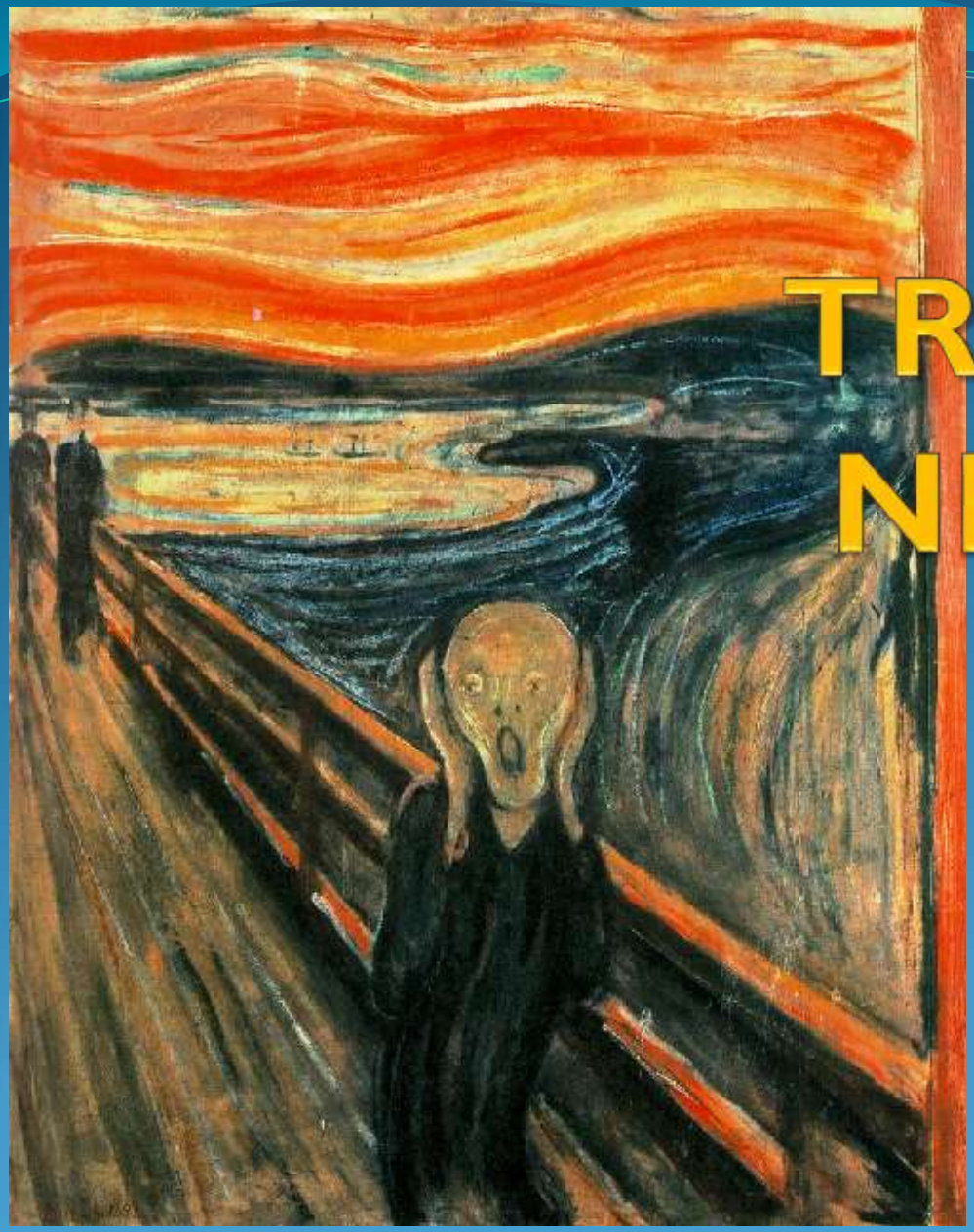
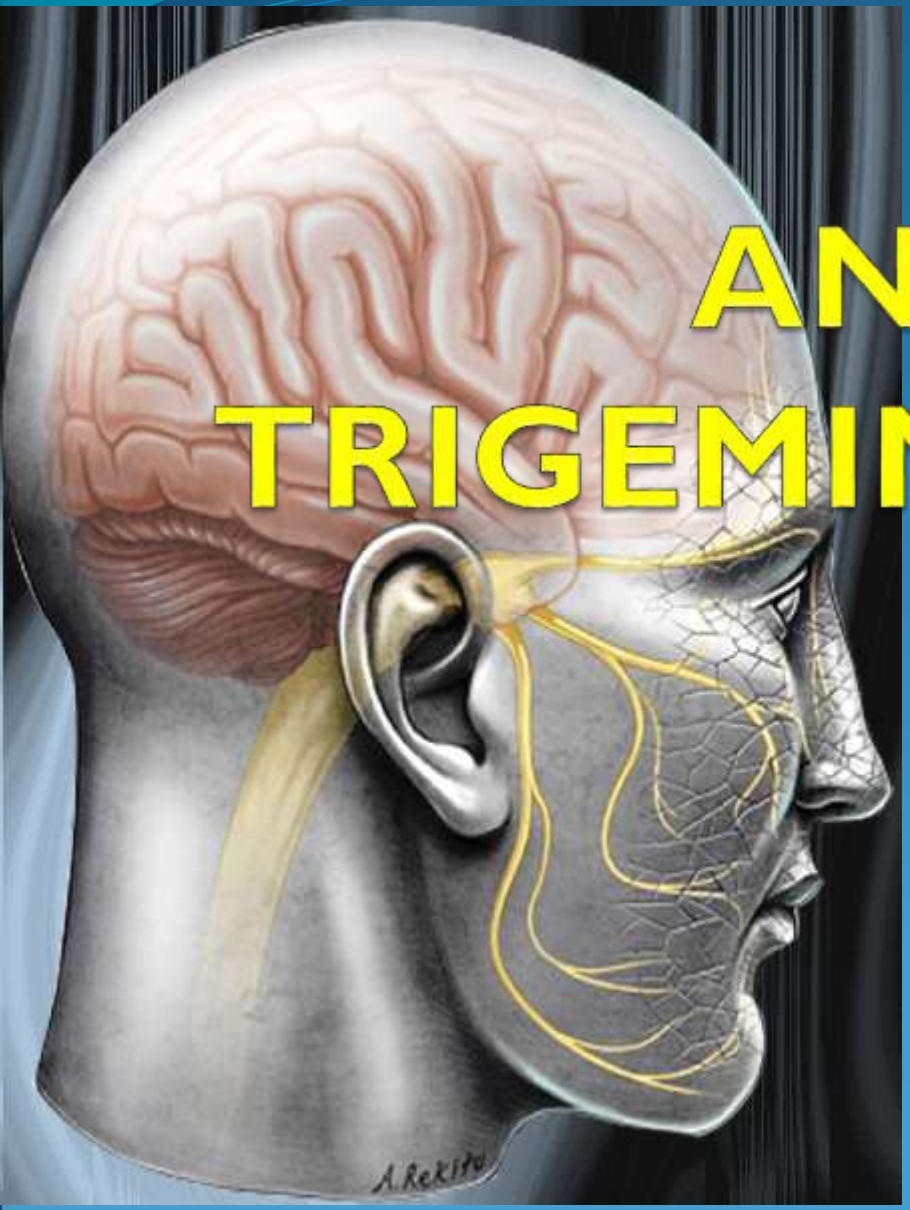


TRIGEMINAL NEURALGIA

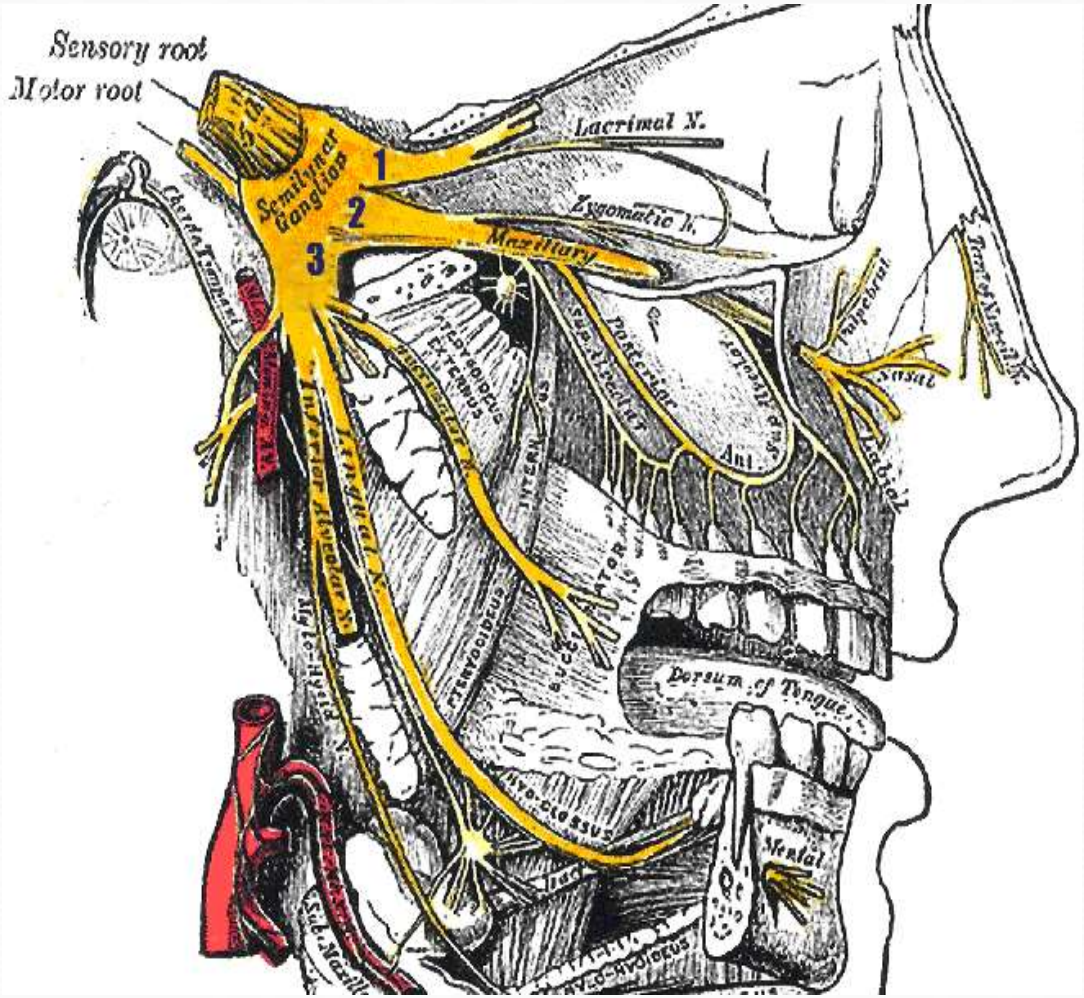
Orofacial pain pathways

- Information from the tissues outside the central nervous system (CNS) needs to be transferred into the CNS and on to the higher centers in the brainstem and cortex for interpretation and evaluation.
- If the information is the result of tissue damage or potential damage (noxious), then the impulses are said to be nociceptive.
- Once this nociceptive input reaches the higher centers for interpretation, it is perceived as pain.
- In the orofacial region, impulses carried by the trigeminal nerve enter directly into the brain stem in the region of the pons to synapse in the trigeminal spinal nucleus.

- The first neuron carrying information is called the primary afferent neuron or the first-order neuron. It receives the stimulus from the sensory receptor.
- The impulse is then carried by the second order neuron across the brain stem to the higher centers.
- There may be multiple interneurons (third order, fourth order etc.) that are involved in the transfer of this impulse to the higher centers.



ANATOMY OF TRIGEMINAL NERVE

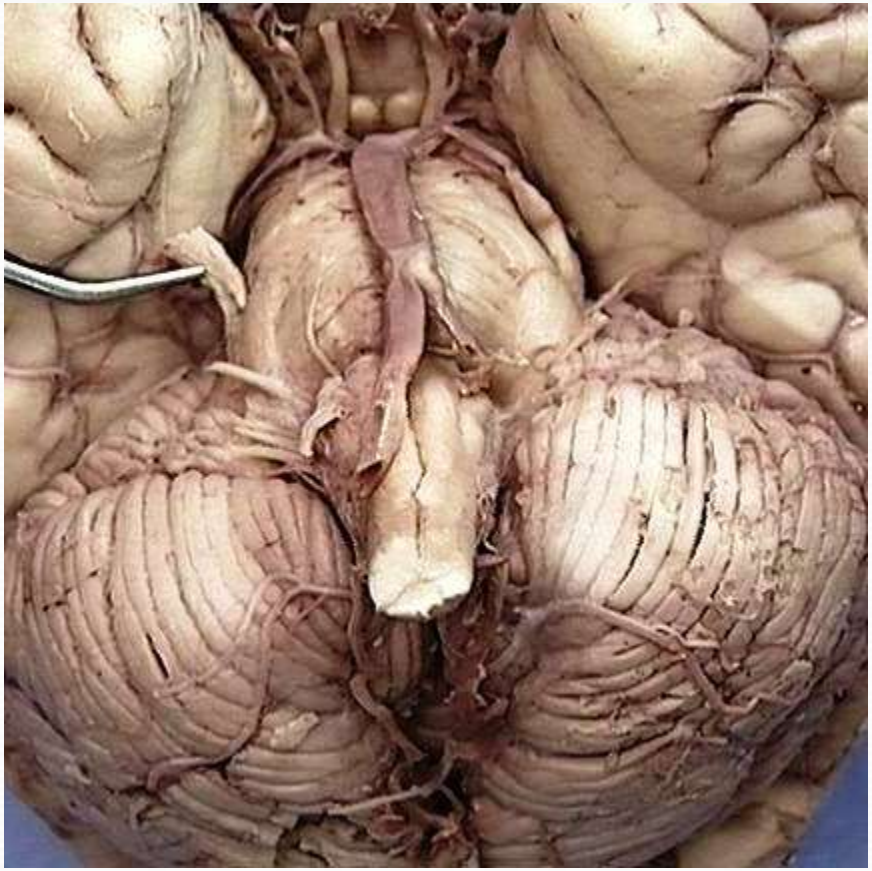


- **The trigeminal nerve with its 3 main divisions and their branches.**
- **1 – Ophthalmic division**
- **2 – Maxillary division**
- **3 – Mandibular division**

Intracranial component

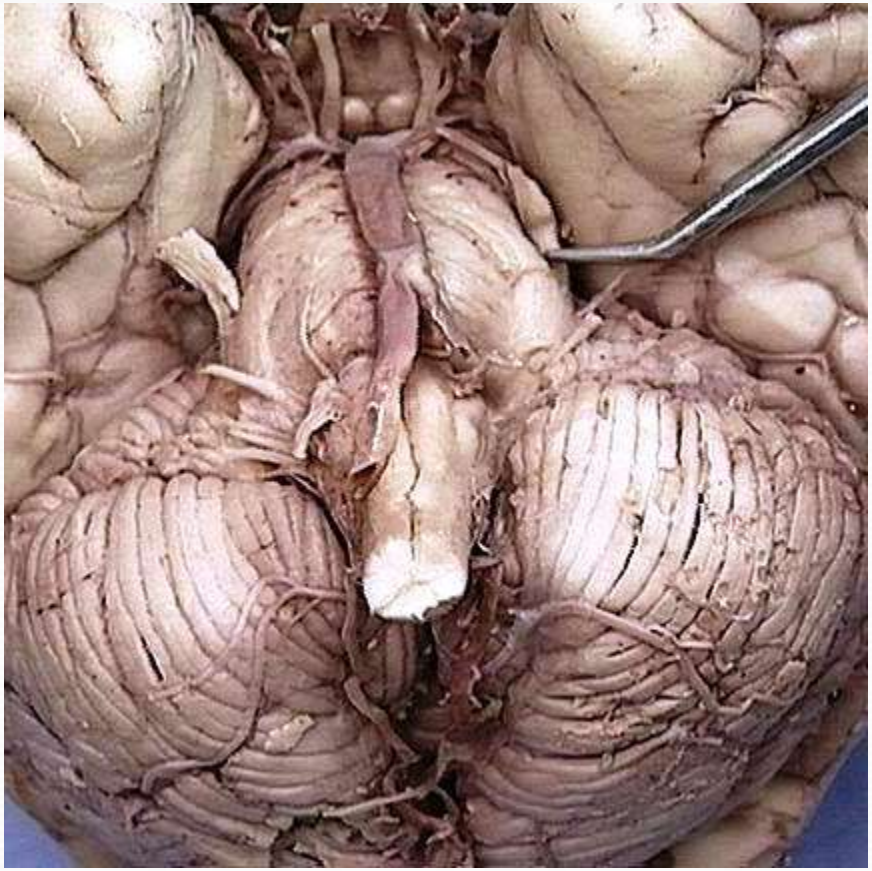
- Consists of the sensory root, the motor root and the 3 nerves upto their exit from the skull via their respective foramina.
- **Sensory root :** The trigeminal nerve emerges from the ventral surface of the trigeminal ganglion (semilunar ganglion) which occupies a recess (trigeminal or Meckel's cave) in the duramater covering the trigeminal impression near the apex of the petrous part of the temporal bone.
- The ganglion consists of unipolar neurons, which form the central and peripheral processes.
- The peripheral branches form the ophthalmic, maxillary and mandibular divisions of the nerve.

Sensory root of the trigeminal nerve

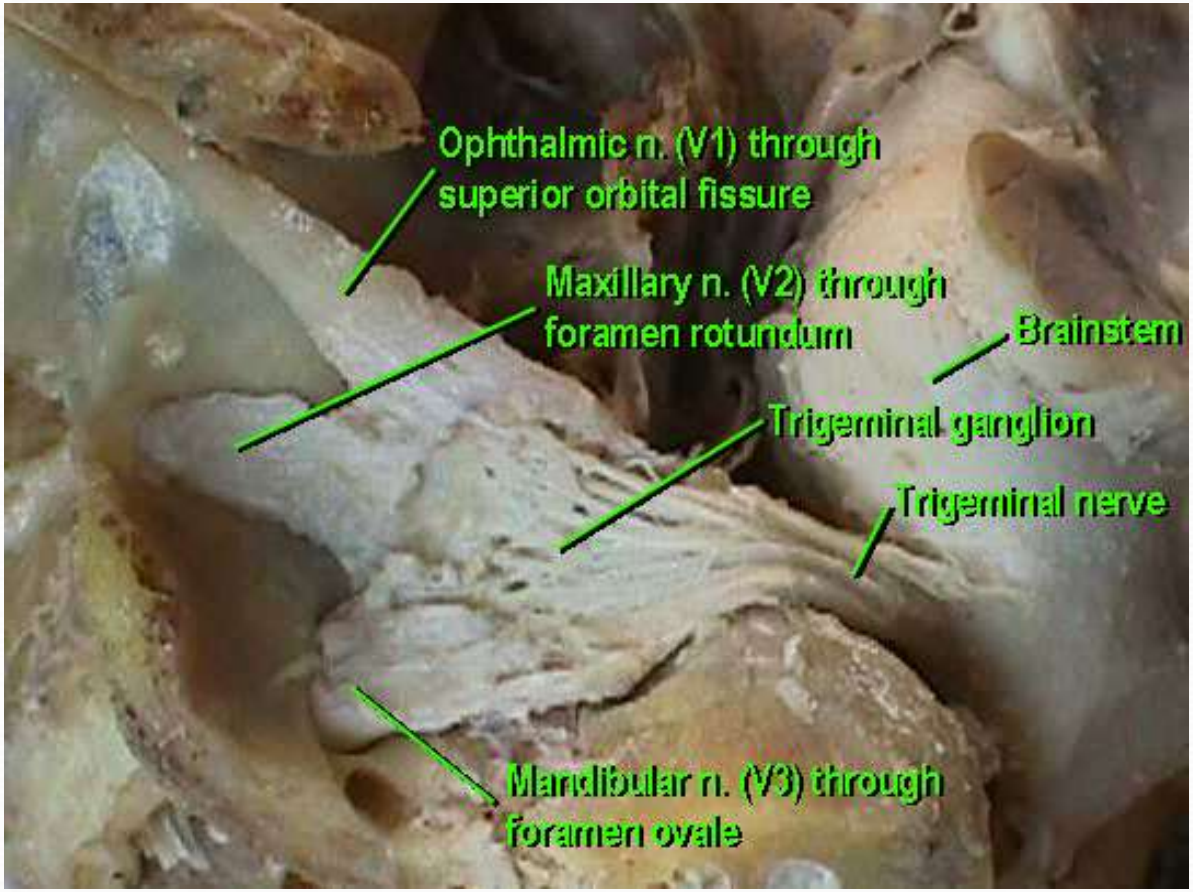


- **Motor root:** The motor root passes below to the foramen ovale, through which it passes to join the mandibular division just below the base of the skull.

Motor root of the trigeminal nerve



Three divisions of the trigeminal nerve



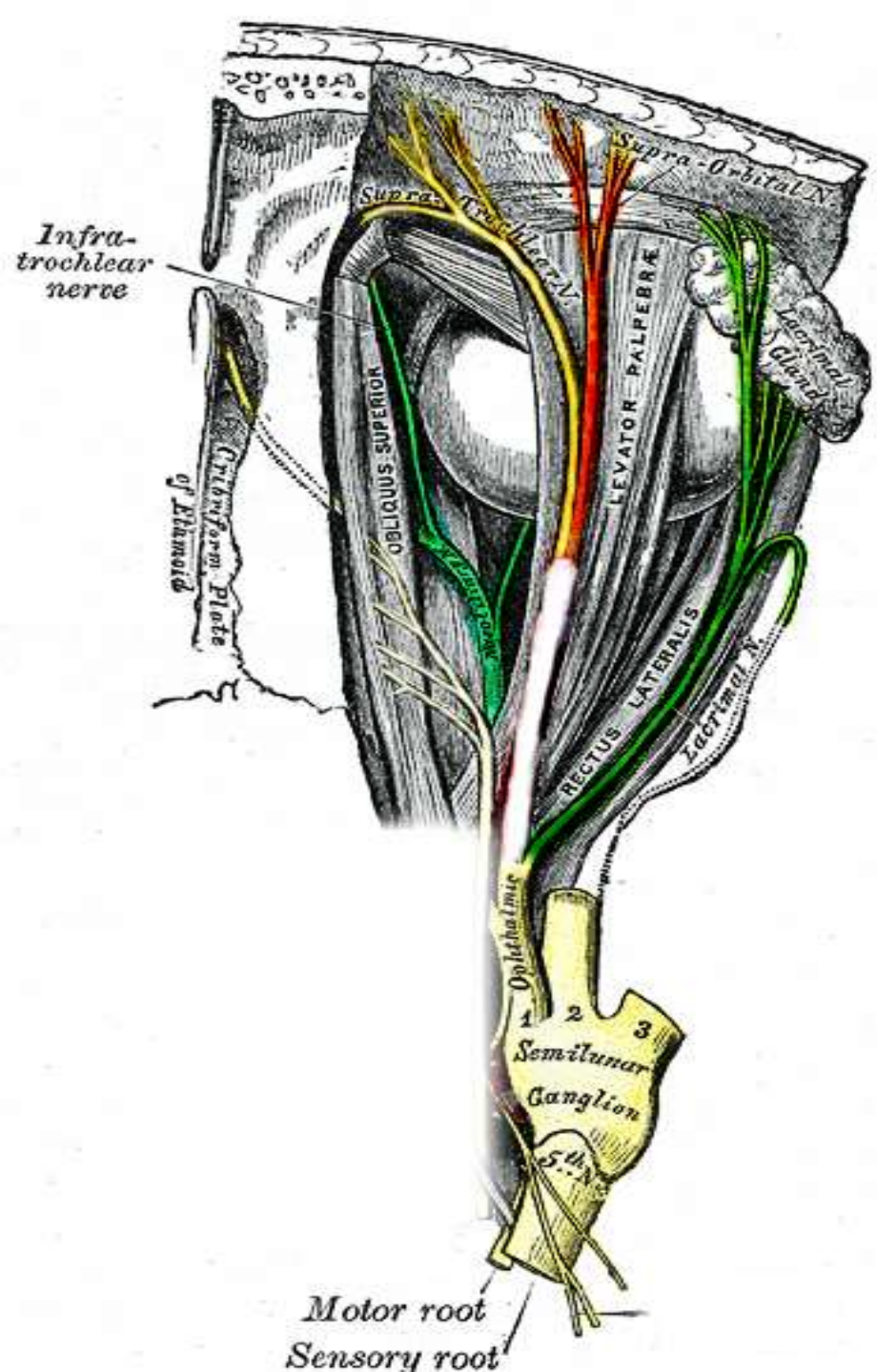


Diagram showing the ophthalmic division and its branches.

Frontal branch in white

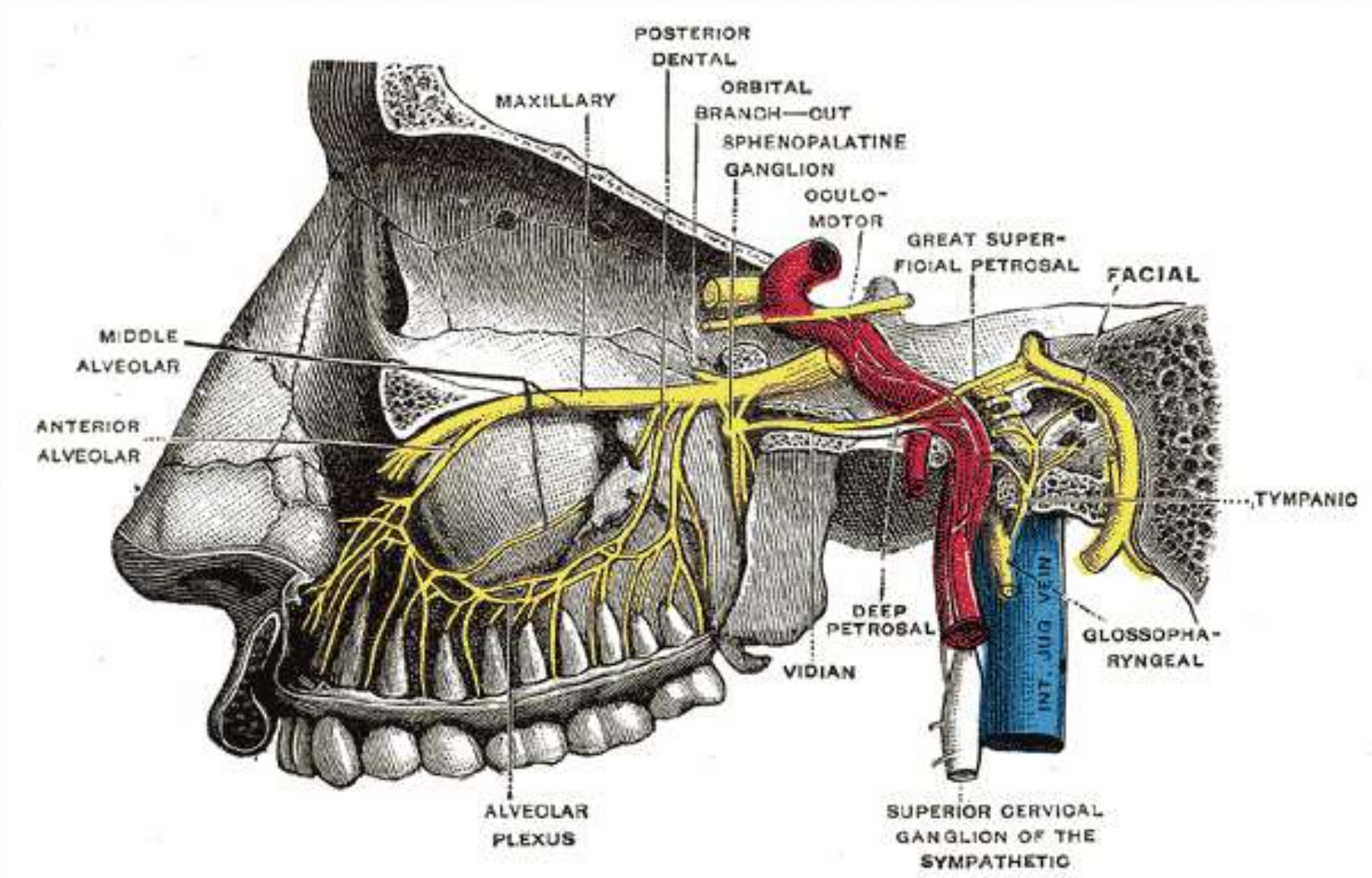
Lacrimal branch in dark green

Nasociliary branch in bluish green

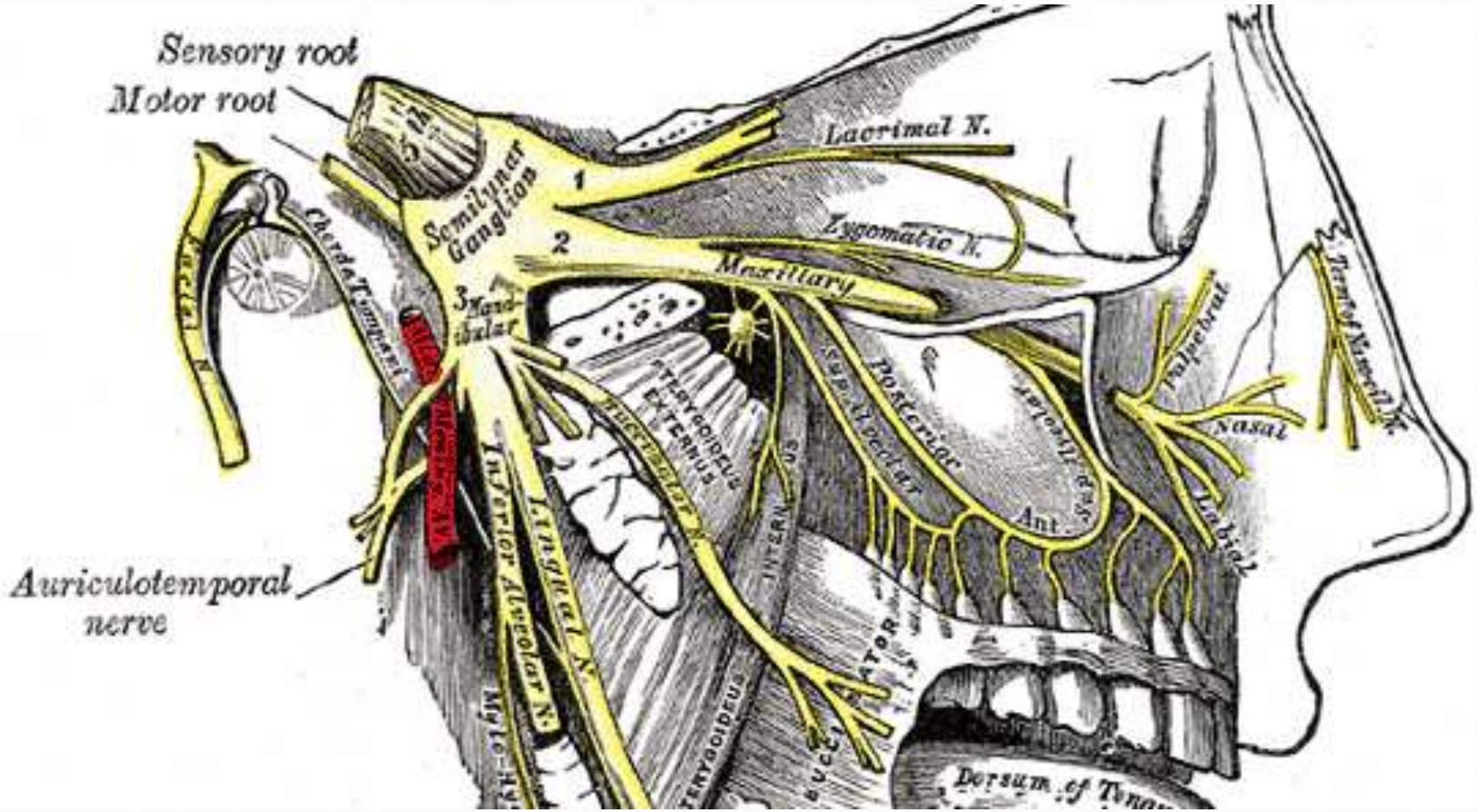
Supraorbital in red

Supratrochlear in yellow

Divisions of the maxillary nerve



Branches of mandibular nerve



Defintion

- The International Association for the Study of Pain (IASP) defines trigeminal neuralgia as ***sudden, usually unilateral, severe, brief, stabbing, and recurrent pains in the distribution of one or more branches of trigeminal nerve.***

- The International Headache Society (IHS) defines classical trigeminal neuralgia as a ***painful unilateral affliction of the face, characterized by brief electric shock-like pains, abrupt in onset and termination, limited to the distribution of one or more divisions of the trigeminal nerve. Pain is commonly evoked by trivial stimuli including washing, shaving, smoking, talking, and/or brushing the teeth (trigger factors), and frequently occurs spontaneously.***

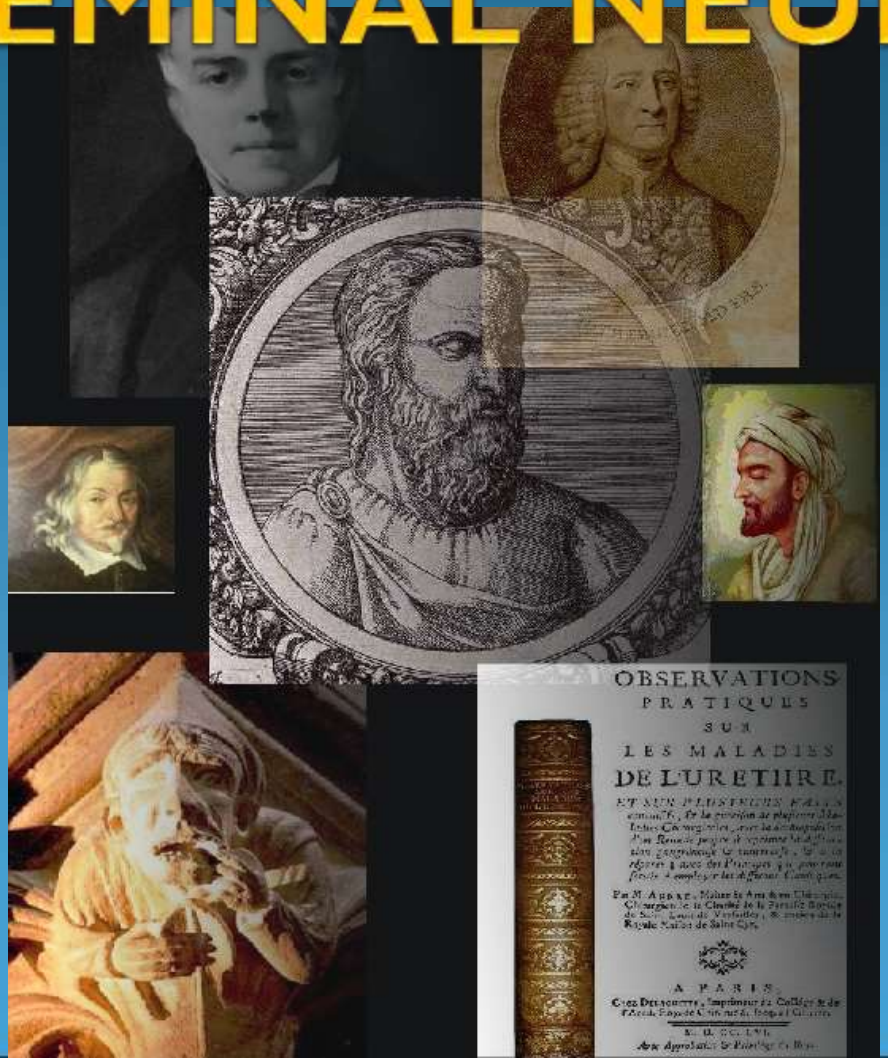
Classification

- From the etiological viewpoint, trigeminal neuralgia is classified into:
- 1) Primary or idiopathic trigeminal neuralgia

- 2) Secondary or symptomatic trigeminal neuralgia

- Primary or idiopathic trigeminal neuralgia does not have a clear cause.
- Secondary or symptomatic trigeminal neuralgia is due to manifest cause, such as tumor, multiple sclerosis or neurovascular compression.

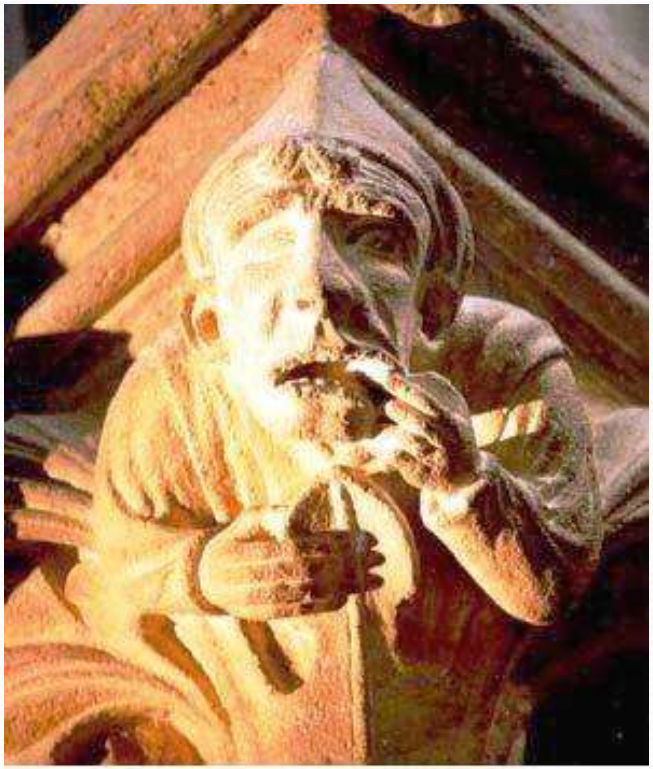
HISTORICAL PERSPECTIVE OF TRIGEMINAL NEURALGIA



- The most dreadful facial pain is that of trigeminal neuralgia which has defied the clinician for centuries.
- This horrible disorder of the trigeminal nerve has been reviewed in the medical literature for centuries.
- The first known description of trigeminal neuralgia, or a similar condition was written by Aretaeus of Cappadocia.

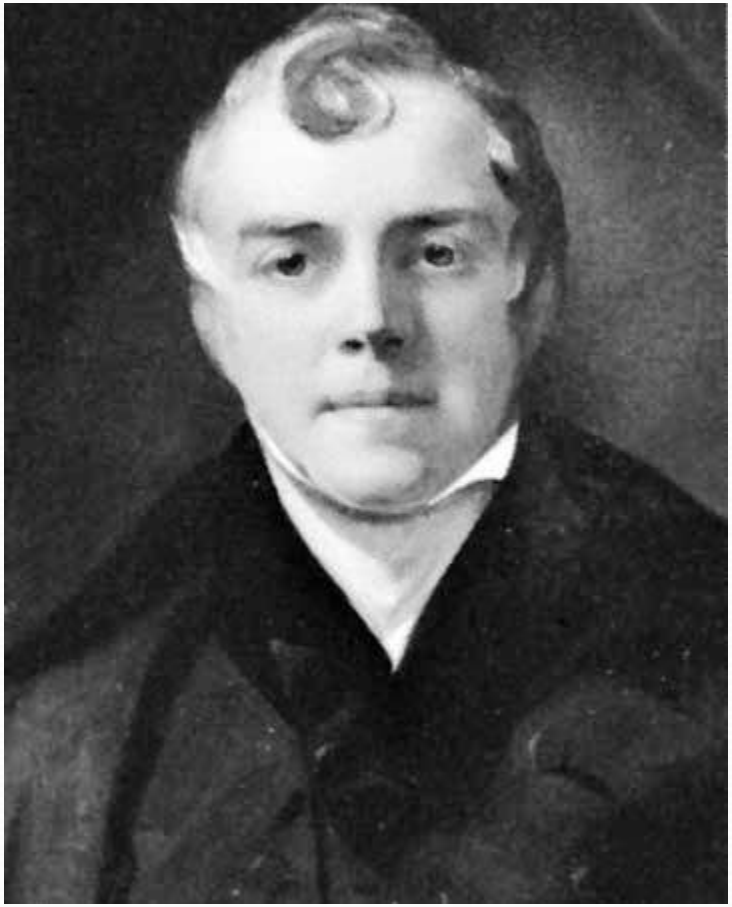
Aretaeus of Cappadocia

- As early as the sixteenth century, stone carvings in the Wells Cathedral depicted pain of trigeminal neuralgia.
- One is famed as toothache figure.
- Because of the surprising rarity of dental caries at that time, it pointed to a probable relevance to trigeminal neuralgia.



The tooth-ache man

- The first written description was reported by physician and sufferer Johannes Bausch in 1672.
- John Locke was a great physician and philosopher who gave an early detailed description of trigeminal neuralgia in 1677.
- The French physician Nicolas André recognized the unique nature of the syndrome, commenting that it was "exclusive and distinctive from all other diseases".
- He invented the term ***tic douloureux*** (literally, "unbearably painful twitch") in 1756, to signify painful affliction of the branches of the fifth nerve.



Sir Charles Bell

- In 1773, John Fothergill, an English physician, presented 14 cases of a painful affliction of the face.
- His description of trigeminal neuralgia has been considered an accurate and clear account.
- It was not until the 1820s, when Charles Bell established the separate functions of the trigeminal and facial nerves, that tic douloureux and Fothergill disease would be truly differentiated from other conditions.
- Bell's contribution enabled tic douloureux to be localized to the trigeminal nerve, which ultimately led to the evolution of the name of the disease to trigeminal neuralgia.

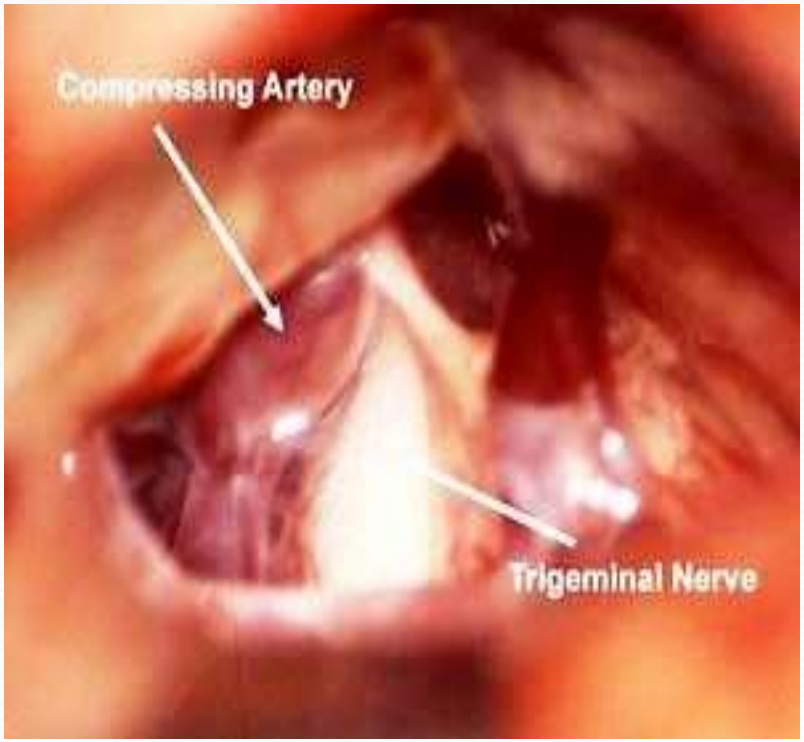
ETIOLOGY OF TRIGEMINAL NEURALGIA

- In the last three decades, however, evidence has been mounting that in a large proportion of cases, compression of trigeminal nerve root at or near the dorsal root entry zone by a blood vessel is a major causative or contributing factor.
- It is believed that compression of the sensory root is influential in the etiology of the disease.

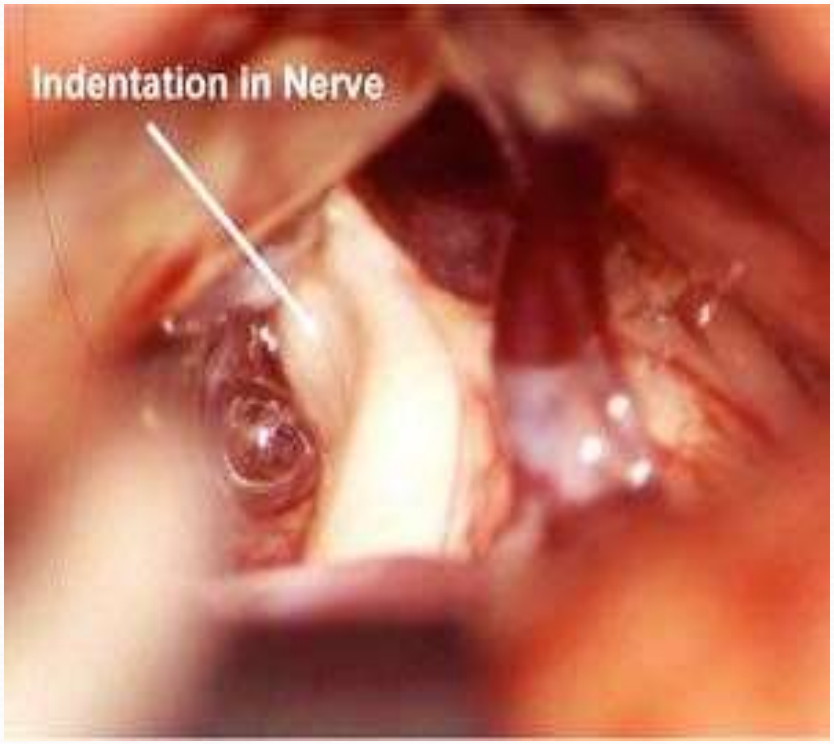
- There are several lines of evidence to support this view:
- Imaging methods like MRI and observations during posterior fossa surgery for trigeminal neuralgia have consistently shown a blood vessel in contact with the nerve root.

- Elimination of the compression leads to long term pain relief in most of the patients.

- The commonest cause of trigeminal neuralgia is focal compression of the trigeminal nerve root, close to its point of entry into the pons, by an aberrant loop of artery or vein.
- The resulting demyelination is postulated to alter the electrical activity of trigeminal neurons.



Trigeminal neuralgia occurs due to compression of the nerve root by an aberrant loop of artery.



Indentation in the nerve seen after retraction of the artery.

- Some interesting minor causes of trigeminal neuralgia include a primitive trigeminal artery or its variants and arteries transfixing the nerve.
- Some other causes of trigeminal neuralgia are known to be masses within the cerebellopontine angle (e.g. meningioma, epidermoid cyst, acoustic neuroma, aneurysm and pituitary adenoma).
- Patients with multiple sclerosis have an extremely high incidence of trigeminal neuralgia.

CLINICAL FEATURES

- Age of onset:
- The peak onset occurs between ages 50 to 70 years.
- The disorder can also occur in children.

- Epidemiology:
- Trigeminal neuralgia is seen commonly in females, with female-to-male ratio 3:2.

- Duration of pain:
- The paroxysms last only for a few seconds, but the actual bout may last for several hours, with varying intervals of freedom from pain between actual paroxysms.
- The pain is short lived and lasts for seconds to minutes.
- Often there is a transitory refractory period after the brief period of torture.
- In the refractory period, the patient may eat, drink or touch the trigger areas without producing the pain.
- Attacks rarely occur at night.

- Character of pain:
- Described by the victim as “lancinating”, “knifelike”, “stabbing”, “jabs with a red-hot poker” or “electric-shock-like” in nature.
- Patients have a characteristic clinical appearance and often immobilize their faces between attacks in the hope of preventing further episodes.
- The patient will have a motionless face – the ‘frozen face’ and will talk little for the fear of triggering the attack.
- The pain is confined to the trigeminal zone, nearly always unilaterally.
- Approximately 95% of the cases affect the second or the third division of the trigeminal nerve.

- Trigger zones:
- A trigger zone is an area of facial skin or mucosa where low intensity mechanical stimulation (such as light touch or air-puff) can elicit a typical trigeminal neuralgia pain attack.
- Characteristically, trigger zones occur around the supraorbital, infraorbital foramina, the inner canthus of the eye, lateral to the ala, and over the mental foramen.
- Small areas in the nasolabial fold and/or chin may be particularly susceptible to the precipitation of pain.
- Facial movements during eating, talking or whistling may initiate pain.
- Some patients demonstrate the "half-inch sign".
- When asked to indicate a trigger zone, they point to it, but stop their finger half an inch away!

DIAGNOSIS

- White and Sweet made a significant contribution by articulating precise and succinct diagnostic criteria for trigeminal neuralgia.
- The criteria emphasize 5 major clinical features that in essence, define the diagnosis of trigeminal neuralgia. These are:

- **1) The pain is paroxysmal (short sudden attacks of pain)**
- **2) The pain may be provoked by light touch to the face (trigger zones)**
- **3) The pain is confined to the trigeminal distribution**
- **4) The pain is unilateral**
- **5) The clinical sensory examination is normal**

DIAGNOSIS

- CT scan
- MRI
- Response to treatment with carbamazepine is almost universal in trigeminal neuralgia but unusual in other types of facial pain.
- Many clinicians therefore use this response as the final step in definitive diagnosis of the condition.

DIFFERENTIAL DIAGNOSIS

- 1) Multiple sclerosis: neuralgia due to multiple sclerosis is occasionally bilateral.
- 2) Post-herpetic neuralgia: this condition is easy to diagnose from a preceding history of facial herpes zoster.
- 3) Neoplasia: Usually careful examination will show other cranial nerve to be involved as well.
- 4) Inflammatory/infective conditions: Dental or temporomandibular joint problems can give rise to pain which is episodic, but its relationship to occlusion or jaw movements is usually obvious. dental infection or sinusitis causes constant throbbing or aching pain.

MEDICAL MANAGEMENT

- **PHENYTOIN:**
- MECHANISM OF ACTION
- Phenytoin has a stabilizing influence on neuronal membrane which prevents repetitive detonation of normal brain cells during "depolarization shift" that occurs in these patients.

- DOSAGE: Orally 300 to 600 mg. per day.

- ADVERSE EFFECTS
- ataxia, mental confusion, nystagmus, gastrointestinal disturbances or a skin rash, thrombocytopenia, aplastic anemia, gingival hyperplasia.

- **CARBAMAZEPINE:**
- MECHANISM OF ACTION: Like phenytoin.

- DOSAGE: An initial dose of 100 mg is given twice daily until pain relief is established. At no time should the daily dose exceed 1200 mg.
- COMBINED THERAPY: a combination of carbamazepine, 200 mg, and phenytoin, 100 mg, thrice daily; to be used if either drug alone be ineffective in an initial or subsequent course

- ADVERSE EFFECTS: drowsiness, dizziness and confusion, vertigo, nausea and vomiting. Hepatotoxic and hematological side effects like agranulocytosis, aplastic anemia, leucopenia or pancytopenia may develop.

- **BACLOFEN:**
- MECHANISM OF ACTION: primary site of action of is considered to be in the spinal cord where it depresses both polysynaptic and monosynaptic reflexes.

- DOSAGE: starting dose of 5-10 mg three times a day. The usual daily maintenance dose is 50-60 mg per day.

- ADVERSE EFFECTS: mental confusion, weakness and ataxia. Sudden withdrawal after chronic use may cause hallucinations, tachycardia and seizures.

- **GABAPENTIN:**
- MECHANISM OF ACTION: enhancement of GABA release in brain and modifies maximal electroshock to inhibit seizures.

- DOSAGE: It is usually effective in doses of 1200 mg to 3600 mg daily in 3 or 4 divided doses.

- ADVERSE EFFECTS: dizziness, ataxia, fatigue, nystagmus, and tremor. These effects are usually mild to moderate in severity, but resolve within two weeks of onset. Overall gabapentin is well tolerated.

- **VALPROIC ACID (SODIUM VALPROATE):**
- MECHANISM OF ACTION: Augmentation of release of GABA, phenytoin like frequency dependant prolongation of sodium channel inactivation.

- DOSAGE: Initially 600 mg per day is used; this dosage can be increased by 100 mg per week as needed for pain control.

- ADVERSE EFFECTS: These include anorexia, vomiting, drowsiness, ataxia, tremors, alopecia, and thrombocytopenia.

SURGICAL MANAGEMENT

- They may be divided into three groups: peripheral procedures, percutaneous procedures and open procedures.

- **PERIPHERAL PROCEDURES:**
- easy to perform and well tolerated with relatively few serious side effects.
- they have limited duration of action and patient must be advised that they may require repeated procedures.
- INCLUDES: anesthetic blocks of peripheral nerves in the region of the perceived pain, cryotherapy, alcohol injection, nerve avulsion.

LOCAL ANESTHETIC AGENTS

- Injection of a local anesthetic agent is often used as a diagnostic procedure for localization of the involved divisions.
- Anesthetic blocks are useful as a palliative procedure to achieve instant relief for a suffering patient
- Palliative blocks with long acting anesthetics such as 0.5% bupivacaine with epinephrine, may be given daily or at longer intervals to control trigeminal neuralgia.
- The pain is relieved for the duration of the anesthesia and the pain free period is short-lived.

ALCOHOL INJECTIONS

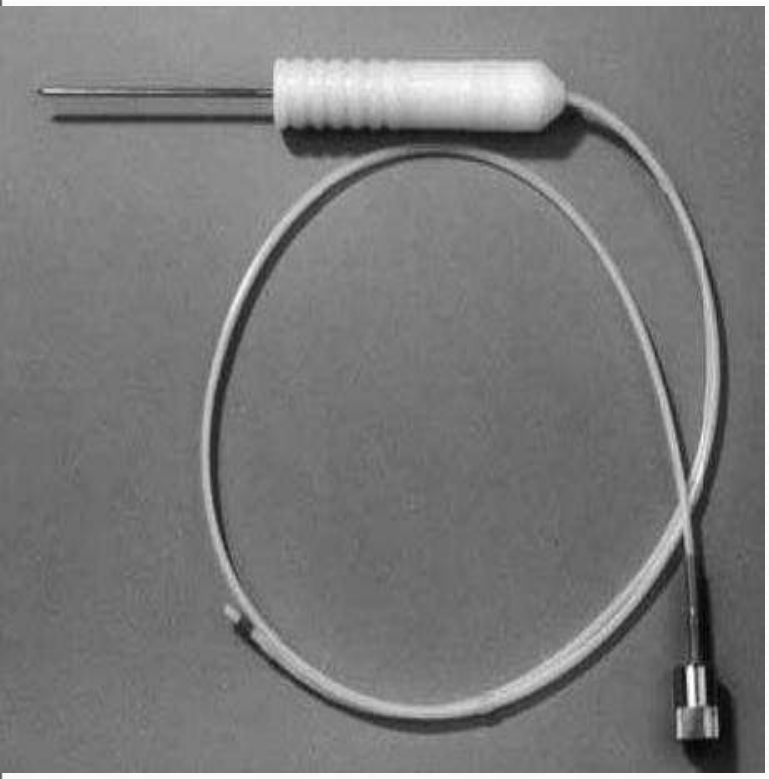
- When alcohol contacts the nerve, neurolysis occurs distal to the injection site.
- Two distinct techniques have been described for peripheral injection of alcohol.
- One involves the transcutaneous injection of the nerve at the skull base.
- The other in the region where the nerve passes recognizable landmarks in the facial skeleton (supraorbital notch, infraorbital foramen, inferior dental foramen or the mental foramen).
- Both techniques involve the injection of the relevant nerve with local anesthetic and once analgesia is confirmed, 0.5 to 1 ml of absolute alcohol is injected.

ALCOHOL INJECTIONS

- Alcohol is highly toxic, hence care must be taken to not inject excess alcohol subcutaneously and one must use an aspiration technique to avoid injecting into the accompanying vessels.

- The main disadvantages of peripheral alcohol injections are:[185]
- 1) Temporary sensory loss or paresthesia
- 2) Eventual recurrence of trigeminal neuralgia as the nerve regenerates
- 3) Temporary weakness of the muscles of mastication because of the close approximation of the motor root to the rest of the mandibular nerve.
- 4) Trismus
- 5) Mucosal ulceration

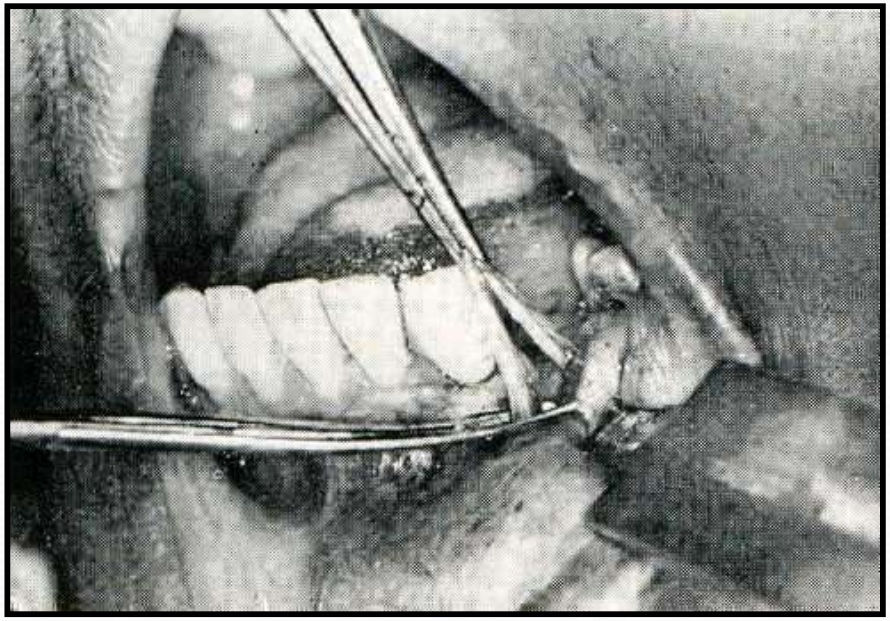
CRYOTHERAPY

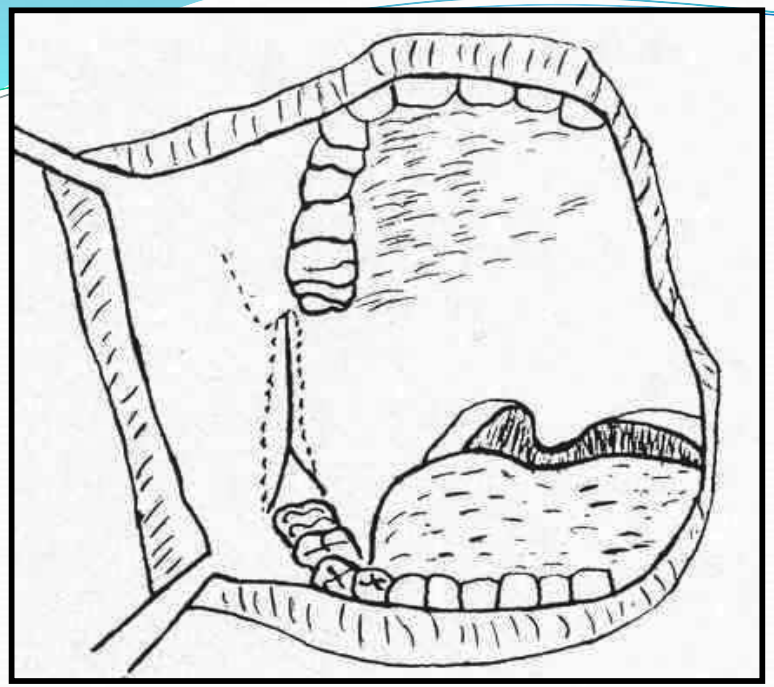


Cryoprobe

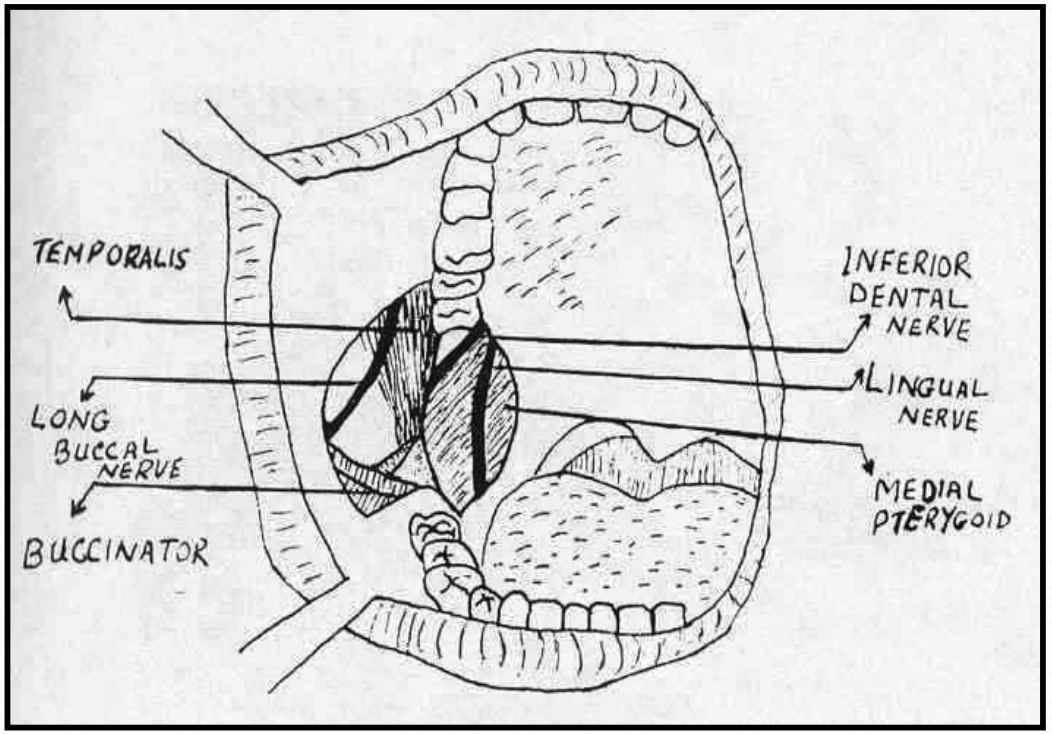
- It involves the use of a cryoprobe.
- The application of cold to tissues creates a conduction block, similar to the effect of local anesthetics.
- At 10^0C, larger myelinated fibers stop conducting, but all nerve fibers stop conducting at -20^0C.
- Long-term pain relief from nerve freezing occurs because ice crystals create vascular damage to the vasa nervosum which produces severe endoneural edema.
- In cryotherapy, a peripheral branch of the three major divisions of trigeminal nerve is exposed and frozen by direct application of a cryoprobe with a tip temperature ranging from -50 to -70^0C.
- The patient requires sedation or general anaesthesia.

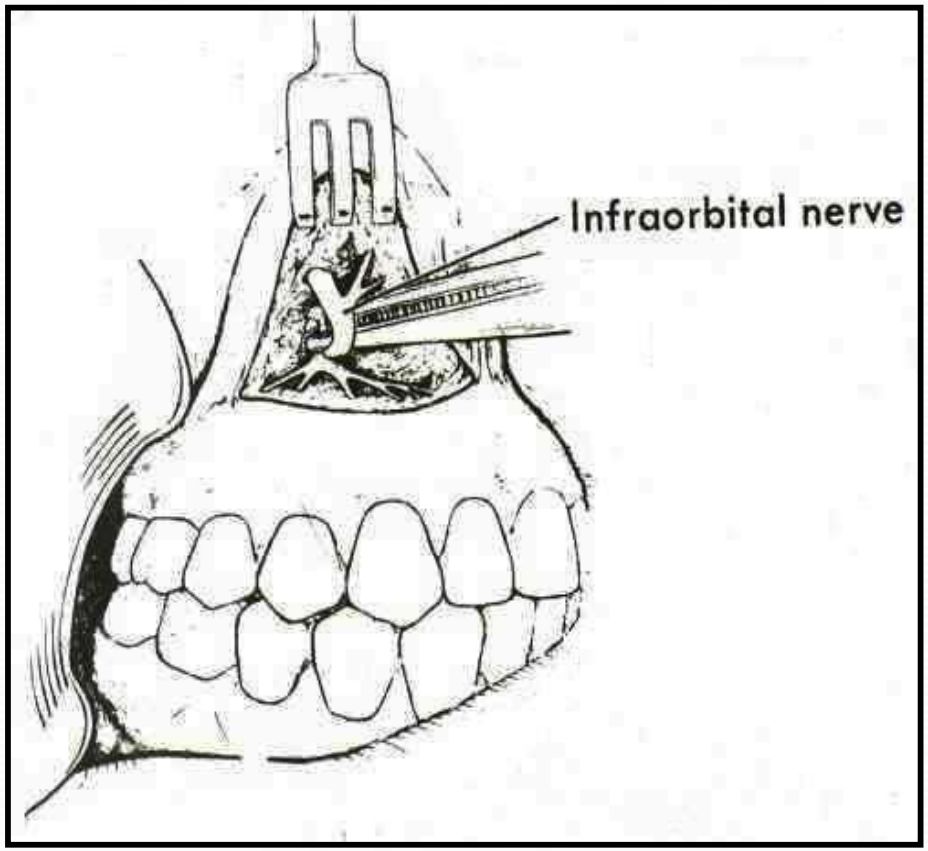
Peripheral Neurectomy



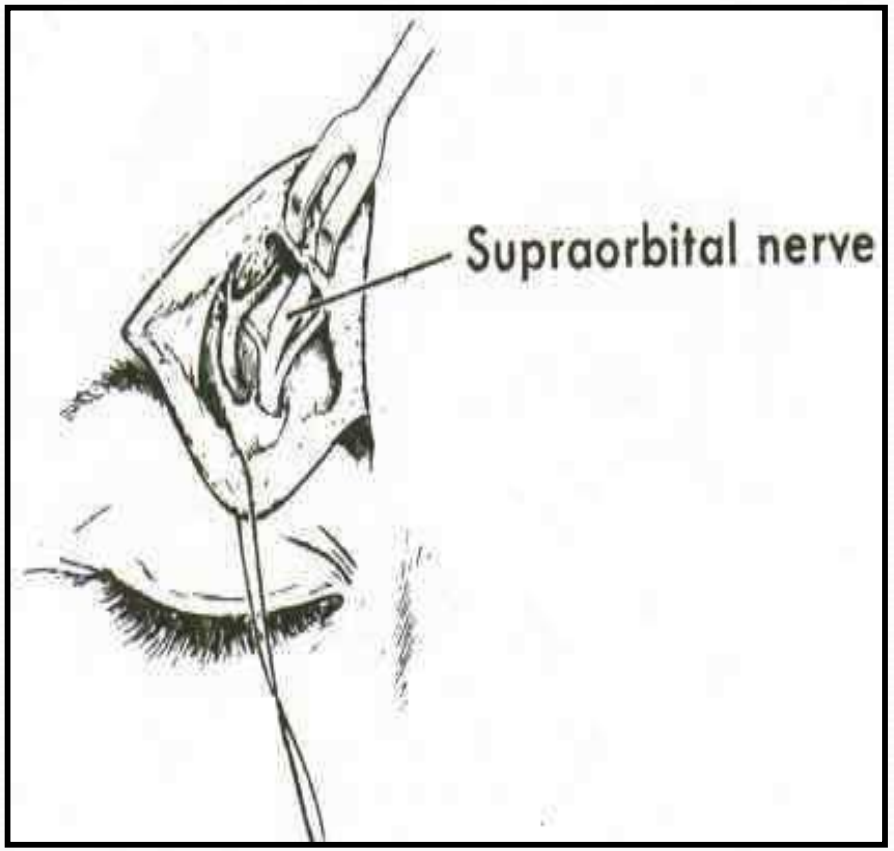


Inverted “Y” shaped incision





Exposure and avulsion of the infraorbital nerve by intraoral incision



Exposure of the supraorbital nerve

RADIOFREQUENCY RHIZOLYSIS

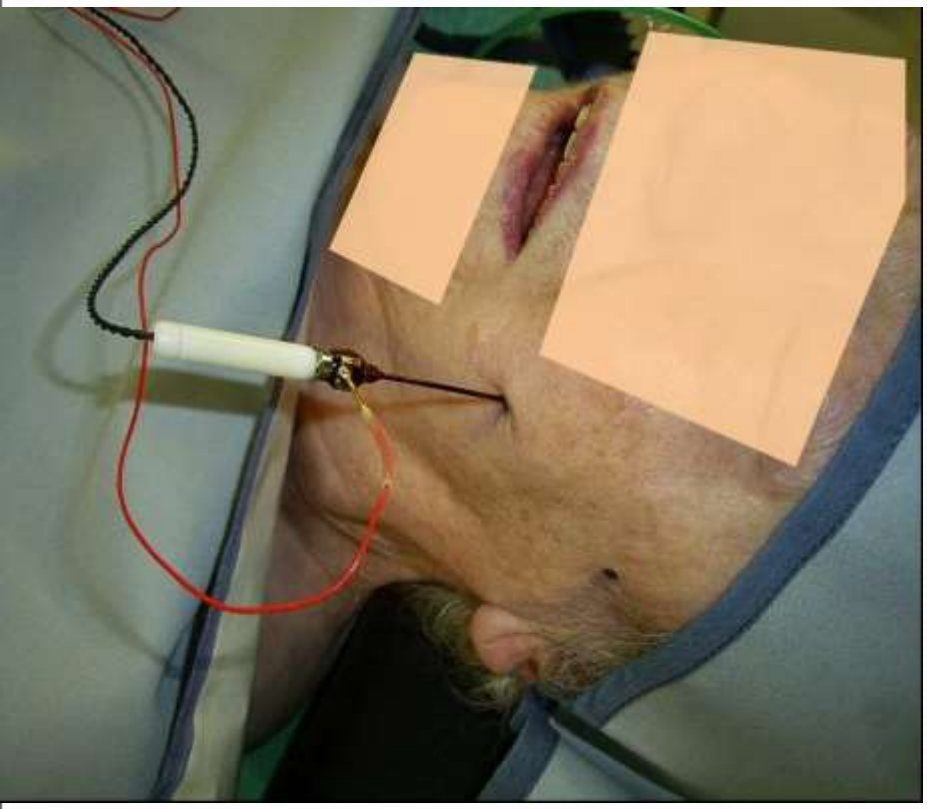
- It involves a selective partial lesioning of the affected ganglion or retrogasserian root.
- Thermocoagulation resulted in selective loss of pain mediating from thinly myelinated or non-myelinated fibres.
- A small insulated electrode is inserted through the skin of the cheek, passed medial to the mandibular ramus and through the foramen ovale to come to rest at the ventral aspect of the trigeminal ganglion in Meckel's cavity.
- Placement is confirmed under fluoroscopic control.
- A current of 0.2 to 0.3 volt at 50 cycles per second and 1msec duration will produce paresthesia in distribution of the involved nerve or trigger zone.

RADIOFREQUENCY RHIZOLYSIS

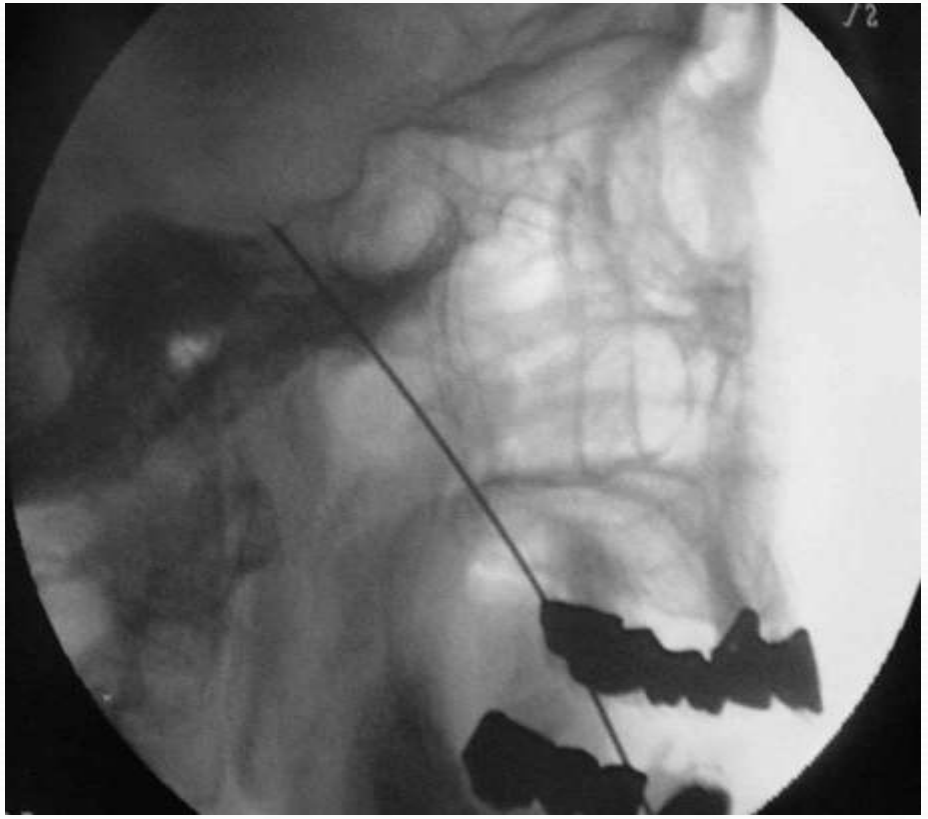
- With a radiofrequency generator, the needle tip is then heated to approximately 40^0C for a short period while the patient is sedated with an ultra short acting sedative.

Device used for radiofrequency trigeminal rhizolysis

Insertion of radiofrequency electrode through the cheek

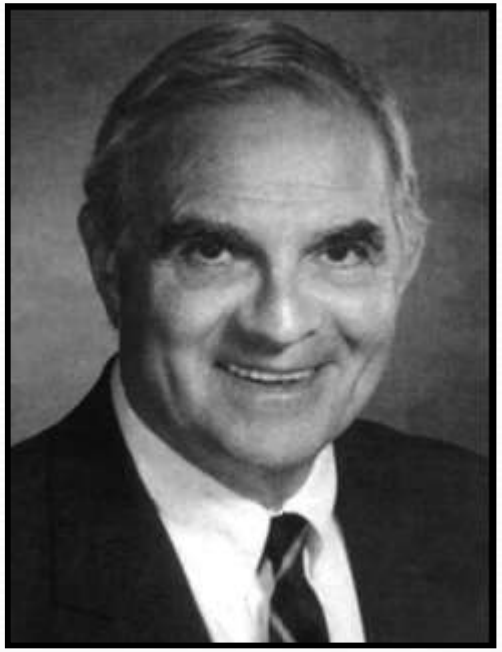


Insertion of the radiofrequency probe through the cheek

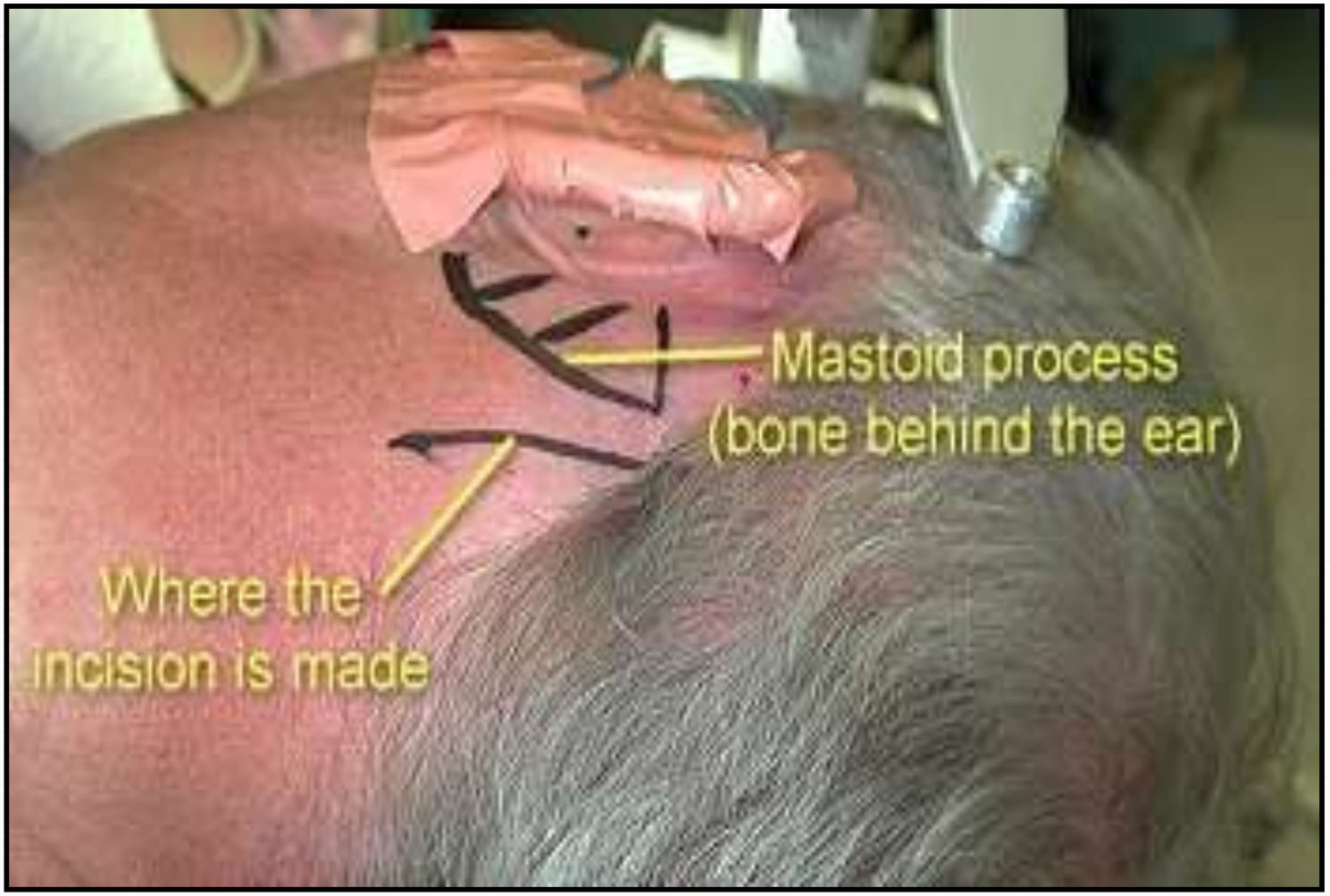


Fluoroscopic image of radiofrequency electrode positioned in the Meckel's cave

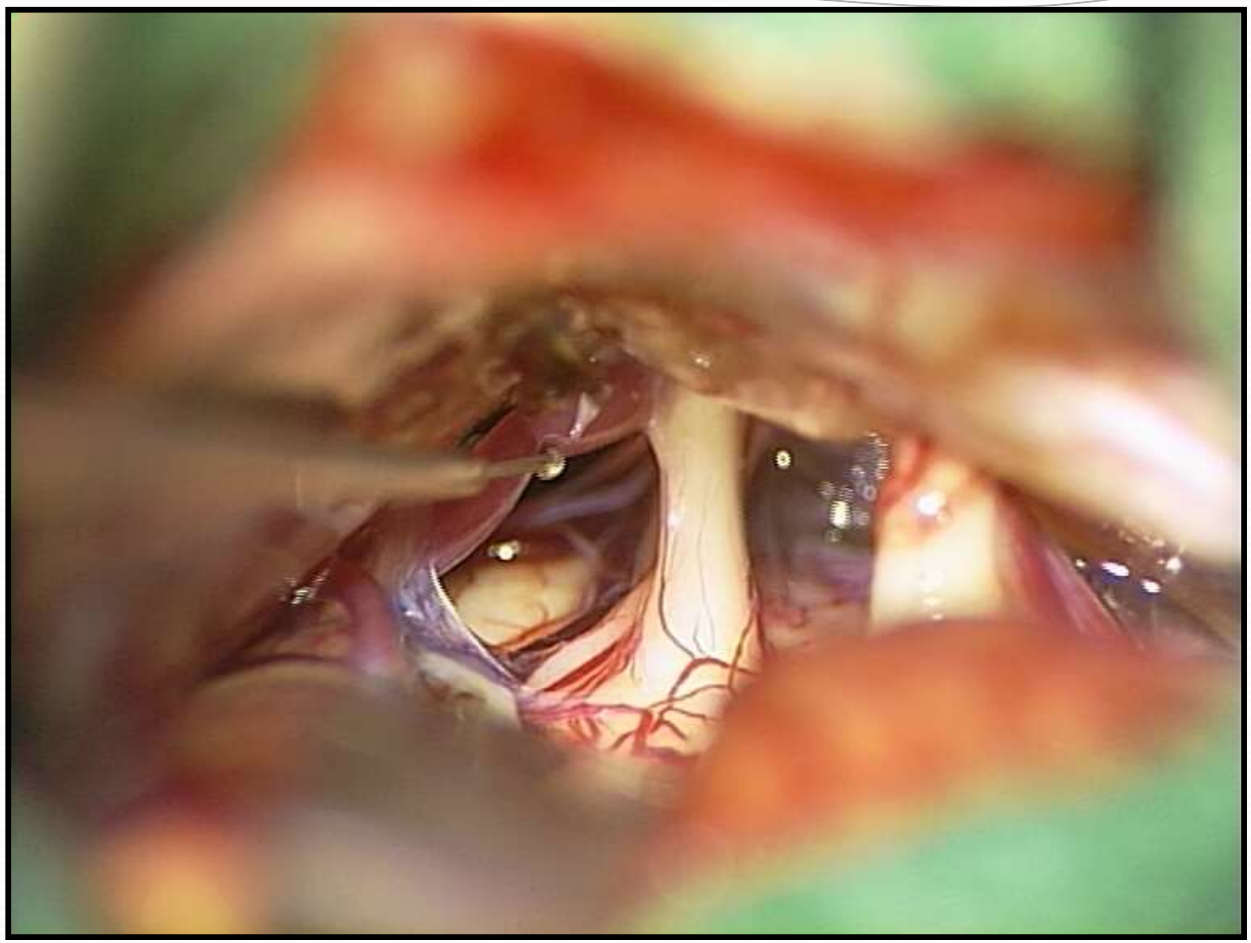
Microvascular Decompression



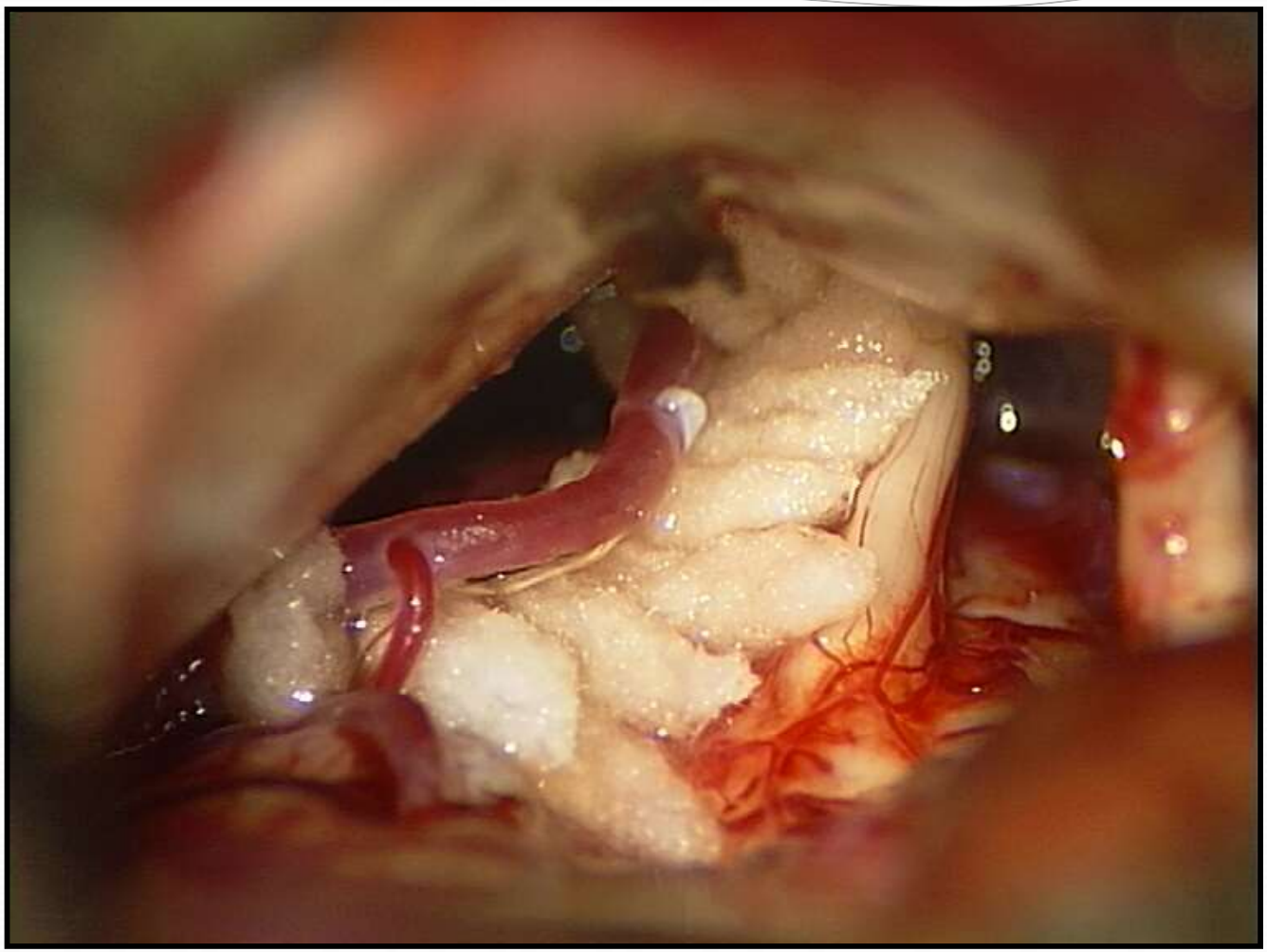
Dr. Peter Jannetta


Mastoid process
(bone behind the ear)
Where the
incision is made

Microsurgical view into the posterior fossa for microvascular decompression. trigeminal nerve and a vessel compressing the nerve close to the brainstem are identified



Mobilisation of the vascular loop away from the nerve



Positioning of Teflon between the nerve and the blood vessel. A new contacting is impossible now.

Complications

- aseptic (noninfectious) meningitis
- hearing loss
- vertigo
- double vision
- facial weakness.
- These problems are usually transient.

- More serious but rare complications include
- intracranial hemorrhage (0.02%)
- infarction (0.1%)
- cranial nerve deficits (1%) and
- postoperative seizures.