

Hemifacial Spasm

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This is a condition in which there is involuntary twitching of muscles that are innervated by the seventh cranial nerve. The condition has two forms: typical and atypical. In the typical case, the twitching starts around the eye, usually the lower lid. As time progresses, the twitching spreads to include the whole lid, then the cheekbone area, then the lower jaw. As the progress is usually inexorable, the muscles in the neck become involved. In the atypical cases, the twitching starts in the lower face and progresses to involve the remainder of the facial muscles. It can sometimes be triggered by volitional contraction of certain facial muscles, especially puckering the lips or forcefully closing the eyes. Stressful situations or fatigue may also worsen the spasms. Estimates suggest that one in ten thousand people have hemifacial spasm.

A common feature is the development of a twitch that does not stop: **tonus**. In the eye area, this causes the eye to close, a prolonged wink, which the sufferer cannot usually oppose. The tonus phenomena lasts a few seconds, but can be prolonged.

As the twitching increases, and especially after the onset of tonus, there is often some underlying muscle weakness seen during periods of muscle relaxation.

The twitching is usually described as a sense of the lower lid moving, but may not be visible at all times. The patient usually experiences a feeling the muscles are always moving, to some degree.

The differential diagnosis of Hemifacial spasm helps to demonstrate how to make a secure diagnosis.

Blepharospasm - Bilateral blepharospasm (eyelid spasm) is most commonly the result of dry eyes, corneal irritation or benign essential blepharospasm (BEB). The prevalence of blepharospasm in the general population is approximately 5 in 100,000.

Meige's syndrome - Meige's syndrome is a rare neurological movement disorder (dyskinesia) characterized by spasms of the muscles of the eyelids and associated loss of tone in these eyelid muscles. Symptoms may include excessive blinking (blepharospasm) or involuntary eyelid closure. In time, the lower facial muscles become involved with yawning, jaw opening, and abnormal tongue movements. This is frequently bilateral. The exact cause of Meige Syndrome is not known. This disorder generally affects people during late middle age.

Facial myokymia - Myokymia is the spontaneous, fine fascicular contractions of muscle without muscular atrophy or weakness. Eyelid myokymia typically involves the orbicularis oculi muscle of one of the lower eyelids; occasionally, the upper eyelids also can be affected. In most cases, eyelid myokymia is benign, self-limited, and not associated with any disease.

Bilateral facial spasms - These can be seen in hemifacial spasm. The characteristic pattern is for unilateral involvement initially. Then, the opposite side of the face starts to spasm, but is usually not in step with the initial side. This is to differentiate from blepharospasm, where the squeezing of the eyelids and other muscles is usually symmetrical. In the myokymias, the movement is also sinuously symmetrical. Bilateral chronic motor tics (sometimes called habit tics) are preceded by an urge to move the face, and can be suppressed by the sufferer.

Etiology

Hemifacial spasm is generally recognized as resulting from compression of the seventh cranial nerve as it exits the brainstem. The compression can be from a blood vessel (common) or a tumor (rare). This can result in the common hyperactive (spasm) form or the less appreciated hypoactive (palsy) form of nerve compression.

Testing/Examinations

The diagnosis of hemifacial spasm is made by taking a history and doing a complete office exam. The facial electromyography study offers insight to phenomena labeled "lateral spread" This test identifies excessive activity in the nerves of the face, related to the hyperactivity found in the controlling cells in the brainstem (nucleus). It can be useful postoperatively, to help decide if the nerve has been well decompressed. A Magnetic Resonance Imaging (MRI) study is useful to delineate the anatomy in this disorder, but does not make the diagnosis. The rare occurrence of a tumor compressing the nerve does make the radiologic study useful.

Treatments

There are no medicines that can effectively control the spasm. Several classes of medicines (anti-spasmodics, anticonvulsants) will reduce some of the symptoms, for a time.

Botulinum toxin has been advocated for symptom control. This agent is a synthetic form of a biotoxin. It affects the muscle nerve connections, making it difficult for the nerve to excite the muscle into activity. It usually lasts for 11 weeks median effect, and has been associated with some waning effects with prolonged use. There are two forms of the agent at present. Neither is superior to the other for this application.

Microvascular decompression (MVD) was described by Dr. Peter Jannetta. This operation is directed to the cause of the problem, vascular compression. The goal is to move the vessel (artery or vein) away from the vulnerable site on the nerve and provide a pad to prevent future compression. Complications can include infection 1%, brain fluid leak 3%, facial weakness 1.4%, hearing loss 0.86%, and stroke <0.5%. Successful spasm relief ranges from 79% to 95%. The difference in the groups relates in part to patients in the former group who had undergone a previous procedure. Recovery from the procedure usually takes six weeks. Patients are usually out of the hospital on the second or third postoperative day. They are fatigued, however. The spasm can persist in 44% of patients, taking up to 18 months for complete resolution. 90% of patients are spasm free by 12 weeks after surgery.