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## Disclaimer
Facial pain syndrome in the distribution of ≥1 divisions of the trigeminal nerve.

Diagnosis is clinical, with a history of paroxysms of sharp, stabbing, intense pain lasting up to 2 minutes.

First-line therapy is medical, to which the majority of patients are partially responsive.

Surgical/ablative therapies are reserved for refractory cases.

Ablative therapies can cause facial sensory loss and are associated with a high recurrence rate; however, they are relatively non-invasive and carry minimal risk of severe morbidity/mortality.

Microvascular decompression has few long-term sequelae and well-established efficacy but has a potential risk of major morbidity.

More research on neurostimulation is needed to define its role in the treatment of trigeminal pain.
Definition
Trigeminal neuralgia (TN) is a facial pain syndrome in the distribution of ≥1 divisions of the trigeminal nerve. It is characterised by some combination of paroxysms of sharp, stabbing, intense pain lasting up to 2 minutes and/or a constant component of facial pain, without associated neurological deficit. The pain can be precipitated by trigger areas or factors, and repeat attacks are typically stereotyped in the individual.

Epidemiology
The incidence of trigeminal neuralgia is generally thought to be from 4 to 13 per 100,000 based on US and UK population studies.[4] [5] This means that 12,000 to 40,000 new cases are diagnosed in the US annually. There is a slight female predominance at all ages, and rates appear to increase with age, with reported incidence rates as high as 74 to 88 per 100,000 in elderly (>75 years of age) populations.[6]

Aetiology
Aetiology is associated with trigeminal nerve compression in the majority of patients. Curiously, many patients have similar compression on the asymptomatic, contra-lateral side, and asymptomatic patients without trigeminal neuralgia (TN) may demonstrate vascular compression:

- Compression: 80% to 90% of patients have demonstrable focal compression of the trigeminal nerve root at the root entry zone by an aberrant vascular loop (typically the superior cerebellar artery).[7] Reports of trigeminal nerve compression by true vascular malformations (aneurysms or arteriovenous malformations) exist but are rare.[8] [9] Posterior fossa tumours can also produce symptoms mimicking TN.[10]
- Demyelinating disease: TN is 20 times more prevalent in multiple sclerosis (MS) sufferers compared with the general population.[11] MS patients with TN usually demonstrate demyelinating plaques in the pons that encompass the root entry zone of the trigeminal nerve.[12] [13] However, one study suggests that MRI-positive root entry zone abnormalities, which are more prevalent than previously thought in the MS population, often occur in the absence of trigeminal symptoms.[14]
- Other brainstem lesions: rare cases of TN have been reported with brainstem infarcts and amyloid or calcium deposition along the trigeminal sensory pathway.[7]

Pathophysiology
Focal demyelination and the resultant conduction aberrations (ephaptic transmission) are thought to represent the pathophysiological mechanism of the neuropathic pain of trigeminal neuralgia (TN). Pathological examinations of post-mortem patients with TN secondary to an aberrant vessel loop demonstrate focal, chronic myelin loss at the root entry zone in close proximity to the region of vascular compression. Pathological specimens have minimal, if any, evidence of ongoing inflammation. MS plaques (foci of demyelination) encompassing the root entry zone have also been described in pathological specimens of MS patients with TN.[15] [16]

Classification
Burchiel classification[1] [2] [3]

Idiopathic trigeminal neuralgia type I (TNI) ‘classic trigeminal neuralgia’ (CTN)

- Sharp, shooting, electric shock-like pain. Pain should be episodic >50% of the time.

Idiopathic trigeminal neuralgia type II (TNII) ‘atypical TN’

- Aching, throbbing, burning pain >50% of the time with a constant background.

Trigeminal neuropathic pain

- Secondary to unintentional trigeminal injury (facial trauma, oral surgery, ear, nose and throat surgery, skull base surgery, posterior fossa surgery, stroke).

Trigeminal deafferentation pain ‘anaesthesia dolorosa’

- Secondary to intentional denervating procedure (e.g., neurectomy, gangliolysis, rhizotomy, nucleotomy, tractotomy).

Symptomatic TN

- Name derives from the fact that pain is a symptom of underlying pathology (tumour, inflammatory demyelination, etc.).
- Associated with multiple sclerosis or compression secondary to local tumour.
- Younger age of onset, involvement of the first division of trigeminal nerve, unresponsiveness to treatment, and abnormal trigeminal evoked potentials should be disregarded as useful for disclosing symptomatic TN.

Post-herpetic TN

- Resulting from an outbreak of facial herpes zoster.

Atypical facial pain

- Somatoform pain disorder.
- Requires evaluation with psychological testing prior to confirmation.

Divisions of the trigeminal nerve

V1 - Ophthalmic division

V2 - Maxillary division

V3 - Mandibular division
Primary prevention

No preventive strategies have been identified.
Case history

Case history #1

A middle-aged woman presents with a complaint of frequent (once or twice daily for 3 weeks), brief (lasting several seconds) episodes of intense, sharp left-sided jaw pain. She has experienced these attacks for several years, but they had previously been relatively rare (1 episode daily for several consecutive days followed by months with no attacks). She says that episodes are sometimes brought on by eating but can occur without an apparent stimulus. The patient states that even though the pain is brief, she lives in fear of repeat flares.

Other presentations

Maxillary/mandibular (V2/V3) distribution symptoms occur in the majority of patients. This can present as sharp pain running from the mouth to the jaw (commonly mistaken for dental pain) or less commonly as pain from the upper lip to the orbit. Symptoms in an exclusively ophthalmia (V1) distribution are the least common, and patients typically refer to the pain as headache.

Step-by-step diagnostic approach

Trigeminal neuralgia (TN) is a clinical diagnosis. Distinguishing between classic (idiopathic) trigeminal neuralgia (CTN) and symptomatic TN (STN) is done through history taking, clinical examination (to identify non-trigeminal neurological signs), and neuroimaging, which may identify a cause in up to 15% of patients.[18]

History

For CTN, history typically shows stereotyped, unilateral paroxysms of facial pain lasting seconds to minutes in a distribution along ≥1 divisions of the trigeminal nerve. The pain associated with TN is typically described as intense, sharp, superficial, stabbing or burning in quality. Triggers such as tooth brushing, eating, cold, and touch are common. Although most patients are asymptomatic between episodes and do not complain of neurological deficit, the intensity of attacks keeps patients continually fearful of repeat attacks. An important component of history taking in these patients involves the attempt to elicit information (such as a history of prior oropharyngeal or facial trauma), prior diagnoses (such as prior herpetic outbreak), and description of other symptomatology (such as rash or facial droop) that may direct diagnosis away from TN.[19]

Physical examination

Patients with CTN (TNI) should have a completely unremarkable physical and neurological examination. Important components of the physical examination needed to rule out other facial pain syndromes include examination of dentition and oropharynx, assessment of temporomandibular joint function, skin examination, palpation of temporal arteries, and full neurological examination with particular focus on facial sensory examination. Sensory changes on neurological examination are suggestive of a pathological underpinning to the pain complaints (i.e., STN).
Tests

There are no useful laboratory tests for diagnosing CTN. However, clinical suspicion of STN should prompt further work-up. Evidence indicates that measuring trigeminal reflexes in a qualified electrophysiological laboratory can be used for distinguishing STN from CTN.[18] Imaging tests that can be used to exclude other causes of trigeminal distribution pain include intra-oral x-rays if pain appears to be of dental origin and magnetic resonance imaging if intracranial pathology is a concern: for example, abnormal trigeminal reflexes on electrophysiological testing or clinical suspicion.

Risk factors

Strong

increased age

- In all studies of trigeminal neuralgia (TN), incidence rates increase with the age of population studied.[6]

multiple sclerosis

- TN is significantly more common in study populations with multiple sclerosis.

Weak

female

- TN shows a slight female predisposition at all ages.

hypertension

- A single study has shown a mildly increased risk of TN in women with hypertension (odds ratio 1.96).[17]

History & examination factors

Key diagnostic factors

facial pain (common)

- Required for diagnosis. Pattern is important, as restriction to trigeminal distributions and quality, duration, and consistency of pain are critical for diagnosis of trigeminal neuralgia. Bilateral involvement is more common in patients with symptomatic trigeminal neuralgia.

presence of risk factors (common)

- Key risk factors include increased age and multiple sclerosis.

Other diagnostic factors

prior oropharyngeal or facial trauma (common)

- Points to other causes of facial pain or trigeminal neuropathic pain syndrome.

prior herpetic outbreak (uncommon)
Trigeminal neuralgia

- Suggestive of post-herpetic trigeminal neuralgia.
- Sensory/motor changes (uncommon)
  - Suggestive of pathological cause.

## Diagnostic tests

### 1st test to order

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>no first test</td>
<td>diagnosis is usually clinical</td>
</tr>
</tbody>
</table>

### Other tests to consider

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>intra-oral x-ray</td>
<td>no dental cause of pain demonstrated</td>
</tr>
<tr>
<td>MRI</td>
<td>may demonstrate presence of abnormal vessel loop in association with the trigeminal nerve, presence of other pathologies (e.g., tumour, infarct, multiple sclerosis plaque)</td>
</tr>
<tr>
<td>trigeminal reflex testing</td>
<td>early blink reflex or early masseter inhibitory reflex in symptomatic TN</td>
</tr>
</tbody>
</table>

## Differential diagnosis

<table>
<thead>
<tr>
<th>Condition</th>
<th>Differentiating signs / symptoms</th>
<th>Differentiating tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dental caries</td>
<td>Well localised to a single tooth.</td>
<td>Intra-oral x-rays.</td>
</tr>
<tr>
<td>Dental fracture</td>
<td>Localised to ≥1 teeth; may be provoked by biting.</td>
<td>Intra-oral x-rays.</td>
</tr>
</tbody>
</table>
## Condition | Differentiating signs / symptoms | Differentiating tests |
--- | --- | --- |
Mandibular osteomyelitis | Typically continuous pain along mandible with associated swelling. History of fever, malaise, purulent discharge, mobile teeth. | Intra-oral x-rays. |
Temporomandibular joint syndrome | Often bilateral, jaw opening may be restricted. | No differentiating tests. |
Migraine | Presence of preceding aura. Associated with photophobia, nausea. | No differentiating tests. |
Glossopharyngeal neuralgia | Pain is in the distribution of the glossopharyngeal nerve, classically oropharyngeal and otic. | No differentiating tests. |
Post-herpetic neuralgia | Typically presents as continuous pain in distribution of upper division of trigeminal nerve (V1). Patient often has tactile alldynia. | No differentiating tests. |
Temporal arteritis | Pain is often continuous, can be bilateral, and may be associated with jaw claudication, visual changes, and anorexia. | Erythrocyte sedimentation rate, temporal artery biopsy (gold standard). |
Atypical facial pain | Often is bilateral and/or exceeds the confines of a trigeminal distribution. | Diagnosis is made with formal neuropsychological evaluation. |
Trigeminal autonomic cephalgias (e.g., cluster headache, short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing [SUNCT] syndrome, chronic paroxysmal hemicrania) | Often have prominent autonomic features (e.g., conjunctival injection, lacrimation, nasal congestion, rhinorrhea, ptosis). Pain associated with cluster headaches, typically retro-orbital.[22] | No differentiating tests. |

## Diagnostic criteria

**Headache Classification Committee of the International Headache Society[23]**

For classic trigeminal neuralgia (TN), history should consist of paroxysms of facial pain lasting seconds to minutes with at least 4 of the 8 following characteristics:
1. Distribution along ≥1 divisions of the trigeminal nerve
2. Pain is described as sudden, intense, sharp, superficial, stabbing, or burning in quality
3. Severe intensity
4. Attacks are precipitated by known triggers
5. Patient is asymptomatic between episodes
6. Lack of neurological deficit
7. Attacks are stereotyped in a given patient
8. Other causes of facial pain have been excluded by history/physical examination/studies.
Step-by-step treatment approach

Pain relief is the goal of therapy.

Medical therapy

Anticonvulsants:

- Medical therapy is the initial treatment of choice. The mainstay of medical therapy consists of anticonvulsant medicines, generally carbamazepine\[Evidence\] or oxcarbazepine.\[Evidence\] Carbamazepine is the only anticonvulsant medicine with efficacy proven in randomised controlled trials in trigeminal neuralgia (TN) and is typically first-line therapy.\[24\] Seventy percent to 75% of patients show at least partial improvement on carbamazepine; however, long-term use may be associated with decreased efficacy.
- Oxcarbazepine, a derivative of carbamazepine with fewer drug-drug interactions and a lower adverse effect profile, appears equally efficacious and may even be useful in patients with carbamazepine-resistant TN. Lamotrigine may be considered, although evidence for this is limited, and a Cochrane review suggests that it has little to no role in the treatment of neuropathic pain.\[3\] \[18\] \[25\]
- Development of allergic rash with use of carbamazepine or lamotrigine may be a precursor to the development of potentially serious reactions (e.g., Stevens-Johnson syndrome) and should prompt immediate discontinuation.
- Gabapentin may be more effective for multiple sclerosis-associated TN; however, there is little evidence to support this.\[26\]
- Patients whose TN is partially or completely refractory to anticonvulsant medicines are often tried on other classes of pain medicine, such as baclofen, although data supporting their use in TN are scant.
- There is some evidence for the effectiveness of topiramate.\[27\] Other anticonvulsant drugs, including newer-generation medicines, are being used off-label for TN in the absence of definitive clinical evidence.\[28\]
- Owing to its significant side-effect profile and the lack of data supporting its use in any neuropathic pain syndrome, the use of phenytoin for TN, even in cases of prior medication failure, cannot be supported.\[29\]

Other medicines:

- There are some data in support of baclofen, but they are insufficient to make definitive treatment recommendations.\[Evidence\] Based on practitioner preference, it can be given sequentially as monotherapy or in combination until an effective regimen is established. In the absence of strong clinical data, the deployment of such treatment regimens cannot be endorsed.
- Other medicines such as tizanidine and pimozide have fallen out of favour and are rarely used.\[30\]

Surgical therapy

Failure of medical therapy leads many patients to more invasive treatment options. The choice of procedure is ultimately dependent upon patient preference but should only be undertaken after a complete discussion and understanding of the efficacy, recurrence rates, and potential complications of each method.
Trigeminal neuralgia

Treatment

Microvascular decompression remains the first-line procedural treatment for TN, as it is targeted to the presumptive pathological initiator of TN and has the least long-term neurological sequelae, with rates and durability of symptom improvement that are equal to or better than other surgical/procedural treatments.[31] [4] Evidence In the hands of experienced surgeons, it is a low-morbidity procedure that is appropriate for younger and older populations as long as there is no contraindication to general anaesthesia/surgery.[32] [33]

Current data suggest that ablative procedures are safe (≤1% life-threatening complication rate), relatively short (often performed on an outpatient basis), and can usually be performed without the need for general anaesthesia. However, studies of these procedures are typically retrospective, uncontrolled, and have mixed definitions of TN. Pain relief usually occurs by the first post-procedure week. Post-procedure dysaesthesia is common (up to 25%), with <10% of patients requiring medicine for attempted control. The high recurrence rate, coupled with low complication rate, leads many practitioners to offer this option to older populations of TN sufferers whose greater comorbidities make major surgery more risky. Patients who do not want open surgery or who carry an unacceptably high surgical risk may be offered radiosurgery as a first option, with percutaneous methods typically reserved for patients with V2 and V3 distribution symptoms, as they are less likely to develop post-procedural impairment of corneal sensation.[34] [35] There are a number of ablative techniques available:

- Stereotactic radiosurgery involves targeting the sensory root of the trigeminal nerve stereotactically to deliver high doses of radiation without significant spread to surrounding tissues. It is the least invasive surgical option, and maximum pain improvement occurs at 1 month. Uncontrolled data suggest it is probably less effective, and results are less durable than percutaneous methods[36] [5] Evidence
- Gasserian gangliolysis can be thermal (radiofrequency) or chemical (glycerol or alcohol). More than 75% of TN type I patients experience improvement, although approximately one-third of patients experience recurrence within 3 years. It is associated with relatively high rates of dysaesthesia. For chemical gangliolysis, glycerol is the most commonly used agent, as alcohol is associated with higher recurrence
- Balloon compression of the trigeminal (gasserian) ganglion requires general anaesthesia. It has similar efficacy and recurrence rates as gangliolysis but with decreased rates of severe dysaesthesia and impaired corneal sensation
- Peripheral neurectomy (partial sensory rhizotomy) is an open procedure involving avulsion, excision, and/or thermocoagulation of ≥1 trigeminal nerve branches innervating a trigger zone. Sensory loss is unavoidable and not considered a complication of the procedure. It can be performed under local anaesthesia with sedation and has a 60% to 80% initial response rate. There is <10% morbidity, as the corneal reflex is typically preserved. The 5-year recurrence rate varies between 35% and 75%. Due to its low success rate and high complication rate it should not be used as first-line surgical option and is generally used in patients who have failed multiple other treatments[37]
- Cryotherapy/peripheral neurectomy targets peripheral branches of the trigeminal nerve in the affected distribution. It can be repeated if incomplete or if symptoms develop in a new distribution. Insufficient clinical data exist to recommend either over more established, ganglion-directed procedures. May produce trigeminal deafferentation pain and/or anaesthesia dolorosa.

Neurostimulation is a relatively new treatment option, and few studies of sufficient methodological rigour exist to explicitly define its role in the management of trigeminal pain. Theoretical advantages to stimulation for pain control include its adjustable nature (reflecting fluctuations in pain level) and its
reversibility. Patients who are referred for such procedures have typically failed multiple medical and surgical regimens. Such patients are best served by referral to speciality pain centres where access to and experience with multiple treatment modalities allow for treatment recommendations unencumbered by the availability of a particular technology.

### Treatment details overview

Consult your local pharmaceutical database for comprehensive drug information including contraindications, drug interactions, and alternative dosing. (seeDisclaimer)

#### Acute

<table>
<thead>
<tr>
<th>newly-diagnosed trigeminal neuralgia (TN)</th>
<th>(summary)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1st</td>
</tr>
<tr>
<td>anticonvulsant-unresponsive trigeminal neuralgia (TN)</td>
<td>1st</td>
</tr>
<tr>
<td></td>
<td>anticonvulsants</td>
</tr>
</tbody>
</table>

#### Ongoing

<table>
<thead>
<tr>
<th>medicine-unresponsive trigeminal neuralgia type I (TNI)</th>
<th>(summary)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>1st</td>
</tr>
<tr>
<td>medicine-unresponsive trigeminal neuralgia type I/II (TNI/II)</td>
<td>1st</td>
</tr>
<tr>
<td></td>
<td>ablative surgery</td>
</tr>
<tr>
<td>refractory trigeminal neuralgia type I/II (TNI/II) (medical and surgical failures); trigeminal neuropathic pain; trigeminal deafferentation pain; symptomatic TN</td>
<td>1st</td>
</tr>
<tr>
<td></td>
<td>neurostimulation</td>
</tr>
</tbody>
</table>
## Treatment options

### Acute

<table>
<thead>
<tr>
<th>newly-diagnosed trigeminal neuralgia (TN)</th>
</tr>
</thead>
</table>

### 1st anticonvulsants

#### Primary options

- **Carbamazepine**: 200 mg/day orally initially given in 1-2 divided doses, usual maintenance dose is 400-1200 mg/day given in 2 divided doses

  **OR**

- **Oxcarbazepine**: 300 mg/day orally initially, usual maintenance dose is 600-1200 mg/day, maximum 1200 mg/day given in 2 divided doses

#### Secondary options

- **Topiramate**: 12.5 to 25 mg/day orally initially, increase gradually according to response, maximum 200 mg/day

#### Tertiary options

- **Gabapentin**: 300 mg orally once daily initially, usual maintenance dose is 300-1800 mg/day, maximum 1800 mg/day given in 3 divided doses

  **OR**

- **Lamotrigine**: consult specialist for guidance on dose

*Carbamazepine is the only anticonvulsant medicine with efficacy proven in randomised controlled trials in TN and is typically first-line therapy.*[24] Seventy percent to 75% of patients show at least partial improvement on carbamazepine. Long-term use may be associated with decreased efficacy.

*Oxcarbazepine, a derivative of carbamazepine with fewer drug-drug interactions, appears equally efficacious and may even be useful in patients with carbamazepine-resistant TN.*

*Lamotrigine may be considered, although evidence for this is limited, and a Cochrane review suggests that it has little to no role in...*
## Acute

<table>
<thead>
<tr>
<th>Anticonvulsant-unresponsive trigeminal neuralgia (TN)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1st</strong> baclofen</td>
</tr>
<tr>
<td><strong>Primary options</strong></td>
</tr>
<tr>
<td>- baclofen: 15 mg/day orally initially given in 3 divided doses, increase according to response, maximum 80 mg/day</td>
</tr>
<tr>
<td>- There are some data in support of baclofen, but they are insufficient to make definitive treatment recommendations.</td>
</tr>
<tr>
<td>Based on practitioner preference, it can be given sequentially as monotherapy or in combination until an effective regimen is established. In the absence of strong clinical data, the deployment of such treatment regimens cannot be endorsed.</td>
</tr>
<tr>
<td>- Other medicines such as tizanidine and pimozide have fallen out of favour and are rarely used.</td>
</tr>
</tbody>
</table>

## Ongoing

<table>
<thead>
<tr>
<th>Medicine-unresponsive trigeminal neuralgia type I (TNI)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1st</strong> microvascular decompression</td>
</tr>
<tr>
<td>- Open neurosurgical procedure where the compressive vascular loop is manually separated from the trigeminal nerve pontine entry site.</td>
</tr>
</tbody>
</table>
**Trigeminal neuralgia**

### Treatment

<table>
<thead>
<tr>
<th>Ongoing</th>
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</thead>
<tbody>
<tr>
<td>» May be somewhat less effective in patients with multiple sclerosis-related TN and co-existing vascular compression on preoperative imaging studies.</td>
</tr>
</tbody>
</table>

**medicine-unresponsive trigeminal neuralgia type I/II (TNI/II)**

<table>
<thead>
<tr>
<th>1st ablative surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Primary options</strong></td>
</tr>
<tr>
<td>» stereotactic radiosurgery</td>
</tr>
<tr>
<td>OR</td>
</tr>
<tr>
<td>» gangliolysis (gasserian)</td>
</tr>
<tr>
<td>OR</td>
</tr>
<tr>
<td>» balloon compression</td>
</tr>
<tr>
<td>OR</td>
</tr>
<tr>
<td>» peripheral neurectomy (partial sensory rhizotomy)</td>
</tr>
<tr>
<td>OR</td>
</tr>
<tr>
<td>» cryotherapy/peripheral neurectomy</td>
</tr>
</tbody>
</table>

» There are a number of ablative techniques available. Current data suggest that ablative procedures are safe (≤1% life-threatening complication rate), relatively short (often performed on an outpatient basis), and can usually be performed without the need for general anaesthesia.

» However, studies of these procedures are typically retrospective, uncontrolled, and have mixed definitions of TN.

» Pain relief usually occurs by the first post-procedure week. Post-procedure dysaesthesia is common (up to 25%), with <10% of patients requiring medicine for attempted control. The high recurrence rate, coupled with low complication rate, leads many practitioners to offer this option to older populations of TN sufferers whose greater comorbidities make major surgery more risky.

» Patients who do not want open surgery or who carry an unacceptably high surgical risk may be offered ablative radiosurgery as a first option.5[C]Evidence with percutaneous ablative
## Treatment

### Ongoing

Methods typically reserved for patients with V2 and V3 distribution symptoms, as they are less likely to develop post-procedural impairment of corneal sensation.\(^\text{[34]}\) \(^\text{[35]}\)

Refractory trigeminal neuralgia type I/II (TNI/II) (medical and surgical failures); trigeminal neuropathic pain; trigeminal deafferentation pain; symptomatic TN

<table>
<thead>
<tr>
<th>1st neurostimulation</th>
<th>Primary options</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Primary options</strong></td>
<td></td>
</tr>
<tr>
<td>» motor cortex stimulation</td>
<td>OR</td>
</tr>
<tr>
<td>» trigeminal branch stimulation</td>
<td>OR</td>
</tr>
<tr>
<td>» gasserian ganglion stimulation</td>
<td></td>
</tr>
</tbody>
</table>

- Stimulation of different brain areas depending on aetiology.\(^\text{[38]}\)

- Motor cortex stimulation is mainly used in cases of severe trigeminal deafferentation pain. A craniotomy is required for electrode implantation.

- Trigeminal branch stimulation is used in cases of trigeminal neuropathic pain secondary to nerve damage (trauma or post-procedure). The stimulator placement is performed percutaneously.\(^\text{[38]}\)

- Gasserian ganglion stimulation is almost exclusively used for trigeminal neuropathic pain. The stimulator is placed via image guidance through the foramen ovale. Electrodes tend to migrate.\(^\text{[39]}\) \(^\text{[40]}\)
Emerging

Reversible techniques

Future advances in the field are likely to occur through the expansion of reversible neuromodulatory techniques such as neurostimulation.[28] Of particular importance for advancement in the field is the need for the development of diagnostic and treatment algorithms that can be directly compared.

Botulinum toxin type A

Botulinum toxin type A, a neurotoxin produced by *Clostridium botulinum* that acts to prevent acetylcholine release from nerves, has been used in a variety of conditions including cervical dystonias, temporomandibular joint pain syndromes, and chronic migraine. Because of its increasing use in other types of headache and facial pain syndromes, some have suggested using it in cases of refractory trigeminal neuralgia (TN). Proposed targets include trigger zones, specific trigeminal nerve branches, and various orofacial muscles. Two recent reviews suggested that botulinum toxin type A may be beneficial in the treatment of some patients with TN.[41] [42] All of the studies identified were plagued by single-arm design, small sample sizes, varying definitions of TN, and confounders such as concomitant medical treatment and differing injection targets. Determination of the ultimate value of botulinum toxin in the treatment paradigm of TN will require larger, better-designed studies. For now it remains largely experimental, with limited use in the most refractory cases.

Lidocaine

Intravenous lidocaine has been used for various neuropathic pain syndromes (e.g., diabetic neuropathy, HIV and post-herpetic neuralgias, as well as neuropathies associated with cancer, stroke, spinal cord injury, and amputation), with some evidence of success compared with placebo.[43] A Japanese case series of 9 patients with intractable TN, who were treated with a weekly intravenous infusion of magnesium and lidocaine for 3 weeks, reported “sound pain relief” following treatment.[44] Dose-related central nervous system and cardiac toxicity are serious concerns associated with the use of intravenous lidocaine. More research, including clinical trials, is needed before this can be considered a viable option in treating refractory TN. There is very little evidence that topical forms of lidocaine are effective in the treatment of neuropathic pain, but further studies are required.[45]
**Recommendations**

**Monitoring**

Patients started on medical therapy should be evaluated regularly for adverse reactions, documentation of efficacy of treatment, and/or dose adjustment until adequate pain control is achieved. Following uncomplicated surgical treatment, patients should be re-evaluated about 1 week post-procedure to document resolution of symptoms and evaluate for the presence of complications. Subsequent evaluations are performed at longer time periods based on level of symptomatic relief, pain recurrence, and patient desire to wean medications.

**Patient instructions**

Patients may find keeping a daily 'pain' log during the initial medication treatment phase useful for charting improvement/worsening and recording failed regimens, thereby facilitating communication with their doctor and providing a sense of empowerment. This information can also be used if flare-ups occur at a later date. Patients may also find the following online resource useful. [The Facial Pain Association]

**Complications**

<table>
<thead>
<tr>
<th>Complications</th>
<th>Timeframe</th>
<th>Likelihood</th>
</tr>
</thead>
<tbody>
<tr>
<td>post-operative complications</td>
<td>variable</td>
<td>medium</td>
</tr>
</tbody>
</table>

Operative treatment of trigeminal neuralgia can be complicated by any of the following: hearing loss (1% to 19% in microvascular decompression), facial hypeaesthesia (highest for stereotactic radiosurgery, percutaneous balloon compression, and partial sensory rhizotomy), corneal hypeaesthesia, trigeminal motor weakness, anaesthesia dolorosa, keratitis, cranial nerve palsies, cerebrospinal fluid leak, meningitis, herpes labialis reactivation, and arteriovenous fistula formation.[37]

**Prognosis**

Trigeminal neuralgia (TN) is a chronic affliction. There are few published data regarding the natural history of the condition, but anecdotal data suggest variable periods of remission and relapse. Most patients find at least partial relief with medical therapies or ablative procedures, although patients typically become less responsive or relapse over time. Symptomatic TN (i.e., secondary to another condition such as multiple sclerosis) appears to be less responsive to therapeutic manoeuvres. If vascular compression can be treated intraoperatively, microvascular decompression produces equal or better symptom control with durable treatment efficacy and fewer long-term sequelae.
## Diagnostic guidelines

### Europe

**EFNS guidelines on neuropathic pain assessment, revised 2009**

*Published by:* European Federation of Neurological Societies  *Last published:* 2010

### International

**AAN-EFNS guidelines on trigeminal neuralgia management**

*Published by:* American Academy of Neurology; European Federation of Neurological Societies  *Last published:* 2008

### North America

**ACR Appropriateness Criteria: headache**

*Published by:* American College of Radiology  *Last published:* 2013

**Practice parameter: the diagnostic evaluation and treatment of trigeminal neuralgia (an evidence-based review)**

*Published by:* American Academy of Neurology; European Federation of Neurological Societies  *Last published:* 2008  *(reaffirmed in 2014)*

## Treatment guidelines

### Europe

**Neuropathic pain in adults: pharmacological management in non-specialist settings**

*Published by:* National Institute for Health and Care Excellence  *Last published:* 2014

**EFNS guidelines on the pharmacological treatment of neuropathic pain, 2010 revision**

*Published by:* European Federation of Neurological Societies  *Last published:* 2010

**Spinal cord stimulation for chronic pain of neuropathic or ischaemic origin**

*Published by:* National Institute for Health and Care Excellence  *Last published:* 2008

### International

**AAN-EFNS guidelines on trigeminal neuralgia management**

*Published by:* American Academy of Neurology; European Federation of Neurological Societies  *Last published:* 2008
International

Practice parameter: the diagnostic evaluation and treatment of trigeminal neuralgia (an evidence-based review)

Published by: American Academy of Neurology; European Federation of Neurological Societies

Last published: 2008
(reaffirmed in 2014)
Online resources

1. The Facial Pain Association (external link)
Evidence scores

1. Pain relief: there is poor-quality clinical evidence that carbamazepine for 5-14 days may be more effective at relieving pain compared with placebo but may be less effective at 5-16 years. Carbamazepine is associated with drowsiness, dizziness, constipation, and ataxia. **Evidence level C**: Poor quality observational (cohort) studies or methodologically flawed randomized controlled trials (RCTs) of <200 participants.

2. Pain relief: there is no direct information about oxcarbazepine in terms of its efficacy compared with carbamazepine in the treatment of people with trigeminal neuralgia (TN). Consensus suggests that oxcarbazepine is an effective treatment for TN. **Evidence level C**: Poor quality observational (cohort) studies or methodologically flawed randomized controlled trials (RCTs) of <200 participants.

3. Pain relief: there is no direct information from randomised controlled trials about baclofen in the treatment of people with trigeminal neuralgia (TN). There is consensus that baclofen may be effective in people with multiple sclerosis who develop TN. **Evidence level C**: Poor quality observational (cohort) studies or methodologically flawed randomized controlled trials (RCTs) of <200 participants.

4. Pain relief: consensus suggests that microvascular surgery is effective at reducing symptoms in the long term, although it can lead to ipsilateral hearing loss. However, there is no direct information from randomised controlled trials about microvascular decompression in the treatment of people with trigeminal neuralgia. **Evidence level C**: Poor quality observational (cohort) studies or methodologically flawed randomized controlled trials (RCTs) of <200 participants.

5. Pain relief: there is poor-quality clinical evidence that stereotactic radiosurgery with 1 or 2 isocentres is effective at relieving pain at 26 months. **Evidence level C**: Poor quality observational (cohort) studies or methodologically flawed randomized controlled trials (RCTs) of <200 participants.

6. Pain relief: there is no direct information from randomised controlled trials about lamotrigine in the treatment of people with trigeminal neuralgia (TN). In clinical practice, lamotrigine is often added to carbamazepine when the latter becomes less effective. The dose of lamotrigine must be escalated slowly in order to avoid rashes, and it is therefore not appropriate for acute management of TN. It is most effective when used for long-term control of moderate pain. **Evidence level C**: Poor quality observational (cohort) studies or methodologically flawed randomized controlled trials (RCTs) of <200 participants.
Key articles


References


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