Saudi Journal of Ophthalmology (2015) 29, 39–47

Neuro-ophthalmology Update

Neuro-ophthalmological approach to facial nerve palsy

Joana Portelinha *, Maria Picoto Passarinho, João Marques Costa

Abstract

Facial nerve palsy is associated with significant morbidity and can have different etiologies. The most common causes are Bell's palsy, Ramsay–Hunt syndrome and trauma, including surgical trauma. Incidence varies between 17 and 35 cases per 100,000. Initial evaluation should include accurate clinical history, followed by a comprehensive investigation of the head and neck, including ophthalmological, otological, oral and neurological examination, to exclude secondary causes. Routine laboratory testing and diagnostic imaging is not indicated in patients with new-onset Bell's palsy, but should be performed in patients with risk factors, atypical cases or in any case without resolution within 4 months. Many factors are involved in determining the appropriate treatment of these patients: the underlying cause, expected duration of nerve dysfunction, anatomical manifestations, severity of symptoms and objective clinical findings. Systemic steroids should be offered to patients with new-onset Bell's palsy to increase the chance of facial nerve recovery and reduce synkinesis. Ophthalmologists play a pivotal role in the multidisciplinary team involved in the evaluation and rehabilitation of these patients. In the acute phase, the main priority should be to ensure adequate corneal protection. Treatment depends on the degree of nerve lesion and on the risk of the corneal damage based on the amount of lagophthalmos, the quality of Bell's phenomenon, the presence or absence of corneal sensitivity and the degree of lid retraction. The main therapy is intensive lubrication. Other treatments include: taping the eyelid overnight, botulinum toxin injection, tarsorrhaphy, eyelid weight implants, scleral contact lenses and palpebral spring. Once the cornea is protected, longer term planning for eyelid and facial rehabilitation may take place. Spontaneous complete recovery of Bell's palsy occurs in up to 70% of cases. Long-term complications include aberrant regeneration with synkinesis. FNP after acoustic neuroma surgery remains the most common indication for FN rehabilitation.

Keywords: Facial nerve palsy, Bell's palsy, Lagophthalmos, Acoustic neuroma, Neuro-ophthalmology

http://dx.doi.org/10.1016/j.sjopt.2014.09.009

Introduction

Facial nerve palsy (FNP) can have many different causes. It spans across all races and ages and has significant functional, psychological and social consequences.

Appropriate management is complicated by the wide spectrum of clinical presentation and disease severity. This article reviews the anatomy, the main causes and discusses acute management as well as the long-term options for long-standing FNP. The ophthalmologist plays a pivotal role in the multi-disciplinary team involved in the evaluation and rehabilitation of these patients.

Anatomy

The facial nerve (FN) may become dysfunctional anywhere along its course. The knowledge of its anatomy and origin of its branches may help the clinicians to localize the lesion.

Received 4 August 2014; accepted 9 September 2014; available online 28 September 2014.

Hospital de Egas Moniz, Centro Hospitalar Lisboa Ocidental, Lisbon, Portugal

* Corresponding author at: Rua da Junqueira n° 126, 1349-019 Lisbon, Portugal. Tel.: +351 964109571.
e-mail addresses: joportelinha@gmail.com (J. Portelinha), picoto.maria@gmail.com (M.P. Passarinho), costa.joao@mail.telepac.pt (J.M. Costa).

Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

Access this article online: www.saudiophthaljournal.com www.sciencedirect.com

It is both a motor and sensory nerve with 3 nuclei:

(1) The main motor nucleus controls the muscles of facial expression. It lies deep in the lower part of the pons. Voluntary facial movements originate in the precentral gyrus. White matter tracts pass through the internal capsule and cerebral peduncles along with other corticobulbar fibers. The portion of the nucleus that supplies the muscles of the upper part of the face receives corticonuclear fibers from both cerebral hemispheres and that of the lower half of the face receives fibres only from the contralateral cortex. Therefore, with a lesion involving the upper motor neurons, only the contralateral lower part of the face will be paralyzed (central palsy). However, with a lesion of the main motor nucleus or FN itself (lower motor neuron lesion), all the affected ipsilateral side will be paralyzed (peripheral palsy). Another separate involuntary pathway exists (extrapyramidal pathways) controlling mimetic or emotional changes in facial expression and is largely responsible for involuntary blinking.
(2) The parasympathetic nuclei are the superior salivatory nucleus, which sends fibers for salivary secretion and the lacrimal nucleus, which supplies the lacrimal gland. It receives afferent fibers from the hypothalamus for emotional responses and from the trigeminal sensory nuclei for reflex lacrimation secondary to irritation of the cornea and conjunctiva.
(3) The sensory nucleus receives taste fibers from the anterior two-thirds of the tongue.

The FN has both a motor root and a sensory/parasympathetic root (the intermediate nerve). They emerge between the pons and the medulla oblongata. They pass laterally in the posterior cranial fossa and in the cerebellopontine angle, with the vestibulocochlear nerve, and enter the internal acoustic meatus of the temporal bone, where it traverses the fallopian canal. The fallopian canal has 3 portions: the labyrinthine, the tympanic and the mastoidal. The geniculate ganglion is located between the labyrinthine and the tympanic portion. It is an important anatomical landmark since the great superficial petrosal nerve (responsible for lacrimal secretion) and the small petrosal nerve (which carries secretory fibers to the parotid gland) emerge from it. Therefore, FN lesions above the geniculate ganglion classically cause more severe ophthalmic symptoms because lacrimal secretion and orbicularis closure are involved. On the other hand, the nerve to the stapedius muscle and the chorda tympani (responsible for taste sensation from the anterior two-thirds of the tongue and salivary secretion) branch out at the mastoid segment. The main branch of the FN exists through the stylomastoid foramen. It runs through the parotid gland to innervate the facial musculature through five terminal branches: temporal, zygomatic, buccal, mandibular and cervical.

Epidemiology

Incidence of FNP varies between 17 and 35 cases per 100,000.[1,2] There is no sexual preponderance.[1]

The incidence in neonates varies from 0.6 to 1.8 per 1000 live births, 91% due to forceps delivery.[1]

Bell's palsy (BP) is the most common disorder and affects 11–40 persons per 100,000 each year, with peak incidence between 15 and 50 years old.[3] In pregnancy, especially during the third trimester and early postpartum, there is a 3-times greater incidence.[1,3]

Ramsay–Hunt syndrome (RHS), one main cause of FNP, presents in only 0.2% of all Varicella Zoster Virus (VZV) cases.[4]

Etiology (Table 1)

Etiology of FNP varies according to the published series. As reported by Rahman (2007), the most common causes include BP (51%), trauma (22%) and RHS (7%).[1] Peitersen (2002) found 38 different etiologies of peripheral FNP (1701 cases of BP, 116 RHS, 76 diabetic, 46 pregnant and 169 neonates).[5] According to Hohman (2014), BP accounted for 38% of cases, acoustic neuroma resections 10%, cancer 7%, iatrogenic injuries 7%, RHS 7%, benign lesions 5%, congenital palsy 5%, Lyme disease 4%, and other causes 17%.[6] One analysis of 40 pediatric patients with peripheral FNP found 65% of BP, 37.5% infection, 2.5% tumor lesion and 2.5% suspected chemotherapy toxicity.[7]

FNP can occur with supranuclear, nuclear or infranuclear lesions and may be grouped into idiopathic (1), infectious (2), traumatic (3) and neoplastic (4) (Table 1).[8,9]

Supranuclear lesions may be caused by a lesion in the motor cortex, the subcortex or corticobulbar tracts. Commonly, the etiology is vascular, but may be demyelinating or tumoral.

Lower motor neuron lesions can be categorized anatomically[10]:

(1) Nuclear: Tumoral, inflammatory or ischemic pathology. It is usually associated with ipsilateral 6th nerve palsy and may also affect the descending corticospinal tracts causing contralateral limb weakness (*Millard–Gubler* syndrome).
(2) Cerebellopontine angle: Its contents include the CN V superiorly, the CN IX and X inferiorly and the CN VII and VIII in between. One of the first signs of this syndrome is the loss of corneal reflex on the ipsilateral side. Usually caused by an acoustic neuroma, it can also be caused by meningiomas, metastases, cholesteatomas or aneurysms. It is suspected in the case of impairment associated with CN VIII (deafness, vertigo, hyperacusis, tinnitus).
(3) Facial canal: The proximal part of the canal is particularly prone to ischemia and compression. BP, fractures of the temporal bone, malignant otitis externa or suppurative otitis media, RHS and neoplastic processes can affect the FN here.
(4) Parotid: A parotid mass with FNP is in general malignant. Other etiologies include inflammatory parotitis from infection or granulomatous conditions (sarcoidosis).

Pathophysiology

Idiopathic

BP is an acute paralysis of one side of the face of unknown etiology, which remains a diagnosis of exclusion.[11]