

TITLE: Bell's Palsy

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Bell's palsy is the most common diagnosis given to patients with acute facial palsy. Despite substantial effort to study its disease process, the management of Bell's palsy remains controversial. This manuscript serves not as a treatment protocol for Bell's palsy, but a review of literature on the subject.

Bell's palsy was named after Sir Charles Bell (1774-1842). His most notable contribution is arguably his description of the facial nerve as the "respiratory nerve of the face" that controls our facial expression. The motor nucleus of the facial nerve lies deep within the reticular formation of the pons where it receives input from the precentral gyrus of the motor cortex, which innervates the ipsilateral and contralateral forehead. The cerebral cortical tracts also innervate the contralateral portion of the remaining face. This accounts for the sparing of the forehead motion in supranuclear lesions of the facial nerve.

The parasympathetic secretory fibers of the nervous intermedius arise from the superior salivatory nucleus. These preganglionic fibers travel to the submandibular ganglion via the chorda tympani nerve to innervate the submandibular and sublingual glands, and to the sphenopalatine ganglion via the greater superficial petrosal nerve to innervate the lacrimal, nasal, and palatine glands. The secretory fibers of the lesser superficial petrosal nerve traverse the tympanic plexus, synapse in the otic ganglion, and travel via the auriculotemporal nerve to innervate the parotid gland. Taste fibers from the anterior 2/3 of the tongue reach the geniculate ganglion via the chorda tympani nerve and from there travel to the nucleus of the tractus solitarius.

The facial nerve and nervus intermedius exit the brain stem at the pontomedullary junction and travel laterally 12 - 14 mm with cranial nerve VIII to enter the internal acoustic meatus. The meatal segment of the nerve then travels 8 - 10 mm within the internal auditory canal (anterosuperior quadrant) to the meatal foramen where the canal narrows from 1.2 mm in diameter to 0.68 mm in diameter (the narrowest part of the canal). The labyrinthine segment then runs 2 - 4 mm to the geniculate ganglion. Here the greater superficial petrosal nerve exits to carry parasympathetic secretomotor fibers to the lacrimal gland. Just distal to this branch, the lesser superficial petrosal nerve exits to supply parasympathetic secretomotor fibers to the parotid. The

tympanic segment begins just distal to the geniculate ganglion where the nerve turns 40 to 80 degrees (first genu) and runs posteroinferiorly 11 mm across the tympanic cavity to the second genu. A branch leaves the segment near the pyramidal eminence to supply the stapedius muscle. The nerve then turns about 90 degrees at the second genu inferiorly where the mastoid segment travels for 12 - 14 mm inferiorly in the anterior mastoid to exit the stylomastoid foramen. The terminal branch of the nervus intermedius, the chorda tympani, leaves the mastoid segment 5 mm proximal to the foramen and travels lateral to the incus, medial to the malleus to exit at the petrotympanic fissure. The extratympanic segment is composed entirely of motor fibers and enters the parotid gland after giving off the posterior auricular branch and a branch to the posterior belly of the digastric muscle. The pes anserus forms 20 mm from the stylomastoid foramen and further divides the nerve into the upper (temporal, and zygomatic) and lower (buccal, mandibular, and cervical) branches.

Although Bell's palsy is a diagnosis of exclusion, it is the most common diagnosis given for acute facial palsy (> 60%). It causes peripheral facial neuropathy that tends to be unilateral and has a rapid onset. Its incidence is about 30 per 100,000. There is an equal male to female ratio and a 3.3 times greater incidence in pregnant females. The left and right sides of the face are equally involved, and less than 1% of cases are bilateral. The recurrence rate is about 10% and can be ipsilateral or bilateral. Patients with diabetes have 4 - 5 times more risk of developing the disease. A family history is positive in about 10% of patients with Bell's palsy.

In 1982, Peitersen et al. published an article on the natural history of Bell's palsy based on more than a thousand Danish patients. This study is unique in that it evaluates the spontaneous course of the disease without medical or surgical intervention. He found that Bell's palsy occurred in every decade of life, with a mean age of between 40 and 44 years. It was less common before the age of 15 and after the age of 60 years. Total unilateral facial paralysis occurred in 71%. Common symptoms included reduced stapedial reflex, postauricular pain, dysgeusia, reduced lacrimation, and phonophobia. The prognosis for Bell's palsy is generally good with 85 % of patients recovering completely within one month. The remaining 15% progress to complete degeneration and will not usually show signs of recovery for three to six months. The longer the time needed for recovery, the greater the probability of sequelae. Patients with incomplete paralysis will recover with no sequelae 95% of the time. Based on this study, poor outcome of Bell's palsy is associated with advanced age, late return of muscular function or beginning of remission, complete palsy, abnormal taste, stapedial reflex, and lacrimation.

Although Bell's palsy has been described as idiopathic facial paralysis, there are increasing pieces of evidence suggesting a viral etiology. A study by Murakami et al. strongly suggests that herpes simplex virus type 1 (HSV-1) is active in idiopathic facial paralysis. DNA fragments of HSV-1 were exclusively found in the perineural fluid of Bell's palsy patients who underwent surgical decompression. An Iowa group has also identified HSV-1 DNA in a temporal bone section of a patient dying 6 days after developing Bell's palsy. These two independent pieces of evidence strongly support the concept that the facial paralysis associated with Bell's palsy is the result of a viral inflammatory response that induces edema within the facial nerve.

The first step in evaluating any patient who presents with facial nerve paralysis involves taking a careful and thorough history. It is important to determine the onset of the paralysis (sudden vs delayed), the duration, and the rate of progression. It is especially important to

determine whether the paralysis is complete versus incomplete. Patients should be questioned regarding previous episodes, family history, associated symptoms (hearing loss, otorrhea, otalgia, vertigo, headaches, blurred vision, parasthesias), associated medical illnesses (diabetes, pregnancy, autoimmune disorders, cancer), history of trauma (recent or remote), and previous surgery (otologic, rhytidectomy, parotidectomy).

A complete head and neck examination must be performed, including microscopic examination of the ears, careful palpation of the parotid glands and neck, ophthalmologic examination (r/o papilledema), auscultation of the neck (r/o carotid bruits), and a thorough neurological examination. It is important to assess the degree of voluntary movement present in order to document the grade of facial paralysis as described in the House classification system:

Grade	Degree	Description
I	Normal	Normal facial movements; No synkinesis
II	Slight	Mild deformity, mild synkinesis, good forehead function, slight asymmetry
III	Moderate	Obvious facial weakness, forehead motion present, good eye closure, asymmetry, Bell's phenomenon present
IV	Moderately	Obvious weakness, increasing synkinesis; no forehead motion
V	Severe	Very obvious facial paralysis, some tone present, cannot close eye
VI	Total	Complete facial paralysis, absent tone

Any patient presenting with facial paralysis should undergo formal audiological testing, including pure tone, air and bone conduction, speech discrimination, reflexes, and tympanometry. If asymmetry is found on the audiogram, an ABR and/or MRI should be obtained.

The most likely site of lesion in Bell's palsy is the meatal foramen (junction of the internal auditory canal portion of the nerve and the labyrinthine segment of the nerve), which is considered to be the narrowest portion of the fallopian canal. MRI with gadolinium will usually show enhancement of the labyrinthine portion of the nerve. As the edema within the nerve increases, axonal flow and circulation are inhibited resulting in varying degrees of nerve injury (first, second, and third degree). Patients who are most severely affected develop a high level of third degree injury that can result in the loss of endoneural tubules and misdirected axonal regeneration. Histological studies from patients with Bell's palsy who died of nonrelated causes reveal diffuse demyelination of the facial nerve with lymphocytic infiltrates.

The principle behind topognostic testing is that lesions distal to the site of a particular branch of the facial nerve will spare the function of that branch. Moving distally from the brainstem, these tests include: the schirmer test for lacrimation (GSPN), the stapedial reflex test (stapedial branch), taste testing (chorda tympani nerve), salivary flow rates and pH (chorda tympani).

Although these tests are of historical interest, they have not been found to be of much use clinically for determining the site of the lesion in facial paralysis or for predicting the outcome. Marked discrepancies are often seen. For example, patients may exhibit a marked decrease in lacrimation with a normal stapedial reflex and intact taste, or they may have absent lacrimation and an absent stapedial reflex with normal salivation. These discrepancies are easily explained in

Bell's palsy, where there can be multiple sites of inflammation and demyelination from the brainstem to the peripheral branches of the nerve.

Electrical tests are useful for patients with complete paralysis for determining prognosis for return of facial function and the endpoint of degeneration by serial testing. They are most useful when considering decompression surgery and are of no value in patients with incomplete paralysis.

The nerve excitability test (NET), maximal stimulation test (MST), and electroneuronography (ENoG) are most useful in the degenerative phase. These tests will give normal results during the first 72 hours after injury due to the stimulating and recording electrodes both being distal to the site of the injury. After 3 - 4 days, the nerve degeneration reaches the site of stimulation and useful results will be obtained. These tests can only be used for unilateral paralysis because all three involve comparison to the contralateral side that must be normal for valid results.

ENoG and electromyography (EMG) are employed more often than NET and MST as the latter two modalities rely on subjective evaluation of muscular response, whereas the former two quantitatively measure compound muscle action potential.

For ENoG, the facial nerve is stimulated with an impulse transcutaneously at the stylomastoid foramen using bipolar electrodes. The muscular response is then recorded using bipolar electrodes placed near the nasolabial groove. The amplitude of the evoked compound action potential is considered proportional to the number of intact axons. The two sides are then compared with the response on the paralyzed side of the face expressed as a percentage of the response on the normal side of the face. A reduction in amplitude on the involved side to 10% or less of the normal side indicates a poor prognosis for spontaneous recovery. Fisch et al. notes that a maximal reduction of less than 90% within 3 weeks of onset gives an expected spontaneous rate of recovery of 80 - 100%. Disadvantages of ENoG include discomfort, cost, and test-retest variability owing to positioning of the electrodes and excitation of the muscles of mastication (V).

EMG is of limited value early in the evaluation of facial paralysis because fibrillation potentials indicating axonal degeneration do not appear until 10 to 14 days post onset. However, EMG becomes important for assessing reinnervation potential of the muscle two weeks after onset. By using needle electrodes placed transcutaneously into the muscles of facial expression, muscle action potentials generated by voluntary activity can be recorded. Electrical silence can indicate normal muscle in a resting state, severe muscle wasting and fibrosis or acute facial paralysis in the early stages. During normal voluntary contraction organized diphasic or triphasic potentials are seen. Fibrillation potentials indicate degeneration of the neural supply to the muscle in question. Polyphasic potentials indicate reinnervation. These are important because they usually appear 6 - 12 weeks before clinical return of function. It is generally obtained if ENoG displays more than 95% degeneration.

Sunderland's classification describes five degrees of nerve injury. The first degree (neuropraxia) involves a localized conduction block in the nerve with the nerve fibers responding

to electrical stimuli proximal and distal to the lesion, but not across the injured segment. Axonal continuity is preserved, Wallerian degeneration does not occur, and recovery is usually complete.

The second degree of nerve injury is called axonotmesis. This refers to disruption of the axon into proximal and distal portions with interrupted axoplasmic flow and Wallerian degeneration. The third, fourth, and fifth degree of nerve injury are grouped together as neurotmesis, and subdivided depending on the integrity of perineurium and epineurium. Wallerian degeneration occurs at the faster rate than axonotmesis and prognosis is the poorest. The rate of nerve degeneration in Bell's palsy falls in between axonotmesis and neurotmesis. However, there is no electrical test to-date that can quantitatively differentiate the two subclasses of injury.

Treatment options of Bell's palsy range from observation, medical treatment, surgical decompression, to facial rehabilitation. The efficacies of oral prednisone and anti-viral agents have been studied extensively, yet there is no consensus among experts on ideal regimen and dosage. Cochrane review summarizes four trials (179 patients) that compare steroid to placebo, saline, and vitamin solutions. Its results show no significant benefit of giving steroid to Bell's palsy patients. A similar conclusion is reached by Turk Boru's randomized controlled trial (56 patients). Ramsey, however, points out in his meta-analysis that the rate of complete recovery (Grade VI \rightarrow I) is 17% higher in the steroid group, provided the total prednisone dose is at least 400 mg and that the treatment is initiated within a week of onset of paralysis (3 trials, 230 patients). The addition of anti-viral agent such as acyclovir is advocated by Adour, whose double-blind randomized controlled trial shows that the combined therapy of prednisone and acyclovir results in better return of muscle function and prevention of partial nerve degeneration. This result is challenged by another prospective trial by Kawaguchi who illustrates that valacyclovir plus prednisone have no advantage over prednisone alone. Regardless of medical regimen, early administration within 3 days of onset appear to have superior effect on outcome, as demonstrated in a study by Hato et al.

Eye care is of utmost importance in facial nerve paralysis due to the risk of exposure keratitis. Artificial tears and lacrilube ointment should be prescribed. Taping of the eye lids during sleep may be helpful as well as the use of a moisture chamber. Patients should avoid contact lens, fans and dust, and should have eye protection when outside in the wind. Gold weight implant to the upper eyelid should be considered in patients with long-standing facial paralysis.

Surgical decompression for Bell's palsy has evolved in many ways throughout the years. The emphasis has shifted from focusing on the stylomastoid foramen in the 1930s to decompressing the meatal foramen nowadays, the narrowest portion of the facial canal. Proponents of surgical decompression such as Gantz believe in early decompression within 2 weeks of onset of paralysis if ENoG demonstrates more than 90% degeneration and EMG shows no voluntary muscle potential. Those who are against decompression argue that the Bell's palsy is a result of viral demyelination and that increased intracanal pressure may not be the primary cause of Bell's palsy. Although the issue of surgical decompression remains highly controversial, factors such as age, medical comorbidities, endpoint and rate of progression of ENoG, days from onset of paralysis, and when muscle function begins to return likely influence the outcome and allow us to predict the success of surgery.

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