DIAGNOSIS AND TREATMENT OF GLOSSOPHARYNGEAL NEURALGIA

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GLOSSOPHARYNGEAL NEURALGIA (GlN)

Glossopharyngeal Neuralgia (GlN) is a rare cranial nerve (CN) syndrome that presents with severe, unilateral (right or left sided) lancinating pain involving the throat (especially lateral posterior portion), posterior tongue (base of tongue) and occasionally the ear and jaw. Discomfort may be spontaneous in onset, but more commonly develops when swallowing liquids (including saliva) or solids, chewing and/or talking. These actions are termed "triggers" and the pain they generate may lead to drooling (due to inability to swallow saliva), weight loss, dehydration and reluctance to speak.

In the author's experience, GlN is more common in woman (67%) than men (33%) with average age at the time of treatment being 50 years with the average period between pain onset and surgical treatment being 5-6 years. Pain occurs on the right or left side in an almost 1:1 distribution.

ETIOLOGY

The definitive cause for GlN is unknown. The widely held theory, however, for GlN and other cranial nerve neuralgias is that injury to a nerve's insulating coating (aka: myelin) is responsible. Myelin is felt to be damaged by the incessant injurious pulsations imposed upon it by arteries or veins that rest against the nerve. Damaged myelin in turn provides reduced insulation around each nerve fiber thus permitting electrical discharge from one fiber to affect neighboring fibers. This abnormal spread of electrical energy between different nerve fibers is termed "ephaptic transmission" (ET). ET results in adjacent nerve fibers discharging when they should be inactive.

To be more specific, imagine if a nerve fiber that transmits the sensation of light touch is in close proximity to a nerve fiber that transmits the sensation of sharp pain. Normally these two types of nerve fibers do not communicate with one another. If, however, the electrical insulative coating (myelin) around these fibers is damaged, then electrical transmissions traveling in one fiber might leak out and inappropriately stimulate the other fiber. As a result, a fiber transmitting the sensation of light touch might inadvertently activate an adjacent nerve fiber that transmits the sensation of pain. If the mucosa of the throat is now stimulated with food during swallowing, the nerve fiber that detects the light touch of food against the mucosa can activate the adjacent nerve fiber that normally only responds to painful stimuli. If the pain sensing nerve activates, then the brain will receive a pain signal from that nerve that was brought about by the simple act of food touching the mucosa. Hence results the inappropriate sensations experience by those who suffer from various cranial nerve neuralgias. The concept of myelin damage being the underlying cause for cranial neuralgias is supported by the finding

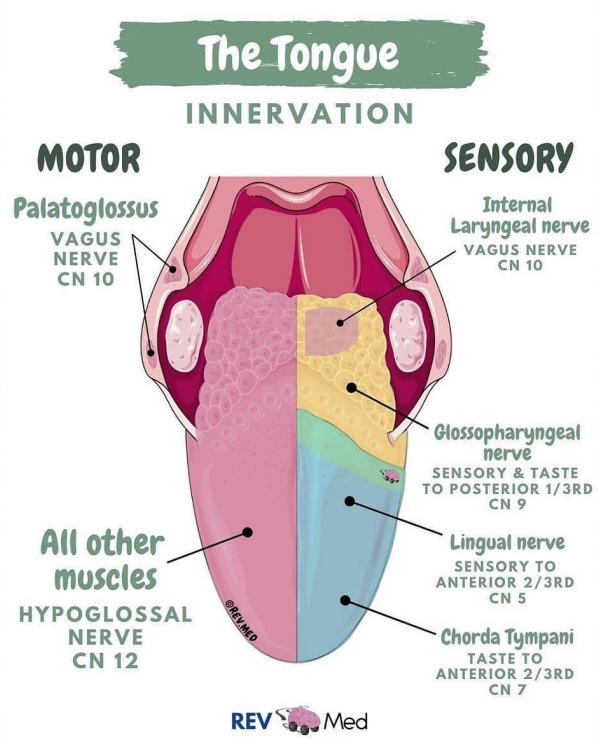
that patients with myelin degenerating disease such as multiple sclerosis can develop cranial nerve neuralgias without the presence of arterial or venous induced nerve injury.

NEUROANATOMY

Tongue:

General sensory information (touch, pain) is transmitted from the right and left sides of the tongue to the brain via the right and left Glossopharyngeal Nerve (CN 9), Vagus Nerve (CN 10), and the Trigeminal Nerve's V3 segment and its Lingual Nerve branch (CN 5) (Figure 1).

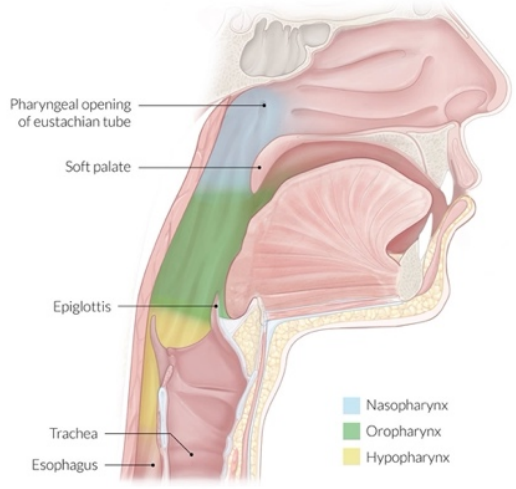
Figure 1:

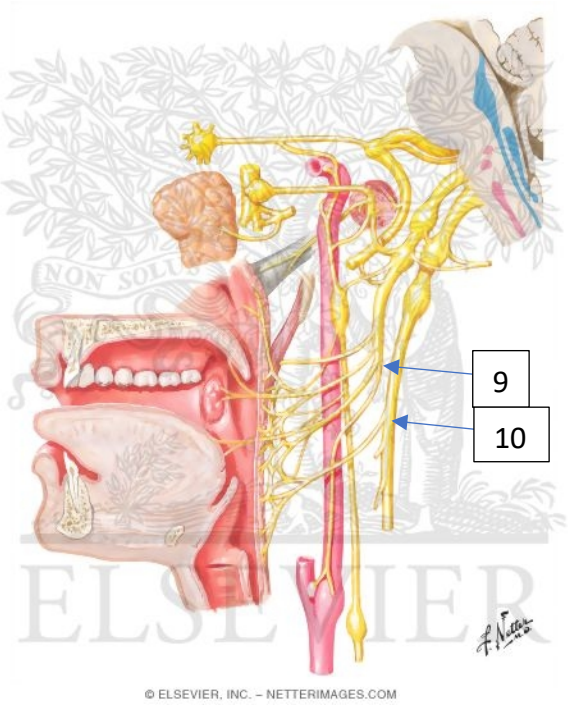


Throat (aka: Pharynx);

General sensory information is transmitted from the right and left sides of the pharynx to the brain via CN 9 and CN 10. The portion just anterior to the epiglottis is primarily serviced by CN 9 while the tissue extending from the epiglottis to the larynx (distal pharynx) is innervated by CN 10 (Figure 2).

Figure 2: (Left) The area shaded blue and green is primarily innervated by CN 9 and the area shaded yellow is primarily innervated by CN 10. (Right) Innervation of throat by CN 9 and CN 10.





The Vagus (CN 10) and Glossopharyngeal Nerves (CN 9) arise from the lateral medulla while the sensory portion of the Trigeminal Nerve (CN 5) arises from the pons. CN 9 is composed of one large exiting nerve fiber and CN 10 is composed of several exiting nerve fibers that ultimately converge and form a single larger structure (Figures 3,4)

Figure 3: (Left) CN 9, 10 arising from the medulla. (Right) Magnified view of CN9, 10 arisifrom the medulla

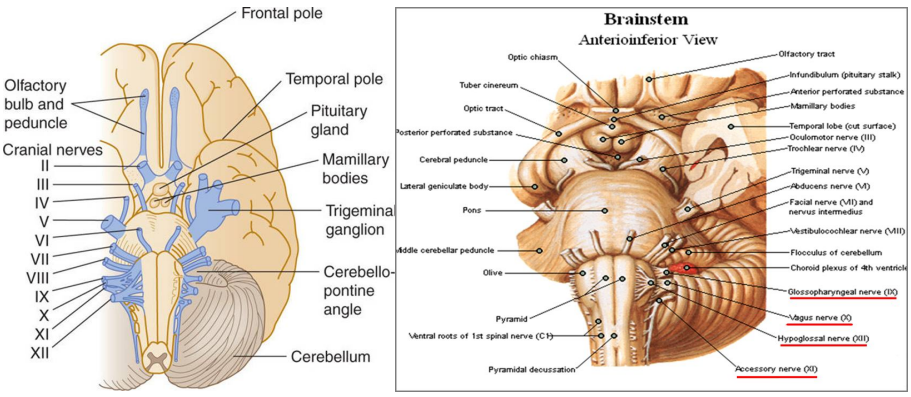
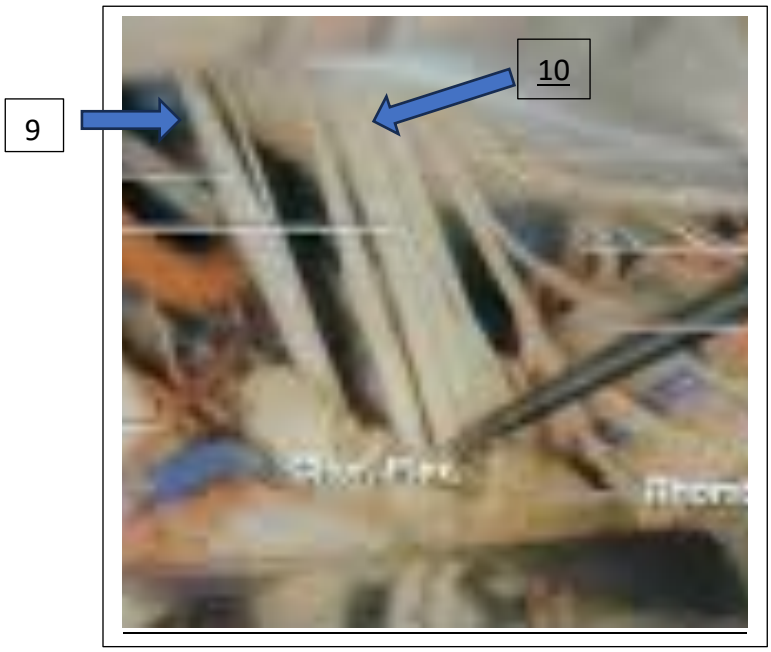


Figure 4: Magnified image of the actual CN 9, 10 exiting the medulla. Notice CN 9 is composed of a single fiber while CN 10 is composed of several fibers.



CLINICAL EVALUATION

The clinical evaluation of patient's suffering from posterior tongue and pharyngeal pain should begin with a thorough history that includes pain location, pain intensity, pain frequency and triggers. Once this information is obtained patients should undergo head CT and MRI with and without Gadolineum. Head CT should be performed with special attention to the skull base and styloid process (see below). All patients should undergo dental and ENT evaluations as well to rule out causes for discomfort aside from GlN. Because GlN is such a rare entity, the author considers it a "diagnosis of exclusion" meaning all other etiologies should be "ruled out" before GlN is "ruled in".

Differential Diagnosis

Because GlN is exceedingly rare (2-7/1,000,000 incidence) it is imperative that patients with severe, lancinating unilateral oropharyngeal pain during swallowing and speaking be evaluated for other potential etiologies. These may include:

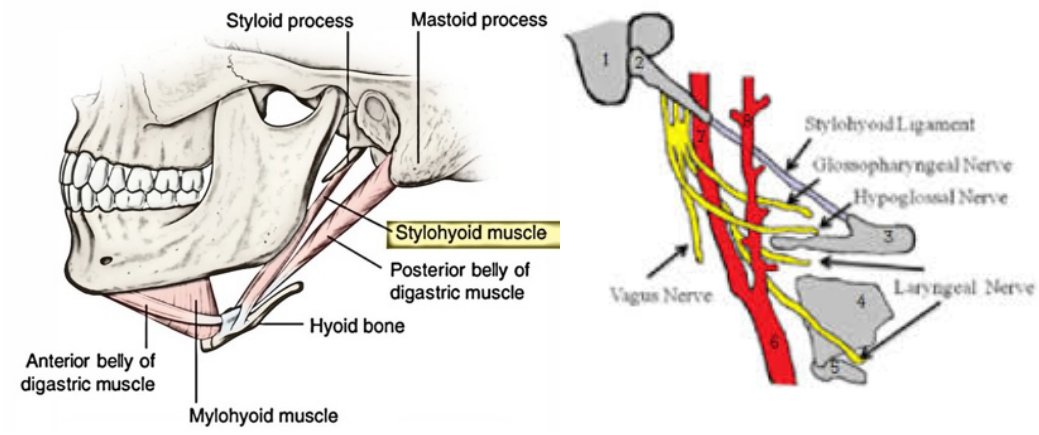
1. Infection
2. Ulcerations
3. Oral/pharyngeal Cancer
4. Trigeminal Neuralgia (TN)

5. Multiple Sclerosis (MS)
6. Eagle Syndrome

The presence of infection, ulceration and cancer can often be made using a combination of visual inspection, radiographic imaging, culture and biopsy/histologic evaluation of suspicious tissue. Examination for these disease processes is best performed by a combination of dentists, oral surgeons and otolaryngologists (aka: ENT surgeons). If this evaluation has not been performed prior to Neurosurgical consultation, GlN diagnosis and treatment should be deferred until a complete examination has been performed. TN can generally be diagnosed based on medical history while MS can be diagnosed using medical history, MR imaging, cerebrospinal fluid analysis and neurophysiologic electrical evaluation of the Optic Nerves using Visual Evoked Responses (VER).

Eagle Syndrome (ES) is a pain condition similar to GlN although it is generally constant and dull as opposed to intermittent and lancinating. Discomfort may involve the neck, ear lobe or lower face (jaw region). GlN may be exacerbated by moving the head or yawning. The etiology for ES is most often a congenitally elongated bony spicule, the styloid process, that protrudes bilaterally from the skull base. An ossified stylohyoid ligament may also be involved. Either of these anomalies may compress the GlN. Once diagnosed using clinical history and CT scanning of the skull, ES is treated by through resection of the styloid process and/or calcified stylohyoid ligament (Figure 5).

Figure 5: Styloid process. Elongation can impinge on CN 9



TREATMENT

Medical

As a result of abnormal communication (aka: ephaptic transmission) between pain fibers located in CN 9 and 10, oropharyngeal movements along with benign stimulation of the oropharyngeal mucosa are misinterpreted by the brain as extremely painful. These misinterpretations can also occur without notable mucosal stimulation. Medical management of GLN utilizes drugs that prevent the nerve fibers from firing inappropriately and stimulating other nerve fibers to do the same. These medications include anticonvulsants.
[carbamazepine (Tegretol), gabapentin (Neurontin), phenytoin (Dilantin)] and others that inhibit nerve excitability [pregabalin (Lyrica), oxcarbazepine (Trileptal) and baclofen (Lioresal).

Surgical

To watch a narrated surgical video showing the surgical procedure (Microvascular Decompression for Glossopharyngeal Neuralgia) the reader may go to michaelhorowitzmd.com and search the Video section of the web site

Medications may not alleviate GlN or may cause unacceptable side effects such as rash, liver injury, lowered blood cell counts, cognitive difficulties and gait instability. If medications fail to provide relief, patients are generally referred to a qualified neurosurgeon for a retromastoid craniectomy (removal of skull behind the ear) and microvascular decompression of CN 9, 10 and in some cases CN 5. To accomplish this surgical procedure the surgeon exposes cranial nerves 5, 9, 10 at the point where they enter the brainstem. CN 5 enters the lateral pons while cranial nerves 9 and 10 enter the lateral medulla. Using a surgical microscope and fine instruments, the surgeon searches for arteries or veins that are compressing the cranial nerves and stimulating/injuring the nerves with each cardiac pulsation (the arteries and to a lesser degree veins pulsate against the nerves causing damage to the insulating layers (myelin) that protect the nerve from abnormal stimulations. Blood vessels that are seen compressing the nerves at the point where they exit the brain (Root Entry Zone; REZ) are elevated from the nerve and Teflon felt padding is inserted between the blood vessel and nerve so that the vessel rests on a “Teflon pillow” rather than directly on the nerve itself. Absence of direct pulsation on the nerve is felt to alleviate the abnormal nerve signals that cause GlN. While compression of CN 9 and 10 may be from small arterial branches or larger arteries such as the Posterior Inferior Cerebellar Artery, Vertebral Artery, and/or Anterior Inferior Cerebellar Artery, venous compression may also b e a factor. The most common area for such venous compression is at the REZ between CN 9 and the first two fascicles (small nerve fibers) that compose CN 10. This. Vein generally cannot be elevated and decompressed using Teflon, but is instead sealed off and then cut using a surgical instrument called a bipolar cautery followed by microsurgical scissors.

Once CN 9 and 10 are treated, the author generally exposes the REZ for CN 5 and if it is compressed by arteries or veins, decompression of CN 5 is carried out as well using Teflon pillows. While some surgeons treat GlN by cutting CN 9 and the upper two fascicles of CN 10, this author does not treat GlN using destructive techniques. In almost all cases, vascular compression can be identified and relieved.

Surgical Outcomes:

Two articles published by the author and his colleagues are included below for the reader's review. The 2002 review of 217 cases is retrospective and as such has attendant limitations inherent to such types of investigations.

Patel A, Kassam AB, **Horowitz M**, Chang YF. Microvascular decompression in the management of glossopharyngeal neuralgia: An analysis of 217 cases. Neurosurgery50:705-711, 2002.

Horowitz MB, Horowitz M, Ochs M, Carrau R, Kassam AB. Trigeminal Neuralgia and Glossopharyngeal Neuralgia. JADA 135: 1427-1433, 2004.

In this author's experience, MVD for typical GlN provides an approximate 80% chance of immediate post-surgical pain relief and a 20% chance of immediate post-surgical partial or no relief. Long term complete pain relief 4 years following surgery drops to 58% with 18% reporting partial relief (76% pain elimination or significant reduction 4 years after surgery).

As stated earlier, patients with GlN may report pain involving the tongue, superficial throat, deep throat, jaw and ear. Patients who presented with primarily throat pain (aka: Typical GlN), displayed greater than 90% post-surgical long term pain relief/reduction while patients who had throat pain along with significant pain in another location had long term pain relief/reduction closer to 60%.

No surgical procedure involving the brain is risk free. Risks of MVD for GlN include, but are not limited to, stroke, death, cranial nerve injury, spinal fluid leak, infection, and difficulty swallowing. In this author's experience there have been no deaths or strokes and total complication rate remains $\leq$ 5%.

Summary:

If you or someone you know or care for suffers from pain that resembles Glossopharyngeal Neuralgia you may contact First Coast Neurosurgery, Orange Park, Florida at 904-276-7336. Additional information is available at michaelhorowitzmd.com