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Surgical treatment of myopic strabismus fixus: a graded approach

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Abstract
Background Surgical treatment of myopic strabismus fixus is challenging. Options for its correction range from conventional combined recession-resection surgery to innovative surgical procedures aiming to correct the deviated muscle paths. In this report we review our experience and compare the results of various surgical options for treatment of strabismus fixus.
Methods We report the surgical outcomes of nine adults with acquired strabismus fixus due to myopia with a follow-up of 1 year. Patients were enrolled between May 2003 and April 2007 in this retrospective study. The surgical procedure was determined depending on the angle of deviation and extent of motility impairment. A new transposition technique was performed in one patient who had an extreme variant of strabismus fixus.
Results Combined recession-resection surgery was performed in four patients with resulting small-angle esotropia. In patients with both esotropia and hypotropia due to muscle alignment, we performed an additional upward displacement of both horizontal recti muscles combined with a myopexy of the lateral rectus muscle. The results were satisfying; in particular in one patient who had a transposition procedure a significant improvement was achieved.
Conclusions For treatment of myopic strabismus fixus, a graded approach seems advisable. Combined recession-resection surgery yields good results for smaller deviations with mildly impaired motility, additional fixation techniques need to be applied once the horizontal muscle paths are deviated, and in extreme cases, a transposition procedure is required.

Keywords Convergent strabismus fixus · Acquired progressive esotropia · High myopia · Combined recession-resection surgery

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Introduction

Convergent strabismus fixus is a rare ocular motor abnormality in which the affected eye is more or less fixed in extreme adduction [1]. In the involved eye, the predominantly affected medial rectus muscle (MR) behaves like a fibrous band causing the eye to turn inwards [2, 3]. Different causes of acquired convergent strabismus fixus have been reported such as intraocular tumor or amyloidosis [4, 5]. Occurrence secondary to abducens nerve palsy has also been suggested [6]. More frequently convergent strabismus fixus is associated with high myopia [2, 7–12]. This condition was first described by Hugonnier and Magnard, who emphasized the disturbed ocular movements [7]. Patients with unilateral or bilateral high myopia tend to develop esotropia with limited abduction which is often accompanied or followed by hypotropia and restricted elevation in the involved eye. Typically this type of strabismus is progressive, with a gradual increase in deviation and restriction. Vertical deviation is very likely to occur in the later stages, coincident with development of displacement of the extraocular muscle paths [11, 13]. The hypotropic component has been termed "heavy eye phenomenon" [14].

Surgical treatment of myopic strabismus fixus is complex and the palette of surgical options to choose from is considerable. Conventional maneuvers are resection-reces-

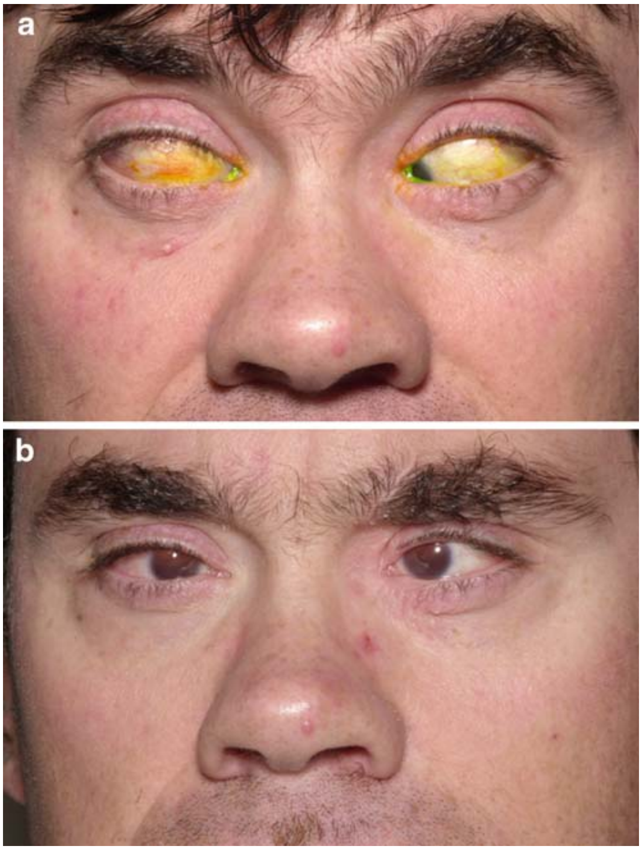

Fig. 1 **a** Patient No. 8 with an extreme form of strabismus fixus leading to functional blindness. **b** Postoperatively, a middle-angle esotropia (20°) resulted after a muscle transposition surgery (see text for details)

sion procedures that mainly affect muscle forces [2, 6, 8, 9, 13, 15]. Modified procedures include transposition techniques that alter the muscle paths [15–22].

We report the surgical outcomes of nine adults with acquired strabismus fixus due to myopia with a follow-up of 1 year. The surgery performed was adjusted individually and included a new transposition procedure in one of the patients.

Material and methods

In a retrospective study, nine consecutive patients with myopic strabismus fixus were included. Patients (seven females, two males) were enrolled between May 2003 and April 2007; their age ranged from 18 to 78 years (mean: 55.2 years). All patients were referred for strabismus correction procedure. All patients had routine ophthalmological and orthoptic examinations. Deviations were measured with the prism cover test at 5 m and at 0.33 m or with the Krimsky method. History was obtained from each patient, and the underlying pathology was supposed to be unilateral or bilateral myopia. A- and B-scan echography was performed in 18 eyes to determine their axial length. Either photographs or history (or both) were used to ascertain that the angle of deviation in each patient had not been present since childhood. Magnetic resonance imaging (MRI) was performed preoperatively in seven patients to exclude other causes of motility restriction and to search for displacement of muscle paths. MRI was not considered in two patients because of the clear clinical picture. Fresnel prisms on spectacles were used whenever possible to simulate the desired surgical outcome and to evaluate the risk of de novo diplopia. The surgical procedure was determined depending on the amount of esotropia and extent of motility impairment. Motility impairment was divided into four grades (-1 to -4) [20, 22]. Procedures for correcting the muscle paths were considered in cases presenting with additional hypotropia and displaced lateral rectus muscle (LR). A novel transposition technique was performed in a patient who presented with an extreme variant of strabismus fixus. This patient (No. 8) is described separately below. All patients were examined on the first postoperative day, 3 months and 1 year after surgery. Postoperative results (angle of deviation, motility) were taken from the 1-year follow-up visit.

Report of patient No. 8

A 38-year-old autistic man was referred from a home for disabled people with both eyes fixed in an extreme adduction and depression, which had led to functional blindness. The cornea was completely hidden in the right eye and only 2 mm of the superotemporal cornea was detectable in the left eye (Fig. 1a). On extreme left gaze, the left cornea could be exposed by 1 more millimeter, but visual axis remained obstructed. The patient could hardly perceive light.

Previous records revealed a diagnosis of bilateral congenital aniridia, glaucoma, and cataract. On earlier photographs, large-angle esotropia was seen, but both corneas had still been visible 12 years previously. Visual acuity could not be measured reliably in the past, but the patient could navigate freely. Over time, the right eye was the first to "disappear" and during the last 2 years the left eye followed this course, becoming anchored in depression and adduction. Echography of both eyes revealed significant axial myopia of 30 mm in the right eye and 32 mm in the left eye. MRI demonstrated herniation of the enlarged globe superotemporally through the muscle cone (Fig. 2). Displacement of the extraocular muscles was noted with the LR deviated inferiorly and the superior rectus muscle (SR) shifted nasally. Surgery was performed in the left eye first in order to restore vision in the slightly less affected eye. A hemitransposition of the SR and the LR combined with an

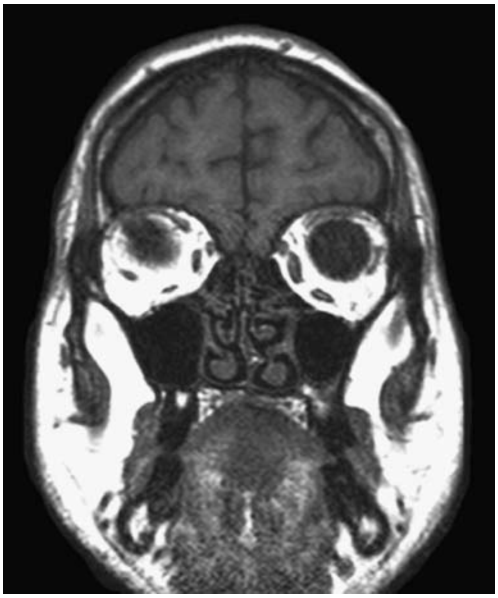

Fig. 2 Coronal MRI after muscle transposition surgery in the left eye of patient 8. In the unoperated right eye, herniation of the enlarged globe superotemporally through the muscle cone is demonstrated. Displacement of the LR inferiorly and the SR nasally is clearly visible. In the operated left eye, normalization of the muscle paths and repositioning of the globe has occurred

MR recession of 10 mm was performed. During surgery, the insertions of these muscles were found to be located as expected. Both the SR and the LR were longitudinally divided in half for approximately 15 mm from their insertions. A new insertion for the lateral half of the SR and the superior part of the LR was created between the SR and the LR, 7 mm posterior from the limbus as described by Yamada et al. [18]. Additionally, the translocated muscle halves were tied together and secured to the superotemporal sclera in 15 mm distance from the new insertion. The same procedure was performed on the right eye 3 months later.

Results

The pre- and postoperative data for each patient are summarized in Table 1.

Clinical features

All nine patients had acquired progressive esotropia that was not noticed in childhood. However, the exact age of onset could not be determined. Axial length was measured in all eyes and revealed an average of 29.6 mm. The average length for the affected, and therefore operated, eyes (patient 8 had bilateral surgery) was 30.6 mm. Four patients had an almost equal degree of high myopia in both eyes (≤1.5 mm difference) while five patients had an anisometropia (>1.5 mm). Eight patients had bilateral high myopia and one patient (No. 2) had a severe anisometropia with an unaffected fellow eye. MRI demonstrated the enormous axial elongation of the globes and displacement of the muscle paths in five patients. Typical findings were an inferiorly deviated LR and a posterior globe partially dislocated into the superotemporal orbital quadrant. In patient No. 8, MRI showed a staphylomatous globe with downward displacement of the LR and nasal displacement of the SR bilaterally. Both globes were herniated superotemporally and retroequatorially through the muscle cone. The prism adaption test was applied in seven patients (No. 2–7 and No. 9) preoperatively. None of them experienced diplopia.

Surgical results

Before surgery, all patients had large-angle esotropia and marked limitation of abduction (-2 or worse) in the affected eyes. Vertical deviation was noted in six patients with corresponding limited elevation (-2 or worse). None of the patients complained of diplopia preoperatively. Only one patient (No. 4) had temporary diplopia after surgery, which resolved by 2 weeks postoperatively.

Three patients with either purely horizontal (No. 2 and 4) or predominantly horizontal deviation (No. 7) had conventional combined recession-resection surgery in the affected eyes. Insertions and muscle paths were intraoperatively found to be normal, which was confirmed by MRI in one patient (No. 2). The extent of MR recession and LR resection in mm was higher than in eyes with concomitant esotropia (overall surgical distance of 17 mm in patient No. 2, 15 mm in patient No. 4 and 12.5 mm in patient No. 7), but extreme values were not applied because of the moderate degree of deviation and moderate limitation of abduction. Results were excellent in all three patients with only a small-angle esotropia remaining (reduction of esotropia between 14° and 32° with a remaining esotropia of 5–6°). No limitation of abduction was subsequently detectable and no adduction deficit occurred. Patient No. 9 was a rather special case because of accompanying hypotropia without muscle displacement. This could not be demonstrated in MRI or intraoperatively. Therefore, a conventional inferior rectus muscle (IR) recession was performed in addition to another MR recession after an initial recession-resection surgery. The result was good, with only a small-angle esotropia and hypotropia postoperatively. Minor limitation of abduction remained.

In four patients (No. 1, 3, 5, 6) with severe preoperative limitation of both abduction and elevation (abduction -3 or worse, elevation -3), a recession-resection procedure was combined with readjustment of the lateral muscle path. The insertion site of the horizontal recti muscles was found to

Table 1 Clinical characteristics of patients

Patient	Age at surgery (years)	Axial length of the globes (mm)		Deviation degrees (°) Esotropia (ET), Hypotropia (HT) (prism cover test or Krimsky method) [if no eye is indicated, patients had an alternating squint]		Limitation of motility		MRI findings	Surgical procedure
				Before surgery	After surgery	Before surgery	After surgery		
1 *	55	30.2	29.1	OD: 60 ET, 15 HT OS: 60 ET, 12 HT	OD: 17 ET, 5 HT OS: unchanged	OD/OS: abduction (-4) elevation (-3)	OD: abduction (-2) elevation (-1) adduction (-1) OS: unchanged	OD/OS: inferiorly displaced LR	OD: MR recession 13 mm (hang back sutures), upwards 3 mm, LR resection 12 mm, upwards 6 mm and equatorial myopexia
2	46	28.1	28.8	38 ET	6 ET	OD/OS: abduction (-2)	OD/OS: non	OD/OS: no displacement	OD: MR recession 5 mm OS: MR recession 5 mm, LR resection 7 mm
3	62	32.0	24.3	OD: 35 ET, 14 HT	OD: 6 ET, 4 HT	OD: abduction (-4) elevation (-3)	OD: abduction (-1) elevation (-1)	OD: inferiorly displaced LR	OD: MR recession 6 mm, upwards 5 mm, LR resection 10 mm, upwards 5 mm and equatorial myopexia
4	66	28.7	28.1	32 ET	5 ET	OD: abduction (-2) OS: abduction (-1)	OD: non OS: unchanged	-(no LR displacement intraoperatively)	OD: MR recession 7 mm, LR resection 8 mm
5	72	31.0	31.3	OS: 55 ET, 10 HT	OS: 15 ET, 4 HT	OD: abduction (-1) OS: abduction (-3) elevation (-3)	OD: unchanged OS: abduction (-1) elevation (-1) adduction (-1)	OS: inferiorly displaced LR	OS: MR recession 10 mm, upwards 5 mm, LR resection 12 mm, upwards 5 mm and equatorial myopexia
6	78	32.1	34.3	OS: 38 ET, 10 HT	OS: 3 ET, 6 HT	OD: abduction (-1) OS: abduction (-3) elevation (-3)	OD: unchanged OS: abduction (-1) elevation (-2) adduction (-1)	OS: inferiorly displaced LR	OS: MR recession 9 mm, upwards 5 mm, LR resection 11 mm, upwards 5 mm and equatorial myopexia, inferior rectus partial (2/3) disinsertion
7	18	29.2	26.3	OD: 20 ET, 2 HT	OD: 6 ET, 3 HT	OD: abduction (-2)	OD: non	-(no LR displacement intraoperatively)	OD: MR recession 5.5 mm, LR resection 7 mm
8**	38	30.1	31.5	OD/OS: both eyes fixed in extreme adduction and depression	20 ET	OD/OS: abduction (-4) elevation (-4)	OD: abduction (-3) elevation (-1) adduction (-1) OS: abduction (-2) elevation (-1) adduction (-1)	OD/OS: inferiorly displaced LR, nasally displaced SR	OD/OS: MR recession 10 mm, hemitranspositions of SR and LR as described above (operation for the right eye was performed 3 months later)
9***	62	27.0	30.0	OS: 10 ET, 9 HT	OS: 4 ET, 3 HT	OS: abduction (-2) elevation (-2)	OS: abduction (-1)	OD/OS: no displacement	OS: further MR recession 4 mm, IR recession 6 mm

ET esotropia; *HT* hypotropia; *OD* oculus dexter; *OS* oculus sinister

*A suggested similar procedure on the left eye could not be performed because of cardiovascular reasons.

** This patient is presented separately as a case report.

*** This patient had been operated for a 15° left esotropia 6 years previously. Procedure was a recession-resection procedure for esotropia (MR recession 4 mm, LR resection 6 mm). Angle of deviation after first surgery was 1° left hypotropia at follow-up examination 3 months after surgery and 2° left esotropia and 2° left hypotropia after 1 year lie in the physiological meridian, but downward displacement of the muscle paths found intraoperatively was consistent with preoperatively determined MRI findings. During surgery, the inferiorly displaced and resected LR was shifted superiorly and fixed at the equator in the physiological meridian using non-resorbable sutures (modified Faden procedure). The recessed MR was also elevated, albeit without further scleral fixation, in an intent to increase the vertical correction. Results were satisfactory but not as good as in the three patients described above. Marked reduction of esotropia and improvement of motility was achieved (reduction of esotropia between 29 and 43° with a remaining esotropia of 3–17°, abduction deficit found postoperatively was -1 in three patients and -2 in one patient). Hypotropia was also reduced, although the success was less predictable: In three patients it was reduced considerably (6–11° with a remaining hypotropia of 3–5°), but in patient No. 6 the effect was insufficient (reduction of hypotropia of 2° only). A second procedure of additional IR weakening was declined by the patient. Elevation improved in all eyes (elevation deficit found postoperatively was -1 in three patients and -2 in one patient), but remained somewhat restricted. Slight limitation of adduction (-1) resulted in large MR recessions (>9 mm).

Patient No. 8, who is reported in detail, was treated using a novel technique of muscle transposition. The result was very good, considering the preoperative situation (Fig. 1b). An esotropia of 20°, a markedly improved, though still limited, abduction and elevation, and an acquired slightly limited adduction, probably due to the large MR recession (10 mm). Visual acuity improved from light perception to hand movements in 3 m in each eye.

No surgical complications occurred in any treated eye. Even though the sclera was thin in nearly all operated eyes due to high myopia, all sutures including those placed at the equator could be placed.

Discussion

Recent reports substantiate the theory that myopic strabismus fixus can be interpreted as a progressive condition with a gradual increase in deviation and restriction. Vertical deviation is very likely to occur in the later stages, coincident with development of displacement of the extraocular muscle paths. Intraoperatively, Herzau and Ioannakis detected an abnormal LR path, although neither CT nor MRI had suggested this observation in their patients [16]. In contrast, based on CT and MRI findings, Kolling demonstrated medialwards transposed SR and IR in esotropia, and downwards transposed MR and LR in hypotropia [23]. Demer et al. recently took into account new aspects of the functional anatomy of the orbit, showing that the extraocular muscles pass through connective tissue sleeves in the posterior Tenon's fascia termed "pulleys" [24]. On the one hand they act as functional origins of the muscles and on the other hand they are supposed to constrain muscle paths during gaze shifts. Krzizok et al., interpreting MRI findings, postulated the downward displacement of the LR, reducing the abduction effect and leading not only to an esotropia but also to hypotropia, as major pathophysiological factor in myopic strabismus fixus [11, 17, 25]. The loss of abduction function of the LR is transferred to an unphysiological depression and excyclorotation force [26].

In general, there are two main surgical techniques for myopic strabismus fixus, which have a different approach. Conventional recession-resection surgery mainly alters muscle forces [2, 6, 8, 9, 13, 15, 24]. Innovative surgery procedures have been recently performed to correct deviated muscle paths [15–22], e.g., a normalized alignment of the muscle course was achieved by myopexy of the LR at the equator [16, 17] or by transposition surgery [15, 18–22].

Generally, our study confirmed both techniques as suitable procedures in the treatment of acquired myopic strabismus fixus. As described above, we performed both procedures with further modifications adapted individually to the specific scenarios. Excellent results were achieved by conventional recession-resection surgery. All patients had only a small-angle esotropia remaining (≤6°). Limitation of abduction was no longer detectable, and no adduction deficit occurred. The sizes of recessions carried out were larger than normally performed, but were not extreme; conventional-size MR recessions alone have not generally been felt to be adequate [6]. The success of this procedure might be explained generally in relation to conventional strabismus surgery for esotropia, where normal-size MR recessions alone or recession-resection surgery have frequently been shown to be adequate. This holds true since myopic strabismus is regarded as a progressive disease with its earlier stage not yet necessarily accompanied by any muscle path displacement [11]. Similar results have been reported by others; Krzizok et al. have shown conventional recession-surgery to be satisfactory in 20 patients [17]. Mohan et al. performed large MR recession, with or without LR resection, successfully in two patients with strabismus fixus [6]. Bagheri et al. successfully combined bilateral recession-resection surgery with bilateral medial conjunctival recessions and transitory traction sutures directed temporally [13].

Considering our group of six patients with large-angle esotropia (≥30°), who all had recession-resection procedures (partially in combination with a Faden operation), we have calculated a dosage-effect curve. We determined a mean corrected angle of deviation of 1.79°/mm muscle correction (range 1.7–1.9). This might help in assessing how to dose in cases of convergent deviation alone or in earlier stages in which transposition surgery is not yet required. These values are slightly lower than those reported by Krzizok et al. [17]. Considering their patients reported with an angle of squint of ≥30° (nine patients), they achieved a somewhat higher dosage-affect with 1 mm of muscle correction neutralizing 2.41°, but their range was larger (range 1.6–3.9) too. Another procedure affecting the horizontal deviation by altering the muscle forces was MR tenotomy reported by Bagolini et al. in two patients [8]. In addition, disinsertion and myectomy of 5 mm of each MR, combined with 8-mm resection and advancement of the LR has been suggested [2].

A different approach takes the muscle alignment into consideration. We performed recession-resection surgery in a modified manner in four patients. Because of the observed downward displacement of the LR (seen in MRI and confirmed intraoperatively), we particularly paid attention to the vertical deviation. We performed recession-resection surgery combined with superior displacement of both horizontal recti muscles. Repositioning of the LR was secured by an equatorial myopexy with a nonresorbable suture. In one case we additionally performed a partial two-thirds disinsertion of the IR. In the first three cases (patients No. 1, 3, and 5), results were promising, neutralizing 6–11° of vertical deviation, even though the range was large. Surprising was the remaining 6° of hypotropia postoperatively in the patient in whom an additional IR disinsertion had been performed (patient No. 6); only 4° of hypotropia had thus been eliminated. Thus, we conclude that the results regarding correction of hypotropia are less predictable. Fortunately, all patients treated with modified Faden procedure showed marked motility improvement. Comparing our results to the seven patients treated with a "guide pulley" from Krzizok et al. confirms the limited predictability of this procedure [17]. They performed recession-resection surgery and LR myopexy without routine vertical displacement of the horizontal recti muscles. Herzau and Ioannakis achieved satisfying results in six patients with a similar procedure, shifting the horizontal recti upwards (myopexy of the LR in four cases), but noticed themselves that there was no clear dosage-effect curve [16].

The patient presented here as a case report had a new modification of transposition surgery on both eyes: In addition to a large MR recession (10 mm), the neighboring muscle halves of LR and SR were abutted one to another in the superotemporal quadrant. A new insertion was built in the middle of the original insertions 7 mm posterior to the limbus. Additionally, the muscle halves were tied together posteriorly. In view of the enormous degree of deviation preoperatively the result may be considered very favorable. Despite the fact that marked limitation of abduction persisted a satisfying repositioning with 20° esotropia was achieved, finally enabling the patient to make use of his eyes again. Elevation deficit was remarkably reduced, but a new slight adduction deficit occurred bilaterally due to the large MR recession.

A successful outcome with surgical correction of muscle paths was also reported by others. Yamada et al. presented a transposition of half of the SR and half of the LR combined with a large recession of the MR in one patient [18]. Postoperatively, the patient was able to fixate in primary gaze, and ocular motility had markedly improved in all directions. Hayashi at al. performed a modified Jensen's procedure with transposition of the SR and the IR combined with MR recession in eight patients with severe limitation of abduction [15]. The percentage reduction rate for this procedure was 91.8–100%. A partial, superior half Jensen's procedure was described by Larsen and Gole [20]. At follow-up (after nine months), the patient had regained significant ability to abduct and elevate the eye. Loop myopexy of LR and SR was introduced by Yokoyama et al [21]. Eye movements of six treated patients had improved. Wong et al. successfully performed loop myopexy in two patients [22]. For one of these patients, an initial large-angle recession-resection surgery showed no effect. Loop myopexy was therefore performed as a second procedure, slinging the LR and SR together and tightening the loop with a silicon sleeve. This modification was suggested because the anterior ciliary circulation would be less compromised as the muscles are not cut or split and less strangulation is caused by a silicon loop than by a suture. Furthermore, this procedure can be easily reversed by cutting and removing the band.

For surgical treatment of myopic strabismus fixus, a graded approach seems advisable. The surgical concept should be based on deviation of squint, limitation of motility and anatomic situation. In patients who did not undergo an imaging procedure consideration of the anatomic situs found intraoperatively is mandatory for surgical strategy. In addition, suture mechanisms might have to be considered before planning the surgical procedure. Placing a suture into the thin sclera of high myopes should be regarded as a potential challenge. In particular this applies to those sutures placed at the equator as described in patient No. 8 but even applies to resection procedures in which considerable traction is exerted on the sclera. However, neither bulbus perforations nor any retinal complications were seen in our patients. Recession-resection surgery has shown to be effective in the earlier stages of strabismus fixus. Once muscular alignment has occurred a transposition procedure is required even though the effect seems less predictable. Transposition techniques alter the muscle paths thereby creating new directions of muscle force. A depressed and excyclorotated globe that is fixed in adduction due to the inferiorly displaced LR can be relieved by upwards orientated maneuvers. Thereby the impaired abduction function can partly be restored by a Faden procedure or real transpositions. In addition, narrowing or closure of the superotemporal locus minoris resistentiae can be achieved. Therefore, procedures like loop myopexy [21, 22], partial Jensen's procedures as described by Larsen and Gole [20] and our modification are particularly effective. Moreover, the created muscle cuff may be especially stable and therefore preventing recurrent posterior globe herniation.

Proprietary interest statement None

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