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452 Trigeminal Neuralgia, Bell’s Palsy, and Other Cranial Nerve Disorders

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Trigeminal Neuralgia,

Bell’s Palsy, and Other

Cranial Nerve Disorders The cranial nerves consist of 12 paired nerves that mediate variable combinations of motor, sensory, and autonomic functions. They are considered as a group because of their close anatomic relationship to the brainstem (Fig. 452-1) and to one another, and tendency to be involved together in a variety of disease states. Nine cranial nerves connect directly with brainstem nuclei; the exceptions are cranial nerves 1 (olfactory) and 2 (optic) that are more accurately considered fiber tracts of the brain, and cranial nerve 11 (spinal accessory) whose motor neurons reside largely in the upper cervical cord. Analogous to spinal nerves (Chap. 453), motor fibers of the cranial nerves have their origin in the brainstem or upper cervical cord, while sensory nerves are pseudounipolar, with ganglia outside the central nervous system and a synapse with second-order fibers in the brainstem. Symptoms and signs of cranial nerve pathology are common

in internal medicine. They may develop in the context of a widespread neurologic disturbance, and in such situations, cranial nerve involve ment may represent the initial manifestation of the illness. In other disorders, involvement is largely restricted to one or several cranial nerves; these distinctive disorders are reviewed in this chapter. Disor ders of olfaction are discussed in Chap. 35, vision and ocular move ment in Chap. 34, hearing in Chap. 36, and vestibular function in Chap. 24.
FACIAL PAIN OR NUMBNESS ■ ■ANATOMIC CONSIDERATIONS The trigeminal (fifth cranial) nerve supplies sensation to the skin of the face, anterior half of the head, and the nasal and oral mucosa (Fig. 452-2). The motor part innervates the muscles involved in chewing (including masseter, temporalis, and pterygoids) as well as the anterior belly of the digastric, mylohyoid, tensor veli palatini, and the tensor tympani (hear ing especially for high-pitched tones). It is the largest of the cranial nerves. It exits in the lateral midpons and traverses the middle cranial fossa to the semilunar (gasserian, trigeminal) ganglion in Meckel's cave, where the nerve splits into three divisions (ophthalmic [V1], maxillary [V2], and mandibular [V3]). V1 and V2 traverse the cavernous sinus to exit in the superior orbital fissure and foramen rotundum; V3 exits through the foramen ovale. The trigeminal nerve is predominantly

Frontal lobe Olfactory bulb and peduncle Pituitary gland Cranial nerves Mamillary bodies X IX VIII VII VI V IV III II Temporal lobe CHAPTER 452 Trigeminal ganglion Cerebellopontine angle Trigeminal Neuralgia, Bell's Palsy, and Other Cranial Nerve Disorders
Cerebellum XII XI FIGURE 452-1 Ventral view of the brain, illustrating relationships between the 12 pairs of cranial nerves and the brainstem. (Adapted from SG Waxman: Clinical Neuroanatomy, 29th ed. http://www.accessmedicine.com.) sensory, and motor innervation is exclusively carried in V3. The cor nea is primarily innervated by V1, although an inferior crescent may be V2. Upon entering the pons, pain and temperature fibers descend ipsilaterally as far as the upper cervical spinal cord as the spinal tract of V, before synapsing with the spinal nucleus of V; this accounts for the facial numbness that can occur with spinal cord lesions above C2. In the brainstem, the spinal tract of V is also located adjacent to crossed ascending fibers of the spinothalamic tract, producing a "crossed" sen sory loss for pain and temperature (ipsilateral face, contralateral arm/ trunk/leg) with lesions of the lateral lower brainstem. CN V is also ensheathed by oligodendrocyte-derived, rather than Schwann cell– derived, myelin for up to 7 mm after it leaves the brainstem, unlike just a few millimeters for other cranial and spinal nerves; this may explain some instances of trigeminal neuralgia in multiple sclerosis (MS) (Chap. 455), a disorder of oligodendrocyte myelin. ■ ■TRIGEMINAL NEURALGIA (TIC DOULOUREUX) Clinical Manifestations Trigeminal neuralgia is characterized by excruciating paroxysms of pain in the lips, gums, cheek, or chin and, very rarely, in the distribution of the ophthalmic division of the fifth nerve. The pain seldom lasts more than a few seconds or a minute or two but may be so intense that the patient winces, hence the term tic. The paroxysms, experienced as single jabs or clusters, tend to recur frequently, both day and night, for several weeks at a time. They may occur spontaneously or be brought on with movements of affected areas by speaking, chewing, or smiling. Another characteristic feature is the presence of trigger zones, typically on the face, lips, or tongue, that provoke attacks; patients may report that tactile stimuli—e.g., washing the face, brushing the teeth, or exposure to a draft of air— generate excruciating pain. An essential feature of trigeminal neural gia is that objective signs of sensory loss cannot be demonstrated on examination. Trigeminal neuralgia is relatively common, with an estimated annual incidence of 4–8 per 100,000 individuals. Middle-aged and

KEY Ophthalmic (V1) Maxillary (V2) Mandibular (V3) Supraorbital nerve Anterior ethmoidal nerve Posterior ethmoidal nerve PART 13 Neurologic Disorders Frontal branch of frontal nerve Supratrochlear nerve Infratrochlear nerve Internal nasal rami Infraorbital nerve External nasal rami a m l ri a c l a l i x L Ma Nasal and labial rami of infraorbital nerve Anterior superior alveolar nerves n l ua ng Li ol ve al or eri Submandibular ganglion Inf Submandibular and sublingual glands Mental nerve FIGURE 452-2 The trigeminal nerve and its branches and sensory distribution on the face. The three major sensory divisions of the trigeminal nerve consist of the ophthalmic, maxillary, and mandibular nerves. (Reproduced with permission from Waxman SG: Clinical Neuroanatomy, 26th ed. New York, McGraw-Hill, 2009.) elderly persons are affected primarily, and ~60% of cases occur in women. Onset is typically sudden, and bouts tend to persist for weeks or months before remitting spontaneously. Remissions may be longlasting, but in most patients, the disorder ultimately recurs. Pathophysiology Symptoms result from ectopic generation of action potentials in pain-sensitive afferent fibers of the fifth cranial nerve root just before it enters the lateral surface of the pons. Com pression or other pathology in the nerve leads to demyelination of large myelinated fibers that do not themselves carry pain sensation but become hyperexcitable and electrically coupled with smaller unmy elinated or poorly myelinated pain fibers in close proximity; this may explain why tactile stimuli, conveyed via the large myelinated fibers, can stimulate paroxysms of pain. Compression of the trigeminal nerve root by a blood vessel, most often the superior cerebellar artery or on occasion a tortuous vein, is believed to be the source of trigeminal neu ralgia in most patients. In cases of vascular compression, age-related

C2 C3 C4 Frontal nerve Mesencephalic nucleus of V Ophthalmic nerve Semilunar ganglion Main sensory nucleus of V Main motor nucleus of V Nucleus of spinal tract of V Mandibular nerve Anterior and posterior deep temporal nerves (to temporal muscles) y r Pterygopalatine ganglion Otic ganglion Auriculotemporal nerve e v er Lateral pterygoid muscle Chorda tympani nerve e v er n ar Buccinator nerve Medial pterygoid muscle Masseter muscle Mylohyoid nerve Anterior belly of digastric muscle brain sagging and increased vascular thickness and tortuosity may explain the prevalence of trigeminal neuralgia in later life. Differential Diagnosis Trigeminal neuralgia must be distin guished from other causes of face and head pain (Chap. 17) and from pain arising from diseases of the jaw, teeth, or sinuses. Pain from migraine or cluster headache tends to be deep-seated and steady, unlike the superficial stabbing quality of trigeminal neuralgia; rarely, cluster headache is associated with trigeminal neuralgia, a syndrome known as cluster-tic. Other rare, paroxysmal headache disorders such as short-lasting unilateral headache attacks with conjunctival injec tion and tearing, short-lasting unilateral headache attacks with cranial autonomic symptoms, and paroxysmal hemicrania (Chap. 441) are distinguished by the frequency and duration of attacks and associated autonomic symptoms. In temporal arteritis, superficial facial pain is present but is not typically shocklike, the patient frequently complains of myalgias and other systemic symptoms, and an elevated erythrocyte

sedimentation rate (ESR) or C-reactive protein (CRP) is usually present (Chap. 375). When trigeminal neuralgia develops in a young adult or is bilateral, MS is a key consideration, and in such cases, the cause is often a demyelinating plaque near the root entry zone of the fifth nerve in the pons, especially when there is evidence of superimposed facial sensory loss, which can be subtle. Cases that are secondary to mass lesions—such as aneurysms, neurofibromas, acoustic schwannomas, or meningiomas—usually produce objective signs of sensory loss in the trigeminal nerve distribution (trigeminal neuropathy, see below). Laboratory Evaluation An ESR or CRP is

indicated if temporal arteritis is suspected. Neuroimaging studies are often necessary to exclude secondary causes and help assess overlying vascular lesions in order to plan for decompression surgery. TREATMENT Trigeminal Neuralgia Drug therapy with carbamazepine is effective in ~50–75% of patients. Carbamazepine should be started as a single daily dose of 100 mg taken with food and increased gradually (by 100 mg daily in divided doses every 1–2 days) until substantial (>50%) pain relief is achieved. Most patients require a maintenance dose of 200 mg four times daily. Doses >1200 mg daily provide no additional benefit. Dizziness, imbalance, sedation, and rare cases of agranulocytosis are the most important side effects of carbamazepine. If treatment is effective, it is usually continued for 1 month and then tapered as tolerated. Oxcarbazepine (300–1200 mg bid) is an alternative to carbamazepine that has less bone marrow toxicity and probably is equally efficacious. If these agents are not well tolerated or are inef fective, phenytoin (300–400 mg daily) is another option. Lamotrigine (400 mg daily), baclofen (10–20 mg tid), or topiramate (50 mg bid) may also be tried. Gabapentin, up to 3600 mg daily in divided doses, may occasionally provide relief. If drug treatment fails, surgical therapy should be offered. The most widely used method is currently microvascular decompres sion to relieve pressure on the trigeminal nerve as it exits the pons. This procedure requires a suboccipital craniotomy. This proce dure appears to have a >70% efficacy rate and a low rate of pain recurrence in responders; the response is better for classic ticlike symptoms than for nonlancinating facial pains. High-resolution magnetic resonance angiography is useful preoperatively to visual ize the relationships between the fifth cranial nerve root and nearby blood vessels. Gamma knife radiosurgery of the trigeminal nerve root is also used for treatment and results in complete pain relief, sometimes delayed in onset, in approximately one-half of patients and a low risk of persistent facial numbness; the response is sometimes longlasting, but recurrent pain develops over 2–3 years in one-third of patients. Compared with surgical decompression, gamma knife surgery appears to be somewhat less effective but has few serious complications. Another procedure, radiofrequency thermal rhizotomy, creates a heat lesion of the trigeminal ganglion or nerve. Short-term relief is experienced by >95% of patients; long-term studies indicate that pain recurs in up to one-third of treated patients. Postoperatively, partial numbness of the face is common, masseter (jaw) weakness may occur especially following bilateral procedures, and corneal denervation with secondary keratitis can follow rhizotomy for firstdivision trigeminal neuralgia. Percutaneous balloon compression of the trigeminal ganglion is an alternative approach performed under general anesthesia that results in similar rates of short- and longterm pain relief and is also commonly complicated by ipsilateral facial numbness. ■ ■TRIGEMINAL NEUROPATHY A variety of diseases can affect the trigeminal nerve (Table 452-1). Most present with sensory loss on the face or with weakness of the jaw

TABLE 452-1 Trigeminal Nerve Disorders Nuclear (Brainstem) Lesions Multiple sclerosis Stroke Syringobulbia Glioma Lymphoma Preganglionic Lesions Acoustic neuroma Meningioma Metastasis Chronic meningitis Cavernous carotid aneurysm Semilunar Ganglion Lesions CHAPTER 452 Trigeminal neuroma Herpes zoster Infection (spread from otitis media or mastoiditis) Cavernous Sinus Lesions (see Table 452-2) Peripheral Nerve Lesions Trigeminal Neuralgia, Bell’s Palsy, and Other Cranial Nerve Disorders
Tumor (e.g., nasopharyngeal carcinoma, squamous cell carcinoma, lymphoma) Trauma Guillain-Barré syndrome Sjögren’s syndrome Collagen-vascular diseases Sarcoidosis Leprosy Drugs (stilbamidine, trichloroethylene) Idiopathic trigeminal neuropathy muscles. Deviation of the jaw on opening indicates weakness of the pterygoids on the side to which the jaw deviates. Some cases

are due to Sjögren’s syndrome or a collagen-vascular disease such as systemic lupus erythematosus, scleroderma, or mixed connective tissue disease. Among infectious causes, herpes zoster (acute or postherpetic) and lep rosy should be considered. Tumors of the middle cranial fossa (menin giomas), of the trigeminal nerve (schwannomas), or of the base of the skull (metastatic tumors) may cause a combination of motor and sen sory signs. Lesions in the cavernous sinus can affect the first and sec ond divisions of the trigeminal nerve, and lesions of the superior orbital fissure can affect the first (ophthalmic) division; the accompanying corneal anesthesia increases the risk of ulceration (neurokeratitis). Isolated sensory loss over the chin (mental neuropathy) can be the only manifestation of systemic malignancy. Rarely, an idiopathic form of trigeminal neuropathy is observed. It is characterized by numb ness and paresthesias, sometimes bilaterally, with loss of sensation in the territory of the trigeminal nerve but without weakness of the jaw. Gradual recovery is the rule. Tonic spasm of the masticatory muscles, known as trismus, is symptomatic of tetanus (Chap. 157) or may occur in patients treated with phenothiazines. FACIAL WEAKNESS ■ ■ANATOMIC CONSIDERATIONS (Fig. 452-3) The seventh cranial nerve supplies all the muscles con cerned with facial expression, as well as the stapedius, stylohyoid, and posterior belly of the digastric. The sensory and parasympathetic com ponents (the nervus intermedius) convey taste sensation from the ante rior two-thirds of the tongue, cutaneous impulses from the anterior wall of the external auditory canal, and preganglionic parasympathetic signals to the pterygopalatine and submandibular ganglia, stimulat ing lacrimation, rhinorrhea, and salivation. The cell bodies of pseu dounipolar sensory neurons lie in the geniculate ganglion. The motor nucleus of the seventh nerve lies anterior and lateral to the abducens nucleus. After leaving the pons, the seventh nerve enters the internal auditory meatus with the acoustic nerve. The nerve continues its course

Superior salivatory nucleus Geniculate ganglion Trigeminal ganglion Motor nucleus VI n. V n. Motor nucleus VII n.

Nucleus fasciculus solitarius C VII n. B A PART 13 Neurologic Disorders Fasciculus solitarius Chorda tympani Lingual nerve Submandibular gland Submandibular ganglion FIGURE 452-3 The facial nerve. A, B, and C denote lesions of the facial nerve at the stylomastoid foramen, distal and proximal to the geniculate ganglion, respectively. Green lines indicate the parasympathetic fibers, red line indicates motor fibers, and purple lines indicate visceral afferent fibers (taste). (Reproduced with permission from MB Carpenter: Core Text of Neuroanatomy, 2nd ed. Williams & Wilkins, 1978.) in its own bony channel, the facial canal, and exits from the skull via the stylomastoid foramen. It then passes through the parotid gland and subdivides to supply the facial muscles. A complete interruption of the facial nerve at the stylomastoid fora men paralyzes all muscles of facial expression. The corner of the mouth droops, the creases and skinfolds are effaced, the forehead is unfur rowed, and the eyelids will not close. Upon attempted closure of the lids, the eye on the paralyzed side rolls upward (Bell’s phenomenon). The lower lid sags and falls away from the conjunctiva, permitting tears to spill over the cheek. Food collects between the teeth and lips, and saliva may dribble from the corner of the mouth. The patient com plains of a heaviness or numbness in the face, but sensory loss is rarely demonstrable and taste is intact. If the lesion is in the middle-ear portion, taste is lost over the ante rior two-thirds of the tongue on the same side. If the nerve to the sta pedius is interrupted, there is hyperacusis (sensitivity to loud sounds). Lesions in the internal auditory meatus may affect the adjacent audi tory and vestibular nerves, causing deafness, tinnitus, or dizziness. Intrapontine lesions that paralyze the face usually

affect the abducens nucleus as well, and often the corticospinal and sensory tracts. If the peripheral facial paralysis has existed for some time and recov ery of motor function is incomplete, a continuous diffuse contraction of facial muscles may appear. The palpebral fissure becomes narrowed, and the nasolabial fold deepens. Facial spasms, initiated by movements of the face, may develop (hemifacial spasm). Anomalous regeneration of seventh nerve fibers may result in other troublesome phenomena. If fibers originally connected with the orbicularis oculi come to innervate the orbicularis oris, closure of the lids may cause a retraction of the mouth (synkinesis), or if parasympathetic fibers originally connected with salivary glands later innervate the lacrimal gland, anomalous tear ing (“crocodile tears”) may occur with eating. Another facial synkinesia is triggered by jaw opening, causing closure of the eyelids on the side of the facial palsy (jaw-winking). ■ ■BELL’S PALSY The most common form of facial paralysis is Bell’s palsy. The annual incidence of this idiopathic disorder is ~25 per 100,000 annually, or

Major superficial petrosal nerve Lacrimal gland

Pterygopalatine ganglion To nasal and palatine glands Sublingual gland about 1 in 60 persons in a lifetime. Risk factors include pregnancy and diabetes mellitus. Clinical Manifestations The onset of Bell’s palsy is fairly abrupt, with maximal weakness being attained by 48 h as a general rule. Pain behind the ear may precede the paralysis for a day or two. Taste sensa tion may be lost unilaterally, and hyperacusis may be present. In some cases, there is mild cerebrospinal fluid lymphocytosis. MRI may reveal swelling and uniform enhancement of the geniculate ganglion and facial nerve and, in some cases, entrapment of the swollen nerve in the temporal bone. Approximately 80% of patients recover within a few weeks or months. Electromyography may be of some prognostic value; evidence of denervation after 10 days indicates there has been axonal degeneration, that there will be a long delay (3 months as a rule) before regeneration occurs, and that it may be incomplete. The presence of incomplete paralysis in the first week is the most favorable prognostic sign. Recurrences are reported in ~7% of cases. Pathophysiology In acute Bell’s palsy, there is inflammation of the facial nerve with mononuclear cells, consistent with an infectious or immune cause. Herpes simplex virus (HSV) type 1 DNA was fre quently detected in endoneurial fluid and posterior auricular muscle, suggesting that a reactivation of this virus in the geniculate ganglion may be responsible for most cases. Reactivation of varicella-zoster virus is associated with Bell’s palsy in up to one-third of cases and may represent the second most frequent cause. A variety of other viruses including SARS-CoV-2 have also been implicated less commonly, and Bell’s palsy can be observed in the setting of human immunodeficiency virus (HIV) seroconversion. Differential Diagnosis There are many other causes of acute facial palsy that must be considered in the differential diagnosis of Bell’s palsy. Lyme disease can cause unilateral or bilateral facial palsies; in endemic areas, ≥10% of cases of facial palsy are likely due to infection with Borrelia burgdorferi (Chap. 191). Ramsay Hunt syndrome, caused by reactivation of herpes zoster in the geniculate ganglion, consists of a severe facial palsy associated with a vesicular eruption in the external auditory canal and sometimes in the pharynx and other parts of the

A B FIGURE 452-4 Axial and coronal T1-weighted images after gadolinium with fat suppression demonstrate diffuse smooth linear enhancement of the left facial nerve, involving the genu, tympanic, and mastoid segments within the temporal bone (arrows), without evidence of mass lesion. Although highly suggestive of Bell’s palsy, similar findings may be seen with other etiologies such as Lyme disease, sarcoidosis, and perineural malignant spread. cranial integument; often the

eighth cranial nerve is affected as well. Facial palsy that is often bilateral occurs in sarcoidosis (Chap. 379) and in Guillain-Barré syndrome (Chap. 458). Leprosy frequently involves the facial nerve, and facial neuropathy may also occur in diabetes mel litus, connective tissue diseases including Sjögren's syndrome, and amy loidosis. The rare Melkersson-Rosenthal syndrome consists of recurrent facial paralysis; recurrent—and eventually permanent—facial (particu larly labial) edema; and, less constantly, plication of the tongue. Its cause is unknown. Acoustic neuromas frequently involve the facial nerve by local compression. Infarcts, demyelinating lesions of MS, and tumors are common pontine lesions that interrupt the facial nerve fibers; other signs of brainstem involvement are usually present. Tumors that invade the temporal bone (carotid body, cholesteatoma, dermoid) may produce a facial palsy, but the onset is insidious and the course progressive. Facial palsy after temporal bone fracture can present acutely or after a delay of several days; blunt head injury without temporal bone fracture may also trigger facial palsy. All these forms of nuclear or peripheral facial palsy must be dis tinguished from the supranuclear type. In the latter, the frontalis and orbicularis oculi muscles of the forehead are involved less than those of the lower part of the face, since the upper facial muscles are inner vated by corticobulbar pathways from both motor cortices, whereas the lower facial muscles are innervated only by the opposite hemisphere. In supranuclear lesions, there may be a dissociation of emotional and voluntary facial movements, and often some degree of paralysis of the arm and leg or an aphasia (in dominant-hemisphere lesions) is present. Laboratory Evaluation The diagnosis of Bell's palsy can usually be made clinically in patients with (1) a typical presentation, (2) no risk factors or preexisting symptoms for other causes of facial paralysis, (3) absence of cutaneous lesions of herpes zoster in the external ear canal, and (4) a normal neurologic examination apart from the facial nerve. Particular attention to the eighth cranial nerve, which courses near to the facial nerve in the pontomedullary junction and in the temporal bone, and to other cranial nerves is essential. In atypical or uncertain cases, an ESR or CRP, testing for diabetes mellitus, a Lyme titer, HIV serologies, angiotensin-converting enzyme and chest imaging studies for possible sarcoidosis, a lumbar puncture for possible Guillain-Barré syndrome, or MRI scanning may be indicated. MRI often shows swelling and enhancement of the facial nerve in idiopathic Bell's palsy (Fig. 452-4). TREATMENT Bell's Palsy Symptomatic measures include (1) the use of paper tape to depress the upper eyelid during sleep and prevent corneal drying, (2) arti ficial tears, and (3) massage of the weakened muscles. A course of glucocorticoids, given as prednisone 60–80 mg daily during the first

 5 days and then tapered over the next 5 days, modestly shortens recovery and improves the functional outcome. Although large and well-controlled randomized trials found no added benefit of the antiviral agents valacyclovir (1000 mg daily for 5–7 days) or acyclovir (400 mg five times daily for 10 days) compared to gluco corticoids alone, either of these agents should be used if vesicular lesions are observed in the palate or external auditor canal. For patients with permanent paralysis from Bell's palsy, a number of cosmetic surgical procedures have been used to restore a relatively symmetric appearance to the face. Trigeminal Neuralgia, Bell's Palsy, and Other Cranial Nerve Disorders

■ ■OTHER MOTOR DISORDERS OF THE FACE Hemifacial spasm consists of painless irregular involuntary contrac tions on one side of the face. Most cases appear related to vascular compression of the exiting facial nerve in the pons. Other cases develop as a sequela to Bell's palsy or are secondary to compression and/or demyelination of the nerve by tumor, infection, or MS. Local injec tions of botulinum toxin into affected muscles can relieve spasms for 3–4 months, and the injections can be repeated. Refractory cases due to vascular compression usually respond

to surgical decompression of the facial nerve. Anecdotal reports describe success using carbamaze pine, gabapentin, or baclofen. Blepharospasm is an involuntary recur rent spasm of both eyelids that usually occurs in elderly persons as an isolated phenomenon or with varying degrees of spasm of other facial muscles. Severe, persistent cases of blepharospasm can be treated by local injection of botulinum toxin into the orbicularis oculi. Clonaz epam, baclofen, and trihexyphenidyl have also been used to treat this disorder. Facial myokymia refers to a fine rippling activity of the facial muscles; it may be caused by MS or follow Guillain-Barré syndrome (Chap. 458). OTHER CRANIAL NERVE DISORDERS ■ ■GLOSSOPHARYNGEAL NEURALGIA The ninth cranial (glossopharyngeal) nerve (Fig. 452-5) conveys somatic sensation from the pharynx, middle ear, tympanic membrane, eustachian tube, and posterior third of the tongue to the spinal trigemi nal nucleus. It also relays taste from the posterior third of the tongue and information about blood pressure from baroreceptors in the carotid sinus to the nucleus solitarius, which also serves as the sensory nucleus for the vagus nerve. Motor function originates in the nucleus ambiguus and is limited to the stylopharyngeus muscle. Parasympa thetic fibers from the medullary inferior salivatory nucleus synapse in the otic ganglion with postganglionic fibers that innervate the parotid gland. Glossopharyngeal neuralgia resembles trigeminal neuralgia in many respects but is much less common. Sometimes it involves por tions of the tenth (vagus) nerve. The pain is intense and paroxysmal; it originates on one side of the throat, approximately in the tonsillar fossa.

Inferior salivatory nucleus (parasympathetic) Petrous portion of temporal bone Ambiguus nucleus (motor) Nucleus of solitary tract (sensory) Superior or jugular ganglion Jugular foramen VII Petrous ganglion PART 13 Neurologic Disorders Communication with auricular branch of X Nodose ganglion X Superior cervical sympathetic ganglion Carotid body Sinus nerve Carotid sinus and nerve plexus Common carotid artery Sensory branches to soft palate, fauces, and tonsils Sympathetic root (vasomotor) IX (Sensory) Vagal root (motor and sensory) Pharyngeal plexus To muscles and mucous membrane of the pharynx and soft palate Taste and sensation to posterior third of tongue FIGURE 452-5 The ninth cranial (glossopharyngeal) nerve. FO, foramen ovale; FR, foramen rotundum; TP, tympanum plexus. (Reproduced with permission from SG Waxman: Clinical Neuroanatomy, 29th ed. New York, McGraw Hill, 2020.) In some cases, the pain is localized in the ear or may radiate from the throat to the ear because of involvement of the tympanic branch of the glossopharyngeal nerve. Spasms of pain may be initiated by swallowing or coughing. There is no demonstrable motor or sensory deficit. Cardiac symptoms—bradycardia or asystole, hypotension, and fainting—

have been reported. Glossopharyngeal neuralgia can result from vascu lar compression, MS, or tumors, but many cases are idiopathic. Medical therapy is similar to that for trigeminal neuralgia, and carbamazepine is generally the first choice. If drug therapy is unsuccessful, surgical procedures—including microvascular decompression if vascular com pression is evident—or rhizotomy of glossopharyngeal and vagal fibers in the jugular bulb is frequently successful. ■ ■DYSPHAGIA AND DYSPHONIA The tenth cranial (vagus) nerve (Fig. 452-6) carries somatic sen sation from the posterior aspect of the external auditory canal,

Geniculotympanic nerve Pterygopalatine ganglion Great petrosal nerve Nerve of the pterygoid canal Facial nerve Otic ganglion Parotid gland FO Small petrosal nerve FR TP Auditory tube (eustachian) Deep petrosal nerve (sympathetic) Tympanic nerve (of jacobson) to tympanic plexus Internal carotid artery Tympanic nerve (of jacobson) to tympanic plexus Parasympathetic nerves

Styloglossus muscle Sensory nerves Communication with facial nerve Motor nerves Sympathetic nerves Stylopharyngeal muscle Tonsils laryngopharynx, superior larynx, and meninges of the posterior fossa to the spinal trigeminal nucleus, as well as taste from the epiglottis and pharynx and visceral sensation from chemoreceptors and barorecep tors in the aortic arch, heart, and gastrointestinal tract to the splenic flexure to the nucleus solitarius. The motor part originates in the nucleus ambiguous and innervates most muscles of the oropharynx and soft palate as well as all laryngeal muscles. Parasympathetic fibers originate in the dorsal motor nucleus of the vagus nerve and decrease the heart rate through action at the sinoatrial and atrioventricular nodes; others promote peristalsis and secretion of the alimentary tract from the esophagus to the splenic flexure. When the intracranial por tion of one vagus (tenth cranial) nerve is interrupted, the soft palate droops ipsilaterally and does not rise in phonation. There is loss of the gag reflex on the affected side, as well as of the “curtain movement” of the lateral wall of the pharynx, whereby the faucial pillars move medi ally as the palate rises in saying “ah.” The voice is hoarse and slightly

Nucleus of solitary tract Nucleus of spinal tract of V Dorsal motor nucleus of vagus Ambiguus nucleus XI C1 Spinal roots of accessory nerve Superior laryngeal nerve Sternocleidomastoid muscle C5 Trapezius muscle Arytenoid, thyroarytenoid, and cricoarytenoid muscles Esophagus Glottis Right subclavian artery Cardiac nerves Cardiac plexus Pulmonary plexus Esophageal plexus Celiac plexus Liver Gallbladder Right kidney Small intestine FIGURE 452-6 The vagus nerve. J, jugular (superior) ganglion; N, nodose (inferior) ganglion. (Reproduced with permission from SG Waxman: Clinical Neuroanatomy, 29th ed. New York, McGraw Hill, 2020.) nasal, and the vocal cord lies immobile midway between abduction and adduction. Loss of sensation at the external auditory meatus and the posterior pinna may also be present. The vagus nerve may be involved at the meningeal level by neo plastic and infectious processes and within the medulla by tumors, vascular lesions (e.g., the lateral medullary syndrome), and motor neuron disease. The nerve may be involved by infection with varicellazoster virus. Injury to the vagus nerve in the carotid sheath can occur with carotid dissection or following endarterectomy. The pharyngeal branches of both vagal nerves may be affected in diphtheria; the voice has a nasal quality, and regurgitation of liquids through the nose occurs during swallowing. Polymyositis and dermatomyositis, which cause hoarseness and dysphagia by direct involvement of laryngeal and pha ryngeal muscles, may be confused with diseases of the vagus nerves. Dysphagia is also a symptom in some patients with myotonic dystro phy. Nonneurologic causes of dysphagia are discussed in Chap. 47.

Meningeal branch to posterior fossa Auricular branch to posterior auricle and part of external meatus VII IX X J Muscles to palate and pharynx N Sensation to lower pharynx CHAPTER 452 Epiglottic and lingual rami Inferior pharyngeal constrictor Trigeminal Neuralgia, Bell’s Palsy, and Other Cranial Nerve Disorders
Cricothyroid muscle Right recurrent laryngeal nerve Left recurrent laryngeal nerve Left vagus nerve Aortic arch Diaphragm Stomach Spleen Pancreas Left kidney Sensory nerves Parasympathetic nerves Motor nerves The recurrent laryngeal nerves, especially the left, are most often damaged as a result of intrathoracic disease. Aneurysm of the aortic arch, an enlarged left atrium, and tumors of the mediastinum and bronchi are much more frequent causes of an isolated vocal cord palsy than are intracranial disorders. However, a substantial number of cases of recurrent laryngeal palsy remain idiopathic. When confronted with a case of laryngeal palsy, the physician must attempt to determine the site of the lesion. If it is intramedullary, there are usually other signs, such as ipsilateral cerebellar dysfunction, loss of pain and temperature sensation over

the ipsilateral face and contra lateral arm and leg, and an ipsilateral Horner's syndrome. If the lesion is extramedullary, the glossopharyngeal and spinal accessory nerves are frequently involved (jugular foramen syndrome). If it is extracranial in the posterior laterocondylar or retroparotid space, there may be a combination of ninth, tenth, eleventh, and twelfth cranial nerve palsies and Horner's syndrome (Table 452-2). If there is no sensory loss over

TABLE 452-2 Cranial Nerve Syndromes SITE CRANIAL NERVES USUAL CAUSE Orbital apex II, III, IV, first division V, VI Invasive fungal infections, amyloidosis, granulomatous disease Sphenoid fissure (superior orbital) III, IV, first division V, VI Invasive tumors of sphenoid bone; aneurysms Lateral wall of cavernous sinus III, IV, first division V, VI, often with proptosis Infection, thrombosis, aneurysm or fistula of cavernous sinus; invasive tumors from sinuses and sella turcica; benign granuloma responsive to glucocorticoids Retrosphenoid space II, III, IV, V, VI Large tumors of middle cranial fossa Apex of petrous bone V, VI Petrositis; tumors of petrous bone Internal auditory meatus VII, VIII Tumors of petrous bone (dermoids, etc.); infectious processes; acoustic neuroma PART 13 Neurologic Disorders Pontocerebellar angle V, VI, VII, VIII, and sometimes IX Acoustic neuroma; meningioma Jugular foramen IX, X, XI Tumors and aneurysms Posterior laterocondylar space IX, X, XI, XII Tumors of parotid gland and carotid body and metastatic tumors Posterior retroparotid space IX, X, XI, XII, and Horner's syndrome Tumors of parotid gland, carotid body, lymph nodes; metastatic tumor; tuberculous adenitis the palate and pharynx and no palatal weakness or dysphagia, the lesion is below the origin of the pharyngeal branches, which leave the vagus nerve high in the cervical region; the usual site of disease is then the mediastinum. ■ ■NECK WEAKNESS The eleventh cranial nerve (spinal accessory) is a pure motor nerve arising from the nucleus ambiguus and the ventral horn of the spinal cord from C1–C6. The nerve travels superiorly through the fora men magnum and exits through the jugular foramen to innervate the ipsilateral sternocleidomastoid and trapezius muscles. Isolated involvement of the accessory (eleventh cranial) nerve can occur any where along its route, resulting in partial or complete paralysis of the sternocleidomastoid and trapezius muscles. Spinal accessory nerve palsy does not result in significant neck weakness because several other muscles also turn the head and flex the neck; therefore, detection of accessory nerve injury relies on palpating the absence of sternoclei domastoid contraction during head turning. Similarly, shoulder shrug is only slightly impacted by trapezius weakness, although the affected shoulder is lower at rest, scapular winging occurs, and the arm cannot abduct beyond 90°. Isolated spinal accessory nerve palsy is often iat rogenic due to neck surgery or jugular vein cannulation, or traumatic. An idiopathic form of accessory neuropathy, akin to Bell's palsy, has been described, and it may be recurrent in some cases. Most but not all patients recover. ■ ■TONGUE PARALYSIS The twelfth cranial nerve (hypoglossal) supplies the ipsilateral muscles of the tongue. Nerve lesions cause the tongue to deviate toward the ipsilateral side during protrusion due to ipsilateral genioglossus weak ness, in addition to weakness of tongue movements toward the affected side to weakness of ipsilateral intrinsic tongue musculature. Atrophy and fasciculations of the tongue develop weeks to months after inter ruption of the nerve. The nucleus of the nerve or its fibers of exit may be involved by intramedullary lesions such as tumor, poliomyelitis, or most often motor neuron disease. Lesions of the basal meninges and the occipital bones (platybasia, invagination of occipital condyles, Paget's disease) may compress the nerve in its extramedullary course or as it exits the skull in the hypoglossal canal. Isolated lesions of unknown cause can occur.

MULTIPLE CRANIAL NERVE PALSIES When multiple cranial nerves are affected by a disease process, the clinical approach begins by determining whether the pathology lies within or outside of the

brainstem. Lesions that lie on the surface of the brainstem are characterized by involvement of adjacent cranial nerves (often occurring in succession) and late and rather slight involvement of the long sensory and motor pathways and segmental structures lying within the brainstem. The opposite is true of primary lesions within the brainstem. Extramedullary lesions are more likely to cause bone erosion or enlargement of the foramens of exit of cranial nerves. By contrast, intramedullary lesions involving cranial nerves often produce a crossed sensory or motor paralysis (cranial nerve signs on one side of the body and tract signs on the opposite side). Involvement of multiple cranial nerves outside the brainstem is frequently the result of trauma, localized infections including varicellazoster virus, infectious and noninfectious (especially carcinomatous) causes of meningitis (Chaps. 143 and 144), granulomatous diseases such as granulomatosis with polyangiitis (Chap. 375), Behçet’s dis ease (Chap. 376), vascular disorders including those associated with diabetes, enlarging aneurysms, or locally infiltrating tumors. Among the tumors, nasopharyngeal cancers, lymphomas, neurofibromas, meningiomas, chordomas, cholesteatomas, carcinomas, and sarcomas have all been observed to involve a succession of lower cranial nerves. Owing to their anatomic relationships, the multiple cranial nerve palsies form a number of distinctive syndromes, listed in Table 452-2. Sarcoidosis (Chap. 379) is the cause of some cases of multiple cranial neuropathy; tuberculosis, the Chiari malformation, platybasia, and basilar invagination of the skull are additional causes. Cavernous sinus syndrome (Fig. 452-7) is a distinctive and fre quently life-threatening disorder. It often presents as orbital or facial pain; orbital swelling, chemosis due to occlusion of the ophthalmic veins; fever; oculomotor neuropathy affecting the third, fourth, and sixth cranial nerves; and trigeminal neuropathy affecting the ophthal mic (V1) and occasionally the maxillary (V2) divisions of the trigemi nal nerve. Cavernous sinus thrombosis, often secondary to infection from orbital cellulitis (frequently Staphylococcus aureus), a cutaneous source on the face, or sinusitis (especially with mucormycosis in diabetic patients), is the most frequent cause; other etiologies include aneurysm of the carotid artery, a carotid-cavernous fistula (orbital bruit may be present), meningioma, nasopharyngeal carcinoma, other tumors, or an idiopathic granulomatous disorder (Tolosa-Hunt syn drome, discussed below). The two cavernous sinuses directly commu nicate via intercavernous channels; thus, involvement on one side may extend to become bilateral. Early diagnosis is essential, especially when due to infection, and treatment depends on the underlying etiology. Ant. cerebral a. Int. carotid a. Ant. clinoid process Subarachnoid space Optic chiasm Oculomotor (III) n. Trochlear (IV) n. Hypophysis Ophthalmic (V1) n. Maxillary (V2) n. Sphenoid sinus Pia Arachnoid Dura Abducens (VI) n. FIGURE 452-7 Anatomy of the cavernous sinus in coronal section, illustrating the location of the cranial nerves in relation to the vascular sinus, internal carotid artery (which loops anteriorly to the section), and surrounding structures.

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