REVIEW

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When is facial paralysis Bell palsy? Current diagnosis and treatment

ABSTRACT

Bell palsy is largely a diagnosis of exclusion, but certain features in the history and physical examination help distinguish it from facial paralysis due to other conditions: eg, abrupt onset with complete, unilateral facial weakness at 24 to 72 hours, and, on the affected side, numbness or pain around the ear, a reduction in taste, and hypersensitivity to sounds. Corticosteroids and antivirals given within 10 days of onset have been shown to help. But Bell palsy resolves spontaneously without treatment in most patients within 6 months.

KEY POINTS

As many as two thirds of patients with Bell palsy spontaneously recover and achieve near-normal function. Many patients show signs of improvement as early as 10 days after the onset, even without treatment.

No readily identifiable cause for Bell palsy has been found, but clinical and experimental evidence suggests herpes simplex type 1 infection may play a role.

As Bell palsy resolves, autonomic synkinesis—involuntary movement accompanying a voluntary one—can develop. Corticosteroid therapy may reduce the chance of this occurring and may also improve the rate of recovery in adults.

Adding acyclovir to prednisone therapy may improve recovery rates compared with prednisone alone.

WHEN SHOULD WE SUSPECT Bell palsy in patients with facial paresis or paralysis? Which clinical signs and tests help us narrow the differential diagnosis?

Bell palsy is a lower motor neuron disease of the facial nerve (cranial nerve VII) characterized by acute unilateral peripheral facial weakness involving muscles innervated by the facial nerve. Bilateral weakness occurs in fewer than 1% of patients.

In this article, I present a brief overview of a systematic approach to diagnosis and treatment of this condition.

INCIDENCE AND RISK

The annual incidence of Bell palsy is about 20 per 100,000, and the incidence increases with age. It is seen as often in men as in women. Those at high risk include pregnant women and people with diabetes mellitus.[1] About 10% of those with Bell palsy have a family history of the condition.

PATHOPHYSIOLOGY

The left and right facial nerves control a number of functions, such as blinking and closing the eyes, smiling, frowning, lacrimation, and salivation. They also innervate the stapedial (stapes) muscles of the middle ears and carry taste sensations from the anterior two thirds of the tongue.

Each facial nerve travels through a narrow bony canal in the skull beneath the ear to the muscles on each side of the face. Acute inflammation and edema of the facial nerve are thought to lead to entrapment of the nerve in the bony canal, which leads to compression ischemia. An

TABLE 1

Facial nerve grading system

GRADE	DEGREE OF DYSFUNCTION	MEASUREMENT*	PERCENT OF FUNCTION	ESTIMATED PERCENT OF FUNCTION
I	—	8/8	100	100
II	Slight	7/8	76–99	80
III	Moderate	5/8–6/8	51–75	60
IV	Moderately severe	3/8–4/8	26–50	40
V	Severe	1/8–2/8	1–25	20
VI	Total paralysis	0/8	0	0

*A centimeter is divided into four equal parts. On the affected side of the face, maximal voluntary lateral movement of the corner of the mouth is measured 0-4, and elevation of the eyebrow is measured 0-4, resulting in a sum from 0/8 to 8/8

REPRINTED FROM HOUSE JW, BRACKMANN DE. FACIAL NERVE GRADING SYSTEM. OTOLARYNGOL HEAD NECK SURG 1985; 93:146–147, WITH PERMISSION FROM THE AMERICAN ACADEMY OF OTOLARYNGOLOGY—HEAD AND NECK SURGERY FOUNDATION, INC.

observation that supports the hypothesis of an inflammatory etiology is that the facial nerve shows enhancement on magnetic resonance imaging (MRI) in patients with acute Bell palsy.[2]

Evidence that herpes simplex virus plays a role

Although no readily identifiable cause for Bell palsy has been found, accumulating clinical and experimental evidence suggests that herpes simplex type 1 infection may play a role: polymerase chain reaction assays have identified herpes simplex virus in the endoneurial fluid, posterior auricular muscle, and saliva in patients with Bell palsy.[3,4]

CLINICAL FEATURES

Clinical characteristics of Bell palsy include:

- Peripheral dysfunction of the facial nerve, involving all distal branches
- Abrupt onset (over hours), with maximal facial weakness at 24 to 72 hours
- Unilateral facial weakness that is complete in most patients, but may be partial in as many as one third of patients
- Numbness or pain around the ear on the affected side
- A reduction in taste on the affected side; altered taste on the anterior two thirds of tongue
- Hypersensitivity to sounds (hyperacusis) on the affected side, found in one third of patients
- Spontaneous improvement within 6 months in most cases[5]
- Incomplete healing in 15% of patients, resulting in residual nerve dysfunction, including partial palsy and motor synkinesis (involuntary movement accompanying a voluntary one).
- No history of trauma, local infection, tumor, or central nervous system disease.

The cause of Bell palsy is not known, but recent work points to herpes simplex virus

PHYSICAL EXAMINATION

A focused physical examination can help rule out other conditions and narrow the differential diagnosis of Bell palsy. Things to observe:

How many branches are involved? In Bell palsy, all branches of the facial nerve are diffusely affected. Involvement of only one or two branches suggests trauma or a parotid gland tumor.

Are there signs of skin rash, trauma, or systemic illness? Check for lesions on the skin of the ear and inside the ear: vesicular lesions due to herpes zoster virus are usually seen in the external ear or ear canal and sometimes on the soft palate. Check for middle ear infection, as chronic middle ear infection has been associated with Bell palsy.

Is Bell phenomenon present? Check for the Bell phenomenon: ie, in a patient with Bell palsy who attempts to close his or her eyes, the normal upward movement of the eye can be noted on the affected side, as the eyelid remains open.

Check for abnormal facial sensation. If facial paresis is noted, the House-Brackmann facial nerve grading system[6] (TABLE 1) can be used to quantify the damage.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of Bell palsy is broad. While Bell palsy is a common cause of facial nerve paralysis, it is not synonymous with it. Bell palsy is a diagnosis of exclusion. The following are some of the conditions that should be ruled out.

Unilateral central facial weakness (lower face muscles) may be due to a lesion of the contralateral cortex, subcortical white matter, or internal capsule. In addition to facial weakness, symptoms may include hemiparesis, hemisensory loss, or hemineglect (severe impairment of spatial perception).

Ramsay Hunt syndrome is acute peripheral facial paralysis due to reactivation of varicella-zoster (chickenpox) virus and its spread to the facial nerves. Vesicular lesions are usually seen in the external ear or ear canal and sometimes on the soft palate.

Diagnosis depends on clinical signs, symptoms, and exclusion of other causes of facial paralysis

Lyme neuroborreliosis. The spirochete *Borrelia burgdorferi* can affect central nervous system tissues. Lyme neuroborreliosis should be suspected in a patient who presents with isolated facial weakness and who has a history of tick bite with rash or who lives in an area where Lyme disease is endemic.

Tumors involving the facial nerve account for fewer than 5% of all cases of facial nerve paralysis. A tumor should be suspected if weakness progress over weeks, if a mass is present in the ear, neck, or parotid gland, and if no functional improvement is seen within 4 to 6 weeks.

Diabetes mellitus and sarcoidosis can cause facial neuropathy with sudden weakness. A history of diabetes and respiratory symptoms in sarcoidosis provide clues to this diagnosis.

Weight loss suggests a systemic illness such as neoplasm, metastasis, or infection.

Visual changes, vertigo, and weakness or numbness may suggest a brain stem lesion such as demyelination.

DIAGNOSIS

The history and physical examination provide the information key to the diagnosis of Bell palsy. Most patients do not require any laboratory testing or imaging studies. However, patients who have persistent weakness without significant improvement, involvement of other cranial nerves, or a second episode of palsy require further investigations.

Imaging

Computed tomography (CT) or MRI is indicated in the following cases:

- No improvement in facial paresis after 1 month
- Hearing loss
- Multiple cranial nerve deficits
- Signs of limb paresis or sensory loss.

MRI with gadolinium is the test of choice to rule out cerebellopontine angle tumor, stroke, multiple sclerosis, or other structural lesions. CT is recommended if a temporal bone fracture is suspected.

Hearing testing

If hearing loss is suspected, then audiologic testing can be performed to measure hearing loss and to help to rule out acoustic neuroma.

Laboratory testing

Laboratory testing is necessary if the patient has signs of systemic involvement, such as fever, weight loss, rash, or progressive facial weakness without significant improvement over more than 4 weeks. A number of tests may be helpful:

Complete blood count with differential helps rule out lymphoreticular malignancy, the first manifestation of which may be peripheral facial palsy.[7]

Blood glucose should be measured if diabetes mellitus is suspected.

Serum antibodies against herpes zoster and *B burgdorferi* (the agent of Lyme disease) can be checked if the patient has signs such as vesicular lesions on the external ear or lives in an area where Lyme disease is endemic.

Serum calcium and angiotensin-converting enzyme levels should be tested if sarcoidosis is suspected; these levels are high in sarcoidosis.

Cerebrospinal fluid testing is helpful if infection or malignancy is suspected; however, cerebrospinal fluid taken from patients with Bell palsy tends to show mild and inconsistently elevated cell counts and protein levels, but is otherwise not helpful in identifying the cause.

Electrodiagnostic testing is not routinely done in Bell palsy. It is not very reliable when Bell palsy is in the initial stages; however, after 2 weeks, it may detect denervation and demonstrate nerve regeneration.

TREATMENT IS STILL CONTROVERSIAL

Treatment of Bell palsy is still controversial. Therapy is difficult to evaluate, because as many as two thirds of patients with Bell palsy spontaneously recover and achieve near-normal function. Many patients begin to improve as early as 10 days after the onset, even without treatment.

Older patients are less likely to recover completely. In series of 250 patients at an ear, nose, and throat clinic in Greece, the percentage of patients who recovered completely varied from 74% to 83% in patients ages 4 to 50, whereas the percentage was less than 54% at age 80.[8]

Protect the eyes

To prevent exposure keratitis, artificial tears should be used frequently during the day in the affected eye to keep it moist. Sunglasses should be worn outside. Areas with air contaminated by excessive particulate matter or noxious fumes (construction areas, textile factories) should be avoided. To protect the cornea during sleep, an ophthalmic ointment should be used along with an eye patch. Any corneal abrasion or infection should be treated immediately to avoid possible visual complications. Ophthalmologic consultation is recommended if any deterioration in visual function is noted.

Drug therapy

Drug therapy mainly consists of corticosteroids with or without an antiviral (acyclovir). These drugs hasten the recovery and lessen the ultimate degree of dysfunction.[9–11]

Corticosteroids alone. A study of 239 patients showed improved rates of synkinesis (involuntary movement accompanying a voluntary one) after treatment with prednisone 60 mg/day for 10 days and then tapered over 1 week, compared with placebo.[12]

Corticosteroids plus acyclovir. Adour and colleagues[11] found that 92% of patients regained normal facial motion when given a 10-day course of prednisone (60 mg a day orally for 5 days, tapered by 10 mg a day for 5 days) and acyclovir (400 mg by mouth five times daily). The outcome of patients treated with prednisone plus acyclovir was superior to the outcome of those treated with prednisone plus placebo, and prednisone plus acyclovir was "statistically more effective in producing return of volitional muscle motion to a recovery profile of 10" (92% vs 76%, $P = .02$).[11]

Corticosteroids or acyclovir. A study comparing prednisone vs acyclovir found that patients treated with prednisone had better complete recovery rates, 93.6% vs 77.7%.[13]

Further study needed

The results of these studies are difficult to interpret. A larger multicenter double-blind placebo-controlled trial in the future may shed more light on the efficacy of steroids and antiviral therapy.

Recommendations

Drug treatment options for Bell palsy are limited to steroids and acyclovir

Corticosteroid therapy reduces autonomic synkinesis and possibly improves the rate of recovery in adults. Adding acyclovir to prednisone therapy may improve recovery rates compared with prednisone alone.

A practice parameter from the American Academy of Neurology states that corticosteroids are safe and probably effective, and that acyclovir is safe and possibly effective.[14] Early treatment (ie, within 3 days after the onset of Bell palsy) is necessary for acyclovir-prednisone therapy to be effective.[15] However, drug therapy may also be effective if started within 10 days of symptom onset. If the patient presents 10 days after the onset of symptoms, no drug treatment is necessary.

A patient who presents within 1 week of the onset of facial weakness and is not diabetic or pregnant and has no signs of infection should receive prednisone (60 to 80 mg/day for 10 days and then a slow taper) plus acyclovir (400 mg five times per day for 10 days). Oral valacyclovir, which is converted in the body to acyclovir, has greater bioavailability than oral

acyclovir and yields similar acyclovir plasma concentrations with only twice-daily dosing.[16]

Surgical treatment

We do not have enough evidence to recommend surgical decompression of the facial nerve to hasten the recovery in patients with Bell palsy. Surgical decompression for indications other than trauma is controversial.[10,17] However, some surgeons still offer middle fossa decompression if electromyography shows a 90% reduction in compound muscle action potential in patients with facial weakness lasting less than 3 weeks.[18]

For a patient with permanent facial paralysis despite medical and surgical treatment, many surgical options are available to improve facial function and appearance. These include static sling procedures with facia lata or alloplastic strips; dynamic procedures with transposition of the temporalis or masseter muscle; hypoglossal-facial nerve anastomosis; cross-facial nerve grafting; free muscle grafting; and microvascular free nerve muscle grafting.

Electrotherapy

The use of electrotherapy in the treatment of Bell palsy remains controversial.

Facial exercise

Facial exercises can be beneficial in patients with Bell palsy. They should be performed while standing in front of a mirror and include trying to raise the eyebrows, opening and closing the eyes, blowing, and whistling. These exercises can be performed a few times daily.[19,20] The efficacy of exercise has not been formally evaluated.

Botulinum toxin injections

Motor synkinesis can occur during recovery from Bell palsy, due to aberrant regeneration of the facial nerve. It can be grossly deforming. Injection of botulinum toxin into the involved muscles is an effective treatment.

Hemifacial spasm occasionally appears after Bell palsy, and blepharospasm can be seen in rare cases.[21] Both can be treated with injections of botulinum toxin. The time from the resolution of Bell palsy to the subsequent development of blepharospasm ranges from a few weeks to 24 years.[21]

Hyperlacrimation secondary to aberrant regeneration of the seventh nerve has also been reported after Bell palsy. Botulinum toxin injection has been used in these patients

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