

## Bilateral facial nerve palsies: Groote Schuur Hospital experience

P. J. WORMALD, M.B., Ch.B.,\* S. L. SELLARS, F.R.C.S.,\* J. C. DE VILLIERS M.D., F.R.C.S.†

### Abstract

Bilateral facial nerve palsies are rare. This article details the Cape Town experience of 24 patients with this condition seen over the past 20 years and highlights the three main groups in which these bilateral facial nerve palsies occurred. These are Bell's palsy, fracture of the temporal bones and sclerosteosis. A variety of other rare causes was also found. Bell's palsy remains a diagnosis of exclusion though there has been some evidence to suggest implication of the *Herpes simplex* virus. The mechanism whereby temporal bone fractures cause bilateral facial nerve palsies is discussed. The clinical presentation and management of sclerosteosis is also discussed.

### Introduction

Facial nerve palsies are common in otolaryngology practice. However, bilateral synchronous facial nerve palsies (BSFNP) are relatively rare (McGovern, 1965; Adour *et al.*, 1978; May and Hardin, 1978; Prescott, 1988). Two major groups of BSFNP (skull trauma and bilateral synchronous Bell's palsies) have been seen in the Department of Otolaryngology. The commonest cause in facial nerve palsy remains the undiagnosed or idiopathic (Bell's) palsy. An incidence of 0.8–1 per cent for BSFNP amongst unilateral palsies is our experience (Prescott, 1988) and this seems to be in keeping with the literature (Adour *et al.*, 1978; May and Hardin, 1978).

In the neurosurgical experience of BSFNP, sclerosteosis and skull trauma form two major groups. Bilateral facial nerve palsies associated with skull trauma usually result from extensive skull base fracture. They are however rare and because of the severity of the injury, death probably occurs in many without identification of the facial weakness. The fracture lines involve both temporal bones and the mechanics of this 'fracture dislocation' of the petrous-temporal bone have been well described by de Villiers (1971). Sclerosteosis is found in the local community because of its autosomal recessive inheritance in the Afrikaner population of South Africa in which its incidence is 1:60,000. This condition is rare outside of the Afrikaner community. It is a cranio-tubular dysplasia of bone with bony overgrowth and thickening throughout the body, especially of the skull and long bones.

### Materials and methods

The patients were taken from the records of the Otolaryngology Facial Nerve Clinic and from the records of hospital admissions to the otolaryngology and neuro-

surgical wards of Groote Schuur Hospital over the 20 year period from 1969 to 1990. Only patients with bilateral synchronous facial nerve palsies (occurring simultaneously in the same patient) were included. However, seven of these patients had an incomplete palsy on one side.

The age of the patients ranged from 5 months to 56 years (mean = 23.7 years), with 15 male and 9 female patients. Sclerosteosis occurred exclusively in the Afrikaner section of the community, while in the other two major categories of BSFNP there was no racial preponderance. In the sclerosteosis group of patients, six patients underwent cranio-cervical decompression for raised intra-cranial pressure. No facial nerve decompressions were done.

One unilateral facial nerve exploration and reanastomosis was undertaken in the temporal bone fracture group and this resulted in partial recovery of function. The patient with tuberculous mastoiditis is currently on medication and may yet fully recover.

### Results

One patient was lost to follow-up and his final outcome could not be determined. The results are summarized in Tables I and II.

### Discussion

The facial nerve is of primary interest to the otolaryngologist because of its anatomical path through the petrous temporal bone and parotid gland.

Analysis of this hospital's experience of BSFNP shows the three distinct major groups of sclerosteosis, trauma and Bell's palsy. Two-thirds of the palsies have a definite identifiable aetiology indicating that a primary pathol-

TABLE I  
BILATERAL SYNCHRONOUS FACIAL NERVE PALSIES

Idiopathic	8
Sclerosteosis	7
Temporal bone fracture	6
Bilateral tuberculous mastoiditis	1
Lymphosarcoma	1
Guillain-Barré	1
Total	24

ogy should always be considered (Sellars, 1983). May and Hardin (1978) found a cause for all of the six bilateral facial nerve palsies seen in their study and stated that if the facial palsy was bilateral then it should not be assumed to be a Bell's palsy. Rare conditions that can present with facial diplegia include Moebius' syndrome which is due to a congenital aplasia or hypoplasia of the seventh nerve nuclei (Masaki and Matsumoto, 1971); Melkerson's syndrome which consists of facial paralysis, facial oedema and a deeply furrowed tongue; and Heerfordt's disease which consists of sarcoidosis, iridocyclitis and paralysis of one or more cranial nerves (McGovern, 1965). None of these conditions presenting with facial diplegia occurred in this twenty year series.

Bell's palsy has in the past been considered a mono-neuropathy, but recent reports (Rauchbach *et al.*, 1975; Adour *et al.*, 1978) indicate that in more than two-thirds of patients a cranial polyneuropathy involving the trigeminal and glossopharyngeal nerves exist. In the aetiology of Bell's palsy there are multiple implicated factors, the most significant of which is the association with *Herpes simplex* viral infections (Adour *et al.*, 1978). One of the patient's with bilateral facial nerve palsies in Adour's series had a significant *Herpes simplex* viral antibody titre in the cerebrospinal fluid. There is no epidemiologically significant relationship to age, sex, or seasonal variation, and diabetes mellitus and hypertension are unrelated. Pregnancy, however, does appear to be related to the incidence of Bell's palsy (Prescott, 1988).

The significance of determining whether a *Herpes simplex* infection is involved is debatable. As yet there have been no trials on the effect of anti-viral agents on the prognosis of Bell's palsy and their use is not advocated at present. Surgical decompression of the facial nerve in Bell's palsy is no longer considered as a treatment option by many otologists (Stein and Tønning, 1973; Adour *et al.*, 1978).

Bilateral traumatic facial nerve palsies associated with a head injury usually result from a longitudinal petrous fracture across the skull base and are thus tangential injuries to the facial nerves.

The petrous bone is unique in that its connections to the surrounding bones are fibrous in nature while the apex of the petrous bone is connected to the basilar process by the strong petro-basilar ligament. The slope and bevel of the squamous-temporal bone as it articulates with the surrounding calvarial bones makes impression of this bone into the skull extremely difficult. A force applied to the lateral surface of the temporal bone, which is supported by two buttresses, the mastoid and the zygomatico-temporal, causes a longitudinal fracture of the petrous bone due to a separation of the buttresses and a backward displacement of the petrous apex. Where petro-basilar ossification and fusion has

TABLE II

Recovery of VII Palsy	Complete	Partial	Nil	Lost to follow-up
Idiopathic	7	1	0	0
Sclerosteosis	0	0	7	0
Temporal bone fracture	2	2	1	1
TB mastoiditis	0	1	0	0
Lymphosarcoma	1	0	0	0
Guillain-Barré	1	0	0	0

occurred, the body of the sphenoid will split coronally through the sphenoid sinus and this results in a mirror image fracture in the opposite temporal bone (de Villiers, 1971). Cranial nerve injury (V, VI, VII and VIII) occurs as a consequence of the longitudinal fracture and backward displacement of the petrous bones. Transverse fractures of the petrous bone will not involve both facial nerves. If the palsy is of delayed onset, the prognosis is good. Conductive or sensorineural deafness which can also result in an ossicular chain disruption may need to be attended to.

Sclerosteosis is characterized by syndactyly of the second and third fingers. Deafness and bilateral facial nerve palsies are also frequently encountered (Beighton *et al.*, 1977; Beighton and Hamersma, 1979). These patients can often be recognized early in life by syndactyly, and by a facial nerve palsy that often occurs before overt bony overgrowth is evident. Conductive deafness due to bony overgrowth usually occurs in mid-childhood and this is often followed by sensorineural deafness in the third decade (Beighton and Hamersma, 1976). There is no treatment other than surgical decompression of entrapped structures.

The management of sclerosteosis consists primarily of decompression operations for raised intra-cranial pressure (craniotomy) or a compressed medulla (cranio-cervical decompression). Decompression of the facial nerves resolves the facial paralysis and in some patients may prevent further attacks. However, because of the thickness of the bone these operations are technically difficult and time consuming. Surgery for ossicular fixation can also be effective but reversal of the hearing loss is not necessarily permanent (Beighton and Hamersma, 1979).

#### References

- Adour, K., Byl, F., Hilsinger, R., Kahn, Z., Sheldon, M. (1978) The true nature of Bell's palsy: analysis of 1000 consecutive patients. *Laryngoscope*, **88**: 787-801.
- Beighton, P., Davidson, J., Durr, L., Hamersma, H. (1977) Sclerosteosis—an autosomal recessive disorder. *Clinical Genetics*, **11**: 1-7.
- Beighton, P., Hamersma, H. (1976) The clinical features of sclerosteosis: A review of the manifestations in twenty-five affected individuals. *Annals of Internal Medicine*, **84**: 393-397.
- Beighton, P., Hamersma, H. (1979) Sclerosteosis in South Africa. *South African Medical Journal*, **55**: 783-788.
- de Villiers, J. (1971) Fracture-dislocation of the petrous temporal bone. *Journal of Neurology, Neurosurgery and Psychiatry*, **34**: 104-109.
- Masaki, S., Matsumoto, M. (1971) Congenital bilateral facial paralysis. *Archives of Otolaryngology*, **94**: 260-263.
- May, M., Hardin, W. (1978) Facial palsy: interpretation of neurological findings. *Laryngoscope*, **88**: 1352-1362.
- McGovern, F. (1965) Bilateral Bell's palsy. *Laryngoscope*, **70**: 1070-1079.
- Prescott, C. (1988) Idiopathic facial nerve palsy. *Journal of Laryngology and Otolaryngology*, **102**: 403-407.

- Rauchbach, E., May, M., Stroud, M. (1975) Vestibular involvement in Bell's palsy. *Laryngoscope*, **85**: 1396–1398.
- Sellars, S. (1983) Idiopathic facial nerve palsy. *South African Medical Journal*, **64**: 379–381.
- Stein, R., Tinning, F.-M. (1973) Acute peripheral facial palsy. *Archives of Otolaryngology*, **98**: 187–190.

Address for correspondence:  
Dr P. J. Wormald,  
Department of Otolaryngology,  
Groote Schuur Hospital,  
Observatory 7925,  
South Africa.

**Key words: Facial paralysis; Sclerosteosis**