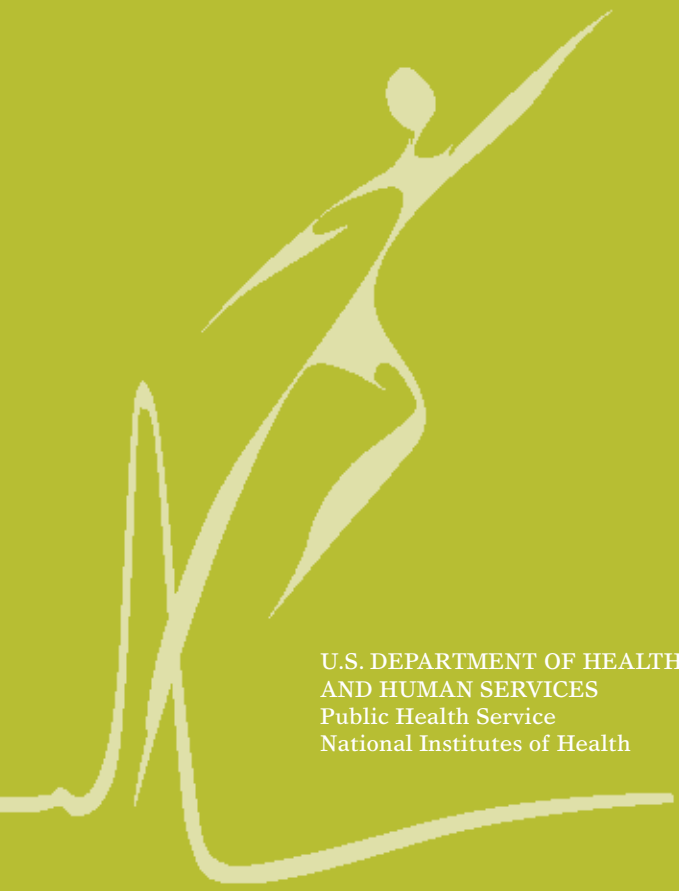
Trigeminal Neuralgia



U.S. DEPARTMENT OF HEALTH
AND HUMAN SERVICES
Public Health Service
National Institutes of Health

Trigeminal Neuralgia

What is trigeminal neuralgia?

Trigeminal neuralgia (TN), also called *tic douloureux*, is a chronic pain condition that affects the trigeminal or 5th cranial nerve, one of the most widely distributed nerves in the head. TN is a form of neuropathic pain (pain associated with nerve injury or nerve lesion.) The typical or "classic" form of the disorder (called "Type 1" or TN1) causes extreme, sporadic, sudden burning or shock-like facial pain that lasts anywhere from a few seconds to as long as two minutes per episode. These attacks can occur in quick succession, in volleys lasting as long as two hours. The "atypical" form of the disorder (called "Type 2" or TN2), is characterized by constant aching, burning, stabbing pain of somewhat lower intensity than Type 1. Both forms of pain may occur in the same person, sometimes at the same time. The intensity of pain can be physically and mentally incapacitating.

The trigeminal nerve is one of 12 pairs of nerves that are attached to the brain. The nerve has three branches that conduct sensations from the upper, middle, and lower portions of the face, as well as the oral cavity, to the brain. The ophthalmic, or upper, branch supplies sensation to most of the scalp, forehead, and front of the head. The maxillary, or middle,

branch stimulates the cheek, upper jaw, top lip, teeth and gums, and to the side of the nose. The mandibular, or lower, branch supplies nerves to the lower jaw, teeth and gums, and bottom lip. More than one nerve branch can be affected by the disorder. Rarely, both sides of the face may be affected at different times in an individual, or even more rarely at the same time (called bilateral TN).

What causes trigeminal neuralgia?

TN is associated with a variety of conditions. TN can be caused by a blood vessel pressing on the trigeminal nerve as it exits the brain stem. This compression causes the wearing away or damage to the protective coating around the nerve (the myelin sheath). TN symptoms can also occur in people with multiple sclerosis, a disease that causes deterioration of the trigeminal nerve's myelin sheath. Rarely, symptoms of TN may be caused by nerve compression from a tumor, or a tangle of arteries and veins called an arteriovenous malformation. Injury to the trigeminal nerve (perhaps the result of sinus surgery, oral surgery, stroke, or facial trauma) may also produce neuropathic facial pain.

What are the symptoms of trigeminal neuralgia?

Pain varies, depending on the type of TN, and may range from sudden, severe, and stabbing to a more constant, aching, burning sensation. The intense flashes of pain can be triggered by vibration or contact with the cheek (such as when shaving, washing the face, or applying makeup), brushing teeth, eating,

drinking, talking, or being exposed to the wind. The pain may affect a small area of the face or may spread. Bouts of pain rarely occur at night, when the affected individual is sleeping.

TN is typified by attacks that stop for a period of time and then return, but the condition can be progressive. The attacks often worsen over time, with fewer and shorter pain-free periods before they recur. Eventually, the pain-free intervals disappear and medication to control the pain becomes less effective. The disorder is not fatal, but can be debilitating. Due to the intensity of the pain, some individuals may avoid daily activities or social contacts because they fear an impending attack.

Who is affected?

Trigeminal neuralgia occurs most often in people over age 50, although it can occur at any age, including infancy. The possibility of TN being caused by multiple sclerosis increases when it occurs in young adults. The incidence of new cases is approximately 12 per 100,000 people per year; the disorder is more common in women than in men.

How is TN diagnosed?

TN diagnosis is based primarily on the person's history and description of symptoms, along with results from physical and neurological examinations. Other disorders that cause facial pain should be ruled out before TN is diagnosed. Some disorders that cause facial pain include post-herpetic neuralgia (nerve pain following an outbreak of shingles), cluster headaches, and

temporomandibular joint disorder (TMJ, which causes pain and dysfunction in the jaw joint and muscles that control jaw movement). Because of overlapping symptoms and the large number of conditions that can cause facial pain, obtaining a correct diagnosis is difficult, but finding the cause of the pain is important as the treatments for different types of pain may differ.

Most people with TN eventually will undergo a magnetic resonance imaging (MRI) scan to rule out a tumor or multiple sclerosis as the cause of their pain. This scan may or may not clearly show a blood vessel compressing the nerve. Special MRI imaging procedures can reveal the presence and severity of compression of the nerve by a blood vessel.

A diagnosis of classic trigeminal neuralgia may be supported by an individual's positive response to a short course of an antiseizure medication. Diagnosis of TN2 is more complex and difficult, but tends to be supported by a positive response to low doses of tricyclic antidepressant medications (such as amitriptyline and nortriptyline), similar to other neuropathic pain diagnoses.

How is trigeminal neuralgia treated?

Treatment options include medicines, surgery, and complementary approaches.

Medications

Anticonvulsant medicines—used to block nerve firing—are generally effective in treating TN1 but often less effective in TN2. These drugs include carbamazepine, oxcarbazepine,

topiramate, gabapentin, pregabalin, clonazepam, phenytoin, lamotrigine, and valproic acid.

Tricyclic antidepressants such as amitriptyline or nortriptyline can be used to treat pain. Common analgesics and opioids are not usually helpful in treating the sharp, recurring pain caused by TN1, although some individuals with TN2 do respond to opioids. Eventually, if medication fails to relieve pain or produces intolerable side effects such as cognitive disturbances, memory loss, excess fatigue, bone marrow suppression, or allergy, then surgical treatment may be indicated. Since TN is a progressive disorder that often becomes resistant to medication over time, individuals often seek surgical treatment.

Surgery

Several neurosurgical procedures are available to treat TN, depending on the nature of the pain; the individual's preference, physical health, blood pressure, and previous surgeries; presence of multiple sclerosis, and the distribution of trigeminal nerve involvement (particularly when the upper/ophthalmic branch is involved). Some procedures are done on an outpatient basis, while others may involve a more complex operation that is performed under general anesthesia. Some degree of facial numbness is expected after many of these procedures, and TN will often return even if the procedure is initially successful. Depending on the procedure, other surgical risks include hearing loss, balance problems, leaking of the cerebrospinal fluid (the fluid that bathes the brain and spinal cord), infection, anesthesia dolorosa (a combination

of surface numbness and deep burning pain), and stroke, although the latter is rare.

A *rhizotomy* (rhizolysis) is a procedure in which nerve fibers are damaged to block pain. A rhizotomy for TN always causes some degree of sensory loss and facial numbness. Several forms of rhizotomy are available to treat trigeminal neuralgia:

- *Balloon compression* works by injuring the insulation on nerves that are involved with the sensation of light touch on the face. The procedure is performed in an operating room under general anesthesia. A tube called a cannula is inserted through the cheek and guided to where one branch of the trigeminal nerve passes through the base of the skull. A soft catheter with a balloon tip is threaded through the cannula and the balloon is inflated to squeeze part of the nerve against the hard edge of the brain covering (the dura) and the skull. After about a minute the balloon is deflated and removed, along with the catheter and cannula. Balloon compression is generally an outpatient procedure, although sometimes the patient may be kept in the hospital overnight. Pain relief usually lasts one to two years.
- *Glycerol injection* is also generally an outpatient procedure in which the individual is sedated with intravenous medication. A thin needle is passed through the cheek, next to the mouth, and guided through the opening in the base of the skull where the third division of the trigeminal nerve (mandibular) exits. The needle is moved into the pocket of spinal fluid (cistern) that

surrounds the trigeminal nerve center (or ganglion, the central part of the nerve from which the nerve impulses are transmitted to the brain). The procedure is performed with the person sitting up, since glycerol is heavier than spinal fluid and will then remain in the spinal fluid around the ganglion. The glycerol injection bathes the ganglion and damages the insulation of trigeminal nerve fibers. This form of rhizotomy is likely to result in recurrence of pain within a year to two years. However, the procedure can be repeated multiple times.

- *Radiofrequency thermal lesioning* (also known as "RF Ablation" or "RF Lesion") is most often performed on an outpatient basis. The individual is anesthetized and a hollow needle is passed through the cheek through the same opening at the base of the skull where the balloon compression and glycerol injections are performed. The individual is briefly awakened and a small electrical current is passed through the needle, causing tingling in the area of the nerve where the needle tips rests. When the needle is positioned so that the tingling occurs in the area of TN pain, the person is then sedated and the nerve area is gradually heated with an electrode, injuring the nerve fibers. The electrode and needle are then removed and the person is awakened. The procedure can be repeated until the desired amount of sensory loss is obtained; usually a blunting of sharp sensation, with preservation of touch. Approximately half of the people have symptoms that reoccur three to four years following RF

lesioning. Production of more numbness can extend the pain relief even longer, but the risks of anesthesia dolorosa also increase.

- *Stereotactic radiosurgery* (Gamma Knife, Cyber Knife) uses computer imaging to direct highly focused beams of radiation at the site where the trigeminal nerve exits the brain stem. This causes the slow formation of a lesion on the nerve that disrupts the transmission of sensory signals to the brain. People usually leave the hospital the same day or the next day following treatment but won't typically experience relief from pain for several weeks (or sometimes several months) following the procedure. The International RadioSurgery Association reports that between 50 and 78 percent of people with TN who are treated with Gamma Knife radiosurgery experience "excellent" pain relief within a few weeks following the procedure. For individuals who were treated successfully, almost half have recurrence of pain within three years.
- *Microvascular decompression* (MVD) is the most invasive of all surgeries for TN, but also offers the lowest probability that pain will return. About half of individuals undergoing MVD for TN will experience recurrent pain within 12 to 15 years. This inpatient procedure, which is performed under general anesthesia, requires that a small opening be made through the mastoid bone behind the ear. While viewing the trigeminal nerve through a microscope or endoscope, the surgeon moves away the vessel (usually an artery) that is compressing

the nerve and places a soft cushion between the nerve and the vessel. Unlike rhizotomies, the goal is not to produce numbness in the face after this surgery. Individuals generally recuperate for several days in the hospital following the procedure, and will generally need to recover for several weeks after the procedure.

A *neurectomy* (also called partial nerve section), which involves cutting part of the nerve, may be performed near the entrance point of the nerve at the brain stem during an attempted microvascular decompression if no vessel is found to be pressing on the trigeminal nerve. Neurectomies also may be performed by cutting superficial branches of the trigeminal nerve in the face. When done during microvascular decompression, a neurectomy will cause more long-lasting numbness in the area of the face that is supplied by the nerve or nerve branch that is cut. However, when the operation is performed in the face, the nerve may grow back and in time sensation may return. With neurectomy, there is risk of creating anesthesia dolorosa.

Surgical treatment for TN2 is usually more problematic than for TN1, particularly where vascular compression is not detected in brain imaging prior to a proposed procedure. Many neurosurgeons advise against the use of MVD or rhizotomy in individuals for whom TN2 symptoms predominate over TN1, unless vascular compression has been confirmed. MVD for TN2 is also less successful than for TN1.

Complementary approaches

Some individuals manage trigeminal neuralgia using complementary techniques, usually in combination with drug treatment. These therapies offer varying degrees of success. Some people find that low-impact exercise, yoga, creative visualization, aroma therapy, or meditation may be useful in promoting well-being. Other options include acupuncture, upper cervical chiropractic, biofeedback, vitamin therapy, and nutritional therapy. Some people report modest pain relief after injections of botulinum toxin to block activity of sensory nerves.

Chronic pain from TN is frequently very isolating and depressing for the individual. Conversely, depression and sleep disturbance may render individuals more vulnerable to pain and suffering. Some individuals benefit from supportive counseling or therapy by a psychiatrist or psychologist. However, there is no evidence that TN is psychogenic in origin or caused by depression, and persons with TN require effective medical or surgical treatment for their pain.

What research is being done?

The National Institute of Neurological Disorders and Stroke (NINDS), a part of the National Institutes of Health, is the federal government's leading supporter of biomedical research on disorders of the brain and nervous system. NINDS-funded projects are exploring the mechanisms involved with chronic pain and trigeminal neuralgia, as well as novel diagnostic methods and treatments. Other research addresses

TN through studies associated with pain research. Additional NIH research on TN is being funded by the National Institute of Dental and Craniofacial Research.

One NINDS-funded study for people with post-herpetic neuralgia of the trigeminal nerve uses a nasal spray applicator to deliver a drug to the tissue that lines the nasal cavity (nasal mucosa) Current drug therapy is absorbed through the body, which may lead to adverse effects such as drug interactions. The local drug delivery affects nerve endings and suppresses the activity of neurotransmitters (which help cells communicate with each other), which makes the trigeminal nerve less able to transmit pain. The study will monitor people's daily assessment of overall pain and note any adverse effects.

Little is known about how the nervous system becomes closely aligned with the vascular system during development. Scientists are using a mouse model to understand this interaction, which may lead to better diagnosis, therapy, and prevention of several neurological diseases, including diabetic neuropathy and TN.

Women are at a greater risk for pain in many acute and chronic pain conditions (including TN), but the reasons behind this aren't well understood. Researchers are looking at the role estrogens may play in affecting nerve pain activity. Understanding estrogen activity on pain nerves may increase the knowledge of why women are at risk for pain and possibly lead to the development of compounds that dampen the activity of estrogen on nerves that send pain signals to the brain and spinal cord.

Where can I go for more information?

For more information on neurological disorders or research programs funded by the National Institute of Neurological Disorders and Stroke, contact the Institute's Brain Resources and Information Network (BRAIN) at:

BRAIN
P.O. Box 5801
Bethesda, MD 20824
800-352-9424
www.ninds.nih.gov

For more information on disorders or research programs funded by the National Institute of Dental and Craniofacial Research, contact:

National Institute of Dental and Craniofacial Research
National Institutes of Health, DHHS
31 Center Drive, Room 5B55
Bethesda, MD 20892-2190
301-496-4261
800-232-4528
www.nidcr.nih.gov

Information on trigeminal neuralgia also is available from the following organizations:

TNA – The Facial Pain Association
408 W. University Avenue, Suite 602
Gainesville, FL 32601
352-384-3600
800-923-3608
www.fpa-support.org/

International RadioSurgery Association
P.O. Box 5186
Harrisburg, PA 17710
717-260-9808
www.irsa.org

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Prepared by:
Office of Communications and Public Liaison
National Institute of Neurological
Disorders and Stroke
National Institutes of Health
Department of Health and Human Services
Bethesda, Maryland 20892 2540

NIH Publication No. 13 5116 August 2013