# Trigeminal Neuralgia

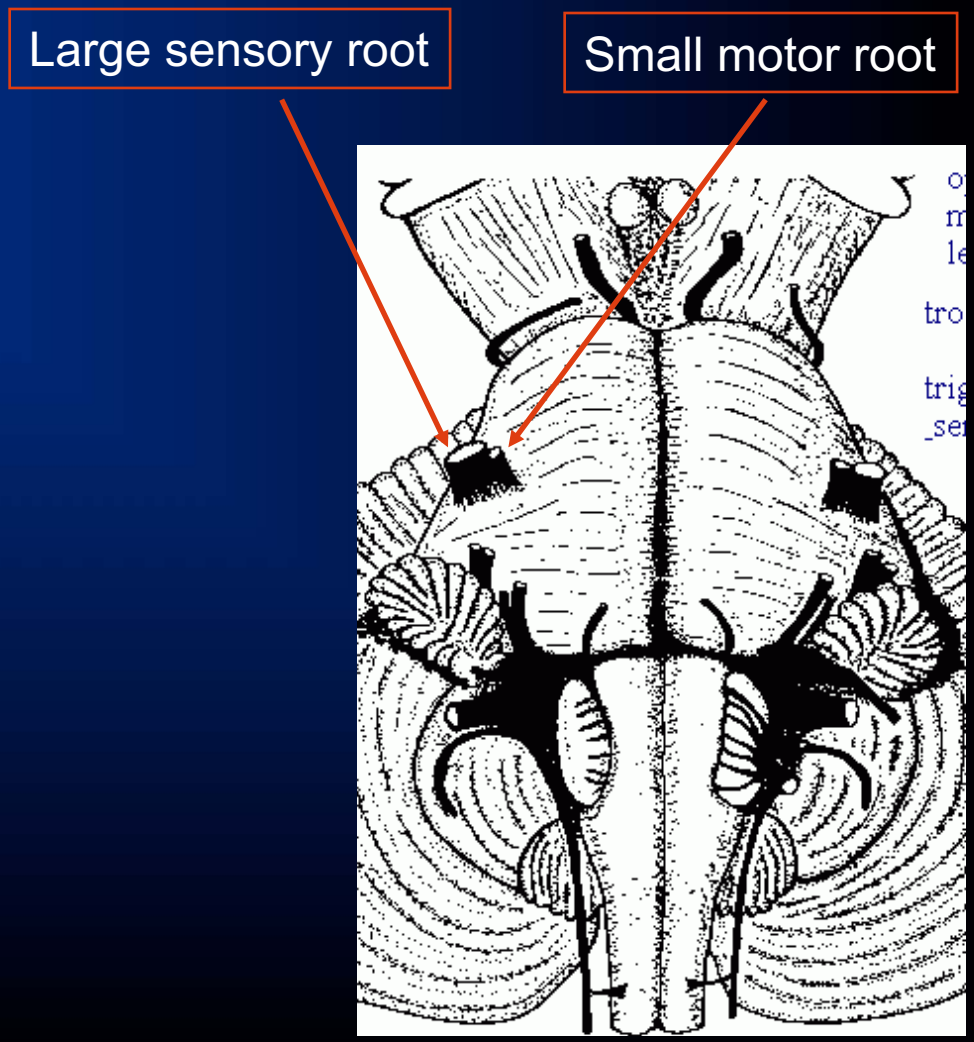
Neurosurgery Academic Half Day
January 17, 2011

# Outline

- CN V Anatomy
- TGN – Clinical Presentation
- TGN – Management
- Other CN Compressive Neuropathies

# Trigeminal Nerve

- "*three twins*"
- Largest CN
- Embryologically derived from 1st branchial arch
- Sensory nerve of face
- Motor nerve for muscles of mastication
- 3 major divisions
  - Ophthalmic V1
  - Maxillary V2
  - Mandibular V3



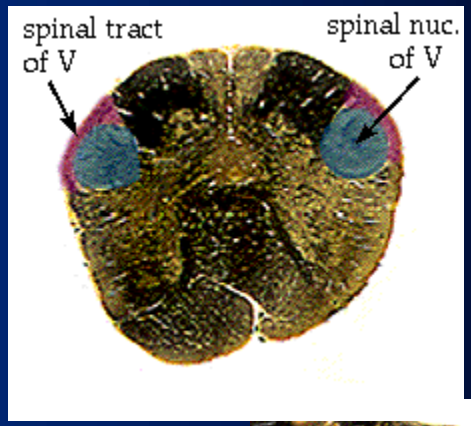
# Trigeminal Nerve

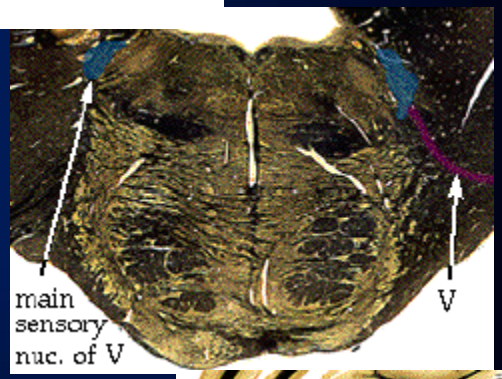
- General Sensory
  - Spinal trigeminal nucleus
    - Pain, temperature
    - Simple touch
  - Pontine trigeminal nucleus
    - Discriminitive touch
  - Mesencephalic nucleus
    - Proprioception
    - Vibration sense
- Branchial Motor
  - Motor (masticator) nucleus, medial to pontine trigeminal nucleus
    - Masseter
    - Temporalis
    - Medial/lateral pterygoid
    - Tensores tympani
    - Tensores veli palatini
    - Mylohyoid
    - Anterior belly of digastric

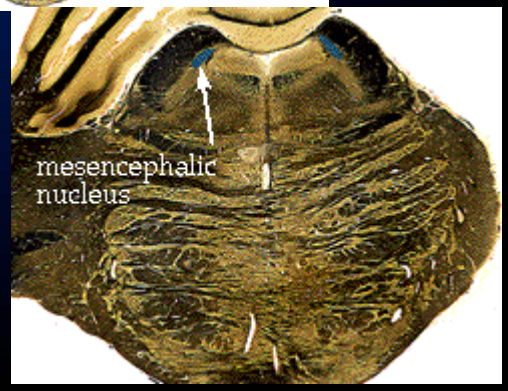
# Trigeminal Nerve Nuclei and Function

Sensory nuclei

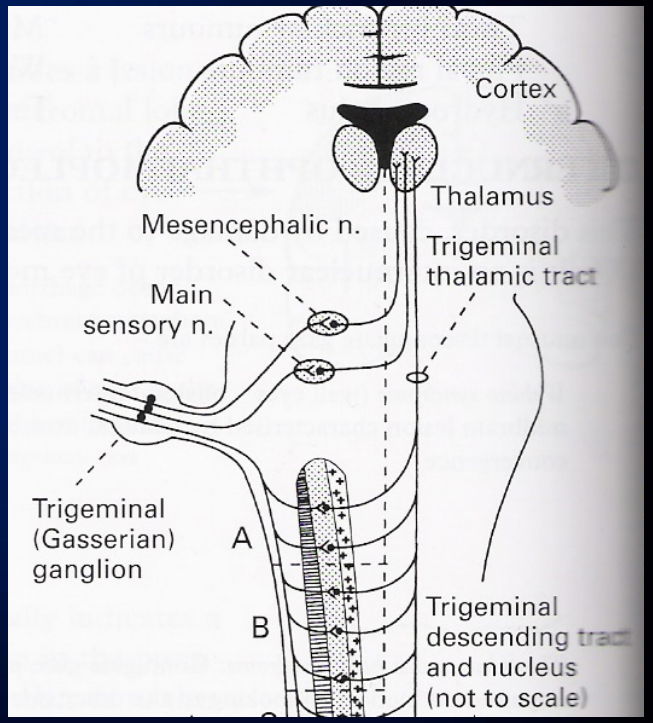
- Spinal trigeminal:
  Pain and temperature
  Simple touch
- Pontine trigeminal:
  discriminative touch
- Mesencephalic:
  Proprioception
  Vibration sense



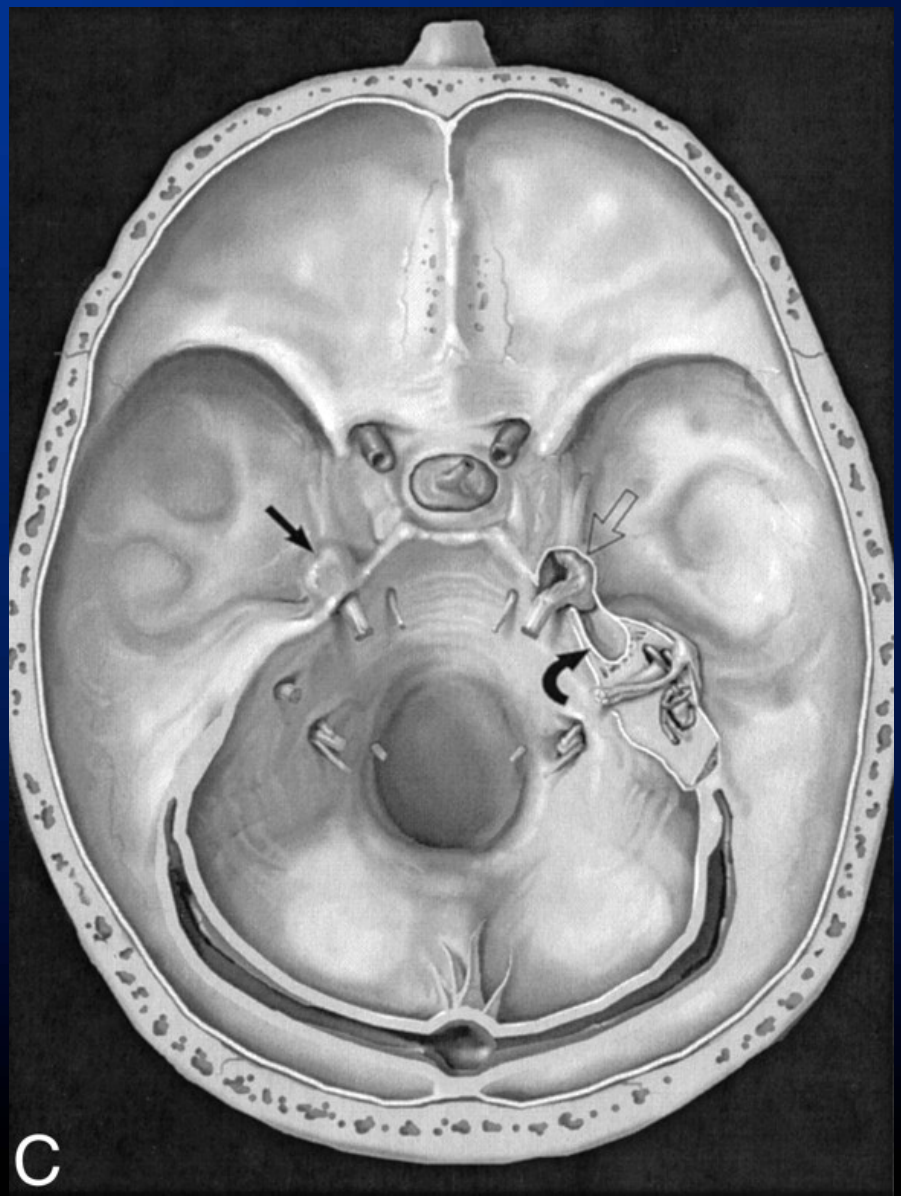



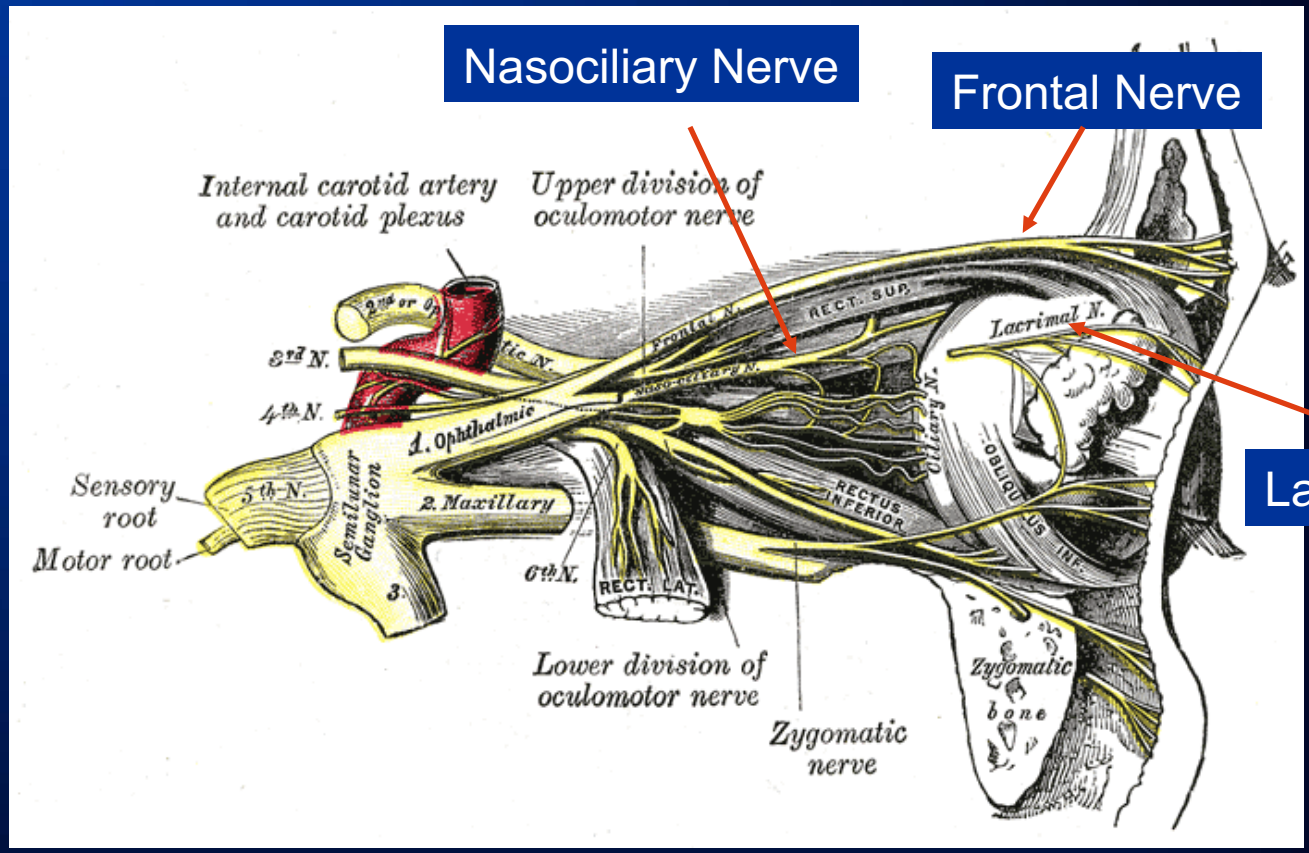
- Gasserian ganglion aka semilunar ganglion residing in Meckel's cave



# Meckel's Cave



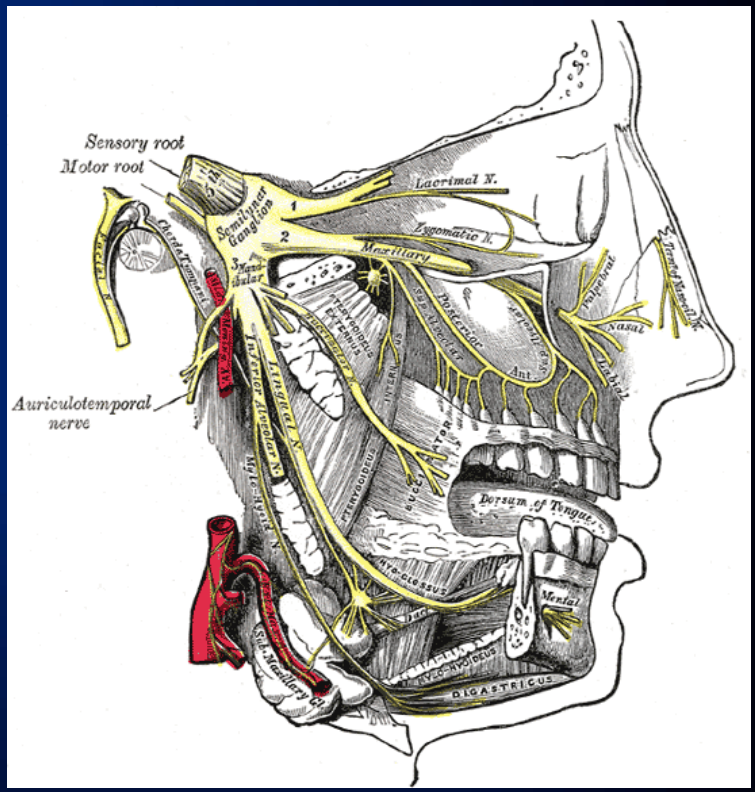
# Ophthalmic Nerve (V1)




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.



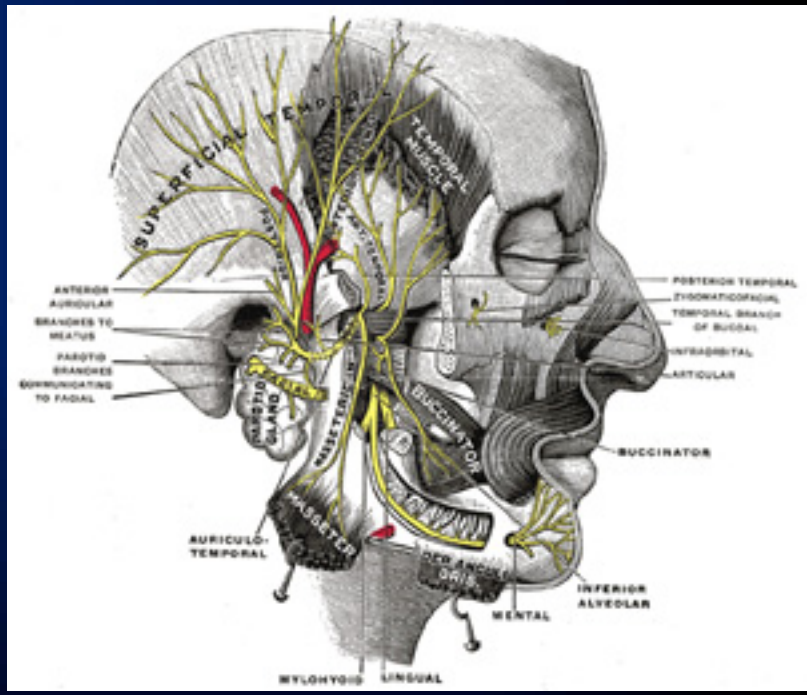

Branches

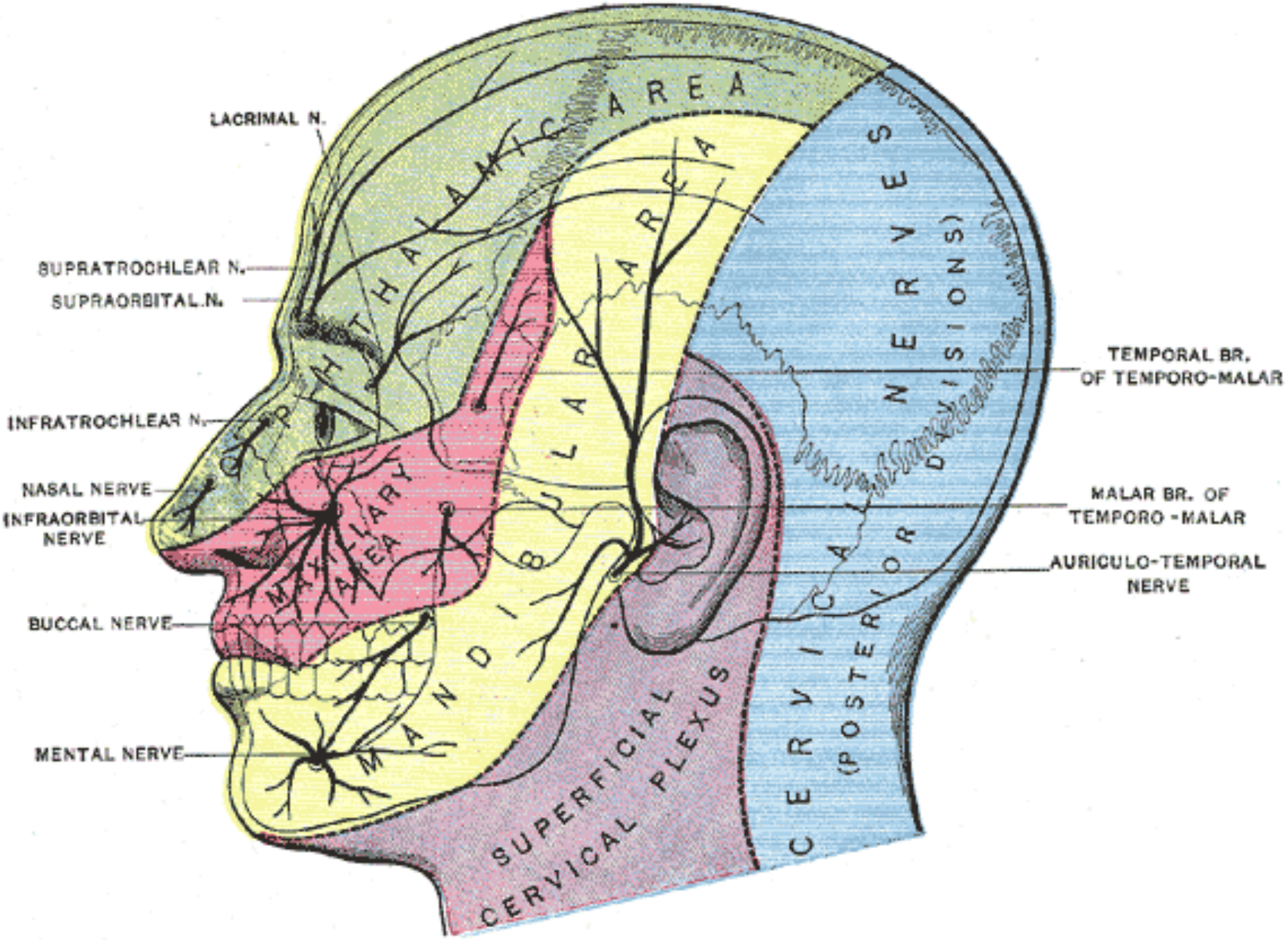
In the Cranium……Middle meningeal.
In the Pterygopalatine Fossa…Zygomatic. Sphenopalatine. Posterior superior alveolar.
In the Infraorbital Canal……Anterior superior alveolar. Middle superior alveolar.
On the Face……Inferior palpebral. External nasal. Superior labial.

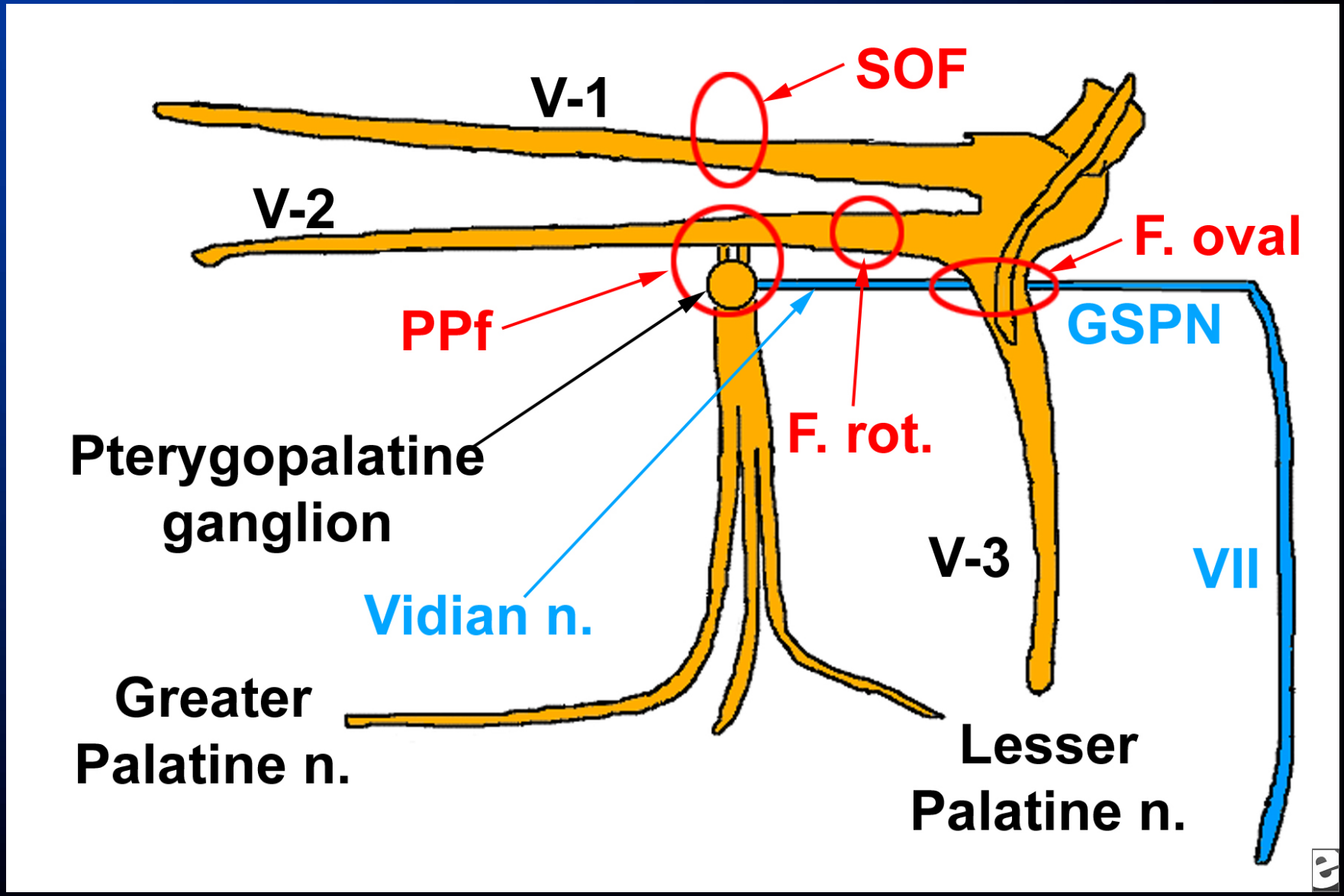
# Mandibular Nerve (V3)

It supplies

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




LACRIMAL N.
SUPRATROCHLEAR N.
SUPRAORBITAL N.
INFRATROCHLEAR N.
NASAL NERVE
INFRAORBITAL NERVE
BUCCAL NERVE
MENTAL NERVE
OPHTHALMIC AREA
MAXILLARY AREA
MANDIBULAR AREA
SUPERFICIAL CERVICAL PLEXUS
CERVICAL NERVES (POSTERIOR DIVISIONS)
TEMPORAL BR. OF TEMPORO-MALAR
MALAR BR. OF TEMPORO-MALAR
AURICULO-TEMPORAL NERVE


SOF
V-1
V-2
F. oval
PPf
GSPN
F. rot.
Pterygopalatine ganglion
V-3
VII
Vidian n.
Greater Palatine n.
Lesser Palatine n.

# CN Compressive Neuropathies

- Trigeminal neuralgia
- Hemifacial spasm
- Glossopharyngeal neuralgia
- Geniculate neuralgia
- Spasmodic torticollis

# Trigeminal Neuralgia

What is it?

# TGN – Historical Perspective

- Fallopius: documented trigeminal nerve in 16th century in an anatomical study
- John Locke: American physician in Paris – gave a full detailed description of TGN. The patient was the Countess of Northumberland, wife of British Ambassador to the French Court
- Nicolas Andre: called the clinical entity "Tic Douloureaux" in 1756 describing 5 patients who suffered from a "*cruel and obscure illness which causes in the face some violent motions, some hideous grimaces, which are an insurmountable obstacle to the reception of food, which putt off sleep*"

# TGN – Historical Perspective

- Fothergill's disease: after eponymous London physician in 1776
- Trousseau: 1853 suggested that the paroxysmal nature of TGN was due to abnormal conduction and called it '*neuralgia epileptiform*'

# TGN - Clinical Features

- Female:Male 2:1
- Age: > 50; mean age 63
- Incidence: 4 per 100,000
- Paroxysmal recurrent pain on one side of face of short duration (few seconds), "*electric*"
- Triggered by sensory stimuli
- Recurrent episodes of pain with progressively shorter periods of remission
- Pain worse in morning, absent in sleep

# TGN - Clinical Features

- Association between MS and TGN
  - 2% of patients with MS have TGN
  - 18% of patients with bilateral TGN have MS
  - Related to MS plaque
  - Typically less responsive to procedures
- Usually some relief with carbamazepine
- Rarely, status trigeminus – rapid succession of tic-like spasms triggered by seemingly any stimulus

# TGN – Clinical Features

- Distribution of pain:
  - Localized to 1 or more branches of CN V
  - V2/V3 combination most common (45%); V1 alone rare (2%)
  - R (60%) >L (39%); bilat 1%
- Clinical examination: usually normal
  - May have mild sensory loss
  - CPA tumours and MS can present with TGN

# TGN – Clinical Features

- Classical TGN or “idiopathic” TGN
  - No structural lesion
  - May have vascular contact
- Symptomatic TGN
  - Tumour
  - MS plaque
  - Skull base structural abnormalities

# TGN - DDx

- Dental/sinus-related pain
- Cluster h/a, migrainous neuralgia
- Atypical facial pain
- Post-herpetic neuropathy
- TMJ dysfn (degenerative vs. RA)
- Trigeminal neuropathy
  - Demyelination i.e. MS
- Facial myalgia

# TGN

What causes it?

# Pathophysiology

- Ephaptic transmission from large myelinated A fibers to poorly myelinated A delta and C (nociceptive) fibers

# Pathophysiology

- Compression at root exit zone: at junction of central myelin (oligodendroglial cells) and peripheral myelin (Schwann cells) aka the Obersteiner-Redlich zone; spontaneous discharge at site of compression
  - Vascular cross compression of Root Exit Zone (classically SCA 80%); atherosclerotic degeneration can cause elongation and tortuosity; venous compression; persistent primitive trigeminal artery, dolichoectatic basilar artery
  - Neoplastic compression (<0.8%) i.e. meningioma, schwannoma, neuroma, epidermoid
- MS plaque/demyelination in brainstem

# Preop imaging: detecting vascular compression, r/o tumour/plaque

- How accurate is MRA in predicting neurovascular compression in patients with TGN?
  - 92 patients with TGN – MRI/MRA
  - Radiologist blinded to side of pain
  - Imaging results compared to operative findings

| | Definite vascular compression | No compression |
|---|---|---|
| MRA : Positive | 76 | 0 |
| MRA : Negative | 8 | 8 |

Specificity of MR :100%

No false positives

Detects *only* cases

Sensitivity of MR : 90.5%

8 false negatives

Does not detect *all* cases of compression

Patek NK, Aguilina K, Clarke Y, Renowden SA, Coakham H. Br J Neurosurg. 2003;17:60-64

# AAN-EFNS Guidelines*

- Question: For patients with TGN without non-TGN neurological S/S, how often does imaging identify a cause (excluding vascular contact)?
- Answer: up to 15%

Cruccu G et al. AAN-EFNS guidelines on TGN management. European Journal of Neuro

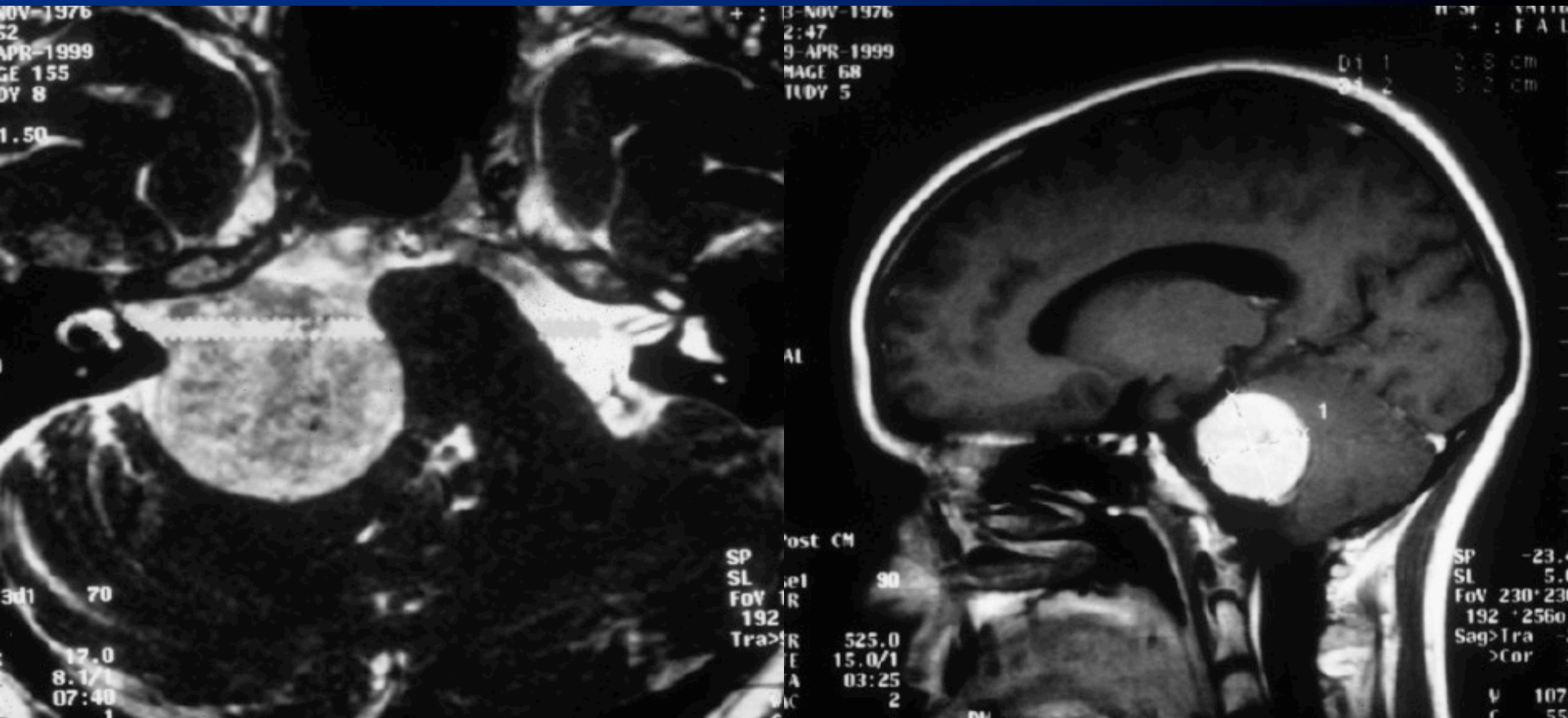
# AAN-EFNS Guidelines*

- Question: For patients with Classical TGN, does high resolution MRI accurately identify patients with neurovascular compression?
- Answer: Insufficient evidence to support or refute usefulness of MRI to identify vascular contact; suggest patients suitable for MVD to undergo high-resolution MRI

Cruccu G et al. AAN-EFNS guidelines on TGN management. European Journal of Neuro

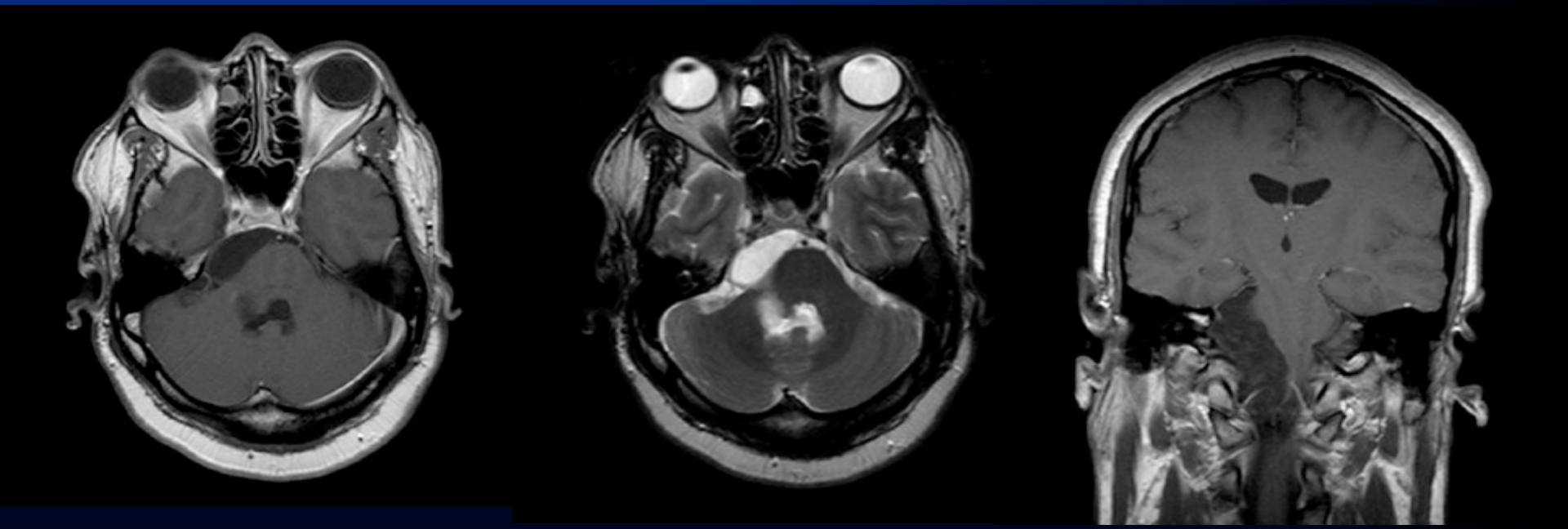
# TGN examples

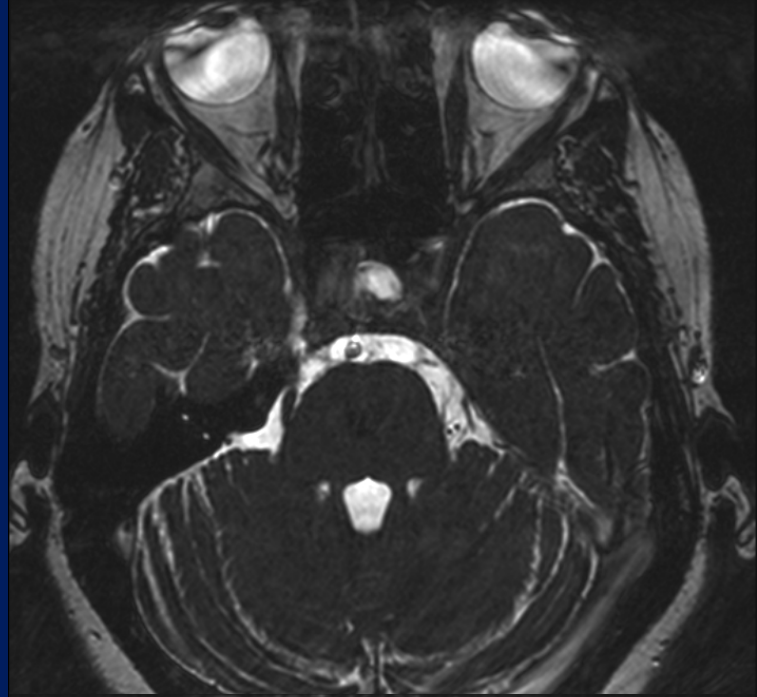
- 30 y.o. male with 10 year hx R-sided deafness and TGN



# TGN examples

- 45 y.o. male with 3 years of classic TGN



- 59F with classic TGN

# TGN

What happens if you have it?

# Natural History

- What we know
  - No prospective natural history studies in literature
  - Surgical series suffer from lack of class 1 evidence
- What we think we know
  - Progress is one of increasing severity with diminishing intervals of remission
  - Pain becomes more constant and atypical
  - Pain becomes resistant to medical tx

# TGN

What can be done about it?

# Quality of reporting in evaluations of surgical treatment of trigeminal neuralgia: Recommendations for future reports*

- Objectives
  - To set criteria and quality standards for reporting outcome of surgical tx
  - Identify published studies and evaluate against these criteria
  - Propose protocol for data collection and reporting outcome
- Results
  - 281 studies identified; 222 scored
  - 32% (71 studies) reached minimal set standards
  - 13% could be assessed for pain outcome
- Conclusion
  - Quality of current reporting is poor for all Tx modalities
  - Non-uniform outcome measures results in difficulty with comparability of studies

Zakrzewska J, Lopez B.
Neurosurgery. 2003;53:110-122.

# TGN – Medical Options

- Carbamazepine (first line)
- Baclofen
- Phenytoin
- Oxcarbazepine
- Gabapentin/Pregabelin/Lamotrigine
- Amitryptiline
- Capsaicin
- Clonazepam
- Botulinum toxin

# Carbamazepine (1st line)

- Periodic pain relief when treated with an adequate trial (pain relief in 69%)
- If no relief from CBZ, consider alternate dx
- 100mg po bid, increase by 200mg/day; max dose 1200mg/day
- S/E: drowisness, rash 5-10% with possible SJS, leukopenia

# Baclofen

- Fewer side effects
- Teratogenic (in rats)
- Don’t stop abruptly (hallucinations, sz)
- Start 5mg po tid, increase 3d by 5mg/dose, max 20mg qid

# Oxcarbazepine (Trileptal)

- Metabolized into carbamazepine
- Useful because patients can tolerate higher doses
- Start 300mg po bid, increase by 600mg/day q1wk; max dose 2400mg/day
- S/E: hyponatremia

# AAN-EFNS Guidelines*

- Question: Which drugs have shown efficacy in treatment of classical TGN?
- Answer:
  - CBZ effective for controlling pain in patients with TGN (Multiple Class I/II evidence)
  - Oxcarbazepine is probably effective (Meta-analysis, one Class II study)
  - Baclofen, lamotrigine, pimozide possibly effective (single Class II study)

Cruccu G et al. AAN-EFNS guidelines on TGN management. European Journal of Neuro

# AAN-EFNS Guidelines*

- Question: Which drugs have shown efficacy in treatment of symptomatic TGN?
- Answer: Insufficient evidence to support one drug over another.

Cruccu G et al. AAN-EFNS guidelines on TGN management. European Journal of Neuro

# TGN – Indications for Surgical Tx

- Failure of medical management
  - No or poor response
  - Unacceptable side effects
    - Ataxia
    - Memory disturbance
    - Skin rash
    - Neutropenia
- Patient choice

# Surgical Options

- Percutaneous Rhizotomy
  - Meckel's cave Glycerol injection
  - Radiofrequency thermocoagulation
  - Balloon microcompression
- Microvascular decompression
- Stereotactic radiosurgery
- Peripheral neurectomy
  - Nerve avulsion
  - Cryotherapy
- Partial sensory nerve section
  - Infratemproal approach
  - Posterior fossa
- Medullary tractotomy

# Microvascular Decompression (MVD)

# MVD – Historical Perspective

- Dandy: 1934 suggested causal relationship between vascular cross compression and TGN
- Gardner & Miklos: 1959 positioned gelfoam between the artery and nerve
- Jannetta: “popularized” the concepts of vascular compression and surgical treatment

# Surgical Treatment

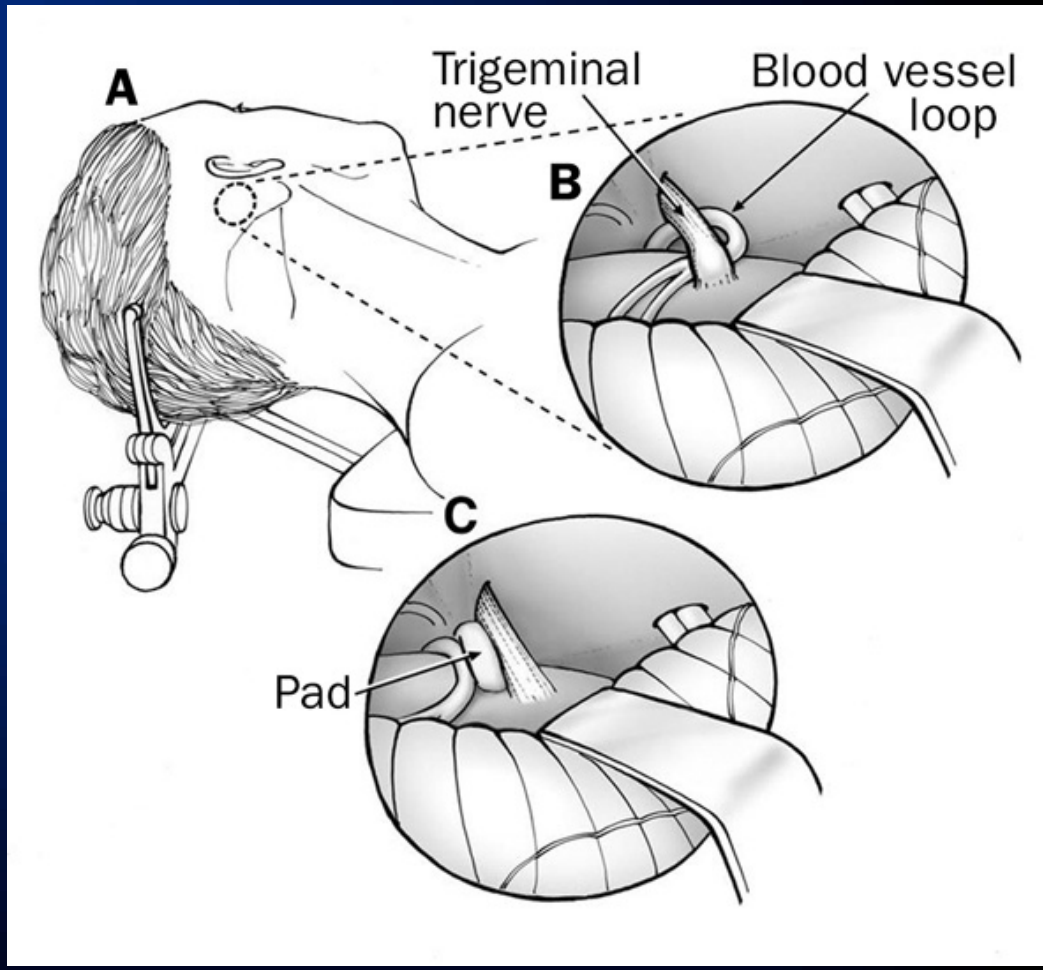
- What we know
  - No prospective RCT
  - Retrospective data
  - Difficult to compare results between centres
- What we think we know
  - MVD
    - Lowest rates of recurrence and longest lasting relief
    - Lowest rates of sensory loss
    - Addresses apparent major cause
    - Low mortality (0.3%)
    - Neurologic morbidity 1-10%

# MVD

- Age <= 65
- Does not produce anaesthesia dolorosa
- Mortality rate 0.3%
- 1-10% of neurologic morbidity
- Aseptic meningitis 20%
- Failure of 20-25%
- Not for MS patients

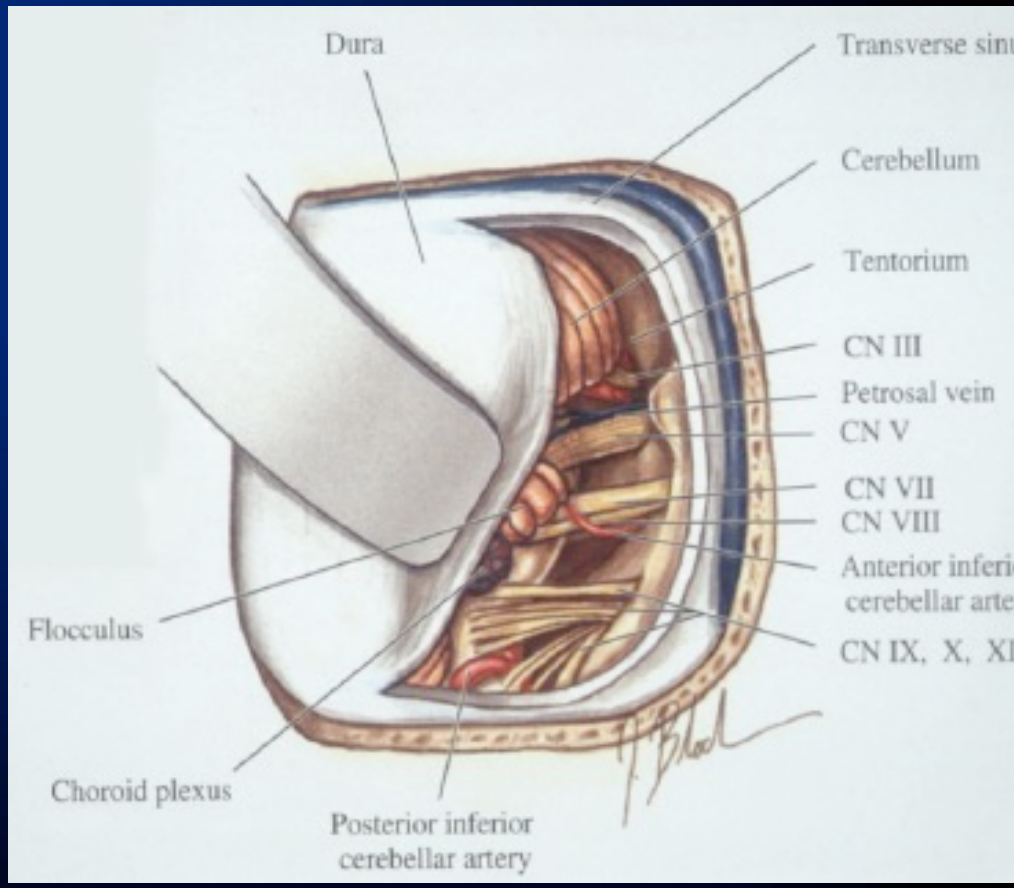
# MVD: Operative approach

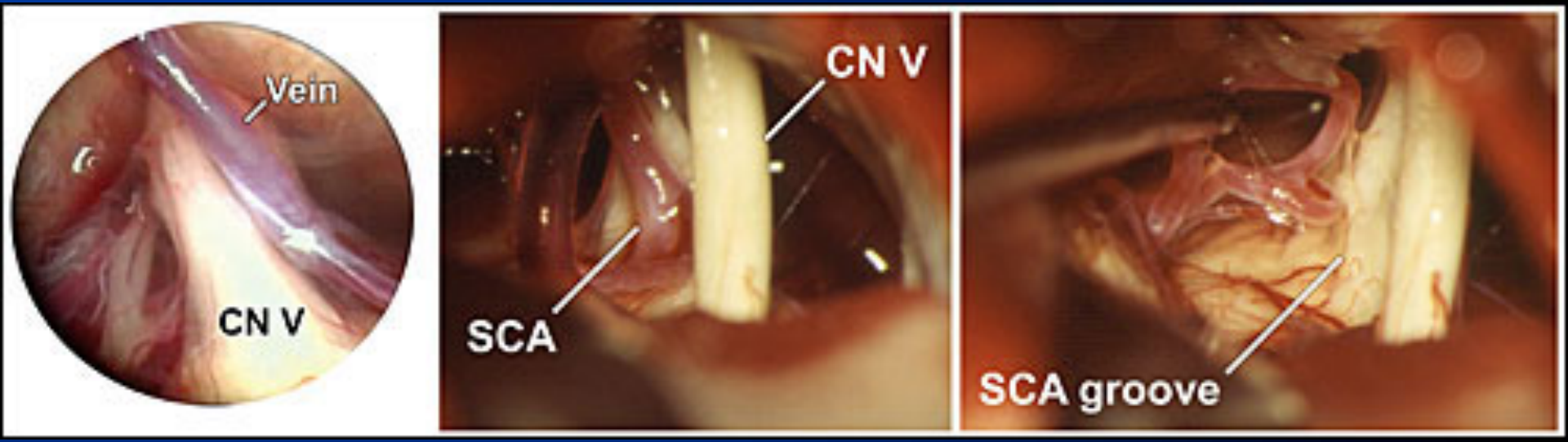
- GA
- Park bench or lateral oblique position
  - Symptomatic side up, axillary roll
  - Thorax elevated 10-15 deg
  - Head rotated slightly away from affected side with lateral head tilt (parallel to floor) and neck flexion (2 fingerbreadths between china and sternum)
  - Upper shoulder retracted caudally with tape
- +/- lumbar drainage
- Retromastoid craniectomy with exposure of sigmoid/transverse sinus



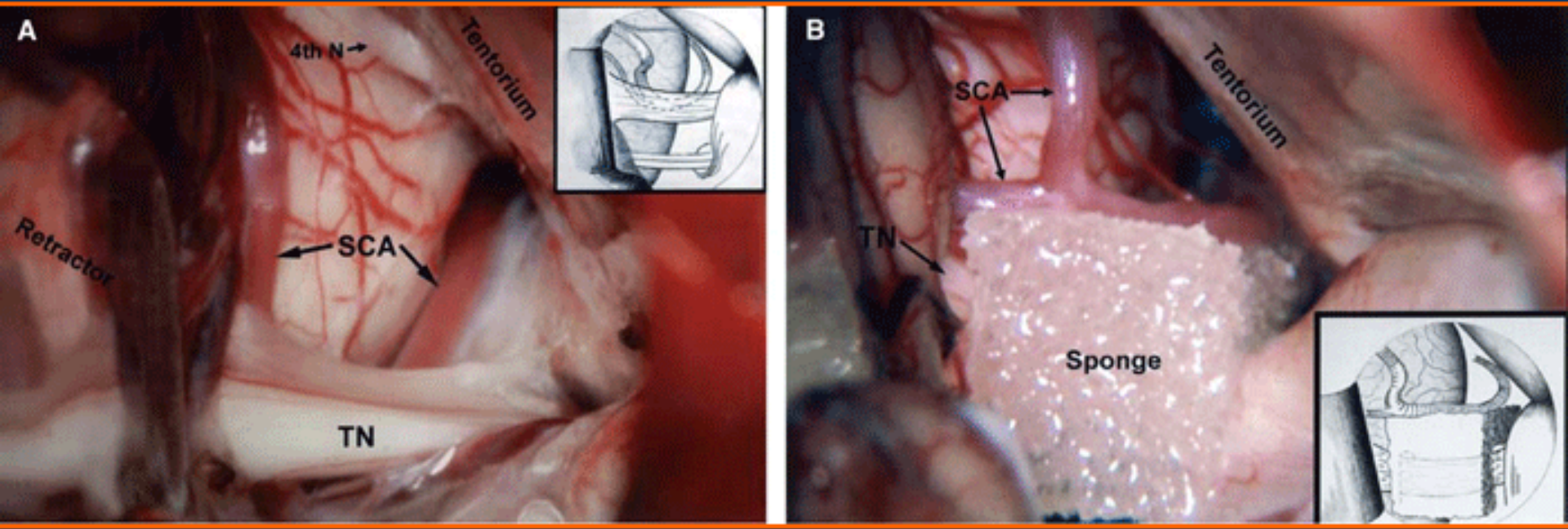
# MVD: Operative approach

- Dural opening
  - Janetta (curvilinear each end at a sinus, convexity away from junction)
  - Inverted T (one incision toward each sinus, third toward junction)
- Retraction of cerebellar hemisphere
  - Opening of arachnoid of superior cerebellar pontine cistern
  - Division of Vein of Dandy from superior petrosal sinus
- MVD/Nerve section
  - Non-absorbable insulator (Ivalon sponge or shredded Teflon felt)
- Standard closure
- Home 5-7 days postop







The New England
Journal of Medicine



Volume 334 APRIL 25, 1996 Number 17

**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.

- N=1185, MVD over 20yrs
  - 1155 clinical f/u after 1yr
  - Median f/u 6.2yrs
- 70% pain free at 10yrs
  - 11% of recurrence underwent 2nd procedure

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- Factors associated with pain recurrence
  - Female
  - Sx duration > 8yrs
  - Venous compression
  - Persistent pain following MVD
- Complications
  - Death 0.2%
  - Cerebellar hematoma 0.2%
  - CSF leak 1%
  - Hearing loss 1%

# Evaluation of MVD and partial sensory rhizotomy in 252 cases of TGN*

- Retrospective study, mean f/u 5.1yrs
  - Pt with extrinsic compression: MVD
  - Pt with no compression: partial sensory rhizotomy
  - Pt with vascular contact but no distortion: MVD & partial sensory rhizotomy

Bederson JB, Wilson CB. J Neurosurg. 1989;71:359-367.

# Evaluation of MVD and partial sensory rhizotomy in 252 cases of TGN*

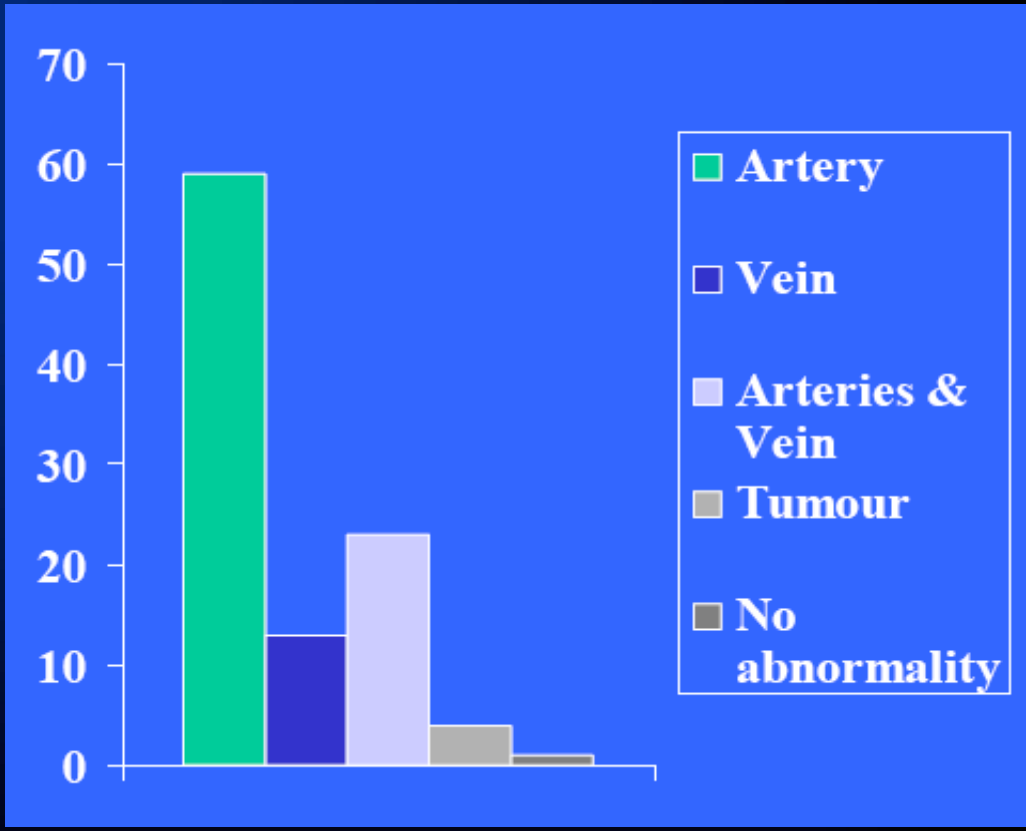
- Results
  - Excellent: 75%, Good: 8% [208]
  - Persistent pain: 5% [13]
  - Recurrent pain: 12% [31] at mean of 1.9yrs postop
    - Recurrence rate 2%/yr thereafter
  - Re-operation (rhizotomy): 85% successful

# Evaluation of MVD and partial sensory rhizotomy in 252 cases of TGN*

- Adverse prognostic factors
  - Percutaneous radiofrequency rhizolysis
  - Absence of vascular compression
  - Long duration of symptoms in pts with vascular compression

# TGN treated by MVD: Long-term follow-up study*

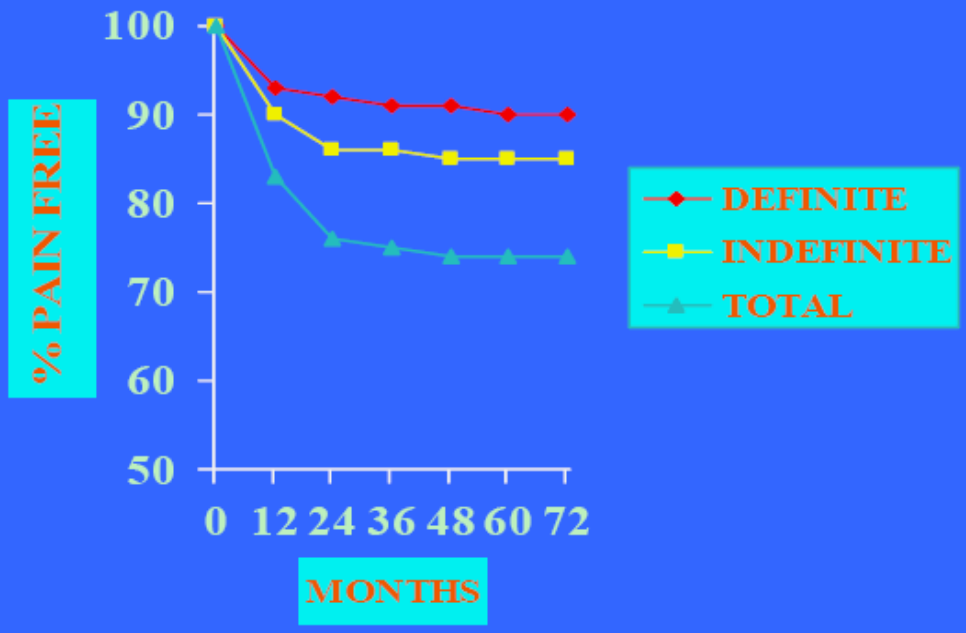
- Vascular compression
  - SCA (61%)
  - AICA
  - Vein
- Tumour
  - Meningioma
  - AN
  - Dermoid




Mendoza N, Illingworth R. British J of Neurosurg. 1995;9:13-19.

# TGN treated by MVD: Long-term follow-up study*

- Recurrence of pain in relation to definite or indefinite operative findings




Recurrence of pain in relation to definite or indefinite operative findings

| Operative findings | Definite | Indefinite |
|---|---|---|
| Pain free | 79 | 16 |
| Pain recurrence | | |
| Major | 7 | 11 |
| Minor | 13 | 8 |

$P < 0.001$

# Serious complications of MVD operations for TGN and HFS*

- N=278, MVD
- 9 serious complications
  - 1 intracerebellar ICH, acute HCP
  - 1 supratentorial ASDH
  - 2 status epilepticus
  - 1 BS infarct
  - 1 SAT due to traumatic aneurysm
  - 1 PCA infarct
- 2/9 → death
- Operative mortality 0.7%

Hanakita J, Kondo A.
Neurosurgery 1988;22:348-352.

# Microvascular decompression in the US, 1996-2000: mortality rates, morbidity rates, and the effects of hospital and surgeon volumes*

- Retrospective study
- N=1326 MVD for TGN
- Mortality 0.3%
- Neurological complications 1.7% (ICH, CNVII palsy, EVD)
- Median annual case load: 5 cases/hospital (1-195), 3 cases per surgeon (1-107)

Kalkanis S, Eskander E, Carter B, Barker F. Neurosurgery. 2003;6:1251-1261

# Microvascular decompression in the US, 1996-2000: mortality rates, morbidity rates, and the effects of hospital and surgeon volumes*

- After adjustment for age, gender, race, diagnosis; outcomes at d/c were superior and complications were less at higher volume hospitals and higher volume surgeons
- Volume and mortality rate not significantly related

# Percutaneous Procedures

# Principle of Percutaneous Procedures

- Radiofrequency Lesioning
  - Destroy nociceptive fibers (A delta and C)
  - Preserve touch fibers (A alpha and beta)
- Balloon Microcompression (opposite of above)
- More effective in MS patients (especially balloon percutaneous microcompression)
  - 50% recurrence in 3 years for MS patients
- Recurrences easily treated by rpt procedures
- Advantages
  - Avoids major surgery
  - No reports of intracranial hemorrhage

# Meckel's Cave Glycerol Injection – Historical Perspective

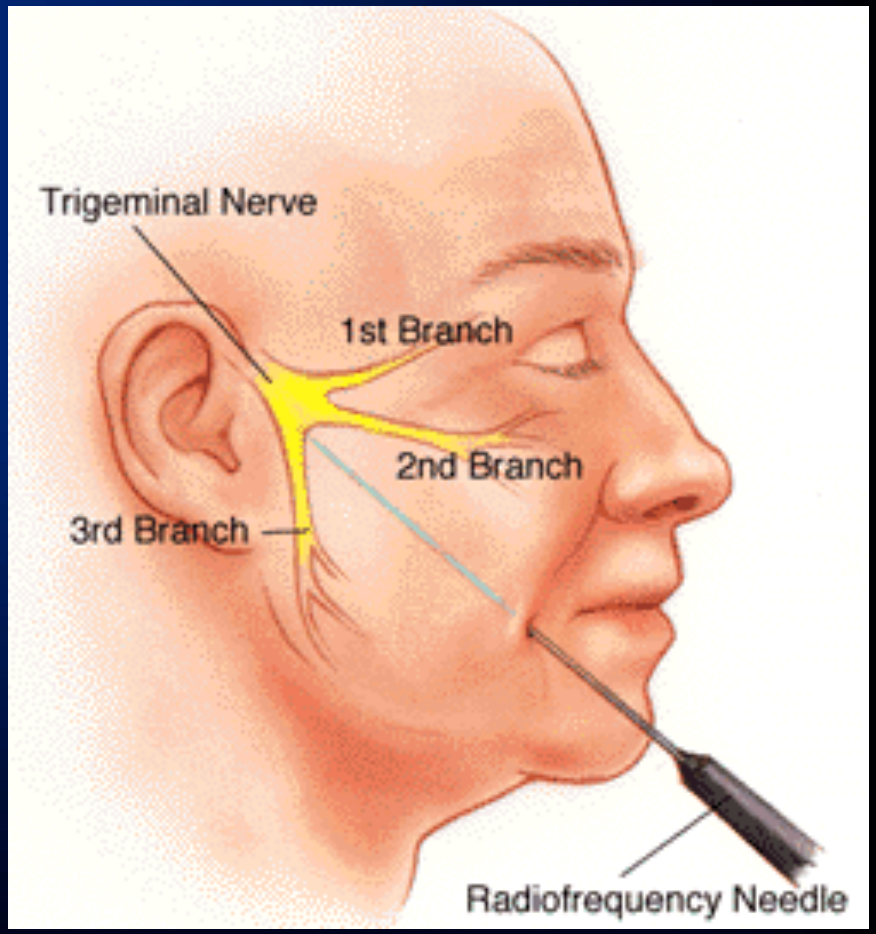
- Hartel: 1912 pioneered the percutaneous transovale approach to the Gasserian Ganglion using absolute alcohol
- Sweet: 1974 described glycerol injection (and radiofrequency) lesioning to the ganglion
- Hakanson: 1981 discovered glycerol relieved tic pain when injecting the retrogasserian space

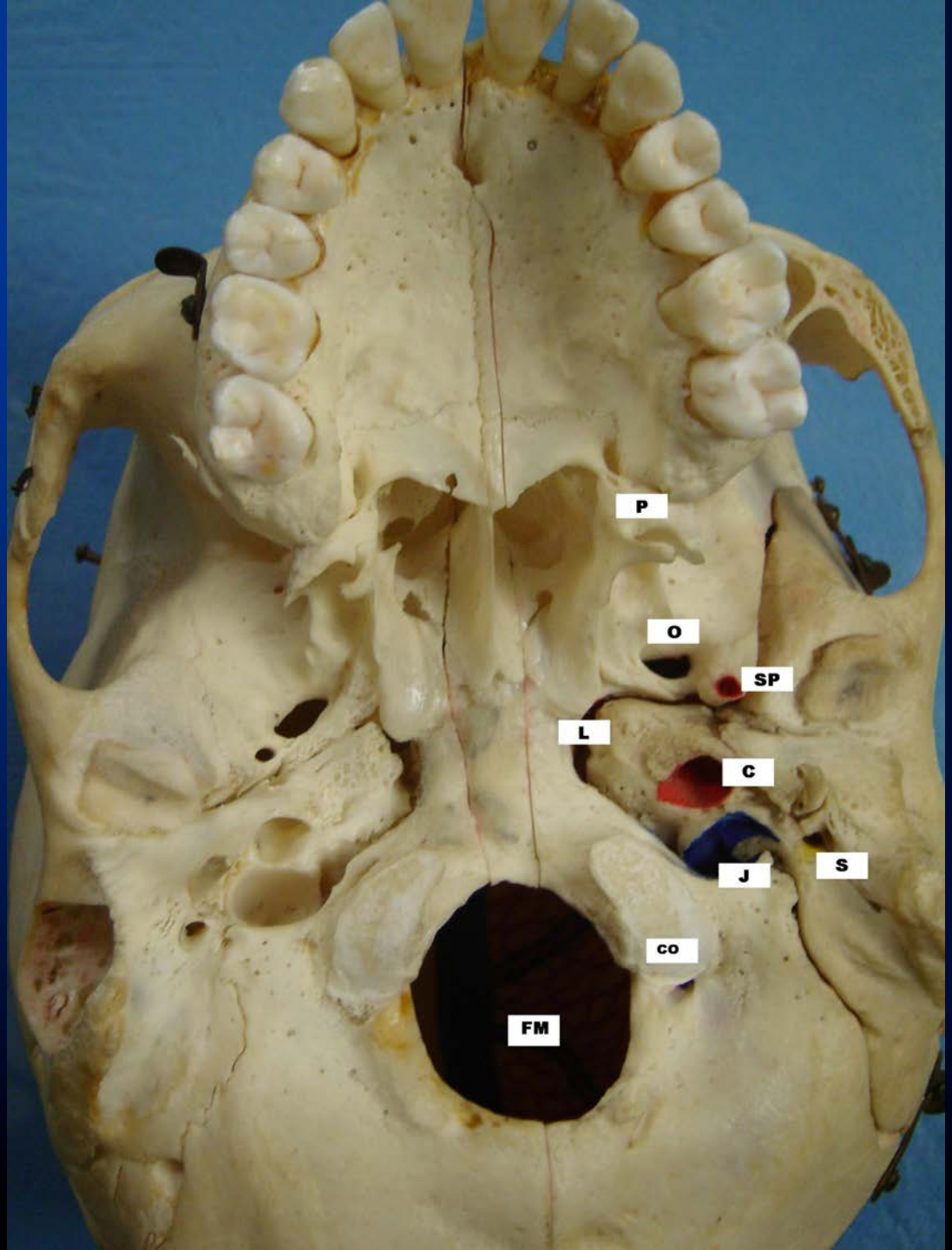
# MCGI

- Possibly lower incidence of sensory loss and anaesthesia dolorosa than with RF lesion

# MCGI – Operative approach

- Supine
- 22G spinal needle
- Positioning
  - 2-3cm lateral to lip insertion site, one hand palpate buccal mucosa, keep needle deep to oral mucosa and medial to coronoid process of mandible
  - Initial trajectory toward intersecting planes 3cm anterior to EAM and medial aspect of pupil
- Fluoro Landmarks
  - AP-submental vertex, aim for foramen ovale
  - Lateral, aim for 10mm below floor of sella along clivus

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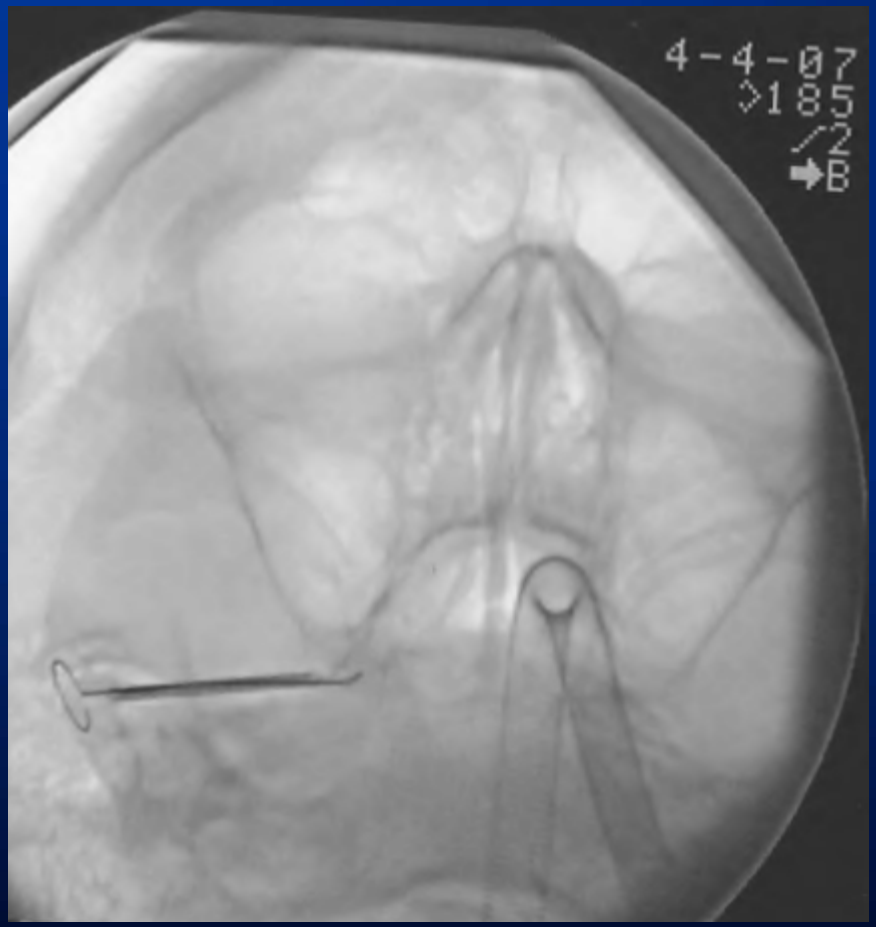
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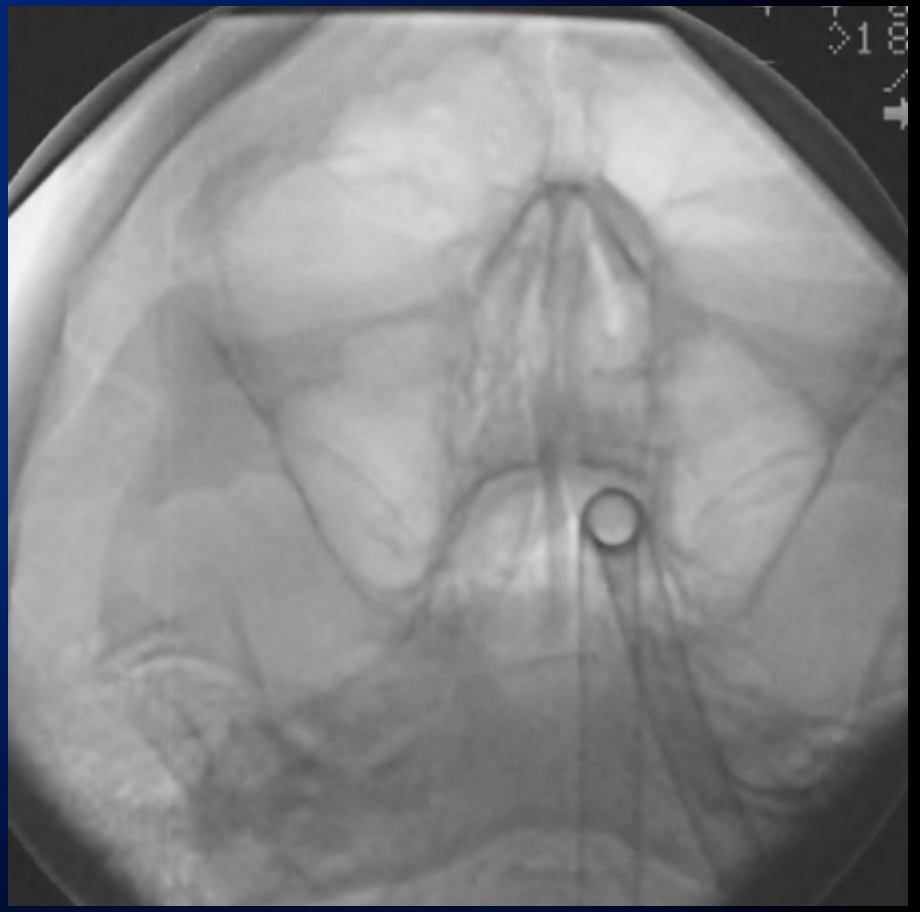
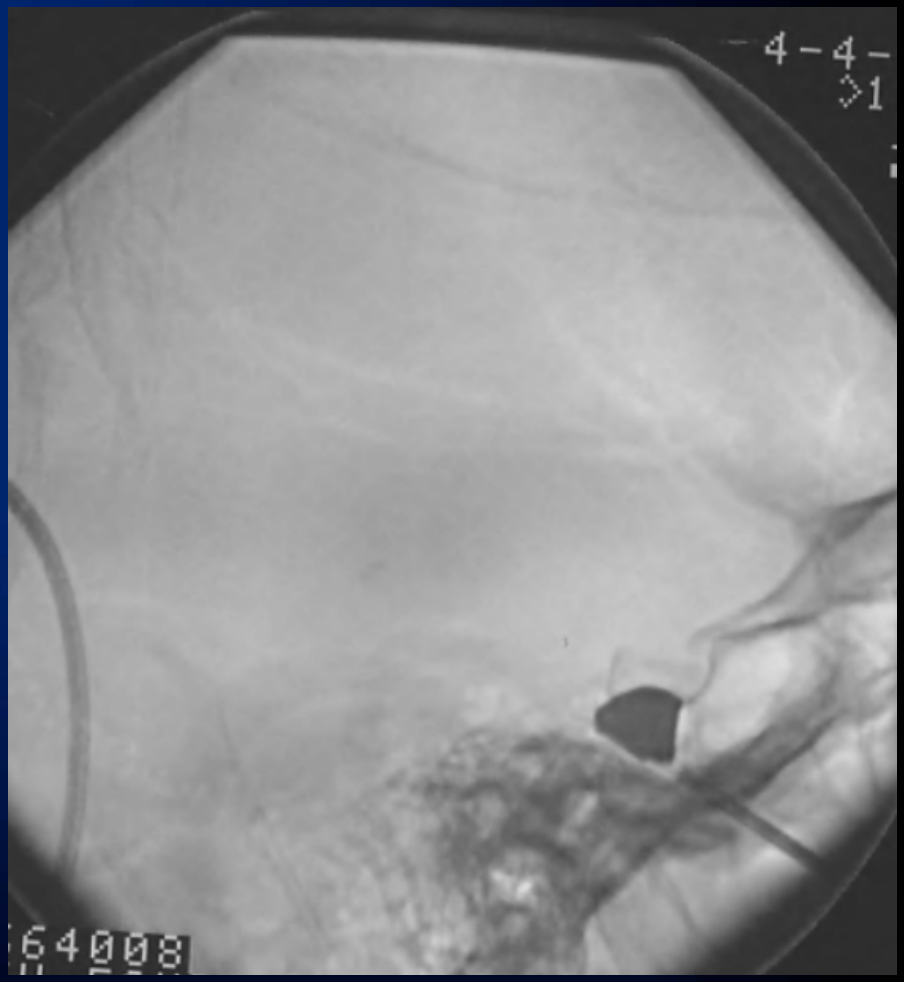




# MCGI – Operative approach

- Clenching of jaw due to motor part of V upon entering FO, remove stylet and see CSF
- Image intensifier is used to identify the needle tip in the Meckel's cave
- Cisternography by injecting dye to delineate Meckel's cave
- Patient made to sit up and inject 1cc of anhydrous glycerol (10%)


# Outcomes in MCGI

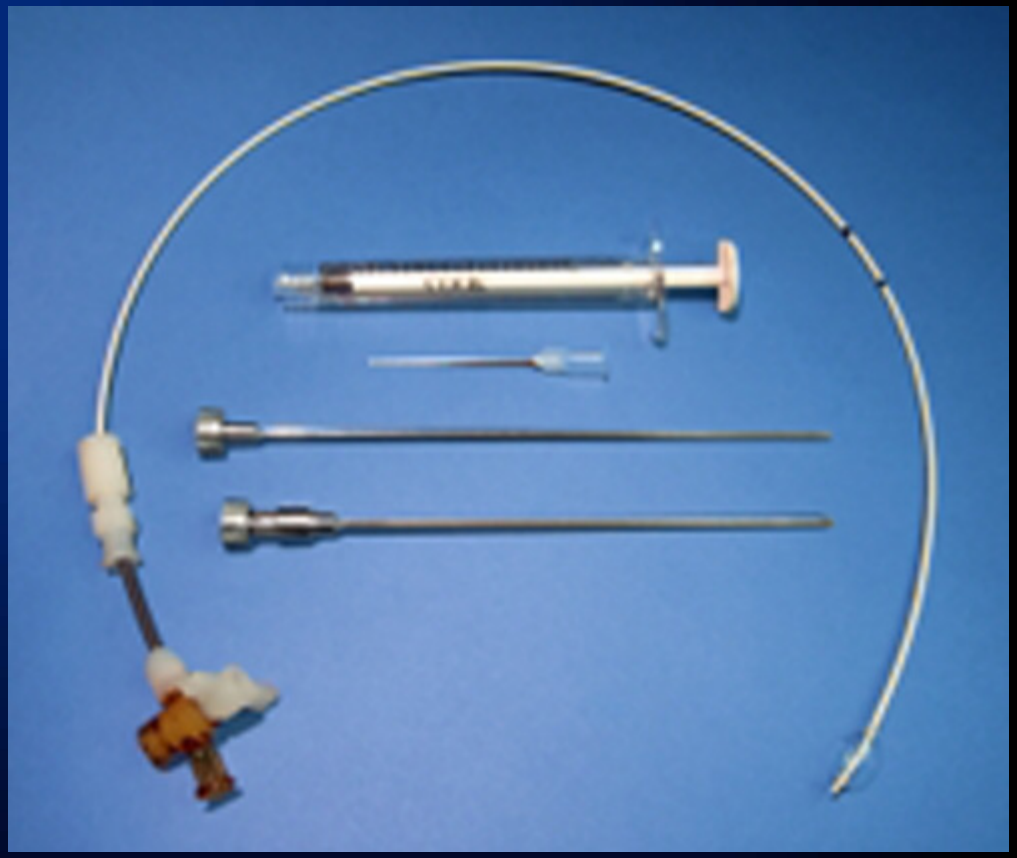
- N=201
  - 176 immediate effect (139 pain free, 37 pain reduced)
  - 24 gradual change in pain (11 resolved completely, 13 partial)
  - 25 persistent pain (1 worse)
- N=141 pts at 3yrs
  - 71 pain free
  - 42 pain controlled
  - 28 uncontrolled pain

# Radiofrequency Trigeminal Rhizotomy

- Uses RF energy to thermocoagulate pain fibers; requires patient to be awake at intervals during procedure
- Ideal for V3 neuralgia, can select out without affecting other divisions
- Patient needs to be cooperative
- Complications
  - Masseter, pterygoid weakness 24%
    - Open jaw, chin deviates to side of weak pterygoid
  - Anaesthesia dolorosa 4%
  - Neuroparalytic keratitis 4%
  - Oculomotor paresis 2%

# Percutaneous Microcompression Rhizolysis

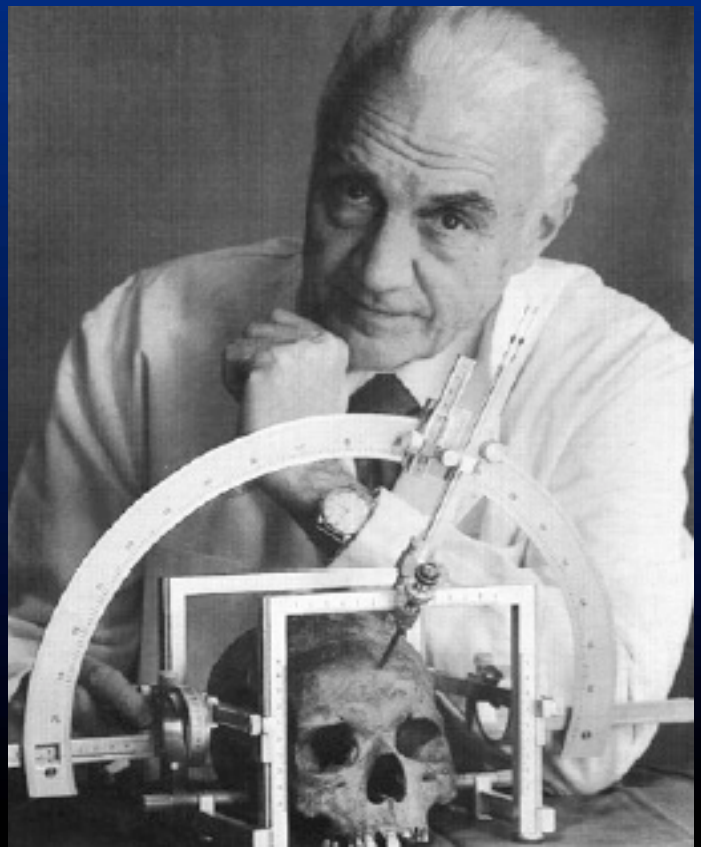
- Inflation of No.4 Fogarty catheter balloon
- Patient not awake



# Stereotactic Radiosurgery (SRS)

# Stereotactic Radiosurgery

- Lars Leksell: 1953 irradiated two patients with TGN



# SRS

- Least invasive
- Traditionally for refractory cases following multiple procedures
- Complete pain relief 65%
- Significant pain reduction in 15-30%
- Ideal for high-risk medical patients, older age, symptoms not too severe, V1 pain
- Not ideal for patients requiring immediate pain relief due to latency of full effect (mean latency 3-4 weeks, can be months)

# Stereotactic radiosurgery for the treatment of trigeminal neuralgia*

- Initiated in 1992
- N=220
- Median SRS dose was 80 Gy (60-90)
- 61.4% had prior procedures
- Median f/u 2yrs

Kondziolka D, Lunsford L, Flickinger JC. Clin J Pain. 2002;18:42-47

# Stereotactic radiosurgery for the treatment of trigeminal neuralgia*

- Complete/partial pain relief in 85.6% at 1yr, 55.8% at 5yrs
- Complete pain relief in 70% at 1yr
- Patients with atypical pain had lower rate of pain relief
- Patients without pre-tx sensory loss and those with no previous surgical tx had better outcomes
- 10% developed facial paresthesias/numbness

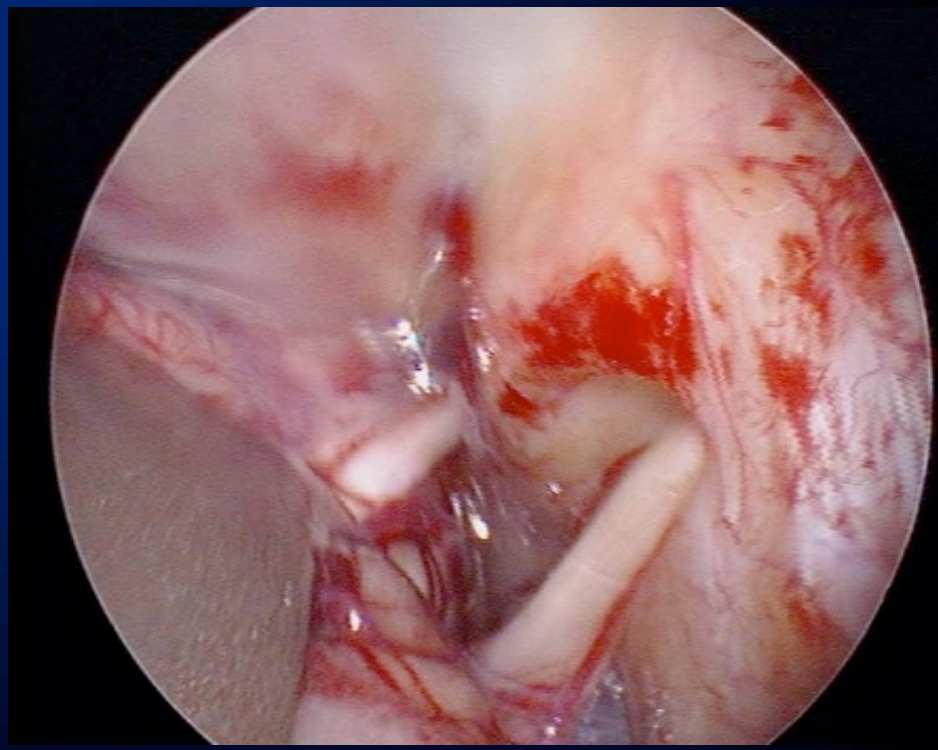
# Stereotactic radiosurgery for trigeminal neuralgia: a multi-institutional study using the gamma unit*

- 4mm single isocenter targeted 2-4mm anterior to junction of pons and CN V
  - 30% isodose deliver to BS
- Dosage 70-80 Gy
- Median time to response: 4 wks
- Median f/u: 18 months
- Facial numbness in 10%


Kondziolka et al. J Neurosurg. 1996;84:940-945

# Endoscopic-assisted MVD

- Utilize rigid endoscope
- Advantage – smaller exposure and less retraction minimizing neurovascular injury
- Disadvantage – learning curve
- No current evidence of improved results



# Simplified Management Algorithm

- In general, if age > 65 or unfit, or afraid of operation, or not worried about possibility of numbness and its effects, have MS, <5yr life expectancy:
  - Percutaneous procedure
- If age <= 65 and fit, worried about possibility of numbness, desire for long-lasting relief despite small potential for life-threatening complications,>5yr life expectancy:
  - MVD
- High medical risk
  - SRS

# Hemifacial Spasm

- Intermittent unilateral painless contractions of facial muscles
- Starts with orbicularis oculi and spreads to lower half of face
- Along with palatal myoclonus – only movement disorder that persists in sleep
- Typically caused by compression of CN VII by AICA
- Risk of hearing loss ~ 20%
- More frequent in female, young adult
- Responds well to MVD
- Unlike TGN, CBZ/dilantin not generally effective

# Glossopharyngeal Neuralgia

- Pain is located in base of tongue ("glosso"), throat ("pharyngeal"), ear, neck
- Other symptoms
  - Hypotension (vagus)
  - Syncope
  - Cardiac arrest
- Incidence 1 per 1,775,000 (1/70 of TGN)

# Glossopharyngeal Neuralgia

- Treatment
  - Medicine: Cocainization of tonsillar pillars and fossa
  - Surgery:
    - MVD
    - Sectioning of preganglionic CN IX and upper 1/3 or 2 fibers (whichever is larger) of CN X

END