

The 12 Cranial Nerves

- ON I Olfactory Nerve
- ON II Optic Nerve
- ON III Oculomotor Nerve
- CN IV Trochlear Nerve
- ON V Trigeminal Nerve
- CN VI Abducens Nerve
- CN VII Facial Nerve
- CN VIII Vestibulocochlear Nerve
- CN IX Glossopharyngeal Nerve
- CN X Vagus Nerve
- ON XI Spinal Accessory Nerve
- ON XII Hypoglossal Nerve

Trigeminal Neuralgia (TN), Hemifacial Spasm (HFS), and Glossopharyngeal Neuralgia (GPN), are cranial nerve hyperactivity syndromes that are usually caused by compression of the cranial nerve (5 - Trigeminal, 7 - Facial, or 9 - Glossopharyngeal) near the nerve root by a small blood vessel. The three syndromes have many similarities in presentation and treatment.

Management of **Neuropathic Face Pain Other Than Trigeminal Neuralgia**

By Michael H. Brisman, M.D., F.A.C.S.

In managing pain in the face or in the "trigeminal distribution" one must first try to distinguish whether such pain is "nociceptive" or "neuropathic" in nature. That is to say, is there pain because there is some abnormal process that is causing the pain, or is there pain, because a nerve or the brain is just not functioning properly and is transmitting pain signals that are not indicative of underlying disease.

For example, dental disease, such as a cavity, could certainly cause facial pain. So could an infection, such as a sinus, or ear, or tooth infection. So could an injury, like a corneal abrasion. So could a tumor in the brain or face regions. In these cases, treatment of the underlying disease, with the help of an appropriate specialist (such as, for example, a dentist, an otolaryngologist, a neurologist, an ophthalmologist, or a neurosurgeon), is the most appropriate method for eliminating the pain.

Sometimes however, patients experience pain in the face / trigeminal region, that is neuropathic in nature, that conveys no useful information. The pain itself is the problem. One specific case of such pain is Trigeminal Neuralgia. In these cases patients experience pains on one side of the face that are intermittent, sudden, brief, severe, sharp pains, that are usually triggered by light touch and respond to seizure medicines, especially carbamazepine. When medicines fail, procedures can be considered such as a Microvascular Decompression (MVD) where the blood vessel that compresses the nerve is moved off the nerve, or nerve injuring procedures such as percutaneous rhizotomy (injuring the nerve a bit through a needle placed into the nerve through the cheek) or Gamma Knife (super focused radiation aimed briefly at the nerve).

But how can other neuropathic facial pain syndromes be managed?

again, the first step is to make sure there is no underlying disease causing the pain. This can include evaluation with appropriate specialists as well as imaging studies, such as an MRI of the head.

Two specific facial pain syndromes are worthy of mentioning because they, like trigeminal neuralgia, have some very specific and effective treatments. The first is Glossopharyngeal Neuralgia. This pain syndrome is similar to Trigeminal Neuralgia, except that the syndrome involves the 9th cranial nerve, rather than the 5th. These patients get intermittent, sudden, brief, severe sharp pains that occur on one side of the deep ear or deep throat regions. Like Trigeminal neuralgia, these pains are often triggered by light touch of the affected areas, and usually respond to seizure medicines, especially carbamazepine. If medicines fail, MVD surgery to move a blood vessel off the 9th cranial nerve (the glossopharyngeal nerve), or Gamma Knife, can be effective at eliminating the pain.

Another important neuropathic pain syndrome that is important to identify is Chronic Paroxysmal Hemicrania. Patients with this condition, like patients with trigeminal neuralgia, experience intermittent severe facial pain episodes. Features that suggest this diagnosis include a focus of the pain in and around the eye, as well as "autonomic" features (such as redness of the eye, tearing of the eye, drainage from the nose, nasal congestion, and drooping of the eyelid). Chronic Paroxysmal Hemicrania is important to identify because the severe pain can be completely relieved with the anti-inflammatory medicine Indocin.

Other types of neuropathic face pain syndromes can be categorized as either having no clear cause, or having some known insult to the nerve. The first type of pain is sometimes called "atypical facial pain" (AFP) or "persistent idiopathic facial pain" (PIFP). The second type of neuropathic pain syndromes may be summarized with the term "trigeminal nerve injury pain", that is pain that results from a nerve injury of

some kind. Such an insult or injury can result from a procedure (such as a dental procedure or deliberate injury to the nerve to treat trigeminal neuralgia, or some other facial surgery) or some other insult to the nerve (such as a shingles outbreak in the area of the face). Shingles outbreaks in the face occur more in the elderly and more in the first division of the trigeminal nerve. This would be called "post herpetic neuralgia in the trigeminal distribution".

It is management of these other neuropathic facial pain syndromes that we will try to address here.

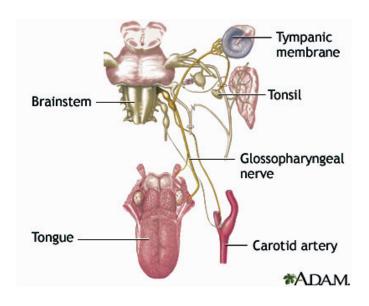
PIFP and Trigeminal nerve injury pain generally share certain features. The pain is often constant, though there can be times when it is worse. The pain is usually described as dull, achy, gnawing, or burning, though some intermittent sharp components are possible. There may be associated numbness, especially in the case of nerve injury pain. If the pain is nerve injury pain, it should develop soon after the injury or insult has occurred. Also, particularly in cases of injury, the face may be hypersensitive to painful stimuli ("hyperalgesia") or hypersensitive to non-painful stimuli ("allodynia").

Sometimes these problems are more mild, and easier to treat. Sometimes they can be quite challenging and difficult. One important point to keep in mind is that the operations to treat Trigeminal neuralgia, such as trigeminal nerve injury procedures, are not likely to work in these cases, as these patients already have some kind of nerve dysfunction. It is also important to recognize that PAIN is influenced by many different factors including MOOD, ANXIETY, STRESS, and SOCIAL SUPPORT. As such, psychiatrists and psychologists may also have a role in helping people with chronic pain. PAIN may also have peripheral nerve generators, central brain generators, or muscular generators. As such, a multimodal approach is often best for more difficult to treat cases. Also, it is important to keep in mind that many pain syndromes will improve with time.

A) Non-medical, non-invasive treatments:

- 1. Addressing depressed mood: Anything that can be done to help with a person's mood and avoid a depressed mood can be helpful. Sometimes just letting patients know that there are many options available and that there is good reason for hope, can improve how a patient feels.
- 2. Addressing anxiety: Anything that can be done to alleviate a patient's anxiety can also help. Sometimes just taking the time to rule out other diseases and giving the patient reassurance about their condition may be helpful. Letting the patient understand that there is no underlying disease present and that the condition may well improve with time, may also help.
- 3. Addressing stress: Anything that can be done to lessen a patient's stress levels can help. Often other family members need to be more sensitive to the patient's pain condition and help them at home so their stress levels are less. Meditation or Yoga may also help with this.
- 4. Addressing support: Anything that can improve a patient's support structure can help. This includes not just family and other treating physicians, but can also include participation in face pain support groups. Some of these are available online. Facial Pain support groups let people share their experiences, discuss things that worked for them, and help people know that they are not alone.
- 5. Application or Hot or Cold to the face.

 Sometimes patients may experience relief from periodic application of hot things to the face (like a warm washcloth or a heating pad) or cold things to the face (like an icepack wrapped in a towel). In some cases, these treatments may ease the pain the patient is experiencing.
- 6. Massage Treatments/Physical Therapy:
 Such treatments can be performed by family members, masseuses, or physical therapists.
 Some patients may find this helpful.



The Anatomy of Glossopharyngeal Neuralgia

B) Medical Management:

Medical Management is usually the mainstay of treatment for most neuropathic facial pain.

- 1. Over the counter (OTC) pain medicines:
 - a) Tylenol (acetaminophen): This is an effective pain reliever for mild to moderate pain.
 - b) Non-Steroidals: These medicines can also be effective for mild to moderate pain. These include common medicines like Aspirin (acetylsalicylic acid), Advil, Motrin (ibuprofen) and Aleve(naproxen sodium). Excedrin is another commonly used medicine that contains aspirin (a non-steroidal) as well as acetaminophen and caffeine.
 - c) Numbing medicines: Sometimes people may benefit from topical, over the counter, numbing medicines like Benzocaine which can be used as a topical pain reliever in the mouth (e.g. Oragel), as a throat lozenge (e.g. Cepacol), or as a skin cream (e.g. Lanacane).

2. Antiseizure medicines (anticonvulsants):

- a) Neurontin (gabapentin) and Lyrica (pregabalin) are usually the first choice for idiopathic and traumatic neuropathic pain, particularly achy, constant pain. People usually start with gabapentin because it is usually less expensive. These drugs are usually the second choice medicines for trigeminal neuralgia and glossopharyngeal neuralgia.
- b) Tegretol (carbamazepine) and Trileptal (oxcarbazepine) are the first choice medicines for Trigeminal Neuralgia and Glossopharyngeal neuralgia. They can also be effective for occipital neuralgia. (an intermittent sharp pain in the back of the head). Both medicines can cause some hyponatremia, particularly at higher doses. These medicines can also be useful if there is a "sharp" component to any type of neuropathic facial pain. Dosing is titrated not based on therapeutic seizure levels but rather on pain relief and the development of side effects.
- c) Topamax (topiramate) is another antiseizure medicine that may help with various craniofacial pain syndromes and headache syndromes.
- d) Depakote (sodium valproate) is another antiseizure medicine that may be used after other medicines have been tried. It can also help prevent migraine headaches.
- e) Lamictal (lamotrigine) is another antiseizure medicine that may be used after other medicines have been tried.
- f) Dilantin (phenytoin) is a useful medicine particularly for people who have responded to antiseizure medicines in the past and need an urgent escalation of antiseizure medicine with some other agent. For example, for patients with trigeminal neuralgia who are maxed out on carbamazepine and gabapentin and come into the Emergency Room with extreme uncontrolled pain, a quick

intravenous loading dose of Dilantin can often provide relief.

3. Anti-Depressants:

- a) Elavil (amitriptyline): This is a tricyclic antidepressant that is also one of the first choice medicines for chronic neuropathic pain. It is usually taken once a day, before bedtime. After gabapentin, Elavil is probably the second most common medicine given for refractory neuropathic pain.
- b) **Cymbalta** (duloxetine): This is a Serotonin and Norepinephrine reuptake inhibitor (SNRI). It can also be effective for alleviating neuropathic pain.
- c) Other anti-depressants can also be used, particularly for treating a patient's depression.

4. Anti-inflammatories:

Indocin (Indomethacin) is a prescription non-steroidal anti-inflammatory that is the treatment of choice for paroxysmal hemicrania. It may also be helpful for other types of refractory severe facial pain syndromes.

5. Prescription topical pain creams:

These topical creams often contain multiple ingredients, and are therefore often referred to as "compound pain creams". There is some controversy as to who exactly may benefit from these, but they are a low risk option to consider for patients who continue to suffer from significant pain.

6. Steroids:

Steroids can help reduce inflammation, and are the first choice for certain inflammatory facial pain conditions, such as temporal arteritis. However, long term use has significant side effects. A one week course of steroids, such as with a Medrol Dosepak (a tapering dose of methylprednisolone), may be worth trying for patients with

neuropathic facial pain to see if there is a response. If there is, a short term course of steroids can be used for flare-ups, or a long term use can be considered, but only at very low doses. A positive response to steroids might also suggest an alternate diagnosis.

7. Skeletal muscle relaxants:

Lioresal (baclofen), Flexiril (cyclobenzaprine), and Robaxin (methocarbamol). Sometimes, chronic pain syndromes have a significant component of muscle spasm or muscle related pain. For this reason, sometimes muscle relaxants can be helpful. These medicines are addictive, so they should only be used if needed, and ideally for short periods. If they are used for a prolonged period, patients would need to taper them off gradually if a time came when they were no longer necessary.

8. Benzodiazepines: Valium (diazepam), Klonopin(clonazepam), Xanax (alprazolam).

These medicines have sedative, antianxiety, and muscle relaxant properties. This category of medicine is also habit forming, and should be used only in very refractory cases of chronic pain. These medicines can help through muscle relaxation, as well as reduction of stressful feelings, tension, and anxiety that often contribute to a chronic pain condition. Again, ideally if these medicines are used at all, they would be used for a short period. Also, when coming off these medicines, a gradual tapering is needed.

9. Barbiturates: Butalbital.

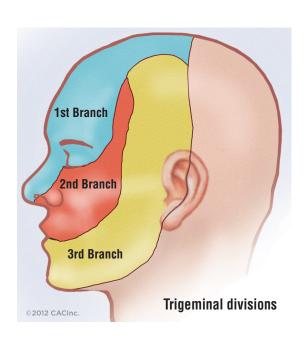
This is most often used as a component of the medicine Fiorecet (Bultalbital/acetaminophen/caffeine). Fiorecet is sometimes used for craniofacial pain syndromes. However, anything containing barbiturates can also be habit forming. Ideally this could be tried for just a short period if other options fail.

10. Marijuana / Cannabis:

While there are definitely downsides to the use of Cannabis, for some patients with chronic, refractory pain, this may be considered to try to make the pain less bothersome.

11. Opioids: (Tramadol, Oxycontin, Dilaudid, Nucynta).

Opioids are really the absolute last resort medicine for the management of chronic pain. They are highly addictive and subject to abuse. The preferred use of opioids for chronic neuropathic pain, if used at all, is for a rare severe "flare up" of the pain. Opioids are also a reasonable choice for severe neuropathic pain in patients with limited life expectancy, for example, with pain related to end stage cancer. Also, Opioids are often not an effective treatment for most people with chronic neuropathic pain. That having been said, it is possible that there is a small subgroup of people with chronic severe neuropathic pain that is refractory to all other measures, who might benefit from opioids. Patients who are being managed on long term opioids for chronic pain are usually under the care of a pain management specialist.



C) Procedures:

- **1. Acupuncture:** Some people find that this can be helpful, although multiple treatments are usually required.
- 2. Nerve Blocks: Sometimes people may experience some relief from various "nerve blocks". These are usually injections of numbing medicines, like lidocaine, sometimes mixed with steroids. If these do help, they are often short lived. They may need to be repeated, and it is possible, that sometimes these injections can "break the pain cycle". Sometimes, a nerve block may also convey useful information for treating the pain, that is, does the pain respond to temporarily blocking a particular nerve. This may be useful information in terms of other future treatments.
- 3. Botox injections: Botulinum toxin, when used in very tiny doses, in a very superficial manner, can cause temporary weakness of various muscles. Because some neuropathic pain is mediated by subtle muscle spasm or tension, Botox may therefore help with some chronic pain syndromes. It may have to be repeated every few months, as the effects are not permanent. Also, Botox may act to relieve chronic pain in other ways that aren't fully understood. Regardless, this is a low risk intervention that can help with certain craniofacial pain syndromes. It can take 1-2 weeks to give pain relief.
- 4. Peripheral neuro-stimularors: These are very small soft wire electrodes that can be placed under the skin in the superficial tissues to stimulate various peripheral nerves, such as the supraorbital nerve (the sensory nerve above the eye), the infraorbital nerve (the sensory nerve below the eye), and the occipital nerves (the sensory nerves in the back of the head). The implant takes only a few minutes to place, and is then attached to an external power supply and regulator for a few days that allows the patient to adjust the settings to see if the electrode is helping.

- If it does help, an electrode can then be placed on a more permanent basis and attached to a battery that is internalized under the collarbone, like a pacemaker battery. This is a minimally invasive procedure that can help some people with chronic severe neuropathic face pain. Patients who are candidates for this procedure have pain that has lasted at least 6 months and have failed multiple other treatments.
- stimulators that are place epidurally, just above the spinal cord. They are more commonly used for chronic pain in the legs or arms, but can be used for refractory face pain as well. For such a treatment, the stimulator is placed at the very top of the cervical spinal cord (as the very top of the spinal cord does contain trigeminal fibers that supply the face). First a trial stimulation would be performed, and if successful, a permanent implant could be placed. Again, this procedure would only be considered after other treatments had failed. This is also a procedure that we offer.

6. Hyperbaric Oxygen Therapy (HBOT):

This treatment involves exposing patients to high levels of oxygen under increased pressure in special chambers. The treatments usually are done over several weeks. The higher oxygen levels are thought to enhance healing of tissues and may have a role in treating chronic pain conditions. It is unclear whether certain patients with refractory facial pain might benefit from this or not.

7. Ketamine Injections: Ketamine is an anesthetic. When administered in low infusion doses it may help to reduce the pain associated with some chronic facial pain syndromes. Several outpatient treatments are required. There may be a role for this treatment in patients who have chronic facial pain.

Conclusions:

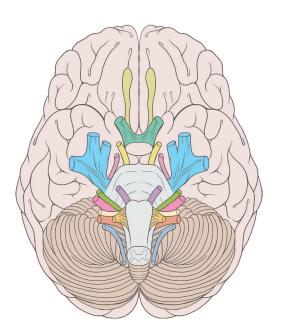
There are many options for treating neuropathic facial pain. These mostly involve medicines and non-surgical treatments. For refractory patients, there are some procedures that may offer relief. Treating physicians must be very patient and willing to try a host of treatments for these often difficult to manage cases. The best chance of success comes with a willingness to try different treatments, including multiple medicines at different doses, and treatments in different combinations. Multimodality efforts (using multiple treatments options) are usually more likely to succeed.

Treatments that we offer for specific facial pain syndromes:

- Comprehensive management of Trigeminal Neuralgia
- Comprehensive management of Glossopharyngeal Neuralgia

Treatments that we offer for other types of chronic neuropathic facial pain:

- Medicines
- Peripheral Nerve Stimulators
- Spinal Cord Stimulators



Facial Pain Syndromes can be complex and challenging to manage, and our role in the diagnosis and management of these disorders is complementary to the role of other specialists, including family practice doctors, internists, neurologists, ophthalmologists, otolaryngologists, pain management doctors, dentists, physiatrists, psychiatrists, and psychologists.



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Board certified by the American Board of Neurological Surgeons and a Fellow of the American College of Surgeons, Dr. Brisman specializes in the treatment of Trigeminal Neuralgia and Brain Tumors. He serves as the Co-Medical Director of the Long Island Gamma Knife Center at Mount Sinai South Nassau Hospital in Oceanside and has served as the Chief of Neurosurgery at NYU-Winthrop Hospital in Mineola. In



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Dr. Brisman Treats:

- Trigeminal Neuralgia
- Gliomas
- Brain Tumors
- Brain AVMS
- Pituitary Tumors
- Hemifacial Spasm
- Meningiomas
- Glossopharyngeal Neuralgia
- Acoustic Neuromas
- Skull Tumors
- Brain Metastases
- Chiari Malformation

Dr. Brisman is proficient in the use of minimally invasive neurosurgical procedures including:

- Stereotactic Radiosurgery
- Gamma Knife®
- CyberKnife®
- Novalis Tx®
- Microvascular Decompression
- Neuroendoscopy
- Transsphenoidal Surgery
- Stereotactic-guided Craniotomy
- Percutaneous Trigeminal Rhizotomy