

SPECIAL POINTS OF INTEREST:

- Trigeminal neuralgia is a type of severe facial pain
- The cause is debated, but often seems to be caused by a blood vessel irritating the trigeminal nerve near the brain.
- The treatment is initially with medication.
- If medication fails there are a number of neurosurgical procedures which can usually give good relief

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Trigeminal Neuralgia

VERSION 1.3

JUNE 2017

What is Trigeminal Neuralgia?

Trigeminal neuralgia (also sometimes known as *Tic Doloureaux*) is a particular type of very severe facial pain. The pain is in the distribution of part of the trigeminal nerve, which is the nerve that supplies feeling to the face and front of the scalp and gums (see trigeminal nerve anatomy below). The pain does not cross the midline.

Typically the pain is lancinating, which means it has a sudden onset, and is usually relatively brief, lasting a few seconds (although in severe cases it may occur again almost immediately, again and again). Sufferers often describe the pain like a lightening-bolt, or stabbing pain, or sometimes as burning, or like a surge of electricity in that part of the face. Repeated attacks

are usually felt in the same area of the face each time. Usually



the patient is pain-free between each attack, although rarely they may have an underlying dull ache between attacks. The pain is usually extreme, and untreated trigeminal neuralgia is usually intolerable.

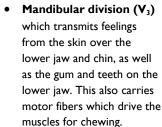
A very common feature is that most sufferers have noticed that there are often things that trigger the sudden onset of the pain. These triggering events usually involve something touching the area of the face or gum where the pain occurs. This touch may be minor, even a breath of wind in some cases. Eating, cleaning the teeth, and even talking can sometimes trigger the pain.

Although it can occur at any age in men and women, it increases in frequency with advancing years, and most people are middle-aged or elderly when it first occurs.

Trigeminal Nerve Anatomy

The trigeminal nerve is the 5th cranial nerve (there are 10 pairs of cranial nerves, each of which is connected to the brain; other nerves connect to the spinal cord). Like all cranial nerves, there are two trigeminal nerves (right and left), each transmitting feeling (sensations) from one side of the face. Each trigeminal nerve has three parts:

- Ophthalmic division (V₁)
 which transmits feelings
 from the skin of the eyelids,
 forehead, and front of scalp
 as well as the surface of the
 eye:
- Maxillary division (V₂)
 which transmits feelings
 from the cheek and upper
 lip, and also the upper gum
 and teeth:

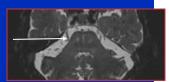


Each of these three divisions pass though a separate hole in the base of the skull before



The three divisions of the trigeminal nerve join at the ganglion (arrow), inside the skull, near the carotid artery

"There is no
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trigeminal
neuralgia."



Trigeminal nerve on MRI (arrow)

Anatomy (continued from page 1)

coming together to form the trigeminal ganglion, which lies just inside the skull, behind the eye and very near the carotid artery.

The main trigeminal nerve then runs backward from the trigeminal ganglion, though the fluid (CSF) around the brain, and joins the brain at the pons, which is the middle part of the brainstem. This region where the trigeminal nerve lies in the CSF between the ganglion and the pons is in

front of the cerebellum, (part of the brain involved with balance and coordination), and the region is often referred to as the cerebellopontine angle (CPA). Also lying in the CSF near the



Trigeminal nerve

trigeminal nerve in the CPA are arteries which supply blood to the brain, veins which drain blood away from the brain, and also the 4th, 6th, 7th and 8th cranial nerves. These nerves are involved with eye movements, movements of the facial muscles and hearing and

Diagnosing trigeminal neuralgia

Trigeminal neuralgia is a clinical diagnosis, and ideally needs to be made by an experienced neurologist or neurosurgeon. Essentially it is the patient's description of the pain (the symptoms) that are used to make the diagnosis, and examination shows no abnormal nerve function (i.e. no abnor-

mal signs).

There is no particular nerve test, x-ray, scan or blood test that can confirm the diagnosis. Never-the-less, these are often done to exclude other unusual conditions which can mimic trigeminal neuralgia, such as tooth problems, or tumours irritating the nerve.

Before considering any surgical procedure I usually perform an MRI scan to exclude problems other than trigeminal neuralgia. The MRI might show a vessel pressing on the trigeminal nerve, but that can also be seen in people without trigeminal neuralgia.

What causes trigeminal neuralgia?

The cause of trigeminal neuralgia is not precisely known, and possibly there are different causes in different people.

The most widely accepted cause is microvascular compression, which is pressure on the trigeminal nerve by an artery in the cerebellopontine angle. The operation of MVD (page 3) in which the neurosurgeon to identifies this artery and lifts it off the nerve is usually very successful at relieving trigeminal neuralgia. As we age our arteries stretch a little and tend to be more

tortuous, and it is this process that is often said to explain why trigeminal neuralgia is more common in older people.

However, in some cases no arterial compression of the nerve is found. Occasionally there is compression by a vein, but in other cases there is no particular abnormality found at surgery.

Other theories suggest that the underlying problem is a "short-circuit" between the nerve fibers either in the trigeminal nerve itself or in the brainstem. This may account for the finding that people with multiple sclerosis, which is a condition where there is loss of insulation (demyelination) around the nerves in the brain and spinal cord, are more likely to develop trigeminal neuralgia than the general population. However, most people with trigeminal neuralgia do not have MS, and only about 1% of people with MS develop trigeminal neuralgia.

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Medical treatment of trigeminal neuralgia

The first line of treatment of trigeminal neuralgia is with medication. As the pains are sudden onset and relatively brief, standard pain killers are often not particularly useful, and the best medica-



tions seem to be certain types of antiepilepsy medications. These stabilize the nerves and help prevent

abnormal impulses, and in many cases totally relieve trigeminal neuralgia.

The most commonly used medication is carbamazepine (also known by its trade-name Tegretol). A similar by newer medication is oxcarbazepine (Trileptal), and this is being used with increasing frequency and good results. Secondline drugs include baclofen (Lioresal), gabapentin (Neurontin), clonazepam (Klonopin), sodium valporate (Depakote), lamotrigine (Lamictal), and topiramate

(Topamax).

All the medications have sideeffects which can include tiredness, confusion, unsteadiness, allergies and abnormalities in liver function and blood-tests. They therefore need to be prescribed and monitored by a neurologist or other physician with expertise in the area.

Patients are usually only referred to neurosurgeons for consideration of surgical treatments if medications fail or the side-effects are intolerable.

" The primary treatment of trigeminal neuralgia should always be with medication. Surgery should be reserved for cases where medication fails or has intolerable side



Rhizotomy

Rhizotomy means making a lesion in a nerve. There are a lot of different techniques, but in general they are all done through a small stab incision under a brief anaesthetic. Although they are relatively quick, rhizotomies do have a small but definite risk of serious complications including stroke, doublevision, heart and anaesthetic problems. Also, because a lesion is made in the nerve there is always a moderate risk of permanent numbness of the face. If the eye

becomes numb this can predispose it to ulceration. Rarely following rhizolysis, people can have numbness associated with persisting severe pain. This devastating complication, called "anaesthesia dolorosa", is very difficult to treat.

In a rhizotomy, a needle is passed under xray control through the cheek, up through the hole in the skull base, and into the ganglion. When the needle is in the correct position inside the skull a lesion is made in the trigeminal ganglion by

either injecting glycerol, or heating the nerve with a radiofrequency (RF) probe, or passing a small tube through the needle with a balloon on the end, and inflating the balloon in the ganglion and causing a pressure injury to the nerve.

Less commonly, stereotactic radiosurgery can also be used to make a lesion in the nerve using a high dose of focused radiation.



RF Rhizotomy being performed (above), and xray showing needle (left)

Microvascular decompression (MVD)

MVD requires a general anaesthetic, and a craniotomy (opening the

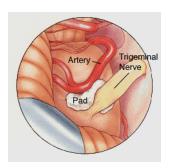


skull). Using microscopic techniques the surgeon identifies the trigeminal nerve in the cerebellopontine angle, and identifies any artery or vein compressing the nerve. The offending vessel is lifted off the nerve and a small pad put between them.

Obviously this is major surgery. However with stereotactic localization and drainage of spinal fluid (CSF) through a spinal drain, I have found that it can be performed with "keyhole" surgery, using a small opening and scar behind the ear, and most people are up and walking the day of

surgery and home a couple of days later.

The risks of this surgery include stroke, facial numbness, doublevision, hearing loss in one ear, CSF leak, infection, heart and lung problems and anaesthetic risks. The risk in a healthy person is small with usually less than 2% of cases having a serious complication.





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Surgery, if appropriate, is usually performed using the most minimally invasive microsurgical techniques appropriate to the case. All surgical procedures are performed at The Royal Melbourne and Melbourne Private Hospitals.

All consultations (insured or uninsured patients, Parkville or Geelong rooms, or Royal Melbourne Hospital clinics) should be arranged through the Parkville rooms.

Selecting the right type of surgery

All the surgical procedures (Rhizotomies and MVD) show varying degrees of immediate success and periods of long-term relief from pain, with none being 100% effective in all cases. Usually about 85% of cases get good symptomatic relief with the procedures, but up to 25% may have some degree of recurrence within the next 5 years. Also, as described, there is a small risk of serious complications with each procedure.

The decision as to what type of procedure is performed must be made according to the patient's preference and expectations, level of fitness and other medical conditions (including MS), previous procedures and treatments, and the specific distribution of the patient's pain. Patients therefore need to be well informed. Information from treating doctors is the most useful source for most patients. However, patient support groups can also be a valuable source of information for patients, and I encourage patients to contact the trigeminal neuralgia association

(www.tnaaustralia.org.au).

Of course recommendations by the treating doctors and neurosurgeon strongly influence the patient's decision-making process. We need also to consider that many doctors are strong proponents of specific proce-



dures, and this is often related to their expertise or familiarity with a particular procedure, as well as their own results, and other potential biases. Patients need therefore to make sure they ask questions of their neurosurgeon, and have these satisfactorily answered and have time to consider the options before committing to any surgery.

When I was training in neurosurgery I was advised that young fit patients with trigeminal neuralgia should have MVD, but older or less fit patients should have a Rhizotomy which is less technically demanding. However, with the advent of minimally-invasive "keyhole" techniques and stereotactic localization, I now consider MVD to be my primary operation of choice in most trigeminal neuralgia patients who have failed drug treatment. The reasons for this are that I have found somewhat better pain relief and significantly less facial numbness with MVD than rhizotomies, and importantly I have found the risk of a serious complication to be very small (depending on the patient's health, usually 2-5%). However, I have had extensive experience with rhizotomies (particularly radiofrequency, but also balloon and glycerol), and still recomend these procedures in selected patients.