The Journal of Laryngology and Otology
October 1984. Vol. **98.** pp. 977–979

Familial Bell's palsy

by

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Introduction:

Bell's palsy is defined as a peripheral facial palsy of sudden onset and unknown aetiology. The above condition is the most commonly encountered cause of lower motor facial nerve paralysis.

A variety of aetiological factors have been mentioned as a possible cause of Bell's palsy, such as viral infection, vascular ischaemia, changes in temperature, narrowing of the Fallopian canal and immunological factors.

Hereditary facial palsy has so far been ignored and some of the basic ENT textbooks do not mention it at all (Ballenger, 1977, Scott-Brown, 1979).

We would like to present an isolated family with an unusual number of recurrent episodes of Bell's palsy.

Case Report:

A 14-year-old Indian female presented with a typical Bell's palsy on the right side of her face. She had the attack two weeks prior to the visit and it was improving gradually.

The past history revealed that she had often developed these attacks. The first attack was in 1974 at the age of 6 years when the right side of her face was involved. Two years later the left side was paralysed. In 1980, the third attack occurred and affected the left side of her face again and her present attack was the fourth episode of Bell's palsy.

The duration of the attacks ranged from 3–6 weeks with full recovery. An upper respiratory infection was noted prior to each attack. The attacks started suddenly without any pain in the face or ear; no swelling of the face or the lips was noted.

The family was not worried about the girl's condition because the father had had similar attacks of facial paralysis that had recovered completely (a slight synkinesis was noted during smiling).

The father was 41 years old and he developed the first attack at the age of 15 years. Since then, he had five attacks of right-sided Bell's palsy. The last episode was 10 years ago. He was a mild diabetic and was hypertensive, being well controlled with Diabenese and Aldomet daily.

In the family there were another three boys and two girls, in good health. The family's traceable history showed no other reports of Bell's palsy.

On examination there was loss of tone on the right side of the face, with impaired movement of the facial muscles but a fair closure of the eyelid and no oedema of the face or lips and no fissuring of the tongue were noted. The facial nerve function tests, including nerve excitability test, lacrimation, gustatory sensation, and stapedial reflex tests were normal. The rest of the ENT examination was normal.

Further investigations carried out on both the girl and her father included: full blood count, urea and electrolytes, erythrocyte sedimentation rate, calcium and phosphate, protein electrophoresis, VDRL, all of which were within normal limits. The girl was subjected to a glucose tolerance test which was normal. X-rays of the mastoids showed no abnormality, with normal aeration.

Polytomography of the facial nerve

https://doi.org/10.1017/S0022215100147826 Published online by Cambridge University Press

showed no abnormality and no difference between the sides. Computerised tomography was normal.

The girl recovered two weeks later and only a slight synkinesis of the right face remained.

Discussion:

The facial nerve is one of the most frequently paralysed nerves in the human body. Hauser *et al* (1971) estimated the annual incidence of Bell's palsy to be 22.8 per 100,000 population but in other authors' reported data it varied from 0.16 per 1,000 to 1.4 per 1,000 population (Gregg, 1961; Melotte, 1961; Stocks, 1944–1947).

The risk of developing Bell's palsy is much higher in a family where one of the individuals has suffered from the disorder.

There have been few reports of a familial Bell's palsy in the literature and one gets the impression that the above condition is quite rare.

Johnson and Stoesser (1937) described a family in which two brothers had Bell's palsy. Stone (1950) reported a father and three sons suffering from Bell's palsy. Cawthorne and Haynes (1956) mentioned two brothers, one of whom had five episodes and the other three episodes of facial palsy. De Santo and Schubert (1969) reported the occurrence of 10 cases of Bell's palsy in a family, spread over a period of 83 years. Kakar *et al.* (1966) came across a family with four members suffering from Bell's palsy. The highest incidence was published by Wilbrant *et al.* (1974) where a family with 29 cases during 40 years was reviewed.

It is evident that Bell's palsy in more than one member of a family is not a simple coincidence.

It is astonishing to note the scarcity of reports published in the literature of familial Bell's palsy, while some authors have found a high frequency in their own, e.g., in 1963, Alter stated that 28.6 per cent of the 105 patients with Bell's palsy had a positive family history; Adour *et al.* (1978) reported eight per cent family history in a study of 1,000 patients, and Willbrand *et al.* (1974) reported an incidence of six per cent familial Bell's palsy.

It may be that many ENT practitioners are not aware of the hereditary factor as a possible aetiology of this condition and therefore no accurate family history is obtained. On the other hand, the episodes are simply forgotten, being of a transitory nature.

Some authors (Alter, 1963; De Santo and Schubert, 1969), stressed that the inheritance is autosomal dominant and the family reported by Willbrand *et al.* (1974) is an excellent example of this kind of inheritance.

Kakar *et al.* (1966) suggested a recessive gene with uncertain, inherited aetiological factors. He discourages marriage between individuals with a history of Bell's palsy. An inherited anatomical variation of the Fallopian canal must be taken into consideration with such a high familial incidence. De Santo and Schubert (1969) discussed this possible inherited factor as a predisposing trigger.

Surgical evidence for a proximal lesion confined between the beginning of the Fallopian canal and the geniculate ganglion was found by Fisch (1973).

When operating on patients with Bell's palsy, swelling proximal to the stylomastoid foramen has been observed (Cawthorne, 1946). The facial nerve normally occupies a maximum of 53 per cent of the cross-section of the canal (Lindeman, 1960). However, in patients with a congenitally narrowed facial canal, this percentage could be much greater.

A combination of congenital constricture of the foramen with a slightly wider nerve in an individual with a predisposition to viral infection or an ischaemia could easily cause a recurrent neuropraxia of the nerve, as seen in the case presented.

Cawthorne (1956), reporting two cases of recurrent Bell's palsy, suggested the possibility that highly cellular mastoids are prone to thermal changes and therefore, to recurrent episodes, evidence that was not supported by others (Adour *et al.*, 1978; Alter, 1963).

It is known that there is a familial tendency in Melkerson's syndrome. In this

https://doi.org/10.1017/S0022215100147826 Published online by Cambridge University Press

syndrome, the facial palsy is associated with ipsilateral oedema of the soft tissues of the face and lips. In 1931, Rosenthal described lingua plicata as a feature of the syndrome.

There is a family history of Bell's palsy in 25 per cent of patients suffering from diabetes (Cecil and Loeb, 1955).

It can be said from the literature review that familial Bell's palsy is not a negligible entity.

Conclusion:

1. The presented case of recurrent familial Bell's palsy is most unusual as a result of the multiple episodes (five times the father and four times the daughter) and the remarkable recovery each time.
2. Summarising the literature, it seems that familial Bell's palsy is not a rare condition but an overlooked one. The few reports of isolated familial Bell's palsy do not reflect the real extent of the disease, which ranged from 6 to 28.6 per cent.
3. The family history in an individual suffering from Bell's palsy is mandatory and the patient must be carefully questioned.
4. The aggregation of cases found makes heredity an important aetiological factor besides the ischaemic and viral theories.
5. Further studies will be necessary to elucidate the theories reviewed.

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https://doi.org/10.1017/S0022215100147826 Published online by Cambridge University Press

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