diabetes and hypertension are common comorbid conditions. Cranial nerves I, VI, and VII are the most frequently affected after minor head trauma.\(^1\) Trigeminal neuralgia is a common cause of facial pain that affects approximately 4.5 per 100,000 individuals; women are affected twice as often as men, and it is more common in those older than 60 years.\(^2\) Trigeminal neuralgia can be severely debilitating and has been termed the “suicide disease.”\(^3\) Bell palsy is the most common cause of acute facial paralysis worldwide. The peak age at incidence has been reported to be between 15 and 45 years,\(^4\) but other investigators have noted an increased incidence in individuals older than 70.\(^5,6\) Pregnant women and patients with diabetes have an associated increased incidence of the disease. A familial association of Bell palsy is noted in 4% of cases,\(^4\) and it can cause both significant psychologic and physical morbidity.

**PATHOPHYSIOLOGY**

**CRANIAL NERVE I (OLFACTORY NERVE)**

**Anatomy**
Cranial nerve I is a special sensory nerve that provides the sense of smell. Inhaled scents are detected by the olfactory epithelium lining the nasal cavity and transmitted to the olfactory bulb, which lies adjacent to the cribriform plate of the ethmoid bone. Olfactory sensations are relayed from the olfactory bulb to the brain via the olfactory tract.

**Presenting Signs and Symptoms**
The patient should be questioned about a history of head trauma. An anteroposterior skull fracture parallel to the sagittal suture or an anteroposterior shearing injury can tear the olfactory fibers traversing the cribriform plate and lead to disruption of the synapses from the olfactory epithelium to the olfactory bulb.

A frontal lobe mass such as a tumor, meningioma, or abscess can compress the olfactory bulb as well, but the signs and symptoms associated with such masses tend to be more subacute.

**Treatment**
Treatment depends on the presence of concomitant injury. Basilar skull fracture and cerebrospinal fluid rhinorrhea associated with trauma require immediate neurosurgical consultation. A subacute mass or abscess should be managed in...
consultation with neurosurgery, depending on the acuity of the findings. Patients with anosmia secondary to trauma and normal findings on head computed tomography (CT) can referred to neurology or neurosurgery for outpatient follow-up.

CRANIAL NERVE II (OPTIC NERVE)

Anatomy

Visual stimuli are transmitted from the retina to the optic nerve through the optic chiasm to the lateral geniculate nucleus in the thalamus, where they synapse. From there, impulses are transmitted along the optic radiations (geniculocalcarine tracts, including the Meyer loop) to the primary visual cortex in the occipital lobes.

Presenting Signs and Symptoms

Unilateral loss of vision is most common with injuries to the optic nerve. Patients with bilateral visual loss may not be aware of any such injury until an examination is performed. Acute visual loss is often of vascular origin, including central retinal arterial or venous occlusion and cerebrovascular disease. Neurologic causes, such as multiple sclerosis, may be suggested by progression of the visual loss over a period of hours or days, pain, and a history of additional neurologic complaints with a recurrent waxing and waning pattern. Inflammatory processes such as optic neuritis may be the initial symptom of multiple sclerosis.

Neuropathy from temporal arteritis usually occurs in elderly patients and is associated with progressive loss of vision (unilaterally or bilaterally), constitutional symptoms, jaw claudication, and headache.

Idiopathic intracranial hypertension should be considered in patients with a history of headache, visual scotomata, and visual changes. The typical patient is a young, heavy-set woman who is taking oral contraceptives. The headache and visual changes are typically worsened by coughing, bending over, or performing techniques such as the Valsalva maneuver.

Orbital compressive tumors or aneurysms cause mass effects that compromise optic nerve function.

Differential Diagnosis

The differential patterns of visual loss are described in Box 95.1.

Treatment

Treatment depends on the cause. Emergency ophthalmologic consultation is essential for vascular causes. Treatment of central retinal artery occlusion should focus on lowering intraocular pressure. Inpatient evaluation for neurologic causes is warranted depending on the clinical findings. Temporal arteritis requires high-dose steroid therapy. Idiopathic intracranial hypertension requires urgent diagnostic and therapeutic lumbar puncture.

CRANIAL NERVE III (OCULOMOTOR NERVE)

Anatomy

The oculomotor nerve is a pure motor nerve that works in conjunction with cranial nerves IV and VI to coordinate extraocular movements. The oculomotor nerve controls the superior rectus (globe elevator), medial rectus (globe adductor), inferior rectus (globe depressor), and inferior oblique (globe elevator) muscles. It also controls the levator palpebrae superioris muscle (upper eyelid elevator) and the intrinsic visceral motor function of the sphincter pupillae muscles and the ciliary muscles, which perform pupillary constriction and accommodation, respectively.

Presenting Signs and Symptoms

The patient typically complains of double vision or difficulty seeing out of the affected eye. There may be mild photophobia in bright light. The patient may also complain of an inability to raise the eyelid (ptosis).

Cranial nerve III palsy is more common in patients older than 60 years and in those with diabetes or hypertension (Fig. 95.1).

Patients with herniation syndromes will have a history of trauma (Fig. 95.2), tumor, or other neurologic findings.

Pain associated with unilateral mydriasis should alert the emergency physician (EP) to look for an aneurysm involving the terminal internal carotid artery. Computed tomographic
Patients with an abscess or cavernous sinus thrombosis may have headaches, altered mental status, and seizures. This diagnosis should be considered in patients with signs and symptoms in the contralateral eye, previous sinus or midface infection, fever, chemosis, eyelid or periorbital edema, and exophthalmos. Extension of internal carotid artery dissection intracranially into the cavernous sinus can result in third, fourth, and sixth cranial nerve palsies.\(^9\)

**Presenting Signs and Symptoms**

Patients with a fourth cranial nerve palsy have double vision exacerbated by looking downward. The classic complaint is difficulty going down stairs. Most commonly, a history of trauma is reported. On physical examination the patient may unconsciously tilt the head away from the affected side (Fig. 95.3). Etiologic mechanisms are similar to those for the third cranial nerve and include inflammatory processes, trauma, and vascular causes.\(^10\)

**Treatment**

Treatment of isolated fourth nerve palsy is generally conservative, and the patient should be referred to neurology or neurosurgery as appropriate.\(^10\) CT, MRI, and neurology consultation are warranted if multiple cranial nerves are involved.

**CRANIAL NERVE IV (TROCHLEAR NERVE)**

**Anatomy**

The trochlear nerve innervates the superior oblique muscle of the eye and causes inward rotation and downward and lateral movement of the globe. It is the smallest cranial nerve but has the longest intracranial course.

**Presenting Signs and Symptoms**

Patients with trigeminal nerve dysfunction have either sensory or motor deficits. Sensory dysfunctions include paroxysmal pain, paresthesias (abnormal sensations such as burning, pricking, tickling, or tingling), dysesthesias (disagreeable, unpleasant, or painful sensations produced by ordinary stimuli), and anesthesia (loss of sensation). The motor dysfunction is usually described as difficulty chewing and difficulty swallowing.

Peripheral lesions cause loss of sensation or pain in only one division. Positive findings in two or more divisions (e.g., loss of light touch in one division and loss of sensitivity to pain, temperature, or pinprick in another division) should raise suspicion for a central cause.

**CRANIAL NERVE V (TRIGEMINAL NERVE)**

**Anatomy**

The trigeminal nerve is a mixed motor and sensory nerve. It provides motor innervation to the muscles of mastication, as well as sensation from the face, scalp, conjunctiva, globe, mucous membranes of the sinuses, tongue, teeth, and part of the external tympanic membrane.

The trigeminal sensory ganglion is located in the middle cranial fossa and branches into three divisions: the ophthalmic nerve (V1), the maxillary nerve (V2), and the mandibular nerve (V3).

**Presenting Signs and Symptoms**

Patients with trigeminal nerve dysfunction have either sensory or motor deficits. Sensory dysfunctions include paroxysmal pain, paresthesias (abnormal sensations such as burning, pricking, tickling, or tingling), dysesthesias (disagreeable, unpleasant, or painful sensations produced by ordinary stimuli), and anesthesia (loss of sensation). The motor dysfunction is usually described as difficulty chewing and difficulty swallowing.

Peripheral lesions cause loss of sensation or pain in only one division. Positive findings in two or more divisions (e.g., loss of light touch in one division and loss of sensitivity to pain, temperature, or pinprick in another division) should raise suspicion for a central cause.
The presence of associated cranial nerve deficits (III, IV, V, or any combination of these nerves) suggests cavernous sinus involvement. In the setting of trauma, if a bruise over the orbit can be detected, a carotid–cavernous sinus fistula may be present. Associated involvement of cranial nerve VII or VIII or gait ataxia should raise suspicion for a cerebellopontine angle or lateral pontine tumor (Table 95.1).

Associated Horner syndrome may indicate a cervical or lateral brainstem lesion.

The main categories of trigeminal nerve dysfunction are trigeminal neuralgia and trigeminal neuropathy. A sudden onset of symptoms should raise suspicion for a vascular, traumatic, or demyelinating cause, whereas a more indolent course suggests tumor or inflammation (Table 95.2).

**TRIGEMINAL NEUROPATHY** Causes include compression by an extrinsic mass, trauma, and vascular, inflammatory, or demyelinating disorders.

Symptoms include neuralgia or paresthesia (or both) involving half of the face. Unlike trigeminal neuralgia, the pain with trigeminal neuropathy is more constant. Loss of the corneal reflex is evident. The patient’s mouth may become more oval.

### Table 95.1 Clinicoanatomic Correlation of Localization of Lesions of Cranial Nerve V

<table>
<thead>
<tr>
<th>ANATOMIC SITE OF DAMAGE</th>
<th>CLINICAL FINDINGS</th>
<th>OTHER NEUROLOGIC AND MEDICAL FINDINGS</th>
<th>COMMON CAUSES</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supranuclear</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sensory cortex</td>
<td>Facial numbness, paresthesias</td>
<td>Neglect, apraxia, aphasia</td>
<td>Stroke, tumor, hemorrhage</td>
</tr>
<tr>
<td>Internal capsule</td>
<td>Hemifacial sensory loss</td>
<td>Hemiparesis of the arm</td>
<td>Stroke, tumor, hemorrhage, MS</td>
</tr>
<tr>
<td>Corona radiata</td>
<td>Central seventh cranial nerve paresis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VPM thalamus</td>
<td>Facial numbness, paresthesias, pain; cheirooral syndrome</td>
<td>Anosmia, hemisensory deficit</td>
<td>Stroke, tumor, hemorrhage</td>
</tr>
<tr>
<td>Midbrain</td>
<td>Facial numbness, paresthesias, pain</td>
<td>Ophthalmoparesis</td>
<td>Stroke, MS, tumor, aneurysm</td>
</tr>
</tbody>
</table>

**Nuclear**

| Pons                     | Facial numbness and weakness, paresthesias, pain; trigeminal neuralgia | Ophthalmoparesis; CN VI, CN VII, CN VIII palsies; Horner syndrome | Stroke, tumor, hemorrhage; MS, syringobulbia, abscess, trauma |
| Medulla                  | Facial numbness, paresthesias, pain; trigeminal neuralgia | Ataxia, CN X palsy, ophthalmoparesis, nystagmus, Horner syndrome, Wallenberg syndrome | Stroke, MS, tumor, aneurysm, abscess, vasculopathy |

**Preganglionic**

| Cerebellopontine angle | Facial numbness | CN VII, CN VIII palsies; headache, cerebellar dysergia | Neuroma, meningioma, meningitis (bacterial, TB, cancer), aneurysm, trauma |
| Middle cranial fossa   |                 |                                                       |               |
| Gasserian ganglion      | Facial numbness and weakness | Gradenigo syndrome; CN VI, CN VII palsies | Tumor, infection, trauma |
| Skull base              | Facial numbness and weakness | Headache, meningismus | Meningitis (bacterial, TB, cancer, sarcoid) |

**Trigeminal Nerve Branches**

| V1: cavernous sinus     | Facial numbness, pain | Headache, ophthalmoparesis; Horner syndrome | Tumor, thrombosis, infection, trauma |
| V1: Carotid-cavernous fistula | Facial numbness | Proptosis, bruit, ophthalmoparesis | Trauma |
| V2: Maxillary region    | Facial numbness; numb cheek syndrome |                                      | Tumor, infarct, vasculopathy, trauma |
| V3: Mandibular region   | Weakness of mastication; numb chin syndrome |                                      | Tumor, trauma, infarct |

CN, Cranial nerve; MS, multiple sclerosis; TB, tuberculosis; VPM, ventroposteromedial.
Table 95.2  Selected Specific Causes Associated with Trigeminal Nerve Disorders

<table>
<thead>
<tr>
<th>ETIOLOGIC CATEGORY</th>
<th>SELECTED SPECIFIC CAUSES</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Structural Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Developmental</td>
<td>Brainstem vascular loop, syringobulbia</td>
</tr>
<tr>
<td>Degenerative and compressive</td>
<td>Paget disease</td>
</tr>
<tr>
<td><strong>Hereditary and Degenerative Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Chromosomal abnormalities, neurocutaneous disorders</td>
<td>Hereditary sensorimotor neuropathy type I, neurofibromatosis (schwannoma)</td>
</tr>
<tr>
<td>Degenerative motor, sensory, and autonomic disorders</td>
<td>Amyotrophic lateral sclerosis</td>
</tr>
<tr>
<td><strong>Acquired Metabolic and Nutritional Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Endogenous metabolic disorders</td>
<td>Diabetes</td>
</tr>
<tr>
<td>Exogenous disorders (toxins, illicit drugs)</td>
<td>Trichloroethylene, trichloroacetic acid</td>
</tr>
<tr>
<td>Nutritional deficiencies, syndromes associated with alcoholism</td>
<td>Thiamine, folate, vitamin B12, pyridoxine, pantothenic acid, vitamin A deficiencies</td>
</tr>
<tr>
<td><strong>Infectious Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Viral infections</td>
<td>Herpes zoster, unknown</td>
</tr>
<tr>
<td>Nonviral infections</td>
<td>Bacteria, tuberculous meningitis, brain abscess, Gradenigo syndrome, leprosy, cavernous sinus thrombosis</td>
</tr>
<tr>
<td>HIV infection, AIDS</td>
<td>Opportunistic infection; abscess, herpes zoster Stroke, hemorrhage, aneurysm</td>
</tr>
<tr>
<td><strong>Neurovascular Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Neoplastic Disorders</td>
<td></td>
</tr>
<tr>
<td>Primary neurologic tumors</td>
<td>Glial tumors, meningioma, schwannoma</td>
</tr>
<tr>
<td>Metastatic neoplasms, paraneoplastic syndromes</td>
<td>Lung, breast; lymphoma, carcinomatous meningitis</td>
</tr>
<tr>
<td><strong>Demyelinating Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Central nervous system disorders</td>
<td>Multiple sclerosis, acute demyelinating encephalomyelitis</td>
</tr>
<tr>
<td>Peripheral nervous system disorders</td>
<td>Guilain-Barré syndrome, chronic inflammatory demyelinating polynuropathy, Tolosa-Hunt syndrome, sarcoidosis, lupus, orbital pseudotumor</td>
</tr>
<tr>
<td><strong>Autoimmune and Inflammatory Disorders</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Traumatic Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Carotid-cavernous fistula, cavernous sinus thrombosis, maxillary/mandibular injury</td>
<td></td>
</tr>
<tr>
<td><strong>Epilepsy</strong></td>
<td></td>
</tr>
<tr>
<td>Focal seizures</td>
<td></td>
</tr>
<tr>
<td><strong>Headache and Facial Pain</strong></td>
<td></td>
</tr>
<tr>
<td>Raeder neuralgia, cluster headache</td>
<td></td>
</tr>
<tr>
<td><strong>Drug-Induced and Iatrogenic Neurologic Disorders</strong></td>
<td></td>
</tr>
<tr>
<td>Orbital, facial, dental surgery</td>
<td></td>
</tr>
</tbody>
</table>

AIDS, Acquired immunodeficiency syndrome; HIV, human immunodeficiency virus.
and oblique in appearance, and because of loss of masseter muscle strength, the chin may be deviated toward the affected side.

Until proved otherwise, neuropathies of cranial nerve V, the chin (numb chin; V3), and the suborbital region (numb cheek) should be presumed to be due to malignancies.11

TIC DOULOUREUX The term tic douloureux was coined by Nicolaus André, a French surgeon, in 1756. Its mechanism is probably compression of the trigeminal nerve root within millimeters of entry into the pons.12 The maxillary and mandibular divisions are most commonly affected, either alone or in combination. In one longitudinal case series, no cases of trigeminal neuralgia affecting both the ophthalmic and mandibular divisions were reported.2 Causes of tic douloureux are listed in Box 95.2.

The International Association for the Study of Pain defines tic douloureux as “a sudden usually unilateral, severe, brief, stabbing, recurrent pain in the distribution of one or more branches of the fifth cranial nerve.” The pain is classically precipitated by normal activities such as eating, talking, washing the face, or cleaning the teeth.

Diagnostic Testing
The presence or absence of a corneal reflex should be checked. An intact reflex indicates normal function of the afferent V1 division, as well as normal cranial nerve VII motor efferent function. Absence of a corneal reflex can be caused by tumors in the posterior fossa or cerebellopontine angle, multiple sclerosis, brainstem strokes (Wallenberg or lateral medullary syndrome), and Parkinson disease.

Motor function is evaluated by having the patient open and close the mouth and laterally deviate the jaw against resistance. Loss of muscle bulk or the presence of fasciculations in the temporalis or masseter musculature indicates a lower motor neuron lesion.

The jaw jerk reflex test determines the integrity of the V3 division. The examiner places a thumb on the patient’s chin, after which the patient is instructed to relax the jaw completely with the mouth closed, and the examiner then taps the chin to elicit the jaw jerk reflex. The reflex will be diminished in patients with a lower motor neuron lesion and accentuated in patients with a supranuclear lesion.

Treatment
A trial of carbamazepine can be therapeutic as well as diagnostic because failure to improve with carbamazepine suggests some other cause. Treatment options are listed in Box 95.3.3 Surgical approaches are considered when medication cannot control the pain or pain medication is not tolerated.13

CRANIAL NERVE VI (ABDUCENS NERVE) Anatomy
The abducens nerve is a pure motor nerve that supplies the ipsilateral lateral rectus muscle of the eye and controls globe abduction.

Presenting Signs and Symptoms
Patients with an abducens nerve palsy usually complain of double vision. The head may be turned away from the affected side to maintain binocularity. Diabetes and hypertension are common risk factors. Another common sign is “crossed eyes” (esotropia or strabismus) (Fig. 95.4).14

Differential Diagnosis
Children are more likely to have a tumor as the principal cause, and older individuals are more likely to have an ischemic cause such as temporal arteritis.

An abducens nerve palsy occurring in isolation is rare. Usually, the seventh and eighth cranial nerves are also involved, which signals a central cause. Causes of abducens nerve palsy are listed in Box 95.4.

BOX 95.2 Causes of Tic Douloureux

| Vascular compression by an artery or vein |
| Saccular aneurysm |
| Arteriovenous malformation |
| Vestibular schwannomas |
| Meningioma |
| Epidermoid cyst |
| Tumor |
| Primary demyelinating disorders |
| • Multiple sclerosis |
| • Charcot-Marie-Tooth disease (rare) |
| Infiltrative disorders |
| • Trigeminal amyloidoma |
| Nondemyelinating lesions |
| • Small infarct or angioma in the brainstem |
| Familial |

BOX 95.3 Treatment of Trigeminal Neuralgia

| First-Line Agent |
| Carbamazepine (Tegretol)—Start at 150 mg daily and increase by 100 mg every 3 days as needed to a total daily dose of 800 to 1600 mg divided into three doses. |
| Second-Line Agents |
| Oxcarbazepine (Trileptal)—Start at 300 mg daily and increase by 300 mg every 3 days as needed to a total daily dose of 1200 to 1800 mg divided into two doses. |
| Gabapentin (Neurontin)—Start at 300 mg three times daily and increase as needed to a total daily dose of 3600 mg divided into three doses. Also commonly used as first-line therapy. |
| Phenytoin (Dilantin)—Start at 300 mg daily and increase as needed, divided into two or three doses. |
| Third-Line Agents (Add-On Therapy or Monotherapy) |
| Lamotrigine (Lamictal)—Start at 25 mg daily and increase by 25 mg every 7 days as needed to a total daily dose of 200 to 400 mg divided into two doses. |
| Baclofen (Lioresal)—Start at 15 mg daily and increase by 5 mg every 3 days as needed to a total daily dose of 60 to 80 mg divided into three doses. |

The palsy is often preceded by a viral syndrome, and a correlation has been noted with herpes simplex virus (HSV). Its association with shingles and the characteristic blistering (from varicella-zoster virus [VZV]) is given the designation Ramsay Hunt syndrome. Reactivation of VZV has also been theorized as a cause. In addition, Bell palsy may be seen in patients with Lyme disease in places where the disease is endemic. Diabetes, hypertension, human immunodeficiency virus infection, sarcoidosis, Sjögren syndrome, parotid nerve tumors, eclampsia, amyloidosis, and the intranasal influenza vaccine have been associated with the development of Bell palsy.

Other common triggers include stress, trauma, fever, tooth extraction, and a chilling episode from exposure to drafts and cold.

Complete facial weakness, severe non–ear-related pain (e.g., retroauricular, cheek), late onset of recovery or no recovery by 3 weeks, diabetes, pregnancy, age older than 60 years, hypertension, and Ramsay Hunt syndrome are risk factors for incomplete recovery.5,18

Electroneurographic studies demonstrate a steady decline in electrical activity on days 4 to 10. When excitability is retained, 90% of patients recover fully, but when excitability diminishes to absence, only 20% of patients recover completely.

The pathophysiology of Bell palsy has not been clearly established. Several theories have been proposed, including infectious or ischemic inflammation leading to nerve compression within the narrow canal as the nerve exits the stylomastoid foramen. Because the nerve is encased in a tight dural sheath within the temporal bone, this edema then causes additional compression of the vascular supply to the nerve.15

The cause of Bell palsy is most commonly idiopathic (66%).4 Numerous observed associations have been described in the literature. The palsy is often preceded by a viral syndrome, and a correlation has been noted with herpes simplex virus (HSV). Its association with shingles and the characteristic blistering (from varicella-zoster virus [VZV]) is given the designation Ramsay Hunt syndrome. Reactivation of VZV has also been theorized as a cause. In addition, Bell palsy may be seen in patients with Lyme disease in places where the disease is endemic.

Diabetes, hypertension, human immunodeficiency virus infection, sarcoidosis, Sjögren syndrome, parotid nerve tumors, eclampsia, amyloidosis, and the intranasal influenza vaccine have been associated with the development of Bell palsy.

Other common triggers include stress, trauma, fever, tooth extraction, and a chilling episode from exposure to drafts and cold.

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Electroneurographic studies demonstrate a steady decline in electrical activity on days 4 to 10. When excitability is retained, 90% of patients recover fully, but when excitability diminishes to absence, only 20% of patients recover completely.5

**Anatomy**

Cranial nerve VII is a mixed motor and sensory cranial nerve, which accounts for the varied symptoms. It travels adjacent to cranial nerves V, VI, and VIII as it traverses the cerebellar pontine angle, the internal auditory meatus, and the temporal bone.

Motor function involves the muscles of facial expression, the posterior digastic muscle, the stylohyoid muscle, and the stapedius muscle of the inner ear.
Parasympathetic innervation includes the lacrimal glands, the mucous membranes of the nose, the hard and soft palate, and the submandibular and sublingual glands.

The geniculate ganglion contains the nerve cell bodies of the sensory taste fibers of the anterior two thirds of the tongue.\(^5\)

**Presenting Signs and Symptoms**

To the patient, the most alarming symptom of Bell palsy is the abrupt onset of unilateral facial paralysis. Approximately 50% of patients believe that they have suffered a stroke, 25% think that they have an intracranial tumor, and the remaining 25% have no clear conception of what is wrong but are extremely anxious.\(^6\)

The EP may note drooping of the eyebrow or the corner of the mouth (or both) and loss of wrinkles on the forehead or the nasolabial folds (or both). Inability to raise the eyebrow and furrow the forehead is a cardinal sign of Bell palsy (Fig. 95.5). Preservation of forehead motor neuron innervation should raise suspicion for a central cause.\(^6\) Because the forehead receives bilateral upper motor neuron innervation, a central stroke will spare the forehead and allow the patient to raise the eyebrow. If the patient can do this, it is not Bell palsy.

Loss of nasolabial fold and nasal flaring is common. Loss of buccinator strength causes an inability to blow out the cheeks. An inability to close the eye on the affected side is a hallmark of Bell palsy. Speech is affected and may sound slurred or garbled, similar to dysarthria from a stroke. An asymmetric smile is often noted on examination (Fig. 95.6).

The signs and symptoms vary depending on the site of the affected nerve. They are listed in Box 95.5.

**Diagnostic Testing and Differential Diagnosis**

The “blow out your cheeks” test (Fig. 95.7) demonstrates loss of buccinator function. A sensitive variation of this test is to ask patients to hold water in their mouth and contract the buccal muscles. The water will either dribble out of the corner of the mouth or shoot across the room.

On testing of hearing, hyperacusis may be observed on the affected side because of denervation of the stapedius. The patient should have no hearing loss.

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**BOX 95.5 Signs and Symptoms of Bell Palsy**

- Ipsilateral tongue numbness
- Loss of taste or a dull taste
- Overt paralysis preceded by a sensation of subjective numbness or weakness on the affected side
- Ear pain in the external auditory canal
- Retroauricular pain
- Occipital headache
- Hyperacusis
- Fullness or snapping sound in the affected ear
- Tinnitus
- Drooling
- Inability to keep liquids in the mouth or chew
- Noticeable dryness of the oral and nasal mucous membranes on the affected side
- Anxiety
To evaluate taste sensation, a few granules of sugar are placed on the tip of the patient’s tongue on the affected side. Decreased taste sensation may be noted.

Other cranial nerves should be normal. The abducens nucleus lies at the level of the genu of cranial nerve VII; infarction in the area can cause concomitant palsy of cranial nerve VI, which signals an upper motor neuron lesion rather than Bell palsy. No evidence of expressive or receptive aphasia should be present.

The presence of vesicles on the tympanic membrane or in the oropharynx (Fig. 95.8) or the presence of grouped vesicular lesions on the face or around the ear (Fig. 95.9) suggests a diagnosis of Ramsay Hunt syndrome.

Residual synkinesis can result from abnormal regeneration of nerve fibers. This can be manifested as abnormal motor function (e.g., blinking causes involuntary contracture of the risorius); as abnormal parasympathetic function, which is classically accompanied by “crocodile tears”—lacrimation after a salivary stimulus; or as hemifacial spasm, which can be bothersome, especially when the patient is tired.

**Treatment**

The algorithm shown in Figure 95.10 outlines the treatment of patients with Bell palsy.

Patients can be discharged home with oral medication, instructions for eye care, and expedited follow-up with a neurologist. Additional investigation for Lyme disease may be indicated for patients at risk.

The evidence available indicates that steroids are safe and effective in shortening the course of the neurologic deficit and improving facial function. Patients receiving steroid therapy are up to 1.2 times more likely to attain good functional outcomes than untreated patients are. Corticosteroids reduce the risk for unsatisfactory recovery by 9%, with the number needed to benefit (NNTB) being 11. Corticosteroids were also associated with a 14% absolute reduction in risk for synkinesis and autonomic dysfunction, with an NNTB of 7.

No studies have demonstrated significantly worse facial functional outcomes in patients treated with steroids.

The most commonly reported treatment regimen is oral prednisone, 1 mg/kg up to 70 mg/day for a 10-day course. Dosing can be once daily or split into twice daily. The starting dose is continued for 6 days and tapered over the next 4 days. Alternatively, prednisone, 1 mg/kg/day, may be given for 7 days without a taper.

Recent studies and metaanalyses have questioned the benefit of using antivirals for the treatment of Bell palsy. Antiviral agents used alone did not provide any benefit over placebo, and their use as the sole therapeutic agent is not recommended. When combined with corticosteroid therapy, antiviral therapy may have incremental benefit, but this remains to be shown conclusively. Therefore, until definitive studies are performed, clinical judgment will probably guide the use of antiviral therapy in cases in which a viral cause is strongly suspected (i.e., patients in whom HSV or VZV is

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**Fig. 95.7** “Blow out your cheeks.” Loss of buccinator function prevents pursing of the lips and allows air (and food and liquids) to escape.

**Fig. 95.8** Buccal herpetic lesions in an individual with Ramsay Hunt syndrome.

**Fig. 95.9** Characteristic auricular rash of Ramsay Hunt syndrome.
Algorithm outlining the treatment of patients with Bell palsy. CSF, Cerebrospinal fluid; ESR, erythrocyte sedimentation rate; HIV, human immunodeficiency virus; MRI, magnetic resonance imaging.

**Prognosis**

In a prospective study describing the spontaneous untreated course of idiopathic peripheral nerve palsy in patients with diabetes, 38% of patients had complete palsies, and only 25% regained normal facial muscle function. This is significantly worse than the observed rate of spontaneous full recovery in nondiabetic patients. Recurrence is rare (6.3%) and should prompt a work-up for other causes such as myasthenia gravis, lymphoma, sarcoidosis, Lyme disease, and rarely, Guillain-Barré syndrome. Although the prognosis for recovery is good, the psychologic consequences can be long-lasting and are perhaps more significant than the physical disability. Patients report self-consciousness about the facial disfigurement, fear of permanent disfigurement, loss of self-esteem, and social ostracism.

**CRANIAL NERVE VIII (VESTIBULOCOCHLEAR NERVE)**

**Anatomy**

The vestibulocochlear nerve is a special sensory nerve that transmits auditory signals from the cochlea (hearing) and signals from the semicircular canals (balance). The vestibular apparatus also sits in the petrous temporal bone and is composed of a body consisting of the saccule and utricle and three semicircular canals aligned in three different planes. Hair cells within the endolymp of the canals detect angular movement and transmit the impulses to the vestibular nuclear complex in the floor of the fourth ventricle. The hair cells collectively combine to form the vestibular ganglion.

**Presenting Signs and Symptoms**

Patients with vestibulocochlear nerve dysfunction usually exhibit various degrees of hearing loss, tinnitus, vertigo, falling, and imbalance. The mechanism is asymmetric integration of vestibular input to the central nervous system or asymmetric disruption of sensory input from the vestibular organs. If the vertigo is severe, nausea and vomiting also occur. Symptoms may be constant or episodic. Vestibular neuritis causes vertigo that lasts for weeks, and central vertigo may persist for years.

Patients should be asked about triggers, particularly positional triggers because this may indicate benign paroxysmal positional vertigo. Recent viral and upper respiratory tract infections may be significant because they predispose to vestibular neuritis. The history should also include the use of medications such as anticonvulsants, antihypertensives, sedatives, and ototoxic drugs.

The examination should be focused on determining reproducibility of the symptoms, gait, balance, and ataxia; on evaluation of possible acute stroke symptoms; and on the character of the nystagmus and severity of the ataxia. The presence or absence of associated cerebellar signs such as lateralizing dysmetria, motor weakness, sensory loss, and abnormal reflexes should be noted, as well as the Babinski reflex and cranial nerve abnormalities such as ophthalmoplegia, dysarthria, and Horner syndrome. Abnormalities in cerebellar function should prompt consideration of a central cause. Patients should also be examined for vertical and rotary nystagmus, which are not typically present in patients with peripheral vertigo; their presence warrants imaging and neurologic evaluation.

**Diagnostic Testing and Differential Diagnosis**

The Dix-Hallpike maneuver is commonly used to elicit positional nystagmus (see Fig. 96.1), which is associated with benign paroxysmal positional vertigo and usually lasts 5 to 60 seconds. Prolonged nystagmus is unlikely to be a result of this disorder. Gait and balance can be assessed with tandem walking and the Romberg test. Ataxia and lateralizing dysmetria can be assessed with finger-to-nose and heel-knee-shin testing. Hearing can be evaluated with the finger rub or finger snap, the Weber test, and the Rinne test. The ear and external auditory canal should be examined for evidence of cerumen.
oitis media, perforation of the tympanic membrane, and mass lesions.

CT lacks sensitivity in the evaluation of cranial nerve VIII disorders but may be useful in evaluating the bony temporal region. MRI with gadolinium enhancement is useful in identifying acoustic neuroma.

When a central cause is suspected because of abnormalities on cerebellar testing or clinical suspicion, MRI or magnetic resonance angiography (or both) should be performed to rule out a posterior circulation stroke as a central cause of the vertigo.

The differential diagnosis should include other cranial nerve deficits that are not typically present in benign causes of cranial nerve VIII dysfunction. Acoustic neuromas may compress the trigeminal nerve when they attain a size of 3 cm or greater; patients with complaints of facial numbness should therefore be evaluated for trigeminal neuropathy, as well for a mass lesion. Because large tumors can affect cranial nerves IX, X, and XI, these nerves should also be tested.

**Treatment**

Some patients who come to the emergency department with sudden or severe symptoms may not be able to comply with testing because the severity of the symptoms limits the ability to open their eyes and turn their head without experiencing nausea and vomiting or exacerbating the symptoms. In these cases it is appropriate to treat the patient symptomatically, initiate a work-up, and reassess clinically for improvement before attempting to move the patient or perform provocative testing.

**CRANIAL NERVE IX (GLOSSOPHARYNGEAL NERVE) Anatomy**

The glossopharyngeal nerve provides branchial motor function to the stylopharyngeus muscle; visceral motor function to the otic ganglion and parotid gland; visceral sensory function from the carotid body; somatic sensory function to the posterior third of the tongue, the skin of the external ear, and the internal surface of the tympanic membrane; and the special sensory function of taste sensation from the posterior third of the tongue.

**Presenting Signs and Symptoms**

Patients with glossopharyngeal nerve palsy usually have associated symptoms involving other cranial nerves, most commonly cranial nerves X and XI. The most common symptoms are dysphagia and choking. If the vagus nerve is involved, the patient complains of hoarseness and demonstrates ipsilateral paralysis of the soft palate. Head, neck, and oral trauma or surgery can cause acute dysfunction of cranial nerve IX. Glossopharyngeal nerve palsy is a known complication of tonsillectomy surgery.³³

Glossopharyngeal neuralgia is a rare disorder consisting of paroxysms of pain in the back of the throat and tongue. The pain is similar to that of trigeminal neuralgia in that the attacks are brief, lasting seconds to minutes. It is unilateral and usually triggered by chewing, swallowing, coughing, or sneezing.

**Treatment**

CT scanning is warranted to evaluate for a cerebrovascular event or tumor. Rarely, vasovagal syncope can result from bradycardia or asystole caused by vagus nerve cardioinhibitory input. Medical management is similar to that for trigeminal neuralgia. If involvement of other cranial nerves is evident on examination, the patient should be admitted for further evaluation and neurologic consultation.

**CRANIAL NERVE X (VAGUS NERVE) Anatomy**

The vagus nerve is a mixed motor and sensory nerve that provides motor function to striated muscle of the pharynx, tongue, larynx, and tensor veli palatini, as well as motor function to smooth muscle and glands of the pharynx, larynx, and thoracic and abdominal viscera. Cranial nerve X provides general sensation from the skin at the back of the ear, the external auditory meatus, the pharynx, and part of the external surface of the tympanic membrane, as well as visceral sensation from the larynx, trachea, esophagus, and thoracic and abdominal viscera; from chemoreceptors in the aortic bodies; and from stretch receptors in the walls of the aortic arch.

**Presenting Signs and Symptoms**

Patients with palsies of the vagus nerve generally have hoarseness or difficulty swallowing. A history of recent carotid or thyroid surgery should prompt suspicion for a recurrent laryngeal nerve injury. The patient may also complain of regurgitation of food and liquid into the nose. Oropharyngeal examination usually reveals a drooped arch of the soft palate and uvular deviation away from the affected side.

**Treatment**

A CT scan of the head without contrast enhancement should be performed to evaluate for a cerebrovascular accident (hemorrhagic or ischemic) or skull-based lesions. Further inpatient evaluation may include MRI of the head and neck and work-ups for metabolic, infectious, or inflammatory disorders as warranted.

**CRANIAL NERVE XI (ACCESSORY NERVE) Anatomy**

The accessory nerve provides motor function to the sternocleidomastoid and trapezius muscles.

**Presenting Signs and Symptoms**

Patients with accessory nerve palsies have neck and shoulder weakness on the affected side. Inspection may reveal a “dropped” shoulder—that is, the affected shoulder lying downward and in lateral rotation. Testing of the sternocleidomastoid reveals weakness when turning the head against resistance to the contralateral side. Because of the proximity of cranial nerves IX and X, particular attention should be paid to these nerve functions on examination. The most common causes are postoperative trauma (e.g., from cervical lymph node dissection) and a cerebrovascular accident.

**Treatment**

Treatment and disposition are similar to that for cranial nerves IX and X.
CRANIAL NERVE XII (HYPOGLOSSAL NERVE)

Anatomy
The hypoglossal nerve provides motor function to all the intrinsic tongue muscles and three of the four extrinsic tongue muscles: the genioglossus, styloglossus, and hypoglossus.

Presenting Signs and Symptoms
Patients with hypoglossal nerve palsies usually have unilateral tongue weakness.

Differential Diagnosis
The primary diagnostic consideration is distinguishing an upper from a lower motor neuron lesion. An upper motor neuron lesion causes contralateral tongue deviation and fasciculations, and tongue atrophy is absent. A lower motor neuron lesion causes ipsilateral tongue deviation and fasciculations, and tongue atrophy is present. A 26-year review of 100 cases of hypoglossal nerve palsy revealed that tumors, predominantly malignant ones, produced nearly half of the palsies. Only 15% of patients made a complete or nearly complete recovery.

External lesions that cause compression or stretching of the nerve include internal carotid artery dissection or aneurysm, intracranial tumor, abscess, and other pharyngeal space tumors.

Treatment
Treatment and disposition are similar to that for cranial nerves IX, X, and XI. If there is concern for a cerebrovascular accident or space-occupying lesion, the patient should be admitted for evaluation.

SUGGESTED READINGS


REFERENCES
References can be found on Expert Consult @ www.expertconsult.com.
REFERENCES