

# Trigeminal Neuralgia

Trigeminal neuralgia or tic douloureux is sometimes described as the most excruciating pain known to humanity. The pain typically involves the lower face and jaw though sometimes it occurs around the nose and above the eye. This intense, stabbing, electric shock-like pain is caused by irritation of the trigeminal nerve, which sends branches to the forehead, cheek, and lower jaw.

Although trigeminal neuralgia cannot always be cured, there are treatments available to alleviate the excruciating pain. Anti-convulsive medications are normally the first therapeutic choice. Surgery can be an effective option for those who become unresponsive to medications or those who suffer serious side effects from the medications.

## The Trigeminal Nerve

The trigeminal nerve is one of 12 pairs of cranial nerves in the head. It is the nerve responsible for providing sensation to the face. One trigeminal nerve runs to the right side of the head and the other to the left. Each of these nerves has three distinct branches. ("Trigeminal" derives from the Latin word "tria," which means three, and "geminus," which means twin.) The first branch controls sensation in the eye, forehead, and nose; the second governs the lower eyelid, side of the nose, upper teeth, gums, lip, and cheek; and the third supplies the lower teeth, gum, lip, and jaw. Trigeminal nerve pain can involve one or more of these branches.

## Causes of Trigeminal Neuralgia

The pain associated with trigeminal neuralgia represents an irritation of the nerve. The most frequent cause is a blood vessel (usually an artery but sometimes a vein) pressing on the nerve near its exit from the brain stem. Trigeminal neuralgia usually strikes people older than 50. Over time, changes in the blood vessels of the brain can result in a redundant loop of vessel rubbing against the trigeminal nerve root. The constant rubbing with each heartbeat wears away the insulating membrane of the nerve. This causes the nerve to be irritated. Even light stimulation of the face can lead to bouts of intense "electrical" pain.

Most patients report that their pain emerged spontaneously out of nowhere one day. Other patients say that their pain followed a car accident, a blow to the face or dental surgery. Most physicians and dentists do not believe that dental work can cause trigeminal neuralgia. Instead, the disorder likely was already developing, and the dental work brought on the initial symptoms coincidentally.

Patients often first experience pain along the upper or lower jaw and hence assume they have a dental abscess. Many visit a dentist and undergo a root canal and other procedures. When the pain persists, they finally realize the problem is not dental.

The pain of trigeminal neuralgia is defined as either "classic" or atypical. With classic pain, there are definite periods of remission. The pain is intensely sharp, throbbing and shock-like. Atypical pain is more of a sharp, stabbing pain as well, but patients often describe it as burning, crushing, or pulsating. With atypical trigeminal neuralgia, some patients do not have remission, and symptoms are more difficult to treat than the classic variant.

Trigeminal neuralgia tends to run in cycles. Patients often suffer long stretches of frequent attacks followed by weeks, months or even years of little or no pain. Even so, the usual pattern is for the attacks to intensify over time with shorter pain-free periods. Attacks for both kinds of trigeminal neuralgia are often triggered by a light touch of the skin, washing, shaving, brushing the teeth, blowing the nose, drinking hot or cold drinks, a light breeze and even smiling. Some patients suffer less than one attack a day while others experience a dozen or more every hour. The pain typically begins with a sensation of electrical shocks that culminates in less than 20 seconds with an excruciating stabbing pain. The pain often leaves patients grimacing, hence the nickname for the disorder, tic douloureux or just plain tic.

The pain is intermittent because the irritation to the nerve varies with the length of damaged nerve. Under the microscope, the trigeminal nerve shows signs of dying. Also observable may be dead axons (the part of the nerve fiber that transfers impulses) and malformed or missing myelin (the nerve's protective coating). After a mild stimulus such as washing or shaving, the naked axons, which end up touching one another, cause the abnormal episode of firing of the nerve that can be compared to a short-circuited electrical wire.

Other causes of trigeminal neuralgia include pressure of a tumor on the nerve or multiple sclerosis, which damages the myelin sheaths. A high quality MRI of the brain can usually detect when trigeminal neuralgia is caused either by a tumor or multiple sclerosis. However, unless a tumor or multiple sclerosis is the cause, imaging of the brain will seldom reveal the precise reason why the nerve is being irritated. The vessel abutting the nerve root is difficult to see even on a high quality MRI.

### Susceptibility

Advanced age is a major risk factor for trigeminal neuralgia. The age of onset typically is the 60s or 70s, though younger people can suffer from the disorder, too. Women are 1.5 times more likely than men to have the disorder. Hypertension and multiple sclerosis are also risk factors. Trigeminal neuralgia is relatively rare. An estimated 40,000 people in the United States suffer from trigeminal neuralgia.

To a certain extent, trigeminal neuralgia runs in families. About five percent of patients have a family history of it. Presumably, this is because we inherit our parents' blood vessels as well as other physical features.

### Diagnosis

A high quality MRI can detect if a tumor or multiple sclerosis is irritating the trigeminal nerve. Otherwise, no test can definitively determine the presence of trigeminal neuralgia. Tests can help rule out other causes of facial disorders. Trigeminal neuralgia is usually diagnosed based on the description of the symptoms provided by the patient.

The medication used for trigeminal neuralgia normally brings quick relief, and the medication itself can be crucial in making a diagnosis. When a patient shows no relief from the medication, a physician has cause to doubt whether trigeminal neuralgia is present.

The symptoms of several pain disorders are similar to those of trigeminal neuralgia. Temporal tendinitis involves cheek pain and tooth sensitivity as well as headaches and neck and shoulder pain. Ernest syndrome refers to the injury of the styomandibular ligament, which connects the

base of the skull with the lower jaw. The injury produces pain in regions of the face, head and neck. Finally, occipital neuralgia involves pain in the front and back of the head (the occipital areas) that sometimes extends into the facial region.

## Treatment

Years ago trigeminal neuralgia was not well understood and treatment was nearly non-existent. Some patients, unable to withstand the pain, succumbed to despair and suicide. Today, there are several effective ways to alleviate the pain.

The first line of treatment is usually an anti-convulsant drug such as Tegretol (carbamazepine) that slows the function of the irritated nerve and consequently relieves the pain. Many patients find significant or total relief from pain with these drugs. The drugs can have several drawbacks, however. Some patients may need a relatively high dose to alleviate the pain and the side effects at high doses include blurred vision, drowsiness, memory problems and balance problems. Secondly, the anticonvulsant may lose its efficacy over time. Patients may need a higher dose to reduce the pain or may need a second anticonvulsant, which can lead to adverse drug reactions.

The drugs can have a toxic effect on some patients, particularly people with a history of bone marrow suppression and kidney and liver toxicity. Such patients must have their blood monitored to ensure their safety.

Patients for whom drugs aren't effective can opt for surgery. Twenty-five to 50 percent of patients will eventually stop responding to drug therapy and may need surgery. Fortunately, about 85 percent of patients who undergo surgery experience significant pain relief. Several types of surgical procedures are available. The two most common are microvascular decompression and percutaneous neurolysis.

Vascular decompression provides many years of pain relief but, as a more invasive procedure, carries more risks. Percutaneous neurolysis with mechanical radiofrequency or chemical lesioning can produce numbness of the face and brief symptom relief. These techniques are preferable for older patients and those whose health problems argue against an invasive surgery and general anesthesia.

Vascular decompression relieves trigeminal neuralgia by placing a small pad between the nerve and the adjoining blood vessels. A neurosurgeon makes an incision in the back of the head to expose the trigeminal nerve at the base of the brain. The pad is inserted between the nerve and the blood vessels to alleviate compression. Pain relief is realized in about 90 percent of patients.

There is a small risk of facial numbness, facial weakness, double vision, infection or deficiencies in hearing or balance. The operation requires a general anesthetic and about a five-day stay in the hospital following the procedure. Most patients need to take two weeks off from work to recover.

Vascular decompression generally provides long-term pain relief. It can be repeated if necessary. Often, as with other types of surgical treatments for trigeminal neuralgia, many patients will continue to require some pain medication but usually at a lower dose.

Percutaneous techniques are performed with a needle passed through the skin. The needle is directed by X-ray control. The trigeminal nerve is "lysed" or lesioned through the needle in one or several ways. Under x-ray control, a needle is passed into the cheek on the side of the face where the patient feels pain and through a small, natural opening in the base of the skull (the

foramen ovale) into the trigeminal nerve. Radiofrequency energy can be applied to burn a portion of the nerve. A small balloon can be used to compress the nerve. Or lastly, a chemical such as glycerol can be injected into the nerve to damage the nerve.

Relief of trigeminal neuralgia through percutaneous techniques results from making the region of the pain permanently numb. The procedure takes less than an hour and patients can usually go home the same day. Most can return to work in a day or two.

An undesirable side effect is that the numbness of the face may produce an unpleasant sensation such as a "tingling" or "stiffening." Other side effects are chewing muscle weakness and, if the pain had been near the eye and treatment targeted that area, numbness of the eye. That in itself is not harmful but such patients cannot feel a foreign body in their eye and can develop serious corneal abrasions. They must inspect the eye in the mirror regularly to ensure that there is no irritation.

Patients who undergo percutaneous treatment can expect to have relief from pain for three to four years after which the procedure can be repeated. This technique is well tolerated even by patients who are 80 or older. Many surgeons will refrain from performing a microvascular decompression for people over 70, though the general health of the patient takes precedent over actual age in choosing a surgical treatment.

Both surgical procedures are more effective when used in the early stages of the disease. Surgical treatment also produces better results with classic trigeminal neuralgia than with atypical trigeminal neuralgia.

#### Role of Neurosurgeons

Neurosurgeons are medical specialists trained to help patients suffering from trigeminal neuralgia. Neurosurgeons provide the operative and non-operative (prevention, diagnosis, evaluation, treatment, critical care and rehabilitation) care of neurological disorders. Neurosurgeons undergo six to eight years of specialized training following medical school, one of the longest training periods of any medical specialties.