

TREATMENT OF GENICULATE NEURALGIA

Geniculate Neuralgia (GN):

Geniculate Neuralgia (aka: Nervus Intermedius Neuralgia (NIN)) is an extremely rare cranial nerve disorder that presents with intermittent, severe, paroxysmal, sudden onset, deep ear pain (aka: otalgia). Many state that GN resembles a knife or pick being thrust into their ear down towards and deep to the tympanic membrane (aka: ear drum) with occasional involvement of the external ear (aka: auricle; pinna; ear canal), eye, cheek, deep face, and hard palate. Patients describe their discomfort as 11/10 in intensity (greater than the worst pain of their lives). GN differs from Trigeminal Neuralgia as the latter involves electrical shocking type of pain in the face without deep ear involvement. GN patients rarely state that a particular action or event incites their pain although touching the ear canal along with swallowing might induce symptoms. It is unusual for any actions or medications to provide relief.

Neuroanatomy:

When surgically treating GN, four different cranial nerves require attention:

1. Trigeminal Nerve (CN 5)- sensation from the face
2. Nervus Intermedius (sensory branch of CN 7)- sensation from the tympanic membrane (ear drum)
3. Glossopharyngeal Nerve (CN 9)- sensation from the eustachian tube, middle ear cavity and throat
4. Vagus Nerve (CN 10)- sensation from the ear canal

The Nervus Intermedius (NI) (aka Nerve or Wrisberg; Glossopalatine Nerve), is a smaller than 1mm diameter fiber that exits the brainstem and travels between the more dorsally located Acoustic-Vestibular Nerve (CN 8) which is involved with hearing and the ventrally located Facial Nerve (CN 7) which is involved with facial movements. On occasion, the NI can consist of 2-3 small fibers each measuring less than 0.5 mm in diameter. While the NI has long been considered the sensory branch of the Facial Nerve (CN 7), some consider the NI to be a truly separate cranial nerve.

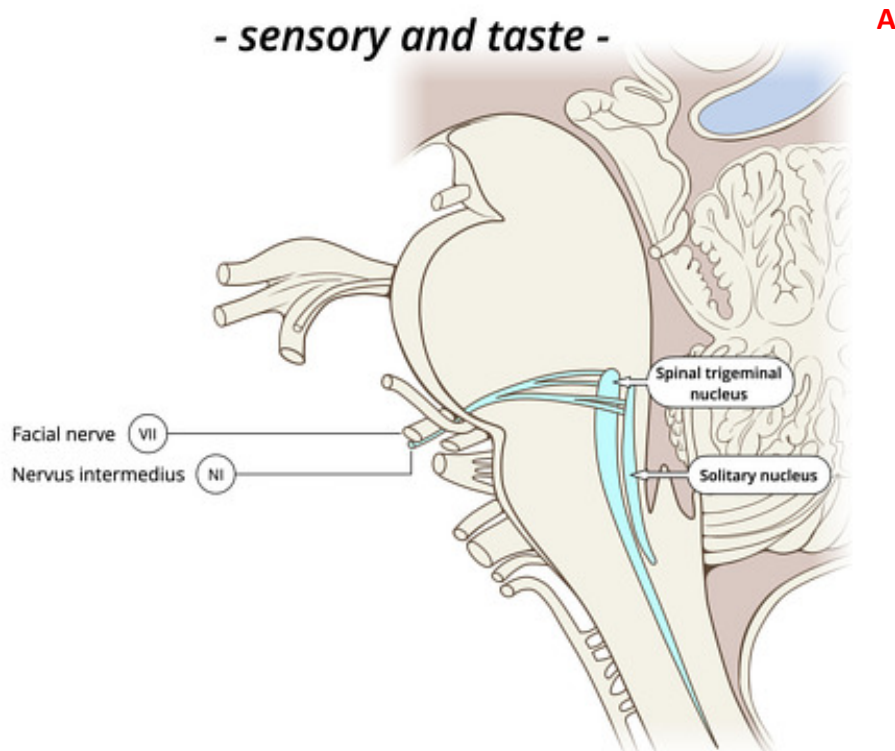
Despite its diminutive size, the NI serves several purposes:

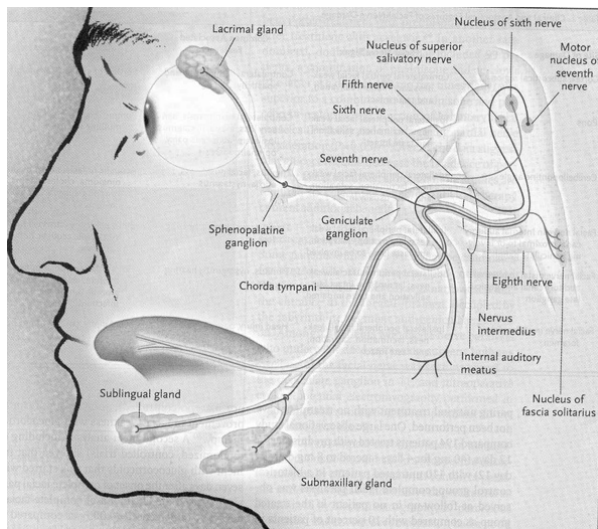
1. Sensory information (pain and touch) from the lateral tympanic membrane (ear drum) projects via the NI to the brainstem's Trigeminal Nucleus which in turn relays information to the sensory cortex.
2. Fibers project from the taste receptors in the anterior 2/3 of the tongue via the NI to the brainstem Solitary Nucleus (SN) from which information is then relayed to the sensory cortex.

3. Parasympathetic fibers from the Superior Salivatory Nucleus (SSN) project via the NI to the sublingual and submaxillary salivary glands to initiate release of oral saliva.
4. Parasympathetic nerve fibers from the Solitary Nucleus (SN) project via the NI to the Greater Petrosal Nerve which in turn innervates the lacrimal gland to stimulate tear release in the eye.

Figure 1 illustrates how the NI serves its multiple functions.

FIGURE 1 A, B, C





B



C

Figure 1:

(A) Anatomic sketch showing (i) NI carrying sensory fibers from the tympanic membrane to the Trigeminal Nucleus in the brainstem; (ii) NI carrying taste fibers from the anterior 2/3 of the tongue to the Solitary Nucleus in the brainstem

(B) Anatomic sketch showing parasympathetic fibers traveling from the Nucleus of the Superior Salivatory Nerve through the NI to the oral salivary glands

(C) Anatomic sketch showing sensory fibers traveling from the lateral tympanic membrane through the NI to the brainstem Trigeminal Nucleus from which sensory information is sent to the sensory cortex.

Differential Diagnosis:

When a patient presents with deep ear pain, clinicians should consider multiple potential causes for discomfort. These may include:

1. Geniculate Neuralgia (GN)
2. Otitis media (middle ear infection)
3. Otitis externa (external ear infection)
4. Herpetic infection of the Geniculate Ganglion (aka: Ramsay Hunt Syndrome, RHS)
5. Trauma
6. Ear drum perforation
7. Foreign body including insects

Alternative diagnoses (2-7 above) from GN can usually be excluded by examining the external ear, ear canal and ear drum. Pain secondary to these other etiologies generally do not present with intermittent and lancinating, but rather with more continuous discomfort. They may also be associated with decreased hearing, tinnitus (ringing in the ears), vertigo, and, in the case of RHS, facial weakness/paralysis.

Treatment:

GN's rarity means it often goes unrecognized, undiagnosed and untreated. Diagnosis is understandably difficult because many physicians, including Neurosurgeons and Ear, Nose and Throat Surgeons (ENT), have never encountered the malady. When it is encountered it is often confused with Trigeminal Neuralgia or considered a psychosomatic phenomenon.

Medical

GN rarely responds to medical . Some individuals, however, do respond to carbamazepine (Tegetol) and, for this reason, a trial of oral therapy may be warranted prior to offering a surgical procedure.

Surgical

The reader may go to michaelhorowitzmd.com where narrated videos are available showing the surgical procedure for treating GN and other cranial nerve disorders.

In summary, surgery for GN involves performance of a suboccipital retromastoid craniotomy (removal of a 3 cm diameter piece of the skull behind the ear) through a 5 cm skin incision that is hidden behind the hairline. Sectioning of the NI at the brainstem exit site is then performed so the nerve no longer functions. Care must be taken to identify the presence of more than one NI fiber because all fibers must be cut to achieve a cure. In addition to cutting the NI, it is recommended that the surgeon also inspect the Trigeminal Nerve (CN 5) where it enters the pontine section of the brainstem, the Glossopharyngeal (CN 9) and Vagus Nerve (CN 10) where they enter the lateral medulla. As mentioned earlier, these nerves also provide sensory information from the ear. If arteries or veins are compressing CNs 5, 9 and 10 at the brainstem

they should be separated from the nerve and brainstem by placing small pads between the vessels and nerve tissue (aka: microvascular decompression). Successful decompression eliminates contact between the pulsing vessels and nerve which hopefully allows the nerves to return to normal function.

Surgical Outcomes:

In this author's experience personally treating over 90 patients (1994 – 2023) with GN using microvascular decompression and NI sectioning (aka: cutting), the cure rate for elimination of ear pain is greater than 90%. This is a very high success rate for the surgical treatment of a pain syndrome and represents the highest cure rate of all the cranial nerve pain disorders. The high success rate may relate to sectioning of the NI. The author also believes that the diagnosis of GN is quite accurate when provided by an individual well versed with the disease. For that reason, there is a low probability of performing surgery on a patient with a pain syndrome that mimics GN but is not responsive to the surgical procedure described above. This differs from Trigeminal Neuralgia which can be misdiagnosed and which involves a more complex sensory apparatus.

In terms of patient experience, most individuals are admitted to the hospital the day of surgery. The surgical procedure takes approximately 2 hours to complete after which the patient is observed in the Intensive Care Unit until the following day. Most patients are discharged home the day after surgery although some may remain longer due to headache and/or nausea. Seven to ten days after surgery the staples or sutures are removed from the incision and the patient needs no further follow up unless they develop surgical complications such as infection, cerebrospinal fluid leak, problematic incision healing, or pain recurrence. If the patient is using pain relieving medications prior to surgery and their surgery is successful they can be weaned over 1-2 weeks from those medications after hospital discharge.

No surgical procedure involving the brain is risk free. All patients undergoing microvascular decompression for a CN disorder are informed that they can suffer a stroke, infection, spinal fluid leak, weakness, unilateral (one sided) facial paralysis, unilateral hearing reduction or loss, double vision, difficulty swallowing, facial sensory loss and potentially death. Surgical failure to eliminate pain is also possible. While these complications are concerning, the risk of any complication is less than 10%. Because the NI rests against the undersurface of the hearing nerve (CN 8), the risk of post-surgical unilateral hearing reduction/loss is unavoidable but unlikely. Interestingly, all patients I have operated on have stated that they would be willing to lose hearing in one ear in an attempt to rid themselves of their severe pain. I personally will not operate on a patient who is already deaf in the ear opposite the side I am treating as I do not think total deafness is an acceptable outcome unless a patient states that they will commit suicide if relief cannot be obtained. One such GN patient that I have seen with suicidal ideation was first referred for psychiatric evaluation, further pain management evaluation and repeat Ear Nose and Throat (ENT) evaluation prior to my agreeing to proceed with surgery. This patient, fortunately, was pain free after surgery and had retained hearing. The author is also

hesitant to operate on professional musicians or others who state their livelihood depends upon retention of normal hearing in both ears.

As stated above, the NI carries innervation to the lacrimal gland and salivary glands and from the anterior 2/3 of the tongue for taste. In all surgical procedures the NI is cut and for that reason patients should expect that they will quite likely note some unilateral changes in taste and unilateral eye dryness. These changes are not considered a surgical complication but rather an expected deficit based on the need to section the NI. In the author's experience, however, no patients find these changes to be life altering or even markedly bothersome.

Summary:

Geniculate Neuralgia (GN) is a rare but debilitating facial pain syndrome that primarily, but not exclusively, involves the deep ear canal. Medications rarely provide relief and most patients require surgery in order to obtain pain relief. Cure rate after surgery exceeds 90% and the complication rate is less than 10%.

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