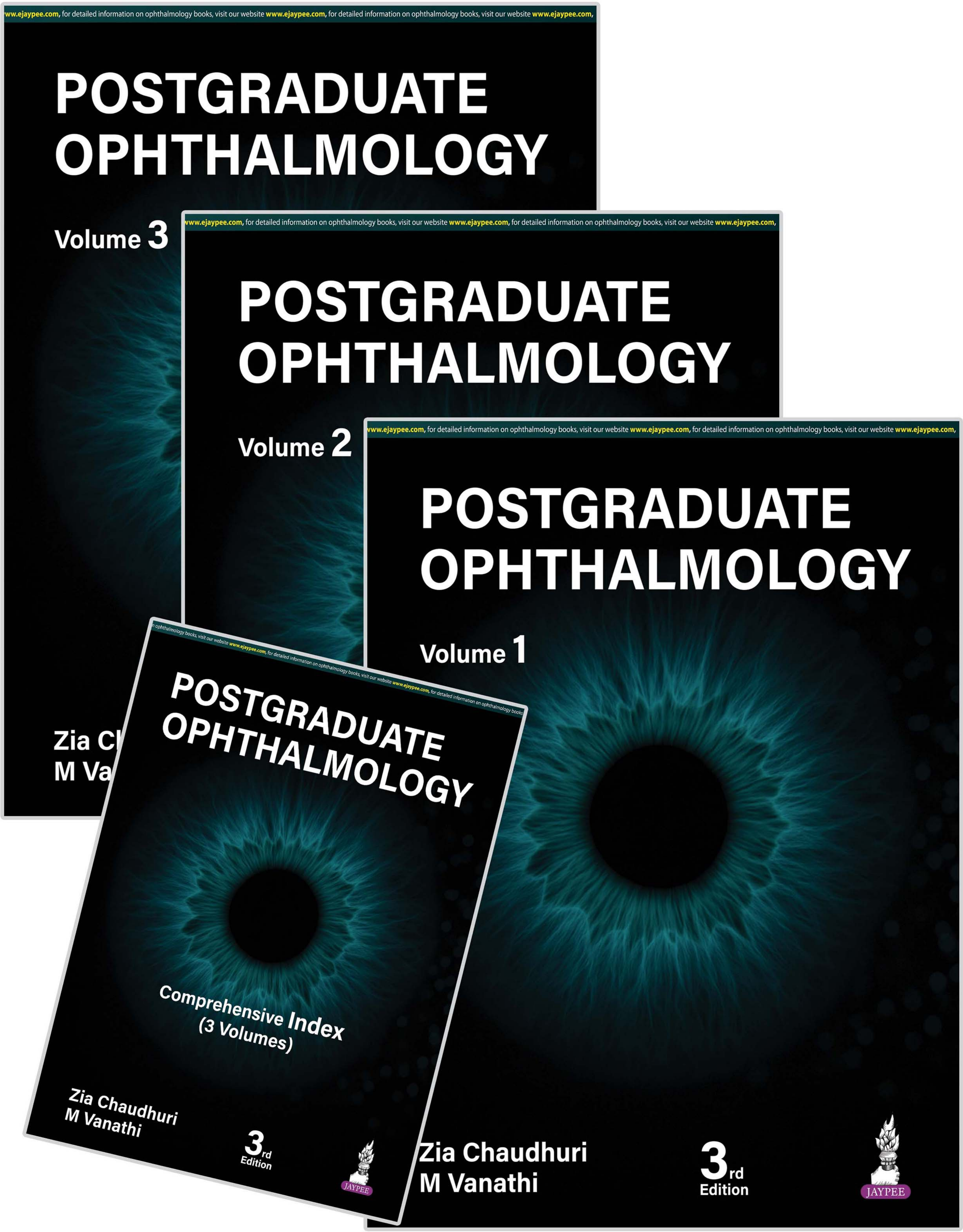
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CHAPTER 11.4

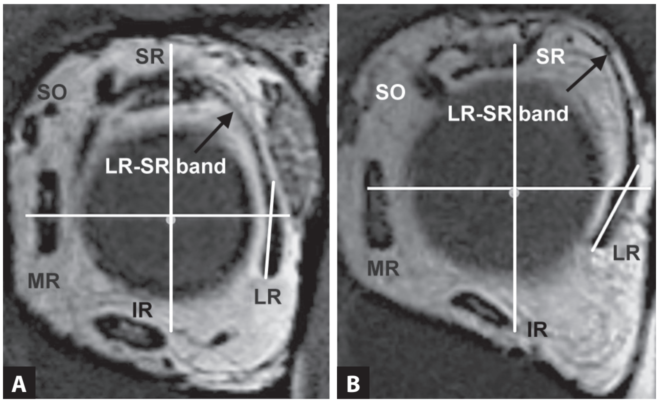
Neuroanatomical Strabismus and Pulleopathies

Zia Chaudhuri, Joseph L Demer

Ocular misalignment may result from primarily neurogenic, myogenic or restrictive origins. However, strabismus may also arise from identifiable structural anomalies of extraocular muscle (EOM) paths through their pulleys within the orbit, as may be objectively identified by direct imaging through high resolution and surface coil MRI. Abnormalities of EOM paths may produce strabismus by altering the pulling directions of the EOMs. These pulley positions may be determined quantitatively by direct imaging of the orbit and the EOMs. These congenital and acquired "pulley" abnormalities can now be classified a "pulleopathies". Identification of these conditions has translational implications for the appropriate surgical management of patients. This chapter overviews some of these recently identified conditions.

SAGGING EYE SYNDROME

Sudden-onset diplopia in adults has classically considered an ominous neurological event necessitating extensive and economically burdensome investigations.[1,2] This section emphasizes the etiologic importance of orbital tissue degeneration during aging, which can result in the resultant sagging eye syndrome (SES), an important, non-neurological cause of acquired diplopia in older people.[3,4] The lateral rectus–superior rectus (LR-SR) band is a ligament interconnecting connecting the lateral border of the SR pulley with the superior border of the LR pulley. Degeneration of the LR-SR band permits inferior sag of the LR pulley. Large or asymmetrical sage of the LR path can cause esotropia, or cyclovertical strabismus (CVS), or both **(Figs. 1A and B)**.[5-9] Common clinical correlates of SES include aponeurotic blepharoptosis, lower eyelid bags and laxity, and limited supraduction due to inferior displacement of the horizontal rectus pulleys.[3,4,10] Bilaterally symmetrical inferior shift of LR path may mechanically cause divergence paralysis esotropia (DPE), also termed "divergence insufficiency", "divergence insufficiency esotropia", "divergence paresis esotropia", and "age-related distance esotropia (ARDE)". Under any of these names, this disorder is characterized by esotropia at distance, fusion of little or no esophoria at near, normal abduction range, and normal horizontal saccadic velocities.[6,7,11-13] Asymmetrical inferior shift of the LR pulley may cause CVS **(Fig. 2)**.[8,9,14] The mechanical etiology of strabismus caused by SES presents a more favorable prognosis for surgical treatment that would a neurological cause and is devoid of ominous neurological portent.[6,7,9]



Figs. 1A and B: T1-weighted quasi-coronal MRI through the anterior left orbit. (A) Young subject demonstrating a sturdy LR-SR band and the LR muscle in normal position. A line passing horizontally through the center of the globe bisects the LR muscle center. The long axis of the LR muscle is nearly vertical and parallel to a vertical line passing through the center of the globe; (B) Elderly subject without strabismus demonstrates a thin, elongated LR-SR band with the LR muscle sagging beneath a horizontal line passing though the center of the globe. The long axis of the LR muscle is oblique to a vertical line passing through the center of the globe. (LR: lateral rectus; MR: medial rectus; SO: superior oblique; SR: superior rectus)

Recent studies have identified the mechanisms and clinical associations of SES. All patients with SES, whether pure esotropia as in DPE, combined esotropia and CVS, or pure CVS, exhibited inferior displacement of one or both LR pulleys.[7,8] In each case, the eye with the greater inferior LR pulley displacement exhibits greater hypotropia and excyclotropia than the hypertropic fellow.[8]

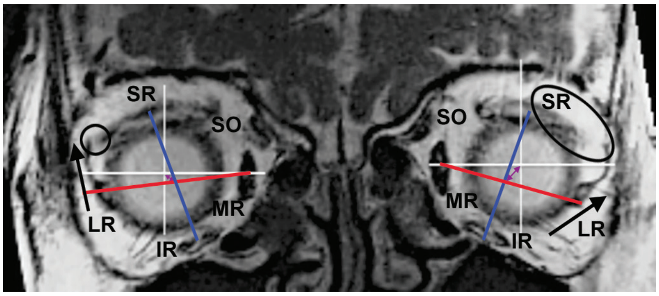
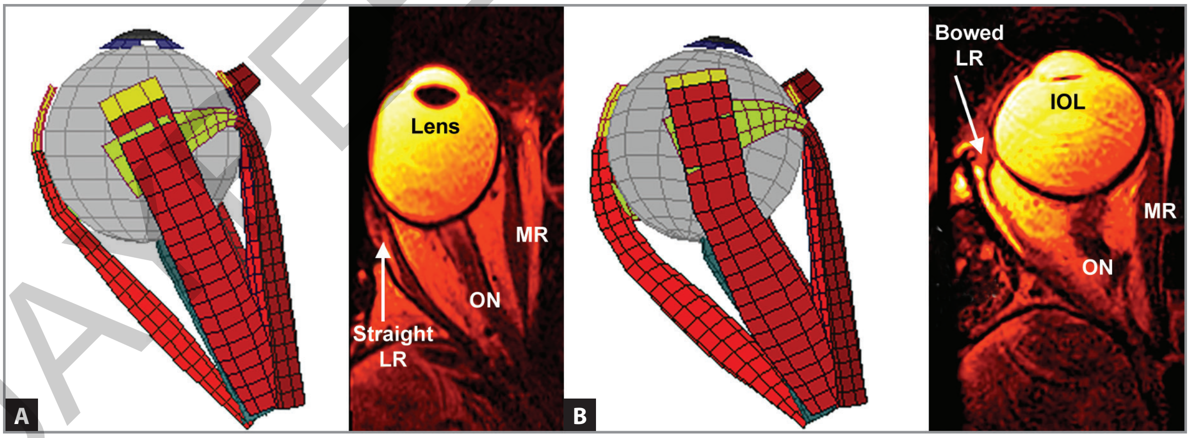


Fig. 2: Quasi-coronal, T2-weighted MRI of an elderly subject demonstrating asymmetrical LR sag greater in the left than right eye. Although there is rupture of the right LR-SR band (circle) with inferior LR, it is still apposed to the globe. There is complete rupture of the left LR-SR band (oval ring) with inferior LR sag. There is increased obliquity associated with SES of the LR muscle's long axis on the left side versus the right side with respect to a vertical line passing through the center of the globe on both sides (black arrows). This subject presented with LE hypotropia and excyclotropia, characteristic of cyclovertical strabismus. (LE: left eye; LR: lateral rectus; MR: medial rectus; SO: superior oblique; SR: superior rectus; SES: sagging eye syndrome)

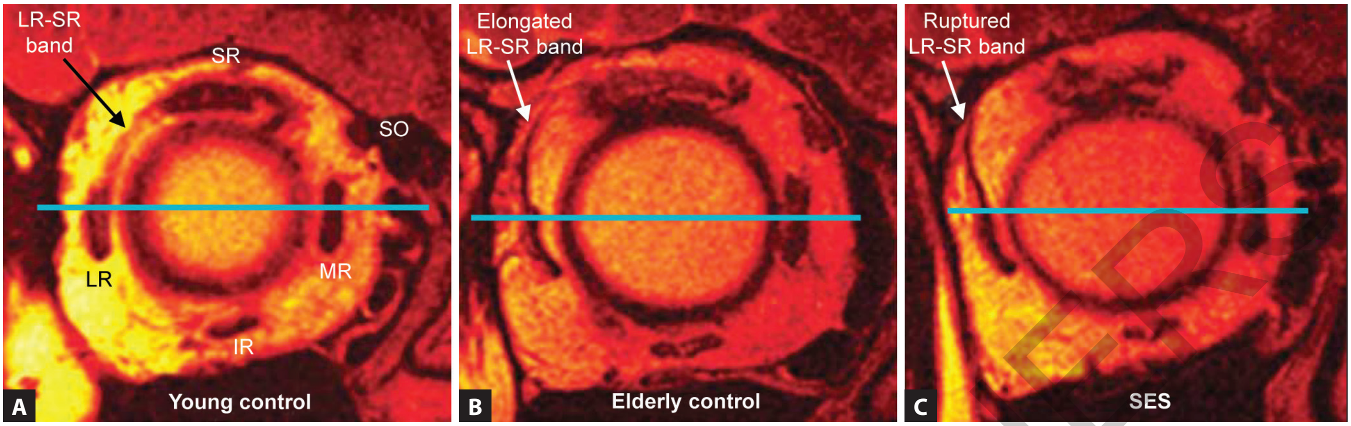
MRI studies have documented the pathophysiological mechanisms of SES. Every case of SES, whether pure esotropia as in DPE, combined esotropia and CVS, or pure CVS, also exhibited striking elongation of the rectus EOMs **(Figs. 3A and B)**.[8] Although even nonstrabismic older subjects exhibited a roughly 50% lengthening of the otherwise-intact LR-SR band, the ligament was ruptured in 64% of orbits with DPE and 91% of orbits with CVS.[8,14] This high prevalence of LR-SR band rupture supports reports of sudden, painful onset of both horizontal and vertical diplopia in SES, and suggests a mechanical mechanism for the associated oblique angulation of the LR orientation relative to the vertical in SES **(Figs. 1A and B)**. Presumably, the LR suddenly slips downward and its tendon tilts into an oblique orientation when the LR-SR band catastrophically ruptures. The amount of hypertropia in CVS was correlated with the amount of horizontal rectus pulley sag.

The MRI investigations have also showed displacement of all four rectus pulleys away from the orbital center in in SES, by amount ranging from 2 to 14 mm, and were much greater than in affected people than healthy people of similar age **(Figs. 4 to 6)**.[8] The inferior rectus (IR) pulley was also displaced more laterally in both SES and in nonstrabismic older subjects. Lateral displacement of the IR pulley could be due to age-related degeneration of the insertion upon it of the orbital layer of the



Figs. 3A and B: (A) Orbit 1.8 computational simulation of a normal globe and EOMs with corresponding axial MRI of the orbit demonstrating normal length and contour of the LR and MR muscles with central gaze position; (B) Orbit 1.8 computational simulation of SES incorporating elongated and lax EOMs with esotropic gaze. This corresponds to MRI at right showing elongated and bowed LR and MR muscles. Computer simulations of the changes in SES observed by MRI thus predict esotropia. (EOM: extraocular muscle; IOL: intraocular lens; LR: lateral rectus; MR: medial rectus; ON: optic nerve; SES: sagging eye syndrome)

Source: MRI photographs reproduced with permission from American Medical Association, March 2020. Chaudhuri Z, Demer JL. Sagging eye syndrome: Connective tissue involution as a cause of horizontal and vertical strabismus in older patients. JAMA Ophthalmol. 2013;131(5):619-25.



Figs. 4A to C: Fast spin-echo T2-weighted sequence quasi-coronal plane magnetic resonance imaging. (A) Younger control participant showing lateral rectus–superior rectus (LR-SR) band. Note the normal morphology of LR muscle with respect to a horizontal reference line drawn through the globe center; (B) Elderly control participant demonstrated marked elongation of LR-SR band associated with LR muscle sag; (C) Rupture of LR-SR band in sagging eye syndrome (SES) with resultant LR sag. (IR: inferior rectus; MR: medial rectus; SO: superior oblique)

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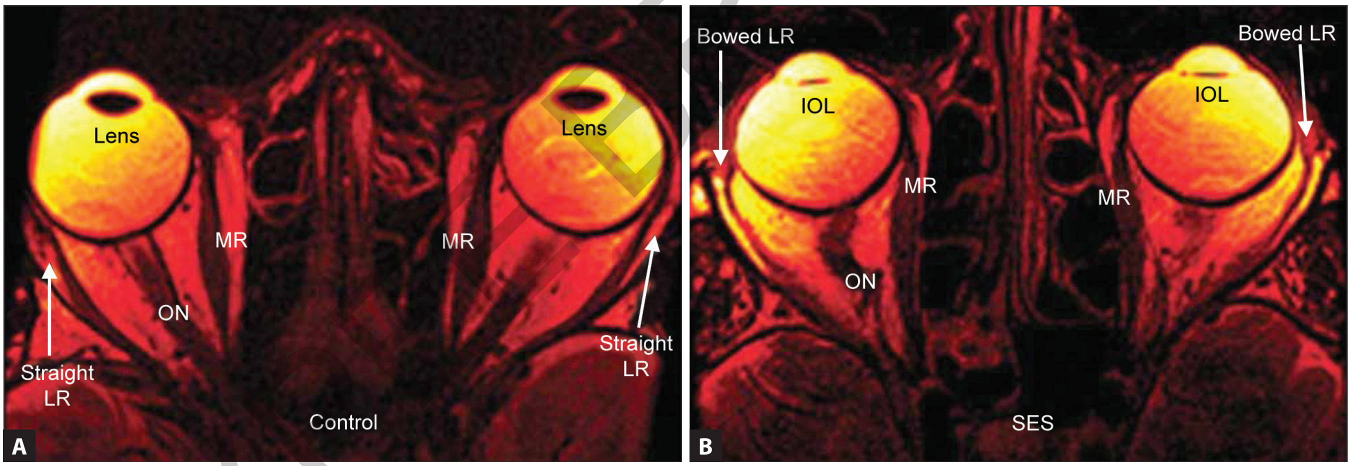

Figs. 5A and B: Fast spin-echo T2-weighted sequence axial magnetic resonance imaging. (A) Control participant illustrates medial rectus (MR) and lateral rectus (LR) configuration; (B) Marked elongation and thinning of LR more than in sagging eye syndrome (SES). (IOL: intraocular lens; ON: optic nerve)

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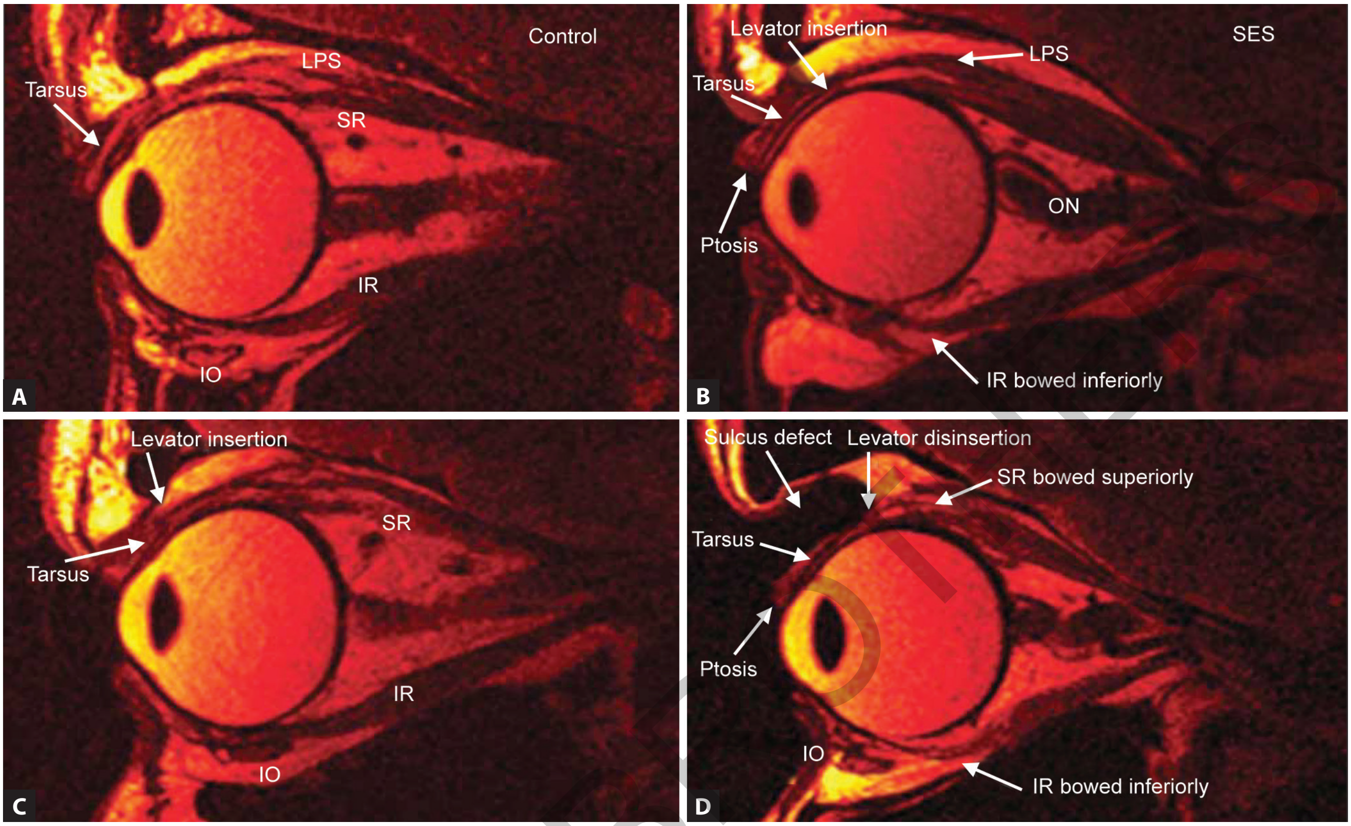
inferior oblique (IO) muscle, as histologically demonstrated.[15,16] **Tables 1 and 2** demonstrate the pulley displacements, EOM muscle lengths and the LR-SR band length observed in subjects with SES causing DPE and CVS as well as in healthy young and older controls.

Centrifugal displacements of rectus pulleys in SES lengthen EOM paths from origin to scleral insertion and explain elongation of horizontal EOM lengths in SES, which were up to approximately 40%, or 14 mm, longer than normal **(Figs. 5 and 6)**. These elongations of rectus EOMs in SES are substantial relative to the resections and recessions of rectus EOMs performed in typical strabismus surgery. Rectus EOM elongation in DPE probably explains the requirement for augmented dosages of



Figs. 6A to D: Fast spin-echo T2-weighted sequence quasi-sagittal magnetic resonance imaging. (A and B) Morphology and length of the superior rectus (SR) and inferior rectus (IR) muscles in a young control participant. The insertion of the levator palpebrae superioris (LPS) to the tarsus and normal eyelid anatomy is visible; (C) Aponeurotic ptosis and bowing of the IR muscle in sagging eye syndrome (SES). The LPS is minimally attached to the tarsus. The optic nerve (ON) is convoluted; (D) Another case of SES showing levator disinsertion, ptosis, and marked superior sulcus defect. The SR and IR are markedly elongated and bowed. (IO: inferior oblique)
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TABLE 1: Rectus pulley positions relative to globe center.

	Rectus length, Mean (SD), mm							
	Medial		*Superior*		*Lateral*		*Inferior*	
Group	*Lateral*	*Superior*	*Lateral*	*Superior*	*Lateral*	*Superior*	*Lateral*	*Superior*
DPE	–12.15 (0.69)[a]	–3.51 (2.0)[a]	–2.31 (3.52)	10.18 (0.69)	14.13 (1.56)[a]	–6.2 (1.6)[a]	1.36 (2.27)[a]	–15.17 (1.17)[a]
CVS:								
• Hypertropic	–12.8 (1.39)	–2.2 (2.03)	–2.8 (2.6)	10.9 (1.2)	14.4 (1.49)[b]	–6.1 (3.7)[b]	0.2 (1.68)[b]	–15.6 (2.27)[b]
• Hypotropic	–12.8 (1.51)[c]	–2.7 (1.6)[cc]	–2.8 (3.23)	10.5 (1.23)[c]	14 (1.54)	–8.8 (3.5)[c]	0.8 (1.88)[c]	
Controls:								
• Older	–14.5 (0.9)[a]	–1.05 (1.11)[a]	–2.06 (1.85)	11.5 (1.11)	10.13 (0.8)[a,b,c]	–2.03 (1.7)[a,c]	–5.14 (0.9)[a,b,c]	–12.6 (0.83)[a,b,c]
• Younger	–14.4 (0.01)[a,c]	–0.1 (1.24)[a,c]	–2.3 (0.9)	11.8 (0.84)[c]	10.1 (0.6)[a,b,c]	–0.3 (1.03)[a,b,c]	–5.4 (0.9)[a,b,c]	–12.2 (1.01)[a,b,c]

[a]Significant differences between DPE and control groups ($p < 0.005$)
[b]Significant differences for the hypertropic eye of CVS versus controls
[c]Significant differences for the hypotropic eye of CVS versus controls
(CVS: cyclovertical strabismus; DPE: divergence paralysis esotropia)
Source: Reproduced with permission from American Medical Association, March 2020. Chaudhuri Z, Demer JL. Sagging eye syndrome: Connective tissue involution as a cause of horizontal and vertical strabismus in older patients. JAMA Ophthalmol. 2013;131(5):619-25.

TABLE 2: Rectus muscles and LR-SR band.

Group	Rectus length, Mean (SD), mm				LR-SR band		
	Medial	Superior	Lateral	Inferior	Length, Mean (SD), mm[a]	Angle, Mean (SD), deg[a]	Rupture (%)
DPE	37.8 (16.4)	40.7 (1.2)	45.3 (16.0)	37.0 (4.7)	12.7 (5.6)	22.4 (5.6)	64
CVS	39.4 (6.2)	43.4 (3.9)	46.8 (5.8)	42.1 (5.7)	14.1 (5.6)	23.6 (13.1)	91
Older controls	29.2 (7.2)	37 (4.0)	30.9 (14.2)	39.3 (4.3)	12.4 (2.9)	17.6 (7.2)	0
Younger controls	31.0 (5.5)	36.1 (2.9)	32.6 (5.6)	39.4 (2.8)	8.5 (1.5)	5.7 (8.9)	0

[a]Values reported only for cases without LR-SR band rupture.
(CVS: cyclovertical strabismus; DPE: divergence paralysis esotropia; LR-SR: lateral rectus–superior rectus)
Source: Reproduced with permission from American Medical Association, March 2020. Chaudhuri Z, Demer JL. Sagging eye syndrome: Connective tissue involution as a cause of horizontal and vertical strabismus in older patients. JAMA Ophthalmol. 2013;131(5):619-25.

medial rectus (MR) recession surgical correction.[7,8] Widespread anatomical changes in the orbits are probably relevant to other forms of strabismus surgery in patients with SES. These changes may result from attenuation of orbital connective tissues that in turn permit both shifts in EOMs and orbital fat that could induce adaptive and pathological effects. Actually, it is remarkable that the extreme EOM elongations and pulley shifts observed in subjects with SES do not more often produce more severe strabismus than usually observed. The potential impact of EOM elongation deserves further consideration in other forms of strabismus. Elongation was not observed in other forms of concomitant horizontal strabismus.[17]

Patients with SES are recognizable from their external appearances and motility patterns, so routine orbital imaging is not clinically warranted. Typical adnexal findings of SES include elongation of the levator aponeurosis, resulting in a superior sulcus deformity, aponeurotic ptosis, and high eyelid creases. An important clinical clue differentiating vertical strabismus in SES from superior oblique palsy (SOP) is that while in SES, the hypotropic eye is excyclotropic, in SOP the hypertropic eye is instead excyclotropic. Since SES is typically associated with clinically obvious adnexal changes, recognition of this SES as the cause of chronic or acute, acquired diplopia can usually avert neurological evaluation and imaging. However, adnexal changes in SES are common, so it is possible that an individual patient might have both these and a second pathology, such as cranial nerve palsy, disorder of neuromuscular transmission such as ocular myasthenia gravis, or mitochondrial myopathy such as chronic progressive external ophthalmoplegia. If the clinical presentation is typical of SES and there is no evidence of associated neurological symptoms, no limitation of duction or saccade slowing, no fatigue variation, or temporal variability of the strabismus, then imaging or other neurological investigations are generally unnecessary. On the other hand, coexistence of these factors may indicate need for appropriate clinical investigations, including neurological consultation if warranted.

Due to the mechanical etiology of both horizontal and vertical strabismus in SES, surgical correction under topical intraoperative adjustment has proven to be of value in ameliorating diplopia. MR recession effectively treats DPE without inducing convergence insufficiency at near.[7] However, greater MR recession is required compared with other common forms of concomitant esotropia.[7] For correction of DPE, by MR recession, conventional surgical tables may be employed, with a target angle equal to twice the largest esotropia angle measured at distance in any gaze direction.[7]

General efficacy of tenotomy in small angle CVS has been described qualitatively.[18-21] We suggested an approximate scheme for graded vertical rectus tenotomy (GVRT) dosage expressed as percentage of tendon width required for measured hypertropia. Correction of 2Δ hypertropia is on average accomplished with about 40% tenotomy, 4Δ with 60%, and 6Δ with 80%. This is similar to the recommendation of Alan B Scott, who suggested that 60–70% tenotomy corrects 4Δ hypertropia.[21] However, because of substantial variability in response, it is prudent to perform surgery be performed in a stepwise manner, considering these recommendations as initial guidelines. Current data are insufficient for quantitative recommendations for hypertropia exceeding 8Δ. While the maximum deviation corrected by this procedure was occasionally as much as 10Δ with 90% tenotomy, hypertropia of this magnitude might preferably be managed using conventional adjustable suture recession of a vertical rectus muscle. It should be noted that intraoperative conversion of GVRT to conventional recession is easily accomplished if necessary, under topical anesthesia.

A significant advantage of GVRT is its technical simplicity and ease, making it suitable for topical anesthesia. Vessel sparing is often possible and may be very useful when surgery on multiple muscles simultaneously or sequentially surgeries is anticipated. Central partial rectus tenotomy has been performed transconjunctivally on an outpatient basis.[18] Advantages of central tenotomy have been postulated to include intact tendon poles, maintaining normal wide rectus insertions and comitant effects.[18,21] However, the incomitant effect of marginal GVRT could be exploited to correct incomitant strabismus.[19]

Management of diplopia due to acquired strabismus in SES includes protocols varying from prismatic spectacles for deviations ≤10Δ, as well as surgeries for which nomograms are now available.[7-9,22] Prismatic spectacles have disadvantages of considerable weight, chromatic aberration, and problematic cosmesis.[7-9] Surgeries for SES have been largely helpful to subjects but undercorrection may develop over time with recurrence of symptomatic diplopia, not because of failure of the surgery per se, but because of the progressive nature of the condition.[22] Of course, progressive connective tissue degeneration may also occur to require progressively greater prism power when nonsurgical management is employed. SES is one of the most common causes of binocular adult diplopia.[23] Demographic statistics in a predominant Caucasian population have shown it to be more common in women with CVS, contributing to about two-thirds cases in both men and women.[23]

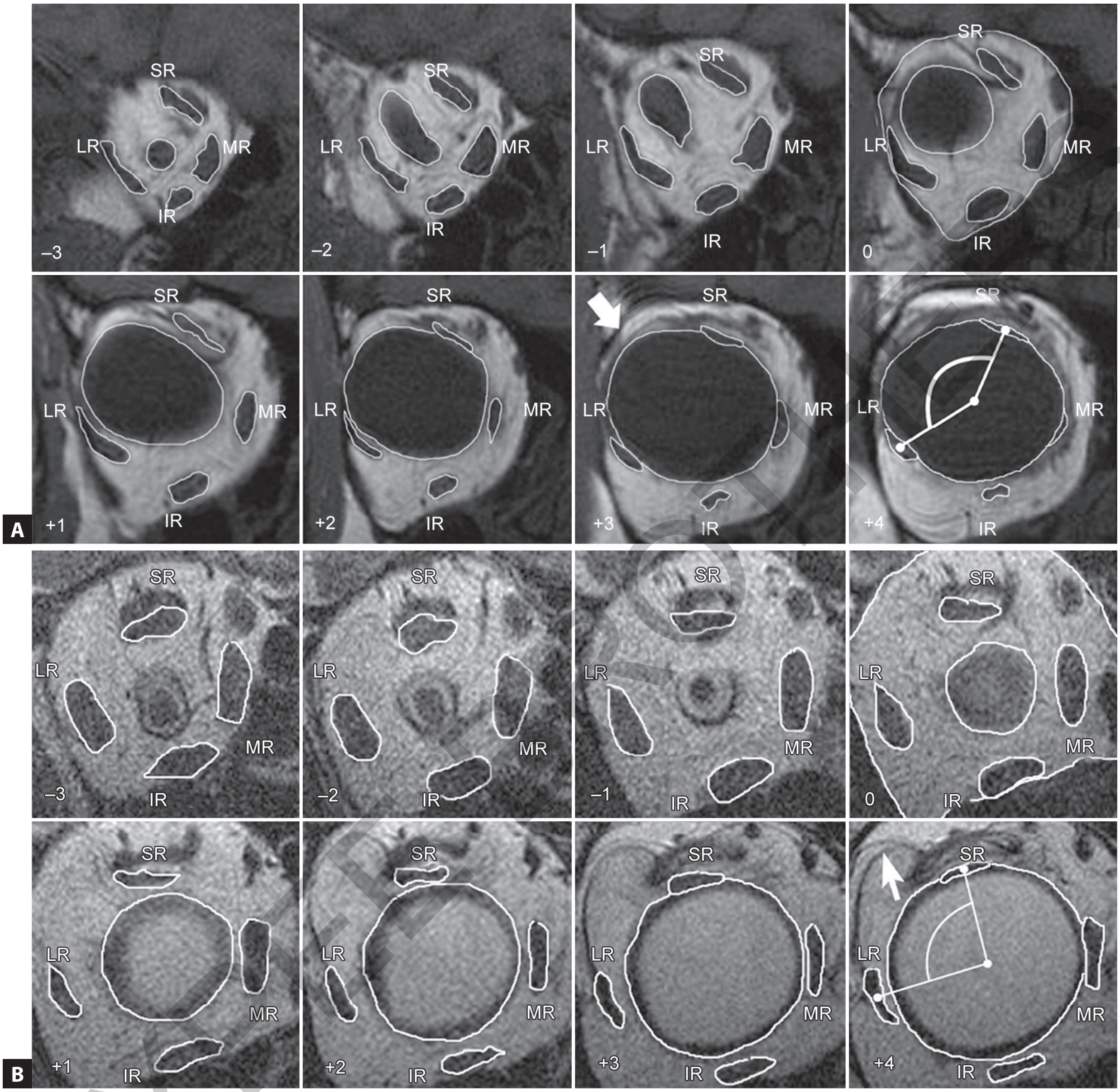
HEAVY EYE SYNDROME

Progressive hypotropia and esotropia with limited abduction and supraduction may occur in highly myopic individuals, classically termed heavy eye syndrome (HES).[24] The term refers to the voluminous, and implicitly "heavy", eyeball associated with high myopia. The condition may be unilateral. Again, MRI has been very useful in elucidating the possible pathophysiology of this condition.[25,26] Krzizok et al. demonstrated inferior displacement of the LR muscle in HES.[25,26] Yokoyama et al. noted superotemporal dislocation of the myopic globe in the orbit, with displacement of both the LR and SR muscle.[27] The existence of HES or myopic strabismus fixus, as it was called, has been long recognized.[24] SES was much later described as having similarities to HES, but in nonmyopic elderly eyes.[6-8] Posterior anastomosis of the LR and SR muscle bellies to shift the myopic globe back into the EOM rectus cone was proposed as treatment for HES, with MR and IR recession as occasionally necessary to augment the effect.[5-31]

Extraocular muscle paths are controlled by connective tissues of the orbital pulley system and progressive age-related connective tissue degeneration resulted in strabismus recurrences after suture repositioning of the LR muscle. Also, surgery in the already degenerated area can be disruptive of the remaining connective tissues in the vicinity of the LR muscle.[22] Surgeries performed in areas anatomically remote from the LR are more likely to be successful than imbrication because of less surgical manipulation of already degenerating connective tissues, and because these alternative surgeries can be performed using adjustable, techniques whereby intraoperative amelioration of diplopia is the endpoint.[7-9,22] This subjective endpoint also helped establish nomograms for the correction of small angle strabismus in SES.[7-9] **Figure 7A and B** demonstrate some of the characteristic features of SES and HES on MRI. **Table 3** presents features that distinguish the two entities.

In an initial study of SES, subjects with high axial myopia were deliberately excluded to avoid confounds.[8] However, the two conditions are not mutually exclusive and may coexist.[31] A MRI study on the anatomical relationship of the globe to the EOM in a highly myopic cohort, predisposed to both SES and HES demonstrated degeneration of the LR-SR band with inferior LR as well as lateral SR displacement in SES, while in HES, there was superotemporal globe shift with inferior displacement of LR and medial displacement of the SR. The mean angle between the SR, globe center, and LR was significantly larger in HES than in SES. No globe prolapse was observed in SES. The LR muscle was tightly opposed to the globe in HES but distracted away from it in SES **(Figs. 7A and B)**. Though clinical presentations may sometimes appear similar, it is relevant to understand these anatomical differences to choose the appropriate surgery.

The term "myopic myopathy (MM)" is an historical term that has often been used in relation to restrictive ocular motility disturbances associated with high myopia. Progressive neurogenic palsy, structural changes of EOM,



Figs. 7A and B: Quasi-coronal MRI at 2 mm intervals in central gaze, arranged from posterior at upper left to anterior at lower right. (A) Heavy eye syndrome (HES) with T1-weighted imaging of the right orbit. Superotemporal prolapse (white arrow) is prominent in the equatorial region (planes +1 to +4), leading to inferior displacement of the lateral rectus muscle and medial displacement of the superior rectus muscle. Note tight apposition of the LR muscle to the globe; (B) Sagging eye syndrome (SES) with T2-weighted imaging. Globe prolapse is absent but there is inferior sag of the lateral rectus muscle due to lateral rectus–superior rectus (LR-SR) band degeneration whose rupture site is indicated by the white arrow. Soft tissue including orbital fat (F) occupies the superotemporal quadrant under the LR-SR band. (IR: inferior rectus; MR: medial rectus)

Source: Reproduced with permission from Elsevier, March 2020. Tan RJD, Demer JL. Heavy eye syndrome versus sagging eye syndrome in high myopia. J AAPOS. 2015;19(6):500-6.

EOM paralysis, and EOM myositis have all been suggested as causes of MM. Bagolini et al. claimed that pressure of the large globe on the LR and its capillary bed resulted in a slightly increased cytoplasmic component of muscle fibers and moderate hypertrophy of the MR in such cases, resulting in esotropia.[32-34] Ruttum et al. have demonstrated flattening of the posteromedial wall of the globe against the medial orbital wall at abduction in a patient with MM.[34]

TABLE 3: Differences sagging eye syndrome and heavy eye syndrome.

Distinctive features	*Sagging eye syndrome*	*Heavy eye syndrome*
Demography	More common in elderly women and in Caucasian and Japanese population. Other populations require study	No age, gender or race predilection, except more common in east Asian population due to their high prevalence of high myopia
Pathophysiology	Age-related degeneration of connective tissues supporting the EOMs, especially the LR-SR band, which shifts rectus pulley position and actions. Rectus muscle elongation	Inferonasal displacement of the lateral rectus path, with relative superotemporal globe dislocation
EOM displacement	LR infraplacement unilaterally or bilaterally. There is often significant inferolateral displacement of the SR and MR muscles. The LR appears "fall away" from the globe (is not tight opposed to it). The LR-SR band is elongated or ruptured	LR infraplacement and globe dislocation and prolapse into the superotemporal quadrant of the orbit. The SR is usually displaced medially. The LR is tightly opposed to the globe. There is LR-SR band rupture due to the globe prolapse but is usually not visible because of the prolapsed globe
EOM elongation and bowing	This has proven to be one of the most characteristic features of sagging eye syndrome	EOM elongation and bowing are not emphasized but probably occur
Clinical features	Small angle esotropia and hypotropia associated with horizontal diplopia at distance and/or vertical diplopia associated with restriction supraduction. There is often a deep superior orbital sulcus with history of blepharoplasty. Myopia is common but not necessary to the diagnosis	High myopia with large angle progressive esotropia and hypotropia associated with significant restriction of abduction and supraduction. Sometimes the affected eye is in adduction, which accounts for the alternate name "myopic strabismus fixus"
Management	Prismatic glasses may be used. Corrective surgery with intraoperative adjustment in an area away from the area of pathology as by bilateral MR recession and GVRT with intraoperative adjustment	LR-SR muscle belly imbrication to relocate the prolapsed globe into the center of the orbit is most effective as it re-established the normal physiology of the globe-EOM system

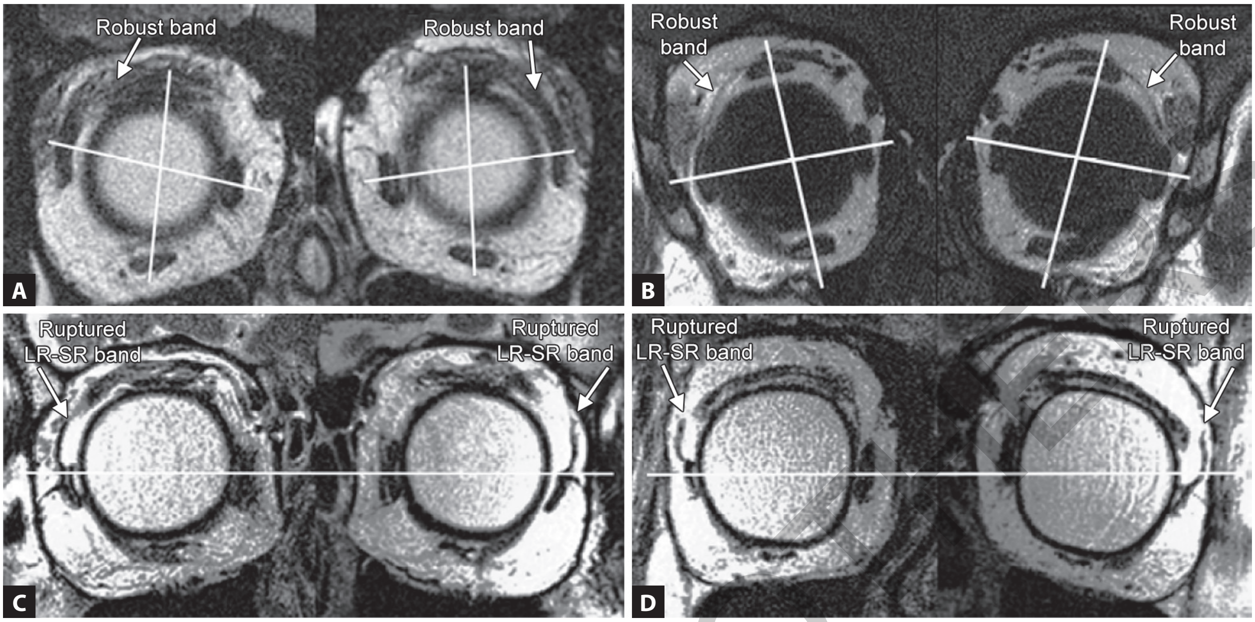
(EOM: extraocular muscle; GVRT: graded vertical rectus tenotomy; LR-SR: lateral rectus–superior rectus; MR: medial rectus)

All these hypothesis regarding the genesis of HES historically preceded clarification of the condition by MRI, and the translation of these findings into appropriate strabismus surgery.

CHILDHOOD-ONSET PULLEY HETEROTOPY

In craniofacial anomalies and congenital orbital dystopia, strabismus has been noted to directly correlate with the position of the EOMs in the orbit, as identified by MRI **(Figs. 8A to D)**.[35,36] More specifically, pulley heterotopy in Crouzon syndrome has been shown to correlate directly with A- and V-pattern strabismus.[37] Orbit 1.8 computational simulation suggests that extorsion of the horizontal and vertical rectus pulley array can reproduced the pattern strabismus observed in Crouzon syndrome. The high correlation between the predicted magnitude of the V-pattern exotropia and the actual observed exotropia in Crouzon syndrome suggests that extorsion of the rectus muscle pulley array primarily accounted for the pattern strabismus.[37] Knowledge of these pulley heterotopy and resultant malposition aids appropriate design of strabismus surgery. In this context, overelevation is adduction if often not due to over contraction of an IO muscle or SOP, but instead to rectus pulley heterotopy. Weakening the IO in this condition is hardly likely to completely ameliorate the incomitance.[38] Holistic evaluation of ocular motility along with observation of facial features including the slant of the palpebral fissures and other orbital and facial dystopias may yield clues to the diagnosis of the condition and should be a part of the workup of these subjects **(Figs. 9 and 10)**.

An interesting case series comprises a set of young children of 2–3 years age with acquired esotropia and minimal cycloplegic refractive error who presented with recurrence of esotropia with 5 PD of preoperative deviation after bilateral MR recession surgery.[39] Inferior displacement of the LR was seen in all these cases on resurgery. LR resection with superior equatorial myopexy stabilized the esotropia, possibly by normalizing the LR path and its direction of action.[39]



Figs. 8A to D: Quasi-coronal plane MRI. (A) Subject with A-pattern exotropia demonstrating robust lateral rectus–superior rectus (LR-SR) band bilaterally, with rectus pulley heterotopy. Lines connect centers of the horizontal and vertical rectus pairs at roughly the pulley locations, showing bilateral superior displacement of the lateral relative to the medial rectus pulley, and lateral displacement of the inferior relative to the superior rectus pulley; (B) Subject with V-pattern exotropia showing robust LR-SR band bilaterally, with rectus pulley heterotopy. Lines connect centers of the horizontal and vertical rectus pairs at roughly the locations of their pulleys, showing bilateral inferior displacement of the lateral relative to the medial rectus pulley, and medial displacement of the inferior relative to the superior rectus pulley; (C) Subject with adult-onset divergence paralysis esotropia. Note bilaterally symmetric infraplacement of the lateral rectus pulleys with bilateral discontinuity of the LR-SR band ligaments. Horizontal line indicates centers of the medial rectus muscles; (D) Subject with adult-onset cyclovertical strabismus causing left hypotropia. Note asymmetric infraplacement of the left lateral rectus pulley and bilateral discontinuity of the LR-SR band ligaments. Line indicates centers of the left horizontal rectus muscles.

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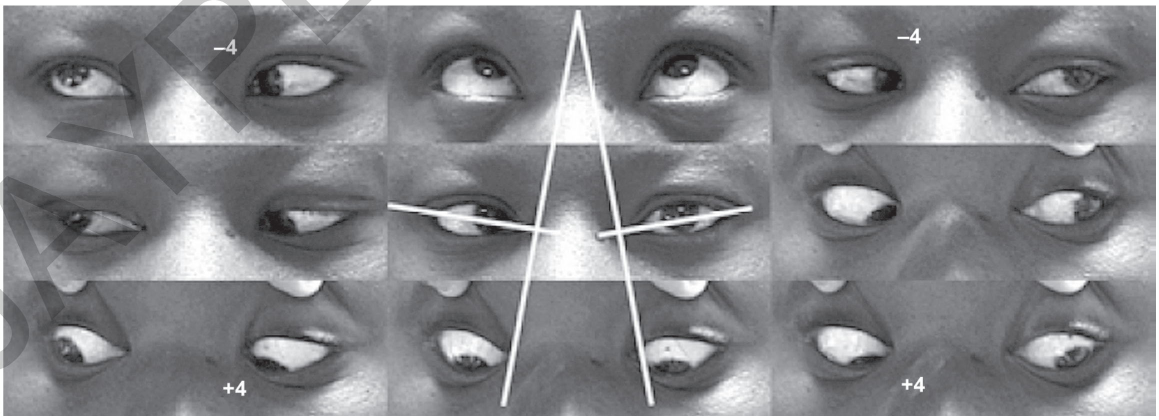

Fig. 9: Versions in subject with A-pattern exotropia demonstrate marked (+4) overdepression in adduction and marked (–4) underelevation in adduction. In central gaze panel, white lines join the medial and lateral canthi; perpendiculars added to these lines inscribe the letter A, correlating with the incomitance.

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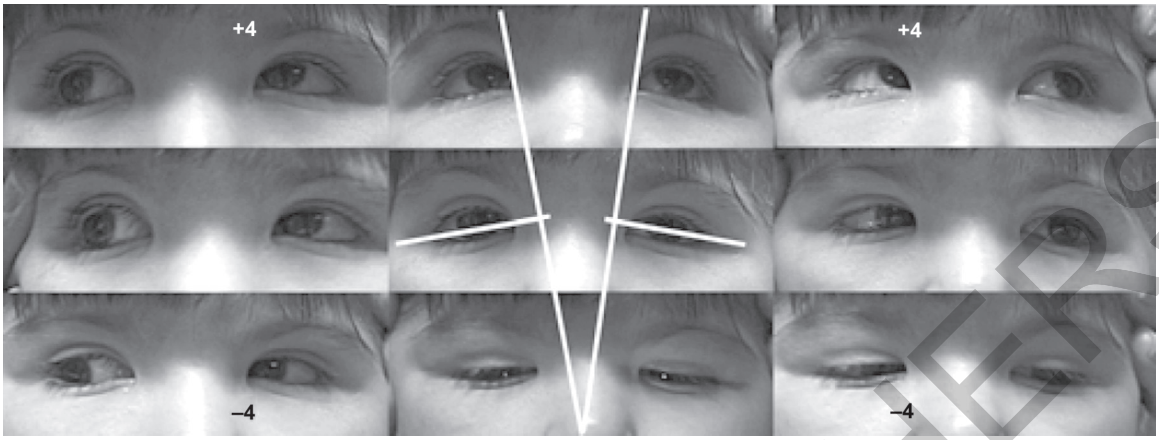

Fig. 10: Versions in subject with V-pattern exotropia demonstrate marked (–4) underdepression in adduction and marked (+4) overelevation in adduction. In central gaze panel, white lines join the medial and lateral canthi; perpendiculars added to these lines inscribe the letter V, correlating with the incomitance.
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CONCLUSION

The anatomy of the EOM pulleys relative to the orbit and globe often determine the way the EOMs act. Many types of ocular misalignment termed previously idiopathic, recalcitrant, complicated, recurrent, as well as strabismus not predictable responsive to conventional modalities of surgical management are good candidates for imaging. Appropriate evaluation of the structural and functional anomalies of EOM pulleys in these conditions will often aid objective interpretation of diagnosis and management of complex strabismus.[36]

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