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Superior oblique tucks for apparent inferior oblique overaction and V-pattern strabismus associated with craniosynostosis

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Abstract

Introduction—Apparent inferior oblique overaction and apparent superior oblique underaction are common in strabismus associated with craniosynostosis, and in many cases are likely due to excyclotorsion of the globes, with the rectus muscles acting with oblique vectors. We present a patient with craniosynostosis who underwent bilateral superior oblique tucks to specifically address the excyclotorsion of the globes.

Methods—A 16 year old male with Saethre-Chotzen syndrome presented with apparent bilateral inferior oblique overaction and apparent bilateral superior oblique underaction. He had 30 prism diopter (PD) esotropia and 5 PD left hypertropia by simultaneous prism and cover test (SPCT) at distance and near fixation. Fundus exam revealed marked excyclotorsion of both globes. The superior oblique tendons were tucked 10 mm bilaterally, using a 6–0 mersilene suture and the medial rectus muscles were recessed 5.5 mm on an adjustable suture with a 10 mm inferior displacement. Surgical results were reviewed at 6 weeks and 2 years post-operatively.

Results—At 6 weeks and two-years postoperatively, apparent inferior oblique overaction, apparent superior oblique underaction, and V-pattern esotropia were markedly improved. At 2 years, SPCT measurements were 8 PD esotropia in primary position at distance and 4 PD esotropia and 3 PD left dissociated vertical deviation at near.

Conclusions—Bilateral superior oblique tucks are useful in addressing the excyclotorsion that leads to apparent inferior oblique overaction and V-pattern strabismus associated with craniosynostosis.

Keywords

craniosynostosis; excyclotorsion; superior oblique tuck; surgery; V-pattern

Introduction

Children with craniosynostosis syndromes commonly have strabismus.(Khan et al., 2003) One of the most challenging types of strabismus to address in such children is apparent inferior oblique overaction and apparent superior oblique overaction, associated with V-pattern horizontal strabismus.(Khan et al., 2003, Morax, 1984) Previously reported approaches include inferior oblique recession, myectomy, anteriorization, denervation and extirpation, and medial rectus infraplacement,(Coats et al., 2000) but results have been disappointing with each of these procedures.(Coats et al., 2000)

One etiological hypothesis for the constellation of ocular motility findings in some cases of craniosynostosis is excyclotorsion of each eye within each orbit, resulting in the rectus muscles having oblique vectors.(Cheng et al., 1993, Morax, 1984, Tan et al., 2005) The superior rectus produces an abduction force, and inferior rectus an adduction force, creating the V-pattern. The medial rectus is higher and lateral rectus lower, producing an upshoot in adduction (apparent inferior oblique overaction).(Guyton & Weingarten, 1994, Kushner, 1985, Weiss, 1966) Findings of excyclotorsion on fundus exam and rotation of the globes on coronal imaging support this hypothesis.(Cheng et al., 1993, Tan et al., 2005)

We performed strabismus surgery on a patient with craniosynostosis to specifically address the excyclotorsion, using superior oblique tucks at the time of horizontal rectus surgery, and we report our findings.

Materials and methods

Institutional Review Board approval was obtained and all data were collected in a manner compliant with the Health Insurance Portability and Accountability Act.

The patient was a 16-year old white male who presented with crossing of the left eye since birth. He had been diagnosed with Saethre-Chotzen syndrome as an infant. His past medical history was significant for cranial surgery and placement of a ventriculoperitoneal shunt for hydrocephalus as an infant, common in Saethre-Chotzen syndrome. His past ocular history was significant for left levator advancement at the age of 3 years. He had not undergone previous strabismus surgery.

On presentation, aged 16 years, his best corrected visual acuity was 20/25 right eye and 20/40 left eye, with myopic astigmatic correction, consistent with mild strabismic amblyopia in the left eye. Assessment of ductions and versions demonstrated apparent bilateral inferior oblique overaction (Figure 1) and apparent bilateral superior oblique underaction (Figure 1). He had a 30 prism diopter (PD) esotropia and 5 PD left hypertropia by simultaneous prism and cover test (SPCT) at distance and near fixation and no measurable stereoacuity by Preschool Randot and Frisby near tests, or by Frisby Davis Distance and Distance Randot distance tests(Hatt et al., 2008) His prism and alternate cover test (PACT) measurements demonstrated a V-pattern, with alternating hypertropia on lateral gazes (Table 1). Fundus examination revealed marked excyclotorsion of both globes (Figure 2), but consistent with infantile onset excyclotorsion, Double Maddox Rod testing revealed no sensory excyclotorsion. A computerized tomography (CT) scan of the orbits confirmed presence of the superior obliques and excyclotorsion of each eye resulting in oblique vectors of the rectus muscles (Figure 3).

Surgical procedure

The superior oblique tendons were tucked 10 mm bilaterally, using a 6-0 mersilene suture as described by Del Monte.(Del Monte, 1993) The medial rectus muscles were recessed 5.5 mm on an adjustable suture with a 10 mm inferior displacement, taking scleral bites 4 mm back from the level of the original insertion to maintain the depression. (Del Monte, 1993) At the time of adjustment, both medial rectus muscles were advanced 0.5mm to reduce the near exotropia and yielded a post adjustment alignment of 8 PD esotropia at distance and 2 PD exotropia at near.

Results

At 6 weeks the patient had marked improvement of his depression in adduction and reduction in his over-elevation in adduction (Table 1). There was a 6PD esotropia in primary

position at distance and a 1 PD XT, 2 PD left hypertropia at near by PACT (Table 1). On right gaze there was a residual left hypertropia, but there was no vertical deviation on left gaze and the V-pattern had been collapsed (Table 1).

At 2 years post-operatively, the patient maintained the marked improvement of depression in adduction and over-elevation in adduction (Figure 4). Angle of deviation by SPCT was 8 PD esotropia in primary position at distance and 4 PD esotropia and 3 PD left dissociated vertical deviation at near. PACT measurements are shown in Table 1: there was no measurable vertical deviation in primary position or on lateral gaze, and a slight recurrence of the V-pattern esotropia in downgaze which was asymptomatic. The patient continued to have no measurable stereopsis as expected in late correction of infantile strabismus. Double Maddox Rod testing revealed no sensory excyclotorsion, consistent with adaptation to the new torsional status of the eyes.

Discussion

We have described using bilateral superior oblique tucks to address the bilateral excyclotorsion of the globes resulting in apparent inferior oblique overaction, apparent superior oblique underaction and V-pattern, commonly associated with some types of craniosynostosis. In our single case, the results appear to be long lasting and provide proof of concept for this novel approach.

Saethre-Chotzen syndrome is characterized by coronal suture synostosis, low-set frontal hair line, ptosis, broad indented nasal bridge, external ear anomalies, and syndactyly. Saethre-Chotzen syndrome shares many features of the more common Crouzon, Apert, and Pfeiffer syndromes (Khan et al., 2003) which also have coronal suture synostosis resulting in excyclotorsion of the globes and orbits, leading to a V-pattern strabismus with apparent inferior oblique overaction and apparent superior oblique underaction. We therefore believe that our approach of directly addressing the excyclotorsion of the globes by using superior oblique tucks will have application to the spectrum of V-pattern strabismus encountered in many craniosynostosis syndromes.

Bilateral superior oblique strengthening procedures directly addresses excyclotorsion of the globes, repositioning the rectus muscles such that their vector forces are normalized. This approach may be preferable to weakening the inferior oblique, which has been reported to be disappointingly ineffective (Coats et al., 2000). Strengthening the superior obliques allows a far greater effect on excyclotorsion than weakening the inferior obliques. An alternative approach to superior oblique tucks would be advancement of the anterior fibers of the superior oblique, as performed in the Fells modification of the Harada Ito procedure (Fells, 1974) (Case reported by M. A. Del Monte MD, Pediatric Ophthalmology sub-specialty day, American Academy of Ophthalmology, Las Vegas, NV, Nov. 2006, subsequent to the date of surgery for the current case report).

The V-pattern strabismus of craniosynostosis syndromes may have analogies to the V-pattern strabismus of more common infantile strabismus. Guyton and Weingarten (Guyton & Weingarten, 1994) described the concept of “sensory torsion” as a cause of A and V patterns, where torsion of the globe results in changes in the vectors of the rectus muscles leading to elevation in adduction and apparent inferior oblique overaction. Simonsz (Simonsz, 1990) and Liesch and Simonsz (Liesch & Simonsz, 1993) described the complex of latent upshoot in adduction, V-pattern, and anatomical excyclotorsion as a common cause of apparent inferior oblique overaction. Abnormal vectors of the rectus muscles causing apparent inferior oblique overaction has also been associated with abnormal pulley position (Clark et al., 1998), but whether or not pulley heterotopia is primary or

secondary remains unclear. We suggest that bilateral superior oblique strengthening surgery, inducing incyclotorsion and changing the vectors of the rectus muscles, may also have a role in V-pattern strabismus associated with infantile esotropia. Advancing the superior oblique tendons in patients with childhood V-pattern esotropia has been reported by Ludwig and Chow (The full tendon advancement of superior oblique. Paper presentation at American Academy of Ophthalmology meeting, October 27, 1999, Orlando FL).

It is possible that our patient also had bilateral 4th nerve palsies, given the history of placement of a ventriculoperitoneal shunt in infancy. Nevertheless, the V-pattern strabismus, over-elevation in adduction, and under-depression in adduction could all be explained by excyclotorsion of the globes associated with the craniosynostosis. Inferior transposition of the medial rectus muscle has been reported ineffective in addressing a V-pattern in the presence of inferior oblique overaction. (Guyton & Weingarten, 1994) Nevertheless, we speculate that addressing the excyclotorsion directly with bilateral superior oblique tucks normalized the vectors of the rectus muscles and allowed infraplacement of the medial rectus muscle to be effective. It is unclear whether infraplacement of the medial rectus muscles was actually needed in our case, because addressing the torsion may have been sufficient to allow simple recession of the medial rectus muscles. In addition, infraplacement of the medial rectus muscles creates an excyclotorsional force, (Guyton & Weingarten, 1994) which may somewhat negate the primary purpose of the procedure which was to directly address the excyclotorsion. Nevertheless, we speculate that the potential excyclotorsional effect of infraplacing the medial rectus muscles would be mitigated by the simultaneous recession. We believe the critical component of our procedure was the correction of bilateral excyclotorsion using superior oblique tucks. The small magnitude of the hyperdeviation in lateral gazes did not dissuade us from tucking both superior obliques because we felt that addressing the bilateral excyclotorsion was the most important component of the surgical plan.

Our approach would not be applicable in cases of craniosynostosis where the superior oblique was completely absent. Preoperative imaging and intraoperative traction testing would be helpful in establishing the presence of the superior oblique before embarking on the approach we suggest. In cases where the superior oblique is abnormally inserted, a resection and advancement under the superior rectus muscle, with restoration of the normal tendon path, may be preferable to tucking.

In summary, bilateral superior oblique tucks may be useful in addressing apparent inferior oblique overaction, apparent superior oblique under-action, and V-pattern strabismus associated with craniosynostosis, and may have broader application to other types of V-pattern strabismus.

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Figure 1.

Pre-operative versions showing apparent bilateral inferior oblique overaction (over elevation in adduction) and apparent bilateral superior oblique underaction (under depression in adduction), greater right eye than left eye.

Figure 2.
Pre-operative fundus photographs showing bilateral exocyclotorsion

Figure 3. Computerized tomography scan of the orbits showing relative excyclotorsion of each globe with medial rectus muscles more superior than normal and inferior rectus muscles more nasal.

Figure 4.

Post-operative versions (2 years following surgery) showing improvement in apparent bilateral inferior oblique overaction (over elevation in adduction) and improvement in apparent bilateral superior oblique underaction (under depression in adduction).

Ocular alignment measured using prism and alternate cover test (PACT) at distance and near pre-operatively and at 6 weeks and 2 years post-operatively.

Table 1

	PACT (prism diopters) at distance in primary position, right, left, up, and down gaze			PACT (prism diopters) at near in primary position
Pre-operatively		35ET, 10LHT		Near: 40ET, 5LHT
	40ET, 20LHT		40ET, 2RHT	
		50ET, 2LHT		
6 weeks post-operatively		6LHT		Near: 1XT, 2LHT
	6LHT	6ET*	0	
		8ET		
2 years post-operatively		8ET, 2LHT		Near: 4ET*
	8ET	8ET*	8ET	
		20ET		

PACT = prism and alternate cover test, ET = esotropia, HT = hypertropia, L = left eye, R = right eye

* patient also demonstrated a small dissociated vertical deviation