BELL'S PALSY: A REVIEW

Bell's palsy can be a frightening condition for the patient, coming on rapidly and resulting in noticeable facial paralysis. However, although most people will make a full recovery, the GP still has an active role in supporting the patient

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CASE STUDY

On a Monday morning, a 45-year-old woman presented to her GP with a left-sided facial weakness. She had difficulty closing her left eye when putting on make-up in preparation for an evening out on Friday; over the weekend the weakness had progressed to involve the whole of the left side of her face. She had been diagnosed with Bell's palsy 10 years previously, so decided not to seek medical attention over the weekend, but to wait until Monday. When seen by her GP, she had a marked left-sided facial paralysis. There was no other limb weakness or focal neurology. A diagnosis of Bell's palsy was made and treatment with prednisolone commenced. When she returned to her GP six weeks later she had made a near, but not complete, recovery. She was pleased with how quickly and her facial paralysis had recovered and not troubled by the small degree of remaining weakness. She was instead more concerned by the cold sore on her upper lip which had become infected, her primary reason for re-attendance.

Bell's palsy is an acute ipsilateral paralysis of the seventh cranial nerve (the facial nerve), characterised by a sudden onset facial weakness that progressively worsens over 48 hours, followed by a gradual recovery, usually over the next three weeks.[1] Bell's palsy affects 11–40 people per 100,000 per year, or 1 in 5,000.[2]

The peak incidence is between the ages of 15 and 50, although younger children can also be affected.[2,3] The incidence is slightly increased in pregnancy (45 per 100,000).[3] The recurrence rate is 12%, with a 36% recurrence rate on the same side.[1] Risk factors include diabetes, hypertension, hypothyroidism,[1] obesity and a compromised immune system.[4] A family history is present in 4-8%.[1] Multiple recurrences are rare, and recurrence does not seem to be correlated with prognosis. Rarely, it can occur bilaterally.[4]

The prognosis is largely favourable. Even without treatment complete recovery will occur in 70% of cases, and near-complete recovery in 85%.[4,5] However, 15% will be left with long-lasting sequelae, which can cause considerable psychological distress. Aberrant neural regeneration can result in regenerating motor neurons innervating inappropriate muscles, resulting in abnormal movements or facial synkinesis. Autonomic dysfunction can occur when fibres intended for salivary glands instead reconnect with the lacrimal ducts, causing lacrimation while eating – the phenomenon nicknamed "crocodile tears".[1,5]

The first documentation of facial nerve paralysis in medical literature was in 1798 by Nicolaus Anton Friedrich, a German pathologist at the University of Wurzburg, who described three cases of acute unilateral facial paralysis. Twenty years later, the Scottish surgeon and anatomist Sir Charles Bell was the first to delineate the anatomy of the facial nerve. In his paper of 1821, titled *On the Nerves: Giving an Account of some Experiments on Their Structure and Functions, Which Lead to a New Arrangement of the System*, he described the disease of unilateral facial paralysis, or Bell's palsy.[1]

Aetiology

Bell's palsy is diagnosed when no other medical aetiology is identified as a cause of the facial weakness.[5] Until recently Bell's palsy has been considered idiopathic, although more recently a viral aetiology has been proposed. The viruses most frequently implicated are Herpes Simplex Virus Isoform -1 (HSV-1) and

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Herpes Zoster Virus (HZV). This theory dates back to 1996 when, in a study of 14 patients who presented with Bell's palsy, Murakami *et al* isolated HSV-1 genomes in the endoneurial fluid of the facial nerve and posterior auricular muscle in 79% of patients.[6] When latent, these viruses can be reactivated from the geniculate ganglion of the facial nerve in the temporal bone.[1,3,6] The mechanism of injury is a viral-mediated inflammation which causes oedema of the nerve

sheath and compression of the nerve, with or without demyelination.[7] Facial anatomy may also play a role. In a study using CT with multiplanar reconstruction (MPR-CT) Murai *et al* demonstrated that a narrow facial nerve canal is an additional risk factor.[7] Certain systemic disease processes can predispose patients towards Bell's palsy, most commonly hypertension and diabetes[2] and less commonly hypothyroidism,[2] sarcoidosis, amyloidosis and Sjogren's syndrome. Pregnancy is also a predisposing factor and a link with hypertensive disease of pregnancy is well-established. Palsies occur more commonly in the third trimester and early post-partum period.[8,9]

Approximately 70% of facial nerve palsies are considered to be Bell's palsy; 30% of patients presenting with facial paralysis have other underlying causes.[4] Varicella Zoster Virus (VZV) in the geniculate ganglion can also cause a facial paresis (in this instance it is known as Ramsay Hunt syndrome and characterised also by pain and vesicles affecting the ear). Other infections such as *Borrelia burgdoferi*, the bacterium causing Lyme disease, and less commonly Rickettsia[10] can also cause a facial paralysis. Lyme disease is more common in children. In a Finnish study of 46 children aged 0-16 years in September 2013 Kanerva *et al* noted that Borreliosis was the single most common cause of facial palsy with VZV the second. In that study, the study hospital was located in a region

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endemic for Lyme disease.[11] Otitis media has also been implicated, especially in children.[2]

Other non-infective causes of facial nerve paralysis should be considered, especially in the recurring case. Benign or neoplastic tumours of the head and neck, specifically those of the parotid gland, posterior fossa or nerve sheath itself, should be looked for and excluded. A longitudinal study performed in Taiwan which followed 2,618 patients with Bells Palsy for five years after diagnosis demonstrated a statistically significant increased risk of oral cancer in patients with Bells palsy.

[12] Cholesteatoma should be considered in those whose hearing is affected.[3] Bilateral facial nerve paralysis is uncommon, accounting for less than 1% of all cases. In these cases Lyme disease,[3,10] Guillian-Barre and sarcoidosis should all be considered.[13]

In general practice the diagnosis remains clinical. The important differentiation to make is between a peripheral motor neuron palsy or a central upper motor neuron lesion as the management of each is substantially different. The differentiation can be made through a careful examination of the cranial nerves, in particular cranial nerve VII, the facial nerve. Upper motor neuron cortical and sub-cortical damage, commonly caused by a middle cerebral artery stroke causes a contralateral lower two-thirds facial palsy which does not affect the frontalis muscle of the forehead to to bilateral cortical innervation of this muscle: the forehead is "spared" the paralysis. A lower motor neuron facial palsy causes an ipsilateral flaccid palsy which affects both the frontalis muscle of the forehead and the lower facial muscles.[14] In the diagnosis of Bell's palsy the facial weakness affects the forehead, and the patient loses the ability to raise an eyebrow or wrinkle the forehead, whereas in a cortical stroke the forehead is unaffected. Careful clinical assessment is required to make the distinction.

The anatomy[14,15]

The facial nerve (CN VII) is largely composed of motor fibres. It also carries sensory fibres from the anterior two thirds of the tongue and parasympathetic secretomotor fibres to the parotid, sublingual and lacrimal glands, via the greater superficial petrosal nerve, and submandibular glands via the chorda tympani, which conveys taste from the anterior two thirds of the tongue.

The cranial nerve originates from the facial nerve nucleus in the brainstem. It takes a winding route through the temporal bone, to emerge from the base of the skull at the stylomastoid foramen via the fallopian canal. In Bell's palsy the facial nerve is most commonly compressed at the narrowest segment of the fallopian canal. When the facial nerve emerges from the stylomastoid foramen, it gives off a number of branches which innervate most of the muscles of the face. The posterior auricular nerve supplies part of the occipitofrontalis muscle of the scalp and the posterior auricular muscle of the ear. A further branch supplies several muscles of the throat. The nerve then enters the parotid gland, one of the main salivary glands which sit underneath the ear. Five terminal branches of the facial nerve emerge from the parotid gland to supply most of the muscles of the face, including muscles of the forehead, temple, supra-orbital region, cheek, upper and lower lips, corner of the mouth, chin and neck.

Clinical features

It follows that in Bell's palsy most movements of the face are affected. There is an ipsilateral facial droop of the affected side of the face, with drooping of the eyebrow and corner of the mouth. There is difficulty closing the eyes and blinking, smiling and frowning. The nasolabial fold is typically absent, and the mouth may be drawn toward the unaffected side on smiling.[1,14] Hearing can be affected due to paralysis

of the stapedius muscle; the most common finding is hyperacusis. Taste may also be altered.[1,3]

Diagnosis

A consultation in general practice should include an examination of the cranial nerves, paying attention to the extent of facial weakness especially with regards to the forehead, and examining for the presence of hearing loss. To exclude other diagnoses, the skin of the head and face should be assessed and the ear examined for signs of ear infection. Granulation and pus may indicate cholesteatoma, discharge can point towards otitis media, while a vesicular rash may indicate Ramsay Hunt syndrome.[4]

If HSV1 or VZV are suspected, serology can be sent for confirmation, although this will not change management. Serological tests for Lyme disease (IgM, IgG) should be used in endemic areas to exclude the disease.

A number of scoring systems can be used to grade facial paralysis. The most commonly used is the House-Brackmann Scale which has grades from I (normal facial movements) to VI (complete paralysis).[16]

TABLE 1: HOUSE-BRACKMANN SCALE

Grade	Definition
I	Normal symmetrical function in all areas
II	Slight weakness noticeable only on close inspection Complete eye closure with minimal effort Slight asymmetry of smile with maximal effort Synkinesis barely noticeable, contracture, or spasm absent
III	Obvious weakness, but not disfiguring May not be able to lift eyebrow Complete eye closure and strong but asymmetrical mouth movement with maximal effort Obvious, but not disfiguring synkinesis, mass movement or spasm
IV	Obvious disfiguring weakness Inability to lift brow Incomplete eye closure and asymmetry of mouth with maximal effort Severe synkinesis, mass movement, spasm
V	Motion barely perceptible Incomplete eye closure, slight movement corner mouth Synkinesis, contracture, and spasm usually absent
VI	No movement, loss of tone, no synkinesis, contracture, or spasm

Treatment

The most well-established treatment for Bell's palsy is corticosteroids, in accordance with the idea that the mechanism of injury is an inflammatory-mediated nerve damage. The available evidence shows significant treatment benefits in terms of both gain of complete recovery and reduction of long-term sequelae.[4,5,17,18] Steroid treatment should be commenced within 72 hours of the onset of symptoms.[18] There is no evidence for use of steroids in children under the age of 16.[4] A meta-analysis performed in 2010 looked at seven trials with a total of 1,507 participants. When treated with corticosteroids, 23% of the patients had incomplete facial recovery at six months compared with 33% of controls: a risk reduction of 71%. The number needed to treat (NNT) for corticosteroids to avoid one patient from suffering incomplete recovery of facial function was 10.[5] There is no established treatment regime. NICE recommends prednisolone. Options include 25mg twice daily for ten days or 60mg daily for five days, followed by a daily reduction in dose of 10mg for a total treatment time of 10 days.[19]

A straightforward diagnosis of Bell's palsy does not require a referral to neurology

Even though a viral link is well-established, there is currently no good evidence for antiviral agents, either alone or in addition to a corticosteroid and there have been numerous trials on the subject.[10,20] A large meta-analysis performed in 2011 looked at studies which compared treatment with valaciclovir and aciclovir and prednisolone to prednisolone alone. It illustrated that although treatment with a corticosteroid plus an antiviral agent may lead to slightly higher recovery rates at three and six months compared to treating with a corticosteroid alone, the difference was not statistically significant.[20]

Other treatment

Eye care is important. If the patient cannot close the eye the exposed cornea is prone to drying, infection and trauma. Lubricating drops should be prescribed and taping the eye shut at night (placing the tape over a soft pad) should be recommended, with careful instruction as to how to prevent further damage. If the eye cannot be closed with tape, an urgent referral to ophthalmology is advised.[7,21]

A number of other treatments have been tried. Physiotherapy is not recommended,[2] while surgical decompression of the facial nerve does little to alter the prognosis. However, more recently, some success has been achieved with a small number of well-selected cases who had been shown to have an unfavourable prognosis following electrodiagnostic testing.[22,23] Trials of acupuncture as a treatment for Bell's palsy indicate that while the available evidence is broadly in favour of acupuncture, the evidence is of poor quality and unreliable.[24] Laser therapy has been

shown to improve neural regeneration. A recent small trial using laser therapy on specific points of the face produced promising results, although further research is required.[25]

Referral

A straightforward diagnosis of Bell's palsy does not require a referral to neurology. However, referral should be considered for recurrent or bilateral cases. If the diagnosis is not straightforward, and a tumour is suspected, the patient should be referred for neuro-imaging or referred as a fast-track referral to ear, nose and throat or neurology as appropriate.[2] For the few cases of Bell's palsy which have limited improvement over time, a referral to neurology for electrodiagnostic testing to assess the extend of nerve damage is warranted.

Conclusion

Bell's palsy is not always idiopathic; herpes simplex virus and herpes zoster virus often play a role. HSV-1 and VZV serology can be an aid to diagnosis if a viral aetiology is suspected. Although this does not currently change management, it can help the patient to better understand their disease. Other causes of facial nerve paralysis should be carefully looked for and excluded, while Lyme disease and otitis media should be considered in children. It is worth checking blood pressure and assessing for diabetes in all who present with Bell's palsy. In adults over the age of 16 years, corticosteroid treatment should be commenced within 72 hours of onset of symptoms. Those cases in which the history and examination findings point towards an upper motor neuron facial nerve paralysis, or when there are findings beyond an isolated CN VII involvement, should be referred for neuro-imaging and further assessment. The experience can be extremely frightening for the patient, and reassurance can be given that the majority will make a full recovery.

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SUMMARY OF KEY POINTS

1. Bell's palsy is not always idiopathic: HSV and HZV are the two main viral causes
2. In addition to an examination of the cranial nerves, examine the ear, parotid gland and skin of the face, head and neck
3. Check blood pressure and look for diabetes in all who present with Bell's palsy
4. Exclude Lyme disease and otitis media in children
5. Treat with corticosteroids within 72 hours of onset of symptoms
6. A treatment regime suggested by NICE is: prednisolone 60mg a day for five days followed by a reduction of 10mg a day for a ten day total course
7. Remember eye care: lubricant drops
8. Refer recurrent or bilateral Bell's palsy for neuro-imaging and further assessment

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