

CRANIAL NERVE COMPRESSIVE NEUROPATHIES

TRIGEMINAL NEURALGIA

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Cranial nerve compressive neuropathies

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

Trigeminal Neuralgia

What is it?

TGN-Historical perspective

- **Fallopian** : documented the trigeminal nerve in the 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 douloureux’ 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 put off sleep’
- **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
- Incidence : 4.3 / 100,000
- Age group : 50 - 60 years
- Paroxysmal recurrent pain of short duration
- Trigger point(s), allodynia
- Periods of remission
- Recurrent episodes of pain with progressively shorter periods of remission
- Distribution of pain :
 - localised to Trigeminal nerve
 - 80% Maxillary / Mandibular combination
 - Right side > Left side
- Clinical examination : Usually normal

TGN-Differential diagnosis

- Dental / Sinus related pain
- Cluster headache / Migrainous neuralgia
- Atypical facial pain
- Post - herpetic neuropathy
- TM joint dysfunction
 - Degenerative , Rheumatoid arthritis
- Trigeminal neuropathy
 - Demyelination : Multiple sclerosis
- Facial myalgia

Trigeminal Neuralgia

What causes it

Pathophysiology

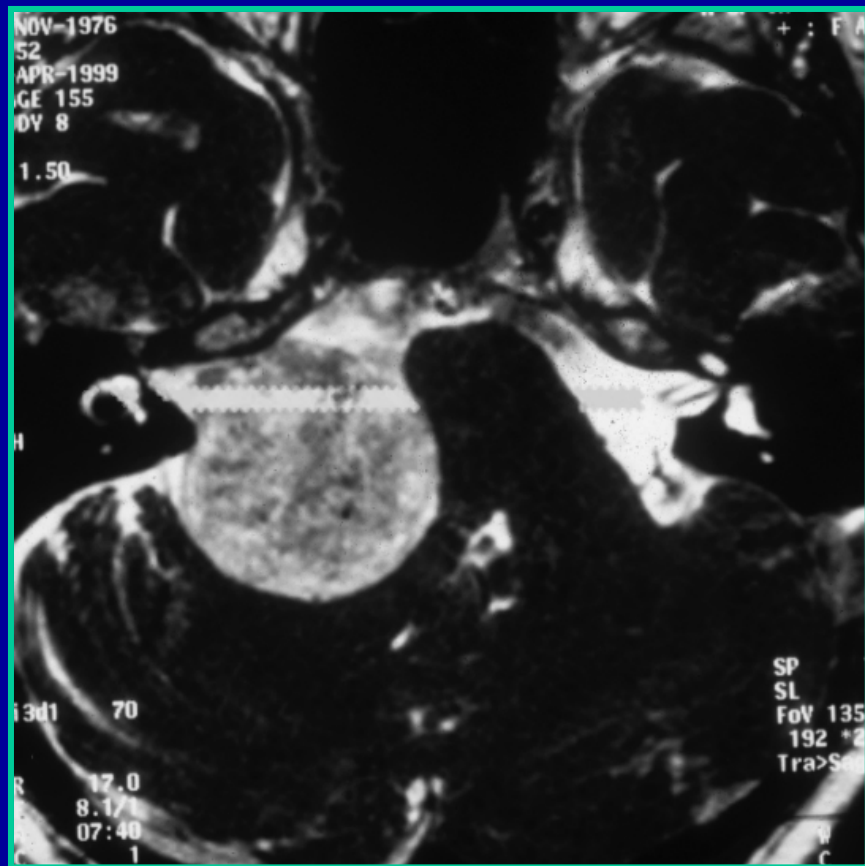
- **Compression at root exit zone** : at junction of central myelin (oligodendroglial cells) and peripheral myelin (Schwann cells) : the **Obersteiner-Redlich** line
 - Vascular cross compression of Root Exit Zone
 - Atherosclerotic degeneration causing elongation and tortuosity
 - Neoplastic compression
 - Meningioma, Schwannoma, Neuroma, Epidermoid
- **Focal segmental axonal demyelination** with aberrant synaptogenesis resulting in transaxonal ephaptic transmission
- **Spontaneous discharge** occur at site of compression
- Evoked potentials studies demonstrate increased latency & threshold consistent with nerve compression

Pathophysiology

- Association between multiple sclerosis and demyelination
 - 4% of patients with MS have TGN
 - *Harris 1950 Rare forms of paroxysmal trigeminal neuralgia and their relation to disseminated sclerosis BMJ 2 1015 - 1019*
 - 2% of patients with TGN have MS

Trigeminal Neuralgia : Acoustic Neuroma

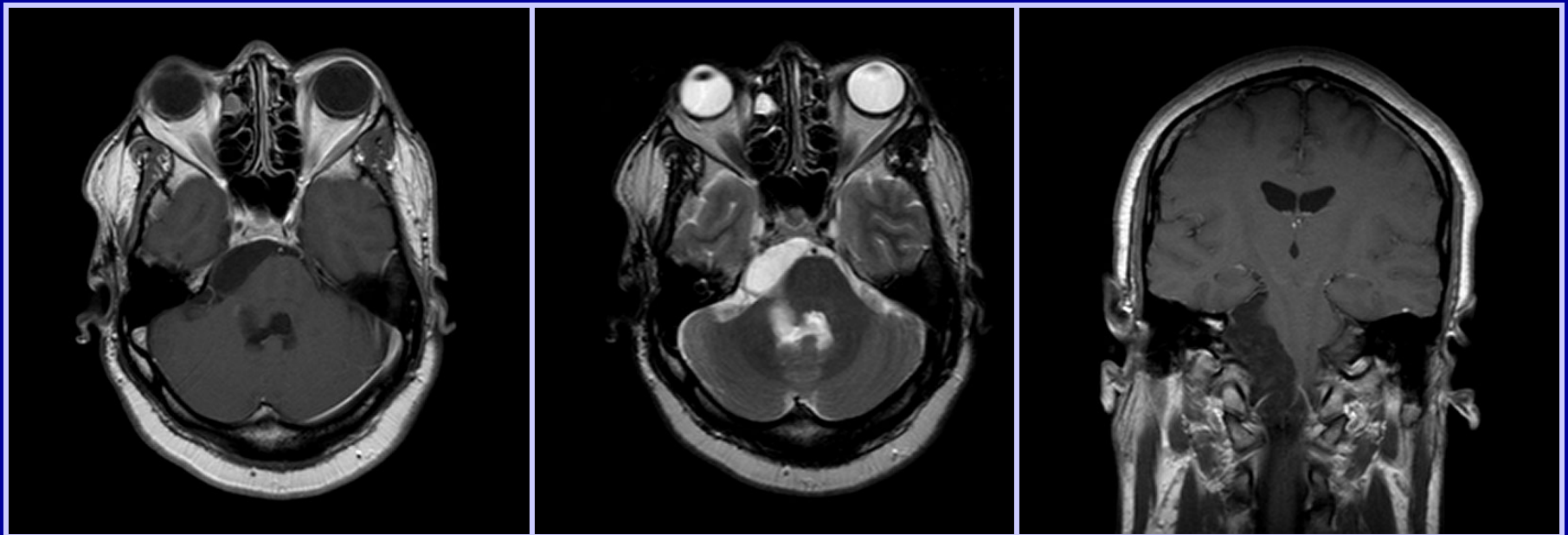
30 yr male with 10 yr history of unilateral deafness and trigeminal neuralgia



Trigeminal Neuralgia : Epidermoid tumour

45 yr male with 3 yr of classic Trigeminal neuralgia

MRI



Trigeminal Neuralgia

What happens if you have TGN ?

Natural history

What we know

No prospective natural history studies in the literature

Surgical series suffer from lack of class 1 evidence

What we think we know

The progress is one of increasing severity with diminishing intervals of remission

Pain becomes more constant and atypical

Pain becomes resistant to medical treatment

TRIGEMINAL NEURALGIA

What can be done about it ?

Levels of Evidence

Level	Type of evidence
1a	Meta-analysis of randomised controlled trial
1b	At least one randomised controlled trial
2a	At least one non randomised controlled trial
2b	At least one type of quasi-experimental trial
3	Non - experimental descriptive study, such as comparative, correlation and case-control studies
4	Expert committee reports/opinions and / or clinical experience of respected authorities

US Agency for Health Care Policy and Research

Quality of reporting in evaluations of surgical treatment of Trigeminal Neuralgia : Recommendations for future reports

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

- **Aims**

- to set criteria and quality standards for reporting outcome of surgical treatment
- 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 treatment modalities
- Non - uniform outcome measures results in difficulty with comparability of studies

TGN

Medical options

- Carbamazepine
 - Blom 1962
 - Initial small dose with incremental increase
 - s/e : Gait ataxia, memory disturbance, nausea, blood dyscrasia
- Oxcarbazine
- Phenytoin
- Gabapentin / Pregabalin /Lamotrigine
- Amitriptyline
- Baclofen

TGN-Indications for surgical treatment

- Failure of medical management
 - no or poor response
 - unacceptable side effects
 - ataxia
 - memory disturbance
 - skin rash
 - neutropeania
- Patient choice

Aims of surgical intervention

- Complete lesion removal
- No mortality
- No neurological morbidity
- No recurrence
- No vascular morbidity
- ‘Results’ are better than natural history

A trade off between simplicity and recurrence

A trade off between advantages and disadvantages

TGN-Surgical options

- **Peripheral neurectomy**
 - nerve avulsion
 - cryotherapy
- **Percutaneous Rhizotomy**
 - Meckels cave Glycerol injection
 - Radiofrequency thermocoagulation
 - Balloon microcompression
- **Microvascular Decompression**
- **Partial sensory nerve section**
 - Infratemporal approach (Frazier approach)
 - Posterior fossa
- **Stereotactic radiosurgery**
- **Medullary tractotomy**

Microvascular decompression : Historical perspective

- **Dandy** : 1934 suggested causal relationship between vascular cross compression and TGN
 - *Dandy WE. Concerning the cause of trigeminal neuralgia. Am J Surg 24 ; 447 -455*
- **Gardner & Miklos** : 1959 positioned gelfoam between the artery and nerve
 - *Gardner and Miklos. Response of trigeminal neuralgia to decompression of sensory root. JAMA 170 ; 1773 - 1776*
- **Jannetta** : ‘popularised’ the concepts of vascular compression and surgical treatment
 - *Arterial compression of the trigeminal nerve at the pons in patients with trigeminal neurlagia. J Neurosurg 1967 : 36 ; 159 - 162*
 - *Treatment of trigeminal neurlgia by suboccipital and transtentorial cranial operations. Clin Neurosurg 1977 : 24 ; 538 - 549*

Surgical treatment

What we know

- No prospective randomised controlled trials
- Data invariable retrospective
- Difficult to compare results between centres

What we think we know

- MVD
 - lowest rates of recurrence, and sensory loss
 - Low mortality (0.3%)
 - Low rate of neurological morbidity
 - 1% deafness

Surgical treatment

What we think we know

- MVD has lowest rate of complications from sensory loss
- MVD produces longest lasting relief
- MVD addresses apparent major cause
- No relationship between sensory disturbance and outcome with MVD

What we observe

- Destructive procedures : Duration of pain relief correlates with degree of numbness produced
- Successive procedures : Pain free interval decreases with pain becoming more atypical / neuropathic

Preoperative imaging : Detecting vascular compression

How accurate is MRA in predicting neurovascular compression in patients with TGN?

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

- 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

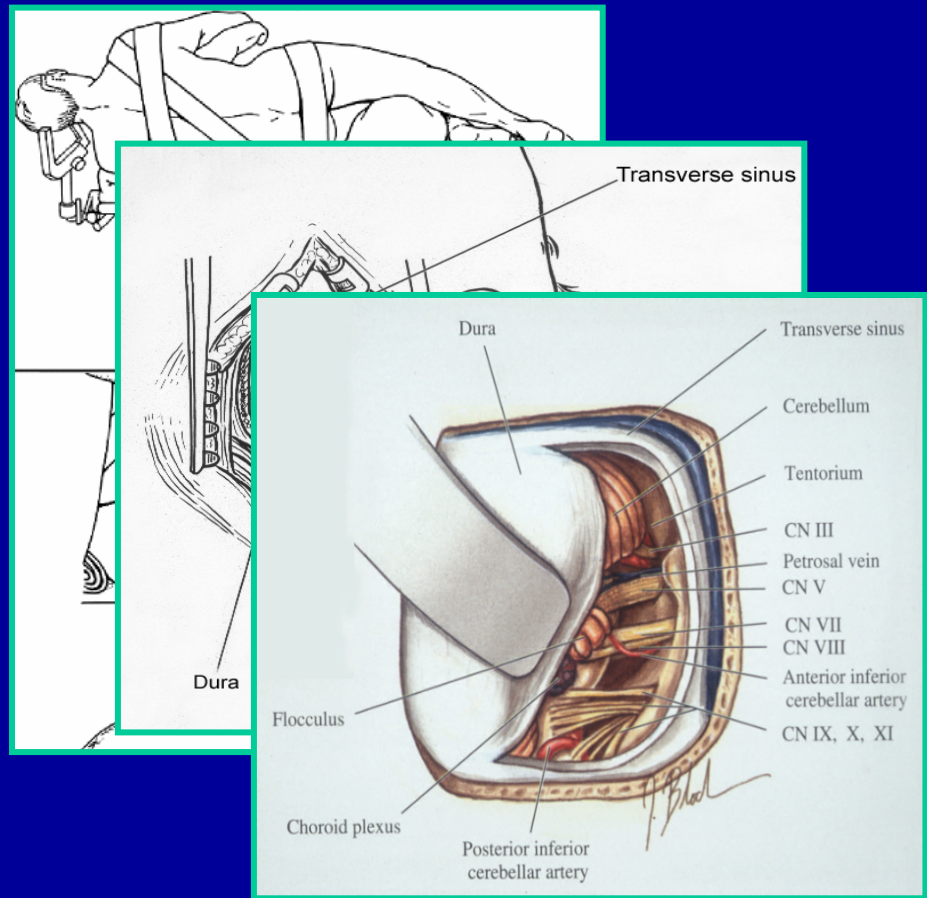
Sensitivity of MR : 90.5%

8 false negatives

Does not detect *all* cases of compression

Operative approach : MVD

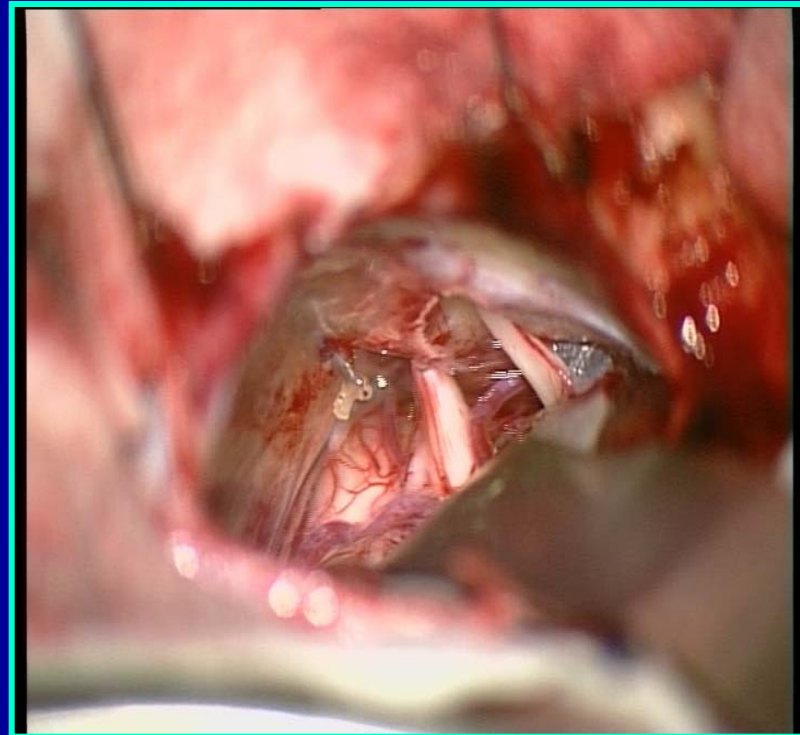
- General anaesthesia
- Park bench position
 - Head position very important
- Lumbar drainage
- Retromastoid craniectomy
 - Exposure of sigmoid and transverse sinus
- Retraction of cerebellar hemisphere
 - opening of arachnoid of superior cerebellar pontine cistern
 - Division of Vein of Dandy from superior petrosal sinus
- MVD / Nerve section
- Standard closure
- Home 5 - 7 days post op



Microvascular Decompression : 1995 - 2005

- 100 cases for TGN
 - 90 MVD
 - 10 Nerve section
- 10 cases for HFS
- Complications:
 - 2 CSF leak
 - 1 wound infection
 - 1 haematoma
 - 1 pneumocephalus

Right side : MVD



The long-term outcome of microvascular decompression for trigeminal neuralgia

Barker F, Jannetta P, Bissonette D, et al

The New England Journal of Medicine 334 ; 1077 - 1083 : 1996

- 1185 patients underwent MVD over 20 years
 - 1155 f/u after 1 year with median f/u of 6.2 years
- 70 % Pain free at 10 years
 - 11 % of recurrences underwent second procedure
- Factors associated with recurrence of pain
 - Female
 - Duration of Sx > 8 years
 - Venous compression
 - Persistent pain following MVD
- Complications
 - Death : 0.2 %
 - Cerebellar haematoma : 0.2 %
 - CSF leak : 1%
 - Hearing loss : 1%

Evaluation of MVD and partial sensory rhizotomy in 252 cases of trigeminal neuralgia

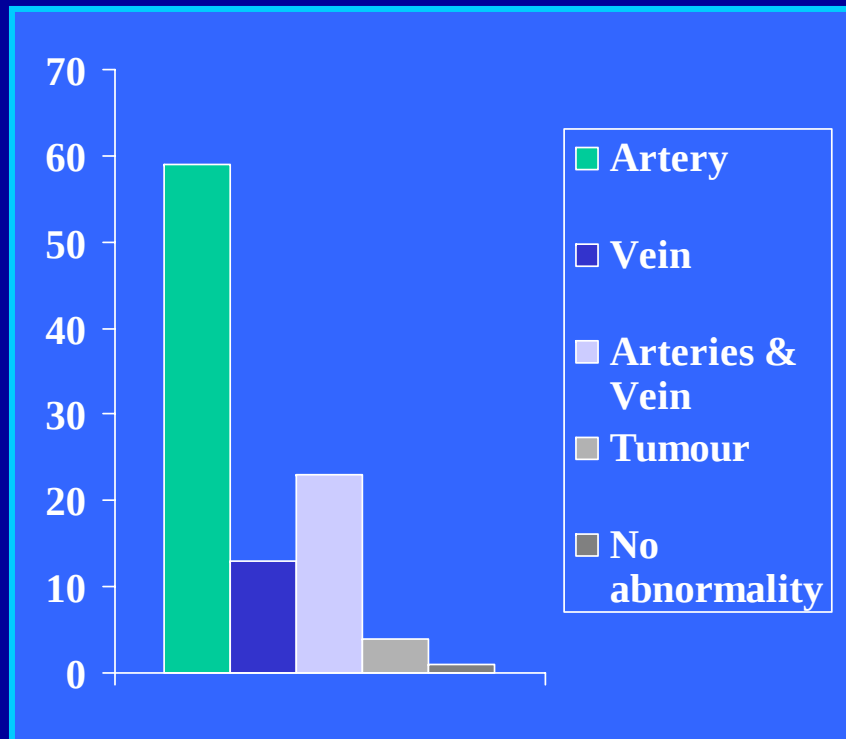
Bederson JB, Wilson CB. J. Neurosurg. 1989 ; 71: 359 - 367

- Retrospective study : Mean F/U of 5.1 years
 - pt with extrinsic compression : MVD
 - pt with no compression : Partial sensory rhizotomy
 - pt with vascular contact but no distortion : MVD + Partial sensory rhizotomy
- Results
 - Excellent : 75% / Good : 8% - 208 pts
 - Persistent pain : 5% - 13 pts
 - Recurrent pain : 12 % - 31 pts at mean of 1.9 years post op.
 - Recurrence rate 2% / year thereafter
 - Re - operation (Rhizotomy) : 85% successful
- Adverse prognostic factors
 - Percutaneous radiofrequency rhizolysis
 - Absence of vascular compression
 - Long duration of Sx in pts with vascular compression

Trigeminal neuralgia treated by microvascular decompression : a long-term follow up study

Mendoza N, Illingworth R. *British Journal of Neurosurgery* 1995 : 9 ; 13 - 19

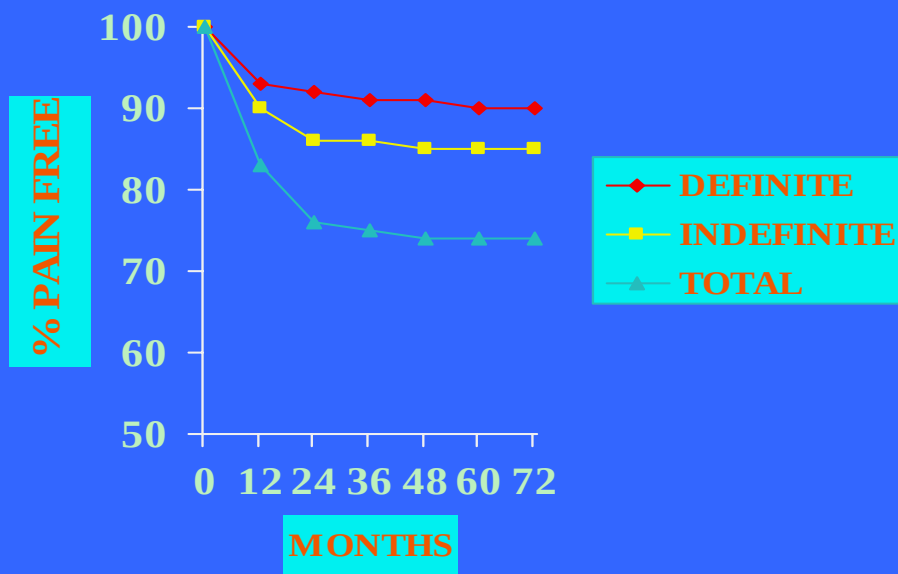
Intraoperative findings



- Vascular compression
 - SCA : 61%
 - AICA
 - PICA
 - V.A
 - Vein
- Tumour
 - Meningioma
 - Neuroma
 - Dermoid

Trigeminal neuralgia treated by microvascular decompression : a long-term follow up study

*Mendoza N, Illingworth R. British Journal of Neurosurgery
1995 : 9 ; 13 - 19*



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

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

- 278 patients underwent MVD
- 9 serious complications
 - 1 Intracerebellar ICH with acute hydrocephalus
 - 1 Supratentorial acute subdural haematoma
 - 2 Status epilepticus
 - 1 Brain stem infarct
 - 1 SAH due to 'traumatic' aneurysm
 - 1 Posterior Cerebral artery territory infarct
- 2 / 9 caused death
- Operative mortality : 0.7 %

Microvascular decompression in the U.S, 1996 -2000: Mortality rates, morbidity rates, and the effects of hospital and surgeon volumes.

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

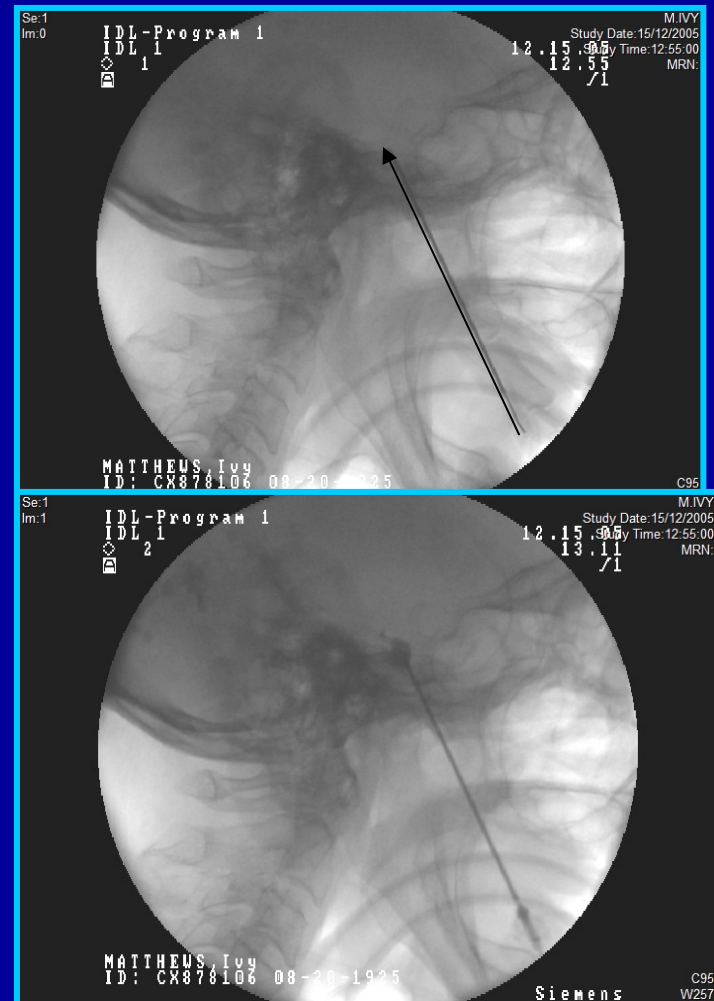
- Retrospective cohort study 1996 - 2000
- 1326 MVD for TGN (237 for HFS, 27 for GN)
- Undertaken in 307 hospitals by 277 identified surgeons
- Mortality rate was 0.3%, Rate of discharge other than home was 3.8%
- Neurological complications recorded in 1.7% (ICH 0.5%, CN7 palsy 0.6%, EVD 0.4%)
- Nerve section : 3.4 % with TGN (Older pt $p=0.08$, Female $p=0.03$, Teaching hosp $p=0.02$
- Median annual case load : 5 cases / hospital (1 - 195), 3 cases per surgeon (1 - 107)
- After adjustment for age, sex, race,diagnosis, outcomes at discharge were superior at higher volume hospitals ($p=0.006$) and higher volume (not fatter!) surgeons ($p=0.02$)
- Complications less frequent at higher volume hospital ($p=0.04$) and higher volume surgeon ($p=0.01$)
- Rate of discharge other than home was 5.1% for lowest volume hospital compared with 1.6% for highest volume hospital
- Volume and mortality rate were not significantly related (3/4 deaths by surgeons only 1 MVD / yr)

Meckels Cave Glycerol Injection

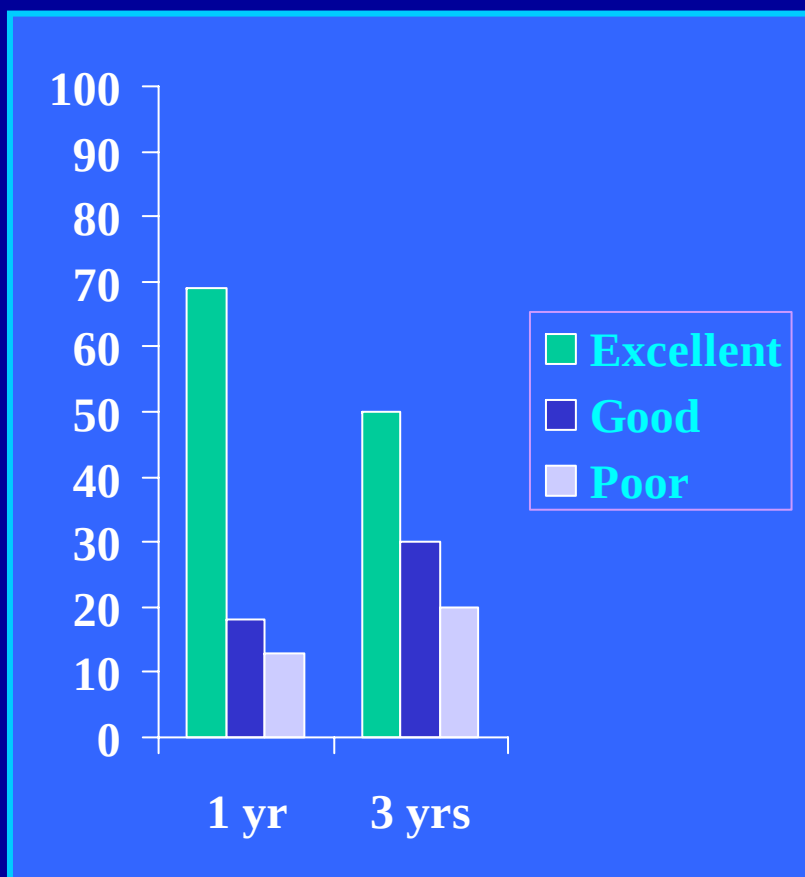
- **Hartel** : 1912 pioneered the percutaneous transovale approach to the Gasserian Ganglion using absolute alcohol
 - *Die Lietungsanasthesie und injektionsbehandlung des ganglion gasseri und der trigeminusstamme. Arch Klin Chir 1012 : 100; 193 - 292*
- **Sweet** : 1974 described glycerol injection (and radiofrequency)lesioning to the ganglion
 - *Treatment of trigeminal neuralgia and other facial pains by retrogasserian injection of glycerol. Neurosurgery. 1981 ; 9 : 647 - 653*
 - *Controlled thermocoagulation of trigeminal ganglion and rootlets for differential destruction of pain fibres. J. Neurosurg . 1974: 39 ; 143 - 156*
- **Hakanson** : 1981 accidentally discovered glycerol relieved tic pain when injecting the retrogasserian space
 - *Trigeminal neuralgia treated by the injection of gylcerol into the trigeminal cistern. Neurosurgery : 9 ; 638 - 646*

MCGI

- General anaesthesia with laryngeal mask.
- Patient is supine on theatre trolley.
- 22G Spinal needle is used.
- Standard landmarks to traverse Foramen Ovale when the tip is in the medial portion of foramen ovale , it lies 3 cms in front of the external auditory meatus on an imaginary line joining it to lateral canthus to the external auditory meatus in the mid-pupillary plane
- Clenching of jaw due to motor part of V.
- 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 & inject 1ml of anhydrous glycerol (10%).



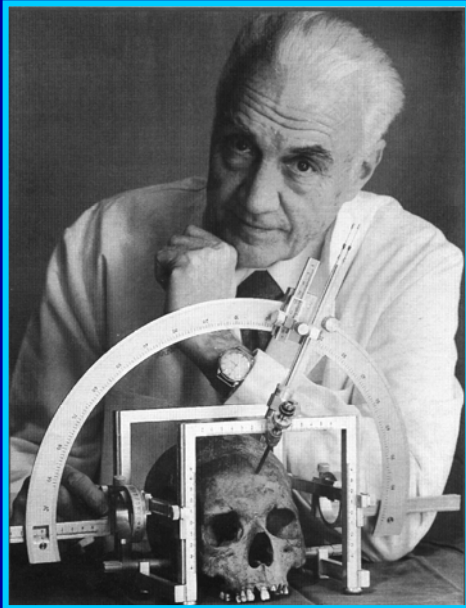
Meckel's Cave glycerol injection



- 201 procedures
 - 176 had immediate effect
 - 139 became pain free
 - 37 had reduction of pain
 - 24 had gradual change in pain (11 pain resolved while 15 had partial resolution)
 - 25 persistent pain (1 worse)
- 141 patients at 3 years
 - 71 are pain free
 - 42 controlled pain
 - 28 uncontrolled pain

Stereotactic Radiosurgery

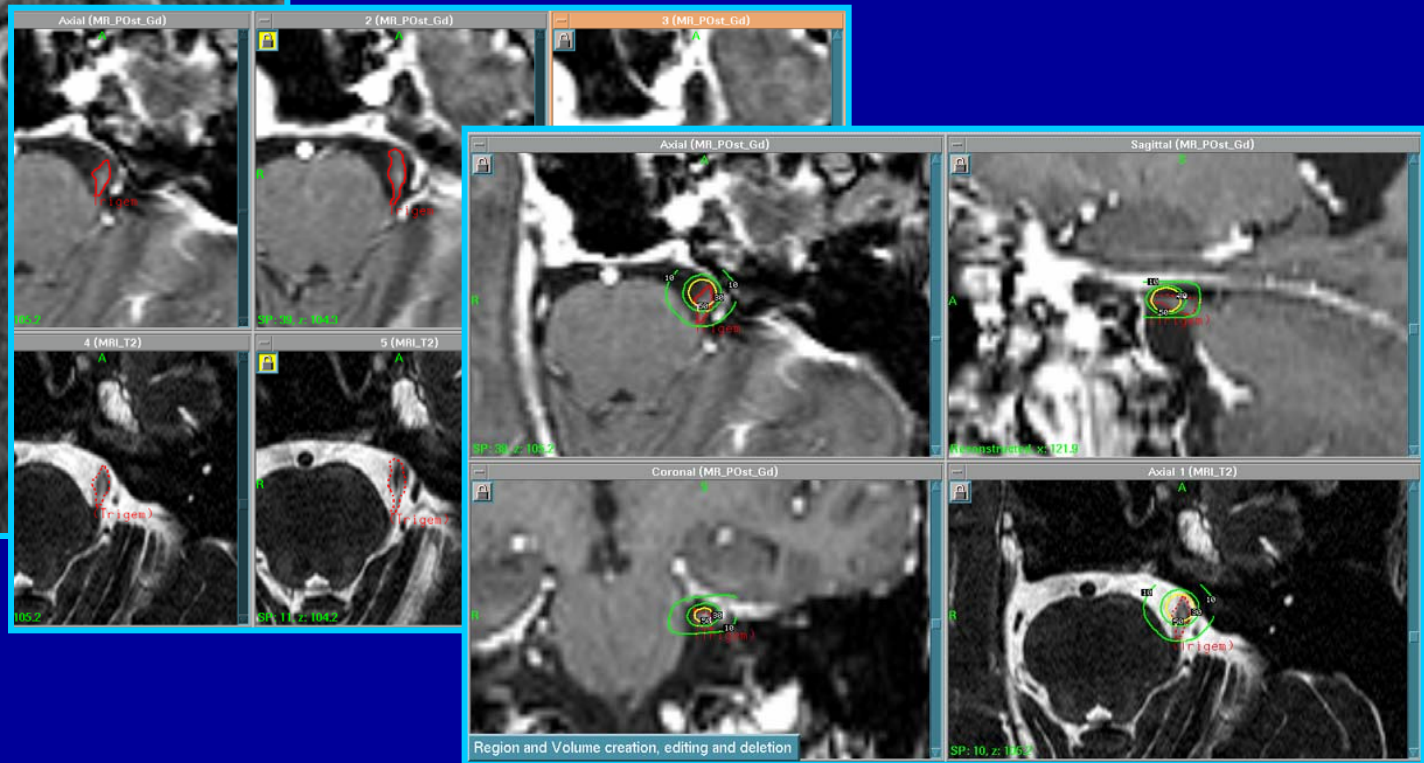
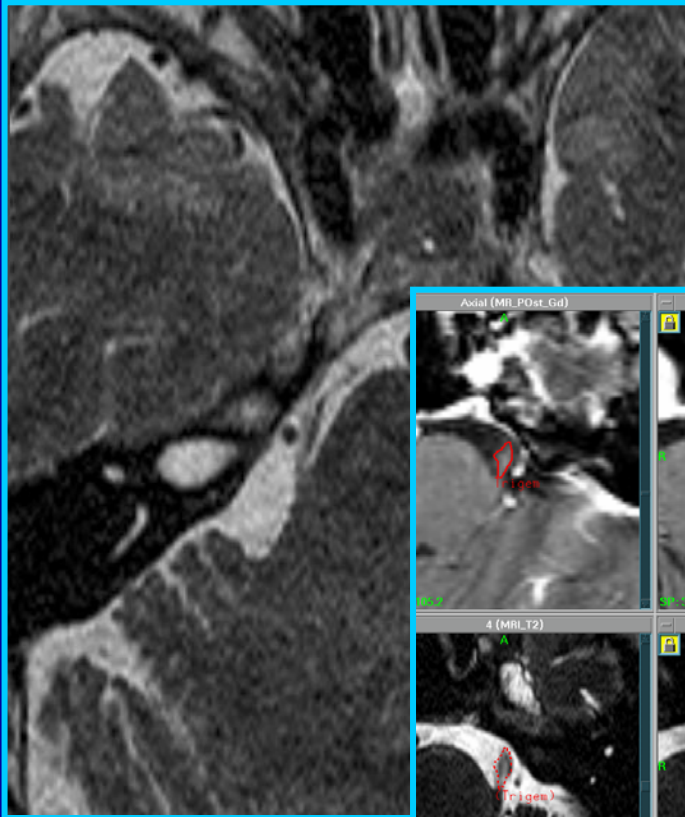
Lars Leksell



1953 irradiated two patients
*Stereotactic radiosurgery in
trigeminal neuralgia. Acta
Chir Scand 1971 : 137 ; 311 -
314*



Stereotactic Radiosurgery : Dose planning



Stereotactic Radiosurgery

Stereotactic radiosurgery for the treatment of trigeminal neuralgia

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

- Commenced 1992
- 220 patients treated : (16 patients (7.3%) had additional atypical features)
- Median radiosurgery dose was 80 Gy (60 - 90)
- 61.4% previous surgery
- Median follow up : 2 years
- Complete / partial pain relief in 85.6% at 1 year, 55.8% at 5 years
- Complete pain relief : 70.3% at 1 year
- Patients with atypical pain had lower rate of pain relief ($p = 0.025$)
- Patients without pre-treatment sensory loss, and those with no previous surgical treatment had better outcome
- 10% developed facial parasthesiae / numbness

Stereotactic Radiosurgery

Stereotactic radiosurgery for idiopathic trigeminal neuralgia

Pollock B, Phuong L, Gorman D, Foote R, Stafford S. J.

Neurosurg.2002;97;347 - 353

- 117 Patients treated, mean age 67.8 years.
- 58% previous surgical treatment
- Excellent outcome was defined as complete pain relief without medication
- Median follow up was 26/12 (range 1 - 48 months)
- At 1 year 57 % had pain relief :
- At 3 years 53% had pain relief :
- **No previous surgery**
 - At 1 and 3 years : 67 % pain relief
- **Previous surgery**
 - 1 year : 51%, 3 years 47% (Significant difference at $p = 0.04$)
- Trigeminal neuropathy (numbness / tingling) : 37% at dose of 90 Gy
 - Excellent outcome in patients with post Rt neuropathy : 76% at 1 year, 74% 3 years (46% at 1 year, 42 % 3 years in patients without Trigeminal dysfunction ($p < 0.01$)

Stereotactic Radiosurgery

Stereotactic radiosurgery for trigeminal neuralgia: a multi-institutional study using the gamma unit. Kondziolka et al. J. Neurosurg 1996 ; 84 : 940 - 945



- 4mm single isocenter targeted 2 -4mm anterior to junction of pons and trigeminal nerve
 - 30% isodose delivered to brain stem
- Dosage 70 - 80Gy.
- Median time to response : 4/52
- Median follow-up :18 months.
- Facial numbness : 10%

Endoscopic assisted MVD

Adjunctive Use of Endoscopy during Posterior Fossa Surgery to Treat Cranial Neuropathies; King, Wesley A., M.D.; Wackym, Phillip A., M.D.; Sen, Chandranath, M.D.; Meyer, Glenn A., M.D.; Shiau, John, M.D.; Deutsch, Harel, M.D. ; July 2001, Volume 49, Number 1; 108 - 116

Pernecky A, Fries G . Endoscopic assisted brain surgery. Neurosurgery 1998 ; 42 : 219 - 225

Jarrah R, Eby J, Cha S, Shahinian H. Fully endoscopic vascular decompression of the Trigeminal Nerve . Minim Invasive Neurosurg. 2002 ; 45: 32 - 35

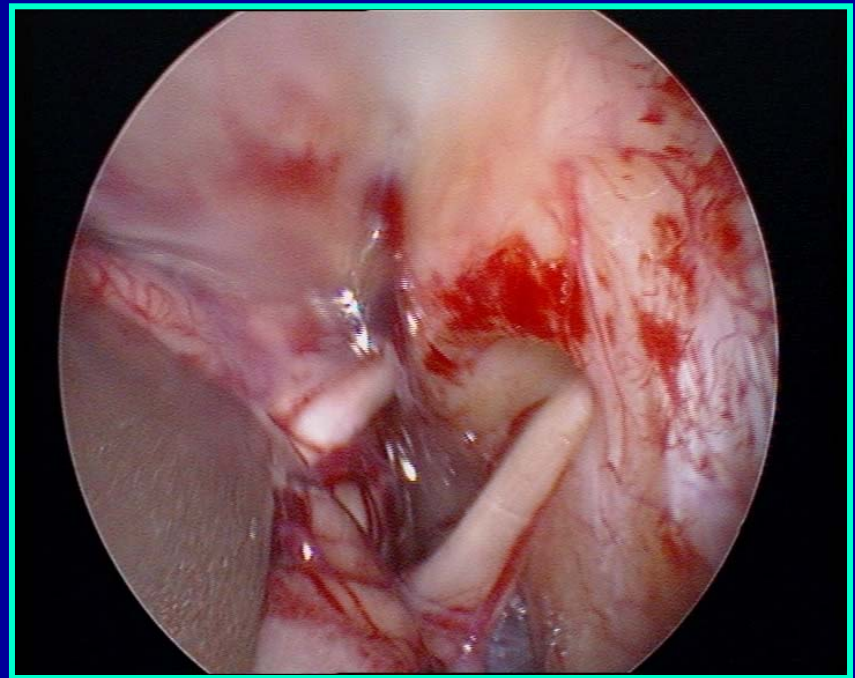
2.5 - 4.5 0° and 30° rigid endoscope

Advantages :

Smaller exposure

Less retraction minimising neurovascular injury

Improved results : Not yet demonstrated



THANK YOU

Any questions!