Aetiology, signs, symptoms and treatment of trigeminal neuralgia

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Trigeminal neuralgia is one of the most common causes of facial pain. It can be excruciating and can severely incapacitate patients and negatively affect their quality of life. First-line treatment is anticonvulsant drugs but other pharmacological options are available. If drug therapy is ineffective, a range of surgical options exist.

Trigeminal neuralgia (TN) is a painful facial condition of unknown aetiology (Olesen, 1988) and one of the most common causes of facial pain (Theodosopoulos et al, 2002). It was originally known as tic douloureux (tic) and is also termed idiopathic and primary trigeminal neuralgia. This article discusses the condition, its epidemiology and prognosis, causes, diagnosis and assessment, and interventions used for its management and treatment.

Symptoms

The primary symptom of TN is pain of a neuropathic nature (Box 1) with both chronic and acute elements (Crombie and Davies, 1999). The clinical hallmark is a sudden, excruciating, repetitious attack of unilateral facial pain – known as a paroxysm – in the distribution of one or more branches of the trigeminal nerve (Fig 1, p38). It is often precipitated by trivial mechanical stimulation such as touch, eating, washing, shaving, brushing the teeth, or talking (Meglio et al, 2000; Olesen, 1988). The condition is chronic, and over time the pain intensifies, episodes become longer and more frequent, and the pain spreads to larger areas of the face (Kitt, 1999). There are few epidemiological studies of TN, and its long-term prognosis, impact on psychological well-being, morbidity and comorbidity for patients is therefore largely unknown (Zakrzewska and Patsalos, 2002).

The aetiology and pathophysiology of TN remain unknown. It is hypothesised that the pain is caused by nociceptors in the V1, V2 or V3 branches of the trigeminal nerve (Crombie, 1999). Trigeminal neuralgia (TN) is one of the most common causes of facial pain. It can be excruciating and can severely incapacitate patients and negatively affect their quality of life. First-line treatment is anticonvulsant drugs but other pharmacological options are available. If drug therapy is ineffective, a range of surgical options exist.

Aetiology and epidemiology

The trigeminal nerve, or fifth cranial nerve, emerges from the base of the brain (pons) and possesses both sensory and motor fibres. It is one of the largest nerves in the head and has three main branches, which are distributed to different areas of the face (Fig 1, p38), and is responsible for sending the impulses of touch, pain, pressure, and temperature from the different areas of the face to the brain (National Institute of Neurological Disorders and Stroke, 2001). The paroxysm of pain usually occurs unilaterally, most commonly on the right-hand side of the face, although in 3–4 per cent of patients it can occur on both sides simultaneously (Jensen et al, 2003). The pain is usually localised along the maxillary or mandibular branch or both, and is occasionally felt along the ophthalmic branch (Costa, 1998).

It is estimated that the prevalence of TN is one per 1,000 of the population (Jensen et al, 2003), with an average age of onset of 50. The condition appears to be more common in women, and incidence increases with age (Kitt, 1999). There are few epidemiological studies of TN, and its long-term prognosis, impact on psychological well-being, morbidity and comorbidity for patients is therefore largely unknown (Zakrzewska and Patsalos, 2002).

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compression of the trigeminal nerve at its entry into the pons, and that this leads to demyelination – the loss of the myelin sheath (Jensen et al, 2003). In turn, demyelination results in the transmission of abnormal pain messages so that the patient experiences neuropathic pain.

Compression of the trigeminal nerve can be due to a number of factors including tumour (10 per cent of patients), pressure from blood vessels, aneurysm or multiple sclerosis, while causes of damage to the myelin sheath include nerve injury, shingles or post-herpetic neuralgia, trauma, infection, degeneration of the teeth or jaw, and the ageing process (Jensen et al, 2003). If the patient has a lesion, multiple sclerosis or other conditions that can cause neuropathic pain in the trigeminal nerve, the TN is said to be secondary or symptomatic, rather than primary or idiopathic (Melzack, 1992).

Although little research has been undertaken on the subject to elicit a comprehensive list, a range of possible risk factors has been associated with TN, including:
- Multiple sclerosis;
- Non-smoker;
- Non-alcohol user;
- Hypertension;
- Not having undergone tonsillectomy;
- Familial clustering, although reports are contradictory for this (Jensen et al, 2003; Kitt et al, 2000).

### Assessment and diagnosis

There are no objective measures, such as laboratory or pathological tests, available to diagnose TN, and physical and neurological examinations and investigations often yield normal results (Kitt et al, 2000). However, some can be helpful in establishing whether the TN is idiopathic or secondary, and/or determining the extent and location of any nerve compression (Jensen et al, 2003):
- Computerised tomography (CT);
- Magnetic resonance imaging (MRI);
- Magnetic resonance angiography (MRA).

The diagnosis of TN is therefore based on history-taking and assessment. The International Headache Society (Olesen, 1988) suggests at least four of the clinical criteria in Table 1 need to be present to make a diagnosis of TN.

The pain of TN occurs in paroxysms, which can last from several seconds to two minutes and can vary in frequency from several hundred a day to periods of remission lasting weeks or years (Kitt, 1999).

Typically, the pain is sudden in onset, severe in degree and short in duration. Patients use terms such as ‘sharp’, ‘stabbing’, ‘burning’, ‘shooting’ or ‘like an electric shock’ to characterise the pain, and describe it as ‘unbearable’, ‘terrible’, ‘excruciating’, and ‘intense’ (Sjaastad et al, 1997). The pain is so severe that it causes patients to wince, which is why TN is also known as tic. As time progresses the paroxysms can become more frequent and severe.

Patients tend to be asymptomatic between the paroxysms, and are generally pain-free at night. However, some experience a background pain that is dull, aching and continuous (Loeser, 1994).

### Management and treatment

The treatment of patients with TN is often challenging (Van Zundert et al, 2003). Medical management remains the first-line treatment for most patients but if this fails to control the pain, or intolerable side-effects occur, surgical management needs to be considered (Van Zundert et al, 2003; Orlandini, 2002).

### Anticonvulsants

Anticonvulsant drugs are effective in reducing TN pain for many patients. Their mechanism of action remains unclear but it has been proposed that they decrease pain impulses and produce pain relief by:
- Blocking sodium channels (Jensen, 2002);
- Acting as antagonists on the N-methyl-D-aspartate (NMDA) mechanism (McQuay and Moore, 1998);
- Suppressing the release of glutamate (Jensen, 2002);
- Raising gamma aminobutyric acid (GABA) levels throughout the central nervous system (Teng and Mekhail, 2003).

In the UK carbamazepine is the only anticonvulsant drug licensed for the management of the paroxysmal pain of TN (McQuay and Moore, 1998). It is an established, effective, evidence-based treatment and it is the drug of choice (McQuay and Moore, 1998; Berde, 1997). However, there are side-effects (Jensen et al, 2003; Teng and Mekhail, 2003).

### TABLE 1. DIAGNOSTIC CLINICAL CRITERIA

<table>
<thead>
<tr>
<th>1 Site</th>
<th>Trigeminal area, usually unilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>2 Radiation</td>
<td>Trigeminal area, other divisions beyond area</td>
</tr>
<tr>
<td>3 Characteristic</td>
<td>Sharp, shooting, electric</td>
</tr>
<tr>
<td>4 Severity severity</td>
<td>Moderate to suicidal</td>
</tr>
<tr>
<td>5 Duration</td>
<td>Each pain episode is a few minutes but there can be many throughout the day</td>
</tr>
<tr>
<td>6 Periodicity</td>
<td>Sudden bursts of pain, periods of no pain</td>
</tr>
<tr>
<td>7 Provoking factors</td>
<td>Light touch</td>
</tr>
<tr>
<td>8 Relieving factors</td>
<td>Antineuralgic drugs</td>
</tr>
<tr>
<td>9 Associated factors</td>
<td>Trigger points, weight loss</td>
</tr>
</tbody>
</table>

The McGill pain questionnaire (Melzack, 1992) is suggested as an assessment tool for TN, as it has been shown to differentiate between TN pain and atypical facial pain, and also assesses pain intensity and quality, and psychological distress.

A pain diary can also be used to assess the frequency and severity of attacks, the ability of the patient to perform normal activities, and for monitoring the response to treatments and any adverse effects (Zakrzewska and Lopez, 2003).

### REFERENCES


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However, it is associated with additional side-effects to carbamazepine and there is no evidence to support its long-term use to treat TN (Jensen et al, 2003). Phenytoin has been replaced with lamotrigine and gabapentin in clinical practice (Jensen, 2002).

Lamotrigine is used as an adjunct to carbamazepine for TN and has been found to be effective with few side-effects. It can also be used as a monotherapy to treat mild pain (Jensen et al, 2003).

Gabapentin is licensed in the UK for generalised neuropathic pain and has been shown to be an effective treatment for other neuropathic pains (Jensen et al, 2003). Some studies of its use with TN have been positive, although the evidence is not conclusive. The main advantage of gabapentin is its relatively benign side-effects, which are usually well tolerated if doses are increased gradually, following a graduated protocol (Jensen, 2002).

Other drugs

Baclofen is used effectively in some patients, either as an adjunct to carbamazepine or as a monotherapy, although there is insufficient evidence for its use in TN (Zakrzewska and Lopez, 2003). It is not licensed for use in TN but has fewer side-effects than carbamazepine (Jensen et al, 2003).

If carbamazepine cannot be tolerated, is ineffective or the patient is awaiting neurosurgery, tricyclic antidepressants (TCA) such as amitriptyline can be used. The side-effects of amitriptyline, which include dry mouth, constipation, blurred vision, tachycardia, urinary hesitancy, orthostatic hypotension, dizziness and tachycardia can be minimised by slow titration of the dosage (Teng and Mekhail, 2003).

Topically administered capsaicin has also been used. However, there is no sound evidence to support its use. Its side-effects include transient burning and erythema and it initially causes a burning sensation followed by anaesthesia. The cream needs to be applied three to five times per day, and this may negatively affect compliance (Teng and Mekhail, 2003).

Other drugs that are sometimes used with the drugs

### REFERENCES


### TABLE 2. SIDE-EFFECTS ASSOCIATED WITH CARBAMAZEPINE

<table>
<thead>
<tr>
<th>COMMON SIDE-EFFECTS</th>
<th>OTHER SIDE-EFFECTS</th>
<th>OLDER PEOPLE</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fatigue* Dizziness* Ataxia* Drowsiness* Blurred vision Diplopia Nausea and vomiting Unsteadiness/gait abnormalities</td>
<td>Haematological changes: Leukopenia Aplastic anaemia Thrombocytopenia Severe hepatic reaction</td>
<td>Side-effects are more common. There is an increased frequency of: Cardiac disease Cardiac conduction defects Decreased osmolarity Water retention Hyponatraemia</td>
</tr>
<tr>
<td>(seven per cent) that presents as a skin rash</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>DISADVANTAGE High drug interaction profile</td>
<td></td>
</tr>
</tbody>
</table>

*Affect >10 per cent of patients, tend to be dose-related, and usually abate within a few days – spontaneously or after dose reduction.
marily through a specialist unit or pain service, which is patients with TN, their pain is likely to be managed pri
table their use, some patients report that other treatment
 approach. Although there is little or no evidence to sup
Management of TN requires a multidisciplinary
Other interventions mentioned above include mexiletine, clonazepam, local anaesthetics, non-steroidal anti-inflammatory drugs, and opioids. But no sound evidence supports their use in TN.
Surgical intervention
If medical intervention is ineffective or cannot be tolerat ed by the patient, a number of surgical options exist (Jensen et al, 2003; Kalkanis et al, 2003; Van Zundert et al, 2003; Zakrzewska and Lopez, 2003; Day, 2001; Tronnier et al, 2001; Meglio et al, 2000; Zakrzewska et al, 1999; Costa, 1998), although there is currently no evi
dence on when surgery should be considered. Surgical intervention can be aimed at three levels (Table 3). However, only microvascular decompression (MVD) causes no destruction of the trigeminal nerve.
Other interventions
The management of TN requires a multidisciplinary approach. Although there is little or no evidence to support their use, some patients report that other treatment options offer some relief. These include acupuncture, distraction techniques, relaxation training, biofeedback, meditation, and physical therapy.
The nurse’s role in assessing and managing patients’ pain is vital and will vary enormously depending on where the patient is being managed. In the case of patients with TN, their pain is likely to be managed primarily through a specialist unit or pain service, which is multidisciplinary. The nurse is therefore usually a specialist or consultant nurse and will have additional skills to be able to assess and manage pain of a complex nature. The specialist nurse role will vary from service to service but may include specialist assessment techniques, advising on pharmacological interventions and reviews, offering a variety of psychological and physical therapies, and being an educator. It will involve liaison with other health professionals and services.

**Conclusion**
The diagnosis, assessment and management of TN is a complex process requiring a multidimensional approach. There are profound gaps in our knowledge of TN and insufficient evidence to support many interventions used to provide pain management in clinical settings. Future research is imperative if the quality of care for patients with TN is to improve.

**REFERENCES**

**PATIENT SUPPORT GROUPS**
Trigeminal Neuralgia Association UK: www.tna.org.uk
Trigeminal Neuralgia Association: www.tna-support.org

**TABLE 3. SURGICAL INTERVENTIONS FOR TRIGEMINAL NEURALGIA PAIN**

<table>
<thead>
<tr>
<th>1. PERIPHERAL NERVE BRANCHES</th>
<th>2. GASSERIAN GANGLION</th>
<th>3. POSTERIOR FOSSA</th>
</tr>
</thead>
<tbody>
<tr>
<td>● Cryotherapy: the trigger nerve is exposed and then frozen.</td>
<td>● Radiofrequency thermo-coagulation: the gasserian ganglion is heated to 60–80°C.</td>
<td>● Microvascular decompression (MVD): all blood vessels or arteries compressing cranial nerves are moved but there is no destruction of the trigeminal nerve.</td>
</tr>
<tr>
<td>● Injection: alcohol, phenol or streptomycin is injected into the trigger nerve.</td>
<td>● Glycerol injection: small doses of glycerol are injected to destroy the areas of the nerve responsible for triggering the pain.</td>
<td>This is the most commonly performed (open) surgical treatment and shows good long-term pain control with reduced mortality and morbidity. However, strong evidence to support its use is lacking. Pain may recur after 10 years and hearing loss may occur in two per cent of patients.</td>
</tr>
<tr>
<td>● Peripheral laser treatment: skin overlying the trigger nerve is subjected to laser irradiation.</td>
<td>● Microcompression: a balloon is inserted into the ganglion then inflated and the nerve is compressed.</td>
<td>● Gamma knife (stereotactic neurosurgery): a beam of gamma rays is directed at the trigeminal nerve.</td>
</tr>
<tr>
<td>● Peripheral neurectomy: the peripheral nerve is excised.</td>
<td>All these interventions are neurodestructive and may be associated with side-effects such as sensory loss, dysthesia, anaesthesia dolorosa, and corneal anaesthesia. Pain may recur in three to five years. Radiofrequency thermocoagulation is one of the most commonly used surgical procedures. However, there is insufficient evidence to support its use in trigeminal neuralgia.</td>
<td>Pain may recur in three to five years. There is a delayed onset of relief and sensory loss develops over two years. Long-term complications of radiation have not been reported.</td>
</tr>
</tbody>
</table>

There is insufficient or no evidence to support the above interventions in TN. Pain may recur 10–24 months after surgery but in a different branch. Mild sensory loss can occur, which may be permanent.