Lateral rectus sag and recurrent esotropia in children

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PURPOSE To describe the clinical and intraoperative findings of an anatomic abnormality in children that resembles sagging eye syndrome documented in older adults and that led to recurrent esotropia after surgery.

METHODS We reviewed records of 4 patients with substantial recurrent esotropia after bilateral medial rectus recession who required subsequent surgery combining lateral rectus resection with correction of the anatomic abnormality affecting the lateral rectus path. Binocular alignment was sequentially analyzed.

RESULTS Three young patients (2-3 years of age) presented with acquired esotropia but minimal cycloplegic refractive error. The fourth patient (14 years of age) initially had moderate hyperopia and partially accommodative esotropia, but subsequently developed marked bilateral overelevation in adduction. In all patients, esotropia recurred within 5^{Δ} of preoperative deviation after bilateral medial rectus recession. Surgical exposure demonstrated that bilateral lateral rectus paths were inferiorly displaced more than one-half tendon width from their normal paths near the globe's equator, despite normal scleral insertions. Equatorial myopexy and lateral rectus resection resulted in stable esotropia correction.

CONCLUSIONS Lateral rectus sag in children creates a type of acquired esotropia and overelevation in adduction poorly responsive to standard surgery but correctable with lateral rectus resection and equatorial myopexy that normalizes the lateral rectus path through permanent scleral fixation. (J AAPOS 2019;23:81.e1-5)

Surgery to correct esotropia in children must sometimes be repeated. Many studies have examined clinical parameters that may predispose to esotropia recurrence after surgery, including poor sensory status,[1-3] high ratio of accommodative convergence to accommodation,[4] undercorrected hyperopia,[5] or incomitance in oblique gaze directions,[4] sometimes with contradictory results.[2-4] Other studies have examined the efficacy of various surgical approaches to reoperation, such as medial rectus re-recession,[6] lateral rectus resection,[7,8] or medial rectus posterior fixation,[9,10] without considering potential individual sensory or anatomic abnormalities.

Orbital anatomy is important in both the etiology and the response to strabismus surgery.[11-14] In particular, the lateral rectus muscle, which combines a long course from orbital apex to insertion with relatively tenuous connective tissue pulley support,[15] appears most susceptible to displacement or an unstable path.[16,17] In patients with esotropia and high myopia (heavy eye syndrome), for example, magnetic resonance imaging (MRI) demonstrates abnormal inferior displacement of the lateral rectus pulley and nasal displacement of the superior rectus pulley to cause the strabismus.[18-20] Lateral rectus sag also causes distance esotropia in older adults with sagging eye syndrome.[21,22] In such cases, correction of the lateral rectus path by equatorial myopexy can restore normal orthotropia.[23]

Within our clinical practice, we recognized a small group of younger strabismic children who share two common characteristics. First, each patient experienced almost complete recurrence of preoperative esotropia after large bilateral medial rectus recessions. Second, each patient shared a common anatomic abnormality of lateral rectus pulley position. The purpose of this study is to describe this anatomic abnormality and to investigate clinical factors that might allow preoperative identification of esotropic patients likely to have a poor response to standard strabismus surgery.

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Subjects and Methods

After obtaining approval from the Institutional Review Board at Long Beach Memorial Medical Center, a retrospective review of medical records was conducted to identify patients with nearly complete (within 5^{Δ}) recurrence of preoperative esotropia following bilateral medial rectus recession. Patients with developmental delay, structural eye abnormalities, or uncorrected amblyopia were excluded, as were patients who underwent initial strabismus surgery concurrently involving other rectus or oblique muscles.

Results

Four patients met inclusion criteria. Each patient had esotropia onset after 5 months of age. Clinical parameters were recorded before and after each strabismus surgery: corrected visual acuity, eye alignment at distance and near, versions, stereoacuity when possible, cycloplegic refraction, slit-lamp examination, and ophthalmoloscopy. In addition, operative reports were reviewed for reported findings.

In each case, initial surgical doses for bilateral medial rectus recession were targeted to the distance esotropia using published tables,[24] except in 1 case augmented by medial rectus pulley posterior fixation for excess near esotropia.[10] During reoperation, the surgical dosages for lateral rectus resections were modified from published tables based on intraoperative findings, sometimes resulting in a reduction of resection.

Case 1

This 2-year-old boy presented with an intermittent left esotropia of 15^{Δ} at near only, normal versions, and spherical equivalent (SE) refraction of −0.25 D in each eye. Control of esotropia initially improved with patching, but the esotropia at near progressed, and by the time the patient was 3 years old, it could not be controlled with bifocal spectacles. Repeat cycloplegic refraction confirmed absence of hyperopia (−0.50 D [SE] bilaterally), good visual acuity in each eye, normal ocular versions, and orthophoric distance alignment but esotropia at near of 30^{Δ}.

The child underwent uncomplicated bilateral 2 mm medial rectus recessions with pulley posterior fixation.[10] One week after surgery, the patient remained orthophoric at distance and had an intermittent esotropia of 8^{Δ} at near. Two weeks later, he presented with complete recurrence of near esotropia, measuring 30^{Δ}, and over the next month newly developed 15^{Δ}-20^{Δ} distance esotropia. He also exhibited mild left medial rectus overaction and left lateral rectus underaction.

Three months after initial surgery, the patient underwent a second strabismus surgery planned as bilateral lateral rectus resections for esotropia 20^{Δ} at distance and 30^{Δ} at near. Intraoperatively, both lateral rectus insertions were in normal positions, but retroequatorial muscle paths were markedly displaced inferiorly in each eye, so that each

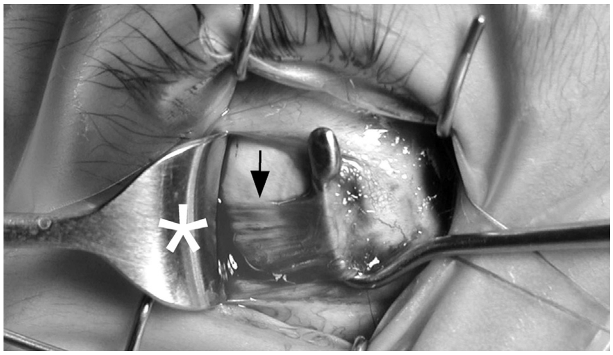


FIG 1. Case 1. The right lateral rectus muscle has been isolated through a limbal incision. The insertion, on a Jameson muscle hook, is in the correct temporal anatomic location, but the posterior muscle belly in the equatorial region is inferiorly displaced by one-half tendon width (black arrow). The posterior muscle belly should be centered onto the center of the retractor blade, marked with the white asterisk, to be in the correct temporal location.

lateral rectus muscle had an inferotemporal rather than normal temporal course (Figure 1). Surgery was modified to resect each lateral rectus 4.5 mm with equatorial myopexy by permanent sutures placed 8 mm posterior to the superior margins of the muscle insertions through the equatorial sclera and adjacent superior third of the lateral rectus belly to elevate posterior lateral rectus paths into vertical alignment with their insertions (Figure 2).[23] More than 3 years after reoperation, the patient has remained orthophoric at both distance and near, with normal versions.

Case 2

This boy presented at age 2.5 years with an intermittent left esotropia of 2^{Δ}-4^{Δ} at distance and 10^{Δ} at near. He demonstrated fusion by Worth 4-dot test. Versions were normal and hyperopia of $< +0.50$ D (SE) bilaterally. Over the next year, esotropia slowly increased to 18^{Δ} at distance and near. Also, for the first time more than 1 year after initial presentation, he demonstrated overelevation of both eyes in adduction without fundus excyclotorsion. Repeat cycloplegic refraction confirmed low hyperopia with +1.00 D (SE) bilaterally.

He underwent uncomplicated bilateral 4 mm medial rectus recessions. One week postoperatively, he was orthophoric at near and had an esotropia of 2^{Δ} at distance. Four weeks later, he had 30^{Δ} left esotropia at distance, exceeding preoperative measurement, and 16^{Δ} at near, with mild LR underaction bilaterally. The patient began atropine penalization of his right eye. A brain MRI scan was normal.

Six months after initial surgery, both distance and near esotropia had increased to 35^{Δ}. Mild bilateral lateral rectus underaction persisted. At his second surgery, both lateral rectus insertions were found to be in the normal positions relative to the limbus. The retroequatorial lateral rectus paths, however, were markedly displaced inferiorly in

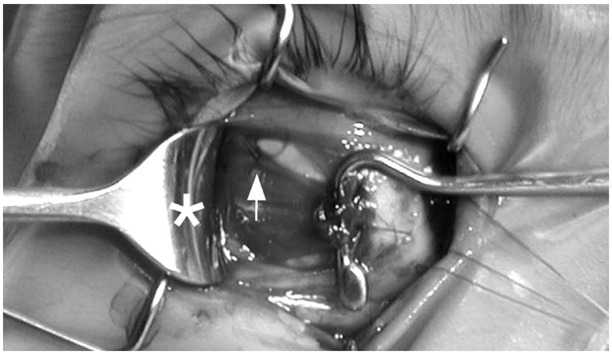


FIG 2. Case 1. A single 6-0 polyester monofilament suture is placed partial thickness into the equatorial sclera and adjacent superior lateral rectus belly (white arrow) to shift the posterior belly into vertical alignment with its insertion. Now the muscle belly is in the correct temporal location and is centered relative to the center of the retractor (white asterisk).

each eye, giving each lateral rectus an inferotemporal trajectory. The surgical plan for 6.5 mm bilateral lateral rectus resections was modified to include equatorial myopexies to elevate the lateral rectus paths into vertical alignment with their insertions.[23] More than 8 months after his second surgery, the patient has remained orthophoric at both distance and near with normal versions and recovery of fusion on the Worth 4-dot test. Overelevation in adduction resolved.

Case 3

This boy presented at age 18 months to another pediatric ophthalmologist with a complaint of poor vision and was noted to have high myopia of −12.50 D (SE) in each eye but normal binocular alignment and motility. Our subsequent evaluation confirmed these findings at age 2 years. Over the next year, myopia progressed by slightly more than 1 D bilaterally. At age 3 years, the patient was noted to have a new-onset left esotropia of 25^{Δ} at distance and 30^{Δ} at near. Occlusion therapy was initiated for mild left amblyopia. Over the next 3 months, the amblyopia resolved, but the esotropia increased to 40^{Δ} (corrected for prismatic artifact in the highly myopic spectacles), with mild left medial rectus overaction and left lateral rectus underaction.

The patient underwent uncomplicated bilateral 6 mm medial rectus recessions. One day following surgery, the patient had a residual esotropia of 15^{Δ} at distance and 30^{Δ} at near. Over the next 6 weeks, the esotropia increased to 30^{Δ} at both distance and near. There was mild-to-moderate lateral rectus underaction bilaterally.

Immediately prior to a second surgery, the patient underwent 2-hour prism adaptation, increasing the esotropia to 45^{Δ} at both distance and near. Two months after his initial surgery, he underwent planned bilateral lateral rectus resections intended to correct 45^{Δ} esotropia. Intraoperatively, both lateral rectus insertions were isolated in the normal position in the temporal quadrant posterior to the limbus, but the retroequatorial lateral rectus paths had slipped inferiorly to lie adjacent to the inferior rectus muscles bilaterally. In addition, the posterior lateral rectus bellies were noted to be very unstable, sliding between the normal temporal position and the inferiorly displaced position with minimal transverse force. Because of the severely abnormal lateral rectus position, the planned resection was abandoned and replaced by bilateral lateral rectus equatorial myopexy.[23]

On the first postoperative day, the boy was orthophoric at distance, with an esotropia at near of 8^{Δ}. Six weeks after surgery, he was orthophoric at distance and near and exhibited normal ocular versions. Over the next 6 months, however, esotropia recurred to 25^{Δ} at distance and 30^{Δ} at near, with mild lateral rectus underaction bilaterally.

Eight months after his second operation, the patient underwent a third strabismus surgery, consisting of bilateral 5.5 mm lateral rectus resections with revision of the lateral rectus equatorial myopexy fixation sutures to maintain normal lateral rectus paths. During surgical exploration, the posterior lateral rectus bellies were observed in correct temporal anatomic position, stabilized by the original posterior fixation sutures. More than 6 months after his third surgery, the patient was orthophoric at near with a residual esotropia of 8^{Δ} at distance and normal ocular versions.

Case 4

This girl presented at age 6 months to another pediatric ophthalmologist with new-onset esotropia. She had 45^{Δ} alternating esotropia at both distance and near, with mild bilateral lateral rectus underaction and moderate hyperopic spherical equivalent of +3.75 in the right eye and +3.50 in the left eye. With spectacles correcting full hyperopia, there was 35^{Δ} alternating esotropia at both distance and near.

The patient underwent uncomplicated bilateral 5 mm medial rectus recession. Two days after surgery she was orthophoric at distance and near. She remained in good alignment, with esotropia of $<6^{\Delta}$ at distance and near, for the following year, until she first exhibited marked overelevation in adduction in both eyes. She also developed an esotropia of 14^{Δ} at near only. Bifocal spectacles were prescribed that controlled the near esotropia, and the inferior oblique muscles were recessed 10 mm bilaterally.

For more than 12 years after the second surgery, the patient maintained good binocular alignment at both distance and near with bifocals. She developed fusion responses on the Worth 4-dot and the Titmus tests. Despite normal versions, during the 2 years following replacement of bifocal spectacles by contact lenses, she developed progressive esotropia ultimately measuring 20^{Δ} at distance and 25^{Δ} at near that no longer responded to bifocals.

The patient underwent a third strabismus surgery, consisting of bilateral 5 mm lateral rectus resections with one-half tendon width infraplacement. Two days postoperatively, there was 4^{Δ} esotropia at distance and 6^{Δ} at near with mild left MR underaction. Four weeks later, there was 16^{Δ} esotropia at distance and 20^{Δ} at near, with normal versions. Three months later, there was 25^{Δ} esotropia at distance and 30^{Δ} at near, exceeding the angle before the third surgery and associated with right lateral rectus underaction. Cycloplegic refraction was unchanged. Over the next 6 months, the esotropia increased to 40^{Δ} at both distance and near.

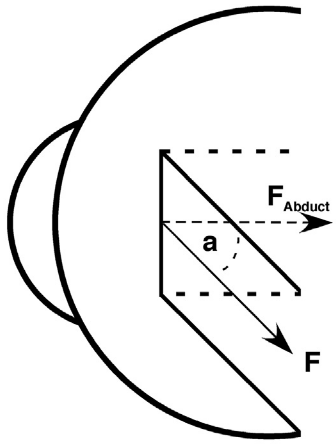
The patient underwent a fourth strabismus surgery, consisting of bilateral medial rectus marginal myotomies. On postoperative day 1, the patient was orthophoric at distance and near, with normal versions. For the next 3 years, she maintained good eye alignment at both distance and near, with a maximum esotropia of 8^{Δ}. During this period, our examination confirmed good ocular alignment and stable cycloplegic refraction but demonstrated mild-to-moderate bilateral lateral rectus underaction and marked right medial rectus overaction. Subsequently, she presented with 25^{Δ} esotropia at both distance and near that was unresponsive to an increase in hyperopic spectacle correction, with marked bilateral medial rectus overaction.

A fifth strabismus surgery was performed for recurrent esotropia. Extensive scar tissue was excised from both medial rectus insertional sites. The left lateral rectus was isolated and its posterior path was found displaced a full tendon width inferiorly. In addition, the left inferior oblique muscle was found to be adherent to the inferior border of the posterior lateral rectus belly and was released; the lateral rectus was resected and advanced 2.0 mm. The posterior lateral rectus belly was then fixated with permanent equatorial scleral sutures 10 mm posterior to the superior margin of the original lateral rectus insertion and through the adjacent superior third of the lateral rectus belly to shift the posterior lateral rectus muscle into the normal vertical location. The posterior muscle path of the right lateral rectus was found displaced one-half to three-quarters tendon width inferiorly. The right lateral rectus was resected and advanced 2.0 mm and its posterior belly similarly fixated with scleral equatorial sutures into the normal vertical location.

One day after surgery, the patient was orthophoric at distance and near with a minimal dissociated vertical deviation (DVD). More than 7 years after surgery, there was esotropia of 6^{Δ}-8^{Δ} at distance and near, minimal DVD, and bilateral medial rectus overaction without lateral rectus underaction.

Discussion

These 4 children experienced recurrence of esotropia despite seemingly appropriate surgical management. Recognition of the abnormal anatomy of the lateral rectus path allowed effective treatment of the cause of the recurrent esotropia. For the first 3 patients, who lacked significant hyperopia, the primary esotropia was likely caused by abnormal inferior sag of the lateral rectus pulleys: there is no other plausible explanation for the near complete recurrence of esotropia mere weeks after medial rectus recession. This type of abnormal lateral rectus anatomy has been well described in adults with high myopia[18] and sagging eye syndrome[21] and may also underlie the reported poor response of infantile esotropia with myopia to medial rectus recession (Demer JL, et al. J AAPOS 2013;17: Abstract 121).[25]



$$F_{Abduct} = \cos(a) \times F = \cos(51°) \times F = 0.62 \times F$$

FIG 3. Using force vector analysis, the abduction force (F_{Abduct}) equals force of muscle contraction (F) multiplied by the cosine of the angle of displacement (a). For a full tendon width lateral rectus inferior displacement at the globe equator, angle (a) is approximately 51°, resulting in only 62% of muscle force directed toward abduction.

In the fourth patient, the inferior displacement of the lateral rectus pulleys may have been iatrogenic. She initially presented with the more common clinical scenario of partially accommodative esotropia with moderate hyperopia and maintained good eye alignment for years after standard medial rectus recessions followed by inferior oblique recessions. After late decompensation of the esotropia, she underwent infraplacement of the lateral rectus insertions. During our reoperation, the left inferior oblique was found to be fused to the inferior border of the left lateral rectus muscle belly, and both lateral rectus posterior bellies were inferiorly displaced. The inferior transposition of the lateral rectus insertion, possibly in combination with scarring from the prior inferior oblique recession, may have created the abnormal orbital anatomy the led to an esotropia recurrence.

Simple quantitative vector analysis demonstrates the effect of inferior displacement of the lateral rectus pulley on its contractile abducting force (Figure 3). Full-tendon-width inferior displacement of the lateral rectus path near the globe equator would result in an approximately 40% loss in the abducting component of lateral rectus force acting on a normal insertion. The residual 60% abducting force may suffice to maintain a near-normal abduction range when the medial rectus muscle relaxes, but it may not suffice to balance primary gaze alignment. Surgery to restore normal lateral rectus pulley position increases the abducting component of the lateral rectus force even without resection, as shown by case 3, where the primary gaze alignment markedly improved after normalization of lateral rectus path. It is often necessary to resect the lateral rectus, however, to augment its force, particularly if distance esotropia is $\geq 20^{\Delta}$.

There is no data on how many patients with esotropia might have similar anatomic abnormalities of the lateral rectus pulleys; strabismic patients do not typically undergo preoperative orbital imaging. Our results suggest two preoperative clinical findings that should raise suspicion of abnormal orbital anatomy that could affect surgical results. The first is the absence of hyperopia $> +2.00$ D in acquired esotropia. In addition to neurological lesions, abnormal orbital anatomy should be included in the differential diagnosis when no clear etiology for esotropia is evident, and lateral rectus pulley infraplacement should be a strong consideration in patients with high myopia. The second clinical finding is overelevation in adduction, linked by previous studies to inferior lateral rectus pulley displacement.[16,26] Two of our patients developed overelevation in adduction. Marked overelevation in adduction should at least arouse suspicion of an abnormal lateral rectus pulley, especially in the absence of fundus torsion. Of note, none of our 4 patients had anomalous positions of the medial rectus insertion or pulley, excluding torsion of the entire rectus pulley array as the basis for the lateral rectus pulley malposition.

The study is retrospective and limited by the small number of patients. Postoperative follow-up was variable. Equatorial myopexy of the lateral rectus may not permanently maintain the lateral rectus pulley in the correct anatomic position, although the 2 subjects with long follow-up maintained alignment years after surgery. The lone reoperation on the lateral rectus muscle demonstrated that permanent sutures maintained normal lateral rectus paths well beyond the healing period.

Inferior displacement of the lateral rectus pulley should be suspected in patients with acquired esotropia without significant hyperopia as well as in patients with partially accommodative esotropia who develop overelevation in adduction. Surgical correction with lateral rectus tightening combined with equatorial myopexy to restore the normal lateral rectus pulling direction can result in stable correction of the esotropia.

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