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

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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 causing squeezing of the eyelids. Pure apraxia of lid opening (which is not associated with blepharospasm) is very rare. However, apraxia of lid opening is commonly associated with blepharospasm. The specific cause or control center for these disorders is poorly understood, but must be somehow intertwined. A blepharospasm patient with associated apraxia of lid opening will typically have spasms closing the eyelids and then for seconds after the spasm stops, the patient is still unable to open the eyelids. The eyelids may open almost normally for a time period and then, without warning, either droop shut or are closed by spasm. Apraxia of lid opening patients typically elevate their brows in an attempt to open their eyelids until the eyelids eventually open. Patients may use a finger to help open the eyelids and have difficulty maintaining open eyelids.

To adequately treat apraxia of lid opening, all blepharospasm or squeezing in the upper eyelids must be relieved. In some apraxia of lid opening patients, it has been shown by EMG that simultaneous impulses to the squeezing muscles and opening muscles occur. Under normal conditions, antagonistic muscle groups cannot contract at the same time. Patients cannot voluntarily squeeze the eyelids closed and open the eyelids simultaneously and if upper eyelids have even minimal spasm, opening cannot occur in some cases. Botulinum toxin, myectomy, or a combination thereof is required to completely relieve upper eyelid orbicularis muscle spasm and allow the levator muscle to contract and elevate the eyelid. Unfortunately, the orbicularis muscle in the central upper eyelid cannot be fully weakened with BOTOX without inducing a ptosis or droopy eyelid in many patients.

In botulinum toxin failures it is important to differentiate whether the patient has failed treatment because of the inability of botulinum toxin to relieve squeezing, or whether opening the eyelids is the problem. This can be determined by having the patient vigorously squeeze the eyelids closed while the examiners fingers attempt to pry the eyelids open. If the patient has markedly weakened eyelid squeezing, then botulinum toxin is working. We have shown that in blepharospasm patients who appear to be botulinum toxin failures, the incidence of apraxia of lid opening approximates 50%. It is important for physicians and patients to diagnose and understand apraxia of lid opening, as it is the most common cause of failure or inadequate response with botulinum toxin therapy. Increasing the dose of botulinum toxin beyond what is necessary to relieve spasm frequently makes ptosis (droopy eyelids). After apraxia of lid opening the next most common cause of inability to adequately open the eyelids is ptosis, brow ptosis, and dermatochalasis. This is simply droopy eyelids and brows or excess baggage and skin in the eyelids. Ptosis (droopy eyelids) can easily be corrected by tightening the tendon of the muscle that raises the eyelids. Dermatochalasis (baggy eyelids) can be corrected by removing the excess baggage and skin in the eyelids. These operations are referred to as

a ptosis repair and blepharoplasty. Removing excess tissues in the brow or elevating the forehead corrects brow ptosis. In blepharospasm sufferers we frequently combine these surgeries with removal of the squeezing muscles in the upper eyelids, a procedure called an upper myectomy. Following myectomy, both the function and cosmesis of the eyelids is usually greatly improved. The amount of botulinum toxin required is decreased and the effect and duration of botulinum toxin are increased. Blepharospasm sufferers who have the combined advantages of myectomy and are benefited by botulinum toxin are the most satisfied patients in our practice. They have improvement in function and cosmesis from both the myectomy and the Botulinum toxin only. A few patients are true failures of botulinum toxin (botulinum toxin provides no weakness when injected in muscles). These patients require both an upper and lower myectomy.

Blepharospasm patients with apraxia of lid opening are treated by an upper 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 myectomy, residual squeezing that is not completely relieved by botulinum toxin is eliminated. Most patients with apraxia of lid opening are markedly improved with a combination of myectomy and ptosis repair. In patients with apraxia of lid opening, nearly every fiber of orbicularis muscle in the central portion of the eyelid must be removed. Orbicularis muscle is meticulously removed overlying the eyelash follicles at the lid margin. Care must be taken not to damage the eyelash follicles and brow hair follicles. If residual muscle is left in the upper eyelid, apraxia of lid opening patients cannot open the eyelids. BOTOX is still used in the lower eyelids after an upper myectomy. If BOTOX provides no weakness in these muscles then a lower myectomy is performed approximately 6 months after the upper myectomy. Even after myectomy surgery and BOTOX, there remains a small group of patients with apraxia of lid opening who are unable to adequately function. In this group, frontalis suspension (frontalis sling) is performed at a second operation. In a frontalis sling, the forehead muscle is used to raise the eyelids by running a permanent suture material from this muscle into the eyelids. We recommend a 0-Gore-Tex suture for frontalis suspensions. This suture is readily obtainable and allows vascular ingrowth. The myectomy operation should be performed before a frontalis sling, which is the last resort for apraxia of lid opening. Unfortunately, drugs have provided little or no improvement in apraxia of lid opening.

In summary, apraxia of lid opening is the most difficult problem associated with blepharospasm. Pure blepharospasm responds remarkably well to botulinum toxin and/or myectomy. The “failures” of botulinum toxin have a high incidence of apraxia of lid opening. It is important for physicians and patients to understand, diagnose, and treat apraxia of lid opening appropriately. The specific causes of blepharospasm and apraxia of lid opening are unknown but these two conditions frequently co-exist and make this multifactoral and multifaceted disease that we call blepharospasm more difficult to treat.

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