

## 2 - The Intelligent Patient Overview

# TRIGEMINAL NEURALGIA

### Introduction to Trigeminal Neuralgia

*Trigeminal neuralgia* (TN), also called *tic douloureux*, is the most common facial pain syndrome and is often described as "the most terrible pain known to man". It is characterized by a sudden (paroxysmal) attack of facial pain described as intense, sharp, like an electric shock, or stabbing. The pain is most commonly felt in the cheekbone, most of the nose, upper lip and upper teeth and, in some people, it also extends to the lower lip, teeth, and chin. Pain is usually felt on one side of the face (*unilateral*) and lasts from a few seconds to two minutes. People with TN report that the intermittent pain attacks and the anticipatory anxiety from not knowing when they will occur result in a significant deterioration of their quality of life and interfere with daily activities such as eating and sleeping. Trigeminal neuralgia is the most frequently occurring nerve pain disorder. The most common cause of trigeminal neuralgia is compression of the trigeminal nerve by a blood vessel near the brainstem. However, many cases remain idiopathic and a specific cause cannot be identified.

There are 12 pairs of nerves, called *cranial nerves*, which innervate (provide the nerve supply to) the right and left side of the head and neck. The *trigeminal nerve*, designated as cranial nerve number V, is the fifth and largest of the cranial nerves. It provides nerve sensation to the face, mouth, and the front of the scalp, as well as controlling the muscles involved in chewing (mastication). The trigeminal nerve is also responsible for sensations such as taste, touch, and pain to the face.

The trigeminal nerve has three branches.

- The *ophthalmic branch* runs through the eye, forehead and nose.
- The *maxillary branch* runs through the upper teeth, gums, lips, cheek, lower eyelid and side of the nose.
- The *mandibular branch* runs through the lower teeth, gums and lip. It also controls jaw movement for mastication or chewing.

The three branches of the trigeminal nerve come together in an area called the *Gasserian ganglion*. From there, the trigeminal nerve root continues along the side of the brain stem and inserts into a portion of the brainstem called the *pons*.

Trigeminal neuralgia most often affects the maxillary branch or the mandibular branch of the trigeminal nerve. One study reported the division of pain in patients with TN to be:

- 44% in the maxillary branch

- 35% in the mandibular branch
- 19% in the ophthalmic branch

Most patients complain of pain in both the maxillary and mandibular branches.

## ***Classification of Trigeminal Neuralgia***

The International Headache Society (IHS) has identified two types of trigeminal neuralgia: *classical TN* and *symptomatic TN*.

*Classical trigeminal neuralgia*, also referred to as *idiopathic trigeminal neuralgia*, is typically caused by a blood vessel (usually the superior cerebellar artery) compressing the trigeminal nerve as it exits from the brainstem. The constant compression of the nerve root, compounded by each heartbeat that causes momentary widening of the artery, results in additional momentary rubbing, and leads to loss of the *myelin sheath*, the covering or insulation that surrounds the nerve fibers. The loss of the protective nerve covering is called *demyelination*. Without proper insulation, the nerve cells become hyperexcitable and begin to fire in an erratic and disorganized manner, resulting in significant pain. This is believed to be the cause of up to 90% of cases of idiopathic trigeminal neuralgia. In young adults, the most common cause of compression is thought to be a vein that compresses the trigeminal nerve, either alone or in addition to arterial nerve compression.

Many patients describe a specific event (*trigger*) that may precipitate an attack of classical trigeminal neuralgia (TN). These triggers include:

- Chewing
- Talking
- Swallowing
- Brushing teeth
- Face washing
- Shaving
- Vibration
- Exposure to cold or a cool breeze across the face
- Light touch to the specific areas of the face, or any vibration

The triggers are usually on the same side of the face as the pain (ipsilateral).

*Symptomatic TN* includes all cases where trigeminal neuralgia is secondary to an underlying, demonstrable medical condition that is not related to vascular compression, including:

- Multiple sclerosis - a chronic autoimmune disorder caused by the destruction (demyelination) of the myelin sheath of nerve fibers in the brain and spinal cord. Multiple sclerosis may also result in demyelination of one or more branches of the trigeminal nerve and lead to trigeminal neuralgia. It is estimated that 3-5% of patients with trigeminal neuralgia have multiple sclerosis. These patients are generally younger and may experience their first incident in their mid-40's. They are also more likely to have pain on both sides of their faces and often suffer from other neurological symptoms such as weakness in the arms

or legs.

- Other diseases which cause damage to the myelin sheath such as *leukodystrophy*, a group of diseases that are related to imperfect growth or development of the myelin sheath, chronic inflammatory demyelinating disease, or Guillain-Barre Syndrome.
- A tumor compressing the trigeminal nerve may also cause trigeminal neuralgia.
- Abnormalities at the base of the skull.
- Arteriovenous malformations - abnormal connection between veins and arteries.

Approximately 5-10% of cases of trigeminal neuralgia are caused by one of these underlying medical conditions.

It should be noted that it is important to distinguish *trigeminal neuralgia* from *trigeminal neuropathy*, which refers to a pain syndrome resulting from a more significant injury to the trigeminal nerve. Frequent causes of trigeminal neuropathy are surgical procedures involving the teeth and sinuses, as well as trauma or infections of the face and head.

## ***Incidence of Trigeminal Neuralgia***

Trigeminal neuralgia is considered by the National Organization of Rare Diseases (NORD) to be a rare condition that affects approximately 1.7 million people in the U.S., although estimates vary widely. It is estimated by some studies that approximately four to five in 100,000 people develop TN each year in the United States. Other estimates report that approximately 14,000 people in the U.S. develop TN each year. In addition:

- The average age of onset is between 50 and 70 years old, though it can be seen in younger people as well.
- The incidence of TN gradually increases with age.
- TN is uncommon in people younger than 30 years of age and only 1% of cases are reported to occur in people younger than 20 years of age.
- TN affects almost twice as many women as men.
- Some studies report pain to be more common on the right side of the face.

## ***Risk Factors for Trigeminal Neuralgia***

A risk factor is anything that increases the chances that a person will develop a particular disease or condition. Risk factors for trigeminal neuralgia (TN) include:

- Age - Average onset of TN occurs most frequently in people 50 years of age or older.
- Gender - Women are 1.5 times more likely to develop TN than men.
- Family history of facial pain - Approximately 5% of patients report a family history of facial pain, perhaps because an inherited pattern of blood vessel formation may increase the likelihood of irritation the trigeminal nerve.
- Multiple sclerosis - Approximately 3-5% of patients with TN also suffer from multiple sclerosis.