Congenital Absence of Lateral Rectus Muscle

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Congenital absence of one or more extraocular muscles is a well-recognized, but rare, occurrence. A recent report by Wong and Jampolsky\(^1\) states that there have been six reported cases of a single missing horizontal muscle. Duke-Elder\(^2\) mentions five cases of absence of the lateral rectus muscle, however, three of these reports are in the German literature (two in the 19th century). Investigation of the American literature\(^3\) does not reveal any surgically documented cases of absence of a single lateral rectus muscle.

The purpose of this paper is to report a case of congenital absence of a lateral rectus muscle in association with Axenfeld’s anomaly. Surgical repair of the congenital esotropia is described.

CASE REPORT

A four-year-old white female was initially seen at the University of Michigan Medical Center Ophthalmology Clinic at the age of seven months. There was a history of a marked left esotropia since birth with no evident abduction OS. Previous patching of OD had not elicited any abduction OS. Examination at that time revealed a left esotropia of approximately 60 PD and a probable Axenfeld’s anomaly OU. An examination under anesthesia was performed. Forced ductions revealed a resistance to abduction OS, grade 4 angles, multiple prominent iris processes which extended to Schwalbe’s line OU, and intraocular pressures of 21 OD and 28 OS. Fundus examination revealed pale flat discs and retinal pigment mottling OS.

At the age of 10 months, repeat examination under anesthesia was done and Schiotz tonometry revealed pressures of 18 OD and 30 OS. A goniotomy of the left eye was performed. After initial follow-up examination, the patient was then followed elsewhere for two and one half years.

The patient returned to the University of Michigan Medical Center in February of 1974, and underwent another examination under anesthesia. Corneal diameters and Schiotz tonometry were normal OU and there were no tears in Descemet’s membrane. The discs were not felt to show glaucomatous damage, although they did appear mildly atrophic OU. There was pigmentary degeneration of the left macula. Diagnoses at this time were: Axenfeld’s anomaly OU, mild optic atrophy OU, macular degeneration OS, and marked left esotropia with contracture of the left medial rectus secondary to a congenital left lateral rectus paralysis. Measurements in the orthoptics clinic revealed a 60 PD left esotropia (Fig. 1). The patient was unable to abduct OS to the midline.

It was decided that strabismus surgery should be performed for cosmetic reasons. The decision between a recess/resect operation or a muscle union was postponed until the time of surgery, when the muscles could be directly examined. The medial rectus muscle was found to be markedly fibrotic; it was recessed 7 mm. Numerous attempts to hook the lateral rectus muscle were unsuccessful. Complete exploration of the lateral half of the globe revealed absence of the left lateral rectus. The conjunctiva was then closed without further surgery. Postoperatively, the patient continued to have a 60 PD left esotropia.

Six months after the first operation, the patient returned for a second procedure. At the time of operation, a large limbal peritomy was performed, exposing the superior, lateral and inferior quadrants. Again, complete absence of the lateral rectus muscle was noted (Fig. 2). The superior rectus, inferior rectus, and inferior oblique muscles were identified and isolated; the course of the inferior oblique was carefully examined to insure that this was not a misplaced lateral rectus muscle. After tagging the vertical rectus muscles, a small spot of cautery was
Fig. 1. Preoperative photograph showing 60 prism diopters left esotropia in primary position.

Fig. 3. New position of superior and inferior rectus muscles.

Fig. 2. Complete absence of the lateral rectus muscle.

Fig. 4. Postoperative, primary position.

placed on the lateral aspect of the sclera midway between the insertions of the superior and inferior rectus muscles, 6 mm posterior to the limbus. The inferior and superior rectus muscles were then detached from the globe and reattached 1 mm inferior and superior to the previously marked spot, respectively (Fig. 3). The muscles were reattached so that the insertions were parallel to the limbus.

On follow-up examination, one month postoperatively, there was a left esotropia of 8 prism diopters in the primary position (Fig. 4). Ductions OS were full (Figs. 5-8). Visual acuity OS was 10/200, with poor fixation. The patient was started on full-time occlusion of the right eye. After three months of patching the vision improved to only 20/200 and occlusion was discontinued. One year after surgery the vision OS remains at 20/200 with unsteady fixation. Muscle measurements, versions, and ductions were unchanged.

DISCUSSION
The first successful treatment of paralysis of
the lateral rectus muscle is generally attributed to Hummelsheim; he suggested splitting the superior and inferior rectus muscles and transferring the lateral halves to a point near the insertion of the lateral rectus. O'Connor modified the basic Hummelsheim procedure by transplanting the nasal halves of the muscles laterally rather than the temporal halves. A further modification was introduced by Berens and Girard, who transplanted the lateral halves of the superior and inferior recti to the insertion of the lateral rectus, and moved the nasal halves of the vertical recti to the position previously occupied by the lateral halves. For cases in which the involved eye could not be abducted to the midline preoperatively, Krewson carried these modifications even further by attaching the nasal portion of the split vertical rectus muscles to a point just lateral to the temporal edge of the original insertions.

Each of the above modifications serve to produce more lateral displacement of the vertical muscles, thereby giving them power of abduction. It was once thought that the displaced vertical recti were incapable of acting as true abductors, but produced outward movement of the eye through the formation of cicatricial adhesions. However, this was neatly disproved by Hildreth in an experiment on a patient undergoing a recess/resect operation of the horizontal rectus muscles under local anesthesia; while the horizontal rectus muscles were detached, Hildreth passed a loop of suture around the superior and inferior rectus muscles and by tightening it, caused the muscles to be displaced laterally; the patient was then able to voluntarily abduct the eye. The amount of abduction that was possible was directly related to the amount of lateral displacement of the vertical rectus muscles.
In addition to its use in the treatment of paralysis of the lateral rectus muscle, the Hummelsheim procedure and its variations have been used for the repair of cases of surgical, traumatic, and congenital absence of the medial rectus muscle.\textsuperscript{12-15}

A slightly different approach to the problem of lateral rectus muscle paralysis was taken by O'Conner in 1935, when he reported the lateral transplantation of the entire tendon of the superior and inferior rectus muscles in six cases of abducens paralysis. The results in these cases were variable and O'Connor\textsuperscript{17} abandoned the procedure. Schillinger\textsuperscript{18} reintroduced this procedure in 1959, with a report of two cases in which good abduction and adduction were obtained, without any vertical imbalance. Uribe\textsuperscript{17} reported a large series of total tendon transplantations with good results in 1968. Knapp\textsuperscript{19} applied this technique to double-elevator paralysis and in 1969, reported a series in which he averaged 38 prism diopters of correction of hypotropia. In his procedure, Knapp moves the entire tendon of the medial and lateral rectus to the corners of the insertion of the superior rectus, yet most cases still exhibited grossly normal abduction and adduction.

A third approach to the problem of lateral rectus muscle paralysis was taken by Jensen.\textsuperscript{20} In 1969, he reported a series of 18 cases in which he performed a rectus muscle union procedure. In this procedure, the superior, lateral, and inferior rectus muscles are split lengthwise from the insertions to a point beyond the equator, without disinserting the muscles. The lateral half of the superior rectus is then united with the superior half of the lateral rectus at the level of the equator of the globe, by using a suture loop to draw the muscles together. The inferior half of the lateral rectus is united with the lateral half of the inferior rectus in a similar fashion. Jensen felt that this procedure produced superior results in terms of restoring function to the field of the action of the paralyzed muscle, with less chance of producing a vertical imbalance. In addition, since the muscles are not detached from the globe, there is minimal interference with the blood supply to the anterior segment. Others have also reported good results utilizing the muscle union procedure.\textsuperscript{20-21}

In the case which is the subject of this report, it was apparent that in order for there to be any chance of an acceptable cosmetic result, the potential abducting power of the vertical rectus muscles would have to be utilized as fully as possible, since the medial rectus had already been recessed with no change in the large esotropia. A modification of Knapp's procedure seemed to offer the best chance of success. Our procedure is similar to Knapp's in that the muscles were reattached parallel to the limbus rather than perpendicular, producing more lateral displacement of the muscles and therefore more power of abduction. The results were gratifying.

It would seem that the occurrence of an Axenfeld's anomaly in this case is probably a coincidence. Axenfeld's has been reported in association with cornea plana, corectopia, aniridia, and other changes of the anterior chamber,\textsuperscript{22} but not in association with congenital absence of extraocular muscles. Although both of these defects are of mesodermal origin, they take place at very different times during the development of the eye. The paraxial mesoderm which will form the extraocular muscles shows signs of cleavage at the 9 mm stage of the embryo, and the muscles are quite separated by 20 mm; differentiation of anterior chamber structures occurs much later in fetal life.\textsuperscript{23}

**SUMMARY**

A four-year-old female presented with a 60 PD left esotropia and Axenfeld's anomaly OU. Cosmetic strabismus surgery was undertaken, at which time it was discovered that the left lateral rectus was absent. The medial rectus was recessed, without change in the esotropia. At a second procedure six months later, the superior and inferior rectus muscles were transposed to the area of the missing lateral rectus. Follow-up examination revealed an 8 PD left esotropia with full ductions.

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**REFERENCES**

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