LEARNING OBJECTIVES

1. Describe the pathogenesis of cyclovertical strabismus in the sagging eye syndrome
2. Recognize the clinical differences between neuropathic strabismus and strabismus caused by sagging eye syndrome
3. Describe the surgical and non-surgical management of strabismus caused by sagging eye syndrome

CME QUESTIONS

1. Degeneration of what connective tissue structure is mainly responsible for age-related distance esotropia?
   a. Lateral canthal tendon
   b. LR-SR band
   c. Levator aponeurosis
   d. Peritoneal ligament
   e. Medial canthal tendon

2. Which clinical feature differs between hypertropia due to sagging eye syndrome, rather than due to superior oblique palsy?
   a. Excyclotropia in the hypotropic eye
   b. Vertical incomitance
   c. Excyclotropia in the hypertropic eye
   d. Vertical nystagmus
   e. Acute onset of diplopia

3. Which anatomic features are characteristic of the heavy eye syndrome, but not of sagging eye syndrome?
   a. High axial myopia
   b. Inferonasal displacement of the inferior rectus muscle
   c. Inferior displacement of the lateral rectus muscle
   d. a, b, and c
   e. a and b

KEYWORDS

1. Strabismus
2. Esotropia
3. Hypertropia
4. Magnetic Resonance Imaging
5. Orbit

INTRODUCTION

Purpose: Sagging eye syndrome (SES) causes strabismus because orbital connective tissue degeneration allows inferior shift of the lateral rectus (LR) extraocular muscle pulleys. This paper reviews this relatively common, non-neurological cause of diplopia in older adults.

Methods: High-resolution MRI of the orbits in patients with SES has been quantitatively compared with normal subjects age-matched to SES, and normal young adults. MRI has defined rectus pulley locations comparable to age-matched norms, and has quantified lengths of the LR-SR band ligament and rectus EOMs. These data have been correlated with facial and adnexal features, and ocular motility. Results of strabismus surgery have been systematically assessed in age-related distance esotropia (ARDE, “divergence paralysis”) and cyclovertical strabismus (CVS) due to SES.

Results: Patients with SES commonly exhibit blepharoptosis and superior sulcus defect, and are likely to have previously undergone blepharoplasty or facelift surgery. The MRI hallmark of SES is significant inferolateral LR pulley displacement, with peripheral displacement of all other rectus pulleys, and lateral displacement of the inferior rectus pulley. Rectus muscles are generally elongated. Symmetrical LR sag is associated with ARDE, and asymmetrical LR sag >1 mm with CVS in which the hypotropic eye is also excyclotropic. The LR-SR band ligament is ruptured in the great majority of patients with SES. While there is no horizontal duction limitation or saccade slowing, patients typically have limited sursumversion due to medial rectus (MR) and LR sag. ARDE responds well to MR recession with augmented dosing, or to LR resection or plication. CVS may be treated by graded vertical rectus tenotomy or more conventional strabismus surgeries.
**Conclusions:** Rectus pulley displacement and EOM elongation, associated with connective tissue involution including LR-SR band rupture, causes acquired cyclovertical and horizontal strabismus in older patients.

**Clinical Relevance:** Small angle esotropia, hypertropia, and cyclotropia may result from common involutional changes in EOMs and orbital connective tissues that may be suspected from adnexal features.

**INTRODUCTION**

As at other anatomical sites, the orbital connective tissues change with aging,\(^1,2\) causing among other untoward effects aponeurotic blepharoptosis and limited supraduction due to inferior displacement of the horizontal rectus pulleys.\(^3,5\) Similar to the way that the externally visible adnexa age, the internal connective tissues in the orbit also involute. Not all such involutional changes are homogeneous, however. A particularly significant involution occurs in the LR-SR band, a ligament originating on the lateral border of the superior rectus (SR) pulley and terminating on the superior border of the lateral rectus (LR) pulley. The LR-SR band vertically supports the LR pulley against the downward force of the inferior oblique muscle, whose orbital layer inserts on the LR pulley.\(^6\) Degeneration of the LR-SR band permits inferior sag of the LR pulley, causing esotropia, or cyclovertical strabismus, or both.\(^2,7,8\) Bilateral inferior shift of the LR pulley may mechanically produce age-related distance esotropia (ARDE), also variously known as divergence paralysis esotropia, “divergence insufficiency,” “divergence insufficiency esotropia,” and “divergence paresis esotropia.” ARDE is characterized by esotropia at distance fixation, orthotropia or esophoria at near fixation, normal horizontal duction range, and normal horizontal saccadic velocities.\(^7,8\) Asymmetrical inferior shift of the LR pulley has been postulated to produce cyclovertical strabismus (CVS) because of unbalanced conversion of some of the forces of the two LR muscles from abduction to infraduction. Clinical strabismus patterns correlate closely with anatomical changes observable by orbital imaging. It is important to understand SES as a cause of acquired diplopia, because this diagnosis is not indicative of an acute neurological event, and does not require extensive or costly investigations as might be typically performed for acutely acquired diplopia due to neurological lesion.\(^9,10\) Moreover, the mechanical etiology of SES presents a favorable prognosis for appropriately designed surgical treatment.\(^11,12\)

This presentation reviews the quantitative anatomy of SES as revealed by high resolution magnetic resonance imaging (MRI) of the rectus EOMs, pulleys, and the LR-SR band ligament. Details of the studies have been published previously.\(^3,8,13,14\)

**METHODOLOGY**

Detailed quantitative results were obtained by surface coil orbital MRI in 28 subjects (11 males, 17 females) of average age 69±12 years who had acquired horizontal or cyclovertical diplopia. Age-related distance esotropia (ARDE), defined as orthophoria or asymptomatic esophoria of ≤10° at near, with distance esotropia measuring double or one and a half times the measured near esophoria, existed in 11 subjects.\(^12\) CVS, defined as hypertropia exceeding 2° with or without cyclotropia, existed in 17 subjects. Patients were excluded if they had superior oblique (SO) palsy, thyroid eye disease, trauma, prior strabismus surgery, or myopic degeneration suggestive of the “heavy eye” syndrome.\(^4\)

Patients had attained mature age, with mean age for ARDE (3 males, 8 females) 72±11 years, and for CVS (7 males, 10 females) 68±2 years. Mean esotropia in ARDE was 12±11° at distance and 1±3° at near. ARDE was vertically comitant. Ten subjects had pure CVS, and 7 more had CVS with associated esotropia. Average hypertropia of these subjects was 10±10°. Fundus torsion was objectively determined by slit lamp measurement of the fovea to optic disc angle.\(^15,16\)

High resolution MRI was performed using surface coils in target-controlled central gaze by each scanned eye.\(^17,18\) Using 2 mm thick quasi-coronal images perpendicular to the orbit, cross sections of the rectus EOMs and the SO were analyzed in standard, oculocentric coordinates to identify rectus pulley positions for comparison with norms.\(^17,18\) Maximum SO cross sections were determined to exclude neurogenic SO atrophy due to SO palsy. Horizontal rectus EOM path lengths were determined from axial images, and vertical rectus EOM path lengths from quasi-sagittal images. Lengths of the superior (SR) and inferior rectus (IR) muscles were determined in 18 orbits of 9 patients with SES (average age 64±4 years), and compared to 64 orbits of 34 normal young (average age 24±4 years) and 15 orbits of 9 normal older controls (average age 62±5 years).

**QUANTITATIVE FINDINGS**

**EXTERNAL FEATURES IN SES.** Adnexal sag was highly prevalent in SES. Retraction of the upper eyelid into the superior orbit (superior sulcus deformity) was evident in 64% of subjects with SES; 29% exhibited aponeurotic blepharoptosis and high upper eyelid crease. Previous blepharoplasty, brow or facelift surgeries had been performed in 29% of patients with SES.

**RECTUS PULLEYS IN ARDE.** Both the MR and LR pulleys were both significantly displaced centrifugally from the orbital center (P<0.005). The LR pulley was 4 mm more lateral than in younger control orbits, while also being bilaterally symmetrically inferolocated by a significant average of 5.9 mm. The MR pulley was about 2.4 mm more medial than in younger control orbits, while also

---

\(^1\) References omitted for brevity.
being significantly inferoplaced by 3.4 mm. The IR pulley was displaced about 5 mm temporally and about 3 mm inferiorly than in young and older normal subjects.

**RECTUS PULLEY POSITIONS IN CVS.** The MR pulley of the hypotropic eye was 2.6 mm inferior and 1.6 mm temporal to younger normal (P< 0.005), but not significantly different from older normal subjects. The SR pulley in CVS was about 1 mm inferior to that of normal younger subjects (P<0.005), but similar to normal older subjects. The MR and SR pulleys in the hypertropic eyes of subjects with CVS were normally located. The LR pulley in the hypotropic eye in CVS was about 4 mm temporal and 6 – 8 mm inferior to that of younger and older controls (P<0.005 for both), while the LR pulley in the hypertropic eye was only about 4 mm inferior. The LR pulley in the hypotropic eye in CVS was about 4 mm more lateral than in both younger and older controls (P<0.005). The inferior rectus (IR) pulley was displaced temporally from both younger and older controls by 4–5 mm in both the hypotropic and the hypertropic eyes in CVS (P<0.005).

As noted above, inferior LR pulley displacement was bilaterally symmetrical in ARDE, differing from right to left on average only 0.3±0.1 mm (P=0.8). Quite different was CVS, in which binocular LR displacement asymmetry averaged 2.6±1.8 mm (P=0.04). For individuals with ARDE, asymmetry of LR sag was always less than 0.5 mm, while in CVS, LR pulley infraplacement in the hypotropic orbit always exceeded that in hypotropic orbit by at least 1 mm. No other difference in pulley positions between hypertropic and the hypotropic eyes was statistically significant (P>0.05). This difference is mechanistically consistent with hypotropia in the eye with greater LR pulley infraplacement, and also consistent with observations of fundus cycloposition. The eye that exhibited greater hypotropia and greater LR sag exhibited 12±6° excycloposition, more than the hypotropic fellow eye that exhibited 7±5° excycloposition (P=0.01.)

**RECTUS MUSCLE PATH LENGTHS.** The LR path was markedly elongated in SES. In younger controls, LR path length averaged 33±6 mm, similar to 31±14 mm in older people without strabismus. The LR path was approximately 40% longer, at 45–47 mm, in both ARDE and CVS (P<0.005). The MR path was not as elongated as the LR in SES (P < 0.000002), but nevertheless was about 25% longer, at 38–39 mm, in both ARDE and CVS than the length of 31±6 mm in younger and older controls (P<0.005). Path length of the SR in both ARDE and CVS was 3–6 mm longer than in younger and older controls, but IR length was not abnormal.

**PATHOLOGY OF LR-SR BAND.** Reflecting age-related degeneration, the LR-SR band was elongated by about ~12 mm in non-strabismic older than younger controls (P < 0.005), but no control exhibited rupture of the LR-SR band. The superior pole of the LR was angulated laterally from the vertical by 6±9° in younger controls, but significantly more so at 18±7° in older normal subjects (P=2.1 x 10^9). Patients with SES exhibited LR-SR band attenuation, stretching, or often even rupture. There was superotemporal bowing of the LR-SR band in milder cases, and abrupt termination of an attenuated remnant in severe cases. The LR-SR band was ruptured in 14/22 orbits with ARDE, and in CVS, it was ruptured in 31/34 orbits; these proportions are statistically similar. In subjects with ARDE having intact LR-SR bands, band length and LR angulation were all significantly exceeded by values observed in normal young controls. From these findings it is inferred that LR-SR band ligament rupture is associated with asymmetrical LR sag that causes hypotropia, because the difference in combined LR plus MR sag in the hypotropic and hypertropic eye in CVS is significantly correlated with the magnitude of hypotropia (P<0.02).

**TREATMENT OF ARDE.** Base-out spectacle prism has been the historical mainstay for treatment of ARDE, particularly for small angles of esotropia in patients whose refractive errors demand corrective lenses for adequate visual acuity. Since convergence fusional amplitudes are typically robust in ARDE, base-out prism in the entire spectacle lens typically does not induce convergence insufficiency during near viewing. It often does not even induce exophoria at near. However, continuing refinement of refractive and cataract surgical technique has now created a population of patients with ARDE who are not dependent on corrective lenses for visual acuity, and who are loathe to wear spectacles merely for their prismatic effects. Spectacle prisms also suffer from disadvantages of weight and optical aberrations, and cannot be tailored to lateral incomitance. Strabismus surgery, probably of any sort that reduces esodeviation, is effective for ARDE as long as convergence fusional amplitudes at near exceed the surgically-induced exoshift; this criterion is very commonly fulfilled in clinical practice. While LR resection has been preferred historically based upon the now-discredited presumption that LR surgery is more effective for distance than near viewing, comparative study demonstrates that MR recession is effective, and equally so to LR resection if performed using modestly augmented dosage targeted, using conventional tables, to twice the esotropia angle measured at distance. Lateral incomitance deserves therapeutic consideration. Even historical descriptions of ARDE noted that distance esotropia might be present mainly or even exclusively in lateral gaze; the angle need not be the same in both dextroversion and levoversion. Because recession of one MR has maximal effect on distance esotropia in contralateral gaze, with little or no effect in ipsilateral gaze, most cases of ARDE treated by MR recession require the procedure bilaterally.

**TREATMENT OF CVS.** Of course, vertical spectacle prism has long been effectively employed for cases of small vertical heterophoria due to SES, and this remains a reasonable approach. Lens weight is seldom a serious disadvantage. A more serious problem is vertical gaze incompatibility, with
heterotropia often varying significantly with vertical gaze direction. Vertical rectus recession or resection, even with adjustable sutures, typically produces larger changes in vertical alignment than the heterophorias associated with SES. A more suitable therapy is partial vertical rectus tenotomy at the insertion. When performed in progressively graded fashion under topical anesthesia, partial vertical rectus tenotomy at the insertion can precisely connect hypertropia of 1 - 8D. Moreover, partial IR tenotomy from the temporal side produces incycloduction helpful to treat the exyclotropia usually encountered in the hypotropic eye in CVS due to SES; the mechanism can be considered similar to nasal transposition of the IR insertion, since only the nasal IR tendon fibers remain intact. Partial SR tenotomy from the nasal side also produces incycloduction in a manner analogous to temporal transposition of the insertion, since only the temporal SR tendon fibers remain intact. Of course, for hypertropia exceeding 8D, more conventional oblique or vertical rectus muscle surgery may be effective.

DISCUSSION
Key to the pathogenesis of SES is degeneration of the LR-SR band ligament interconnecting the SR and LR pulleys so as normally to maintain the LR pulley’s vertical position against inferior traction exerted by the inferior oblique (IO) muscle, whose orbital layer inserts on the LR pulley. In serially-sectioned autopsy orbits, quantitative densitometry has demonstrated progressive collagen and elastin loss, associated with attenuation and rupture of the LR-SR band ligament with age. This phenomenon is associated with gross LR pulley sag, presumably because downward IO force becomes unopposed after LR-SR band rupture. In a 93-year old post-mortem orbit, the LR-SR band was ruptured and its remnants severely attenuated. Rutar and Demer first proposed that three elderly, non-myopic patients who presented with unilateral hypertropia and divergence paralysis type esotropia exhibited a previously unrecognized entity they termed SES, and noted association with orbital connective tissue degeneration manifested by prominent eyelid fat pads, superior sulcus deformity, aponeurotic blepharoptosis, and proclivity towards blepharoaplasty or similar surgery. Subsequent quantitative imaging has confirmed and extended original suppositions about SES. Inferior LR pulley displacement is a universal feature of SES. All subjects with CVS, with or without associated esotropia, exhibited highly significant asymmetrical inferior LR pulley displacement, characteristically greater in the hypertropic and more exyclopositioned eye. A useful additional observation is that the rectus muscles are elongated in SES. Although even non-strabismic older subjects exhibit appreciable lengthening of the otherwise-intact LR-SR band, the ligament was ruptured in two-thirds of orbits with ARDE and nearly all with CVS. This high prevalence for LR-SR band rupture supports occasional patient accounts of sudden, painful onset of diplopia in SES. Presumably, catastrophic LR-SR band rupture creates a sudden horizontal and/or vertical LR force imbalance sufficient to cause diplopia. The inferred causal connection is supported by the quantitative observation that magnitude of vertical strabismus is correlated with magnitude of horizontal rectus pulley sag.

Muscle length, as embodied in the classical length-tension curve, is a parameter implicit to all physiologic studies of muscle behavior, yet with the only very recent application of in vivo imaging has extraocular muscle length received scientific or clinical attention. Rectus muscle length, as uniformly assessed in central gaze position, does not change appreciably in concomitant esotropia and exotropia. However, rectus muscle lengths are abnormal in SES, corresponding to displacement of the rectus pulleys. All four rectus pulleys are significantly displaced by 2 to 14 mm from the orbital center in SES. Although normal aging is also associated with some centrifugal pulley displacement, the changes in SES greatly exceed those in non-strabismic subjects of comparable age.

Since the shortest distance between points is a straight line, centrifugal pulley displacements necessarily imply that the muscles passing through the pulleys from origin to scleral insertion must traverse greater than normal paths, and at least in this sense these muscles are abnormally long. Thus, MRI measurements demonstrate horizontal EOM lengths in SES that are ~40%, or 14 mm, longer than the EOMs of normal young and older controls. These elongations are large in comparison to resections and recessions performed typically performed for strabismus surgery. What would happen if otherwise normal muscles were suddenly stretched to the lengths observed here? Computational modeling using the Orbit 1.8 program predicts significant exotropia, unless muscle rest length is commensurately increased to conform to increased muscle path length. One can suppose that many of the changes in pulley position and corresponding changes in muscle lengths develop gradually over a long period of time by chronic remodeling, perhaps including addition of sarcomeres, or insertion of passive connective tissue in series with sarcomeres. This remodeling would likely have other consequences for treatment of strabismus. It is possible that the marked rectus EOM elongation in ARDE explains the observed requirement for atypically large dosages of MR recession for surgical correction. Widespread anatomical changes in the orbits in SES probably also impact the effects of other forms of strabismus surgery. Adaptive remodeling may at least in part result from orbital connective tissues thinning that allows both pulley shifts and orbital fat migration, as well as secondary adaptive and pathological changes in muscles and central innervation patterns. It is indeed remarkable that the extreme EOM elongations and pulley shifts observed in normal older subjects do not more frequently cause strabismus, and that

456 | North American Neuro-Ophthalmology Society
patients with SES do not exhibit more severe strabismus than typically observed. Fusional vergence mechanisms are likely protective against symptomatic diplopia, as evidenced by a longitudinal observational study demonstrating progressive increase in distance esophoria in ARDE, but with development of diplopia only when esophoria exceeded fusional divergence amplitude.19

The foregoing studies of SES excluded well-understood causes of strabismus such as orbital trauma, thyroid ophthalmopathy, restriction, and EOM paralysis. After these clinical exclusions, the remaining cases proved to be SES. In one specialty strabismus practice in Florida, for example, 17% of 200 cases of adult acquired esotropia were attributed to ARDE, and 96% of these were successfully treated by strabismus surgery33. The clinician can thus infer that many cases of similar acquired adult DPE and CVS are likely to be caused by SES, and are likely to exhibit the characteristic adnexal signs including blepharoptosis and superior sulcus defect. However, many subjects with SES had previously undergone blepharoplasty or facelift, so some of the orbital findings may have been partially iatrogenic.

Patients with SES are recognizable from their external appearances and motility patterns. Typical findings include generalized elongation of the levator aponeurosis, resulting in a superior sulcus deformity, aponeurotic ptosis, or lid crease elevation compensated by frontalis recruitment. Most patients with SES exhibit symmetrically limited supraduction, although all exhibit full horizontal ductions and normal horizontal saccadic eye velocities that should be clinically required to exclude abducens palsy. If these conditions are met, do patients with clinical evidence of SES require orbital imaging? If such patients present with horizontal binocular diplopia for distant but not near targets, and report no associated neurological complaints, further etiological investigations are probably unnecessary even if the onset of diplopia is acute. Patients presenting with acute or chronic onset of vertical binocular diplopia in clinical circumstances suggestive of SES may also be spared neuro-ophthalmological investigations when no evidence exists of cranial neuropathy or other acute neurological deficit.

Neuro-ophthalmologists should be aware that CVS due to SES can mimic features of SO palsy. The 3-step test, heretofore considered the gold standard for diagnosis of SO palsy, has now been demonstrated to have only about 70% sensitivity34 and 50% specificity for deficits of SO structure or function that can be demonstrated by high resolution MR[35]. Since even normal subjects adapted to wearing of a vertical prism develop an hyperphoria that varies with head tilt36, head tilt dependence of hypertropia should probably be regarded as a non-specific concomitant of any vertical heterophoria, not as the signature of SO palsy34. Because all subjects with CVS contributing to the foregoing data exhibited normal SO size by MRI, SO palsy due to denervation could not have been a confounding factor here. The busy clinician might well be faced with a more difficult diagnostic dilemma. A clinical finding that may be helpful is that while in SES, the hypertropic eye is excyclotropic, in SOP the hypertropic eye is excyclotropic. If cycloptropia is not a sufficiently clear diagnostic sign and the clinical situation warrants, then high resolution orbital imaging may be helpful in confirming or excluding SO palsy based upon the presence or absence of SO muscle atrophy, and in confirming or excluding SES based upon LR position.

CME ANSWERS
1. b. LR-SR band
2. a. Excyclotropia in the hypertropic eye
3. e. a and b

REFERENCES


