

# Craniofacial Pain: Trigeminal Neuralgia




# 49

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## Introduction

- Trigeminal neuralgia is a neuropathic pain in the face and characterized by recurrent **brief episodes of unilateral electric shock-like** [1] pains along the distribution of one or more divisions of the trigeminal nerve. Pain is usually triggered by innocuous stimuli.



-  Ophthalmic branch
-  Maxillary branch
-  Mandibular branch

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- **Trigeminal nerve:** provides sensory supply to the face and the sensory and motor supply to the muscles of mastication. It has three major

divisions: Ophthalmic (V1), Maxillary (V2), and Mandibular (V3) branches

- These three peripheral branches join at the **Gasserian ganglion** (a.k.a. trigeminal ganglion, semilunar ganglion, or Gasser's ganglion) ⇒ trigeminal nerve root ⇒ trigeminal nerve nucleus in the pons

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## Clinical Manifestations

- Most common cause of chronic facial pain after age 50
- Incidence: females > males
- Usually **unilateral**, if bilateral, exclude multiple sclerosis
- Ophthalmic branch is **least affected**
- Pain is paroxysmal, **electric shock** like, stabbing, lasts few seconds to few minutes, and occurs repetitively up to 50–60 times a day. Pain can also present as continuous dull pain with paroxysms of pain. Pain can be reproduced by light touch on the affected area (trigger zone), chewing, talking, contact with a brush, cold air, smile etc. Pain may be accompanied by salivation, lacrimation or rhinorrhea.
- Primary Trigeminal Neuralgia
  - Usually due to compression of the trigeminal nerve root during its entry into the pons by an aberrant loop of an artery or vein, most commonly the **superior cerebellar artery**
- Secondary Trigeminal Neuralgia
  - vestibular schwannoma (acoustic neuroma), meningioma, multiple sclerosis, brain stem lesions or AV malformation.
- Differential diagnosis
  - Postherpetic neuralgia
  - Dental pain (usually continuous; **no paroxysm**)
  - Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
  - Cluster headache
- Imaging (MRI head) identifies neurovascular compression or a structural brain lesion (eg, tumor in the cerebellopontine angle,

demyelinating lesions including multiple sclerosis).

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## Treatment

- First line agents:
  - **Carbamazepine** [2]
    - NNT = 4
    - 200–1200 mg/day
    - SE: low Na, low WBC (agranulocytosis), aplastic anemia, high LFTs (hepatotoxicity)
  - Oxcarbazepine
    - SE: similar to carbamazepine but less severe.
    - 600–1800 mg/day
- Second line agents:
  - Gabapentin/Pregabalin
  - Lamotrigine – need to titrate the dose over many weeks, given the risk of rash and other serious adverse effects
  - Baclofen –The starting dose of baclofen is 5 mg TID, with gradual titration to a maintenance dose of 50–60 mg per day. The drug should be discontinued slowly to avoid withdrawal.

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## Interventions

- Trigeminal nerve blocks
  - Target depends on pain distribution, may include a single peripheral branch or Gasserian ganglion block
  - Targets for peripheral branch blocks
    - Ophthalmic pain = **V1 = supraorbital foramen**
    - Maxillary pain = **V2 = foramen infraorbitale**
    - Mandibular pain = **V3 = mandibular notch or foramen mentale**
- Radiofrequency ablation
  - May be performed at Gasserian ganglion or over peripheral branches of Trigeminal nerve
  - SE: **anesthesia dolorosa** (or deafferentation pain) is neuropathic pain in an area

(usually in the face) that is numb to touch. Most commonly associated with treatment for trigeminal neuralgia.

- Methods (approach for Gasserian ganglion block and RFA): patient supine, submentovertex position, needle insertion ~3 cm from ipsilateral angle of the mouth, needle with 30-degree caudal incline, advance towards **foramen ovale**, confirm the position with fluoroscopy, motor testing, and sensory testing.
- Microvascular decompression: major neurosurgical procedure with craniotomy and the removal of various vascular structures, e.g., superior cerebellar artery, away from the trigeminal nerve.
- Gamma Knife radiosurgery: radiation beams are aimed at trigeminal nerves and the procedure is carried out with a stereotactic frame MRI. The beams cause axonal degeneration and necrosis.

### Clinical Pearls

- The trigeminal nerve is a mostly sensory cranial nerve with motor innervation only to the muscles of mastication
- Trigeminal neuralgia is **usually unilateral**, affects females > males; bilateral symptoms should raise concern for MS
- MRI of the head is recommended to identify neurovascular compression or brain lesion
- First-line treatment is Carbamazepine (200–1200 mg/day). Know side effects.
- Trigeminal nerve branch blocks and ablation, as well as **Gasserian ganglion blocks** and ablation, are indicated for cases refractory to medical management
- Microvascular decompression at the Gasserian ganglion is the **most successful** surgical treatment [3]
- Anesthesia dolorosa is a relatively rare side effect of trigeminal nerve trauma or surgery resulting in spontaneous pain signals without nociceptive stimuli

### Questions

1. Which of the following branches of the trigeminal nerve is least likely to be affected in cases of Trigeminal Neuralgia?
  - A. V1
  - B. V2
  - C. V3
  - D. Each branch is equally affected
2. A 50-year-old woman presents reporting many weeks of short bursts of facial burning and tingling affecting the right face. She is unsure as to whether or not she has symptoms on the left side of her face as well. Her neurological exam is normal. What is the next best step?
  - A. Refer for electrodiagnostic testing of the trigeminal nerve
  - B. Order MRI of the head
  - C. Trial of oral Carbamazepine
  - D. Diagnostic block of the maxillary branch of the trigeminal nerve
3. The same woman returns to your clinic after a full work-up and has failed treatment with Carbamazepine. She elects for a trigeminal nerve branch block. Given the distribution of her symptoms, you recommend blocking only the maxillary branch. What is the anatomical target for this block?
  - A. Supraorbital foramen
  - B. Foramen infraorbitale
  - C. Mandibular notch
  - D. Foramen mentale
4. The same woman has not had any long-standing relief despite medical treatment and trigeminal nerve branch blocks. She elects for a surgical referral. Which surgical technique is most likely to confer the longest benefit in cases of idiopathic trigeminal neuralgia?
  - A. Gamma-knife surgery
  - B. Partial sensory rhizotomy
  - C. Balloon compression
  - D. Microvascular decompression
5. After successful surgical treatment of her symptoms, the same woman now presents with intermittent pain on the right side of her

face, though the skin itself is numb. What is the most likely diagnosis?

- A. Post-herpetic neuralgia
- B. Temporomandibular disorder
- C. Anesthesia Dolorosa
- D. Multiple Sclerosis

- 2. Zakrzewska JM, Linskey ME. Trigeminal neuralgia. *BMJ*. 2014;348. <https://doi.org/10.1136/bmj.g474>.
- 3. Bendtsen L, et al. European Academy of Neurology guideline on trigeminal neuralgia. *Eur J Neurol*. 2019; 26(6):831–49. <https://doi.org/10.1111/ene.13950>.

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## Correct Answers

1. A, 2. B, 3. B, 4. D, 5. C

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## References

- 1. Trigeminal neuralgia. [http://www.openanesthesia.org/aba\\_trigeminal\\_neuralgia/](http://www.openanesthesia.org/aba_trigeminal_neuralgia/). Accessed 6 May 2020.

## Recommended Reading

- Bendtsen L, Zakrzewska JM, Abbott J, et al. European Academy of Neurology guideline on trigeminal neuralgia. *Eur J Neurol*. 2019;26(6):831–49. <https://doi.org/10.1111/ene.13950>.
- Wall PD, McMahon SB, Koltzenburg M. Wall and Melzack's textbook of pain. Philadelphia: Elsevier/Churchill Livingstone. 2006.