Surgical correction of ptosis in ocular fibrosis syndrome

Christopher Liu, Rita Ohri, Giovanni Frongia, Richard Collin

Abstract

The surgical management of ptosis is reported in seven patients suffering from the ocular fibrosis syndrome. Satisfactory results were obtained with bilateral Crawford type brow suspension with autologous fascia lata in six patients and bilateral Fox type brow suspension with stored fascia lata in a young child. As patients with ocular fibrosis syndrome usually exhibit little or no Bell's phenomenon, corneal exposure can become a problem after brow suspension. It was recommended that the lids are left just closed on the operating table at the end of the operation. None of the patients required a subsequent procedure to lower an overcorrection of the ptosis. The routine prescription of ocular lubricants for 2 months after ptosis correction is advocated. Urgent brow suspension in young children using non-autologous materials should only be considered if there is a risk of amblyopia.

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Generalised fibrosis syndrome of the extraocular muscles (also known as the ocular fibrosis syndrome,¹ congenital fibrosis of the extraocular muscles,²⁻⁵ and exaggerated A-pattern⁶) is a rare,⁷ congenital anomaly of unknown aetiology affecting the extraocular muscles and their fascial sheaths. The syndrome may occasionally be

familial when transmission is autosomal dominant.^{1,4,5,8,9} The anomaly is characterised by normal extraocular muscle tissues being replaced by primary fibrosis leading to a mechanical restriction of eye movements accompanied by ptosis.^{4,5,7,9,10} The eyes are in a hypotropic position to a varying degree because the inferior rectus muscles are the most severely involved.^{4,5,9} The patient adopts a head posture with chin elevation.^{4,5,7,9} The diagnosis is made clinically from the unique eye movement pattern, the presence of ptoses, and is confirmed by a positive forced duction test.⁹

Since there is minimal or no levator function in these patients,⁷ the choice of surgical procedure for the correction of ptosis is limited. Ideally, any extraocular muscle surgery to align the eyes in as near the primary position as possible for the elimination of chin elevation should be completed before ptosis surgery is considered.⁷ We undertook ptosis surgery on seven patients with ocular fibrosis syndrome in whom other causes for squint and ptosis had been excluded (for example, brain stem anomaly, third nerve palsies, etc).

Patients and methods

We carried out bilateral ptosis surgery in seven patients with ocular fibrosis syndrome. There were six females and one male. The average age at

Lid Clinic, Moorfields Eye Hospital, City Road, London
C Liu
R Ohri
G Frongia
R Collin

Correspondence to:
Mr C S C Liu, Department of Ophthalmology, Addenbrooke's Hospital, Hills Road, Cambridge CB2 2QQ.

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Table 1 Patient characteristics

Case No	Age at presentation (years)	Sex	Presenting complaint	Family history	No of procedures for strabismus and ptosis before presentation	Further procedures planned for strabismus	Ptosis: palpebral aperture (mm)		Ptosis: skin crease (mm)		Ptosis: levator function (mm)		Frontalis overaction	Bell's phenomenon
							R	L	R	L	R	L		
1	2	M	Bilateral congenital ptoses; bumping into things	Brother mother	Bilateral IR recession 6 months after ptosis surgery	None	2-3	2-3	None	None	Very poor	None	Marked	?
2	17	F	Eyelid levels getting lower interfering with vision	None	Bilateral Everbush (ptosis surgery) as small child	None	3	3	None	None	None	None	Marked	Poor
3	1 yr 4 mths	F	Bilateral ptosis; strabismus	None	2 previous strabismus procedures	None	5	6	None	None	<1	<1	Marked	None
4	10	F	Couldn't see through ptotic lids and lashes; problems with ball games at school	None	1 previous strabismus procedure	None	3	3	None	None	None	None	Marked	None
5	5	F	Bilateral ptosis	None	1 previous strabismus procedure	None	6	6	6	5	None	None	Marked	None
6*	24	F	Lids dropping again	Strong family history	2x strabismus ops. Bilateral ptosis surgery at 16 years	Awaiting correction of L exotropia	8	6	Incomplete (4)	(6)	1 to 2	1	Marked	Poor
7*	22	F	Bilateral ptosis	Strong family history	1 previous strabismus procedure	None	5	5	Hint of skin crease at (5)	(5)	None	None	Marked	Poor

All the above patients had bilateral ptosis since birth. All had chin elevation. Five patients had a positive traction test recorded at the time of surgery. *Cousins.

Table 2 Type of surgery and results

Case No	Indication(s)	Age at surgery (years)	Type of surgery (all bilateral)	Material	Result: palpebral aperture (in mm) (Pre-op)		Result: skin crease (mm)		Result: chin elevation	Result: complications* (including corneal exposure)	Result: patient satisfaction	Follow up (months)
					R	L	R	L				
1	Amblyopia despite occlusion	2	Fox brow suspension	Stored fascia lata	7 (2-3)	5 (2-3)	Good	None	Moderate residual chin elevation	Lid asymmetry	No longer falling over and bumping into things	15
2	Cosmesis; vision	18	Crawford brow suspension	Autogenous fascia lata	7 (3)	5 (3)	Not recorded		Much reduced	Leg hernia; on the brink of corneal exposure	Patient pleased	6
3	Cosmesis; head posture	4.5	Crawford brow suspension	Autogenous fascia lata	6-7 (5)	8-9 (6)	5	5	No head posture	None	Parents pleased; good cosmesis	22
4	Couldn't see through ptotic lids and lashes; problems with ball games at school	12	Crawford brow suspension	Autogenous fascia lata	6 (3)	7 (3)	None		Much reduced	None	Seeing better; performance with ball games improved	4
5	Cosmesis	6.5	Crawford brow suspension	Autogenous fascia lata	6 (6)	6 (6)	5	5	Much reduced	Mild left corneal stain	Father happy with appearance	9
6	Cosmesis	25	Crawford brow suspension	Autogenous fascia lata	8 (8)	8 (6)	4	6	Much reduced	R<L lagophthalmos; left corneal stain	Patient pleased	11
7	Cosmesis	22.5	Crawford brow suspension	Autogenous fascia lata	9 (5)	9 (5)	Not recorded		Much reduced	None	Patient pleased	5

All cases were done under general anaesthesia. In all cases there was much reduced frontalis overaction following brow suspension. None of the patients required further eyelid surgery for postoperative corneal exposure.

presentation was 11-62 years (range 16 months to 24 years). The presenting complaint, family history, surgical history, and further surgery planned for strabismus and ptosis and the pre-operative assessment of ptosis are summarised in Table 1. Since all the patients effectively had no levator function, they all underwent brow suspension ptosis operations.

The indication(s) for ptosis surgery, the age at which surgery was carried out, the type of surgery and material used, the result of ptosis surgery including complications and patient satisfaction, and the follow up period are summarised in Table 2.

Results

We achieved satisfactory results in all seven patients (Figs 1 to 4). All the patients found it

less of an effort to open their eyes post-operatively. The palpebral apertures of six patients widened after surgery. Case 5 had palpebral apertures of 6 mm in each eye both before and after brow suspension, but had good skin creases postoperatively as had the other patients and she was happy with the result. Skin creases have the effect of improving cosmesis as patients 'look more awake' (Fig 2). Five patients had their chin elevation head posture much reduced. One had no awkward head posture and one had moderate residual chin elevation. Three patients developed mild corneal exposure but were controlled with simple lubricant ointment at night. One patient developed a leg hernia which healed after some months. One patient had asymmetrical lids which may be related to his having bilateral vertical rectus muscle surgery after ptosis correction (case 1). All the patients and/or their parents were pleased with their surgery. Two patients saw better and performed better. Case 1 no longer fell over and bumped into things. Case 4 performed better with ball games in the school playground.

Discussion

We have carried out ptosis surgery in seven patients with ocular fibrosis syndrome. In five patients, a full assessment of their strabismus and its treatment was made before any surgical intervention for the ptosis. Ideally, any extra-ocular muscle surgery should be completed several months before ptosis surgery is carried out. This is because the position of the lid may be affected by the repositioning or resection of extraocular muscles. Case 1 developed amblyopia despite treatment with occlusion and urgent bilateral brow suspension before strabismus correction was considered necessary. He went on to have bilateral inferior rectus recession 6 months later. He retained a good result for ptosis correction although there was

Figure 1 Preoperative photograph of a patient with ocular fibrosis syndrome demonstrating bilateral ptosis despite marked frontalis overaction and virtually no skin creases.

Figure 2 Postoperative photograph of the same patient demonstrating good elevation of both upper lids and reasonable skin creases. The eyebrows are now at a much lower level.

Figure 3 Postoperative photograph of the same patient demonstrating the use of her frontalis muscles to elevate her eyebrows and upper lids while the ocular position remains static due to ocular fibrosis.

Figure 4 Postoperative photograph of the same patient with both eyes closed demonstrating that eyelid movement is dependent on the eyebrows which are lowered allowing full closure.

some lid asymmetry. With case 6, surgery on the horizontal rectus muscles to correct her exotropia should not affect the lid position afterwards. In patients with ocular fibrosis syndrome, extra-ocular muscle surgery aims to improve cosmesis and head posture by centring the eyes within the palpebral apertures. Improvement in ocular motility cannot be expected.

The indications for ptosis surgery may be considered under the following groups:

Functional. Urgent ptosis surgery should only be carried out if there is a risk of amblyopia in young children. Very occasionally, this situation may be brought about by surgical correction of hypotropia. Ptosis surgery in older children and in adults may be carried out if vision is compromised despite using maximum tilted head posture. Paediatric patients should be refracted at regular intervals as the incidence of refractive errors is high.¹¹

Cosmetic. In adult patients cosmetic surgery is carried out primarily to reduce awkward head posture and create skin creases.

Not every patient with ocular fibrosis syndrome requires ptosis surgery.

Patients with ocular fibrosis syndrome have good frontalis muscle action.¹⁷ The purpose of brow suspension is to improve the efficiency of the frontalis action in lifting the lids while not limiting eyelid closure. The slings should also create a lid crease thereby improving cosmesis. Levator resection by either an anterior or posterior approach is not thought to be suitable because there is little or no levator function preoperatively.^{6 10 12} Even more importantly, the already fibrosed levator muscle, if resected, would induce lagophthalmos leading to corneal exposure. Our procedure of choice is a Crawford type brow suspension using autogenous fascia lata as first preference.^{4 5 9 12-14} The use of this autologous material is associated with the least risk of postoperative inflammation, rejection, and necessity for repeat surgery. The size of the leg is more important than the age of the patient in determining whether fascia lata can be harvested. In very small children the leg may not be long enough to allow a substantial graft to be taken and the underlying muscle may also be damaged. If non-autogenous material is required we currently prefer Mersilene mesh.^{15 16} Case 1 had bilateral Fox type brow suspensions in 1986 using stored fascia lata as we did not develop the use of Mersilene mesh until 1989.¹⁵

Corneal exposure is a very real risk with this group of patients.^{4 5 7 9 12} There is usually little or no Bell's phenomenon exhibited^{6 12} and the hypotropia adds to the problem. Brow suspension has to be carried out cautiously. At the end of the procedure, the position of the lids should be set so that they are just closed on the table with the patient asleep. We believe that this is the maximum amount of surgery that can be done without causing serious problems with corneal exposure. We routinely prescribe tear supplements during the day and an ocular lubricant at night for a period of 2 months postoperatively. If there is minor corneal exposure, these should be continued indefinitely. The upper lid may be dropped surgically if there is a serious problem with exposure. Asymmetry of the upper lid

positions may also be adjusted by way of further surgery.

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