Facial Palsy

Techniques for Reanimation of the Paralyzed Face

Chieh-Han John Tzou
Andrés Rodríguez-Lorenzo
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Preface

> "The facial expressions of human beings fascinate me because they convey both the lowest, most bestial pleasures and the strongest and gentlest emotions of the spirit."—Sir Charles Bells (1774–1842)

Facial paralysis is a devastating condition, which substantially affects a patient's quality of life. The face is one of the most visible parts of the body, and facial expressions are critical in human communication, social interaction, and self-esteem. Injuries to the facial nerve have important functional consequences, such as corneal damage from the impariment of blinking and the derangement of speaking and eating from the loss of oral continence.

Facial paralysis is a fascinating and dynamic field, full of constant innovations, with a vast variety of surgical approaches and techniques. Reanimation of the paralyzed face is considered by many to be one of the most challenging endeavors in reconstructive surgery, as it requires knowledge of multiple complex surgical techniques and disciplines, such as peripheral nerve surgery, microvascular surgery, and aesthetic facial surgery. As reconstructive surgeons with interest in facial palsy, we are passionate and enthusiastic about new technical innovations and surgical approaches to improve the quality of life of patients with paralyzed faces. Success in providing the best clinical outcomes in the rapidly changing field of facial reanimation relies on multidisciplinary collaborations. In this book, we wanted to honor our precious multidisciplinary collaborations and bring up all aspects of therapies by including experts from various disciplines and specialties.

This work, *Facial Palsy: Techniques for Reanimation of the Paralyzed Face,* is a collaborative effort among international experts aiming to provide the most comprehensive and detailed overview of the most pertinent surgical procedures, including a wide spectrum of innovative and cutting-edge approaches and an up-to-date guide to every aspect of reanimating the paralyzed face.

This book is structured in six parts and 38 chapters. The first part focuses on general principles in facial paralysis, including diagnosis, nonsurgical treatments, and documentation of patient outcomes. The second part is related to facial nerve anatomy and reconstruction techniques, including nerve repair, nerve grafts, vascularized nerves, and nerve transfers. The third and fourth parts address the surgical management of long-standing facial palsy, including smile reanimation techniques and rehabilitation of the paretic eye. The fifth part relates to symmetrization of the paralyzed face and ancillary proce-

dures. The sixth and final part focuses on current new frontiers and innovations in reconstructing the paralyzed face.

We are deeply grateful to all authors who devoted an enormous amount of time to this comprehensive coverage of current state-of-the-art knowledge. Your time is a most precious gift—to this book, to the medical society, and to our patients—a gift that can never be returned. Thank you! Moreover, we heartily thank the Springer Nature team—von Behrens Inga, Sasirekka Nijanthan, Martina Himberger, and Daniela Heller—for their continuous professional assistance throughout this project.

Andrés: I would like to dedicate this book to my mentors throughout my career: my first chief and mentor, Dr. Francisco Martelo, for introducing me to plastic surgery; Dr. Pedro Cavadas for showing me what is possible in reconstructive microsurgery; Professor Fu-Chan Wei, for your example of leadership and excellence; and Professor David Chuang, for encouraging me and inspiring me to go into the fascinating field of facial paralysis. I also dedicate this book to my wonderful wife, Debora, and my daughters, Nadia and Olivia, for your endless love, support, and patience. You are the biggest success of my life.

John: I dedicate this book and offer my deepest gratitude to my mentors: Professor Manfred Frey, who introduced me to his beloved field of the reanimation of the paralyzed face; Professor Rafael Leopold Walzer, for showing me amazing and exciting insights in your trick box of plastic and reconstructive microsurgery; Professor Thomas Rath, for globally broadening my view of plastic surgery; Professor Fu-Chan Wei, for your care, leadership, and excellence; and Professor David Chuang, for your amazing inspirations to think outside the box. Moreover, I dedicate this book to my family for your unfailingly support, endless love, and everlasting belief in me.

Last but not least, we would like to express our most sincere gratitude to all patients who have made this book possible by sharing and permitting the publications of photographs related to their treatments. You are a true source of inspiration for all of us.

It is an honor to serve you.

With us, reconstructive surgery has not been a purpose, but a passion.

Uppsala, Sweden — Andrés Rodríguez-Lorenzo
Vienna, Austria — Chieh-Han John Tzou
2020

Contents

About the Editors

Chieh-Han John Tzou completed his medical education and plastic surgery residency at the Medical University of Vienna, Austria, followed by a 1½-year microvascular fellowship at the Chang Gung Memorial University Hospital in Taiwan. Currently, he is the director of the Plastic Reconstructive Surgery Clinic of the Hospital of the Divine Savior and TZOU MEDICAL, Facial Palsy and Lymphedema Center, and is affiliated as an Associate Professor for Plastic and Reconstructive Surgery at the Medical Faculty of the Sigmund Freud University, Vienna, Austria. His focus in clinical work and research is microsurgical and supermicrosurgical reconstruction in facial paralysis, lymphedema, nerve, and tumor patients. He is the co-director of the biennial course "Facial Palsy: A Hands-on Course—www.facialpalsy.eu" in Vienna, Austria.

Andrés Rodríguez-Lorenzo completed his plastic surgery residency and PhD studies at the University Hospital of A Coruña, Spain, followed by a 2-year microvascular fellowship at the Canniesburn Plastic Surgery Unit in Glasgow and the Chang Gung Memorial University Hospital in Taiwan. Currently, he is the Chief and Associate Professor in Plastic Surgery and the program director of the International Fellowship Program in Microsurgery at the Uppsala University Hospital, leading one of the most active microsurgical centers in Scandinavia. His main research focus is head and neck reconstruction, including preclinical studies of face transplantation, virtual planning, and facial paralysis. He is the co-director of the biennial course "Facial Palsy: A Hands-on Course—www.facialpalsy.eu" in Vienna, Austria, and the biannual Scandinavian Facial Nerve Meeting in Uppsala, Sweden.

Part I

Facial Paralysis

1 Facial Paralysis: Etiology, Diagnosis, and Medical Treatment

Lars Jonsson

1.1 The Facial Nerve

The main portion of the facial nerve, cranial nerve VII (CN VII), consists of motor fibers innervating the facial musculature for voluntary and involuntary emotional movements. This is its most important function. The nerve also carries motor fibers to the stapedius, posterior auricular, occipital belly of occipitofrontalis, posterior belly of the digastric and stylohyoid muscles. It also comprises parasympathetic and sensory components that are carried by the intermediate nerve of Wrisberg. The long and inextricable motor pathway of the facial nerve from the cortical region through the brain, brain stem, temporal bone, and parotid gland to the facial muscles makes it susceptible to injury and disease. Depending on the location of injury and origin of disease, facial nerve disorders will present differently in the clinic. Knowledge of nerve anatomy as well as clinical signs of diseases affecting the nerve is thus essential in diagnosing and treating facial nerve dysfunction. This chapter reviews the more common causes of facial paralysis and discusses relevant early medical treatment strategies.

1.2 Facial Nerve Pathways

The control of facial muscle movements has a complicated and spread central neural representation. Movements are dually controlled, voluntary and/or involuntary emotional. There are at least five bilateral cortical representations that project to the four paired facial subnuclei in the pons—these subnuclei selectively innervate various facial muscle groups. The primary motor cortex (M1) and the ventral lateral premotor cortex on the lateral side and the supplementary motor region (M2) on the medial aspect of each hemisphere are important for the facial voluntary control. The rostral (M3) and caudal cingulate cortex (M4) on the medial side of the hemispheres receive information from the limbic system and are essential for involuntary emotional facial movements, for overview see Müri [1]. The central innervation of the upper facial muscles is bilateral while the regulation of the lower part of the face mainly is contralateral. One vital clinical sign is therefore the preservation of forehead and eye closure function in a central (contralateral) lesion contrary to the reduced tone and weakness seen in these muscles on the affected side of a peripheral facial lesion (Figs. 1.1 and 1.2). A central (supranuclear) facial injury is often accompanied by tongue weakness and hemiparesis starting with thumb, finger, and hand movements on the ipsilateral side to facial paralysis due to the close relation of these motor centers in

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Fig. 1.1 Central facial paralysis on the right side. Central lesion on the left side and contralateral (right) lower facial musculature affected (drooping corner of the mouth and less pronounced nasolabial fold). Upper facial musculature (eye and forehead) unaffected

Fig. 1.2 Left-sided peripheral facial paralysis. Ipsilateral lower and upper facial musculature affected with drooping corner of the mouth, less pronounced nasolabial fold, lagophthalmos/wide eye and less pronounced forehead wrinkles. Published with oral and written permission from the patient

the cortex and internal capsule. Signs and appearance of central and peripheral facial palsy (Bell's palsy) are illustrated in Table 1.1. From a clinical viewpoint, the disparity of the central pathways between the voluntary corticofacial projections and involuntary emotional projections explains why emotional responses are intact (except for cases of severe unilateral brain damage) in upper motor neuron (supranuclear) facial paralysis affecting the corticobulbar tract responsible for voluntary facial movements.

A lesion affecting the pontine facial motor nucleus and/or the facial nerve peripheral to the nucleus usually affects both the upper and lower facial muscles. If the peripheral facial palsy originates from a central lesion of the nucleus or fascicle, other signs from nearby structures most often will be present. The palsy is usually accompanied by other nearby neurological signs with gaze palsy/internal strabismus on the side of facial paralysis, plus contralateral hemiparesis, ataxia, and cerebellovestibular signs [2]. Preservation of the forehead, which is a sign of a central (supranuclear) lesion, may, however, appear with a selective lesion in the nucleus as well as intra- or extratemporal portion of the facial nerve.

Before the facial nerve emerges from the pons, fibers loop around the nuclei of CN VI (abducens nerve). The nerve accompanies the CN VIII (vestibulocochlear nerve) in the cisternal and auditory internal meatus pathways. After entering its own bony canal (the fallopian canal in the temporal bone), the nerve makes two bends and divides in labyrinthine, horizontal tympanic, and vertical mastoid segments before leaving the skull base through the stylomastoid foramen. The labyrinthine portion ends at the geniculate ganglion where the nerve makes its first external bend. At this point, the greater petrosal nerve exits with parasympathetic motor fibers to the sphenopalatine ganglion and supply the lacrimal, nasal and palatinal glands. In the horizontal tympanic portion, a second nerve branch exits for the stapedial muscle, which protects the inner ear from loud noise. In the vertical mastoid segment, visceral branches forming the chorda tympany provide submandibular and sublingual gland innervation as well as taste to the anterior two-thirds of the tongue (Fig. 1.3).

Table 1.1 Characteristics of central and peripheral facial palsy (Bell's palsy)

	Central	Peripheral facial palsy
Forehead wrinkle	Normal	Weakened/flaccid
Eye closure	Normal	Lagophthalmos/wide eye
Nasolabial fold/oral commissure	Less pronounced/drooping	Less pronounced/drooping
Onset	Rapid (seconds to minutes)	Hours to 2–3 days
Associated symptoms	Weakness arm/leg, slurred speech, double vision, ataxia vertigo (cerebellovestibular symptoms), problems swallowing	Periauricular and/or neck pain, hyperacusis, taste disturbance, reduced tearing
Age (years)	> 60	20–60

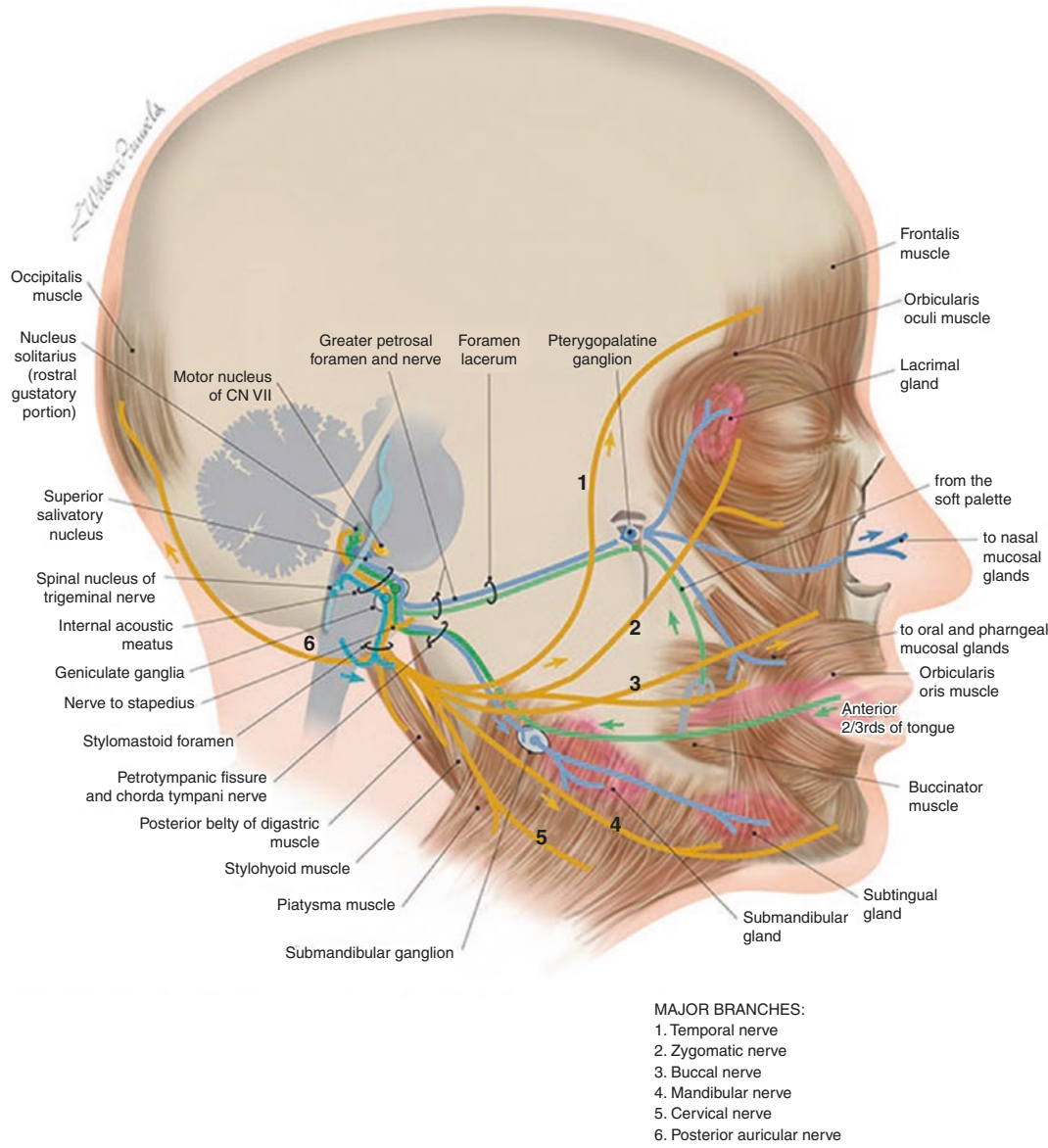

Fig. 1.3 Overview of the facial nerve components (parotid gland removed). (By permission from Cranial Nerves 3rd Ed. © 2010 Wilson-Pauwels, Stewart, Akesson, Spacey, PMPH-USA)

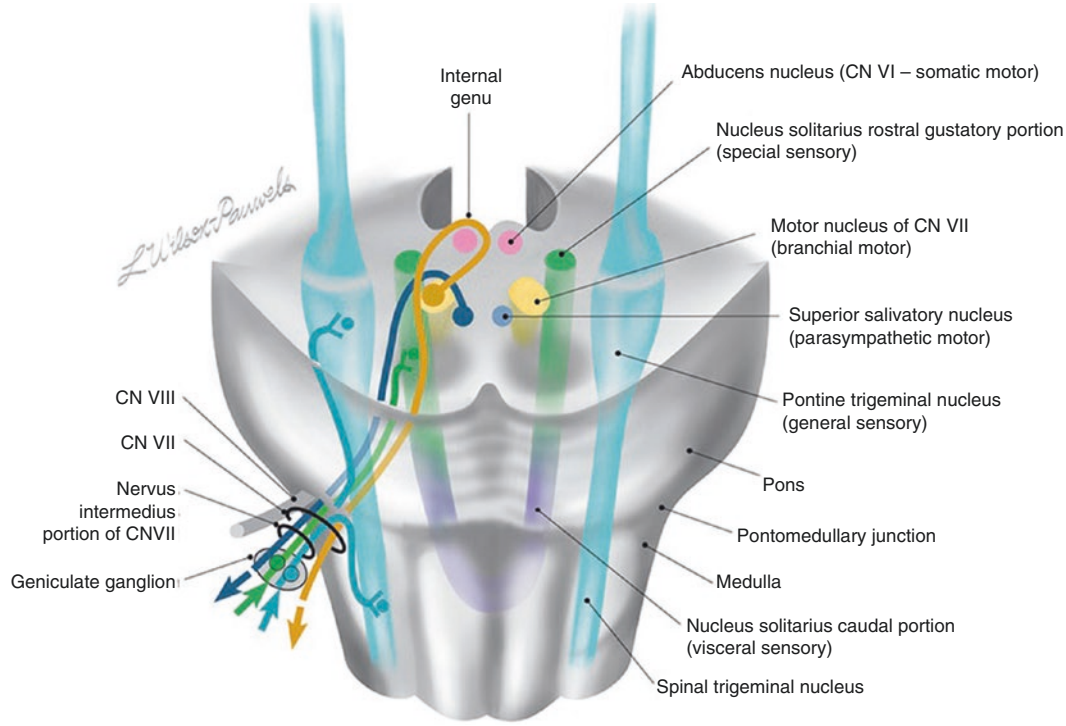

Fig. 1.4 Facial nerve motor, parasympathetic and sensory nuclei in the brain stem. (By permission from Cranial Nerves 3rd Ed. © 2010 Wilson-Pauwels, Stewart, Akesson, Spacey, PMPH-USA)

In the internal auditory canal, the facial nerve is accompanied by the intermediate nerve of Wrisberg, which contains sensory and parasympathetic fibers. Sensation of the external auditory canal, pinna, mastoid, and palate mucosa is carried by afferent fibers, as is taste in the anterior two-thirds of the tongue. The parasympathetic fibers emerge in the superior salivatory nucleus while taste fibers end in the nucleus of tractus solitarius. The sensory afferent fibers terminate in the nucleus of the spinal tract of the trigeminal nerve, CN V (Fig. 1.4).

The facial nerve exits the skull base through the stylomastoid foramen. It gives off the posterior auricular nerve for sensation of the periauricular area before traveling anteriorly through the parotid gland and dividing into five terminal branches that supply the muscles of facial expression; the temporofrontal, zygomatic, buccal, marginal mandibular, and cervical branches.

1.3 Terminology and Grading of Facial Palsy

The literature uses different terms to describe the severity of a peripheral palsy, which can be confusing. The term palsy can be divided into paresis, which indicates some facial muscle function, whereas paralysis indicates a complete loss of function (flaccid face). Palsy may also be described as incomplete or complete, the former meaning still some facial muscle function, while the latter means total loss of function with a flaccid face. The terms incomplete and complete are most often used in the early stage of palsy, but also in the recovery stage of palsy. A better terminology for this would instead be incomplete or complete recovery. In summary, the term palsy respective paralysis is often assimilated with each other in the literature.

Evaluating and describing facial function requires an objective and reliable measurement

system that can be used both in the acute stage and to assess the course of recovery over time. Grading facial function is also an instrument for clinical prediction of non-recovery in Bell's palsy [3]. The most widely used systems today are the gross scaled House–Brackmann [4] system and the regional weighted Sunnybrook facial grading systems [5]. Grading scales in facial palsy are described in Chaps. 2, 3.

1.4 Evaluation of Peripheral Facial Palsy

At the initial examination, first determine if the facial weakness is of central or peripheral origin. A central lesion with thrombosis, hemorrhage, tumor, or trauma affecting the voluntary corticofacial projections may cause weakness of the lower contralateral face with flattened nasolabial fold and dropped corner of the mouth. Forehead wrinkle and eye closure is, contrary to a peripheral palsy, intact due to bilateral cortical representation of the upper face. Facial emotional involuntary expressions are not affected. Loss of involuntary emotional facial movements indicates involvement of cingulate corticofacial projections. Acute ischemic cerebral stroke from an arterial obstruction, contrary to the peripheral type, results in an immediate onset of palsy.

1.5 Causes of Peripheral Facial Palsy

A number of diseases are related to/or cause peripheral facial palsy. A lesion that involves the facial nerve in the cerebellopontine angle often affects CN VIII with sensorineural hearing loss, tinnitus, and vertigo. Most common are vestibular schwannomas and meningiomas. Acute idiopathic facial palsy (Bell's palsy), most probably located to the meatal segment and facial hemangiomas, neuromas, acute and chronic (cholesteatomas) ear infection, temporal bone fractures involving the facial canal are other conditions affecting the facial nerve. Topognostic testing may help locate the level of intratemporal lesion. Decreased lacrimation indicates a lesion proximal to the branch of the greater petrosal superficial nerve, hyperacusis, and loss of gustation proximal to the stapedial respective chorda tympani branches.

The proportion of causes of facial nerve disorders varies in the literature. One reason is that referral centers may differ when comparing facial nerve disorders that are examined and treated at the respective institutions. Furthermore, standards for reporting the different causes of facial nerve disorders are lacking. Table 1.2 summarizes the most common main causes of facial disorders from the results of three large studies including 8291 cases. The studies included cases examined in 1963 and 1996 with 3721 cases—3650 patients— [2], 2570 patients seen over 25 years (year of examination not stated) in the study of Peitersen published 2002 [6], and 2000 cases of facial palsy between 2003 and 2013 by Hohman and Hadlock 2014 [7]. The predominating causes are idiopathic facial palsy (53%),

Table 1.2 Proportion (percent) of the main causes of peripheral facial palsy based on three studies including 8291 facial nerve disorders, Schaitkin and colleagues 2000 [2], Peitersen 2002 [6] and Hohman & Hadlock 2014 [7]

Main cause of peripheral facial palsy	Proportion (percent)
Idiopathic including Bell's palsy	53
Trauma (including iatrogenic)	14
Tumor (including benign tumor, malignancy, cholesteatoma and postresection tumor)	9
Herpes zoster oticus	6
Congenital/birth/neonatal	5
Infection (excluding herpes zoster)	4
Stroke, central nervous disease (lesion) and vascular brainstem	2
Facial hyperkinesis/spasm	1
Other—Not classified	6

Reference [2] by permission from Thieme Publishers, New York, United States, reference [6] copyright © Acta Oto-Laryngologica AB (Ltd) by permission Taylor & Francis Ltd., Abingdon, United Kingdom, http://www.tanfonline.com on behalf of Acta Oto-Laryngologica AB (Ltd) and the late author's son Claus Peitersen and reference [7] by permission from the publisher Wiley, United States, © 2013 The American Laryngological, Rhinological and Otological Society, Inc.

trauma (14%), tumor (9%), herpes zoster (6%), congenital palsy (5%), and infection (4%). It should however be noted that the etiological proportions varied between studies. In the study of Peitersen [6], idiopathic palsy had a percentage of 66 compared with 38 percent in the study of Hohman and Hadlock [7]. May reported as many as 849 of 3721 (23%) traumatic cases compared with 95 of 2570 (4%) reported by Peitersen [6].

1.6 Differential Diagnosis of Peripheral Facial Palsy

Lower motor neuron lesion is the most frequent cause of facial nerve disease. The most common clinical sign is ipsilateral facial muscle weakness and, more seldom, stimulation. In rare cases, both sides of the face may be affected [8]. As previously mentioned, the acute idiopathic form of palsy (Bell's palsy) is the commonest type. Nevertheless, a number of differential diagnoses (discussed in Sect. 1.7) have to be excluded when a patient presents with a facial palsy. The comprehensive book on the facial nerve by May and Schaitkin reviews the causes of facial paralysis and classifies them in groups that have since been widely used in the literature [2]. Table 1.3 presents a differential diagnosis of facial palsy and attempts to grade the most common etiological factors with the help of two large studies published during this century, 2500 cases by Peitersen published in 2002 [6] and 2000 cases of facial palsy seen between 2003 and 2013 by Hohman and Hadlock [7].

1.7 Bell's Palsy

Bell's palsy, named after the Scottish surgeon and anatomist Sir Charles Bell, presents as rapid (usually maximal development within 48 h) unilateral weakness or paralysis of the face due to acute dysfunction of the peripheral facial nerve with no readily identifiable cause and some recovery within 3–6 months [9]. Bell's palsy accounts for 50–70% of peripheral facial palsies and the annual incidence is about 30 per 100,000 individuals. There is no difference in gender or side of the face, and no seasonal clustering. Peak incidence is between ages 30 and 60. Diabetes mellitus, hypertension, and pregnancy (third trimester and 2 weeks postpartum) are considered as risk factors. Ipsilateral pain around the ear or in the neck (50–60%), taste disturbance (35%), and/or hyperacusis (30%) are concomitant symptoms.

The rate of misdiagnosis of Bell's palsy by the initial consulting clinician has been reported to be approximately 1% but also as high as 10.8% [10]. Approximately 7% of Bell's palsy patients will have a recurrence—either ipsilateral or contralateral—to previous palsy. Contralateral (alternating) recurrent palsy is usually benign and the need for extended investigation is not so urgent [10]. An ipsilateral (recurrent palsy) recurrence, however, requires a meticulous search for another etiology. Searching for a neoplasm of the facial nerve (schwannoma) or temporal bone and parotid gland is also required. The Melkersson–Rosenthal syndrome with recurrent facial palsy, fissured tongue and periodic lip or facial swelling as well as sarcoidosis is another differential diagnosis to be considered. The diagnosis of Bell's palsy should be reevaluated if there is deviation in history, new or worsening neurologic findings, new symptoms and/or clinical findings or if there are no signs of recovery within 3–4 months [11, 12] (Table 1.4).

The etiology and pathogenic mechanism in Bell's palsy is by definition not known. A possible cause of nerve injury is inflammation of the facial nerve, which might be related to herpes simplex virus type 1 and/or varicella zoster reactivation (zoster sine herpete). The virus theory is supported by the presence of herpes simplex virus type 1 DNA in facial nerve endoneural fluid and/or tissue from posterior auricular muscle in Bell's palsy patients [13]. Edema of the facial nerve within the fallopian canal has been noticed during decompressive surgery for Bell's palsy and the finding of facial nerve contrast enhancement (mainly the meatal/labyrinthine segment) on MRI may be explained by inflammatory damage to the blood-nerve barrier, with increased vascular permeability and/or venous congestion [14].

Table 1.3 Cause of peripheral facial palsy and approximate frequency

Cause of peripheral facial palsy	Approximate frequency Hohman and Hadlock [7]	Approximate frequency Peitersen [6]
Bell's palsy—idiopathic (54%)	761	1701
Neoplastic (10%)	355	105
Acoustic neuroma/vestibular schwannoma[a] (includes neurofibromatosis type 2)		
Head and neck cancer[a]		
Facial nerve tumor		
Parotid tumor lesions (benign and malignant)		
Metastatic carcinoma		
Cholesteatoma		
Leukemia		
Lymphoma		
Meningioma		
Glomus jugulare tumor		
Hemangioblastoma		
Sarcoma		
Anomalous sigmoid sinus		
Carotid artery aneurysm		
Hemangioma of tympanum		
Hidradenoma		
Teratoma		
Histiocytosis		
Fibrous dysplasia		
Temporal bone myeloma		
Carcinomatous encephalitis		
Endolymphatic sac tumors		
Fibrosarcoma		
Ossifying hemangioma		
Granular cell myoblastoma		
Infection (7%)	151	148
Herpes zoster		
Lyme neuroborreliosis		
Acute and chronic otitis/mastoiditis		
HIV		
Mononucleosis		
Poliomyelitis		
Meningitis/encephalitis		
Tuberculosis		
Mumps		
Malignant external otitis		
Chickenpox		
Coxsackie virus		
Influenza		
Acute suppurative parotitis		
Leprosy		
Malaria		
Sinus thrombosis		
Syphilis		
Scleroma		

(continued)

Table 1.3 (continued)

Cause of peripheral facial palsy	Approximate frequency Hohman and Hadlock [7]	Approximate frequency Peitersen [6]
Botulism		
Mucormycosis		
Leptospirosis		
Zika virus		
Acute hemorrhagic conjunctivitis		
Gnathostomiasis		
Mucormycosis		
Cat scratch disease		
Congenital/birth/neonatal age (6%)	99	169
Non-syndromic		
Congenital unilateral lower lip paralysis		
Möbius syndrome		
Forceps delivery		
Myotonic dystrophy		
Trauma (5%)	113	95
Skull base fracture		
Facial soft tissue and bone injuries		
Middle ear injury		
Genetic and metabolic (4%)	82	80
Diabetes		
Pregnancy associated		
Hyper/hypothyroidism		
Hypertension		
Alcoholic neuropathy		
Autoimmune/neurologic/others (3%)	32	124
Melkersson–Rosenthal syndrome		
Sarcoidosis		
Multiple sclerosis		
Collagenosis		
Guillain–Barré syndrome		
Myasthenia gravis		
Amyloidosis		
Hereditary hypertrophic neuropathy (Charcot–Marie–Tooth disease)		
Dejerine–Sottas disease		
Temporal arteritis		
Thrombotic thrombocytopenic purpura (TTP)		
Periarteritis nodosa		
Osteopetrosis		
Osteogenesis imperfecta		
Kawasaki disease		
Iatrogenic (3%) (not including postresection or postradiation acoustic neuroma/head and neck cancer)	143	0
Oral surgery		
Head and neck surgery		
Otologic surgery		
Cosmetic surgery		
Neurosurgery		

Table 1.3 (continued)

Cause of peripheral facial palsy	Approximate frequency Hohman and Hadlock [7]	Approximate frequency Peitersen [6]
Temporal artery biopsy		
Mandibular block anesthesia		
Antitetanus serum		
Rabies vaccination		
Dental		
Sagittal split osteotomy		
Embolization		
Central lesion (2%)	79	34
Central nervous lesion and disease/stroke		
Vascular brainstem lesion/stroke		
Millard–Gubler syndrome (lesion in base of pons with abducens palsy and contralateral hemiplegia)		
Toxicity (0%)	0	0
Thalidomide		
Tetanus		
Diphtheria		
Carbon monoxide		
Lead intoxication		
Ethylene glycol		
Arsenic intoxication		
Alcoholism		
Anticancer drugs		
Not classified (6%)		

Modified after Schaitkin and colleagues 2000 [2]. Approximate frequency according to average numbers given by Peitersen 2002 [6] including 2570 patients and Hohman and Hadlock 2014 [7] including 2000 patients

[a]Includes postresection and postradiation. Reference [2] by permission Thieme Publishers, New York, United States, reference [6] copyright © Acta Oto-Laryngologica AB (Ltd) by permission Taylor & Francis Ltd., Abingdon, United Kingdom, http://www.tanfonline.com on behalf of Acta Oto-Laryngologica AB (Ltd) and the late author's son Claus Peitersen and reference [7] by permission from the publisher Wiley, United States, © 2013 The American Laryngological, Rhinological and Otological Society, Inc.

Table 1.4 History and findings/symptoms that should reevaluate the diagnosis of Bell's palsy

History	Findings/symptoms
Gradual onset of palsy	Vertigo, hearing loss, tinnitus
No improvement within 3–4 months	Bilateral facial palsy
Living in a Borrelia-endemic area	Other cranial nerve involvement
Risk factors for HIV	Febrile illness, fatigue, malaise, radicular pain, and general headache (borreliosis)
Facial skin cancer	Limb or bulbar weakness
Systemic cancer	Parotid gland enlargement
	Otitis media
	Vesicles in external ear/ear canal, tympanic membrane and/or oropharynx
	Cervical adenopathy
	Facial swelling/fissured tongue

Medical treatment of Bell's palsy is based on the theory that inflammation and edema produced by herpes virus of the facial nerve is implicated in its cause. Many trials, limited in size, studying the efficacy of corticosteroids in Bell's palsy had been carried out before two large Class I randomized and placebo-controlled multi-center Bell's palsy trials were published in 2007 and 2008: a Scottish study including 551 patients treated with prednisolone and/or aciclovir [15] and a Scandinavian study including 829 patients treated with prednisolone and/or valaciclovir [16]. In both trials, evidence for the efficacy of oral corticosteroid treatment (compared with no prednisolone) started within 72 h after onset of palsy was demonstrated. In the study of Sullivan and co-workers [15], early treatment with prednisolone 25 mg twice per day

for 10 days significantly improved the chances of complete recovery at 3 and 9 months. In the trial of Engström and colleagues [16], patients given prednisolone 60 mg daily for 5 days then tapering to 10 mg daily (total treatment time 10 days) had a significantly shorter time to complete recovery and a significantly higher rate of recovery at 3, 6, and 12 months. With these two Class I studies as base, the guideline development group in the clinical practice guidelines of the American Academy of Otolaryngology—Head and Neck Surgery Foundation published in 2013 recommends a 10-day course of either prednisolone 50 mg for 10 days or prednisolone (the guideline says prednisone) 60 mg for 5 days with a 5-day taper initiated within 72 h of symptom onset [11]. The efficacy of corticosteroids was also stated in the Cochrane Database of Systematic Reviews published in 2016 in which the authors concluded that the available moderate- to high-quality evidence from randomized controlled trials shows significant benefit from treating Bell's palsy with corticosteroids [17]. The benefit of corticosteroid treatment for Bell's palsy after 72 h is less clear since Class I studies with treatment started after this time point are lacking [11]. In his guidelines based on a combination of data and theoretical mechanism of action, Vrabec discusses that the prognosis for those with complete palsy (compared with incomplete) is much poorer, and that treatment is advocated even if presentation is delayed for more than 1 week. After 2 weeks from onset, it is unlikely that any treatment will impact the recovery [18]. With caution regarding side-effects taken into consideration, this reasoning seems logical until further knowledge on the effect of delayed corticosteroid treatment is obtained.

The theory that Bell's palsy is related to a viral herpes infection has led to numerous treatment trials on the effect of antivirals of which aciclovir and valaciclovir are most studied. Including the abovementioned Class I studies [15, 16] both guideline development groups in the clinical practice guidelines of the American Academy of Otolaryngology—Head and Neck Surgery Foundation published in 2013 and the Cochrane Database of Systematic Reviews published in 2015 concluded that antiviral therapy alone in Bell's palsy is no better than placebo with respect to facial recovery [11, 19]. Antiviral treatment in combination with corticosteroids was studied in the two Class I trials by Sullivan and co-workers [15] and Engström and colleagues [16] and the addition of antivirals to corticosteroids was not proven to be of benefit in Bell's palsy. Several Bell's palsy studies of lower methodological class have reported minor improvements in facial nerve recovery with the addition of antivirals to corticosteroids so a small benefit with combination therapy cannot be excluded [11]. The guideline development group in the clinical practice guidelines of the American Academy of Otolaryngology—Head and Neck Surgery Foundation published in 2013 concluded that patients may be offered combination therapy if treated within 72 h of palsy onset, with a large role for shared decision-making. Gagyor and colleagues [19] concluded in the Cochrane Database of Systematic Reviews published in 2015 that low-quality evidence from randomized controlled trials shows a benefit from the combination of antivirals with corticosteroids compared with corticosteroids alone for the treatment of Bell's palsy of various degrees of severity. If the patient is given combination therapy, peroral antiviral valaciclovir 1000 mg 3 times daily for 7 days can be recommended.

In general, mild palsy and signs of improvement within the first 2 weeks indicate a shorter time for recovery. Patients with poor improvement after 8 weeks are at high risk for suffering sequelae. Recovery has generally reached its maximum after 9 or 12 months. In severe cases, synkinesis develops after approximately 3–4 months and may deteriorate up to 12 months after onset of palsy [20]. Recurrent Bell's palsy, on the same or opposite side, occurs in about 7% of cases. About 70% of patients with Bell's palsy recover completely within 6 months without treatment [6]. The remainder will suffer sequelae that consist of residual palsy, contracture, and synkinesis.

Due to inadequate eye closure and/or reduced tearing in most Bell's palsy patients, topical eye care to protect the cornea from dryness and abra-

sion needs to be initiated as soon as possible: lubricating eye drops at least 6–8 times daily, frequent closure of the upper eye lid with a finger by the patient and tight glasses (sun- or sports glasses) at day time, eye ointment at night, lateral eye lid cross-taping, patching and, in severe cases, a moisture chamber. In case of ocular symptoms such as pain, irritation, or itching, the patient should immediately be referred to an eye specialist. Patients with severe, persistent lagophthalmos also should have an ophthalmologic evaluation [11].

The effect of surgery for treatment of Bell's palsy is dubious and the literature gives no clear recommendations. Since the initial injury is thought to be located in the meatal/labyrinthine portion of the facial nerve only, transmastoid decompression of the facial nerve is insufficient. Middle fossa decompression, including the meatal portion, in patients diagnosed with a severe palsy (House–Brackmann score VI, electroneurography >90% nerve degeneration, and no response on voluntary electromyography) and operated on within 2 weeks after onset was reported to improve outcome compared with corticosteroid treatment only in a case control study by Gantz and colleagues [21]. The Cochrane Database of Systematic Reviews published in 2013 by McAllister and co-workers concluded that only very low quality evidence has emerged from randomized controlled trials and that this is insufficient to decide whether surgical intervention is beneficial or harmful in the management of Bell's palsy [22]. The clinical practice guidelines of the American Academy of Otolaryngology-Head and Neck Surgery Foundation from 2013 state that limited data support surgical decompression of the meatal facial nerve segment in patients with complete palsy and demonstrating severe denervation on electrodiagnostic testing within 2 weeks. It also discussed the substantial costs and rare but serious risks (including seizures, unilateral hearing loss, cerebrospinal fluid leak, and injury to the facial nerve) with this type of surgery. The subject still remains controversial and in their conclusion, the panel could make no recommendation regarding surgical decompression for Bell's palsy [11].

Acupuncture by needle insertion alone or combined with drugs is a treatment modality used for Bell's palsy. The Cochrane Database of Systematic Reviews by Chen and colleagues published in 2010 [23] concluded that the quality of the six included randomized clinical trials was inadequate to allow any conclusion about the efficacy of acupuncture for Bell's palsy. The clinical practice guidelines of the American Academy of Otolaryngology—Head and Neck Surgery Foundation from 2013 summarized that randomized clinical trials do support the use of acupuncture but these trials were downgraded due to poor quality. The panel was unable to determine the ratio of benefit to harm and could make no recommendation regarding acupuncture in Bell's palsy [11]. Li and colleagues, who included 14 randomized controlled trials involving 1541 Bell's palsy individuals in their meta-analysis from 2015, reported that there is insufficient evidence to support the efficacy and safety of acupuncture [24]. To summarize, evidence-based recommendations for acupuncture for Bell's palsy treatment are currently lacking.

Sequelae from severe Bell's palsy are persistent facial muscle weakness, facial muscle contracture, and synkinesis. In the acute stage of severe palsy, the face is flaccid with drooping corner of the mouth, less pronounced nasolabial fold, and a wide eye with lagophthalmos. In the recovery phase starting after 3–6 months, contracture of the facial muscles develops with pulled up corner of the mouth (shortening of lip elevator muscles), deepening of the nasolabial fold, and a smaller eye with contraction of the orbicularis oculi muscle. Synkinesis, which develops 3–6 months after onset of palsy, is an involuntary movement in one region of the face produced during intentional movement in another facial region (Fig. 1.5). The prevailing theory for its mechanism of development is that injured axons undergo aberrant regeneration resulting in innervation of facial muscles other than those originally innervated [25]. Mild synkinesis is present in about 15% and moderate to severe synkinesis in around 7% of Bell's palsy patients 1 year after onset [20]. The most common and troublesome synkinesis affects the muscles of

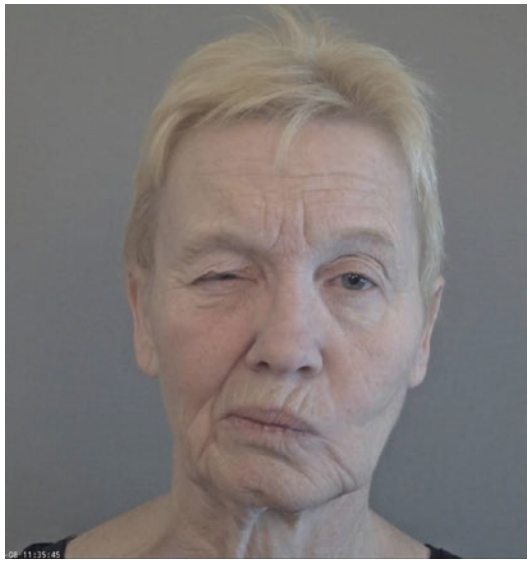

Fig. 1.5 Contracture of the facial muscles with pulled up corner of the mouth, deepening of the nasolabial fold, smaller eye and involuntary facial movements (synkinesis). (Published with oral and written permission from the patient)

the eye and mouth. Rehabilitation and management of synkinesis in facial palsy are discussed in Chaps. 4 and 31, respectively.

1.8 Acquired Peripheral Facial Palsy in Children

Bell's palsy is the leading etiology of acquired acute peripheral facial palsy in children (about 40–50%) but is 2–4 times less frequent than in adults. Its estimated yearly incidence has been reported as about 6 cases per 100,000 in ages 1–15 years, but in a large population-based epidemiologic study it was reported as 18.8 per 100,000 person-years with an increase by age [26]. There are no large randomized controlled Bell's palsy treatment trials on the effect of corticosteroids and/or antivirals in the pediatric population. Etiological factors that can cause peripheral palsy in children resemble those in adults. Infectious causes from otitis media and Lyme disease (especially in borrelia-endemic areas) are more common in children than in adults. Other infectious causes in children (as well as adults, see Table 1.3) are herpes zoster (Ramsay Hunt syndrome), Epstein–Barr virus, cytomegalovirus, adenovirus, rubella, mumps, HIV, Haemophilus influenzae, Mycoplasma pneumoniae, tuberculosis, leprosy, and syphilis. Kawasaki disease (mucocutaneous lymph node syndrome) is a rare acute systemic vasculitic condition that mainly affects children under the age of five and may be accompanied by peripheral facial palsy. The association between severe hypertension and peripheral facial palsy is mainly described in children. Facial palsy can occur in head trauma with fracture of the temporal bone, blunt cheek injury, parotid- and ear surgery, and in vestibular schwannoma surgery for neurofibromatosis type 2. Cholesteatoma of the ear may also cause peripheral facial palsy in children. Tumors and hemopathies as a cause are rhabdomyosarcoma of the temporal bone, Langerhans cell histiocytosis, vestibular and facial schwannoma, parotid tumor, leukemia, and hemophilia. Slow progress (beyond weeks) after onset of palsy or lack of improvement after 3 months suggests a neoplastic or neurological cause. Recurrent facial palsy may relate to Bell's palsy or Melkersson–Rosenthal syndrome (recurrent or persistent orofacial edema, fissured tongue, and relapsing peripheral facial palsy). Alternating recurrent palsy seldom is secondary to causal neoplasm but ipsilateral recurrent palsy needs more comprehensive investigation. Acquired bilateral facial palsy may be due to neurologic disease such as Guillain–Barré syndrome, Lyme borreliosis, most of the other above mentioned viral and bacterial infections (including meningitis), Kawasaki disease, and sarcoidosis.

The role of corticosteroid treatment for Bell's palsy in children is inclusive due to lack of controlled trials. The clinical practice guidelines of the American Academy of Otolaryngology—Head and Neck Surgery Foundation from 2013 note that despite the absence of quality trials supporting corticosteroid use in children, and given the presumed similar disease process and benefit-harm ratio of Bell's palsy in adults and children, corticosteroids may be considered in pediatric patients with a large role for caregiver

participation in the decision [11]. Prednisolone may be given in doses of 1 mg/kg (maximum 60 mg/day) per day for 7 days tapered over the following days, started within a week of onset (ideally 72 h) [27]. There is no evidence supporting the use of antiviral therapy alone [11], but addition of antivirals to prednisolone (especially in severe palsy) has been suggested as treatment in children with Bell's palsy. As for adults, it is important to protect the eye to prevent corneal ulcers in children with peripheral facial palsy.

Simultaneous bilateral peripheral facial palsy can be seen in diseases of infectious, neurological, idiopathic, neoplastic or traumatic, autoimmune, iatrogenic, and toxic origin. This is a very uncommon form of palsy and accounts for less than 1% of peripheral facial palsy patients. It requires a careful diagnostic evaluation in order to find the appropriate therapy [2]. The most common causes are Guillain–Barré syndrome, Lyme disease, "bilateral Bell's palsy-herpes simplex type 1," Melkersson–Rosenthal syndrome, sarcoidosis (Heerfordt's syndrome), Epstein–Barr and cytomegalovirus, HIV, meningitis, leukemia, and lymphoma. Trauma with skull base fracture and congenital diplegia seen in Möbius syndrome are other causes for bilateral peripheral facial palsy (Table 1.5).

Table 1.5 Underlying causes of simultaneous bilateral facial palsy

Guillain–Barré syndrome
Borreliosis
Bilateral Bell's palsy
Melkersson–Rosenthal syndrome
Sarcoidosis (Heerfordt's syndrome)
Bacterial meningitis
Epstein–Barr virus
Leukemia
HIV
Lymphoma
Syphilis
Chickenpox
Zika virus
Herpes zoster oticus
Rabies immunization
Granulomatosis with polyangiitis
Botulism
Pontine infarction
Brain stem encephalitis
Kawasaki disease
Amyloidosis
Leptospirosis
Others
Skull base fracture
Mandibular fractures
Congenital diplegia (Möbius syndrome, thalidomide toxicity)
Diabetes mellitus
Myotonic dystrophy
Myasthenia gravis
Bone disorders (sclerosing bone dysplasia, osteogenesis imperfecta, cleidocranial dysostosis)
Acute porphyria
Hypothyroidism
Pregnancy
Benign intracranial hypertension
Bilateral neurofibromas
Prepontine or intrapontine tumor
Charcot–Marie–Tooth syndrome
Bilateral acute or chronic otitis media

1.9 Herpes Zoster Oticus or Ramsay Hunt syndrome

Herpes zoster oticus or Ramsay Hunt syndrome is an acute peripheral facial palsy accompanied by ipsilateral vesicular eruptions in the ear canal, auricle, or mucous membrane of the oropharynx. About 50–75% of patients experience ipsilateral vestibulocochlear symptoms with vertigo and/or sensorineural hearing loss, more severe in the high frequency range. The annual incidence of the disease is about 4 per 100,000 individuals. Herpes zoster oticus is the most common infectious cause of acute peripheral facial palsy in adults and accounts for around 5–10% of peripheral facial palsy cases. Its etiopathology includes the reactivation of latent varicella zoster virus in the geniculate ganglion of the facial nerve. The diagnosis of herpes zoster oticus is mainly clinical via the auricular vesicular herpetic eruptions (Fig. 1.6) that appear in approximately 85% of cases, but in unclear situations, skin/blister analysis by PCR and serologic and/or cerebrospinal examination may add information. Since the palsy may precede vesicular eruptions, herpes

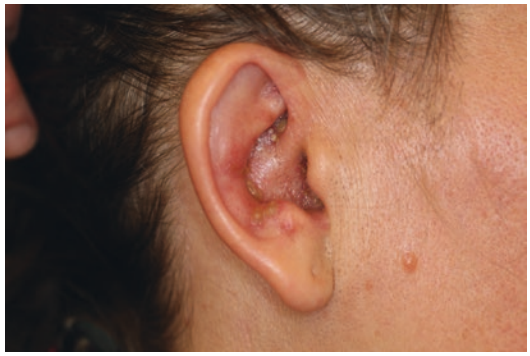

Fig. 1.6 Auricular vesicular herpetic eruptions in a patient with herpes zoster oticus (Ramsay Hunt syndrome). (Published with oral and written permission from the patient)

zoster oticus may be misdiagnosed with Bell's palsy. Even with eruptions present, a Bell's diagnosis with external otitis is sometimes incorrectly made. The facial palsy in herpes zoster oticus may be accompanied by multiple cranial nerve involvement and generally is more severe, which, even with treatment, results in a higher number of patients with sequelae (50%) compared with Bell's palsy. In contrast to Bell's palsy, herpes zoster oticus with facial palsy never occurs simultaneously on both sides and recurrence is extremely rare. Large prospective randomized controlled studies are lacking but the suggested treatment for herpes zoster oticus includes peroral antiviral valaciclovir (500 mg two times daily for 1 week) and peroral corticosteroids in doses as for Bell's palsy. In severe cases, peroral valaciclovir may be replaced with intravenous antivirals. For individuals 60 years or older, vaccinating against varicella zoster virus decreases the risk of herpes zoster.

1.10 Lyme Disease (Neuroborreliosis)

The most common manifestation of Borrelia infection is erythema migrans (25–89%), neuroborreliosis (16–29%), and arthritis (3–41%). Other less common manifestations are acrodermatitis chronica atrophicans, lymphocytoma, and carditis. Lyme neuroborreliosis is caused by a central nervous system infection due to the tick-borne spirochete *Borrelia burgdorferi*. Borrelia may invade the central nervous system directly via tissues, along peripheral nerves or by hematogenous dissemination. Neuroborreliosis is often manifested by cranial nerve engagement. Peripheral facial palsy is the most common neurologic manifestation (approximately 10%) and is often accompanied by radicular pain. Peripheral facial palsy caused by neuroborreliosis is more common in children and increasing risk factors are onset during peak season in an endemic area and a history of an erythema migrans lesion. Patients may have a preceding or concomitant febrile illness, fatigue, malaise, radicular pain, and general headache, in contrast to the more localized periauricular pain that occurs in approximately 50% of Bell's palsy patients. In areas endemic for *Borrelia burgdorferi*, Lyme neuroborreliosis is estimated to cause up to 25% of peripheral facial palsies and should therefore always be considered as a cause of palsy in these regions, especially in bilateral cases and/or if the patient is a child. Borrelia is found in the Northern temperate zone in Europe, Central and North America, and parts of Northern Asia. In peripheral facial palsy cases in endemic areas, serologic testing for immunoglobulin G and M antibodies to *Borrelia burgdorferi* antibodies should be performed in the acute stage and at follow-up within 4–6 weeks. In patients with clinical signs and/or increasing risk factors (especially children) for Borrelia, cerebrospinal examination for mononuclear pleocytosis and Borrelia antibodies should always be considered. Treatment consists of peroral doxycycline 100 mg two times daily for 14 days. Children aged ≥8 years are treated with peroral doxycycline 4 mg/kg once daily for 10 days and children <8 years are given intravenous ceftriaxone 50–100 mg/kg once daily for 10 days. In addition to antibiotics, extra corticosteroid treatment for peripheral facial palsy caused by Borrelia has been advocated, but larger randomized controlled studies to prove this are currently lacking. Findings supporting caution in using corticosteroids in this type of palsy have also been reported.

1.11 Sarcoidosis

Sarcoidosis is a systemic granulomatous disease of unknown etiology but similarities with the two other granulomatous disorders tuberculosis and leprosy have been put forward. It is more common in females than males and peak age is between 20 and 40 years. The most affected organs are the lungs (85%) followed by lymph nodes (65%), liver, skin, and eyes. Radiology and strong clinical evidence of multisystem involvement, together with biopsy findings of non-caseating epitheloid cell granulomas in addition to negative cultures for bacteria, mycobacteria and fungi, support a diagnosis of sarcoidosis.

The central nervous system is involved in sarcoidosis in approximately 10% of cases. The clinical picture involves a peripheral neuropathy or chronic basal meningitis (rarely acute meningitis) with multiple cranial nerves. Peripheral facial palsy, which may be unilateral alternating or recurrent, as well as simultaneous bilateral, is the most common neurological manifestation of sarcoidosis. In Heerfordt's syndrome, which includes peripheral facial palsy, uveitis, enlarged parotid gland, and fever, the underlying disease is sarcoidosis. Radiological diagnosis for sarcoidosis includes chest CT for showing hilar lymphadenopathy and positron emission tomography (PET)/CT/MRI for identifying sites suitable for biopsy and assessing inflammatory active sarcoidosis. Biopsy of lymph node, skin, brain, meninges, or muscle demonstrating non-caseating epitheloid cell granulomas strongly indicates a diagnosis of sarcoidosis. Serum ACE is significantly higher in most (approximately 80%) patients with active sarcoidosis. Serum angiotensin-converting enzyme (ACE) levels are significantly higher in active sarcoidosis patients compared with controls. MRI is a sensitive tool for detecting neurological lesions. Cerebrospinal fluid findings include lymphocytic pleocytosis (which may also be found in borreliosis and viral disease), elevated protein levels and, in some patients, low glucose levels. Cerebrospinal fluid should also be analyzed for ACE levels. Corticosteroids are the first-line medical treatment in neurosarcoidosis. To lower corticosteroid doses, immunosuppressive drugs may be added. Treatment with tumor necrosis factor alfa blockers has also been described.

1.12 Guillain–Barré Syndrome

Guillain–Barré syndrome is an acute to subacute inflammatory polyradiculoneuropathy. It is characterized by the development of bilateral weakness or paralysis, areflexia, paresthesia, pain, and autonomic dysfunction, thus representing a heterogeneous group of immune-mediated neuropathies. In most cases, a demyelinating process with a relatively short recovery period is found, but in its severe form, secondary axonal injury occurs. Severe sequelae from Guillain–Barré syndrome are seen in about 20% of patients. The annual incidence is about 1–2 per 100,000 individuals and its origin is an autoimmune reaction directed against peripheral nerves and nerve roots. An association with vaccines, but also with infection by cytomegalovirus, Epstein–Barr virus, Influenza A virus, *Mycoplasma pneumoniae*, and *Campylobacter jejuni* has been reported. Bilateral facial palsy is common and occurs in about 50% of cases, often with one side affected before the other. Miller Fischer Syndrome, a triad of ataxia, areflexia, and ophthalmoplegia is a variant of Guillain–Barré syndrome that also may occur with facial palsy. Neurophysiological examination shows multifocal demyelination/axonal injury. Cerebrospinal fluid findings with raised protein content and elevated albumin quotient strengthen the diagnosis. Pleocytosis is not prevalent in Guillain–Barré syndrome and if it occurs, Borrelia or HIV infection must be considered. Medical treatment with immunomodulation is performed by plasmapheresis or intravenous immunoglobulin G therapy.

1.13 Melkersson–Rosenthal Syndrome

Melkersson–Rosenthal syndrome is a rare multidisciplinary disease characterized by a triad of symptoms with recurrent orofacial edema

(mainly upper lip), facial palsy, and lingua plicata (a deeply fissured tongue). Its annual *incidence* is approximately 0.3 per 100,000 individuals. The cause of the disease is unknown. Typical histology of the orofacial edema shows a non-caseating granulomatous infiltration demonstrating similarities with that of Crohn disease and sarcoidosis. Melkersson–Rosenthal syndrome affects all age groups, but onset is most often seen in young adulthood. The most frequent sign is lingua plicata and the most common initial symptom is edema, which eventually affects most patients and gradually becomes persistent. Facial palsy appears in approximately 30–50% of cases but the complete triad form is only seen in about 20–25%. Onset of palsy is usually associated with ipsilateral orofacial swelling. Relapsing (recurrent or alternating) facial palsy may occur as well as bilateral symptoms (bilateral facial palsy and bilateral orofacial edema). Medical treatment for Melkersson–Rosenthal syndrome is symptomatic. Non-steroid anti-inflammatory drugs and systemic and/or intralesional corticosteroids have remained a mainstay of therapy. Oral antimicrobials have also been reported effective. Other less evaluated treatments include thalidomide, tumor necrosis factor alfa blockers, hydroxychloroquine, methotrexate, dapsone, azathioprine, and mycophenolate mofetil. Plastic surgery of disfiguring refractory orofacial edema may be considered. Total facial nerve decompression has been suggested to prevent further attacks of facial paralysis and its sequelae.

1.14 Ear Infection

The incidence of peripheral facial palsy associated with ear infection, including cholesteatoma, accounts for about 2–3% of all peripheral palsies [2, 6, 7]. Acute otitis media is an infection localized to the mucosa or mucoperiosteum of the middle ear and may be complicated by an acute peripheral facial palsy. Other concomitant complicating symptoms may occur from the inner ear with sensorineural hearing loss and vertigo. A pre-existing dehiscence of the tympanic segment of the fallopian canal is a predisposing factor for facial palsy. Literature shows that the majority of cases are children and that the palsy may be the first sign of acute otitis media with or without mastoiditis. Before antibiotics, otitis media was responsible for a greater proportion of facial palsies. Intravenous antibiotic therapy should be initiated and acute myringotomy drainage with bacterial culture performed. With treatment, the prognosis for facial recovery in general is favorable. Mastoidectomy is performed in cases of complicating acute mastoiditis and/or subperiosteal abscess. In cases of suspected complications (cholesteatoma/bone destruction/sigmoid sinus thrombosis), computed tomography (thin-sliced) is mandatory. Adding corticosteroids to the medical therapy may be considered but convincing reports for their effectiveness are lacking.

Two forms of chronic otitis media associated with peripheral facial palsy are found: chronic suppurative otitis media without cholesteatoma and chronic otitis with cholesteatoma. In contrast to the acute form of otitis media, which mainly affects children, peripheral facial palsy in chronic otitis media mostly affects youths and adults. The palsy in chronic suppurative otitis media with perforation of the tympanic membrane may present as an acute or slowly progressing facial palsy. Prompt treatment with intravenous broad-spectrum antibiotics after bacterial culture from middle ear discharge as well as from blood must be commenced. Computed tomography is needed to evaluate for dehiscence and/or bone destruction within the temporal bone. Pathologic tissue with destruction, progressing disease, and deterioration of facial function demands explorative surgery and facial nerve decompression with elimination of pathological tissue. Cholesteatoma is a non-malignant tumorous process with focal regions of keratinizing squamous epithelium mostly located to the middle ear or mastoid, which may erode surrounding bone. Congenital cholesteatoma occurs in children and acquired cholesteatomas, which are in the majority, are primarily found in adults. Peripheral facial palsy

associated with chronic otitis with cholesteatoma needs immediate management in the same manner as for chronic suppurative otitis media. MRI may add further information (cholesteatoma shows characteristic reduced diffusion). Surgery includes facial nerve decompression and removal of cholesteatoma.

1.15 Intratemporal Facial Nerve Trauma

Trauma accounts for about 5% of all peripheral facial palsies. Facial palsy due to intratemporal injury is often associated with external trauma. The most common cause of temporal bone fracture is motor vehicle accidents [7] although seat belts and air bags have reduced these injuries. Computed tomography with thin slices is the radiological method for mapping the extent of injury. It is important to look for cerebrospinal fluid leakage. Temporal bone fracture lines are classically described in relation to the long axis of the petrous pyramid as longitudinal, oblique, and transverse. Transverse fractures account for about 10% of the total but facial palsy is observed in 40–50% of them as opposed to longitudinal fractures (80–90% of total) in which palsy occurs in just 20%. Peripheral facial palsy is twice as common in fractures that cross the otic capsule (also results in sensorineural hearing loss) compared to fractures that do not intersect the capsule. An immediate palsy from head injury will be apparent as soon as the patient is examined in the hospital. Delayed onset of palsy can occur within an interval of 4–5 days. Delayed palsy has a better prognosis than the immediate type and it is generally agreed that delayed palsies should be conservatively treated with corticosteroids as medical treatment. Treatment of immediate palsy depends on the patient's general neurologic condition after the head injury, severity of palsy on electroneuronography as well as computed tomography findings. Surgical exploration of the facial nerve has to be considered in cases with poor prognosis preferably within 2 weeks after onset.

1.16 Extratemporal Facial Nerve Trauma

Injury to the face may wound the facial nerve main trunk or its branches. These injuries account for around 1.5% of peripheral facial palsies. The most common trauma is penetrating, laceration, slash, and facial fracture to the soft tissue. Careful examination of the separate facial nerve branches is important since edema and bruising may mask palsy. Penetrating nerve injuries need prompt exploration and identification of cut nerve branches. Primary nerve repair results in better recovery compared with rerouting or grafting (see Chap. 6).

1.17 Iatrogenic Facial Nerve Trauma

Iatrogenic injury to the facial nerve causes 3% of all peripheral facial palsies. Patients operated for removal of vestibular schwannoma at the cerebellopontine angle comprise one of the largest iatrogenic groups and in larger acoustic neuroma referral centers, these patients account for up to 9% [7]. Facial palsy also occurs after tumor resection alongside any part of the intratemporal or extratemporal facial nerve, including temporal bone tumors, parotid tumors, and head and neck cancer. In tumor surgery, volitional sacrificing of the facial nerve frequently must be performed to obtain radicality. Other causes of iatrogenic injury to the facial nerve are mastoid surgery, middle ear surgery, oral surgery, cosmetic surgery (face lift), neurosurgery, and temporal biopsy surgery [7]. If unexpected facial palsy occurs after surgery and the intactness of the nerve is unclear, facial nerve exploration has to be carried out.

1.18 Tumor

Benign and malignant tumors account for around 5% of peripheral facial palsies [2, 6, 7]. Clinical suspicion of a tumor as cause of peripheral facial palsy is raised with gradual onset, fluctuating

or recurrent palsy, hearing loss on the palsy side, and ipsilateral parotid or ear tumor mass. Benign tumors as cause of facial palsy are vestibular schwannoma (often starts with hearing loss) at the cerebellopontine angle and facial neuroma and hemangioma in the intratemporal portion. Malignant tumors in the temporal bone consist of squamous cell carcinomas (primary or metastatic) and basal cell carcinomas. Malignant tumors located in the region of the parotid gland as cause of palsy are mucoepidermoid carcinomas, adenoid cystic carcinomas, adenocarcinomas, and squamous cell carcinomas. Tumor resection (in some cases also reconstructive surgery) and reanimation surgery followed by oncological treatment is the option for these malignant tumors.

1.19 Peripheral Facial Palsy in Newborn, Congenital Facial Palsy

The incidence of peripheral facial palsy at birth, so-called congenital facial palsy, is between 1 and 2 per 1000 newborns. The palsy is categorized as traumatic or developmental and distinguishing between the two is important since prognosis and therapy differ. The spontaneous recovery rate in congenital traumatic palsy is approximately 90%, while developmental facial palsy in general will not improve. The most common cause of unilateral congenital palsy is birth-related trauma. Primipara, birth weight more than 3500 g, prolonged labor, and forceps delivery are risk factors for trauma. Signs of birth-related injury include face or skull lacerations or hematomas as well as mastoid hematomas and hemotympanum. Other anomalies, bilateral palsy or cranial deficits indicate developmental background. Except for history and clinical findings, early-stage computed tomography and/or MR imaging may help differentiate traumatic (fracture of the skull base) from developmental palsy (ear anomalies). Furthermore, congenital traumatic palsy, in contrast to developmental palsy, initially shows normal electroneurographic and electromyographic responses at birth. In severe traumatic palsies, which also contrast to developmental palsies, synkinesis will develop after 3–4 months.

The causes of developmental facial palsy may be isolated (non-syndromic) or related to syndromes. Family history of congenital facial palsy indicates a developmental origin. In Möbius syndrome with an incidence of approximately 1 per 50,000 births, bilateral facial nerve palsy and unilateral or bilateral abducens palsy are the most typical features. Other cranial nerves (especially hypoglossal) may also be affected and deformities of the head, trunk (pectoral muscles), and limbs (syndactylism and club foot) may accompany the cranial nerve defects. The pathogenesis of facial palsy in Möbius syndrome is thought to be caused by a nuclear or peripheral lesion. Oculoauriculovertebral syndrome is a spectrum of craniofacial disorders with unilateral abnormalities related to first and second branchial arches. Hypoplasia of the maxilla and mandible, microtia, and abnormally small mouth opening are clinical features of this syndrome. In addition to facial palsy, sensorineural and/or conductive hearing loss is frequent in hemifacial microsmia. Goldenhar syndrome, a variant of oculoauriculovertebral syndrome, also includes cleft palate, vertebral anomalies and epibulbar dermoids, as well as the above mentioned malformations. Symptoms affect both sides of the face in 10–30% of cases, with the right side usually more severely involved. Heart defects, kidney problems as well as central nervous system abnormalities may be present in this syndrome.

Congenital unilateral lower lip palsy, also called asymmetric crying facies, is confined to the lower lip depressor muscles innervated by the mandibular branch of the facial nerve. At rest, the face is symmetric but when crying or laughing drooping of the lower lip towards the unaffected side is seen. Congenital unilateral lower lip palsy is one of the commonest forms of congenital palsies. Sometimes this type may be associated with other malformations of which heart disease is most common. Other grounds or related disorders to congenital facial palsy are CHARGE syndrome (colobomata, heart defects, atresia of choanae, retarded growth, genital hypoplasia, and ear abnormalities), facioscapulohumeral muscu-