Cranial Nerves of Facial Asymmetry

Subjects: Neuroimaging

Contributor: Justyna Chojdak-Łukasiewicz ,

Symmetry of the face is the one of the most important features for the perception of attractiveness. The word “symmetry” derives from Greek and comes from “syn” (together) and “metron” (meter). Symmetry means that both sides of the face, right and left, are alike. The term “asymmetry” refers to dissimilarity between components, altering the balance between structures. Cranial nerve damage, especially the affection of oculomotor, trochlear, trigeminal, and facial nerves, may occur in many neurological conditions. The most common acquired causes of cranial nerve damage are cerebrovascular events, such as ischemic or haemorrhagic stroke.

facial asymmetry nerves palsy

1. Nerve III (Oculomotor Nerve)

The third cranial nerve is a mixed nerve and provides motor, parasympathetic, and sympathetic fibres. The oculomotor nerve innervates the extraocular muscles (levator palpebrae superioris, superior rectus, inferior rectus, medial rectus, and inferior oblique), which are responsible for eyeball and upper eyelid movements [1]. Injury to the oculomotor nerve is characterised by ptosis (a drooping upper eyelid) secondary to paralysis of the levator palpabrae superioris and unopposed activity of the orbicularis oculi muscle [2]. Ptosis leads to a characteristic “sleepy” appearance and asymmetry in both unilateral and bilateral cases. The most common causes of third nerve palsy are presumed to be microvascular trauma and compression resulting from a neoplasm or an aneurysm [3][4][5].

2. Nerve IV (Trochlear Nerve)

The trochlear nerve is a motor nerve responsible, together with the oculomotor nerve and the abducens nerve, for eye movement. It is the smallest of the cranial nerves and has the longest course. The trochlear nerve innervates the superior oblique muscle and controls the abduction and intorsion of the eye [6]. Palsy of the trochlear nerve is characterised by vertical diplopia exacerbated when looking downwards and inwards. A patient can also develop compensatory head positioning, tilting away from the affected side. The three most common causes of trochlear palsy are trauma (especially to the occiput), ischemic or vascular damage (microvascular lesions associated with hypertension, diabetes mellitus, or vasculitis), and congenital disorders [7].

Congenital trochlear palsy typically presents with a long-standing compensatory head tilt and facial asymmetry from the head tilt. Congenital superior oblique palsy (SOP) is one of the most common causes of ocular torticollis in children, leading to asymmetric development of the face [8]. Congenital palsy of the trochlear nerve may be

sporadic or genetic. This familiar condition is a rare, autosomal-dominant inherited disease characterised by congenital fourth cranial nerve palsy, facial asymmetry, and superior oblique tendon abnormalities (absence, redundance, misdirection) [9]. Children with torticollis tilt their heads to use both eyes together, a strategy that is associated with progressive facial asymmetry. The symptoms of superior oblique palsy are hypertropia in the affected eye, an abnormal head position, and excyclotorsion [10]. Deviation of the nose and mouth toward the hypoplastic side can be observed.

3. Nerve V (Trigeminal Nerve)

The trigeminal nerve is a mixed nerve and consists of a sensory and a motor part. The three sensory branches innervate the face, mucous membranes, and sinuses. The third branch (mandibular) of the trigeminal nerves has motor fibres. It innervates the muscles of mastication (medial pterygoid, lateral pterygoid, masseter, and temporalis) and the anterior belly of the digastric, mylohyoid, tensor veli palatini, and tensor tympani. Pure unilateral trigeminal motor neuropathy (UTMN) is a rare condition resulting from paralysis of the motor branch of the trigeminal nerve without sensory disturbances or lesion of other cranial nerves [11]. The first description was published in 1988 by Chia et al. [12]. The most common postulated aetiological factors of UTMN include viral infection, tumours, head trauma (skull base fracture of the oval foramen or pontine haemorrhage), autoimmune-inflammatory causes (e.g., Sjögren's syndrome), and factors of vascular origin. In some cases, the condition is idiopathic [11][13][14][15]. A common clinical presentation is unilateral asymmetry of the face preceded by a feeling of wasting and weakness of the masticatory muscles. In addition, chewing problems and deviation of the jaw can be observed. The typical finding from magnetic resonance imaging (MRI) of the brain is replacement of the muscle by fat tissue [16].

4. Nerve VII (Facial Nerve)

The facial nerve is a mixed nerve containing both motor and sensory components [17]. The nerves emerge from the pons and carry motor fibres, which control all the muscles of the face. Anatomically, the upper part of the face (the eyes and forehead) is innervated from both hemispheres, while the lower part of the face receives innervations from the contralateral hemisphere [18]. Symptoms of facial nerve palsy depend on the location of injury. Supranuclear lesions affect upper motor neurons and manifest as central facial nerve palsy. Intranuclear locations of lesions, distal to the facial nerve nucleus, display signs of peripheral facial nerve palsy [19]. Disorders of the facial nerve are relatively common and have many possible causes, each resulting in varying levels of facial asymmetry due to paralysis of the structures innervated by the facial nerve. Congenital facial palsy is present at birth and can be caused by abnormal developmental processes, disturbances of the neuromuscular junction (congenital myasthenic syndrome 9–11), myopathies, or trauma (resulting from perinatal injury, especially the use of forceps during delivery). Acquired facial nerve palsy is a result of infection, neoplasms, or neurodegeneration [18].

Bell's Palsy

The most common acute neurological cause of facial asymmetry is Bell's palsy. This is the most frequent condition of "peripheral lesion" of the facial nerve and accounts for about two-thirds of unifacial nerve disorders. The annual incidence rate ranges from 13 to 34 cases per 100,000 with no differences between sexes [20]. The aetiology is unknown, but in some cases it is caused by a viral infection (especially herpes simplex virus type 1 (HSV-1)) [21]. Other possible causes of Bell's palsy are anatomical, ischemic, or inflammatory in nature or due to cold stimulation [21]. Bell's palsy is characterised by specific clinical features: weakness involving the mouth, eye, and forehead. The symptoms often are accompanied by earache, dysgeusia, or altered facial sensation. The symptoms develop within hours to days. Steroids are recommended as a therapeutic tool. It is a benign condition, with complete or near-complete recovery in the majority of cases [19].

Central Palsy

Central facial palsy occurs as a result of damage to the cortico-nuclear tract above the nucleus of Nerve VII. The clinical manifestation includes weakness in the lower part of the face, problems with smiling, and flattening of the nasolabial fold. The most common causes of central palsy are stroke, multiple sclerosis, and tumours [22]. Stroke-related oro-facial impairment depends on the location and size of vascular lesions. Cortical lesions affecting the precentral gyrus are connected to contralateral impairment of the motor and sensory functions of the face. A stroke in the middle cerebral artery (MCA) region manifests with multiple symptoms, depending on which branches and structures are affected [23]. Isolated central facial palsy has been observed in the case of lacunar stroke located in the corona radiata or pons [24][25].

Moebius Syndrome

Moebius syndrome (MS), sometimes called the Moebius sequence, is a congenital, non-progressive uni- or bilateral facial nerve palsy associated with dysfunction of other cranial nerves, especially nerve abducens [26][27]. Both the aetiology and pathology are unknown. Some theories of the underlying intrauterine environmental factors and genetic causes have been considered. The prevalence of MS is estimated to be 1:250,000 newborns, with an equal incidence between women and men [28]. The characteristic clinical feature is secondary to the failure of the abducens and the facial nerves. Facial asymmetry and dysmorphic symptoms (microencephaly, epicanthal folds, flat nasal bridge, micrognathia, defects of the external ear, dental deficits, clinodactyly low-set thumbs) are typical features of Moebius syndrome [29][30].

Ramsay Hunt Syndrome

Ramsay Hunt syndrome (or herpes zoster oticus) is a peripheral facial nerve palsy related to a vesicular rash in the external auditory canal and the mouth [31][32]. This is the second most common atraumatic lesion of the nerve palsy. Its incidence is about 5:100,000 with a predominance in women. The condition is connected to reactivation of a latent infection of the Varicella zoster virus (VZV) in the geniculate ganglion of the facial nerve [33][34]. According to anatomical associations with abducens, vestibulocochlear, or glossopharyngeal nerves or cervical spinal nerves, other clinical features can be present, such as tinnitus, hearing loss, nausea, vomiting, vertigo, and nystagmus [34][35].

Melkersson–Rosenthal Syndrome

Melkersson–Rosenthal syndrome is a rare condition characterised by three symptoms: recurrent peripheral facial nerve palsy (uni- or bilateral), recurrent or persistent oedema of facial soft tissue, and lingua plicata (scrotal tongue) [36][37]. The symptoms may appear singly or together. The aetiology is unknown, and various factors, including genetic, inflammatory, allergic, and microbial factors, are involved. In some cases, the syndrome is genetically determined, inherited autosomal dominant, and associated with a defect in chromosome 9p11 [38]. The disease affects people in the second and third decades of life [39].

Hemifacial Spasm

Hemifacial spasm (HFS) is a neuromuscular disorder manifesting in unilateral, involuntary contractions of the facial muscles of one side of the face, innervated by the ipsilateral facial nerve [40][41]. The estimated prevalence is about 11 cases per 10,000, with a predominance in women. HFS is caused by irritation of or damage to the facial nerve of varying aetiology [42]. The most common cause is vascular compression of the facial nerve root in the brainstem. The neurovascular compression is produced by the anterior inferior arteries (AICAs), posterior inferior cerebellar arteries (PICAs), and the vertebral arteries (VAs). A hemifacial spasm can be secondary to trauma, demyelinating lesion, stroke, or mastoid or ear infection [43]. The clinical features include irregular muscle contractions in the area of the lower facial muscles, such as the orbicularis oris, mentalis, zygomaticus major, and platysma [40].

Miller Fisher Syndrome

Miller Fisher Syndrome (MFS) is an immune-related polyneuropathy, a rare variant of Guillain–Barre syndrome [44]. The estimated annual incidence is 1:1,000,000. Common clinical features include the following three symptoms: gait ataxia, areflexia, and ophthalmoplegia [45][46]. Facial nerve palsy appears in half of all patients with MFS, typically in the early phase of the disease [47]. In 85% to 90% of MFS cases, antibodies against GQ1b (a ganglioside component of the nerve) are present [48].

References

1. Finsterer, J. Ptosis: Causes, Presentation, and Management. Aesthetic Plast. Surg. 2003, 27, 193–204.
2. Bacharach, J.; Lee, W.W.; Harrison, A.R.; Freddo, T.F. A review of acquired blepharoptosis: Prevalence, diagnosis, and current treatment options. Eye 2021, 35, 2468–2481.
3. Kim, T.; Nam, K.; Kwon, B.S. Isolated Oculomotor Nerve Palsy in Mild Traumatic Brain Injury. Am. J. Phys. Med. Rehab. 2020, 99, 430–435.
4. Kim, K.; Noh, S.R.; Kang, M.S.; Jin, K.H. Clinical Course and Prognostic Factors of Acquired Third, Fourth, and Sixth Cranial Nerve Palsy in Korean Patients. Korean J. Ophthalmol. 2018, 32,

221.

5. Kung, N.; Van Stavern, G. Isolated Ocular Motor Nerve Palsies. Semin. Neurol. 2015, 35, 539–548.
6. Laine, F.J. Cranial Nerves III, IV, and VI. Top. Magn. Reson Imaging 1996, 8, 111.
7. Morillon, P.; Bremner, F. Trochlear nerve palsy. Br. J. Hosp. Med. 2017, 78, 38–40.
8. Akbari, M.R.; Nejad, M.K.; Askarizadeh, F.; Pour, F.F.; Pazooki, M.R.; Moeinitabar, M.R. Facial asymmetry in ocular torticollis. J. Curr. Ophthalmol. 2015, 27, 4–11.
9. Harris, D.J.; Memmen, J.E.; Katz, N.N.K.; Parks, M.M. Familial Congenital Superior Oblique Palsy. Ophthalmology 1986, 93, 88–90.
10. Tollefson, M.M.; Mohney, B.G.; Diehl, N.N.; Burke, J.P. Incidence and Types of Childhood Hypertropia: A Population-Based Study. Ophthalmology 2006, 113, 1142–1145.
11. Kang, Y.-K.; Lee, E.-H.; Hwang, M. Pure trigeminal motor neuropathy: A case report. Arch. Phys. Med. Rehab. 2000, 81, 995–998.
12. Chia, L.-G. Pure trigeminal motor neuropathy. BMJ 1988, 296, 609–610.
13. Wilson, M.; Hodgson, E.; Felstead, A. Focal atrophy of the masticatory muscles caused by pure trigeminal motor neuropathy: Case report. Br. J. Oral Maxillofac. Surg. 2016, 54, e13–e14.
14. Braun, J.S.; Hahn, K.; Bauknecht, H.-C.; Schielke, E. Progressive Facial Asymmetry due to Trigeminal Motor Neuropathy. Eur. Neurol. 2006, 55, 96–98.
15. Andonopoulos, A.P.; Lagos, G.; Drosos, A.A.; Moutsopoulos, H.M. The Spectrum of Neurological Involvement in Sjögren’s Syndrome. Rheumatology 1990, 29, 21–24.
16. Kämppi, A.; Kämppi, L.; Kemppainen, P.; Kanerva, M.; Toppila, J.; Auranen, M. Focal atrophy of the unilateral masticatory muscles caused by pure trigeminal motor neuropathy: Case report. Clin. Case Rep. 2018, 6, 939–943.
17. Takezawa, K.; Townsend, G.; Ghabriel, M. The facial nerve: Anatomy and associated disorders for oral health professionals. Odontology 2018, 106, 103–116.
18. Williams, O.; Ulane, C. Facial Nerve (Cranial Nerve VII). In Encyclopedia of the Neurological Sciences; Elsevier: Amsterdam, The Netherlands, 2014; pp. 263–268. Available online: https://linkinghub.elsevier.com/retrieve/pii/B9780123851574006576 (accessed on 28 November 2021).
19. George, E.; Richie, M.B.; Glastonbury, C.M. Facial Nerve Palsy: Clinical Practice and Cognitive Errors. Am. J. Med. 2020, 133, 1039–1044.
20. Myers, E.N.; De Diego, J.I.; Prim, M.P.; Madero, R.; Gavil$Aan, J. Seasonal Patterns of Idiopathic Facial Paralysis: A 16-Year Study. Otolaryngol. Neck Surg. 1999, 120, 269–271.

21. Zhang, W.; Xu, L.; Luo, T.; Wu, F.; Zhao, B.; Li, X. The etiology of Bell's palsy: A review. J. Neurol. 2020, 267, 1896–1905.
22. Ahdab, R.; Saade, H.; Kikano, R.; Ferzli, J.; Tarcha, W.; Riachi, N. Pure ipsilateral central facial palsy and contralateral hemiparesis secondary to ventro-medial medullary stroke. J. Neurol. Sci. 2013, 332, 154–155.
23. Schimmel, M.; Ono, T.; Lam, O.L.T.; Müller, F. Oro-facial impairment in stroke patients. J. Oral Rehabil. 2017, 44, 313–326.
24. Sands, K.A.; Shahripour, R.B.; Kumar, G.; Barlinn, K.; Lyerly, M.J.; Haršány, M.; Cure, J.; Yakov, Y.L.; Alexandrov, A.W.; Alexandrov, A.V. Acute Isolated Central Facial Palsy as Manifestation of Middle Cerebral Artery Ischemia: Isolated Central Facial Palsy with MCA Ischemia. J. Neuroimaging 2016, 26, 499–502.
25. Wolf, M.E.; Rausch, H.-W.; Eisele, P.; Habich, S.; Platten, M.; Alonso, A. Acute Corticonuclear Tract Ischemic Stroke with Isolated Central Facial Palsy. J. Stroke Cerebrovasc. Dis. 2019, 28, 495–498.
26. Picciolini, O.; Porro, M.; Cattaneo, E.; Castelletti, S.; Masera, G.; Mosca, F.; Bedeschi, M.F. Moebius syndrome: Clinical features, diagnosis, management and early intervention. Ital. J. Pediatr. 2016, 42, 56.
27. Rucker, J.C.; Webb, B.D.; Frempong, T.; Gaspar, H.; Naidich, T.P.; Jabs, E.W. Characterization of ocular motor deficits in congenital facial weakness: Moebius and related syndromes. Brain 2014, 137, 1068–1079.
28. Kulkarni, A.; Madhavi, M.R.; Nagasudha, M.; Bhavi, S. A rare case of Moebius sequence. Indian J. Ophthalmol. 2012, 60, 558–560.
29. Ali, M.H.; Jamal, S.; Rashid, M.A.; Javaid, U.; Butt, N.H. Moebius Syndrome with Hypoglossal Palsy, Syndactyly, Brachydactyly, and Anisometropic Amblyopia. Cureus 2018, 10, e2334.
30. Domeshek, L.F.; Zuker, R.M.; Borschel, G.H. Management of Bilateral Facial Palsy. Otolaryngol. Clin. N. Am. 2018, 51, 1213–1226.
31. Ananthapadmanabhan, S.; Soodin, D.; Sritharan, N.; Sivapathasingam, V. Ramsay Hunt syndrome with multiple cranial neuropathy: A literature review. Eur. Arch. Oto-Rhino-Laryngol. 2021, 1–6. Available online: https://link.springer.com/10.1007/s00405-021-07136-2 (accessed on 1 November 2021).
32. Jeon, Y.; Lee, H. Ramsay Hunt syndrome. J. Dent. Anesth. Pain Med. 2018, 18, 333.
33. Sweeney, C.J. Nosological Entities?: Ramsay Hunt syndrome. J. Neurol. Neurosurg. Psychiatry 2001, 71, 149–154.

34. Crouch, A.E.; Andaloro, C. Ramsay Hunt Syndrome. In StatPearls; StatPearls Publishing: Treasure Island, FL, USA, 2021. Available online: http://www.ncbi.nlm.nih.gov/books/NBK557409/ (accessed on 22 November 2021).
35. Ostwal, S.; Salins, N.; Deodhar, J.; Muckaden, M. Management of ramsay hunt syndrome in an acute palliative care setting. Indian J. Palliat. Care 2015, 21, 79–81.
36. Casper, J.; Mohammad-Khani, S.; Schmidt, J.J.; Kielstein, J.T.; Lenarz, T.; Haller, H.; Wagner, A.D. Melkersson–Rosenthal syndrome in the context of sarcoidosis: A case report. J. Med. Case Rep. 2021, 15, 1–6.
37. Jamil, R.T.; Agrawal, M.; Gharbi, A.; Sonthalia, S. Cheilitis Granulomatosa. In StatPearls; StatPearls Publishing: Treasure Island, FL, USA, 2021. Available online: http://www.ncbi.nlm.nih.gov/books/NBK470396/ (accessed on 22 November 2021).
38. Scully, C.; Langdon, J.; Evans, J. Marathon of eponyms: 13 Melkersson-Rosenthal syndrome: Marathon of eponyms. Oral Dis. 2010, 16, 707–708.
39. Ziem, P.E.; Pfrommer, C.; Goerdt, S.; Orfanos, C.E.; Blume-Peytavi, U. Melkersson-Rosenthal syndrome in childhood: A challenge in differential diagnosis and treatment: Melkersson-Rosenthal Syndrome in Childhood. Br. J. Dermatol. 2000, 143, 860–863.
40. Vaughan, C.L.; Goetz, C.G. Hemifacial Spasm. In Encyclopedia of the Neurological Sciences; Elsevier: Amsterdam, The Netherlands, 2014; p. 545. Available online: https://linkinghub.elsevier.com/retrieve/pii/B9780123851574000221 (accessed on 1 November 2021).
41. Chopade, T.R.; Bollu, P.C. Hemifacial Spasm. In StatPearls; StatPearls Publishing: Treasure Island, FL, USA, 2021. Available online: http://www.ncbi.nlm.nih.gov/books/NBK526108/ (accessed on 22 November 2021).
42. Lefaucheur, J.-P.; Ben Daamer, N.; Sangla, S.; Le Guerinel, C. Diagnosis of primary hemifacial spasm. Neurochirurgie 2018, 64, 82–86.
43. Hermier, M. Imaging of hemifacial spasm. Neurochirurgie 2018, 64, 117–123.
44. Rocha Cabrero, F.; Morrison, E.H. Miller Fisher Syndrome. In StatPearls; StatPearls Publishing: Treasure Island, FL, USA, 2021. Available online: http://www.ncbi.nlm.nih.gov/books/NBK507717/ (accessed on 23 November 2021).
45. Berlit, P.; Rakicky, J. The Miller Fisher syndrome. Review of the literature. J. Clin. Neuroophthalmol. 1992, 12, 57–63.
46. Al Othman, B.; Raabe, J.; Kini, A.; Lee, A.G. Update: The Miller Fisher variants of Guillain–Barré syndrome. Curr. Opin. Ophthalmol. 2019, 30, 462–466.

47. Tan, C.-Y.; Yuki, N.; Shahrizaila, N. Delayed facial palsy in Miller Fisher syndrome. J. Neurol. Sci. 2015, 358, 409–412.
48. Willison, H.J.; Veitch, J.; Paterson, G.; Kennedy, P.G. Miller Fisher syndrome is associated with serum antibodies to GQ1b ganglioside. J. Neurol. Neurosurg. Psychiatry 1993, 56, 204–206.