotropia was less than 5 prismatic dioptres.

RESUMEN

Objetivo: Se han descrito numerosos tratamientos quirúrgicos para el síndrome de Duane (SD) tipo I. El objetivo de este trabajo es comunicar los resultados obtenidos en pacientes diagnosticados de SD tipo I sometidos a retroinserción del recto medial (RM) del lado afecto y operación de faden del RM contralateral.

Métodos: Estudio retrospectivo sobre once pacientes con SD tipo I a los que se les realiza una retroinserción del RM del lado afecto de entre 4 y 7 mm y faden a 13 mm del RM contralateral para resolver el torticolis y la endotropía presentes en posición primaria.

Resultados: Tras la cirugía, el torticolis desapareció en el 81,8% de los pacientes, siendo menor de 10º en el resto. En todos los pacientes la endotropia postquirúrgica fue menor de 5 dioptrías prismáticas.
Conclusion: This is a safe and effective procedure in Duane’s syndrome type I to treat moderate esotropia and torticollis (Arch Soc Esp Oftalmol 2008; 83: 113-116).

Key words: Duane Syndrome type I, Faden, torticollis, esotropia, surgical treatment.

INTRODUCTION

The Duane Syndrome (DS) is an alteration of ocular motility characterized by a retraction of the ocular globe and a narrowing of the palpebral slit upon adduction. It is associated to restricted abduction, adduction or both. A number of theories have been postulated to explain the etiology of DS, but most authors agree that it is a result of a congenital alteration of the 6th cranial pair with an aberrant enervation of the lateral rectus of the 3rd cranial pair (1). DS is the most frequent cause of congenital aberrant ocular enervation.

Hubber classified els in three types (2). In type 1 there is a limitation or total absence of abduction as a normal or slightly altered function of adduction. In type 2 adduction is limited or absent, with abduction being normal or slightly deficient. Type 3 is characterized by a limitation of abduction and adduction with retraction of the globe and narrowing of the palpebral slit during adduction attempts by the ocular globe. Type 1 corresponds to the characteristic and most frequent form of DS.

The majority of patients with this syndrome exhibit good binocular vision (3) by assuming a torticollis towards the action field of the deficient muscle. Surgery is indicated when the compensating torticollis is unacceptable or there is strabismus in the primary position.

The literature describes a variety of surgical options, including retro-insertion of the medial rectus (MR) of the affected side (4), transposition of the vertical straight muscles (5,6), retro-equatorial miopexia (faden) (7), retro-insertions of both medial recta (MRMR) and retro-insertion of the MR and the lateral rectus of the affected eye (8).

The purpose of this paper is to communicate the result of a series of patients diagnosed with DS type 1 submitted to MR retro-insertion of the affected eye associated to Faden operation of the healthy eye MR.

SUBJECTS, MATERIAL AND METHODS

A retrospective study of patients diagnosed with DS type 1 and surgically intervened since 1987 to 2005 in a University Hospital and in the private clinic of one of the authors. The patients diagnosed with DS type 2, 3 or bilateral were excluded, as well as the DS type 1 associated to oculomotor paresis, hyper-function of inferior obliques, Brown syndrome, myopic restrictions or who had strabismus surgery or botulin toxin injections.

We collected data related to the age and gender of patients at the time of surgery, personal history including the presence of congenital malformations and familial history of strabismus.

The reference ophthalmological explorations were taken two weeks prior to surgery, and the latest one included in the medical history. All patients had a postop follow-up of at least one year.

The visual acuity of patients was taken with the Snellen letters or Pigassou test depending on their age. Refraction was explored by means of retina shadow techniques (esquiascopia) and subjectively when the patient cooperated. Before and after the surgery the torticollis was assessed, dividing the patients in three groups depending on the degrees of deviation: 0 -15º (slight), 16 - 30º (moderate) and > 30º (severe). Primary gaze position strabismus was measured by means of the cover test, fixing the patient with the healthy eye and expressed in prismatic dioptres (PD).

The MR of the affected side was retro-inserted following the therapeutic approach described in Table 1, associated to retro-equatorial miopexia (faden) of the contralateral MR at 13 mm of the original MR insertion.

The surgery was performed by the same surgeon and under general anesthesia with double anchoring of the rectus muscles in the case of retro-insertion. In this technique, after fixing the suture to the mus-
the suture tunnelizes the sclera at the desired position for the new insertion (first anchor), placing the muscle at that point and fixing in the original position (second anchor). When the patient cooperated, the surgery was carried out with topical anesthesia and intra-op adjustment of sutures.

The results of the post-surgery torticollis were classified in four groups: very good (no torticollis), good (1 - 15º), flawed (16 - 30º) and deficient (>30º).

The post-surgery residual strabismus was classified as successful if below 5 DP, acceptable between 5 and 10 DP, flawed from 11 to 15 DP and deficient if over 15 DP.

The statistical analysis was performed utilizing the SPSS 12.0 for Windows (Chicago, Illinois).

### RESULTS

The series comprised 11 patients with a mean age of 4 (range 1-44). The proportion of males and females was of 4:7. All the patients had a visual acuity of at least 0.5 or, when they did not cooperate for obtaining visual acuity, a central and maintained fixation pattern in each eye. The mean follow-up time was of 18.23 months with a SD (standard deviation) of 20.4 months. The proportion of affected right and left eyes was of 3:8.

27.3% of patients exhibited a congenital anomaly such as ptosis or congenital obstruction of the lachrymal pathway. 18.2% of cases exhibited family strabismus antecedents, mainly severe hypermetropy and amblyopia with congenital endotrophy.

Eight patients (72.7%) exhibited pre-surgery torticollis classified as moderate, with 18.2% (two patients) with severe torticollis and one patient (9.1%) had slight torticollis (fig. 1).

The pre-surgery mean deviation was of 23.8 DP of esotropy SD 10.32. The mean MR retro-insertion value was of 5.4 mm SD 1.04.

In what concerns the post-surgery results, two of the patients (18.2%) exhibited slight torticollis and the rest did not exhibit residual torticollis (81.8%).

The mean post-surgery deviation was of 2.61 DP (SD 1.31) and the mean deviation improvement was of 22.19 DP (SD 9).

None of the patients exhibited severe surgical complications.

### DISCUSSION

After a comprehensive search of the literature we could not find studies in which DS type 1 was treated by recession of the MR and retro-equatorial mioptexia in the healthy eye. Even so, we did find some proposals for surgery on the contralateral eye. Saunders et al (7) defended the idea of operating on the healthy eye on the grounds that large retro-insertion and posterior myoscleropexia in the healthy eye cause similar adduction limitations in the affected eye, allowing for a larger visual field in the absence of diplopia. Jampolsky recommends asymmetric retro-insertion of the MR of both eyes in patients with severe torticollis when the retro-insertion of a single MR would not be sufficient. On the other hand, Greenberg et al (9) concluded that in patients with DS type 1 and small angle endotrophy, healthy eye surgery could reduce the positive effects of the MR retro-insertion in the affected side vis-à-vis torticollis and an increased risk of consecutive exotrophy risk. Other papers indicate that the MR retro-insertion of the affected eye yields the same abduction improvement but without less limitation of adduction than when associated to surgical intervention of the healthy eye (8,10).

On the basis of the above references, we propose a Faden-type retro-equatorial mioptexia of the healthy eye MR associated to MR recession of the affected eye in esotrophy and moderate torticollis cases. Thus, we aim to avoid the possibility of consecutive exotrophy and achieve the least degree of

<table>
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<tr>
<th>Deviation Angle</th>
<th>Recommended procedure</th>
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<tr>
<td>20-30 DP</td>
<td>MR reversed 4-5mm</td>
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<tr>
<td>30-40 DP</td>
<td>MR reversed 5-6mm</td>
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<td>40-50 DP</td>
<td>MR reversed 6-7mm</td>
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post-surgery torticollis. On the other hand, said ipsilateral MR retro-insertion, combined with the Faden-type contralateral intervention gives rise to a limitation of the compensatory adduction of the healthy eye, which may improve the abduction in the affected one.

We consider there are clinical entities such as DS type 1 which require weakening effects in a specific position of eye gaze, without altering the balance between the agonist and antagonist muscle in the other positions. The aim of the proposed retro-equatorial myoscleroexia is to produce a slight weakening effect of the paretic eye joint muscle in order to diminish or eliminate the strabismus angle in the paretic field of gaze (11).

In our study we found a larger proportion of females and left eyes in comparison to other publications (10). DS is frequently associated to congenital malformations (12,1). It is believed this is due to an alteration of normal embryogenesis between the 4th and 10th week of gestation. Marshman et al (12) found that over half of his DS patients had associated congenital anomalies and 46% had a family history of said anomalies. In our series we found a lower percentage, probably due to the absence of this type of data in clinical records, a consequence of the retrospective nature of the study.

With the proposed technique and according to the classification of results described above we have achieved «success» in correcting endotrophy in all cases and «very good» results in 81.8% of torticollis and «good» results in 18.2%. Accordingly, said technique seems to be an option to be considered with DS patients having torticollis and strabismus. Unilateral Duane type 1 syndrome surgery required a personalized approach based on two main parameters: horizontal torticollis and endotrophy in the primary gaze position. The technique proposed in this study is focused on moderate endotrophies because it adequately resolves both clinical symptoms and, in addition, it seems to avoid consecutive exotrophy.

REFERENCES