

WHAT IS APRAXIA OF EYELID AND HOW DO YOU TREAT IT?

Richard L. Anderson, M.D.

Apraxia of lid opening is a condition in which patients who have otherwise normal eyelids have difficulty opening the eyelids. This is a problem in the circuitry for opening the eyelids, much like blepharospasm is a problem in the circuitry allowing squeezing of the eyelids. Pure apraxia of lid opening (which is not associated with blepharospasm) is very rare, but apraxia of lid opening is commonly associated with blepharospasm. The specific cause or control center for both of these diseases is poorly understood, but must be somehow intertwined. The blepharospasm patient with apraxia of lid opening will typically have spasms squeezing the eyelids shut, and then for seconds to minutes after the spasm stops, the patient is unable to open the eyelids. The eyelids may then come open almost normally for a time period and then, without warning, slowly droop shut again or be drawn shut by spasm. The apraxia of lid opening patient can be seen raising their brows and trying to open their eyelids as strongly as possible without elevation of the eyelids.

To make the diagnosis of apraxia of lid opening, all blepharospasm or squeezing in the eyelids must be relieved. In rare apraxia of lid opening patients, it has been shown that a simultaneous firing of the squeezing muscles as well as the opening muscles of the eyelids occurs. However, under normal conditions, antagonistic muscles in the eyelids cannot contract at the same time. Therefore, if the eyelids have even minimal squeezing, opening cannot occur. Botulinum toxin, myectomy, or a combination thereof must completely relieve muscle squeezing before a diagnosis of apraxia of lid opening can be confirmed.

We examined 100 consecutive blepharospasm patients presenting to our clinic and found the incidence of apraxia of lid opening to be 7% in the general population of blepharospasm sufferers. However, in patients who are *botulinum toxin failures* the incidence of apraxia of lid opening is approximately 50%. It is very important for physicians and patients to be aware of apraxia of lid opening, as it is the most common cause of *failure* with botulinum toxin therapy, and increasing the dosage of botulinum toxin beyond what is necessary to adequately relieve squeezing or spasm in the eyelids frequently makes ptosis associated with apraxia of lid opening even worse.

In *botulinum toxin failures* of blepharospasm treatment, it is important to differentiate whether the patient has failed treatment because of the inability of botuli-



Figure 1.
Blepharospasm



Figure 2.
Blepharospasm but inability to open eyelids for seconds to minutes. Note elevation of brows in an attempt to open eyelids.

num toxin to relieve squeezing in the eyelids, or whether failure to adequately open the eyelids is the problem. This can be determined by having the patient vigorously squeeze the eyelids shut. If the patient has markedly weakened eyelid squeezing, then the botulinum toxin is working and doing all it can. If the patient still cannot open the eyelids, the most common cause of this problem is apraxia of lid opening.

The next most common cause of inability to adequately open the eyelids in the absence of spasms is ptosis and dermatochalasis. This is simply droopy eyelids or excess baggage or skin in the eyelids. Ptosis can easily be corrected by tightening the tendon of the muscle that raises the eyelids and dermatochalasis can be corrected by removing the excess baggage and tissues in the eyelids. These operations are referred to as a ptosis repair and blepharoplasty. However, in blepharospasm sufferers we combine these surgeries with removal of the squeezing muscles in the upper eyelids and call this procedure a limited myectomy. Following a limited myectomy, both the function as well as the cosmesis of the eyelids is greatly improved. The amount of botulinum toxin required is decreased and the effect and duration of the botulinum toxin are increased. Blepharospasm sufferers who have the combined advantages of success with limited myectomy and botulinum toxin are the happiest patients in our practice. They have improvement in function from both the limited myectomy and the botulinum toxin as well as cosmetic improvement.

Apraxia of Eyelid continued from page 8

Blepharospasm patients with apraxia of lid opening are treated by a limited myectomy associated with tightening of the levator tendon that raises the eyelids (aponeurotic ptosis repair). By tightening the tendon of the muscle that raises the eyelids, patients can more effectively open their eyelids. By excising the squeezing muscles in the upper eyelids via a limited myectomy, residual squeezing that is not completely relieved by botulinum toxin is improved. Most patients with apraxia of lid opening can be markedly improved with a combination of limited myectomy, ptosis repair, and botulinum toxin. Unfortunately, drugs have provided little or no improvement. However, despite these treatments, there remains a small group of patients who are unable to function adequately. In this group, a frontalis sling or frontalis suspension is performed as a second operation. The frontalis muscle is the forehead muscle that raises the eyebrows. In a frontalis sling, the frontalis is used to raise the eyelids by running a material from this muscle into the eyelids. We have recently found that a thick Gore-Tex suture is the best material available for frontalis suspensions in apraxia of lid opening. This suture material is readily obtainable and allows vascular ingrowth as opposed to other synthetic materials. Fascia lata, or tendons from elsewhere in the body, can also be used for frontalis suspension, but they require an additional surgical site for obtaining this material and they are not as easily adjustable if the patient's eyes are open too much or do not close adequately after the surgery.

In summary, apraxia of lid opening remains the most difficult problem to treat in blepharospasm sufferers. Pure blepharospasm sufferers respond remarkably well to botulinum toxin and/or myectomy. The failures of these forms of therapy have a 50% incidence of apraxia of lid opening. It is important for physicians and patients to be aware of apraxia of lid opening to appropriately diagnose and treat this condition. Increasing doses of botulinum toxin in a patient who is suffering from apraxia of lid opening frequently makes the condition worse by inducing further ptosis or droopiness to the eyelids. Unfortunately, drugs have been of little use in this condition. Limited myectomy and ptosis repair improves most patients but a few patients require frontalis suspension. The specific cause of apraxia of lid opening as well as blepharospasm is unknown but these two conditions are intertwined in this multifactorial and multifaceted disease process that we refer to as blepharospasm.
