Orbicularis Myectomy in Blepharospasm With Apraxia of Eyelid Opening Refractory to Botulinum Toxin A

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**Aim:** To discuss surgical management in Blepharospasm with Apraxia of eyelid opening (BEB with ALO), refractory to Botulinum toxin injection A (Botox). **Methods and Materials:** A series of 3 patients of BEB with ALO unresponsive to Botox, underwent partial Orbicularis myectomy. Patients were evaluated for eye opening, spasms, lagophthalmos and response to Botox. Eye opening and spasms improved vastly, requirement of Botox was reduced and response to Botox augmented. Conclusion: Partial Orbicularis myectomy for BEB with ALO is a safe and effective modality of treatment in cases refractory to Botox.

Benign essential blepharospasm (BEB) is a focal dystonia of unknown cause manifested by eyelid closure which is bilateral, involuntary and persistent, resulting in visual disability. Significant psychological and socioprofessional distress accompany this disorder. Along with the typical eyelid squeezing seen in BEB, patients may also experience an associated apraxia of lid opening (ALO). ALO is described as inability to open the eyes at will in the absence of visible contraction of the orbicularis oculi (OO) muscle. ALO results from involuntary levator palpebrae superioris inhibition and pretarsal OO contraction. Although all forms of BEB are distressing for the patient, those associated with ALO are the most debilitating because available treatments like botulinum toxin A (Botox) are usually ineffective. The aim is to discuss surgical management of BEB with ALO, refractory to Botox.

**MATERIALS AND METHODS**

A series of 3 patients of BEB with ALO unresponsive to Botox underwent partial OO myectomy. The surgery involved removal of the upper eyelid pretarsal, preseptal and orbital orbicularis oculi muscle (including the lateral raphae and temporal lower eyelid orbicularis), through an upper-lid crease incision. Patients were evaluated for eye opening, spasms, lagophthalmos and response to Botox injection, post surgery.

**Case 1**

A 52 year old female suffering from BEB with ALO since 3 years, showed poor response to Botox. She underwent partial OO myectomy surgery. Post surgery, she noticed good eye opening, an absence of eyelid spasms and no
lagophthalmos. She underwent upper lid skin blepharoplasty 6 months post myectomy surgery. 14 months post surgery, she had not received botox injection.

**Case 2**
A 70 year old female of BEB with ALO since 4 years, responded to Botox initially. Gradually the response weakened and she required 50-60 units BOTOX injection every 2 months. The poor control of spasms prompted her to undergo partial OO myectomy surgery. 1 year post surgery, she has good eye opening, infrequent and weak spasms, no lagophthalmos and requires 15-20 units Botox at every 8-9 months intervals with good response.

**Case 3**
A 67 year old female suffering from BEB with ALO since 6-7 years, had received multiple Botox injections with waning response. She required 70-80 units of Botox every 6-8 weeks and ultimately even this was insufficient to enable her to open her eyes. She opted for partial OO myectomy surgery. 1 year post surgery, she reports good eye opening, few weak spasms and no lagophthalmos. She has not received any further Botox for 1 year.

**DISCUSSION**
BEB with ALO is a debilitating condition, affecting the patient’s day to day activities as he is rendered visually handicapped at frequent intervals. Modalities like oral medications (Benzodiazepine, Antipsychotic, GABA B Receptor Agonist) and Botox are usually ineffective in controlling the spasms. OO myectomy appears to be effective in treating this condition and in improving quality of life. Upper eyelid myectomy is an integrated procedure that involves removal of the orbital, preseptal, and pretarsal OO (including the lateral raphae and temporal lower eyelid orbicularis) through an upper eyelid crease incision.

Our patients showed a vast improvement in eye opening and spasms, requirement of Botox was reduced and response to Botox augmented. In patients who continued Botox treatment after surgery, fewer units were required and the effect lasted longer. These results have even more importance considering that no other treatment has improved this debilitating problem significantly.

We conclude that partial OO myectomy for BEB with ALO is a safe and effective modality of treatment in cases refractory to Botox.