Clinical review

Recent developments in Bell's palsy

N Julian Holland, Graeme M Weiner

General practitioners in the United Kingdom will see about one patient with Bell's palsy every two years. Increasing evidence shows that the way the patient is managed has an important effect on outcome. Untreated Bell's palsy leaves some patients with major facial dysfunction and a reduced quality of life. Of patients with Bell's palsy registered by general practitioners between 1992 and 1996 a fifth were referred for specialist opinion, just over a third received oral steroids, and 0.6% received aciclovir.[1] Improving outcomes requires coordination between specialists and general practitioners so that patients are treated during the critical first 72 hours. We outline recent developments in Bell's palsy and current best evidence in its management.

Recent developments

Bell's palsy is probably caused by herpes viruses, mainly herpes simplex virus type 1 and herpes zoster virus

Facial palsy improves after treatment with combined oral aciclovir and prednisone

Treatment of partial Bell's palsy is controversial; a few patients don't recover if left untreated

Treatment is probably more effective before 72 hours and less effective after seven days

A fifth of cases of acute facial palsy have an alternative cause that should be managed appropriately

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Sources and selection criteria

We canvassed specialists with an interest in acute facial palsy and incorporated the latest consensus from key publications and systematic reviews. We performed a hierarchical literature search through Medline, CINAHL, SUMSearch, bmj.com, Lancet Neurology Network, Bandolier, Health Technology Assessment, Clinical Evidence, and the Cochrane Library. Both authors are otolaryngologists with an interest in neurotology and facial palsy.

Incidence and pathophysiology

Bell's palsy accounts for almost three quarters of all acute facial palsies, with the highest incidence in the 15 to 45 year old age group (table 1).[2] The annual incidence in the UK population is around 20 per 100 000, with one in 60 people being affected during their lifetime. Men and women are equally affected, although the incidence is higher in pregnant women (45 cases per 100 000).

Table 1 Causes and incidence of acute facial palsy[2]

Causes	Incidence (%)
Bell's palsy	72
Herpes simplex virus type 1	84
Herpes zoster virus:	
Zoster sine herpete	16
Ramsay Hunt syndrome	7
Trauma	4
Tumour	4
Otitis media or cholesteatoma	3
Neonatal conditions	6
Rare and unusual conditions	4

Increasing evidence implies that the main cause of Bell's palsy is latent herpes viruses (herpes simplex virus type 1 and herpes zoster virus), which are reactivated from cranial nerve ganglia. Sensitive polymerase chain reaction techniques have isolated herpes virus DNA from the facial nerve during acute palsy.[3] Inflammation of the nerve initially results in a reversible neurapraxia, but ultimately Wallerian degeneration ensues. Herpes zoster virus shows more aggressive biological behaviour than herpes simplex virus type 1 because it spreads transversely through the nerve by way of satellite cells.

Symptoms

The most alarming symptom of Bell's palsy is paresis; up to three quarters of affected patients think they have had a stroke or have an intracranial tumour. The palsy is often sudden in onset and evolves rapidly, with maximal facial weakness developing within two days. Associated symptoms may be hyperacusis, decreased production of tears, and altered taste (table 2).

Patients may also mention otalgia or aural fullness and facial or retroauricular pain, which is typically mild and may precede the palsy. Severe pain suggests herpes zoster virus and may precede a vesicular

P+ *Further information and description of levels of evidence are on bmj.com*

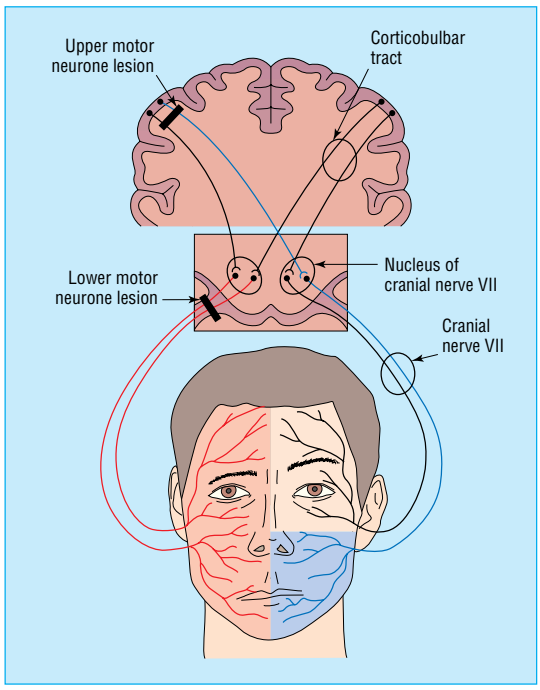

Fig 1 Lesion of right upper motor neurone causes central pattern of facial weakness on left. Lesion of right lower motor neurone causes facial palsy on right. Adapted from Dresner[6]

eruption and progression to Ramsay Hunt syndrome. Features may be consistent with a mild polyneuropathy. A slow onset progressive palsy with other cranial nerve deficits or headache raises the possibility of a neoplasm.

Examination

Bell's palsy causes a peripheral lower motor neurone palsy, which manifests as the unilateral impairment of movement in the facial and platysma muscles, drooping of the brow and corner of the mouth, and impaired closure of the eye and mouth. Bell's phenomenon—upward diversion of the eye on attempted closure of the lid—is seen when eye closure is incomplete.

A central upper motor neurone deficit causes weakness of the lower face only (fig 1). More complex segmental deficits may be caused by peripheral facial nerve lesions. Patients with facial palsy require careful examination of the other cranial nerve and cerebellar function. The modified House-Brackmann facial grading scale allows consistent documentation of facial palsy (see table on bmj.com).[5]

Table 2 Polyneuropathy in Bell's palsy[4]

Symptoms	Incidence (%)
Altered glossopharyngeal or trigeminal nerve sensation	80
Facial or retroauricular pain	60
Altered taste	57
Hyperacusis	30
Reduced sensation C2 dermatome	20
Vagal motor weakness	20
Decreased tearing	17
Trigeminal motor weakness	3

Assessment of the ear should include pneumatic otoscopy and tuning fork tests. Polyposis or granulations in the ear canal may suggest cholesteatoma or malignant otitis externa. Vesicles in the conchal bowl, soft palate, or tongue suggest Ramsay Hunt syndrome (see fig A on bmj.com).

The examination should exclude masses in the head and neck. A deep lobe parotid tumour may only be identified clinically by careful examination of the oropharynx and ipsilateral tonsil to rule out asymmetry (fig 2). Erythema migrans on the limbs or trunk with a history of tick bite implies Lyme disease, which may cause facial palsy.[7]

Investigations

Serum testing for rising antibody titres to herpes virus is not a reliable diagnostic tool for Bell's palsy. Salivary polymerase chain reaction for herpes simplex virus type 1 or herpes zoster virus is more likely to confirm virus during the replicating phase, but these tests remain research tools. Serological tests for Lyme disease (IgM, IgG) are essential to exclude this disease in endemic areas, and magnetic resonance imaging has revolutionised the detection of tumours. Typically, the hearing threshold is not affected in Bell's palsy, but stapedius reflexes may be reduced or absent. Topognostic tests and electroneurography may give useful prognostic information but remain research tools.[8]

Zoster sine herpete

Herpes zoster virus has traditionally been associated with Ramsay Hunt syndrome, with typical cutaneous vesicles and cochleovestibular dysfunction. Vesiculation may not necessarily appear (zoster sine herpete) or may be delayed in up to half of patients. Dermatomal pain and dysaesthesia before vesiculation is termed preherpetic neuralgia and may be the only clinical indicator that herpes zoster virus is involved. Zoster sine herpete is thought to be the cause of almost a third of facial palsies previously diagnosed as idiopathic.[9]

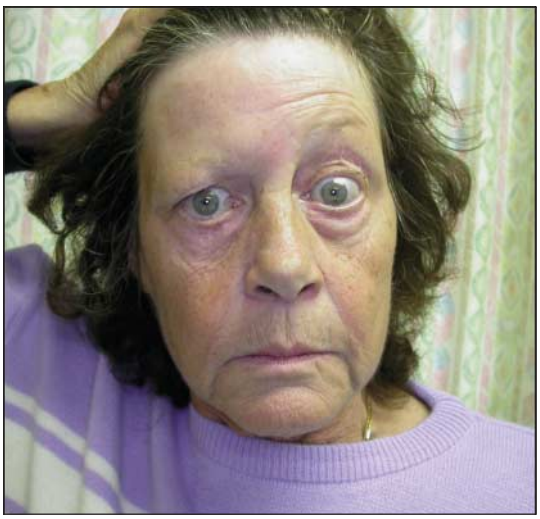

Fig 2 Woman with segmental facial palsy showing reduced elevation of right eyebrow and closure of right eye. A malignant mass was found in the right parotid gland. Reproduced with patient's permission

Box 1: Indicators of poor prognosis in Bell's palsy

- Complete facial palsy
- No recovery by three weeks
- Age over 60 years
- Severe pain
- Ramsay Hunt syndrome (herpes zoster virus)
- Associated conditions—hypertension, diabetes, pregnancy
- Severe degeneration of the facial nerve shown by electrophysiological testing

Bell's palsy in children

Bell's palsy is a much less common cause of facial palsy in children under 10 years of age. These children therefore merit careful review to identify an alternative cause, including acute suppurative ear disease. Lyme disease may be responsible for as many as half the cases in endemic areas.

Outcomes

Overall, Bell's palsy has a fair prognosis without treatment, with almost three quarters of patients recovering normal mimetical function and just over a tenth having minor sequelae. A sixth of patients are left with either moderate to severe weakness, contracture, hemifacial spasm, or synkinesis. Patients with a partial palsy fair better, with 94% making a full recovery. The outcome is worse when herpes zoster virus infection is involved in partial palsy. In patients who recover without treatment, major improvement occurs within three weeks in most. If recovery does not occur within this time, then it is unlikely to be seen until four to six months, when nerve regrowth and reinnervation have occurred. By six months it is clear who will have moderate to severe sequelae. Box 1 lists the poor prognostic indicators of Bell's palsy.

In facial palsies caused by herpes simplex virus or herpes zoster virus there remains a strong correlation between the peak severity of the palsy and the outcome. As yet there is no reliable investigation or test at presentation that can indicate who will make a full recovery.

Treatment

The main aims of treatment in the acute phase of Bell's palsy are to speed recovery and to prevent corneal complications. Treatment should begin immediately to inhibit viral replication and the effect on subsequent pathophysiological processes that affect the facial nerve. Psychological support is also essential, and for this reason patients may require regular follow up.

Eye care

Eye care of patients with Bell's palsy focuses on protecting the cornea from drying and abrasion due to problems with lid closure and the tearing mechanism. The patient is educated to report new findings such as pain, discharge, or change in vision. Lubricating drops should be applied hourly during the day and a simple eye ointment should be used at night.

Corticosteroids

Two recent systematic reviews concluded that Bell's palsy could be effectively treated with corticosteroids in the first seven days, providing up to a further 17% of patients with a good outcome in addition to the 80% that spontaneously improve (see also fig B on bmj.com).[10 11] Other studies have shown the benefits of treatment with steroids; in one, patients with severe facial palsy showed a significant improvement after treatment within 24 hours.[12 13] Recovery rates in patients treated within 72 hours were enhanced by the addition of aciclovir.[14]

A randomised controlled trial of patients treated with high dose parenteral steroids within 72 hours compared with placebo found a significant improvement in recovery rate and time to return to work but no statistical difference in final outcome.[15] More randomised controlled trials are needed, but at least 200 patients would be required in each arm.[16 17]

Given the existing evidence (see bmj.com for description of grades (A) to (D)), we support the use of oral prednisone with aciclovir in patients presenting with moderate to severe facial palsy, ideally within 72 hours. Immunocompetent patients without specific contraindications are prescribed prednisone at 1 mg/kg/d (maximum 80 mg) for the first week, which is tapered over the second week.(B) Around a fifth of patients will progress from partial palsy, so these patients should also be treated.[11](C)

Antiviral agents

Treatment with antivirals seems logical in Bell's palsy because of the probable involvement of herpes viruses. Aciclovir, a nucleotide analogue, interferes with herpes virus DNA polymerase and inhibits DNA replication. Because of aciclovir's relatively poor bioavailability (15% to 30%),[18] newer drugs in its class are being trialled. Better bioavailability, dosing regimens, and clinical effectiveness in treating shingles have been shown with valaciclovir (prodrug of aciclovir), famciclovir (prodrug of penciclovir), and sorivudine.[19]

Box 2: Evolving treatments for Bell's palsy

Some evidence of effect

Methylcobalamin—an active form of vitamin B-12

Hyperbaric oxygen—may be useful in patients who show degeneration despite maximal therapy

Facial retraining—"mime therapy"

Botulinum toxin for synkinesis and hemifacial spasm

Uncertain effect

Transcutaneous electrical stimulation

Acupuncture

Current research

Multicentre, randomised, double blind, placebo controlled trials on steroid and antiviral therapy are being carried out in Sweden and France

New antivirals—for example, famciclovir, sorivudine

Vaccination against herpes zoster virus and herpes simplex virus types 1 and 2

Neurotrophic growth factors, neuroprotective agents—for example, nimodipine, glial cell derived neurotrophic factor

Additional educational resources

Book

Pensak ML. *Controversies in otolaryngology*. New York: Thieme, 2001:218-31—three chapters presenting current perspectives on acute facial palsy

Key papers

Peitersen E. Bell's palsy: the spontaneous course of 2,500 peripheral facial nerve palsies of different aetiologies. *Acta Otolaryngol Suppl* 2002;549:4-30—key text on the epidemiology and outcomes of untreated Bell's palsy

Morrow MJ. Bell's palsy and herpes zoster oticus. *Curr Treat Options Neurol* 2000;2:407-16—excellent review and evidence based treatment guidelines

Sweeney CJ, Gilden DH. Ramsay Hunt syndrome. *J Neurol, Neurosurg Psychiatry* 2001;71:149—relevant publication from active researchers in this subject

Internet resources

emedicine.com—has several well structured articles on Bell's palsy

Information for patients

The official patient's sourcebook on Bell's palsy: a revised and updated directory for the internet age publisher. San Diego, CA: Icon Health, 2003—several books are available to patients, which tend to present the authors' viewpoints

Patient information. Bell's palsy. *J Fam Pract* Feb 2003;52:160—useful information leaflet

Bell's palsy information site (www.bellspalsy.ws/links.htm)—a structured website with information about acute treatment and rehabilitation

Bell's palsy association (www.bellspalsy.org.uk/links.htm)—UK based site providing information and support for patients

Open directory project (http://dmoz.org/)—access to a huge number of links of variable quality

Aciclovir compared with prednisone

Aciclovir has been compared with prednisone.[20] Prednisone has been shown to be more effective in producing good recovery at three or more months, but despite flaws in this study, we would not recommend using aciclovir (or any antiviral) without steroids unless steroids are contraindicated.[19](B)

Aciclovir with prednisone

A recent systematic review found that patients treated with combined aciclovir and prednisone had a better outcome than those treated with prednisone alone.[10] However, a Cochrane review at that time concluded that more studies were required.[21] More recently, a study of patients with severe palsies found better recovery with combined aciclovir and prednisone than with prednisone alone. The main determinate of the difference was treatment within three days of the onset of palsy.[14]

A prospective case controlled study showed that patients treated with valaciclovir and prednisone (86% within 72 hours) had better recovery rates than patients treated with prednisone alone. A noticeable benefit was seen in elderly patients, a group that is often overlooked for maximal treatment.[22] A study of systemic therapy found no difference between oral aciclovir with prednisone and intravenous aciclovir with prednisone.[23] Systemic treatment should be considered in immunocompromised patients or for widespread zoster involving the central nervous system.

We support the use of oral aciclovir or valaciclovir with prednisone in patients presenting within a first week (ideally within 72 hours) with moderate to severe facial palsy.(B)

Treatment in children

Studies have found that children with complete facial palsies and major degeneration have poor outcomes as often as adults. However, no supportive evidence has been found for use of steroids or antivirals in children with Bell's palsy (see fig C on bmj.com).[24](D)

Zoster sine herpete

Although 2000 mg/d of aciclovir would not be adequate for Ramsay Hunt syndrome with vesicles, it seems to be effective in patients with zoster sine herpete.[14] On the basis of current evidence, in the absence of major pain or evidence of vesicles, this dose would be adequate with steroids for treating Bell's palsy associated with herpes zoster virus.(C)

Future research may indicate that patients with severe post auricular pain, dense palsy, or herpes zoster virus do better with higher dose antiviral therapy from the outset.(D)

Surgery

Surgical intervention decompresses the facial nerve.[25] However, middle fossa craniotomy carries risks, including seizures, deafness, leakage of cerebrospinal fluid, and facial nerve injury. Hence decompression surgery for Bell's palsy is not routinely offered in the United Kingdom.(D)

Physical therapies

Several physical therapies, including massage and facial exercises, are recommended to patients, but there are few controlled clinical trials of their effectiveness.(D) Some recent evidence supports facial retraining (mime therapy) with biofeedback.[26](C)

Follow up

Patients with Bell's palsy should start treatment immediately and be referred to a specialist as soon as possible. In a few cases the diagnosis may be subsequently reassigned.[2] Patients should receive psychological support and eye care during follow up. Long term sequelae may be missed if patients are not monitored for a full year.

A multidisciplinary team approach (general practitioners, otolaryngologists, ophthalmologists, plastic surgeons, physiotherapists, and psychologists) is essential when there is no prospect of further recovery of facial nerve function. Synkinesis and facial spasm, common features of partially recovered deficits, can be effectively managed with subcutaneous or intramuscular injections of botulinum toxin. Facial reanimation may be possible by a combination of static and dynamic surgical techniques and may result in functional as well as cosmetic improvements. Weighting of the upper lid improves eye closure.

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Lesson of the week

Rare causes of haemoptysis in suspected pulmonary embolism

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When a patient presents with haemoptysis and pleuritic chest pain, a pulmonary embolism is an important and common diagnosis to consider. There is a tendency in busy medical admissions units to start treatment of certain conditions without thorough investigation, with the intention of reducing delays in starting treatment. Patients are usually treated for suspected pulmonary embolism with heparin early to reduce mortality and morbidity. It is important, however, to remember other less common causes of haemoptysis.

Case report

A 59 year old woman was admitted to our medical assessment unit with chest pain and haemoptysis. She had experienced pleuritic left sided, chest wall pain intermittently for the previous week, with gradually increasing intensity. On the day of admission she had also produced about a cupful of bright red blood while coughing. She had no medical history of note, except that she was a smoker.

On admission the patient was in distress but not objectively dyspnoeic—her respiratory rate was not raised and her oxygen saturations on air were 97%. She did not show any signs of shock; she had no tachycardia and her systolic blood pressure remained around 120 mm Hg throughout. Chest examination showed some decreased air entry at her left lung base with a loud pleural rub. She had no cardiac murmurs, and no clinical evidence was found of a deep venous thrombosis in her legs or pelvis.

The admitting doctor's differential diagnosis included pulmonary embolism, pneumonia, and carcinoma of the lung; a plain chest radiograph and routine blood tests were ordered including a D-dimer assay.

The chest x ray film (fig 1) showed moderate left basal shadowing with blunting of the right costophrenic angle. There was no cardiomegaly, and, importantly, the mediastinum appeared normal.

Blood tests indicated a mild anaemia (haemoglobin 96 g/l), a raised white cell count (28.5, neutrophils 25.9)

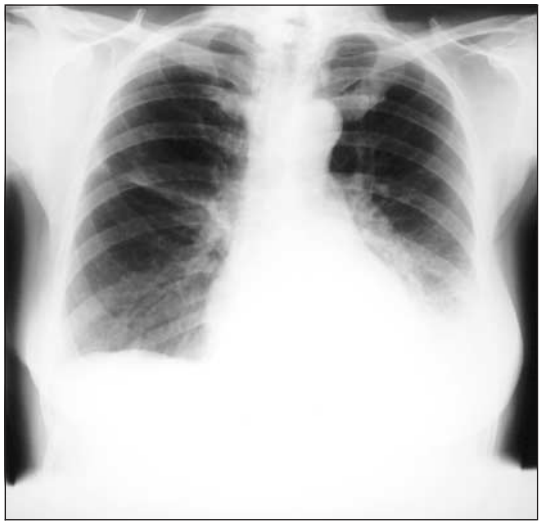

Fig 1 Chest x ray film showing moderate left basal shadowing with blunting of the right costophrenic angle

Do not automatically treat a suspected pulmonary embolism with heparin; consider other diagnoses first

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