Blepharoptosis surgery in patients with myasthenia gravis

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ABSTRACT

Background/aims To review our approach of cautious surgical correction of blepharoptosis in patients with myasthenia gravis (MG) to minimise risk of exposure complications.

Methods Retrospective case note review of 30 patients with symptomatic eyelid concerns despite appropriate medical treatment, who underwent eyelid surgery. The mean age at diagnosis was 47 years. 13/30 patients had systemic MG, 14/30 ocular MG and 3/30 congenital MG. The main outcome measures were improvement in eyelid height and/or position, duration of a successful postoperative result, need for further surgical intervention, and intraoperative or postoperative complications.

Results 38 blepharoptosis procedures were performed on 23 patients. Mean age at time of surgery was 62 years, with an average follow-up of 29 months. 10 patients (16 eyelids) underwent anterior approach levator advancement, 4 patients (5 eyelids) posterior approach surgery and 8 patients (15 eyelids) brow suspension. One patient (2 eyelids) had tarsal switch surgery. An average improvement in eyelid height of 1.9 mm was achieved. Postoperative symptoms or signs of exposure keratopathy occurred in 17% of patients. This necessitated lid lowering in one eyelid of one patient. During follow-up, 37% of eyelids required further surgical intervention to improve the upper eyelid height, after an average of 19 months (range 0.5–49 months).

Conclusions Over a third of patients in our series required repeat surgery, which would be expected when the initial aim was to under-correct this group. In contrast to previous commentaries, the amount of eyelid excursion was not the main factor used to guide the surgical approach.

INTRODUCTION

Myasthenia gravis (MG) is a term derived from the Greek word 'myasthenia' meaning 'muscle weakness' and the Latin word 'gravis' meaning 'serious'. Blepharoptosis and diplopia are common manifestations of MG,¹ with approximately half of all patients first presenting to an ophthalmologist.² When suspected, MG can be readily diagnosed, but the diagnosis is often missed.³ While medical therapy remains the first line approach for management of these patients,² ¹ it is thought that up to a third will require surgical intervention.⁵-8

It is now 15 years since the largest review of patients with MG who underwent blepharoptosis surgery.⁸ At that time, the authors demonstrated large gains in palpebral aperture (PA) height could be achieved, with levator function guiding the choice of surgical procedure performed. The aim

of this paper was to review the value of more cautious correction of these patients and whether levator function still guided treatment decisions for this challenging group.

MATERIALS AND METHODS

A retrospective case note review of 30 patients (15 female) with MG operated upon between 1992 and 2012 was performed. Institutional Review Board approval was obtained and a standardised template was used to collect the data. Patients of interest were either still symptomatic despite, or intolerant of, medical treatment and underwent eyelid surgery for blepharoptosis. Details of patients with MG undergoing other eyelid procedures were also collected.

The diagnosis of MG was confirmed by one or more of the following investigations: edrophonium chloride testing, acetylcholine receptor antibody assay, or repetitive stimulation single fibre electromyography. For a diagnosis of ocular MG, it was sufficient if blepharoptosis was present in one or both upper lids, which was not due to local eyelid disease and demonstrated fatigue (worsening of the ptosis after up gaze for 30–60 s) with recovery or improvement after a period of eyelid closure or after placement of a small ice-pack onto the upper eyelid.

Levator function was measured from full down gaze to full up gaze with the frontalis muscle prevented from contributing to eyelid movement by means of a thumb applied to the frontal area just above the eyebrow. The patient's PA was measured with the patient fixed in primary gaze with no abnormal head posture. The upper marginal reflex distance (MRD) before and after surgery was recorded where this was available. Outcome measures included change in the patient's PA or MRD, duration of a successful postoperative result, requirement for further surgical intervention, and intraoperative or postoperative complications.

RESULTS

Thirty patients with MG underwent eyelid surgery during the study period (table 1). Thirteen of 30 patients had systemic MG, 14/30 ocular MG and 3/30 congenital MG. The median age at diagnosis was 47 years (range 3 months to 78 years) and the median follow-up was 29 months (range 3–112 months). All patients were euthyroid, with two having a history of hyperthyroidism.

A total of 38 blepharoptosis procedures were performed on 23 patients (table 2) at a median age of 62 years (range 3 months to 80 years). Follow-up measurements were missing for nine



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Table 1 Diagnostic features of patients with myasthenia gravis undergoing surgery for blepharoptosis

Patient no.	Age at diagnosis (years)	Sex	Туре	Edrophonium	Anti ACh-receptor antibody	EMG	Thymectomy	Muscle biopsy	Time from diagnosis to surgery (years)
1	64	F	Ocular	+					8
2	67	M	Ocular	+		Equivocal	_		2
3	72	M	Systemic						3
4		M	Ocular		+				
5	47	F	Ocular	_	+				1
6	63	M	Systemic				+		2
7	53	F	Ocular	+	_		+	Normal	9
8	38	M	Systemic	+			+		20
9	34	F	Systemic	+			+		2
10	62	M	Ocular	+	_	+	_	Normal	1
11	3	F	Congenital	+			+	Normal	5
12	43	F	Ocular	_					34
13	69	M	Ocular	+	+	_	_		1
14	65	M	Systemic	_			_		1
15	45	F	Ocular						6
16	20	F	Systemic	+			+		4
17	56	F	Ocular		+				0
18	66	F	Systemic	+					7
19	38	F	Ocular	_	_	_	_		4
20	66	M	Systemic	+	+		+		4
21	48	F	Ocular	+			_		0
22	8	M	Systemic	+					1
23	0.5	M	Congenital	+	+	+			2
24	52	M	Systemic	+					2
25	62	F	Systemic	+			-		2
26	78	М	Ocular	+					2
27	32	F	Ocular	+					2
28	46	F	Systemic	+	+				3
29	57	М	Systemic	+	+				6
30	0.25	М	Congenital	+		+			0

⁺ represents a positive test; – represents a negative test; blank spaces indicate not performed or unknown. EMG, electromyography.

patients (16 procedures), who were excluded from the numerical analysis.

Postoperative symptoms or signs of exposure keratopathy occurred in 4/23 (17%) patients, necessitating lid lowering in one case. During follow-up, 14/38 (37%) eyelids required further surgical intervention (table 3) to improve the upper eyelid height after a mean of 19 months (range 2 weeks to 49 months).

Seven other patients underwent surgeries for reasons other than blepharoptosis (table 4). These included eyelid lowering in three patients, two lower lid malposition corrections, one epiblepharon procedure and one four-lid blepharoplasty. Three of the four eyelid lowering procedures were after blepharoptosis surgery performed elsewhere. No other complications were reported.

DISCUSSION

Ophthalmologists appear to be less apprehensive about performing blepharoptosis correction on patients with MG since Bradley and Bartley published their series 15 years ago. However, these are still high-risk cases with complications continuing to present to the authors' practices. Impaired or variable Bell's

Table 2 Details of patients with myasthenia gravis undergoing surgery for blepharoptosis

Type of surgery	Number of eyelids	Levator function (mm)	Over-corrected requiring surgery	Under-corrected requiring surgery	PA, MRD preoperatively (mm)	Mean gain postoperatively (mm)	Follow-up (months)
Anterior approach	16	11.3	0	31%	5.7, 0	1.5	33
Posterior approach (WLA)	4	12.5	0	50%	5.25, -0.1	1.6	7
Posterior approach (excisional)	1	18	100%	0	4, -1	6	37
Brow suspension	15	5.4	0	13%	4.7, -0.6	2.6	39
Tarsal switch procedure ⁹	2	10	0	0	7.5, -0.5	2.5	14
Mean	_	10	2.6%	37%	5.4, -0.2	1.9	29

MRD, upper marginal reflex distance; PA, palpebral aperture; WLA, white line advance.

Patient no.	Time after initial surgery (months)	Side	Procedures	Total follow-up (months)	Comments
1	6	L	Initial posterior approach surgery (WLA). Anterior approach performed as secondary procedure	37	1 mm lagophthalmos and slight recurrence, patient not keen on further surgery
2	49	L	Repeat posterior approach ptosis correction	52	'Excellent correction of eyelid height and contour'
6	0.5	L	Anterior approach, sutures adjusted in office at first postoperative visit	6	Continues to have asymmetry of eyelid position despite revision once in the operating room without much impact or eyelid position
8	5	R L	Adjusted—patient would like eyelids raised as no lagophthalmos. Sling placement adjusted on tarsus with permanent suture	93	Informed by referrer of 'good outcome'
	87	R L	Removal of old slings, new silicone slings inserted		
9	8	R	Repeated anterior approach ptosis correction	32	'Good outcome'
	32	L			
	11	R	Müller's muscle-conjunctival resection, 6 mm		
12	37	L	Initial posterior excisional surgery. Left levator	47	Exposure symptoms persist despite absence of
	46	L	then recessed on two occasions, both posterior approach		lagophthalmos, so eyelid lowered further
20	8	R	Initial anterior approach ptosis surgery redone with Gortex frontalis slings	24	Bell's phenomenon now noted to be fair to poor. Managing with topical drops, ointment and lower lid punctual plugs. Examination reveals mild inferior exposure changes.
21	39	R	Conservative redone anterior approach levator advancement because of dryness and mild lower lid retraction	50	Left pseudoretraction noted. Dryness present in both eyes and patient uses drops and nocturnal ointment

phenomenon, reduced ocular motility, incomplete or infrequent blink, a weakened orbicularis and risk of frank postoperative lagophthalmos are reasons to hesitate before recommending surgery. ¹² In this paper, we have demonstrated a shift in trend toward less ambitious gains in eyelid height in favour of functional results with a preference to repeat surgery if indicated.

This change is apparent when comparisons are made with previous series. In the largest group of 10 patients with predominantly ocular MG and good levator function, a single procedure corrected the blepharoptosis in 87% of eyelids, with only 20% of patients requiring additional surgery. The increase in PA with anterior-approach levator advancement averaged 4.1 mm. In this series, the gain was only 1.5 mm, with 37% of patients requiring repeat surgery.

The current goal of surgery must be to lift the eyelid to a better functional level while not causing unmanageable corneal

exposure. 13 Numerous procedures have been recommended for myogenic ptosis, with the choice historically being based upon the severity of ptosis and the degree of levator function. 12 14 15 The range of procedures these patients underwent appears to be largely surgeon dependent, as all the authors placed less reliance on actual measurements when managing this cohort. The old practice of elevating the upper eyelid by maximal levator advancement or levator muscle resection until loss of function necessitates a sling, may put patients at risk of severe lagophthalmos and exposure keratopathy.⁹ ¹⁵ One reason for this is that variation in patients with MG is the norm. 16 Fluctuation can mean that patients with MG often show a surprisingly good degree of levator function, even in the setting of severe blepharoptosis which, confusingly, does not necessarily correspond with a good surgical outcome.⁸ Even though levator function varies, many patients with MG can remain stable for long periods. 17-20

Patient	Age at surgery		Follow-up after surgery		
no (years)	Procedure	(months)	Comments		
14	65	Left lower lid tightening for epiphora. Lateral tarsal strip and tarsoconjunctival retractor plication	1	Epiphora resolved 4 weeks after surgery	
15	51	Right lid lowering due to eyelid retraction from previous ptosis surgery. Pericardial graft used as a spacer	4	3 earlier ptosis surgeries performed elsewhere, last one 4 years previously	
16	24	Bilateral entropion correction by retractor reinsertion	1		
17	56	Bilateral upper and lower eyelid blepharoplasty	25	Performed elsewhere before diagnosis of myasthenia gravis made	
18	73	Bilateral lid lowering due to lid retraction from previous ptosis surgery. Supraclavicular skin used as a full thickness graft to anterior lamellar	47	4 previous ptosis surgeries performed elsewhere	
19	40	Left lid lowering due to eyelid retraction	84	Thought to be secondary to Grave's disease	
29	63	Bilateral endoscopic brow lift and blepharoplasty	25	Bilateral lower eyelid elevation planned but patient is otherwise happy	

Clinical science

The authors also found that early levator resection does not necessarily correct eyelid position for the long term. Frontalis slings were therefore used in patients with MG at an earlier stage than for other patients (even when 8 mm of levator function was present, rather than the 4 mm suggested in standard texts). So 13 15 14 Apart from three of the earliest procedures, there was a preference for using more readily reversible and adjustable silicone slings. When placing a sling for these patients, the aim was to have the eyelids just closed at the end of the procedure; postoperatively, with brow elevation the visual axis could then be cleared.

Other surgeries were performed on some of the patients in the cohort, highlighting some additional learning points. Any lower eyelid malposition should be assessed and corrected first, in order to maximise corneal protection and reduce the risk of postoperative problems. Several blepharoplasties were performed without any detrimental sequelae. Some have tried to create a physiological sling between the eyebrow and eyelid simply through blepharoplasty for patients with myogenic ptosis, ²² although if too much skin or muscle is excised, blink and eyelid closure will still be compromised. ¹⁴ Three additional patients required eyelid lowering after ptosis surgery performed at other centres. These patients highlight the importance of stressing that the goals of surgery in patients with MG must be functional and not aesthetic.

This study, like others addressing blepharoptosis surgery in the setting of MG, is a retrospective review. The intention was not to imply a genuine temporal or geographical series, but to identify current practice and knowledge gained from managing these cases. As such, and similar to previous series, the review is limited by missing data for up to a third of patients, there are no controls, there is a high risk of potential observer bias, and there is a lack of agreed standardised outcome measures. In defence of this, as highlighted in the discussion, applying numerical values to patients with MG during follow-up can often be less useful than assessing how the patient is actually managing, if they are running into difficulties or if they require repeat surgeries. Follow-up on MG patients is difficult and statistical analysis of such cases is not going to provide hard-and-fast rules. ¹⁶

CONCLUSION

Although 37% of patients in our series needed adjustment, this reflects the safer approach to under-correct these patients in order to guard against exposure-related complications. In contrast to previous commentaries, the amount of eyelid excursion is not the main factor used to guide the surgical approach in these patients. Repeated operations are frequently required, and indeed expected, in the cautious management approach we describe.

Contributors RM and ASL contributed to the conception and design of the work. All authors were involved in the acquisition of data. All authors reviewed and revised the manuscript and gave final approval of the version to be published.

Competing interests None.

Ethics approval Queen Victoria Hospital NHS Foundation Trust IRB approved this study.

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Data sharing statement Anonymous case details are available from the corresponding author on request.

REFERENCES

- Roh HS, Lee SY, Yoon JS. Comparison of clinical manifestations between patients with ocular myasthenia gravis and generalized myasthenia gravis. Korean J Ophthalmol 2011;25:1–7.
- Benatar MK. Medical and surgical treatment for ocular myasthenia. Cochrane Database Syst Rev 2006;(2):CD005081.
- 3 Cogan D. Myasthenia gravis: a review of the disease and a description of lid twitch as a characteristic sign. Arch Ophthalmol 1965;74:217–21.
- 4 Kupersmith MJ, Ying G. Ocular motor dysfunction and ptosis in ocular myasthenia gravis: effects of treatment. *Br J Ophthalmol* 2005;89:1330–4.
- 5 Hardie D. A case of myasthenia gravis operated on for ptosis by Hess's method. Br J Ophthalmol 1928;12:31–3.
- 6 Kapetansky DI. Surgical correction of blepharoptosis in myasthenia gravis. Am J Ophthalmol 1972;74:818–20.
- 7 Bradley EA, Bartley GB, Chapman KL. Surgical correction of blepharoptosis in patients with myasthenia gravis. *Trans Am Ophthalmol Soc* 2000;98:173–80; discussion 180–1.
- 8 Bradley EA, Bartley GB, Chapman KL, et al. Surgical correction of blepharoptosis in patients with myasthenia gravis. Ophthal Plast Reconstr Surg 2001;17:103–10.
- 9 Demartelaere SL, Blaydon SM, Shore JW. Tarsal switch levator resection for the treatment of blepharoptosis in patients with poor eye protective mechanisms. *Ophthalmology* 2006;113:2357–63.
- 10 Eshaghian J, Anderson RL, Weingeist TA, et al. Orbicularis oculi muscle in chronic progressive external ophthalmoplegia. Arch Ophthalmol 1980;98:1070–3.
- 11 Lynch DR, Farmer J. Practical approaches to neurogenetic disease. *J Neuroophthalmol* 2002;22:297–304.
- 12 Lane CM, Collin JR. Treatment of ptosis in chronic progressive external ophthalmoplegia. Br J Ophthalmol 1987;71:290–4.
- Holck DE, Dutton JJ, DeBacker C. Lower eyelid recession combined with ptosis surgery in patients with poor ocular motility. Ophthalmology 1997;104:92–5.
- 14 Wong VA, Beckingsale PS, Oley CA, et al. Management of myogenic ptosis. Ophthalmology 2002;109:1023–31.
- 15 Anderson RL. Predictable ptosis procedures: do not go to the dark side. Ophthal Plast Reconstr Surg 2012;28:239–41.
- 16 Eliasoph I. RE: Surgical correction of blepharoptosis in patients with myasthenia gravis. Ophthal Plast Reconstr Surg 2002;24:312–13.
- 17 Ortiz S, Borchert M. Long-term outcomes of pediatric ocular myasthenia gravis. Ophthalmology 2008:115:1245—8.e1.
- 18 Kupersmith MJ, Latkany R, Homel P. Development of generalized disease at 2 years in patients with ocular myasthenia gravis. Arch Neurol 2003;60:243–8.
- 19 Conti-Fine BM, Milani M, Kaminski HJ. Myasthenia gravis: past, present, and future. J Clin Invest 2006;116:2843–54.
- Shorr N, Christenbury JD, Goldberg RA. Management of ptosis in chronic progressive external ophthalmoplegia. Ophthal Plast Reconstr Surg 1987;3:141–5.
- 21 Lamont M, Tyers AG. Silicone sling allows adjustable ptosis correction in children and in adults at risk of corneal exposure. *Orbit* 2010;29:102–5.
- 22 Burnstine MA, Putterman AM. Upper blepharoplasty: a novel approach to improving progressive myopathic blepharoptosis. *Ophthalmology* 1999;106:2098–100.