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PRACTICE

SUMMARIES OF BMJ CLINICAL EVIDENCE

Trigeminal neuralgia

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This series comprises summaries of BMJ Clinical Evidence, a database of systematic overviews of the best available evidence on the effectiveness of commonly used interventions (available at http://clinicalevidence.bmj.com/).

Trigeminal neuralgia is a characteristic pain in the distribution of one or more branches of the fifth cranial nerve. Diagnosis is made on history alone: pain occurs in paroxysms, each pain lasts from a few seconds to several minutes, and no pain is experienced in between. The frequency of attacks ranges from four or five to hundreds a day, attacks can occur daily for weeks or months, or there can be months or years of pain remission. Pain is severe and described as intense, sharp, stabbing, or shooting—often like an electric shock. It can be triggered by light touch in any area innervated by the trigeminal nerve. Triggers include eating, talking, washing the face, wind, and cleaning the teeth. The condition can impair activities of daily living and lead to depression. Some people experience background pain of lower intensity for 50% of the time; this form of the disease is termed atypical trigeminal neuralgia, type 2 trigeminal neuralgia, or trigeminal neuralgia with concomitant pain.[1] Neurological examination is usually normal but sensory and autonomic symptoms may be reported,[1] and people with a long history of the disease may show subtle sensory loss.

Key findings: efficacy of interventions

The table⇓ lists the evidence for the effects of ongoing treatments in people with trigeminal neuralgia.

Clinical guide to treatments

- Carbamazepine: Guidelines from the National Institute for Health and Care Excellence (NICE) recommend offering carbamazepine as the initial drug treatment for trigeminal neuralgia.[2 3] Start or stop treatment by changing the dose in increments over several days to reduce common adverse effects. It is associated with a risk of potentially life threatening skin reactions, including Stevens-Johnson syndrome.[4]
- Oxcarbazepine: One non-systematic review found that oxcarbazepine and carbamazepine were associated with similar reductions in attacks of trigeminal neuralgia.[5] Although oxcarbazepine is more expensive than carbamazepine in countries where it is still under patent, many clinicians favour this drug for its lower toxicity profile.
- Lamotrigine: There is insufficient evidence from randomised controlled trials (RCTs) to judge the effectiveness of lamotrigine in people with trigeminal neuralgia.[5] Clinicians often use this drug in people who cannot tolerate carbamazepine (for example, because of allergy) or in addition to carbamazepine when it becomes less effective. The dose must be escalated slowly to avoid rashes, and it is therefore not appropriate for acute management, but is most effective for long term control of moderate pain.[5]
- Gabapentin: Although effective in treating many neuropathic pain conditions, particularly post-herpetic neuralgia, evidence for its use in trigeminal neuralgia is lacking. One RCT showed that gabapentin plus ropivacaine (injected into trigger points) compared with gabapentin alone can improve pain and functional health status in trigeminal neuralgia with little or no side effects.[5]
- Baclofen: Consensus suggests that this may be useful in people with multiple sclerosis who develop trigeminal neuralgia. Such patients are often taking baclofen already and may achieve control of symptoms without having to add carbamazepine. Baclofen is associated with transient sedation and loss of muscle tone. Abrupt discontinuation may cause seizures and hallucinations.
- Although combinations of drugs can be considered, there is no good evidence that patients will achieve durable pain control once an adequate trial of a single first line agent (such as carbamazepine or oxcarbazepine) at maximum tolerable dosage has failed.[5 6] Surgical consultation should be considered at this point.

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The bottom line

- Trigeminal neuralgia is characterised by sharp stabbing pains that are usually unilateral, last for less than a minute, and occur within the distribution of the fifth cranial nerve
- Few high quality, large randomised control trials are available to guide practice
- Initial treatment is usually medical with a single first line agent (such as carbamazepine or oxcarbazepine)
- Consider surgical treatment if an adequate trial of a single first line agent at maximum tolerable dose has failed

- Microvascular decompression: No RCTs exist but observational evidence suggests that pain relief after this procedure is usually immediate,[5] and it is the operation of choice in patients whose pain is no longer controlled by drugs and whose quality of life has markedly deteriorated. The procedure requires general anaesthesia, and its main adverse effect is ipsilateral hearing loss (in less than 5%), which is usually permanent. Its large therapeutic effect size, with 70-80% of people achieving immediate complete pain relief and 60-70% remaining pain free at 10-20 years after surgery,[5] means there are ethical concerns with using RCTs to compare microvascular decompression with medical therapies. It also suggests that microvascular decompression should be considered as first line treatment in certain circumstances (for example, in younger patients, those with major side effects from antiepileptic drugs, and those unable or unwilling to tolerate the potential side effects of antiepileptic drugs). Microvascular decompression has a lower treatment success rate in patients with trigeminal neuralgia related to multiple sclerosis, and it is generally not considered first line surgical treatment in these patients, whose pain may be caused by a different mechanism.[5]
- Stereotactic radiosurgery: This is performed using technologies such as the Gamma Knife, CyberKnife, and linear accelerators with multileaf collimator capabilities (LINAC-MLC). It does not require general anaesthesia (or sedation), but its pain relieving effects are not immediate.[5] Therefore, it is not considered an option for emergency management. Adverse effects include facial numbness, although this is less common than with other palliative destructive procedures. Deafferentation pain after stereotactic radiosurgery is rare but may be a problem with repeat procedures.
- Percutaneous destructive neurosurgical techniques (radiofrequency thermocoagulation, glycerol rhizolysis, or balloon compression): Similar to microvascular decompression, but unlike stereotactic radiosurgery, these techniques can achieve immediate pain relief and can therefore be considered for emergency management.[5] However, the duration of response is shorter than is seen with microvascular decompression,[5] and a brief pulse of heavy sedation and sometimes general anaesthesia is needed. Adverse effects include a small risk of facial numbness, corneal insensitivity, and deafferentation pain, particularly with repeat procedures. In addition, all three procedures carry a small risk of trigeminal-vagal reflex effects on the heart during lesioning and a very small risk of carotid injury or intracranial infection. Balloon compression carries a risk of temporary trigeminal motor dysfunction.
- Before repeating any surgical techniques for recurrent trigeminal neuralgia it is crucial to go through the pain history again and determine whether drugs might be sufficient. Repeated stereotactic radiosurgery requires a much lower dose of radiation, to minimise numbness as a complication. Percutaneous destructive neurosurgical techniques can also be repeated. However each re-treatment with this or with stereotactic radiosurgery is associated with a cumulative risk of trigeminal de-afferentation.[5]
- People with trigeminal neuralgia should be managed in specialist centres with multidisciplinary teams. Contact with other people with the condition through approved support groups can also be beneficial.

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Table

Table 1| Evidence for the effects of ongoing treatments in people with trigeminal neuralgia

Effects	Treatment
Likely to be beneficial	
Systematic reviews, randomised controlled trials, or the best alternative source of information have shown some effectiveness, although this has not been fully established; benefits are likely to be greater than harms	Carbamazepine
	Oxcarbazepine*
	Baclofen (in people with multiple sclerosis who develop trigeminal neuralgia)*
Trade off between benefits and harms	
Clinicians and patients should weigh up beneficial and harmful effects according to individual circumstances and priorities	Microvascular decompression*
	Non-percutaneous destructive neurosurgical techniques (stereotactic radiosurgery)*
	Percutaneous destructive neurosurgical techniques (radiofrequency thermocoagulation, glycerol rhizolysis, and balloon compression)*
Unknown effectiveness	
Data are currently insufficient or of inadequate quality	Lamotrigine
	Gabapentin

*Categorisation based on observational studies or consensus, or both.