

# Trigeminal Neuralgia and Endoscopic Microvascular Decompression Surgery

September 23, 2017

Garni Barkhoudarian, MD

Assistant Professor of Neuroscience and Neurosurgery

Pacific Neuroscience Institute

John Wayne Cancer Institute at Saint John's Health Center



# Disclosures

- VTI - Consultant
- Anatomy lab receives financial or material support from:
  - Karl-Storz Endoskope America, Inc
  - Stryker Corporation
  - Mizuho America, Inc
  - Surgical West, Inc

# Trigeminal Neuralgia



# Trigeminal Neuralgia

- A rare clinical entity characterized by severe, sudden, brief, mostly unilateral, stabbing pain in the distribution of the 5<sup>th</sup> cranial nerve



# Trigeminal Neuralgia

- Condition of hyperexcitation of the trigeminal nerve causing:
  - Severe stabbing or lancinating pain
  - Triggered by light touch, chewing, brushing teeth, wind
  - Typically responsive to carbamazepine or other anti-epileptic agents
- Differential diagnosis includes:
  - Multiple Sclerosis
  - Cerebello-pontine angle tumor
  - Herpes Zoster
  - Dental caries

# History

- Aretaeus of Cappadocia
  - Rome and Alexandria
  - 2<sup>nd</sup> Century AD
  - Headache with “spasm and distortion of the countenance”



Aretaeus, the Cappadocian.



# History

- Jurjani (Zayn al-Din Isma'il ibn al-Husayn)
- 11<sup>th</sup> Century physician
- Persian province of Khvarazm
- “type of pain which affects the teeth on one side and the whole of the jaw on the side which is painful”



# History

- Wells Cathedral
  - Somerset, England
  - The Tomb of Bishop Button, 1274
  - The toothache figure

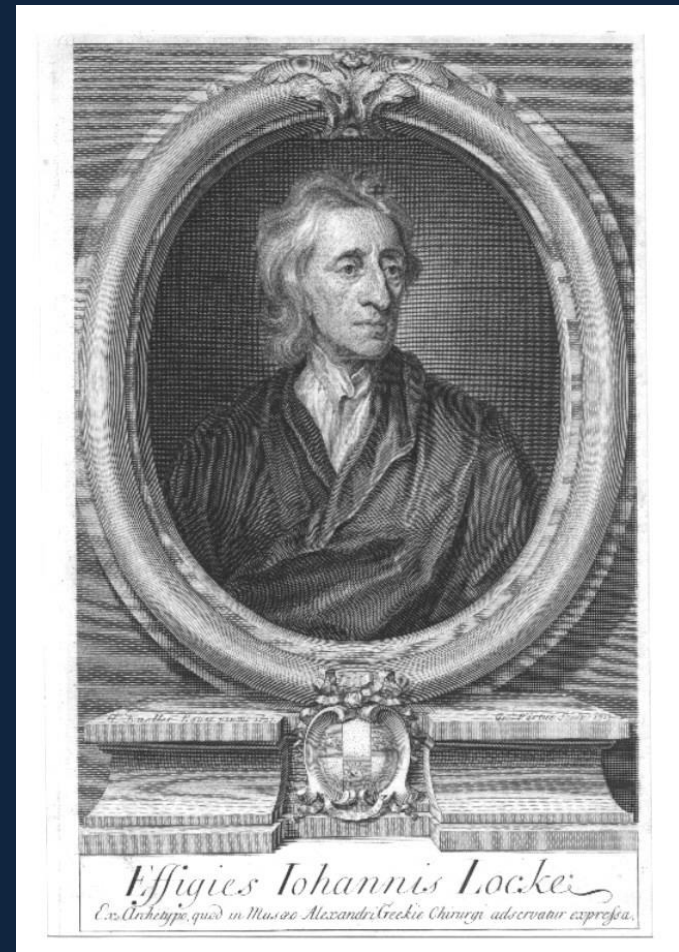


(C) 1997 Pete Harlow



# History

- John Locke
- 17<sup>th</sup> Century Philosopher and Physician
- Described the facial pain of the Countess of Northumberland, wife of the English Ambassador to France in 1677



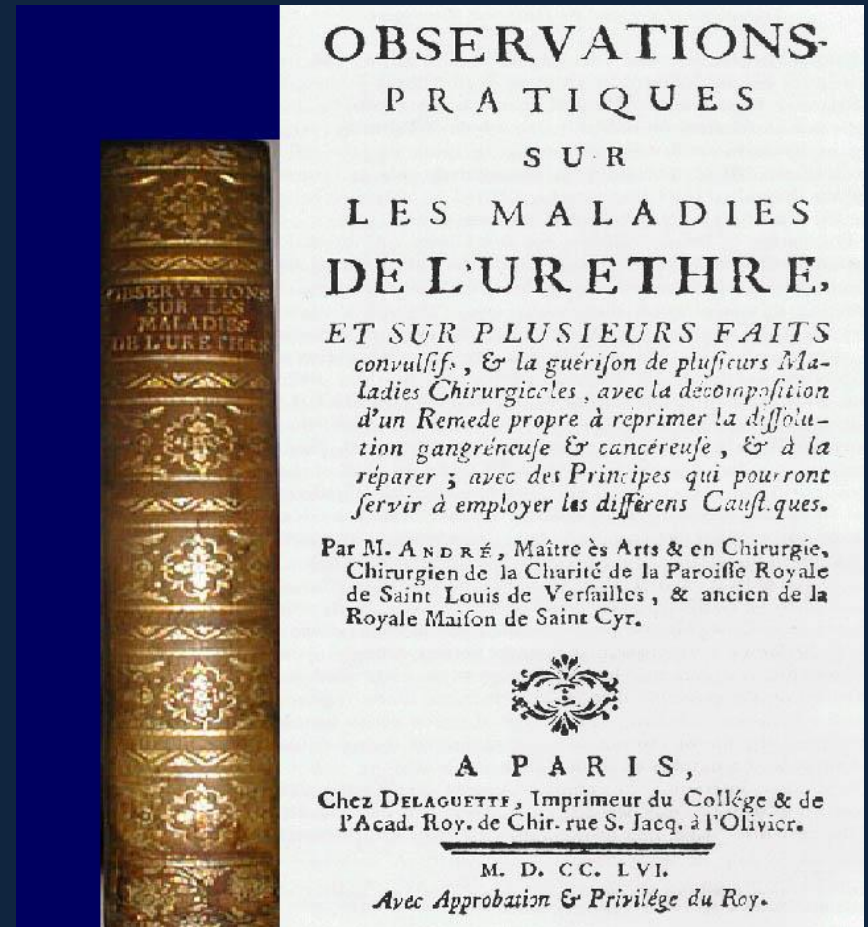
# History

- “...I found in a fit of such violent and exquisite torment... it forced her to such cries and shrieks as you would expect from one upon the rack... her mouth was constantly drawn on the right side towards the right eare... These violent fits terminated on a suddaine, and then my Lady seemed to be perfectly well... there was not the least appearance of any alteration in her face... speaking was apt to put her into these fits... or touching her gums”



# History

- Nicholas Andre
- *Tic douloureux*
- 1756
- *Observations pratiques sur les maladies de l'urethre et sur faits convulsifs*



# History

- James Ewing Mears
- Study of the pathological changes occurring in Trifacial Neuralgia.  
*Medical News of Philadelphia* 1884
- First suggested Gasserian ganglionectomy as treatment

# History

- Hartel
- 1914
- First detailed account of injecting (procaine) into Meckel's cave percutaneously

# History

- Walter Dandy
- 1934
- While performing partial sectioning of the trigeminal sensory root first described the presence of vascular compression of nerve root
- 50% of his cohort





# History

- Gardner and Miklos
- First described decompressing vascular contact with sponge pledget
- 1959

# History

- Peter Jannetta
- University of Pittsburgh
- Defined modern technique of MVD
- 1967



# History

- Lars Leksell
- Stereotactic Radiosurgery
- 1971



**"The tools used by the surgeons must be adapted to the tasks, and where the human brain is concerned they cannot be too refined."**

*-Lars Leksell, M.D., Ph.D.  
Gamma Knife Developer, 1971*

# Epidemiology

- Annual Incidence 4-5 in 100,000
- Age: > 50 (avg 63)
- Female to male 1.8-1
- Laterality:
  - Right: 60%
  - Left: 39%
  - Both: 1%

# Epidemiology

- Division:
  - V1 – 2%
  - V2 – 20%
  - V3 – 17%
  - V1 & V2 – 14%
  - **V2 & V3 – 42%**
  - all 3 – 5%



# Pathophysiology

- Focal demyelination of root of trigeminal nerve
- At the level of the junction between central myelin (from oligodendroglial cells) and peripheral myelin (from Schwann cells)
- Called the Obersteiner-Redlich Zone (ORZ); or Root Entry Zone (REZ)



# Pathophysiology

- Ephaptic transmission in trigeminal nerve from large diameter myelinated A fibers to poorly myelinated A-delta and C (nociceptive) fibers
- May explain why innocuous stimulation of trigger zones within distribution of same trigeminal branch causes attack

# Pathogenesis

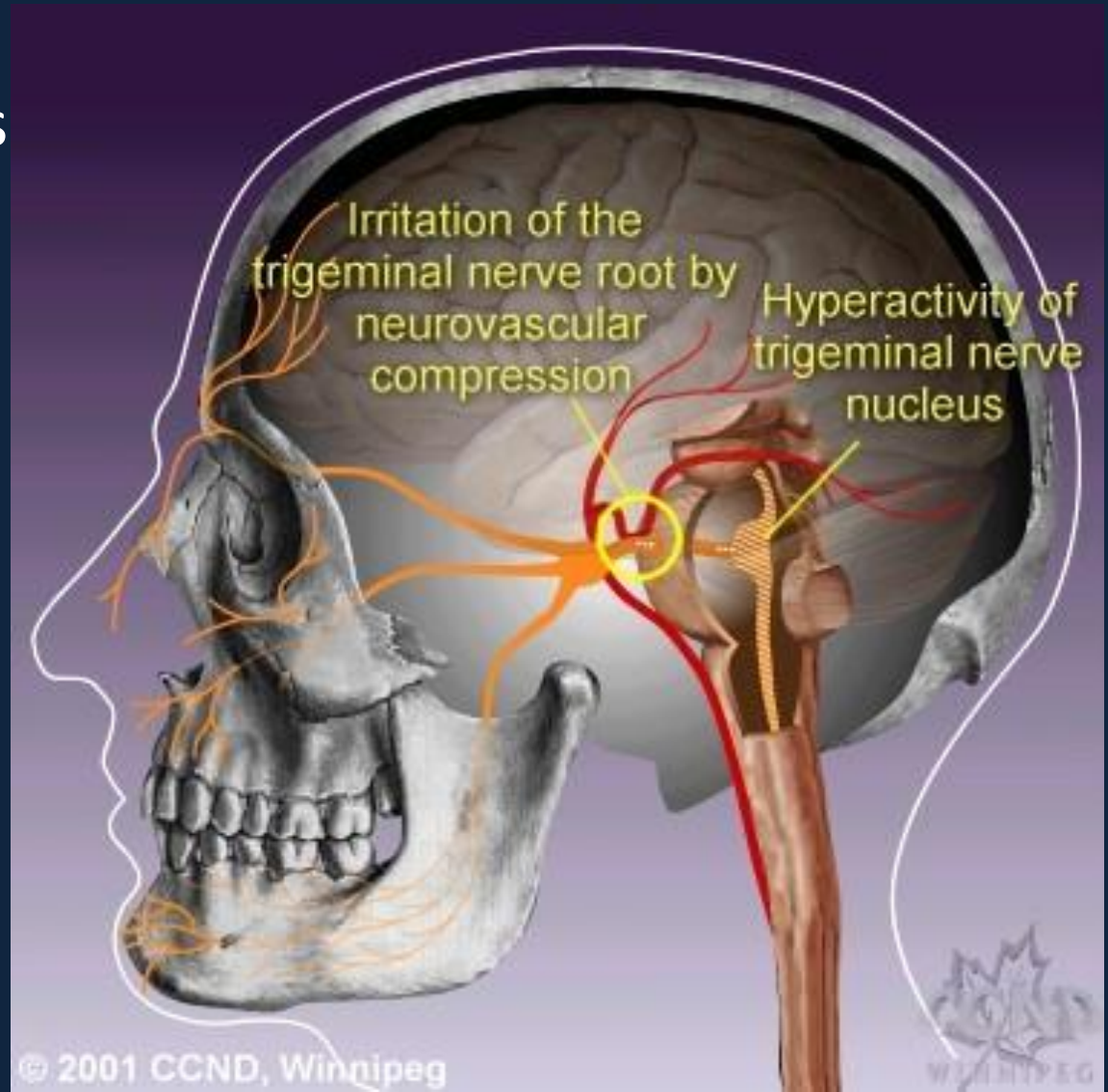
- Vascular Compression
- MS
- Posterior fossa tumor

# Pathophysiology

- Vascular compression of the trigeminal nerve at the REZ
  - Most commonly by SCA (80%)
  - Persistent primitive trigeminal artery
  - Note: vascular compression may be seen in up to 50% of autopsies in patients **WITHOUT** TGN

# Pathophysiology

- Vascular Compress

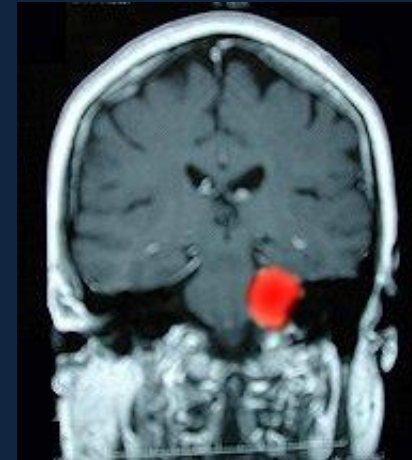
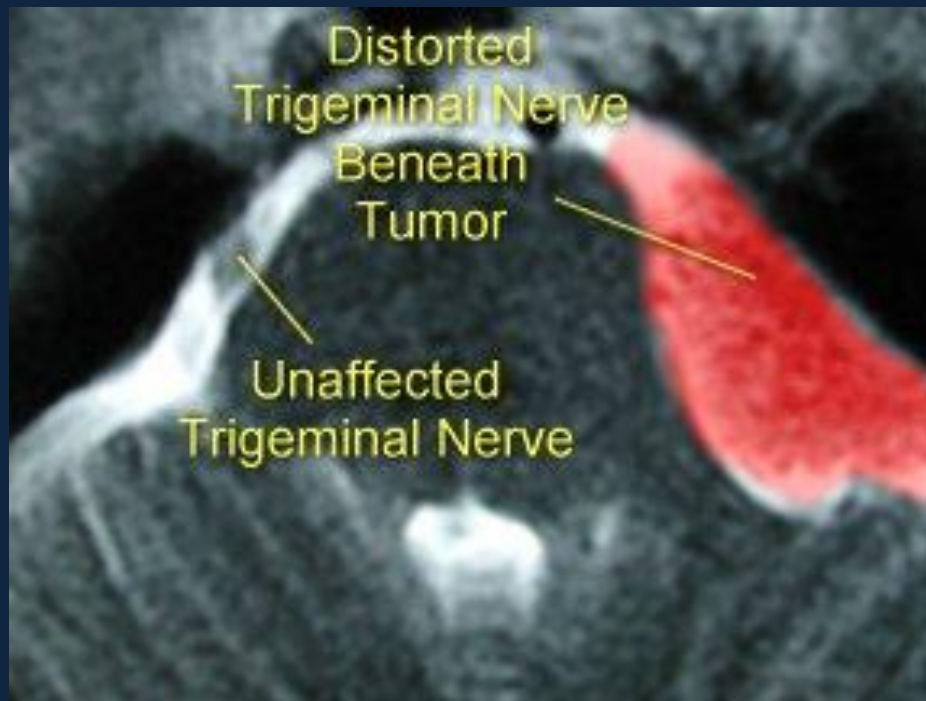


# Pathophysiology

- MS: plaque within brainstem
  - poorly responsive to MVD
  - 2% of patients with MS have TGN
  - 18% of patients with bilateral TGN have MS

# Pathophysiology

- Posterior fossa tumor





# Diagnostic Criteria

- 1) Paroxysmal attacks (< 2 min)
- 2) Pain characteristics (at least 4)
  - Distribution along 1 or more divisions of CN V
  - Sudden, intense, sharp, superficial, stabbing, burning
  - Severe intensity
  - Trigger areas or activities (eating, talking, etc)
  - Asymptomatic between attacks
- 3) No neurologic deficit
- 4) Stereotyped in individual patient
- 5) Exclusive of other causes by history, physical

(adapted from International Headache Society)

# Classification of Facial Pain

- The surgeon, however, is chiefly concerned with the question: “What cases of neuralgia are suited for operative treatment, and what are the best methods to employ?” The answer, obviously, should depend upon a scientific classification, based solely upon the causes of neuralgia; at present such a classification is impossible.
  - J. Hutchinson (1905)

# Classification

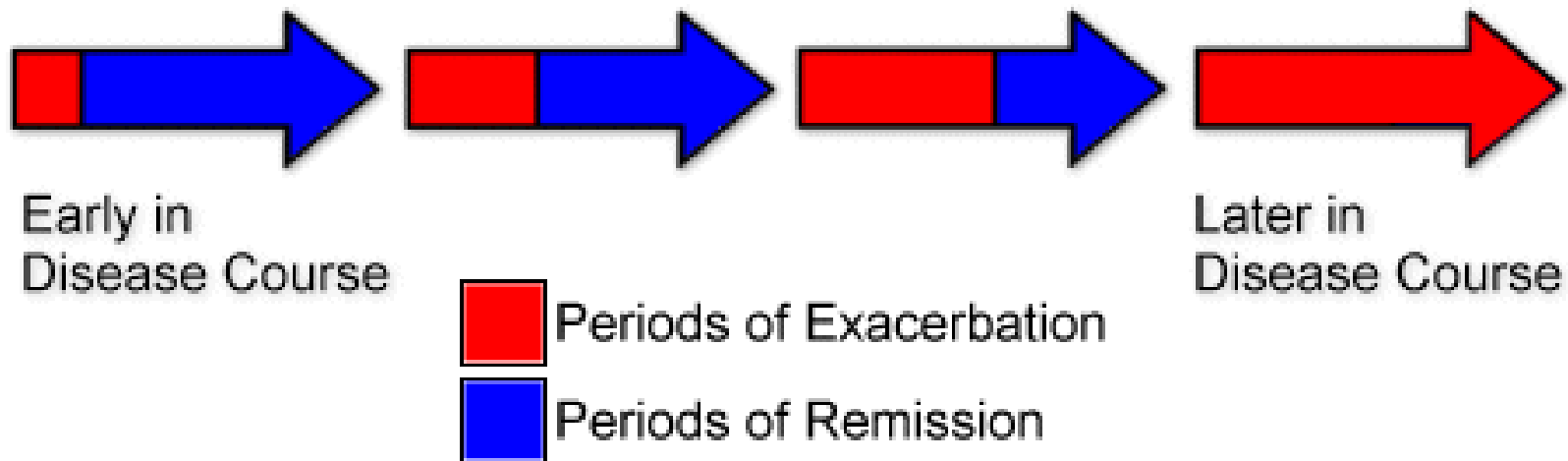
- Typical trigeminal neuralgia
- Atypical trigeminal neuralgia
- Atypical facial pain (AFP)

# Classification

- TGN type I: episodic
- TGN type II: constant
- Trigeminal neuropathic pain: unintentional injury
- Trigeminal deafferentation pain: intentional injury (gangliolysis, or rhizotomy)
- Postherpetic neuralgia
- Symptomatic trigeminal neuralgia: MS
- Atypical facial pain: 2<sup>nd</sup> to somatoform disorder
  - Kim Burchiel, 2003

# Natural History

## Progression of Trigeminal Neuralgia Over Time



# Differential Diagnosis

- Herpes Zoster – pain is not paroxysmal
- Dental disease
- Orbital disease
- Temporal arteritis – tenderness over STA
- Intracranial tumor – usually has sensory deficit

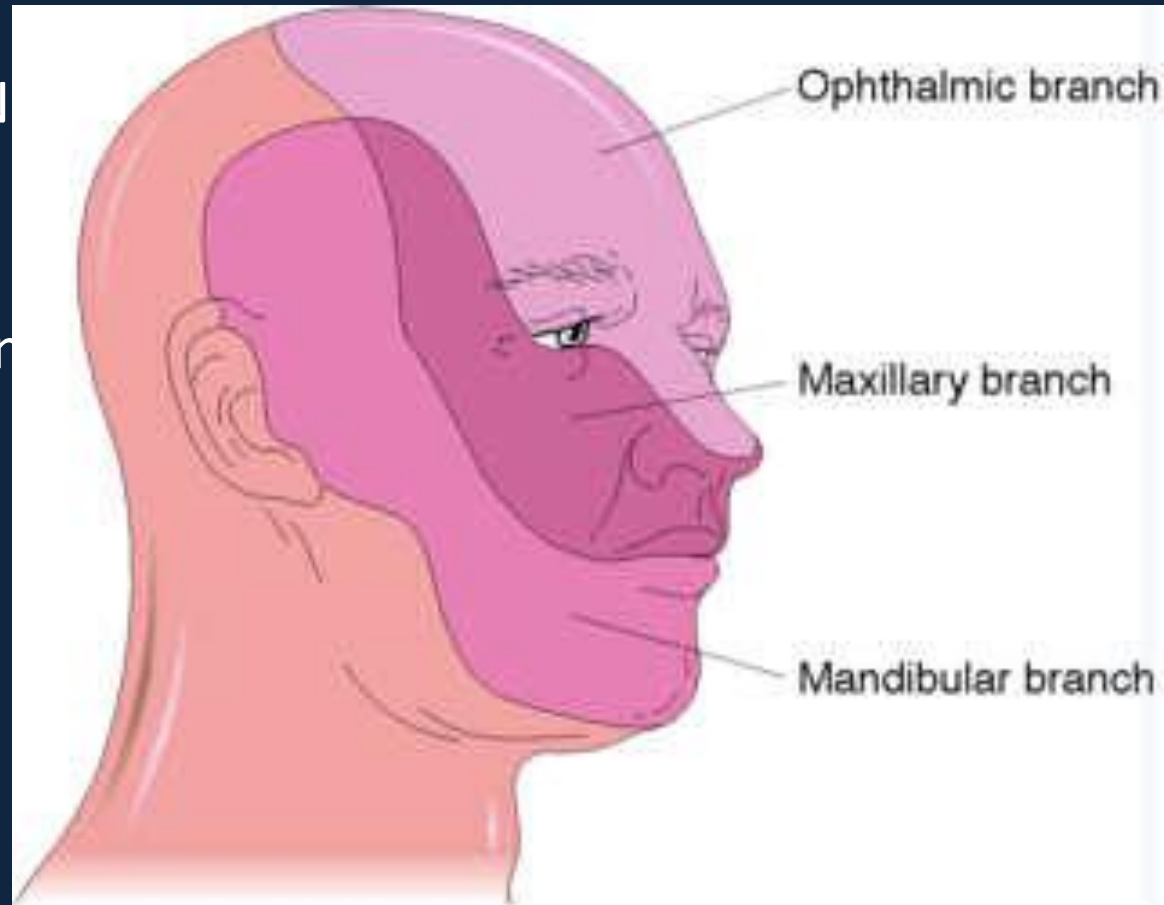


# History and Physical

- Physical exam: should be normal in TGN – any deficit in previously un-operated patient should prompt search for tumor or other lesion

## History and Physical

- Assess all divisions of CN V – including corneal reflexes
- Assess masseter function (bite) and pterygoid function (on opening mouth the chin deviates to the weak side)



# Imaging

- Old teaching: imaging required only when atypical features are present
- Current wisdom: Advanced imaging may be able to identify, pre-operatively, vascular compression affecting the trigeminal nerve

# Medical Management

- Carbamazepine (Tegretol): complete or acceptable relief in 69%
  - If 600-800 mg/d are tolerated and give no relief, the diagnosis of TGN should be questioned
- Lyrica
- Neurontin

# Medical Management

- Dilantin
  - may use in IV form for patients in too much pain to open mouths for oral Tegretol
- Capsaicin (Zostrix)
  - Red Hot Chili Pepper
- Clonazepan (Klonopin)
  - Works in 25%
- Amytriptyline

# Medical Management

- Status Trigemini: a rare manifestation of TGN characterized by rapid succession of tic-like spasms triggered by seemingly any stimulus
  - IV Tegretol (if available) or IV Dilantin

# Percutaneous Ablation

- Most employ the technique/landmarks initially described by Hartel
  - 1) Beneath the medial aspect of the pupil
  - 2) 3 cm anterior to the external auditory meatus
  - 3) 2.5 cm lateral to the oral commissure

# Percutaneous Ablation

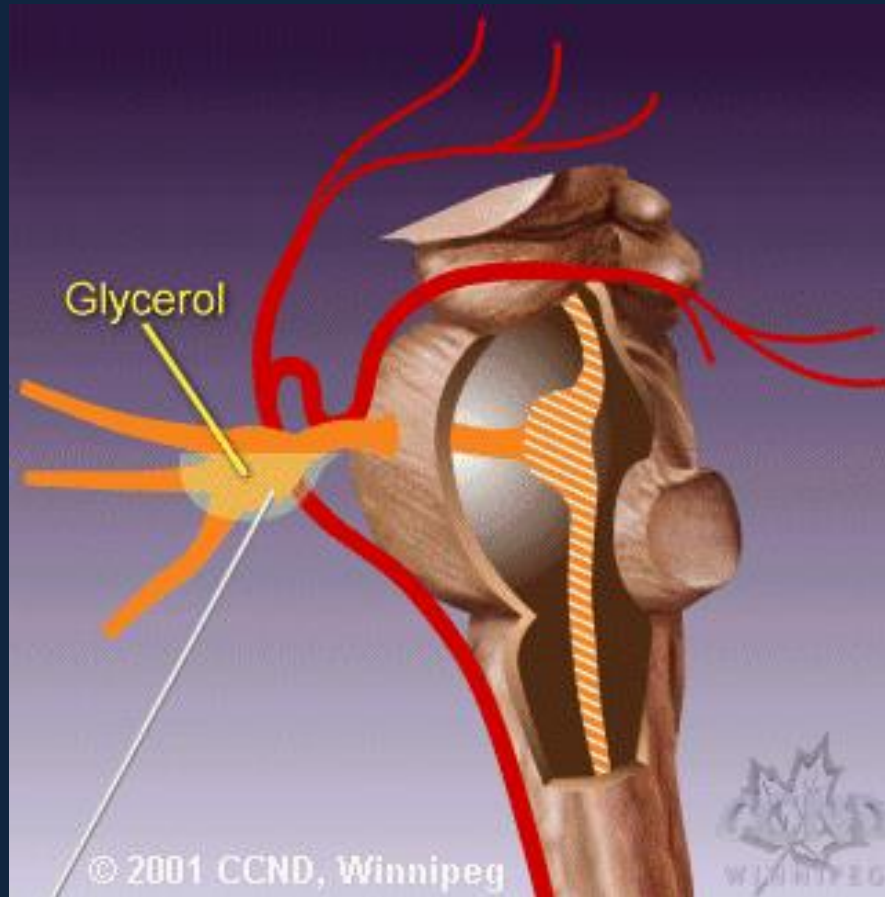
- Finger intraorally and inferior to lateral pterygoid wing, 18 guage needle introduced at the point 2.5 cm lateral to oral commissure.
- Trajectory approximates intersection of coronal plane passing through the point 3cm anterior to tragus and sagittal plane passing through medial aspect of pupil.
- Lateral fluoroscopy used to direct needle into foramen ovale



# Glycerol Rhizolysis

- Development of stereotactic technique to deliver gamma irradiation to the trigeminal ganglion
- Tantalum dust was injected into the trigeminal cistern using glycerol as a carrier
- Even without irradiation, patients had resolution of their pain

# Glycerol Rhizolysis



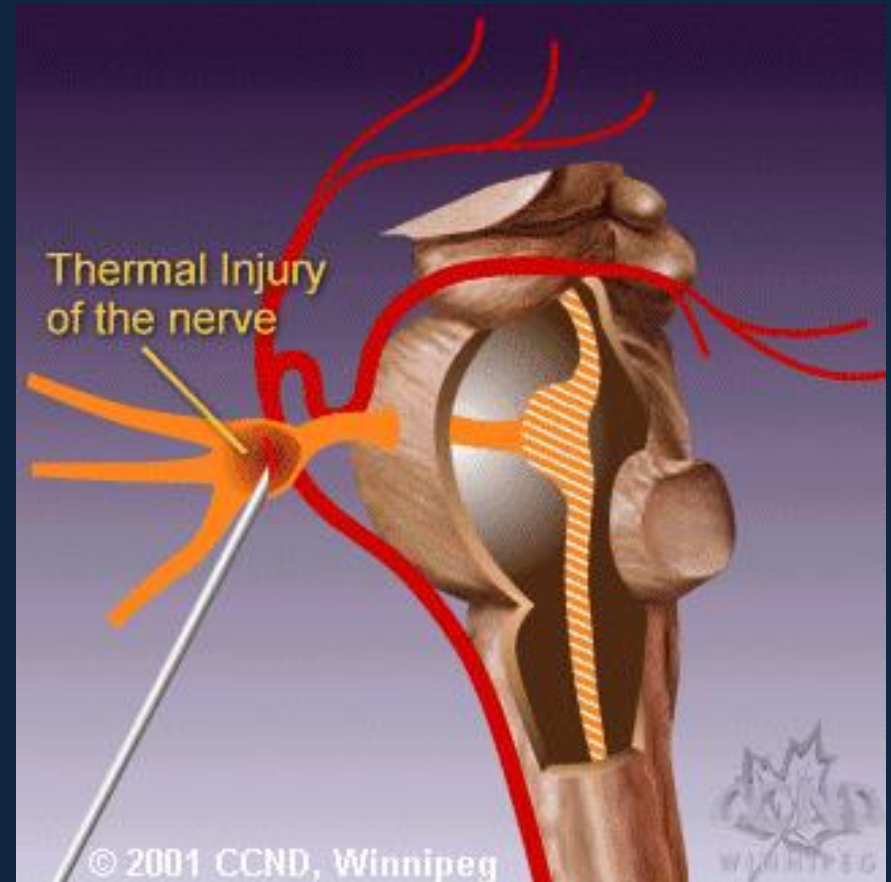
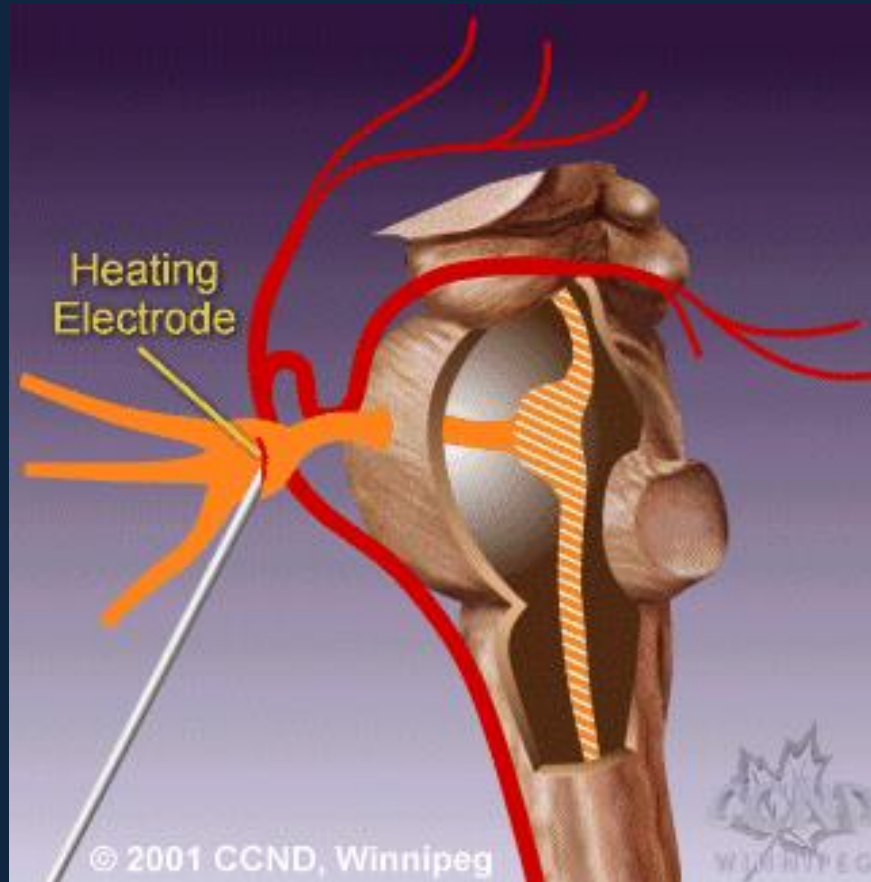
# Glycerol Rhizolysis

- Initial pain relief: 72-96%
- Recurrence rate: 10-92% (longer follow-up periods identify higher recurrence)
- The largest study of glycerol rhizolysis (N=522)
  - Recurrence at 2 years: 41%
  - Recurrence at 5 years: 83%
  - Recurrence at 6 years: 92%
- Median pain-free interval: 16-32 months

# Glycerol Rhizolysis

- Complications:
  - Hypesthesia: 46-63%
  - Dysesthesia:
    - Minor: 10-15%
    - Major: 0-3%
  - Corneal anesthesia: 2-8%
  - Anesthesia dolorosa: 0-2.5%

# Radiofrequency Thermocoagulation



# Radiofrequency Thermocoagulation

- High rates of initial pain relief: 97-100%
- Recurrence rate: 13-37%
- Early studies showed a high rate of complications (dysesthesia and masseter weakness), and the procedure fell out of favor
- Subsequent modifications have reduced morbidity

# Radiofrequency Thermocoagulation

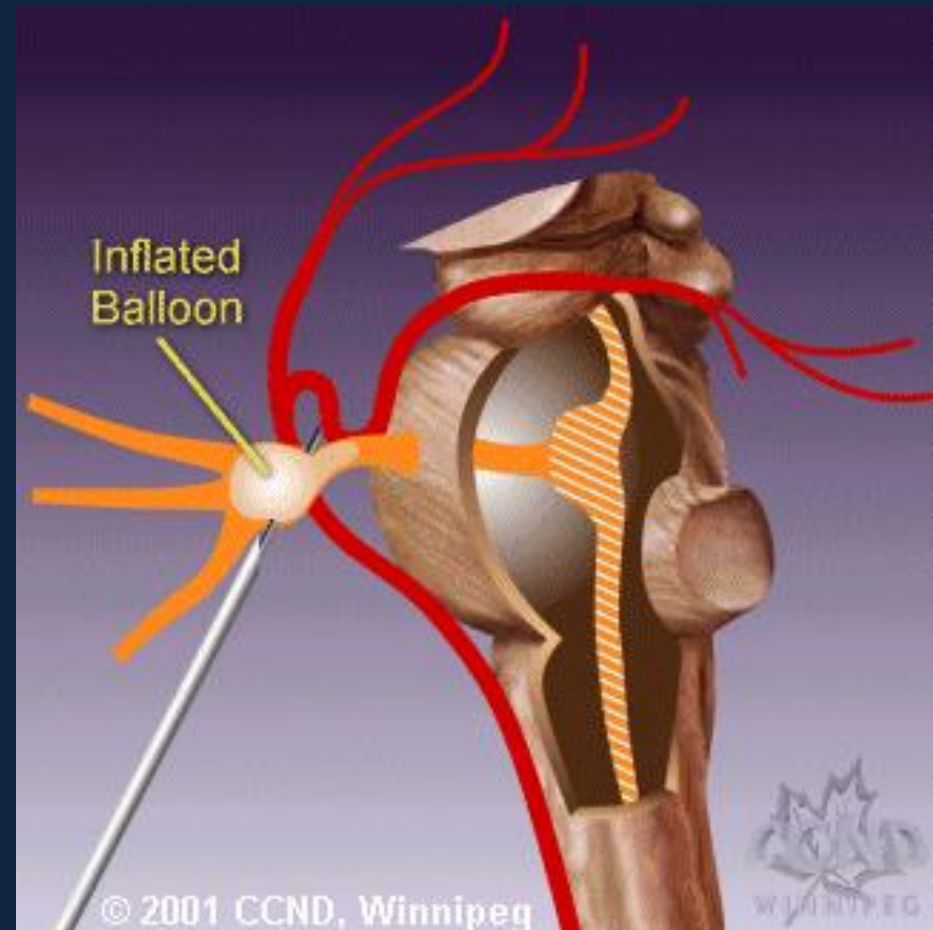
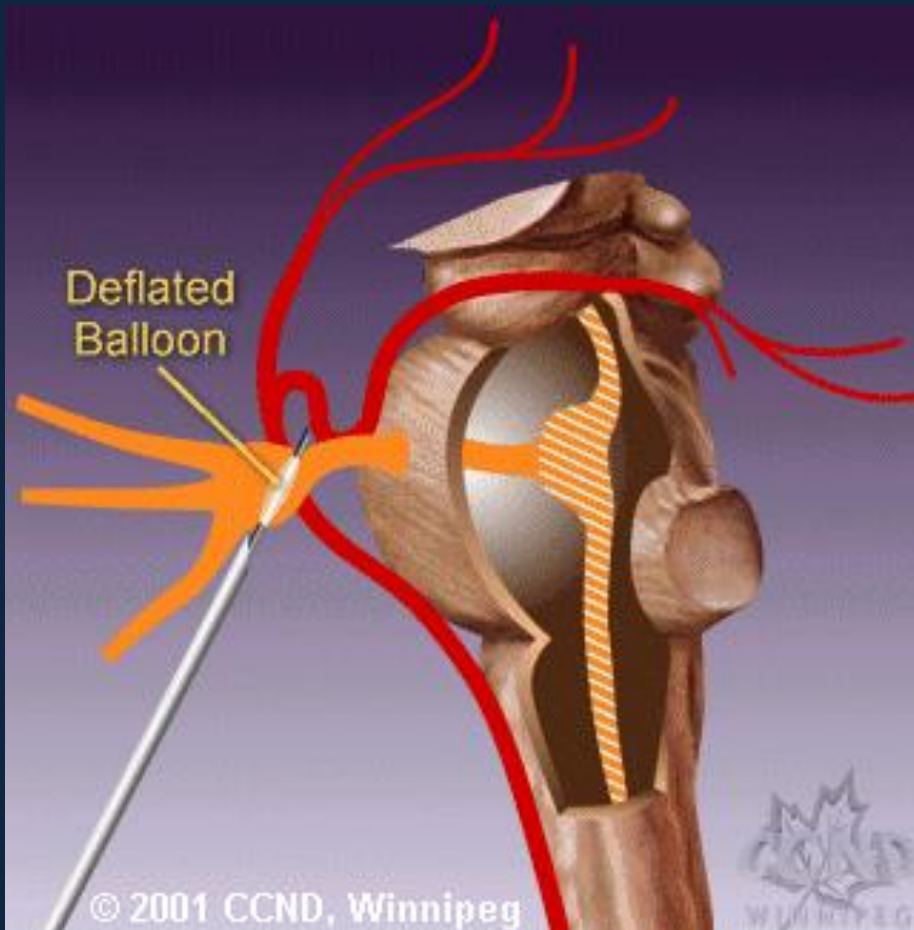
- Using a curved-tip electrode in 500 patients treated with RT, only 9% of patients reported intermittent dysesthesia that did not require treatment
- The largest series in the literature (N=1600) showed an overall recurrence rate of 25%
  - 719 patients followed 5 years, recurrence: 43.3%
  - 365 patients followed 10 years, recurrence: 47.7%
  - Improved outcome with multiple procedures
  - After 5 yrs, 10 yrs, 92% and 94% reported absence of neuralgia with 1 or more RT procedures

# Radiofrequency Thermocoagulation

- Radiofrequency Thermocoagulation requires the cooperation of the patient
- This should be considered when deciding to proceed with this technique



# Balloon Compression



# Balloon Compression



# Balloon Compression

- Initial relief: near 100%
- Recurrence rates: 10-33% (most recur in the 2<sup>nd</sup> year of follow up)
- Complications:
  - Dysesthesia: 4%
  - Bradycardia/hypotension: used by some as an indicator of penetration into the foramen ovale
    - Need for atropine administration rare

# Stereotactic Radiosurgery

- Used to lesion CN V at the REZ
- Retrospective single institution reviews with little long term follow-up
- Patients described are typically older and have failed at least 1 other surgical intervention

# Stereotactic Radiosurgery

- Largest study (N=220) from University of Pittsburgh
- Mean age 70 (29-92)
- >60% had undergone previous surgery
- >30% had a sensory disturbance
- Most treated with single 4mm isocenter with maximum dose of 70-80 Gy
- Median follow-up 2 years

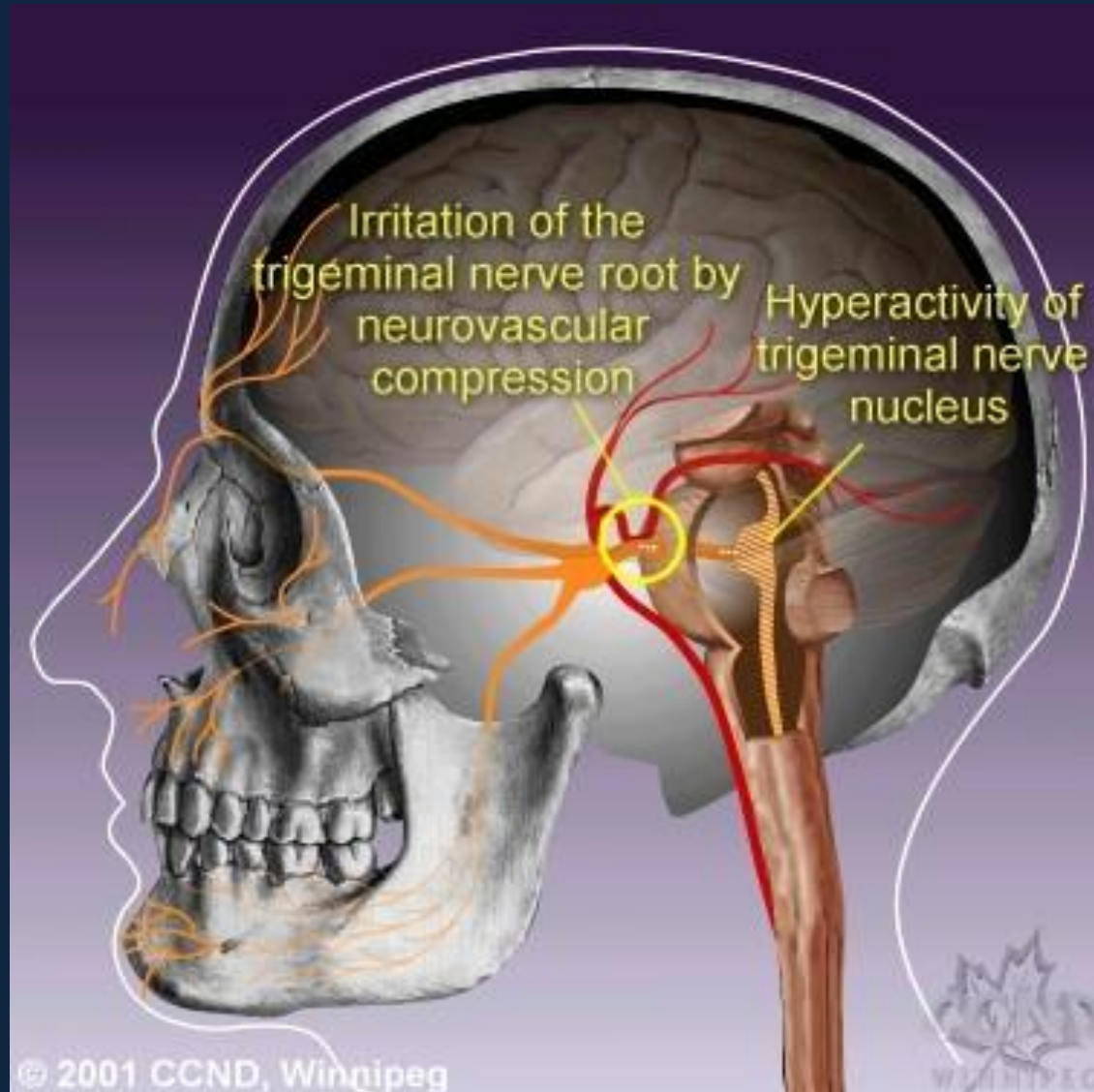
# Stereotactic Radiosurgery

- Median time to initial response: 2 months
- At 1 year complete or partial relief: 85%
- At 5 years complete or partial relief: 56%
- 10% experienced new facial numbness or paresthesias at 2 years (most complications observed within the first year)

# Stereotactic Radiosurgery

- Another large cohort study (N=117)
- Similar patient profile
- Higher dose (4mm isocenter, 90 Gy)
- Complete pain free medication free:
  - 1 year: 57%
  - 3 years: 55%
- Slightly higher complication rate: 24%

# Neurovascular Compression Syndromes





# Neurovascular Compression

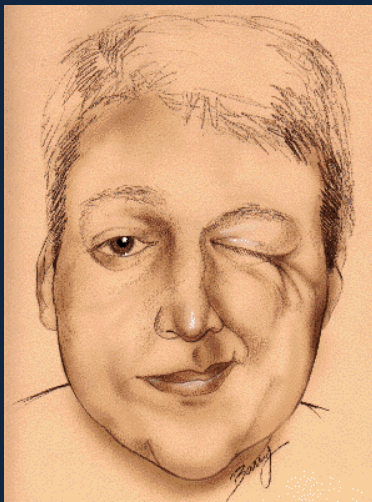
## Syndromes

### Pathophysiology

- Ephaptic transmission in trigeminal nerve from large diameter myelinated A fibers to poorly myelinated A-delta and C (nociceptive) fibers
- May explain why innocuous stimulation of trigger zones within distribution of same trigeminal branch causes attack

# Neurovascular Compression Syndromes

- **Trigeminal neuralgia**
  - “tic douloureux”
  - Superior cerebellar artery (SCA) typically culprit vessel
  - MVD 80-97% effective for typical TN
- **Hemifacial spasm**
  - Culprit vessels:
    - Anterior inferior cerebellar artery (AICA), Posterior inferior cerebellar artery (PICA), Vertebral artery, Venous structures
  - MVD 85-93% effective for classic HFS



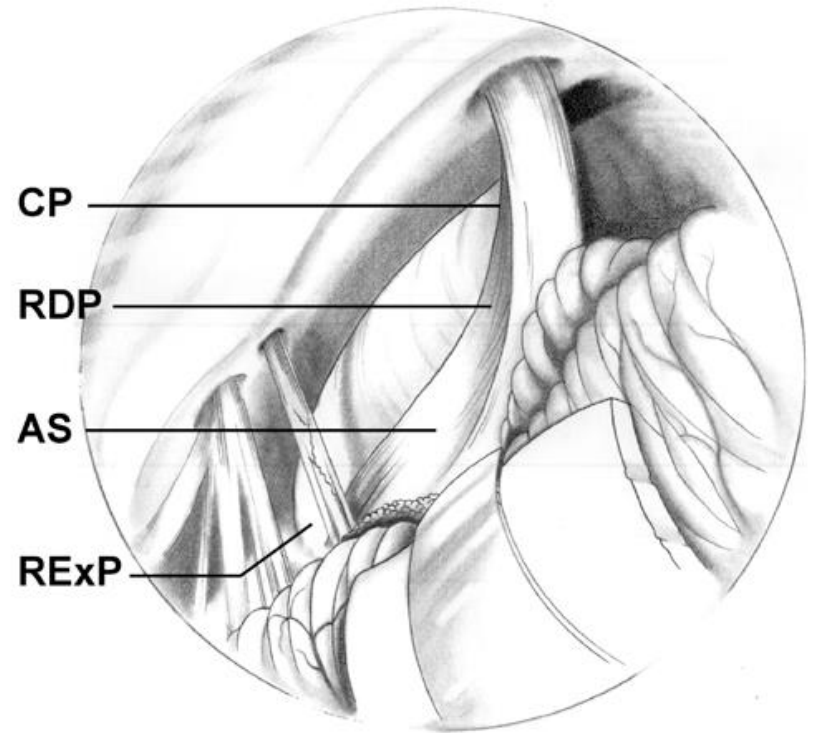
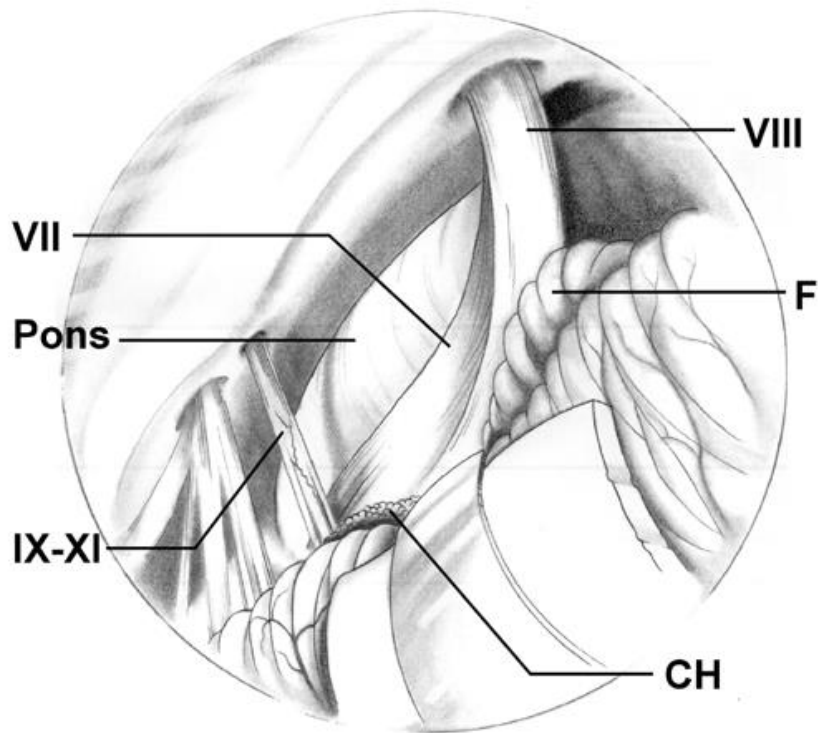
# Hemifacial spasm

- A syndrome of facial muscle spasms that typically begin with blepharospasm and progress to involve the lower facial muscles.
- Differential diagnosis for blepharospasm:
  - Tardive dyskinesia
  - Meige syndrome
  - Facial Myokymia
  - Benign essential blepharospasm
  - Dopamine agonists

# Hemifacial spasm

- Etiology:
  - Vascular compression of the facial nerve.
  - Tumor compressing facial nerve
  - Post-Bell's palsy
- Treatment:
  - Oral medications typically ineffective
  - Can be managed temporarily with botulinum (BoTox) injections of facial muscles
  - Definitive therapy: Microvascular decompression (MVD) of the facial nerve
    - 85-96% efficacy
    - Can take up to 3 years for full resolution

# Hemifacial spasm



*Neurovascular compression: primary culprit vessel, location, and severity\**

Vessel	Location				Severity		
	RExP	AS	RDP	CP	Mild	Moderate	Severe
AICA	4	26	16	3	12	30	7
PICA	3	25	7	1	10	23	3
VA	5	20	1	0	6	17	3
vein	0	3	1	0	3	0	1
total	12	74	25	4	31	70	14

# Neurovascular Compression Syndromes

- **Glossopharyngeal neuralgia**
  - Culprit vessels: AICA or PICA
  - MVD 90% effective
- **Disabling positional vertigo (DPV)**
  - Constant positional vertigo without hearing loss or vestibular dysfunction
  - Associated with compression of the vestibulocochlear nerve
  - Can respond to MVD

# Neurovascular Compression Syndromes

- **Geniculate neuralgia**
  - “Hunt’s neuralgia”
  - Facial nerve neuralgia
    - paroxysmal otalgia
    - Prosopalgia (deep facial pain)
  - Can be caused by herpetic ganglionitis (Ramsay-Hunt Syndrome)
  - Can be associated with hemifacial spasm
    - “tic convulsif”
    - AICA typical culprit vessel

# Intra-operative Considerations

- Positioning
- Anesthesia
- Neuromonitoring
- Microscopy
- Endoscopy
- Nerve decompression
- Closure



# Beach Chair Position



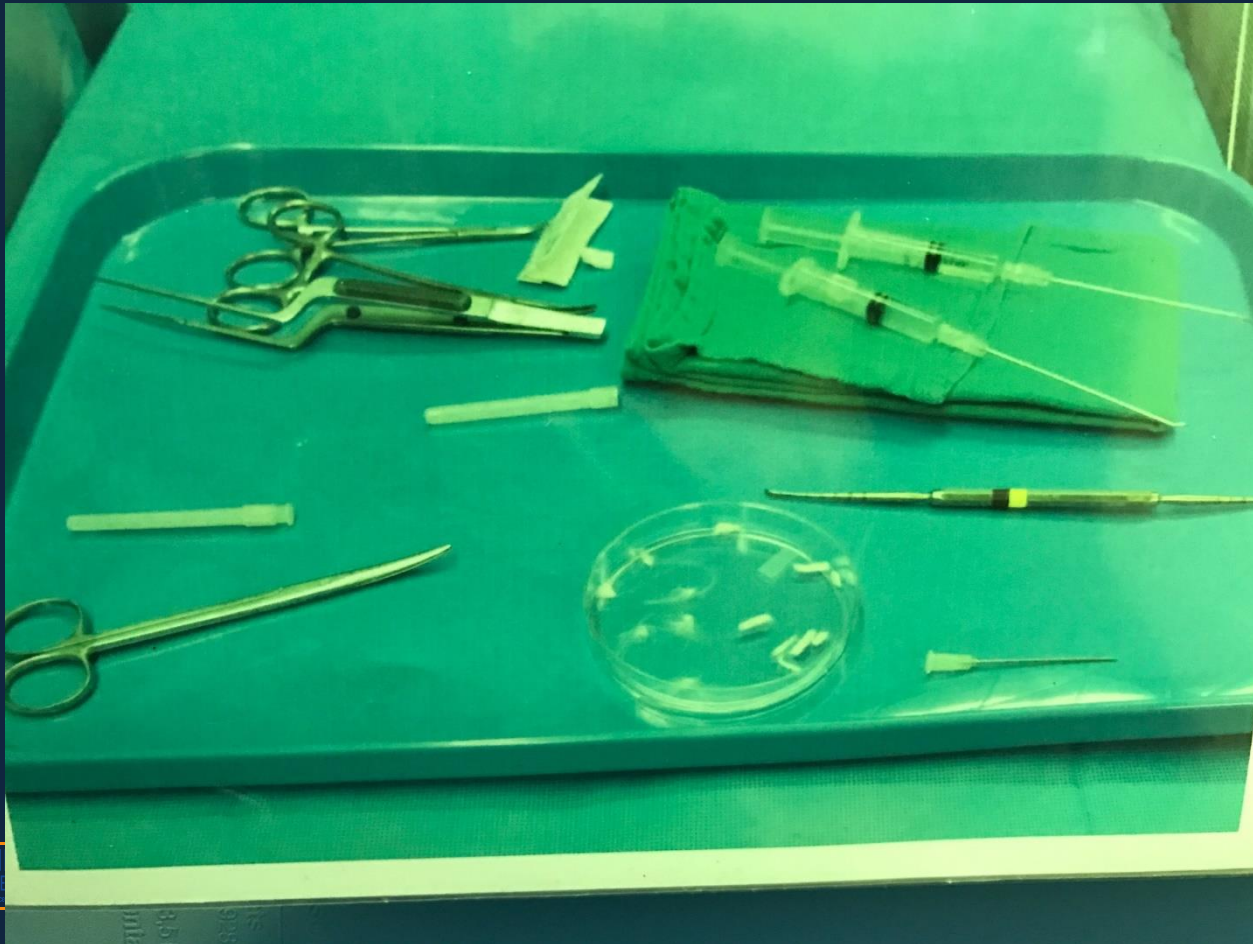
# Beach Chair Position







# Teflon Back-table



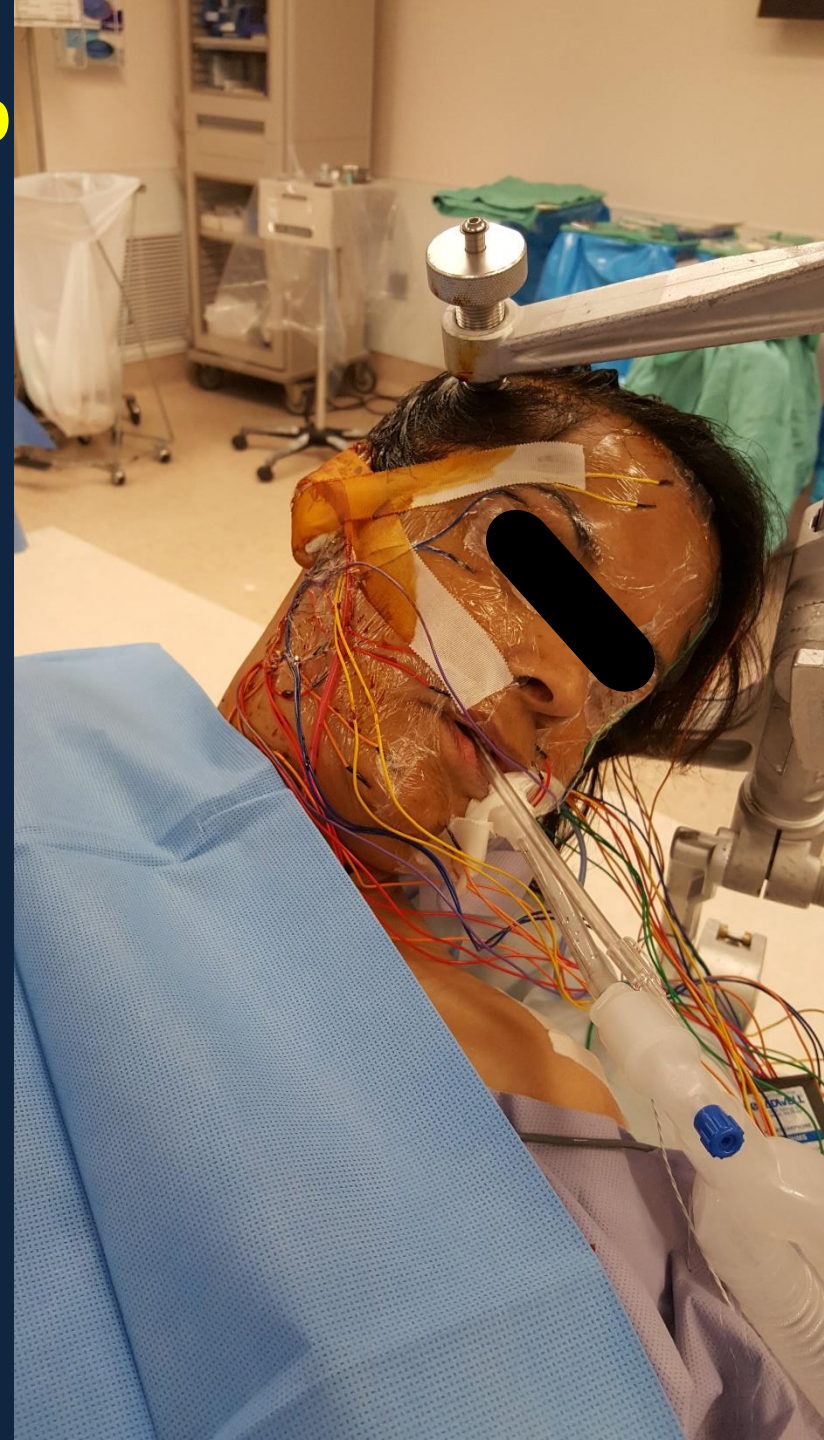
# Anesthesia

- **Total IV Anesthesia (TIVA)**
- **Mannitol 25g**
- **Mild hyperventilation**
- **Dexamethasone – to prevent reaction to Teflon or glue**
  - Continued for 12 days slow taper
- **Antibiotics**
- **NO: Keppra necessary**



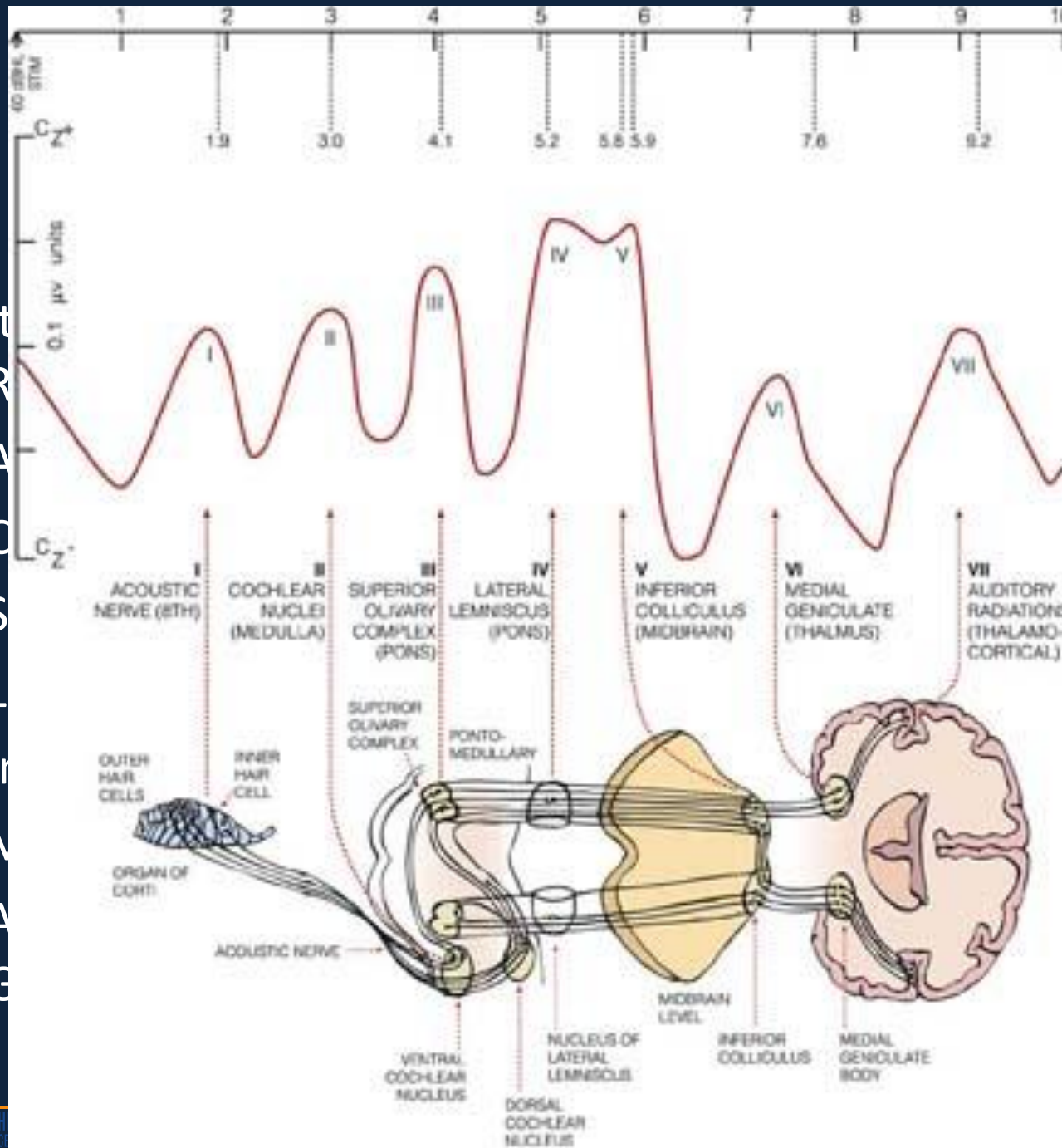
# Neuromonito

- Brainstem Auditory Evoked Response (BAER)
- Somatosensory Evoked Potentials (SSEP)
- Cranial nerve V and VII



# Brainstem

1. Auditory
2. Cochlear
3. Superior
4. Lateral
5. Inferior
6. Nucleus
7. Auditory



14:41:43

ms	Diff:	ms
uv	Diff:	uv
nHL	Delay:	0ms

Latency and Amplitude				
LAT	LAT	AMP	AMP	
ms	ms	uv	uv	
II	V	1.11	1.11	
III	V	5.68	0.260	



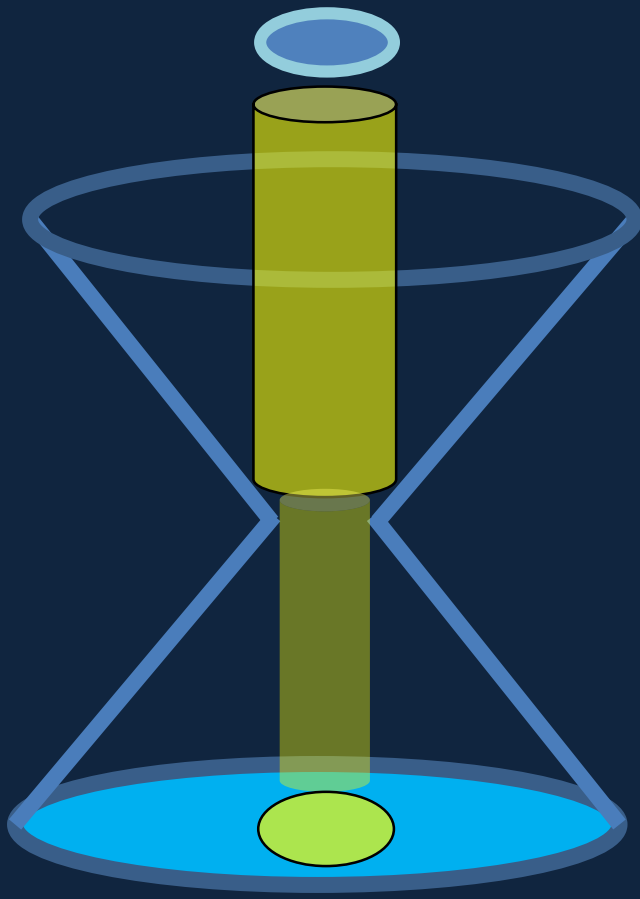
# Keyhole Surgery



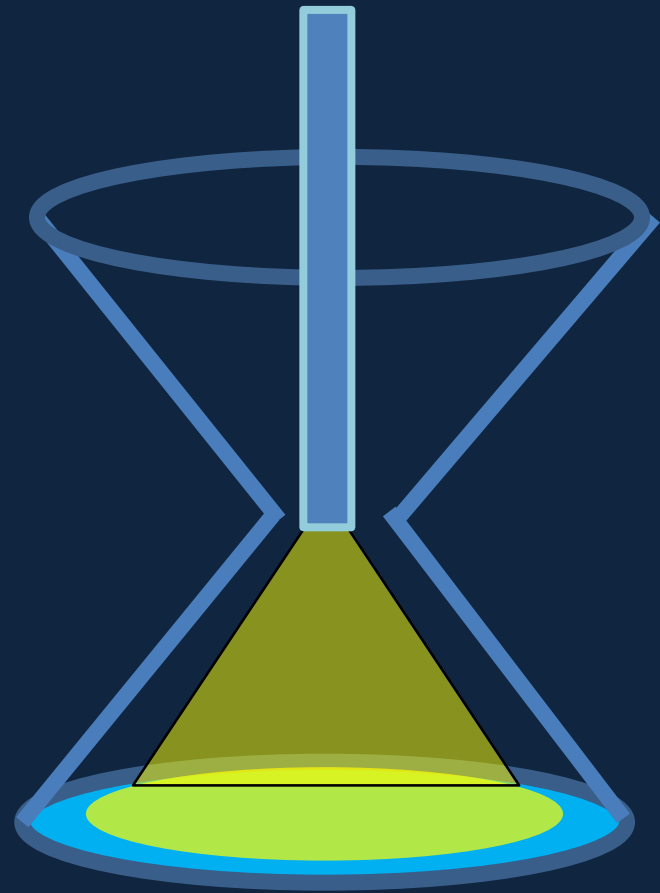


# Microvascular Decompression





Microscope



Endoscope







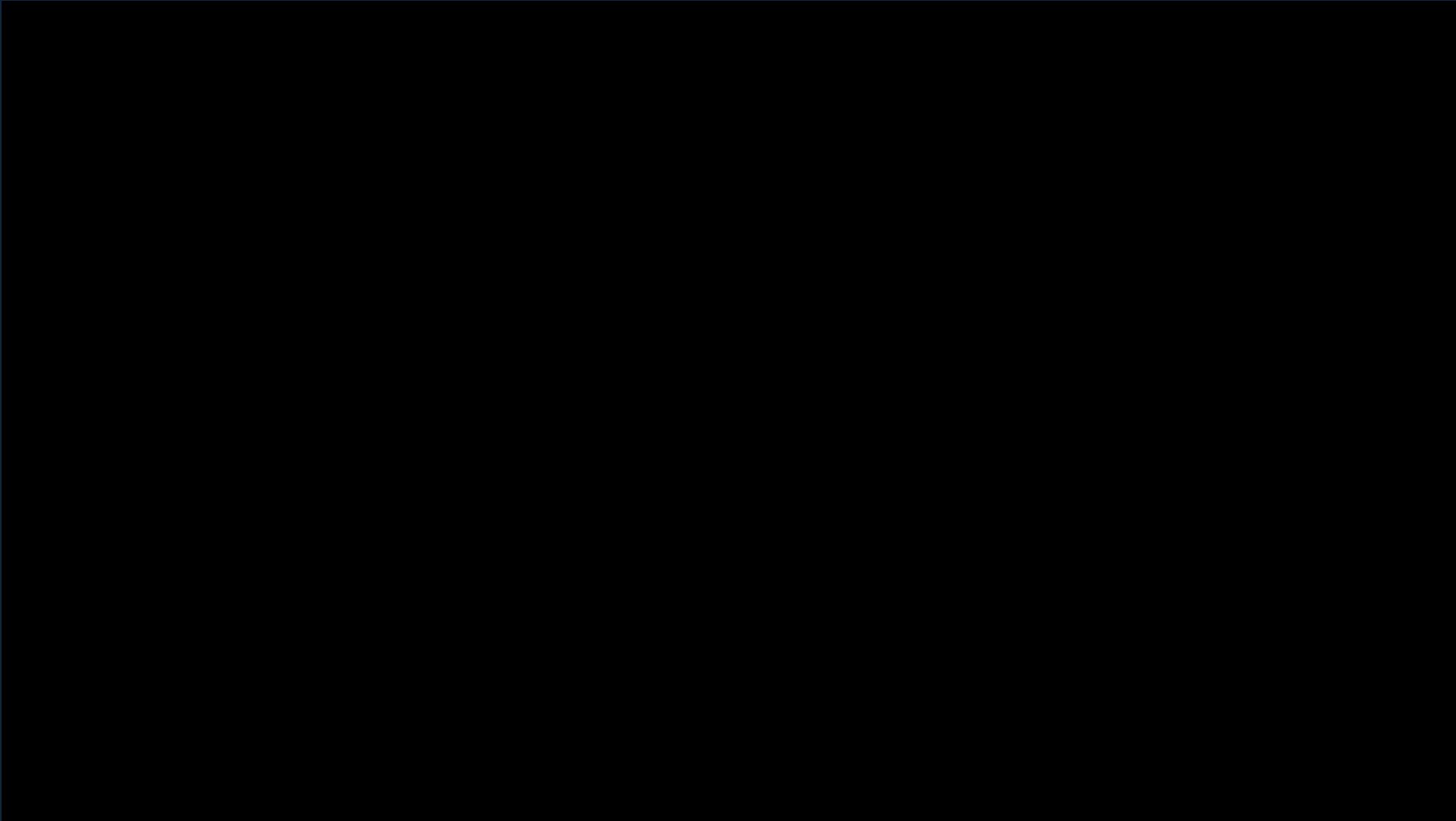








# Endoscopic MVD





# Post-operative Considerations

- **Blood-pressure management**
- **Delayed Hydrocephalus Development**
- **Cerebrospinal Fluid Leakage**
- **Facial nerve weakness**
- **Hearing loss**
- **Failure to improve pain or spasms**

# Pacific Facial Pain Center

## Facial Pain Neurology



Marisa Chang, MD

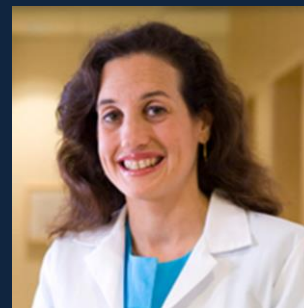


Daniel Franc, MD, PHD

## Neurosurgery



## Radiation Oncology



Lisa Chaiken, MD



Robert Wollman, MD

# Thank You

