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Long-term Surgical Outcomes in the Sagging Eye Syndrome

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Abstract

Background/aims—Strabismus due to sagging eye syndrome (SES) caused by age-related connective tissue involution is now an established cause of diplopia in older people. High suspicion of the condition results in early recognition, often obviating extensive neurological investigations and enabling surgical correction of the strabismus.

Methods—This retrospective study reviewed surgical results in 93 patients (40 males) of average age 68 ± 12 years, who had small-angle strabismus due to SES, during the 20-year period 1994–2014.

Results—In central gaze, mean distance pre-operative esotropia was 4.2 ± 7.5 (mean, SD), while mean hypertropia was 4.7 ± 5.9 . Surgeries comprised medial rectus (MR) recession; lateral rectus (LR) resection; plication, imbrication, and superior transposition of the LR to the superior rectus (SR) for esotropia; and graded vertical rectus tenotomy (GVRT) or vertical muscle recession for hypertropia. Mean post-operative immediate and long-term deviation after 316 $\pm$ 265 months average follow-up was 0.2 ± 1.2 and 1.1 ± 2.7 esotropia (both distance), respectively, and 0.00 and 1.1 ± 2.7 , respectively, for hypertropia. Strabismus recurred in 19 cases.

Conclusions—Progressive connective tissue involution in SES may occasionally result in symptomatic recurrences of the small-angle strabismus in about 20% of patients, irrespective of surgical procedures performed, possibly because of progressive involutional changes. This risk should be disclosed pre-operatively.

Keywords

Diplopia; sagging eye syndrome; strabismus

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Declaration of Interest

The authors report no conflicts of interest.

Introduction

The sagging eye syndrome (SES), comprising both age-related distance esotropia (ARDE) and cyclovertical strabismus (CVS), is now considered an established cause of strabismus in the elderly, arising largely due to degeneration of the connective tissues supporting the extraocular muscles (EOMs).¹ Along with secondary anomalies in rectus EOM pulley positions, rectus EOM muscle elongation also occurs in SES,¹ but has not been observed in other types of strabismus.² These changes have implications for strabismus surgical planning in SES.^{1,3} The lateral rectus (LR) muscle is most vulnerable to changes in its position within the orbit due to these age related changes.^{1,4} The LR exhibits the greatest average elongation (40%) of all EOMs affected by SES, but dehiscence of the LR–SR band, the primary connective tissue ligament supporting the LR path, results in its eponymous “sag” and the associated signs of SES.¹ Since SES is due to connective tissue degeneration with aging, and since these changes could progress with further passage of time, it seems possible that after surgical correction, further connective tissue degeneration might eventually reinstate strabismus in some patients.^{1,3}

Mindful of this ominous prospect, the present study was conducted retrospectively to evaluate outcomes of surgeries performed for patients with SES during the 20-year interval from 1994 to 2014. The surgeries that were performed were considered to be the appropriate in their era and were not designed for comparative trial; hence, surgical heterogeneity is inevitable. However, this aids understanding of the evolution of both, the etiopathogenesis of the condition as well as differing management strategies over a period of two decades. Surgeries comprised equatorial myopexy of LR muscles to their correct positions, 10 mm behind its insertion with imbrication of the LR muscle to the superior rectus (SR) muscle, bilateral LR resection or plication, bilateral medial rectus (MR) recession, vertical EOM recessions, and graded vertical rectus partial tenotomy (GVRT).^{1,3,5,6}

Methods

With Institutional Review Board approval, adhering to tenets of the Declaration of Helsinki and compliant to the USA Health Insurance Privacy and Portability Act (HIPAA), retrospective analysis was performed of consecutive patients with SES who were operated for comitant or incomitant small-angle horizontal or vertical strabismus between 1994 and 2014 at Stein Eye Institute, University of California, Los Angeles, USA, by a single surgeon (JLD). Currently, the diagnosis of SES is supported by clinical findings of adnexal laxity, blepharoptosis, limited supraduction, and greater excycloposition in the hypotropic than hypertropic eye. In the absence of associated neurologic signs and symptoms, these findings are considered sufficient to establish the diagnosis of SES.¹ In cases where other causes of hypertropia were clinically plausible or concurrent pathologies were suspected; the existence of SES is confirmed by high-resolution, surface coil magnetic resonance imaging (MRI) for demonstration of the characteristic inferior displacement of the LR muscle in the hypotropic than hypertropic orbit.¹ However, subjects included in this retrospective study were labeled as cases of “small angle” vertical or horizontal strabismus for a period of 20 years; much of this interval occurred before recognition of the cause of SES. All of these patients had undergone MRI. While evaluating them retrospectively, many cases were re-diagnosed as

having SES from clinical records, and re-interpretation of orbital MRI. Other clinical data that were abstracted included best-corrected visual acuity, refractive error, stereopsis (Titmus Fly), motility examination including alternate cover prism testing, Hess screen testing, fundoscopic evaluation, and clinical evaluation of saccades. Heterotropia was measured at distance and near-by alternate prism and cover testing. Fundus torsion was objectively determined by slit-lamp measurement of angle between the fovea and center of the optic disc.⁷⁻⁹

A total of 103 cases of SES were identified: of these 93 cases were operated for symptomatic small-angle strabismus and 10 cases declined surgery in favor of spectacle prisms. The operated cohort comprised 40 males and 53 females of average 68 ± 12 years (mean, SD). Surgeries performed were MR recession (28 cases)³; LR resection (8 cases)³; LR plication (1 case)¹⁰; imbrication and superior transposition and myopexy of the LR to the SR (6 cases); GVRT (41 cases)⁵; and vertical rectus muscle recession (9 cases).⁵ The average distance esotropia was 4.2 ± 7.5 (the near horizontal deviation being close to 0), while mean hypertropia with the higher eye fixing was 4.7 ± 5.9 .

It is relevant to emphasize that the understanding of the neuro-anatomical basis of SES, based on MR imaging, has been relatively recent.¹ These patients were previously recorded as having small-angle incomitant or comitant horizontal or vertical deviation, variably termed as divergence paralysis esotropia, CVS, heavy eye syndrome without myopia, etc., in the absence of any pattern of strabismus conforming to a neurological or restrictive cause.^{3,4,6} While the imaging analysis of the condition was ongoing that ultimately leads to the establishment of SES as a distinct clinical entity, the patients were treated as per their clinical findings, surgically or non-surgically resulting in inevitable heterogeneity of surgical procedures. After 2014, and the establishment of a definitive evidence-based clinical criteria for SES as well as associated clinical features that should arouse a high index of clinical suspicion of the condition, especially if in the geriatric age group, MRI of the orbits is not considered mandatory in SES.¹

Patients were considered eligible for surgery under topical anesthesia if, in the office after topical application of 0.5% proparacaine hydrochloride, they tolerated moderate preoperative pressure over the bulbar conjunctiva by a cotton swab-stick. An anesthesiologist monitored surgery, and upon surgeon request administered minimal doses of propofol or alfentanil so that patients remained alert and cooperative during intra-operative adjustment.¹¹⁻¹³ Recession of the MR and GVRT were performed under 1% lidocaine topical anesthesia, with intra-operative adjustment, as published,^{3,5} immediately correcting diplopia because alternate cover testing was performed in diagnostic positions in the operating room with the patient seated upright wearing the required refractive correction. If diplopia persisted following initial surgical dose, further 5–10% increments of GVRT up to a maximum of 90% and further MR recession were performed with interval intraoperative alignment monitoring by cover testing. Ciliary vessels were mobilized by blunt dissection and spared in selected cases.

Strabismus recurrence was defined as development of symptomatic diplopia, persistent at 3 or more months after the last strabismus surgery. Considering the relatively small pre-

operative deviation, no specific objective value was as a cutoff point for determining recurrence. Percentages of success were evaluated using Students *t*-test with a 0.05 level of statistical significance.

Results

It is notable that 84 of the 103 cases had employed prismatic correction prior to surgery. The subjects had diplopia for an average of $1,602 \pm 223$ days (approximately 4.5 years). The average prismatic correction used for esotropia was 4.4 ± 2.0 , and was 3.0 ± 2.0 for hypertropia. Mean stereopsis in these 84 subjects was 374 ± 712 arcsec (Titmus fly). Ten subjects refused surgery and preferred to retain spectacle prisms. Table 1 elucidates surgical procedures performed on the remaining 93 subjects. It is impossible to re-examine some of these subjects who were already in the eighth and nine decades of life when operated, as they have not followed up for various reasons, including death.

Table 2 elucidates the follow-up data of the immediate and late post-operative strabismus deviation in the 87 patients (besides the six patients who had undergone LR imbrication with superior transposition) who were followed post-operatively for almost one year (317 ± 265 days). Of these, 6/37 (16%) with ARDE and 9/50 (18%) with CVS had symptomatic recurrence of diplopia. The average distance horizontal deviation in these six patients with recurrence of distance esotropia was 10.7 ± 7.5 while the average vertical deviation in nine patients with vertical deviation was 5.1 ± 4 , This implies that both horizontal and vertical components of SES were equally prone to recurrences. Of the remaining 72 patients with orthotropia at last follow-up, 37 demonstrated stereopsis of 77 ± 67 arc seconds.

Except for imbrication of the LR muscle to the SR muscle combined with superior LR transposition, all other procedures had a recurrence rates between 14 and 25%. Imbrication of LR to the SR was performed in the earliest cases identified as being SES in the 1990s when the etiology of this form of strabismus was obscure. The average age of these six subjects in whom imbrication was performed was older than the overall average at 78 ± 8 years; five were women, three of them with a previous history of blepharoplasty. The average pre-operative distance esotropia was 12 ± 8 and hypertropia was 7 ± 6 , respectively. Four of these six subjects had symptomatic esotropia of 5.7 ± 4.8 at 120 ± 111 days (approximately 4 months) after surgery. As initial deviation in these six cases was less than 10 , all these patients had been treated by spectacle prisms for an average of about 5 years before the prisms were became ineffective in controlling diplopia due to gradually or suddenly increasing esotropia. One of these patients, in fact, reported the classical history of sudden pain over the supra-orbital area followed by onset of sudden diplopia probably marking acute rupture of the LR–SR band. This progression prior to surgery suggests the progressive nature of the condition, as now understood. All of these patients underwent extensive but non-revealing neurological investigations.

Table 3 elucidates the subsequent management in patients with SES who developed recurrences of symptomatic diplopia after their first surgery. The average age of the 15 patients with recurrences was 72 ± 7.5 years (five males), significantly higher than the 72 patients who maintained orthotropia at 66 ± 12 years ($p = 0.02$). Four of the 10 female

patients in this group had previously undergone blepharoplasty, but this prevalence did not significantly differ from patients without recurrences. However, the average follow-up period of subjects with recurrence was 635 ± 111 days, significantly longer than the period of 229 ± 183 days ($p < 0.05$) patients without recurrence. None of the subject with recurrences underwent post-operative MRI, but 11 of them underwent further strabismus surgery with good results. In all reoperated cases, intra-operative adjustment was performed with excellent immediate intra-operative alignment. Four patients refused surgery and were managed with prisms.

Discussion

Diplopia due to acquired strabismus in SES can be difficult to manage. Traditionally, prismatic spectacles have been preferred for deviations $\leq 10^\circ$ because recessions and resections are commonly expected to overcorrect the strabismus. However, prismatic spectacles have disadvantages including weight, chromatic aberration, and problematic cosmesis.^{1,3} One of the early fears deterring surgery for patients with small-angle strabismus was the absence of routine dose tables for angles $\leq 10^\circ$. It is interesting to note that nomograms were developed for surgeries on patients with SES with intra-operative adjustment and topical anesthesia.^{1,3,5} This approach has proven useful in providing precise alignment ameliorating diplopia in the immediate post-operative period while avoiding risks of general anesthesia in elderly patients.

It is probable that recurrence of post-operative diplopia in patients with SES was due to progression of the age-related dehiscence of orbital connective tissue, rather than surgical overcorrection or undercorrection, as surgical error would have been immediately evident in the post-operative period. In fact, there were no cases of surgical overcorrection, only undercorrection, manifesting as symptomatic diplopia over a period of time. Another important observation was that the patients in whom strabismus recurred were older than the group without recurrence. Moreover, the older patients also had a longer post-operative follow-up in this series. All of these suggest that the recurrences were due to progression of the orbital connective tissue degeneration rather than the surgery itself. Additional surgeries, all under topical anesthesia with intra-operative adjustment, succeeded in ameliorating the diplopia in patients who consented for it.

While LR superior transposition with imbrication to the SR theoretically should be the ideal physiological surgical corrective measure for this condition in light of its etiology as LR–SR band degeneration causing infradisplacement of the LR muscle, this powerful operation is not readily adjustable and may be subject to overcorrection. There have been only a few reports of this procedure's effectiveness in SES.¹⁴ Paths of the EOMs are controlled by connective tissues of the orbital pulley system, and progressive connective tissue degeneration might cause strabismus recurrences even after suture repositioning of the LR muscle, which is relatively disruptive of remaining connective tissues in the vicinity of the LR. In the current series, surgeries performed in areas anatomically remote from the LR had better success rates than imbrication, perhaps because of less surgical manipulation of already degenerating connective tissues, or perhaps because alternative surgeries are readily adjustable.

It may be concluded that surgical repair provides long-term relief of diplopia in the great majority of patients with SES, and at least a substantial interval of relief for the rest. Strabismus surgery can readily and safely be repeated in the event of strabismus recurrence. These likely outcomes should be disclosed to patients as part of pre-operative informed consent in order to maintain realistic expectations.

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Table 1

Surgical procedures performed for sagging eye syndrome.

Surgical procedure	Surgeries performed	Recurrences	Percentage recurrence
MR recession	28	4	14
LR resection	8	2	25
LR plication	1	0	0
Imbrication of LR to SR with superior LR transposition	6	4	67
GVRT	41	7	17
Vertical muscle recession	9	2	22
Total	93	19	20

Table 2

Pre- and post-operative strabismus after surgery for SES.

	Pre-operative deviation	Early post- operative deviation	Final post-operative strabismus (317 days)
Distance Esotropia	4.2 ± 7.5	0.4 ± 2.4	1.2 ± 3.6
Hypotropia	4.7 ± 5.9	0.2 ± 0.9	1.1 ± 2.1

* ($p < 0.05$) for all differences between the pre-operative deviation, early post-operative, and late post-operative deviations for both distance esotropia and hypotropia.

Table 3

Surgeries for strabismus recurrences in SES.

Initial surgical procedure	Number of recurrences	Subsequent procedures
MR recession	4	(1) LR resection in 2 cases (2) LR plication in 1 case (3) Prism management in 1 case
LR resection	2	MR recession in 2 cases
GVRT	7	(1) Contralateral SR GVRT in 4 cases (2) Ipsilateral IR in 1 case (3) Prism management in 2 cases
Vertical rectus recession	2	(1) Ipsilateral SR plication in 1 case (2) Prism management in 1 case