

Apraxia of eyelid opening: Clinical features and therapy

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PURPOSE. *Botulinum toxin injection is the treatment of choice in cases of benign essential blepharospasm. However, about 10% of the patients do not get sufficient effect from this treatment, and many of them have concomitant apraxia of lid opening.*

METHODS. *Over a 3-year period we treated 12 patients. Three had pure apraxia of lid opening and in the other nine it was associated with blepharospasm. All patients were initially treated with botulinum toxin injections with poor results. They underwent surgical treatment like blepharoplasty, limited myectomy, aponeurosis repair, and/or frontalis suspension. Some of them needed postoperative botulinum toxin injections in the pretarsal part of orbicularis oculi muscles.*

RESULTS. *This combined therapy gave good functional and aesthetic results.*

CONCLUSIONS. *The specific causes of blepharospasm and apraxia of lid opening are unknown, but these two conditions coexist in some patients and can be difficult to treat. It is important to make a correct diagnosis, and a combined surgical and botulinum toxin treatment can be very effective. (Eur J Ophthalmol 2006; 16: 204-8)*

KEY WORDS. *Apraxia of eyelid opening, Blepharospasm, Botulinum toxin*

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INTRODUCTION

Disturbance of eyelid movements may result in involuntary eyelid closure and make the patient practically blind.

Blepharospasm (BSP) is a focal dystonia, characterized by excessive involuntary closure of eyelids, due to spasm of the orbicularis oculi muscles, in the absence of an ocular pathology (1). This condition can be associated with apraxia of lid opening (ALO), a non-paralytic inability to open the eyes at will in the absence of visible contraction of the orbicularis muscles (2).

Use of botulinum toxin injection is the treatment of choice in BSP; its efficacy is high and it can be used for many years without side effect or loss of efficacy. However, some patients do not get sufficient effect with this treatment.

Ophthalmologists, who treat blepharospasm patients, have to know about ALO, because it is the most frequent cause of a missing effect of the botulinum toxin treatment.

We report our experience of using combined botulinum toxin and surgical therapy in cases of ALO.

MATERIALS

Over a 3-year period we treated 12 patients (7 women, 5 men, age 50 to 75 years, median 61 years) (Tab. I). Three had pure ALO and in the others it was associated with blepharospasm.

One patient had Parkinson disease, one had progressive supranuclear palsy, two had Meige syndrome, and in the other eight no underlying disease was detected.

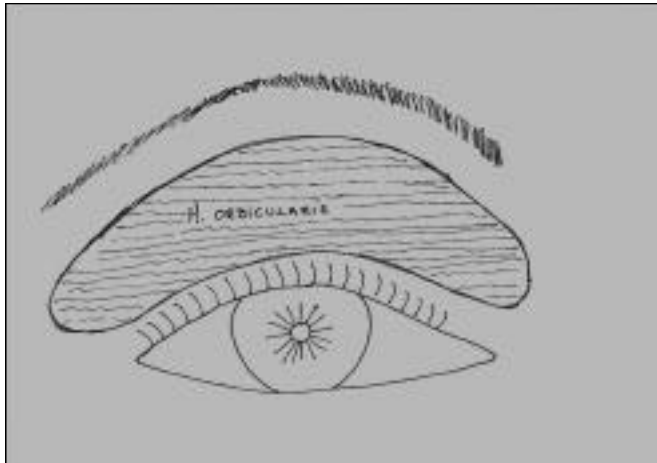


Fig. 1 - Limited myectomy: After removing the skin, the orbicularis muscle is dissected. Limited myectomy surgery of preseptal part of the upper eyelids. Excision of the preseptal part and upper part of the pretarsal orbicularis muscle, but leaving the lower part to maintain closure.

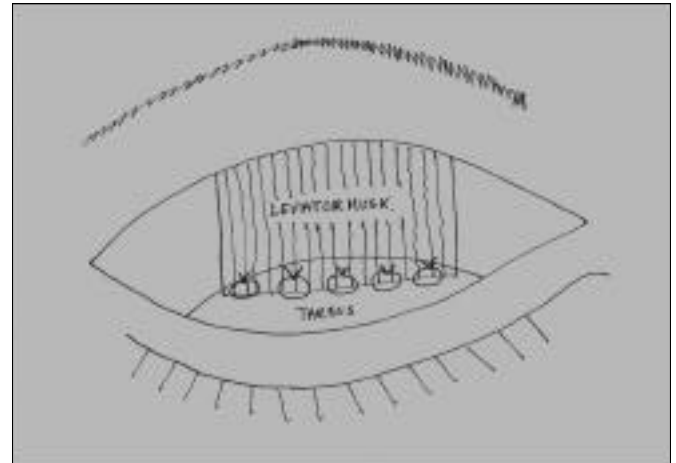


Fig. 2 - Aponeurosis repair: After orbicularis muscle is removed the levator aponeurosis is dissected and inspected. If there is a disinsertion it is repaired with plication, three to five sutures of Prolene 6-0 sutured to tarsus. If there is not, septum is dissected to the aponeurosis and repaired with plication. The skin is closed with 6-0 Prolene sutures.

Symptom durations before start of treatment varied greatly, from 1 year up to 10 years, median 4 years.

METHODS

All patients were initially treated with botulinum toxin injections with little or no effect.

In the nine patients who had a combination of blepharospasm and apraxia the starting total dose of BTX was 20 U (Botox, Allergan, dilution 100 µg/mL), and gradually increased to maximum dose of 40 U, injected at three points (in the upper eyelid laterally and medially upper and in the lower eyelid laterally only) into the orbital/preseptal part of the *musculus orbicularis oculi*.

After at least three inadequate treatments at 3-monthly intervals all these patients additionally received pretarsal treatment. For the other three patients, who had pure eyelid apraxia, we started directly with pretarsal BTX injections.

The average dose was lower in these cases, 28.4 (9.2) U. After at least two unsatisfactory BTX treatments, a combination of surgical treatments using different techniques was performed.

These were blepharoplasty, limited myectomy (Fig.1), aponeurosis repair (Fig. 2), or frontalis suspension (Fig. 3). The operations performed are shown in Table I.

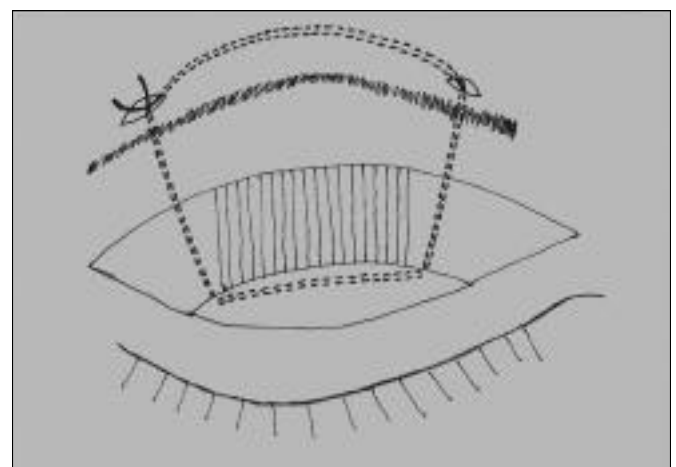


Fig. 3 - Frontalis suspension: Silicone rod or Gortex strip is just for suspension. Lid crease incision and one small skin incision lateral and one medial over the eyebrow. The sling is placed from tarsus in the eyelid to the frontalis muscle over the eyebrow. We used a single rombe technique.

RESULTS

Excellent or good and long-lasting functional and esthetic results were obtained in nine patients. Three of them needed additional BTX injections postoperatively for their blepharospasm and with this combined therapy an excellent functional result was obtained in these patients also. Three patients with moderate results after operations also needed BTX injections and were satisfied with



Fig. 4 - Patient with apraxia of lid opening. The patient is elevating the brows in an attempt to open the eyes.

the results. The average observation time was 14 months, range 5 to 28 months, and the results are still stable.

DISCUSSION

BTX is very efficient, simple, has few, if any, side effects, and can be used for a long time without loss of effect (3). However, in some patients one cannot obtain satisfactory effect, and in these cases one should consider that ALO may be associated. On the other hand, a few patients can have ALO alone, without BSP.

A patient who has a combination of blepharospasm and lid opening apraxia will typically have spasms closing the eyelids. But seconds after the spasm stops the patient will still be unable to open the eyes spontaneously or at command. They use the frontalis muscle to elevate the eyebrows. Despite the elevation of eyebrows, the eyes remain closed until the upper eyelid slowly elevates.

Pure apraxia of lid opening can occur, but is very rare as an isolated abnormality in association with extrapyramidal diseases, Parkinson disease, atypical parkinsonism, progressive supranuclear palsy, or after cerebral vascular events.

The prevalence of blepharospasm has been determined as 4.7 per 100,000 in Oslo, Norway (4), and ALO 59 per million (5). About 10% of the BSP patients had a concomitant ALO (6) and atypical parkinsonism in 25% of the cases (5).

The pathophysiology of BSP and ALO is not well known. ALO may be a result of involuntary levator palpebrae inhibition (7), pretarsal orbicularis oculi con-

traction (8), or prolonged orbicularis oculi activity (9).

Lid movement recording confirmed the clinical impression that patients with ALO show significant delay in completely opening their eyes (9). In these patients the time it takes to inhibit the orbicularis oculi activity is significantly longer than in the control subjects. The common association with extrapyramidal diseases suggests that dysfunction of basal ganglia may play a role in the pathogenesis of ALO.

EMG studies have shown that persistent activity of the pretarsal orbicularis oculi is a major factor interfering with eyelid opening in most patients with ALO although this is undetectable during clinical examination.

There are many clinical similarities between the two conditions.

The clinical characteristic of BSP consists of repeated involuntary forceful bilateral eye closure. The spasms are associated with lowering the brows below the superior orbital rim, the so called Charcot sign, indicating contraction of the pretarsal, preseptal, and orbital fibers of orbicularis oculi muscles. Often the first symptoms are increased blinking, photophobia, and foreign body sensation. The spasm increases in frequency and duration, making the patient functionally blind. When the spasm stops, the patient is able to open the eyes. BSP can start unilaterally, but always becomes bilateral.

ALO patients have transitory inability to initiate lid opening with no evidence of ongoing orbicularis oculi contraction. The ALO patient contracts the frontalis muscles causing elevation of the eyebrows—the inverse Charcot sign (Fig. 4). After a period with closed eyes, the eyelids slowly elevate and the patient can see again. The blinking rate is often reduced when the eyes are open. There is no photophobia, and the eye closure could be partial and asymmetric.

A common feature of the two conditions is using a sensory trick to open the eyes, such as light touch to the forehead or the eyebrow, which can help to open the eyes. Both conditions show diurnal variation, being better in the morning and worsening in the evening. The condition is worse during outdoors activities and in bright light.

Anderson et al (10) suggested that myectomy should always be the first surgical intervention, and in the absence of satisfactory effect, a frontalis suspension should be the second operation. According to our experience the treatment should be individually tailored to obtain the greatest benefit for the patients. Almost all patients with blepharospasm and apraxia have dermatochalasis. The eye-

lid skin becomes less elastic after the frequented manual traction attempt to open the eyes. We therefore performed blepharoplasty by removing excess skin in the upper eyelids in all patients. The cases with blepharospasm-associated apraxia of eyelid opening were treated by an upper myectomy and aponeurosis repair. When the patients had pure apraxia, without blepharospasm, we performed frontalis suspension.

De Groot et al presented 13 patients with ALO, treated

primarily with frontalis suspension: good and excellent results were obtained in 10 of them (11). They did not report whether the patients needed additional BTX injections postoperatively. Six of our patients still needed additional BTX treatment.

Previous authors reported different side effects of frontalis suspension, like numbness of the forehead, chronic lymphoedema of the periorbital region, ptosis, or eyelid retraction (10).

TABLE I - DEMOGRAPHY, CLINICAL SYMPTOMS, AND SURGICAL PROCEDURES IN PATIENTS WITH APRAXIA OF LID OPENING

Patient	Age, yr/ sex	Diagnosis	Duration, yr	BTX effect preop.	Operation	Results	Postop. treatment	Side effect	Follow-up mo.
1	74/F	Blepharospasm and apraxia Mb. Parkinson	9	None	BP, limited M, AR	Good	BTX		28
2	69/M	Blepharospasm and apraxia	6	Insufficient	BP, limited M, AR	Good	BTX		23
3	74/F	Blepharospasm and apraxia	4	Insufficient	BP, limited M, AR	Good	BTX		18
4	50/F	Pure apraxia	1	None	BP, FS	Moderate		Dry eyes	8
5	64/F	Blepharospasm and apraxia, Meige syndrome	4	Insufficient	BP, limited M, AR	Good	BTX		15
6	56/M	Pure apraxia	1	None	BP, FS	Good			7
7	52/F	Blepharospasm and apraxia	7	Insufficient	BP, limited M, AR	Good			21
8	61/M	Blepharospasm and apraxia	5	Insufficient	BP; limited M, AR 3 years later: FS	Excellent			8
9	75/M	Blepharospasm and apraxia Meige syndrome	10	None	BP, limited M, AR	Moderate	BTX		11
10	64/F	Blepharospasm and apraxia	3	None	BP, limited M, FS	Good			12
11	59/M	Blepharospasm and apraxia	2	None	BP, limited M, AR	Excellent			5
12	58/F	Pure apraxia Progressive supranuclear palsy	4	None	BP, FS	Moderate	BTX		16

BTX = Botulinum toxin; BP = Blepharoplasty; Limited M = Limited myectomy; AR = Aponeurosis repair; FS = Frontalis suspension

We did not observe any of these complications. This combined therapy – surgery and BTX injections – can give good functional and aesthetic results in patients with blepharospasm and lid opening apraxia when BTX injections alone are unsatisfactory.

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