Bell’s palsy — a neglected condition?

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The facial nerve is the nerve most often paralysed in the body. Of the 82 published aetiologies, May found that the ‘idiopathic’ group called Bell’s palsy was the commonest (60-70%), followed by trauma (15%) and herpes zoster oticus (7%).

Bell’s palsy is universally regarded as a benign condition with good recovery. Unfortunately this is not true. The majority of patients will have either a partial paralysis (paresis) or a total paralysis lasting 2-3 weeks only. Unfortunately those with a total paralysis have a 6-30% chance of staying paralysed for 3 months and will have an incomplete recovery. In older people this may be acceptable but for a young person planning a career as a model it is disastrous.

The usual modus operandi of attending physicians throughout the world is to give a favourable prognosis and the majority prescribe steroids and vitamins. Sometimes the patient is referred to a specialist who confirms that recovery will take a few weeks only. Regular electrodiagnosis is seldom used to monitor the condition of the nerve. Physiotherapy is advised as well as a follow-up examination after a month. If the patient enquires about possible surgery it is usually condemned outright.

Patients who are still paralysed after 1 month are told that they are exceptions. The specialist cannot offer a definite time when recovery can be expected and occasionally refers the patient to a neurosurgeon or otologist for possible surgery. On rare occasions plastic surgery is eventually recommended; this involves an error of judgement because all Bell’s palsies will have a moderate recovery. Non-recovery of Bell’s palsy after 6 months is proof of a wrong diagnosis and warrants serious investigation for other causes.

This article presents the latest views on Bell’s palsy and herpes zoster of the facial nerve. The author had the privilege of being the moderator at the last World Congress of Otorhinolaryngology where the experts debated the controversy regarding medical and surgical treatment. It became quite clear that early and frequent electrodiagnosis was the only way the condition of the nerve could be monitored.

In First World medicine it has become necessary to keep patients informed, especially if any controversy exists about treatment. It is therefore wise to use all available investigations in such cases and to alert the patient if a critical stage of nerve degeneration develops. Involving the patient in decision-making protects the doctor from possible medicolegal problems later on.

Facial nerve anatomy

The facial nerve is a mixed motor-sensory cranial nerve. It contains approximately 10 000 fibres of which 7 000 are myelinated motor axons that reach the facial muscles. The nucleus in the pons also receives some uncrossed fibres. A lesion central to the nucleus, which occurs in approximately 1% of cases of facial palsy, results in incomplete paralysis with maximal involvement of the musculature of the middle and lower face and a tendency towards sparing of the frontal and upper zygomatic regions. Peripheral (lower motor neuron) lesions are distal to the nucleus and involve all regions of the face.

Secretory fibres reach the lacrimal gland via the greater superficial
Fig. 1. Intra-osseous course of the facial nerve.
petrosal nerve which leaves the main trunk at the geniculate ganglion, situated in the attic region of the middle ear. Another set of fibres to the submandibular and sublingual salivary glands leaves the main trunk in the mastoid via the chorda tympani, which traverses the middle-ear space close to the ear-drum.

Afferent fibres conveying taste sensation from the anterior two-thirds of the tongue follow the pathways of the lingual nerve and the chorda tympani. Another set of afferent fibres mediates sensation from the posterior external auditory canal, external ear and deep parts of the face; these are prominently involved in herpes zoster oticus (blisters and severe otalgia) and Bell’s palsy (pain).

The facial nerve exits from the pons, from where four portions of the nerve are distinguished (Fig. 1). The intracranial portion, 12-14 mm long, passes through the cerebellomedullary angle to the internal auditory canal. The mastication, 8-10 mm long, is inside the internal auditory canal together with the vestibulocochlear nerve. The intratemporal portion, 28-30 mm long, is inside the fallopian canal, the longest bony conduit through which a peripheral nerve passes. The diameter of the fallopian canal varies most often involved in skull fractures and surgical mishaps.

- The mastoid segment, 13 mm long, passes inferiorly to the stylomastoid foramen.
- The extratemporal portion of the nerve leaves the temporal bone.

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Crucial to any consideration of facial nerve paralysis is the determination of the site, nature, and severity of the lesion responsible for the condition from an average of 0.68 mm at its beginning to 1.50 mm at its widest. Three segments of the nerve are distinguished inside the temporal bone as the nerve has two knee-bends:

- The labyrinthine segment, 2-4 mm long, passes anterolaterally to the geniculate ganglion through the narrowest part of the fallopian canal.
- The tympanic segment, 11 mm long, passes posteriorly from the geniculate ganglion through the middle ear to the mastoid and is forms the pectoralis minor and supplies the muscles of the face.

The topographical anatomy helps in localising a lesion, but unfortunately it has limited value in case of infection of the nerve.

### Axonal block, degeneration and regeneration

As stated by Sunderland (quoted by Coker and Fisch) ‘Crucial to any consideration of facial nerve paralysis is the determination of the site,
nature, and severity of the lesion responsible for the condition, together with some estimate of the prospects for spontaneous recovery. Various degrees of damage are described of which the most important are:

- **Neuropraxia** — a localised physiological conduction block with nerve fibres responding to electrical stimulation proximal and distal to the lesion but not across the affected segment. Axonal continuity is preserved and no wallerian degeneration occurs. Following a latent period complete function is restored. In the case of the facial nerve this takes from 1 to 2 weeks.

- **Axonotmesis** — the axons are separated into proximal and distal parts but the endoneurial sheaths are preserved. Distal parts undergo wallerian degeneration; however, each axon retains the ability to regenerate along its endoneurial tube to its original end organ, with the potential for complete restoration of function. The facial nerve takes 10-16 weeks to recover.

- **Endoneuromatosis** — the nerve fibre and endoneurial sheath are destroyed but the perineurial sheath survives. Regenerating axons may be blocked by scar tissue, resulting in partial reinnervation, or upon entering the damaged distal segment they may enter functionally different endoneurial tubes with inappropriate end organ reinnervation. Synkinesis, the unintentional movement of one part of the face when another moves voluntarily, is the functional expression of misdirection. In the face this recovery also takes 10-16 weeks.

- **Perineuromatosis** and
- **Neurotmesis** — further stages of discontinuity which can only be repaired by surgical suturing or grafting.

The terms endoneuromatosis and perineuromatosis are new definitions suggested by Fisch.³

**Definitions**

1. **Palsy** is a nonspecific term which includes both partial (pareisis) and total (paralysis) loss of neural function.

2. **Peripheral and central palsy** have been discussed in the section on anatomy.

3. **Bell’s phenomenon** is the involuntary upward tilting of the eyeball on attempts to close the eye. In case of a paralysis the white sclera remains visible.

4. **Bell’s palsy** is the name for idiopathic facial palsy. One often hears the erroneous expression that a patient 'has a Bell', when the speaker actually means that the patient has a facial palsy. The incorrect use of the name of Sir Charles Bell stems from the historical background. In 1829 Sir Charles distinguished between the sensory innervation of the face by the fifth nerve and the motor innervation of the muscles of expression by the seventh nerve. For some time the seventh nerve was known as 'Bell's nerve' and all cases of peripheral facial paralysis were called 'Bell's palsy'. Only later, as specific lesions were shown to involve the nerve as a result of tumour growth, trauma or infection, was the term Bell's palsy narrowed down to those cases coming on abruptly for no apparent reason in an otherwise healthy person.

**Electrodiagnosis**

Many tests are available but it is recommended that the maximal nerve stimulation test (MST) described by May⁴ in 1971 be used in all cases. If a myograph is available, the more accurate electroneurography (ENoG) method described by Essen⁵ in 1973 can be employed in addition to the MST.

For the MST a Hilger nerve stimulator is used to deliver the maximal stimulation the patient can tolerate but any standard stimulator may suffice. Surface electrodes are used. The test is not painful but in children a light halothane or ketamine anaesthesia may be needed. The stimulation is over the main trunk of the nerve in front of the ear and the contraction of the face is assessed as a percentage of the result obtained on the normal side. The stimulation is a square wave impulse of 0.6 millisecond duration. Because of the fact that the stimulation is applied distally to lesions inside the temporal bone, there is an actual delay of 24-48 hours. i.e. the result shows what the nerve was like 1-2 days previously. Therefore the speed at which a nerve degenerates is important and daily tests are needed in the acute phase.

In ENoG a square wave impulse of 0.2 millisecond with a voltage varying from 60 to 120 volts is used. The contraction is observed but the summation action potential of the nerve is also measured by means of surface electrodes in the nasolabial fold (normal 2-6 millivolts).

Both tests are quick and easy to perform. When carried out by the doctor himself, they will help him to become experienced in measuring nerve degeneration. There is no other method by which the condition of a totally paralysed nerve can be assessed. 'Monitoring' has become standard practice in anaesthesia, myocardial infarcts and fetal distress, and omitting it constitutes negligence in First World medical practice. Similarly a patient with a paralysed face can now expect accurate monitoring of the condition of the...
nerve. After all, a skew face in a young person causes as severe stress as does a cardiac condition in an elderly patient!

**Idiopathic facial nerve palsy (Bell's palsy)**

The name idiopathic indicates the fact that this diagnosis is based on exclusion and is therefore a dangerous diagnosis. May reported that 10% of referrals originally diagnosed as Bell's palsy had treatable, progressive or life-threatening disorders, e.g. tumours. Sir Terence Cawthorne warned: 'All that palpises is not Bell's.' Jongkees said that every facial palsy should be treated as an emergency. Adour advised that each patient with facial palsy should have a general neurological as well as a thorough ear, nose and throat examination.

For many years it was thought that Bell's palsy was caused by vascular spasm because it was found that the acute palsy sometimes followed exposure to cold air. The names ischaemic or rheumatic facial palsy or 'paralysis faciale a frigore' are well known. Surgical exploration usually revealed swelling of the nerve and the entrapment theory was developed. It was reasoned that swelling of the nerve resulted from vascular spasm. This swelling then closes the venous channels resulting in a vicious circle inside the narrow bony canal. That such a mechanism could be possible is demonstrated by the fact that in sclerosing bone dysplasias like marble bone disease, where the fallopian canal does stenose, the face never paralyses slowly. It always presents as acute paralysis followed by total degeneration and partial recovery. Further attacks are the rule, resulting in progressive loss of nerve function. Some cases have even presented with the first attack of paralysis at birth, thereby confirming a non-infective aetiology.

Because Bell's palsy is not a lethal condition, few histological studies are available. Those published point to an inflammatory cause complicated by swelling of the nerve resulting in pressure on the vascular supply to the nerve tissue. Surgical decompression of the mastoid and tympanic portions of the nerve was often done in past decades but despite early surgery the results were not encouraging. In 1961 House developed the technique of decompressing the labyrinthine portion of the facial nerve. This difficult procedure, entirely safe in well-trained hands, requires a small craniotomy in the temporal region, extradural approach to the roof of the temporal bone and drilling away the bone over the internal auditory canal, the epitympanum and then the short and very thin labyrinthine portion of the fallopian canal. The findings at the entrance of the fallopian canal were dramatic. Coker and Fisch reported severe swelling of the nerve inside the internal auditory canal and dramatic compression of the nerve at the 0.68 mm entrance to the canal. Intra-operative evoked electromyography showed that this area was the pathological site in 94% of cases.

According to Adour clinical, epidemiological and laboratory data have shown that Bell's palsy is an acute, benign cranial polynuereitis that is probably caused by reactivation of the herpes simplex virus. The primary disease is a sensory ganglionitis of the central nervous system with secondary motor nerve palsy. The muscle paralysis is caused by inflammation and demyelination rather than ischaemic compression. Involvement of the trigeminal, glossopharyngeal, vagal and second cervical nerves is common. Taste disturbance due to chorda tympani involvement is frequently accompanied by numbness of the tongue and the skin of the cheek due to trigeminal nerve involvement.

**The natural course of untreated Bell's palsy**

Peitersen reported on 2,255 patients with facial palsy. Of these 67% had Bell's palsy. Incomplete paralysis (paresis) occurred in 30% and they all recovered within 3 - 4 weeks. Of the remaining patients with total paralysis, 75% recovered within 4 weeks without sequelae. The remaining 25% (17% of all Bell's palsies) had a paralysis lasting 11 - 16 weeks and recovered incompletely (but none remained permanently paralysed). Owing to the damage to the endoneurial tubes, the regeneration of the nerve fibres was only partially successful, resulting in various degrees of paresis, contractures and associated movements (synkinesis).

In the author's experience recovery of a totally degenerated nerve will result in approximately 70 - 80% maximal movement capacity in a young person and 50 - 70% in an adult.

**Herpes zoster oticus**

This was first described by Körner in 1904 and the full syndrome detailed by Ramsay Hunt in 1908. Apart from the blisters at the external ear, which sometimes appear after a few days only, these patients have more pain than is experienced with Bell's palsy. Vestibular and cochlear symptoms often occur. The damage to the vestibular nerve is permanent but the patient recovers very well by means of adaptation. Hearing loss, when it occurs, is usually permanent.
Dramatic swelling of the nerve inside the internal auditory canal was found by the author in a patient during exploration on the 7th day. The labyrinthine portion of the nerve and the geniculate ganglion were pale and the colour returned dramatically on relief of the pressure on the petrosal vessels anterior to the geniculate ganglion. Small vessels filled with blood just like a collapsed canvas fire-hose fills with water when the tap is opened. This was witnessed by the attending general practitioner. This elderly patient's recovery was markedly better than the poor recovery of degenerated nerves usually found in herpes zoster infection.

In general the recovery of movement in cases of total degeneration in herpes zoster infection is less than 50%, in contrast with a better recovery in Bell's palsy. This is due to the severe damage to the endoneurial tubes by the more severe infection by the zoster virus.

**Treatment of Bell's palsy and herpes zoster oticus**

Time is essential in the management of facial palsy. Not only must the correct clinical diagnosis be established but a course of action must be instituted before irreversible damage to the nerve occurs. Following Jongkees' advice, treat every facial palsy as an emergency. The first days are the most important. The follow-up must be an accurately planned and strictly adhered to schedule.

Adour found the only proven effective treatment by medical means to be steroid treatment. Prednisone 60 mg daily from day 1 is given for at least 5 days and continued in lower doses for at least another 7 days. In patients with total paralysis steroids reduced the percentage of total degeneration from 30% to only 6% in his series. This treatment is also recommended for herpes zoster oticus but it has been found that the treatment should be continued for a longer period because the chances of total degeneration are significantly higher, even as high as 70%. Acyclovir can be used but no statistical proof of efficacy for Bell's palsy or herpes zoster oticus has yet appeared. Whether it is necessary to give steroids in cases of incomplete paralysis (paresis) is not certain. It may possibly prevent a paresis from progressing to a paralysis. Physiologists have recommended that hyperbaric treatment could possibly help, but this has not yet been tried by the author.

As stated previously, electrodiagnosis is the only method which will monitor the nerve degeneration once the palsy is total.

**A practical suggestion is that the MST be used if a myograph is not available. If the contraction of the muscles decreases to less than 50% of that on the normal side, ENoG is essential.**

This is recommended from day 3 (because of the built-in error of the test) on a daily basis until the condition has stabilised. Usually some degeneration occurs in all cases of total palsy. Only when the degeneration exceeds 70% does one need to be alarmed. In the author's experience the condition becomes serious if the summation potentials drop to a measurement of less than 1.0 millivolt or more than 80% degeneration. Twice daily tests are then ideal. A decompression operation becomes a serious consideration at 0.5 millivolt or 90% degeneration and the patient should be informed about the crisis situation.

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Lacrimation should be monitored by means of Schirmer's test. A bilateral temporary decrease of lacrimation is often found. This is due to a central shutdown mechanism by the reticular formation in the pons and need not be an unfavourable sign. On the other hand a unilateral decrease will correlate with the electrical tests and be a sign of nerve degeneration. If the decrease is more than 70% it indicates serious degeneration and supports the plea for mandatory electrodiagnosis.

Unfortunately the lacrimation test cannot be used as the only test for nerve degeneration, just as the test of submandibular salivary secretion has also proved to be impractical. Topographical diagnosis should be of value in determining the level of disease involvement in the tortuous course of the facial nerve, but unfortunately has not proved to be reliable in the case of infections. It has important value when fractures and tumours are present, however.

Statistical proof of the efficacy of surgical decompression of the labyrinthine portion of the nerve was published by Fisch. Reports from more researchers are awaited. If the guidelines set out above are followed, the reports can be compared with the findings of Fisch. This surgery is difficult and special training is required to decompress the labyrinthine portion of the nerve which is less than 1.0 mm in diameter. This surgery should not become the victim of disrepute as a result of lack of surgical skill. Fortunately in this country we already have a few otologists trained in the technique.

The value of physiotherapy of the facial muscles is uncertain. Faradic stimulation during the first 3 weeks is only used as a test and has no validity as a treatment. In favourable cases the movement
returns within that period and no damage occurs to the muscles as a result of the rest period. In the unfortunate cases who have to wait another 2 - 3 months for movement to come back, galvanic stimulation of the muscles to prevent excessive wasting is often recommended. There is no proof of its value except as supportive treatment for the patient's depression, which is always present because of the disfigurement. Cawthorne once warned against this treatment because he had the impression that it increased contractures (personal communication). Whether this is true is not certain.

When the movement returns after 3 months, active exercises by the patient without any electrical stimulation are very valuable as demonstrated by Brook and Devries8 from Amsterdam. Synkinesis and contractures always occur to a varying degree. The mime therapy they demonstrate on their videotape not only helps the patient to exercise the muscles, but also helps in reprogramming the cortical centres because some of the regrown nerve fibres invariably end up in the wrong endoneurial tubes because of endoneurotmesis. The mass movements occurring after regeneration of the nerve make selective movement of a part of the face impossible and can lead to embarrassing moments, e.g. smiling and winking at the boss's wife when only a smile is intended! The mime therapy helps to correct this and may 'save the day' for someone who has to appear in front of an audience.

For a dry eye it is necessary to prescribe artificial tears and ointment. Consultation with an ophthalmologist is advised as temporary tarsorrhaphy may become necessary to prevent corneal ulceration.

Coker and Fisch9 reported that recurrent facial palsy occurred in 10% of idiopathic cases. In their experience surgical decompression prevented recurrences in Bell's palsy as well as in Melkersson-Rosenthal syndrome. May10 found contralateral recurrences were twice as common as ipsilateral recurrences in Bell's palsy. He did not find recurrences in herpes zoster oticus. He remarked about the possibility that herpes simplex type 1 infection could be the cause of Bell's palsy because of these findings. May also reported that other causes of facial palsy should be ruled out in all cases of a recurrent palsy. In 6 of 20 patients with a recurrence on the same side a tumour was discovered.

Sclerosing bone dysplasias should also be ruled out especially if a recurrence is found in a child. A radiological examination of the skull base and of the dorsal spine is obligatory in all facial palsies occurring in children because marble bone disease usually has recurrent facial palsy as its first symptom.11
Conclusion

- Treat all cases of facial palsy as an emergency. The actions taken during the first week will determine the prognosis.
- Make sure of the cause of a facial palsy before labelling it a 'Bell'.
- Steroid treatment is recommended for all patients suffering from Bell's palsy. In herpes zoster oticus acyclovir as well as steroids are recommended.
- Electrical monitoring of the nerve is mandatory in all cases of total paralysis to detect the small but significant number of patients in whom total degeneration of the nerve with subsequent incomplete recovery will occur.
- Surgical decompression of the labyrinthine portion of the facial nerve is recommended in selected cases. The decompression should be carried out before total degeneration has occurred. If total degeneration has already occurred, the value of surgery can only be to assist recovery of the nerve. Obliteration of the endoneurial tubes by scar tissue may possibly be reduced if the pressure on the nerve is relieved within a few days of the tragic event of total degeneration.

The closing remarks of Professor Fisch12 of Zürich at the International Congress summarised the situation very well: 'Give steroids if you wish but remember that this does not prevent total degeneration in all cases. If you want to do more than just watch the nerve go out on steroids, and if you feel trained to do the proper surgery, decompress the mental foramen and labyrinthine portion of the nerve but without doing harm to the patient.'

REFERENCES

12. Bronk J, Devries P.P. Mimo and mimo-therapy. 1984, Videotape from Audiovisual Centre, Free University of Amsterdam, P O Box 7161, 1007 MC Amsterdam, Netherlands.

OPSOMMING
Bell se verlamming
Bell se verlamming en die nou verwante herpes zoster oticus (Ramsay Hunt-sindroom) word bespreek. N Pleidool vir vroeër elektdiagnose word gemaak, want dit is die enigste manier waarop die toestand van die senuwee gemonitoeer kan word wanneer die verlamming totaal geword het. Die waarde van mediese en chirurgiese behandeling word bespreek.