Chapter 24: Disorders of the facial nerve

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The material presented is based on the author's experience in managing over 2000 patients over a period of 20 years. The emphasis is on management in terms of diagnosis, prognosis, and treatment. The presentation begins with applied basic science and progresses to clinical evaluation, stressing pathophysiology, differential diagnosis, special tests, natural history, and treatment of specific disorders. For more details the reader is referred to May (1986).

Embryology

Normal and abnormal presentations of the facial nerve can best be understood through an awareness of its embryonic development (Gasser, 1967a, b). The main pattern of the nerve's complex course, branching pattern, and relationships is established during the first 3 months of prenatal life. During this period the muscles of expression also differentiate, become functional, and actively contract. Important steps in facial nerve development occur throughout gestation and the nerve is not fully developed until approximately 4 years after birth (Table 24.1).

<table>
<thead>
<tr>
<th>Week of gestation</th>
<th>Structures noted</th>
</tr>
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<tbody>
<tr>
<td>Week 3</td>
<td>Collection of neural crest cells to become seventh cranial nerve identifiable</td>
</tr>
<tr>
<td>Week 5</td>
<td>Chorda tympani, greater petrosal, VII motor nucleus</td>
</tr>
<tr>
<td>Week 6</td>
<td>External genu, postauricular branch, branch to posterior belly digastric</td>
</tr>
<tr>
<td>Week 7</td>
<td>Geniculate ganglion, nervus intermedius</td>
</tr>
<tr>
<td>Week 8</td>
<td>Stapedius nerve, temporofacial and cervicofacial part of extracranial facial nerve becomes apparent</td>
</tr>
<tr>
<td>End of week 8</td>
<td>Rest of terminal branches of VII form</td>
</tr>
<tr>
<td>Week 7-8</td>
<td>Myoblasts that will form the facial muscles are noted</td>
</tr>
<tr>
<td>Week 12</td>
<td>All facial muscles are identifiable.</td>
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</table>

Congenital anomalies can be understood by relating them to embryological development. The facial nerve develops within the second pharyngeal arch during the time that closely adjacent derivatives of the first arch and first external groove and internal pouch are forming the external and middle ear regions. Anomalies of the facial nerve within the temporal bone should therefore be anticipated whenever there is an associated malformation of the external or middle ear. If the stapes or incus is deformed the surgeon should be on guard for a possibly misplaced and exposed facial nerve; a soft tissue mound over the footplate of the stapes or the promontory may actually be the facial nerve (Jahrsdoerfer, 1981).
A great variety of facial nerve arrangements have been encountered within the temporal bone (Proctor and Nager, 1982). The nerve may course with the chorda tympani nerve, bifurcate, trifurcate, or take innumerable other aberrant pathways within the temporal bone. When a large chorda tympani nerve is encountered it may be carrying motor fibres to the face. In such instances, the vertical segment of the facial nerve just distal to the point where the chorda tympani nerve branches off may dwindle to a fibrous strand and lie in a narrowed fallopian canal. This condition has been encountered in children born with facial paralysis. The nerve may be dehiscent and it may herniate into the middle ear cavity (Johnson and Kingsley, 1970). This unusual presentation of the facial nerve, when encountered during otological surgery, must not be confused with a facial nerve schwannoma. Excision or biopsy of such a structure would cause iatrogenic facial paralysis which would have to be repaired by surgery.

Anatomy

A general knowledge of the anatomy of the seventh cranial nerve is essential for diagnosis and treatment of facial nerve disorders. For example, specific differential diagnostic possibilities can be derived by localizing the site of the lesion (Table 24.2) and, in the event that surgical therapy is inappropriate, defining the level of facial nerve involvement is critical.

Table 24.2. Signs indicating probable diagnosis of lesions of the facial nerve at various levels

<table>
<thead>
<tr>
<th>Levels</th>
<th>Signs</th>
<th>Probable diagnosis</th>
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<tbody>
<tr>
<td>(I) Supranuclear Cortex</td>
<td>Tone and upper face intact, loss of volitional movement with intact spontaneous expression, slurred speech (tongue weakness), hemiparesis (arm greater than leg) on side of facial involvement.</td>
<td>Lesion of motor cortex or internal capsule on opposite side of facial involvement.</td>
</tr>
<tr>
<td>Cortex and internal capsule</td>
<td>Paresis of upper extremity begins with involvement of thumb, finger and hand movement.</td>
<td>Paresis upper extremity usually middle cerebral artery.</td>
</tr>
<tr>
<td></td>
<td>Lesion of motor cortex or internal capsule on opposite side of facial involvement.</td>
<td>Paresis lower extremity usually anterior cerebral artery.</td>
</tr>
<tr>
<td>Opercular syndrome</td>
<td>Voluntary facial and lingual movements impaired, emotional and automatic movements preserved or exaggerated. speech is dysarthric, laryngeal, sternocleidomastoid, and trapezius muscles involved.</td>
<td>Weakness of the tongue, pharynx, jaws, neck muscles, and upper extremity may occur.</td>
</tr>
<tr>
<td></td>
<td>Weakness of the tongue, pharynx, jaws, neck muscles, and upper extremity may occur.</td>
<td>EEG may not be abnormal because of depth of lesion in operculum (insula or insland of Reil complex).</td>
</tr>
<tr>
<td></td>
<td>Upper face usually not spared as with other motor cortex lesions.</td>
<td>Vascular, neoplastic, encephalitic or traumatic lesions.</td>
</tr>
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2
Extrapyramidal

Increased salivary flow, spontaneous facial movement impaired, volitional facial movement intact.

Masked face of parkinsonism or dystonia, progressive hemifacial spasm.

Grimacing and choreiform movements.

- Tumour or vascular lesion of basal ganglia.
- Parkinsonism.
- Meige's syndrome (cervical facial dystonia).

Midbrain

Involvement of face and oculomotor roots; loss of pupillary reflexes, external strabismus, and oculomotor paresis on opposite side of facial paresis.

- Unilateral Weber's syndrome (vascular lesion).

Bilateral facial paresis with other cranial nerve deficits, emotional liability, hyperactive gag reflex, marked hyperflexia associated with hypertension.

- Pseudobulbar palsy associated with multiple infarcts.

Pontine nucleus

Involvement of cranial nerves VII and VI on side of lesion with gaze palsy on side of facial paresis.

- Contralateral hemiparesis, ataxia, cerebellovestibular signs.

- Involvement of pons at level of VII and VI nuclei by pontine glioma, multiple sclerosis, encephalitis, infection, or polio.

- Contralateral hemiplegia with ipsilateral facial palsy.

- Possible lesion just above pontine facial nucleus, below decussation of corticobulbar tract.

- Internal strabismus may be present on side of facial palsy.

- Millard-Gubler syndrome, Foville's syndrome.

- Deficits of cranial nerves VII and VI noted from time of birth with or without other congenital anomalies.

- Facial motor involvement usually incomplete sparing of corner of mouth or lower lip common.

- Another type of presentation is involvement of the lower lip with complete or partial sparing of upper face.

- Anomalies of the pinna, canal or mandible associated with facial palsy indicate developmental defect of facial nerve.

- Developmental facial palsy (noted at birth).

- Oculofacial syndrome or Moebius' syndrome.

- Thalidomide toxicity.

- Non-developmental facial palsy due to facial or abducens nerve anomalies are most often due to infranuclear lesions.

(II) Infranuclear intracranial

Cerebellopontine angle (CPA)

Impairment of hearing, especially discrimination out of proportion to pure tone scores.

- Possible ataxia, abnormalities of tearing or taste, stapes reflex decay, decreased corneal sensation.

- Facial motor deficit (late sign).
Prolongation of the latency of waves III-V of auditory brainstem response (ABR).
Anomalies on CT scan (usually not enhanced with contrast).
   Acoustic neuroma (schwannoma).
Abnormalities in trigeminal, acoustic-vestibular and facial nerve function, starting with facial pain or numbness.
Lesion noted on CT (enhancement with contrast).
   Meningioma
Abnormalities of facial and acoustic-vestibular nerve function.
May start with facial twitching.
Erosion or lytic area evident on plain radiographs of temporal bone.
   Cholesteatoma or facial schwannoma arising in temporal bone.
Abnormalities of cranial nerves VII, VIII, IX, X, XI, and XII.
Pulsatile tinnitus and purple-red pulsating mass bulging through the tympanic membrane.
   Glomus jugulare tumour.
Abnormalities of abducens nerve in addition to above.
   Glomus jugulare tumour extending to petrous apex to involve middle fossa.

**Skull base**
Conductive or sensorineural hearing loss, acute or recurrent facial palsy.
Positive family history, abnormalities of bone density on skull radiograph.
   Osteopetrosis
Multiple cranial nerve involvement in rapid succession.
   Carcinomatous meningitis, leukaemia, Landry-Guillain-Barré, mononucleosis, diphtheria, tuberculosis, sarcoidosis, malignant external otitis.

**Transtemporal bone**
Internal auditory canal and labyrinthine segment of facial nerve
Ecchymosis around pinna and mastoid prominence (Battle's sign).
Haemotympanum with sensorineural hearing loss (tuning fork lateralizes to normal side), vertigo, nystagmus (fast component away from involved side).
Sudden complete facial paralysis following head trauma.
 Usually associated with basilar skull fracture, loss of consciousness and CSF leak.
Transection of facial nerve more likely with this injury compared to longitudinal fracture.
   Temporal bone fracture (transverse, longitudinal or combination).

Geniculate ganglion
Dry eye, decreased taste and salivation. Erosion of geniculate ganglion area or middle fossa demonstrated by pluridirectional tomography and CT scan of temporal bone.
Schwannoma, meningioma, cholesteatoma, haemangioma, A-V malformation.

Ear pain, vesicles on pinna, dry eye, decreased taste and salivary flow.
Sensorineural hearing loss, nystagmus, vertigo, red chorda tympani nerve.
Facial palsy may be complete, incomplete, or progress to complete over 14 days.

Herpes zoster cephalicus (Ramsay Hunt syndrome).
Same as above without vesicles, no other cause evident.
Facial palsy may be complete, incomplete, or progress to complete over 10 days.

Idiopathic (Bell's) palsy, (viral inflammatory immune disorder).
Same as above but no recovery in 6 months.

Tumour
Ecchymosis around pinna and mastoid (Battle's sign), haematotympanum.
Conductive hearing loss (tuning fork lateralizes to involved ear).
No vestibular involvement unless stapes subluxed into vestibule (causes fluctuating sensorineural hearing loss and vertigo with nystagmus).

Longitudinal fracture of temporal bone.
May be proximal or at geniculate ganglion (dry eye), or distal to geniculate ganglion (tearing symmetrical).
(Tear test valid only in acute injury).

Tympanomastoid

Decreased taste and salivation, loss of stapes reflex and symmetrical tearing.
Sudden onset facial palsy which may be complete or incomplete or may progress to complete.

Pain, vesicles, red chorda tympani.

Herpes zoster cephalicus.

Pain without vesicles, red chorda tympani.

Bell's palsy.

Red, bulging tympanic membrane, conductive hearing loss.
Usually history of upper respiratory tract infection.
Lower face may be involved more than upper.

Acute suppurative otitis media.

Foul drainage through perforated tympanic membrane.

History of recurrent ear infection, drainage, and hearing loss.

Chronic suppurative otitis media, most likely associated with cholesteatoma.

Pulsatile tinnitus, purple-red pulsatile mass noted through tympanic membrane.

Glomus tympanicum or jugulare.

Recurrent facial paralysis, positive family history, facial oedema, fissured tongue.
May present with simultaneous bilateral facial paralysis.
Melkersson-Rosenthal syndrome. 

(IV) Extracranial 
Incomplete facial nerve paresis.
Hearing, balance, tearing, stapes reflex, taste, salivary flow spared.
Penetrating wound of face; sequelae of parotid surgery; malignancy of parotid, tonsil or oronasopharynx; rarely, with benign lesion of parotid gland compressing facial nerve.
Uveitis, salivary gland enlargement, fever.
Sarcoidosis (Heerfordt's syndrome), lymphoma.

(V) Sites variable 
Bilateral facial paralysis from birth.
Moebius' syndrome.
Bilateral facial paralysis, acquired.
Landry-Guillain-Barré syndrome, sarcoidosis, mononucleosis, leukaemia, idiopathic (Bell's) palsy.
Facial paralysis, especially simultaneous bilateral facial paralysis with symmetrical ascending paralysis, decreased deep tendon reflexes, minimal sensory changes.
Abnormal spinal fluid (protein and few cells, albumino-cytological dissociation).
Landry-Guillain-Barré syndrome.
Deficits of cranial nerves VI and VII or VII, VI, and III, possibly in association with other neurological signs.
Carcinoma of nasopharynx, metastatic carcinoma from breast, ovary, prostate, meningitis, leukaemia, diabetes mellitus.

(VI) Pseudobulbar palsy 
Inappropriate or exaggerated laughing or crying.
May be associated with marked increase in jaw jerk, or gag reflex.
Polyneuritis.
Toxic, viral or vascular lesion involving bilateral corticobulbar pathways.

Cortex and internal capsule

Anatomy of the pyramidal system from the cortex to the pontine nucleus is illustrated. Facial motor nerves are represented with the forehead uppermost and the eyelids, midface, and lips located sequentially below the representation of the forehead. Note that the tracts to the lower face are crossed while innervation to the forehead is both crossed and uncrossed. Sparing of the forehead movement is considered to be characteristic of a cortical lesion. However, it is also possible to have forehead sparing with a lesion of the pontine facial nucleus, with selective lesions within the temporal bone, or even in association with an injury to the nerve in its distribution in the face. Since preservation of forehead function is not sufficient to make a diagnosis of a central lesion, other neurological signs must be looked for (Table 24.3).
Table 24.3. Signs differentiating supranuclear from infranuclear lesions

<table>
<thead>
<tr>
<th>Supranuclear</th>
<th>Infranuclear</th>
</tr>
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<tbody>
<tr>
<td>Forehead intact bilaterally.</td>
<td>Total facial palsy (usually unilateral).</td>
</tr>
<tr>
<td>Emotion intact</td>
<td>Emotion impaired.</td>
</tr>
<tr>
<td>If patient has volitional smile, but not spontaneous smile, a deeper basal ganglion lesion should be suspected.</td>
<td></td>
</tr>
<tr>
<td>If patient coughs and cries appropriately but has marked increase in jaw jerk and gag reflex in response to minor stimulus involvement of both corticobulbar tracts (pseudobulbar palsy) should be suspected.</td>
<td></td>
</tr>
<tr>
<td>Deficit of tongue, thumb, fingers, hand.</td>
<td>No deficit.</td>
</tr>
<tr>
<td>Hemiplegia on side of facial palsy.</td>
<td>Ipsilateral facial palsy with contralateral hemiplegia suggests pontine lesion near facial nucleus.</td>
</tr>
<tr>
<td>Ataxia.</td>
<td>No ataxia.</td>
</tr>
<tr>
<td>Reflexes intact (can be decreased with acute lesion).</td>
<td>Flaccid.</td>
</tr>
<tr>
<td>Drooping corner of mouth.</td>
<td>Not an isolated finding.</td>
</tr>
<tr>
<td>Slight flattening of nasolabial fold.</td>
<td>Not an isolated finding.</td>
</tr>
<tr>
<td>No muscle atrophy.</td>
<td>Muscle atrophy.</td>
</tr>
<tr>
<td>No muscle fasciculations.</td>
<td>Muscle fasciculations.</td>
</tr>
<tr>
<td>Electrical tests.*</td>
<td></td>
</tr>
<tr>
<td>MST responses equal bilaterally.</td>
<td>MST responses decreased or absent.</td>
</tr>
<tr>
<td>EEMG responses normal.</td>
<td>EEMG responses decreased or absent.</td>
</tr>
<tr>
<td>EEMG responses normal.</td>
<td>EEMG fibrillations.</td>
</tr>
<tr>
<td>MUAP present.</td>
<td>MUAP decreased or absent.</td>
</tr>
</tbody>
</table>

* MST, Maximal stimulation test; EEMG, evoked electromyography; EMG, needle electromyography; MUAP, motor unit action potential. Presence or absence of signs listed depend upon level and completeness of injury. The following alterations in function can occur with supranuclear as well as infranuclear lesions: autonomic - tearing, salivary flow; special
sensory - taste; motor function - stapes reflex (stapes nucleus separate from pontine facial motor nucleus).

**Extrapyramidal system**

The extrapyramidal system consists of the basal ganglia and the descending motor projections other than the fibres of the pyramidal or corticospinal tracts. This system provides for automatic associated movements and spontaneous, emotional, mimetic human facial language which accompanies the more precise voluntary responses. The interplay between the pyramidal and extrapyramidal system accounts for tonus and stabilizes the motor responses. The affect of parkinsonism is known to be the result of extrapyramidal pathway destruction, and the facial dystonia of Meige's syndrome, a rare clinical entity, is thought to be due to basal ganglion disease. The severe progressive hemifacial spasm that accompanies Meige's syndrome will be discussed further under central nervous system facial nerve disorders.

Emotion is another function of the extrapyramidal cortical system and is mediated by discharges passing through the cingulate, orbital, and other frontal cortical areas and the basolateral portion of the amygdala.

**Upper midbrain**

A lesion in the upper midbrain will involve the oculomotor pathways and result in ipsilateral loss of direct and consensual pupillary light reflexes, ipsilateral external strabismus, and oculomotor paresis. In addition, paresis of contralateral muscles of the head and body will be noted. This symptom complex is referred to as unilateral Weber's syndrome.

**Lower midbrain**

A lesion in this region that is above the facial nerve nucleus involves the tracts of the abducens and may cause contralateral paresis of the face and muscles of the extremities, ipsilateral abducens paresis, and internal strabismus. A lesion that extends far enough laterally to include the emerging facial nerve fibres may present as peripheral ipsilateral facial paralysis associated with loss of taste and papillae on the anterior two-thirds of the tongue, and a dry eye on the same side. In addition, salivary flow from the submaxillary gland on the side of the lesion may be greatly diminished or absent.

It is important to emphasize that the peripheral topognostic tests for tearing, taste, and lacrimal flow can be altered by supranuclear lesions. However, a lesion in this region of the brainstem would involve other neural functions as well, and would be highly unlikely to involve only facial function.

**Pontine nucleus**

The facial motor nucleus contains approximately 7000 neurons and is seated in the lower third of the pons, beneath the fourth ventricle. The neuronal processes that leave the nucleus pass around the abducens nucleus (cranial nerve VI) before emerging from the brainstem. A peripheral seventh nerve paralysis, an internal strabismus on the same side, and inability to turn the non-paralysed eye toward the nose when asked to look toward the
paralysed side of the face, suggest a single lesion near the floor of the fourth ventricle involving the sixth and seventh cranial nerves. A lesion near the ventricle at the level of the superior salivary nucleus causes peripheral facial paralysis, a dry eye, paralysis of voluntary muscles, loss of following gaze toward the side of the facial paralysis, and often vertical or rotatory nystagmus.

**Cerebellopontine angle**

The facial nerve emerges from the brainstem with a more slender nerve, the nerve of Wrisberg or nervus intermedius. Because of the association of the facial nerve with the nervus intermedius and the vestibuloacoustic nerve at the level of the cerebellopontine angle and in the internal auditory canal, tearing, taste, submandibular salivary flow, and hearing and balance may be disturbed with a facial nerve lesion at this level. Large lesions filling the cerebellopontine angle may compress other cranial nerves and cause deficits of the fifth cranial nerve and later the ninth, tenth, and eleventh cranial nerves. Lesions that may occur in the area include temporal bone fractures, acoustic neuromata (schwannomata), meningiomata, primary cholesteatomata, and perhaps hyper- and hypokinetic disorders from vascular cross-compression of cranial nerves.

**Transtemporal bone portion of the facial nerve**

An understanding of the gross and microscopic anatomical relationships between the facial, acoustic, and vestibular nerves, described by Silverstein and Norrell (1980), is essential for performing a retrolabyrinthine vestibular neurectomy. The intracranial segment of the facial nerve from the brainstem to the fundus of the internal acoustic meatus is covered only by a thin layer of glia, which makes it quite vulnerable to any type of surgical manipulation but also quite resistant to a slow process of stretching or compression. Thus, the facial nerve in this region can become quite elongated and spread out over the surface of a sizeable but slow-growing vestibular nerve schwannoma without any gross evidence of facial weakness.

**Fallopian canal**

The course of the facial nerve through the fallopian canal is unique. No other nerve in the body covers such a long distance through a bony canal. The nerve is also remarkable for the Z shape of its infratemporal portion, in that it has a ganglion, and that the length of its course is 28-30 mm. The nerve in the fallopian canal can be divided into three segments: labyrinthine, tympanic, and mastoid. The labyrinthine segment is the thinnest part of the facial nerve within the fallopian canal. The narrowest part is at its entrance, where it averages 0.68 mm in diameter (Fisch and Esslen, 1972). Fisch (1977) feels that this bottleneck at the entrance of the fallopian canal predisposes the nerve to strangulation in cases of oedematous swelling. The observation is supported by post-mortem findings reported by Fowler (1963) and by Proctor, Corrigill and Proud (1976). The blood supply to the nerve in this region is unique; this is the only segment of the facial nerve in which there are no anastomosing arterial arcades.

The labyrinthine segment of the facial nerve includes the geniculate ganglion. The somatosensory (pain), and special sensory (taste) fibres are afferent fibres that synapse in the geniculate ganglion, while the autonomic secretomotor fibres to the lacrimal gland pass
through the geniculate ganglion and form the first branch of the facial nerve, the greater petrosal nerve. The secretory fibres to the parotid gland are carried with the ninth cranial nerve. They travel through the tympanic plexus and form the lesser petrosal nerve. There are communications with the nervus intermedius, which provides an alternate route for the parasympathetic fibres to reach the parotid, thus bypassing the tympanic plexus and the ninth cranial nerve branch of Jacobson. This might explain why sectioning Jacobson's nerve, in many cases, have little effect on parotid salivary flow.

In the region of the geniculate ganglion there are ample alternative pathways and connections for parasympathetic fibres to reach their terminations. Such alternative pathways explain how lacrimal flow may be unaffected by slow-growing lesions at or proximal to the geniculate ganglion, and the spontaneous recovery of tearing following resection of the geniculate ganglion or nervus intermedius, such as might occur with posterior fossa surgery. The geniculate ganglion lacks a bony covering in approximately 15% of temporal bones, an arrangement which makes the facial nerve quite vulnerable to injury during surgery involving the middle cranial fossa, especially in children. Further, the bone of the tegmen tympani and middle fossa plate over this region may be quite thin.

In the author's experience with temporal bone fractures, this is the area of the facial nerve most often compressed by crushed, thin, bony fragments. The change in direction taken by the facial nerve at the genu is another reason why this site is the most common focus of injury when severe traction is applied to the nerve along the axis of its tympanic segment, as may occur in longitudinal fracture of the petrous pyramid. The fact that the arachnoid pia mater extends to the geniculate ganglion, as well as the complex embryological development of this portion of the nerve, may explain why this area of the facial nerve is so often the site of primary cholesteatomata, vascular malformations, meningiomata, and schwannomata (Fisch, 1977).

The geniculate ganglion marks the proximal end of the tympanic portion, and from this point the nerve courses 3-5 mm, before passing just behind the cochleariform process and the tensor tympani tendon. The cochleariform process is a useful landmark to find the facial nerve when other landmarks are obscured by granulation tissue or cholesteatoma, or in cases of trauma. The entire tympanic segment is approximately 8-11 mm long and the tympanic wall of this part of the fallopian canal is thin and easily fractured. In addition, dehiscences occur frequently, allowing the uncovered nerve to prolapse into the oval window niche, partly or completely concealing the footplate of the stapes; this makes the nerve subject to trauma during stapes surgery. The tympanic segment is divided from the mastoid portion by the pyramidal eminence.

At this point the fallopian aqueduct makes another turn downward, forming the second genu. The latter is another area where the facial nerve is vulnerable to injury during mastoid surgery. The distal aspect of the tympanic segment is found by the surgeon through the mastoid approach by entering the suprapyramidal recess (retrofacial recess). Here, the facial nerve is lateral and distal to the pyramidal process. In the presence of chronic infection, care must be taken not to confuse a pathological dehiscence of the facial nerve in this region with a mound of granulation tissue. The best way to avoid this is to identify the nerve proximal and distal to the area that looks suspicious. The second genu, which marks the beginning of the mastoid segment, is lateral and posterior to the pyramidal process, which houses the
stapedius muscle that lies on the deep side of the facial nerve; this explains the fact that the facial nerve lies lateral to the pyramidal process. The nerve continues vertically down the anterior wall of the mastoid process to the stylomastoid foramen. The distance from the beginning of the second genu to the stylomastoid foramen varies between 10 and 14 mm. This segment of the facial nerve has three branches:

1. the nerve to the stapedius muscle
2. the chorda tympani nerve
3. the nerve from the auricular branch of the vagus.

The nerve to the stapedius muscle arises from small neurons within the pons, located outside the main facial nerve nucleus, which interface with the rostral end of the facial nucleus and the caudal end of the lateral superior salivatory nucleus (Lyon, 1978; Joseph et al, 1985). Although Lyon (1978) studied cats and Joseph et al (1985) studied rabbits to determine the location of the motor neurons relative to the stapedius muscle, it is quite likely that these neurons lie in a similar location in man. If so, this may help to explain why alterations in the middle ear reflex occur when a brainstem lesion is present. Further, the separate nucleus for the stapedius muscle innervation provides the anatomical basis for sparing of the stapedius muscle in patients with congenital facial palsy such as Moebius' syndrome.

**Surgical landmarks to identify the facial nerve**

The facial nerve will usually be found just deep to the short process of the incus, in a line between the short process of the incus and the anterior extent of the digastric ridge. The facial nerve is thus posterior to the chorda tympani nerve and just lateral to the ampullary end of the posterior semicircular canal. Skeletonizing the posterior canal is helpful in order to avoid fenestrating this part of the labyrinth. The tympanomastoid suture line is another useful landmark since it lies just anterior to the facial nerve and close to the course of the chorda tympani nerve. The chorda tympani nerve and facial nerves are deep to this suture. The facial nerve lies anterior to the sigmoid sinus and leaves the temporal bone through the stylomastoid foramen just anterior and lateral to the sigmoid sinus, where the digastric ridge turns and runs in the direction of the stylomastoid foramen.

**Facial nerve sheath**

The sheath that surrounds the facial nerve through its course in the fallopian canal consists of periosteum, epineurium, and perineurium. Although surgical decompression and opening of the perineurium of the facial nerve are controversial in the management of Bell's palsy and herpes zoster cephalicus, opening the sheath is imperative in cases of suspected tumour or trauma. A tumour of the facial nerve may be discovered when the sheath is opened, or a traumatic haematoma may be found compressing the nerve deep to the sheath. Finally, when the nerve has been disrupted, it is necessary to open the sheath to find the proximal and distal ends for repair.
**Spatial orientation**

Agreement is lacking, in spite of efforts to determine it, as to whether or not the facial nerve is spatially oriented in its extra-axial course from the brainstem to the periphery, as it is in the cortex and pontine nucleus. Evidence against topographical organization of the facial nerve fibres has come from several investigators who have found that the fibres destined for each peripheral branch are diffusely located in the facial nerve trunk (Sunderland and Cossar, 1953; Harris, 1968; Sade, 1975; Thomander, Aldskogius and Grant, 1982).

Thomander, Aldskogius and Grant (1982) exposed the individual peripheral facial nerve branches to horseradish peroxidase, permitting retrograde transport of the tracer to demonstrate the location of these fibres in the cat facial nerve trunk. The study indicated that the fibres to each peripheral branch were diffusely arranged in the facial nerve trunk at least as far proximally as the tympanic segment.

Gacek and Radpour (1982) studied the cross-sectional anatomy of the facial nerve through its course in the temporal bone by making discrete lesions in the facial nerve of the cat proximal to the geniculate ganglion and documenting anterograde wallerian degeneration. Gacek and Radpour (1982) discovered degenerated myelin sheaths in all three of the peripheral branches studied, regardless of whether the lesion involved the rostral, caudal or middle fascicles of the facial nerve. They concluded that small fascicles of the facial nerve at the level of the internal auditory meatus carried motor fibres to all peripheral branches, and that motor axons of the facial nerve in the cat are not topographically arranged in the facial nerve trunk, as had previously been proposed. Jannetta (1975) described 31 patients with hemifacial spasm treated by removing a vessel compressing the facial nerve in the cerebellopontine angle. In those cases where the compressing vessel was found on the cephalic aspect of the nerve, the spasm was more severe in the upper part of the face. In cases where the vessel compressed the caudal aspect of the nerve, the spasm began in the lower face in an atypical fashion. This observation lends support to the existence of spatial orientation of the nerve in its most proximal intracranial portion.

Considering all the evidence, it is likely that there is some degree of spatial organization of facial nerve fibres, especially at the level at which the axon processes leave the brainstem nucleus and course toward the periphery. Accepting the fact that the peripheral facial nerve is at best only partially topographically oriented, with some axons carried with the upper division terminating in muscle groups of the lower face and vice versa, it is understandable that regeneration following facial nerve injuries usually results in some degree of mass movement and synkinesis.

**Blood supply**

The nerve receives its nourishment from the anterior inferior cerebellar artery, which enters the internal auditory meatus in close association with the seventh and eighth cranial nerves, the petrosal branch of the middle meningeal artery which runs along with the greater petrosal nerve, and the stylomastoid branch of the postauricular artery, which enters the facial canal at the stylomastoid foramen. The territories supplied by the three arteries tend to overlap at any given level. As mentioned previously, the anastomosis between the arterial systems is immediately proximal to the geniculate ganglion, making this segment of the facial nerve
vulnerable to ischaemia from oedema. This might have bearing on the pathogenesis of facial paralysis following embolization of the middle meningeal artery (Metson and Hanson, 1983).

**Extracranial segment of the facial nerve**

The facial nerve leaves the fallopian canal at the stylomastoid foramen. In newborns and in children up to 2 years of age, the facial nerve as it exits the skull is just deep to the subcutaneous tissue underlying the skin. After 2 years of age, as the mastoid tip and tympanic ring form, the facial nerve takes a deeper position and, in an adult, it may be up to 5 cm below the level of the skin. Beyond the age of 2 years, the facial nerve is protected by the tympanic bone, the mastoid tip, the ascending ramus of the mandible, and the fascia between the parotid and cartilaginous external canal.

The position of the facial nerve in the young child must be kept in mind by the otologist and head and neck surgeon. To avoid unintentional injury to the facial nerve, a postauricular incision should be modified to avoid coursing near the junction of the tympanic ring and mastoid tip, and this area should be protected by placing a finger over the area at the time the incision is made. The surgeon is cautioned not to depend upon a nerve stimulator to find the facial nerve in the region of the stylomastoid foramen. A muscle response may be noted in spite of the fact that the stimulator is not directly on the facial nerve, or the stimulator may give no response when on the facial nerve, if a thin layer of connective tissue is insulating the nerve. The nerve must therefore be identified by its anatomical location and appearance.

The main trunk may be identified entering the substance of the parotid and then bifurcating into an upper and a lower division. The facial nerve passes through the parotid gland and emerges over the fascia of the masseter muscle. There are communications between the upper and lower divisions in the majority of patients, and these form a variety of patterns. The rich plexus of nerve filaments that forms in the peripheral zone, just before entering the undersurface of the facial muscles, provides for free intermingling between branches carried by the upper and lower divisions, which may explain the diffuse distribution of axons within the main trunk of the facial nerve throughout its course from the brainstem.

**Communications of the facial nerve**

There are diffuse intra-axial connections within the central nervous system and, in addition, the facial nerve communicates with the vestibulocochlear nerve within the internal auditory meatus, with the otic ganglion and sympathetic fibres in the area of the geniculate ganglion and, just before it leaves the stylomastoid foramen, with the auricular branch of the vagus nerve. Outside the stylomastoid foramen the facial nerve communicates with the glossopharyngeal nerve, the vagus nerve, the great auricular nerve, and the auriculotemporal nerve. The peripheral branches communicate behind the ear with the lesser occipital, on the face with branches of the trigeminal, and in the neck with the cervical cutaneous nerve. These relationships have been documented by the meticulous dissections of Bischoff (1977). The fact that myriads of strands of the facial nerve interconnect with the fifth, seventh, eighth, ninth, tenth, eleventh, and twelfth cranial nerves, and with the cervical cutaneous nerves, may help to explain the symptoms of many syndromes; head and face pain, and ear, throat, eustachian tube, and neck pain. These syndromes are extremely hard to treat when the cause
is malignant disease or a functional imbalance such as that which causes cluster headaches or atypical facial neuralgia. These interconnections also explain mastoid, ear, face or neck pain associated with Bell's palsy and herpes zoster cephalicus, the presence of residual facial sensation after the trigeminal nerve has been cut, preservation of taste and tearing after facial nerve severance, and the occurrence of pain with skull base cancer after resection of the fifth, seventh, ninth, or tenth cranial nerve, the first or second cervical nerve, or the nervus intermedius.

**Spontaneous recovery of facial nerve function**

This free intermingling fibres of the facial nerve with fibres of other neural structures (particularly the fifth cranial nerve) has been proposed as the mechanism of spontaneous return of facial nerve function after peripheral injury to the nerve. Although spontaneous recovery of facial function was noted in approximately 25% of patients studied by Martin and Helsper (1957), this potential should not be relied upon for spontaneous reanimation of the face following resection of the facial nerve. There is no question that appropriate nerve repair at the earliest possible time following injury yields the best results. Nevertheless, spontaneous recovery does occur and may play a part in some of the cases in which the results of surgical reanimation are superior.

One other mechanism for the spontaneous recovery of facial function should be discussed. The plasticity hypothesis was first proposed by Cajal (1894), and was discussed in detail by Kandel (1977). This hypothesis offers the most plausible explanation, not only for spontaneous recovery of facial function following facial nerve sectioning, but also for repair after interruption of infranuclear pathways. The plasticity hypothesis, according to Cajal, is based on pre-existing connections between groups of cells that are reinforced by multiplication of terminal branches of protoplasmic appendices and nerve collateral, thus bringing about functional transformations in particular systems of neurons as the result of appropriate stimuli or their combinations.

**Neuropathophysiology**

**Nerve injury**

The facial nerve carries approximately 10,000 fibres, of which 7000 are myelinated motor axons that reach the facial muscles (Van Buskirk, 1945). It must be understood that non of the various injuries and disorders involving the facial nerve causes an all-or-nothing lesion, but rather each of the fibres is capable of being spared or injured to a different degree at any one time.

**Classification of injury and recovery**

Sunderland (1978) described five possible degrees of injury that a peripheral nerve fibre might undergo. This classification system is depicted diagrammatically and is more comprehensive than the classification of Seddon (1943), which described only neuropraxia, axonotmesis, and neurotmesis. The Table 24.4 shows the pathological changes that occur in the nerve and the anticipated responses of the nerve to electrical testing, as well as the type of recovery that might be expected with the various types of injuries. The span of possibilities
in terms of electrical responses, as well as recovery, reflects the possible mixtures of degree of injury which might occur. The five degrees of injury suggested by Sunderland describe very nicely the pathophysiological events associated with all types of disorders that afflict the facial nerve. The first three degrees of injury can occur with the viral inflammatory immune disorders, such as Bell's palsy and herpes zoster cephalicus. The fourth and fifth degrees of injury occur when there is disruption of the nerve, as in transection, which might occur during surgery, as a result of a severe temporal bone fracture, or from a rapidly growing benign or malignant tumour.

Fortunately, the pathological processes causing facial paralysis in patients with Bell's palsy and herpes zoster cephalicus usually do not progress past the first or second degree of injury, which accounts for the fact that most individuals recover satisfactorily. A similar process causes facial paralysis due to acute suppurrative otitis media, chronic otitis media associated with a cholesteatoma, slow-growing benign neoplasms, and temporal bone fractures. In each of these disorders, the nerve is usually not transected, but rather compressed. In acute otitis media and trauma, compression may be sudden or slowly progressive, evolving over 5-10 days, just as is noted with Bell's palsy and herpes zoster cephalicus. However, unlike the process that occurs with Bell's palsy or herpes zoster cephalicus, and these other disorders pressure is exerted on the nerve from without rather than from within the intraneural space; nevertheless, the results of compression of the nerve are the same. Eventually, axoplasm is dammed up, compression of venous drainage leads to further compression of the nerve and loss of axons, and eventually loss of endoneural tubes which leads to third-degree injury. In fourth- or fifth-degree injury, since most or all of the endoneural tubes have been disrupted, as well as the perineurium in the fourth-degree injuries and the perineurium and epineurium in the fifth-degree injuries, recovery even under ideal conditions is never as good as with the first three degrees.

**Correlation of degree of injury, morphological changes in the nerve, and expected type of recovery.**

- First degree: compression.

- Second degree: interruption of axoplasm and myelin.

- Third degree: disruption of endoneurium.

- Fourth degree: disruption of endoneurium and perineurium.

- Fifth degree: transection of nerve.

- Regeneration: as the degree of injury becomes more severe the quantity and quality of recovery become worse.

(From May (1986)).
**Altered function of the facial nerve following injury**

Three major changes that occur in the axon following regeneration may contribute to a combination of hypo- and hyperkinesis:

1. The distance between the nodes of Ranvier is altered.
2. The newly formed axons are covered with myelin that is much thinner than the normal axon.
3. There is a splitting and crossing of axons that reinnervate denervated muscle groups without necessarily corresponding to the cell body-motor unit arrangement that was present prior to degeneration.

As a result of these factors a tic or involuntary twitching occurs. In addition, inappropriate movement may be noted, such as movement of the mouth with blinking, or closing of the eye with smiling. Another cause of abnormal facial movements following regeneration may be changes that occur at the myoneural junction. In addition to these factors, it is quite likely that there are changes within and around the facial nerve nucleus in the brainstem, as well as alterations in central connections to the cell body. The combination of these factors may lead to spasms that occur on the involved side of the face, causing the eye to close and the corner of the mouth to pull. These spasms may be quite painful.
Table 24.4. Neuropathology and spontaneous recovery correlated with degree of facial nerve injury

<table>
<thead>
<tr>
<th>Degree of injury</th>
<th>Pathology of injury</th>
<th>Clinical recovery begins</th>
<th>Spontaneous recovery - result one year postinjury</th>
</tr>
</thead>
<tbody>
<tr>
<td>EEMG (evoked electromyography) MST (maximal stimulation test) Neurobiology of recovery</td>
<td>Normal.</td>
<td>No morphological changes noted.</td>
<td>1-4 weeks.</td>
</tr>
<tr>
<td>Neurobiology of recovery</td>
<td>Grade I: Complete: without evidence of faulty regeneration.</td>
<td>Normal.</td>
<td>1-4 weeks.</td>
</tr>
<tr>
<td>Neurobiology of recovery</td>
<td>Grade II: Fair: some noticeable difference with volitional or spontaneous movement, minimal evidence of faulty regeneration.</td>
<td>Normal.</td>
<td>1-2 months.</td>
</tr>
<tr>
<td>Neurobiology of recovery</td>
<td>Grade III-IV: Moderate to poor: obvious incomplete recovery to crippling deformity with moderate to marked complications of faulty regeneration.</td>
<td>Normal.</td>
<td>2-4 months.</td>
</tr>
<tr>
<td>Neurobiology of recovery</td>
<td>Grade V: Motion barely perceptible.</td>
<td>Normal.</td>
<td>4-18 months.</td>
</tr>
<tr>
<td>Neurobiology of recovery</td>
<td>Grade VI: None.</td>
<td>Normal.</td>
<td>Never.</td>
</tr>
</tbody>
</table>

Classification by groups I-VI modified from House and Brackmann (1985).
Facial hyperkinesis may be due to another mechanism referred to as ephaptic transmission. This term describes facial hyperkinesis or a hemifacial spasm that seems to occur spontaneously, without any discoverable cause. It is theorized that depolarization at the site of injury acts as a stimulus to the intact portion of the fibre, and that the action potential in one fibre is capable of exciting adjacent fibres in the area of injury. Granit, Leksell and Skoglund (1944) demonstrated ephaptic transmission at the site of compression in a nerve that was still capable of transmitting impulses across the site, and Kugelberg and Cobb (1951) demonstrated an acute, reversible phenomenon in the peripheral nerve of man. After producing ischaemia by means of pneumatic cuffs, Kugelberg demonstrated the development of foci of spontaneous, repetitive, and synchronized discharges, both during the ischaemia and after release of the cuff.

**Synkinesis**

This is an abnormal synchronization of movement, occurring with voluntary and reflex activity of muscles that normally do not contract together. This phenomenon may be grossly deforming and debilitating. In its worst form, mass movement of all parts of the involved side of the face occurs; the patient is unable to move each part of the face separately. In its subtlest form it may consist of no more than a tiny twitch of the chin accompanying blinking on the side of the involvement. This may be the only sign of previous facial paralysis, and to detect it requires very close observation.

**Crocodile tears**

Increased unilateral lacrimation on the involved side associated with eating may occur with a severe denervating lesion when it involves the facial nerve at or above the site of the geniculate ganglion or along the greater petrosal nerve. This phenomenon is probably the result of faulty regeneration of parasympathetic fibres, which innervate the lacrimal gland instead of the salivary glands.

**Stapedius tendon contraction**

This is a hyperkinetic syndrome which occurs with faulty regeneration and causes fullness or roaring in the ear. The complaint is noted with facial movements and often coincides with facial spasm. The diagnosis can be confirmed on tympanometric recordings employing the electroacoustic bridge; sectioning the stapedius tendon through a tympanotomy approach has been effective in relieving the spasm.

**Hemifacial spasm**

Unilateral facial nerve hyperactive dysfunction is characterized by the onset of mild intermittent spasms in the orbicularis oculi muscle that gradually increase in severity and frequency and spread downward to include all of the muscles of facial expression, including the platysma. The most common cause is cross-compression from vessels in the posterior fossa. On rare occasions this syndrome may be mimicked by benign lesions in the parotid gland (Horne, Crumley and Schindley, 1981), temporal bone (Brooken, Pulec and Haleberg, 1969; Jackson et al, 1980), or cerebellopontine angle tumours such as meningiomata, cholesteatomata, and schwannomata (May and Hardin, 1977). The most effective treatment
of hemifacial spasm is vascular decompression of the nerve at its root entry zone by the retromastoid approach (Jannetta et al., 1977).

**Facial myokymia**

A continuous fine fibrillar or undulating movement of the facial muscles gives the face an appearance suggesting a 'bag of worms'. This condition has been associated with multiple sclerosis and intrinsic tumours of the brainstem.

**Blepharospasm**

Involuntary spasmodic eye closure may start on one side but classically it is a bilateral disorder. It is characterized by symmetry and the electromyogram shows that individual contractions are asynchronized. This condition has not been noted in children. Treatment involves selective neurolysis or myolysis or, more recently, injections of botulinum A toxin (Biglan, May and Walden, 1986).

**Psychogenic or habit tic**

This condition is usually noted in children. The movements are repetitive and may involve muscles outside the distribution of the seventh nerve. There is a compulsion to perform facial movements and they are under voluntary control. These movements are not observed during sleep, as are the movements of hemifacial spasm, facial myokymia, and blepharospasm. Psychological evaluation and treatment are indicated for this disorder.

**Focal cortical seizures**

These movements involve the face and are usually tonic, often spreading beyond the distribution of the seventh nerve. After a seizure, there may be transient postictal facial paralysis of the supranuclear type that spares the forehead muscles. The results of electroencephalographic recording during a seizure are diagnostic of this condition.

**Evaluation of facial nerve function**

**Differential diagnosis**

Peripheral facial paralysis is a diagnostic challenge. Every effort must be made to determine the aetiology, since often a treatable cause can be found. The differential diagnostic possibilities are numerous (Table 24.5). However, diagnostic clues are obtained from a carefully taken history, from the findings upon physical examination, and from the results of special tests (Tables 24.6 and 24.7). The relative incidence of the variety of causes in the author’s experience can be noted in Table 24.8. In spite of the fact that, in the majority of patients, a cause cannot be found and their condition is labelled idiopathic (Bell's palsy), the clinician must not be discouraged from taking the time required to make an accurate diagnosis, since without this approach a treatable, progressive, or life-threatening disorder may be overlooked. It must be emphasized that Bell's palsy is a diagnosis by exclusion (Table 24.9).
Table 24.5. Causes of facial palsy identified in a review of medical literature (1900-1983).

<table>
<thead>
<tr>
<th>Category</th>
<th>Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Birth</td>
<td>Birth Moulding, Forceps delivery, Dystrophia myotonica, Moebius’ syndrome (facial diplegia associated with other cranial nerve deficits)</td>
</tr>
<tr>
<td>Trauma</td>
<td>Basal skull fracture, Facial injuries, Penetrating injury to middle ear, Altitude paralysis (barotrauma), Scuba diving (barotrauma), Lightning</td>
</tr>
<tr>
<td>Neurological</td>
<td>Opercular syndrome (cortical lesion in facial motor area), Millard-Gubler syndrome (abducens palsy contralateral hemiplegia due to lesion in base of pons involving corticospinal tract)</td>
</tr>
<tr>
<td>Infection</td>
<td>External otitis, Otitis media, Mastoiditis, Chicken pox, Herpes zoster cephalicus (Ramsay Hunt syndrome), Encephalitis, Poliomyelitis (type I), Mumps, Mononucleosis, Leprosy, Coxsackievirus, Malaria, Syphilis, Scleroma, Tuberculosis, Botulism, Acute haemorrhagic conjunctivitis (enterovirus 70), Gnathostomiasis, Mucormycosis, Lyme disease</td>
</tr>
<tr>
<td>Metabolic</td>
<td>Diabetes mellitus, Hyperthyroidism, Pregnancy, Hypertension, Acute porphyria</td>
</tr>
<tr>
<td>Neoplastic</td>
<td>Cholesteatoma, Seventh nerve tumour, Glomus jugulare tumour, Leukaemia, Meningioma, Haemangioblastoma, Sarcoma, Carcinoma (invading or metastatic), Anomalous sigmoid sinus, Haemangioma of tympanum, Hydradenoma (external canal), Facial nerve tumour (cylindroma), Schwannoma, Teratoma, Hand-Schüller-Christian disease, Fibrous dysplasia, von Recklinghausen's disease</td>
</tr>
<tr>
<td>Toxic</td>
<td>Thalidomide (Miehlke syndrome, cranial nerves VI, VII with congenital malformed external ears and deafness), Tetanus, Diphtheria, Carbon monoxide</td>
</tr>
<tr>
<td>Iatrogenic</td>
<td>Mandibular block anaesthesia, Antitetanus serum, Vaccine treatment for rabies, Postimmunization, Parotid surgery, Mastoid surgery, Post-tonsillectomy and adenoidectomy, Iontophoresis (local anaesthesia), Embolization, Dental</td>
</tr>
</tbody>
</table>
**Idiopathic**
Bell's, familial
Melkerson-Rosenthal syndrome (recurrent alternating facial palsy, furrowed tongue, faciolabial oedema)
Hereditary hypertrophic neuropathy (Charcot-Marie-Tooth disease, Déjérine-Sottas disease)
Autoimmune syndrome

**Temporal arteritis**
**Thrombotic thrombocytopenic purpura**
**Polyarteritis nodosa**
**Landry-Guillain-Barré syndrome** (ascending paralysis)
**Multiple sclerosis**
**Myasthenia gravis**
**Sarcoidosis (Heerfordt syndrome - uveoparotid fever)**
**Osteopetrosis.**

**History**

The type of onset of facial palsy is not diagnostic, whether incomplete, complete, sudden, or delayed. All of these patterns of onset have been noted with idiopathic (Bell's) palsy, as well as with other conditions in which the facial nerve may be compressed or invaded within its anatomical course from the brainstem to the parotid. These other conditions include herpes zoster cephaliicus, temporal bone fractures, parotid or otological surgery, infections, and neoplasms. However, the type of onset may have prognostic significance. Complete recovery will most likely occur in cases of incomplete palsy that do not progress to complete palsy. The exception is the patient who does not begin to recover in 3-6 weeks or if the paresis progresses for more than 3 weeks; in such cases a tumour must be considered as the underlying cause. Although slow progression beyond 3 weeks is diagnostic of a tumour, progression that occurs within the first 10 days of onset has been noted with idiopathic (Bell's) palsy, herpes zoster cephaliicus, external blunt trauma, and surgical trauma to the facial nerve within the parotid, temporal bone, or posterior fossa.

Half of the patients with Bell's palsy present with a sudden complete onset of facial paralysis. In spite of this, it is not diagnostic of Bell's palsy since the onset was noted to be sudden and complete in 40% of patients with confirmed tumours involving the facial nerve. In half of these patients the tumour was malignant. A sudden complete onset associated with trauma may indicate that the facial nerve has been transected, while a history of a delayed onset or a slowly progressive onset would rule out nerve transection.

Facial paralysis has been noted to recur with idiopathic (Bell's) palsy, Melkerson-Rosenthal syndrome, and tumours. The incidence of recurrence in the author's experience with Bell's palsy was 12%, with 36% on the same side and 64% on the opposite side. The incidence of patients with idiopathic (Bell's) palsy who had ipsilateral recurrence was 4%. Of the total number of patients in this study who had ipsilateral recurrent facial palsy, 17% had tumours. Thus, the onset of facial palsy is not, of itself, diagnostic; tumours, like Bell's palsy, can present with incomplete, complete, sudden, delayed, or recurrent ipsilateral peripheral facial palsy.

In contrast to recurrent facial paralysis on the same side, recurrence involving the opposite side is almost always diagnostic of idiopathic (Bell's) palsy, since alternating recurrent facial paralysis has been noted only rarely with other disorders.
Melkersson-Rosenthal syndrome is the most common example of a rare disorder that is characterized by recurrent alternating facial palsy. This syndrome is characterized by:

1. recurrent alternating facial palsy
2. recurrent oedema of the lips, face, and eyelids
3. cheilitis
4. fissured tongue.

Most authors agree that the presence of any two of these four manifestations permits the diagnosis. The syndrome may be accompanied by migraine phenomena (Stevens, 1965).

**Malignancies**

A history of cancer, particularly involving the breast, lung, thyroid, kidney, ovary, or prostate, associated with a facial paralysis suggests that a metastatic lesion is causing the palsy. Appropriate radiographic and laboratory studies are indicated to search for the primary site as well as to localize the site of facial nerve involvement. In some cases, surgical exploration of the temporal bone and extracranial course of the facial nerve is recommended to locate the lesion.

**Bilateral simultaneous palsy**

Bilateral facial nerve paresis may be a medical emergency and presents a special diagnostic and therapeutic challenge. The therapeutic challenge is early diagnosis and appropriate treatment of a potentially progressive and life-threatening disorder. The most common cause of acute simultaneous bilateral palsy in the author's series was Guillain-Barré syndrome. Other less common causes included idiopathic (Bell's) palsy, leukaemia, bulbar palsy, sarcoidosis, skull fracture, Moebius' syndrome, and myotonic dystrophy. Guillain-Barré syndrome, acute leukaemia, and bulbar palsy due to rabies immunization presented as life-threatening medical problems.

**Differential diagnosis of bilateral facial palsy by physical findings**

**Guillain-Barré syndrome**

Guillan-Barré syndrome is an acute inflammatory polyradiculoneuropathy evolving as a paralytic disease of unknown cause. The characteristic pathological feature of Guillain-Barré syndrome is a lymphocytic cellular infiltration of peripheral nerves and destruction of myelin. The major complaint is weakness with the severity of the motor weakness covering a wide continuous spectrum from mild ataxia to total paralysis of every motor and cranial nerve. In most instances it is noticed first in the legs, but can begin in the arms. Tendon reflexes are abolished in the affected areas and facial diplegia is seen in at least half of the cases. Weakness can evolve to total motor paralysis and, when respiratory muscles become involved, respiratory embarrassment may lead to death. Abnormal cerebrospinal fluid findings are characteristic of this disorder, although in the first few days cerebrospinal fluid may be normal. After several days, the cerebrospinal fluid protein begins to rise and may become very high and peak at approximately 4-6 weeks after the onset of clinical symptoms. Cells in the cerebrospinal fluid are not prominent. The absence of cells in conjunction with an
elevated protein level is the 'albumino-cytological dissociation' which at one time was thought to be characteristic of the disease. Guillain-Barré syndrome is a recognizable disease entity; its diagnosis is based on clinical, laboratory, and electrodiagnostic findings. In the author's experience, the prognosis for spontaneous recovery in Guillain-Barré syndrome is the same as for idiopathic (Bell's) palsy.

**Infectious mononucleosis**

Infectious mononucleosis is characterized by fluctuating fever, sore throat, and lymphadenopathy. Uncommonly, unilateral, recurrent, and simultaneous bilateral facial paralysis has been caused by this disorder. The syndrome of infectious mononucleosis, caused by Epstein-Barr virus, has a classical presentation and can often be diagnosed on clinical grounds. The prodrome lasts from 3 to 5 days, and consists of headache, malaise, myalgia, and fatigue. Sore throat occurs in the first week and is the most common feature of infectious mononucleosis. A greyish-white exudative tonsillitis is practically pathognomonic, persists for 7-10 days, and is present in approximately 50% of cases. Petechiae located near the border of the hard and soft palates are observed in about one-third of patients towards the end of the first week of illness. Lymph node enlargement is a hallmark of infectious mononucleosis. The onset is gradual, and anterior and posterior cervical lymph node chains are the most commonly involved. Infectious mononucleosis resembles a number of febrile disorders characterized by fever, sore throat, adenopathy, and lymphocytosis. It may be difficult to distinguish from the early stages of other forms of febrile exudative pharyngotonsillitis, such as streptococcal infections, and exudative tonsillitis of viral aetiology. The differentiation depends upon the results of throat cultures as well as haematological and serological features characteristic of infectious mononucleosis.

**Sarcoidosis**

A patient presenting with bilateral facial paralysis and uveitis should be suspected of having sarcoidosis. Sarcoidosis is a granulomatous disease of undetermined origin that involves multiple systems. Although there is no single laboratory test that is absolutely diagnostic, sarcoidosis is characterized by an elevation in serum and urinary calcium levels, an increase in serum globulin, and an elevated serum angiotensin-converting enzyme level. A chest X-ray may demonstrate hilar adenopathy or diffuse pulmonary infiltrates, and examination of the eye grounds may indicate uveitis, supporting the diagnosis of sarcoidosis. The diagnosis is made on the basis of clinical findings together with biopsy of tissue involved by the sarcoid. Such tissue will contain a non-caseating granuloma with giant cells. Facial palsy is the most commonly seen clinical neurological deficit to accompany sarcoidosis. Uveitis occurs four times more commonly in patients with neurological symptoms than in those without. The peripheral neuropathy associated with sarcoidosis has been shown to be due to perineural inflammatory changes, with the nerve fibres themselves undamaged. This might account for the favourable prognosis with steroid therapy.

**Lyme disease**

Lyme disease has also been reported to cause bilateral facial paralysis (Clark et al, 1985). This disease is characterized by erythema chronicum migrans, tick-borne meningopolyneuritis, myocardial conduction abnormalities, and Lyme arthritis. The disorder
was first recognized in 1975 by close geographical clustering of children with arthritis in the small community of Lyme, Connecticut. This spirochaete disorder is transmitted by an arthropod vector. The disease should be suspected if the patient has been along the north-eastern coast in the USA, in the mid-west (Wisconsin and Minnesota), or in California or Oregon during the summer or early autumn months. These are the geographical locations where the tick vector is found. People are most likely to be out of doors and thus exposed to a tick bite in the warmer months of the year. This disorder has been recognized in Europe and Australia as well. In Europe, the disease complex is referred to as Bannwarth's syndrome.

The disease is characterized by a skin lesion that begins as a red macule or papule and expands to form a large red ring with partial central clearing. This lesion typically lasts about 3 weeks. Associated symptoms include malaise, fatigue, chills and fever, headache, stiff neck, backache, myalgias, nausea, vomiting, and sore throat. Some patients may develop a spectrum of neurological symptoms. The diagnosis can be confirmed by sending a blood sample for serological examination to detect characteristic cryoglobulins and circulating immune complexes. In the report by Clark et al (1985), the incidence of facial palsy was over 10% of all patients with Lyme disease and one-quarter of these patients had bilateral paralysis. The prognosis for recovery was excellent. Only one of the 124 palsies in this series had significant sequelae. Tetracycline is considered the drug of choice, with penicillin and erythromycin as acceptable alternatives. The antibiotic therapy is directed at concurrent symptoms and to prevent serious late complications of Lyme disease. The antibiotics did not alter the course of the paralysis.

**Idiopathic (Bell's) palsy**

One must consider a diagnosis of idiopathic (Bell's) palsy for those patients in whom no cause of facial palsy can be found. If vesicles are present, herpetic neuropathy may be the cause. Other physical findings which may help to define the cause of facial palsy as Bell's palsy include the presence of a red chorda tympani nerve or vascular flaring in the posterior superior aspect of the tympanomeatal area, pain and numbness, hyperacusis, dizziness, loss of tearing, and taste.

**Significance of special tests**

Trying to localize the site of a lesion using the results of tests for tearing (Zilstorff-Pedersen, 1965), taste (Kvarup, 1958), and salivary flow (Blatt, 1965), popularized by Tschiassny (1953), has been found to be of limited value when the lesion is acute and of little or no value in long-standing facial paralysis. This is true for the prognostic value of these tests as well, in contradistinction to a previous report by the author (May, Blumenthal and Taylor, 1981).

The lack of correlation between test results and the location of the lesion is related to a number of variables:

1. the anatomy of the facial nerve and its branches is quite variable, allowing for a variety of alternate pathways for the axons to reach their termination
the lesion responsible for the paralysis may not be sharply localized to a particular level, since a lesion may affect different components of the nerve at various levels and with different degrees of severity.

recovery of the various components may occur at different times.

the techniques used to measure the various facial nerve functions may not be completely reliable.

**Electrical tests**

Whereas tearing, salivary flow, and taste have not been useful as diagnostic and prognostic tests, the prognosis in acute facial palsy can be accurately determined by serial electrical testing. The time course of the degree of loss of response can be plotted. The steeper the line within the first 10 days the poorer the prognosis. Therefore, prognosis is based upon not only the absolute level in 10 days, but also the acceleration of the loss within that period of time (Fisch, 1984). The response to electrical tests has been found to be most helpful in the first 5 days after onset (Esslen, 1977). A study by May, Klein, and Taylor (1985) showed that, if a response to maximal stimulation or evoked electromyography (EEMG) of 25% of normal or greater is maintained up to the tenth day after onset, the patient has a 98% chance of having a satisfactory recovery. If the response remains at 11-24% within the first 10 days, there is an 84% chance of having a satisfactory recovery when the response to maximal stimulation or evoked electromyography drops to 0-10% within the first 10 days.

**Reporting results - facial function recovery**

A standardized, internationally acceptable system for reporting recovery of facial function after injury to the facial nerve has been established (House and Brackmann, 1985) (Table 24.10). From a clinical point of view, patients who fell into grades I and II were considered to have a satisfactory recovery compared to those who fell into grades III and IV. The latter group was considered to have an unsatisfactory recovery. Patients with recovery grades I or II can be separated easily from those in grades III and IV by the absence of the ability to lift the eyebrow or the presence of obvious synkinesis on the involved side.

This five degrees of injury suggested by Sunderland (Table 24.4) describe very nicely the pathological events associated with all types of disorders that afflict the facial nerve. Further, the five degrees of injury fit in very nicely with the clinical classification of recovery reported by House and Brackmann (1985).

**General management of facial palsy**

*Office (outpatient) medical management of acute facial palsies*

Patients and their families were satisfied if answers could be provided to three questions:

1. What is the cause (diagnosis)?
2. When can recovery be expected (prognosis)?
What can be done to promote recovery (treatment)?

In most patients who present with an acute facial palsy these three questions can be answered after a thorough evaluation is performed during the initial office visit. When no specific cause such as trauma, infection, or tumor can be identified and the patient's symptoms fit the picture of idiopathic (Bell's) palsy as described previously (see Table 24.6), the patient is told that the facial nerve weakness was most probably caused by a viral inflammatory immune disorder often referred to as Bell's palsy. The prospects for recovery from this disorder are excellent, and the patient should be reassured that he or she has not had a stroke, and will not be permanently deformed. Next, the time and degree of likely recovery are predicted by evaluating:

1. the completeness of the palsy
2. the response to the maximal stimulation test or evoked electromyography
3. the time recovery first begins.

The degree of recovery can be categorized (Table 24.10) as grade I (complete, with no detectable difference between the normal and the involved side), grade II (a very subtle deficit remains), or grade III or IV (incomplete recovery marred by more or less severe signs of faulty regeneration, such as synkinesis and spasm as well as facial weakness). Almost every patient with idiopathic (Bell's) palsy or acute facial palsy due to trauma or infection who maintains some facial movement beyond 14 days after onset will have a satisfactory recovery from this disorder (grade I or II recovery).

Nevertheless, patients must be followed carefully, both in order to document recovery and to watch for signs of progression that indicate a worse prognosis. The prognosis in acute facial palsy can be accurately determined by serial electrical testing, as noted previously.

Management plan

As long as patients maintain an incomplete palsy, and have been evaluated within the first 14 days of onset, they can be given an appointment to return in 3 weeks for further evaluation. However, they should be told to return sooner if the palsy progresses as determined by daily evaluation of facial movement. This can be accomplished by the patient standing in front of a mirror or having a family member observe the effects of raising the eyebrows, squeezing the eyes closed, wrinkling the nose, attempting to whistle, blowing out the cheeks, and grinning so as to show the teeth. As long as facial function does not worsen, the patient should have satisfactory return of function with no further treatment. However, if the patient with persistent incomplete palsy does not begin to recover in 6 weeks or the paresis worsens rather than shows improvement, a tumour should be suspected.

When a patient presents with a complete facial motor deficit one must rely upon the response to maximal stimulation or evoked electromyography and the time post-onset that beginning of facial recovery is first noted to determine prognosis and develop a management plan. Early recovery of facial function, within the first 3 weeks, is a reliable indication that recovery will be satisfactory, but this prediction should be supported by electrical tests performed every other day up to the tenth day. If facial paralysis persists and response to evoked electromyography remains above 11% of normal, the patient is re-evaluated every
other day up to the fourteenth day post-onset. If on the fourteenth day the response to maximal stimulation persists or evoked electromyography remains above 11% of normal, the patient is informed that the prognosis for early and ultimately satisfactory recovery is excellent. On the other hand, if the response to evoked electromyography drops below 11% of normal or is lost completely within the first 14 days, then the prognosis for satisfactory recovery drops to 21%.

**Table 24.6. Differential diagnosis of aetiology of facial palsy by history and physical findings**

<table>
<thead>
<tr>
<th>Bell's palsy</th>
<th>(6) Mass in parotid, submandibular gland, or neck</th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Acute onset of unilateral facial palsy</td>
<td>(7) Mass between ascending ramus and mastoid tip</td>
</tr>
<tr>
<td>(2) Numbness or pain of ear, face, neck, or tongue (50%)</td>
<td>(8) Progression of other motor cranial nerve deficits</td>
</tr>
<tr>
<td>(3) Viral prodroma (60%)</td>
<td>(9) Some of branches of facial nerve spared</td>
</tr>
<tr>
<td>(4) Recurrent facial palsy (12%)</td>
<td>(10) History of cancer</td>
</tr>
<tr>
<td>(ipsilateral 36%, alternating 64%)</td>
<td></td>
</tr>
<tr>
<td>(5) Positive family history (14%)</td>
<td></td>
</tr>
<tr>
<td>(6) Loss of ipsilateral tearing and/or submandibular salivary flow (10%)</td>
<td></td>
</tr>
<tr>
<td>(7) Decrease in or loss of ipsilateral stapes reflex (90%)</td>
<td></td>
</tr>
<tr>
<td>(8) Red chorda tympani nerve (noted in 40% of patients evaluated in first 10 days in whom the chorda tympani could be seen; also noted with herpes zoster cephalicus and Guillain-Barré syndrome)</td>
<td>(11) Bell's - herpes simplex</td>
</tr>
<tr>
<td>(9) Self-limiting and spontaneously remitting.</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Herpes zoster cephalicus</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Same as for Bell's, except pain more common and severe</td>
<td></td>
</tr>
<tr>
<td>(2) Vesicles on pinna, face, neck, or oral cavity (100%)</td>
<td></td>
</tr>
<tr>
<td>(3) Sensorineural hearing loss and/or vertigo (40%)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Tumour</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Sudden complete onset similar to Bell's; EEMG results abnormal (10% within 5 days)</td>
<td></td>
</tr>
<tr>
<td>(2) Recurrent same side (17%)</td>
<td></td>
</tr>
<tr>
<td>(3) Slowly progressive weakness beyond 3 weeks (59%)</td>
<td></td>
</tr>
<tr>
<td>(4) No recovery after 6 months</td>
<td></td>
</tr>
<tr>
<td>(5) Twitching with paresis</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Bilateral simultaneous facial palsy</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Guillain-Barré</td>
<td></td>
</tr>
<tr>
<td>(2) Moebius' syndrome</td>
<td></td>
</tr>
<tr>
<td>(3) Sarcoidosis</td>
<td></td>
</tr>
<tr>
<td>(4) Myotonic dystrophy</td>
<td></td>
</tr>
<tr>
<td>(5) Skull trauma</td>
<td></td>
</tr>
<tr>
<td>(6) Infectious mononucleosis</td>
<td></td>
</tr>
<tr>
<td>(7) Cytomegalovirus</td>
<td></td>
</tr>
<tr>
<td>(8) Acute porphyrias</td>
<td></td>
</tr>
<tr>
<td>(9) Botulism</td>
<td></td>
</tr>
<tr>
<td>(10) Lyme disease</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Birth</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Congenital diplegia (Moebius' syndrome, thalidomide toxicity)</td>
<td></td>
</tr>
<tr>
<td>(2) Lower lip palsy (developmental)</td>
<td></td>
</tr>
<tr>
<td>(3) Trauma</td>
<td></td>
</tr>
<tr>
<td>(4) Tumour</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Trauma</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Skull fracture (acute or delayed)</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Infection</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>(1) Bulbar palsy (viral meningitis, encephalitis, or immune reaction)</td>
<td></td>
</tr>
<tr>
<td>(2) Postinfluenza, rabies, or poliomyelitis immunization</td>
<td></td>
</tr>
</tbody>
</table>
(3) Infectious mononucleosis  
(4) Botulism  
(5) Tetanus  
(6) Syphilis  
(7) Malaria  
(8) Lyme disease  
(9) Herpes zoster cephalicus  
(10) Otitis media (acute or chronic, with or without cholesteatoma)  
(11) Leprosy  

Metabolic  
Acute porphyria  

Neoplastic  
Acute leukaemia  

Iatrogenic  
Bilateral arterial embolization  
Idiopathic  

(1) Guillain-Barré syndrome  
(2) Sarcoidosis (Heerefordt syndrome - uveoparotid fever)  
(3) Polyarteritis nodosa  
(4) Bell’s palsy  

Melkersson-Rosenthal syndrome  
(1) Recurrent alternating facial palsy  
(2) Fissured tongvue  
(3) Labial-periorbital facial oedema  
(4) Non-specific labial granuloma  
(5) Positive family history.

Once the prognosis has been established, patients are asked to return in 3 months, 6 months, and finally one year for final evaluation of facial function employing the system of House and Brackmann (1985). However, while waiting for recovery to begin, medical treatment is recommended, and precautions must be taken to prevent possible sequelae of facial nerve paralysis.

**Medical treatment**

There are three main types of treatment for acute facial palsy: physical, pharmacological, and psychophysical.

Physical therapy includes heat, massage, and exercises performed twice a day. Patients are advised to wet a Turkish towel with hot water, wring it out, and place the hot towel on the face until the towel cools. Then the patient should massage facial cream into the skin around the eyes and mouth and over the midface for a few minutes, ideally using an electric vibrator. Finally, the patient should stand in front of a mirror and watch the face while raising the eyebrows, squeezing the eyes closed, wrinkling the nose, whistling, blowing out the cheeks, and grinning. Even though no facial movement may be noted, intact nerve fibres will be activated, and the exercises will help to maintain muscle tone.

Although several medications, including steroids, have been used to treat facial paralysis, none has been shown to be efficacious.

Psychophysical modalities such as motor sensory re-education have been useful (Schram and Burres, 1984; Balliet, 1984). In the acute phase, integrated electromyographic tracings of motor strength can often be displayed on an oscilloscope, offering a patient significant encouragement at a time when no visible movement can be seen. The course of the recovery can be followed since there is a relationship between the response of voluntary effort recorded on the oscilloscope and actual recovery. During the post-acute phase, when recovery has begun, the patient can benefit from a combination of strategies using biofeedback, working in front of a mirror, and touching the face while attempting movements.
These strategies are particularly useful in the post-acute phase once the patient has plateaued in terms of facial recovery. Further improvement can be achieved using these strategies.

**Table 24.7. Special diagnostic tests to evaluate patients with facial palsy**

<table>
<thead>
<tr>
<th>Topognostic tests</th>
<th>Electrical tests</th>
<th>Radiographic studies</th>
<th>Surgical exploration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hearing and balance tests</td>
<td>Maximal stimulation test (MST)</td>
<td>Plain views of mastoid and internal auditory canal</td>
<td>Lumbar puncture (cerebrospinal fluid) to detect meningitis, encephalitis</td>
</tr>
<tr>
<td>Schirmer test</td>
<td>Evoked electromyography (EEMG)</td>
<td>Pluridirectional tomography of temporal bone</td>
<td>Guillain-Barré syndrome, multiple sclerosis, meningeal carcinomatosis</td>
</tr>
<tr>
<td>Stapes reflex</td>
<td>Electromyography (EMG)</td>
<td>Computerized tomography of brainstem, cerebellopontine angle, temporal bone, skull base; contrast sialography of parotid</td>
<td>Complete white blood cell count and differential to detect infectious mononucleosus, leukaemia</td>
</tr>
<tr>
<td>Submandibular flow test</td>
<td></td>
<td>Magnetic resonance imaging</td>
<td>Monospot test to detect infectious mononucleosis</td>
</tr>
<tr>
<td>Taste test</td>
<td></td>
<td>Chest radiographic survey to detect sarcoidosis, lymphoma, carcinoma</td>
<td>Heterophil titre to detect infectious mononucleosis</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Topognostic tests</th>
<th>Electrical tests</th>
<th>Radiographic studies</th>
<th>Surgical exploration</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Urine and faecal examinations:</td>
<td></td>
<td>Lumbar puncture (cerebrospinal fluid) to detect meningitis, encephalitis</td>
</tr>
<tr>
<td></td>
<td>Acute porphyria - elevated porphyrins and urinary porphobilinogen</td>
<td></td>
<td>Guillain-Barré syndrome, multiple sclerosis, meningeal carcinomatosis</td>
</tr>
<tr>
<td></td>
<td>Botulism - <em>C. botulinum</em> toxin in stool specimen</td>
<td></td>
<td>Complete white blood cell count and differential to detect infectious mononucleosus, leukaemia</td>
</tr>
<tr>
<td></td>
<td>Sarcoidosis - urinary calcium</td>
<td></td>
<td>Monospot test to detect infectious mononucleosis</td>
</tr>
<tr>
<td></td>
<td>Serum cryoglobulins and immune complexes to detect Lyme disease</td>
<td></td>
<td>Fluorescent treponemal antibody titre to detect syphilis</td>
</tr>
<tr>
<td></td>
<td>Serum globulin level to detect sarcoidosis</td>
<td></td>
<td>Erythrocyte sedimentation rate to detect sarcoidosis, collagen vascular disorders</td>
</tr>
<tr>
<td></td>
<td>Serum and urine calcium determinations to detect sarcoidosis</td>
<td></td>
<td>Serum and urine calcium determinations to detect sarcoidosis</td>
</tr>
<tr>
<td></td>
<td>Serum angiotensin-converting enzyme level to detect sarcoidosis</td>
<td></td>
<td>Serum antinuclear antibody test (ANA), and rheumatoid factor (RF) to detect collagen vascular disorders (polyarteritis nodosa)</td>
</tr>
<tr>
<td></td>
<td>Serum globulin level to detect sarcoidosis</td>
<td></td>
<td>Bone marrow examination to detect leukaemia, lymphoma</td>
</tr>
<tr>
<td></td>
<td>Serum and urine calcium determinations to detect sarcoidosis</td>
<td></td>
<td>Glucose tolerance test to detect diabetes mellitus.</td>
</tr>
</tbody>
</table>
Depression

Patients who suddenly suffer complete facial paralysis of acute onset initially fear that they have a permanent deformity or have suffered a stroke. Once patients have been reassured that they did not have a stroke, the obvious facial deformity often leads to depression. If the prognosis is favourable for early recovery, the patient should be encouraged by this news, but if recovery is not expected for 2-4 months, the patient should be informed of this openly and supported sympathetically. Patient counselling and group therapy have been effective in helping patients to deal with this deformity, especially when patients are selected to be of the same sex and similar age, and the patient counsellor has had a satisfactory recovery or learned to deal with the problem in a positive way.

Table 24.8. Causes of facial nerve disorders in 1989 patients seen over 20 years (1963-1985) by one clinician

<table>
<thead>
<tr>
<th>Cause</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>No</td>
</tr>
<tr>
<td>Bell's palsy</td>
<td>1082</td>
</tr>
<tr>
<td>Herpes zoster cephalicus</td>
<td>146</td>
</tr>
<tr>
<td>Trauma</td>
<td>375</td>
</tr>
<tr>
<td>Tumour</td>
<td>127</td>
</tr>
<tr>
<td>Infection</td>
<td>78</td>
</tr>
<tr>
<td>Birth</td>
<td>62</td>
</tr>
<tr>
<td>Hemifacial spasm</td>
<td>45</td>
</tr>
<tr>
<td>Central nervous system (axial) disease</td>
<td>21</td>
</tr>
<tr>
<td>Other</td>
<td>41</td>
</tr>
<tr>
<td>Tumour suspect</td>
<td>12</td>
</tr>
<tr>
<td>Total</td>
<td>1989</td>
</tr>
</tbody>
</table>

Physical pain

Approximately half of patients with acute idiopathic (Bell's) palsy and almost all with herpes zoster cephalicus have pain. In most cases pain can be controlled with a non-narcotic analgesic, although in rare instances a narcotic may be required.

Eye care

Efforts should be directed towards keeping the globe moist to prevent keratitis and corneal breakdown. The patient should voluntarily close the eyelids on the involved side whenever the eye feels irritated or burns, about two to four times a minute, and drops should be used during the day and ointment at night. In addition, a moisture chamber should be worn over the involved eye whenever the patient is out of doors or the eye becomes irritated. Surgery to reanimate the paralysed eyelids should be considered if medical treatment is ineffective, in particular when patients lack Bell's phenomenon, have corneal Anaesthesia, and lack tears or have a Dry eye - the BAD syndrome. A tarsorrhaphy should be a last resort and revised later using one of the more effective reanimation techniques. A tarsorrhaphy produces
a cosmetic blight, limits vision, and often does not protect the exposed cornea. In the event that the tarsorrhaphy can be reversed, sequelae often result including notching of the lid margin and trichiasis. Implantation of a gold weight or eyelid spring for the upper lid and lower lid tightening procedures have been so effective that a tarsorrhaphy is rarely indicated.

**Table 24.9. Bell's palsy - diagnosis of exclusion**

The palsy is not Bell's if one of the following is present

- Signs of tumour
- Bilateral simultaneous palsy
- Vesicles
- Involvement of multiple motor cranial nerves
- History and findings of trauma
- Ear infection
- Signs of central nervous system lesion
- Facial palsy noted at birth
- Triad of infectious mononucleosis (fever, sore throat, cervical lymphadenopathy).

**Surgical management of facial nerve paralysis**

**Indications for surgery**

The benefit of facial nerve surgical decompression through the transmastoid or middle fossa route has not been established for idiopathic (Bell's) palsy, or herpes zoster cephalicus. Further, surgical decompression for acute suppurative otitis media, necrotizing external otitis, or facial paralysis following iatrogenic or external temporal bone trauma is indicated only in selected cases (Maiman et al, 1985). However, facial paralysis due to an ongoing process such as chronic suppurative otitis media with or without cholesteatoma can only be relieved by eradicating the primary process. It should be performed prior to electrical denervation to give the most satisfactory facial function recovery, and must not be delayed if the palsy has progressed from incomplete to complete over a period of hours or days and if the response to evolved electromyography is less than 25% of normal or dropping precipitously after the third day following onset.

In addition, there are two situations where surgery is absolutely indicated in managing facial nerve disorders: facial nerve transection and tumour infiltration. Further, there are times when nerve transection or tumour infiltration can only be established by surgical exploration, in particular when the temporal bone has fractured or a tumour is suspected.

**Technique of transmastoid surgical exploration of facial nerve - labyrinthine segment to stylomastoid foramen**

The postaural approach offers direct access to the tympanomastoid, geniculate, and distal labyrinthine segments of the facial nerve. The technique for this approach preserves hearing. When exposure of the geniculate ganglion and labyrinthine segment is required and preservation of hearing need not be considered, the translabyrinthine route is preferred.
Table 24.10. Classification system for reporting results of recovery from facial paralysis

<table>
<thead>
<tr>
<th>Degree of injury</th>
<th>Grade</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal (1°)</td>
<td>I</td>
<td>Normal symmetrical function in all areas</td>
</tr>
<tr>
<td>Mild dysfunction noticeable) (1°-2°)</td>
<td>II</td>
<td><em>Slight weakness noticeable</em> only on close (barely inspection. Complete eye closure with minimal effort. Slight asymmetry of smile with maximal effort. Synkinesis barely noticeable, contracture, or spasm absent.</td>
</tr>
<tr>
<td>Moderate dysfunction (obvious difference) (2°-3°)</td>
<td>III</td>
<td><em>Obvious weakness</em>, but not disfiguring. May not be able to lift eyebrow. Complete eye closure and strong but asymmetrical mouth movement with maximal effort. Obvious, but not disfiguring synkinesis, mass movement or spasm.</td>
</tr>
<tr>
<td>Moderately severe dysfunction (3°)</td>
<td>IV</td>
<td><em>Obvious disfiguring weakness</em>. <em>Inability to lift brow</em>. Incomplete eye closure and asymmetry of mouth with maximal effort. <em>Severe synkinesis, mass movement, spasm.</em></td>
</tr>
<tr>
<td>Severe dysfunction (3°-4°)</td>
<td>V</td>
<td><em>Motion barely perceptible</em>. Incomplete eye closure, slight movement corner mouth. <em>Synkinesis, contracture, and spasm usually absent.</em></td>
</tr>
<tr>
<td>Total paralysis</td>
<td>VI</td>
<td><em>No movement</em>, loss of tone, no synkinesis, contracture, or spasm.</td>
</tr>
</tbody>
</table>

The transmastoid approach to the facial nerve consists of a preliminary mastoidectomy with removal of air cells from the antrum downward to the mastoid tip, and defining the ridge of the digastric groove. Further, cells are removed from the antrum forward to the root of the zygoma until the upper edge of the incus and the prominence of the bony horizontal canal are identified. Care is taken not to disturb the ossicles. The bony meatal wall, while thinned, is left intact. The landmark for the vertical mastoid portion of the facial nerve is the posterior tip of the incus above and the anterior end of the digastric groove below. Under the operating microscope, the periosteum of the digastric groove is exposed and followed forward and upward until the stylomastoid foramen is exposed. Then the bone between the foramen and the horizontal semicircular canal is thinned with a diamond burr, used parallel to the course of the nerve, under continuous irrigation with Ringer’s solution or Tis-U-Sol to remove bone dust and blood and prevent overheating. As the nerve is approached, it begins to appear through the paper-thin bone as a pink streak. Brisk bleeding may be encountered from the artery that enters the fallopian canal at the pyramidal bend. With the diamond burr used gently, the final thinning of bone over the nerve is accomplished. Thus, the fallopian canal is exposed from its tympanic portion to the stylomastoid foramen, with care being taken not to disturb the incus or to open the horizontal semicircular canal. With right and left dental
curettes the thinned bone covering the facial epineurium is lifted off, exposing the contents of the fallopian canal. With magnification provided by the operating microscope, this can be accomplished safely without injuring the nerve.

When the horizontal segment is involved, decompression is carried out through a triangle bounded by the facial nerve medially, chorda tympani and tympanic annulus laterally, and short process of the incus superiorly which provides the surgical guidelines for this approach. In constructing this triangular window into the facial recess, it is advisable to leave a small pillar of bone over the fossa incudis to prevent accidental brushing of the incus by the burr, which could result in serious, irreversible acoustic trauma. When the hearing is normal and the entire horizontal segment must be decompressed, it may be necessary to disarticulate the incus. In most cases this manoeuvre can be done quite safely through the facial recess by gently separating the capsules of the incudostapedial and malleoincudal joint leaving the short process of the incus attached to the fossa incudis. The incus can then be rotated toward the middle ear to facilitate dissection over the proximal tympanic, geniculate, and distal labyrinthine segments. The incus can then be rotated towards the mastoid so that the mid-tympanic portion of the facial nerve can be dissected without concern for transmitting vibrations from the incus to the stapes into the inner ear. Following decompression, the incus is replaced in its natural anatomical relationship where it will remain providing the fossa incudis has been preserved.

Decompression of the facial nerve is completed by slitting the sheath vertically on its posterior aspect with a disposable Beaver knife. If bleeding is troublesome it should be controlled with Surgical. Use of electric cauterity is discouraged, as even a wet-field bipolar cautery may create an unwanted injury by the transmission of heat at the site of application. If it is absolutely necessary to use the bipolar wet-field cautery, it must be done while the area is being irrigated.

*Exploration and decompression or repair for traumatic facial palsy*

When facial paralysis follows trauma, either surgical or otherwise, the site of injury must be exposed. Since this is most often in the tympanic or pyramidal portion of the nerve, it is possible to expose and decompress this segment by following the nerve into the facial recess through the postauricular approach. In some cases it may be necessary to take down the posterior canal wall for adequate exposure. The tympanic portion of the fallopian canal is examined under the operating microscope to determine the site and extent of injury. If the ossicles are intact, the incus may need to be dislocated from the stapes and later replaced to gain access to the tympanic fallopian canal, as described previously. The nerve is exposed at the site of injury and for at least 5 mm in both directions until normal-appearing nerve is encountered. If the nerve is intact but swollen or compressed by a depressed bone fragment, the latter is removed and the sheath slit. If the nerve has been partially torn, the intact fibres are carefully preserved and the torn fibres approximated or replaced by a small free graft if they cannot be approximated. If the nerve is completely severed, it must be repaired by approximation or by a free nerve graft as described later.
Repair of severed facial nerve by approximation

Theoretically, it might seem that regeneration of the facial nerve would be more satisfactory across a single junction than across two junctions at either end of a free graft. For this reason it is tempting to reapproximate the facial nerve when there are just a few millimetres between the two ends. However, re-routing the facial nerve to gain length to accomplish an end-to-end approximation in the horizontal and vertical segments by mobilizing the proximal and distal ends is not the procedure of choice. Often, the more the nerve is freed up, the more it seems to shorten and the more the blood supply to the nerve is jeopardized.

Additional length can be obtained for end-to-end approximation if the injury to the facial nerve is proximal to the geniculate ganglion and hearing and balance function have been destroyed. In this case the facial nerve can be freed from its first genu, separating the nerve from the geniculate ganglion and mobilizing the vertical and horizontal segments posteriorly towards the internal auditory canal. Length can also be achieved for the purpose of an end-to-end anastomosis with injuries of the facial nerve in the region of the parotid gland. However, as a general rule, it is much better to put a nerve graft between severed portions rather than to try to mobilize the ends of the nerve to accomplish an end-to-end anastomosis. Based upon the author's experience, the results were as good or better when a free graft was introduced as when a nerve was re-routed. If the ends cannot be brought together without tension, then a free graft must be inserted. Lack of tension at the site of approximation is the best guarantee that the repair will be a success.

Repair of a severed facial nerve by graft

When there is a gap between the cleanly cut ends of a severed facial nerve, so that the distal segment of the nerve cannot be brought up to establish contact with the proximal end without tension, a nerve graft should be inserted. The great auricular and sural cutaneous nerves are most suitable for facial nerve grafting. The great auricular is ideal for grafts up to 10 cm in length and the sural cutaneous for longer grafts.

A segment of nerve, measured so as to be slightly longer than the gap to be bridged, is removed, and its ends are cut sharply at right angles with a safety razor blade against a wooden tongue depressor. The nerve graft is handled carefully to avoid pinching or other trauma. Under a microscope, the nerve graft is carefully approximated to the distal and proximal stumps using 10-0 monofilament suture, employing the technique illustrated. By accomplishing a fascicular anastomosis, the graft need not be protected by covering it with a vein graft or silastic tubing. As long as the two ends of the graft lie within the temporal bone and do not extend outside the stylomastoid foramen or into the internal auditory canal suturing has not been necessary.

Results of facial nerve graft repair

The best results were achieved when the central nerve stump was connected to the peripheral system of the facial nerve within one year of injury. When the central stump was not available or the time between injury and repair was between one and 2 years, the procedure of choice was a hypoglossal-facial anastomosis. When repair was performed between 2-4 years after injury, the distal stump of the facial nerve was biopsied and, if it was
fibrotic, a muscle transposition procedure was performed. If injury had occurred more than 4 years previously, or if the facial nerve and muscle system were not suitable for the procedures just described, temporalis muscle transposition was the preferred reanimation technique for the mouth, and separate eye reanimation techniques were combined with mouth reanimation.

Knowing the cause of the facial paralysis may be critical in determining how best to restore function. Most facial palsies evaluated for possible rehabilitation will be the result of trauma, either surgical or accidental. If the injury was acute and the nerve was severed, the best results are achieved when repair is performed within 30 days of injury.

The time following onset of the paralysis must be taken into account. No irreversible procedure such as a nerve graft, facio-facial cross-face graft, or hypoglossal-facial anastomosis should be undertaken while there remains the possibility of spontaneous recovery. If the facial nerve was spared following acoustic tumour surgery, it is advisable not to perform a procedure that interrupts the integrity of the facial nerve less than 12 months from the time of injury, in order to allow adequate time for evidence of spontaneous recovery to occur. Twelve months is a good waiting period before performing such nerve repair procedures, since it has been the author’s experience that if no recovery has been noted in this period of time spontaneous recovery of useful function is unlikely.

However, time is of the essence in achieving the best possible results when the nerve has been injured, since an eightfold decrease in axon diameter occurs over 3 months (Sunderland and Bradley, 1950b). The decrease is due to shrinkage and later gradual thickening of the collagen of the endoneurial sheath (Sunderland and Bradley, 1950b). This suggests that nerve repair be undertaken without delay, so that axons which regenerate early can grow into the collapsing tubes, thus re-inflating them and suppressing collagenization before it progresses too far. In general, the sooner re-innervation begins, the better. The ideal time for nerve grafting is within 30 days and not later than one year following injury; beyond one year the results of nerve grafting by any technique have been disappointing. The best results with a hypoglossal-facial anastomosis were achieved when surgery was performed within the first 2 years after injury, although satisfactory results were noted following surgery performed up to 4 years after injury (Conley, 1975). Recovery has been noted with later repairs in cases where part of the peripheral system was spared or spontaneously regenerated.

There are a number of factors that influence results following nerve repair. Technical flaws that might downgrade recovery include tension at the suture line, residual tumour, lack of suitable nerve ends, and the presence of infection. Other factors, such as the timing of the surgery following injury, the cause of the injury, the site of the injury, the number of anastomotic sites and the length of the graft, have been considered as variables influencing results. Nevertheless, the author’s experience has shown that the most important factor is timing after injury.

The first sign of returning function is improving tonus of the paralysed side of the face, actually before there is any voluntary movement. Even in a long graft from the internal auditory meatus to the extracranial segment of the facial nerve, returning motion has been detected as early as 4 months, but it may be delayed as long as 24 months, the average interval being 10 months. In cases of nerve repair, maximum recovery requires 2 years and
improvement may continue over a period of 5 years. Under ideal conditions satisfactory recovery following nerve repair can be expected in over 90% of cases.

Other techniques for facial reanimation

Results following facial nerve grafting or hypoglossal nerve anastomosis can be augmented employing reanimation techniques directed to the eye and mouth. Brow lift, gold weight or eyelid spring implantation and lower lid tightening combined with temporalis muscle transposition provide immediate eye reanimation with mouth symmetry and voluntary movement within 3-6 weeks after the procedure. Free muscle neurovascular repair techniques should be reserved for cases where the techniques already mentioned are not possible. The free muscle techniques are still evolving and greater experience is required before precise indications and anticipated results can be proposed.

Management of idiopathic (Bell's) palsy, herpes zoster cephalicus and other facial nerve disorders of viral origin

Bell's palsy

Bell's palsy is a term used to designate acute peripheral facial palsy of unknown cause, although accumulating evidence supports a viral inflammatory immune mechanism. The disorder is self-limiting, non-progressive, non-life-threatening, spontaneously remitting, and at this time can be neither prevented nor cured. The incidence varies between 15 and 40 per 100,000 population (Hauser et al, 1971; Adour et al, 1978; Peitersen, 1982).

Clinical features

Bell's palsy is characterized by a viral prodrome (60%), which is accompanied by pain around the ear (50%), facial numbness (40%), changes in taste (50%), and numbness of the tongue (20%) (May and Hardin, 1977). A positive family history was obtained in 14% of patients, and the syndrome recurred in 12%. Of those with a history of recurrence, the same side was involved in 36%, while in the remaining 64% the palsy recurred on the other side. The common involvement of stapedius reflex, and salivary flow indicates that the segment most often involved is the tympanomastoid portion of the facial nerve (May and Hardin, 1977).

Predicting outcome

By studying the results of evoked electromyography and evaluating the completeness of the palsy, the patient's prognosis for recovery of facial function can be predicted with a high degree of accuracy. More than 90% of patients will have a satisfactory recovery, provided the palsy is incomplete and response to evoked electromyography remains greater than 10% beyond the first 14 days after onset. Patients with a complete palsy and response to evoked electromyography of 10% or less within the first 5-10 days have an 80% chance of an unsatisfactory recovery. It is this latter group that requires the greatest attention in terms of treatment directed toward improving the natural history of facial palsy and preventing complications of nerve degeneration.
Peitersen (1982) studied the natural history of over 1000 patients with Bell's palsy seen over a 15-year period, and found that in 84% recovery was satisfactory; 71% recovered without sequelae, and 13% had defects that were barely noticeable. The other 16% of patients had obvious incomplete recovery of facial function, but sequelae were crippling in only 4%, and there was not a single patient who did not have some recovery. Peitersen noted that 85% of the patients in his study began to recover facial function within 3 weeks of the palsy, which in 31% was incomplete at onset. Peitersen concluded that there is a relationship between the degree of injury and ultimate recovery and the time that recovery is first noted; the earlier recovery is noted, the better the prognosis for a satisfactory and speedy recovery.

Treatment for Bell's palsy is supportive, and involves eye care, pain control, reassurance, heat, massage, and facial exercises. Steroids and surgery have not been shown to alter the natural history.

**Herpes zoster cephalicus (Ramsey Hunt syndrome)**

Hunt first described a syndrome, now called herpes zoster cephalicus, or herpes zoster oticus, which is characterized by a viral prodrome, with severe pain in and around the ear, and vesicles involving the pinna (Hunt, 1907, 1908). In its mildest form there may not be any neurological signs, but in a more severe form it may be accompanied by a sensorineural hearing loss and disturbed vestibular function, and even viral encephalitis. The vesicles in Ramsey Hunt syndrome may occur over the ear, face, and neck down to the shoulder, and may also involve the tongue, larynx, or buccal mucosa. The distribution of the vesicles depends on which sensory afferent fibres are involved by the viral eruption, but all of the nerves that communicate with the facial nerve may be involved, including cranial nerves V, VII, VIII, IX, and X and the cervical plexus arising from cervical nerves 2, 3, and 4. The sign common to all forms of herpes zoster cephalicus, and necessary to establish the diagnosis, is the vesicles.

Groves (1976) has presented a comprehensive review of facial nerve disorders, including a review of the literature on the history, aetiology, and treatment of Ramsey Hunt syndrome. Herpes zoster cephalicus is similar to Bell's palsy, except that in the former the vesicles are present, and there is a higher incidence of auditory vestibular involvement, postherpetic pain, and a poorer prognoses. Response to maximal stimulation or evoked electromyography may remain in the normal range beyond 10 days, then be lost by the fourteenth to the twenty-first day. This is in contrast to Bell's palsy, in which electrical response may become abnormal by the tenth day. A sensorineural hearing loss was noted in 10% of patients and a decreased response to electronystagmography in 40% of patients with herpes zoster cephalicus. Bilateral vestibular suppression has been noted with this disorder, perhaps an indication of brainstem involvement.

The natural history of herpes zoster cephalicus differs from that of Bell's palsy in several ways, perhaps reflecting the difference in behaviour of herpes simplex type I and varicella-zoster viruses. Bell's palsy recurs in 12% of cases, but herpes zoster cephalicus
rarely recurs. In addition, the acute phase of the infection, as measured by electrical response and progression of palsy, peaks in 5-10 days with Bell's palsy but in 10-14 days with herpes zoster cephalicus. Lastly, 84% of individuals suffering from Bell's palsy have a satisfactory recovery of facial function, but only 60% of those with herpes zoster cephalicus recover a satisfactory degree of facial function.

**Treatment**

Treatment of herpes zoster is similar to that for Bell's palsy, with the exception that greater attention must be devoted to control of pain and the vesicular eruption. Often narcotics are required and a steroid antibiotic cream is effective for treating the vesicular eruption. Use of antiviral agents may hold promise, but must be subjected to a prospective, controlled randomized study (see Chapters 13 and 17). The availability of a vaccine to prevent chicken pox has been announced but has not yet been released. Prevention of chicken pox would eliminate herpes zoster since it is a reactivation of chicken pox virus.

**Other viral disorders**

Other viruses in the herpes virus group can cause facial nerve disorders. The Epstein-Barr virus is the known cause of infectious mononucleosis, and has also been isolated in cases of Guillain-Barré syndrome. The cytomegalovirus has also been isolated in patients with Guillain-Barré syndrome, suggesting that multiple viral agents are capable of producing this disorder. The Melkersson-Rosenthal syndrome is still another clinical entity that may masquerade as Bell's palsy. However, although it is possible that a viral agent may play a role in this last disorder, there is strong evidence that it is actually a hereditary autoimmune disorder. It is characterized by recurrent alternating facial palsy, tongue plication, facial swelling, granulomatous labial submucosal lesions, a positive family history, and migraine headaches. The presence of two of these factors associated with recurrent alternating facial palsy satisfies the necessary criteria for diagnosis.

**Facial nerve disorders in the newborn and children**

Facial disorders in children can be due to a variety of causes and should not be assumed to be of the Bell's type. Further, the type of treatment and ultimate outcome depend on early, accurate diagnosis of the cause of the palsy. The principles of managing facial paralysis in children are the same as those for adults with a few exceptions, and these will be noted. The information presented is based on the diagnosis and management of facial paralysis in 332 patients, from newborn to 18 years, seen between 1963 and 1985. The causes of facial palsy in these children were similar to those in adults, with the exception of paralysis noted at birth and the number of cases due to acute otitis media (*Table 24.11*).

**Facial paralysis noted at birth**

The differential diagnosis and treatment of facial paralysis in the newborn has been reviewed by May et al (1981) and Harris et al (1983). The two main differential diagnostic possibilities are developmental and traumatic; the factors that aid in differentiating between them are listed in *Table 24.12*. 
The most common finding associated with congenital facial paralysis was the presence of one or more other anomalies. Weakness of the lower lip has particular significance in that it may be associated with multiple congenital anomalies (Pape and Pickering, 1972). Developmental bilateral facial palsy is frequently incomplete, with the lower portion of the face usually less affected than the upper part. This distinguishes it from facial palsy due to trauma, which is rarely bilateral and in which the upper and lower parts of the face are equally involved. Bilateral immobility of the face may not be apparent at birth and may be manifested by incomplete eyelid closure when asleep, an open mouth, and/or inability to suck.

**Syndromes associated with congenital facial paralysis**

**Moebius' syndrome**

This is a rare congenital disorder which usually includes bilateral facial palsy, unilateral or bilateral abducens palsy, anomalies of the extremities, absence of various muscles, particularly the pectoral group, and involvement of other cranial nerves, particularly the last four and especially the hypoglossal.

**Dystrophia myotonica**

This disorder is a steadily progressive familial distal myopathy with associated weakness of the muscles of the face, jaw, neck, and levators of the eyelids. Children with muscular dystrophy usually present at birth with congenital facial diplegia, although without abducens paralysis, and only later evidence the progressive nature of the myopathy (Hanson and Rowland, 1971). Congenital facial diplegia associated with dystrophia myotonica that appears at birth is the earliest manifestation of the disease in its severest form. Unlike Moebius' syndrome, there is muscle wasting, particularly of the sternocleidomastoid, temporal, and facial muscles, creating an expressionless face which is so characteristic that it is referred to as the myopathic facies. Extramuscular dystrophies such as cataract, premature frontal baldness, and testicular atrophy are also present, and the neck is usually described as swan-like. This latter defect is due to wasting of the muscles of mastication and the sternocleidomastoid muscle.

**Thalidomide embryopathy**

Phocomelia (seal-like limbs) has apparently been known since Babylonian times, and was described by Ballantyne (1904), but the sudden increased incidence of this rare deformity between 1958 and 1962 focused attention on the disorder. Investigations discovered that the sedative thalidomide taken by the mother between the twenty-eighth and forty-second day of pregnancy led to thalidomide embryopathy with associated arrested development of the ear and abducens paralysis (Miehlke, 1965).
**Table 24.11. Causes of facial palsy in 339 patients newborn to 18 years old evaluated between 1963 and 1985**

<table>
<thead>
<tr>
<th>Causes</th>
<th>Number</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bell's palsy</td>
<td>133</td>
<td>39</td>
</tr>
<tr>
<td>Herpes zoster cephalicus</td>
<td>13</td>
<td>4</td>
</tr>
<tr>
<td>Birth</td>
<td>62</td>
<td>18</td>
</tr>
<tr>
<td>Developmental</td>
<td>(46)</td>
<td></td>
</tr>
<tr>
<td>Traumatic</td>
<td>(16)</td>
<td></td>
</tr>
<tr>
<td>Traumatic</td>
<td>62</td>
<td>18</td>
</tr>
<tr>
<td>Accidental</td>
<td>(33)</td>
<td></td>
</tr>
<tr>
<td>Iatrogenic</td>
<td>(24)</td>
<td></td>
</tr>
<tr>
<td>Surgical</td>
<td>(5)</td>
<td></td>
</tr>
<tr>
<td>Infection</td>
<td>38</td>
<td>11</td>
</tr>
<tr>
<td>Acute otitis media</td>
<td>(28)</td>
<td></td>
</tr>
<tr>
<td>Chronic otitis media</td>
<td>(4)</td>
<td></td>
</tr>
<tr>
<td>Chicken pox</td>
<td>(3)</td>
<td></td>
</tr>
<tr>
<td>Mononucleosis</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Mumps</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Diphtheria</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Tumour</td>
<td>19</td>
<td>6</td>
</tr>
<tr>
<td>Other</td>
<td>12</td>
<td>4</td>
</tr>
<tr>
<td>Melkersson-Rosenthal syndrome</td>
<td>(4)</td>
<td></td>
</tr>
<tr>
<td>Poliomyelitis</td>
<td>(3)</td>
<td></td>
</tr>
<tr>
<td>Guillain-Barré syndrome</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Hypothyroidism</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Sickle-cell crisis</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Myotonic dystrophy</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>(1)</td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>339</td>
<td>100</td>
</tr>
</tbody>
</table>

* From May (1986)

**Osteopetrosis (malignant variant)**

Malignant osteopetrosis is a rare cause of facial paralysis at birth, although it may be present later in childhood (*see Table 15.2*).
Table 24.12. Facial palsy at birth: differential diagnosis*

<table>
<thead>
<tr>
<th>Developmental</th>
<th>Traumatic</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>History</strong></td>
<td></td>
</tr>
<tr>
<td>No recovery of facial function after birth</td>
<td>Total paralysis at birth with some recovery noted subsequently</td>
</tr>
<tr>
<td></td>
<td>family history of facial and other anomalies</td>
</tr>
<tr>
<td><strong>Physical</strong></td>
<td></td>
</tr>
<tr>
<td>Other anomalies, bilateral palsy, lower lip or upper face palsy</td>
<td>Haemotympanum, ecchymosis, tics, synkinesis</td>
</tr>
<tr>
<td>Other cranial nerve deficits</td>
<td></td>
</tr>
<tr>
<td>Radiograph of temporal bone</td>
<td>Fracture</td>
</tr>
<tr>
<td>Anomalous external, middle, or inner ear; mandible; or vertical segments of facial nerve</td>
<td></td>
</tr>
<tr>
<td>Maximal stimulation / evoked electromyography</td>
<td></td>
</tr>
<tr>
<td>Response decreased or absent and without change on repeat testing</td>
<td>Normal at birth, then decreasing to possible loss of response</td>
</tr>
<tr>
<td>Electromyography</td>
<td></td>
</tr>
<tr>
<td>Reduced or absent response, no evidence of degeneration</td>
<td>Normal at birth, then loss of spontaneous motor units and 10-21 days later appearance of fibrillations and giant motor unit potentials</td>
</tr>
<tr>
<td>Abnormality in waves III-V</td>
<td>Auditory brain stem response</td>
</tr>
<tr>
<td></td>
<td>Normal, providing hearing is normal.</td>
</tr>
</tbody>
</table>

* From May (1986)

Management of developmental facial paralysis

At the present time, with the exception of free muscle neurovascular transplantation which is still investigational, there is no effective way to restore facial function in the newborn or young child with facial paralysis due to a congenital anomaly. It is in the child's best interest to delay reanimation surgical procedures until the patient reaches his or her adolescent years. Therefore, management of the newborn or young child with a congenital facial paralysis should be directed towards preventing complications and performing animation techniques which have very low morbidity. The main area of concern is the eye. Children with facial paralysis from birth usually do not have problems with keratitis and corneal scarring. However, this may occur, particularly if the child has poor Bell's phenomenon, decreased tearing, or entropion with irritation of the globe from eyelashes rubbing against the cornea. The child should be evaluated periodically by an ophthalmologist and, if there is any evidence of irritation or corneal keratitis, medical and perhaps surgical measures should be considered to correct the deformities.

Trauma

Facial nerve trauma can be either accidental (external trauma), surgical (unavoidable injury during surgery), or iatrogenic (unintentional surgical injury); the incidence of each is
approximately equal in the author's experience. Facial paralysis has been noted following infiltration of local anaesthetics to the ear, face, and oral cavity. The mechanism may be either direct infiltration of the trunk or branch of the facial nerve with the anaesthetic or precipitation of an inflammatory immune disorder, as noted with Bell's palsy. The time of onset after injection is most helpful from a differential diagnostic point of view. An unresolved facial paralysis following injection is most probably related to the needle penetrating the nerve; while when a patient recovers from the anaesthesia and then days later develops a palsy, the most likely type is Bell's (idiopathic). Treatment for a patient with paralysis following local infiltration involves all of the recommendations made for Bell's palsy. Prognostic signs include the completeness of the palsy, electrical test results, and the time that recovery is first noted. If the injury is incomplete (first through third degree), then the treatment is the same as that described for Bell's palsy.

**Surgical management**

There is only one absolute indication for surgical exploration and repair in acute traumatic facial paralysis: when the nerve has been transected. In cases of temporal bone fracture in the author's series, the nerve was most likely to be transected if there was marked displacement of the temporal bone fragments seen on the computerized tomographic (CT) scan and electrical response to stimulation was lost by the fifth day.

If it is known that the nerve has been disrupted, it should be repaired at the earliest possible moment; if the injury occurred during surgery, the ideal time for repair is at the time of operation. However, repair may need to be delayed if the patient's general condition is unstable or the wound is contaminated, although it should be performed within 30 days after injury for best results. The ideal choice is nerve repair, when possible, since this is the only technique that will provide any semblance of mimetic movement. The hypoglossal-facial nerve anastomosis is the procedure of choice when direct nerve repair is not possible, providing it is done within 2-4 years following the injury. Both nerve repair and hypoglossal-facial anastomosis techniques provide useful functional recovery when applied under the appropriate circumstances. However, both may result in mass movement and the inability to separate eye from mouth movement.

Regional reanimation techniques may be used as primary or secondary procedures to restore facial expression when the other two techniques are unsuitable, when they have failed, or when further animation is required. Although spontaneous or mimetic facial language cannot be restored with regional reanimation techniques, they have the advantage over nerve repair and hypoglossal-facial anastomosis of:

1. giving immediate results
2. separating eye from mouth movement
3. being independent of the cause and location of injury and between injury and reconstruction.

By applying the principles just discussed, it is possible to restore facial function to some degree for all patients with facial paralysis following trauma.
Tumours involving the facial nerve

Tumour types

Tumours of the head and neck may lie in close proximity, envelop, or invade the facial nerve as it courses from the brainstem through the temporal bone and parotid gland to reach the facial muscles. The benign lesion most frequently seen to involve the facial nerve is a schwannoma. Half of these tumours are acoustic neuromata (schwannomata) and are located in the cerebellopontine angle or internal auditory canal and, in the author's series, half were found to involve the facial nerve or chorda tympani nerve within the temporal bone. Vascular lesions such as meningiomata, angiomata, interosseous haemangiomata, and arteriovenous malformations were the second most common types of benign tumours causing facial palsy and, with few exceptions, involved the facial nerve extraneurally at or proximal to the geniculate ganglion. Unlike schwannomata, these lesions often could be separated from the facial nerve with preservation of residual function.

Malignant tumours may involve the facial nerve. The most common types of malignancy affecting the nerve in the author's series of patients have been adenoid cystic and mucoepidermoid carcinomata, and the most common site of origin of these lesions has been the parotid gland.

Reanimation immediately after tumour resection

Since brow function is rarely restored by grafting of the facial nerve or cranial nerve substitution, a brow lift is often performed immediately after resection of a tumour mass. Closure of the upper eyelid may be restored by implantation of a gold weight or a spring, and the lower eyelid may be tightened by a Bick (lid-tightening) procedure to protect the cornea.

The mouth region can be reanimated by transposing the temporalis muscle to the corner of the mouth. By performing this procedure in conjunction with eye reanimation procedures, the mouth and eye can move separately, which is not always possible with nerve grafting or cranial nerve substitution techniques. An additional advantage of this technique is that the results are immediate.

When facial paralysis has been long-standing preoperatively, or excision of the tumour mass leaves a facial nerve not amenable to grafting or even hypoglossal-facial nerve anastomosis, the techniques described for reanimating the eye and mouth may still be performed.

Facial paralysis in acute and chronic otitis media

In spite of the frequency of acute otitis media, particularly in children, associated facial paralysis is quite uncommon. A spontaneous satisfactory recovery is the usual course following treatment with an appropriate antibiotic and a myringotomy. Surgical therapy is indicated if the infection does not respond to these measures. Surgical decompression, even with loss of electrical response, has not been shown to improve recovery of facial function.
Chronic suppurative infection of the middle ear has a different natural history and does call for immediate surgical intervention when associated with a peripheral facial paralysis. Often the pathological process involves compression of an exposed nerve by cholesteatoma or chronically infected granulation tissue. Abscess and osteitis are not unusual findings at the time of surgery. Surgery, for the best possible recovery of facial function, should be carried out within 24 hours, provided that the patient's general condition permits.

**Hyperkinetic disorders**

The variety of hyperkinetic disorders has been mentioned under faulty regeneration. The three most common are essential hemifacial spasm, blepharospasm, and hemifacial spasm of the mass type or synkinetic variety following faulty regeneration. Primary hemifacial spasm is most effectively treated by retromastoid vascular decompression of the nerve. In cases that fail or in patients who are not candidates for an intracranial procedure, selective peripheral neurolysis may be tried, with resulting weakness. Another alternative is injections of botulinum A toxin. The effects of one injection last from 6 weeks to 6 months, and these may have to be repeated. The injections relieve the spasm, but are also associated with weakness, although this is not so marked as with selective neurolysis. Blepharospasm does not respond to retromastoid vascular decompression but has been effectively relieved by peripheral selective neurolysis, or selective resection of muscles in and around the eyelids (Anderson, 1982). Botulinum A toxin injections offer significant relief and are a much more conservative approach (Biglan, May and Walden, 1986). Severe hemifacial spasm secondary to faulty regeneration does not respond to retromastoid vascular decompression but can be relieved to some degree by selective myectomy, neurolysis, or injections.