DISEASES OF THE NERVES AND MUSCLES

DEPARTMENT OF ORAL AND MAXILLOFACIAL PATHOLOGY & ORAL MICROBIOLOGY

Neuralgia (Greek *neuron, "nerve" + algos, "pain") is pain in the distribution* of a nerve or nerves, as in intercostal neuralgia, trigeminal neuralgia, and glossopharyngeal neuralgia.

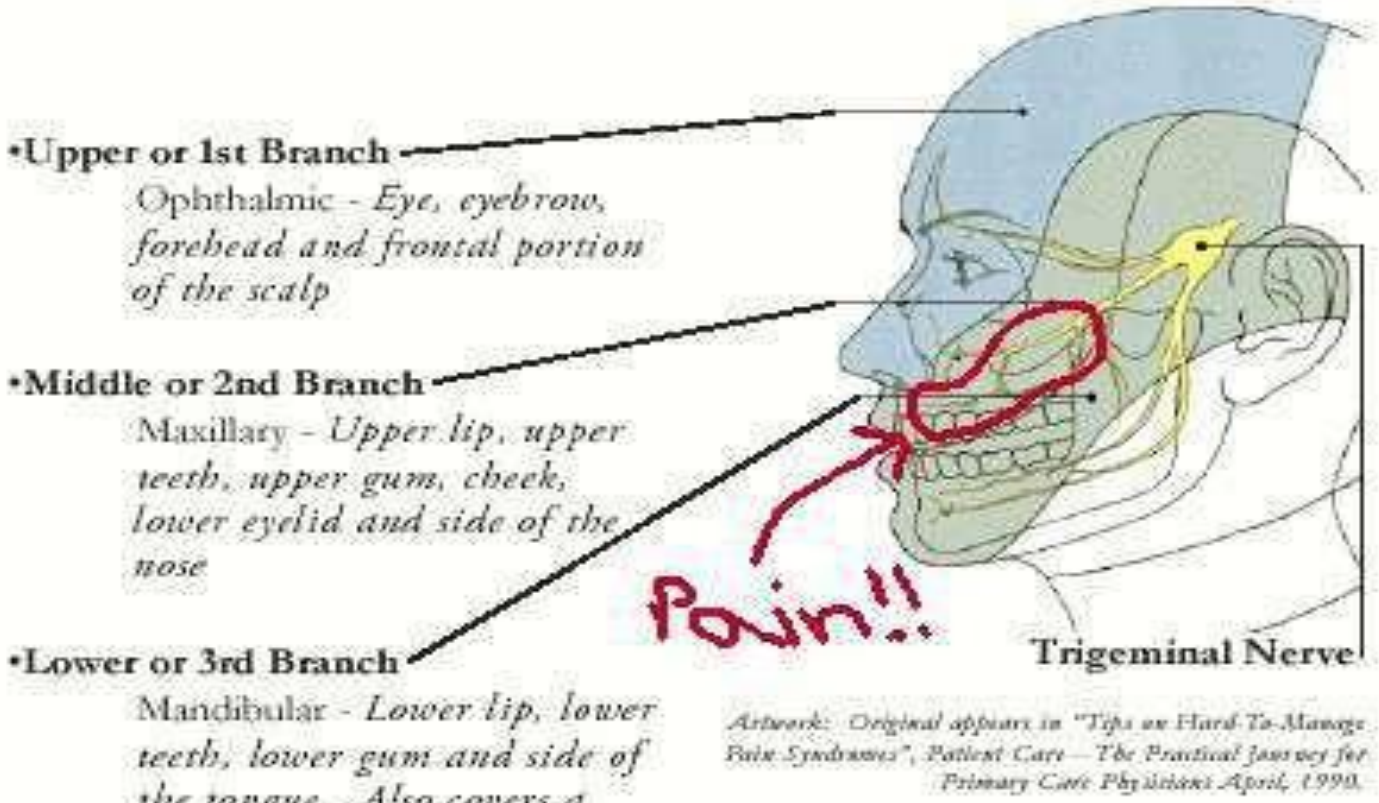
- **CLASSIFICATION:**

Under the general heading of neuralgia are:

- Trigeminal neuralgia
- Occipital neuralgia
- Glossopharyngeal neuralgia
- Postherpetic neuralgia
- Intercostal neuralgia

- **TRIGEMINAL NEURALGIA ((** TIC DOULOUREUX,
- TRIFACIAL NEURALGIA,
- FOTHERGILL'S NEURALGIA)

TRIGEMINAL NERVE



★ TRIGEMINAL NEURALGIA

- It is the most debilitating form of neuralgia that affects the sensory branches of the Vth cranial nerve.
- ▫ It is a disorder of the peripheral or central fibres of the trigeminal nerve in which the dominant symptom is pain in the anterior half of the head

DEFINITION

- It is defined as sudden, usually unilateral, severe,brief, stabbing, lancinating, recurring pain in the distribution of one or more branches of the Vth cranial nerve
- Trigeminal neuralgia also known as prosopalgia or fothergill's disease is aneuropathic disorder characterized by episodes of intense pain in the face, originating from trigeminal nerve

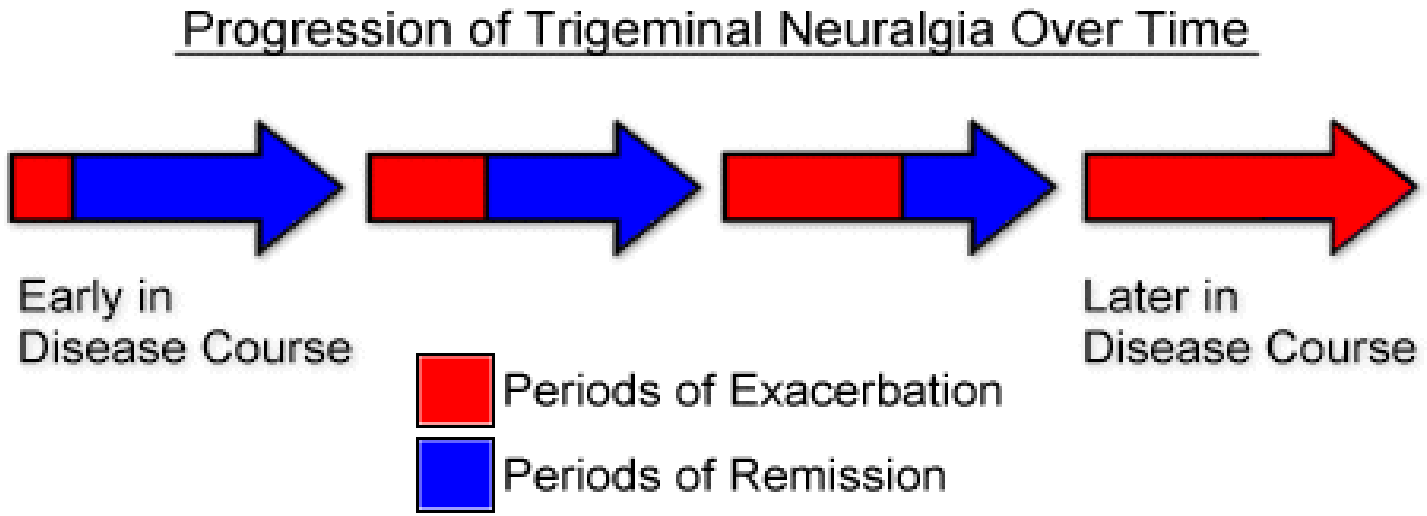
TIC DOULOUREUX

- TiC DOULOUREUX painful jerking.
- It is a truly agonizing condition, in which the patient may clunch the hand over the face & experience severe, lancinating pain associated with spasmodic contractions of the facial muscles during attacks- a feature that led to use of this term

ETIOLOGY:

- Usually idiopathic
- Demylination of the nerve
- Multiple sclerosis
- Petrous ridge compression
- Post - traumatic neuralgia
- Intracranial tumors
- Intracranial vascular abnormalities
- Viral etiology

PATHOGENESIS



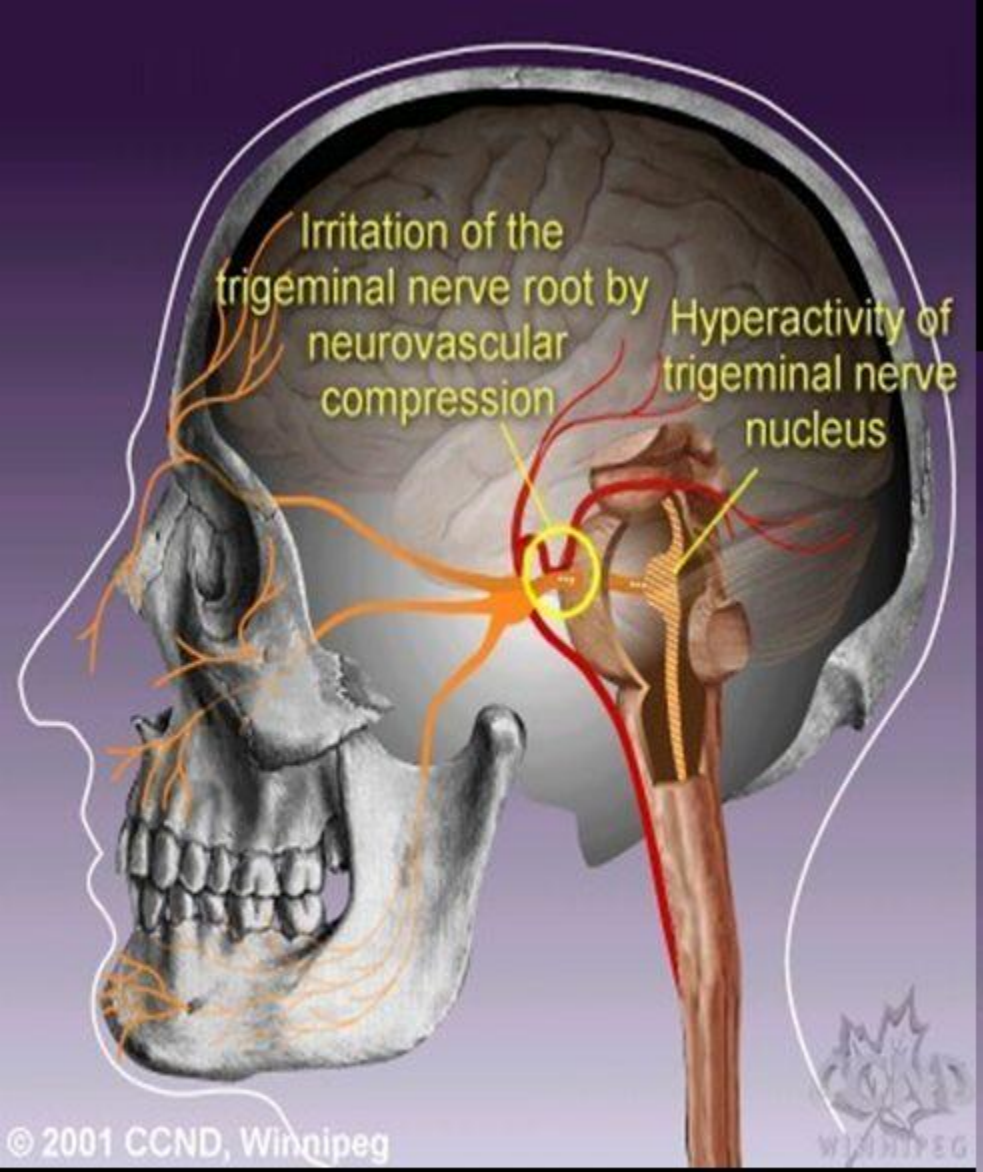
TYPES OF TRIGEMINAL NEURALGIA AND THEIR CAUSES:

- TYPICAL TRIGEMINAL NEURALGIA
- ATYPICAL TRIGEMINAL NEURALGIA
- PRE- TRIGEMINAL NEURALGIA
- MULTIPLE SCLEROSIS RELATED TRIGEMINAL NEURALGIA
- SECONDARY OR TUMOR RELATED TRIGEMINAL NEURALGIA
- TRIGEMINAL NEUROPATHY OR POSTTRAUMATIC TRIGEMINAL NEURALGIA
- FAILED TRIGEMINAL NEURALGIA

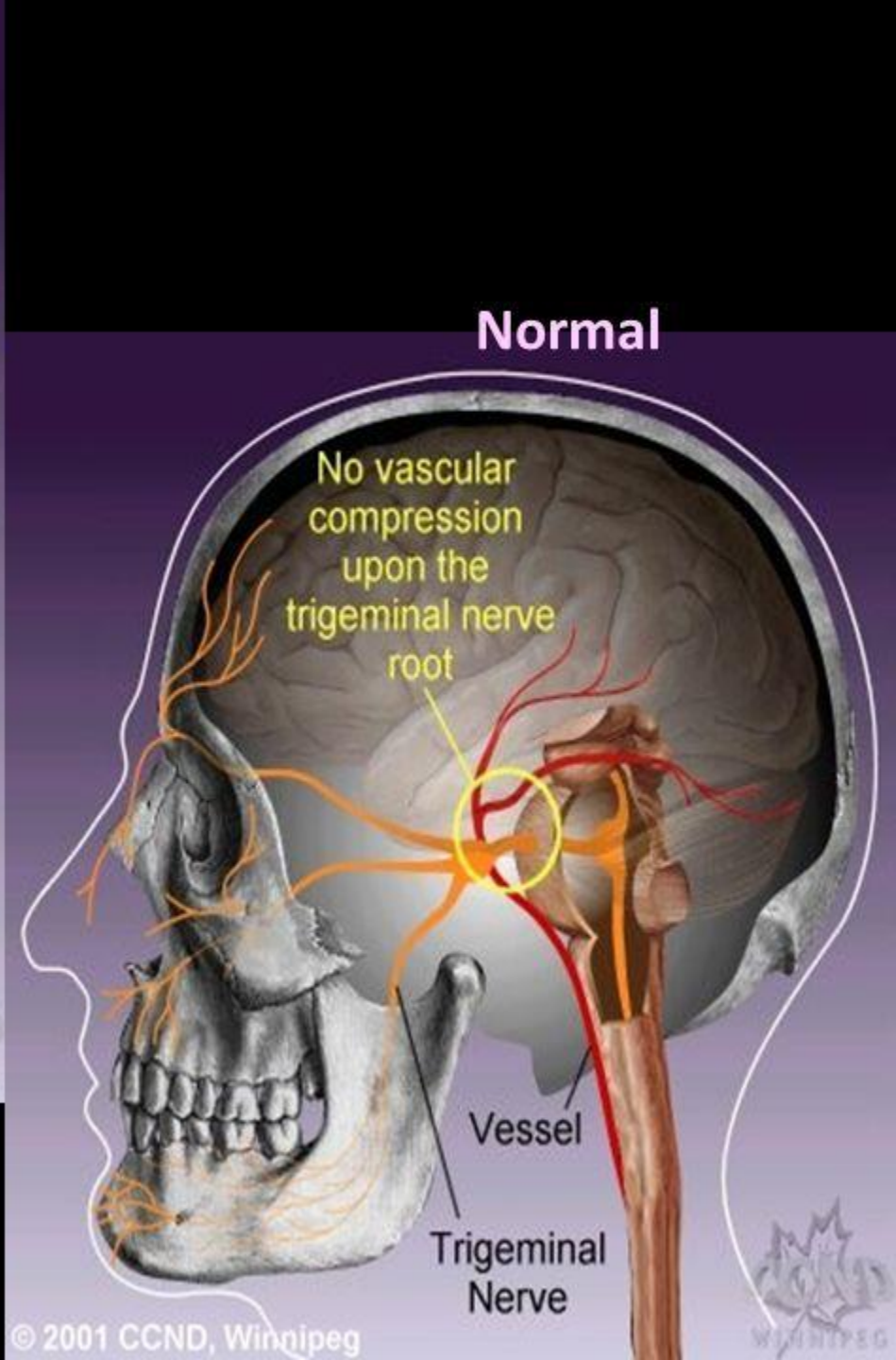
TYPICAL TRIGEMINAL NEURALGIA

- most common form, previously termed CLASSICAL,IDIOPATHIC and ESSENTIAL TN. Nearly all cases oftypical TN caused by blood vessel compressing the trigeminal nerve root.
- pulsation of vessels upon thetrigeminal nerve root do not visibly damage the nerve. However irritation from repeated pulsations may lead to

  changes of nerve function, delivery of abnormal signals to the trigeminal nerve nucleus , this causes hyperactivity of trigeminal nerve root leading to trigeminal nerve pain.



Neurovascular compression



ATYPICAL TRIGEMINAL NEURALGIA

- it is characterized by a unilateral,prominent constant and severe aching and burning pain superimposed upon otherwise typical symptom.
- Some believe that atypical TN is due to vascular compression upon specific part of the trigeminal nervewhile other theorize atypical TN as more severe progression of typical TN

SECONDARY OR TUMOR RELATED TN:

- TN pain caused by a lesion, such as a tumor. Tumor that severely compresses or distorts the trigeminal nerve may cause numbness, weakness of chewing muscles or constant aching pain

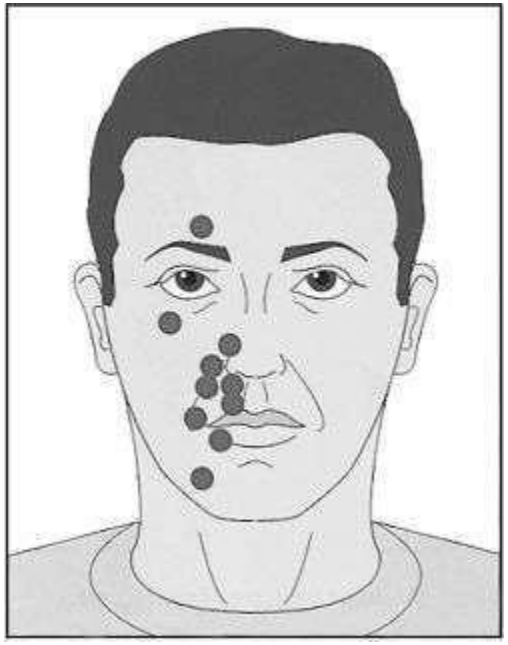
6. FAILED TRIGEMINAL NEURALGIA:

- In a very small proportion of suferres, all medications, surgical procedures prove ineffective in controlling TN pain
- Such individual also suffer from additional trigeminal neuropathy as a result of destructive intervention they underwent.

GENERAL CHARACTERISTICS

- INCIDENCE- 8: 1,00,000
- AGE- 5th-6th decade of life
- SEX- female> male
- AFFLICTION FOR SIDE- right> left
- DIVISION OF TRIGEMINAL NERVE
- INVOLVEMENT- V3>V2>V1
- TRIGGERING

TRIGGER ZONE



CLINICAL CHARACTERISTICS

- Manifests as a sudden, unilateral, intermittent paroxysmal,sharp, shooting, lancinating , shock like pain, elicited by slight touching superficial ‘trigger points’ which radiates from that point, across the distribution of one or more branches of the trigeminal nerve
- Pain is usually confined to one part of one division of trigeminal nerve
- Pain rarely crosses the midline
- Attacks do not occur during sleep

- Pain is of short duration, but may recur with variable frequency.
- In extreme cases, the patient will have a motionless face – the‘frozen or mask like face’.
- Common trigger zone include- cutaneous(corner of the lips,cheek, ala of the nose, lateral brow); intraoral(teeth, gingivae,tongue). Trigger area on the face are so sensitive that touching

  or even air currents can trigger an episode.
- 10-12% of cases are bilateral, or occurring on both sides. Thismainly seen in cases with systemic involvement include multiple

  sclerosis or expanding cranial tumor

DIAGNOSIS

- From a well taken history
- CT- scan
- MRI
- Diagnostic nerve block

DIFERENTIAL DIAGNOSIS

- MIGRAINE- severe type of periodic headache is persistent, at least over a period of hours and it has notrigger zone.
- SINUSITIS- pain is not paroxysmal, in this pain is persistent, associated nasal symptoms.
- DENTAL PAIN- localized, related to biting or hot or cold foods, visible abnormalities on oral examination.
- Tumors of nasopharynx - in this similar type of pain is produced, manifested in the lower jaw, tongue and side of the head with associated middle ear deafness. This complex lesion is called TROTTER'S syndrome.

  Patient exhibit asymmetry and defective mobility of the

- Soft palate and affected side. As the tumor progresses,trismus of internal pterygoid muscle develops, and patient is unable to open the mouth. Here actual cause of pain is involvement of mandibular nerve in the foramen ovale.
- Post herpetic neuralgia- pain is usually involved in ophthalmic division. The history of skin lesion prior to onset of neuralgia, pain is persistent, associated nasal symptoms.

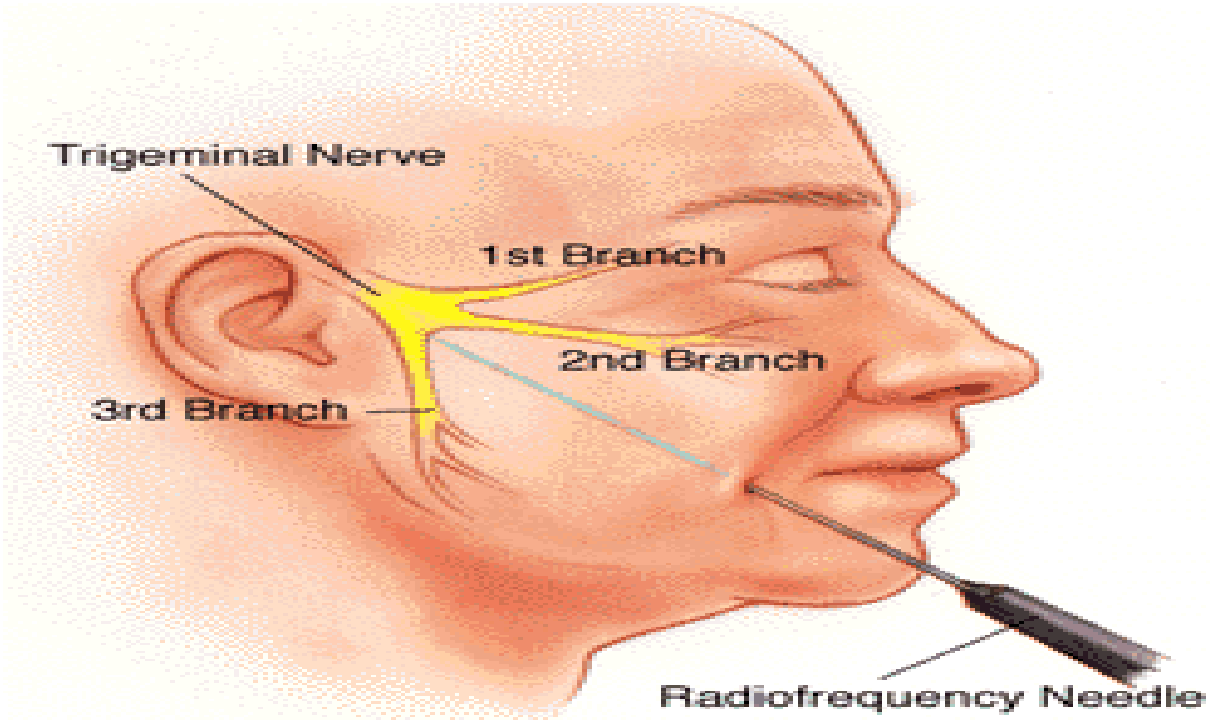
TREATMENT

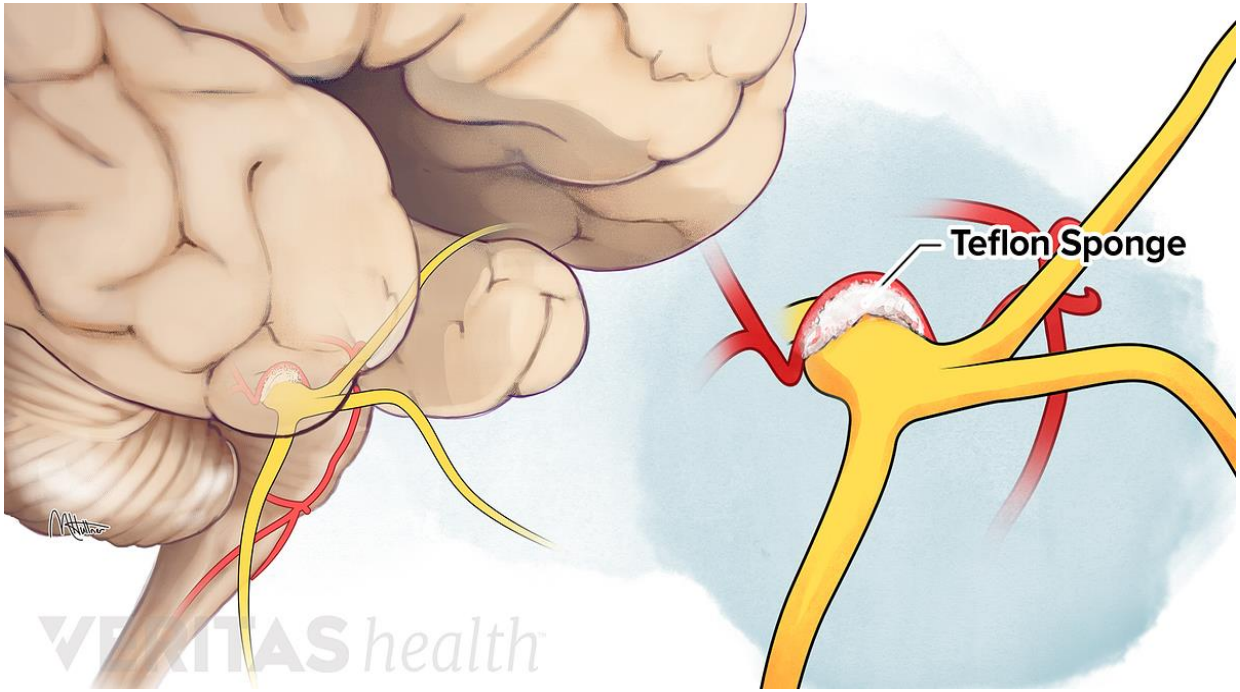
1. MEDICAL TREATMENT

- First line of treatment is: CARBAMAZIPINE (anticonvulsant)
- Second line of treatment is: BACLOFEN, LAMOTRIGINE,OXCARBAZEPINE, PHENYTOIN, GABAPENTIN, PREGABALIN,SODIUM VALPROATE
- Low dose of Antidepressants such as AMITRYPTILINE are thought to be effective in treating neuropathic pain. Antidepressant are also used to counteract a medication side effect.
- DULOXETINE is helpful where neuropathic pain and depression are combined.
- Opiates such as MORPHINE and OXYCODONE, there is evidence of their effectiveness on neuropathic pain, especially if combined with gabapentin, gallium maltoate in a cream or ointment base has been reported to relieve refractory postherpetic TN

2. SURGICAL TREATMENT

- INJECTION OF NERVE WITH ANESTHETIC
- AGENT
- Long acting anesthetic agents
- Alcohol injection
- PERIPHERAL GLYCEROL INJECTION
- PERIPHERAL NEURECTOMY(NERVE AVULSION)
- OPEN PROCEDURES (INTRACRANIAL PROCEDURES)
- MICROVASCULAR DECOMPRESSION
- PERCUTANEOUS RHIZOTOMIES
- GAMMA KNIFE RADIOSURGERY



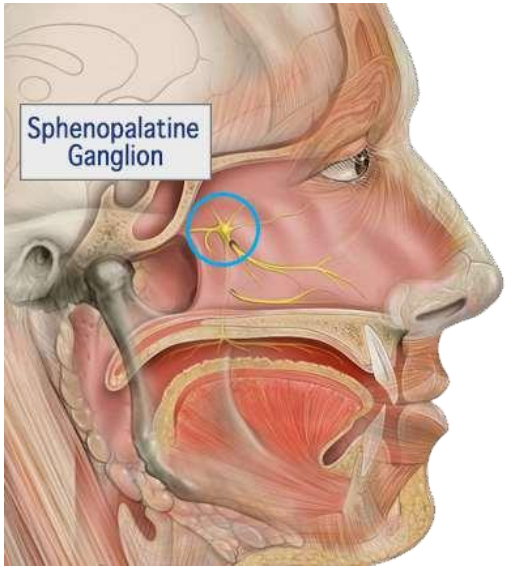


SPHENOPALATINE NEURALGIA

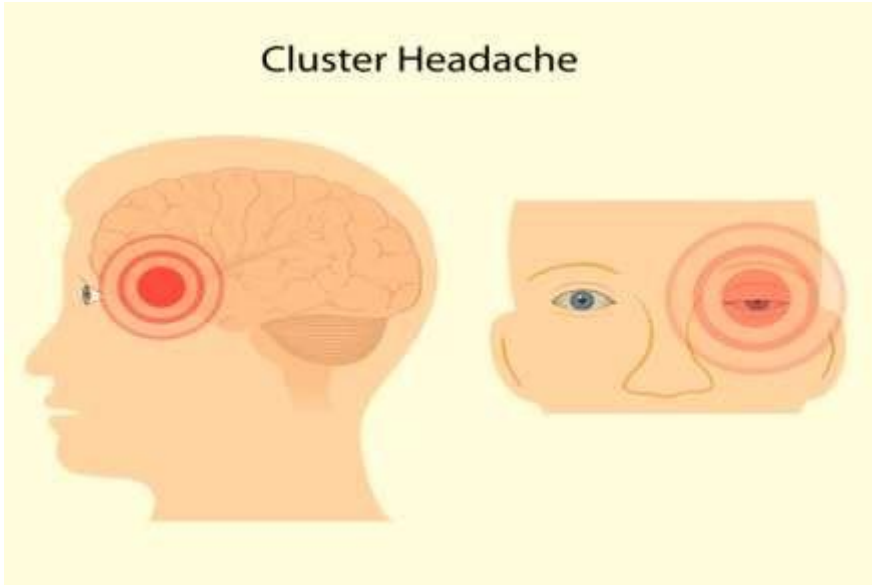
OTHER NAMES

- CLUSTER HEADACHE
- LOWER-HALF HEADACHE
- SLUDER'S HEADACHE
- VIDIAN NERVE NEURALGIA
- ATYPICAL FACIAL NEURALGIA
- HISTAMINE CEPHALGIA
- HORTON'S SYNDROME
- SUICIDE HEADACHE
- PERIODIC MIGRANOUS NEURALGIA
- ALARM CLOCK HEADACHE

- A pain syndrome originally described by **Sluder** as a symptom complex referable to the nasal ganglion.
- The sphenopalatine ganglion neuralgia is a type of headache or neuralgia, which was once believed to be attributed to the irritation of the sphenopalatine ganglion.



- A painful affliction of the mid face and upper face, particularly in and around the eye.
- The name is derived from the fact that the headache ‘clusters’ groups or occurs in periods of with extended remisssion between attacks.



ETIOLOGY

- The exact cause is not yet known.
- However, evidence strongly suggests that

- abnormalities in the hypothalamus may play a major role in cluster headaches.
- Also related to the body's sudden release of histamine (chemical in the body released during an allergy response) or serotonin (chemical made by nerve cells).

- By some not completely understood mechanism, the trigeminal nerve is also involved.

- May be caused by blood vessel dilation in the eye area (caused by excessive release of histamine). Inflammation of nearby nerves may give rise to the distinctive stabbing, throbbing pain usually felt in one eye.
- Hormones - researchers have found that many people who suffer from cluster headaches have unusual levels of melatonin and cortisol during their attacks.

Trigger Factors

Alcohol , cigarette smoking	High altitudes	Bright light
Exertion (physical activity)	Heat (hot weather, hot baths)	Foods high in nitrites (such as bacon and preserved meats)
Certain medicines	Cocaine	

CLINICAL FEATURES

May occur at any age, although it usually affects persons in the 3rd and 4th decade of life .

M > F (6:1)

They tend to run in families, passed down through genes.

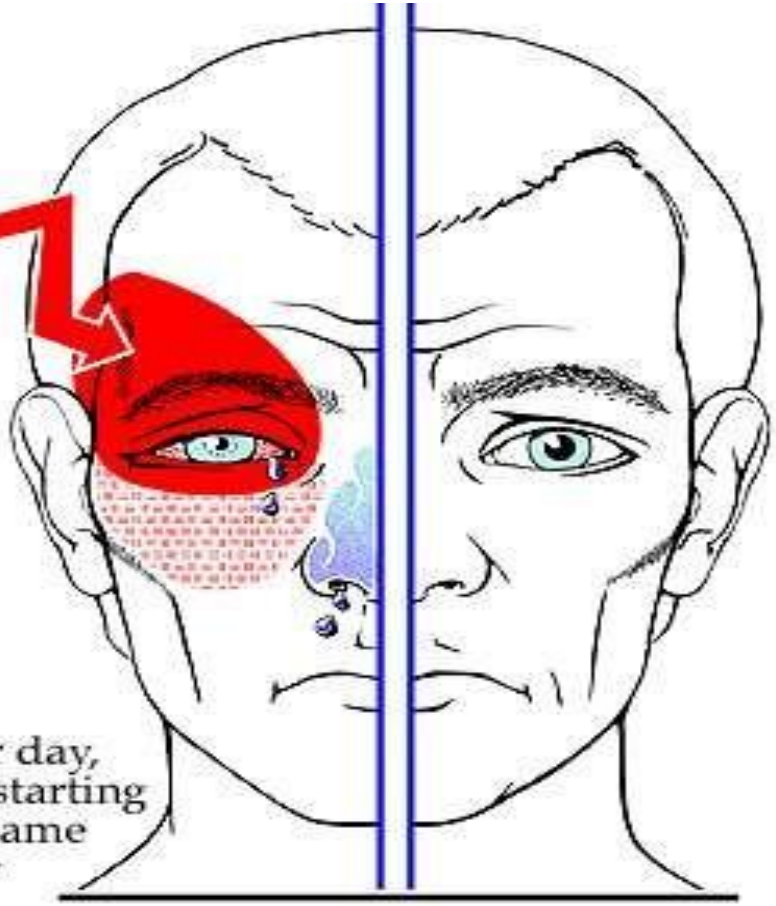
The pain is described as paroxysmal (abrupt onset) and intense, with a burning or lancinating quality and without a trigger zone.

- The pain occurs on one side of the head.
- It may be described as:
  - **Burning**
  - **Sharp**
  - **Steady**

- The pain may occur in, behind, and around one eye.
- May involve one side of the face from neck to temples.
- In some cases, the pain is so intense in and around the eye that the patient feels he should get rid off that eye.

- The pain often begins at the same time and in a given 24-hour period **(Alarm clock headache)**
- The attacks may last from **15 minutes- 3 hours if left untreated.**
- During an active cycle, people can experience as few as 1 attack every other day to as many as 8 (mostly in the night)
- Attack cycles typically last 6 - 12 weeks with remissions lasting up to 1 year.

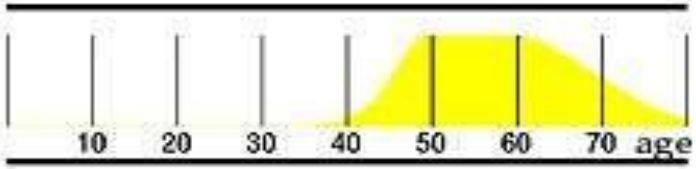
Maximum in 20 minutes



Lasting for 1 to 2 hrs,
of extreme intensity

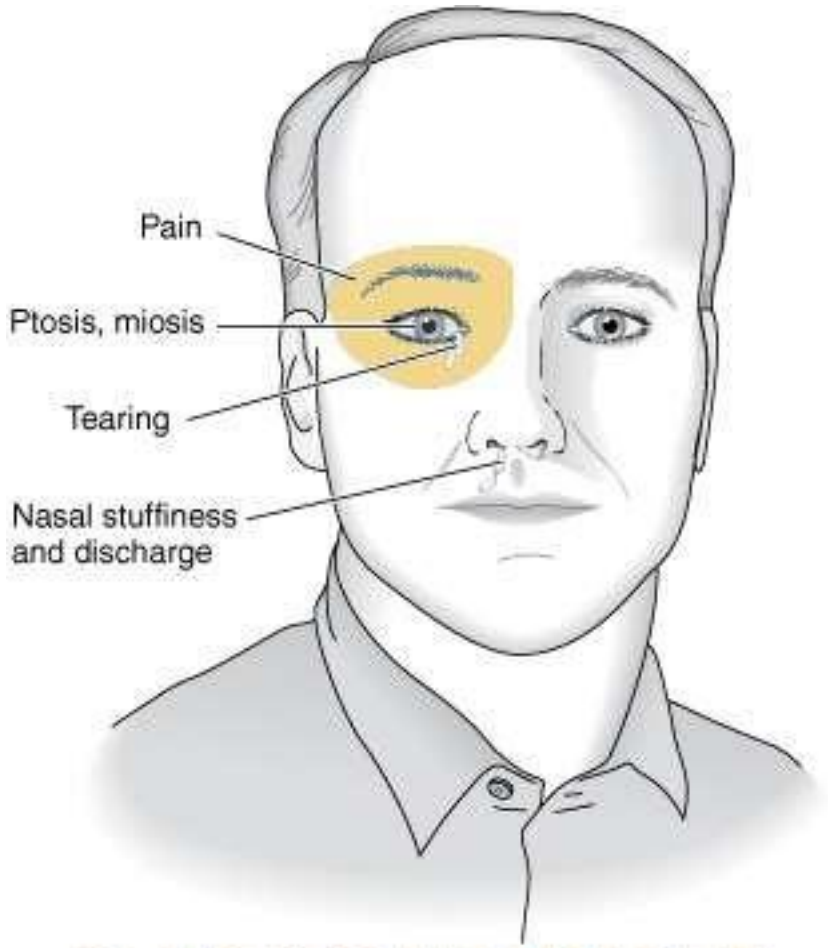
1 to 3 times per day, often at night, starting usually at the same hours each day

♂ ♀



SIGNS AND SYMPTOMS

Conjunctival injection	Excessive tearing	Lacrimation
Forehead/ facial sweating	Rhinorrhea	Ptosis
Miosis	Eyelid edema	Red flushed face



CLUSTER HEADACHE

Diagnosis

- Made from the history.
- The International Headache Society (IHS) guidelines have suggested the following diagnostic criteria:

1. At least five attacks fulfilling the criteria below:
2. Severe, or very severe, unilateral orbital, supraorbital and/or temporal pain lasting 15 to 180 minutes if untreated.
3. Headache accompanied by at least one of: ipsilateral conjunctival injection and/or lacrimation; ipsilateral nasal congestion and/or rhinorrhoea;

ipsilateral eyelid oedema; ipsilateral forehead and facial sweating; ipsilateral

miosis and/or ptosis; a sense of restlessness or agitation.

4. Attacks occur from one every other day to eight times daily.
5. Not attributable to another disorder.

Investigations:

- While no imaging study or specific blood test can confirm the diagnosis of cluster headache, an MRI or CT scan of the brain may be ordered to confirm that there are no other contributing factors that may mimic it.

Differential Diagnosis

- Headaches that may be most commonly confused with CH include:

1. **Chronic paroxysmal hemicrania** (CPH)
2. **Migraine**

3. **Trigeminal neuralgia**

TYPES OF HEADACHE

MIGRAINE

Where the pain is - Usually on one side of your head with pressure behind one eye.
Things that make it worse - Pain gets worse with normal physical activity.

SINUS

Where the pain is - Around your eyes, cheekbones, forehead, and bridge of the nose.
Things that make it worse - Leaning over, sudden movement, or exercising may make the headache worse.

TENSION

Where the pain is - On both sides or all over your head.
Things that make it worse - Physical activity does not make headache worse.

CLUSTER

Where the pain is - On one side of the face, head, or neck; does not switch sides.
Things that make it worse - Lying down makes it worse.

Medical Observer | Source: Everydayhealth / A.D.A.M

Management

Divided into three primary categories:

- ABORTIVE
- TRANSITIONAL
- PREVENTIVE

Abortive therapy

- Goal- fast, effective and consistent relief.
- Should work within 10-15 minutes to be considered adequate therapy.

1. **Oxygen**

- Excellent abortive for cluster
- Safe and easy to use
- Typical dosing — 100% oxygen given via face mask (nasal cannula not effective) at 7-10 liters/minute for 20 minutes. Pain relief typically occurs after 10-20 minutes
- **Can be used daily**

2. Sumatriptan

- Has been effective in cluster headache.
- Injectable form — most effective, often giving complete relief within 15 minutes after injection
- **Because triptans are not appropriate for daily use and certainly not multiple times each day, this treatment must be reserved for only the most serious attacks**

3. Dihydroergotamine (DHE)

- Available in injectable and nasal spray preparations
- Usually given intravenously; relief is slower with intramuscular or subcutaneous formulations
- **Can be used for several days in a row but not endlessly**

Transitional therapy

- Short-term preventive treatment that bridges the time between cluster diagnosis and when a true preventive agent becomes effective.

- When the transitional agent is tapered off (typically in one to two weeks) the maintenance preventive will have kicked in, thus the patient will have no gap in headache prevention.

1. **Steroids (e.g., prednisone, dexamethasone)**

- best transitional therapy
- Effective within 24 to 48 hours of administration
- Usually discontinued after 8-10 days of treatment when main preventive agent has started to become effective
- Long-term use not recommended

2. Dihydroergotamine (DHE)

- Can be used here also
- Best given intravenously .
- Typically relieves pain in 1-2 days of repetitive treatment; pain may not return for days to months which allows time for a preventive(s) to become effective.

3. Naratriptan

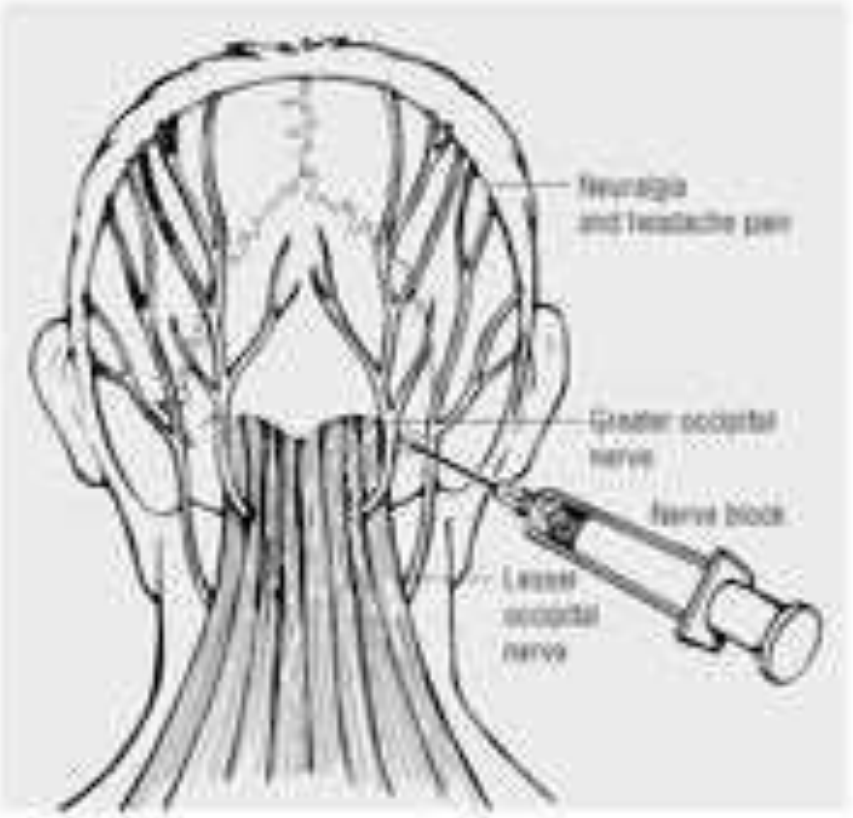
- Dose — 7 days at 2.5 mg twice daily while transitioning to a preventive program

Drawback — if an attack occurs when a cluster patient is on naratriptan, sumatriptan cannot be used as an abortive; however, oxygen therapy can be used in this case

4. Occipital nerve blockade

- Injection of anesthetic agent and a small dose of steroid into the region of the greater occipital nerve (base of skull) can provide relief averaging 13 days
- Can be performed in an outpatient setting with minimal discomfort for the patient.

Figure 3. Occipital nerve block. Via a needle inserted at the base of the skull, an anesthetic agent is injected around the origin of the greater occipital nerve.



Preventive therapy

- Starts at the onset of the cluster episode with the goal of suppressing attacks.

1. Calcium channel blockers.

- Verapamil is often the first choice
- Often used in conjunction with other medications.
- Occasionally, longer term use is needed to manage chronic cluster headache.

2. Corticosteroids. Inflammation-suppressing drugs called corticosteroids, such as prednisone, are fast-acting preventive medications

- Effective if the attack has started recently

3.Melatonin

- A safe and promising addition to the list of prophylactic agents for nocturnal attacks.

4. Lithium carbonate.

- When other medications fail.
- While you're taking this medication, your blood will be checked regularly for the development of more-serious side effects, such as kidney damage.

5. Ergots.

- Ergotamine, available as a tablet that you place under your tongue, can be taken before bed to prevent night time attacks.

6. Sodium valproate –

- Has also been used for prophylaxis, usually in chronic CH

BURNING MOUTH SYNDROME

- Burning sensations accompany many inflammatory or ulcerative diseases of the oral mucosa, but the term BMS is reserved for describing oral burning that has no detectable cause.
- In burning mouth syndrome, burning sensation of the oral mucosa with no clinically apparent alterations.
- Burning sensation with no mucosal lesions or neurological disorders to explain the symptoms.

CLASSIFICATION

- BMS has been subdivided into three general types, with
- TYPE 2 being the most common and TYPE 3 being the least common
- Type 1: symptoms not present upon waking, and then increases throughout the day
- Type 2: symptoms upon waking and through the day
- Type 3: no regular pattern of symptoms

ETIOPATHOGENESIS

- The cause remains unknown, but a number of factors have been suspected;
- hormonal and allergic disorders
- salivary gland hypo function
- chronic low-grade trauma
- psychiatric abnormalities
- Complication of therapy with ACE inhibitors

CLINICAL FEATURES

- Mucosal pain
- Burning dorsum of the tongue- highest at the anterior 1/3
- Irritated or raw feeling
- Dysgeusia (loss of taste)
- dysesthesia (abnormal sensation)
- other causes of burning symptoms of the oral mucosa must be eliminated by examination and laboratory studies before the diagnosis of bms can be made

TREATMENT

- once the diagnosis of BMS has been made by eliminating the possibility of detectable lesions or underlying medical disorders, the patient should be reassured of the benign nature of the symptoms
- Patients with symptoms that are more severe often require drug therapy.
- The drug therapies that have been found to be the most helpful are low doses of TCAS, such as amitriptyline and doxepin, or

- A 2-month course of 600 mg daily of alpha-lipoic acid has been shown to reduce BMS pain
- systemic capsaicin (0.25% capsule 3/d for 30 days) demonstrated some positive effects on bmS pain intensity.
- burning of the tongue that results from parafunctional oral habits
- may be relieved with the use of a splint covering the teeth and/or the palate.

AURICULOTEMPORAL SYNDROME (FREY'S SYNDROME ,GUSTATORY SYNDROME)

It is an unusual phenomenon ,which arises as a result of damage to the auriculotemporal nerve and subsequent reinnervation of sweat glands by parasympathetic salivary fibers.

ETIOLOGY

- Some surgical operation i.e removal of parotid tumor or the ramus of the mandible ,or a parotitis of some type that has damaged the auriculotemporal nerve .
- After a considerable amount of time following surgery ,during the damaged nerve regenerates ,the parasympathetic salivary nerve supply develops,innervating the sweat glands ,which then function after salivary ,gustatory or psychic stimulation .

CLINICAL FEATURES

- Patient typically exhibits flushing and sweating of the involved side of the face ,chiefly in the temporal area ,during eating .
- Profuse sweating may often be evoked by the parenteral administration of pilocarpine or eliminated by the administration of atropine or by procaine block of auriculotemporal nerve .
- Gustatory sweating which occurs in otherwise normal individuals when they are eating certain foods , particularly spicy or sour ones.

TREATMENT

- Intracranial division of the auriculotemporal nerve

MIGRAINE

- Migraine Is A Dominantly Inherited Disorder Characterized By Varying Degrees Of Recurrent Vascular Headache, Photophobia, Sleep Disruption, And Depression.
- Headache During A Migraine Attack Was Thought To Be A Reactive Hyperaemia In Response To Vasoconstriction-induced Ischaemia During Aura
- Throbbing Quality Of The Headache

CLINICAL FEATURES

- Second decade of life and is especially common in professional persons.
- Women more than men
- May occur at frequent intervals over a period of years
- Site-
- Temporal, frontal and retro-orbital areas,
- Parietal, postauricular, occipital or suboccipital

MIGRAINE CHARACTERISTICS

- HEADACHE ATTACKS LASTING 4-72 HOURS
- UNILATERAL LOCATION
- PULSATING SENSATION
- MODERATE OR SEVERE PAIN INTENSITY
- AGGRAVATED BY ROUTINE PHYSICAL ACTIVITY
- ACCOMPANIED BY NAUSEA AND/OR VOMITING
- SENSITIVITY TO LIGHT AND/OR SOUND

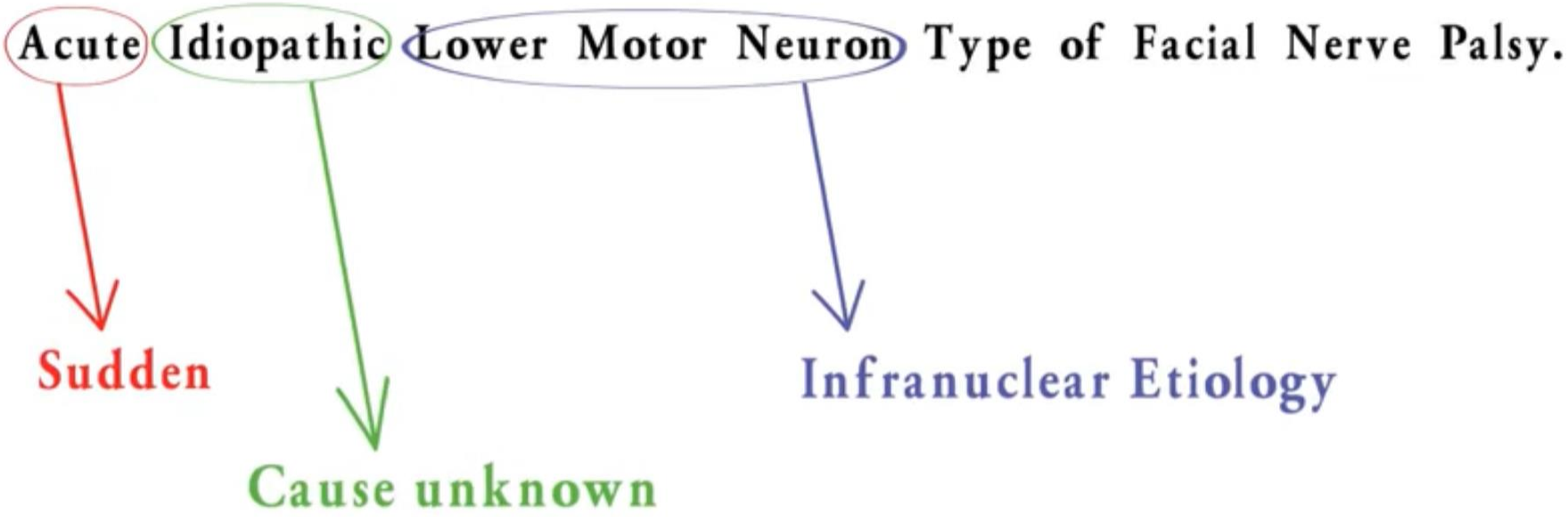
TENSION-TYPE HEADACHE CHARACTERISTICS

- LASTING FROM 30 MINUTES TO 7 DAYS
- BILATERAL LOCATION
- PRESSING OR TIGHTENING SENSATION AROUND THE HEAD
- MILD OR MODERATE PAIN INTENSITY
- NOT AGGRAVATED BY ROUTINE PHYSICAL ACTIVITY
- NO NAUSEA OR VOMITING
- SENSITIVITY TO EITHER LIGHT OR SOUND

BELL'S PALSY (SEVENTH NERVE PARALYSIS ,FACIAL PARALYSIS)

◉ INTRODUCTION:

- ◉**Bell's palsy is a form of facial paralysis resulting from a dysfunction of the** cranial nerve VII (the facial nerve) causing an inability to control facial muscles on the affected side
- ◉Several conditions can cause facial paralysis eg. Brain tumor, stroke,myasthenia gravis.
- ◉if no specific cause can be identified, the condition is known as Bell's palsy
- ◉DEFINITION: - Bell's palsy is defined as an idiopathic unilateral facial nerve paralysis, usually self-limiting.
- ◉The hallmark of this condition is a rapid onset of partial or complete paralysis that often occurs overnight.


Acute Idiopathic Lower Motor Neuron Type of Facial Nerve Palsy.
Sudden
Cause unknown
Infranuclear Etiology

Clinical Features

Onset - Acute

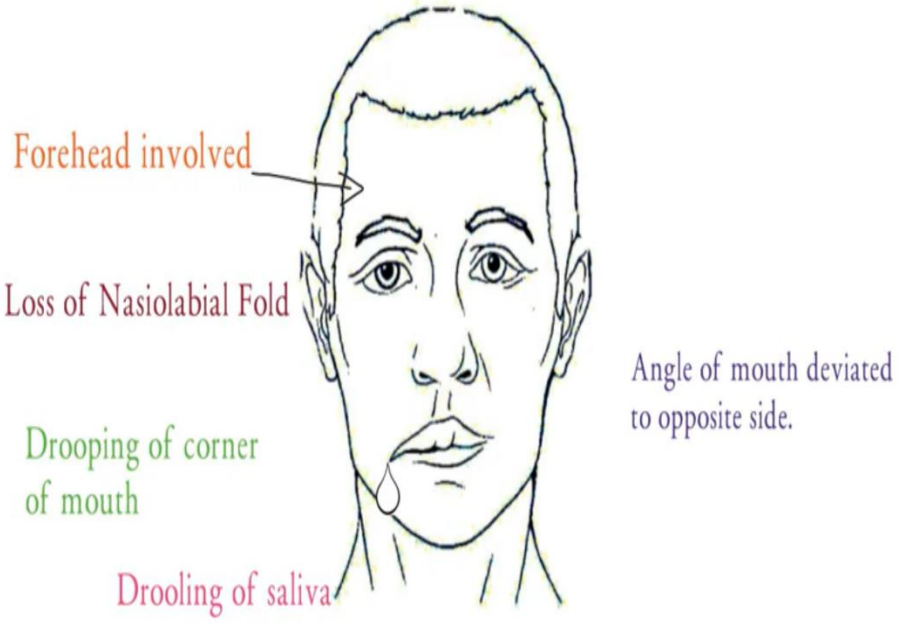
Most of the cases attain paralysis in 48 hours.

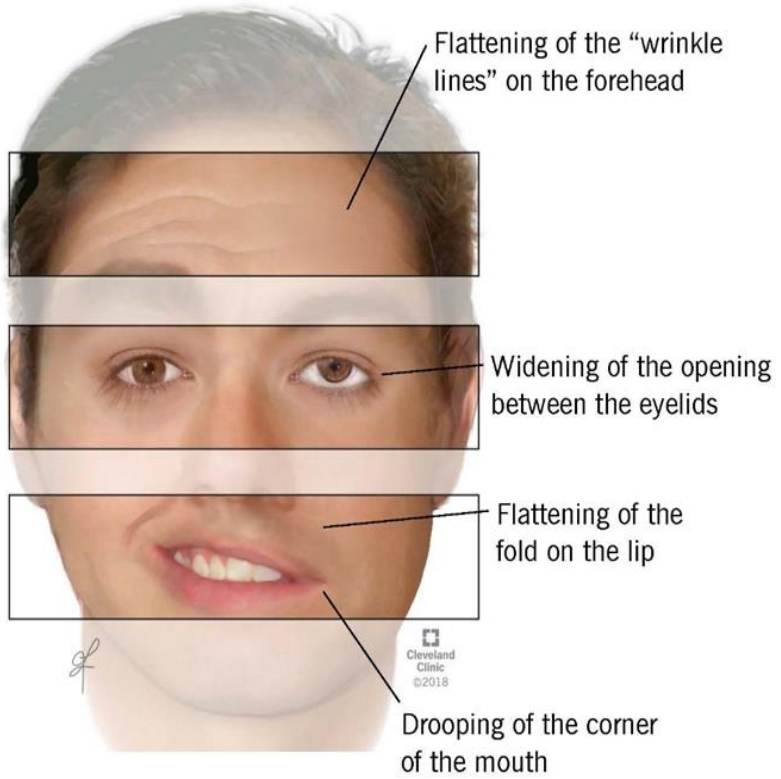
Preceded by pain behind the ear.

Impairment of taste sensations.

Hyperacusis - Involvement of Stapedius Muscle.

Features of LMN type of facial neve palsy.



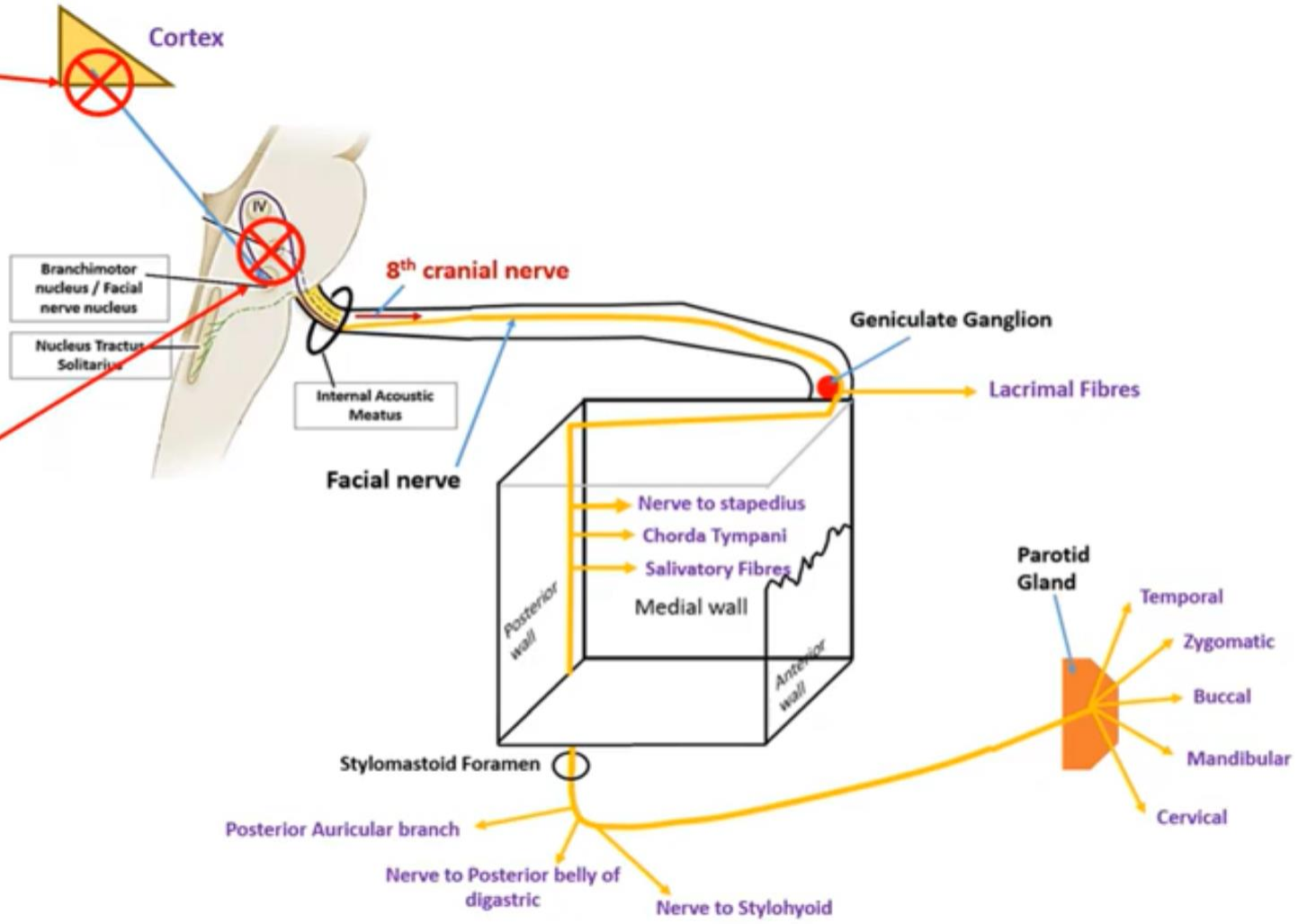


1. Supranuclear lesions:

- Contralateral UMN Type of Facial Nerve Palsy.
- Contralateral hemiplegia.

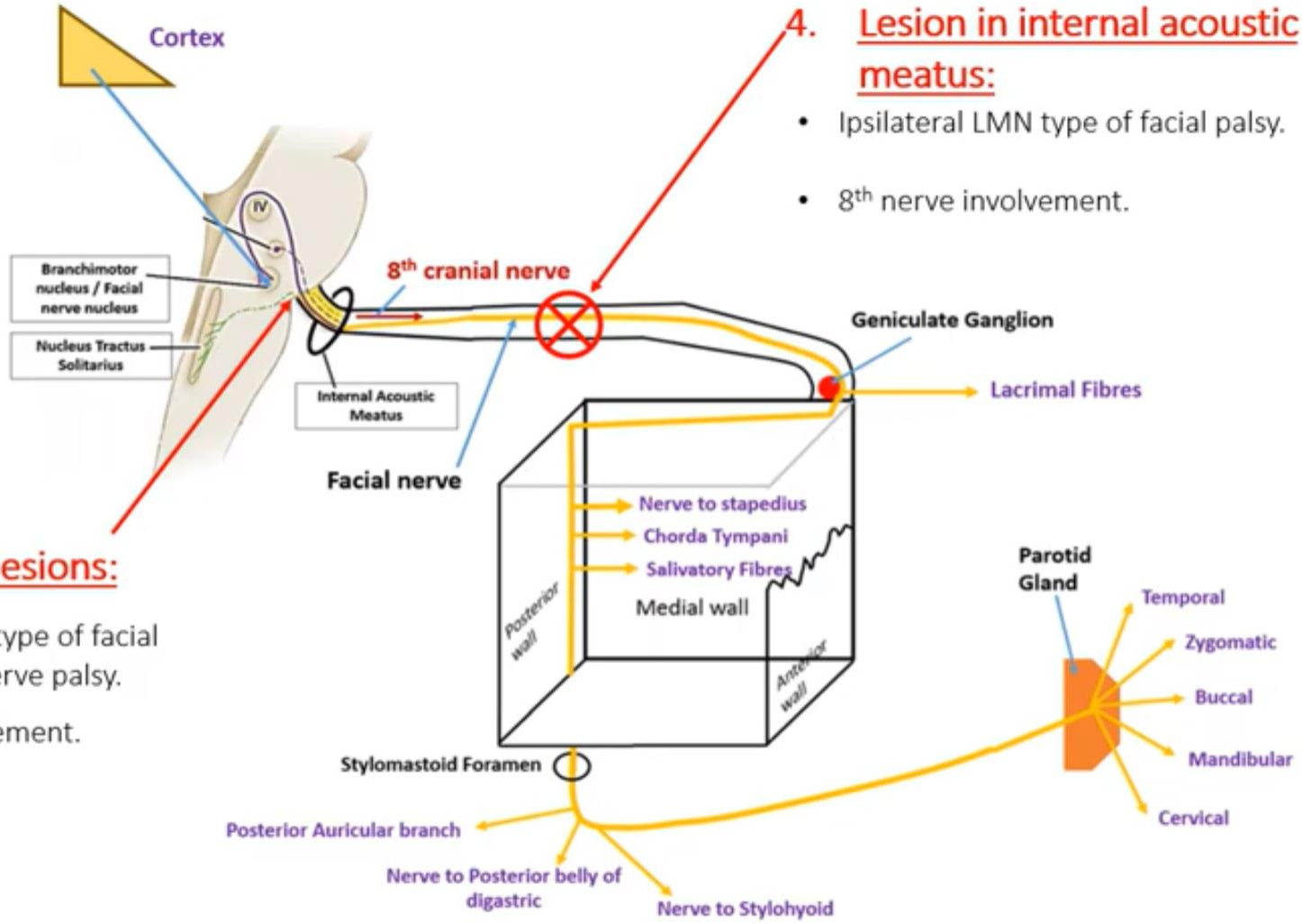
2. Nuclear lesions:

- Ipsilateral LMN Type of Facial Nerve Palsy.
- Contralateral hemiplegia.
- 6th Nerve palsy: Failure of Abduction of an ipsilateral eye.



4. Lesion in internal acoustic meatus:

- Ipsilateral LMN type of facial palsy.
- 8th nerve involvement.

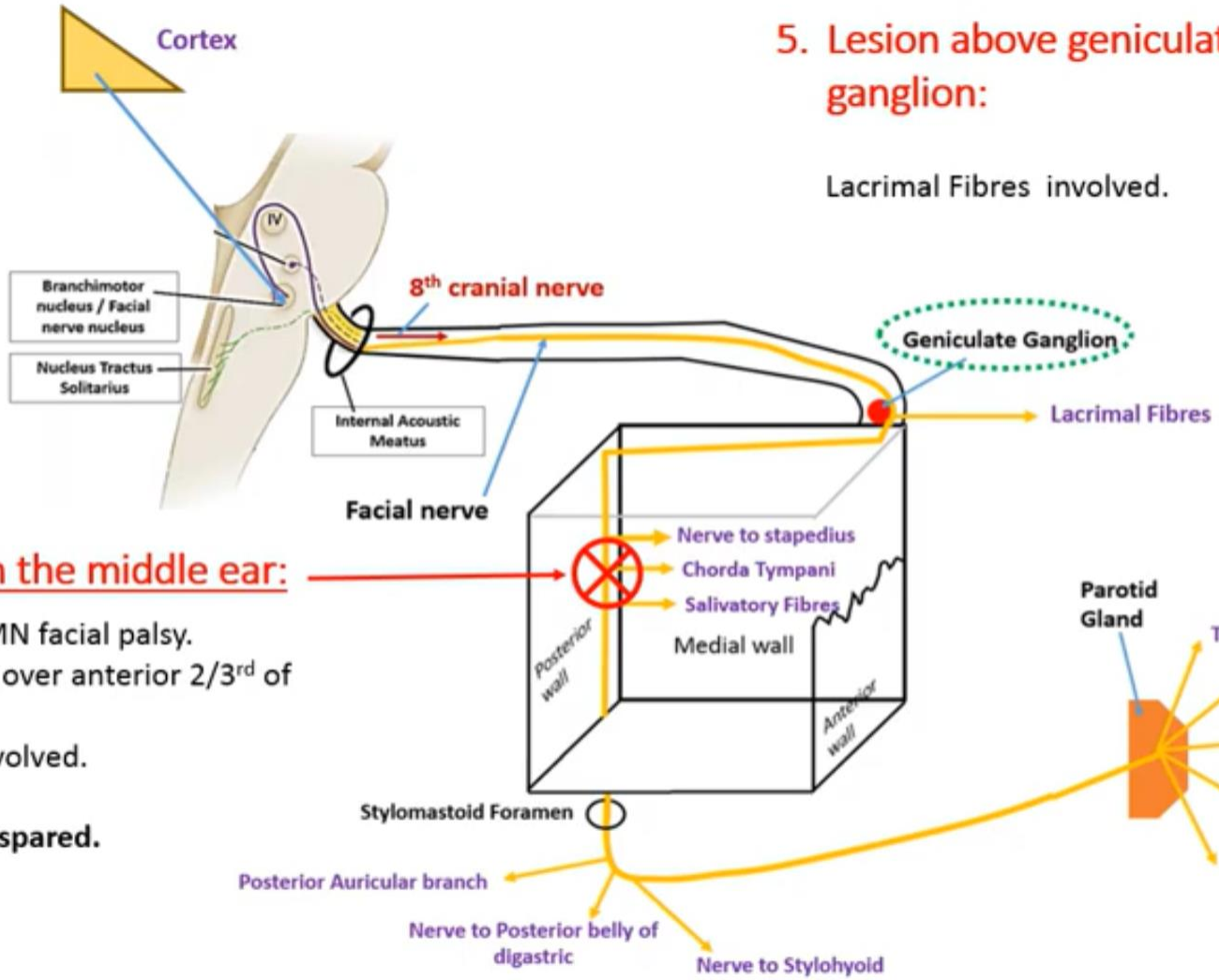


3. CP angle lesions:

- Ipsilateral LMN type of facial palsy with 6th nerve palsy.
- 8th nerve involvement.

5. Lesion above geniculate ganglion:

Lacrimal Fibres involved.

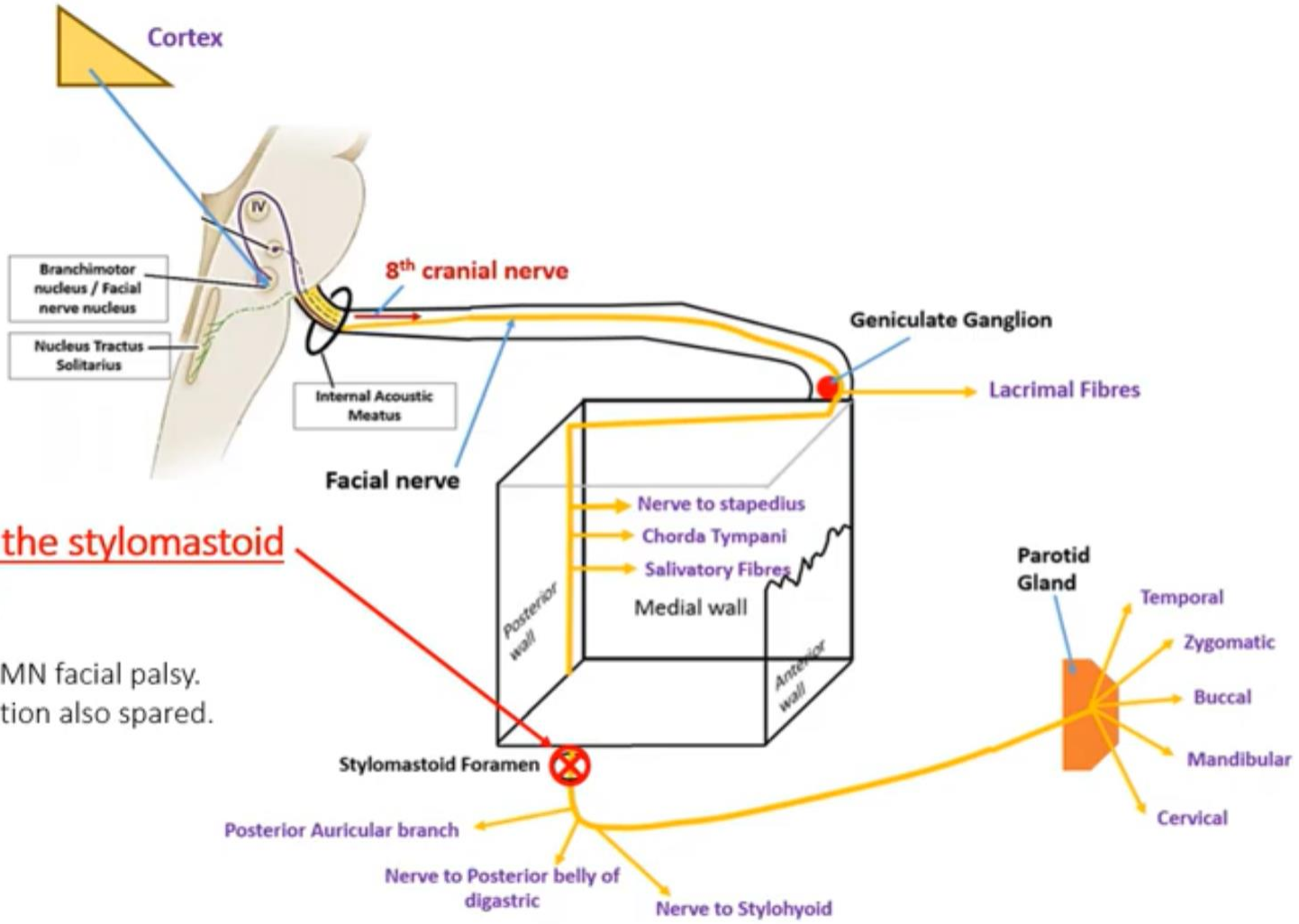


6. Lesion in the middle ear:

- Ipsilateral LMN facial palsy.
- Loss of taste over anterior 2/3rd of the tongue.
- Salivation involved.
- Hyperacusis.
- **Lacrimation spared.**

7. Lesion in the stylomastoid foramen:

- Ipsilateral LMN facial palsy.
- Taste sensation also spared.

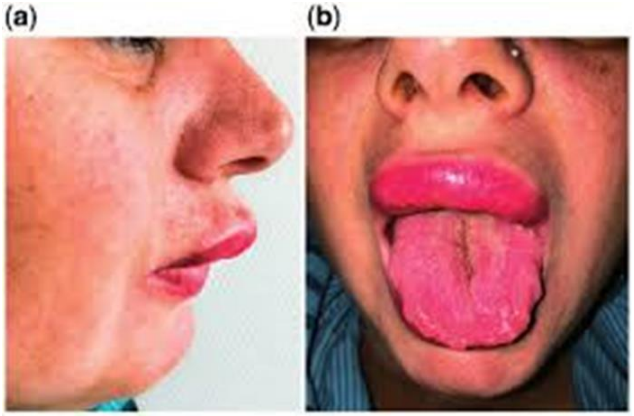


Site/Lesion	Features
Supranuclear Lesions	• Contralateral UMN Type of Facial Nerve Palsy. • Contralateral hemiplegia.
Nuclear Lesions	• Ipsilateral LMN Type of Facial Nerve Palsy. • Contralateral hemiplegia. • 6th Nerve palsy: Failure of Abduction of an ipsilateral eye.
CP Angle Lesions	• Ipsilateral LMN type of facial palsy with 6th nerve palsy. • 8th nerve involvement.
Lesion in Internal Acoustic Meatus	• Ipsilateral LMN type of facial palsy. • 8th nerve involvement.
Lesion in Middle Ear	• Ipsilateral LMN facial palsy. • Loss of taste over anterior 2/3rd of the tongue. • Salivation involved. • Hyperacusis. • Lacrimation spared.
Lesion near Stylomastoid Foramen	• Ipsilateral LMN facial palsy. • Taste sensation also spared.

ASSOCIATED SYNDROME

- 1. MELKERSON ROSENTHAL SYNDROME(a triad of fissured tongue, persistent or recurring lip or facial swelling and cranial nerve 8th paralysis)
- 2. CROCODILE TEAR SYNDROME(Due to injury to facial nerve proximal to the genicular ganglion, there may be misdirection of the nerve fibers to the lacrimal gland instead of going to the submandibular through greater petrosal nerve. As a result the patient lacrimates while

  eating. This is treated by dividing the greater petrosal nerve.
- 3. RAMSAY HUNT SYNDROME(Severe facial paralysis with vesicles in the ipsilateral pharynx and external auditory canal may be due to herpes zoster of the geniculate ganglion of the facial nerve.)
- **BILATERAL FACIAL PARALYSIS is rare may be due to acute idiopathic polyneuritis,**sarcoidosis, post cranial fossa tumors.



GENRAL CHARACTERISTICS

- INCIDENCE- 20: 1,00,000
- AGE- middle age group
- SEX- female> male

SIGN AND SYMPTOM

- This is characterized by unilateral paralysis of all muscles of facial expression for both voluntary and emotional movements.
- Forehead is unfurrowed.
- Patient is unable to cross eye on that side, any attempted closure causes rolling of eye upwards (Bell's sign).
- Tears tend to overflow (epiphora). Tears fail to enter the lacrimal puncta because they are no longer in contact with the conjunctiva. Conjunctival reflex is absent.
- Corner of the mouth droops and nasolabial fold is obliterated. Saliva dribbles and food collects in thevestibule because of paralysis of buccinator.
- The lips remain in contact and cannot be pursued,in attempting to smile the angle of mouth is not drawn up on the affected side. The mouth takes a triangular form.

TREATMENT

◉**Medications**

Commonly used medications to treat Bell's palsy include:

•**Corticosteroids,** such as prednisone, are powerful anti-inflammatory agents.

•**Antiviral drugs**

valacyclovir (Valtrex) or acyclovir (Zovirax) is sometimes given in combination with prednisone in people with severe facial palsy.

Physical therapy

Surgery

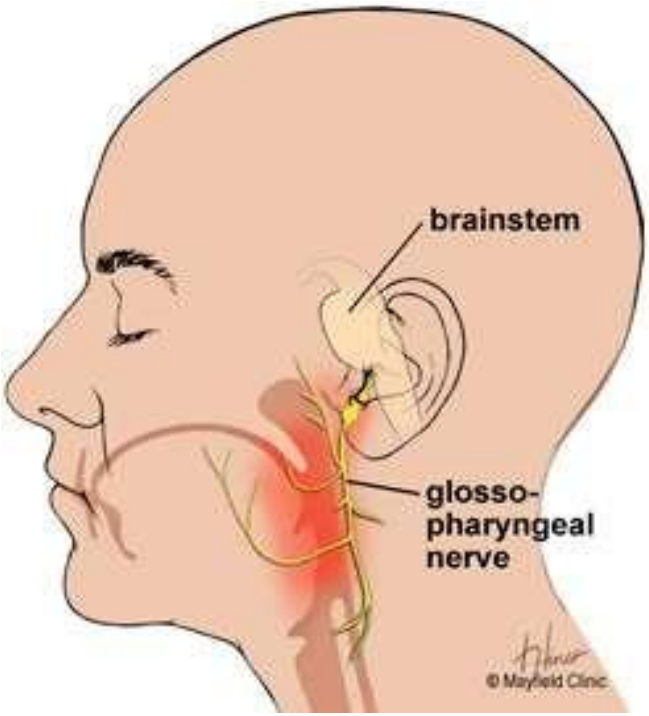
decompression surgery was used to relieve the pressure on the facial nerve by opening the bony passage that the nerve passes through.

Glossopharyngeal neuralgia

- Similar to trigeminal neuralgia except the location.
- Pain is present on tonsil and ear.
- Pain is radiating from throat to ear because of tympanic branch of IX nerve.
- It is two types

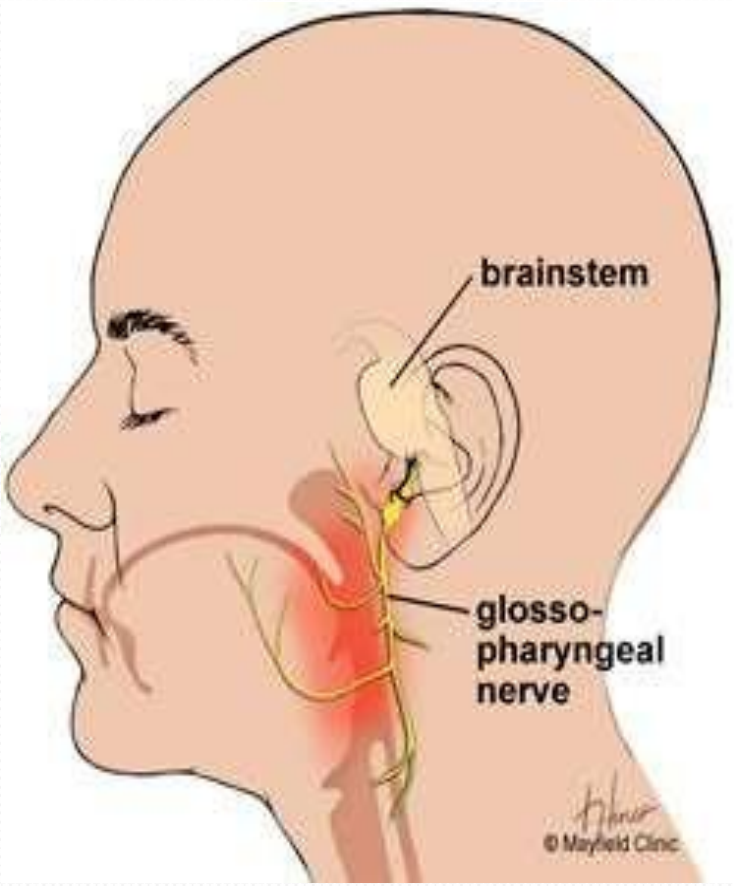
  1.Classical

  2.Symptomatic



- Classical type is obtained by arterial compression of the nerve at courses through jugular foramen

- Symptomatic type is obtain by under lying cause such as oropharyngeal tumors pagetic bone or calcified stylohyoid ligament.



Clinical features

- Rarely bilateral involvement.
- Episodic pain ,sharp, lancinating, and extreme intense.
- Pain during talking ,chewing, yawning, swallowing, touching a blunt instrument to tonsil.
- No definite trigger zone.
- Pt difficulty to localize the pain in oropharynx.

Diagnosis

- MRI scan of head
- CT scan of head
- X rays of arteries with dye(conventional angiography)

Treatment

Medical :

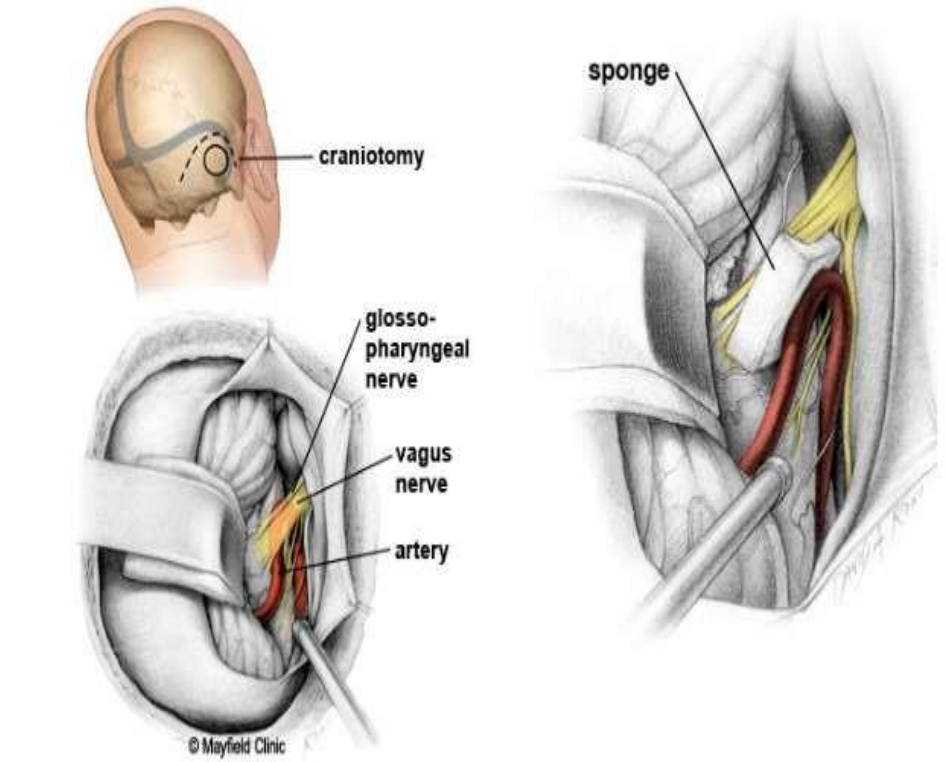
Anticonvulsant medications-

Carbamazepine,
Oxcarbazepine,
Baclofen,
Phenytoin,
Lamotrigine

Surgical

1.Micro vascular decompression

2.Surgical sectioning of glossopharyngeal nerve.



Glossopharyngeal paralysis

Etiology –

Diphtheria may cause IX nerve paralysis(Reversible)
Diseases to lower brainstem
Surgical procedures to tonsil.

Symptoms –

Loss of sensation from pharynx
Decreased salivation
Loss of taste sensation at post 1/3rd of the tongue.

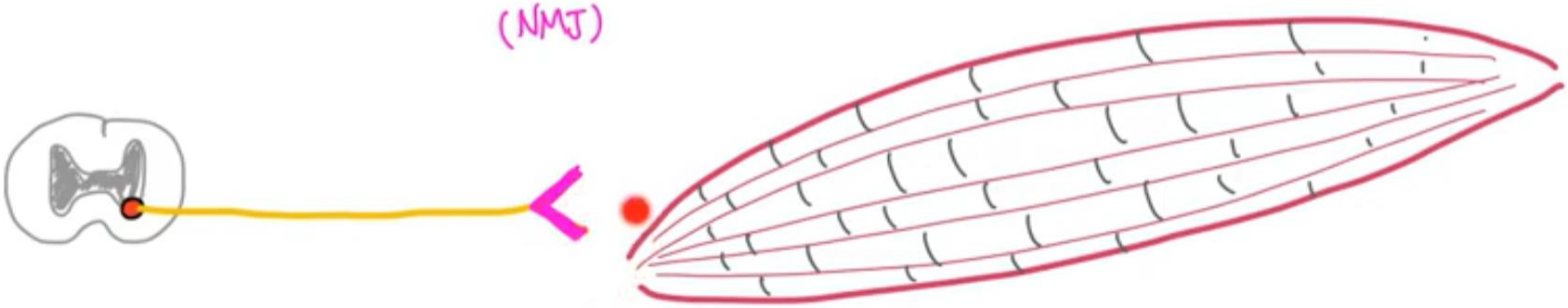
Horner's Syndrome

- Miosis, or contraction of the pupil of the eye due to paresis of the dilator of the pupil.
- Ptosis, or drooping of the eyelid due to paresis of the smooth muscle elevator of the upper lid.
- Anhidrosis and vasodilatation over the face

•DISEASE OF MUSCLES

Neuromuscular diseases (NMDs) of childhood

- Anterior horn cell (A.H.C) disorders
- Peripheral neuropathy
- Diseases of the neuromuscular junction (NMJ)
- Muscular dystrophies
- Metabolic myopathies
- Congenital myopathies



Spinal cord

Peripheral nerve

Muscle

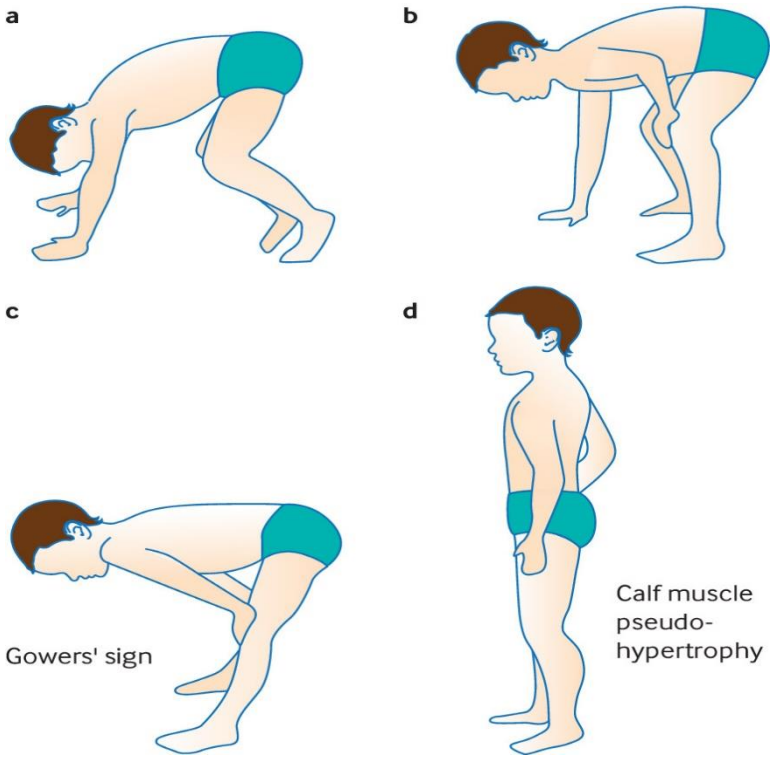
Muscular dystrophy-

a group of inherited diseases that damage and weaken **muscles** over time.

This damage and weakness is due to the lack of a protein called **dystrophin**, which is necessary for normal **muscle** function

Duchenne muscular dystrophy

This type of muscular dystrophy is the most common among children. The majority of individuals affected are boys.

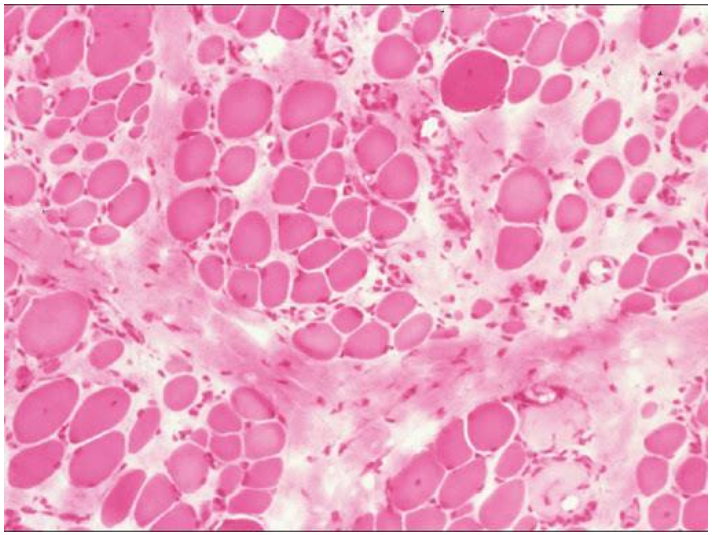


symptoms include:

- trouble walking
- loss of reflexes
- difficulty standing up
- poor posture
- bone thinning
- scoliosis, which is an abnormal curvature of your spine
- mild intellectual impairment
- breathing difficulties
- swallowing problems
- lung and heart weaknes

Histologic Features

- gradual disappearance of muscle fibres as the disease progresses, until ultimately no fibres may be recognized, being replaced entirely by connective tissue and fat.
- Persistent fibres show variation in size in earlier stages of the disease, some being hypertrophic, but others atrophic.



Laboratory Findings

- serum creatine phosphokinase (CPK) level & aldolase is elevated

Becker muscular dystrophy

- Becker muscular dystrophy is similar to Duchenne muscular dystrophy, but it's less severe. This type of muscular dystrophy also more commonly affects boys. Muscle weakness occurs mostly in your arms and legs, with symptoms appearing between age 11 and 25.

- walking on your toes
- frequent falls
- muscle cramps
- trouble getting up from the floor

Duchenne & Becker muscular dystrophies		
Diagnosis	**Duchenne**	**Becker**
Genetics	X-linked recessive deletion of dystrophin gene on chromosome Xp21	
Clinical presentation	• Onset: Age 2-3 • Progressive weakness, Gower sign, calf pseudohypertrophy • Cardiomyopathy • Scoliosis	• Onset: Age 5-15 • Variable weakness • Cardiomyopathy
Diagnosis	• ↑ Serum creatine phosphokinase • Genetic testing: Dystrophin deletion • Electromyography: Myopathic changes • Muscle biopsy: Significant fibrosis, fat, muscle degeneration & absent immunochemistry staining of dystrophin	
Western blot	<5% dystrophin	20%-50% dystrophin
Treatment	Physical therapy, glucocorticoids	Physical therapy
Prognosis	• Wheelchair-dependent by adolescence • Death by age 20-30 from respiratory or heart failure	Death by age 40-50 from heart failure

Congenital muscular dystrophy

- Congenital muscular dystrophies are often apparent between birth and age 2.

- muscle weakness
- poor motor control
- inability to sit or stand without support
- scoliosis
- foot deformities
- trouble swallowing
- respiratory problems
- vision problems
- speech problems
- intellectual impairment

Facioscapulohumeral

- This type of muscular dystrophy affects the muscles in your face, shoulders, and upper arms. FSHD may cause:
- difficulty chewing or swallowing
- slanted shoulders
- a crooked appearance of the mouth
- a wing-like appearance of the shoulder blades
- A smaller number of people with FSHD may develop hearing and respiratory problems.

MYOTONIAS

- **Myotonia** is a symptom of a small handful of certain neuromuscular disorders characterized by delayed relaxation (prolonged contraction) of the skeletal muscles after voluntary contraction or electrical stimulation.

•Dystrophic Myotonia

- Dystrophic myotonia is a steadily progressive, familial, distal myopathy with associated weakness of the muscles of the face, jaw and neck, and levators of the eyelids
- It is inherited as an autosomal dominant characteristic.

Clinical Features

- Atrophy of muscles (manifested first in the muscles of the hands and forearms.)
- third decade of life
- facial muscles involve
- ptosis of the eyelids
- atrophy of the masseter and sternocleidomastoid muscles
- masseteric atrophy produces a narrowing of the lower half of the face
- myopathic facies & swan neck'.
- Pharyngeal and laryngeal muscles in patients with dystrophic myotonia also exhibit weakness manifested by a weak, monotonous, nasal type of voice and subsequent dysphagia
- Recurrent dislocation of the jaw

Histologic Features

- Enlargement of scattered muscle fibres
- centrally placed muscle nuclei in long rows
- nuclear proliferation, intense basophilic cytoplasmic staining and phagocytosis
- In advanced muscular atrophy, fibres appear small and there may be interstitial fatty infiltration.

MYASTHENIA GRAVIS

Myasthenia Gravis

Myasthenia gravis (MG) is an autoimmune disorder affecting the myoneural (neuro – muscular) junction characterized by the fluctuating weakness of certain skeletal muscle groups..

Women are affected more frequently than men, and they tend to develop the disease at an earlier age (20 to 40 years of age, versus 60 to 70 years for men).

Depolarisation wave of Action potential

↓

Voltage Gated Ca^{++} channel open

↓

Ca^{++} ion Influx

↓

Fuse with Protein Receptor
on Presynaptic Membrane

↓

Help to fuse Ach Vesicles with
Nerve Membrane

Release of Ach in Synaptic cleft.

(Exocytosis)

↓

Ach diffuse from High concentration to lower

(presynaptic membrane → post synaptic membrane)

↓

Ach Bind with Ach Recep.

↓

Na^+ Ion channel open → Na^+ influx

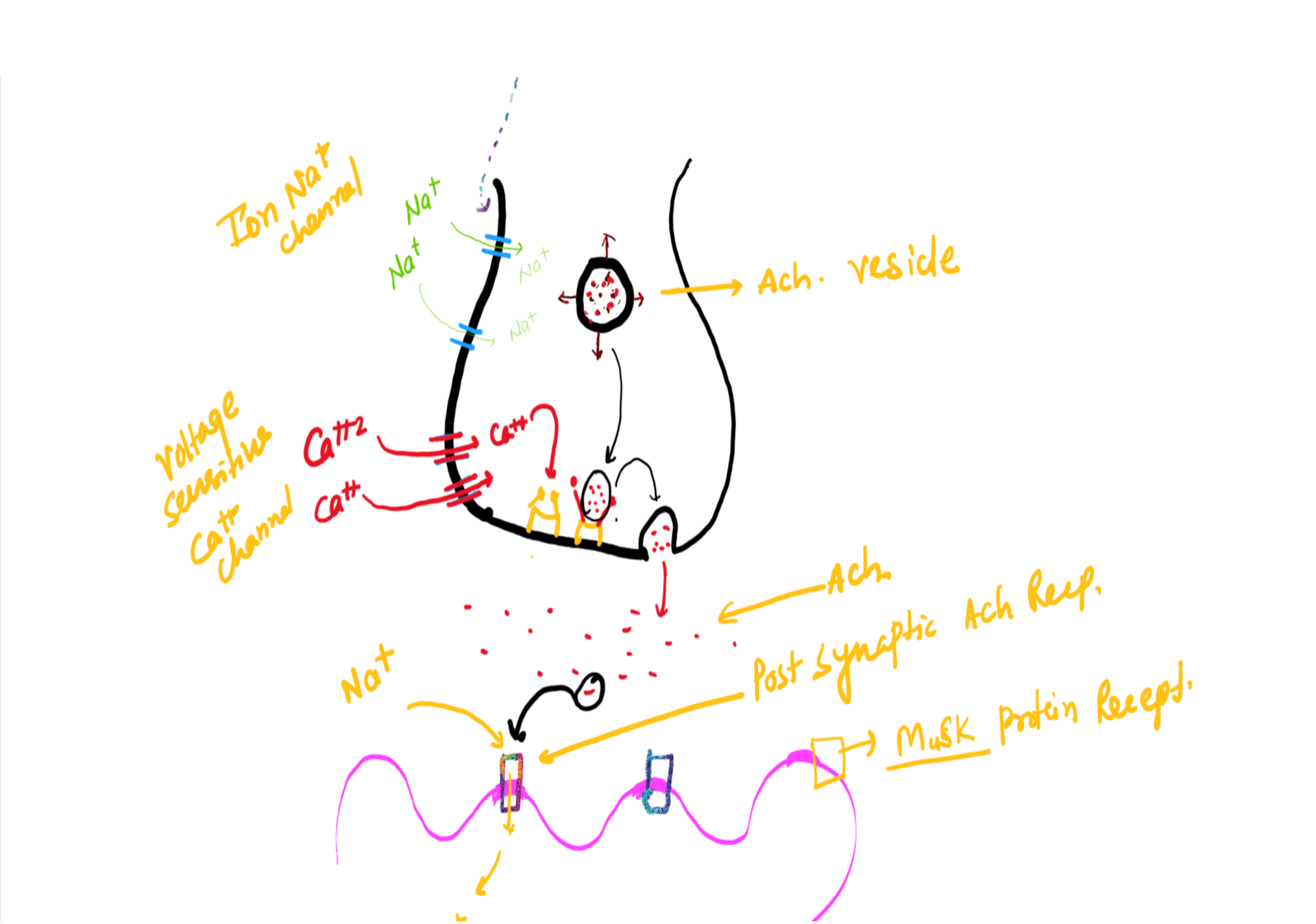
(End plate potential)

↓

Voltage Sensitive Na^+ channel open

↓

Depolarization of muscle membrane.

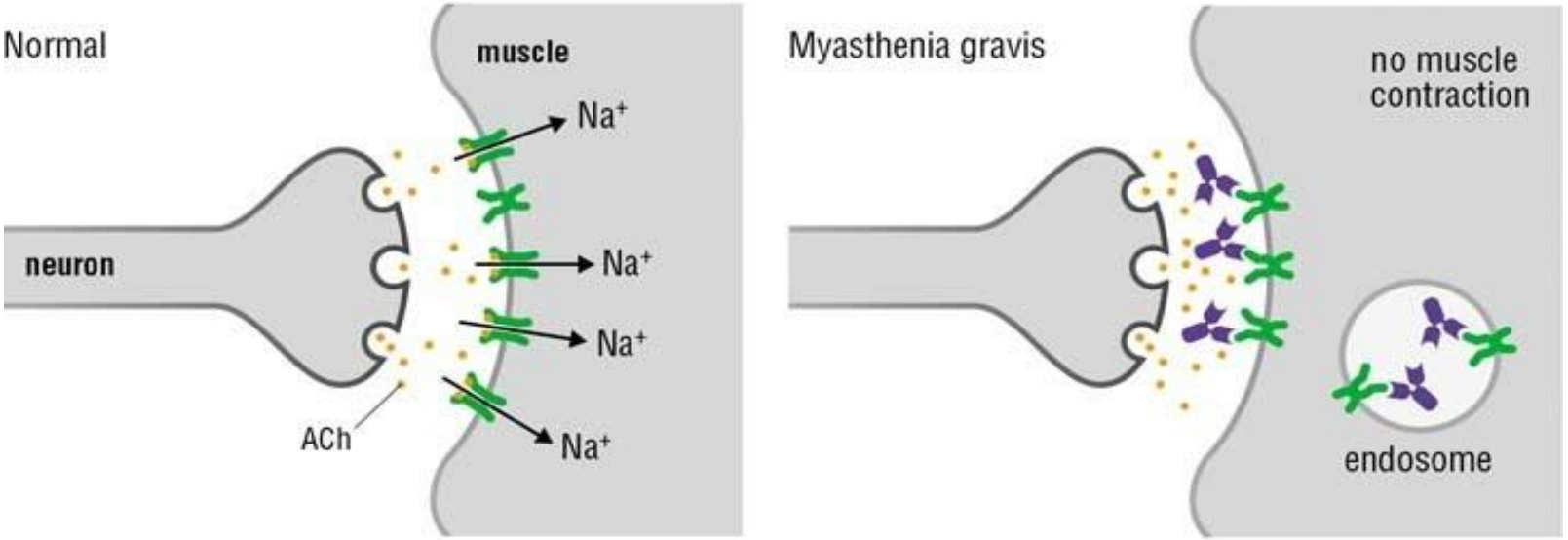

Ion Na+ channel
Na+
Na+
Na+
Na+
Ach. vesicle
Voltage sensitive Ca++ channel
Ca++2
Ca++
Ca++
Ach.
Post Synaptic Ach Recp.
Na+
MuSK protein Recepd.

Pathophysiology

- Antibodies directed at the acetylcholine receptor sites impair transmission of impulses across the neuromuscular junction. Therefore, fewer receptors are available for stimulation, resulting in voluntary muscle weakness that escalates with continued activity.

Pathophysiology

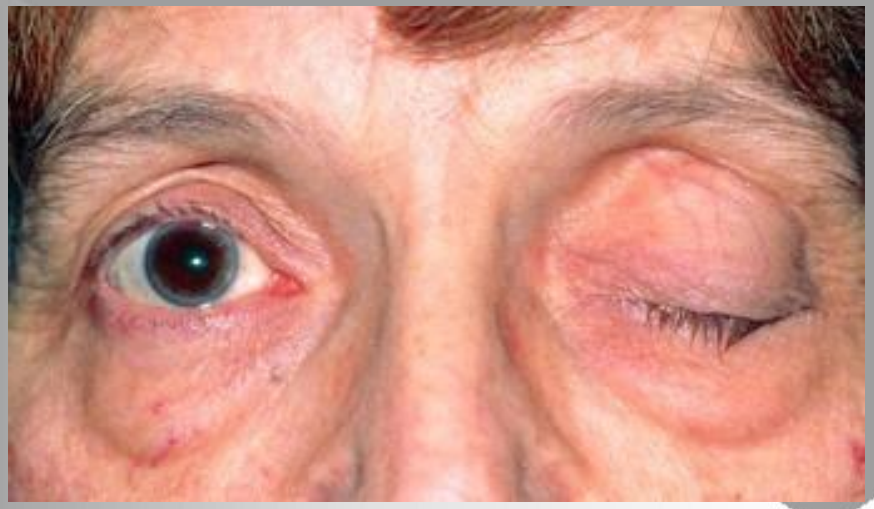
From **Immunity: The Immune Response in Infectious and Inflammatory Disease**
by DeFranco, Locksley and Robertson

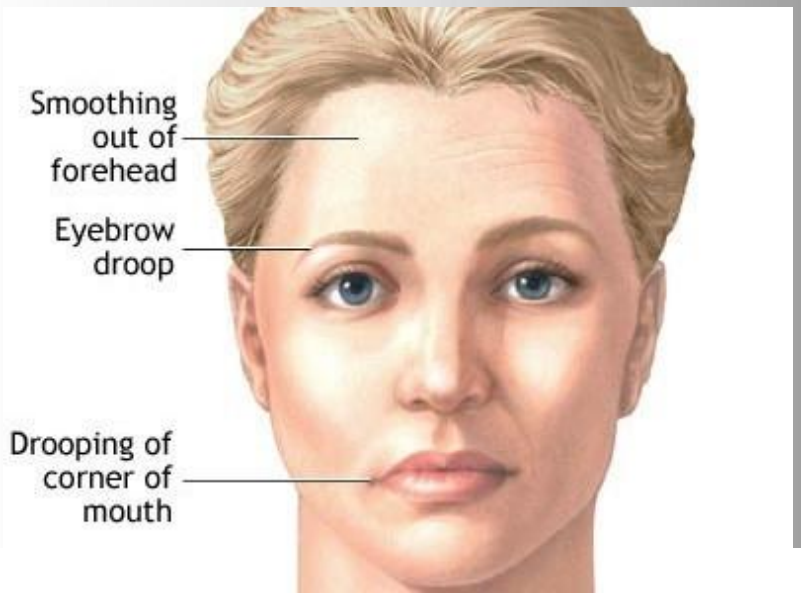


Clinical Manifestations

MG is purely a motor disorder with no effect on sensation or coordination.

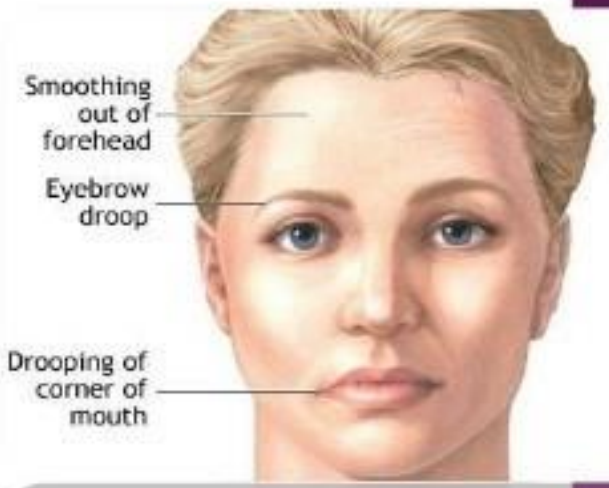
- Initial manifestation involves ocular muscles (eg, diplopia and ptosis)
- Weakness of the muscles of the face (resulting in a bland facial expression) and throat (bulbar symptoms) and generalized weakness
- Laryngeal involvement: dysphonia (voice impairment) and increases the risk of choking and aspiration
- Generalized weakness that affects all extremities and the intercostal muscles, resulting in decreasing vital capacity and respiratory failure




Smoothing out of forehead
Eyebrow droop
Drooping of corner of mouth

SYMPTOMS

- **Eye muscles**
- Drooping of one or both eyelids (ptosis).
- Double vision (diplopia)
- **Face and throat muscles**
- Altered speaking(dyasarthria)
- Difficulty swallowing(dysphagia)
- Problems chewing
- Limited facial expressions
- **Neck and limb muscles**
- Weakness in arms, legs, neck, fingers etc.
- Weakness in the chest muscles sometimes
- occurs. If this is severe, myasthenic crisis
- may result .



Assessment and Diagnostic Findings

- Injection of edrophonium / Tensilon (anticholinesterase) is used to confirm the diagnosis (have atropine available for side effects). Improvement in muscle strength represents a positive test and usually confirms the diagnosis.
- MRI may demonstrate an enlarged thymus gland.
- Tests include serum analysis for acetylcholine receptor and electromyography (EMG) to measure electrical potential of muscle cells.

Medical Management

- Management of MG is directed at improving function and reducing and removing circulating antibodies. Therapeutic modalities include administration of
  - anticholinesterase medications
  - Corticosteroids
  - immunosuppressive therapy,
  - plasmapheresis,
  - thymectomy.(since the presence of thymus gland in MG patients appears to enhance the production of ACh Receptor Antibodies .)

Pharmacologic Therapy

1.Anticholinesterase agents

- Neostigmine, Pyridostigmine

2. Corticisteroids

- Prednisolone

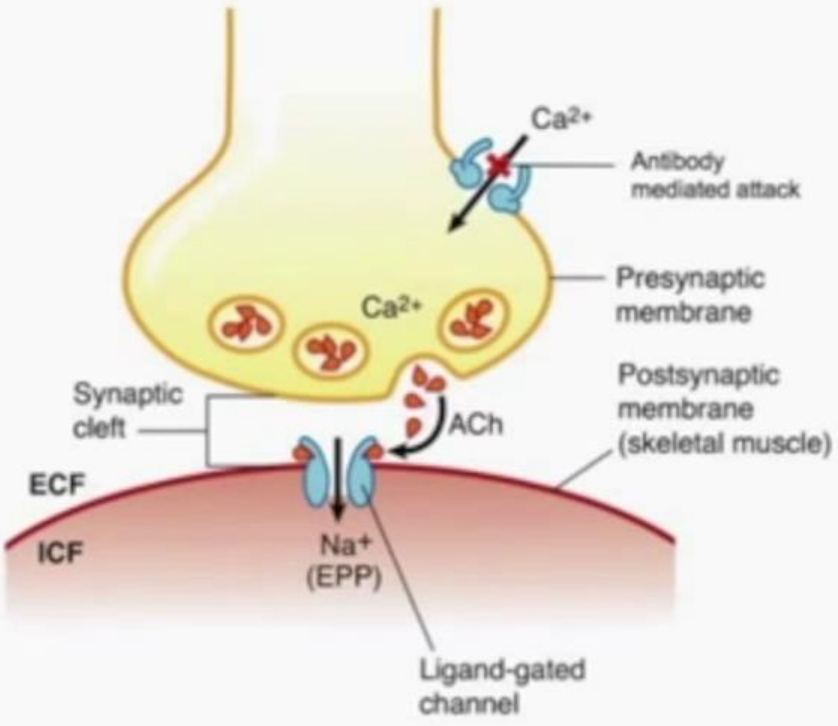
3. Immunosuppressants

- Azathioprine, Cyclophosphamide

LAMBERT EATON SYNDROME

Lambert-Eaton syndrome

- Etiology:
  - Antibodies against the presynaptic calcium channels of the neuromuscular junction
  - Decreased acetylcholine release with neuronal transmission
- Signs/symptoms:
  - Proximal muscle weakness that improves with repeated use
- Other characteristics:
  - Associated with malignancy, occurring as a paraneoplastic syndrome (e.g. small cell lung cancer)



Myositis ossificans

Myositis ossificans describes a heterotopic bone formation within a muscle.
Thereby **myositis ossificans** is classified in two different groups: **myositis ossificans** progressiva (MOP) which describes a genetic autosomal dominant rare disease and **myositis ossificans** traumatica (MOT).

- Generalized myositis ossificans usually occurs in young children or adolescents with the development of soft, fluetuant or firm nodular swellings anywhere on the body but frequently on the neck or back

- usually painless
- trunk and proximal limbs are most frequently involved
- Ultimately, entire groups of muscles become transformed into bone with resulting limitation of movement
- The masseter muscle is often involved so that fixation of the jaw occurs
- The patient becomes transformed into a rigid organism sometimes encountered in circuses as the 'petrified man'.

Histologic Features

- Osteoid formation
- Cartilage formation may also be evident.
- Characteristically, intact muscle fibres may be found within the bony tissue.

Traumatic Myositis Ossificans

- Traumatization of the periosteum of an adjacent bone with the displacement of osteoblasts into the muscle and subsequent formation of bone
- Metaplasia of the pluripotential intermuscular connective tissue into bone
- Metaplasia of fibrocartilage, a normal constituent of many muscle tendons, into bone

Clinical Features

- Myositis ossificans developing after a single acute traumatic injury usually manifests as a firm, painful mass in the injured muscle within 1-4 weeks.
- In some cases, motion is limited by the lesion.
- Chronic cases of myositis ossificans are usually asymptomatic

Oral Manifestations

- Mainly involve masseter and temporal muscles
- Difficulty in opening the mouth

Radiographic Features

- Myositis ossificans may appear either as a feathery type of calcification in muscle, following ossification of a haematoma which dissected along muscle bundles, or as a solitary irregular calcified mass occurring in a simple haematoma
- The radiopaque calcification may be first seen within two to three weeks of the traumatic experience and show a progressive increase in radiodensity

CONGENITAL FACIAL DIPLEGIA (MOBIUS SYNDROME)

A nonfamilial deficient development of cranial muscles consisting of facial diplegia with bilateral paralysis of the ocular muscles, particularly the abducens.

Clinical Features

manifested in infancy during the first few days of life by failure to close the eyes during sleep

Because of the partial or complete facial paralysis, the infant exhibits no change in facial expression even when crying or laughing

- The prominent lips are often everted, and the mouth may remain partially opened
- Difficulty in mastication
- Saliva frequently drools from the corners of the mouth, and speech is severely impaired.
- Deformity of the external ears, deafness
- Defects of the pectoral muscles
- Paresis of the tongue, soft palate or jaw muscles
- Mental defects
- Epilepsy.

REFERNECES

- GRAY'S ANATOMY
- TEXTBOOK OF ORAL SURGERY NEELIMA MALIK
- TEXT BOOK OF ORAL PATHOLOGY SHAFER'S
- TEXTBOOK OF ORAL PATHOLOGY NEVILE
- TEXTBOOK OF LOCAL ANESTHESIA MONHIMS
- TEXTBOOK OF ORAL MEDICINE- ANIL GHOM'S