HEMIFACIAL SPASM: DIAGNOSIS AND TREATMENT

Facial movement disorders:

It is not unusual for patients to report to their primary care physician with twitching on one side (unilateral) of their face. These muscle contractions may involve the eye, side of the face and/or neck. The diagnosis of the cause for such movements can often be made based on location and character. Listed below are common different diseases along with their distinguishing characteristics:

1. Blepharospasm: Bilateral, uncontrolled closure of both eyelids (blinking) secondary to contraction of the *orbicularis oculi* muscles which surround the eye. Causes include dry eyes, inflammation, bright light, stress, sleep deprivation, underlying neurologic disorder, and medications.
2. Facial myokymia: Bilateral or unilateral facial spasm that is continuous in nature. This disorder may be due to facial nerve nucleus injury from a brainstem tumor or from multiple sclerosis.
3. Hemifacial Spasm (HFS): Unilateral, uncontrolled, intermittent and sudden facial muscle contractions that usually begin around the eye causing eye closure. Over time, muscles of the mid and lower face may become involved as might the superficial muscle of the neck (aka: platysma muscle).

This newsletter will discuss Hemifacial Spasm (HFS) and its treatment.

<u>The Facial Nerve (Cranial Nerve 7; CN7; CNVII):</u>

The Facial Nerve (CN 7) is one of the twelve cranial nerves. Anatomically, CN7 arises from a nucleus located in the brainstem at the junction between the pons and medulla (Figures 1,2). The right CN7 innervates the right side of the head while the left CN7 innervates the left side of the head. While CN7 provides sensory (ear canal, tympanic membrane), special sensory (taste anterior 2/3 of the tongue) and parasympathetic (lacrimal gland and salivary gland) innervation, it is primarily a motor nerve that innervates the facial muscles and the stapedius muscle of the ear. The former control facial expression, while the latter protects hearing by stabilizing the middle ear's stapes bone when loud sounds are experienced.

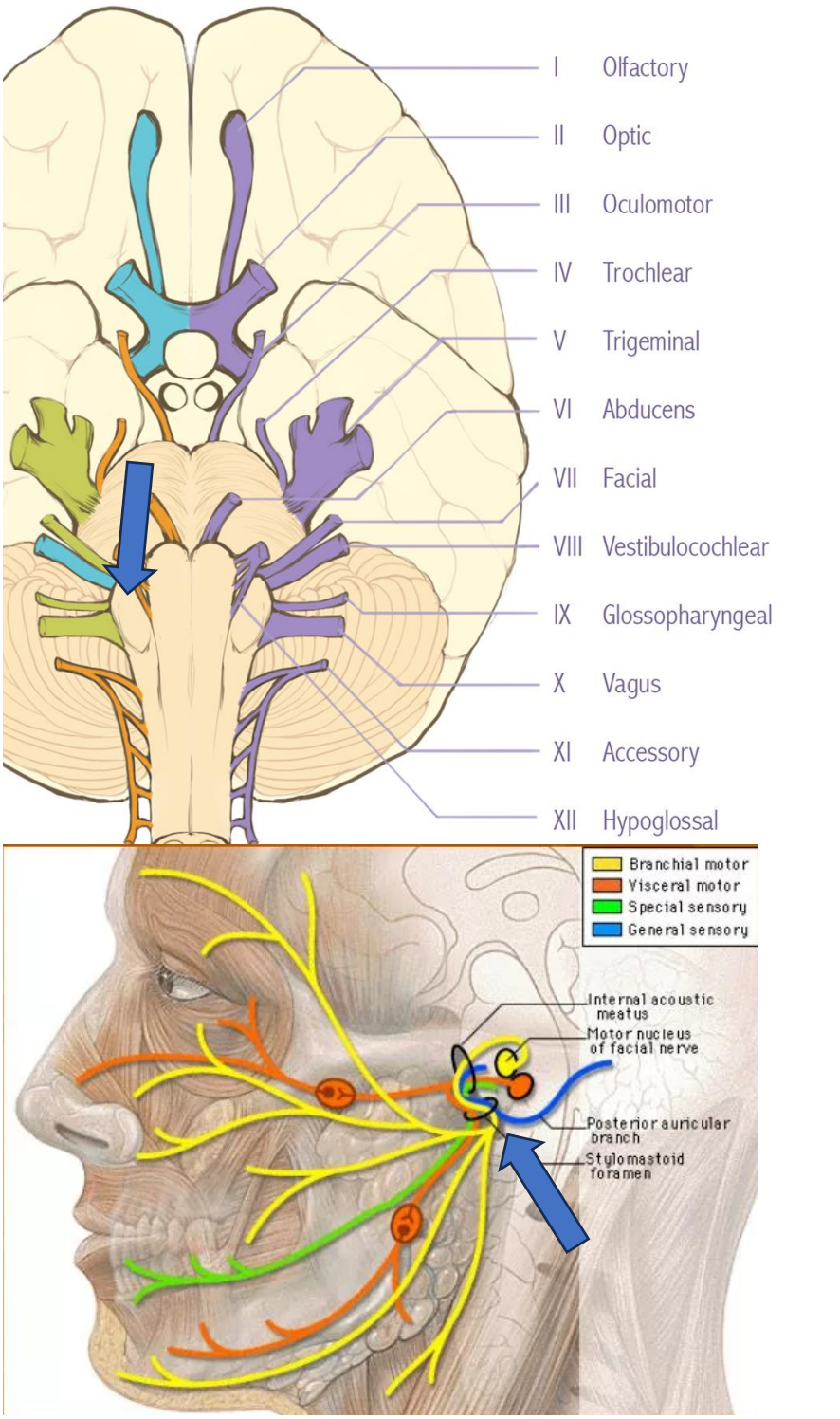
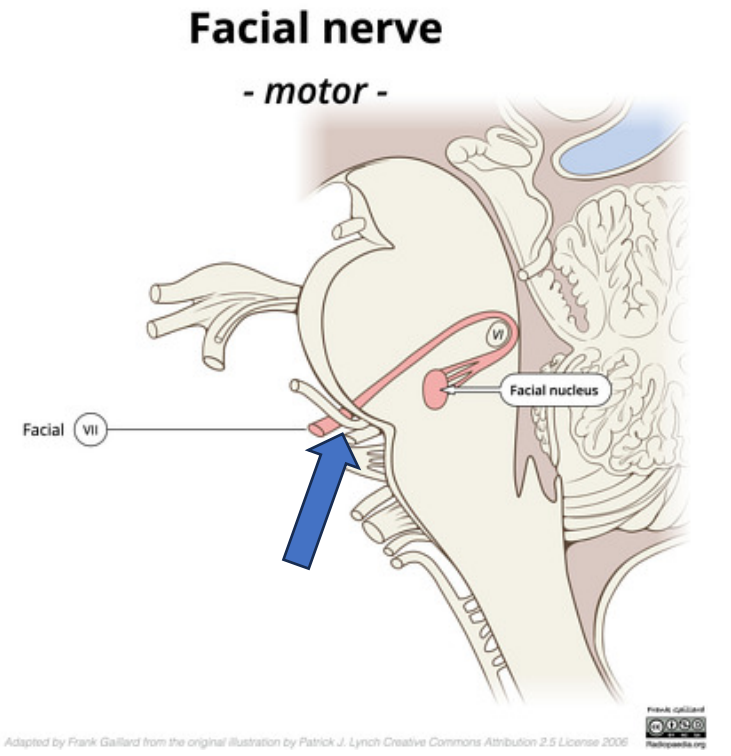


FIGURE 1: (Top) The 12 Cranial Nerves (Bottom) Cranial Nerve 7 anatomy and innervation. Yellow represents motor innervation of the facial muscles. Large blue arrows point to surgical target site for decompression.



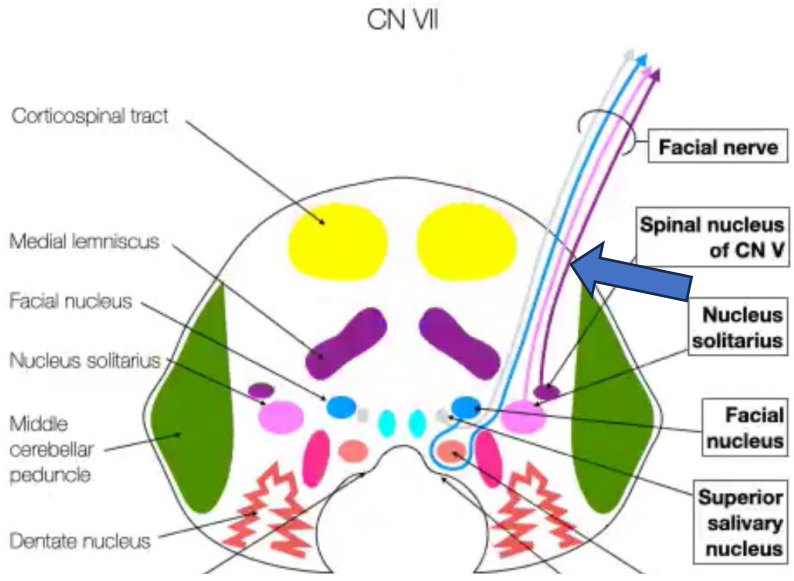


Figure 2: (Top) Sagittal view of the brainstem showing the location of the CN7 nucleus and CN7 at the level of the pontomedullary junction (Bottom) Axial view of the brainstem showing the CN7 nucleus location and the course CN7 as it exits the brainstem. Large blue arrows point to surgical target site for decompression

What are the hallmarks of HFS?

As stated above, HFS is a painless, involuntary movement disorder that presents with contractions of the facial muscles on one side of the face and neck. While symptoms generally begin around the eye, they usually progress to involve the muscles of the mid and lower face. Contractions are intermittent and spasmodic and may be worsened by stress or fatigue. In some instances, the face may temporarily lock in a contracted state with the eye lid shut and the mouth drawn upward. These findings are due to spasmodic contractions of the *orbicularis oculi* (muscle that closes the eye lids) and the *orbicularis oris* (muscle that controls the shape and movement of the lips). One HFS' interesting distinguishing factors is that it persists during sleep. Aside from palatal myoclonus (rhythmic involuntary jerking of the soft palate, throat muscles and diaphragm) no other movement disorder persists in such fashion. Less commonly, HFS can be accompanied by increased eye tearing or facial pain (aka: *tic convulsif*). Patients who have suffered from HFS for long periods of time may also present with underlying facial muscle weakness due to facial nerve injury. This weakness may be difficult to appreciate before HFS is treated because increased muscle tone that results from abnormal nerve stimulation may mask the underlying weakness which can only be observed when the nerve is completely inactive.

The involuntary and unpredictable character of HFS is responsible for social awkwardness as the face appears contorted and asymmetric. Frequent spasmodic unilateral eye closure can hamper an individual's ability to read, drive and carry out other activities of daily living. Many patients report being accused of inappropriate winking which makes interpersonal interactions difficult. When *tic convulsif* is present patients suffer from simultaneous movement disorder and severe facial pain which can make existence unbearable leading to self-harm and suicide.

Diagnostic criteria

HFS is primarily diagnosed using the above-mentioned criteria. Having an individual's sleeping partner witness facial contractions during sleep confirms all suspicions. If patients do not spontaneously demonstrate contractions during a physical examination/office consultation, instructing them to voluntarily hyperventilate("pant like a dog") while they also voluntarily squeeze their eyelids shut often elicits HFS when the patient is allowed to suddenly stop hyperventilating and voluntarily contracting the *orbicularis oculi* muscle.

While HFS is primarily identified based on history and physical exam, neurophysiologic examination of the facial nerve (electrical stimulation of the nerve and subsequent recording of the nerve's electrical reaction to that stimulation) yields signature recordings that are particular to the disease. This objective diagnostic tool, termed Lateral Spread Testing (LST), is based on synkinesis which occurs when one branch of the facial nerve is electrically stimulated and discharge of another facial nerve branch is recorded approximately 11 msec afterward. LST is not required to identify HFS, but it can be useful in cases where the physician questions the accuracy of their diagnosis. LST is extremely useful during surgery to help confirm that HFS has

been appropriately treated (Thirumala PD, Shah AC, Nikonow TN, Habeych ME, Balzer JR, Crammond DJ, Burkhart L, Chang Y, Gardner P, Kassam AB, **Horowitz MB**. Microvascular Decompression for Hemifacial Spasm: Evaluating Outcome Prognosticators Including the Value of Intropeative Lateral Spread Response Monitoring and Clinical Characteristics in 293 Patients. J Clinical Neurophysiology, Volume 28 (1) February 2011).

What causes HFS?

HFS is caused by a blood vessel (artery and/or vein) compressing the pontomedullary junction in a region that is adjacent to the CN7 motor nucleus. The most common offending vessel is the Posterior Inferior Cerebellar Artery (PICA) but the author has encountered compression from large veins, the Anterior Inferior Cerebellar Artery (AICA), the Vertebral Artery (VA) and the Basilar Artery (BA). With each heartbeat the blood vessels swell and pulse against the brainstem and exiting nerve. This brainstem compression causes CN7 misfiring which in turn results in involuntary spasmodic contraction of the facial muscles on the corresponding side of the head.

Treatment of HFS

Two treatments are currently offered to reduce or eliminate the signs and symptoms of HFS. These include:

1. Botulinum toxin (aka: (BT; Botox) injections (BTI) into the *orbicularis oculi* and *orbicularis oris* muscles.
2. Craniectomy and microvascular decompression of CN7 at the brainstem and nerve exit site from the brainstem (Root Exit Zone; REZ) .

Botulinum toxin Injection (BTI)

BTI into the muscles of facial expression works by chemically paralyzing the muscles so that they no longer normally contract. Reduction in muscle contraction means less spasmodic activity. This therapy uses Botulinum toxin one of the most poisonous biological substances known which is produced by the bacterium *Clostridium botulinum*. This substance blocks the release of acetylcholine, the principal neurotransmitter at the neuromuscular junction, causing muscle paralysis. The weakness induced by injection with botulinum toxin A usually lasts about three months (Nigam PK, et al. Botulinum Toxin. Indian Journal of Dermatology. 2010 Jan-Mar; 55(1):8-14).

BTI may help mask the signs and symptoms of HFS, but it does not cure the root cause of this movement disorder. Because it completely paralyzes muscle fibers, spasms cannot be completely eliminated unless the entire muscle is paralyzed which would lead to facial paralysis and drooping. For this reason, muscles are injected with lower doses of BT so that their contractile activity is lessened but not eliminated. Hence, spasms still occur but are less pronounced. BT's effect on muscle is temporary and injections must be repeated at regular

intervals to maintain spasm control. Often increased dosages are required over time. Long term, chronic use of BT may lead to permanent irreversible muscle weakness and facial droop. Taking the above into consideration, the author currently recommends BTI for HFS only in the very elderly and in those patients who are to infirm to tolerate general anesthesia.

Microvascular Decompression of Cranial Nerve 7 (MVD CN7)

MVD provides definitive cure for HFS because it targets the movement disorder's root cause, vascular (arterial and/or venous) compression of the facial nerve nucleus and the proximal facial nerve at the point where it exits the brainstem. This procedure is performed with the patient asleep under general anesthesia. Through a small hole placed in the skull behind the ear, the surgeon is able to access the patient's brainstem. Using a surgical microscope and fine instruments, the surgeon locates CN7 REZ and identifies blood vessels compressing the nerve. These vessels are elevated off the brainstem and soft padding made of Teflon is placed between the nerve and vessel so that the vessel no longer beats against the nerve and nucleus. Following surgery, patient's recover for 1-2 days and are then discharged home to be seen 10 days later for suture removal. While surgery sounds invasive, the incidence of any complications is less than 7%. These include stroke, neurologic injury, hearing loss, double vison, difficulty swallowing and infection. Cure rates after surgical decompression approach 85% (Thirumala PD, Shah AC, Nikonow TN, Habeych ME, Balzer JR, Crammond DJ, Burkhart L, Chang Y, Gardner P, Kassam AB, **Horowitz MB**. Microvascular Decompression for Hemifacial Spasm: Evaluating Outcome Prognosticators Including the Value of Introperative Lateral Spread Response Monitoring and Clinical Characteristics in 293 Patients. J Clinical Neurophysiology, Volume 28 (1) February 2011) and can be higher with two surgical procedures (Engh JA, **Horowitz M**, Burkhart L, Chang YF, Kassam A. Repeat microvascular decompression for hemifacial spasm. Journal of Neurology, Neurosurgery and Psychiatry 76: 1574-1580; 2005).

Of note, cure rates in patients who have received prior BTI is a bit lower than in those who have never engaged in such therapy. For best results, the author recommends that patient who have had BTI should not have surgery sooner than 3-6 months after their most recent treatment (Habeych, ME, Shah AC, Nikonkow TN, Balzer JR, Crammond DJ, Thirumala PD, Kassam A, **Horowitz M**. Effect of Botulinum Neurotoxin Treatment in the Lateral Spread of Monitoring of Microvascular Decompression for Hemifacial Spasm. Muscle & Nerve. August 2011).

NOTE: To watch a narrated video of surgical microvascular decompression for HFS, the reader may go to the Video Section at michaelhorowitzmd.com and select the appropriately labeled video from that section.

Conclusion

HFS is a facial motor movement disorder caused by vascular compression of CN 7 as it exits the brainstem. HFS that can be surgically treated with a high cure rate and low complication rate.