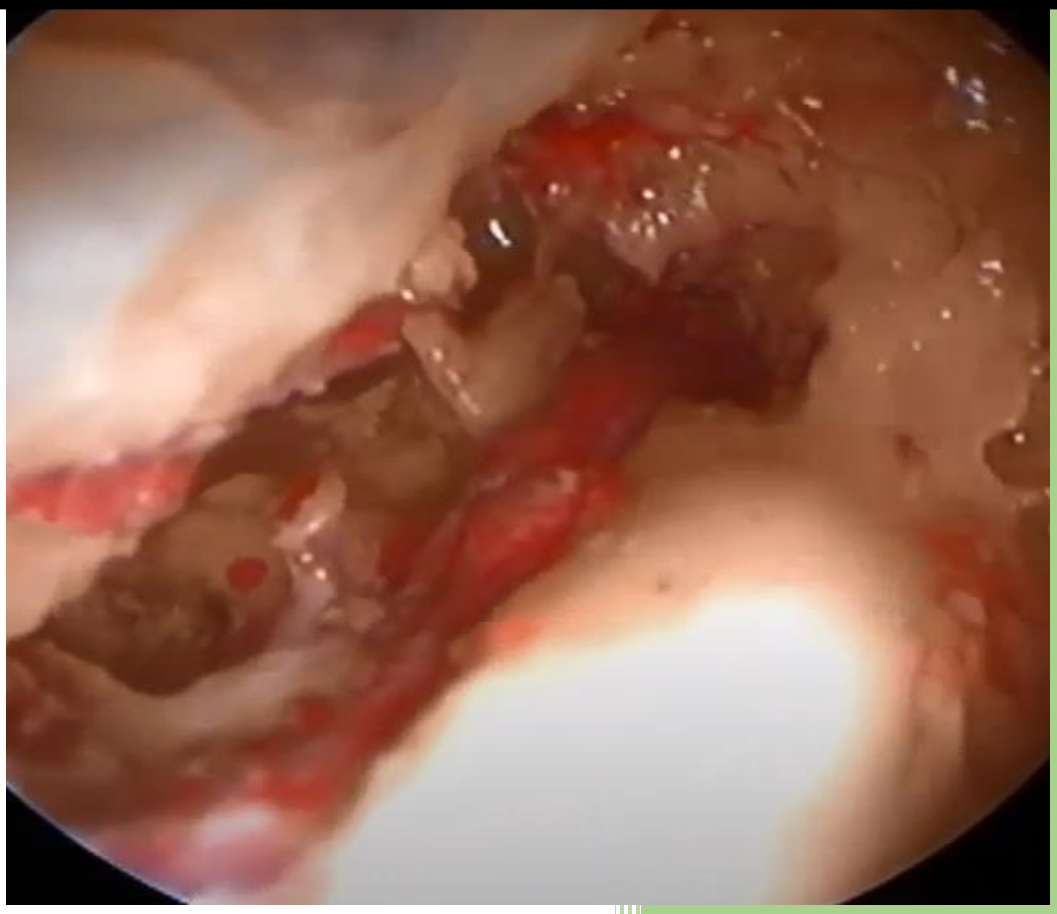
2020

Facial Nerve and Its Disorders



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Preface:

Facial musculature plays an important role in mastication, facial expression, emotions, and speech. Facial musculature is supplied by the facial nerve. Disorders affecting facial nerve would cause facial paralysis causing asymmetry of the face due to paralysis of muscles of the face.

This manuscript discusses clinical anatomy of facial nerve, pathologies affecting this nerve, its clinical manifestations and various management modalities available in managing these conditions. This book has been authored from the perspective of otolaryngologist. Otolaryngologists commonly encounter facial nerve lesions since this nerve courses through the temporal bone. The intratympanic portion of facial nerve comes under the domain of otolaryngologist.

Lesions of facial nerve in this book has been arranged under two headings:

1. Non neoplastic disorders of the facial nerve
2. Neoplastic disorders of the facial

It should be pointed out facial nerve has excellent regenerative capacity and it is never late to attempt decompression / trying out various anastomotic techniques in managing patients with facial nerve paralysis. Rehabilitation process in facial nerve paralysis should focus on managing and protecting the eyes of the patient. In facial paralysis patients are unable to close their eyelids due to paralysis of the muscles. Various rehabilitation procedures have been developed to manage these patients. Imaging of facial nerve has undergone tremendous advancement in recent times. A chapter on imaging of facial nerve has also been added to this manuscript to make it complete.

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Introduction

Facial palsy is a devastating handicap that could occur in patients. The is the foremost complication the otologist and parotid surgeons try to avoid. In addition to medico legal issues involved facial paralysis leaves behind a huge negative impact on the quality of life of the patient.

Currently otological surgery has expanded into the cerebellopontine angle areas. It is thus imperative for the surgeon to be familiar with management of facial nerve in the angle as well as the temporal bone and neck.

Peripheral facial paralysis resulting from affection of the 7th cranial nerve is the most common cranial nerve lesion. Its incidence ranges from 20-30 cases per 100,000 people. Commonest cause for facial nerve paralysis happens to be the idiopathic Bell's palsy. Trauma is the second most frequent cause.

A surgeon should guard against idiopathic causes of facial paralysis which include middle ear surgical procedures, and parotid surgeries. It should also be stressed that a surgeon should not fear the nerve but instead should attempt to use it as a road map during surgeries. With the easy availability of operating microscopes and magnification loups things have become easy for the surgeon. Identification of the location of the nerve becomes easier under good illumination and magnification. In addition, the widespread availability of facial nerve monitors has helped the surgeon in protecting this vital nerve during the surgical procedure.

Facial paralysis in Ramsay Hunt syndrome is caused by reactivation of latent herpes zoster virus in the geniculate ganglion causing acute facial nerve paralysis followed by severe pain and vesicular eruptions in the external auditory canal.

Acute otitis media could present with facial nerve paralysis as a complication. The incidence is rather high in children. This can commonly occur in patients with dehiscent facial nerve (facial nerve within the temporal bone without bony covering).

The degree of paralysis of the nerve has a bearing on the completeness of its recovery. Worse the grade of paralysis at the initial onset poorer is the prognosis. Yet Bell's palsy has an excellent recovery rate (nearly 99%) of these patients recover facial nerve function completely. Even though the diagnosis of Bell's palsy is a diagnosis of exclusion, a surgeon can ideally wait for a minimum period of 6 weeks before taking up the patient for surgical management of facial paralysis.

Electrophysiological examination is very important for the prognosis and serves as an indication for more aggressive therapy. Tests such as Hilger, Electroneuronography and Electromyography are largely used every day.

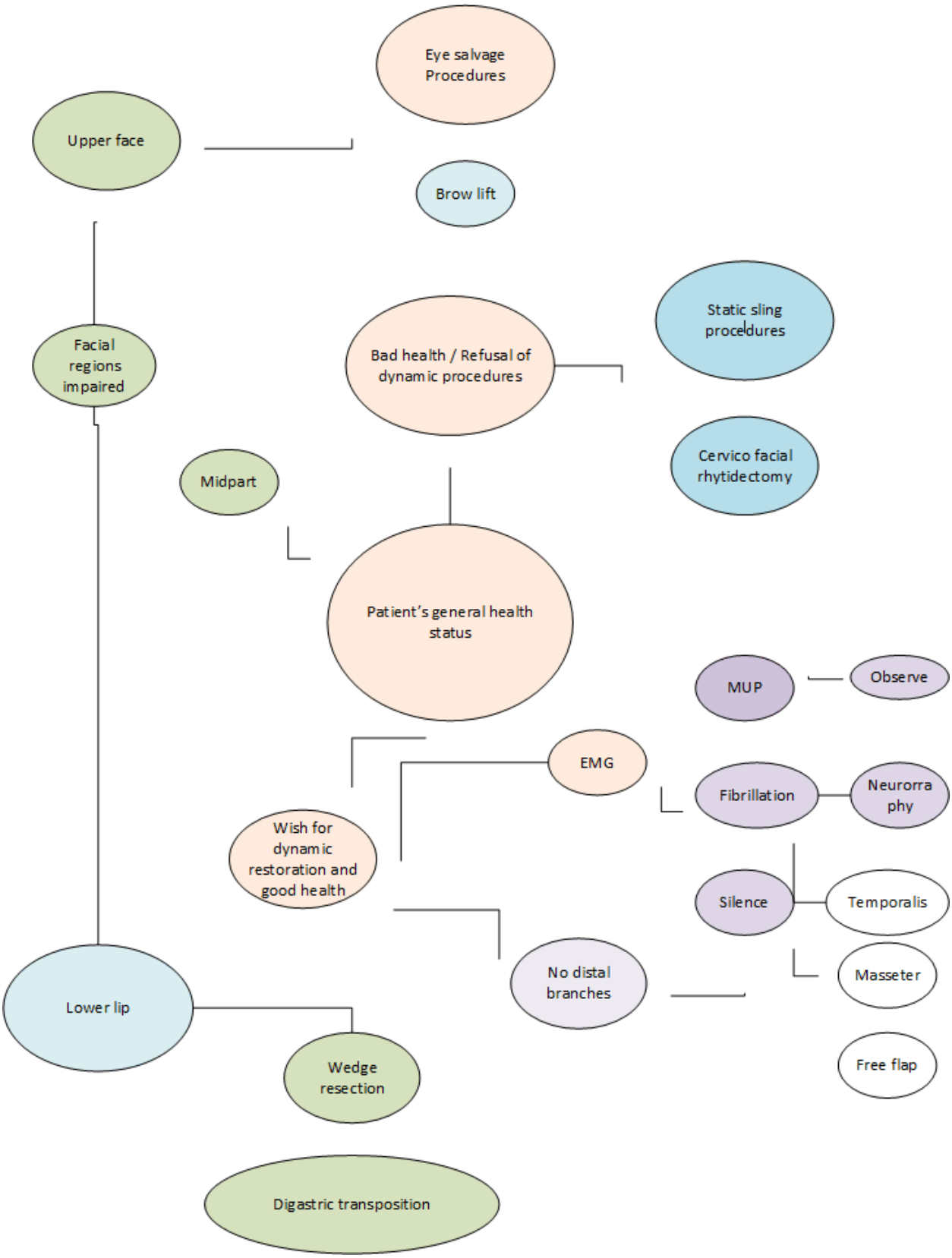

Image showing the algorithm used in the management of facial nerve lesions

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Surgical Anatomy:

Anatomically the course of the facial nerve is rather complex. It has many branches that transmits a combination of sensory, motor and parasympathetic fibers. Cadaveric studies reveal that the total number of myelinated axons ranges between 7000-9000 in the motor root of the facial nerve and between 3000-5000 in the nervus intermedius. Both these nerves also contain unmyelinated axons.

Special visceral / branchial efferent (SVE) motor axons supply muscles derived from the second arch, namely the muscles of facial expression, buccinator, stapedius, platysma, the posterior belly of digastric and stylohyoid. The axons arise from neurons in the facial nuclear complex in the lateral part of the central tegmen of the pons just rostral to the pons medulla transition. It runs posteromedially through the pons, arching over the abducens nerve nucleus to raise the facial colliculus on the floor of the IV ventricle (this could be seen in axial MRI sections), before turning anterolaterally to leave the brain stem.

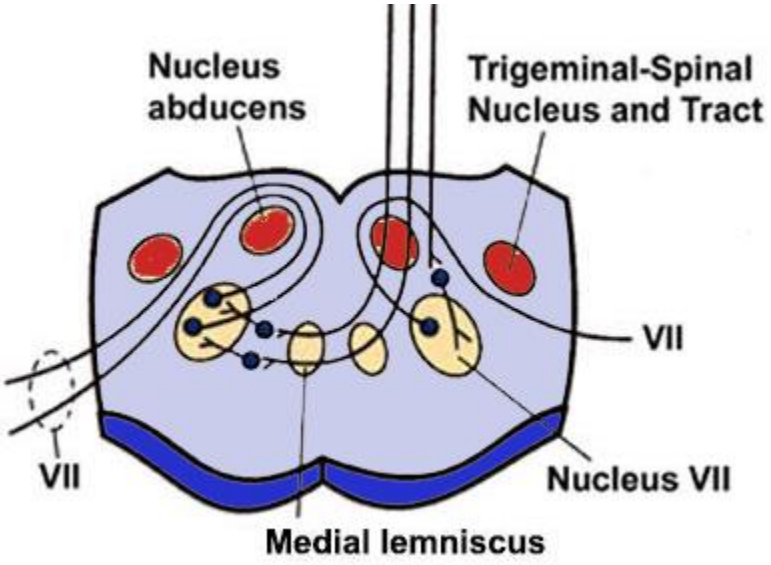
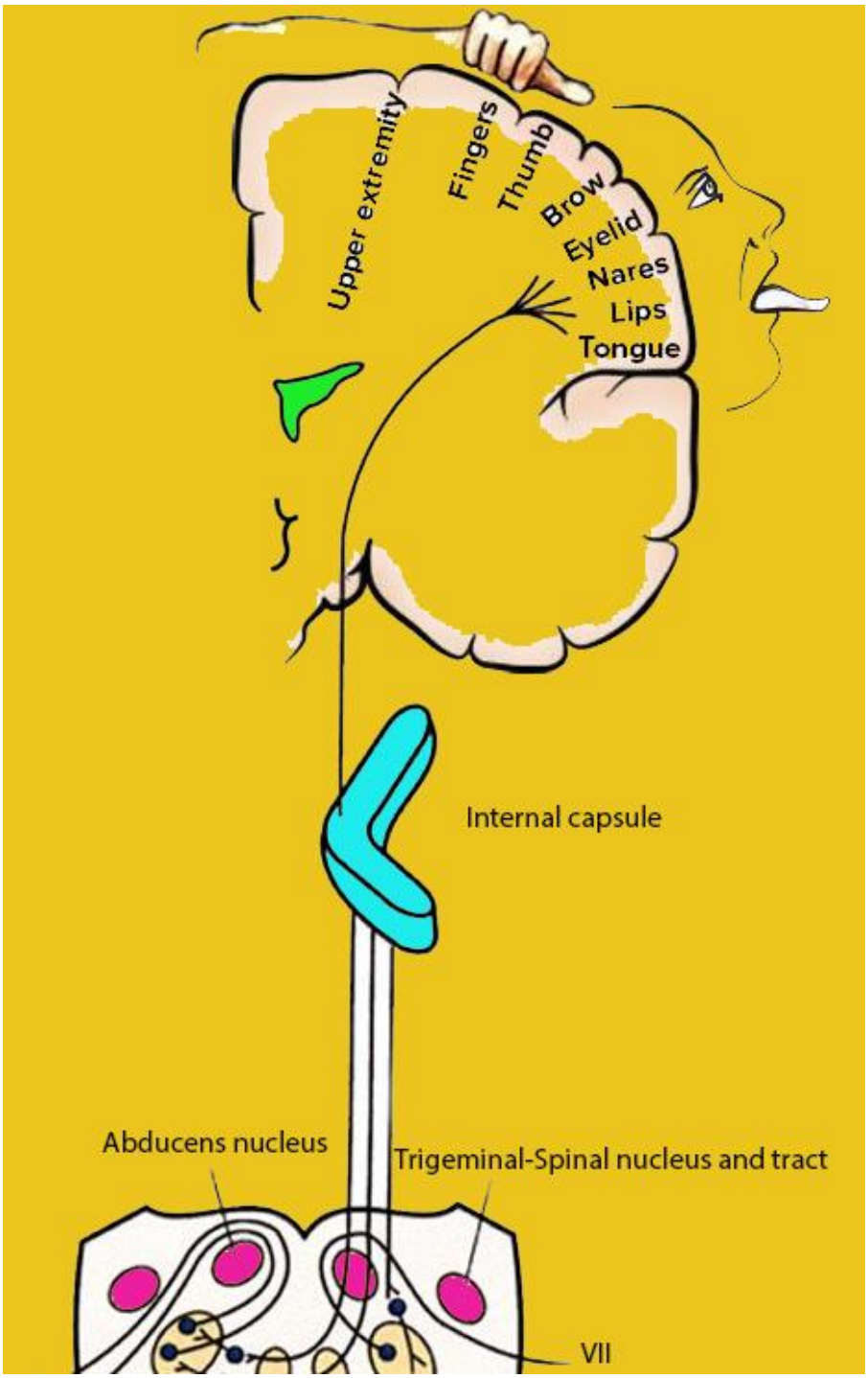


Image showing facial nerve nucleus

Neurons of the face area of the motor cortex (supranuclear neurons) project bilaterally to facial motor neurons that control muscles in the upper face (frontalis, orbicularis oculi) but contralaterally to facial motor neurons that innervate the muscles in the middle and lower face.

Sparing of forehead muscles in upper motor neuron type of facial paralysis does not occur always. Although theoretically sparing of forehead muscles is common in UMN type of facial palsy. Axons supplying orbicularis oculi constitute the efferent limb of the corneal reflex.

Studies also reveal that nerve to stapedius could arise from neurons that lie outside the main facial nerve nucleus. Stapedial reflex is used to test for topographical assessment of facial nerve lesion and to assess cochlear function as it is elicited by a sound impulse of threshold +80 dB.



Facial nerve pathway

As the neurons pass through the anterolateral part of pons the special visceral efferent motor axons are joined by general visceral efferent (preganglionic parasympathetic axons from the neurons in the superior salivary nucleus) which will ultimately synapse on post ganglionic neurons in either the pterygopalatine ganglion or the submandibular ganglion.

The facial nerve contains two / three types of sensory axons. They are:

1. Special visceral afferent (SVA) carrying sensation of taste from anterior two thirds of the tongue via chorda tympani nerve.

2. General sensory afferent (GSA) axons carrying cutaneous sensation which include pain from the posterior aspect of external auditory canal. The innervation of this area is overly complex and is largely contributed by Arnold's nerve which is a branch of auricular branch of vagus nerve. Arnold's nerve is responsible for producing cough during ear wax removal and this is also the site for vesicular eruptions in Ramsay Hunt syndrome. The sensory innervation of this area contributed by facial nerve is responsible for the production of Hitzelberger sign in acoustic neuroma (Hypoesthesia of posterior canal wall).

3. Some authors suggest that nervus intermedius also contains general visceral afferent (GVA) axons mediating pain from the tongue and oropharynx. This is the reason why in some patients with Ramsay Hunt syndrome rashes could be seen in the region of soft palate, in the anterior pillar of tonsils and anterior two thirds of the tongue.

The craniofacial muscles innervated by the facial nerve typically lack muscle spindles and proprioceptive information from these muscles may be mediated by modified cutaneous mechanoreceptors or capsular corpuscle like structures within the muscles.

The facial nerve has intra and extracranial connections with the cutaneous branches of all three divisions of the trigeminal nerve (including branches of the auriculotemporal, buccal, mental, lingual, infraorbital, zygomatic and ophthalmic nerves), with branches of vestibulocochlear, glossopharyngeal and vagus nerves; and with branches of cervical plexus (including great auricular, greater and lesser occipital and transverse cervical nerves).

The cutaneous connections are significant in facilitating the perineural spread of tumors that arise either within the parotid or on the face and explains why some patients with perineural spread from cutaneous or parotid malignancies may present with vocal cord paralysis.

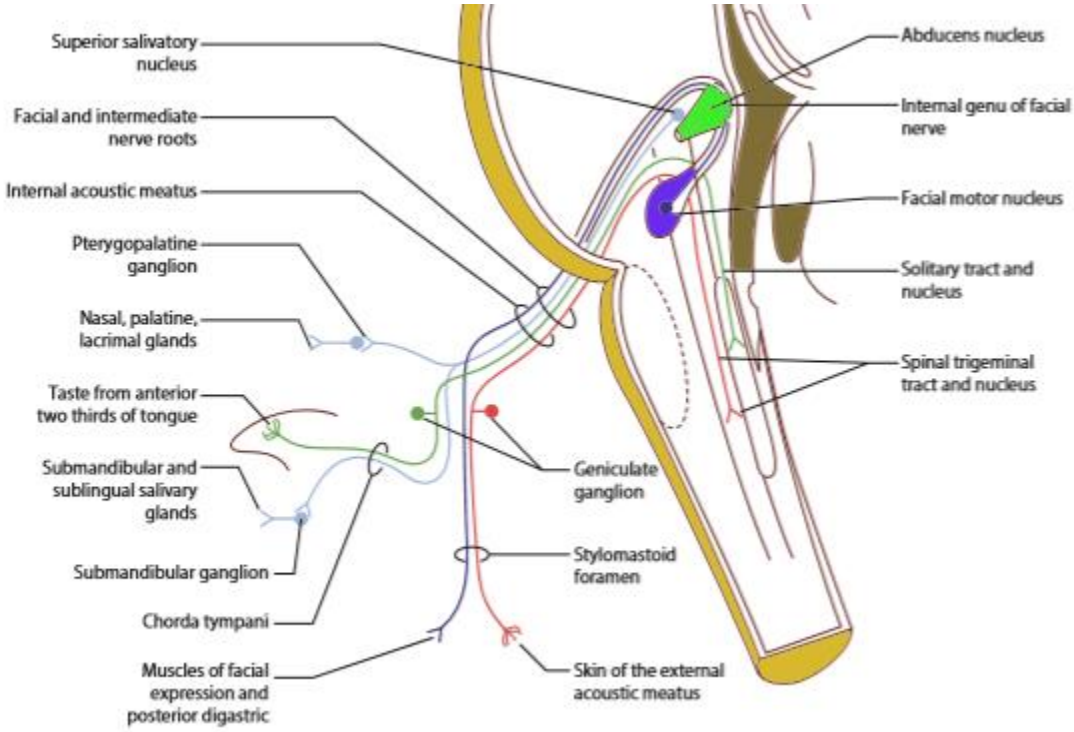


Image showing facial nerve and its branches

The GVE, SVA, GSA and GVA axons from the nervus intermedius of Wrisburg. This nerve lies between the motor root of facial nerve and the vestibulocochlear nerve as it exits the brain stem.

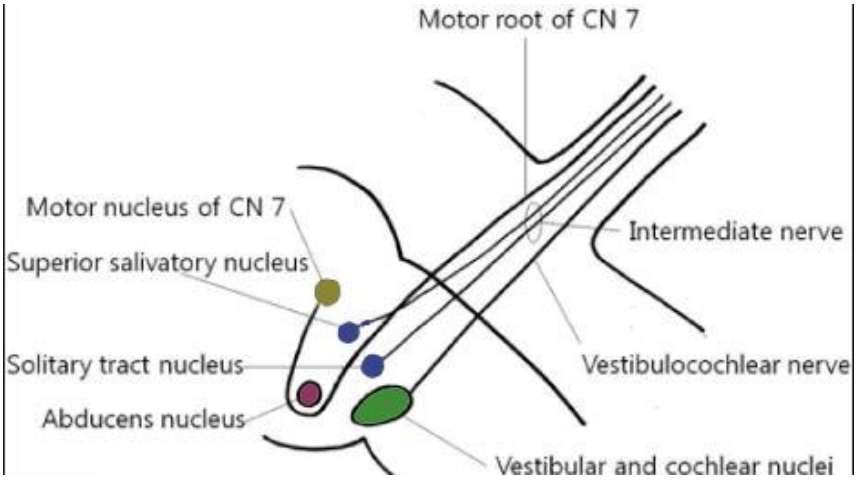


Image showing Nervous intermedius of Wrisburg

The axons of the motor root of the facial nerve and the nervous intermedius intermingle within the substance of pons but emerge separately onto the surface of the brain stem, deep in the pontomedullary sulcus.

In retrosigmoid approach, visualization of the root emerging point is impaired by the lower cranial nerves. The root entry/exit zones (REZ) of the facial nerve and nervus intermedius and the more distal transition zones of these nerves (Obersteiner-Redlich zones) of these nerves, where central myelin (produced by oligodendrocytes) abuts the peripheral myelin (produced by Schwann cells), are both targets in the microvascular decompression for hemifacial spasm.

Surgical landmarks to the REZ on the brainstem are the pontomedullary sulcus; the junction of the glossopharyngeal and vagus nerves with the medulla; the foramen of Luschka and choroid plexus; the cerebellar flocculus (the last three structures are related to the lateral recesses of the fourth ventricle).

The subsequent course of the facial nerve is conventionally divided into intracranial (cisternal), Intratemporal and extratemporal portions.

Cisternal portion of facial nerve:

This portion is about 24 mm long. The motor root of facial nerve and the nervous intermedius course anterolaterally through the CPA cistern to the porus of the internal auditory meatus, accompanied by and usually anterior to the cisternal segment of the vestibulocochlear nerve. The nervous intermedius may adhere to the vestibulocochlear nerve for a variable distance. The nerves are ensheathed only by a delicate layer of arachnoid mater; the absence of more robust connective tissue covering renders them not only extremely vulnerable in CPA tumor surgery but also difficult to identify during dissection of a schwannoma.

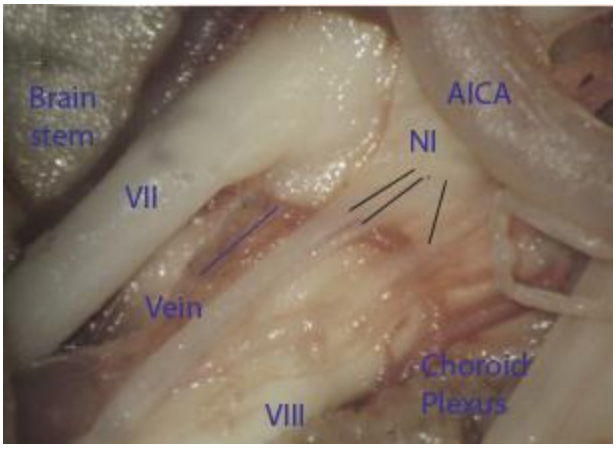


Image showing the cisternal portion of facial nerve

Intratemporal portion:

This portion is about 28-30 mm long and is subdivided into meatal, labyrinthine, tympanic (also known as horizontal) and mastoid (vertical) segments. The meatal segment runs in the internal acoustic meatus; the labyrinthine segment runs laterally to the geniculum (first turn or genu) and the geniculate ganglion; the tympanic segment runs backwards from the geniculate ganglion to the second genu; the vertical segment runs from the second genu to the stylomastoid foramen; the extratemporal segment runs forwards from the stylomastoid foramen into the parotid gland where it usually divides into temporofacial and cervicofacial branches.

The first three segments do not display any obvious fascicular organization, which may explain why a very selective partial lesion in this part of the nerve is likely to produce a slight overall paresis of the face and also why regeneration following injury usually results in some kind of synkinesis (reinnervation of inappropriate facial muscle groups by regenerating facial nerve axons). These proximal segments are invested by arachnoid mater but lack epineural and perineural sheaths, rendering suture technically challenging if not contraindicated in favor of glue.

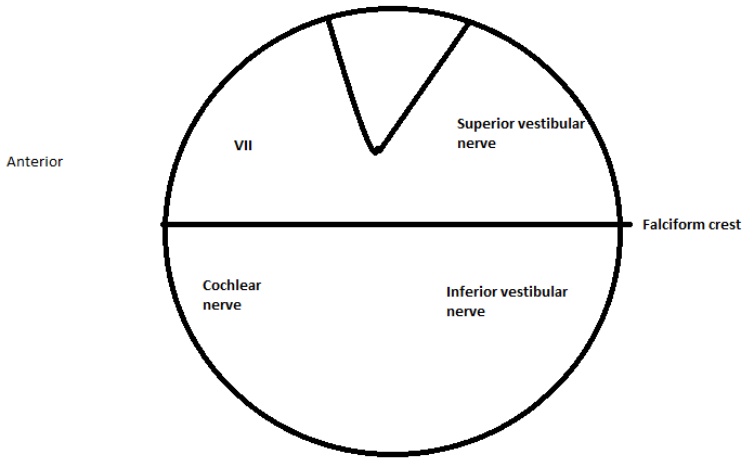
Arachnoid cysts have been reported within the internal acoustic meatus and facial nerve canal, where they are known to compress adjacent nerves. Clearly identifiable fascicles defined by perifascicular, interfascicular and intrafascicular connective tissue only appears distal to the geniculate ganglion. The number of fascicles increases proximodistally as axons are progressively segregated into specific named nerves. Even though the spatial relationship of the fascicles is highly variable, a very selective partial lesion affecting the lower part of the mastoid segment will produce total paralysis of the denervated muscles.

Meatal segment:

This segment is about 5-12 mm long. The motor root of the facial nerve and the nervus intermedius run through the internal acoustic meatus from its porus (medial) to its fundus (lateral wall), accompanied by the cochlear nerve and the superior and inferior vestibular nerves and the labyrinthine artery. The nerve bundles that make up the nervus intermedius join the facial nerve within the Internal auditory meatus, more or less 3 mm from the porus. The components of the vestibulocochlear nerve rotate 90° as they travel from the brainstem to the inner ear. The facial nerve remains anterior in its course to the porus acusticus and within the internal auditory meatus and so lies anterior to the superior vestibular nerve in the lateral end of the meatus.

On cross section the fundus is divided into four quadrants. It is divided horizontally by a thin bony septum, the crista falciformis and vertically into anterior and posterior compartments above the crista by an important landmark, the Bill's bar. The motor root of the facial nerve and the nervus intermedius lie anterior to the Bill's bar in the anterior superior quadrant; the superior vestibular nerve lies posterior to the Bill's bar in the posterior superior quadrant; the cochlear nerve and the inferior vestibular nerve lie below the crista, the cochlear nerve lying anteriorly and the inferior vestibular nerve posteriorly.

The close anatomical relationship between the motor root of the facial nerve, nervus intermedius and vestibulocochlear nerve along their cisternal and meatal portions explains the disturbances in lacrimation, taste, salivary flow, hearing, balance or facial motor control that could result from either the cerebello pontine angle lesions or internal acoustic meatus lesions.



Relationship between facial nerve and other nerves at the level of internal acoustic meatus

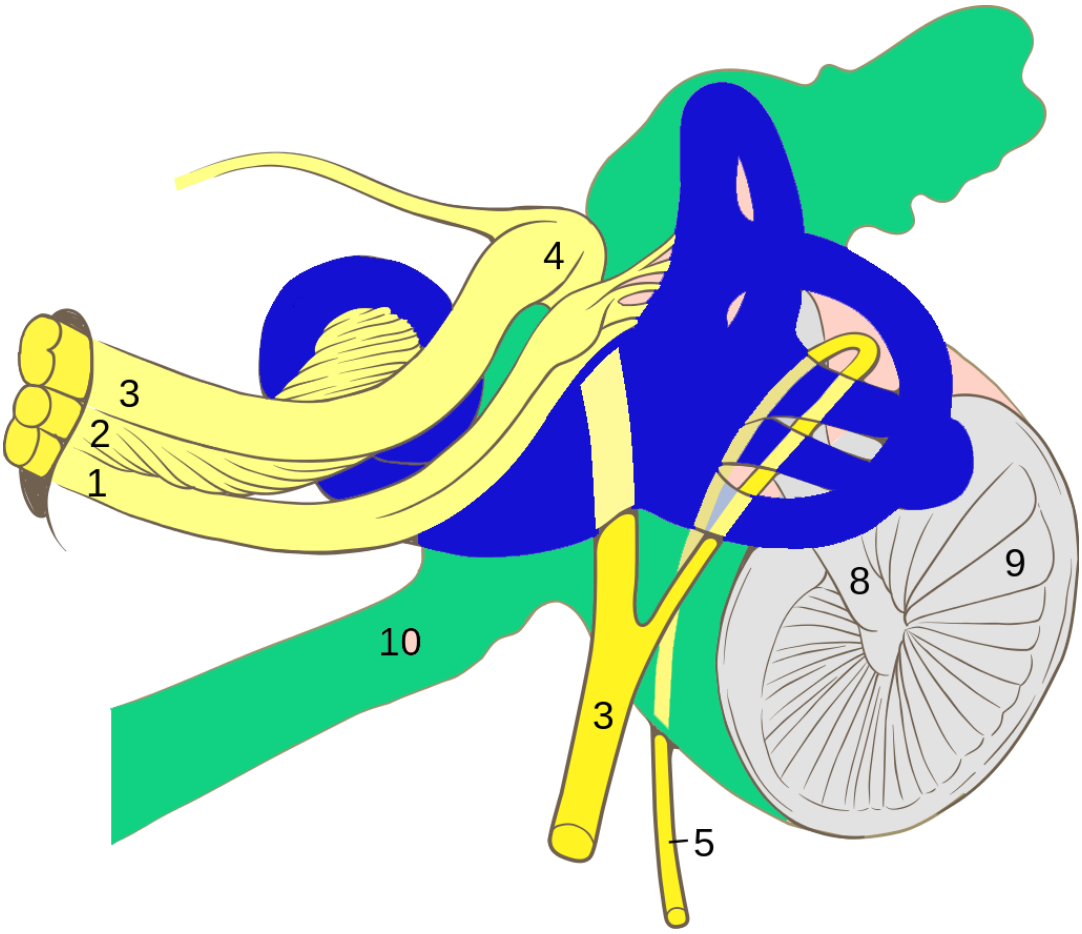
Labyrinthine segment:

This is the shortest segment of the facial nerve (3-5 mm) and the narrowest part of the facial nerve (average diameter of 0.68 mm). It is the only segment of the facial nerve that lacks anastomosing arterial cascades and is hence vulnerable to embolic phenomena, low flow states and vascular compression. This segment is the most common portion to be affected by ischemia in the event of oedema following trauma / inflammation.

At the distal end of the labyrinthine segment, the geniculate ganglion forms part of a sharp hairpin bend, the geniculate or the first genu of the facial nerve. This marks the start of the tympanic segment of the facial nerve. At the geniculum, the nerve is cradled by the superior canal posteriorly and the cochlea anteriorly and inferiorly. The nerve may be accompanied by the subarachnoid space as far as the geniculate ganglion.

Geniculate ganglion:

This is a sensory ganglion. It does not contain synapses. The central processes of GSA (General sensory afferents) neurons in the geniculate ganglion carry pain from the external auditory canal and terminate somatically in the spinal tract of V. The central processes of the SVA (somatic visceral afferents) neurons in the geniculate ganglion that carry taste from the anterior two thirds of the tongue via the corda tympani nerve terminate somatotopically on second order neurons in the gustatory nucleus (rostral end of the tractus solitarius). The chorda tympani nerve Is thought to modulate trigeminal and glossopharyngeal sensitivity.



Parts shown in relation to the facial nerve:

1. Vestibular nerve
2. Cochlear nerve
3. Nervus intermedius
4. Geniculate ganglion
5. Chorda tympani
6. Cochlea
7. Semicircular canal ducts
8. Malleus

9. Tympanic membrane
10. Eustachean tube

Preganglionic parasympathetic axons destined to synapse on post ganglionic neurons in either the pterygopalatine ganglion or submandibular ganglion pass through the geniculate ganglion without synapsing. Those destined for the pterygopalatine ganglion form the greater petrosal nerve, the first branch of the facial nerve. The greater petrosal nerve is an important landmark in directing operative approaches through the middle cranial fossa to the internal acoustic meatus, petrous apex, trigeminal nerve and petrous portion of the internal carotid artery. It runs anteriorly and medially from the geniculate ganglion, receives a branch from the tympanic plexus and traverses a hiatus on the anterior surface of the petrous part of temporal bone to enter the middle cranial fossa, where it runs forwards in a groove on the bone above the lesser petrosal nerve and then passes beneath the trigeminal ganglion to reach the foramen lacerum. Here it is joined by the deep petrosal nerve which carries post ganglionic sympathetic axons from the internal carotid sympathetic plexus, to become the nerve of the pterygoid canal (Vidian nerve) destined for the pterygopalatine ganglion.

The geniculate ganglion lies in a fossa covered by a very thin layer of bone that separates it from the floor of the middle cranial fossa. Dehiscences in this area are rather common. When this area is dehiscent, they could render the nerve vulnerable during middle fossa surgery and bring it into direct contact with the meninges. The geniculate ganglion may be injured by bony spicules or directly contused by a shock wave. An enlarged fossa on temporal bone CT strengthens a diagnosis of geniculate ganglion fossa fracture in patients with traumatic facial nerve paralysis.

During a translabyrinthine surgery the labyrinthine segment is at risk while drilling along the superior semicircular canal. The ampullated ends of the superior and the lateral canals (the cat's eyes) should be identified; the facial nerve lies just anterior to this area. The labyrinthine segment of the facial nerve is most likely to be injured in temporal bone fractures when it is likely to be compressed by bony fragments.

Tympanic segment:

This segment is about 8-11 mm long. It runs the length of the superior edge of the medial wall of the tympanic cavity, perpendicular to the long axis of the petrous portion of the temporal bone, taking a posterior path that inclines downwards and laterally from the geniculate ganglion to the second genu at the level of pyramidal eminence.

Proximally it passes just above and medial to the posterior edge of the cochleariform process and the tendon of tensor tympani. The processes cochleariformis is a reliable landmark for locating the tympanic segment of the facial nerve because it is not destroyed by the disease processes involving the middle ear including cholesteatoma.

Distally the nerve lies above the oval window niche, just anterior and inferior to the prominence of the lateral semicircular canal. This relationship with the prominence of the lateral canal is also constant. It

bends at the second genu to enter the bony mass of the styloid complex. The pyramidal eminence is a useful landmark for the second genu of the facial nerve. The retro tympanum contains several small sinuses around the facial canal. The sinus tympani lie medial and anterior to the facial canal and the facial and lateral tympanic sinuses lie lateral and posterior to the canal.

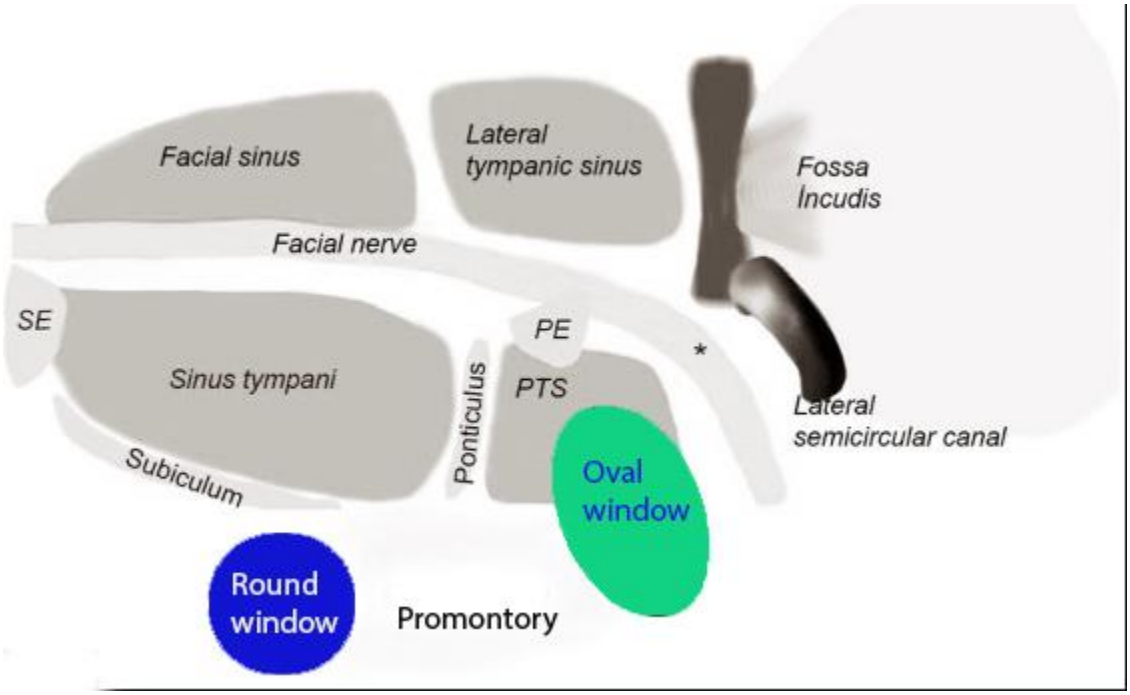
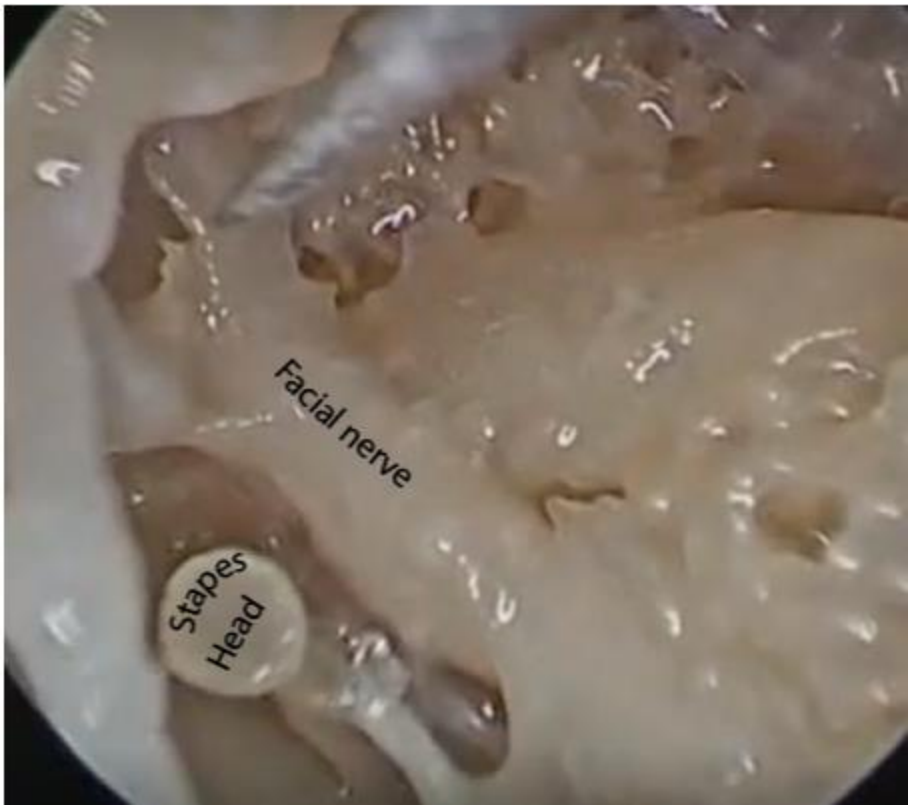


Image showing tympanic segment of facial nerve



Facial nerve seen above the oval window

The distal aspect of the tympanic segment of the facial nerve can be located surgically via a facial recess approach. The second genu of the facial nerve runs inferolateral to the lateral canal (this is a constant relationship). The posterior canal lies just posterior to the second genu; it also marks the superior end of the retrofacial air cells, which are helpful in delineating the medial aspect of the facial canal.

Developmental Dehiscences are common at this level hence the nerve is vulnerable during middle ear surgery especially around the oval window niche. A dehiscent or prolapsed facial nerve should always be anticipated in the tympanic segment especially in patients with congenital deformities. This segment is also vulnerable to traction along its axis in longitudinal fractures of the temporal bone.

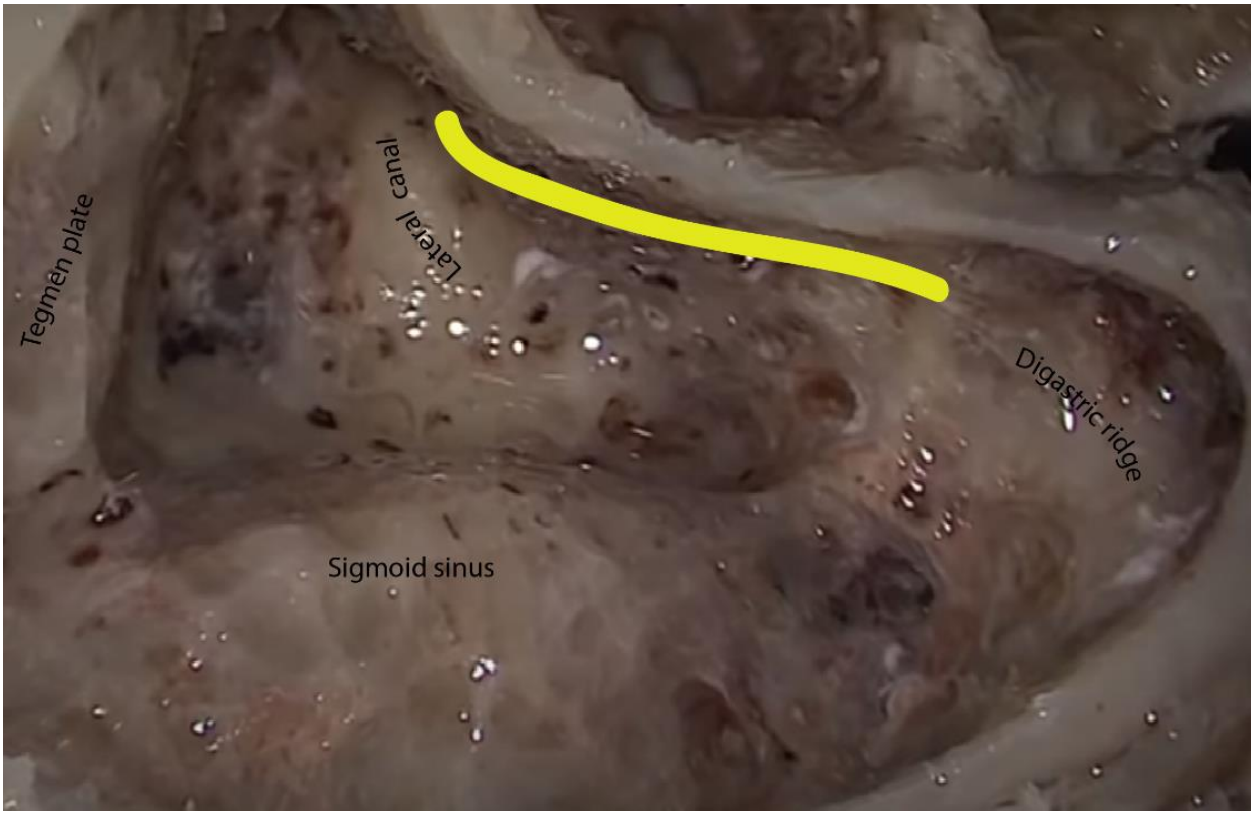


Image showing the relationship of facial nerve (Yellow) with the digastric ridge

Mastoid segment:

The mastoid also known as the vertical segment of the facial nerve is the longest of the petrous segments (10-14 mm). It runs vertically downwards in the posterior wall of the tympanic cavity and the anterior wall of the mastoid, from the second genu just distal to the pyramidal process to the stylomastoid foramen on the skull base. The digastric ridge (seen in well pneumatized bones) as the medial aspect of the mastoid tip and it points to the lateral and inferior aspect of the mastoid segment. A line drawn between the anterior end of digastric ridge and the tip of the short process of the incus marks the path of the mastoid segment. The nerve can easily be identified by removing the mastoid tip, skeletonizing the digastric ridge and outlining the inferior part of the vertical segment of the facial nerve. This procedure could be of help when the extracranial trunk of a functioning nerve is encased in benign tumor recurrences at the stylomastoid foramen.

The nerve travels from the second genu posteromedially to the stylomastoid to the stylomastoid foramen anterolaterally, which means that expanding a posterior tympanotomy inferiorly in the same plane as it was fashioned could risk injury to the facial nerve. The plane of the facial nerve is almost always lateral to the level of the tympanic annulus, but it may cross it, or lie medial to it in its lower half.

The nerve may be damaged when elevating the tympanic annulus or when lowering the mastoid ridge overlying the nerve, as in a modified radical mastoidectomy.

It should be remembered that most commonly, iatrogenic injury to facial nerve is caused by failure to identify the mastoid antrum properly or identification of antrum is impeded by an overhanging tegmen plate. It should also be remembered that the positions of both the tympanic annulus and the mastoid segment of the facial nerve change during childhood.

The mastoid division of facial nerve has three branches. They include:

1. Nerve to stapedius - Supplying the stapedius muscle. The nerve to stapedius is a small twig given off behind the pyramidal eminence that passes forwards through a small canal to reach the muscle.

2. Chorda tympani nerve

3. Sensory auricular branch

Chorda tympani nerve:

The chorda tympani nerve is a branch of facial nerve. It exits the facial nerve just before it exits via the stylomastoid foramen. It is one of the three cranial nerves that is involved in transmission of taste fibers from the tongue. Chorda tympani nerve conveys taste fibers from the anterior 2/3 of tongue. Mechanism of taste sensation is rather unique in that it involves a complicated feedback loop with each nerve acting to inhibit the signal of other nerves. The chorda tympani exert strong inhibitory influence on other taste fibers as well as pain fibers from the tongue. When chorda tympani nerve is damaged its inhibitory function is disrupted, causing the other taste fibers to act in an uninhibited manner.

The chorda tympani nerve carries with it two types of fibers which traverse via lingual nerve to reach their destination. These fibers include:

Special sensory fibers providing taste sensation from anterior 2/3 of tongue.

Presynaptic parasympathetic fibers to submandibular ganglion providing secretomotor fibers to submandibular and sublingual salivary glands

Presynaptic parasympathetic fibers is also supplies the blood vessels of the tongue. When stimulated the chorda tympani nerve causes dilatation of blood vessels of the tongue.

Central connections:

Chorda tympani nerve contains fibers from two brain stem nuclei. They are:

Superior salivatory nucleus: This nucleus houses cell bodies of secretomotor preganglionic parasympathetic neurons

Nucleus of tractus solitarius: The superior portion contributes to chorda tympani fibers. It receives central processes of taste neurons which have their cell bodies in the ganglia of the three cranial nerves conveying taste sensation. After synapsing in this nucleus secondary axons ascend in the lateral lemniscus to relay in the thalamus. This pathway then passes through the posterior limb of internal capsule to reach the primary gustatory cortex.

Connections seen within the facial canal:

Sensory branch of facial nerve Nervus intermedius of Wrisburg joins the facial nerve here. It conveys special sensory fibers from taste buds present in the anterior 2/3 of tongue and soft palate. It also contains secretomotor fibers to salivary glands present below the oral cavity. Nerves intermedius exits the brain stem adherent to the vestibulo cochlear nerve. At the level of internal auditory meatus, it leaves this nerve to merge with that of facial nerve.

Chorda tympani nerve exits from the facial nerve before the facial nerve exits via the stylomastoid foramen. It is the largest branch of facial nerve in its intrapetrous compartment. It arises below the nerve to stapedius. It traverses antero superiorly via the posterior canaliculus usually accompanied by posterior tympanic branch of stylomastoid artery. This canaliculus opens into the middle ear cavity through an aperture situated at the junction of posterior and lateral walls of tympanic cavity. This opening lies just medial to the fibrocartilagenous annulus. The posterior canaliculus is roughly 0.5 mm in diameter. Chorda tympani nerve shows many variations. In some patients the chorda tympani nerve may arise from more proximal portion of facial nerve, even close to the geniculate ganglion. The length of the posterior canaliculus is also highly variable ranging from 3 – 14 mm. In 10% of individuals there may not be a posterior canaliculus at all but could be replaced by a groove.

If the chorda tympani nerve originates outside the temporal bone, then the posterior canaliculus will be separate from that of the facial nerve canal. In fetus and young infants, the chorda tympani nerve leaves the facial nerve outside the skull, but the postnatal growth of mastoid process causes it to migrate to a

more proximal position. Since the facial canal grows more than the mastoid segment of facial nerve the chorda tympani nerve typically diverges from the facial nerve in an infant of 1 yr of age.

Course of chorda tympani in the tympanum:

The chorda tympani arches across pars flaccida medial to the upper part of the handle of malleus and traverses above the insertion of tensor tympani. In patients with congenital anomalies of malleus chorda is also displaced laterally.

Chorda tympani nerve exits the middle ear via a separate bony canal, the anterior canaliculus also known as the canal of Hugier. This canal runs in the medial portion of petrotympanic fissure. Anterior tympanic branch of maxillary artery accompanies this nerve along this canal. Chorda exits the skull through a small foramen behind the base of spine of the sphenoid. At its exit it is closely related to the medial surface of temporomandibular joint.

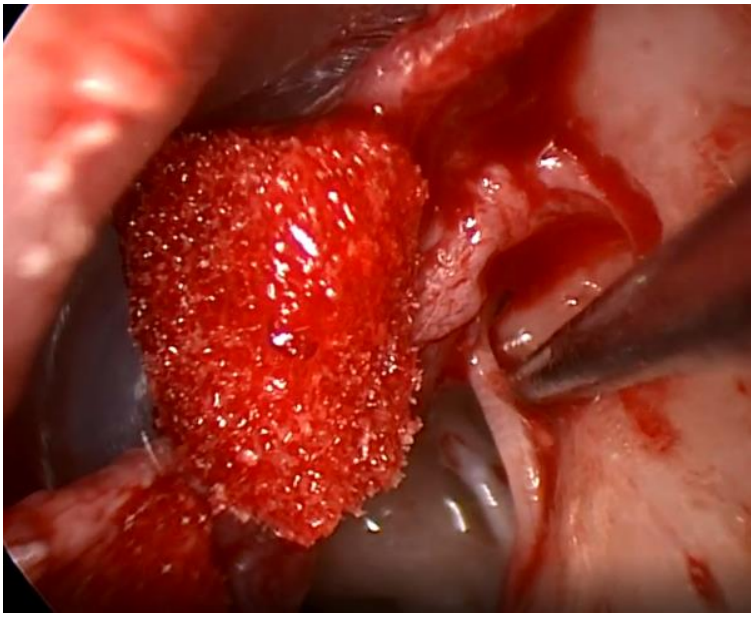
Course of Chorda in the infratemporal fossa:

In the infratemporal fossa the chorda tympani nerve descends medial to the spine of sphenoid and angles forward to join the lingual nerve about 2 cms below the skull base. This junction lies close to the lower border of lateral pterygoid muscle.

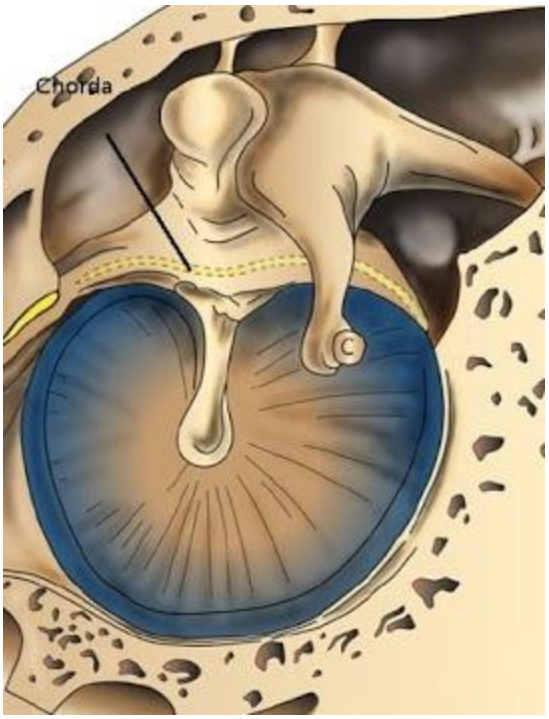
Rarely this nerve could arise extratemporally.

Functions of chorda tympani nerve:

1. It carries taste sensation from the anterior 2/3 of tongue
2. Supplies secretomotor fibers to the salivary glands in the floor of the mouth
3. Conveys general sensation from the anterior 2/3 of tongue which includes pain and temperature
4. Supplies secretomotor fibers to the parotid gland
5. Supplies efferent vasodilator fibers to the tongue



Chorda tympani nerve



Anatomy of chorda tympani nerve

The facial nerve is surrounded by thick fibrous tissue at the stylomastoid foramen. To avoid irrevocable damage to the nerve during anterior transposition of the facial nerve it is advisable to lift the segment enbloc together with the posterior belly of digastric and slivers of bone around the stylomastoid foramen. On leaving the stylomastoid foramen, the facial nerve initially lies below the tympanic plate, lateral to the bases of styloid process and the carotid sheath and posterior to the parotid gland. It immediately gives off the posterior auricular nerve which passes upwards behind the ear between the parotid gland and the anterior border of the sternomastoid muscle and then in the notch between the external auditory canal and the mastoid process to supply the occipital belly of the occipitofrontalis, auricularis superior and the intrinsic auricular muscles. A second muscular branch divides to supply the posterior belly of digastric and the stylohyoid muscle.

Extracranial segment:

The main trunk of the extracranial facial nerve enters the parotid gland high up on its posteromedial surface and passes forwards and downwards behind the ramus of mandible. It divides within the gland, usually just behind and superficial to the retromandibular vein and external carotid artery into the upper temporofacial and a lower cervicofacial trunk. The relationship between the nerve and the vein could vary.

Th length of the facial nerve trunk from the stylomastoid foramen to the initial intraparotid bifurcation ranges between 8 - 22 mm. The temporofacial trunk contains more axons but fewer fascicles than the cervicofacial trunk. the trunks branch further to form a parotid plexus (pes anserinus) from which five main branches emerge. They are:

1. Temporal

2. Zygomatic

3. Buccal

4. Marginal mandibular

5. Cervical

There are considerable number of variations in branching patterns and anastomosis between branches within the parotid and on the face. Branches of facial nerve within the parotid gland are plexiform in nature and can tolerate surgical injury than any other branches of the facial nerve. Accidental division or deliberate sacrifice of a small branch in a plexiform nerve is rarely accompanied by a significant or noticeable facial weakness.

Injury to facial nerve is the most significant complication following parotidectomy. Intraoperative facial nerve monitoring will help in protecting the nerve. A number of landmarks can be used as an identifier of the nerve. They include:

1. Tragal pointer - This is a cartilaginous portion of the tragal cartilage pointing towards the location of the facial nerve. The nerve lies 1 cm antero-infero-medial to the pointer.

2. Tympanomastoid suture line - Facial nerve lies 6-8 mm deep to this suture line

3. Groove between mastoid and the bony external auditory canal - This is bisected by facial nerve

4. Styloid process - Facial nerve lies lateral to the styloid process

5. Posterior belly of digastric - Facial nerve lies superior and parallel to the posterior belly of digastric muscle

Blood supply:

Arterial supply is derived from branches of the vertebrobasilar and external carotid arterial systems. The labyrinthine artery supplies the cisternal, meatal and labyrinthine segments. It usually arises directly from the anterior inferior cerebellar artery as it loops between the cisternal segments of the motor root of the facial nerve, the nervus intermedius and the vestibulocochlear nerves projecting towards and often into the internal auditory meatus. This vessel could also arise from the basilar, vertebral or superior cerebellar arteries. The greater petrosal nerve is supplied by the petrosal branch of the middle meningeal artery which usually passes through the bone enclosing the geniculate ganglion and tympanic segment of the facial nerve. The tympanic and mastoid segments are supplied by the facial arch an anastomotic network formed by the superficial petrosal branch of the middle meningeal artery and the stylomastoid branch of either the occipital or posterior auricular arteries which enters the facial canal via the stylomastoid foramen.

The lowest branch from the stylomastoid artery to the facial nerve is given off at the level of the origin of the chorda tympani nerve. The posterior auricular and the occipital arteries and their branches including the stylomastoid artery supply the facial nerve from the stylomastoid foramen to the parotid gland. Pesanserinus portion of the facial nerve within the parotid gland is supplied by collaterals of the superficial temporal, transverse facial, facial and maxillary arteries.

The labyrinthine portion of the facial nerve is supplied exclusively by the meatal arteries and hence more likely to suffer from ischemic damage than the other segments

Facial nerve injury Pathophysiology

Nerves both peripheral and cranial may be injured in many ways. These injuries can be classified under the following headings:

1. Trauma – can be blunt / penetrating

2. Compression

3. Traction – Chronic / acute stretch

4. Local chemical / freeze injury

5. Systemic causes like immune mediated inflammation, diabetes mellitus, vasculitis and drug induced

6. Ischemia induced nerve damage – This could be caused by mild pressure and initially it could cause transient paresthesia with no obvious structural changes. Recovery is usually rapid and surgical intervention is not needed. Prolonged ischemia / immune mediated insult to the nerve can cause loss of myelin (primary / secondary demyelination), without associated axonal loss of integrity. In this condition the refractory period of nerve conduction is increased with resultant slowing of the conduction mechanism which is followed by total conduction block. If the probable causative agent (a spicule of bone or hematoma is removed) remyelination takes place within 2-4 months. There is a minor residual loss of function, and specific surgical intervention like nerve anastomosis is not needed. On the other hand, any injury that physically separates the axon from its neuronal body triggers a program of molecular and cellular events at the site of the lesion as well as in distant parts of injured neurons and their target organs.

Therapeutic intervention cannot manipulate responses that happen many centimeters from the injury and that may influence functional outcome months or even years later. Centrally, nerve injury induces reprogramming of synaptic connectivity in the cortex and spinal cord as well as responses in affected sensory ganglia that could mediate the development and maintenance of chronic neuropathic pain. Variable number of neurons will die when injury isolates them from their supply of retrogradely transported neurotrophins, particularly if the insult is close to the neuronal cell body. Peripherally, atrophy of chronically denervated sensory end organs and muscles could preclude their reinnervation.

Wallerian degeneration:

This term specifically refers to the process of degeneration that takes place along a nerve distal to an injury and is named after Augustus Waller who first described this process while studying denervated glossopharyngeal / hypoglossal nerves in frogs. The environment of an acutely denervated distal stump facilitates axonal regrowth because it provides a vascularized segment of longitudinally oriented, laminin rich basal lamina tubes filled with axon responsive Schwann cells. Axonal regrowth process is however impeded by progressive endoneural fibrosis and Schwann cell senescence. " Time is Muscle" as per this aphormism early nerve repair really helps in the process of nerve healing. Surgical intervention involves direct coadaptation of stumps by gluing / suturing; bridging the long interstump gap by either grafting a segment of nerve using nerve grafting techniques using nerves harvested from other areas. It should be stressed that mitigation of endoneural fibrosis, muscle and sensory end organ atrophy and Schwann cell senescence remain key clinical challenges.

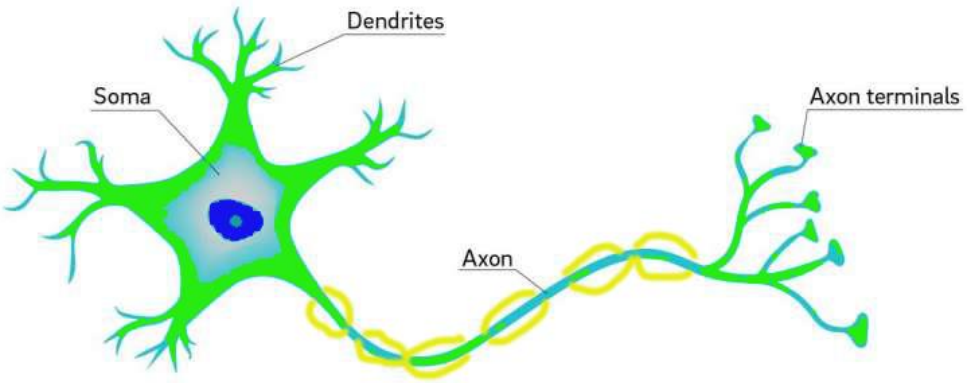


Image of a Neuron

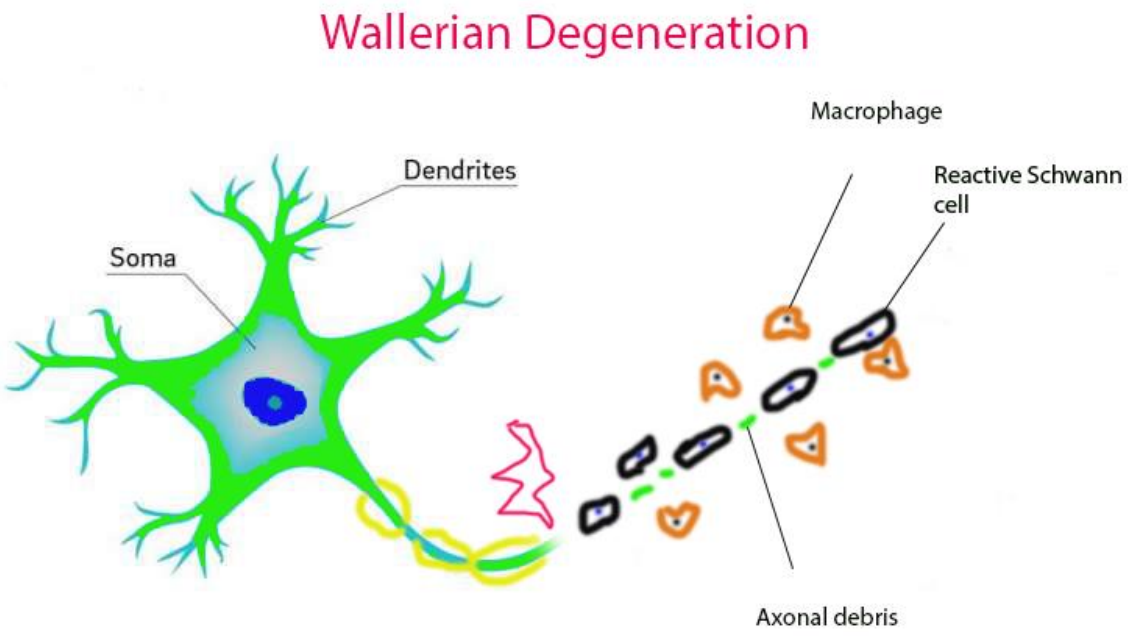
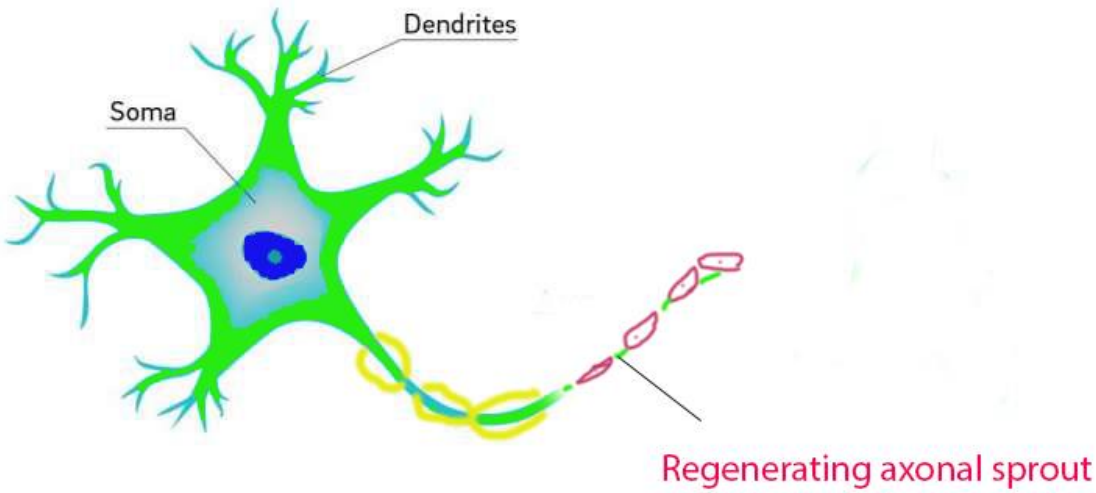
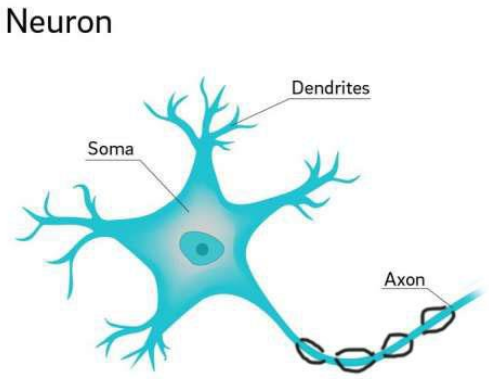


Image showing Wallerian degeneration. Axotomy results in fragmentation of the distal axon and myelin sheaths. Schwann cell proliferates and macrophages invade the distal nerve segment and phagocytose degrading materials.



Schwann cells in the distal segment line up in bands of Bungner. Axonal sprouts advance embedded in the Schwann cells and attracted by gradients of neurotrophic factors



Axonal reconnection with end organs and maturation and remyelination of the nerve fibers

Sir Herbert Seddon distinguished three types of localized injuries to peripheral nerves and he introduced the terms neuropraxia, axonotmesis and neurotmesis.

Neuropraxia:

Occurs when compression / stretch occurs producing an anoxic, physiological block or both axoplasmic transport and ion channel functions along affected axons. Loss of function is usually temporary, and release of the compressive agent usually results in rapid and complete recovery of function and relief of pain. The term neuropraxia is reserved for those situations where electrodiagnosis has conclusively shown that demyelinating conduction block is solely responsible for the neural lesion.

Axonotmesis:

This occurs when a blunt injury to a nerve results in Wallerian degeneration distal to the site of injury, but the connective tissue layers (epi, peri and endo neurium) remain intact. Conduction ceases throughout the distal extent of the nerve within a few days of injury. The distance from the site of injury to the end organ is a determinant for completeness of the recovery. Regrowing axon cell sprouts could extend to the target within the Schwann cell tubes that housed their parent axon. If their progress is unimpeded, they tend to regrow at the rate of 1 mm / day. This fact can be judged by the presence of advancing Tinel's sign. (Tinel sign is elicited by light tapping over the regenerating nerve route elicits a sensation of tingling, or pins and needle sensation in the distribution area of the nerve. Complete recovery of the functions of the end organ is not a certainty in these patients.

Neurotmesis:

Occurs when the nerve is either completely cut or so badly disorganized by the injury process that recovery without some form of surgical intervention is impossible. All the connective tissue layers of the nerve as well as the axons are disrupted at the site of injury, a wide inter stump gap may be produced either by the injury or during subsequent intra-operative wound debridement.

Spontaneous axonal regeneration following this type of injury will be imperfect and disorderly and may not occur at all. Distance from the site of injury is also a significant factor in the recovery process. Changes in the fiber type and loss of striated muscle mass begin within days after denervation; up to 80% of muscle volume may be lost within 4 months and irreversible muscle fibrosis and fatty infiltration occurs after 2 years. Even after nerve repair, muscles usually exhibit weakness, impaired coordination and reduced stamina. Regeneration of the largest diameter axons and coactivation of alpha and gamma efferents may fail.

Cutaneous sensory receptors undergo a slow degenerative change after denervation and may disappear after 3 years. Their reinnervation tends to reverse these changes particularly if the injury occurs close to the location of end organs and the nerves involved are sensory. Longer the period of denervation, the less complete will be the degeneration.

Sydney Sunderland amplified Seddon's category of axonotmeses to describe progressively more invasive levels of injury to the endoneural contents, perineurium and epineurium respectively correlating these new levels with more accurate prognosis of outcomes in axonotmesis injuries. Recovery is typically complete after Sunderland's grade 1 (equivalent to neuropraxia), grade 2 axonal degeneration with intact endoneurium, grade 3 injuries recover partially, while grades 4 and 5 (equivalent to neurotmesis) usually require surgical intervention.

Classification of various types of nerve damages

Seddon	Sunderland	Pathology	Electrophysiological correlate
Neuropraxia	Grade I	A transient light compression causing endoneurial oedema but no significant morphological changes. A more substantial and prolonged mechanical compression or stretch is most likely to cause a focal demyelination that may be paranodal or affect whole internodes. Anoxia plays a role in the pathogenesis	Conduction block ± conduction slowing. Nerve distal to 'lesion' shows normal conduction
Axonotmesis	Grade 2	Axons degenerate distal to the site of the lesion, irrespective of calibre or modality Endoneurium, perineurium and epineurium remain intact. Schwann cell basal lamina tubes either remain continuous across the lesion or are minimally	Fibrillation Mild diminution to complete absence of SNAP and CMAP responses, in proportion to degree of axonal loss ± varying degrees of conduction block and slowing associated with demyelination

		separated within a morphologically intact perineurium and epineurium	
Axonotmesis	Grade 3	Endoneurium disrupted, axons degenerate distal to the site of the lesion	Fibrillations, absent SNAP and CMAP responses
Axonotmesis	Grade 4	Perineurium disrupted, axons degenerate distal to the site of the lesion	Fibrillations, absent SNAP and CMAP responses
Axonotmesis	Grade 5	Epineurium disrupted; axons degenerate distal to the site of the lesion	Fibrillations, absent SNAP and CMAP responses

Depending upon the level of disruption misdirection of regrowing axons into modality-inappropriate Schwann tubes in the distal stump is inevitable.

Etiology of Facial nerve Paralysis

Facial paralysis may be caused by varying pathologies. They are elaborated under.

Facial paralysis at birth:

Facial nerve palsy in the newborn period could be due to:

Congenital causes: Commonly caused by developmental defects. In congenital facial nerve paralysis other cranial nerves like III, IV, V, VIII paralysis are also associated with facial nerve palsy. This syndrome is known as the Mobius syndrome. Prevalence of this syndrome is about 1 in 150000 live births. It is reported to be due to hypoplasia of the motor nuclei of the cranial nerves within the brainstem, probably due to hypoxic-ischemic encephalopathy.

Goldenhar syndrome: This syndrome includes hemifacial microsomia, with a spectrum of congenital malformations involving the structures derived from the first and second pharyngeal arch can also present with congenital facial paralysis.

Congenital pseudobulbar palsy: Also known as syringobulbia, is a condition that clinically manifests with facial paralysis, dysphagia and speech difficulties.

Arnold - Chiari syndrome: Congenital facial paralysis is usually associated to other cranial nerve palsies especially the 6th cranial nerve due to malformations of the posterior fossa that allow herniation of brain structures through the foramen magnum

Traumatic: Is caused by head molding of the fetus, as a sequel to forceps delivery. This is common in infants with birth weight greater than 3500 g. Other risk factors being use of forceps and premature delivery. This condition usually has a favorable prognosis, with infants recovering the full functionality of the 7th nerve within a few months without sequelae.

Traumatic causes of facial paralysis:

Skull base fractures:

Temporal bone fracture is caused by blunt closed head trauma is the most common cause of traumatic facial nerve paralysis. Motor vehicle accidents are the most common mechanism of injury, followed by

assaults, falls and two-wheeler accidents. Blunt extratemporal trauma to the face, is a rare cause of paralysis and can be differentiated from Intratemporal bone trauma by the fact that it often involves only specific branches of the facial nerve.

Penetrating injuries (lacerations, stab wounds) results in lesions to the facial nerve distal to the stylomastoid foramen. Gunshot wounds however can injure both the Intratemporal and extratemporal portions of the facial nerve.

Facial nerve paralysis is more common in transverse fractures of temporal bone.

Facial injuries

Penetrating injury to middle ear

Altitude paralysis (barotrauma)

Scuba diving

Lightning strike

Neurologic causes of facial nerve paralysis:

Opercular syndrome:

Opercular syndrome consists of the clinical syndrome of bilateral corticobulbar involvement with facio-labio-pharyngo-glosso-laryngo-brachial paralysis of voluntary movement of these muscles, but well preserved automatic and reflex movements of the same muscles due to lesions at the cortical and subcortical region involving the anterior opercular area surrounding the insula formed from gyri of the frontal, temporal, and parietal lobes.

Bilateral opercular lesions typically impair the voluntary movements of the facial, lingual, pharyngeal and masticatory with preservation of the automatic emotional movements which is the classical presentation of this syndrome.

Millard Gubler syndrome is also known as facial abducens hemiplegia syndrome or ventral pontine syndrome. It is caused by tumors, infectious diseases (neurocysticercosis and tuberculosis), demyelinating diseases like multiple sclerosis, and viral infections like Rhomb encephalitis. In older individuals, it is frequently caused by vascular events like hemorrhage / ischemia. This syndrome is

caused due to a lesion at ventral part of pons that involves the fibers of cranial nerves VI, VII, and corticospinal tract fibers.

Infections causing facial nerve paralysis

Otitis Externa

Otitis media

Mastoiditis

Chicken pox

Herpes zoster oticus (Ramsay Hunt syndrome)

Mumps

Infectious mononucleosis

Leprosy

Influenza

Coxsackie virus

Malaria

Lyme disease

Cat scratch disease

HIV

Metabolic causes of facial paralysis

Diabetes mellitus

Hyperthyroidism

Pregnancy

Hypertension

Acute porphyria

Vitamin A deficiency

Neoplastic lesions causing facial nerve paralysis

Tumors involving parotid gland

Cholesteatoma

Tumors involving facial nerve

Glomus jugulare

Acoustic neuroma

Leukemia

Meningioma

Hemangioblastoma

Carotid artery aneurysm

Fibrous dysplasia

Toxic causes of facial paralysis

Thalidomide

Ethylene glycol

Alcoholism

Arsenic intoxication

Tetanus

Diphtheria

Carbon monoxide

Iatrogenic causes of facial nerve paralysis

Mandibular block anesthesia

Antitetanic serum

Post immunization

Parotid surgery

Mastoid surgery

Embolization

Idiopathic causes

Bell's palsy:

Bell's palsy is defined as idiopathic lower motor neuron type of facial nerve paralysis. This is in fact the most common type of facial palsy. This condition was first described by Sir Charles Bell one century ago.

This condition is mostly unilateral, and rarely bilateral. Bell's palsy is a diagnosis of exclusion, which must be made only after excluding all the known causes of facial nerve paralysis.

Pathophysiology: Etiology and pathophysiology is highly controversial. The patient gives history of going to bed normally, and waking up with facial palsy, or there is a history or bus / train travel with the patient seated close to the window.

1. Exposure to cold air has been postulated as one of the causes

2. Viral infections involving the nerve sheath

There is inflammation of the facial nerve causing it to swell up. Since it is enclosed inside a rigid bony canal it has virtually no space to expand causing the damage to the nerve. The labyrinthine segment of the facial canal is the narrowest portion of the whole facial canal (about 0.6mm).

Clinical features:

The patient wakes up with lower motor neuron type of facial paralysis.

1. Inability to close the ipsilateral eye

2. Reduction of tearing in the ipsilateral eye

3. Deviation of the angle of the mouth to the opposite side

4. Drooling of saliva

5. Metallic taste in the tongue

6. Inability to wrinkle the forehead

7. Bell's phenomenon (rolling of eyeball upwards)

This condition is exceedingly rare in pregnant women, and if present it tends to be very severe with poor recovery.

Prognosis is excellent. 99% of patients recovering completely.

Melkerson Rosenthal syndrome: Causing recurrent alternating facial palsy, furrowed tongue and faciolabial oedema. Etiology is unknown and this condition is managed symptomatically using anti-inflammatory drugs (NSAIDS).

Landry Guillian Barre syndrome

Myasthenia gravis

Sarcoidosis (Heerfordt syndrome, Uveoparotid fever)

Osteopetrosis

Clinical Evaluation

Detailed history taking coupled with through clinical examination is a must in patients with facial nerve paralysis. No effort should be spared in determining the exact cause of the paralysis. This is particularly so if the patient lands up with facial nerve paralysis with a history that does not quite fit with that of Bell's palsy.

Patients usually demand answers to the following three questions:

1. What was the cause?

2. When can recovery be expected? (prognosis)

3. What can be done to promote recovery? (treatment modality)

Patient history should focus on the following points:

Majority of patients with facial paralysis consult the general practitioner first.

The history of onset of paralysis, whether complete or incomplete, sudden or progressive should be sought from the patient even though they are not diagnostic they could serve as pointers.

All of these patterns have been noted with idiopathic facial nerve palsy as well as with infections / neoplasms. Progressive facial paralysis over a period of more than 3 weeks or an incomplete facial paralysis that does not start the recovery process even after 3-6 weeks, should point towards the diagnosis of neoplasm and investigations should be tailored accordingly.

Ipsilateral recurrent facial nerve paralysis can occasionally be seen in patients with idiopathic palsy (Bell's palsy), Melkerson-Rosenthal syndrome, and tumors. Studies reveal that patients with a recurrence were 2.5 times more likely to have a family history which is positive for this type of facial nerve paralysis. Recurrent facial nerve paralysis is common in herpes simplex type I infection but rare with Herpes zoster.

It is highly recommended that a tumor be suspected in every patient who presented with recurrent facial nerve paralysis. In contrast to ipsilateral recurrence contralateral recurrence of facial nerve paralysis is more sinister.

Alternating recurrent facial nerve paralysis even though rare is a feature of Melkerson-Rosenthal syndrome. This condition is characterized by the presence of facial oedema, fissured tongue and a positive family history. Concurrent bilateral facial nerve paralysis is probably associated with a systemic condition, the most common being Gullain-Barre syndrome. This condition is also seen in cerebral lymphoma, leukemic infiltration, sarcoidosis, Lyme disease, rabies, infectious mononucleosis and Moebius syndrome.

The following things should be looked for while eliciting a patient's history:

1. Presence of pain in the ear: Patients with Bell's palsy frequently complain of pain behind the ear.

2. Paresthesia / Numbness: This can occur on the affected side

3. Taste disturbance: Loss of taste sensation on the anterior two thirds of the tongue on the ipsilateral side

4. Presence of rash: Typically, multiple small vesicles over the ear, external auditory canal and the pharyngeal mucosa may indicate herpes zoster infection. Involvement of facial nerve by herpes zoster is a recognized cause of facial palsy (Ramsay Hunt syndrome).

5. Hyperacusis: Perceiving sound as unduly loud in the ipsilateral ear, occurs in approximately a third of patients and is secondary to weakness of the stapedius muscle.

6. Recent viral infection and history of recent vaccination: This is to rule out viral etiology and autoimmune reaction

7. History pertaining to involvement of other cranial nerves

Physical examination:

A complete head and neck and otological and cranial nerve examination should be done. Facial weakness could be subtle and apparent only to a trained examiner and most certainly to the patient and their close friends and care givers. The degree of facial weakness must be recorded from the onset and every attempt should be made to localize the precise site / cause of the lesion along the course of the facial nerve. Facial nerve paralysis could be the first presentation of a systemic illness.

If symptoms or signs of other cranial nerve deficits are present, a central / systemic cause should be suspected. Sparing of forehead movement is considered to be characteristic of a central lesion but is not always the case. Normal movement can be seen in facial nucleus lesions and even peripheral lesions of the temporal branch of the facial nerve.

A partial facial nerve palsy involving one or two of its branches would be suggestive of a disease localized distal to the sternomastoid foramen and a parotid malignancy should be suspected. If in association with facial nerve paralysis, there is parotid gland enlargement with impaired vision or iritis then sarcoidosis should be seriously considered.

Facial paralysis grading systems:

Currently many systems of clinical grading of facial nerve function have been described. It was in 1983 when House proposed a six-point grading system for reporting the recovery and outcomes of surgery for vestibular schwannomas. This system was later modified and became known as the House-Brackmann staging system and is the most widely used scheme.

In the House-Brackmann system:

Grade I - Normal

Grade II - V - Intermediate

Grade VI - Complete absence of facial nerve function

House-Brackmann system has its own limitations with respect to precision and inter-observer error. It emphasizes the characteristics of facial nerve paralysis from two assessment domains.

Domain I: Gross observation of the facial symmetry or looking out for facial asymmetry. This is also known as static criteria.

Domain II: This domain involves assessment of movements of the forehead, eye and mouth. This is also known as dynamic criteria.

Burres and Fisch method: This method requires multiple measurements of movement in different parts of the face.

Facial nerve dysfunction includes secondary effects that complicate facial nerve injury. They are usually caused by aberrant neural degeneration and include synkinesis, hemifacial spasm, contracture, crocodile tears, epiphora, dysgeusia, pain and hyperacusis. These effects usually don't accompany the primary weakness in an orderly manner, but they significantly affect the patient's quality of life.

Functional impairments caused by facial nerve paralysis include:

Difficulties in

Drinking

Eating

Speaking

Conveying emotions via facial expression

There are also psychological consequences for the patient like lack of self-esteem, anxiety when meeting strangers etc.

House-Brackmann grading system of functional status of facial nerve:

Degree of Injury	Grade	Definition
Normal	I	Normal symmetrical function in all areas
Mild dysfunction (barely noticeable)	II	Slight weakness noticeable only on close inspection. Complete eye closure with minimum effort Slight asymmetry of smile with maximal effort. Synkinesis barely noticeable, contracture or spasm absent.
Moderate dysfunction	III	Obvious weakness but not disfiguring. May not be able to life eyebrow. Complete eye closure and strong but asymmetric movement with maximal effort. Obvious but not disfiguring synkinesis, mass movement or spasm
Moderately severe dysfunction	IV	Obvious disfiguring weakness Inability to life eyebrow Incomplete eye closure and asymmetry of the mouth with maximal effort Severe synkinesis, mass movement, spasm
Severe dysfunction	V	Motion barely perceptible Incomplete eye closure, slight movement of the corner of the mouth Synkinesis, contracture, and spasm usually absent
Total paralysis	VI	No movement, loss of tone, no synkinesis, contracture or spasm

Diagnostic tests

Diagnostic tests that need to be performed in these patients should clear the air any doubts pertaining to the location of the lesion and the probable cause for it.

Topodiagnostic tests:

This battery of tests refers to the functional testing of an individual facial nerve branch in an attempt to locate the anatomical level of dysfunction / injury. These tests have some value but has limited correlation with the precise site of nerve damage.

Test	Nerve branch assessed	Technical considerations	Assessment / Outcome
Schirmer's test	Greater superficial petrosal nerve	Strips of filter paper is placed in the inferior conjunctival fornix for 5 minutes and the length of the paper moistened is compared between two eyes.	➢ 75% unilateral decrease in lacrimation, or a bilateral decrease in lacrimation (less than 10 mm wetted for both sides at 5 minutes)
Stapedial reflex	Nerve to stapedius muscle	Psychoacoustic audiometry	Present / absent
Electrogustometry	Chorda tympani	The tongue is stimulated electrically to produce a metallic taste and the two sides are compared	Threshold of the test is compared between the two sides
Salivary flow testing	Chorda tympani	Wharton duct can be cannulated, and salivary flow is measured over time following a gustatory stimulus. (sweet under the tongue can be used)	A reduction of 25% is considered to be abnormal

Image showing Schirmer's test being performed

Measurement of stapedial reflex thresholds:

Stapedius reflex thresholds can be measured by tympanometry. These measurements provide information about the middle and inner ear, in addition to the 8th and 7th cranial nerve integrity and brainstem function. Dynamic changes that results from contraction of stapedius muscle in response to stimuli of 500, 1000, 2000 and 400 Hz at intensities of 70-115 dB sound pressure level, are measured and the thresholds of activation is documented.

Afferent limb for stapedial reflex includes:

Ear drum

Middle ear

Cochlea

8th cranial nerve

Efferent limb includes:

7th cranial nerve

Middle ear

Ear drum

Cross over in the brainstem at the level of superior olive complex

The stapedius reflex can be measured both ipsilaterally and contralaterally

When presented with a high intensity sound stimulus, the stapedius and the tensor tympani muscles contract. The stapedius stiffens the ossicular chain by pulling the stapes away from the oval window and the tensor tympani muscle stiffens the ossicular chain by loading the ear drum when it pulls the malleus medially towards the middle ear. This decreases the middle ear admittance which can easily be measured using an impedance audiometer.

Electroneuronography:

In this test a supramaximal stimulus is delivered to the facial nerve trunk as it exits the stylomastoid foramen and the evoked biphasic compound muscle action potential is recorded using surface electrodes. The response of the paralyzed side is compared with that of the normal side, which serves as a control and is expressed as a percentage of normal. Studies reveal that reduction in CMAP correlates well with histological axonal loss.

If the CMAP amplitude on the affected side is 10% of the normal side, then it is assumed that 90% axonal loss has been sustained. This test is said to be useful only from the 4^{th} day of facial nerve paralysis. Neural conduction disappears quite rapidly after an injury that transects the axon, because a degenerating axon is incapable of conducting an action potential and majority of these axons have started to degenerate within 3-4 days of such an injury.

Electroneuronography is considered the most valuable prognostic indicator among current electrophysiological tests and its main indication is acute onset of complete facial nerve paralysis. Both the percentage of amplitude reduction and rate of degeneration have been used as prognostic indictors of facial nerve recovery. In idiopathic palsy, degeneration of > 90% within 14 days of complete paralysis indicates a probable poor recovery in >50% of cases. This test is not useful in Ramsay Hunt syndrome because of multiple sites of nerve involvement.

Electromyography:

This test records active motor unit potentials of the orbicularis oculi and orbicularis oris muscles during rest and voluntary contractions. EMG can be used to determine:

1. If a nerve is in continuity (volitional activity recorded)

2. If there is evidence of Wallerian degeneration (fibrillation potentials0

3. If there are early sings of reinnervation (polyphasic innervation potentials)

Fibrillation potentials typically arise 2-3 weeks following injury and polyphasic reinnervation potentials may precede clinical signs of recovery by 6-12 weeks.

In cases of acute onset and complete paralysis with unfavorable ENoG recordings if active motor unit potentials are present it indicates that some fibers are intact and suggests a favorable prognosis. As long as some active motor units can be recorded one week following a complete paralysis, severe degeneration is unlikely to take place. EMG has a role in decision making regarding surgical intervention in long standing paralysis as follows:

1. Polyphasic motor unit potentials indicate regenerative processes and hence surgical intervention is not indicated.

2. Fibrillation potentials indicate lower motor neuron denervation indicate lower motor neuron denervation, but viable motor end plates. Surgical exploration is hence indicated with a view to achieving nerve continuity, either by end to end anastomosis, interposition grafting, re-routing and reinnervation techniques

3. Silence in EMG (no electrical output) indicates long term denervation and suggests that muscle has been replaced by fibrous tissue. In this case, static or dynamic facial reanimation is indicated.

Limitations of electrophysiological testing:

Electrophysiological testing could provide useful prognostic information and would serve as a guide to the management of facial palsy, there are some important shortcomings:

1. Electrical impulse can only stimulate normal / neuropraxic fibers and cannot distinguish whether the remaining fibers are in a state of axonotmesis or neurotmesis. Thus, it cannot distinguish between injuries which have different prognosis.

2. It provides no useful information in cases of incomplete facial paralysis

3. It fails to provide information on the immediate post paralysis period (first 72 hours).

Study	**Measurement**	**When to measure**	**Use in acute onset paralysis**	**Use in long standing paralysis**
ENoG	Evoked CMAP compared to normal side	Between 3 days and 3 weeks	>90% of degenerated fibers suggests poor prognosis	Not useful because of desynchronization
EMG	Active motor unit potentials after voluntary forceful contraction	Complementary to ENoG after 2 weeks	Presence of active motor potentials in response to voluntary contractions indicates good prognosis	Fibrillation potentials suggest Wallerian degeneration
		In long standing paralysis		Prophylactic potentials suggest reinnervation

Transcranial magnetic stimulation:

While the electrodiagnostic tests provide information about the facial nerve distal to the stylomastoid foramen, transcranial magnetic stimulation is able to stimulate the facial nerve within the cranium. Magnetic impulses induce an electric current within the brain and stimulate neuromuscular tissue in exactly the same way as conventional electrostimulation. The design and the type of coil determines the penetration and site of stimulation. A double coil is normally employed for facial nerve studies and has the ability to focus on the infranuclear portion of the nerve, probably at the root of entry zone or temporal bone itself.

Intraoperative facial nerve monitoring

Intraoperative nerve monitoring is widely used in all surgeries where there is a risk of damage to the facial nerve. Many of these devices employ continuous EMG monitoring from peripheral facial muscle groups for spontaneous and electrically evoked stimulation of the facial nerve trunk or its branches. Intraoperative nerve monitoring has proved itself in CPA tumor surgery, revision mastoidectomy, parotidectomy and congenital ear abnormalities.

Pit falls and caveats of intraoperative monitoring:

Always ask the anesthetist to avoid neuromuscular blockade.

The closer the bipolar electrodes are placed the fewer the muscle fibers that will be represented in the EMG response.

Transparent drapes should be used.

Test the integrity of the system with the anesthetic nerve stimulator before starting the operation and check the system periodically during surgery.

Even after nerve transection, the distal segment can still be stimulated for a few days, creating evoked EMG responses that may falsely be interpreted as evidence of continuity.

Mastoid segment is least sensitive to nerve stimulation, so a higher setting is needed (0.5mA in mastoid surgery as opposed to 0.05-0.1 mA in CPA tumor surgery).

Investigations

Blood investigations:

The following blood tests would help to identify / exclude other less common causes of facial paralysis:

Angiotensin converting enzyme titers

Anti-neutrophil cytoplasmic antibody profile

Cytoplasmic antibody profile

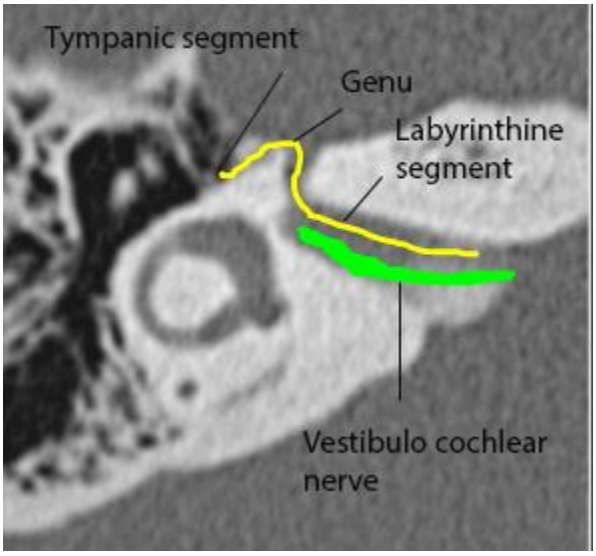
HIV serology

Lyme serology

Syphilis serology

Imaging of facial nerve

CT imaging of temporal bone allows identification of the fallopian canal from the fundus to the internal auditory canal to the stylomastoid foramen. The tympanic portion of the facial nerve is the easiest to identify on axial scans at the level of body of incus and its short process. From here it can be followed proximally and distally towards the labyrinthine and descending segments, respectively. The labyrinthine segment is banana shaped on axial sections.



Axial CT image of temporal bone showing facial nerve

On coronal sections it may be visualized as the medial of the two circular eyes directly above the cochlea. The sulcus for the geniculate ganglion is also well demonstrated in coronal sections. The mastoid segment is well visualized in coronal / sagittal section. In coronal view it is easier to follow the course of the nerve from the stylomastoid for amen to its more proximal part. Facial nerve is difficult to identify in well pneumatized temporal bones. One way to locate the facial nerve in these cases is to locate the pyramid or the stapedius muscle. The facial nerve lies immediately behind it. Bance and Erb have described the 'B-line'; a tangential line extrapolated from the posterior border of the basal turn of cochlea, which falls within 1 mm of the facial nerve average.

The intraparotid facial nerve cannot be identified in CT images.

The rich perineural arteriovenous plexus which surrounds the normal facial nerve facilitates visualization on contrast enhanced T1-weighted MRI. It is usually observed in more than one segment. Intraparotid portion of facial nerve cannot be routinely identified in MRI.

Imaging in facial nerve paralysis:

Imaging strategy depends on the suspected nature of the lesion and level of the lesion. A high-resolution CT scan enables superior visualization of the Intratemporal portion of the facial nerve and is specifically useful in temporal bone trauma, Fallopian canal involvement in CSOM or temporal bone malignancies.

Imaging in idiopathic facial palsy:

Imaging a patient with idiopathic facial paralysis is not a cost-effective option. The degree of enhancement on MRI carries no significant prognostic value and the chance of acute facial nerve palsy in a tumor is remote. Imaging is recommended if the presentation is atypical, an alternative diagnosis is suspected, surgical decompression is planned, or recovery is incomplete at 6 months after the lesion.

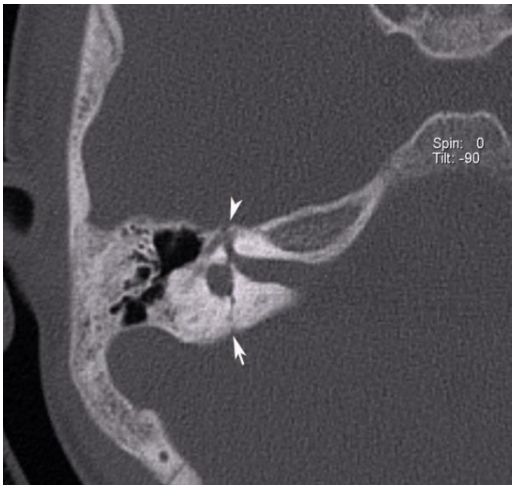
MRI may be normal during the first 10 days following the onset of palsy. Post gadolinium enhancement is characteristically asymmetric, diffuse, intense and linear, it should not be nodular. It can involve the entire temporal segment of the facial nerve and may even persist for several months after full recovery.

Imaging in acute / chronic otitis media:

Imaging in facial nerve paralysis following acute / chronic otitis media is directed towards identifying coexisting Intratemporal / intracranial complications rather than nerve damage. CT imaging of temporal bone can delineate the site of erosion of the Fallopian canal, but MRI cannot differentiate between the increased signal of facial nerve from that of surrounding inflamed granulation tissue.

Imaging in fractures temporal bone:

HRCT is the investigation of choice for temporal bone fractures. Facial nerve injury is usually situated just distal to the geniculate ganglion and the first genu in longitudinal fractures and just proximal to the geniculate ganglion in transverse fractures.



CT image showing temporal bone fracture

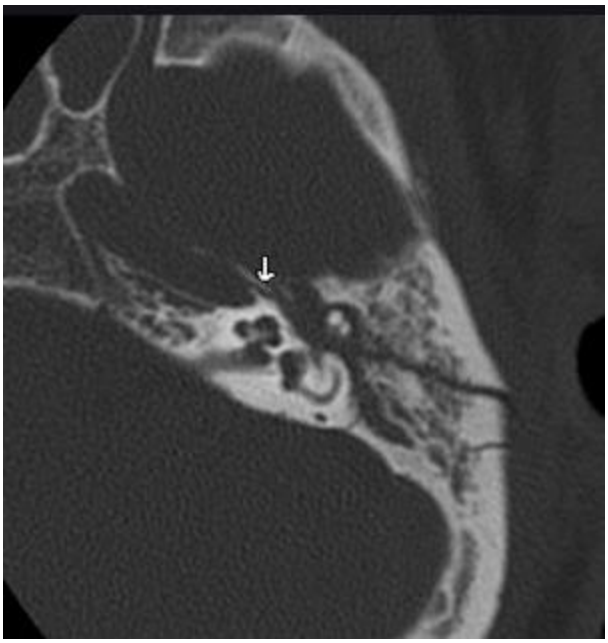


Image showing longitudinal fracture of temporal bone

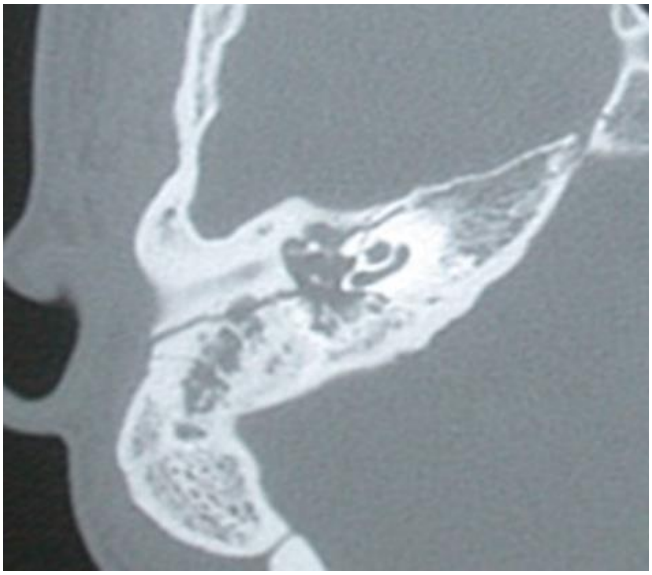


Image showing transverse fracture of temporal bone

Role of imaging in tumors of temporal bone:

MRI is the investigation of choice in these patients. Hemangiomas involving the facial nerve enhance intensely on post contrast T1 weighted MRI. It has a typical honeycomb matrix appearance on CT. Facial nerve schwannoma shows a strong enhancement with gadolinium on T1 weighted images and when they grow to the point of filling the internal acoustic meatus they cannot be distinguished for acoustic neuroma.

Malignant tumors of parotid gland could involve the facial nerve and a soft tissue intensity enhancing mass could be observed extending from the gland through an enlarged stylomastoid foramen to involve the mastoid segment of the nerve.

Management

Management of acute facial nerve paralysis depends to a large extent on the cause of the lesion. Management should also focus on the issues caused by facial paralysis. These issues could be really acute in nature and it is necessary to offer treatment on an immediate basis.

Eye care

Eye care includes corneal protection. Eye closure and lacrimation is affected in patients with facial nerve paralysis. Lack of eye closure could lead to exposure keratitis leading on the blindness. These patients need eye closures / Eye cap to protect the eyes. They should also be advised to wear cornea protecting dark glasses if wearing eye cap is not feasible.

If the nerve injury involves the trunk proximal to the 1st genu before the origin of the greater superficial petrosal nerve the secretomotor supply of the lacrimal gland is impaired thus compounding the problem of corneal dryness. Moisturizing eye drops can be used as frequently as needed during daytime to protect the eyes. Moisturizing ointment is preferred during nighttime.

As time progresses the unopposed action of levator palpebrae superioris (supplied by oculomotor nerve) results in shortening of the upper eyelid. Upper eyelid weighting with skin tone colored weights or levator lengthening procedures are used in long standing cases of facial nerve paralysis. Formerly lateral tarsorraphy was performed as an interim measure.

Image showing right lower motor neuron type facial paralysis

Management of Bell's palsy:

Minimum diagnostic criteria of Bell's palsy include:

1. Paralysis / paresis of all muscle groups on one side of the face
2. Sudden onset
3. Signs of absence of CNS disease
4. Absence of signs of ear disease / CPA lesions
5. Prodromal illness with mild fever and aural pain +
6. Classic history of going to bed normally and waking up with facial nerve paralysis

It should be stressed that prognosis of facial nerve paralysis is better in pregnant women than in non-pregnant women. Clinical signs and symptoms show significant improvement following delivery of the child.

Pathophysiology:

1. Exposure to cold air has been postulated as one of the causes

2. Viral infections involving the nerve sheath

There is inflammation of the facial nerve causing it to swell up. Since it is enclosed inside a rigid bony canal it has virtually no space to expand causing the damage to the nerve. The labyrinthine segment of the facial canal is the narrowest portion of the whole facial canal (about 0.6mm).

Majority of these patients show complete recovery of facial nerve function within 3 months of illness.

Management:

1. Eye care: The patient should wear glasses to protect cornea. (Black glasses are preferable), use of artificial tears.

2. Regular physiotherapy (Balloon blowing)

3. Cheek / eye massage

4. Steroids: Very useful in early stages of the disease

5. Antiviral drugs like acyclovir has been tried with varying degrees of success

6. Facial nerve decompression can be considered in patients who don't show signs of recovery within 6 months

Facial nerve disorders of viral origin

Ramsay Hunt Syndrome:

Ramsay Hunt syndrome is a disease affecting the external auditory canal associated with the following symptom complexes:

1. Lower motor neuron type of facial nerve palsy

2. Herpetic blisters of the skin of the external auditory canal

3. Otalgia

This syndrome was first described by J. Ramsay Hunt in 1907. He described patients with Otalgia associated with cutaneous and mucosal rashes. He attributed it to the infection of geniculate ganglion by Herpes virus type 3.

Pathophysiology:

The primary pathophysiology is located in the geniculate ganglion of the facial nerve. Geniculate ganglion is found to be affected by Human Herpes virus type 3 i.e. (Varicella zoster virus). Varicella zoster virus have been identified from tears of these patients by polymerase chain reaction. In fact Varicella zoster virus have also been identified from tears of patients with Bell's palsy.

These patients have deep seated pain in the affected ear associated with vertigo, tinnitus, ipsilateral transient hearing loss and lower motor neuron type of facial palsy. These symptoms develop due to involvement of the geniculate ganglion of the facial nerve located near the petrous pyramid portion of the temporal bone. The site of rash varies from patient to patient due to individual variations in the areas supplied by the nervous intermedius of wrisburg (sensory branch of facial nerve). Rashes may be present in the anterior 2/3 of the tongue, soft palate, external auditory canal and the pinna.

Morbidity / Mortality:

This disease is usually not associated with mortality. It is a self-limiting disease, with morbidity due to facial nerve palsy. Complete recovery of the nerve is seen only in 50% of patients as compared to more than 90% in Bell's palsy.

Clinical features:

Patient has deep seated pain in the affected ear. The pain is intermittent in nature, radiating towards the pinna of the ear. There is associated diffuse dull aching background pain. Patients also give history of exposure to Varicella virus infections (chicken pox). The classic Ramsay Hunt syndrome is associated with 1. Pain in the ear, 2. Vertigo and ipsilateral hearing loss, 3. Tinnitus, and 4. Facial palsy (LMN type). Rash or blisters can also be seen along the distribution of nervus intermedius. These herpetic blisters in the external auditory canal may become secondarily infected causing cellulitis.

Investigations:

Basic investigations like blood count, ESR and electrolytes estimation must always be done in these patients.

Virology:

1. Varicella virus the causative agent responsible for this syndrome also causes chicken pox in children

2. Serologic tests for Varicella virus is positive

3. Varicella virus can be isolated and cultured form the fluid extruding from the blisters

4. It can also be detected by PCR on samples of tear fluid from these patients.

5. Audiometry demonstrates sensorineural hearing loss

6. Unilateral caloric weakness may be present on electronystagmography (ENG).

Histology:

The affected ganglia are found to be swollen and inflamed. The inflammatory reaction is lymphocytic in nature. Some of the cells in the ganglia may show evidence of degeneration.

CSF analysis is not indicated in these patients.

Management:

1. Steps towards alleviating pain: Carbamazepine can be prescribed in doses of 400 mg / day in divided doses. Temporary relief of Otalgia in geniculate neuralgia may be achieved by applying a local anesthetic or cocaine to the trigger point, if in the external auditory canal.

2. Corticosteroids and oral acyclovir can be administered. Steroids in the form of prednisolone can be administered orally in doses of 10mg twice a day. Steroids should not be stopped abruptly. The dosage needs to be tapered. Acyclovir can be administered in doses of 800 mg orally 5 times a day.

3. Management of vertigo: can be managed using meclizine in doses of 25 mg orally 4 times a day.

4. Care must be taken to prevent exposure keratitis because of the inability to close the eye lids. The patients must wear protective goggles.

Management of facial nerve trauma

Management of facial nerve paralysis following trauma is generally deferred until the patient is both medically and neurologically stable as most of these cases would have sustained more life-threatening injuries. This commonly delays treatment for a large proportion of patients.

Facial nerve injuries can result from stab wounds to the face or mandibular fractures. If it is feasible, it is advisable to explore the region within 3 days so that a nerve stimulator can be used to identify the distal segments of facial nerve branches. Transection of the temporofacial / cervicofacial trunk should be promptly repaired by end-to-end anastomosis. Mobilization to avoid unwanted tension may be necessary if a short segment of the nerve is destroyed. Very rarely graft interposition could be needed. Injuries to branches distal to the lateral canthus or nasolabial fold are too small for anastomosis and are hence better approximated using tissue glue. This procedure offers better chances of recovery without synkinesis. If the wound is dirty, the facial nerve branches should be identified, washed, cleaned and tagged in order to facilitate repair at a later date.

Major problem with knife wounds to the face is the increased incidence of multilevel transections at more than one level. With each transection, the chance of a good functional outcome is diminished.

Facial nerve injuries due to gunshot wounds are tricky. Facial oedema and muscle tone and eye closure appear adequate when in fact they are not. This illusion can last for several days till the oedema actually subsides. The mastoid segment of the facial nerve is commonly involved and that too over a long segment damage. Low velocity bullets can be found lodged at the mastoid tip. Surgical exploration of the facial nerve should be performed immediately. Since there is considerable bony damage, the potential for subsequent cholesteatoma formation is very much real.

Management of facial palsy due to fractures temporal bone

Temporal bone fractures are divided traditionally into longitudinal and transverse. The fracture could be of mixed type in a significant number of these patients. Longitudinal fractures are associated with 20% incidence of complete paralysis of the nerve and the perigeniculate region is commonly involved. Transverse fractures have a higher incidence of facial nerve paralysis nearly (50%) and the labyrinthine and mastoid segments are commonly affected. Longitudinal fractures are commoner than that of transverse fractures.

Issues to consider in the management protocol:

1. Is there an indication for surgical exploration?
2. If there is an indication when to operate?
3. Which approach to follow?

High resolution CT imaging and ENoG are of real help in decision making process.

Goal of surgery:

1. To decompress the nerve in order to prevent ischemia developing
2. To remove bony fragments that could impinge on the nerve
3. To re-establish continuity in case of transection

Patients with normal facial nerve function at presentation, regardless of whether they develop delayed palsy or even paralysis, and those with acute onset incomplete palsy without progression have an excellent prognosis and do not need surgical intervention. Follow up serial study of ENoG is not needed. Timing of surgery if needed depends on the general condition of the patient.

ENoG is not helpful after 3 weeks have elapsed. The decision to explore is largely based on CT finding and electromyographic results. If no return of facial nerve function is observed 6-12 months after the injury and there are no signs of polyphasic potentials that suggest reinnervation, exploration is indicated with an aim to achieve continuity either by end to end anastomosis / nerve interposition. The surgical approach used depends on the site of injury and the hearing status of the patient. Generally, middle cranial fossa approach is preferred for longitudinal fractures in which hearing is preserved. In the presence of severe sensori-neural hearing loss, a translabyrinthine approach is easier to use.

Iatrogenic injury

Middle ear and mastoid surgery:

The most common site of injury during middle ear / mastoid surgery is the distal tympanic segment including the second genu, followed by the mastoid segment. If an injury to the facial nerve is recognized intraoperatively, exploration with decompression of proximal and distal segments of the nerve should be undertaken.

If facial palsy occurs during immediate post op period, then edema could be the cause. Mastoid dressing should be removed and loosened to reduce post op oedema. Delayed facial paralysis in these patients could be due to reactivation of Herpes simplex / Varicella zoster virus.

Facial paralysis in parotid surgery:

The risk of injury to facial nerve depends on the tumor location. All parotid surgery is best undertaken with facial nerve monitoring and the end of the procedure the main trunk should be stimulated to confirm continuity. If there is no response, the nerve and its branches should be closely inspected for areas of discontinuity. Reduced incidence of facial nerve paralysis has been reported if harmonic scalpel is used.

CP angle tumor surgery:

Risk of facial nerve injury in CP angle surgery depends on the size of the tumor and the skill of the surgeon. When loss of facial nerve function is identified during surgery then end to end anastomosis or

cable interposition graft. A subsequent reanimation procedure can be performed if needed at a later date.

Management of neonatal facial nerve injury:

Trauma is the main cause. Other congenital causes should be excluded. Forceps delivery should be avoided in favor of cesarean section. Any child with facial paralysis at birth should also undergo brainstem response audiometry to test hearing as some developmental disorders are associated with cochlear nucleus abnormalities. Prognosis of neonatal facial nerve injury is excellent with more than 90% recovering completely.

Management of facial nerve paralysis as complications of ear infection

Facial nerve paralysis may complicate both ASOM / CSOM. It is caused due to direct involvement of facial nerve by infection through facial canal Dehiscences. Fallopian canal osteitis with bone erosion and nerve involvement, inflammatory edema leading to compression and secondary thrombosis of vasa vasorum with consequential ischemia and infarction of the facial nerve. Mainstay of treatment of facial paralysis associated with acute otitis media is antibiotic therapy. Steroids maybe used although their efficacy is not supported by clinical studies. Mastoid exploration and facial nerve decompression is essential for patients with CSOM.

Physiotherapy

Balloon blowing would help to strengthen cheek musculature.

Nerve stimulation can be used

Chemodenervation

This is useful to manage synkinesis. Botulinum toxin is used as a second line treatment to release the over tightened facial muscles and to facilitate the stretches. It also helps in reducing unwanted muscle movements.

Dynamic reanimation techniques popularized by plastic surgeons can be tried in totally paralyzed facial nerve.

These procedures include:

Static reanimation – Various face lifting procedures belong to this category.

Dynamic reanimation – When active muscle movements are desired then dynamic reanimation procedures can be performed they include the use of muscle transfer techniques like the temporalis transfer.