

Trigeminal Neuralgia basic level

Overview

Trigeminal neuralgia, also known as tic douloureux, is an inflammation of the trigeminal nerve causing extreme pain and muscle spasms in the face. It usually occurs in adults and may affect those with multiple sclerosis. Although the exact cause of trigeminal neuralgia is not understood, several treatments can effectively relieve pain. Each treatment offers benefits, but each has limitations. You and your doctor should determine which treatment is best for you.

What is trigeminal neuralgia?

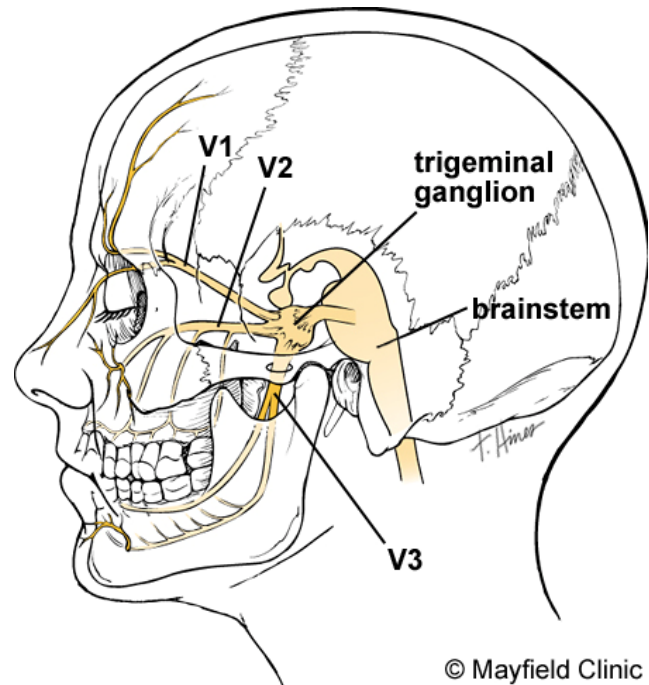
Neuralgia is severe pain caused by injury or damage to a nerve. Trigeminal neuralgia is a painful disorder of the fifth cranial nerve, called the trigeminal nerve. This nerve arises in the brain, supplying feeling and movement in the face.

Also called tic douloureux because of the uncontrollable facial twitching caused by the pain, trigeminal neuralgia is serious because it interferes with many aspects of a person's life. "Typical" trigeminal neuralgia involves brief instances of intense pain, like an electrical shock in one side of the face. This pain comes in repeated waves that last an hour or more. A less common form of the disorder, called "atypical" trigeminal neuralgia, causes a less intense, constant, dull burning or aching pain. This pain sometimes occurs with occasional electric shock-like stabs that may last a day or more.

What triggers a painful attack?

When the trigeminal nerve becomes irritated, an attack of intense pain results. Patients describe an attack as a "pins and needles" sensation that turns into a burning or jabbing pain, or as an electrical shock that may last a few seconds or minutes. In some cases of extreme pain, patients have even considered suicide. Everyday activities can trigger an episode. Some patients are sensitive in certain areas of the face, called trigger zones, which when touched cause an attack (Fig. 1). These zones are usually near the nose, lips, eyes, or ear.

Therefore, some patients avoid talking, eating, kissing, or drinking. Other simple activities, such as shaving or brushing teeth, can also trigger pain.



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Figure 1. The trigeminal nerve is both a sensory and motor nerve supplying feeling and movement to the face. It has three divisions that branch from the trigeminal ganglion: ophthalmic division (V1) provides sensation to the forehead and eye, maxillary division (V2) provides sensation to the cheek, and mandibular division (V3) provides sensation to the jaw.

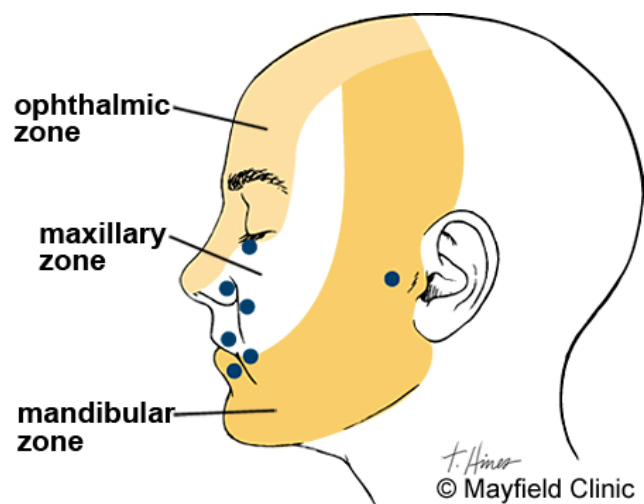


Figure 2. Facial areas of trigger zones. Trigger points (circles) have the greatest sensitivity.

The pain of trigeminal neuralgia usually has the following features:

- 1) Typically affects one side of the face
- 2) Can last several days or weeks, followed by a remission for months or years
- 3) Frequency of painful attacks increases over time and may become disabling

What are the causes?

Many believe that the protective sheath of the trigeminal nerve deteriorates, sending abnormal messages along the nerve. Like static in a telephone line, these abnormalities disrupt the normal signal of the nerve and cause pain. Several factors can cause the deterioration of this protective sheath: aging, multiple sclerosis, tumors, but most doctors agree that it is caused by an abnormal vein or artery that compresses the nerve.

Some types of facial pain can result from an infected tooth, sinus infections, or previous nerve injury. Because the causes of pain vary, each patient should undergo a medical evaluation.

Who is affected?

Trigeminal neuralgia affects 1 in every 25,000 people, and occurs slightly more in women than men. Patients are usually middle age and older. Some people with multiple sclerosis also develop trigeminal neuralgia.

How is a diagnosis made?

When a person first experiences facial pain, the primary care doctor or dentist is often consulted. If the pain requires further evaluation, a consultation with a neurologist or a neurosurgeon may be recommended.

Few causes of trigeminal neuralgia are serious. However, the possibility of a tumor or multiple sclerosis must be ruled out. Therefore, the doctor or neurologist will prescribe an imaging study, such as a computed tomography (CT) or magnetic resonance imaging (MRI) scan. The diagnosis of trigeminal neuralgia is made after carefully assessing the patient's symptoms.

What treatments are available?

A variety of treatments are available, including medications, surgical treatments, and radiosurgery. The first treatment option is usually medical.

Medications

Analgesics such as aspirin and ibuprofen are generally not effective against trigeminal neuralgia.

Anticonvulsants, such as carbamazepine (Tegretol), phenytoin (Dilantin), gabapentin (Neurontin), lamotrigine (Lamictal), oxcarbazepine (Trileptal), and pregabalin (Lyrica) are commonly used because they block firing of the nerve. These medications

are initially effective for pain control in 90% of patients. These drugs can cause side effects (e.g., drowsiness, unsteadiness, nausea, skin rash, blood disorders). Therefore, patients are monitored routinely and undergo blood tests to ensure that the drug levels remain safe and that the patient doesn't develop blood disorders. Medications are used as long as the pain is controlled and the side effects do not interfere with a patient's activities. When medication is no longer effective, surgical procedures may be considered. Approximately 25% of patients achieve long-term relief with medications.

Muscle relaxants such as baclofen (Lioresal) are sometimes effective in treating trigeminal neuralgia. Sometimes multiple drug therapy is necessary to control pain (e.g., Tegretol and Neurontin).

Surgical procedures

Medications sometimes fail to control pain or cause side effects. When this happens, the neurologist may suggest that the patient consult a neurosurgeon, who may recommend one of several surgical procedures. No one surgical procedure is best for everyone and each procedure varies in its effectiveness versus side effects (see Comparison of Treatment Results for Facial Pain). Among the current treatment options, microvascular decompression (MVD) and percutaneous stereotactic radiofrequency rhizotomy (PSR) have comparable rates of pain relief that are highest among the available options. In a review of series of approximately 100 patients or more published in the past 10 years, the rates of pain relief calculated were 77% in 7 years for MVD and 75% in 6 years for PSR rhizotomy.

- **Percutaneous stereotactic radiofrequency rhizotomy (PSR)**, also known as electrocoagulation, produces a heating current to destroy some of the trigeminal nerve fibers that produce pain. PSR eliminates pain but also causes partial numbness of the face. This outpatient procedure is performed under local anesthesia and sedation. While the patient is awake, an electrode is passed through the cheek to stimulate the nerve and locate the pain-causing area. The patient is then anesthetized as the surgeon destroys the portion of the nerve that carries the pain.

PSR is effective because it provides the most lasting pain relief by destroying part of the trigeminal nerve. PSR provides immediate pain relief for 99% of patients. Of the 15% of patients whose symptoms recur within 10 years, medication, repeat PSR, or another surgical procedure will again be considered. PSR can cause minor complications, such as double vision or weakness of the jaw. Partial numbness in the area where the pain existed is a normal

side effect. Other complications, such as blurred vision or chewing problems, are usually temporary (see Percutaneous Stereotactic Radiofrequency Rhizotomy).

- **Percutaneous glycerol rhizolysis** is similar to PSR in that a needle is passed through the cheek to the nerve. However, it uses an injection of glycerol instead of a heating current to damage some of the trigeminal nerve fibers that produce pain. This outpatient procedure is performed under local anesthesia and sedation. The surgeon injects glycerol to damage the portion of the nerve that carries the pain. Because the location of the glycerol cannot be controlled precisely after injection, the results are somewhat unpredictable. As with PSR, partial numbness of the face is expected. Other side effects are similar to PSR. After undergoing percutaneous glycerol rhizotomy, about 70% of patients have immediate pain relief. In about 50% of patients, symptoms recur within 3 to 4 years.
- **Percutaneous balloon compression** is similar to PSR and PGR in that a needle is passed through the cheek to the nerve. The surgeon places a balloon in the trigeminal nerve through a catheter. The balloon is inflated where fibers produce pain. The balloon compresses the nerve, injuring the pain-causing fibers. After several minutes the balloon and catheter are removed.

Complications can be minor numbness, eye infection, chewing problems, or double vision. The procedure provides immediate pain relief for 80% of patients. In about 20% of patients who undergo balloon compression, symptoms recur within 3 years.

- **Microvascular decompression (MVD)** is the only surgical procedure that may actually preserve facial sensation, but is the most invasive. While the patient is under general anesthesia, the surgeon makes a 1 inch circular opening at the back of the skull called a craniotomy. This opening exposes the trigeminal nerve at its connection with the brain. A blood vessel (occasionally a tumor or other abnormality) is often found that compresses the nerve. After the nerve is freed from compression, it is protected with a small sponge.

This procedure is effective for 95% of patients. The major benefit of MVD is that it causes little or no facial numbness. After MVD, 20% of patients have pain recur within 10 years. Major disadvantages are the risks of anesthesia and of undergoing an operation near the brain.

- **Neurectomy** is the irreversible cutting of the trigeminal nerve or one of its branches. Cutting the supraorbital nerve, which supplies sensation to the forehead, may be advised if pain is isolated to the area above the eyebrow. Cutting the infraorbital nerve may be performed if pain is limited to the area below the eye along the upper cheekbone. Cutting the nerve causes permanent numbness of the region that the nerve supplies and should only be considered when all other treatments have failed to control pain. Also, a neurectomy may be performed during an MVD procedure if no vessel is found compressing the nerve. In this case, a portion of the trigeminal nerve root is cut at its connection with the brain.

radiosurgery

Stereotactic radiosurgery is a noninvasive outpatient procedure that uses radiation beams to destroy some of the trigeminal nerve. A metal frame is attached to the patient's head by four pins. While wearing the frame the patient is scanned in the MRI. Using the frame as reference points the exact location of the trigeminal nerve is determined three dimensionally in a computer. Surgeons then use highly focused beams of radiation to damage a site on the trigeminal nerve. In the weeks following the treatment, a lesion (injury) gradually develops where the radiation occurred.

Currently there are two types of radiosurgery technologies: the LINAC and Gamma Knife. The Gamma Knife, a hemispheric device that surrounds the patient's head, targets 201 beams of cobalt-60 radiation on a tiny area of the trigeminal nerve. Each beam travels through a different area of the brain. By itself, it is weak enough not to harm the tissue it passes through. The beams overlap in the target area and where they intersect, they create a lesion. The other type, LINAC, is a typical linear accelerator that rotates around the patient, producing multiple beams of radiation. Each arcs toward the target on the trigeminal nerve through different tissue. Where the beams converge, they cause slight damage.

Complications of this procedure are rare. Facial numbness may develop in 10% of patients. Pain relief may not occur immediately but rather gradually over six months to a year. Patients remain on medication for a period of time following treatment to control the pain while the radiation takes effect. Seventy percent of patients are relieved of pain for 2 years.

Holistic therapies

Some patients want to try holistic therapies such as acupuncture and biofeedback. The effectiveness of these treatments for trigeminal neuralgia is being studied.

Clinical Trials

Clinical trials are research studies in which new treatments - drugs, diagnostics, procedures, vaccines, and other therapies - are tested in people to see if they are safe and effective. Research is always being conducted to improve the standard of medical care and explore new drug and surgical treatments. You can find information about current clinical investigations, including their eligibility requirements, protocol, and participating locations on the web: the National Institutes of Health (NIH) at clinicaltrials.gov, sponsors many trials; private industry and pharmaceutical companies also sponsor trials <http://www.centerwatch.com/>

Sources & Links

If you have more questions, please contact the Mayfield Clinic at 800-325-7787 or 513-221-1100.

Links

Trigeminal Neuralgia Association (TNA),
www.fpa-support.org
American Pain Society, www.ampainsoc.org
Facial Neuralgia Resources, facial-neuralgia.org

Support

Through the Trigeminal Neuralgia Association, local support groups are available. The support group provides an opportunity for patients and their families to share experiences, receive support, and learn about advances in treatments, pain control, and medications. If you would like information about the Greater Cincinnati Trigeminal Neuralgia Support Group, please call the Mayfield Clinic at (513) 475-8646. For support outside Greater Cincinnati, please contact the Trigeminal Neuralgia Association at 800-923-3608.

References

- 1) Taha JM, Tew JM Jr: Comparison of surgical treatments for trigeminal neuralgia: Reevaluation of radiofrequency rhizotomy. *Neurosurgery* 38:865-871, 1996.

Glossary

acupuncture: an ancient Oriental system of therapy that can be used to control pain.

anticonvulsant: a drug that stops or prevents convulsions or seizures. **biofeedback:** a technique of learning how to control some body functions by monitoring brain waves, muscle tension, etc.

glycerol: a sweet, oily fluid that can be injected into a nerve to destroy its pain-producing portion.

multiple sclerosis: a chronic degenerative disease of the central nervous system in which the myelin (sheath) surrounding the nerves is destroyed.

neuralgia: nerve pain.

percutaneous: through the skin

rhizolysis: cutting or destroying of a group of cells (e.g., nerve cells). **percutaneous:** by way of the skin (e.g, injection).

tic douloureux: French for trigeminal neuralgia.

trigeminal nerve: the fifth cranial nerve.

trigger zones: small areas that produce intense pain.

updated > 4.2009

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