Chapter 184: Clinical Disorders of the Facial Nerve

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This chaper describes clinical disorders of the facial nerve. Most of the discussion is devoted to idiopathic paralysis (Bell's palsy); however, disorders of the facial nerve associated with other known disease states are also presented. The usefulness of electrodiagnostic testing and medical and surgical management of idiopathic palsy are discussed, but the methods and surgical techniques are covered in Chapters 149 and 185. Facial paralysis from tumors and trauma is discussed in Chapters 160 and 192.

Bell's Palsy: Spontaneous Idiopathic Facial Paralysis

Introduction and diagnostic criteria

The term *Bell's palsy* should be reserved for those cases of facial paralysis that have signs and symptoms consistent with the disease and in which a diligent search for another etiology is negative. Every experienced otolaryngologist has seen patients whose facial paralysis was caused by malignancy, facial neuroma, intracranial tumor, infection, or some other treatable cause but who were misdiagnosed by another physician. The dictum that "all that palsies is not Bell's" cannot be overemphasized.

Although the older literature has relegated Bell's palsy to a diagnosis of exclusion, May et al (1984) emphasized that it is a positive diagnosis based on specific clinical features. Taverner (1959) outlined the minimum diagnostic criteria for Bell's palsy: (1) paralysis or paresis of all muscle groups of one side of the face, (2) sudden onset, (3) absence of signs of central nervous system (CNS) disease, and (4) absence of signs of ear or cerebellopontine angle disease. The differential diagnosis of facial palsy is shown in Table 184-1.

Table 184-1. Differential diagnosis of facial paralysis

Acute paralysis

Polyneuritis

- Bell's palsy
- Herpes zoster
- Guillain-Barré syndrome
- Autoimmune
- Lyme disease
- HIV
- Kawasaki disease

Trauma

- Temporal bone fracture
- Barotrauma
- Birth trauma

Otitis media

- Acute bacterial
- Chronic bacterial
- Cholesteatoma

Sarcoidosis

Melkersson-Rosenthal

Neurologic disorders

- HIV
- Cerebrovascular - central or peripheral

Chronic/progressive

Malignancies

- Primary parotid
- Metastatic

Benign tumors

- Schwannoma
- Glomus tumor

Cholesteatoma

Incidence

The annual estimated incidence of Bell's palsy is 20 to 30 patients per 100.000 population. When categorized by age groups, the percentage is higher in patients over 65 (59 per 100.000) and lower for children under age 13 (13 per 100.000) (Adour et al, 1978; Hadar er al, 1983; Katusic et al, 1986).

The male/female ratio for Bell's palsy is approximately equal, except for a female predominance under the age of 20 and a slight male predominance over the age of 40 (Adour et al, 1978). The left and right sides of the face are equally involved. Rarely, patients have bilateral paralysis (0.3%) and 9% have a history of a previous paralysis (Adour et al, 1978). A family history of Bell's palsy is present in 8% of patients (Adour et al, 1978).

Etiology

The proposed etiologies of Bell's palsy include microcirculatory failure of the vasa nervorum (Hazama et al, 1972; Devriese, 1974), viral infection, ischemic neuropathy, and autoimmune reactions (Abramsky et al, 1975; McGovern et al, 1977). Of these, the viral hypothesis has been the most widely accepted (Adour et al, 1975a; Adour et al, 1978);

however, no virus has ever been consistently isolated from serum or nerve tissue from patients with Bell's palsy (Palva et al, 1978). Thus the evidence for the viral hypothesis is indirect, relying on clinical observations and changes in viral antibody titers. Furthermore, although there may be an underlying viral etiology, the immediate cause of the paralysis itself is still debated between the viral neuropathy alone or an ischemic neuropathy secondary to the viral infection (Fisch and Felix, 1983).

Acute facial paralysis can occur as part of many viral illnesses including mumps (Saunders and Lippy, 1959), rubella (Fowler, 1963), herpes simplex (McCormick, 1972), and Epstein-Barr virus (Grose et al, 1973, 1975). A viral cause for Bell's palsy is also supported by the finding that it appears to be part of a polyneuritis syndrome in which the facial palsy is the most obvious of the cranial nerves involved. A careful neurologic examination will reveal other cranial nerve weaknesses in over half the patients afflicted with Bell's palsy (Aviel et al, 1983a; Nieuwmeyer et al, 1985). Adour (19760 found several affected cranial nerves in most patients (Table 184-2).

Several serologic studies have demonstrated that a higher percentage of patients suffering from Bell's palsy have antibodies to herpes simplex specific antigen than do matched controls (Adour et al, 1975a; Hadar et al, 1983; Tovi et al, 1980; Vahlne et al, 1981a, 1981b). Adour et al (1975a) detected complement-IgG antibodies in all cases of acute facial paralysis versus 85% in set-matched control subjects. Hadar et al (1983) found IgG antibodies in 92% of patients versus 75% of controls ($P < 0.05$). However, despite these differences in the prevalence in the antiherpes simplex antibodies, the geometric mean of the titers was identical for the two groups.

On the other hand, other studies have failed to demonstrate increased titers to herpes simplex and varicella-zoster (Adour et al, 1978; Aviel et al, 1983a, 1983b; Brackmann, 1974; Njoo et al, 1988; Vahlne et al, 1981). For example, Aviel et al (1983a) found no changes in the level of immunoglobulins (IgG or IgM), complement (C3 or C4), or antiviral antibodies to herpes simplex, herpes zoster, Epstein-Barr virus, cytomegalovirus, adenovirus, influenza, or mumps.

The difficulty in interpreting these studies is that antibodies to these viruses are extraordinarily common in the general population. The constant evolution of technology also makes comparison of the studies more difficult. The study by Vahlne et al (1981) used radioimmunoassay techniques and that by Adour et al (1975a) used the complement fixation test. Hadar et al (1983) used the indirect immunoperoxidase antibody-membrane antigen technique, which avoids cross-reactivity between varicella-zoster and herpes simplex.

Because no convincing evidence for a significant rise of viral antibodies during the course of Bell's palsy has been found, the condition may result from the reactivation of a latent viral infection. This theory is supported by the finding of a modest increase in serum interferon levels during the course of Bell's palsy (Aviel et al, 1983a; Jonsson et al, 1989).

A few animal studies also support a viral etiology for Bell's palsy. Ishii et al (1990) were able to induce facial paralysis in guinea pigs by inoculating the surface of the facial nerve with herpes simplex virus type I. The severity of paralysis and subsequent degeneration were evaluated histologically and appeared to be more severe if the epineurium was removed

inoculation. Interestingly, these authors found greater extension of the virus within the nerve centrally than toward the periphery.

Recent evidence has suggested that Bell's palsy may have an autoimmune basis. Elevation of the serum levels of the complement component C1q, and C3-containing circulating immune complexes have been reported in the acute stage of Bell's palsy (Jonsson et al, 1987). Aviel et al (1983b) found a decrease in peripheral blood T lymphocytes and an increase in the percentage of B lymphocytes during the first 24 days after the clinical onset of facial paralysis. These patients had no change in the total number of peripheral blood lymphocytes, and there was no correlation between these results and the degree of paralysis or ultimate recovery. Both studies concluded that Bell's palsy may be an autoimmune demyelinating polyneuritis caused by a preceding viral infection, not unlike Guillain-Barré syndrome.

Although it is likely that the underlying disease in Bell's palsy is viral polyneuropathy, the question remains as to why there is such a profound effect on the facial nerve while the changes in the other cranial nerves are relatively minor and transient. The major anatomic difference between the facial and other cranial nerves is its long bony canal. Fisch (1981) measured the diameter of the fallopian canal throughout the temporal bone and found that the narrowest point was at the junction of the internal auditory canal and the labyrinthine portion of the fallopian canal, which he called meatal foramen. The meatal foramen averaged 0.68 mm in diameter, whereas the remainder of the fallopian canal ranged between 1.02 and 1.53 mm. He reasoned that edema in this narrow segment of the fallopian canal could cause damming of the axoplasmic flow within the facial nerve. This site of lesion in Bell's palsy has been confirmed by both clinical observation (Jenkins et al, 1985; for photograph see Fisch and Mattox, 1988) and by intraoperative neuronography (Fisch and Esslen, 1972b; Gantz et al, 1982). A similar site of lesion has been identified electrophysiologically in herpes zoster oticus (Fisch, 1977).

Histopathology of the facial nerve in spontaneous paralysis

Fowler (1963) reported the first postmortem autopsy findings in a patient who died shortly after developing Bell's palsy. The entire intratemporal nerve contained dilated and engorged veins and venules. Fresh hemorrhage was found within the internal auditory canal surrounding the facial nerve that extended as far as the geniculate ganglion. Fowler theorized that the ischemia resulted from microthrombi rather than vasospasm.

Reddy et al (1966) examined a facial nerve 17 days after the onset of spontaneous paralysis. The nerve showed scattered degeneration of the myelin sheaths and axons. Ten percent to 30% of facial nerve fibers were surrounded by phagocytic cells. The perivascular areas also showed inflammatory reaction and hemorrhage.

Proctor et al (1976) examined a patient who died from Bell's palsy 13 days after the onset of paralysis. The facial nerve showed lymphocytic infiltration and phagocytosis of myelin by macrophages throughout the infratemporal course of the nerve. Initially, this case was interpreted as evidence for a viral etiology. However, when McKeever et al (1987) reexamined the case, they emphasized that the inflammatory cells were most prominent where the facial nerve was surrounded by bone, especially in the labyrinthine portion of the fallopian

canal, but not within the internal auditory canal. They theorized that the histopathology suggested a compression-type injury with no evidence of vascular occlusion.

O'Donoghue and Michaels (1983) found degeneration of myelin sheaths and axon fibers throughout the intratemporal course of the facial nerve. The nerve was constricted at the meatal foramen, and an osteoclastic giant cell reaction caused resorption of bone around the geniculate ganglion. These authors interpreted their findings as consistent with a viral etiology for the palsy, but, as pointed out by Jenkins et al (1985), these findings do not preclude edema and constriction of the facial nerve as the final even leading to the paralysis.

Podvines (1984) found inflammatory infiltrates in the intratemporal portion of the facial nerve 6 months after the onset of the palsy, as well as signs of wallerian degeneration and regeneration. He suggested that the persistence of infiltrates for this extended time period may result from compromised circulation within the fallopian canal.

Jackson et al (1990) reported the histopathologic findings from the labyrinthine segment of the facial nerve in a patient 1 year after the onset of a total facial paralysis secondary to herpes zoster. A portion of the nerve within the auditory canal was essentially normal; however, at the meatal foramen, the nerve became a mixed fibrotic and necrotic acellular mass.

Michaels (1990) described the histopathologic findings from an autopsy performed 11 days after the onset of Bell's palsy. Like other authors, he found demyelination, compression, bone resorption, and lymphocytic infiltration in the proximal labyrinthine segment of the facial nerve. He cautioned against the overinterpretation of bulging of the nerve in the distal internal auditory canal because it was found in both the normal and pathologic sides.

Another approach to the histologic evaluation of Bell's palsy are biopsies obtained intraoperatively, including biopsies of the chorda tympani and greater petrosal nerve. Biopsies of the chorda tympani in acute Bell's palsy have shown degeneration of myelinated fibers but no inflammatory response (Jongkees, 1954; May and Schlaepfer, 1975). Fisch and Felix (1983) examined biopsies of the greater petrosal nerve obtained during middle cranial fossa facial nerve decompression. Their findings included degeneration and demyelination of large axons and lymphocytic infiltration. They found these results consistent with wallerian degeneration starting proximal to the geniculate ganglion, probably from the meatal foramen.

In summary, most of the histologic studies show diffuse demyelination of the facial nerve throughout its intratemporal course, with the most severe findings in the labyrinthine segment and at the meatal foramen.

CNS changes in Bell's palsy

Auditory symptoms, usually in the form of hyperacusis, may be present in up to 30% of patients with Bell's palsy (Adour et al, 1985). The cause for these auditory symptoms is unknown. McCandless and Schumacher (1979) were unable to find evidence of a lesion affecting the cochlear nerve in patients with facial paralysis. Many authors have attributed the hyperacusis to decreased damping of sound secondary to dysfunction of the stapedius muscle. However, most authors have not found reduced contralateral stapedius reflex thresholds,

suggesting that an absence of stapedial damping is not the cause (Rosenhall et al, 1983).

Based on these observations, many authors have concluded that the auditory dysfunction results from CNS involvement (Adour et al, 1985). A small percentage of patients with Bell's palsy develop auditory brainstem response (ABR) abnormalities when compared to age-matched controls. These abnormalities include an increase in the wave I to V interval and the interaural difference for wave V. These changes are usually present bilaterally and resolve with the recovery of the facial paralysis (Rosenhall et al, 1983). These findings suggested a brainstem abnormality associated with the Bell's palsy that these authors hypothesized is related to the reactivation of the herpes simplex virus. These findings must be interpreted with caution. Other authors have been unable to confirm such brainstem auditory findings (Hendrix and Melnick, 1983; Maurizi et al, 1987), and the findings have not been corroborated by somatosensory evoked potentials or visual evoked potentials.

A number of investigators have looked for CNS changes in acute facial paralysis by studying the cerebrospinal fluid (CSF). Again the results are contradictory. The finding of elevated levels of myelin basic proteins (Edström et al, 1987) and pleocytosis (Sandstedt et al, 1985) in the CSF support CNS involvement in Bell's palsy. Weber et al (1987) found a normal total cell count, total protein concentration, blood-CSF permeability, and CSF/serum immunoglobulin ratios in the majority of patients with Bell's palsy. Only about 10% of these patients had some pathologic feature of the CSF including mild increase in blood-CSF permeability or pleocytosis. Weber et al (1987) concluded that their findnigs did not support the hypothesis that Bell's palsy was a part of a cranial polyneuropathy.

CNS changes in Bell's palsy have also been suggested by magnetic resonance imaging (MRI). Jonsson et al (1990) found brain or brainstem changes in 5 of 19 patients with Bell's palsy. These areas of increased signal did not correlate with the facial nerve brainstem nucleus or the supranuclear pathways and were interpreted as indicative of unrelated vascular disease.

Electrophysiology and testing

The large number of branches from the facial nerve, including the greater petrosal, stapedial nerve, chorda tympani, and the multiple muscular branches, led to the development of topodiagnostic testing to localize the site of a facial nerve lesion. The Schirmer test evaluates greater petrosal nerve function by measuring the lacrimal secretions accumulating on a piece of filter paper placed under the eyelid at the medial canthus (Gontier and Fisch, 1976). Stapedial nerve function can be evaluated with stapedial reflex testing. The chorda tympani nerve can be evaluated with either taste testing or the submandibular salivary flow test described by Magielski and Blatt (1958). Unfortunately, the accuracy of localization using topodiagnostic testing has been disappointing (Gantz et al, 1982). This is not a surprising finding in Bell's palsy hwere the lesion is a diffuse demyelination throughout the nerve. However, topodiagnostic testing has also been disappointing in tumors where one would expect a sharply defined site of lesion (Pulec, 1977).

Electrodiagnostic tests on the motor branches of the facial nerve also have been used to assess function and predict outcome. The techniques for performing these tests are discussed in Chapter 149. Unfortunately, all of these tests have a similar shortcoming: Both stimulation and recording are performed distal to the lesion rather than on opposite sides of

the site of injury (Manni and Stenner, 1984). Therefore, one must wait until nerve degeneration has reached the site of stimulation, usually 4 to 5 days, before electrodiagnosti tests become abnormal.

Minimal nerve excitability test

The use of electricity as a neurodiagnostic test was described by Duchenne in 1892 and was adapted to the facial nerve by Laumans and Jongkees (1963). The Hilger nerve stimulator is the most commonly used method of facial nerve evaluation currently used by otolaryngologists. The extratemporal portion of the nerve is stimulated with a small pulsed DC current. The face is observed for the lowest current that will produce a visible twitch. A threshold difference of more than 3.5 mA between the two sides suggests some nerve degeneration.

Maximal stimulation test

A modification of the minimal excitability test, the maximal stimulation test, attempted to determine the difference between the strength and amount of contraction of the facial musculature caused by a supramaximal electrical stimulus (May et al, 1971). Not surprisingly the maximal stimulation test is difficult to quantitate and is more subject to interobserver variation than is the minimal stimulation test (Manni and Stennert, 1984).

Electroneurography (electroneuronography)

Electroneurography adds recording of the facial muscle action potential with surface or needle electrodes to the stimulation test. Esslen (1977) introduced the use of bipolar surface electrodes for both stimulation and recording of responses. The two electrodes are moved independently to produce the maximum amplitude of response. The response is evaluated by comparing the peak-to-peak amplitude of the maximum response obtained for the two sides of the face.

Electromyography

Electromyography measures muscle action potentials generated by *spontaneous* and *voluntary* activity. It is distinct from all the other electrodiagnostic tests in which the activity is generated by active stimulation of the nerve. Denervation potentials are seen 10 or more days after the onset of the palsy; therefore, they are of limited value in determining early prognosis of facial paralysis. However, the loss of voluntary motor units within the first 3 to 4 days of paralysis suggests a poor prognosis (Manni and Stennert, 1984). Conversely, retention of voluntary motor activity past the seventh day suggests that complete degeneration will not occur.

Interpretation of electrical tests

All of the electrical stimulation tests described previously have the same fundamental weakness; that is, inferences are made based on tests on the nerve distal to the site of injury within the temporal bone. Therefore, there is an inherent delay between the onset of the injury and the development of sufficient degeneration of the distal segment of the nerve to be

discovered by the tests. Only intraoperative monitoring, with stimulation proximal to the lesion, provides any direct information about the state of the nerve at the site of the lesion. New experimental techniques, including antidromic stimulation and magnetic stimulation, hold some promise for circumventing this limitation. For the moment, a combination of neuronography and electromyography appears to give the most accurate picture of the status of the facial nerve.

Electromagnetic stimulation of the facial nerve

Facial nerve testing with electromagnetic stimulation has also been explored. Benecke et al (1988) could stimulate the nerve with cortical (supranuclear) electromagnetic charges, demonstrating that the nerve could be stimulated transsynaptically. Direct stimulation of the facial nerve with electromagnetic stimulation also has been described. Based on latencies of compoung muscle action potentials from both magnetic and electrical stimulation of the facial nerve, Seki et al (1990) concluded that the root entry zone of the facial nerve was the most likely site of excitation with magnetic stimulation. These data are encouraging because the evaluation of Bell's palsy requires an ability to stimulate medial to the presumed site of the conduction block in the labyrinthine segment of the nerve. Meyer et al (1989) found absence of electromagnetic responses to be the earliest sign of nerve conduction block in patients with idiopathic palsy and that electromagnetic stimulation could differentiate between central and peripheral nerve lesions. It is not yet clear, however, whether magnetic stimulation will be a useful predictive test. In patients with Bell's palsy, no short latency responses could be obtained either acutely or after the onset of clinical improvement (Benecke et al, 1988). More data are needed to determine whether electromagnetic stimulation will be a useful prognostic test.

Imaging of the facial nerve

MRI has been used to study the peripheral facial nerve in Bell's palsy. Millen et al (1990) reported that four of five patients with acute facial paralysis showed gadolinium enhancement of the facial nerve within the fallopian canal. Schwaber et al (1990) evaluated 10 patients with acute and 7 patients with chronic Bell's palsy. All but one of these patients showed increased signal intensity with gadolinium enhancement of some portion of the facial nerve. The most common area for enhancement was in the internal auditory canal or labyrinthine segment. Enthusiasm for the use of this modality in the evaluation of Bell's palsy must be tempered by the lack of correlation between the severity of the paralysis and the degree of enhancement and by the high incidence of contralateral enhancement - 11 out of 17 cases in the series by Schwaber et al (1990).

Murphy (1991) reported gadolinium enhancement of the labyrinthine segment of the facial nerve in all patients with Bell's palsy in his study; a minority of patients also had enhancement of the mastoid segment. Those patients with more widespread enhancement had poorer outcomes.

Prognosis and statistics

The prognosis for most patients with Bell's palsy is excellent. Eighty percent to 90% of patients recover completely (Adour et al, 1985; Katusic et al, 1986). Many large series of Bell's palsy patients have been analyzed to identify prognostic factors that have a significant impact on outcome. The most important of these factors is whether the paralysis is incomplete or complete. The prognosis for persons who never develop complete facial paralysis is excellent: 95% to 100% of these patients recover with no identifiable sequelae (Katusic et al, 1986; Smith et al, 1988).

Many reports are available in the literature, but they are difficult to compare because they use different criteria for inclusion and evaluation of outcome. Stankiewicz (1987) collated the outcome from nine reports, including the large series of Park and Watkins (1949) and Peitersen (1982). The overall results from these series was 54% complete recovery, 44% partial recovery, and only 3% no recovery. The results in the general population are probably better than those that filtered through the various facial nerve clinics for inclusion in these studies.

Other factors that have been identified that are associated with poor outcome include hyperacusis; decreased tearing; age more than 60 years; diabetes mellitus; hypertension; and severe aural, anterior facial, or radicular pain (Adour et al, 1972, 1978). Abraham-Inpijn et al (1987), however, looked in detail at 200 patients and compared outcome with a number of potential prognostic factors, including severity of the facial paralysis, mean arterial pressure, age, clinical or chemical diabetes mellitus, and history of hypertension. Only the severity of the paralysis at its maximum extent and the mean arterial pressure at the time the patient was first seen proved statistically significant.

Treatment

The treatment of Bell's palsy has had a long and complex evolution that is not yet complete. Methods currently advocated include observation alone, steroids, surgical decompression, and antiviral agents.

The use of steroids in Bell's palsy was first proposed in a case report by Roghendler (1951). Since then steroids have become the most common treatment for Bell's palsy (AAO, 1982). Adour et al (1964, 1982) stated that while predinisone does not reduce the number of patients with *partial* denervation or contracture or synkinesis, it does reduce the number of patients with *complete* denervation therefore making the disease less severe.

Unfortunately no large randomized, double-blind trials have been performed comparing steroids to placebo in Bell's palsy. Stankiewicz (1987) carefully reviewed the evidence for the effectiveness of steroids. He concluded that although all the studies were statistically flawed or involved small numbers of patients, steroids *may* be helpful in preventing or lessening degeneration, synkinesis, progression of the palsy, and *may* help hasten recovery.

More recent studies have come to similar conclusions. Abraham-Inpijn et al (1987) found no statistically significant beneficial effect from the administration of steroids to a large group of patients suffering from Bell's palsy; however, the trend suggested a positive effect

of prednisone in patients with a more severe paralysis and a short delay preceding the institution of medication. Prescott (1987), however, found no beneficial effect of steroid treatment for Bell's palsy in children.

The use of steroids may be indicated for reasons other than the facial paralysis itself. Adour et al (1980) found that prednisone dramatically relieved the pain of Bell's palsy sufficiently to obviate the need for analgesics. Acyclovir is useful to treat facial paralysis associated with herpes zoster (see later for discussion); however, clinical trials of the usefulness of acyclovir in idiopathic paralysis are not yet available.

Surgical therapy has been even more controversial because, unlike steroids, additional injury may occur. The early enthusiasm for transmastoid decompression of the tympanic and mastoid segments of the facial nerve has waned (Yanagihara et al, 1979) and the procedure has been abandoned (May et al, 1984). The move away from transmastoid decompression has occurred both as a result of randomized trials showing that the procedure was not beneficial (May et al, 1984) and because of evidence that the site of the lesion is in the proximal labyrinthine portion of the facial nerve, which is inaccessible through the mastoid (Fisch, 1981).

The efficacy of surgical decompression of the meatal foramen and labyrinthine segment of the facial nerve in patients with poor prognosis as demonstrated by electroneuronographic testing is promising, but it has been difficult for any group to assemble a large enough series to definitively establish its value in randomized trials. Fisch (1981) compared 14 surgical patients with 90% or more degeneration as demonstrated by electroneuronography within 3 weeks of the onset of paralysis to 13 similar patients who refused surgery. He found a subtle but statistically significant improvement in the long-term facial recovery in the operative group. Graham and Kartush (1989) reported six patients with Melkersson-Rosenthal syndrome and recurrent facial palsy, none of whom had a recurrent facial paralysis after total seventh nerve decompression from the stylomastoid foramen to the internal auditory canal.

Special Cases of Facial Paralysis

Ramsay-Hunt syndrome

Herpes zoster facial paralysis (Ramsay-Hunt syndrome) differs from Bell's palsy because it is associated with varicella-zoster virus (as demonstrated by rising titers of antibodies to the varicella-zoster virus) and the presence of skin vesicles on the pinna, retroauricular area, face, or mouth. Compared to Bell's palsy, Ramsay-Hunt syndrome generally causes more severe symptoms and patients have a higher risk of developing complete nerve degeneration (Adour and Wingerd, 1974).

Varicella, or chickenpox, is the manifestation of the primary infection by varicella-zoster virus, (herpesvirus, varicellae) in a nonimmune host. Herpes zoster is the manifestation of this same virus in a partially immune host. Serologic and epidemiologic data strongly suggest that varicella-zoster represents the reactivation of a latent virus rather than reinfection. After the primary infection the virus probably travels to the dorsal root to extramedullary cranial nerve ganglia where it remains dormant until it is reactivated. Reactivation generally

occurs during a period of decreased cell-mediated immunity.

Varicella-zoster virus is the second most common cause of facial paralysis. The incidence of herpes zoster in patients with peripheral facial palsy has been calculated between 4.5% and 9% (Devriese and Moesker, 1988). A Mayo Clinic study (Ragozzino et al, 1982) estimated the annual incidence of herpes zoster as 130 cases per 100.000. The attack rate increased dramatically over the age of 60, and 10% of this population had identifiable risk factors for decreased cell-mediated immunity including carcinoma, trauma, radiation therapy, or chemotherapy. The increased incidence in the elderly population is explained by a decrease in cellular immune response to varicella-zoster virus with age (Burke et al, 1982).

In comparison with Bell's palsy, the severity of the paralysis is worse and the prognosis poorer in herpes zoster oticus. Peitersen (1977) reported full recovery in only 22% of patients, and Devriese (1977) found complete recovery in only 16%. As in Bell's palsy, the recovery is in part predicted by the severity of the paralysis. Complete recovery occurred in only 10% of patients after complete loss of facial function and in about 66% after incomplete loss (Devriese and Moesker, 1988).

The timing of the appearance of the vestibular eruption may have prognostic significance. In most cases, eruption and paralysis occur simultaneously. In approximately 25% of cases the eruption precedes the paralysis, and the likelihood of recovery is higher in this group (Devriese and Moesker, 1988).

Patients with Ramsay-Hunt syndrome are also more likely to have associated cranial nerve symptoms than patients with Bell's palsy, including hyperacusis, hearing loss, and pain (Robillard et al, 1986).

Severe ocular complications can be seen with herpes zoster ophthalmicus. These complications include uveitis, keratoconjunctivitis, optic neuritis, and glaucoma and are almost always associated with involvement of the ophthalmic division of the trigeminal nerve. Herpes zoster ophthalmicus may be difficult to differentiate from the localized skin rash associated with herpes simplex. Although both of these conditions may cause keratitis, differentiation between them is extremely important because topical steroids are used to treat herpes zoster but are contraindicated in herpes simplex. Opthalmologic consultation for biomicroscopy, staining, cytology, viral isolation studies may be used for the differentiation of these two conditions (Marsh et al, 1976). Adour (1982) stated that aside from concerns about opthalmic involvement, the development of skin vesciles either before or after initiation of prednisone is not a contraindication for steroid use.

The management of patients with herpes zoster, includingt cephalic zoster, is systemic corticosteroids (Reuler et al, 1984). A specific benefit of steroid therapy is a reduction of postherpetic neuralgia. The usefulness of steroids in fostering the recovery of facial paralysis is still controversial; however, early institution of steroids appears to relieve the acute pain, reduce vertigo, and decrease the incidence of postherpetic neuralgia (Adour et al, 1981; Keczkes and Basheer, 1980).

The antiviral agent acyclovir has also been recommended to treat herpes zoster facial paralysis (Adour and Hetzler, 1984; Ivarsson et al, 1987). Acyclovir is a nucleotide analog that interferes with herpes virus DNA polymerase and inhibits DNA replication. The drug is preferentially taken up by herpesvirus-infected cells. Early return of facial function after acyclovir treatment has been reported by some authors (Adour and Hetzler, 1984; Ivarsson et al, 1987), but no beneficial effects were reported by others (Schrader et al, 1989). At the least, treatment with acyclovir appears to lessen pain and promote resolution of the vesicles.

Congenital

The incidence of facial paralysis in newborns has been estimated at 0.23% of live births (McHugh et al, 1969). The first dilemma in managing facial palsy in an infant or young child is differentiating between a true congenital paralysis and birth trauma. The differential diagnosis of congenital facial palsy is shown in the box.

Box: Congenital facial paralysis

Congenital

Mononeural agenesis
- Congenital facial paralysis
- Congenital unilateral lower lip palsy (CULLP)

Facial paralysis with other deficits
- Möbius Syndrome (VI, VII, bilateral)
- Hemifacial microsomia
- Oculoauriculovertebral dysplasia
- Poland's syndrome (agenesis pectoralis major muscle)

Secondary to teratogens
- Thalidomide
- Rubella

Acquired

Birth trauma
- Forceps injury
- Pressure from maternal sacrum
- Pressure from fetal shoulder
- Intracranial hemorrhage

Idiopathic
- Bell's palsy

Systemic disease/Infectious
- Melkersson-Rosenthal syndrome
- Poliomyelitis
- Infectious mononucleosis
- Varicella
- Acute otitis media
- Meningitis.

Birth trauma usually causes an isolated facial paralysis as well as other signs of injury, including facial swellinhg, ecchymosis, or hemotympanum. Abnormalities of other cranial nerves or abnormalities on brainstem audiometry (prolongation of the I to III or I to V interval) suggest that the facial paralysis is congenital and not traumatic (May et al, 1981).

Seventy-eight percent of facial paralyses in infants are related to birth trauma. Surprisingly, these cases are equally divided between forceps deliveries and vaginal deliveries plus cesarean sections (Hepner, 1951; Smith et al, 1981), suggestingt that intrauterine facial nerve injury can occur from pressure on the infant's face by the sacral prominence during birth. Supranuclear palsy secondary to intracranial hemorrhage also has been reported (Paine, 1957).

The mildest form of congenital facial dysfunction is congenital unilateral lower lip palsy (CULLP), in which the defect is limited to an absence of depressor labii inferioris muscle activity. It is associated with a lesion of the brainstem (Portmann, 1977).

Möbius' syndrome represents a broad spectrum of clinical and pathologic findings ranging from isolated unilateral facial paralysis to bilateral absence of facial and abducens nerve function. Multiple other cranial nerves, including the glossopharyngeal, vagus, hypoglossal, and other extraocular motor nerves, may also be affected (Sudarshan and Goldie, 1985). These abnormalities may also be associated with a number of limb and head and neck developmental abnormalities, including congenital absence of the pectoralis major muscle (Poland-Möbius syndrome) (Hopper et al, 1985; Miller et al, 1989). Pathologic examination of the brain and brainstem of patients with Möbius syndrome shows a variety of abnormalities including dysgenesis or hypoplasia of the cranial nerve nuclei, brainstem infarction, and primary hypoplasia of the facial musculature (Harris et al, 1983; Sudarshan and Goldie, 1985).

Dysgenesis of the intratemporal facial nerve can also occur as an isolated abnormality. Computed tomography (CT) and surgical findings show that the lesion occurs most commonly in the distal portion of the mastoid segment of the facial canal. At this site the diameter of the bony fallopian canal becomes very narrow, and the nerve becomes little more than a fibrous band (Harris et al, 1983). Electromyographic studies in these cases usually show a small number of motor units, although there usually is no clinically useful facial function.

Habilitation of the face in congenital facial paralysis is usually with temporalis muscle transfer, tarsorrhaphy, upper eyelid gold weights, and free muscle transfers. Surgical exploration and decompression of the facial nerve is rarely useful (May, 1987).

Spontaneous facial paralysis in children

The incidence of Bell's palsy decreases with age; 8% of the patients in the series by Peitersen (1982) and only 2% in the series by Adour et al (1978) were less than 10 years old. Manning and Adour (1972) and Prescott (1987) found a female preponderance in this group.

The prognosis for children with Bell's palsy is uncertain. Peitersen (1982), Taverner (1959), and Prescott (1987) reported a high rate of spontaneous recovery in children with Bell's palsy, in part related to the high frequency of incomplete paralysis. However, both Alberti and Biagioni (1972) and Manning and Adour (1972) reported that a significant

percentage of children did not recover from their facial paralysis. Jenkins et al (1985) and Prescott (1987) argued that the prognosis was determined by the amount of nerve degeneration revealed by electrophysiologic testing, and that children and adults with similar electrical testing results had similar outcomes.

Familial facial paralysis

Adour et al (1978) reported 8% of patients with Bell's palsy had a positive family history, and Willbrand et al (1974) reported a 6% incidence. In addition there are sporadic reports of families with several affected individuals, usually each experiencing several attacks. Cawthorne and Haynes (1956) reported on two brothers, one of whom had five episodes of Bell's palsy and the other three. DeSanto and Schubert (1969) reported 10 cases of Bell's palsy in a family over an 83-year period and Willbrand et al (1974) reported a family with 29 cases of Bell's palsy over a 40-year period. Samuel (1984) reported a child who experienced four episodes of Bell's palsy involving both sides of the face; the child's father had six episodes of unilateral Bell's palsy. Amit (1987) described a family tree in which only females (the patient, her mother, maternal aunt, and maternal grandmother) all suffered from Bell's palsy. Three of the four had the onset during puberty, suggesting that hormonal influence may have been important. These sporadic cases are insufficient to determine conclusively a genetic influence. It can be said, however, that these cases often have an early age of onset, are recurrent, and have an excellent approach.

Recurrent paralysis

Adour et al (1978) found that 9.3% of patients with Bell's palsy had a history of previous paralysis. Similarly, Hallmo et al (1983) reported that 10.9% of patients had a history of a previous facial paralysis, and roughly equal proportions occurred on the ipsilateral or contralateral sides. Prescott (1987) reported that 20 of 228 children (9%) (< 18 years) with Bell's palsy had a history of a previous paralysis on the same side and another 8 had suffered a contralateral paralysis. The interval between the two attacks was usually more than 1 year (Yanagihara, 1984). Perhaps the record for recurrent facial paralysis was a woman who developed more than 50 episodes of unilateral lower facial paralysis after exposure to the disinfectant chlorocresol (Døssing et al, 1986).

The age distribution of patients experiencing recurrent facial palsy was the same as the overall population suffering from Bell's palsy (Adour et al, 1978), except for those with ipsilateral recurrent palsies, which may be associated with a younger age of onset (Yanagihara, 1984). Several reports note a slight female predominance in recurrent facial palsy (DeVriese and Pelz, 1969; Yanagihara et al, 1984). Diabetes mellitus was present in 39% of patients with recurrent paralysis in one study (Adour et al, 1978) but only in 4% in another (Hallmo et al, 1983).

Controversy continues as to whether the prognosis for recurrent facial palsy is better or worse on subsequent attacks. Several authors have stated that the second attack has a poorer prognosis and thus constitutes a stronger indication for surgical decompression (Auerbach et al, 1981). Most other authors, however, have found no prognostic difference between the primary and subsequent attacks of facial paralysis and no difference whether the second attack occurred on the ipsilateral or contralateral side (Hallmo et al, 1983).

Recurrence of facial palsy should prompt a careful investigation of the patient. May et al (1984) found a tumor in 8 of 40 (20%) of patients with a second palsy on the same side.

Bilateral facial paralysis

Bilateral idiopathic facial paralysis is much less common than unilateral paralysis and occurs in only 0.3% to 2% of patients (Sherwen and Thong, 1987). Bilateral facial paralysis has a much higher incidence of systemic causes than unilateral palsy and should spur a diligent search for an underlying etiology (McGovern, 1965).

The diseases most commonly associated with bilateral facial paralysis are Guillain-Barré syndrome, syphilis, leukemia, sarcoidosis, Lyme disease, and bacterial meningitis (Friedman et al, 1979). Most of these conditions are associated with other systemic or neurologic signs that suggest the appropriate diagnosis.

Guillain-Barré syndrome is a progressive ascending motor paralysis following a viral infection that usually affects the lower limbs. However, rare bulbar, myelitic, and cerebral forms of Guillain-Barré do not affect the limbs. Of the cranial nerves, the facial nerve is the third most commonly affected after the glossopharyngeal and vagus (Haymaker and Kernohan, 1949). The diagnosis is based on the typical clinical picture and the presence of elevated spinal fluid protein but a normal cell count.

Other conditions that have been reported in association with facial diplegia include influenza (Barkas, 1895), infectious mononucleosis (Bonfiglio and Suechting, 1964), other viruses (Schuring and Saunders, 1964), and diabetes (Hattori and Schlagenhauff, 1977). Heerfordt's syndrome (parotid enlargement, iridocyclitis, and cranial nerve palsy) associated with sarcoidosis can cause bilateral facial paralysis. (Sarcoidosis is discussed later.)

The disability caused by bilateral facial paralysis is dramatically more severe than that caused by unilateral paralysis. Aggressive eye care with ointment, taping, or patching is usually required; but this approach significantly interferes with visual function. Bilateral paralysis of the lower lip leads to a characteristic speech impairment as well as oral incompetence and drooling. In severe cases dental sequelae can occur from inadequate circulation of saliva (Tautin and Basasik, 1977). The psychologic impact can be devastating because these patients are incapable of any voluntary or emotional expression (Steenerson, 1986). Recovery in bilateral Bell's palsy is similar to that in unilateral palsy, although one side may recover before the other (Rontal and Sigel, 1972).

Progressive paralysis

Slowly progressive facial paralysis is not Bell's palsy. The differential diagnosis includes primary neuromas of the facial nerve (Fig. 184-1); metastases from squamous cell carcinomas and melanomas of the face and scalp; and (occasionally) distant metastases from kidney, breast, lung, and prostate (Gruber et al, 1989). Progressive facial paralysis can also occur from other primary temporal bone and cerebellopontine angle lesions as well as from carotid artery aneurysms (Brandt et al, 1986).

The presence of a tumor must be excluded in all of these cases, beginning with physical examination for neuroma of the peripheral branches of the facial nerve and continuing with CT and gadolinium-enhanced MRI scanning of the infratemporal and intracranial portions of the nerve. If these studies are negative, some authors argue for exploration of the nerve from the peripheral branches through the temporal bone (May, 1989). At the very least, patients with negative initial evaluations require careful serial observation and imaging.

Facial Paralysis Associated with Other Conditions

Pregnancy

The possible role of hormonal and fluid changes in the pathogenesis of Bell's palsy as been debated since Sir Charles Bell first suggested an association between idiopathic paralysis and pregnancy in 1830 (Bell, 1830). Bell's palsy occurs 3.3 times more frequently during pregnancy than in similarly aged nonpregnant women (Hilsinger et al, 1975) and most commonly occurs in the third trimester or immediately postpartum (Adour et al, 1978; Hilsinger et al, 1975). Recurrent palsy with repeated pregnancies and bilateral facial palsies during pregnancy have also been reported (McGregor et al, 1987).

In a series of 18 patients, Falco and Eriksson (1989) reported that preeclampsia was six times more common among patients with facial palsy than in the general population of pregnant women. Facial palsy in pregnant women cannot be correlated with preterm labor, low birth weight, fetal congenital abnormality, or other perinatal abnormality. The prognosis and outcome seem to be no different between pregnant women and their nonpregnant counterparts (Matthews, 1982).

Melkersson-Rosenthal syndrome

Melkersson-Rosenthal syndrome is a triade of symptoms including recurrent orofacial edema, recurrent facial palsy, and lingua plicata (fissured tongue). Orofacial edema is the defining feature; lingua plicata and peripheral facial paralysis each occur in half the patients. The complete triad is present only in one fourth of patients. The condition generally begins in the second decade of life, and the manifestations usually occur sequentially and seldom appear simultaneously.

The major diagnostic criterion in Meklersson-Rosenthal syndrome is persistent or recurring nonpitting facial edema that cannot be explained by infection, malignancy, or connective tissue disorder. The oral swelling usually involves the lips and buccal area; but the gingiva, palate, and tongue can also be affected. The swelling can extend to the supraorbital and infraorbital tissues of the cheek (Hornstein, 1970). The edematous lips assume a chapped, fissured, red-brown appearance. The swelling is usally transient but may recur at regular intervals.

After numerous recurrences the lips eventually develop permanent deformity. They may also develop chronic fissuring that is slow to heal and painful (Greene and Rogers, 1989). The extent of the facial swelling varies from unilateral involvement of the lower lip to bilateral total facial edema (Greene and Rogers, 1989). Chronic swelling not only causes

a cosmetic problem, but also may interfere with speaking and eating. The chronic and recurrent nature of the edema distinguishes it from transient angioneurotic edema. Biopsies of the lip reveal noncaseating epithelioid cell granulomas surrounded by histiocytes, plasma cells, and lymphocytes (Greene and Rogers, 1989).

The etiology of Melkersson-Rosenthal syndrome is unknown. Some authors have considered it a variant of sarcoidosis, but others dispute this theory. Some have suggested that it is a primary vasomotor disturbance (Streeto and Watters, 1964), whereas others have suggested it has an allergic cause. Elevation of angiotensin-converting enzyme, seen in sarcoidosis as well as some other diseases, has been reported in some cases (Orlando and Atkins, 1990).

Facial paralysis occurs in 50% to 90% of patients with Melkersson-Rosenthal syndrome and it has an abrupt onset, which is identical to that in Bell's palsy (Wadlington et al, 1984). A history of bilateral sequential paralysis and relapse of paralysis after initial recovery are common. The site of the paralysis usually corresponds to the area of facial swelling (Greene and Rogers, 1989). Symptomatic treatment of the facial paralysis is indicated. There are no randomized trials of steroids or surgery, although a cessation of the recurrent facial paralyses has been reported after facial nerve decompression (Graham and Kartush, 1989; Kettle, 1959).

Human immunodeficiency virus

Acute facial palsy can occur at any stage of human immunodeficiency virus (HIV) infection. The palsy can be a direct result of the HIV or secondary to the immunodeficiency, for instance resulting from a secondary infection by the herpes zoster virus. Most cases of facial palsy in early stage HIV-infected individuals resemble Bell's palsy in that they have an abrupt onest and there is no other identifiable etiology. These same individuals are also at risk for craniocervical form Guillain-Barré syndrome.

HIV is neurotropic and has been isolated from the CSF and neural tissues in all stages of HIV infection (Ho et al, 1985). Acute facial palsy in both acute and chronic HIV infection could result from invasion of the facial nerve or the facial ganglion by the virus (Karnes, 1984). Alternatively, it has been suggested that acute facial palsy may be part of an acute inflammatory demyelinating polyneuropathy that is seen in other nerves in early stages of HIV infection (Cornblath et al, 1987).

In later stages of HIV infection, facial nerve involvement may be related to the immunodeficiency resulting in cephalic herpes zoster or systemic lymphoma (Snider et al, 1983). In addition, the facial nerve may be involved with other nerves in a chronic peripheral polyneuropathy (Gray et al, 1988).

The prognosis for recovery from idiopathic paralysis in patients with acquired immunodeficiency syndrome (AIDS) is similar to that seen in the general population (Bélec et al, 1989). The prognosis in facial paralysis from other causes in AIDS patients depends on the underlying pathology.

Lyme disease

Lyme disease (Bannwarth's syndrome in Europe) is caused by the tick-borne spirochete *Borrelia burgdorferi.* The vectors for Lyme disease are several speciaes of *Ixodes* ticks. The primary reservoirs of the infection are the white-footed mouse and the white-tailed deer. The disease is known to occur in all parts of the USA.

Not unlike syphilis, Lyme disease occurs in several stages. It begins with erythema migrans, an influenza-like illness, regional lymphadenopathy, and general malaise. The erythema migrans is an enlarging, annular, erythematous-skin lesion, which may be multiple and is not limited to the site of the bite (Caruso, 1985). The second stage starts several weeks to months later when neurologic abnormalities develop, including meningitis, cranial nerve neuropathies, and other peripheral neuropathies. Early symptoms include headache, neck stiffness, and spinal pain. Stage three occurs months to years later in the form of chronic arthritis, neurologic deficits, recurrent meningitis, and subtle mental disorders (Logigian et al, 1990). Chronic neurologic complications occur in 18% of untreated patients with Lyme disease (Steere et al, 1983).

Ten percent of patients develop facial palsy and hearing loss. The facial paralysis may be unilateral or bilateral and may be the only neurologic abnormality. Facial weakness or paralysis nearly always resolves completely, although there may be mild, or rarely, severe residual facial weakness (Clark et al, 1985). Reik et al (1979) reported that 6 of 18 patients with neurologic complications developed facial palsy. Three of the six residual weakness after 5 months.

Treatment for Lyme disease is with ceftriaxone, 2 g/day intravenously for 14 days. Improvement is not expected for several months after the therapy and recovery is seldom complete (Logigian et al, 1990).

Kawasaki disease

Kawasaki disease, also known as infantile acute febrile mucocutaneous lymph node syndrome, is a multisystem disease primarily occuring in infants and young children. In addition to involvement of the mucous memberanes, skin, lymph nodes, and cardiac system (coronary artery aneurysms), neurologic complications have been reported in up to 30% of patients (Aso and Watanabe, 1984; Teresawa et al, 1983). Aseptic meningitis and irritability are the most common neurologic complications; however, facial palsy has been reported by several authors (Gallagher, 1990; Hattori et al, 1987; Kleiman and Passo, 1988).

In a collation of 17 cases with well-documented facial palsy collected from the literature, Gallagher (1990) reported the median age to be 13 months; none of the patients was older than 25 months. The average day of onset of the paralysis was the sixteenth day of illness, and, except for patients who died from cardiac complications, the palsy resolved in 1 to 12 weeks. Histologic examination of patients dying of Kawasaki disease demonstrated arteritis (Amano and Hazama, 1980), leading Teresawa et al (1983) to conclude that the facial paralysis resulted from ischemia of the nerve. Treatment of Kawasaki disease includes supportive care, appropriate treatment of cardiac failure if it occurs, and high-dose aspirin.

Sarcoid

Sarcoidosis is a chronic noncaseating granulomatous disease. Systemic involvement usually includes the lungs (hilar or peripheral adenopathy), polyarthralgias, anergy, hepatic dysfunction, and elevated serum calcium. Heerford's disease (uveoparotid fever), a variant of sarcoidosis, is characterized by nonsuppurative parotitis, uveitis, mild fever, and cranial nerve paralysis, most commonly of the facial nerve. Whereas only 5% of patients with sarcoidosis have cranial nerve involvement, the facial nerve is the most commonly affected (Delaney, 1977). In contrast, 50% of patients with uveoparotid fever suffer facial paralysis. The paralysis starts abruptly days to months after the onset of the parotitis. The cause of the paralysis is thought to be from direct invasion of the nerve by the granulomatous process (Waldenström, 1937) and not from pressure from the swollen parotid gland (Weiss, 1960). Sarcoidosis should be included in the differential diagnosis when the paralysis is bilateral (Sherwen and Thong, 1987).

The diagnosis can be confirmed by an elevated serum angiotensin-converting enzyme (ACE), in contrast to tuberculosis, cancer and pulmonary fungal diseases, in which ACE levels are low (Lieberman, 1976). Treatment of sarcoidosis is with steroids, after which the ACE levels are expected to return to normal (Cohen et al, 1983).

Otitis media

Facial paralysis from otitis media is rare, but can occur in either acute or chronic otitis media. In a recent series, otitis media accounted for only 3.1% of acute facial palsies (Takahashi et al, 1985). Of these 50 cases, only five were children with acute otitis media. The remaining adult cases were equally divided between chronic purulent otitis media and cholesteatomas. Three additional cases were from tuberculous otitis. The majority of paralyses were incomplete. Nearly all patients explored surgically had a dehiscence of the bony canal of the facial nerve, usually in the tympanic segment, which presumably allowed spread of the inflammation from the middle ear to the nerve.

Facial paralysis associated with acute otitis media, especially in infants and children, should be treated with parenteral antibiotics and a wide myringotomy for drainage. Surgical manipulation of the facial nerve in the face of acute otitis media is not recommended. Adour (1982) also recommended the addition of a 10-day course of corticosteroids in conjunction with myringotomy and antibiotics.

On the other hand, facial paralysis associated with chronic otitis media suggests a high probability of cholesteatoma, and surgical intervention is appropriate. The mechanism of facial paralysis associated with cholesteatoma could be either compression or inflammation. Djeric (1990) studied autopsy specimens from patients who had chronic otitis media but no antemortem evidence of facial paralysis. Two of twenty facial nerves had focal areas of demyelination, suggesting that adjacent inflammation may be more important than pressure.

Barotrauma

A curious phenomenon of recurrent facial paralysis with changes in barometric pressure has been described by several authors. Woodhead (1988) reported a 50-year-old man who developed repeated facial paralysis and ipsilateral loss of taste on ascending to 8.000 to 10.000 feet in a car or airplane. Symptoms reversed on descent. Similar cases have been reported after commercial airline flights (May, 1986) and after scuba diving (Becker, 1983; Edsvik and Molvoer, 1985). A brief facial palsy has also been reported after forceful nose blowing (Onundarson, 1990).

Barometric facial paralysis seems to be related to pressure changes in the middle ear transmitted directly to the facial nerve through natural dehiscences in the fallopian canal. The symptoms are relieved by pressure equalization tubes or other means of improving eustachian tube function (Woodhead, 1988).

Benign intracranial hypertension

The most common symptoms of benign intracranial hypertension are headache and visual disturbances, but occasionally it can be associated with cranial nerve palsies, falsely suggesting a localized lesion. The abducens nerve is the most commonly involved, occurring in 10% to 60% of cases (Johnston and Paterson, 1974). Unilateral, and occasionally bilateral, facial paralysis has been reported in a few cases of benign intracranial hypertension (Kiwak and Levine, 1984). These cranial nerve palsies tend to reverse promptly after reestablishment of normal intracranial pressure, either through medical means or by shunting.

Metabolic

Adour et al (1975b) reported that 17% of patients over the age of 40 with Bell's palsy had abnormal glucose tolerance tests, leading them to calculate that a person with diabetes is 4.5 times more likely to develop Bell's palsy than a person with diabetes. The incidence of diabetes, or at least abnormal glucose tolerance curves, in patients with Bell's palsy has been reported between 10% (Adour et al, 1974) and 56% (Abraham-Inpijn and Devriese, 1982). A case-controlled study of Paolino et al (1985) also found a frequency of abnormal glucose tolerance curves in 24% of patients with Bell's palsy compared to 13% of controls ($P<.005$). These findings warrant caution when prescribing corticosteroids to patients suffering from Bell's palsy.

Reversible facial paralysis has been reported with hypovitaminosis A (Sillman et al, 1985).

Central Facial Paralysis

Central facial paralysis is caused by a lesion in the parietal motor cortex or the connections between the cortex and the facial nucleus. Lesions in these areas are usually accompanied by other neurologic symptoms referable to the CNS. A supranuclear palsy of the face most severely affects the lower half of the face and spares the forehead. Another important sign is that although voluntary motion may be severely affected, emotional facial reaction remains intact.

Hyperkinetic Disorders

Hemifacial spasm

Hemifacial spasm is an involuntary twitching and contraction of one side of the face. The spasm usually starts around the mouth and, as the disease develops, involves the entire face (Podvines, 1984). These uncontrollable spasms cause severe functional, social, and psychologic dysfunction. Although the spasms are usually painless, a rare form of the disease also involves the trigeminal nerve.

The etiology of hemifacial spasm remains a widely debated subject; most believe that the origin of the disorder is somewhere in the peripheral facial nerve. Esslen (1957) hypothesized that pressure or ischemia within the fallopian canal could generate spontaneous action potentials. Jannetta (1975) thought that compression of the seventh nerve by aberrant blood vessels near its exit from the brainstem (root entry zone) was the cause, although others disagree with this conclusion (Adams and Chir, 1989). Jannetta offers electrophysiologic findings in support of his conclusions. Intraoperative facial electromyographic recordings showed that stimulation of the zygomatic branch of the facial nerve would cause immediate and prolonged responses that could be recorded in the mentalis muscle. In the majority of patients, these responses disappeared or decreased when the facial nerve was decompressed, and the amount of decrease roughly correlated with the postoperative result (Møller and Jannetta, 1987).

The best treatment of hemifacial spasm has also been hotly contested. Many surgeons have advocated some form of partial destruction of the peripheral facial nerve, either partial resection of the three main branches (McCabe, 1970; Miehlke, 1960), or resection of involved branches determined by electromyographic monitoring (Fisch and Esslen, 1972a). On the contrary, Jannetta advocated separation of the aberrant vessels from the nerves through a posterior fossa craniotomy (Jannetta et al, 1977; Møller and Jannetta, 1987).

Blepharospasm

Blepharospasm is an idiopathic progressive involuntary spasm of the orbicularis oculi and upper face (corrugator and procerus muscles). Extension of the spasms to include the lower face is not uncommon (Jordan et al, 1989). In advanced cases the eyes may be closed for as much as one third of the individual's waking time, rendering the patient functionally blind (Podvinec, 1984). Blepharospasm is generally considered to be central in origin, although the exact mechanism is not yet determined (Podvinec, (1984).

Treatment is directed toward selective destruction of the peripheral nerve branches innervating the orbicularis oculi muscles. McCabe (1972) recommends exposing the upper branches of the facial nerve, confirmation with nerve stimulation, and resection of all branches to the orbicularis oculi.

Success has been obtained using botulinus toxin. Botulinus A toxin, a neurotoxin produced by *Clostridium botulinum* that interferes with the presynaptic release of acetylcholine, has been used in the treatment of many facial dystonias, especially blepharospasm. Over 90% of patients have experienced some relief of symptoms, although

the average duration of the maximum improvement is only 11 to 14 weeks (Jankovic et al, 1990; Ruusuvaara and Setala, 1990). Systemic complications are usually transient and include lid ptosis, facial weakness, corneal exposure, and diplopia (Kalra and Magoon, 1990). Long-term improvement is prevented by regeneration of axons after treatment (Holds et al, 1990). Periorbital myectomy may also produce acceptable long-term results (Jordan et al, 1989).