Trigeminal Neuralgia

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Trigeminal Nerve Anatomy

- Largest cranial nerve.
- Great sensory nerve of the head and face.
- Motor nerve of the muscles of mastication.



Trigeminal Nerve Nuclei and Function

Motor (masticator) \bullet nucleus: Inervation of the muscles of mastication (ie, masseter, temporalis, medial and lateral pterygoid muscles, plus tensores tympani, tensores veli palatini, mylohyoid, and anterior belly of the digastric muscles



 Sensory roots arise from the trigeminal ganglion (gasserian ganglion, semilunar ganglion)



Trigeminal Nerve Nuclei and Function

Sensory nuclei

- Spinal trigeminal: Pain and temperature Simple touch
- Pontine trigeminal: discriminative touch
- Mesencephalic: Propioception Vibration sense



Meckel's Cave





Ophtalmic Nerve (V1)



Sensory nerve.

It supplies branches to the cornea, ciliary body, and iris; to the lacrimal gland and conjunctiva; to the part of the mucous membrane of the nasal cavity; and to the skin of the eyelids, eyebrow, forehead, and nose.

Maxillary Nerve (V2)

- Sensory nerve
- It leaves the skull through the foramen rotundum into the pterygopalatine fossa.
- It enters the orbit through the inferior orbital fissure.
- It appears upon the face at the infraorbital foramen.



Branches

In the Cranium......Middle meningeal. In the Pterygopalatine Fossa...Zygomatic.Sphenopalatine.Posterior superior alveolar. In the Infraorbital Canal......Anterior superior alveolar.Middle superior alveolar. On the Face.....Inferior palpebral.External nasal.Superior labial.

Mandibular Nerve (V3)

It supplies

- Teeth and gums of the mandible
- Skin of the temporal region, the auricula, the lower lip, the lower part of the face.
- Muscles of mastication.
- Mucous membrane of the anterior two-thirds of the tongue.





Clinical Presentation

- Paroxysmal, "electric" pain in trigeminal distribution on one side of the face.
- Presence of trigger areas.
- Periods of remission and exacerbation.
- Pain is more severe in the morning and absent during sleep.
- Periodic pain relief when treated with an adequate trial of CBZ.

Pathophysiology

- Demyelination of large-diameter A fibers at the trigeminal root entry zone.
- Ephaptic transmission from these fibers to poorly myelinated Aδ and unmyelinated C fibers, which results in paroxysmal facial pain.
- Demyelinitation may come from compression of the CN5 by an artery or vein at the root entry zone.

Epidemiology

- Female-to-male ratio 2:1
- Age: \geq 50 years.
- Incidence: 4 per 100,000 people per year.
- More common location: V2, V3 distribution.

Neurological Exam

- It is usually normal.
- Remember: Patients with MS or PC angle tumors can present with classic features of trigeminal neuralgia.

Facial Pain Etiologies

Structure	Examples
Nerve	Trigeminal neuralgia Postherpetic neuralgia Trigeminal neuropathic pain Glossopharyngeal neuralgia Sphenopalatine neuralgia Geniculate neuralgia (Ramsay Hunt)
Teeth and Jaw	Dentinal, pulpal, or periodontal pain Temporomandibular disorders
Sinuses and aerodigestive tract	Sinusitis Head and neck cancer
Eyes	Tolosa-Hunt syndrome Optic neuritis Iritis Glaucoma
Vessels	Giant cell arteritis Migraine Cluster headache
Psyche	Psychogenic facial pain Atypical facial pain

Diagnostic Clues in Facial Pain

Diagnosis	Pain Character	Pain Distribution	Pain Triggers	Other clues
Trigeminal neuralgia	Paroxysmal, lancinating	Trigeminal only V2 most frequent	Touch, chewing, talking, etc	
Glossopharyn geal neuralgia	Paroxysmal, lancinating	Ear, throat	Swallowing	
Trigeminal neuropathic pain	Constant, burning, dull throbbing	Trigeminal only	None	Hx of trigeminal nerve injury
Postherpetic neuralgia	Constant, crawling May have paroxysmal component	Trigeminal only V1 most frequent	Touch	Hx of herpes zoster ophtalmicus
Anesthesia dolorosa	Constant, burning, itching in an insensate region	Trigeminal only	None	Hx of trigeminal nerve lesion
Malignancy	Constant May have paroxysmal component	In area of neoplasm or referable to nerve compression	Possible if trigeminal nerve involved	Head/neck neoplasm
Atypical facial pain	Constant	Nonanatomic, often bilateral	None	Prominent psychiatric component

Consider the difference between....

- Typical trigeminal neuralgia.
- Atypical trigeminal neuralgia.
- Atypical facial pain.

Medical Management

The initial treatment is usually medical

Carbamazepine:

- Initial response of TN to CBZ is virtually universal. Lack of response Reassessment
- A small percentage of patients cannot tolerate its side effects.
- Efficacy may have a gradual decline.
- Dosing: Starting dose: 100-300 mg/day Therapeutic range: 800-1200 mg/day
- Before initiating administration, a baseline complete CBC and liver and renal function tests should be obtained.
- Side effects: 20-40% of patients. Early dose-related side effects: somnolence, dizziness, nausea, and nystagmus. Dermatologic reactions: 5%-10% of patients. Rash, erythema multiforme, and Steven-Johnson syndrome. Hemathologic: 2%-6% Aplastic anemia. Others: hepatotoxicity, hyponatremia, and CHF.
- Interactions: other AEDs. No significant interaction with baclofen.

Phenytoin

- Pain relief starts within 2 days of onset of therapy.
- Dose: 5-7 mg/Kg/day
- No controlled trials comparing CBZ to phenytoin.
- Response rate: 25%-60%.
- Side-effects: ataxia, drowsiness, nystagmus, and diplopia.
- Other side effects: gingival hyperplasia, acne, hirsutism, morbilliform rash, Steven-Johnson syndrome, hepatitis, lupus-like syndrome, and folate-responsive megaloblastic anemia.
- Drug interactions: loosely bound to hepatic cytochrome P-450 → susceptible to competitive displacement → monitoring of serum levels.

Baclofen

- GABA analogue
- Rapid gastrointestinal absorption: serum peak achieved in 2-3 hours.
- Half life 3-4 hours.
- Dose: initial dose: 10 mg TID. Incrementally increased until pain relief/toxicity. Usual maintenance dose in TN: 50-60 mg/day.
- Synergism of baclofen with CBZ and phenytoin supports the use of combination therapy.
- Side effects: somnolence, dizziness, and gastrointestinal distress.
- Baclofen does not have the life-threatening side effects of CBZ or phenytoin.
- Withdrawal of medication: should be gradual to prevent seizures and hallucinations.
- Interactions: Not known.

Oxcarbazepine

- Keto derivative of CBZ.
- Metabolized to an active compound whose halflife is 14 to 26 hours.
- Small series have shown benefit in patients refractory to CBZ.
- Less significant toxicity compared to CBZ.
- Dosing is similar to that for CBZ.

Zakrzewska JM et al. J Neurol Neurosurg Psychiatry 52:472-476, 1989.

Surgical Treatment Options



Radiosurgery

History

- 1950s Taarnhoj, Shelden, and Pudenz: observed that patients with numbness (nerve injury) after middle fossa craniectomy had better results. They proposed middle fossa surgery for compression of the ganglion.
- John Mullan developed the percutaneous approach for compression.

Indications

- •Medically refractory TN
- •Patient suitable for a brief general anesthesia
- •Patient understands and accepts that compression is an ablative procedure that will cause facial numbness

Preoperative considerations

- Brain MRI
- EKG: Identification of patients at risk of intraoperative arrhythmias.
- Appropriate plan to discontinue warfarin and antiplatelet medications.
- Consider preoperative acyclovir for patients with known history of HSV infections.

Anesthesia

- •Avoid atropine before surgery.
- •General anesthesia is induced with propofol and maintained with isoflurane.
- •External pacemaker on the patient's chest.

Positioning and equipment placement

- Neck in neutral position with a soft support.
- Avoid neck manipulation mean age of the patients: 65 year old.
- Keeps the area of the head free to provide space for the imaging unit.

Intraoperative imaging

Three views are used: modified sub-mental, modified anteroposterior (AP), and lateral view.

Operative Procedure

- Entry point: 2.5 cm lateral from the angle of the mouth.
- The cannula is passed percutaneously and directed to the foramen ovale by using fluoroscopic imaging.
- Confirmation that the cannula has engaged the foramen ovale: images, tactile feedback, and depressor response (bradycardia).
- The cannula should not pass beyond the foramen ovale.
- The blunt obturator is replaced by a straight guiding stylet which is pointed to the center for V2, lateral for V3, and medial for V1. The stylet is advanced to the entrance of the Meckel's cave (17 to 22 mm beyond the foramen ovale).
- The stylet is withdrawn and the balloon is inserted.
- The balloon is inflated with 0.7 to 0.75 mL of 180 mg% of iohexol to reach a compression pressure of 1065 to 1215 mmHg. Pear shape is observed. The balloon is left inflated for 1 to 1.5 min.
- The needle and catheter are removed.





Results

- Initial success: 93%
- Preserves poorly myelinated and unmyelinated fibers of the blink reflex: especially indicated in V1 pain distribution.
- Subjective numbness after surgery: 61%
- Jaw weakness: 19%. More common than with thermal rhizotomy or glycerol injection.
- Recurrence at 2 years 21%. Higher risk in patients with little sensory loss after surgery.
- Unusual complications: SAH, carotid-cavernous fistula.

Percutaneous Glycerol Rhizotomy

- Jefferson: First injection of phenol mixed with glycerol for the ablation of TN in 1963.
- Håkanson: injected glycerin and tantalum into the trigeminal cystern as a radiographic marker for subsequent gamma knife radiosurgery. Patients improved before the radiosurgical procedure was performed.
- Mechanism of action: Glycerol injures both myelinated and unmyelinated axons when injected into the nerve.
- Injection of 0.1 to 0.4 mL of glycerol into the trigeminal cistern.

Percutaneous Glycerol Rhizotomy

- Anesthesia: Sedation. GA is required in less than 1%
- Pain relief is immediate or within the next seven days.
- Recurrence is common: 92% at six years.
- Complications: meningitis, sensory loss, anesthesia dolorosa, posterior fossa injury (deafness) and persistent pain.

Percutaneous Radiofrequency Rhizotomy

- RF energy is used to cause a thermal injury in a selected rootlet of the TN.
- Introduced by Sweet 40 years ago.
- Results are better in V2 and V3 distribution cases.
- Avoid in V1 cases higher risk of corneal problems.
- Anesthesia: Sedation. Verbal feedback from the patient is required.

Percutaneous Radiofrequency Rhizotomy



A 20 gauge needle is placed in the foramen ovale following regular landmarks. The electrode is inserted and its tip is placed depending on the targeted division.

Electrical stimulus with the patient awake confirms the selective placement of the electrode.

Lesion: increase of sedation; repeat until the desired effect is achieved. Immediate pain relief: 99%.

Pain recurrence: 15%-20% in 10 to 15 years.

Taha JM et al, J Neurosurg 83:989-993,1995

• Goal: treatment the underlying cause compression of the nerve by arteries or veins from the root entry zone to the exit into Meckel's cave.



Technique

- General anesthesia.
- Monitoring of brain stem and CN 8 function: BSAEPs.
- Positioning:
 - Lat. decubitus
 - Three-point head holder.
 - Pressure points are padded.
 - Axillary roll.

Neck: minimally stretched, mild flexion and rotation (10°), vertex is tilted down (10°).

The point of these maneuvers is to expose the occipitalboss.

Shoulder: taped down and out of the way.



Jannetta PJ, et al. Neurosurg Focus 18(5):E5, 2005

Technique

- Retromastoid region: shaving and cleaning.
- Incision: straight or slightly curved, 5-6 cm, behind the hairline.
- Landmarks to keep in mind: Position of sinuses, digastric groove (intersection of the sinuses).
- Burr hole: a finger-breadth posterior and caudal to the junction of sinuses.
- Craniectomy: Isosceles triangle with the hypotenuse facing away from the sinuses.
- Mastoid air cells should be waxed.
- Durotomy: curvilinear.

Ext. auditory canal



Inion



Technique

- Cerebellar relaxation: opening of lumbar drain, opening of subarachnoid space in the basal cisterns.
- Entering the supralateral angle of the posterior fossa: bridging veins are visualized.
- The superior petrosal venous complex may have to be partially or totally sacrificed to expose the CN5 which is underlying these vessels.
- CN5 should be exposed entirely from pons to the Meckel's cave entry site.
- Identification of arteries and veins that may be compressing the CN5: SCA, veins, ectatic basilar artery.
- Placement of Teflon.
- Hemostasis Valsalva maneuver.
- Dural closure.

Complications

Results

- Complication rate is less than 5%
- Hematoma (0.1%)
- Significant edema (0.4%)
- Infarction (0.1%)
- Hydrocephalus (0.1%)
- Seventh CN paralysis (1.7%)
- Eighth CN injury (1.4%)
- CSF leak (1.6%)
- Meningitis (0.2%)
- Death (0.2%)

- Complete immediate pain relief in 84%
- Partial relief in 13%
- Complete relief at 5 years in 75%.
- Partial relief at 5 years in 8-10%.

Kassam A, and Horowitz M. Posterior fossa procedures for cranial neuralgias. In Follet KA (ed). Neurosurgical Pain Management. 2004,pp 232-236

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THE LONG-TERM OUTCOME OF MICROVASCULAR DECOMPRESSION FOR TRIGEMINAL NEURALGIA

Fred G. Barker II, M.D., Peter J. Jannetta, M.D., D.Sc., David J. Bissonette, P.A.-C., Mark V. Larkins, M.D., and Hae Dong Jho, M.D., Ph.D.

- 1185 patients during a 20 year period (median follow-up 6 years)
- Immediate post-operative results: complete relief in 82%, partial relief in 16%, and absent in 2%.
- One year after surgery: complete relief in 75% (excellent outcome), and partial relief in 9% (good outcome).
- Ten years after surgery: excellent results in 64% and good results in 4%.
- Predictors of recurrence: female gender, symptoms lasting more than eight years, venous compression, and lack of immediate pain relief.

Systematic Review of Ablative Neurosurgical Techniques for the Treatment of Trigeminal Neuralgia

 Comparison of the different ablative techniques: radiofrequency thermocoagulation, glycerol rhizolysis, balloon compression and stereotactic radiosurgery.

Series (ref. no.)	Technique	No. of patients	Follow-up period (mo)	MS (%)	Atypical (%)	Preop. dysf. (%)	Prev. Sx (%)	Ops
Latchaw et al., 1983 (22)	RFT	96	60	5	45	NS	51	NS
Kanpolat et al., 2001 (18)	RFT	1216	60	4.5	NS	NS	25	0.3
Oturai et al., 1996 (36)	RFT	185	96	NS	NS	NS	28	NS
Zakrzewska et al., 1999 (57)	RFT	48	30	0	35.4	0	0	0
North et al., 1990 (34)	GR	85	36	4.7	12.9	NS	38.8	NS
Slettebo et al., 1993 (45)	GR	60	54	10	15	15	65	0.86
Brown et al., 1993 (6)	BC	50	36	10	NS	NS	52	NS
Maesawa et al., 2001 (28)	SRS	220	22	0	7.3	37.8	61.4	1.5
Pollock et al., 2002 (39)	SRS	117	26	0	7	42	58	1.6

TABLE 2. Characteristics of the patient population included in the outcome analysis^a

^a RFT, radiofrequency thermoccagulation; GR, glycerol rhizolysis; BC, balloon compression of the gasserian ganglion; SRS, sterectactic radiosurgery; NS, not stated; follow-up period, median/mean follow-up period for the series; MS, percentage of patients with multiple sclerosis included in the analysis; Atypical, percentage of patients with atypical trigeminal neuralgia included in the analysis; Preop. dysf., percentage of patients with preoperative sensory deficits; Prev. Sx., percentage of previously surgically treated patients; Ops, mean number of previous operations per patient.

Lopez BC et al. Neurosurgery 54:973-983,2004

Systematic Review of Ablative Neurosurgical Techniques for the Treatment of Trigeminal Neuralgia

- RFT provides better rates of complete pain control.
- SRS is associated with the fewest complications.
- Insufficient good quality data on BC



Lopez BC et al. Neurosurgery 54:973-983,2004