

Apraxia of Eyelid Opening

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Apraxia of eyelid opening (AEO) is an eyelid movement disorder related to benign essential blepharospasm (BEB). This article discusses these disorders in five sections: basic background information, how they can be diagnosed, what causes AEO, non-surgical treatment and surgical treatment.

BACKGROUND

The orbicularis oculi muscle closes the eyelids and the levator muscle opens them. AEO and BEB affect the orbicularis muscles on each side of the face. A definition of BEB is involuntary bilateral contractions of the orbicularis that are associated with light sensitivity and eye surface irritation, frequently involving the face and neck muscles. A definition of AEO is the inability to relax the orbicularis muscles and initiate lid opening.

Both BEB and AEO are involuntary disorders, which mean patients cannot control these movements. Most patients with AEO also have BEB. The primary difference is patients with BEB have orbicularis contractions that close the lids, while patients with AEO have orbicularis contractions preventing them from initiating opening the eyelids. In summary, BEB patients cannot stop squeezing their lids and AEO patients cannot initiate opening their eyelids, even though they do not appear to have eyelid spasm.

Sometimes BEB is associated with other diseases, but AEO nearly always occurs in patients with other disorders. By far the most common associated disorder with AEO is BEB. The other two common associations are Parkinson syndrome and Progressive Supranuclear Palsy.

AEO sometimes develops after treatment for a different disorder. Some patients who had stereotaxic brain surgery for Parkinson syndrome have been cured of Parkinson syndrome but developed apraxia. Similarly, some patients who had deep brain stimulation to treat Parkinson syndrome have developed apraxia. AEO has been associated with drugs such as Flunarizine, Lithium and Levodopa. Levodopa, deep brain stimulation and stereotaxic brain surgery have sometimes caused secondary blepharospasm.

AEO is a rare disease affecting about 5 per 100,000 people with a 2:1 female:male ratio. The mean age of onset is 55 to 65 years old. A few patients will have a family history of muscle contraction disorders.

DIAGNOSIS

Untreated, AEO and BEB are visually disabling and may create functional blindness. Despite high availability of effective therapy, the average patient sees several doctors and goes without treatment an average of five years. An informed physician can diagnose both with a careful clinical examination.

The primary test to diagnose AEO is a voluntary lid opening test. This is a simple test where the patient closes the lids and attempts to open them on cue. When AEO patients are told to open the lids, there is a variable delay before they open or they may not open at all. To an untrained observer, it looks as if the patient is attempting to open the lids but has forgotten how to open them. To initiate lid opening, patients classically use tricks like looking up, throwing up the chin, or using fingers to open the eyelids and/or brows. This is different from BEB, where obvious lid squeezing prevents the lids from opening. When the lid squeezing stops, the lids open.

CAUSE

A reflex is an automatic response to a stimulus. A familiar example is a knee jerk when a physician taps the knee with a reflex hammer. Another type of reflex allows opposing muscles to work together to carry out a movement. Imagine a weight lifter curling a barbell to strengthen the biceps. When the athlete contracts the biceps to lift a weight, the triceps muscle must relax. If the triceps did not relax, it would work against the biceps and prevent the weight from rising.

There are many similar pairs of opposing muscles throughout the body. In the eyelid, these are the orbicularis oculi muscle, which closes the eyelids when it contracts and the levator muscle, which opens the lids when it contracts. These muscles work together by an inhibitory reflex. In a normal eyelid, when the orbicularis contracts, the levator relaxes and the lid closes. When the levator contracts, the orbicularis relaxes and the lid opens.

In AEO, there is an abnormality of the eyelid opening inhibitory reflex. When AEO patients try to open their eyelids, a portion of the orbicularis muscle nearest the eyelashes, called the pretarsal orbicularis, refuses to relax and the lid cannot open. AEO is caused by an abnormal reflex between the levator muscle and the persistently contracting pretarsal portion of the orbicularis muscle. The pretarsal muscle is quite small so when this muscle fails to relax and prevents eyelid opening, the muscle contractions are not visible. This is why patients with AEO often appear unable to open their eyes when there is no visible spasm of the eyelids.

NON-SURGICAL TREATMENT

The treatment of choice for patients with AEO is botulinum toxin injections in the pretarsal portion of the orbicularis, the specific muscle fibers responsible for the disorder that are nearest the eyelashes. Pretarsal injections improve AEO in about 70% of patients. In patients who have both BEB and AEO, the BEB is the more apparent

disorder because of the squeezing. After treatment with botulinum toxin the AEO may become more apparent, particularly if pretarsal toxin injections are not given.

Most patients with AEO also have BEB, which requires injections in other parts of the orbicularis muscle. Some physicians do not inject botulinum toxin into the pretarsal muscle for fear the toxin could spread upward and weaken the levator muscle, causing a droopy lid. However, botulinum toxin only spreads about 1 cm from where it is injected and the distance from the center of the pretarsal orbicularis to the levator muscle is about 2 cm. Pretarsal injections have demonstrated improved outcomes in AEO and increase the effectiveness of botulinum toxin in some patients with BEB who do not have AEO.

Oral medications do not work as well and are more toxic than botulinum toxin. Eyelid crutches are another option for AEO patients. Curved struts are attached to spectacle frames. The patient pushes the spectacles and the associated struts into the lids to assist with initiating lid opening. Some patients have used goggles to prop the eyes open instead of relying on the struts.

SURGICAL TREATMENT

Surgery should be reserved for patients who can no longer be managed with botulinum toxin injections or for patients in whom botulinum toxin injections are not effective. Pretarsal botulinum toxin injections should be given a trial prior to surgery in cases of AEO. There are two surgeries. One is myectomy surgery where the squeezing orbicularis muscle is removed. Myectomy surgery is reserved for patients in whom the eyelid spasm cannot be managed with botulinum toxin. The second is frontalis suspension surgery where a large stitch is used to couple eyebrow movements to eyelid movements. Frontalis suspension surgery is less invasive than myectomy surgery and is best for AEO patients who do not have eyelid spasm or those whose eyelid spasms can be managed with botulinum toxin. Frontalis suspension surgery allows AEO patients to initiate eyelid opening with the frontalis muscle of the forehead, as they are not able to do so with the levator muscle because of the abnormal reflex.

CONCLUSION

Apraxia of eyelid opening (AEO) should be differentiated from benign essential blepharospasm (BEB). The diagnosis of AEO rests primarily in the inability of patients to initiate lid opening, even when lid spasm is not evident. The cause of AEO is an abnormal reflex where the pretarsal orbicularis muscle continues contracting and prevents the levator muscle from initiating lid opening. First-line treatment for AEO is pretarsal botulinum toxin injections. In persistently symptomatic patients, myectomy and frontalis suspension surgeries are useful options.

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