Disorder of the facial nerve
Embryology

The main pattern of the nerve complex course, branching pattern and the relationship is established during the first 3 months of gestation. During this period, the muscles of facial expression also differentiated, became functional and actively contracted, and the nerve is not fully developed till the 4 years of age.

The facial nerve develops within the 2nd pharyngeal arch at the same time the external and middle ear developed (1st arch). So, abnormality of the facial nerve should be anticipated whenever there is associated malformation in the external or the middle ear.
Anatomy

1. Nucleus lies deeply within the substance of the pons is that situation it is closely related to the V nucleus
2. Fibers travel a circuitous route at the first backward to encircle the V1 CN nucleus in the floor of the 4th ventricle and then forward through the pons to emerge on its surface then anterolateral to enter the petrous temporal bone
3. Within the cranial cavity it is closely related to the Viii CN
4. In the intrapetrous part, the facial n. and its sensory root accompany the Viii CN in the internal auditory canal her it has anastamotic with the vestibular nerve at the bottom of this canal, it enter the facial canal which at first runs laterally above the vestibule of the labyrinth until it turns backward through a right angle on the medial wall of the promontory and then fenestera vestibuli
At the medial wall of the aditus it curves downward to emerge on the inferior surface of temporal bone at the stylomastoid foramen to run forward within the parotid gland.

The N. supplies the following:

1. **Motor**: muscles of facial expression, stapedius muscle, posterior belly of diagastric muscle, and stylohyoid muscle.
2. **Sensory**: to the concha and to the parts behind the auricles.
3. **Autonomic fibers**: for lacrimal, submandibular and sublingual glands together with glands at the nasal and oral cavities (secretomotor and vasodilator).
4. **Special nerve taste via chorda tympani branch**: to supply anterior 2/3 of the tongue.
Types of nerve injury

1. **Neurapraxia** is defined as a reversible blockage of the transmission of nerve impulses due to pressure on the nerve fibers. Release of the pressure usually results in rapid and complete recovery of the function with no distal Wallerian degeneration.

2. **Axonotmesis** is a more severe injury and involves the blockage of axoplasmic flow. Although endoneurial tubules are preserved, distal Wallerian degeneration occurs.

3. **Neurotmesis** is a total N. transaction.
the history of the onset of palsy, whether complete or incomplete, sudden or progressive progressive facial nerve palsy over a period of more than three weeks, or an incomplete facial nerve palsy that does not start to recover after three to six weeks, should make the clinician suspect an underlying neoplasm as the cause and should dictate the need for further investigations.

Ipsilateral recurrent facial nerve palsy can happen in idiopathic palsy, Melkersson–Rosenthal syndrome and tumours.
In Bell’s palsy recurrence is 13% and family history is 2.5 times more. Melkersson-Rosenthal syndrome, a condition also characterized by alternating recurrent facial nerve palsy associated with facial oedema, fissured tongue and a positive family history.
In contrast to recurrent ipsilateral facial paralysis, contralateral recurrence is almost always benign. Bilateral concurrent facial nerve paralysis is most probably associated with a systemic condition, such as Guillain–Barre´ syndrome (most common), leukaemia, sarcoidosis, Lyme disease, rabies, infectious mononucleosis.

Physical examination thorough head, neck, otological and cranial nerve examination is the absolute minimum required when evaluating facial nerve dysfunction. Complete or incomplete facial nerve palsy localize the lesion intracranial, intratemporal or extratemporal. Facial nerve palsy may be the first presentation of systemic illness. If symptoms or signs of other cranial nerves deficits are present, a central or systemic cause should be suspected. Sparing of forehead movement is considered to be characteristic of a central lesion. However, it should be remembered that normal movement can also be seen in facial nucleus lesions and peripheral lesions of the temporal
Grading of facial nerve palsy. The House–Brackmann system. It has become the most widely used scheme and has been endorsed by the American Academy of Otolaryngology – Head and Neck Surgery. In the House–Brackmann system, grade I is normal function, grade VI is complete absence of facial motor function and grades II–V are intermediate.
<table>
<thead>
<tr>
<th>Degree of injury</th>
<th>Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal ((1^\circ))</td>
<td>I</td>
<td>Normal symmetrical function in all areas</td>
</tr>
<tr>
<td>Mild dysfunction ((barely noticeable) ((1-2^\circ))</td>
<td>II</td>
<td>Slight weakness noticeable only on close inspection</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Complete eye closure with minimum effort</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Slight asymmetry of smile with maximal effort</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Synkinesis barely noticeable, contracture or spasm absent</td>
</tr>
<tr>
<td>Moderate dysfunction ((obvious difference) ((2-3^\circ))</td>
<td>III</td>
<td>Obvious weakness, but not disfiguring</td>
</tr>
<tr>
<td></td>
<td></td>
<td>May not be able to lift eyebrow</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Complete eye closure and strong but asymmetric mouth movement with</td>
</tr>
<tr>
<td></td>
<td></td>
<td>maximal effort</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Obvious, but not disfiguring synkinesis, mass movement or spasm</td>
</tr>
<tr>
<td>Moderately severe dysfunction ((3^\circ))</td>
<td>IV</td>
<td>Obvious disfiguring weakness</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Inability to lift eyebrow</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Incomplete eye closure and asymmetry of the mouth with maximal effort</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Severe synkinesis, mass movement, spasm</td>
</tr>
<tr>
<td>Severe dysfunction ((3-4^\circ))</td>
<td>V</td>
<td>Motion barely perceptible</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Incomplete eye closure, slight movement corner mouth</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Synkinesis, contracture and spasm usually absent</td>
</tr>
<tr>
<td>Total paralysis</td>
<td>VI</td>
<td>No movement, loss of tone, no synkinesis, contracture or spasm</td>
</tr>
</tbody>
</table>
Special investigations
There are 3 important issues when confronted with facial palsy:
The cause
The site of lesion
The prognosis

TOPODIAGNOSTIC TESTING
These tests aim to localize the site but have no prognostic value.
<table>
<thead>
<tr>
<th>Test</th>
<th>Nerve branch assessed</th>
<th>Technique considerations</th>
<th>Assessment/Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schirmer test (Figure 241c.5)</td>
<td>Greater superficial petrosal nerve</td>
<td>Strips of paper are placed in the inferior conjunctival fornix for five minutes and the length of paper moistened is compared between eyes</td>
<td>&gt;75% unilateral decrease in lacrimation, or a bilateral decrease in lacrimation (less than 10 mm for both sides at five minutes)</td>
</tr>
<tr>
<td>Stapedial reflex</td>
<td>Nerve to stapedius muscle</td>
<td>See Chapter 232, Psychoacoustic audiometry</td>
<td>Present or absent</td>
</tr>
<tr>
<td>Electrogustometry</td>
<td>Chorda tympani</td>
<td>The tongue is stimulated electrically to produce a metallic taste and the two sides are compared</td>
<td>Threshold of the test is compared between sides</td>
</tr>
<tr>
<td>Salivary flow testing</td>
<td>Chorda tympani</td>
<td>Warthin's ducts are cannulated and salivary flow is measured over time following a gustatory stimulus (6% citric acid on anterior part of tongue)</td>
<td>A reduction of 25% is considered abnormal</td>
</tr>
</tbody>
</table>
Electrophysiological test

Currently, the two most helpful are the ENoG and EMG

Electroneuronography (ENoG)

Consider the most valuable prognostic indicators among electrophysiological test and the main indication is acute onset complete facial paralysis

Electromyography (EMG)

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

- if a nerve in question is in fact in continuity
- if there is evidence of Wallerian degeneration
- if there are early signs of reinnervation

Intraoperative nerve monitoring

Intraoperative monitoring includes continuous EMG measurement from peripheral facial muscle groups and electrical stimulation of the facial nerve itself or its branches to obtain a CMAP.

It has a place in cerebellopontine angle (CPA) tumour surgery, in revision mastoid and parotid surgery, and in surgery of congenital ear abnormalities. Other issues to consider are medicolegal issues.
Ct scan
The tympanic portion is probably easiest to identify on axial computed tomography (CT) scans at the level of the body of incus and its short process. From there on, it can be followed proximally and distally towards the labyrinthine and descending parts, respectively.

The descending or mastoid segment is best visualized in coronal or sagittal views.

MRI
Owing to the rich perineural arteriovenous plexus which surrounds the facial nerve, enhancement may be observed normally on T1-weighted magnetic resonance imaging (MRI) with the use of contrast agents. It is usually observed in more than one segment, more commonly in the geniculate ganglion and the tympanic segments and it may enhance asymmetrically between right and left.
Causes of facial palsy

**Birth**
- Moulding
- Forceps delivery
- Dystrophia myotonica
- Moebius syndrome (facial diplegia associated with other cranial nerve defects)

**Trauma**
- Basal skull fracture
- Facial injuries
- Penetrating injury to middle ear
- Altitude paralysis (barotauma)
- Scuba diving (barotauma)
- Lightning

**Neurological**
- Opercular syndrome (cortical lesion in facial motor area)
Infection

Otitis externa
Otitis media
Mastoiditis
Chicken pox
Herpes zoster cephalicus (Ramsay Hunt syndrome)
Encephalitis
Poliomyelitis (type I)
Mumps
Infectious mononucleosis (glandular fever)
Leprosy
Coxsackie virus
Malaria
Syphilis
Scleroma
Tuberculosis
Botulism
Acute haemorrhagic conjunctivitis (enterovirus

Mucormycosis
Lyme disease
Metabolic
Diabetes mellitus
Hyperthyroidism
Pregnancy
Hypertension
Acute porphyria

Neoplastic
Cholesteatoma
VIIth nerve tumour
Glomus jugulare tumour
Leukaemia
Meningioma
Haemangioblastoma
Sarcoma
Carcinoma
Anomalous sigmoid sinus
Haemangioma of tympanum
Facial nerve tumour
Schwannoma
Teratoma
Toxic

Tetanus
Diptheria
Carbon monoxide

*iatrogenic*
  Mandibular block anaesthesia
Anti-tetanus serum
Vaccine treatment for rabies
Post-immunization
Parotid surgery
Mastoid surgery
Embolization
Dental

*idiopathic*
  Bell’s, familial
Melkersson–Rosenthal syndrome (recurrent alternating facial palsy, furrowed tongue, faciolabial oedema)
Temporal arteritis
Multiple sclerosis
Myasthenia gravis
Idiopathic (Bell's) palsy
Bell's palsy means facial paralysis that has signs and symptoms consistent with the disease and no cause was found.
It includes paralysis of paresis of all muscle groups on one side of the face; sudden onset; absence of signs of central nervous system disease; and absence of signs of ear or CPA disease.
Male to female ratio is equal.
Recurrence rate 4.5 – 15 %
4.1 % has family history.
The etiology of Bell's palsy remains unclear although microcirculation failure of vasa nervorum, ischemic neuropathy, infection, genetics and immunological causes.
process include herpes simplex virus type 1 (HSV-1), herpes simplex virus type 2 (HSV-2), human herpesvirus, varicella zoster virus (VZV), influenza B, adenovirus, Coxsackie virus and Epstein–Barr virus (EBV).

The majority of patients will recover completely, but a poor prognosis has been related to:

- complete paralysis at onset or incomplete paralysis with late onset of recovery,
- old age,
- a dry eye,
- abolished taste,
- absent stapedius reflex,
- postauricular pain.

Normal function is usually regained within three months in about two-thirds of all patients. No further recovery is expected after a period of six months has elapsed.
Exercises
Predinsone 1mg /kg for 5 days then followed by ten days taper
Acyclovir 200-400 mg 5 times daily for 10 days
Facial nerve disorder of viral origin
VARICELLA ZOSTER VIRUS INFECTION (RAMSAY HUNT SYNDROME)
Ramsay Hunt syndrome is a peripheral facial nerve palsy accompanied by an erythematous vesicular rash on the ear (zoster oticus) or in the mouth, The mechanism of disease is reactivation of the latent VZV virus in the geniculate ganglion
Other cranial nerves are commonly involved. The onset of palsy is preceded by pain which may persist and be excruciating. In a small proportion of patients, the facial palsy is accompanied by a sensorineural hearing loss. The prognosis for Ramsay Hunt is worse than Bell’s palsy. Persistent weakness is observed in 30–50 percent of patients and only 10 percent recover completely after complete loss of function without treatment.

**Treatment**

Same as Bell's palsy (2-3 weeks)

Predison 1 mg / kg / day for 5 days followed by 10 days taper

IV acyclovir 250 mg 3 times daily or 800 mg orally 5 times daily
Facial nerve trauma
management of facial nerve paralysis following trauma is generally deferred until the patient is both medically and neurologically stable

MAXILLOFACIAL TRAUMA
stab wound or mandible fracture
Treatment by end to end anastmosis or interposition graft

TEMPORAL BONE TRAUMA
Longitudinal fracture---- 20 % facial palsy perigenigulate region
Transverse fractures ------ higher incidence of facial nerve paralysis (50 percent) and the labyrinthine and mastoid segments are most commonly involved
Middle ear and mastoid surgery
The most common site of injury during middle ear or 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 more than 50 percent of the circumference has been disrupted, it should be repaired with either direct suture or graft.

Parotid surgery
Cerebellopontine angle tumour surgery
Forceps delivery
more than 90% good prognosis

Facial nerve paralysis as complications of the ear infection

• Otitis media
Facial nerve paralysis may complicate both acute and CSOM due to direct involvement of the nerve by infection through Fallopian canal dehiscence or through Fallopian canal osteitis with bone erosion and nerve involvement

• MALIGNANT OTITIS EXTERNA
Malignant otitis externa is an invasive Pseudomonas or Aspergillus infection of the ear canal which may lead to skull base osteomyelitis
Facial palsy indicates advancing infection and invasion through the bony-cartilaginous junction and the fissures of Santorini, under the tympanic ring and posteriorly to the stylomastoid foramen.
Primary or secondary

Primary facial nerve tumours are rare. Schwannomas and haemangiomas are the most frequent. Any part of the nerve may be involved and multiple segments can be affected simultaneously.

Clinical features slowly progressive for function, recurrent palsy and pain.

Treatment

Poor facial function ------- resection and graft.

Good facial function ------- conservative treatment with regular imagining and clinical evaluation.

Secondary facial nerve tumors

Squamous cells carcinoma or adenoid cystic carcinoma of the parotid gland.

If the facial nerve is functioning preoperatively, the nerve can be preserved in most patients.

The facial nerve should be sacrificed if there is direct invasion of the tumour into the nerve where the tumour cannot be separated from the nerve.
Neurophysiology of pain

Pain as ‘an unpleasant sensory or emotional experience associated with actual or potential tissue damage

Most otalgia is mediated via unmyelinated pain fibres, which characteristically cause a diffuse dull ache. Myelinated fibres, such as supply skin or dental enamel, are associated with much better localization and easier diagnosis.

Pain may be nociceptive or neuropathic
Peripheral nociceptors respond to noxious stimuli, such as physical trauma, thermal or chemical injury or inflammation.

Neuropathic pain results from damage to the peripheral or central nervous systems or from an abnormality in the pain processing system. The resulting sharp, sudden, stabbing, lightning type of pain is typical of neuralgias.

Nerve supply of the ear

1. The auriculotemporal branch of V innervates the anterosuperior external canal and pinna, but also the temporomandibular joint.
2. The facial nerve makes a smaller contribution, providing some sensory input from the posterior tympanic membrane and external canal and the bowl of the concha.
3. Cranial nerve IX innervates the posterior external canal, meatus and tympanic membrane, but also the ipsilateral oropharynx. Its tympanic branch (Jacobson’s nerve) forms the tympanic plexus, innervating the middle ear cleft.
4. The auricular branch of the vagus (Arnold’s nerve) has a similar otologic distribution, but cranial nerve X has a vast dispersion to the viscera of the neck and even mediastinum.
5. The upper cervical nerves C2 and C3, via the great auricular nerve and lesser occipital nerve, supply the cranial surface of the pinna, but also the skin and muscles of the neck and cervical spine.

This rich innervation of the ear allows central misinterpretation of the origin of pain arising from throughout the head and neck and is the basis for referred otalgia.
Causes
1. From the ear
   • From the pinna
   • Trauma: tear, laceration, bite
   • Haemtaoma
   • Infected eczema
   • Perichondritis
   • Infected basal or squamous cell carcinoma

2. Refereed
b) from the meatus
1. impacted wax
2. impacted foreign body
3. otitis externa
4. Herps zoster oticus
5. keratosis obturance
6. furunculosis
7. malignant otitis externa
8. carcinoma
C) middle ear
1. bullous myringitis
2. traumatic perforation
3. OME
4. carcinoma
5. acute om
6. otitis baro trauma
7. hemotympanum
D) mastoid
1. acute mastoiditis
2. zygomatic mastoiditis
3. Bezold's abscess
4. complications of cholesteatoma
5. cholesterol granuloma

E) inner ear
1. noise
2. menieres disease
3. tinnitus
4. vestibular schwannoma
Causes of referred otalgia

• Via the V cranial N
• Lesion of the teeth and jaw
  Impaction of molar tooth, apical abscess, dental caries, malocclusion, TMJ arthritis
• Lesion of salivary gland and duct (acute infection or calculus)
• Sphenopalatine neurologia
• Lesion of the tongue, ulceration, carcinoma
B) via the X and IX CN

1. lesion of the oro and hypopharynx
   . acute pharyngitis and tonsillitis
   . parapharyngeal and retropharyngeal abscess, quinsy
   . tonsillectomy, TB, neoplasm

2. Lesion of the tongue
   Ulceration, neoplasm, infection

3. elongated styloid process causing stretching of the glossopharyngeal CN

4. Glossopharyngeal neuralgia
C) via the 2nd and 3rd cervical spinal nerve
Cervical disc lesions
Arthritis of the cervical spine
Fibrositis of the upper part of sternomastoid m

The most common cause of refereed otalgia are impaction of lower molar tooth, infection or removal of tonsil, and dental malocclusion
How to arrive at diagnosis

**History**

- **Features suggestive of primary otalgia (due to ear disease):**
  - hearing loss;
  - aural discharge;
  - vertigo;
  - unilateral rather than bilateral symptoms.

- **Symptoms suggesting referred otalgia:**
  - pain on chewing/trismus;
  - dysphagia/odynophagia;
  - hoarseness;
  - risk factors (smoking/alcohol history);
  - neck swelling/goitre;
  - cervical musculoskeletal symptoms;
  - dental history/recent treatment.

- **Features of neuropathic pain:**
  - radiation, e.g. to throat;
  - typical time course/duration;
  - quality of pain;
  - trigger zone/precipitating factors, e.g. swallowing.
Examination

- **Primary otalgia:**
  - inspection of ear and otoscopy;
  - palpation for tenderness;
  - aural examination with teleotoscope and microscope;
  - tympanometry.

- **Referred otalgia:**
  - cranial nerve (CN) examination, especially CN V, VII, IX and X;
  - palpation of cervical lymphatic chain;
  - assessment of cervical spine mobility/tenderness;
  - palpation of TMJ and pterygoid muscles;
  - exclude trismus;
  - dental inspection for caries, absent dentition and malocclusion;
  - direct and fibreoptic examination of oropharynx and laryngopharynx;
  - palpation of oropharynx to seek induration, trigger zone or styloid bone
Imaging
– Where diagnosis eludes the examiner, CT will detect skull base erosion, petrous apex disease and otherwise asymptomatic malignancies and demonstrate the styloid process. Enhanced MRI is superior in evaluating soft tissue disease, e.g. cranial nerve lesions, such as vestibular schwannoma or adenoid cystic carcinoma of the infratemporal fossa