Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 1: General Problems

Chapter 14: Facial Nerve Paralysis

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> *Paralysis of the facial nerve on one side only, and without other symptoms, always means disease of the nerve trunk. If it occurs without obvious disease or injury near the nerve after it emerges, it means disease of the nerve during its passage through the bone.*
>
> *Sir William Gowers, 1895*

Every physician should recognize these words of a celebrated neurologist and should know the facial nerve has the longest bony canal of any nerve in the body, that it is paralyzed more often than any other motor nerve, and that in 90 per cent of patients the facial palsy is due to disease within the temporal bone. However, too often a patient is assumed to have self-limiting Bell's palsy, when in fact paralysis of the face is due to cholesteatoma invading the bone, a tumor of the middle ear, or a neuroma of the facial nerve. Because management of lesions of this region should be the province of the otologic surgeon, patients who present with facial palsy should usually be seen by a specialist in such disorders.

Historical Background

The history of our knowledge of facial nerve paralysis is associated with the names of three great British surgeons, each one knighted for his scientific contributions.

In 1829, Sir Charles Bell demonstrated before the Royal Society in London that motor innervation of the muscles of expression is by cranial nerve VII. His vivid and picturesque description of facial paralysis cannot be surpassed: "... and the immediate effect has been the horrible distortion of the face by the prevalence of the muscles of the opposite side, and that distortion is unhappily increased when a pleasurable emotion should be reflected in the countenance".

For a long after Bell's demonstration, all cases of facial nerve paralysis were called Bell's palsy. Only in recent years has the application of this name been confined only to those disorders that come on abruptly and without apparent cause in an otherwise healthy individual.

The second great name associated with facial nerve paralysis is that of Sir Charles Ballance, founder of the Royal Society of Neurosurgery and author of the two-volume text *Surgery of the Temporal Bone.* In 1895, the same year that Sir William Gowers lectured on facial palsy, Ballance described the successful anastomosis of a severed facial nerve to cranial nerve XI. In 1932 at the age of 74 this indomitable surgeon, with Arthur B. Duel of New

York, published a series of articles detailing the success of nerve grafts in restoring continuity of the facial nerve in the fallopian canal, first in monkeys and then in human patients. The resulting restoration of emotional facial expression firmly established the advantage of facial nerve grafts over anastomosis to another nerve.

The third great contribution of facial nerve paralysis was Sir Terence Cawthorne, who in 1938 reported the first use of the operating microscope in an operation on the facial nerve within the temporal bone. This instrument, now used universally in facial nerve surgery, allows the nerve to be explored, decompressed, and grafted with minimal risk of surgical trauma to the nerve. Sir Terence continued to contribute to our knowledge of facial paralysis and presented the Fifth Gowers Memorial Lecture on December 5, 1968, at the National Hospital for Nervous Diseases on the subject "Intratemporal Facial Palsy".

Additional historical events in facial nerve surgery were the first repair of a severed facial nerve by approximation in its bony canal, reported in 1903 by L. Stacke; the first suture of this nerve in its bony canal in 1927 by Sterling Bunnell; and, in 1963, following removal of a facial nerve involved by an acoustic neuroma, successful insertion of a long graft from the stump of the nerve at the porus of the internal acoustic meatus to the nerve at the stylomastoid foramen by N. M. Dott.

Anatomy of the Facial Nerve

A fundamental and detailed knowledge of the anatomy of cranial nerve VII is essential for localizing the level of the lesion. It is necessary for arriving at the diagnosis and critical for the surgical approach in cases of compression by infection, neoplasms, and fractures.

The course of the facial nerve, for the sake of this discussion, is conveniently considered in three segments: supranuclear, nuclear, and infranuclear. The infranuclear segment is further subdivided into the cerebellopontine angle; internal auditory canal; and labyrinthine, tympanic, mastoid, and extracranial segments.

Supranuclear Segment

In the cortex, the tracts to the upper face are crossed and uncrossed. The tracts to the lower face are crossed only; therefore, the forehead is bilaterally innervated, so that if a lesion is present in the facial area on one side of the cortex, the forehead is unaffected. One must not rely solely upon sparing of the forehead to differentiate supranuclear from infranuclear lesions, however, since sparing of the forehead or other parts of the face can occur with lesions involving a more distal portion of the nerve. In addition to no change in function of the upper face, supranuclear lesions are characterized by no deficit in facial tone or spontaneous facial expression, but loss of volitional facial movement. Supranuclear lesions can also be identified by the presence of other neurologic signs of central nervous system involvement. The fact that involuntary movement is spared with supranuclear lesions is thought to be due to sparing of the extrapyramidal system, which is considered responsible for involuntary of emotional facial movement. With nuclear and infranuclear lesions there is loss of both involuntary and voluntary movement.

Nuclear Segment

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 (cranial nerve VI) nucleus before emerging from the brainstem.

Peripheral paralysis of nerve VII, ipsilateral paralysis of nerve VI, and inability to turn the eye on the non-paralyzed side toward the nose suggest a lesion of the fourth ventricle involving nuclei of nerves VI and VII as well as the para-abducens nucleus. 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. The facial motor fibers in the pons are topographically oriented, which may account for their partial sparing in lesions involving this area. This topographical orientation has been confirmed by a number of investigators, more recently by Radpour and Gacek.

Infranuclear Segment

Cerebellopontine Angle

At the cerebellopontine angle, cranial nerve VIII joins the facial nerve, and they course together until they enter the internal auditory canal. Thus, lesions in the cerebellopontine angle may cause vestibular and cochlear, as well as nerve VII, deficits. Large lesions filling the cerebellopontine angle might compress other cranial nerves and cause deficits of cranial nerves V and, later, of cranial nerves IX, X, and XI.

Internal Auditory Canal

The motor facial nerve and sensory intermedius nerve are loosely joined together as they enter the internal auditory meatus, side by side with the acoustic nerve. The sensory intermedius nerve lies inferiorly, while the facial nerve lies superiorly and runs along the roof of the internal auditory canal. The intracranial segment of the facial nerve from the brain stem 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; however, it is 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 sizable but slow-growing vestibular nerve schwannoma without any gross evidence of facial weakness. Although it is unusual to see facial motor involvement with such a lesions, there is often evidence of involvement of tearing, taste, and salivary flow because of compression of the sensory intermedius nerve. If one considers the sensory intermedius nerve to be part of the facial nerve, the facial nerve is the cranial nerve most often affected by vestibular schwannomas.

Labyrinthine Segment

At the fundus of the internal auditory meatus, the facial nerve is physiologically "pressed" into the fallopian canal. This 33-mm-long passage for the facial nerve in the temporal bone was described in the middle of the sixteenth century by the famed anatomist

of Padua, Gabriel Fallopius. Because of its resemblance to a water pipe, Fallopius called the canal an aqueduct. As they enter the fallopian canal the facial and sensory intermedius nerve carry with them a continuation of the dura mater and periosteum, which forms a well-defined and tough fibrous sheath that covers the nerve from the internal acoustic meatus all the way through the fallopian aqueduct to the terminal branches of the facial nerve in the face and neck.

The portion of the facial nerve between its entrance into the fallopian canal and the geniculate ganglion is called the labyrinthine segment because it runs between the cochlear and vestibular labyrinths. It lies beneath the middle fossa and is the shortest and narrowest part of the fallopian canal, averaging 5 mm in length by 0.68 mm in diameter. Since this is the narrowest part of the facial canal, it is reasonable to suspect that the facial nerve in this portion of the canal is the most vulnerable to inflammatory changes. The nerve in this area is further jeopardized by any process that causes compromise of this very limited space, because the blood supply to the nerve in this region is unique: this is the only segment of the facial nerve that has no anastomosing arterial arcades.

The labyrinthine segment includes the geniculate ganglion, from which arises the first branch of the facial nerve, the greater superficial petrosal nerve. This nerve carries secretory motor fibers to the lacrimal gland. The second branch from the geniculate ganglion is a tiny thread that forms the lesser superficial petrosal nerve as it is joined by fibers of the tympanic plexus, contributed by cranial nerve IX. This nerve carries secretory fibers to the parotid gland.

Tympanic Segment

At the geniculate ganglion the facial nerve makes a sharp right-angled turn backward, forming a knee, or genu, to enter the horizontal tympanic portion of the fallopian canal. The proximal end of the tympanic portion is marked by the geniculate ganglion and courses peripherally 3 to 5 mm, passing posterior to the cochleariform process and tensor tympani tendon. The distal end is approximately 12 mm long and lies just above the pyramidal eminence, which houses the stapedius muscle. At the beginning of the tympanic segment, the fallopian canal forms a prominent rounded eminence between the bony horizontal semicircular canal and the niche of the oval window. The tympanic wall of this part of the fallopian canal is thin and easily fractured. In addition, frequent dehiscences are present, allowing contact between the nerve and the tympanic mucoperiosteum. In some patients the uncovered nerve is prolapsed into the oval window niche, partially or completely concealing the footplate of the stapes and, therefore, subject to trauma during stapes surgery.

The surgeon must look for prolapse of the facial nerve when there is a congenital deformity of the incus and stapes superstructure. It is also worthwhile to palpate the horizontal segment of the facial nerve when performing middle ear or tympanomastoid surgery in order to determine whether the nerve is covered by bone, or if there is a dehiscence in the fallopian canal.

Just distal to the pyramidal eminence, the fallopian aqueduct makes another right-angled but more gentle turn downward - the second genu. The second genu is another area where the facial nerve may be injured during mastoid surgery. The nerve emerges from the

middle ear between the posterior canal wall and horizontal semicircular canal, just beneath the short process of the incus. When granulation tissue is present, as occurs with chronic infection, the surgeon must be very careful not to mistake a pathologic dehiscence of the facial nerve in this region for 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 facies nerve gives off its third branch, the motor nerve to the stapedius muscle, at the distal end of the tympanic segment.

Mastoid Segment

The fallopian aqueduct then turns downward to run along the anterior wall of the mastoid process. This mastoid segment ends at the stylomastoid foramen and averages, from the second genu to the foramen, 13 mm in length.

The chorda tympani nerve, the fourth branch of the facial nerve (its last sensory branch and thus the terminal branch of the intermedius nerve), usually arises from the distal third of the mastoid segment of the facial nerve, runs upward and anteriorly over the incus and under the malleus, and crosses the tympanic cavity through the petrotympanic fissure to join the lingual nerve. The chorda tympani nerve carries secretory motor fibers to the submaxillary and sublingual glands and carries fibers responsible for taste from the anterior two thirds of the tongue and information on pain, temperature, and touch from the posterior wall of the external auditory meatus.

Extracranial Segments

The remaining branches of the facial nerve arise outside the fallopian aqueduct, after the nerve has emerged from the funnel-shaped stylomastoid foramen. The foramen lies at the anterior end of the digastric groove, which houses the posterior belly of the digastric muscle. At this point the facial nerve gives off its fifth and sixth branches, the posterior auricular nerve, which ascends the anterior external surface of the mastoid process to supply the small muscles of the auricle, and the branch to the posterior belly of the digastric muscle. Both of these branches can be severed without causing subjective disability, allowing the facial nerve, after being freed from its attachments below the foramen, to be displaced upward several millimeters in the fallopian canal for approximation of the ends when a small segment of the nerve is missing.

The facial nerve sweeps around the styloid process posteriorly, then laterally, and as it passes forward into the substance of the superficial lobe of the parotid gland divides into two main branches, the upper one innervating the forehead, orbicularis oculi, and upper face, and the lower one supplying the muscles of the corner of the mouth, orbicularis oris, chin, and platysma of the neck. These two main branches, still within the parotid gland, divide into several smaller branches, forming the pes anserinus, or goose-foot, of the peripheral facial nerve.

Fallopian Canal Variations and Anomalies

The 33-mm-long fallopian aqueduct containing the facial nerve and its blood vessels is considered one of the most reliable and consistent anatomic landmarks in the temporal

bone. However, minor variations are frequent, and major anomalies, while rare, can lead to unavoidable surgical injury to the nerve by even the most experienced, skillful, and careful surgeon. The most frequent minor variation is dehiscence of the bony canal, most often in the tympanic portion but sometimes in the petrous portion where the nerve is in contact with the middle fossa dura and therefore easily injured during operation on the trigeminal ganglion. Occasionally it is dehiscent in the mastoid portion so that the unprotected nerve lies against the mucoperiosteum of a mastoid cell and is easily injured during mastoidectomy.

Major anomalies of the fallopian canal and facial nerve are most frequent in patients with congenital atresia of the external auditory meatus, but sometimes occur in patients with normal external auditory canals and tympanic membranes. For example, one of the authors (GES) accidentally sectioned a facial nerve while making a routine endaural incision in a patient who proved to have complete absence of a mastoid process, the nerve emerging from the temporal bone at the pyramidal eminence. In a patient undergoing stapedectomy, the uncovered facial nerve was found to cross the tympanic cavity between the oval and round windows; the nerve in this case was not injured. In one reported case, the nerve crossed the promontory inferior to both cochlear windows, whereas in another the nerve divided and embraced the crura of the stapes. A backward "hump" in the pyramidal bend will make the nerve particularly vulnerable during mastoidectomy, whereas if the nerve is divided in its vertical mastoid portion into two or three segments, each in its own bony canal, it will almost assuredly be injured during mastoidectomy.

In addition to the frequent minor variations and infrequent major anomalies that may occur in the fallopian canal, its location may be distorted and obscured by a tumor, suppurative disease, or previous surgery, so that its orientation is decidedly difficult to discern during surgery and accidental injury may be unavoidable. Even the most experienced otologic surgeon has unintentionally injured the facial nerve. According to one survey, the incidence of injury to the facial nerve during ear surgery, including stapedectomy and tympanomastoidectomy, was approximately 1:1000 patients. For these reasons, every patient undergoing ear surgery must be informed of the possibility of facial paralysis, even though it occurs rarely.

Blood Supply to the Facial Nerve

The blood supply to the facial nerve is derived from three sources: the anterior inferior cerebellar artery, which enters the internal auditory meatus in close association with cranial nerves VII and VIII; the petrosal branch of the middle meningeal artery, which runs along with the great 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. In spite of this, the area proximal to the geniculate ganglion is vulnerable to ischemic compression not only because this is the narrowest part of the fallopian canal but also because of the nerve's unique vascular supply at this point. There are no anastomoses between the arterial systems immediately proximal to this ganglion, which could make this segment of the facial nerve vulnerable to ischemia from edema. This might have bearing on the pathogenesis of facial paralysis following ligation or embolization of the middle meningeal artery.

Facial Nerve Sheath and Decompression

The "sheath" that surrounds the facial nerve through its course in the fallopian canal consists of periosteum, epineurium, and perineurium. Because the damage that occurs to the facial nerve with Bell's palsy has been thought to be the result of increased intraneural pressure, surgical decompression has often been performed in patients with Bell's palsy to relieve this pressure. However, it has not been established whether it is the sheath or the bony canal that constricts the nerve. Recently, much attention has been given to the anatomic relationships between the bony canal and the facial nerve as it enters the fallopian canal and makes it course through the labyrinthine segment. Fisch believed that the bottleneck at the proximal end of the fallopian canal could be the site of compression of the facial nerve in cases of acute inflammation such as accompany Bell's palsy and herpes zoster cephalicus. He pointed out that the results of intraoperative electrical testing show the block to be at the entrance of the fallopian canal and proposed that increased intraneural pressure resulting from inflammatory edema associated with Bell's palsy should be relieved by trans-middle fossa meatal decompression. Although attractive in theory, this hypothesis requires clinical confirmation by a prospective study with enough subjects for statistical validation.

Although surgical decompression, whether by opening the perineurium or the meatus, remains controversial in the management of Bell's palsy and herpes zoster cephalicus, opening the sheath is imperative in some cases of suspected tumor or trauma. A tumor may be intraneural and not discovered unless the sheath is opened, and in cases of trauma a hematoma may be deep to the sheath or the nerve may be disrupted and the pathology not appreciated unless the sheath is opened.

Spatial Orientation

Researchers have sought to determine whether the facial nerve is spatially oriented in its extra-axial course from the brain stem to the periphery, as it is in the cortex and pontine nucleus. If the nerve were spatially oriented it would be possible to localize lesions compressing the nerve in the cerebellopontine angle or within the temporal bone by reviewing the results of tests of specific motor functions. In addition, interrupted ends of the facial nerve in its extra-axial course could be re-approximated more appropriately, thus improving regeneration, if specific areas on the nerve could be identified as innervating specific end-organs. However, agreement is lacking on this subject.

Topographic organization of facial nerve fibers is unlikely, based on the work of several investigators who have found that the fibers destined for each peripheral branch are diffusely located in the facial nerve trunk. Gacek and Radpour studied the cross-sectional anatomy of the facial nerve through its course in the temporal bone and discovered degenerative changes in myelin 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 meatus carried motor fibers to all peripheral branches and that the axons of the facial nerve in the cat are not topographically arranged in the facial nerve trunk, as had previously been proposed. Based on these observations it is understandable that regeneration following facial nerve injuries usually results in some degree of mass movement and synkinesis.

Pathology of Axonal Block and Degeneration

Nerve impulses are transmitted by the axons, elongated extensions of the cytoplasm of a neuron or nerve cell. In the case of motor nerves, the parent nerve cell is in the brain stem or spinal cord and has but one axon, whereas sensory cell bipolar neurons that lie in nuclei outside the central nervous system have two axons, one from the peripheral sensory end-organ and one to the central nervous system.

The axon is covered by Schwann cells, which form a simple tunnel for nonmyelinated nerves and which lay down a spiral wrap of insulating myelin for myelinated nerves. At intervals of about 1 mm, the myelin sheath is interrupted at a node of Ranvier, representing the end of one Schwann cell and the beginning of another. A basement membrane is continuous on the outside of the Schwann cells and across the nodes of Ranvier, so that at no point is the axon in contact with extracellular space.

The axon must receive oxygen as well as be insulated from the Schwann cells. Its axoplasm must also be replenished by the parent neuron, because axoplasm is gradually depleted by catabolism at a constant rate that would result in complete loss of the axon in 29 days if it were not replaced. The rate of flow of axoplasm from the parent neuron is 1 mm a day, or approximately an inch a month, and this is the rate of regeneration of the axon when the nerve is cut.

Pressure on a nerve results in damming up of the flow of axoplasm. As long as a little axoplasm can squeeze by, there is a temporary but reversible loss of conductivity that is called *neurapraxia.* Release of pressure when neurapraxia is present results in rapid and complete recovery of function, with no residual effects.

Pressure sufficient to block replenishment of axoplasm completely results, not at once but after several days, in death of the axon, termed *axonotmesis.* If the entire nerve trunk dies by continued severe pressure or as a result of sectioning, *neurotmesis* has occurred. A process of degeneration in the axon or the nerve trunk then takes place, known as wallerian degeneration. The axons of the peripheral segment of a nerve, unable to be replenished by fresh axoplasm because of severe pressure or sectioning, live on for 2 or 3 days with continued electrical excitability but, of course, without conduction of impulses across the site of injury. The axons, isolated from their parent neurons and no longer able to receive axoplasm, then slowly die of starvation, with gradual decline in electrical excitability as the dying axon begins to fragment. Following severing of an axon, and as early as 48 hours after sectioning, the Schwann cells become swollen and phagocytic, breaking down the myelin. Macrophages remove the axonal and myelin debris. The Schwann cells then multiply until they fill the connective tissue tubule that surrounds each nerve fiber. Meanwhile, the parent neuron, deprived of nutrients normally brought to it by the axon, loses Nissl substance and undergoes swelling of its cytoplasm, known as chromatolysis.

At the proximal end of the cut axon, a growth cone soon appears, actively taking in nutrients by pinocytosis while it puts forth fanlike protoplasmatic processes that actively extend and then retract as they seek a favorable pathway for growth. The Schwann cells offer such a pathway, so that the tip of the axon grows into a Schwann cell, then on into the next Schwann cell in the connective tissue tubule, without being exposed to the extracellular space.

Because of the numerous protoplasmatic processes of the growth cone, a single regenerating axon is likely to branch and enter Schwann cells of several tubules, whereas a single Schwann cell may be shared by many small axons. Thus, the invariable result of regeneration of the facial nerve is that an axon that previously supplied a single muscle now supplies widely separated muscles, causing synkinesis or associated movements. When a patient who has experienced such nerve regeneration whistles or smiles, his eye winks, sometimes with embarrassing consequences.

The regenerating axon grows out at the rate of 1 mm a day, starting as a thin protoplasmatic thread less than 1 micron in diameter. As soon as the axon reaches the motor end-plate in the facial muscles, the continuing flow of axoplasm from the parent neuron causes it to begin to thicken. When the axon attains a thickness of 1 to 2 microns, myelin may begin to be formed around it by the Schwann cells; larger axons all become myelinated.

Because of the branching of the growth cone, the regenerated facial nerve has many more axons below the site of sectioning than above it, provided a dense scar has not formed at the site of sectioning to block many of the nerve tubules. Not only are axons more numerous in the regenerated nerve but also many more of them remain small and nonmyelinated than in the normal nerve. The deficient insulation of these many nonmyelinated axons results in mass movement and spasm of the facial muscles, in addition to synkinesis.

Classification of Injury and Recovery

Classification of Injury

Sunderland described five possible degrees of injury that a peripheral nerve fiber might undergo. This classification system is comprehensive and explains the course of physiologic events associated with all types of disorders that affect the facial nerve. The pathologic changes that occur in the nerve, as well as the anticipated responses of the nerve to electrical testing and the type of recovery one might expect with the various types of injuries are described. The first three degrees of injury may result from viral inflammatory immune disorders, such as Bell's palsy and herpes zoster cephalicus. The fourth and fifth degrees of injury may be due to disruption of the nerve during surgery or as a result of temporal bone fracture or tumor involvement.

A *first-degree injury,* referred to as *neurapraxia,* occurs when a physiologic neural block is created by increased intraneural pressure. The nerve will not conduct an impulse across the site of compression. However, the nerve will respond to electrical stimulation applied distal to the lesion. If the compression is relieved, return of facial movement may begin immediately or within 3 weeks.

A *second-degree injury* occurs if the compression is not relieved. The mechanism of injury is thought to be obstruction of venous drainage due to increased intraneural pressure, leading to further damming of axoplasm accompanied by proximal and distal swelling and to eventual interruption of the flow of nutrients to the nerve through the compressed arterioles. The result is loss of axons. This type of injury is referred to as *axonotmesis.* If the process is reversed, there will be complete recovery, although recovery will take longer than

with first-degree injury because it takes time for the degenerated axons to regenerate. The recovery begins 3 weeks to 2 months after injury and will be complete and without any evidence of faulty regeneration, provided that there is no loss of endoneural tubes.

Table 1. Neuropathology and Spontaneous Recovery Correlated with Degree of Facial Nerve Injury

Degree of Injury
Pathology of Injury
EEMG Response
Neurobiology of Recovery
Clinical Recovery Begins
Spontaneous Recovery - Result 1 Year Post

1°
Compression; damming of axoplasm; no morphologic changes (neurapraxia)
Normal
No morphologic changes noted
1-4 weeks
Grade 1 - complete: Without evidence of faulty regeneration

2°
Compression persists; increased intraneural pressure; loss of axons, but endoneurial tubes remain intact (axonotmesis)
25% of normal
Axons grow into intact empty myelin tubes at a rate of 1 mm/day, which accounts for longer period for recovery in 2° injuries compared with 1°; less than complete recovery is due to some fibers with 3° injury
1-2 months
Grade II - fair: Some noticeable difference with volitional or spontaneous movement; minimal evidence of faulty regeneration

3°
Intraneural pressure increases; loss of myelin tubes (neurotmesis) 0-10% normal
With loss of myelin tubes, new axons have opportunity to get mixed and split, causing mouth movement with eye (synkinesis)
2-4 months
Grade III-IV - moderate to poor: Obvious incomplete recovery to crippling deformity, with moderate to marked complications of faulty regeneration

4°
Above, plus disruption of perineurium (partial transection)
No response
In addition to problems caused by 2° and 3° injuries, the axons are

blocked by scarring, which impairs regeneration 4-18 months
Grade V - motion barely perceptible

5°
Above, plus disruption of epineurium (complete transection)
No response
Complete disruption with a scar-filled gap presents insurmountable barrier to regrowth of axons and neuromuscular re-anastomosis
Never
Grade VI - None.

Classification by groups I-IV modified from House, JW, and Brackmann, DE: Otolaryngol Head Neck Surg, 93:146, 1985.

A *third-degree injury*, or *neurotmesis*, occurs if intraneural pressure continues and there is loss of endoneural tubes. At this point, there will be a marked reduction in the response to electrical tests, and spontaneous recovery will not be noted for 2 to 4 months. As the axon regenerates, it is free to find any distal endoneural tube available, and since many axons may enter incorrect endoneural tubes, recovery will be incomplete and accompanied by synkinesis. The degree of faulty regeneration depends directly on the number of endoneural tubes that have been disrupted. It should be kept in mind that the various degrees of injury tend to overlap, since injuries to the nerve are not likely to be of a pure type.

Fortunately, the pathologic processes causing facial paralysis in patients with Bell's palsy and herpes zoster cephalicus usually do not progress beyond the first or second degree of injury, which accounts for the fact that most individuals recover satisfactorily from these disorders.

A similar process may be responsible for facial paralysis associated with acute suppurative otitis media, chronic otitis media and cholesteatoma, tumors, and temporal bone fractures. In each of these disorders, the nerve is usually not transected, but rather is compressed. In acute otitis media and trauma, compression may be sudden or slowly progressive, evolving over 5 to 10 days, just as is noted with Bell's palsy and herpes zoster cephalicus. However, unlike the process that occurs with Bell's palsy and herpes zoster cephalicus, in these 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 and compression of venous drainage leads to further compression of the nerve and loss of axons, and eventually to loss of endoneural tubes, which leads to third-degree injury.

Fourth- or fifth-degree injury results from partial or complete transection of the nerve. Spontaneous recovery should not be expected following fourth- or fifth-degree injury to the nerve; the best results of nerve repair come with surgical repair at the earliest possible moment following injury. Since most or all of the endoneural tubes have been disrupted, as well as the perineurium in fourth-degree injuries and the perineurium and epineurium in fifth-degree injuries, recovery even under ideal conditions is never as good as with the first three degrees of injury.

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 (Table 2). From a clinical point of view, patients who fall into grades I and II are considered to have satisfactory recovery compared with those who fall into grades III and IV, who are considered to have 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 in the latter group. The five degrees of injury described by Sunderland (see Table 1) fit in very nicely with the clinical classification of recovery reported by House and Brackmann (see Table 2).

Table 2. Classification System for Reporting Results of Recovery from Facial Paralysis

Degree of Injury	**Grade**	**Definition**
1°: Normal	I	Normal symmetric function in all areas
1°-2°: Mild dysfunction (barely noticeable)	II	*Slight weakness noticeable* only on close inspection; complete eye closure with minimal effort; slight asymmetry of smile with maximal effort; synkinesis barely noticeable; contracture or spasm absent.
2°-3°: Moderate dysfunction (obvious difference)	III	*Obvious weakness* but not disfiguring; may not be able to lift eyebrow; complete eye closure and strong but asymmetric mouth movement with maximal effort; obvious but not disfiguring synkinesis, mass movement, or spasm.
3°: Moderately severe dysfunction	IV	*Obvious disfiguring weakness; inability to to lift brow;* incomplete eye closure and asymmetry of mouth with maximal effort; *severe synkinesis, mass movement, or spasm.*
3°-4°: Severe dysfunction	V	*Motion barely perceptible;* incomplete eye closure, slight movement of corner of mouth; *synkinesis, contracture and spasm usually absent.*
Total paralysis	VI	*No movement;* loss of tone; no synkinesis, contracture, or spasm.

Recovery results were noted 1 year or less after onset.
Based on the system proposed by House and Brackmann and adopted by the Facial Nerve Disorders Committee of the American Academy of Otolaryngology-Head and Neck Surgery, Sept 16, 1984.

Facial Nerve Testing

Tears, Saliva, and Taste

Attempts to localize the site of a lesion using the results of tests for tearing, taste, and salivary flow popularized by Tshiassny have been of limited value when the lesion is acute, and of little or no value in patients with long-standing facial paralysis. This is true for the prognostic value of these tests as well, in contradiction to a previous report by one of the authors (M.M.).

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

1. The anatomy of the facial nerve and its branches is quite variable, allowing axons to take a variety of alternate pathways to reach their terminations.

2. The lesion responsible for the paralysis may affect different components of the nerve at various levels and with different degrees of severity.

3. Recovery of the various components may occur at different times.

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

Electrical Tests

Whereas tests of tearing, salivary flow, and taste have not been useful in diagnosis and prognosis, the prognosis in acute facial palsy can be determined accurately by serial electrical testing. When the time course of the degree of loss of response is plotted, this can be visualized. The steeper the line (within the first 10 days), the poorer the prognosis. Therefore, prognosis is based not only on the absolute response for the first 10 days but also on the acceleration of the loss within that period of time. The responses to electrical tests performed in the first 5 days after onset of paralysis have been found to be most helpful in prognosis. A study by May and colleagues showed that if a response to maximal stimulation or evoked electromyography (EEMG) of 25 per cent of normal or greater is maintained up to the tenth day after onset, the patient has a 98 per cent change of having a satisfactory recovery. If the response remains 11 to 24 per cent of normal for the first 10 days there is an 84 per cent chance of satisfactory recovery; however, there is only a 21 per cent probability of satisfactory recovery when the response to maximal stimulation or evoked electromyography drops to 0 to 10 per cent of normal within the first 10 days.

The problem for the clinician who sees a patient with an acute facial paralysis, regardless of cause, is to determine whether spontaneous recovery will be as good as or better than that expected after surgical intervention. Three electrical tests are useful in this situation: (1) the maximal stimulation test (MST); (2) evoked electromyography (EEMG), known also as electroneurography (ENOG); and (3) electromyography (EMG). The first two tests are capable of detecting early or ensuing degeneration, whereas the last test is useful when degeneration has occurred. Thus, the first two tests are useful in the first week following

onset, while the last test becomes useful by the tenth to fourteenth day following onset. EMG is also useful in the first 10 days in cases in which there is a question of transection of the nerve, or if the response to MST or EEMG is lost. The presence of voluntary motor unit action potentials in the first instance indicates less than total transection, whereas in the latter case, the nerve has become unblocked, is recovering function, or was not completely interrupted physiologically to begin with.

Maximal Stimulation Test

The principle underlying the maximal stimulation test (MST) is that a motor nerve will conduct in response to an electrical stimulus applied distal to a lesion, even though the lesion blocks volitional movement, provided that the nerve is morphologically intact distal to the lesion. This would occur in a first-degree injury. In a more complete lesion that has caused damage to the axon, as in a second- or third-degree injury, an increase in the intensity of the stimulus is required to cause a muscle to twitch. If the myelin and axon distal to the lesion have degenerated, as in third-degree or greater injuries, then no conduction will occur no matter how intense the stimulus. Because a completely sectioned nerve may continue to conduct distal to the section site as long as 5 days after the injury, the MST is of limited value until 3 to 5 days after the onset of paralysis. In addition, the MST is of value only as long as a response is noted. Once the nerve degenerates and response to electrical excitability is lost, the test is no longer useful. Duchenne, who first suggested the excitability test, stated that when excitability is lost after degeneration it returns in only a minority of cases, even if there is recovery and return of volitional movement. Another limitation of this test is the need to compare the results of the patient's involved side with those of the other side, and in patients who have experienced recurrent palsy or alternating bilateral involvement the other side may have decreased response as well.

The excitability test can be performed using any electrical stimulus that can be varied in strength and duration. The Hilger nerve stimulator is especially designed to test the facial nerve: model 2N is favored by one of the authors (M.M.). The instrument is conveniently portable and thus can be used at the patient's bedside. Stimulus intensity should be set at 5 mA or the highest setting tolerated by the patient without undue discomfort. An area of the patient's skin between the sideburns and the eyebrow and extending down over the cheek, brow, and neck is wiped with electrode contact paste, and the stimulating probe is then passed slowly down over this area. The response over the forehead, eye, nose, mouth, lower lip, and neck on each side is noted, and the response is recorded as equal, decreased, or absent on the involved side compared with the normal side.

The maximal stimulation test performed in the way just described was more reliable and became altered sooner than when performed as originally recommended by Hilger. When the response to maximal stimulation was equal, 12 per cent of the patients had incomplete return of facial nerve function; when the response to MST was decreased, 73 per cent of the patients had incomplete return. The test was most accurate when the response to MST was lost; all such patients had incomplete return of facial nerve function, with marked evidence of faulty regeneration.

When the facial nerve is involved by an acute process, as with paralysis following trauma or infection, the electrical tests should ideally be repeated daily until the response to

MST becomes abnormal or until return of volitional facial movement is noted.

Evoked Electromyography

Evoked electromyography (EEMG), known also as electroneurography, involves the recording of evoked summation potentials. This test was popularized by Fisch and Esslen and uses a principle similar to that of the maximal stimulation test. However, instead of depending on visual observation of the degree of muscle twitch elicited, in EEMG evoked summating potentials (SPs) are recorded on a graph produced by a sophisticated electrodiagnostic apparatus, the direct-recording electromyograph. The amount of degeneration is related to the difference in amplitude of the measured SPs on the normal and involved sides.

The great advantage of EEMG over the simple observation of facial movements used for the maximal stimulation test is the precise quantitative assessment of the response available with EEMG.

Fisch and Esslen recommended surgical exposure of the intratemporal portion of the facial nerve for management of (1) traumatic lesions, when the amplitude of the SP becomes 10 per cent or less of the value on the normal side within 6 days after the onset of the palsy; (2) idiopathic (Bell's) palsy, as soon as the SP is reduced to 10 per cent or less of normal within 2 weeks of onset of the palsy; (3) herpes zoster oticus when inner ear symptoms are present; and (4) acute otitis media, when there is reduction to 10 per cent or less of normal, even after paracentesis and antibiotic treatment.

The experience of one of the authors (M.M.) would modify the recommendations of Fisch and Esslen for surgery based on EEMG results. Surgical exploration of the facial nerve in patients with facial paralysis following temporal bone fracture should only be performed if three criteria are met: (1) onset of paralysis was sudden and complete; (2) there was loss of electrical response by the fifth day; and (3) the fallopian canal had been disrupted, as documented by computerized tomography (CT) or, although no disruption was shown by CT, the patient had no recovery of facial function by 6 months from time of injury.

Furthermore, using reduction in response to evoked testing as an indication for surgery to treat Bell's palsy, or herpes zoster oticus remains controversial, and although the response to evoked electrical testing is of value in predicting the outcome in patients with acute otitis media, it has not been useful as an indication for surgery. However, surgical therapy is indicated if the infection does not respond to medical measures, including appropriate antibiotics and a myringotomy. Nevertheless, surgical decompression, even when electrical response has been lost, has not been shown to improve recovery of facial function.

Electromyography

Denervated muscle, being hyperirritable, produces spontaneous electrical potentials referred to as fibrillation potentials. These fibrillation potentials do not appear until 10 to 21 days after degeneration occurs, but when they do occur they can be detected very reliably by electromyography (EMG), which samples motor unit activity. When the motor unit is intact, a motor unit potential can be detected with volitional movement, and when this is demonstrated following trauma, nerve transection can be ruled out. Further, reappearance of

motor unit potentials after degeneration of a nerve is one of the first signs of regeneration and can often be detected before volitional movement is noted.

Etiology of Facial Paralysis

Differential Diagnosis

Peripheral facial paralysis is a diagnostic challenge because the differential diagnostic possibilities are numerous (Table 3), but every effort must be made to determine the etiology because often a treatable cause can be found. Diagnostic clues are obtained from a carefully taken history and from the findings on physical examination (Table 4), from the results of special tests (Table 5), and from determining the level of involvement by the presence of clinical signs (Table 6). The relative incidence of causes of peripheral facial paralysis in the experience of one author (M.M.) can be seen in Table 7. Although in the majority of patients no cause can be found and the condition is labeled idiopathic (Bell's) palsy, it must be emphasized that Bell's palsy is a diagnosis of exclusion (Table 8). Thus, this diagnosis must be made accurately only after all other possible diagnoses have been eliminated.

Table 3. Causes of Facial Palsy Identified in a Review of Medical Literature (1900-1986)

Birth

Molding
Forceps delivery
Dystrophia myotonica
Möbius syndrome (facial diplegia associated with other cranial nerve deficits)

Trauma

Basal skull fracture
Facial injuries
Penetrating injury to middle ear
Altitude paralysis (barotrauma)
Scuba diving (barotrauma)
Lightning

Neurologic

Opercular syndrome (cortical lesion in facial motor area)
Millard-Gubler syndrome (abducens palsy with contralateral hemiplegia due to lesion in base of pons involving corticospinal tract)

Infection

External otitis
Otitis media
Mastoiditis

Chickenpox
Herpes zoster cephalicus (Ramsay Hunt syndrome)
Encephalitis
Poliomyelitis (type I)
Mumps
Mononucleosis
Leprosy
Coxsackie virus
Malaria
Syphilis
Scleroma
Tuberculosis
Botulism
Acute hemorrhagic conjunctivitis (enterovirus 70)
Gnathostomiasis
Mucormycosis
Lyme disease
Cat-scratch disease

Metabolic

Diabetes mellitus
Hyperthyroidism
Pregnancy
Hypertension
Acute porphyria
Vitamin A deficiency

Neoplastic

Benign lesions of parotid-vascular malformation, cystic hygroma, pleomorphic adenoma
Cholesteatoma
Tumor of cranial nerve VII
Glomus jugulare tumor
Leukemia
Meningioma
Hemangioblastoma
Sarcoma
Carcinoma (invasive or metastatic)
Anomalous sigmoid sinus
Carotid artery aneurysm
Hemangioma of tympanum
Hydradenoma (external canal)
Facial nerve tumor (cylindroma)
Schwannoma
Teratoma
Hand-Schüller-Christian disease
Fibrous dysplasia

von Recklinghausen's disease

Toxic

Alcoholism, ethylene glycol intoxication, arsenic poisoning
Thalidomide (Miehlke syndrome, cranial nerves VI and VII, with congenital malformed external ears and deafness)
Tetanus
Diphtheria
Carbon monoxide

Iatrogenic

Mandibular block anesthesia
Antitetanus serum
Vaccine treatment for rabies
Post-immunization
Parotid surgery
Mastoid surgery
Post-tonsillectomy and adenoidectomy
Iontophoresis (local anesthesia)
Embolization
Dental

Idiopathic

Bell's palsy, familial
Melkersson-Rosenthal syndrome (recurrent alternating facial palsy, furrowed tongue, faciolabial edema)
Hereditary hypertrophic neuropathy (Charcot-Marie-Tooth disease, Déjérine-Sottas disease)
Autoimmune syndrome
Temporal arteritis
Thrombotic thrombocytopenic purpura
Periarteritis nodosa
Guillain-Barré syndrome (ascending paralysis)
Multiple sclerosis
Myasthenia gravis
Sarcoidosis (Heerfordt's disease - uveoparotid fever)
Osteopetrosis
Amyloidosis.

History

Type of Onset

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 it anatomic course from the brain stem to the parotid. These other conditions include herpes zoster cephalicus, 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 to 6 weeks, or in whom the paresis progresses for more than 3 weeks; in such cases a tumor must be considered as the underlying cause. Although slow progression beyond 3 weeks is diagnostic of a tumor, progression that occurs within the first 10 days of onset has been noted with idiopathic (Bell's) palsy, herpes zoster cephalicus, external blunt trauma, and surgical trauma to the facial nerve within the parotid, temporal bone, or posterior fossa.

Table 5. Special Diagnostic Tests to Evaluate Patients with Facial Palsy

Topognostic tests

Hearing and balance tests
Schirmer test
Stapes reflex test
Submandibular flow test
Taste test

Electrical Tests

Maximal stimulation test (MST)
Evoked electromyography (EEMG)
Electromyography (EMG)

Radiographic Studies

Plain views of mastoid and internal auditory canal
Pluridirectional tomography of temporal bone
Computerized tomography of brain stem, cerebellopontine angle, temporal bone, skull base; contrast sialography of parotid gland
Magnetic resonance imaging
Chest radiographic survey to detect sarcoidosis, lymphoma, carcinoma

Surgical Exploration

Special Laboratory Tests

Lumbar puncture (cerebrospinal fluid) to detect meningitis, encephalitis, Guillain-Barré syndrome, multiple sclerosis, meningeal carcinomatosis
Complete white blood cell count and differential to detect infectious mononucleosis, leukemia
Mono spot test to detect infectious mononucleosis
Heterophil titer to detect infectious mononucleosis
Fluorescent treponemal antibody titer to detect syphilis
Erythrocyte sedimentation rate to detect sarcoidosis, collagen vascular disorders

Urine and fecal examinations:

Acute porphyria - elevated porphyrins and urinary porphobilinogen
Botulism - *Clostridium botulinum* toxin in stool specimen
Sarcoidosis - urinary calcium

Serum cryoglobulins and immune complexes to detect Lyme disease
Serum globulin level to detect sarcoidosis
Serum and urine calcium determinations to detect sarcoidosis
Serum angiotensin-converting enzyme level to detect sarcoidosis
Serum antinuclear antibody test (ANA) and rheumatoid factor (RF) to detect collagen vascular disorders (periarteritis nodosa)
Bone marrow examination to detect leukemia, lymphoma
Glucose tolerance test to detect diabetes mellitus.

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

Table 6. Signs and Probable Diagnosis Resulting from Lesions of the Facial Nerve at Various Levels

Level

Signs

Diagnosis

Supranuclear

Good tone; intact upper face; presence of spontaneous smile; neurologic deficits

Cerebrovascular accident, trauma

Nuclear

Involvement of cranial nerves VI and VII; corticospinal tract signs

Vascular or neoplastic disorder, thalidomide toxicity, poliomyelitis, multiple sclerosis, encephalitis, metastatic adenocarcinoma

Cerebellopontine angle

Involvement of vestibular and cochlear portions of cranial nerve VIII; involvement of facial nerve, particularly lacrimation and loss of stapes reflex; taste and salivation may be altered; cranial nerves V and later IX, X, and XI may become impaired

Neuroma, meningioma, cholesteatoma, fracture, metastatic adenocarcinoma

Geniculate ganglion

Facial paralysis; hyperacusis; alteration in taste, lacrimation, and salivation

Herpes zoster oticus, fracture, Bell's palsy, cholesteatoma, schwannoma, meningioma

Tympanomastoid

Facial paralysis; loss of stapes reflex; alteration in taste and salivation; lacrimation intact

Bell's palsy, cholesteatoma, fracture, infection, schwannoma, glomus jugulare, metastatic adenocarcinoma

Extracranial

Facial paralysis (usually a branch is spared); parasympathetic pathways and taste intact; deviation of jaw to normal side; palpable mass in area between ascending ramus of mandible and mastoid tip

Trauma, parotid carcinoma, pharyngeal carcinoma.

Facial paralysis has been noted to recur with idiopathic (Bell's) palsy, Melkersson-Rosenthal syndrome, and tumors. The incidence of recurrence in one author's (M.M.) experience with Bell's palsy was 12 per cent; in 34 per cent of these patients palsy recurred on the same side, and in 66 per cent on the opposite side. Thus, of all patients in whom idiopathic (Bell's) palsy was diagnosed, 4 per cent experienced ipsilateral recurrence. Of 48 patients with Bell's palsy (40) or tumors (8) who had recurrent ipsilateral facial palsy, 17 per cent were found to have tumors. This shows that the onset of facial palsy is not, of itself, diagnostic because tumors, like Bell's palsy, can present with incomplete, complete, sudden, delayed, or recurrent ipsilateral peripheral facial palsy.

Table 7. Causes of Facial Nerve Disorders in 2165 Patients seen over 22 years (1963-1986)

Cause	**Patients**	
	No	%
Bell's palsy	1150	53
Herpes zoster cephalicus	159	8
Trauma	437	20
Tumor	133	6
Infection	86	4
Birth	69	3
Hemifacial spasm	49	2
Central nervous system (axial) disease	23	1
Other	47	2
Questionable	12	1
Total	2165	100.

However, while recurrent facial paralysis on the same side may not be Bell's palsy, 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 disorders other than Bell's palsy.

Table 8. Bell's Palsy - Diagnosis of Exclusion

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

Signs of tumor
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).

Melkersson-Rosenthal syndrome is the most common example of a rare disorder that is characterized by recurrent alternating facial palsy. The classic presentation of this syndrome is recurrent swelling of the lip or face, fissured tongue, and intermittent facial palsy that usually begins suddenly, is of the peripheral type, and is clinically indistinguishable from Bell's palsy. The palsy may be partial or complete and is occasionally bilateral. Relapses are frequent, but the great majority of patients eventually recover. One third of 250 patients with Melkersson-Rosenthal syndrome studied by Hornstein (cited by May and Hughes) had fissured tongues, but a plicated tongue is notable in only 0.5 to 5 per cent of the normal population, according to Gorlin et al. This finding may point to a familial or genetic origin of the syndrome. Other associated findings that have been noted include swelling of the hands, chest, and buttocks; migraine headaches; and ophthalmologic abnormalities, including keratitis, blepharochalasis, retrobulbar neuritis, and retinal vessel abnormalities. Recently, it has been suggested by Minor and colleagues that Melkersson-Rosenthal syndrome can be diagnosed even when the only presenting syndrome can be facial edema.

Despite all that is known about this disorder, the cause and optimal treatment of Melkersson-Rosenthal syndrome have yet to be determined. In a previous report, one of us (M.M.) stated that in his experience there had been no recurrence of facial palsy following surgical decompression of the facial nerve in patients with Melkersson-Rosenthal syndrome. However, since that report was published 10 years ago, recurrence of palsy has been noted in two patients with Melkersson-Rosenthal syndrome after transmastoid facial nerve decompression. Graham and Kemink reported that following total facial nerve decompression using the middle fossa approach to treat recurrent facial paralysis in patients with Melkersson-Rosenthal syndrome, there was no recurrence during a follow-up period of 7 years. Their preliminary report suggests that edematous involvement of the facial nerve in recurrent paralysis can be prevented by transmastoid and middle cranial fossa total facial nerve decompression. Thus, decompression of the facial nerve canal through the middle fossa, as proposed and popularized by Fisch for managing Bell's palsy, may be gaining support for the treatment of recurrent facial paralysis.

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.

Differential Diagnosis by Physical Findings

Bilateral Simultaneous Palsy

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

Guillain-Barré Syndrome

Guillain-Barré syndrome is an acute inflammatory polyradiculoneuropathy evolving as a paralytic disease of unknown cause. The characteristic pathologic feature of Guillain-Barré syndrome is a lymphocyte cellular infiltration of peripheral nerves and destruction of myelin. The major complaint is motor weakness, ranging from mild ataxia to total paralysis of every motor and cranial nerve, which in most instances is noticed first in the legs, although it can begin in the arms. Tendon reflexes are abolished in the affected areas, and facial diplegia is seen in at least half of cases. When weakness evolves to total motor paralysis and respiratory muscles become involved, respiratory embarrassment may lead to death.

Abnormal cerebrospinal fluid (CSF) findings are characteristic of Guillain-Barré syndrome, although cells in the CSF are not prominent. At first, CSF may be normal, but by several days after onset of symptoms the CSF protein level begins to rise and may become very high, peaking in most cases 4 to 6 weeks after the onset of clinical symptoms. The absence of cells in conjunction with an elevated protein level is the albuminocytologic dissociation which at one time was thought to be characteristic of Guillain-Barré syndrome. Now, this recognizable disease entity is diagnosed by clinical, laboratory, and electrodiagnostic findings. In one author's (M.M.) 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; although uncommon, unilateral, recurrent, or simultaneous bilateral facial paralysis has been caused by this disorder. The disease, which is caused by the Epstein-Barr virus, can often be diagnosed by classical clinical presentation. 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 grayish-white exudative tonsillitis is practically pathognomonic, persists for 7 to 10 days, and is present in approximately 50 per cent of cases. Petechiae located near the border of the hard and soft

palates are observed in about one-third of patients toward the end of the first week of illness.

A hallmark of infectious mononucleosis is lymph node enlargement, usually of gradual onset and usually involving the anterior and posterior cervical lymph node chains. However, infectious mononucleosis resembles a number of other disorders; the fever, sore throat, adenopathy, and lymphocytosis of infectious mononucleosis 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 etiology. The differentiation depends on the results of throat cultures as well as on hematologic and serologic 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. The diagnosis is made on the basis of clinical findings and on the finding, in a biopsy specimen from the sarcoid, of a noncaseating granuloma with giant cells.

Facial palsy is the most commonly seen clinical neurologic deficit accompanying sarcoidosis. Uveitis occurs four times more commonly in patients with neurologic symptoms than in those without. The peripheral neuropathy associated with sarcoidosis has been shown to be due to perineural inflammatory changes that leave the nerve fibers themselves undamaged, which may account for the favorable prognosis with steroid therapy.

Lyme Disease

Lyme disease has been reported to cause bilateral facial paralysis. 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 geographic clustering of children with arthritis in the small community of Lyme, Connecticut. This spirochete disorder is transmitted by an arthropod vector, and should be suspected if the patient spent time where the tick vector is found on the northeastern coast of the United States, in the midwest (Wisconsin or Minnesota), or in California or Oregon. Exposure occurs more often during the summer or early autumn months when individuals spend more time outdoors and thus may be exposed to a tick bite. This disorder has been recognized in Australia and Europe, where it is referred to as Bannwarth's syndrome.

The tick bite begin 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 of Lyme disease include malaise, fatigue, chills and fever, headache, stiff neck, backache, myalgias, nausea and vomiting, and sore throat; some patients also develop neurologic symptoms. The diagnosis can be confirmed by sending a blood sample for serologic examination to detect characteristic cryoglobulins and circulating immune complexes.

In a report by Clark and colleagues, the incidence of facial palsy was over 10 per cent in all patients with Lyme disease; 25 per cent of these patients had bilateral paralysis. The prognosis for recovery is excellent, as only one of the 124 patients with 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 the prevention of serious late complications of Lyme disease; it does 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 that 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, and loss of tearing and taste.

Medical Management of Facial Palsy

Patients with acute facial palsy and their families are satisfied if answers can be provided to three questions:

1. What is the cause (diagnosis)?

2. When can recovery be expected (prognosis)?

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

In most patients who present with an acute facial palsy these questions can be answered after a thorough evaluation is performed during the initial office visit.

Diagnosis

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 4), 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 of recovery from this disorder are excellent, and the patient should be reassured that he or she has not had a stroke and probably will not be permanently deformed. Next, the time and degree of likely recovery are predicted by evaluating the following:

1. The completeness of the palsy.

2. The response to the maximal stimulation test or evoked electromyography.

3. The time when recovery first begins.

The degree of recovery can be categorized (see Table 2) as grade I (complete, with no detectable side effects), 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 retains some facial movement beyond 14 days after onset will have a satisfactory recovery from this disorder (grade I or II recovery).

Table 4. Differential Diagnosis of Etiology of Facial Palsy by History and Physical Findings

Bell's Palsy

1. Acute onset of unilateral facial palsy
2. Numbness or pain of ear, face, neck, or tongue (50%)
3. Viral prodrome (60%)
4. Recurrent facial palsy (12%) (ipsilateral 36%, alternating 64%)
5. Positive family history (14%)
6. Loss of ipsilateral tearing and/or submandibular salivary flow (10%)
7. Decrease in or loss of ipsilateral stapes reflex (90%)
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)
9. Self-limiting and spontaneously remitting

Herpes Zoster Cephalicus

1. Same as for Bell's palsy, except pain more common and severe
2. Vesicles on pinna, face, neck, or oral cavity (100%)
3. Sensorineural hearing loss and/or vertigo (40%)

Tumor

1. Sudden complete onset similar to Bell's palsy; evoked electromyography results abnormal (10% within 5 days)
2. Recurrent same side (17%)
3. Slowly progressive weakness beyond 3 weeks (59%)
4. No recovery after 6 months
5. Twitching with paresis
6. Mass in parotid gland, submandibular gland, or neck
7. Mass between ascending ramus and mastoid tip
8. Progression of other motor cranial nerve deficits
9. Some of branches of facial nerve spared
10. History of cancer

Bilateral Simultaneous Facial Palsy

1. Guillain-Barré syndrome
2. Möbius syndrome
3. Sarcoidosis
4. Myotonic dystrophy
5. Skull trauma
6. Infectious mononucleosis
7. Cytomegalovirus
8. Acute porphyrias
9. Botulism
10. Lyme disease
11. Bell's palsy - herpes simplex

Birth

1. Congenital diplegia (Möbius syndrome, thalidomide toxicity)
2. Lower lip palsy (developmental)
3. Trauma
4. Tumor

Trauma

1. Skull fracture (acute or delayed)

Infection

1. Bulbar palsy (viral meningitis, encephalitis, or immune reaction)
2. Following influenza, rabies, or poliomyelitis immunization
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

1. Acute porphyria

Neoplastic

1. Acute leukemia

Iatrogenic

1. Bilateral arterial embolization

Idiopathic

1. Guillain-Barré syndrome
2. Sarcoidosis (Heerfordt's disease - uveoparotid fever)
3. Periarteritis nodosa
4. Bell's palsy

Melkersson-Rosenthal Syndrome

1. Recurrent alternating facial palsy
2. Fissured tongue
3. Labial-periorbital facial edema
4. Nonspecific labial granuloma
5. Positive family history.

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.

Prognosis

As long as patients have 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. This is determined by daily evaluation of facial movement by the patient or a family member. The patient should stand in front of a mirror, or have a family member observe him or her, and evaluate facial function while 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 tumor should be suspected.

When a patient presents with a complete facial motor deficit the physician must rely on 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 about 11 per cent of normal, the patient is reevaluated every other day up to the fourteenth day post-onset. If, on the fourteenth day, the response to maximal stimulation persists of evoked electromyography remains above 11 per cent 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 per cent of normal or is lost completely within the first 14 days, then the prognosis for satisfactory recovery drops to 21 per cent.

Once the prognosis has been established, patients are asked to return in 3 months, 6 months, and finally 1 year for final evaluation of facial function employing the system of House and Brackmann. However, while the patient waits 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 medical treatment for acute facial palsy: physical therapy, pharmacologic treatment, and psychophysical therapy.

Physical Therapy. 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 his or her 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 fibers will be activated, and the exercises will help to maintain muscle tone.

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

Psychophysical Therapy. Modalities such as motor sensory reeducation have been useful. In the acute phase, integrated electromyographic tracings of motor strength can often be displayed on an oscilloscope, offering the patient significant encouragement at a time when no visible facial movements can be seen. The course of the recovery can be followed in this way, 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 including biofeedback, working in front of a mirror, and touching the face while attempting movements. These strategies are particularly useful in the post-acute phase, when recovery has plateau, as further improvement can be achieved using these techniques.

Management of Common Associated Problems

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 favorable for early recovery, the patient should be encouraged by this news, but if recovery is not expected for 2 to 4 months, the patient should be informed of this openly and should be supported sympathetically. Patient counseling and group therapy have been

effective in helping patients deal with this deformity, especially when they are selected to be of the same sex and similar age and when the counselor has had a satisfactory recovery or learned to deal with the problem in a positive way.

Physical Pain

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

Eye Care

Efforts should be directed toward keeping the globe moist to prevent keratitis and corneal breakdown. The patient should voluntarily close the eyelids about 2 to 4 times a minute on the involved side whenever the eye feels irritated or there is a burning sensation; also, 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 outdoors or the eye becomes irritated.

Surgery to reanimate the paralyzed eyelid(s) should be considered if medical treatment is ineffective, in particular when patients lack *B*ell's phenomenon, have corneal *A*nesthesia, and lack tears or have a *D*ry eye - the BAD syndrome. A tarsorrhaphy should be a last resort and, when performed, should be revised later when on of the more effective reanimation techniques is employed. This is because a tarsorrhaphy produces a cosmetic blight, limits vision, and often does not protect the exposed cornea; even when a tarsorrhaphy can be reversed, sequelae such as notching of the lid margin and trichiasis may result. 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.

Surgical Management of Facial Nerve Paralysis

Indications for Surgery

Surgical decompression of the facial nerve through the transmastoid or middle fossa route has not been shown to be an effective treatment for idiopathic (Bell's) palsy or herpes zoster cephalicus. Furthermore, 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. 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. In such cases, surgery should be performed prior to electrical denervation to give the most satisfactory facial function recovery, and it 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 evoked electromyography is less than 25 per cent of normal or is dropping precipitously after the third day following onset.

In addition, there are two situations where surgery is absolutely indicating in managing total facial palsy: (1) facial nerve transection and (2) tumor infiltration. Sometimes nerve transection or tumor infiltration can only be established by surgical exploration; this is true

in particular when the temporal bone has fractured or a tumor is suspected. In one author's experience (M.M.), when the facial nerve was involved by trauma or tumor, the lesion involved the nerve within the temporal bone in 90 per cent of cases. The lesion could be approached, with few exceptions, through a transmastoid route.

Surgical Techniques

Transmastoid Surgical Exploration of 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.

The transmastoid approach to the facial nerve consists of a preliminary mastoidectomy with removal of mastoid air cells from the antrum downward to the mastoid tip and defining the ridge of the digastric groove. 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 short process 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. Continuous irrigation with Ringer's solution or Tis-U-Sol removes bone dust and blood and prevents 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. In this way, 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. When performed under 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 triangular surgical approach bounded by the facial nerve medially, chorda tympani and tympanic annulus laterally, and short process of the incus superiorly. 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 maneuver can be done quite safely through the facial recess by gently separating the capsules of the incudostapedial and malleoincudal joints, leaving the short process of the incus attached to the fossa incudis. The incus can thus be rotated toward the middle ear to facilitate dissection over the proximal tympanic, geniculate, and distal labyrinthine segments. The incus can then be rotated toward 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 anatomic position, where it will remain, provided that the fossa incudis and the lenticular process of the incus have 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 Surgicel. Use of electric cautery is discouraged, as even a wet-field bipolar cautery may cause 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, this segment must be exposed and decompressed, which can be done by following the nerve into the facial recess through the postauricular approach (in some cases it may be necessary to remove the posterior canal wall for adequate exposure). To expose and decompress the tympanic portion of the nerve, 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 be need to be dislocated from the stapes to gain access to the tympanic fallopian canal; this technique, and replacement of the incus, were just described. The nerve is exposed at the site of injury and for at least 5 mm in both directions - until the nerve appears normal. 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 fibers are carefully preserved and the torn fibers 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 millimeters between the two ends. However, mobilizing the proximal and distal ends of the horizontal and vertical segments of the facial nerve to gain length to accomplish an end-to-end approximation is not the procedure of choice. Often, the more the nerve is freed from surrounding tissues, 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 if hearing and balance functions have been destroyed. In this case, the facial nerve can be freed, beginning at its first genu, by dividing the greater superficial petrosal nerve and mobilizing the vertical and horizontal segments posteriorly toward the internal auditory canal. A similar technique may be used to obtain enough length in the facial nerve to perform an end-to-end anastomosis to repair an injury to the nerve in the region of the parotid gland. Nevertheless, it is much better to put

a nerve graft between severed portions rather than try to mobilize the ends of the nerve to accomplish an end-to-end anastomosis. Based upon one author's experience (M.M.), the results were as good when a free graft was introduced as when a nerve was re-routed. Furthermore, if the nerve ends cannot be brought together without tension, then a free graft *must* be inserted, because 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 Grafting

Technique

When there is a gap between the cleanly cut ends of a severed facial nerve such 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 is suitable 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 obliquely with neurorrhaphy scissors. The nerve graft is handled carefully to avoid pinching or other trauma. Under a microscope, the epineurium of the ends of the graft and severed facial nerves are stripped back, and the nerve graft is carefully approximated to the distal and proximal stumps using 8-0 monofilament sutures. When a fascicular anastomosis is performed, the graft need no 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 is not 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 1 year after injury. When the central stump was not available or the time between injury and repair was between 1 and 2 years, the procedure of choice was a hypoglossal-facial anastomosis. When repair was performed between 2 and 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 (these procedures are described later).

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 following 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 during acoustic tumor 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 satisfactory waiting period before performing such nerve repair procedures, since it has been one author's (M.M.) 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 as the result of shrinkage and later gradual thickening of the collagen of the endoneurial sheath. This suggests that nerve repair should be undertaken without delay, so that axons that regenerate early can grow into the collapsing tubes to reinflate them and suppress collagenization before it progresses too far. Thus, in general, the sooner reinnervation begins, the better. The ideal time for nerve grafting is within 30 days and not later than 1 year following injury; beyond 1 year, the results of nerve grafting by any technique have been disappointing. The best results with a hypoglossal-facial nerve 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. Recovery has been noted with later repairs in cases in which part of the peripheral system was spared or spontaneously regenerated.

Many factors influence results following nerve repair. Some causes of less than ideal recovery include tension at the suture line, residual tumor, 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. One author's (M.M.) experience has shown that the most important factor is timing after injury.

The first sign of returning function is improving tonus of the paralyzed side of the face, which occurs before there is any voluntary movement. Even when a long graft was placed from the internal auditory meatus to the extracranial segment of the facial nerve, returning motion has been detected as early as 4 months; however, the average interval is 10 months and return of function may be delayed as long as 24 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 per cent of cases.

Hypoglossal-Facial Nerve Anastomosis

Special problems prevail when the site of damage to cranial nerve VII is in the posterior cranial fossa or the internal auditory canal. Surgical repair may be completed through the posterior fossa when the nerve is injured - for example, in the removal of a cerebellopontine tumor through the suboccipital route. Repair by direct anastomosis or nerve grafting has been described by Drake and Dott and may be done immediately or as a second-stage procedure.

In the event that such a course is not possible, the facial nerve distal to the injury, and usually the main trunk of the facial nerve outside the stylomastoid foramen, should be

anastomosed to the proximal end of the transected hypoglossal nerve. However, this operation leaves the hypoglossal muscles denervated and should only be undertaken after considering (1) the additional deficit that it causes, which is an inevitable result of the procedure, and (2) that the resultant facial nerve function may vary from complete flaccidity (facial paralysis) to restoration of tonus with some voluntary movement, but without emotional expression. A good result in this instance, then, is in distinction contrast to the optimal results following placement of a nerve graft or anastomosis of the cut ends of the facial nerve, which are expected to lead to return of emotional movement of the face. Furthermore, failure of a facial-hypoglossal anastomosis adds significantly to a patient's misery. Loss of the pursing action of the mouth and cheek, coupled with ipsilateral tongue paralysis, makes speech thick and less intelligible and mastication and initiation of swallowing more difficult, and it results in increased difficulty in drinking and trouble in handling normal salivary flow. With the passage of time, reorientation of motor facial function begins, and hemiatrophy reduces the bulk of the paralyzed tongue, which can relieve most of these problems. However, the patient will still have difficulty clearing the buccal gingival sulcus on the paralyzed side and often has to use a finger to move food to the normal side.

Thus, before plans are made to use this technique, the surgeon must fully inform the patient of the potential risks just noted. If cranial nerve X was injured during removal of the tumor, a hypoglossal-facial anastomosis should not be done, since adding a cranial nerve XII deficit to a cranial nerve X deficit may leave the patient unable to swallow without aspirating. Furthermore, the hypoglossal nerve should not be sacrificed in cases of von Recklinghausen's disease because of the possibility of subsequent involvement of other cranial nerves.

Other Techniques for Facial Reanimation

Results following facial nerve grafting or hypoglossal nerve anastomosis can be augmented by performing surgery to reanimate the eye and mouth. Brow lift, gold weight or eyelid spring implantation, and lower lid tightening, combined with temporalis muscle transposition, provide eye reanimation with mouth symmetry and voluntary movement within 3 to 6 weeks after the procedure. These procedures are described later.

Free muscle neurovascular repair techniques should be reserved for patients in whom 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 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, nonprogressive, non-life-threatening, and spontaneously remitting, and at this time the disorder can be neither prevented nor cured. The incidence varies between 15 and 40 per 100.000 population.

Clinical Features

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

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 per cent of patients will have a satisfactory recovery, provided that the palsy is incomplete and response to evoked electromyography remains greater than 10 per cent beyond the first 14 days after onset. Patients with a complete palsy and response to evoked electromyography of 10 per cent or less within the first 5 to 10 days have a 50 per cent 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.

Natural History

Peitersen studied the natural history of over 1000 patients with Bell's palsy seen over a 15-year period, and found that in 84 per cent recovery was satisfactory; 71 per cent recovered without sequelae, and 13 per cent had defects that were barely noticeable. The other 16 per cent of patients had obviously incomplete recovery of facial function; however, sequelae were crippling in only 4 per cent, and there was not a single patient who did not have some recovery. Peitersen noted that 85 per cent of the patients in his study began to recover facial function within 3 weeks of the palsy, which in 31 per cent 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 of 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 (Ramsay Hunt Syndrome)

Hunt first described a syndrome, now called herpes zoster cephalicus or herpes zoster oticus, that is characterized by a viral prodrome, with severe pain in and around the ear and with vesicles involving the pinna. In its mildest form there may not be any neurologic signs; 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 Ramsay 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 fibers are involved by the viral eruption, but all of the nerves that communicate with the facial nerve may be involved, including cranial nerves V, IX, and X and the cervical plexus arising from cervical nerves II, III, and IV. The sign common to all forms of herpes zoster cephalicus, which is necessary to establish the diagnosis, is the appearance of the vesicles.

Groves has presented a comprehensive review of facial nerve disorders, including a review of the literature on the history, etiology, and treatment of Ramsay 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 and postherpetic pain, and there is a poorer prognosis. 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 per cent of patients and a decreased response to electronystagmography in 40 per cent of patients with herpes zoster cephalicus. Bilateral vestibular suppression has been noted with this disorder, perhaps an indication of brain stem involvement.

Natural History

The natural history of herpes zoster cephalicus differs from that of Bell's palsy in several ways, perhaps reflecting the difference in behavior of herpes simplex type I and varicella-zoster viruses. Bell's palsy recurs in 12 per cent 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 to 10 days with Bell's palsy but in 10 to 14 days with herpes zoster cephalicus. Lastly, 84 per cent of individuals suffering from Bell's palsy have a satisfactory recovery of facial function, but only 60 per cent of those with herpes zoster cephalicus recover a satisfactory degree of facial function.

Treatment

Treatment of herpes zoster cephalicus is similar to that for Bell's palsy, with the exception that greater attention must be devoted to control of pain and of the vesicular eruption. Often narcotics are required for pain management, and a steroid antibiotic cream may be needed to treat the vesicular eruption. Use of antiviral agents hold promise but must be examined prospectively in a controlled randomized study. Prevention of chickenpox may eliminate herpes zoster, since it is a reactivation of chickenpox virus; a vaccine to prevent chickenpox has been announced, but the vaccine has not yet been released for clinical use.

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 (discussed earlier in this chapter under Type of Onset) 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 actually has a hereditary autoimmune basis.

Trauma

After Bell's palsy, the second most frequent cause of facial paralysis is injury to the facial nerve. Facial nerve trauma can be accidental (external trauma), surgical (unavoidable injury during surgery), or iatrogenic (unintentional surgical injury); the incidence of each is approximately equal in one author's (M.M.) experience.

Facial paralysis has been noted following infiltration of local anesthetics to the ear, face, and oral cavity. The mechanism may be (1) direct infiltration of the trunk or branch of the facial nerve with the anesthetic or (2) precipitation of an inflammatory immune disorder, as noted with Bell's palsy. The time of onset after injection is very helpful in making a differential diagnosis, as an unresolved facial paralysis following injection is most likely related to the needle penetrating the nerve, whereas when a patient recovers from the anesthesia and then days later develops a palsy, it is most likely to be Bell's (idiopathic) palsy. When a patient experiences paralysis following local infiltration, it may be managed like Bell's palsy, and the prognosis may be made in the same way - that is, based on the completeness of the palsy, electrical test results, and time that recovery is first noted. If the injury is incomplete (first, second, or third degree), then the treatment is the same as that described for Bell's palsy.

External Trauma

Temporal bone fractures may be classified as transverse or longitudinal but are often mixed. The facial nerve is injured more often and more severely with a transverse than with a longitudinal fracture. Transverse fractures across the petrous pyramid perpendicular to its long axis pass through the labyrinth, producing total loss of auditory and vestibular function on the involved side. Longitudinal fractures parallel to the long axis of the petrous pyramid commonly spare the labyrinth but may interrupt ossicular continuity, resulting in conductive hearing loss and facial palsy.

Although a diagnosis of temporal bone fracture is assumed in a patient with a history of head trauma and facial paralysis, in many such patients no fracture of the temporal bone can be established by the most sophisticated imaging techniques or even by surgical exploration. One likely explanation for the palsy that occurs in such cases is stretching of the facial nerve between the brain stem and the periosteal epineurial attachments at the distal end of the labyrinthine segment. Stretching may be followed by secondary edema or hemorrhage within the fallopian canal, causing the nerve to be compressed slowly and thus leading to facial palsy of delayed onset. When the paralysis occurs several days after the traumatic event, it can be assumed that the nerve has not been transected and that satisfactory spontaneous recovery will probably occur without surgical exploration. However, if there is firm evidence that the nerve has been transected, including a sudden complete paralysis, loss of electrical excitability by either maximal stimulation testing or evoked electromyography by the fifth day, and evidence of a displaced fracture through the fallopian canal on CT scanning, surgery is indicated. The transmastoid approach allows adequate exposure in the majority of patients requiring such exploration, although in the event that cochlear function

has been destroyed the translabyrinthine approach can be used to expose the facial nerve in the proximal labyrinthine segment to the internal auditory canal. If this area needs to be exposed in a patient who still has cochlear function and an uninterrupted ossicular chain, the middle fossa route may be used.

Surgical Trauma

Facial paralysis occurring during temporal bone surgery is more likely during reoperations, when the anatomic relationships have been obscured or altered by the preceding surgery. Minor or major anomalies of the facial nerve increase its vulnerability.

Surgical injury occurs, in order of decreasing frequency, in the following areas:

1. The tympanic portion, especially when the fallopian canal is dehiscent so that the nerve protrudes into the oval window niche, where it may be injured during stapedectomy.

2. The pyramidal bend, when the antrum is being sought in a contracted mastoid with a low tegmen tympani.

3. The vertical mastoid portion, when retrofacial cells are being removed.

4. The petrous portion during a middle fossa approach to the internal acoustic meatus or during trigeminal ganglion surgery.

Surgical injury to the facial nerve can also occur during removal of an acoustic neuroma. Injury to the nerve in this location differs from the injuries that may occur in the fallopian canal in that there is no rigid bony canal to press on the swollen nerve, so decompression is never indicated. If the surgeon feels certain that the nerve was not severed during removal of an acoustic neuroma, he or she should be patient and wait as long as 1 year for spontaneous recovery to occur.

While waiting for recovery, the patient should apply heat to the face, massage the facial muscles, and perform facial exercises daily. Electrical stimulation may also be used. Each of these modalities is directed toward improving the status and survival of the denervated muscles until reinnervation occurs. Although it is difficult to prove that ultimate recovery is improved by these forms of therapy, there is no question that the patient is much happier knowing something active is being done.

Surgical trauma to the nerve in the fallopian canal may result in swelling and first-degree injury of the intact nerve, or the nerve may be partly of completely severed with a fifth-degree injury. In some cases the nerve may be exposed deliberately by the surgeon during an ear operation, without visible trauma as viewed through the operating microscope; yet several days to a week later, paralysis may occur.

Evaluation for Intervention

The prognosis and treatment of postoperative facial paralysis are quite different, depending on whether the paralysis is immediate or begins only after an interval of several

days. Jongkees stressed that when postoperative facial paralysis occurs, the facial nerve should be explored and decompressed "before the sun sets". This has been interpreted to mean that re-exploration should occur within 24 hours of the original surgery. However, this is true only if the paralysis occurs following surgery in which the surgeon never identified the facial nerve and had no idea how the injury occurred. In such a case surgery would be indicated only if the palsy was immediate and complete and if response to electrical stimulation was lost by the fifth day. In the event that the patient maintained electrical response to maximal stimulation or evoked electromyography beyond 5 days, it would be unlikely that the nerve was transected or, for that matter, severely injured; however, more important, it is doubtful that surgical exploration would improve the natural history of the palsy. In addition, the adage certainly should not be held to when the surgeon was careful to identify the facial nerve in the course of the surgery and, prior to closure, stimulated the nerve proximal to the area of surgery and noted active facial movement. In such a case, even if immediate complete facial palsy is noted, nothing would be gained by re-exploring the facial nerve.

When complete facial palsy follows surgery immediately, other causes must be considered besides surgical injury. The first action should be to remove any packing that could possibly be compressing a dehisced nerve, such as may occur especially after modified or radical mastoidectomy. Another action is to evaluate what effect local anesthetics may have had on facial nerve function. Most anesthetics lose their effectiveness within 2 hours, but others are longer-acting and one must allow a greater period of time of observation before one can be sure that the paralysis is not due to the anesthetic. In evaluating a patient who experiences sudden complete palsy following surgery, it is important to base the prognosis on the degree of involvement of the nerve. The best place to look for movement is the forehead and midface. If the patient can raise the brow or create a forehead wrinkle or demonstrate lateralization of the corner of the mouth or formation of a lip cheek crease, it is certain that the function of the facial nerve has been spared. Nostril flaring is still another clinical sign of normal facial function and may be evaluated even in the immediate postoperative period. The patient may still be under the influence of general anesthesia and not completely alert; yet, by squeezing the nostrils between the thumb and forefinger for a few seconds and then releasing them, one can observe flaring of the nostrils. If there is flaring of the nostril on the involved side, this is a reliable sign of a less than complete facial nerve lesion.

Once the patient has recovered completely from anesthesia, and certainly within 24 hours, electromyography can be used to determine the presence of voluntary motor unit action potentials and the integrity of the facial nerve. Maximal stimulation and evoked electromyography results may not become altered until the third to the fifth day, even after the nerve is transected, whereas the presence of voluntary motor unit action potentials ensures that impulses arising from the brain are passing across the area of injury and are being detected in the facial muscles. This is a useful application of electromyography and may spare the patient unnecessary surgery and allow the surgeon to forecast a favorable prognosis. Although there may be some loss of volitional motor unit action potentials, the nerve is not necessarily transected or injured severely enough to require surgery; it means, at the minimum, that there is a complete physiologic block that prevents central impulses from passing through the site of injury to the periphery.

Timing of Surgical Re-Exploration

Although operating within the first few days after the nerve transection may prevent connective tissue proliferation, the results of surgery have not been significantly different whether the nerve was repaired immediately or up to 30 days after transection. Thus, there is no need for emergency surgery within 24 hours of the original surgery. In fact, there are many advantages to waiting to repair a transected nerve. It is in the patient's best interest, as well as the surgeon's, to schedule surgery as an elective procedure rather than as an emergency. Elective surgery allows time for proper consultation, appropriate observations, and tests to be performed that can be most helpful in the decision-making process. Perhaps as important is having the time for the patient and family to recover from the initial surgery and to be informed properly so that they can take an active role in the decision-making process. The details of surgical management have been discussed previously in the section "Exploration and Decompression or Repair for Traumatic Facial Palsy".

Patients who do not meet the criteria for nerve re-exploration because they will probably experience spontaneous recovery still need to be kept under observation, given reassurance, and treated medically, as discussed in the section "Medical Management of Facial Palsy". Recovery in these cases may be noted as early as 10 days and as late as 6 months after onset of the palsy. The time and type of recovery can be predicted to some degree by the completeness of the palsy, electrical test results, and the time that recovery is first noted, as discussed previously.

Other Trauma

The principles of management of surgical trauma and temporal bone fractures can be applied to managing intracranial lesions due to knife, glass, or bullet wounds, as well as parotid, submandibular, skull base, and maxillofacial surgery. The only exception would be management of a contaminated wound, such as may occur from a shotgun blast; in such cases the distal ends of the facial nerve should be identified and tagged, and nerve repair delayed up to 30 days if necessary to ensure that the wound is healthy.

Tumors of the Head and Neck

About 5 per cent of cases of facial palsy are due to a tumor. Tumors of the head and neck may lie in close proximity to, envelop, or invade the facial nerve as it courses from the brain stem through the temporal bone and parotid gland to reach the facial muscle.

The benign lesion that most frequently involves the facial nerve is a schwannoma. Half of these tumors are acoustic neuromas (schwannomas) and are located in the cerebellopontine angle or internal auditory canal. In one author's (M.M.) series, half were found to involve the facial nerve or chorda tympani nerve within the temporal bone. Surprisingly, an acoustic neuroma, until of enormous size, rarely causes facial paralysis, even though the nerve is thinned and stretched over the surface of the neuroma.

Malignant tumors may involve the facial nerve. The most common types of malignancy affecting the nerve in one author's (M.M.) series of patients have been adenoid cystic and mucoepidermoid carcinomas, and the most common site of origin of these lesions

has been the parotid gland.

General Principles of Managing Tumors

The primary consideration in resecting a mass lesion that involves the facial nerve is to cure the patient. However, if such resection causes facial paralysis where none existed preoperatively, the surgeon must make every effort to restore facial function. The procedure should be neither too radical for excision of a benign lesion nor inadequate to excise a malignant one completely.

The surgeon must consider not only actual removal of the tumor but also possibilities for facial reanimation at the time that a mass is resected. Even when facial paralysis was present before surgery, if the facial nerve is left intact, some function may be recovered spontaneously. This principle should be followed in some cases (based on the patient's instructions to the surgeon) even if less than total resection of a benign lesion is required to maintain integrity of the facial nerve, since even the best techniques for facial reanimation cannot duplicate natural facial nerve function. In most cases the cosmetic, functional, and emotional costs of damaging the facial nerve are greater than the morbidity of subtotal resection of a benign lesion.

Malignant lesions involving the facial nerve, on the contrary, usually should be resected with a wide margin of normal tissue, including normal facial nerve tissue. En bloc resection of such lesions, with segments of the facial nerve as far proximal and distal from the mass as possible, will ensure the optimal chance for local control and patient survival. Nevertheless, while the patient should be prepared preoperatively for the possibility of total facial paralysis, every effort should be made to restore some facial mobility by reanimation procedures performed either at the time of the resection or as soon postoperatively as possible.

The most desirable reanimation procedure is grafting the facial nerve to restore anatomic integrity. This procedure is usually only possible when the lesion that was removed was benign and is most likely to be successful if the patient had intact facial nerve function preoperatively. When the lesion was malignant or the patient had preoperative facial paralysis, transposition of the temporalis muscle to reanimate the mouth with other techniques for reanimating the eyelids is usually advisable. Table 9 lists guidelines for sparing or sacrificing the facial nerve during surgery to manage different types of benign or malignant tumors that may involve the nerve.

Table 9. Management of the Facial Nerve with Tumors of the Head and Neck

The Facial Nerve May Be Spared
When facial function is normal and tumor is:
Benign
Malignant but low grade and not adherent to nerve or involving deep lobe of parotid gland
Lymphoma
Leukemia

When facial function is impaired but tumor is:

Lymphoma
Leukemia
Schwannoma separable from nerve, or localized compression within tympanic or vertical segment.

The Facial Nerve Should Be Sacrificed

When facial palsy is total and due to:

Malignant tumor
Benign tumor which is invading schwannoma or enveloping (cholesteatoma)

When facial function is normal but tumor is:

Malignant or low grade, recurrent or adherent to nerve or involves deep lobe of parotid
Benign but recurrent and adherent to nerve (inseparable).

Now that the general principles in managing the facial nerve with tumors of the head and neck have been presented, the exceptions must be noted as well.

Sacrificing the Facial Nerve with Resection of a Benign Tumor

The indications for sacrificing the facial nerve when a benign tumor is resected include (1) presence of total facial paralysis for 1 year or longer preoperatively, (2) threat to the integrity of other structures or to life from a tumor located in the posterior fossa, and (3) inability to separate the tumor from the facial nerve, when allowing the tumor to remain in place will jeopardize the patient's life (glomus tumors, cholesteatomas, and recurrent pleomorphic adenomas). In each of these cases the patient is at greater risk if subtotal resection is performed to spare the facial nerve than if the facial nerve is resected to achieve total tumor removal.

Sparing the Facial Nerve When Malignant Tumors Are Resected

Some tumors that affect the facial nerve, although malignant in cell type, behave benignly. Acinic cell tumors and low-grade mucoepidermoid carcinomas may be treated like benign lesions by excising the tumor and sparing the nerve. Although the behavior of any tumor may lead the surgeon to revise his or her decision, most of these types of tumors may be resected without compromising facial nerve integrity as long as there is (1) a sizeable margin of normal tissue between the tumor and the facial nerve, and (2) the deep portion of the parotid gland is uninvolved. When such tumors present peripherally, it may be possible to resect only a portion of the facial nerve and thus preserve a significant degree of facial function.

Options for Reanimating the Face After Tumor Resection

Table 10 outlines the options available for reanimating the face following surgery in which a portion of the facial nerve was resected, and gives the indications for each option. Techniques for reanimating the eye are described. Since brow function is rarely restored by grafting of the facial nerve or by cranial nerve substitution, a brow lift is often performed immediately after resection of a tumor 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.

Table 10. Options for Reanimating the Face Following Resection of Facial Nerve

1. Grafting the facial nerve

Normal facial function prior to surgery
Proximal and distal nerve ends available
Favorable prognosis

2. Hypoglossal-facial nerve anastomosis

Facial palsy present for less than two years
Peripheral stump of facial nerve suitable for anastomosis
No contraindications to sacrificing the hypoglossal nerve

3. Regional reanimation (if first two options are not available)

Restoration of eyelid function
Implantation of gold weight or spring
Lower lid tightening
Temporalis muscle transposition

4. Combination procedures

Option 3 plus either option 1 or option 2
Eyelid reanimation only
Temporalis muscle transposition for immediate results and to augment other techniques that take longer to take effect.

The mouth region can be reanimated by transposing the temporalis muscle to the corner of the mouth. When this procedure is performed 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. Eye and mouth reanimation procedures can be combined readily with nerve grafting or hypoglossal-facial nerve anastomosis to provide greatly augmented facial function. When the temporalis muscle is used for mouth reanimation, it should be passed through a tunnel superficial to the superficial musculoaponeurotic system so that the dissection will be lateral to the distribution of the facial nerve branches; in this way, it will not interfere with other surgical techniques to reanimate the face.

Table 11. Causes of Facial Palsy in 366 Patients (newborn to 18 years old) Evaluated between 1963 and 1986

Causes	**No**	**%**
Bell's palsy	139	38
Tumor	19	5
Birth	69	19
Developmental (53)		
Traumatic (16)		
Trauma	74	20
Accidental (37)		
Iatrogenic (29)		
Surgical (8)		
Herpes zoster cephalicus	14	4
Infection	41	11
Diphtheria (1)		
Chickenpox (3)		
Mumps (1)		
Mononucleosis (1)		
Cholesteatoma (1)		
Otitis media (32)		
Questionable (2)		
Other	10	3
Polio (5)		
Guillain-Barré syndrome (3)		
Hypothyroidism (1)		
Myotonic dystrophy (1)		

Total	366	100.

When facial paralysis has been long-standing preoperatively, or excision of the tumor mass has rendered the facial nerve unsuitable for grafting or even for hypoglossal-facial nerve anastomosis, the techniques described for reanimating the eye and mouth may still be performed.

Management of Patients in Whom a Tumor Is Suspected But Not Found

When the results of physical and radiographic examination show no evidence that a tumor mass is present, but the clinician is still suspicious that a tumor is present (see Table 4), the facial nerve may be explored surgically from the labyrinthine segment to beyond the pes anserinus to complete the diagnostic evaluation. In 4 of 1575 patients with facial nerve disorders who were studied by one author (M.M.), a tumor was suspected but not found. Onset of facial paralysis was sudden in two of the patients and slowly progressive in the other two. Even though the cause of the paralysis was not clear, reanimation procedures were performed and provided some facial function. These patients are re-evaluated yearly, since evidence of a tumor may show-up years after the initial evaluation.

Infection

In spite of the frequency of acute otitis media, particularly in children, associated facial paralysis is quite uncommon (see Tables 7 and 11). 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. However, surgical decompression has not been shown to improve recovery of facial function even when such surgery has been indicated by loss of response to electrical stimulation.

The natural history of chronic suppurative infection of the middle ear is different from that of Bell's palsy, and immediate surgical intervention is indicated when such an infection is accompanied by peripheral facial paralysis. Often the facial nerve has been compressed by a cholesteatoma or chronically infected granulation tissue, and abscess and osteitis are not unusual findings. For the best possible recovery of facial function, surgery should be carried out within 24 hours of onset of paralysis associated with otitis media, provided that the patient's general condition permits.

Facial Nerve Disorders in the Newborn Children

Disorders of facial function in children can be due to a variety of causes and should not be assumed to be of the Bell's type. 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 the few exceptions noted here. The information presented is based on the diagnosis and management of facial paralysis in 366 patients, newborn to 18 years, seen between 1963 and 1986 (Table 1). The causes of facial palsy in these children were similar to those seen in adults, with the exception of paralysis noted at birth and the number of cases due to acute otitis media.

Facial Paralysis Noted at Birth

The differential diagnosis and treatment of facial paralysis in the newborn has been reviewed by May and associates and Harris and colleagues. The two main differential diagnostic possibilities are developmental and traumatic; the factors that aid in differentiating between them are listed in Table 12.

Table 12. Facial Palsy at Birth: Differential Diagnosis

Differential Diagnosis		
Evaluation	***Developmental***	***Traumatic***
History	No recovery of facial function after birth; family history of facial and other anomalies	Total paralysis at birth with some recovery noted subsequently

Physical examination

Other anomalies, bilateral palsy, lower lip or upper face palsy; other cranial nerve deficits

Hemotympanum, ecchymosis, ticks, synkinesis

Radiograph of temporal bone

Anomalous external, middle, or inner ear; mandible; or vertical segment of facial nerve

Fracture

MST/EEMG results

Response decreased or absent and without change on repeat testing

Normal at birth, then decreasing to possible loss of response

EMG results

Reduced or absent response; no evidence of degeneration

Normal at birth, then loss of spontaneous motor units; 10-21 days later, appearance of fibrillations and giant motor unit potentials

Auditory brain stem response

Abnormality in cranial nerves III-V

Normal, provided hearing is normal.

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. 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 inability to suck.

Syndromes Associated with Congenital Facial Paralysis

Möbius Syndrome. This is a rare congenital disorder that usually includes bilateral facial palsy, unilateral or bilateral abducens nerve palsy; anomalies of the extremities; absence of various muscles, particularly the pectoralis group; and involvement of other cranial nerves, particularly cranial nerves VIII through XII, and especially the hypoglossal.

Dystrophia Myotonica. Dystrophia myotonia (muscular dystrophy) 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 nerve paralysis, and only later show evidence of the progressive nature of the myopathy. Congenital facial diplegia associated with dystrophia myotonica that appears at birth is the earliest manifestation of the disease in its severest form. Unlike Möbius 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 blindness, and testicular atrophy are also present, and the neck is usually described as swanlike. This latter defect is due to wasting of the muscles of mastication and of the sternocleidomastoid muscle.

Thalidomide Embryopathy. Phocomelia (seal-like limbs) has apparently been known since Babylonian times, and was described by Ballantyne, 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-eight and forty-second day of pregnancy led to this type of limb deformity and also to associated arrested development of the ear and abducens nerve paralysis. Thus, this acquired congenital deformity is now called thalidomide embryopathy.

Osteopetrosis (Malignant Variant). Malignant osteopetrosis is a rare cause of facial paralysis at birth, although it may be present later in childhood.

Management of Developmental Facial Paralysis

At the present time, with the exception of free muscle neurovascular transplantation, which is still an investigational procedure, 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 child reaches adolescence. Therefore, management of the newborn or young child with a congenital facial paralysis should be directed toward preventing complications and performing animation techniques that 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 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; if there is any evidence of irritation or corneal keratitis, medical and perhaps surgical measures should be considered to correct the deformities.

Birth Trauma

Facial palsy may occur during birth by compression of the diploic bone of the infant's mastoid process, even when forceps are not used. The bone over the mastoid is almost paper thin, and it has been theorized that it can be compressed and push on the facial nerve as a result of the fetal position in utero, and particularly with parapartum molding as the head is pushed through the birth canal. Facial paralysis noted at birth has not often been attributed to this mechanism, but injury to the facial nerve during traumatic delivery does occur. Usually, there is evidence of the trauma - either a forceps bruise over the course of the facial nerve or a hematoma with diffuse ecchymosis behind the ear and blood in the external canal or deep to the drum. However, although the physical findings are most helpful in differentiating a traumatic from a developmental facial palsy at birth, they are often absent if the patient is evaluated weeks, months, or years after birth.

In the past, if the infant was born with a complete facial paralysis with evidence of ecchymosis over the postauricular area or a hemotympanum, immediate surgical exploration

and decompression was recommended. This recommendation has been revised in light of more recent observations. Since complete transection of the facial nerve from the forces applied during birth is so unusual, and the prognosis for spontaneous recovery of a newborn is so favorable, surgical exploration of the facial nerve should be considered only for infants who have clinical and electrophysiologic evidence of complete paralysis and in whom no improvement has been noted by 5 weeks of age. Even if surgical exploration at this time reveals that the nerve has been severely damaged, the ultimate outcome should not be significantly influenced by this delay. Since very few infants show no improvement in function by 5 weeks, there will be little need to explore the facial nerve of a newborn with traumatic facial paralysis.

Hyperkinetic Disorders

The basis for hyperkinetic disorders, faulty regeneration of the facial nerve, was discussed in the sections "Pathology of Axonal Block and Degeneration" and "Classification of Injury and Recovery". The three most prevalent hyperkinetic disorders seen following faulty regeneration are essential hemifacial spasm, blepharospasm, and hemifacial spasm of the mass type or synkinetic variety.

Primary hemifacial spasm is most effectively treated by retromastoid vascular decompression of the nerve. Should this fail, or for patients who are not candidates for an intracranial procedure, selective peripheral neurolysis may be tried. However, this leads to facial muscle weakness. Another alternative is injections of botulinum A toxin. The effects of one injection last from 6 weeks to 6 months, and although these injections are also associated with weakness, the weakness 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 by selective resection of muscles in and around the eyelids. Botulinum A toxin injections also offer significant relief to patients with this disorder and are a much more conservative approach.

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 botulinum A injections.