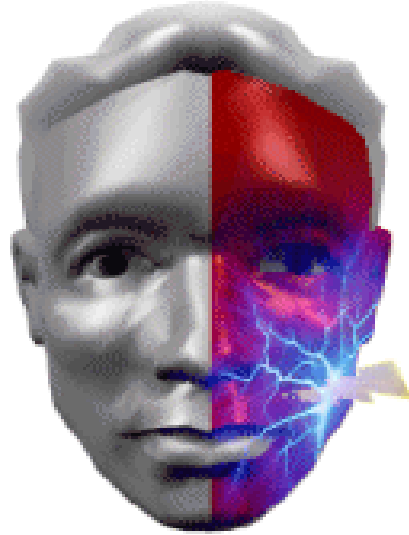


# *TRIGEMINAL NEURALGIA*



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# Demography

- Incidence: 4-5/100,000
- Also known as Fothergill's disease/Tic Douloureux
- Female predominance (m: f = 1:2 ~2:3)
- Mean age: 50 yrs.

# Why old age?

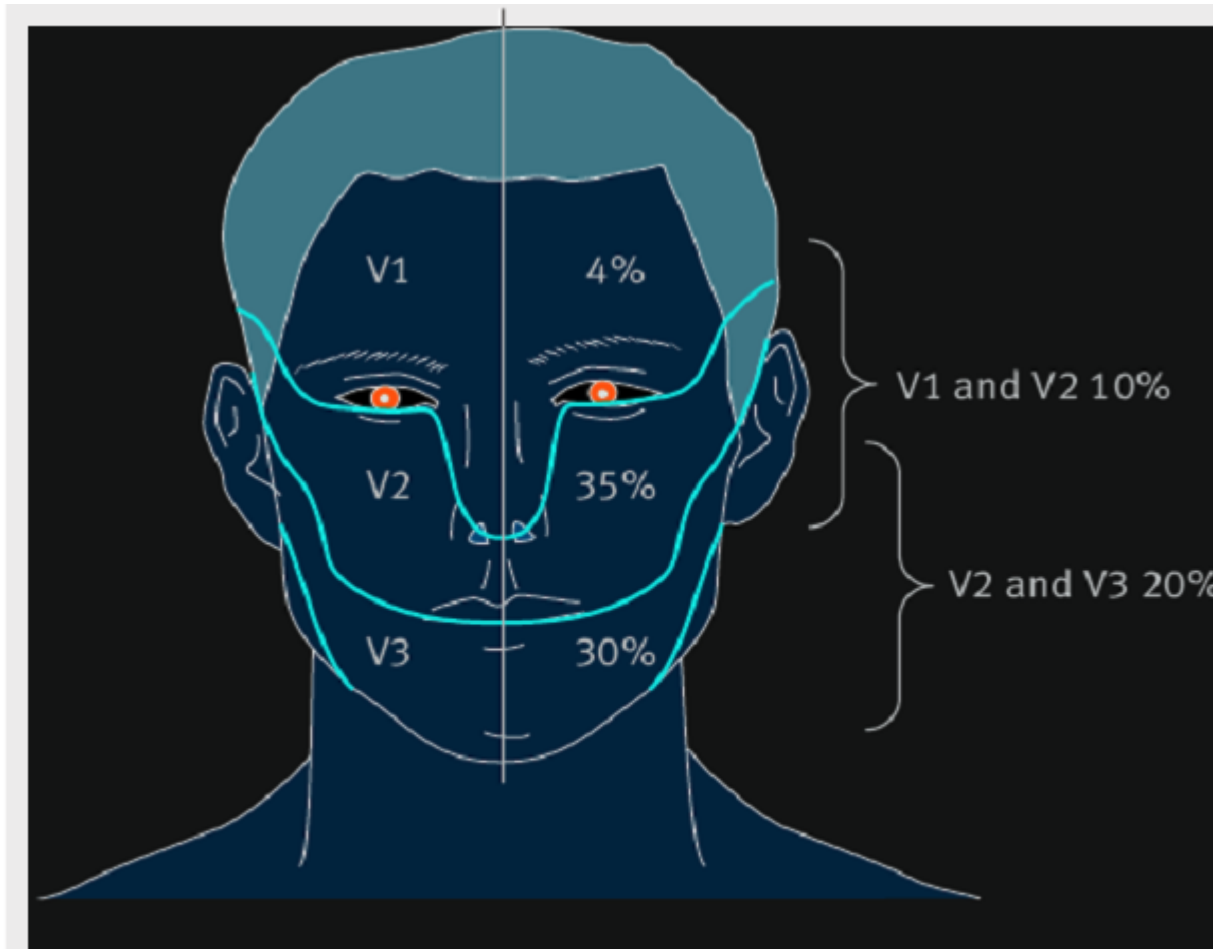
- Probably, with age  
the arteries elongate and become ectatic  
brain sags more within the skull

## Characteristics of pain

- paroxysms of severe, lancinating, electric shock-like bouts of pain restricted to the distribution of the trigeminal nerve
  - Unilaterally (right side)
  - The mandibular (V3) and/or maxillary (V2) branch or, rarely the ophthalmic (V1) branch
- Spontaneous attack or triggered by trigger zones
- Seconds to minutes



# Pain distribution



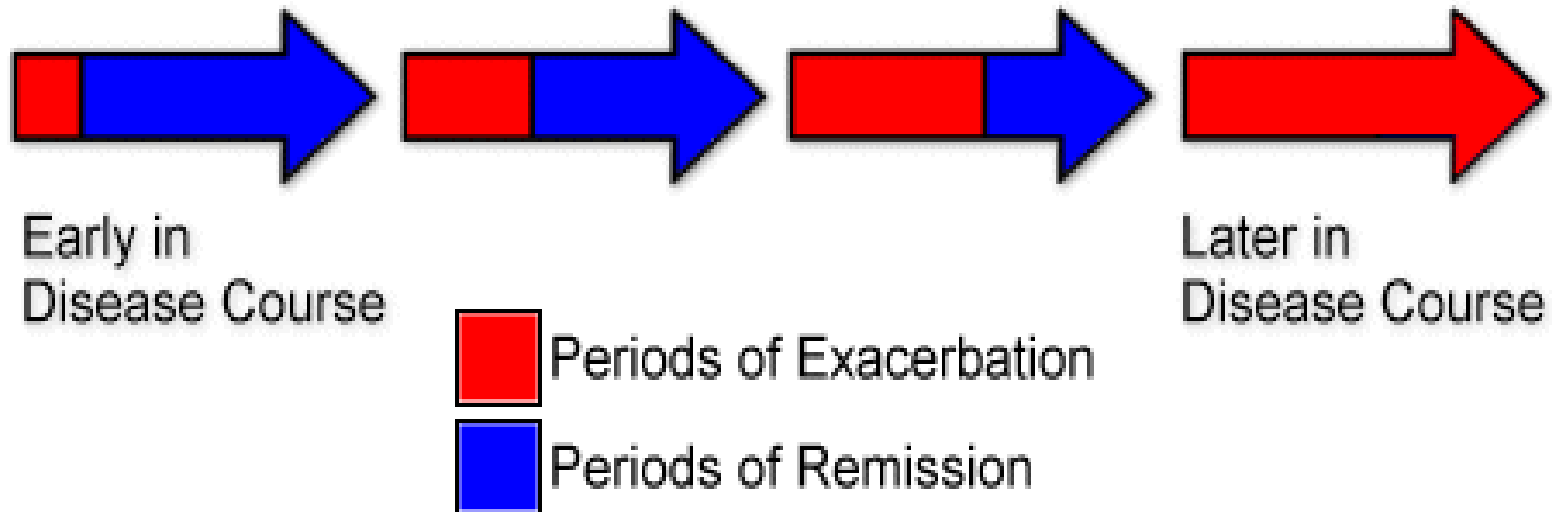
## Trigger zones

Trigger zones most commonly are located on the cheek, lip, nose or buccal mucosa.



- Triggers may be shaving, brushing teeth, drinking, eating or even slight breeze

## Progression of Trigeminal Neuralgia Over Time



## **Diagnostic criteria for classic trigeminal neuralgia (IHS)**

- Paroxysmal attacks of pain lasting from a fraction of a second to two minutes that affect one or more divisions of the trigeminal nerve
- Pain has at least one of the following characteristics
  - intense, sharp, superficial, or stabbing
  - precipitated from trigger areas or by trigger factors
- Attacks are similar in individual patients
- No neurological deficit is clinically evident
- Not attributed to another disorder



## Types of TN

- Classic (typical TN)
- Atypical TN
- Pre-TN
- Post traumatic TN (neuropathy)
- Failed TN

## Differential diagnosis

- Glossopharyngeal neuralgia
- Post herpetic neuralgia
- Sluder's sphenopalatine neuralgia
- Geniculate neuralgia
- TM joint pain
- Cluster headache
- Dental, orbital pain or sinusitis

## Etiology

- Idiopathic (vascular)
- Tumors (V nerve schwannoma, CPA tumors)
- Human herpes simplex virus
- Multiple Sclerosis

4% patients with MS have TN

2% patients with TN have MS

# Vascular

- Aberrant loop of artery or vein found to be compressing the root entry zone of the Vth nerve in 80-90% of patients at surgery
- The nerve is demyelinated next to the compressing vessel
- Eliminating the compression provides long term relief
- Compression by tumours or the demyelinating plaques of multiple sclerosis produce similar lesions of the root entry zone

## Vascular compression (Janetta et al 1996)

Vessel	Total(%)
SCA	909(75.5%)
AICA	116(9.6%)
PICA	8(0.7%)
VA	19(1.65)
BA	9(0.7%)
LA	3(0.2%)
Unnamed artery	186(15.4%)
Vein	822(68.2%)
Vein and artery	671(55.7%)
Vein only	151(12.5%)

# Why is trigeminal neuralgia paroxysmal?

- Compression of the REZ leads to demyelination and axonal damage
- Damaged axons become electrically hyperexcitable, exude neurotransmitters and potassium into the interstitial space, and crucially can repetitively fire at high frequency
- Demyelination leaves bare axons, often subserving different sensory modalities such as light touch and pain in contact with one another; this allows ephaptic transmission directly between them

# Why is trigeminal neuralgia paroxysmal?

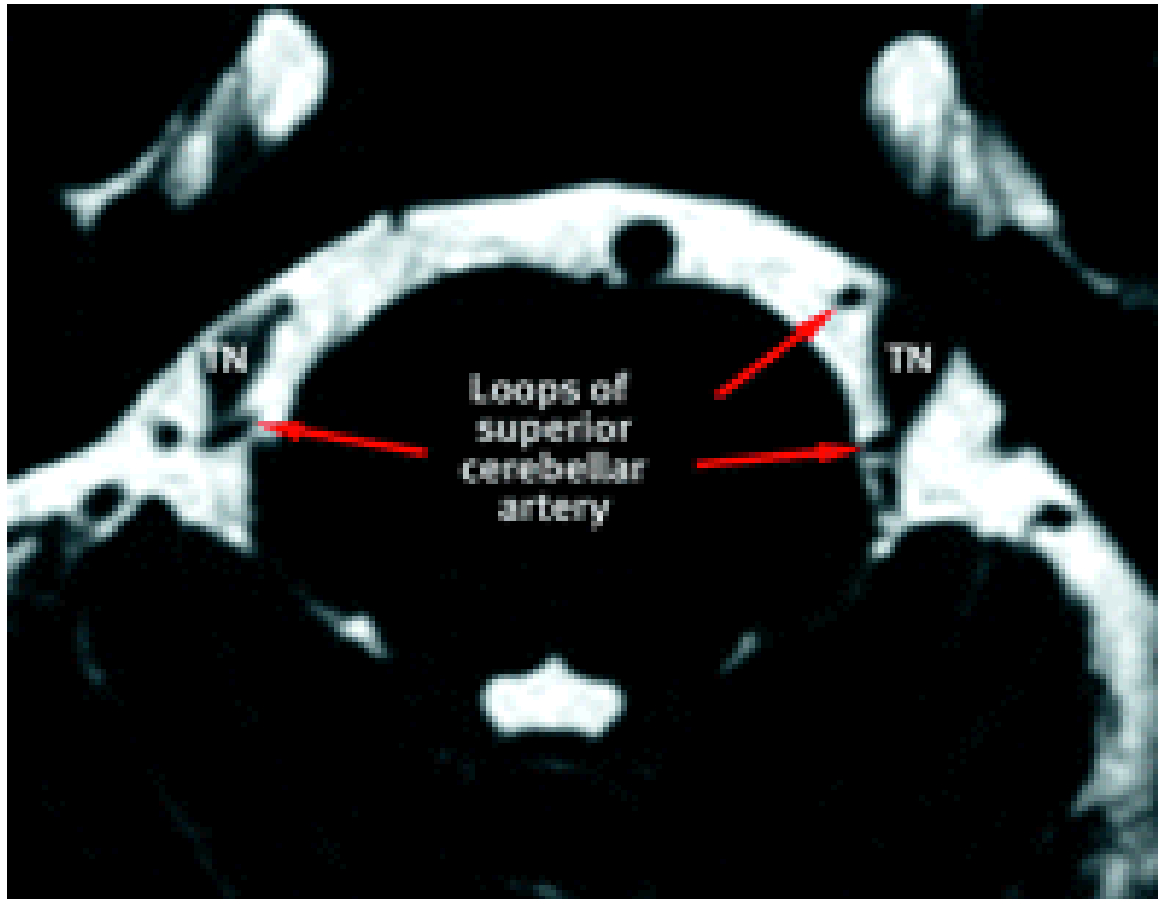
- A "spark"—often from light touch or cold in a trigger zone—"ignites" a self fuelling "inferno" of electrical discharge dependent upon all the above mechanisms. This rapidly burns itself out as axons become hyperpolarized, leading to the sudden and merciful cessation in pain and characteristic refractory period of a minute or so during which further pain cannot be triggered



# Investigations

- Whether there is an identifiable cause of the disease, particularly with a view to surgical cure
- Scan should be obtained in
  1. younger patients
  2. atypical clinical features, including sensory loss or a dull burning pain between paroxysms
  3. patients who do not respond to initial medical therapy.





- 3D CISS with MPR sequences and MRA
- 90.5% sensitivity and 100% specificity

# Management

- Medical treatment
  - Carbamazepine (Tegretol) – first line
  - Oxcarbazepine
  - Gabapentin (Neurontin)
  - Lamotrigine
  - Baclofen
  - Phenytoin
  - Clonazepam
  - Valproate
  - Mexiletine
  - Topiramate

# Carbamazepine (mazelol/tegretol)

- ▶ 1<sup>st</sup> line treatment (1961)
- ▶ Sodium-channel modulator - Antiepileptic drug
- ▶ Initial response is virtually universal
- ▶ By 10 yr it drops to 50%
- ▶ Lack of response   ???? diagnosis
- ▶ Start at 100 mg bid (200 mg/d)  
Add up to 200 mg/d in increments of 100 mg every 12 hr.
- ▶ Maximum dose 1200 mg/day.

# Carbamazepine (mazelol/tegretol)

## *Side effects*

- Dermatological: rash, Steven Johnson, EM
- Idiosyncratic(2-6%): **Aplastic anemia**
- Somnolence/dizziness/nausea/nystagmus/  
hepatic dysfunction/ hyponatremia
- Drug interactions

# Carbamazepine (mazelol/tegretol)

## *Monitoring treatment*

- CBC/LFT/RFT prior to start of treatment
- Every 2 weeks for 2 months
- Every 3 monthly thereafter
- Stop if TLC < 3000/microl

# Oxcarbazepine (oxetal)

- Pain relief similar to carbamazepine but fewer side effects or drug interactions
- Sodium-channel modulator, calcium-channel modulator.
- 300 mg bid. Maximum 2400 mg/day when given as monotherapy
- Dizziness, diplopia, ataxia, nausea, somnolence, headache, hyponatremia (more often than with carbamazepine)
- 300 mg of oxcarbazepine is equivalent to 200 mg of carbamazepine

# Baclofen

- Some people advocate it as 1<sup>st</sup> line
- 5-10 mg tid. Increase by 10 mg every other day until 60-80 mg daily
- Few drug interactions
- Ataxia, lethargy, GI intolerance
- Possibly synergistic with phenytoin/  
carbamazepine
- Benefit shown on randomized controlled trials

# Phenytoin (Dilantin/eptoin)

- ▶ Antiepileptic drug, sodium-channel modulator
- ▶ 100 mg po tid, or 300 mg once per day
- ▶ Comes in an intravenous form for acute exacerbation of pain
- ▶ Many drug interactions ; Ataxia, slurred speech, rash
- ▶ Monitor blood levels
- ▶ Response rate 25-60%



# Surgical treatment

## – *Gasserian ganglion-level procedures*

- Microvascular decompression (MVD)
- Ablative treatments
  - Radiofrequency thermocoagulation (RFT)
  - Glycerol rhizolysis (GR)
  - Balloon compression (BC)
  - Stereotactic radiosurgery (SRS)

## – *Peripheral procedures*

- Peripheral neurectomy
- Cryotherapy (cryoanalgesia)
- Alcohol block

# Radiofrequency thermocoagulation

- 1st explored by Kirchner, modified to produce more precise and safer lesions by Sweet (1974)
- Selective partial lesioning of the affected ganglion or retrogasserian root
- Intermittently anaesthetized patient under fluoroscopic control
- Needle is inserted through the foramen ovale into Meckel's cave using bony landmarks
- Once the needle has travelled the pre-planned distance, the patient is allowed to awake, the stylet replaced by the electrode and stimulation of the nerve root carried out.

# Radiofrequency thermocoagulation

- Once appropriate siting has taken place, the patient is anaesthetized again for thermal lesioning
- This is performed in cycles of 45 to 90 s at temperatures of 60–90°C
- After each lesioning the patient is awakened and manual sensory testing of the face carried out
- Additional thermal lesions performed until clear hypalgesia has ensued

# complications

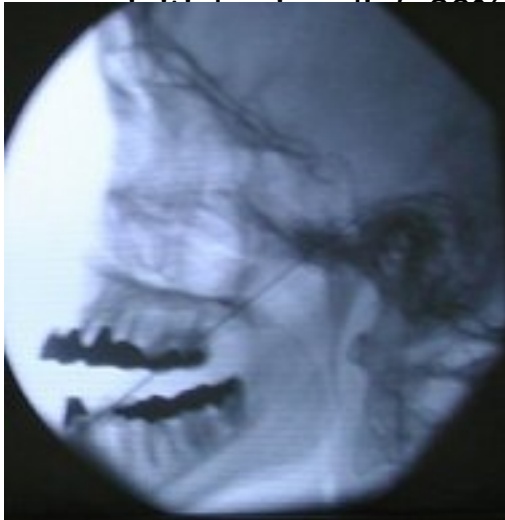
- dysesthesias (20%),
- masticatory muscle weakness (23%)
- absent corneal reflex(6%)
- keratitis (2%),
- anesthesia dolorosa(1%)
- meningitis (0.2%).

# Glycerol rhizolysis

- discovered serendipitously
- Leksell and Häkanson injected tantalum dust mixed with glycerol into the trigeminal cistern as a marker for radiosurgery
- patient cooperation is not necessary
- localization of the target is performed using intraoperative cisternography
- lower risk of facial sensory loss compared with either RFT or BC

Test dose: 0.1-0.15 ml  
0.05~0.1 ml at 3~5 min. intervals  
Total dose: 0.1~0.4 ml  
(average volume of the trigeminal cistern is 0.25 ml, and rarely >0.4ml)

Sensory changes: pain, burning or paresthesia  
recurrences 10-50% at 1 yr

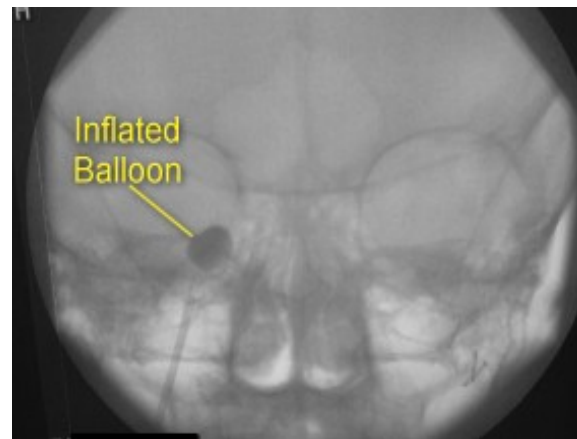
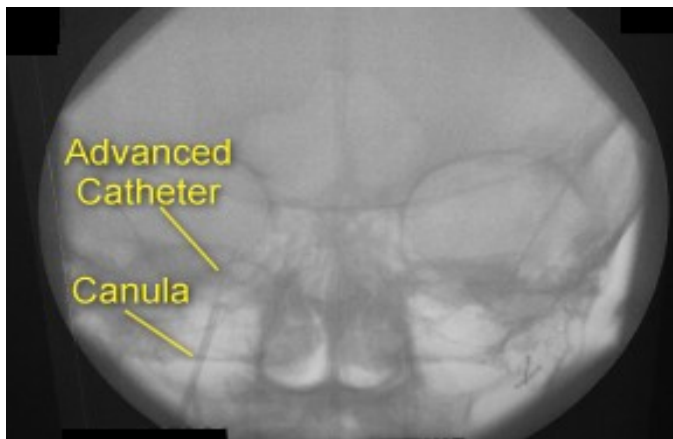


# Balloon compression

- Pear-shape balloon
- Compression time: 1~7 min.
- compresses the retrogasserian fibers against the firm edge of the dura and petrous ridge

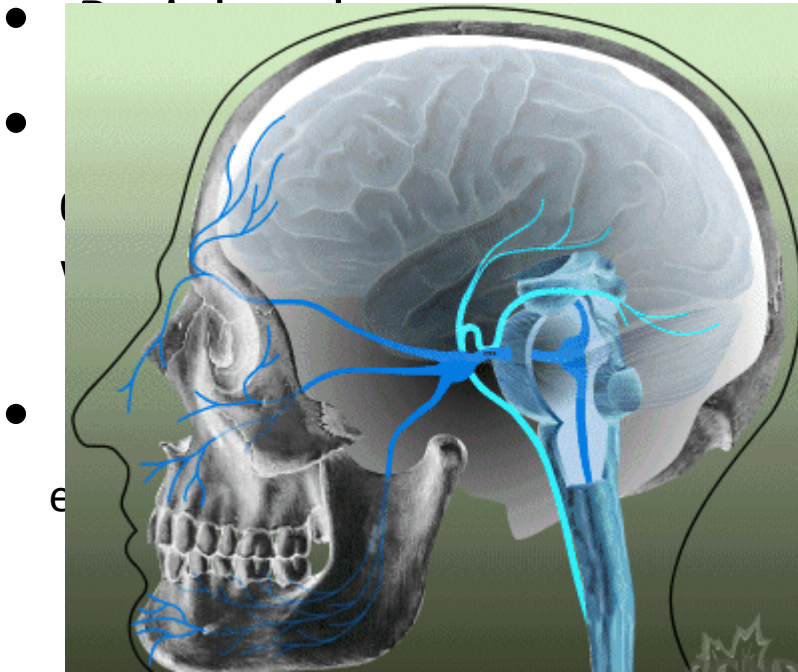
Immediate pain relief in almost all, 6–14% recurrence in the first year

dysesthesias and masseter weakness can occur





# Peripheral procedures



3 cycles of 2 min with a 5 min thaw  
in between

## Ganglion-level procedures vs. Peripheral procedures

- Ganglion-level ablative procedures
  - Similar long-term success rate
  - Varying degrees of sensory loss
  - Balloon compression least likely to impair corneal sensation or to cause anesthesia dolorosa
- Peripheral procedures
  - High recurrence rates
  - No benefit over ganglion-level procedures
  - Reserved for emergency use

# Microvascular decompression (MVD)

- Concept of Dandy(1920)
- Popularized by Janetta
- **Gold standard** as it deals the underlying cause
- Suboccipital craniotomy ,retraction of superolateral margin of cerebellum arachnoid is dissected and vessel freed, piece of shredded teflon felt placed between the vessel and the nerve to separate them.
- Veins, if any, coagulated and divided

## Microvascular decompression (MVD)

- Of 1185 patients treated by Barker et al with mean follow up of 6.5 years, 70% had excellent results at 10 yrs

(Barker FG II, Jannetta PJ, Bissonette DJ, et al: The long-term outcome of microvascular decompression for trigeminal neuralgia. *N Engl J Med* 334:1077–1083, 1996)

- Most recurrences within 1<sup>st</sup> two years.

# Recurrence???

- Female sex
- symptoms lasting >8 years
- venous compression of the trigeminal root entry zone
- lack of immediate postoperative cessation of pain
- Repeat posterior fossa exploration can be attempted

# Stereotactic radiosurgery

- Pioneered by Lars Leksell(1953)
- An ablative procedure
- **Target**- Gasserian Ganglion, Far anterior Target & REZ
- LINAC/GK/Cyberknife
- In GK, dose used is 70-90 Gy
- Mean time to pain relief is approximately one month
- Focal axonal degeneration and necrosis

# Outcome of GK Radiosurgery

Author (yr)	No. of pt.	max.dose(Gy)	Median f/u (m)	Pain free(%)
Kondziolka, '98	106	70-90	18m	60%
Nicol,2000	42	90	14m	74%
Maesawa,2001	220	60-90	22m	55%
Pollock,2002	117	70-90	26m	75%
Brisman,2004	293	75-77	23m	51%
Regis, 2006	100	70-90	NA	82%
Longhi ,2007	160	85	36m	61%

# Positive predictors

- Absence of MS
- Higher dose
- Primary radiosurgery
- Proximity of isocentre to brainstem



# Primary vs. secondary GK

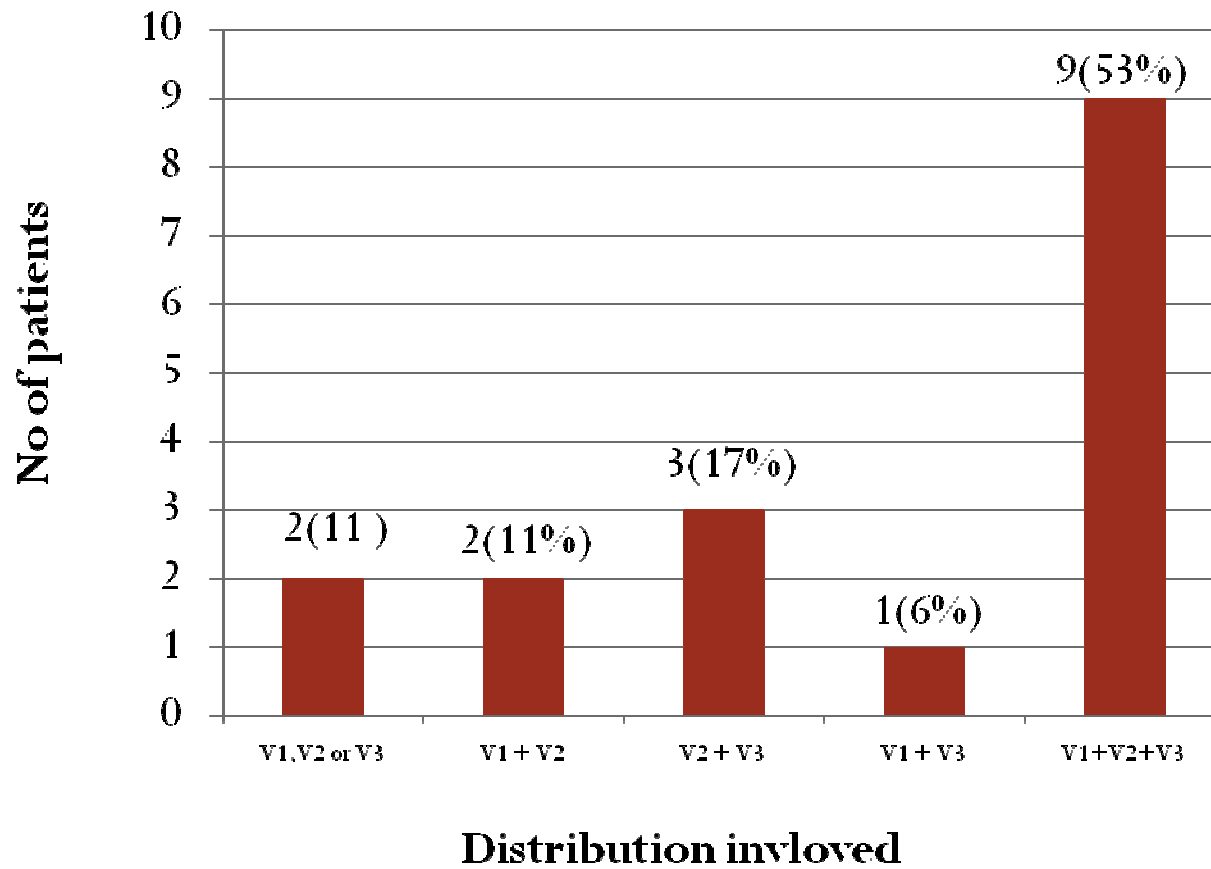
Study	Pt.	1 yr(%)	2yr(%)	3yr(%)
Pollock 2002	117			
	primary	67		67
	secondary	51		46
Fountas, 2006	77			
	primary	81	69	54
	secondary	64	44	12

# Complications

- Facial dysesthesias
- Anesthesia dolorosa (disabling painful facial numbness)
- Isolated reports of dysguesia, facial weakness, dry eye etc

# AIIMS experience

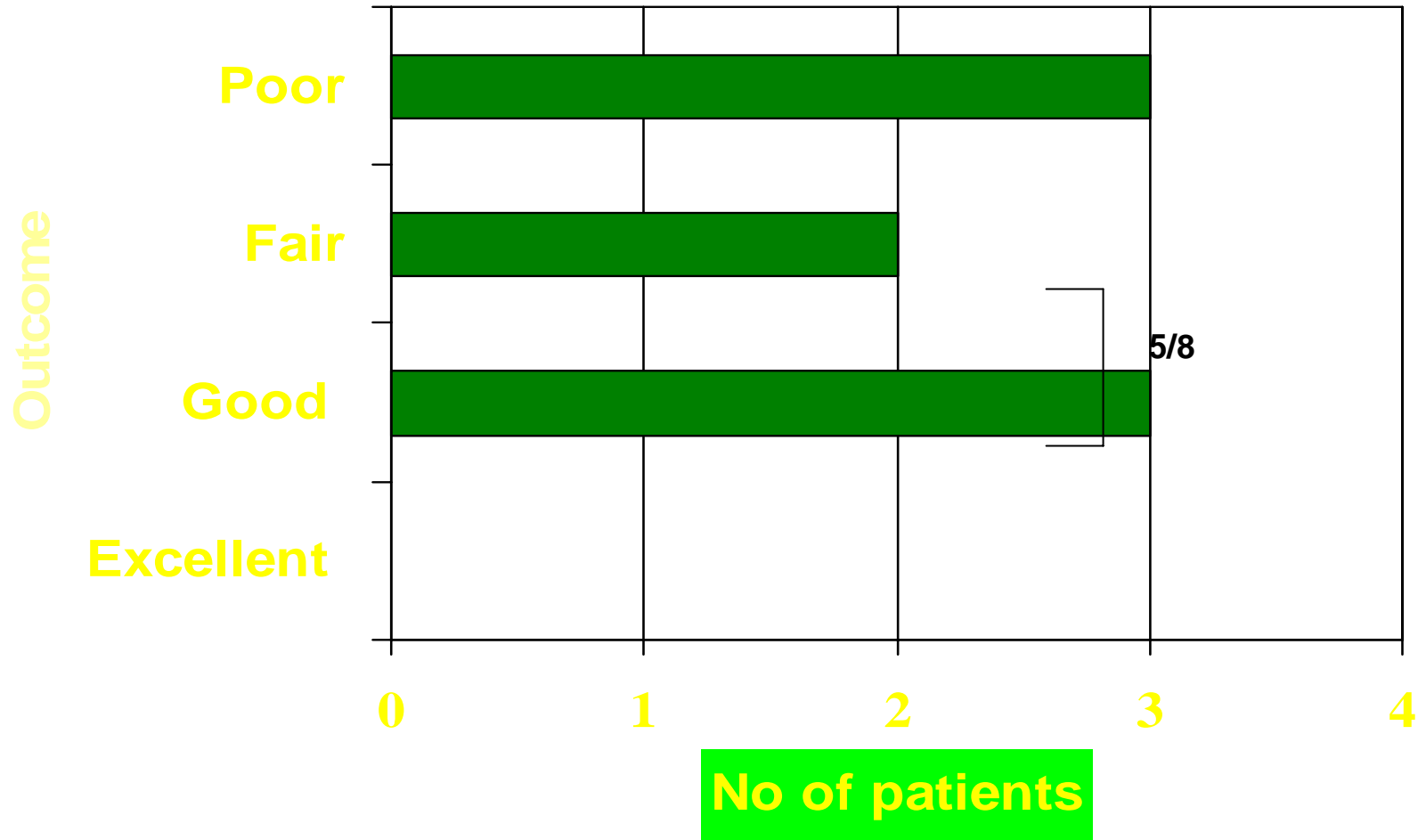
<b>No. of patients</b>	<b>17</b>
Male: female	10:7
Right: left	11:6
Age	32-78yr (mean 56yr)
Mean follow up	15months(3-48months)
Primary/secondary/repeat	6/11/1
Mean pain duration	9.85 yrs( 1-25 yr)



## radiosurgical procedure

- Single isocentre, median dose of 80 Gy (80–90 Gy ) to the 100 % isodose line using a 4 mm collimator
- Target: REZ
- no part of the brainstem received dose of > 12Gy

# Results



# Analysis of results

- None of our patients had excellent result
- 70-80% overall relief in various series
- Why we failed?
  - low dose/more secondary procedures
  - prolonged duration of symptoms/inadequate MR sequences
  - ?shorter Vth nerve in asians / ?resistance to radiation

*THANKS FOR ATTENTION*