Chapter 7

CRANIAL NEUROPATHIES: ELECTRODIAGNOSIS AND MANAGEMENT

MARGARETE DI BENEDETTO, M.D.*

Colonel Medical Corps, US Army (Ret); formerly, Assistant to the Deputy Commanding General, US Army Training Command, Fort Monroe, Virginia; currently, Professor, Physical Medicine and Rehabilitation, University of Virginia, Box 3522 UVA Health Science Center, Charlottesville, VA 22908

INTRODUCTION

Injuries of the cranial nerves (CN) are relatively common, especially secondary to head trauma. Injuries may be direct or indirect, through damage caused by shear, distraction, or compression. Upper cervical lesions can involve the lower CN IX–XII, because the 1st and 2nd cervical vertebrae are in anatomical proximity to the base of the skull and to CN VI, whose intracranial course is precariously long.[1,2] The exact incidence is not known, but numerous reports discuss such injuries in specific patient populations.[3–9]

A large retrospective study by Dobson and associates[10] reviewed head and neck injuries, especially maxillofacial injuries, during 50 military conflicts between 1914 and 1986, and showed an overall mean incidence of 16% of all reported injuries. This exceeds the reported incidence concerning whole body surface area. There was little variation during the period, except for an incidence of 21% in terrorist conflicts.

Blunt trauma to the head mostly affects CN I, VII, and VIII, followed in frequency by injuries to CN III and IV. Injury to CN III is reported especially in pediatric populations.[11,12] Skull crushing injuries have a stretching effect on cranial nerves, with possible avulsion of nuclei from the brainstem and potential disruption of CN II in the optic canal. However, they cause relatively limited brain damage. Penetrating injuries such as gunshot wounds have varying effects, frequently involving CN II or the lower CN X, XI, and XII. CN V is more vulnerable to facial trauma. Orbital or jaw fractures can cause "atypical" (minor or secondary) neuralgia. Atypical neuralgia must be differentiated from "typical" (major or primary) trigeminal neuralgia because of the differing therapeutic approach to each. (See section in this chapter on CN V.)

Surgical trauma of cranial nerves occurs most often during removal of space-occupying lesions at the base of the skull. CN VII and VIII are vulnerable during cerebellopontine angle surgery (acoustic neuroma or meningioma). CN III, IV, V (branches 1 and 2), and VI may suffer injury during exploration of the cavernous sinus. CN III, V, VI, VII, X, XI, and XII are at risk during tumor removal at the petrous ridge. Endartarectomies may be followed by tongue weakness (CN XII) or minor transient facial muscle weakness (CN VII), or both. Vocal cord weakness (CN X) is also possible. Neuromonitoring during surgery employing cranial nerve stimulation and display of evoked potential amplitude changes may lead to decreased morbidity. The most common procedures are facial nerve stimulation, electromyography (EMG) of mimetic muscles, and auditory (brainstem) evoked potentials.[13–16]

If cranial nerves are injured during surgery, reconstruction through end-to-end anastomoses or insertion of nerve grafts may be undertaken. Sekhar and colleagues[17] reported a high success rate for reconstruction of CN III–VI traumatized during surgery.

Nontraumatic lesions can be degenerative, infectious, or vascular (sudden onset), but most frequently they are secondary to tumors (primary or metastatic). Ransom and associates[18] reported a series of patients with prostate cancer and metastases to the base of the skull who presented with symptoms and signs of different cranial nerve lesions.

Prognosis for spontaneous recovery after cranial nerve injury is best for the facial and oculomotor nerves, and poorest for the olfactory, optic, and audiovestibular nerves. Steroid therapy sometimes promotes recovery. Decompression of CN II or VII may result in return of function.[11]

In the comatose patient, early recognition of cranial nerve involvement may be difficult except for CN III, VI, and VII. An incompetent afferent arc of the pupillary reflex indicates a CN II lesion. Due to the effects of cerebral edema or hemorrhagic meningitis, close monitoring for any deterioration in function of CN VI and VII must be maintained, even when initial findings are normal. Transtentorial herniation may cause a delayed CN III lesion, and if this is not recognized early the result can be increased morbidity and mortality. Cranial nerve functions must be reevaluated frequently to ensure an optimal rehabilitation outcome.

The following sections will be concerned with the anatomy and function of the 12 pairs of cranial nerves, and the incidence, diagnosis, prognosis, and therapeutic measures for injuries or diseases affecting them. CN I and II can be regarded as extensions of the brain, and the spinal root of CN XI originating from the cervical spinal cord can be considered a spinal nerve. However, for this discussion these three pairs of nerves will be considered as cranial nerves (Figures 7-1 and 7-2).

Cranial nerves have one or more types of functional components. Special sensory fibers are those

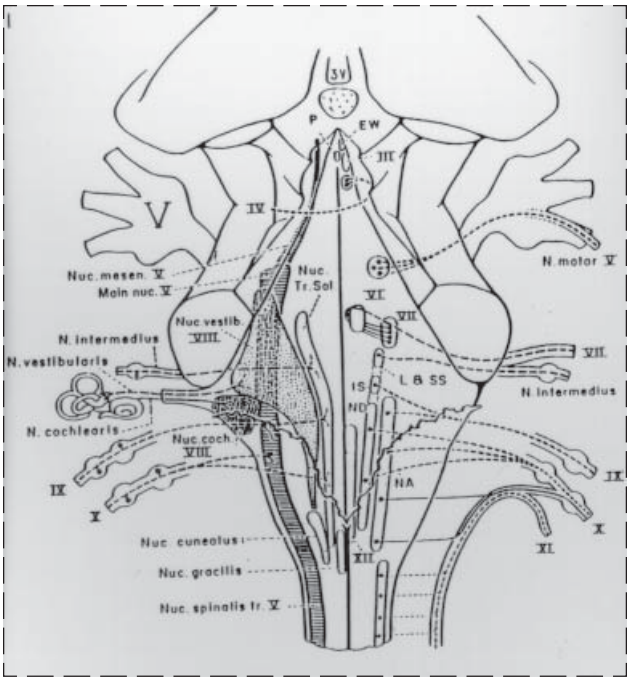

Fig. 7-1. Positions of cranial nerve nuclei III–XII and course of their respective fibers (dorsal view). 3V: Third ventricle; EW: Edinger-Westphal nucleus; P: Perlia's nucleus; Nuc. Tr. Sol.: nucleus tractus solitarius; L and SS: Lacrimal and superior salivatory nuclei; IS: Inferior salivatory nucleus; ND: Dorsal nucleus of vagus nerve; NA: Nucleus ambiguus. Reprinted with permission from Haymaker W, Kuhlenbeck H, Baker AB, Baker LH, eds. Disorders of the brainstem and its cranial nerves. *Clinical Neurology.* Vol 3. Hagerstown, Md: Harper and Row; 1976: 3.

from the special sense organs. Efferent fibers pass from the central nervous system, to skeletal muscle (striated muscle) derived from somites or branchial arches, to visceral (smooth) and cardiac muscle, and to the secretory cells of glands. Afferent fibers transmit sensation from mucous membranes, skin, blood vessels, and internal organs. These cranial nerve functional components are noted below.

1. Special sensory fibers
   - (I, II, VIII): from special sense organs.
2. Efferent fibers
   - Somatic (III, IV, VI, and XII): innervate striated muscle derived from somites;
   - Branchial (V, VII, IX, X, and XI): innervate striated muscle derived from branchial arches;
   - Visceral: parasympathetic preganglionic fibers travel through CN III to smooth muscle within the eye; through CN VII to salivary and lachrymal glands; through CN IX to the parotid gland; and through CN X to the heart, the smooth muscle and glands of the lung and bowel, and to the liver and pancreas.
3. Afferent fibers
   - Visceral: sensation from the heart, lungs, blood vessels, and alimentary tract through CN IX and X; and gustatory fibers through CN VII, IX, and X;
   - Somatic: sensation from skin and mucous membranes of the head through CN V. Some afferent fibers travel with CN VII, IX, and X, but then terminate centrally on the trigeminal nuclei in the brainstem.

Knowing the anatomy and function of each individual cranial nerve is essential for accurate diagnosis and effective treatment of injuries. A detailed discussion follows.

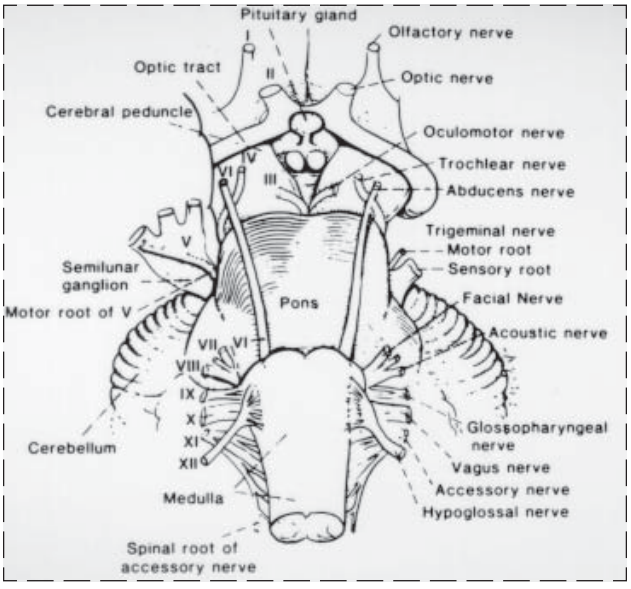

Fig. 7-2. Ventral view of the brain stem with cranial nerves. Reprinted with permission from Brazis P, Masdeu J, Biller JL. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 271.

CRANIAL NERVE I: OLFACTORY

The olfactory nerve is classified as special sensory and its anatomy is depicted in Figures 7-3 and 7-4.

Anatomy and Function

The primary neurons are bipolar sensory nerve cells located in the lateral wall of the nasal cavity and the posterior nasal septum. The dendrites extend to ciliated receptors in the upper part of the nasal mucosa. The unmyelinated axons (central processes) of these bipolar cells are gathered into bundles of approximately 20 filaments each, which then pass through the cribriform plate of the ethmoid bone to the olfactory bulb. There they synapse with secondary neurons and send myelinated processes to form the olfactory tract. At the anterior perforated substance, the tract divides into medial and lateral striae, forming the olfactory trigone. Some striae fibers decussate in the anterior commissure and terminate in the contralateral cerebral hemisphere, so as to provide bilateral cortical representation for smell. Most of the lateral striae fibers pass to the ipsilateral piriform lobe, the primary olfactory cortex (temporal cortex).

Here the secondary neurons synapse again with tertiary neurons that extend to the endorhinal cortex (area 28), the lateral preoptic area, the amygdaloid body, and the hypothalamus. The central connections of the olfactory nerve are complex. Association fibers to the tegmentum and pons pass directly from the anterior perforated substance and indirectly from the hippocampus via the fornix and olfactory projection tracts through the mamillary bodies and anterior nuclei of the thalamus. Close linkage of the prepiriform cortex and amygdala with the thalamus may enable integration of smell with affective behaviors. Certain reflex connections with the nuclei of other cranial and spinal nerves may be functionally significant for swallowing and digestion.

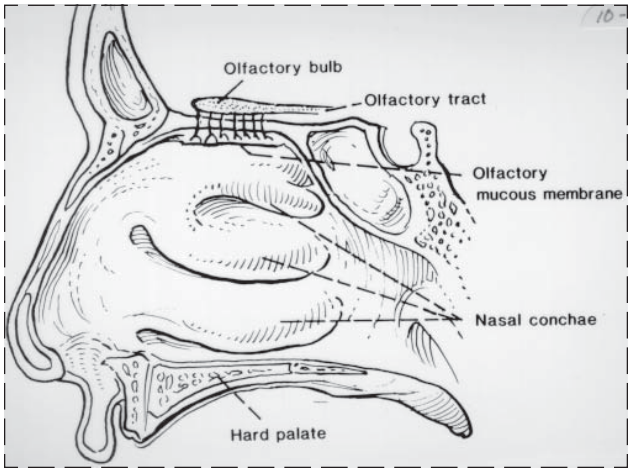

Fig. 7-3. The olfactory nerve (lateral view). Reprinted, with permission from Brazis P,Masdeu J, Biller JL. *Localization in Clinical Neurology.* 2nd ed. Boston, Mass: Little, Brown; 1990: 95.

Injuries and Lesions

The most common cause of injury or lesion is traumatic injury.[19,20] This includes fracture of the cribriform plate, closed head injury without fracture (shearing), or compression of vascular supply by increased intracranial pressure. In some cases a blow to the back of the head (contrecoup coup) may cause injury to CN I.

The incidence of olfactory nerve involvement for all head injuries is approximately 7%, but incidence increases to 20% or 25% following severe head trauma.[6,20] CN I involvement following head trauma is less common in children.[21] Another source of olfactory nerve injury is surgical trauma during procedures in the olfactory region. Nontraumatic lesions include tumors (olfactory groove meningioma may present with anosmia as the sole symptom in the beginning),[22] chronic basilar meningitis,[23] Korsakoff's syndrome,[24] Huntington's chorea,[25] Alzheimer's disease,[25,26] Parkinson's disease,[27] or Foster Kennedy syndrome[28] (ipsilateral anosmia, ipsilateral optic atrophy, and contralateral papilledema due to increasing intracranial pressure). With increasing age, smells are less intense and more difficult to identify and discriminate; the cause of this may be receptor site pathology, or neuronal, hormonal, or neurotransmitter abnormalities.[29]

The causes of loss of smell were studied by Deems and associates[30] in a comprehensive study of 750 consecutively evaluated patients at the University of Pennsylvania Smell and Taste Center. Inflammatory nasal disorders were reported to be responsible for 26%; 15% were due to nasal or paranasal sinusitis; 22% were idiopathic; 18% were due to head trauma; 4% were congenital; 2% were due to toxins; and several other causes each accounted for less than 2%. Intranasal cocaine free-

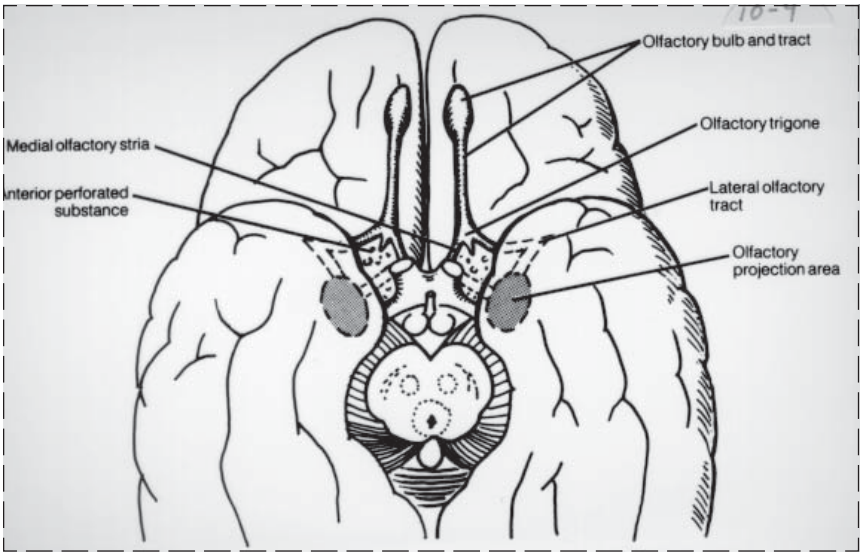

Fig. 7-4. The olfactory nerve (inferior view). Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy*. 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 296.

basing (smoking the extracted and volatile form of the street drug) seems to have produced an increased frequency of bony septal and cartilaginous necrosis with osteolytic sinusitis as a cause for loss of smell in patients.[31]

Symptoms and Signs

Patients present with an altered sense of smell, which may be diminished detection and recognition, or heightened awareness.

1. Dysosmia
   - Anosmia: absence of smell. This is most significant when unilateral.
   - Hyposmia: decreased sense of smell.
   - Parosmia: perversion of smell. In addition to head injury, this is found with schizophrenia, uncinate gyrus lesions, hysteria, and other temporal lobe lesions. During recovery from injury, parosmia is often transitory, although it may become permanent.
   - Cacosmia: unpleasant odors, usually due to decomposition of tissues and noticed by the patient while breathing out. Cacosmia can also occur secondary to head injury.
   - Hyperosmia: increased sensitivity to odorants. This may be present in hysteria, cocaine addiction, or pregnancy.
2. Diminished olfactory recognition
   - This may be present despite a relatively preserved olfactory detection. It may occur with blunt trauma affecting the orbitofrontal and temporal lobes.[32]
3. Olfactory hallucination
   - This may be present in psychoses but also may result from structural nerve damage.

Evaluation

Every patient with mild head injury due to an occipital blow or frontal vault fracture should be evaluated carefully for olfactory dysfunction.[33] Peripheral causes of dysosmia such as fractures, nasal mucosal swelling, and other soft tissue nasal obstruction must be ruled out.

The psychophysical examination is a qualitative and quantitative testing of the ability to perceive different fragrances. There are several approaches of increasing sophistication. Tests 1 and 2 below are mainly qualitative in nature.

1. Presentation of nonirritating odorants (chocolate, vanilla, coffee, and the like) to one nostril at a time, while occluding the other nostril.
2. Odor identification test with forced choice format. The patient is offered a choice of four possible responses for each of 40 odorants located on microencapsulated strips.
3. Threshold tests add a quantitative element to the evaluation by offering known concentrations of odorant.
   The Connecticut Chemosensory Clinical Research Center[34] has developed a thresh-

old test that uses eight plastic squeeze bottles containing graded concentrations of butanol and distilled water.

Threshold tests are further refined in the Medical College of Virginia Olfactory Screening Test (MCVOST).[34] In this test, a stimulus such as chocolate, vanilla, or coffee is presented to each nasal cavity using a 20-mL plastic squeeze bottle. Scoring includes stimulus detection (1 point) and identification (1 point) for each stimulus. The maximum score is 6 points for each side or 12 points in all. Lateralization of olfactory function is considered when there is a difference in left and right scores. In one study[35] of 51 consecutive rehabilitation admissions, 60% of mild head injury patients were found to have normal olfactory function; 20% had impaired function; and 13% were anosmic. Patients with severe head injury showed only 8% with normal olfactory function; 67% were impaired; and 25% were anosmic. Care must be taken not to present strong aromatic odorants like ammonia because they will stimulate trigeminal nerve terminals.

Electrodiagnosis

Olfactory evoked potentials are a study of the late near-field event-related potentials during multichannel electroencephalogram (EEG) recordings.[36,37] The test is performed using an olfactometer, which delivers stimuli having the shape of a square wave (steep onset and decline). The stimulants are presented in a constantly flowing air stream of controlled temperature and humidity. Rise time of the stimulus must not be more than 20 millisecond. Cues from tactile, thermal, or acoustic sensation must be prevented. Total flow rate can be varied within a wide range but is generally about 140 mL/s. Delivery of the stimulus should be independent of breathing. This is done by velopharyngeal closure and avoiding the flow of respiratory air in the nose during stimulation. The stimulus interval should be approximately 6 to 8 millisecond and care should be taken to avoid habituation. The odorant presented is usually vanillin. Evoked responses are recorded from scalp electrodes placed on C_z referenced to A_1, according to *The International Ten-Twenty System of Electrode Placement* (International 10-20), as shown in Figures 7-5 through 7-7.)Normal values are a response latency of 300 to 400 millisecond and a response amplitude of 10 to 20 μV.

With stimulation of the left nostril, the more pleasant an odor is perceived to be, the longer the latencies and the larger the amplitudes observed.

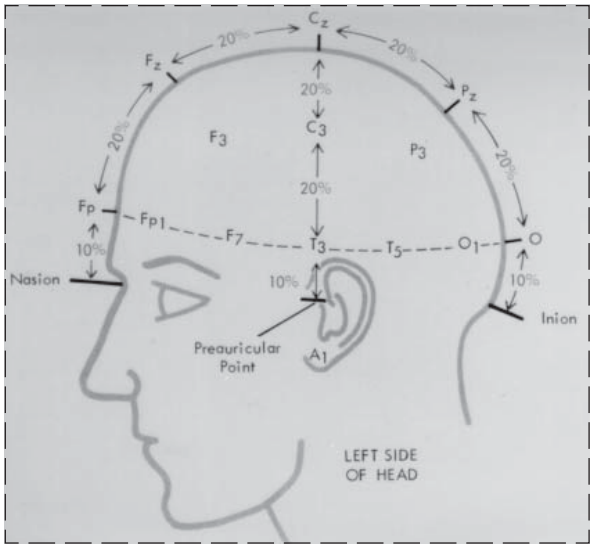

Fig. 7-5. Standard international 10-20 EEG electrode placement system (electrodes are placed either 10% or 20% of the total distance between skull landmarks). Lateral view. Adapted with permission from Grass Medical Instruments. *A Review of The International Ten–Twenty System of Eelectrode Placement*. Quincy, Mass: Grass Instrument Co; 1974: 1.

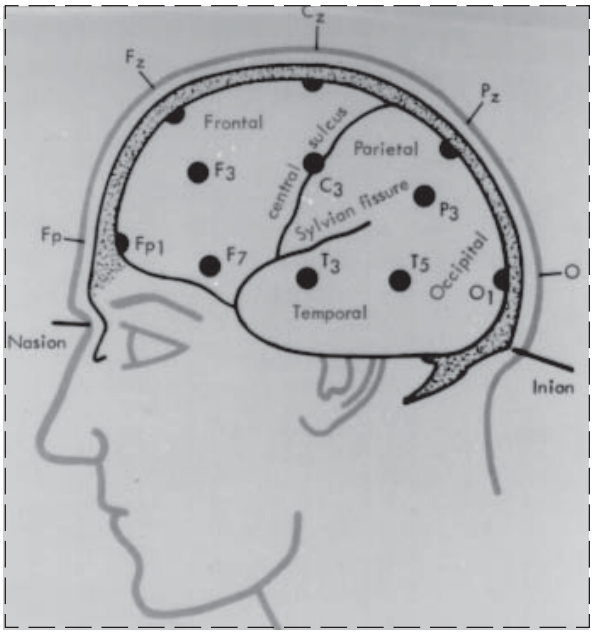

Fig. 7-6. Lateral skull showing location of electrode placement points in relationship to brain. Adapted with permission from Grass Medical Instruments. *A Review of The International Ten–Twenty System of Electrode Placement.* Quincy, Mass: Grass Instrument Co; 1974: 2.

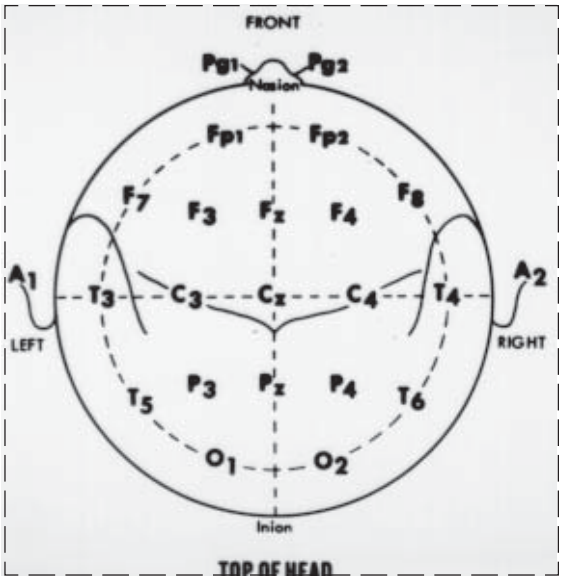

Fig. 7-7. Standard International 10-20 EEG electrode placement system, view from above. Adapted with permission from Grass Medical Instruments. *A Review of The International Ten–Twenty System of Electrode Placement*. Quincy, Mass: Grass Instrument Co; 1974: 19.

This variation is believed to be due to emotional responses to the odorant. Responses to carbon dioxide, menthol, and hydrogen sulfide (trigeminal stimulation) show significantly shorter latencies and smaller amplitudes after stimulation of the left side. Vanillin shows shorter latencies and smaller amplitudes after stimulation of the right side.

Imaging

Lesions identified by computed tomography (CT) and magnetic resonance imaging (MRI) correlate highly with olfactory impairments, especially when primary and secondary olfactory cortical centers are involved.[38]

Prognosis

In patients with CN I involvement secondary to head injury, Costanzo[6] reported a 33% recovery rate, 27% further deterioration, and 40% no change. Improvement may occur as late as 5 years after injury but is unlikely after 1 year.[4] Recent animal studies[39] have shown regeneration and functional reconnection of olfactory nerves, suggesting a similar possibility in humans.

Management

Contrary to common clinical opinion, dysosmias can have a significant impact on function.[6] Inability to smell the "warning signals" of fire, gas, or other dangerous substances may interfere with safety and present problems in some vocations. The American Medical Association[40] impairment rating scale allows a 3% disability for bilateral anosmia.

Rehabilitation strategies to normalize function must take account of the impact of dysosmia on emotions. The emotional component of smell appears to be confirmed by the variability of evoked olfactory responses, and the bilateral differences of latencies and amplitudes. This evidence also tends to support the concept of Dimond and colleagues [41] that pleasant emotions are predominantly processed by the left hemisphere and unpleasant feelings by the right. Many association fibers from the olfactory nucleus course through the hypothalamus to the limbic forebrain, influencing both sexual and nonsexual behaviors.

Therapeutic interventions consist mainly of counseling. They include the following:

- Olfactory evaluation: in the United States this is available in national centers providing chemosensory testing (chemosensory clinics).
- Teaching of hygiene routines.
- Nutritional counseling to avoid medically contraindicated excessive use of spices. Instead, increased attention to the texture, temperature, and visual appeal of foods is emphasized.
- Attention to appropriate food storage when there is a danger that spoilage will not be detected by odor.
- Concentration on visual and auditory cues while cooking.
- Observing fire safety measures, including extra smoke detectors.

CRANIAL NERVE II: OPTIC

Anatomy and Function

The optic nerve is classified as special sensory and its anatomy is shown in Figures 7-8 and 7-9.

The rods and cones are photoreceptors in the retina. They form the deepest layer of the retina and are oriented toward the pupillary opening. The pigment of the rods is rhodopsin, a glycoprotein that

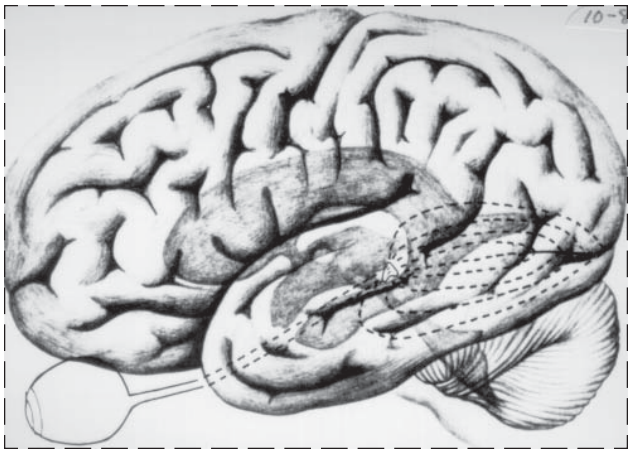

Fig. 7-8. Lateral view of the brain showing optic radiation in the parietal and temporal lobes, lateral to the ventricular system. Reprinted with permission from Brazis P, Masdeu J, Biller J. *Localization in Clinical Neurology*. Boston, Mass: Little, Brown; 1990: 109.

reacts to visible light. The receptors convert light energy to electrical signals. The retina contains approximately 100 million rods and 7 million cones. The cones are of three types, reacting maximally to red, green, or blue light. They are concentrated in the macula region and are especially dense in the fovea centralis. The fovea is the center of the macula and is tightly packed with cones to provide the most acute visual discrimination. The rods and cones connect with bipolar cells, the primary neurons. The primary neurons synapse with the secondary neurons, which are ganglion cells near the surface of the retina. The myelinated axons of ganglion cells form the optic nerve. Postchiasmatic fibers form the optic tract, consisting of ipsilateral temporal fibers of the retina and contralateral nasal fibers. Optic tract fibers pass to the tertiary neurons located in the lateral geniculate bodies of the thalamus. A small number of fibers ascend to terminate in the pretectal region as part of the pupillary light reflex pathway. Axons of the remaining neurons are contained in the geniculocalcarine tract (optic radiations), as seen in Figure 7-8. This tract goes from the lateral geniculate body through the internal capsule to the occipital (calcarine) primary visual cortex. Images from the upper visual field project to the lower area of the calcarine fissure and images from the lower visual field project to the upper area of the calcarine fissure. This compensates for the inverted and reversed image projected onto the retina by light rays converging and passing through the pupil. The central connections are from the pretectal region to the Edinger-Westphal nucleus via the posterior commissure, and from the superior colliculi via tectobulbar and tectospinal tracts to other cranial and spinal nuclei (for visual and body reflexes, such as turning the head in response to light).

Injuries and Lesions

The most common cause of injury or lesion is traumatic injury to the head. These injuries frequently cause basilar skull fractures, frontal lobe lesions, or increased intracranial pressure. Also, temporal bone fractures and blunt trauma to the outer orbital ridge may cause blindness in one eye. Most lesions are in the anterior visual pathways. Injuries can be intraocular, intraorbital, intracanicular, or intracranial. The incidence of CN II injuries in head trauma is reported to be 0.5% to 5.2%, and in children up to 6.0%.[42]

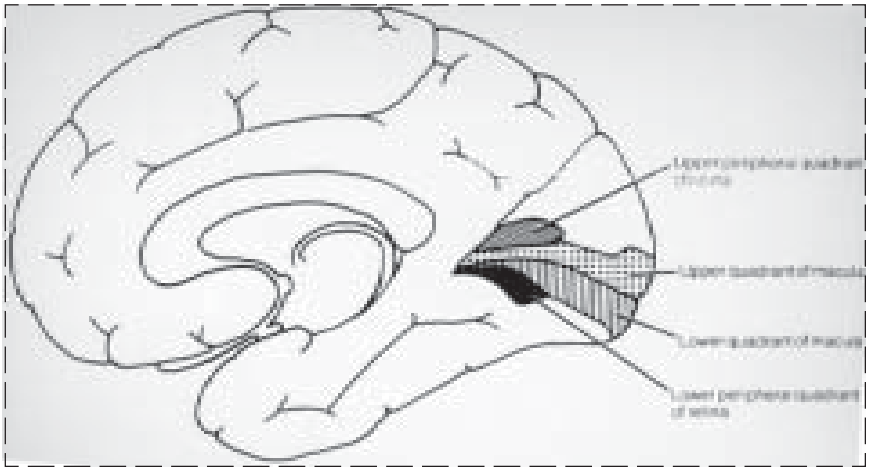

Fig. 7-9. Medial view of the right cerebral hemisphere, showing projection of the retina on the calcarine cortex. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 286.

Penetrating gunshot wounds with entry in front of the ear frequently cause blindness despite often minimal brain damage. LaGrange[43] reported on war injuries with specific attention to the projectile entry site and the resulting orbital trauma. The report noted that entry above the orbit typically caused fractures of the orbital vault, affecting sensory, motor, and optic nerves near the optic foramen. Entry below the eyeball may fail to fracture the orbit at any point, but may still produce concussion of the eye in the macula region. Reduction of central acuity will then follow. If there is fracture of the orbit, findings are significantly more severe. Another observation was that a projectile may traverse the orbit without touching the eyeball, but still divide the optic nerve and avulse the papilla (optic disk).

Lesions of the optic nerve need not necessarily be severe. In a study by Wilbrand and Saenger (cited by LaGrange[44]), 50 out of 100 cases of unilateral optic nerve lesions following cranial nerve trauma had complete and permanent blindness, 4 had total blindness at onset but recovered fully, 17 recovered partially, and 34 had only partial involvement from the start. The incidence of optic nerve trauma through gunshot wounds is 25%. Keane and Baloh[11] report 231 cases in 21 years.

Nontraumatic lesions include tumors of the orbit, disease of the cavernous sinus, and cocaine abuse.[45] Consequences vary according to the location of the lesions, and several syndromes characterized by specific deficits must be recognized. Syndromes involving the optic nerve are as follows:

- Foster Kennedy syndrome: characterized by ipsilateral blindness and anosmia due to atrophy of optic and olfactory nerves and contralateral papilledema, caused variously by tumors at the base of the frontal lobe, arachnoiditis, syphilis, and occult trauma.
- Amaurotic familial idiocy (Tay-Sachs disease): caused by cerebromacular degeneration.
- Holmes-Adie syndrome: presents with tonic pupillary reaction and absence of one or more tendon reflexes. There is abnormal sensitivity to weak solutions of mecholyl (2.5%) instilled into the conjunctival sac. Normal eyes are not affected, but the response of tonic pupils is constriction (Adler-Scheie test).[46]

Symptoms and Signs

Optic nerve lesions are manifested by defects in visual field or acuity, or by objective alterations observed in an ophthalmoscopic examination. Relevant symptoms and signs are as follows:

1. Alteration of visual acuity
   - Amblyopia: markedly reduced vision
   - Amaurosis: complete blindness
   - Hemeralopia: day blindness
   - Fatigue syndrome: vision that is best in dim light
   - Nyctalopia: night blindness, sometimes associated with vitamin A deficiency
2. Visual field defects
3. Papilledema
4. Optic atrophy
5. Cortical blindness due to bilateral occipital infarction, caused by bilateral posterior cerebral artery involvement. The pupillary light reflex remains intact and total blindness is unusual.[47]

Visual field defects are illustrated in Figure 7-10. Signs and symptoms relative to the specific anatomic location of injury or lesion are presented in Table 7-1.

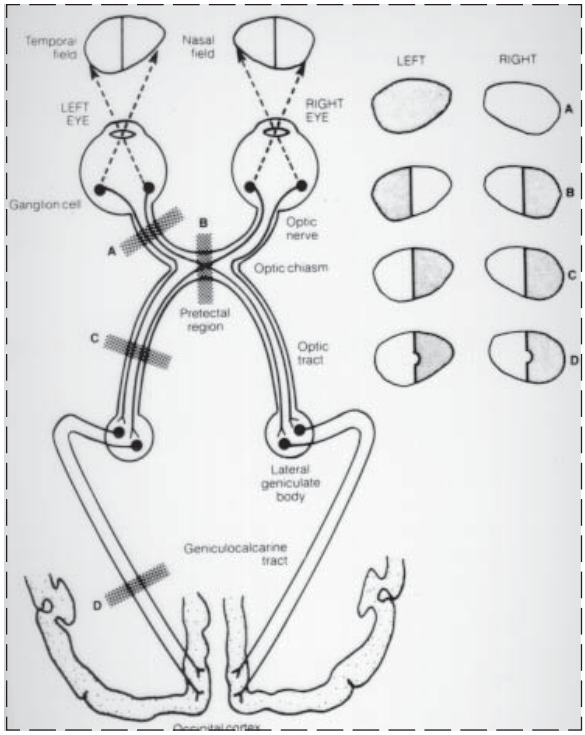

Fig. 7-10. Visual pathways and locations (indicated by letters) causing field defects shown in the diagrams on the right. Reprinted with permission from Ganong WF. *Review of Medical Physiology*. 16th ed. Norwalk, Conn: Appleton & Lange; 1993: 287.

TABLE 7-1

SIGNS AND SYMPTOMS: VISUAL PATHWAYS

	Retina	Optic Nerve	Optic Chiasm
Visual Acuity	Normal, if macula spared	Decreased	Medial chiasm: decreased vision bilateral Lateral chiasm: decreased vision ipsilateral
Visual Field Deficit	Corectopia central, arcuate or sectorial ring	Monocular in unilateral lesion shape as retinal lesions	Anterior angle ipsilateral (temporal or paracentral; contralateral) upper temporal
Pupillary Light Reflex	Unimpaired unless large lesion	Marcus-Gunn-pupil-lesion asymmetric afferent arc deficit	Afferent arc deficit. Ipsilateral impairment in lateral chiasmatic lesions

Adapted with permission from Masdeu GE. The localization of lesions affecting the visual pathways. In: Brazis PW, Masdeu JC, Biller J, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 120–122.

Evaluation

Clinical evaluation consists mainly of visual acuity testing, which can be performed by presenting Snellen charts for distant vision and Jaeger cards for near vision. The patient is tested for ability to recognize the printed material at a specific distance, light, and contrast. If the patient is functioning at an extremely low level, saccadic responses (short, rapid movements of the eye made in order to scan the environment) may be obtained with an optokinetic stimulus. If a large stimulus is successfully followed, the size of the stimulus can be serially decreased, eventually reaching small letters. Convergence and accommodation can also be tested in this manner.

Visual field testing by confrontation can be used as a screening tool. If a defect is apparent, perimetry should be performed, preferably computer automated perimetry. When a cortical lesion is suspected, tests for visual inattention, neglect, and extinction are necessary. A lesion within a visual pathway can be localized by observing the different types of field defects. Monocular lesions are usually evidence of injury to the retina or the optic nerve. Binocular lesions can be localized at or beyond the optic chiasm. The shapes of different visual field defects and localization of the specific lesion are shown in Figure 7-10.

Trauma or infarction in bilateral occipital lesions is commonly responsible for bilateral altitudinal defects, in which the lower half of the visual field is affected and the macula is mostly spared. Such defects may also be caused by bilateral ischemic disease of the retinae or the optic nerves.[48,49] Color vision, often affected in patients with retrobulbar neuritis, is evaluated by testing ability to recognize figures hidden in a pattern of specially colored dots (Ishihara's chart).

Objective signs are observed during fundoscopy. Assessment for papilledema or optic atrophy is most important. Disk pallor will not be present before at least one month following injury. Vessels should be observed for size, regularity, and tortuosity. The examiner should look for hemorrhages and exudates and study the maculae carefully (patients should look at the examiner's light).

Another objective test for evaluating the visual pathways is the pupillary reflex (Figure 7-11; and see the discussion of CN III). The fibers that make up the afferent arc of the pupillary reflex exit the visual pathway before the lateral geniculate body and travel to the dorsal midbrain. Only large retinal lesions impair the light reflex. Asymmetric optic

Optic Tract	Lateral Geniculate	Optic Radiations	Calcarine Cortex
Normal in unilateral lesion	Normal in unilateral lesion	Normal in unilateral lesion	Normal in unilateral lesion
Contralateral homonymous hemianopia; incongruous	Contralateral homonymous may be incongruous; quadruple sectoranopia	Contralateral homonymous hemianopia (total lesion) or quadrantanopia (inferior with parietal lesion; superior with temporal lesion). Macular sparing with purely quadrantic defects	Contralateral homonymous hemianopia, congruous. Macular sparing. Involvement or sparing of contralateral unpaired temporal crescent. Ring shape or altitudinal with "vertical step" in bilateral lesions
Afferent deficit in contralateral eye	Normal	Normal	Normal

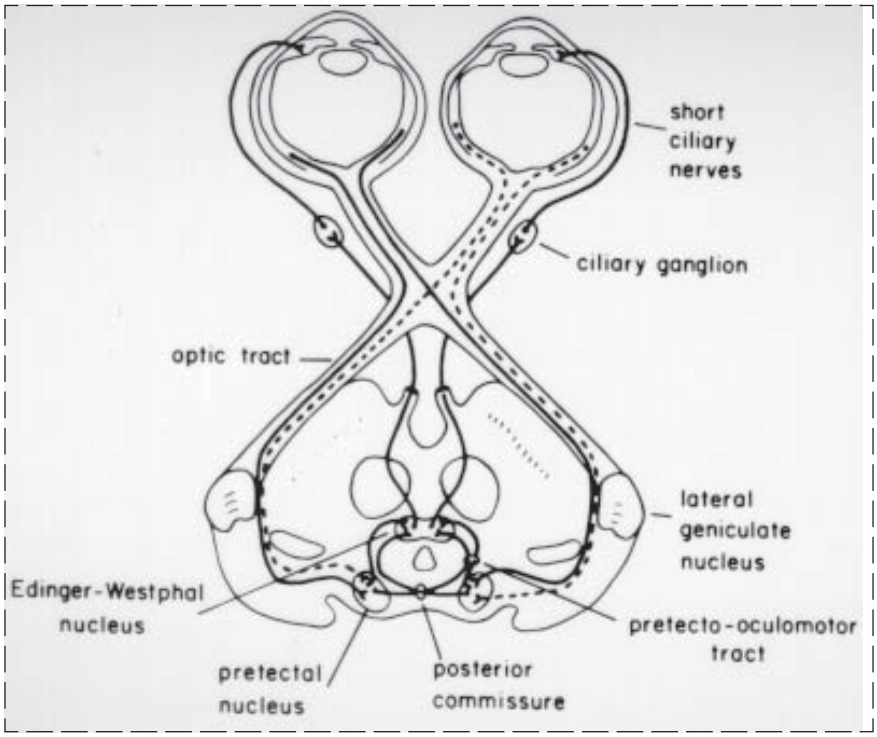

Fig. 7-11. Diagram of the path of the pupillary light reflex. Reprinted with permission from Brazis P, Masdeu J,Biller JL, eds. *Localization in Clinical Neurology.* Boston, Mass: Little, Brown; 1990: 145.

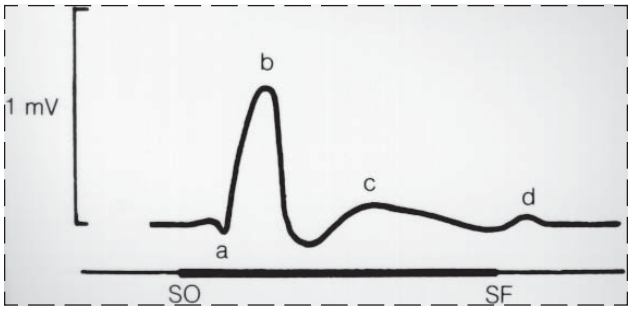

Fig. 7-12. Human electroretinogram (ERG). SO: Light stimulus on; SF: Light stimulus off. Adapted with permission from Ziv B. Electroretinography. *N Engl J Med*. 1961;264(11):546.

nerve or chiasmatic lesions can be detected by the Marcus-Gunn pupil. This is a relative afferent pupillary defect in which the abnormal pupil first dilates paradoxically as light is quickly moved from the normal to the abnormal eye. If one optic nerve is damaged, for example, by a temporal bone fracture, early diagnosis of blindness in that eye can be assessed by the pupillary light response. The pupil will be large and nonreactive to direct light but will respond to a light in the other eye (consensual reflex).

Electrodiagnosis

Two electrodiagnostic tests are available for objective assessment of the visual pathways. Electroretinography tests the function of the retina.[50] In this test, a light stimulus (stroboscope) or pattern reversal (reversing checkerboard) is presented to the eye. For recording, an electrode is placed in a contact lens over the cornea and an indifferent electrode is placed over the scalp. A 6 mV potential difference is observed between the two electrodes at rest. After onset of a light stimulus, a wave form (a, b, c) can be demonstrated, and another wave (d) follows when the light stimulus is turned off (Figure 7-12). Visually evoked potentials evaluate the complete visual pathway, including the retina. The parameters of the test are presented in the following list. Electrode placements are shown in Figures 7-5, 7-7, and 7-13.[51]

Visually Evoked Potentials

Stimulation
- Pattern reversal (reversing checkerboard)
- Size of squares, 30 to 50 minutes of arc
- Rate of change, 1 to 2 reversals per second
- Contrast and brightness held constant

Recording
- Active electrode placed on O_Z, reference on F_Z, ground electrode on F_{PZ} (International 10–20)
- Sensitivity: 10 μV per division
- Filters: low frequency 1 to 2 Hz; high frequency 100 to 200 Hz
- Averaging: 64 to 256 responses
- Analysis time: 300 to 500 millisecond

Measurements
- Absolute peak latencies
- Peak to peak amplitudes
- Duration of P100 (positive) (ie, N75 [nega-

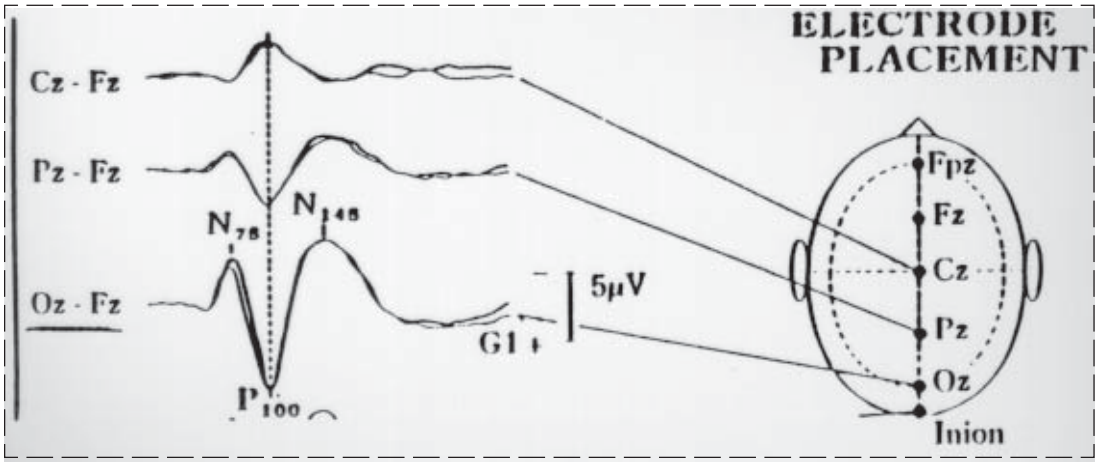

Fig. 7-13. Visual evoked potential electrode placement and evoked response. Reprinted with permission from TECA Corp. *Pattern reversal visual evoked potential VEPs*. In: TECA Applications Bulletin No. 1002. Pleasantville, NY: TECA Corp; 1981.

tive] to N145)
- Interwave latencies
- Interocular variation

Normal values
- Latency of P100: 102.3 ± 5.1 millisecond (range 89–114 ms)
- Mean ± 3 SD upper border of normal 117.6
- Latency difference between two sides: 2.3 ± 2.0 millisecond (range 0–6 ms)
- Amplitude of P100: 10.1 ± 4.2 μV (range 3.21 μV amplitude difference between two sides: 1.6 ± 1.6 μV (range 0–5.5 μV)
- Duration of P100: 63 ± 8.7 millisecond (range 47–86 ms)
- Duration difference between two sides: 2.8 ± 2.9 millisecond

Prognosis and Management

The prognosis for recovery after optic nerve injury is poor. Only one third of patients show significant improvement.[52]

Only in delayed visual loss are possible decompressive procedures indicated. There is hope that high dose steroid therapy may help counteract swelling and compression of the optic nerve in the optic canal. New drugs such as the GM (Monosialotetrahexosyl Ganglioside) gangliosides offer some hope for the future.[53] In case of cortical blindness with some sparing, training (utilizing low vision and other cues) seems to have promise.[47] Strong intensity stimulation of a defective visual field may help to improve function.

CRANIAL NERVES III, IV, AND VI: OCULOMOTOR, TROCHLEAR, AND ABDUCENS

Cranial Nerves III, IV, and VI are all motor cranial nerves. The anatomy is illustrated in Figure 7-14.

Anatomy and Function

Cranial Nerve III. The oculomotor nerve has its nuclei located in the central gray matter at the level of the superior colliculus (mesencephalon). These nuclei are arranged in one unpaired and four paired rostrocaudal columns. The unpaired rostrocaudal column shares bilateral nuclei, rostrally the Edinger-Westphal nuclei, and caudally the subnuclei for the levator palpebrae superioris. The most medially located nuclei of the paired columns are the subnuclei for the superior rectus muscles.

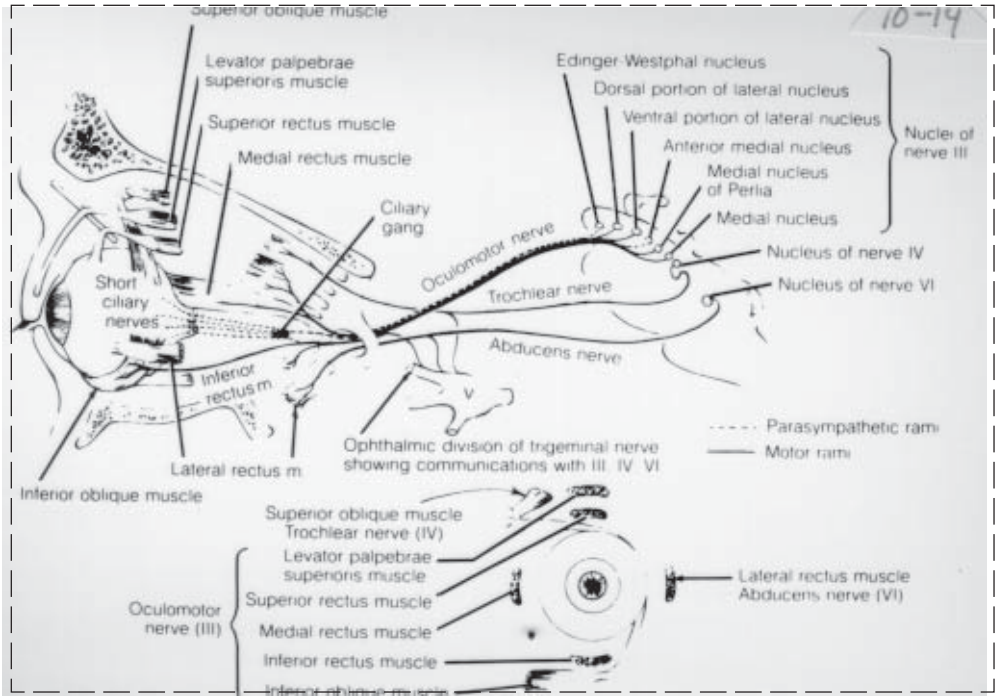

Fig. 7-14. The oculomotor, trochlear, and abducens nerves. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 147.

The motor fibers (somatic efferent) from the superior rectus subnuclei immediately cross, actually through the opposite subnucleus. In the fascicular portion, oculomotor fibers diverge, and those to the levator palpebrae are located laterally. The oculomotor fibers travel through the red nucleus and the inner side of the substantia nigra and emerge on the sella turcica in the outer wall of the cavernous sinus. The fiber tracts then leave the cranium through the orbital fissure, where they separate into superior and inferior divisions. The superior division supplies the levator palpebrae and superior rectus muscles. The inferior division supplies the medial and inferior rectus and the inferior oblique muscles.

There is ipsilateral innervation of the medial rectus, inferior rectus, and inferior oblique muscles (CN III), and of the lateral rectus muscle (CN VI). Contralateral innervation supplies the superior rectus (CN III) and the superior oblique (CN IV) muscles.

The oculomotor nerve also carries parasympathetic fibers (visceral efferent), which arise from the Edinger-Westphal nucleus (preganglionic fibers) as part of the craniosacral division of the autonomic nervous system. These fibers end in the ciliary ganglion, from which postganglionic fibers emerge as short ciliary nerves to supply the ciliary muscle and the sphincter pupillae.

Cranial Nerve IV. The trochlear nerve nucleus is located just caudal to CN III at the level of the inferior colliculus (mesencephalon). The motor fibers (somatic efferent) in their entirety decussate in the anterior medullary velum and wind around the cerebral peduncles. The nerve then follows CN III and is situated in the lateral wall of the cavernous sinus. It enters the orbit through the superior orbital fissure and innervates the superior oblique muscle.

Cranial Nerve VI. The abducens nerve nucleus is located in the floor of the 4th ventricle in the lower portion of the pons near the internal genu of the facial nerve. Its special significance is its internuclear neurons, which send axons across the midline via the medial longitudinal fasciculus to the CN III medial rectus subnuclei. These fibers coordinate horizontal gaze. The abducens nucleus also relays impulses from the contralateral vestibular nucleus, which sends simultaneous impulses to the ipsilateral medial rectus subnucleus of the oculomotor nerve (vestibuloocular reflex).

The motor fibers (somatic efferent) to the lateral rectus muscle emerge anteriorly from the pontomedullary fissure and pass through the cavernous sinus in close proximity to the internal carotid artery. Abducens nerve fibers exit the cranium via the superior orbital fissure and supply the lateral rectus muscle. A few sensory (proprioceptive) fibers from the eye muscles are present in nerves III, IV, and VI. These sensory fibers terminate in the mesencephalic nucleus of CN V.

Central connections of the ocular motor nerve fibers are connected from the pretectal region near the posterior commissure to the Edinger-Westphal nucleus, in order to mediate the ipsilateral and consensual light reflexes. If this pathway is interrupted, an Argyll Robertson pupil occurs. From the supe-

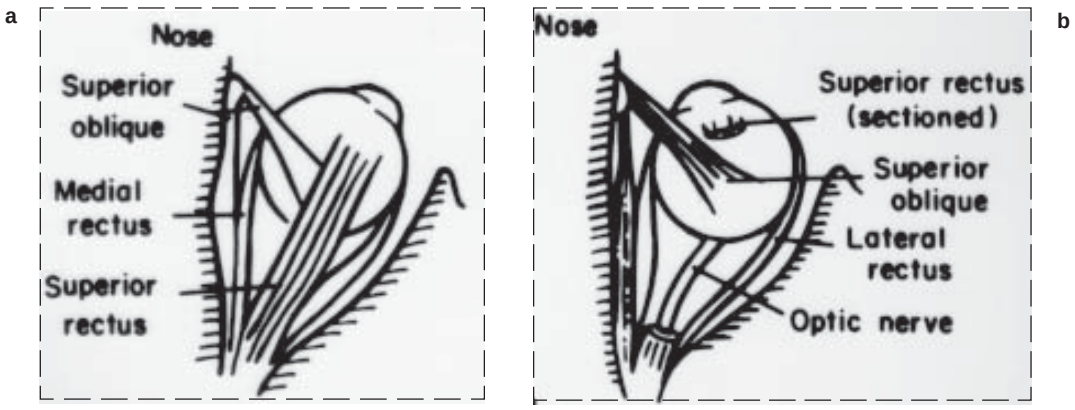

Fig. 7-15. Superior view of the right orbit. In the abduction (**a**), the superior rectus acts as an elevator and the superior oblique intorts the eye (brings the upper pole toward the nose). In adduction (**b**), the superior oblique acts as a depressor and the superior rectus intorts the eye. In (**b**), the superior rectus has been removed to show the position of the superior oblique. Reprinted with permission from Brazis, P, Masdeu, J. and Biller, JL. *Localization in Clinical Neurology.* Boston, Mass: Little, Brown; 1990: 129.

rior colliculi, fibers connect via the tectobulbar tract to nuclei III, IV, and VI, for mediation of accommodation and other reflexes. From the inferior colliculi, fibers connect via the tectobulbar tract to eye muscle nuclei, for reflexes correlated with hearing. From the vestibular nuclei, fibers connect via the medial longitudinal fasciculus, for reflex correlation with balance. From the cortex, fibers connect through the corticobulbar tract, for mediation of voluntary and conditioned movements of the eyes.

It is *imperative to understand fully* the anatomy and function of the ocular motor nerves and the innervated muscles associated with them (Figure 7-15). The recti muscles originate from the ligament of Zinn, which surrounds the circumference of the optic foramen except at the upper and outer part. All muscles attach with tendinous expansions to the sclera of the globe, at points above, below, medial, and lateral, as required.

The superior oblique arises above the inner margin of the optic foramen and in tendinous form passes through a pulley, a cartilaginous loop at the internal angular process of the frontal bone. It then reflects posterior, lateral, and inferior, passing beneath the superior rectus muscle to the lateral aspect of the globe. It inserts between the superior and lateral rectus muscles into the sclera, behind the equator.

The inferior oblique originates from the orbital plate of the maxilla. It passes lateral, posterior, and superior beneath the inferior rectus, and then inserts into the sclera behind the insertion of the superior oblique muscle.

The functions of the oculomotor nerve innervated muscles are illustrated in Figure 7-16 and listed here.

1. The medial and lateral rectus muscles move the globe horizontally.
2. The superior rectus and inferior oblique, as well as the inferior rectus and superior oblique, have complementary actions.
   With the globe in abduction:
   - the superior rectus elevates the globe,
   - the inferior rectus depresses the globe,
   - the inferior oblique extorts the eye (moves the left eye clockwise), and
   - the superior oblique intorts the eye (turns the left eye counterclockwise).

   If the globe is in adduction:
   - the inferior oblique elevates the globe,
   - the superior oblique depresses the globe,
   - the superior rectus intorts the eye (moves the left eye counterclockwise), and
   - the inferior rectus extorts the eye (moves the left eye clockwise).
3. The levator palpebrae elevates the lid. Fibers are intermingled with Müller's muscle (sympathetic innervation) and with the fibers of the orbicularis oculi (CN VII, which closes the lid).
4. The ciliary muscle decreases the tension of the lens capsule, increasing the convexity of the lens and adjusting the eye for near vision.

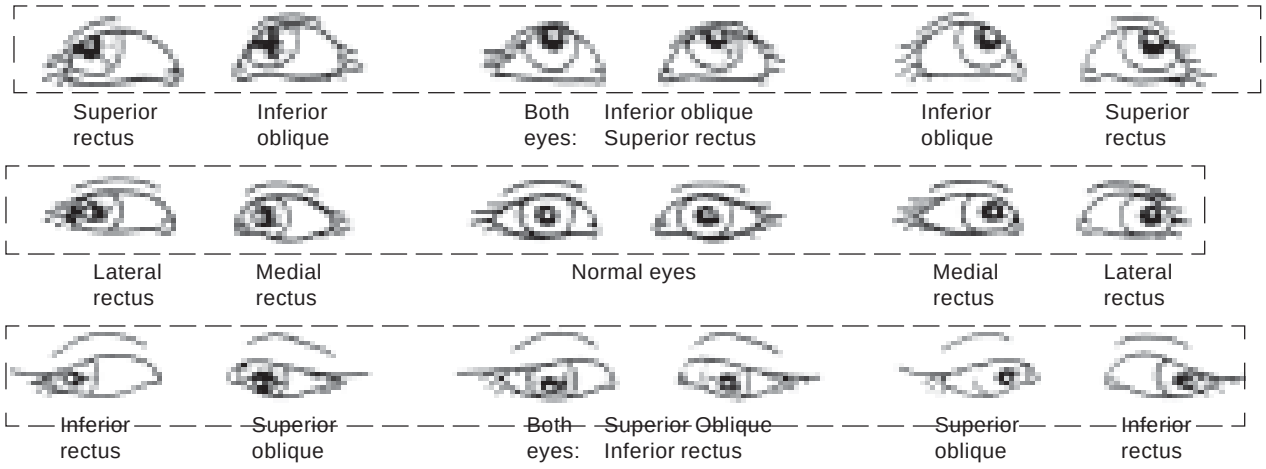

Fig. 7-16. Ocular muscles responsible for eye movement in different gaze positions. Drawing: Courtesy of artist: Dr. Anna Bettendorf, University of Virginia.

5. The sphincter pupillae constricts the pupil in response to a light stimulus and for accommodation.

The supranuclear control system of eye movement is as follows:

1. Vestibular system: The vestibuloocular reflex maintains focus on an object when the head moves; the eye moves within 10 millisecond in the opposite direction from head movement. Fibers from the vestibular nucleus travel to the contralateral abducens nucleus, and to the ipsilateral medial rectus subnucleus (oculomotor).
2. Optokinetic reflex: The optokinetic reflex maintains eyes on target during prolonged head movement in the same direction, after the vestibuloocular reflex has fatigued (ie, after approximately 30 s).
3. Smooth pursuit system: This system keeps a particular image in the fovea involuntarily. Images moving away from the fovea represent strong stimuli for smooth pursuit. The system can follow objects as rapidly as 30° to 40° per second. Ability decreases as a person grows older.[54] As the eyes and head move to follow an object, the vestibuloocular reflex is inhibited.
4. Saccadic system: Fast moving objects require rapid eye movements. These movements are called saccades and are under voluntary control. Objects of interest are registered by the peripheral vision and then focused in the fovea. Alertness is necessary to produce saccades and they are crucial for reading. Abnormal or inaccurate saccades, such as hypermetric (too fast), or hypometric (too slow), or purely initiated (unintentional) saccades, may occur with lesions in the structures that mediate the production of saccades. Elderly subjects and inattentive or medicated subjects have slower saccadic eye movements.[54]
5. Convergence system: Convergence and divergence work in tandem to permit binocular vision. Active eye muscle contraction is required to bring a single point within the visual field into homologous sites of both maculae. If there is a disparity of image registration between the retinas, diplopia will occur. During convergence the pupillary sphincter constricts. In a CN III lesion this accommodation reflex may be affected later than the light reflex because the pupillomotor fibers mediating convergence outnumber the fibers mediating the light reflex.[55]
6. Visual perception: Visual perception involves the interaction and integration of lower level abilities such as oculomotor control, visual field and acuity (the basis for visual attention), scanning, pattern recognition, and memory. Disruption of any of these skills, as is frequently observed in patients with head injury or stroke, causes significant deficits in daily activities. Severe visual scanning difficulties may contribute to language deficits in some aphasics because of the decreased information gathered.[56]

The anatomy and functions of the oculomotor nerves are as follows:

1. Innervation of muscles of the eyelid:
   - Levator palpebrae: CN III.
   - Orbicularis oculi: CN VII.
   - Müller's muscle: sympathetic innervation.
   - Sensation: CN V.
2. Innervation of ocular muscles:
   - Lateral rectus: CN VI.
   - Superior oblique: CN IV.
   - Inferior oblique: CN III.
   - Superior rectus: CN III.
   - Medial rectus: CN III.
   - Inferior rectus: CN III.
3. Innervation regulating size of pupil:
   - Pupillary constrictor: CN III (parasympathetic fibers).
   - Pupillary dilator: sympathetic nerve (from superior cervical ganglion).
   - Ciliary muscle: CN III (relaxes the lens for accommodation).
   - Corneal sensation: CN V (ophthalmic branch-upper cornea; maxillary branch-lower cornea).
4. Innervation of pupillary light reflex:
   - Afferent: CN II to pretectum to Edinger-Westphal nuclei.
   - Efferent: inferior division of CN III. Parasympathetic preganglionic fibers to bilateral ciliary ganglia, then through postganglionic ciliary nerves to constrictor pupillae muscles.
   - Reflex inhibition of dilator pupillae (sympathetic).

5. Innervation of accommodation reflex:
   - Afferent: CN II to visual cortex (occipital lobe), to pretectum.
   - Efferent: CN III, IV, VI (somatic efferents) for convergence by extraocular muscles and CN III (visceral efferents) for pupillary constriction.
6. Innervation of vestibuloocular reflex:
   - Afferent: CN VIII to contralateral nucleus of CN VI and ipsilateral CN III.
   - Efferent: CN VI to contralateral lateral rectus; and CN III to ipsilateral medial rectus.

Injuries and Lesions

The most common cause of injury or lesion is traumatic injury. In blunt head trauma, which causes closed head injuries, all three oculomotor nerves may be involved peripherally or centrally, primarily or secondarily through edema or herniation. Deviation of the eyes is commonly seen in early stages of brain injury, although it is often temporary. The incidence is reported to be 3% to 7% in all head injury populations.[57]Cerebral trauma most commonly affects CN III, especially in children.[42] The superior rectus appears to be most severely involved with blunt trauma. Blunt trauma may also damage the pupillary sphincter directly or through ischemia, causing mydriasis, poor response to light, and poor accommodation. CN IV is less frequently involved but may be affected in mild head injuries. CN VI has the longest intracranial course of all the cranial nerves; it is therefore vulnerable and is frequently involved. Bilateral lesions occur in many cases; often, injury is due to stretching of nerves after broad frontal impact.

Ophthalmoplegia is secondary to orbital fracture, which affects mostly CN II, III, IV, and VI; fracture may also cause a sensory defect by injuring the ophthalmic division of CN V. Ophthalmoplegia secondary to basal skull fracture (sphenoid, petrous ridge) involving the cavernous sinus may involve all oculomotor nerves. Here again, because of its long intracranial course, CN VI is most frequently impaired.

Oculomotor nerve lesions must be distinguished from orbital displacement observed during blowout fracture of the orbit. Entrapment of the inferior rectus muscle may cause restriction of upward gaze.[58] Old trauma or chronic progressive ophthalmoplegia also may limit the range of motion of the globe through shortening or fibrosis of the ocular muscles. These particular causes can be discovered or ruled out by the "forced duction" test, which moves the globe mechanically and, therefore, evaluates range of motion passively.

Penetrating gunshot wounds may involve oculomotor nerves as well as CN II. Injuries to the upper cervical spine may involve CN VI, in addition to CN IX, X, XI, and especially XII.[1]

Nontraumatic lesions include inflammatory cavernous sinus disease (Tolosa-Hunt syndrome), which may involve all oculomotor cranial nerves and branches 1 and 2 of CN V (Figure 7-17). An-

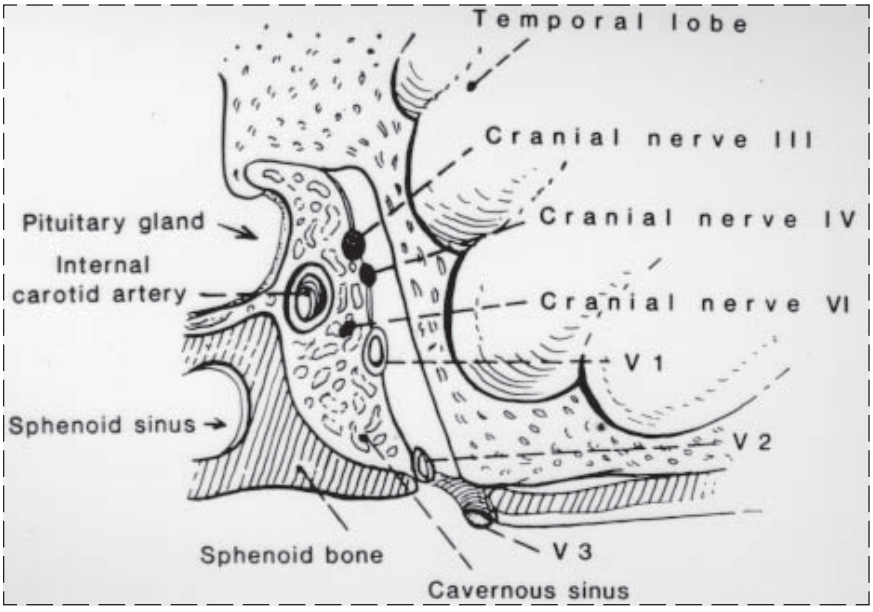

Fig. 7-17. Coronal diagram of the cavernous sinus. V_1: Ophthalmic division of cranial nerve V. V_2: Maxillary division of cranial nerve V. V_3: Mandibular division of cranial nerve V. Reprinted with permission from Brazis P, Masdeu J, Biller JL. *Localization in Clinical Neurology.* Boston, Mass: Little, Brown; 1990: 135.

other cause of lesion is septic thrombosis of the cavernous sinus.[59] In diabetic ischemic neuropathy, CN III and VI are most often affected. Frequently only one nerve is compromised. The pupillary light reflex is often reduced, although at times the pupil may be spared. Sparing can occur because ischemic lesions are frequently confined to the core of the nerve and spare the peripherally situated pupillary motor fibers.[55] In hypertension, CN VI fascicles can be infarcted and present as an isolated cranial nerve lesion.[60]

Inflammation and fibrosis are seen most frequently in thyroid ophthalmopathy, causing vertical diplopia because of asymmetric involvement of muscles with predilection of the inferior or superior rectus muscles. A myositis of the inferior oblique is common. Chronic progressive ophthalmoplegia is called Graefe disease. In myasthenia gravis, early involvement is seen in the medial rectus and levator palpebrae, monocular or binocular. The oculomotor nerve is also frequently involved in multiple sclerosis, more often causing internal rather than external ophthalmoplegia (60%); abducens nerve paresis has an incidence of 20%.[61] Involvement of oculomotor nerves by tumors is observed most commonly with pituitary adenomas that mainly involve CN III but is also common with meningiomas and nasal pharyngeal carcinomas. The most common lesions of the oculomotor nerves due to infections are seen in syphilis, scarlet fever, mumps, whooping cough, chicken pox, tuberculosis, and herpes zoster. Herpes zoster involves especially nerve III and the sphincter pupillae, levator palpebrae, and internal rectus muscles. Nutritional deficiencies, especially thiamin deficiency, may affect the function of the oculomotor nerves.

Other less common causes of ophthalmoplegia are Wernike's encephalopathy, internal carotid artery aneurysm or thrombosis, Paget's disease of the orbit, and Guillain-Barré syndrome. Occasionally there occurs a familial isolated oculomotor nerve lesion in association with Friedreich ataxia or Charcot-Marie Tooth disease.[62] It has been reported[63] that during dental anesthesia there may occasionally be paralysis of ocular muscles secondary to anesthetic injection into the inferior or superior dental artery. The anesthetic agent is carried through the maxillary artery, the middle meningeal artery, the lacrimal artery, and finally to the ophthalmic artery. As a postoperative complication following surgery or radiation therapy, ocular neuromyotonia may occur. The incidence was approximately 0.25% following spinal anesthesia.

Drugs or poisons such as phenytoin or phenobarbital may cause impairment of convergence and accommodation reflexes (watch for a diplopia occurring on near vision only). Lead may cause lateral rectus muscle paralysis, which develops rapidly; internal ophthalmoplegia may be observed. Methyl chloride and sodium fluoride poisoning may mimic botulism.

Syndromes Involving the Oculomotor Nerves

The following are syndromes involving the oculomotor nerves.

- Benedikt's syndrome: Ipsilateral ophthalmoplegia and contralateral hyperkinesia such as tremor, chorea, or athetosis resulting from a lesion of the tegmentum, which destroys the oculomotor nerve and the red nucleus on one side of the midbrain.
- Foville's syndrome: Pontine lesion causing contralateral hemiplegia with ipsilateral palsy of CN VII and ipsilateral paralysis of lateral gaze.
- Weber's syndrome: Ipsilateral ophthalmoplegia and contralateral hemiplegia. Ophthalmoplegia results from oculomotor nerve or nucleus interruption. Hemiparesis results from involvement of the cerebral peduncle with its corticospinal tract.
- Millard-Gubler syndrome: Ipsilateral facial weakness and contralateral hemiplegia, in many cases involving also CN VI, producing an internal strabismus. The lesion is in the pons.
- Duane's retraction syndrome: May follow paralysis of the lateral rectus muscle and is characterized by retraction of the eyeball on adduction of the eye with oblique upward movement of the eyeball and narrowing of the palpebral fissure.[64]
- Gradenigo's syndrome: Pain in the face caused by irritation of the semilumar ganglion; external rectus palsy. The syndrome is caused by meningitis at the tip of the petrous bone, usually secondary to purulent otitis media.[65]
- Wernicke's syndrome: Ocularmotor palsy due to involvement of the CN nuclei III or IV. Ptosis and pupillary changes are frequently observed, due to involvement of the red nucleus. Optic neuritis, retinal hemorrhages, ataxic gait, and muscular weakness may also occur.
- Möbius' syndrome: Ocular palsy in addition to facial palsy.

- Parinaud's syndrome: Conjugate ocular paralysis resulting in paralysis of upward gaze. This is mainly associated with lesions or disorders of the midbrain, especially the superior colliculi. It may be due to compression by pineal body tumor. Section of the posterior commissure can also produce this syndrome.

Symptoms and Signs

The symptoms and signs of oculomotor nerve lesions are described below. In addition, Table 7-2 reports them according to their location.

- Diplopia is the most common complaint associated with oculomotor nerve lesions. It is usually greatest in the direction of the weak muscle. In the position where the unfused images have their greatest separation, the more peripheral image will usually belong to the eye that has decreased mobility.
- Argyll Robertson pupil is miosis with loss of the light reflex and ciliospinal reflex, and with preservation of accommodation. Differential diagnosis includes neurosyphilis, multiple sclerosis, diabetes mellitus, pineal tumor, Wernicke-Korsakoff's syndrome, and midbrain encephalitis.
- Adie's pupil (myotonic pupil) is part of the Holmes-Adie syndrome. It is a benign condition in young women, often associated with absent deep tendon reflexes. It is of major importance to recognize the syndrome and so to prevent unnecessary investigation.[46]
- Pseudo-Graefe's syndrome is due to aberrant innervation (misdirection of regenerating nerve fibers). Most commonly it is observed after CN III and VI lesions, but it also occurs with CN III and X lesions. This is never caused by an ischemic neuropathy, but other lesions may cause nerve fibers to regenerate falsely.
- Exophthalmus may present secondary to extraocular muscle paralysis.
- Ocular neuromyotonia may be observed.

A complete isolated CN III lesion presents with the patient's eye being closed (ptosis). When opened manually the eye is found to be deviated outward and downward and the pupil is dilated. Unilateral, isolated oculomotor nerve palsy is most often related to ischemic diabetic neuropathy or to a lesion in the subarachnoid portion due to compression by an aneurysm. Isolated pupillary dilation is common in early uncal herniation. Migraine headaches may cause unilateral mydriasis lasting several hours. Seizures also can cause temporary unilateral mydriasis.

Other possible presentations of symptoms and signs secondary to CN III lesions are listed below.

- There may be paresis or paralysis of the superior rectus, the inferior rectus, the medial rectus, the inferior oblique muscles, and the levator palpebrae muscles (external ophthalmoplegia). One, a few, or all muscles may be affected.
- There may be dilation of pupils and reduction or absence of pupillary and accommodation reflexes (internal ophthalmoplegia).
- Bilateral large pupils may be seen normally in anxious young adults. Bilateral small pupils may be seen normally in the aged.
- There may be bilateral extraocular muscle weakness with sparing of the levator palpebrae muscle.[66]
- Bilateral ptosis is seen with nuclear involvement of CN III because of the midline position of the nucleus for the levator palpebrae muscle.
- Isolated bilateral ptosis with sparing of extraocular muscles and pupils may occur with encephalitis[67] or stroke.[68]
- Occasionally oculosympathetic spasms are associated with lesions of the cervical cord.
- Intermittent spasm of a portion of the pupillary sphincter may occur with recent onset CN III trauma or aberrant oculomotor reinnervation.

CN IV lesions in isolation are uncommon. The trochlear nerve may be involved in head injury, even in mild trauma. Other lesions occur mainly in conjunction with other oculomotor lesions. A complete CN IV lesion causes the eyeball to be turned upward and outward. Horner's syndrome may appear if the injury is near the sympathetic fibers. Symptoms and signs of CN IV lesions include

- weakness or paralysis of the superior oblique muscle, and
- vertical diplopia, which is greatest in down and inward gaze. The head tilts to the opposite side to compensate for diplopia. This is a characteristic sign.

TABLE 7-2

SYMPTOMS AND SIGNS ACCORDING TO LOCATION OF LESION–CRANIAL NERVES III, IV, VI

	Oculomotor	Trochlear	Abducens
Nuclear	Paresis/paralysis of oculomotor innervated muscles, contralateral superior rectus weakness, bilateral incomplete ptosis; complete one-sided CN III palsy rare; occasional isolated inferior rectus involvement[1]; possible bilateral paresis of CN III muscles. Occasional sparing of levator palpebrae[2]; pupillary constrictor weakness may be bilateral.	Contralateral superior oblique muscle weakness; lesion due to decussation.	Ipsilateral lateral rectus paresis; also ipsilateral gaze palsy (involvement of abducens interneurons), Möbius' syndrome, Duane's retraction syndrome.[3]
Fascicular	Complete CN III involvement unilateral; possible correctopia (irregularity of pupil); possible additional other neurological deficits (see midbrain syndrome).	Same as Nuclear. May in addition cause Horner's syndrome.[4,5]	Ipsilateral lateral rectus palsy and facial weakness and contralateral hemiparesis (Millard-Gubler syndrome); isolated lateral rectus palsy.[6]
Subarachnoid	Isolated unilateral CN III involvement; possible sparing of pupillary sphincter with incomplete lesions (pupillomotor fibers more dorsally located and smaller and more pressure resistant.[7]	None	Ipsilateral lateral rectus palsy; possible concomitant trigeminal nerve involvement (more so if lesion is in petrous bone.) Mostly secondary to chronic otitis media.
Cavernous Sinus	CN III innervated muscle weakness may involve also CN IV and VI and ophthalmic branch of CN V (combined ocular motor paresis, miosis, and poorly reactive pupil[8]); retroorbital pain.	Paresis superior oblique, may involve also CN III and VI and branch of CN V.	Paresis superior oblique, may involve also CN III and V (branch 1 and 2); retroorbital pain; occasional ipsilateral; Horner's syndrome.
Suborbital Fissure	Similar to cavernous sinus lesion; possible proptosis.	Similar to cavernous sinus lesion.	Similar to cavernous sinus lesion, except for *absence* of Horner's syndrome.
Orbit	Involvement of only superior or inferior branch of oculomotor nerve innervated muscles.	Weakness superior oblique muscle may be involved. CN III and VI also involved.	Paresis lateral rectus muscle; may also involve CN III and IV.

CN: cranial nerve

(1) Pusateri TJ, et al. Isolated inferior rectus muscle palsy from solitary metastasis to the oculomotor nucleus. *Arch Ophthalmol.* 1987;105:675. (2) Keane JR, Zaias B, Itabashi HH. Levator sparing oculomotor nerve palsy caused by a solitary midbrain metastasis. *Arch Neurol.* 1984;41:210–212. (3) Masdeu JC, Brazis PW. The localization of lesions in the oculomotor system. In: Brazis PW, Masdeu JC, Biller JL, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown 1990;140–143. Chap 7. (4) Coppeto JR. Superior oblique paresis and contralateral Horner's syndrome. *Ann Ophthalmol.* 1983;15:681–683. (5) Guy J, Day AL, Mickle JP, Schatz NJ. Contralateral trochlear nerve paresis and ipsilateral Horner's syndrome. *Am J Ophthalmol.* 1989;107:73–76. (6) Donaldson D, Rosenberg NL. Infarction of abducens nerve fasicle as cause of isolated sixth nerve palsy related to hypertension. *Neurology.* 1988;38:1654. (7) Nadeau SE, Trobe JD. Pupil sparing in ocularmotor palsy: A brief review. *Ann Neurol.* 1983;13:143. (8) Spector RH, Smith JL, Chavis PS. Charcot-Marie-Tooth disease mimicking ocular myasthenia gravis. *Ann Ophthalmol.* 1978;10:1033–1038.

CN VI lesions are manifested by paresis or paralysis of the lateral rectus muscle. In a complete CN VI lesion the eye is turned inward (internal strabismus). There may be ipsilateral gaze palsy, which may present as conjugate gaze palsies, showing symmetrical restriction of gaze to one side, up, or down. Horizontal gaze palsies are manifested by unilateral restriction to one side, mostly due to contralateral frontal or ipsilateral pontine damage; vertical gaze palsies are due to bilateral involvement of structures in the commissure. Lesions in the upper pontine tegmentum can cause both horizontal and vertical gaze palsies.[69] Skew deviation results from supranuclear derangements.[70] Oculomotor nerve lesions may cause vertical misalignment and nystagmus. Because nystagmus has many different causes, often related to the vestibular system, this impairment will not be discussed here in detail.

Saccadic deficits commonly occur following brain injury.[71] There may be decreased saccadic accuracy, inability to fixate the gaze in the contralateral field, or decreased initiation of saccades toward the contralateral side of the lesion.[72]

Evaluation

Clinical examination observes the movements of the eye by having the patient follow a light stimulus. It is important to observe the reflection of light from the cornea to assess alignment (Hirschberg reflex).[73] Spontaneous voluntary and reflex gaze without a light stimulus must also be assessed. If diplopia is present, it must be determined whether it is a monocular or binocular diplopia. Physical evaluation should begin with examination of the eyelids; they should be checked for ptosis of the lid, upper, lower, or both. Pupils should then be examined for size (normally 2–6 mm in ambient light) and regularity (anisocoria up to 30% is normal).[74] Response to visual stimulus and accommodation must be observed. Further, clinical examination must rule out abnormal movement such as nystagmus.

It is important to look for a misdirection syndrome, which can be observed many months following CN III lesions, secondary to an aberrant regeneration. Fibers from the ocular muscles may regenerate aberrantly to the levator palpebrae, resulting in the pseudo-von Graefe's sign (lid elevation during an attempt to look down or lid winking while chewing). It is important to look for bilateral involvement because it is not uncommon in CN IV and VI lesions. The head tilt test demonstrates a CN IV lesion.[75] The observed degree of diplopia is greatest when looking down, and this causes the compensatory head tilt to the opposite side.

Miosis in darkness represents a paradoxical constriction (Flynn phenomenon) and may be congenital. It is seen in optic atrophy and bilateral optic neuritis.[76] In light, near dissociation, absent pupillary reflex to light and present convergence, is termed an Agyll Robertson pupil and may be seen with syphilis, sarcoidosis, diabetes, myotonic dystrophy, amyloidosis, or in aberrant regeneration.

Examination should include careful evaluation of gaze to rule out conjugate or dysconjugate gaze palsies. The lower motor neuron controls muscles; the upper motor neuron controls movement and gaze. The gaze must further be evaluated by observing automatic and planned eye movements. It is important to look for saccade deficits. These can be measured by having the patient look quickly from one object to another. The test objects should be held 6 in. apart and approximately 15 in. from the patient.[77] Mild traumatic brain injury can cause aberrant saccades and oscillations.[78]

Electrodiagnosis Tests

Electrodiagnostic tests for evaluating oculomotor nerve function utilize electronystagmography (ENG).[79] This is described in the discussion of CN VIII. Electrooculography may be of help in the diagnosis in eye movement disorders.[69] EMG of external ocular muscles is carried out according to standard electromyographic procedures.

Imaging and Management

CT scans and MRI scans are useful especially when delayed diplopia occurs. Spontaneous recovery within 9 to 12 months is not uncommon. In children, recovery up to 80% or 90% is reported.[7]

Diplopia is initially managed with eye patching. The sound eye is patched to encourage full excursion of the involved eye and to increase its function. Disuse amblyopia does not occur in adults and there is no need for alternate patching. However, during critical activities the involved eye should be patched to allow optimal performance. When the patient is able to suppress the second image, patching can be discontinued. Pleoptic exercises as well as stereoscopic training devices may be used to improve muscle excursion (Worth Four Dot flashlight).[80,81] Another recommended intervention is the use of Fresnel lenses[56] to preserve binocular vision. If after a prolonged time of observation (9 to 12 mo)

and appropriate exercises, no significant improvement is noted, surgical procedures may be considered for functional or cosmetic reasons. Surgical procedures frequently show excellent results, especially for persistent trochlear palsy, but less for abducens or oculomotor lesions. Another alternative is injection of botulin toxin into the antagonist of the paralyzed muscle.

Recent research[56] suggests that the best therapeutic approach to visual perceptual dysfunction is to aim at increasing the skills of visual attention, scanning, pattern recognition, visual memory, and ultimately cognition. To accomplish this goal the therapeutic approach employs strategies for remediating and compensating deficits in foundation skills, such as oculomotor control, visual field, and acuity. Visual field deficits are best evaluated with computerized automated perimetry.[82] Limited visual fields can be increased by training with repetitive intensive stimulation of the blind hemifield. Compensation for visual field deficits can also be increased with training. As already mentioned, training exercises can improve oculomotor control. Visual acuity has to be optimized with corrective lenses and improvement of lighting conditions. The patient is then taught an increased awareness of the deficit and how to "intellectually override" by repeated practice and meticulous planning of compensatory techniques for both self-care and academic activities.

CRANIAL NERVE V: TRIGEMINAL

The trigeminal nerve is a mixed nerve that is chiefly sensory. It has three major divisions, shown in Figures 7-18, 7-19, and 7-20.

Anatomy and Function

The three major divisions of the trigeminal nerve are the ophthalmic, the maxillary, and the mandibular (see Figures 7-18 and 7-19).

The *ophthalmic nerve* (CN V_1) supplies sensation to the upper part of the face, including the eyes (upper half of cornea, conjunctiva, and iris), paranasal sinuses, and part of the nasal mucosa and meninges. It is located in the lateral wall of the cavernous sinus near CN III, IV, and VI. The ophthalmic nerve and CN III, IV, and VI enter the orbit together through the superior orbital fissure. The ophthalmic nerve has three major branches: (1) the frontal nerve, which branches into supraorbital and supratrochlear nerves; (2) the lacrimal nerve; and (3) the nasociliary nerve.

The *maxillary nerve* (CN V_2) supplies sensation to the lower half of the cornea, the conjunctiva, and the iris; the upper jaw, teeth, lip, cheeks, and hard palate; and the maxillary sinuses and nasal mucosa. The maxillary nerve is located inferior to the ophthalmic division in the cavernous sinus. It leaves the cranium through the foramen rotundum and enters the orbit through the inferior orbital fissure. Its branches are the infraorbital, superior alveolar, zygomaticofacial and zygomaticotemporal, and the greater and lesser nasopalatine nerves.

The *mandibular nerve* (CN V_3) receives sensation from the lower jaw; teeth, lip, buccal mucosa, tongue; and part of the external ear, auditory meatus, and meninges. It supplies the skin of the chin, lower lip, and lower jaw, except for the area over the mandibular angle, which is supplied by the auricular nerve from the second and third cervical nerve roots. Sensory branches of CN V_3 are the buccal nerve, auriculotemporal nerve, lingual nerve, inferior alveolar nerve, and the meningeal branch of the mandibular nerve.

Trigeminal motor fibers supply the masseter muscle, the temporalis muscle, the medial and lat-

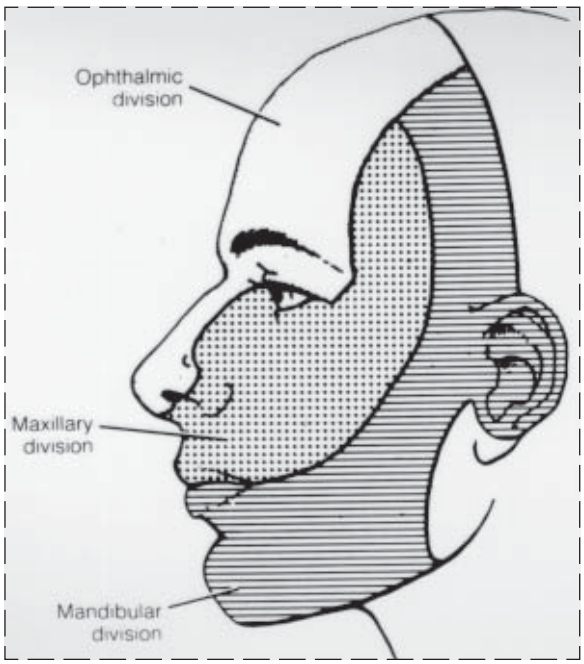

Fig. 7-18. Sensory distribution of cranial nerve V. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 21st ed. Norwalk, Conn: Appleton & Lange; 1991: 154.

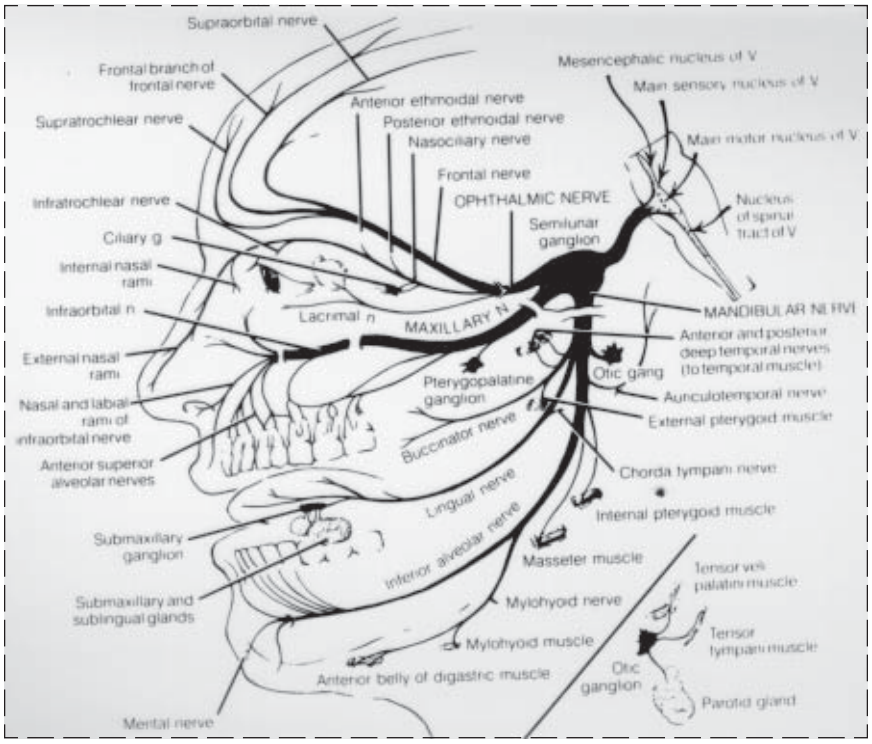

Fig. 7-19. The trigeminal nerve. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 21st ed. Norwalk, Conn: Appleton & Lange; 1991: 153.

eral pterygoid muscles, the mylohyoid muscle, and the anterior belly of the digastric muscle. Branches pass also to the tensor tympani and the tensor veli palatini (see Figure 7-19) The motor nucleus of CN V is located medial to the main sensory nucleus (see Figure 7-20) near the floor of the fourth ventricle. Motor fibers leave the midlevel pons on the ventricle surface, transverse the cranium through the foramen ovale, and join the mandibular nerve to reach and supply the muscles of mastication. Via the optic ganglion, fibers reach the tensor tympani and the tensor veli palatini. Via the mylohyoid nerve, fibers reach the mylohyoid muscle and the anterior belly of the digastric muscle.

Sensory receptors within the trigeminal nerve system include mechanoreceptors (rapidly and slowly adapting), thermoreceptors (warm and cold), nociceptors (responding to painful stimuli, possibly including chemoreceptors), and proprioceptors (responding to muscle or joint position).[83] The primary sensory neurons for tactile sensation (see Figure 7-20) have their cell bodies located in the gasserian (semilunar) ganglion, located in the middle cranial fossa near the cavernous sinus. Fibers terminate in the most lateral of the three sensory nuclei, the main sensory nucleus of the trigeminal nerve. Primary neurons for pain and temperature sensation have their cell bodies also located in the gasserian ganglion, but fibers terminate in the spinal nucleus, the most caudal of the trigeminal nerve. Primary neurons for proprioception (sensory

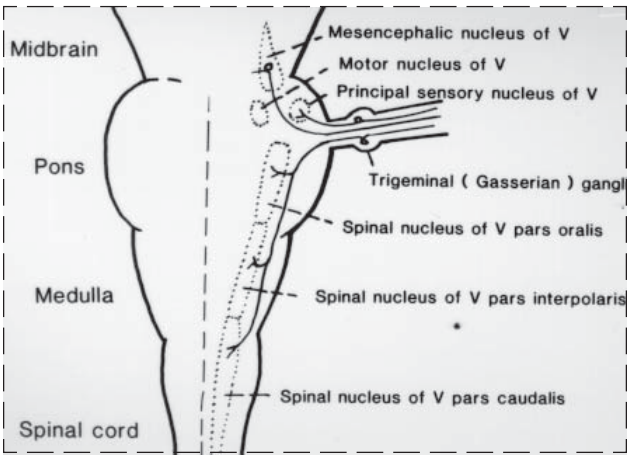

Fig. 7-20. Schematic diagram of the trigeminal nuclei. Reprinted with permission from Brazis P, Masdeu J, Biller JL. *Localization in Clinical Neurology.* Boston, Mass: Little, Brown; 1990: 191.

neurons for spindle stretch receptors in masseter and temporalis muscles) have their cell bodies *not* in the gasserian ganglion, but in the most superior trigeminal nucleus, that is, in the mesencephalic nucleus in the midbrain tegmentum (see Figure 7-20). Cell bodies of the golgi tendon organ for jaw closure muscles are located in the gasserian ganglion.

Central connections for CN V show sensory pathways from the spinal nucleus terminating in the thalamus. Reflex connections pass to the motor nuclei of CN V, VII, and IX. The motor nucleus receives bilateral, mainly crossed, cerebral connections, which originate as corticobulbar fibers in the lower frontal motor cortex and descend through the internal capsule; they decussate in the pons and supply the trigeminal motor nucleus. There also is input from extrapyramidal tracts.

The anatomic substrates for the reflexes involving CN V are as follows:

1. Masseter reflex:
   - Afferent: Muscle spindles to proprioceptive fibers from muscles of mastication through the mandibular nerve to the mesencephalic CN V motor nucleus, then monosynaptically to efferent limb.
   - Efferent: Trigeminal motor nucleus to mandibular nerve to extrafusal fibers in masseter and temporalis muscles.
2. Corneal reflex:
   - Afferent: A-delta fibers from the upper cornea (smaller than the fibers subserving the blink reflex) to the long ciliary nerve, to the ophthalmic nerve to the pons, to the spinal trigeminal nucleus; and A-delta fibers from the lower cornea to the maxillary nerve, to pons and also to spinal trigeminal nucleus; then multisynaptically to efferent limb.
   - Efferent: Direct response: ipsilateral facial nerve nucleus to nerve to orbicularis oculi. Consensual response: contralateral facial nerve nucleus to nerve to orbicularis oculi.
3. Blink reflex: (Figure 7-21)
   - The afferent limb consists of the supraorbital nerve to ophthalmic nerve to main sensory nucleus (CN V), then oligosynaptically to the efferent limbs, for which there are two possible efferent expressions. (1) Efferent: Facial nerve motor nucleus, ipsilateral facial nerve to orbicularis oculi—R1 response (early response) direct and polysynaptically from the spinal sensory nucleus of CN V. (2) Efferent: Bilateral facial nerve nuclei to nerve to orbicularis oculi muscle—R2 response direct and consensual. R2 coincides with eyelid closure.

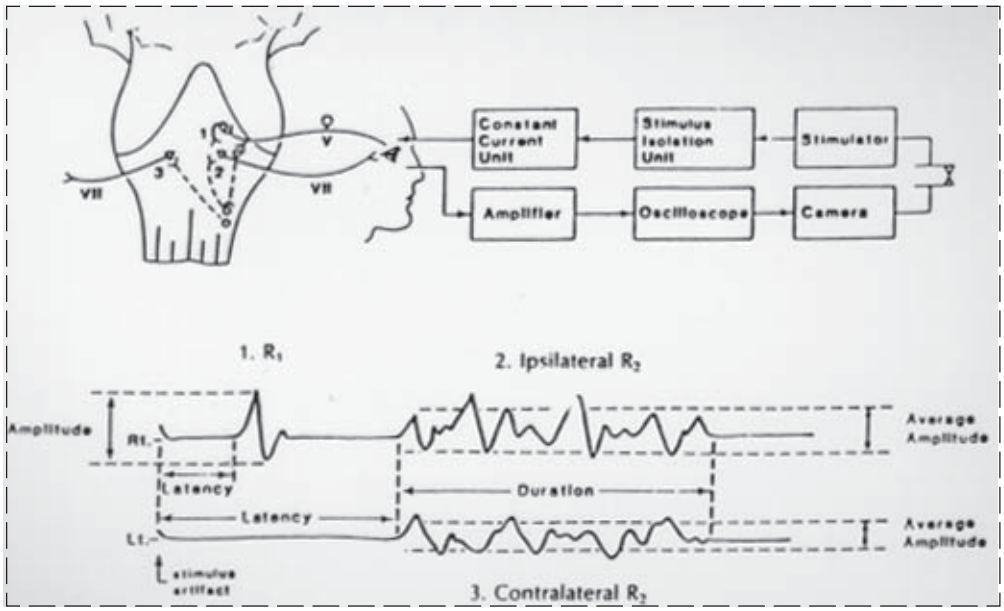

Fig. 7-21. Stimulation and recording arrangements for the blink reflex and its projected pathways. Reprinted with permission from Kimura J. *Electrodiagnosis in Diseases of Nerve and Muscle: Principles and Practice.* 2nd ed.Philadelphia, Pa: FA Davis; 1989: 308.

The corneal reflex differs from the blink reflex. It uses different interneurons; there is no early response (R1) and there is a shorter latency to R2; there is less habituation than with the blink reflex.

Injuries and Lesions

The most common causes of injury or lesion are traumatic injuries, including skull fractures and orbital floor blow-out fracture, mainly injuring the maxillary nerve.[84,85] Blunt trauma often affects the supraorbital notch, injuring the supraorbital and supratrochlear nerves. Penetrating gunshot wounds may directly injure the semilunar ganglion. Nontraumatic lesions of CN V may be due to tumors such as meningioma, schwannoma, metastasis, nasopharyngeal carcinoma, or acoustic neuroma[86]; or to aneurysm, multiple sclerosis, syringobulbia, or infections.

In surgical trauma the most commonly affected nerve is the lingual nerve during lower third molar tooth extraction. Blackburn and Bramley[87] report an incidence of 11%; 0.5% of these patients experienced permanent sensory loss from the ipsilateral tongue.

A significant presentation of CN V involvement is trigeminal neuralgia (tic douloureux or Fothergill's disease), which may be idiopathic or due to multiple sclerosis, cerebellopontine angle tumor, or aberrant blood vessel.[88,89] A trigeminal sensory neuropathy may occur with Sjögren's syndrome. Other causes may be idiopathic, with possible full recovery within several months. Rheumatoid arthritis, systemic sclerosis, systemic lupus erythematosis, dermatomyositis,[90] and temporal arteritis[91] may present as sudden onset of unilateral or bilateral tongue numbness. Herpes zoster may affect the gasserian ganglion.[92]

Syndromes involving the trigeminal nerve are as follows:

- Tic douloureux (Fothergill's disease)[93] presents with severe pain in the trigeminal nerve distribution following irritation of a trigger zone. It is usually confined unilaterally to one division of CN V in adults over 40 years of age. Occasionally it is associated with dental or sinus disease.
- Paratrigeminal syndrome (Raeder's syndrome) is trigeminal neuralgia due to a semilunar (gasserian) ganglion tumor followed by facial anesthesia (rare). It presents also with paresis of muscles of mastication. CN III may be involved and there may be an ipsilateral Horner's syndrome.
- Auriculotemporal nerve syndrome (Frey's syndrome) presents with flushing and sweating during eating in the distribution of the ipsilateral auriculotemporal nerve. This may follow an injury or infection of the parotid gland area.
- Bonnier's syndrome involves CN III, V, VIII, IX, and X, with symptoms of Ménière's disease, contralateral hemiplegia, somnolence, apprehension, and weakness. The lesion is in the lateral vestibular nucleus and adjacent pathways.
- Gradenigo's syndrome presents with CN V_1 (ophthalmic branch) and CN VI deficits secondary to a lesion at the apex of the temporal lobe (osteitis or leptomeningitis secondary to otitis media); it also may be caused by trauma or tumor.
- Cavernous sinus syndrome may be secondary to trauma, carotid aneurysm, carotid-cavernous fistula, or infection. It may involve the ophthalmic as well as the maxillary branch of CN V, and in addition, CN III, IV, and VI.
- Superior orbital fissure syndrome may be caused by tumor, trauma, aneurysm, or infection. It may present as ophthalmoplegia associated with pain, paresthesias, and sensory loss (CN V, ophthalmic branch, and CN III, IV, and VI). There may be exophthalmos due to ophthalmic vein blockage and occasionally the maxillary branch is simultaneously involved. Except for exophthalmos it is difficult to differentiate a cavernous sinus syndrome from a superior orbital fissure syndrome.
- Horner's syndrome consists of miosis (paralysis of pupil dilator) and ptosis (Müller's muscle paralysis). There is often slight elevation of the lower lid because of paralysis of the lower tarsal muscle, which gives the appearance of an enophthalmus. Anhidrosis is possible, but is often not present if the lesion is beyond the carotid bifurcation.
- Neck and tongue syndrome presents with numbness of the tongue. This may be seen in temporal arteritis or it may be due to irritation of the cervical second dorsal nerve root, because proprioceptive fibers from the tongue via the hypoglossal nerve enter through the C2 nerve root.[94]
- Numb cheek syndrome may be caused by a maxillary fracture or lesion such as a recurrent squamous cell carcinoma of the

skin causing injury to the infraorbital nerve.[95] It may actually be the initial presentation of a nasopharyngeal tumor.
- Numb chin syndrome (Roger's sign) is caused by adhesions of the mandibular branch of CN V, which also may cause masticatory paralysis. Symptoms and signs are pain, swelling, and numbness of lower lip and chin (carcinoma of breast and lung, lymphoma).

Different symptoms correspond to different sites of lesions. Lesions located in the supranuclear area present with contralateral paresis of the muscles of mastication, causing deviation of the jaw away from the lesion. Even though innervation is predominantly from the contralateral hemisphere there is some bilateral control of CN V motor function. Usually this means that weakness is not too severe. Bilateral upper motor neuron lesions cause severe weakness of chewing muscles and an exaggerated jaw jerk. Lesions in the nuclear area are diagnosed by "the company they keep" (other cranial nerves or long tracts or both, as described with the anatomy of CN III, IV, VI, and the trigeminal). Cavernous sinus lesions may present with sensory deficits in the first (ophthalmic nerve) and second (maxillary nerve) divisions of CN V. In addition, there may be oculomotor nerve involvement (CN III, IV, and VI). Supraorbital fissure lesions show sensory loss in the ophthalmic nerve distribution (CN V), and oculomotor nerve lesions (specifically CN III, IV and VI).

The symptoms or signs of CN V lesions are pain, sensory loss, weakness of chewing muscles, and reflex changes, and more specifically as follows:

- Facial pain may be significant, especially with gasserian ganglion involvement.
- Loss of sensation may present with early corneal anesthesia. It may occur as isolated impairment of ipsilateral tongue sensation due to lingual nerve (mandibular nerve branch) involvement or lingual nerve injury. It may present with partial or complete loss of sensation of one, two, or all three branches of CN V.
- Dissociated anesthesia (trigeminal spinal tract involvement—syringobulbia) presents with loss of pain but not of touch; it may be present if the spinal tract of CN V is involved. Facial paresthesias, however, may also be seen in anemia or in nervous patients without cranial nerve lesion.
- Paralysis of muscles of mastication with deviation of jaw to the affected side.
- Loss of reflexes: Blink reflex, jaw jerk, sneeze, lid, conjunctiva, and corneal reflexes.
- Reduced hearing may be due to paralysis of the tensor tympani.
- Triasmus (lockjaw) is a tonic spasm of muscles of mastication (eg, in rabies, tetanus, epilepsy, and hysteria).
- Trophic and secondary disturbances such as dryness of nose, ulcerations of face, and loss of teeth may also occur (Herpes simplex, neurokeratitis).

Evaluation

The clinical examination consists of careful evaluation of sensory and motor functions and reflexes. The sensory exam must evaluate pinprick, light touch, and temperature over the three divisions of the trigeminal nerve on each side. The skin over the angle of the jaw is supplied by the auricular nerve.

Special attention must be paid to the sensation of the tongue, especially if the patient's history includes a recent lower third molar tooth extraction. Tongue numbness may cause difficulty in the oral phase of swallowing and in speech articulation, which is often noticed when the patient talks on the telephone. The patient may also accidentally bite the tongue.

To help identify lingual nerve compromise a moving two-point discrimination test was developed and recommended by Blackburn and Bramley.[87] The test is based on the observation of Mountcastle and colleagues[96] and Dellon[97] that touch sensation is mediated through quick and slow adapting fibers. Quick adapting fibers mediate moving touch, and they were found to greatly outnumber slow adapting fibers in the nerve to the tongue. The likelihood of identifying abnormalities is significantly increased by testing a larger number of fibers. Blackburn and Bramley[87] obtained two-point discrimination scores in the tongue of stimuli 1 mm and 3 mm apart. In normals there was a small difference between the two sides, with the threshold on the right being somewhat lower. Blackburn and Bramley also demonstrated that the test could predict recovery or nonrecovery correctly in approximately 90% of patients. Seven test stimuli were presented in a moving fashion. A score of one to three abnormal responses indicated a good prognosis for recovery, while four or more wrong responses indicated a poor prognosis. Test results may be helpful in considering possible surgical exploration.

Reflex testing should include the corneal test, in which a light touch of the cornea causes contraction of bilateral orbicularis oculi muscles. The blink reflex is tested by a tap applied to the glabella, which causes bilateral orbicularis oculi contractions.

Motor evaluation is carried out by testing the strength of jaw opening and lateral deviation. Weakness of one side will cause the open mouth to deviate toward that side. It is important to palpate for the contraction of the temporalis and masseter muscles while the patient bites on a tongue depressor. To test the jaw jerk, tap the mandible. If the jerk is visible or palpable, a bilateral upper motor neuron lesion above the level of CN V motor nucleus is suggested.

Electrodiagnosis

Electrodiagnostic testing can evaluate the masseter reflex,[98,99] the masseter inhibitory reflex,[100–102] the pterygoid reflex,[103] the blink reflex,[104–106] and the corneal reflex,[107] by means of reflex nerve conduction, trigeminal evoked responses, mandibular motor nerve conduction, and EMG of muscles of mastication. The masseter reflex is a myotatic reflex conducted through the midbrain. Afferent proprioceptor fibers innervate cell bodies in the mesencephalic nucleus and have monosynaptic connections with the motor nucleus of CN V for the efferent pathway to the masseter muscle. Unlike the spindle afferents in the limb musculature, it appears that the primary spindle afferents from the masseter muscle exert no direct inhibitory influence on the antagonistic muscles (those opening the jaw). Vibration of the jaw muscles facilitates the masseter reflex, while at the same time such vibration inhibits the "H" reflex.

The procedures for the electrically elicited masseter reflex and for the masseter inhibitory reflex follow.

Electrodiagnostic Evaluation of Masseter Reflex

Stimulation: A mechanical tap is applied with an electronic reflex hammer to the examiner's finger held to the subject's chin. A microswitch triggers the oscilloscope sweep.

Recording: This is accomplished by active surface or needle electrodes placed bilaterally on or in the lower third of the masseter muscles. Responses are recorded simultaneously from both sides. The reference electrodes are placed below the mandibular angle, and the ground is placed on the forehead.

Normal values: Because reflex latencies vary with successive trials, asymmetry of simultaneously recorded right and left values is more meaningful than absolute values.

- Amplitude: This is variable and depends on the weight supported by the mandible. The amplitude ratio between the two sides is clinically meaningful.
- Latency: Approximately 8.4 millisecond (range 6.4–9.2 ms). A difference of more than 0.5 millisecond between the two sides is abnormal, as is bilateral absent responses in persons below the age of 70 years.

The masseter inhibitory reflex is tested by stimulation of the infraorbital or mental nerve during maximal contraction of the masseter muscle (teeth clenching). Two silent periods ($SP_1$and SP_2) can be observed in the EMG interference pattern. This is believed to be a protective mechanism to prevent intraoral injury from excessive jaw movement during speech in patients with movement disorders.

Procedure to Evoke Masseter Inhibitory Reflex

Stimulation of the mental nerve is carried out in the mental foramen with a stimulus of 0.2 millisecond duration and approximately 30 to 40 mA.

Recording is bilateral on the masseter muscles.

Normal values:

- Latency: SP_1 10-15 millisecond, mean 11.4 $\pm$ 1.3 millisecond; SP_2 40-50 millisecond, mean 47 $\pm$ 6 millisecond
- Duration: SP_1 20 $\pm$ 4 millisecond; SP_2 40 $\pm$ 15 millisecond

Electrodiagnostic Evaluation of Blink Reflex

The blink reflex[104,105] (see Figure 7-21) can be elicited mechanically with an electronic hammer tapping the glabella (this is a cutaneous rather than a stretch reflex). A midline tap will cause an R1 component bilaterally. Latencies for R1 are a few milliseconds longer than those elicited with electrical stimulation. The electrically evoked blink reflex procedure is explained below.

Electrodiagnostic Evaluation of Pterygoid Reflex[103]

Stimulation: To elicit the masseter reflex, a mechanical cap is applied with an electronic reflex hammer to the examiner's finger held to the

subject's chin. A microswitch triggers the oscilloscope sweep.

Recording: To record, two monopolar needle electrodes are placed into the medial pterygoid muscle 6 to 10 mm apart, and 22 to 25 mm beneath the skin. Insertion is halfway between the angle of the mandible and the facial artery at its crossing of the mandible. Jaw closure can confirm adequate placement by observing the appropriate EMG signals. Since there is midline stimulation at the identical point for the masseter and pterygoid reflexes, all four can be elicited simultaneously if four channels are available.

Normal values: Values are slightly faster than that of the masseter reflex, which may have to do with the location of the subnuclei within the trigeminal mesencephalic nucleus; a cluster of neurons for the pterygoid muscles being located more caudally than those for the masseter.

- Amplitude: Amplitudes with needle recording are unreliable.
- Latency: 6.9 ± 0.43 millisecond, and side to side difference: 0.29 ± 0.21 millisecond. It is believed that testing both the masseter and the pterygoid reflexes provides a more precise localization of a small pontomesencephalic lesion.

Procedure to Evoke the Blink Reflex

Stimulation: The stimulation to evoke the blink reflex is carried out with cathode placement over the supraorbital fissure. The anode is placed laterally and above the cathode to avoid current spread to the other side. Optimal shock frequency is one shock every 7 seconds to avoid habituation. Stimulation is best applied between blinks.

Recording is best with superficial disks. The active electrode is placed 1 cm below lid margin, below the pupil, or slightly lateral over the orbicularis oculi. The reference is placed over the zygoma, or the nasalis muscle. The ground is placed on the chin.

Normal values:

Latencies in adults[104]:

- R1: 10.5 millisecond $\pm$ 0.8 millisecond (difference between two sides less than 1.2 ms)
- R2: 30.5 millisecond $\pm$ 3.4 ipsilateral (upper limit of normal 40 ms);
  30.5 millisecond $\pm$ 4.4 contralateral (upper limit of normal 41 ms)
- R1-D ratio: 3.6 $\pm$ 0.5 (D = direct facial nerve response)

The difference between R2 ipsilateral and R2 contralateral should not exceed 5 millisecond. The latency difference between R2 evoked by right side stimulation and corresponding R2 evoked by left side stimulation should be less than 7 millisecond.

Latencies in neonates[106]:

- R1: 12.1 millisecond $\pm$ 1.0 millisecond
- R2: 35.9 millisecond $\pm$ 2.5 millisecond ipsilateral;
  Contralateral often absent
- R1-D ratio: 3.7 $\pm$ 0.4

Stimulation: infraorbital fissure
Recording: same as above
Normal values:

Latency in adults:

- R1: none
- R2: 41 millisecond ipsilateral (upper limit of normal);
  42 millisecond contralateral (upper limit of normal)

Physiological variations observed in blink reflex recordings are variations during light sleep such that the R1 amplitude decreases and may disappear, and the R2-D ratio is of prolonged duration. During feelings of apprehension, R1 is decreased in amplitude and R2 is increased in amplitude. With repeated trials R1 is more stable than R2 and therefore better suited for assessing the trigeminal nerve. Analysis of R2 is essential to determine whether the afferent or efferent limb of the reflex is primarily involved. If R1 is unstable, paired shocks with 5-millisecond interstimulus intervals may be utilized to facilitate the response. The first shock should be simply a conditioning stimulus of lower amperage. Late responses are absent in coma.

The corneal reflex[107] and the blink reflex may help in evaluating brainstem interneuronal activity; disease or injury affecting the reflex arc; or lesions, such as compression or hemispheric, that indirectly affect the reflex arc. The procedure for eliciting the corneal reflex follows:

Electrodiagnostic Evaluation of Corneal Reflex

Stimulation is carried out through a thin, saline soaked cotton thread connected to a constant current stimulator. The anode is placed on the ipsilateral earlobe. A stimulus of 1 millisecond duration is employed, and responses are usually observed with a stimulus of 0.1 to 3 mA.

Recording is carried out from bilateral orbicularis oculi muscles.

Normal values are latencies of 35 to 50 millisecond.

The difference between the direct and consensual response is 5 millisecond or less. The difference between responses evoked from both corneas is 8 millisecond or less.

Sensory nerve conduction[108] can be determined by *stimulating* the supraorbital nerve at the supraorbital fissure and *recording* from a superficial electrode over the forehead 4 cm distal to the point of stimulation. Normal values are latencies from 1.3 to 1.5 millisecond and amplitudes of 5 to 8 μV.

Trigeminal evoked responses[109] (Figure 7-22) can be elicited over the scalp when superficially *stimulating* the lips (on the right and left sides independently), the gingiva, or the infraorbital nerve, with a current strength approximately 3-fold that of the sensory threshold. An averaging procedure is necessary as follows.

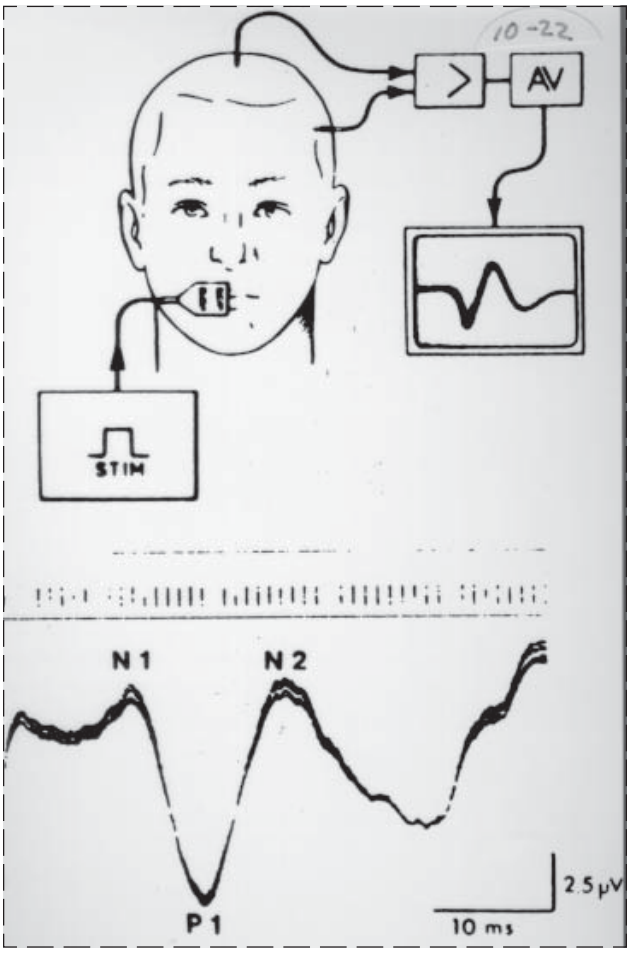

Fig. 7-22. A cortical sensory evoked potential of the trigeminal nerve elicited with stimulation of the lips. Reprinted with permission from Kimura J. *Electrodiagnosis in Diseases of Nerve and Muscle: Principles and Practice*. 2nd ed. Philadelphia, Pa: FA Davis; 1989: 396.

Procedure for Trigeminal Evoked Potential Studies[109]

Stimulation: This is supplied to lips, gingiva, or infraorbital nerve. Shocks between 1 and 2 Hz are averaged.

Recording: Electrodes are applied 1 or 2 cm behind C_5 or C_6, that is, between C_3 and T_3 or between C_4 and T_4 respectively (according to International 10-20; see Figure 7-5). The reference electrode is placed at F_Z.

Normal values: N1 is the first small negative response, often not easily distinguishable. P1 (second wave) can be used for measuring latency.

- Normal latency is 18.5 $\pm$ 1.5 millisecond. The mean difference between the two sides is 0.55 $\pm$ 0.55 millisecond.
- Amplitude N1 to P1 is 2.6 $\pm$ 1.05 μV. The difference between the two sides is 0.51 $\pm$ 0.54 μV.

Mandibular motor nerve conduction can be studied through magnetic stimulation to the central cortex, with recording electrodes placed over the muscles of mastication. This method can detect nerve conduction abnormality, but at present the appropriate equipment is not available for use in most laboratories.

Peripheral mandibular motor nerve conduction was reported by Dillingham and associates[110] Stimulation is carried out intraorally where the deep temporal nerve branches from the CN V_3. Recording is carried out from the temporalis muscle. The mylohyoid nerve (another branch of CN V_3) is also stimulated with recording from the mylohyoid muscle. Latencies for the deep temporal nerve are reported to be 2.1 $\pm$ 0.2 millisecond, and for the mylohyoid nerve they are 1.8 $\pm$ 0.2 millisecond.

EMG of the masseter muscles can easily be performed and evaluated in the usual manner. During voluntary jaw clenching, the elicited jaw reflex will cause a brief pause in the electromyographic activity of the masseter muscle. In normal subjects this pause lasts approximately 13 millisecond. It is prolonged in temporomandibular joint syndrome and decreased or absent in patients with tetanus.

The prognosis for tic douloureux is relatively good for pain relief with medications or surgery or

both. For secondary neuralgias the results are less satisfying.

Management

Facial pain or tic douloureux is successfully managed using the medications phenytoin, carbamazepine, and more recently also baclofen, chlorphenesin, and mephanesin. If no relief is obtained, surgery may have to be considered. Procedures most successful for relief of trigeminal pain are gangliolysis or microvascular decompression. Retrogasserian neurotomy or peripheral neurectomy may also be considered.[93]

Ablative trigeminal surgery is sometimes followed by anesthesia dolorosa, which is manifested by severe constant pain in the anesthetic area. Unfortunately, this condition is very resistant to treatment.[111,112]

CRANIAL NERVE VII: FACIAL

The facial nerve is mixed, but chiefly motor. It contains both somatic and visceral efferent components, and the afferents likewise are somatic and visceral. The anatomy is depicted in Figures 7-23 and 7-24.

Anatomy and Function

Motor branchial efferents are fibers from the motor nucleus of CN VII, located in the reticular formation of the caudal pons. The fibers loop around the nucleus of CN VI and then are joined by the visceral efferent fibers of the nervus intermedius of Wrisberg. The nervus intermedius carries sensory fibers to the nucleus tractus solitarius, and parasympathetic fibers from the superior salivatory nucleus. CN VII (motor) and nervus intermedius (the parasympathetic and sensory part of CN VII) emerge together from the brainstem in the region of the cerebellopontine angle just medial to the acoustic nerve (CN VIII). They cross the posterior cranial fossa and enter the internal auditory meatus of the temporal bone. Within the temporal bone the facial nerve can be divided into four parts: (1) the meatal segment, (2) the labyrinthine segment, (3) the horizontal segment, and (4) the mastoid segment.[113]

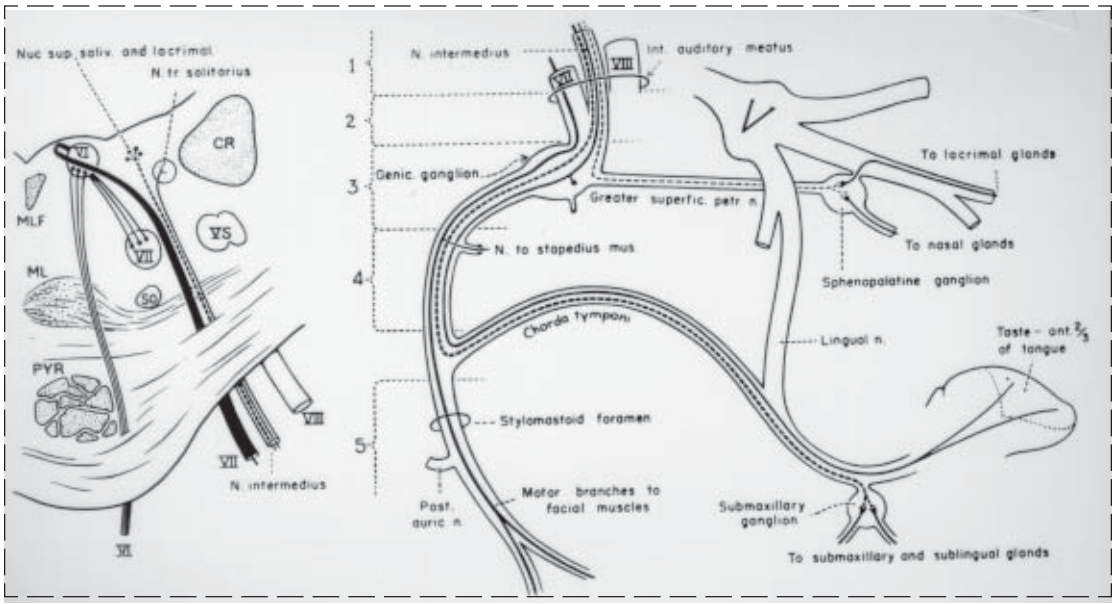

Fig. 7-23. Origin and distribution of various components of the facial nerve. The facial, intermedius, and acoustic nerves illustrated on the left continue in the drawing on the right. CR: Corpus restiforme; MLF: Medial longitudinal fasciculus; ML: Medial lemniscus coursing vertically through corpus trapezoideum; PYR: Pyramidal bundles in pars basilaris pontis; SO: Superior olivary nucleus; VS: Nucleus of spinal tract of the trigeminal nerve. Reprinted with permission from Haymaker W, Kuhlenbeck H. Disorders of the brainstem and its cranial nerves. In: Baker AB, Baker LH, eds. *Clinical Neurology.* Vol 3. Hagerstown, Md: Harper and Row; 1976: 28. (Used with permission from J.B. Lippincott Co.)

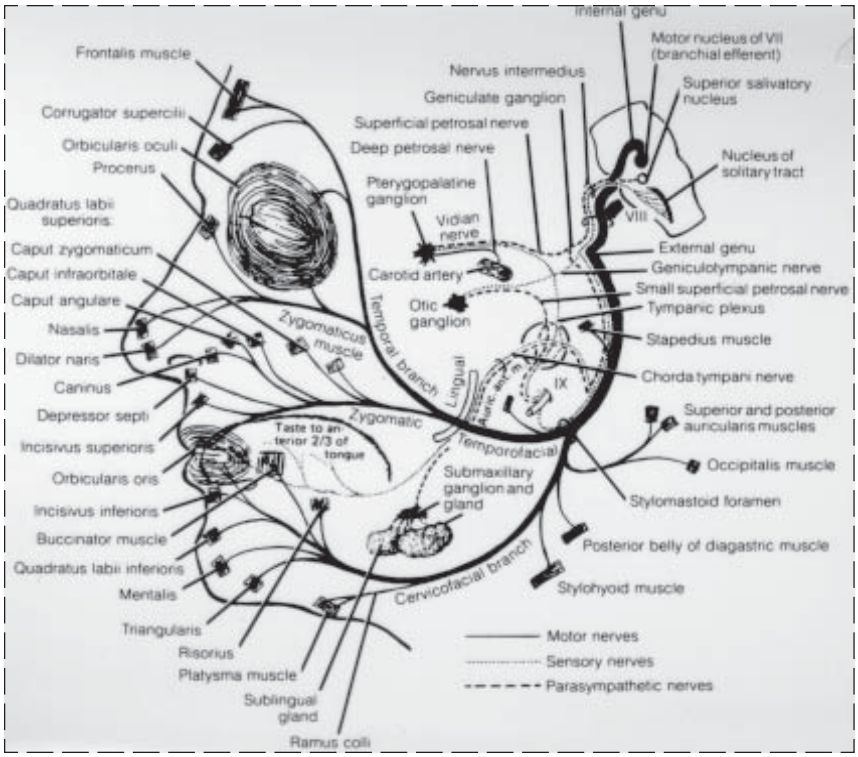

Fig. 7-24. The facial nerve. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy***.** 21st ed. Norwalk, Conn: Appleton & Lange; 1991: 155.

The *meatal segment* (see Figure 7-23, area 1) gives off no major branches, but there is a close relationship with the position of CN VIII. The motor fibers of CN VII are superior and anterior; and the nervus intermedius, coming from the superior salivatory lacrimal nucleus and solitary tract nucleus, is between them.

In the *labyrinthine segment* (see Figure 7-23, areas 2 and 3), the motor division of CN VII and the nervus intermedius enter the fallopian (facial) canal in the petrous bone. They reach the geniculate ganglion from which the greater superficial petrosal nerve originates. This nerve contains preganglionic, parasympathetic efferents that innervate the lacrimal, palatal, and nasal glands via the sphenopalatine ganglion. Sensory cutaneous afferent fibers from the skin of the external auditory meatus, lateral pinna, and mastoid also travel through the greater superficial petrosal nerve (somatic afferents).

The *horizontal segment* (see Figure 7-23, area 3) distal to the geniculate ganglion) gives off no major branch. The *mastoid segment* (see Figure 7-23, areas 4 and 5) gives off the nerve to the stapedius muscle at the upper end of the segment. Further down, the chorda tympani branches off and joins the lingual nerve. The chorda tympani contains preganglionic parasympathetic fibers originating in the superior salivatory nucleus. The parasympathetic fibers innervate the submandibular and sublingual glands via the submaxillary ganglia. Afferent taste fibers from the anterior two thirds of the tongue also travel in the chorda tympani to reach the nucleus of the solitary tract.

The facial nerve fibers exit at the stylomastoid foramen (see Figure 7-24). The branches given off are the posterior auricular nerve, the digastric nerve (to posterior belly of digastric muscle), and the stylohyoid nerve. The facial nerve pierces the parotid gland and divides into temporofacial and cervicofacial branches. Its terminal branches are the temporal, zygomatic, and mandibular nerves supplying the facial mimetic muscles and the platysma.

The visceral efferent (parasympathetic) fibers of the nervus intermedius of Wrisberg supply all the major glands of the head except the parotid gland and glands of the skin. From the superior salivatory lacrimal nucleus, preganglionic fibers divide in the facial canal into the greater petrosal nerve and the chorda tympani. In the pterygoid canal, the greater petrosal nerve is joined by the deep petrosal nerve

(CN V_2). Together they reach the pterygopalatine (sphenopalatine) ganglion. Postganglionic fibers from the sphenopalatine ganglion supply the lacrimal, palatal, and nasal glands. The chorda tympani joins the lingual nerve (CN V_3) to reach the submaxillary ganglion. Postganglionic fibers subserve the submandibular and sublingual glands.

The visceral afferent primary neurons are located in the geniculate ganglion. The fibers of these neurons carry taste sensation (gustatory) from the anterior two thirds of the tongue. These gustatory afferents then travel in the nervus intermedius and end in the tractus solitarius and its nucleus in the medulla. Taste information is received from fungiform papillae on the anterior two thirds of the tongue and is transmitted via the chorda tympani (CN VII[114]; Figure 7-25). Taste information from the circumvallate papillae at the junction of the posterior third and anterior two thirds of the tongue, and from the foliate papillae at the rear edge of the tongue, is mediated by the lingual branches of CN IX. Fibers of both cranial nerves terminate centrally in the gustatory nucleus. Taste buds on the laryngeal surface of the epiglottis transmit impulses through the superior laryngeal nerve, a branch of CN X.

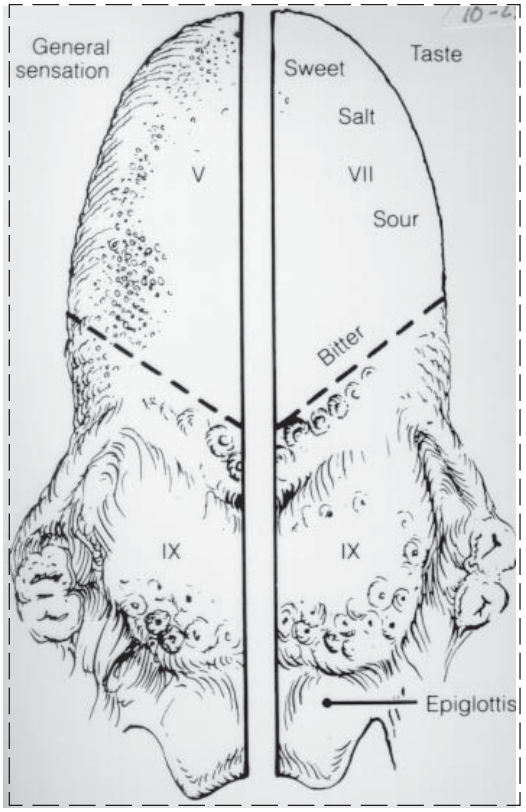

Fig. 7-25. Sensory innervation of the tongue. Roman numerals represent the cranial nerves innervating that area. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 169.

The palatal taste buds located on the margin between the hard and soft palates transmit taste through the greater superficial prostrosal nerve, which lies deep to the semilunar ganglion and traverses the petrous portion of the temporal bone. The nerve exits the middle cranial fossa at the facial hiatus and joins the anterior margin of the geniculate ganglion. From that point fibers run in the nervus intermedius. Taste projections from the medulla terminate in the ventroposteromedial nucleus of the thalamus. Whether the projections are ipsilateral or contralateral is not known, for certain.

The somatic afferent primary neurons are located in the geniculate ganglion in the petrous temporal bone. They receive afferent fibers from the mucosa of the pharynx, nose, and palate, and from the skin of the external auditory meatus, lateral pinna, and mastoid. The sensory fibers enter the brainstem via the nervous intermedius, then descend in the spinal tract of the trigeminal nerve to synapse in the trigeminal spinal nucleus.

Proprioceptive fibers from the facial muscles travel in CN VII. They terminate in the mesencephalic nucleus of CN V.

Central connections for control of innervation of facial movements have fibers arising in the lower third of the precentral gyrus. The fibers course downward through the genu of the internal capsule (as the corticopontine tract) to the base of the peduncle, and terminate in the facial nucleus. The ventral part of the facial nucleus innervates the lower two thirds of the face. For the most part it has crossed supranuclear control. The dorsal portion supplies the upper third of the face and is controlled by bilateral supranuclear input. It is believed that the upper facial motor neurons controlling upper facial movements receive little direct cortical input, while the lower facial motor neurons have significant cortical innervation. Therefore, in supranuclear lesions there is sparing of the upper face.[115]

Ipsilateral innervation of the facial nucleus is also received from the extrapyramidal system originating cortically, that is, from areas anterior to the central sulcus and from areas behind the sulcus. From there, fibers proceed to the basal ganglia (mainly the putamen), then to the brainstem tegmental nuclei, and then to the facial nucleus. As the integrating center of the autonomic nervous system, the

hypothalamus has much influence on the parasympathetic superior salivatory nucleus. Impulses from the limbic system (emotional behavior) and the olfactory area reach the lacrimal nucleus via the dorsal longitudinal fasciculus. Visceral reflexes, such as salivation in response to smell, are mediated through these pathways.

With respect to motor innervation, the pathways for emotional movements may be different from those for voluntary movements. It appears that emotional pathways do not descend through the internal capsule. It was reported by Borod and colleagues[116] that the right hemisphere may be especially significant for emotional expressions of the face.

Sensory connections from the facial nucleus are to the thalamus and postcentral gyrus. The gustatory center is believed to be in the parietal sensory cortex. There is stimulation of the lacrimal nucleus by the gustatory nucleus to produce secretion from oral glands.

Taste information is received from fungiform papillae on the anterior two thirds of the tongue and is transmitted via the chorda tympany (CN VII). Taste information from the circumvallate papillae at the junction of the posterior third and anterior two thirds of the tongue and the foliate papillae at the rear edge of the tongue are mediated by the lingual branches of CN IX. Fibers also terminate in the gustatory nucleus. Taste buds on the laryngeal surface of the epiglottis transmit impulses through the superior laryngeal nerve, a branch of CN X. The palatal taste buds located on the margin between the hard and soft palate transmit taste throughout the greater superficial petrosal nerve, which lies deep to the semilunar ganglion and traverses the petrous portion of the temporal bone. It exits the middle cranial fossa at the facial hiatus and joins the anterior margin of the geniculater ganglion. From there on, fibers run in the nervus intermedius. Taste projections from the medulla terminate in the ventroposteromedial nucleus of the thalamus. Whether the projections are ipsi- or contralateral is not well known.

Injuries and Lesions

Taste awareness is present at birth and is not significantly reduced by aging. There are genetic differences in abilities to taste certain bitter or sweet substances. Taste may be lost partially (hypogeusia) or totally (ageusia), and there can occur taste phantoms (dysgeusia). The taste system functions by a balance of excitatory and inhibitory neuromessages. Loss of taste on one side of the tongue may cause hypersensitivity to taste on the other side. For this reason, total loss of taste due to unilateral nerve injury is rare.

The most common cause of injury or lesion of CN VII is trauma such as head injury, which very commonly affects CN VII.[114] Gustatory fibers may be involved. Central lesions may be bilateral,[117–119] but peripheral nerve damage is mostly unilateral. The incidence of involvement in head injuries is 0.5%, but 5% of patients with posttraumatic anosmia also have some deficit in taste sensation.[6]

An incidence of 10% to 15% facial paralysis was reported for patients with longitudinal fractures of the temporal bone, and 30% to 50% was reported for those with transverse fractures. The lesion is most often located at the geniculate ganglion. Fisch[120] reported that for 50% of the fracture patients facial palsy was due to an intraneural hematoma, 26% showed evidence of transection, and 17% bone impingement. Injuries to the ear or to the side of the face (mandible, parotid gland) also frequently result in peripheral facial nerve injury.

Nontraumatic lesions such as Bell's palsy are quite common. The incidence is clustered. Thirty percent of patients with Bell's palsy have a close family member with a history of Bell's palsy. Other nontraumatic lesions are neoplasms (especially cerebellopontine angle tumor), infections, cerebrovascular accident, and idiopathic. Bilateral Bell's palsy is suggestive of Lyme's disease.

Syndromes involving CN VII are mostly due to tumors, multiple sclerosis, infarction, poliomyelitis, or congenital abnormalities. Syndromes include the following:

- Millard-Gubler syndrome is caused by a lesion in the ventral pons. It presents with ipsilateral peripheral facial palsy, ipsilateral rectus palsy, and contralateral hemiplegia.
- Foville's syndrome is caused by a lesion in the pontine tegmentum and manifested by ipsilateral peripheral facial palsy, paralysis of conjugate gaze to the side of the lesion, and contralateral hemiplegia.
- Möbius' syndrome is congenital absence of facial muscles or nerves. It presents with facial diplegia, abducens palsy, and sometimes deafness.
- Ramsay-Hunt syndrome is caused by an injury to the geniculate ganglion or infection by herpes zoster. Symptoms and signs are ipsilateral facial paralysis, pain in the eardrum region, hyperacusis due to lack

of the stapedius muscle function that dampens sound, and loss of taste. In herpes there will be herpetic vesicles on the eardrum, external auditory meatus, and palate, in addition to the geniculate neuralgia.
- Bilateral facial palsies may be congenital, or due to infections such as *Mycoplasma pulmonalis*, infectious mononucleosis, or Lyme's disease; granuloma (sardoidosis); or Guillain-Barré syndrome. Other possible causes are allergic vascular disease, trauma, or birth injuries (forceps).
- Gustatory hyperhidrosis syndrome (auriculotemporal syndrome) is common after parotiditis, trauma, or surgery. It manifests as hemifacial redness and sweating during eating.
- Chorda tympani syndrome has the same expression as gustatory hyperhidrosis but spreads to involve the neck, back, and chest. Possible misdirected regeneration of nerve fibers is the cause.
- Crocodile tears are paroxysmal lacrimation during gustatory stimulation. Fibers that normally reach the submaxillary gland are traumatized and during regeneration are misdirected to the lacrimal gland.

The symptoms or signs of CN VII lesions are mainly mimetic facial muscle weakness or paralysis. Depending on the location, different afferent fibers may also be affected. Possible dysfunctions are

- Dysfunction of lacrimation.
- Hearing impairment (hyperacusis) due to loss of stapedius muscle function. Normally, strong acoustic stimuli cause stapedius muscle contraction that pulls the stapes out of the round window and so attenuates the loud sound. If this reflex is impaired, hyperacusis results.
- Loss of salivation.
- Loss of taste from the anterior two thirds of the tongue. From 10%[121] to 52%[122] of patients with Bell's palsy have gustatory problems.
- Minor sensory deficit at the external ear.
- Retroauricular pain (somatic pain fibers from the external ear canal and the skin between mastoid and pinna).
- Dysarthria, pooling of saliva, and excessive tearing. Food may collect between gum and cheek due to buccinator weakness.
- The jaw may deviate to the sound side when the mouth is opened wide because CN VII innervates the posterior belly of the digastric muscle.[123] In contrast, the jaw deviates to the involved side in CN V lesions, due to weakness of the pterygoid muscles.
- Mimetic muscle weakness.

In upper motor neuron lesions there may be weakness of lower facial muscles of expression with intact automatic or emotional movements. This may be possible through bilateral subcortical innervation from deep gray nuclei.[124] Abnormal facial movements may be classified as oral facial dyskinesis. Such movements include facial grimacing, twitching, puckering, lip smacking, protrusion of the tongue, or oromandibular dystonia. The cause may be extrapyramidal disease or it may be idiopathic. Other abnormal movements are hemifacial spasm or postparalytic spasm and synkinetic movements, mostly due to aberrant reinnervation or ephaptic transmission. Symptoms and signs according to the location of the lesion are reviewed in Table 7-3 and Figure 7-23.

Evaluation

There are several clinical tests available to localize a facial nerve lesion.[125] The *Schirmer lacrimation test* measures the production of tears and saliva. Filter papers 5.0 x 0.5 cm are bent on one end and inserted into each lower lid to collect tears. Normally the moisture migrates approximately 3.0 cm in 5 minutes.

The *stapedius muscle reflex* is examined when the patient complains of hyperacusis, especially for low tones (this suggests stapedius muscle weakness). A stethoscope is placed into the patient's ear with a gentle vibrating tuning fork on its bell. Normally the sound is heard equally in both ears. If stapedius reflex is absent the sound will be lateralized to the involved side.

The *salivary flow meter* measures production of saliva. *Gustometry* examines the sense of taste. A cotton swab is moistened with a dilute solution of sugar (4% glucose); salt (2.5% sodium chloride); vinegar; 1% citric acid solution; or 0.075% quinine hydrochloride. The solution is applied to the tongue outside, not inside, the mouth, separately to each side. Patients should drink water between stimuli to remove any traces of the previous application. The anterior two thirds of the tongue is tested.

Motor testing of the mimic facial musculature is done by clinical observation of possible asymme-

TABLE 7-3

SYMPTOMS, SIGNS, AND CAUSES ACCORDING TO LOCATION OF FACIAL NERVE LESION

	Symptoms and Signs	Possible Causes
Supranuclear	Paralysis of contralateral limbs and lower face No reaction of degeneration Taste and salivation not affected	CVA, head injury, trauma
Nuclear	May present with paralysis of conjugate gaze to ipsilateral side and contralateral hemiplegia Involvement of other cranial nerves (V, VI) Deafness (CN VIII) Impairment of lacrimations and salivation (nervus intermedius) Hyperacusis Mimic facial muscle paralysis with abnormal EMG	Pontine lesion, trauma, cerebello-pontine angle tumor
Meatal Segment	Impairment of lacrimation and salivation (nervus intermedius) Hyperacusis Loss of taste, anterior two thirds of tongue Mimic facial muscle paralysis with abnormal EMG	Fracture of petrous temporal bone; or pressure transition between meatal and labyrinthine segment may cause facial nerve compromise
Labyrinthine and Horizontal Segments	Often tympanic membrane pain Hyperacusis Loss of taste, anterior two thirds of tongue Mimic facial muscle paralysis with abnormal EMG	Trauma, mastoiditis, neoplasm (usually benign), herpes zoster
Suprachordal Mastoid Segment	Loss of taste, anterior two thirds of tongue Mimic facial muscle paralysis with abnormal EMG	Trauma, middle ear infections, measles, mumps, chicken pox, sarcoiditis
Infrachordal Mastoid Segment	Mimic facial muscle paralysis with abnormal EMG	Tissue swelling within canal of undetermined origin

CVA: cerebrovascular accident
CN: cranial nerve
EMG: electromyogram
Localization my not always be as precise as mentioned above. A general rule is that lesions proximal to or at the geniculate ganglion produce taste, lacrimation, and stapedial reflex abnormalities; lesions distal to the ganglion cause only muscle weakness.[1]
(1) Bartoshuk LM, Kveton JF, Karrer T. Taste. In: Bailey BJ, ed. *Head and neck surgery. Otolaryngology*. Philadelphia, Pa: JB Lippincott; 1993: 520–529.

try of the face during smiling, speaking, raising eyebrows, wrinkling forehead, and resistance given to grimacing. Upper motor neuron lesions spare the forehead and mostly the eyes. Lower motor neuron lesions may involve only the lower face if the lesion is restricted to the caudal portion of the nucleus, as may happen in polio.

Observation of any *reflex phenomenon* is important. Reflex eye closure is observed in response to threatening movements of a hand in front of the patient's face (menace reflex). A glabellar tap over the bridge of the nose elicits reflex blinking. Care is taken to hold the hand over the top of the head to avoid eliciting a visual blink reflex. After about three taps, blinking ceases. In patients with Parkinson's disease, blinking continues.

Testing of the *auditory palpebral reflex* is accomplished by observing the reflex bilateral eye closure secondary to presentation of a sudden loud noise. It is important to look for the possible Elve's phenomenon, which shows upward turning of the eyeball under the closed lid. In facial palsy the lid may not be fully closed and the turning of the eyeball will be visible. Facial muscles are observed also in

response to emotions. Occasionally in lower facial palsy there is a dissociation of volitional and emotional mimetic muscle function.[124]

Electrodiagnosis

Electrodiagnostic studies include electrogustometry, gustatory evoked potentials, nerve excitability studies, nerve conduction evaluation, reflex studies, and EMG.

Electrogustometry is a test of an anodal current threshold. The patient experiences a sour metallic taste. This test investigates loss or decrease of "sour" taste only (ageusia), not loss of other tastes, and not the distortion of taste (dysgeusia). These are better evaluated with clinical chemical taste testing.[126]

Gustatory evoked potentials[127] can be performed but, at the present time, these tests are mainly performed for purposes of research. The procedure is explained below.

Stimulation is achieved when an electrical stimulus or certain volatile tastants are presented to the taste buds. Because of habituation, presentation must be at random, with at least 5 seconds between each stimulus.

Recording electrodes are placed over C_7 and A_1 (International 10-20, as seen in Figures 7-5, 7-6, 7-7).

Normal values are a response in approximately 300–400 millisecond.

Nerve excitability studies have been performed often for early detection testing for threshold stimulation. Even after complete facial nerve section, the distal excitability remains normal for up to 4 days. There will no longer be any response at the end of the first week. If excitability remains, prognosis is good. The facial nerve is stimulated at the stylomastoid foramen with an electrical current of 1 millisecond duration. A minimal amount of current is applied to cause a minimal twitch in a facial muscle. The normal threshold is between 3 and 8 mA. There should be less than 2 mA difference between the two sides of the face.

Nerve conduction studies are the most commonly employed electrodiagnostic evaluations of the facial nerve. They are sensitive tests for evaluating axonal loss when the response amplitudes from side to side are compared. Some investigators recommend retesting every other day for the first 2 weeks for best prognostication.[128] If the evoked motor amplitude on the involved side is smaller than 10% of that on the good side, the prognosis for recovery is poor. The procedure for direct facial nerve conduction[129] is given below.

Procedure of Direct Facial Nerve Conduction Studies

Stimulation is carried out at the stylomastoid foramen.

Recording electrodes are placed in or over facial muscles. Preferred location is over the nasalis muscle. The lower orbicularis oculi muscle may be tested also when studying the blink reflex. The orbicularis oris, mentalis, or frontalis muscles may also be used for recording.

Normal values:

- Latency: 3.2 ± 0.34 millisecond (range, 2.2 to 4.0 ms)
- Amplitude: 2.0 ± 0.9 mV (range, 1 to 5 mV measured baseline to peak)
- Duration: 12.8 ± 2.9 millisecond (range, 6.2 to 22 ms)

Reflex studies can be utilized for indirect facial nerve conduction, especially to evaluate the proximal conduction velocity. This can be estimated by eliciting the blink reflex[106] (see trigeminal nerve). The clinical value of the blink reflex in Bell's palsy is used mainly for prognostication. If R1 has been absent, its return suggests a reasonable prognosis, even though the R1 may be delayed during the first 4 weeks. Delay indicates demyelination rather than physiological (functional) nerve block. The R1-D ratio (R1 vs direct facial response) is helpful to delineate whether a lesion is more proximal or distal. For example, the R-D ratio is slightly increased in Guillain-Barré Syndrome and multiple sclerosis, but decreased in Charcot-Marie-Tooth disease and diabetic polyneuropathy.

EMG is carried out in the usual fashion. A needle recording electrode is inserted into any one of the mimetic muscles. Evaluation is by observation of activity at rest, motor unit parameters during minor activation, and especially recruitment. The most commonly tested muscles are the orbicularis oris and oculi. The number of muscles having voluntary action potentials preserved for the first week provide an additional index for prognostication. Muscle over activity can present as:

- Facial myokymia, where motor units of normal shape and duration appear in a single, double, or group discharge about every 100 -to 200 millisecond, firing asynchronously. Continuous and discontinuous, but definitely rhythmic, patterns can be observed. Causes may be fatigue, mul-

tiple sclerosis, brainstem tumors, or injury. Myokymia is not affected by sleep or facial movements.
- Synkinesis, in which there is simultaneous motor unit activity in different facial muscles. Simultaneous activation of muscles innervated by different nerve trunks is also observed when a stimulus is given to one branch, or during a blink reflex study. This phenomenon is associated with regeneration or degeneration of the facial nerves (aberrant reinnervation or ephaptic transmission).
- Hemifacial spasm, as manifested by high frequency (150 to 400 Hz) rhythmic discharges with synchronization. The most common cause is compression of CN VII by an aberrant arterial loop. Therefore, for this lesion, decompression may be of help.[130] Hemifacial spasm is not of cortical origin and cannot be abolished by sleep or cerebral infarction. The most likely cause is ephaptic transmission.

The prognosis for recovery of facial nerve injuries as well as upper motor neuron facial palsy is in general good, without intervention.[131–135] Within 2 to 6 months, 60% to 70% of all Bell's palsy patients recover fully or have only minimal residual deficit. Thirty percent to 40% have slow or defective motor recovery after more than 6 months.

In a patient with incomplete facial palsy, if the evoked motor amplitude of an affected facial muscle is 10% or more of the amplitude of the unaffected side in the first 14 days after onset, the prognosis is good. Ninety percent of these patients will recover satisfactorily. By contrast, patients who show clinically complete palsy and an evoked response smaller than 10% within the first 14 days have an 80% chance of poor recovery.[128] Other criteria for possibly good prognosis reported in the literature are the presence of voluntary action potentials on EMG registered in two or more muscles on or after the third day, and return of the previously absent R_1 (or R_2) of the blink reflex.[131]

Management

Most clinicians promote early administration of prednisone, 1 mg/kg of body weight daily in two divided doses for 4 days, then a quick taper to a total of 5 mg/d within 10 days.[136] Dexamethasone, 10 mg intravenously every 12 hours, is also recommended, especially for facial palsy after head trauma.[137] This regimen may reduce edema and therefore compression of the facial nerve in the bony canal and may reduce synkinetic movements. If pain is present, steroids relieve symptoms promptly. If severe facial nerve paralysis persists, surgical intervention may be indicated. Hypoglossal facial nerve anastomosis may restore tone and function. A study[138] of 22 cases (out of a study sample of 245 surgical cases) of complete facial nerve palsy that occurred during cerebellopontine angle tumor removal reported good results in 63.6%, fair in 13.6%, and poor in 18.2%. One case (4.5%) was a failure, following hypoglossal-facial nerve anastomosis.

Electrical motor point or muscle stimulation to retard atrophy and promote early regain of function has been advocated and widely used. However, the effectiveness of this mode of treatment has not been proved scientifically and is not clinically convincing.

To prevent complication from possible corneal irritation secondary to incomplete eye closure, administration of artificial tears during the day and ophthalmic ointment at night, such as Lacrilube, are recommended. A protective plastic wrap taped around the eye may be helpful.

CRANIAL NERVE VIII: ACOUSTIC

The acoustic nerve is a composite sensory nerve with two separate parts, the cochlear (auditory) nerve and the vestibular nerve. The anatomy is shown in Figures 7-26 and 7-27.

Anatomy and Function

The cochlear nerve

The cochlear nerve is a special sensory nerve. The primary neurons are bipolar cells in the spiral ganglion of the cochlear nerve. The peripheral branches of the cells end in auditory receptors (hair cells) located in the cochlear duct. The entire sensory structure is called the organ of corti. Hair cells in the cochlear apex are stimulated by low frequencies, and basilar hair cells respond to high frequencies. The mechanical deformation of the hair cells is transduced into electrical signals. The central branches of the bipolar cells terminate in the ventral (low frequency) and dorsal (high frequency) cochlear nuclei (see Figure 7-27).

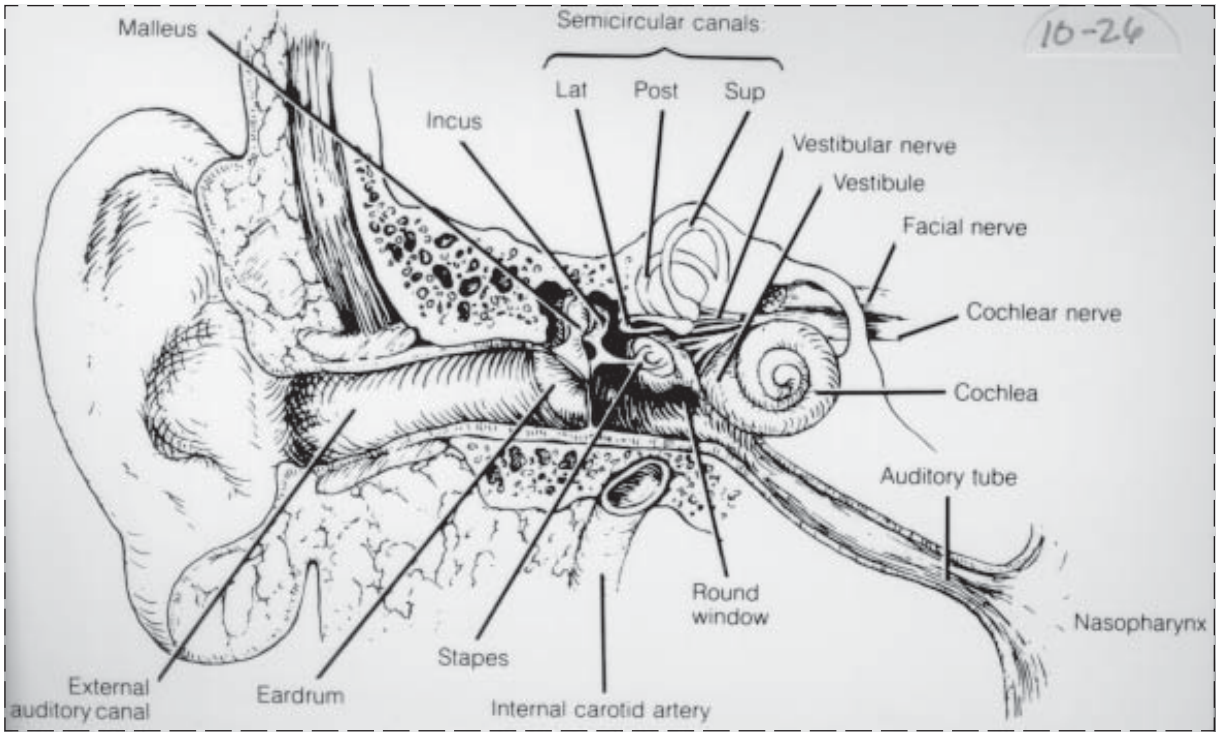

Fig. 7-26. The human ear. Middle ear muscles omitted. Reprinted with permission from Ganong WF. *Review of Medical Physiology.* 16th ed. Norwalk, Conn: Appleton & Lange; 1993; 157.

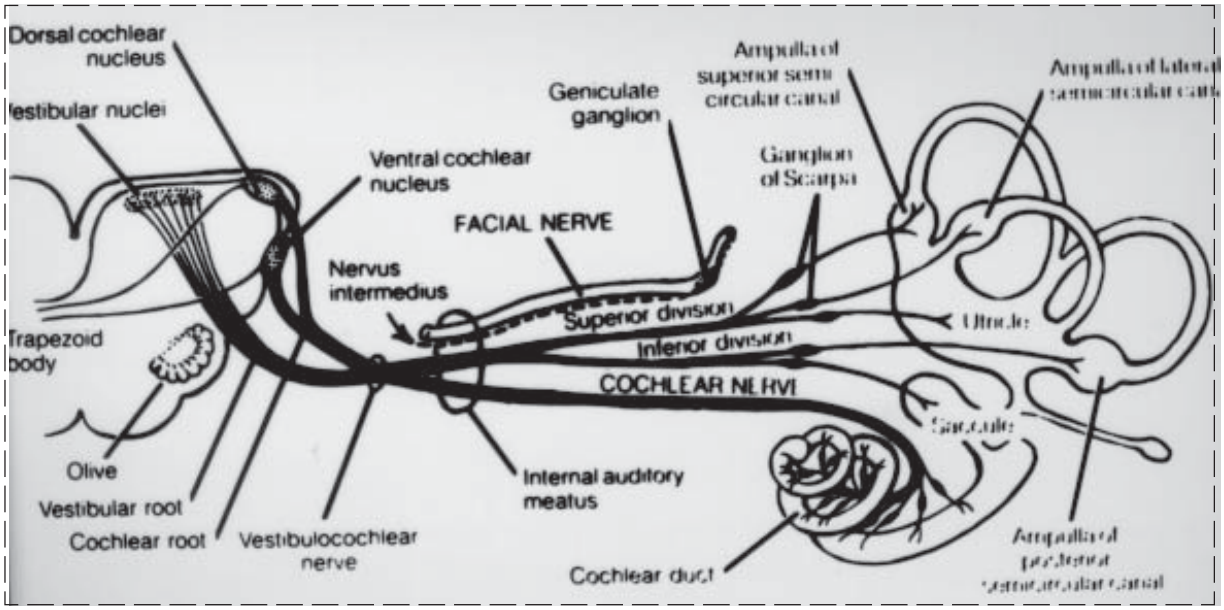

Fig. 7-27. Vestibulocochlear nerve. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 157.

The secondary neurons are in the dorsal, anteroventral, and posteroventral cochlear nuclei. Innervation is tonotopically organized; the dorsal nuclei receive high frequency stimuli and the ventral aspects receive fibers conveying low frequency signals. From the dorsal and ventral cochlear nuclei nerve impulses are projected to the contralateral brainstem. Here they ascend in fibers of the lateral lemniscus to terminate in the inferior colliculus.

The tertiary neurons are in the inferior colliculus (midbrain tectum). Again, the fibers are located in tonotopic organization. From the inferior colliculus, fibers terminate in the medial geniculate body (thalamus). Fibers responding to low frequencies end in the apical lateral areas, and those responding to high frequencies end in the medial portions.

The central connections arise from the medial geniculate body. Auditory radiations are projected to the auditory cortex (area 41) located in the contralateral temporal gyri, and others terminate in the auditory association cortex (area 42). Reflex connections pass to the eye muscle nuclei and other motor nuclei of the cranial and spinal nerves via the tectobulbar and tectospinal tracts. Some fibers project bilaterally to the facial nucleus for reflex contractions of the stapedius muscle to dampen loud sounds.

There are also some inhibitory efferent axons with cell bodies in the superior olivary complex in the brainstem. A feedback loop function to suppress unwanted auditory signals is suggested.[139]

The vestibular nerve

The vestibular nerve also carries special sensory fibers. The primary neurons are in bipolar cells in the vestibular ganglion (ganglion of scarpa; see Figure 7-27). Peripheral fibers pass to the neuroepithelium in the ampullae of the semicircular canals and in the maculae of the utricle and saccule. The sensory receptors in the maculae are ciliated hair cells covered by gelatinous material. Otoliths (calcium carbonate crystals) are located in the gel and stimulate the hair cells in response to movement. Central branches enter the brainstem medial to the restiform body and end in the vestibular nuclei.

The vestibular system is concerned with maintaining equilibrium. The utricle, saccule, and semicircular canals make up the labyrinth. The main function of the utricle and the saccule is to detect the position of the head relative to gravity. Special receptors (hair cells) in the maculae of the utricle and saccule monitor linear acceleration, while angular acceleration is monitored by the cristae in the ampullae of the semicircular canals. Horizontal head movements stimulate the utricle, whereas tilting of the head activates the saccule. The superior portion of the vestibular nerve conducts impulses from the anterior and horizontal semicircular canals and from the utricle. The inferior portion of the nerve transmits information from the posterior semicircular canal and the saccule. The vestibular nuclei exert an influence on conjugate eye movements and on head and neck movements through the medial longitudinal fasciculus.

Pathways from the lateral vestibular nucleus project to the ipsilateral spinal cord. These fibers facilitate extensor trunk tone and the action of antigravity axial muscles.

Central connections of the vestibular system are mainly projections to the cerebellum. However, the medial longitudinal fasciculus projects ipsilaterally from the superior vestibular nucleus and contralaterally from the medial vestibular nucleus to the eye muscle nuclei. The cortical representation of the vestibular system is located in the postcentral gyrus (near areas 2 and 5).

Injuries and Lesions

The most common causes of injury and lesions are traumatic injuries, especially head trauma. It is reported[140] that the sensory organ most commonly injured is the ear. Blunt trauma causes inner ear concussion with symptoms of high frequency sensorineural hearing loss, transient positional nystagmus, and vertigo.[141] If the temporal bone is fractured, significant injury to the auditory and vestibular systems results. Eighty percent to 90% of patients with longitudinal fractures of the petrous portion of the temporal bone experience conductive hearing loss due to disruption of the ossicular chain in the middle ear, often due to dislocation of the incus. Ten percent to 20% of fractures of the temporal bone are transverse, causing sensorineural hearing loss and often involvement of the facial nerve. The vestibular system is frequently involved, and as many as 65% of traumatic brain injury patients are reported to demonstrate symptoms of vestibular dysfunction some time during their recovery.[142–145]

Nontraumatic lesions are mostly caused by tumors. Cerebellopontine angle tumors cause mostly unilateral sensorineural hearing loss with the main deficit in the high frequencies. The most important lesion to rule in or out with CN VIII testing is a cerebellopontine angle tumor, which is most often a vestibular schwannoma. The types of tumors ob-

served are mainly acoustic neuromas (mostly originating in the vestibular portion of CN VIII), schwannoma, meningioma, cholesteatoma, metastatic lesions, and posterior fossa lesions (possibly cerebellar junction hematoma). Other causes of CN VIII lesions are cranial neuropathy in association with systemic disorders; arteriovenous malformations; congenital and familial disorders; demyelinating diseases (multiple sclerosis); drug intoxications; excessive noise exposure; presbycusis (hearing loss secondary to aging); atherosclerosis; and viral diseases (mumps). Ménière's disease (fluctuating sensorimotor hearing loss, ear pressure, and tinnitus) may be a delayed consequence of conclusive injury.

Symptoms and Signs

The symptoms and signs observed with cochlear nerve lesions are mainly tinnitus, hearing loss, or both. Tinnitus is seen in up to 32% of adults visiting the audiologist or otologist.[146] Many patients experiencing tinnitus sustained a minor head injury or other injuries or diseases resulting in hearing loss. Tinnitus may precede, follow, or coincide with the onset of hearing loss. It may actually be an early symptom of a tumor in the internal auditory meatus or the cerebellopontine angle.[147] Tinnitus is characterized by humming, whistling, hissing, roaring, or throbbing auditory sensations.

Patients may hear these noises in one or both ears, or localized in the head. The most common cause is cochlear disease, but tinnitus can also result from a lesion in the external ear canal, tympanic membrane, ossicles, cochlea, auditory nerve, brainstem, or cortex. Six million persons in the United States reportedly experience tinnitus, which is often as disabling as hearing loss. Patients so afflicted are often willing to undergo multiple surgical procedures even if hearing in the involved ear may have to be sacrificed.[148] The examiner may be able to hear tinnitus caused by abnormal muscle contractions such as palatal myoclonus, tensor tympany, or stapedial myoclonus. Such mechanical tinnitus is an objective find.

Hearing impairment is the most prevalent chronic physical disability in the United States. Health statistics estimate that by the year 2050 more than 10% of the population will be hearing impaired.[148] The degree of hearing impairment as correlated with the inability to understand speech is shown in Table 7-4. Some of the causes of impairment and the corresponding effects are listed below.

- Conductive hearing loss due to lesions of the sound conducting apparatus of the middle ear affects mostly the lower frequencies.
- Sensorineural hearing loss is due to lesions central to the oval window, and affects mostly the higher frequencies.
- A lesion in the central auditory pathways beyond the cranial nerve causes central auditory dysfunction, but not complete deafness.

TABLE 7-4

AVERAGE HEARING THRESHOLD LEVEL FOR 500, 1,000, AND 2,000 Hz IN THE BETTER EAR

Class	Classification Category of Hearing Loss	More Than (dB)	Not More Than (dB)	Ability to Understand Speech
A	Within normal limits		25	No significant difficulty with speech
B	Slight or mild	26	40	Difficulty only with faint speech
C	Moderate	41	55	Frequent difficulty with normal speech
D	Moderately severe	56	70	Frequent difficulty with loud speech
E	Severe	71	90	Can understand only shouted or amplified speech
F	Profound	91		Usually cannot understand even amplified speech

Adapted with permission from Davis H. Guide for the classification and evaluation of hearing handicapped. *Trans Am Acad Ophthalmol Otolaryngol.* 1965;69:740–751.

- Hearing scotomas may be caused by hysteria, multiple sclerosis, schizophrenia, and at times trauma. This condition reveals deafness only to certain frequencies and noises.
- Auditory hallucinations may occur as an epileptic aura or during drug intoxications or psychoses.
- Hearing impairment may be secondary to lesion of CN V because of its innervation of the tensor tympany.
- Hearing impairment may be secondary to lesion of CN VII, because this nerve innervates the stapedius muscle. It supplies the efferent limb of the stapedial reflex.

Presbycusis may present with sensorineural hearing loss caused by loss of neurons in the central nervous system or by degeneration of the sensory and supportive cells in the cochlea. Presbycusis may also present with conductive hearing loss. This may be due to metabolic changes caused by atrophy of the stria vascularis (the site of endolymph production), or it may be mechanical, resulting from stiffness of the basilar membrane in the cochlea.

Vestibular nerve dysfunction symptoms and signs include dizziness, vertigo, nystagmus, ataxia, and dysequilibrium. The main symptoms and signs are dizziness, vertigo, and nystagmus, in any combination.

Dizziness is described as lightheadedness, faintness, dysequilibrium, or disturbance of consciousness. It is a disturbance in perception of bodily position in space. Dizziness may be confused with vertigo. Common systemic causes are cardiovascular disease, hematologic disorders, hypoglycemia, hypothyroidism, hyperventilation syndrome, ocular disorders, drugs, and psychiatric disorders.

Vertigo is the experience of movement of the self or the environment and is caused by lack of integration of information received by the visual, auditory, and vestibular systems. The basic functions of the vestibular system are spatial orientation at rest and during acceleration, visual fixation during head movement, body movement (vestibuloocular reflex), and feedback control of muscle tone to maintain posture. Vertigo reflects a disturbance in one or more of these systems.

Vertigo can have peripheral causes. Idiopathic (positional) vertigo is usually of short duration and not accompanied by cochlear or neurological symptoms, although nystagmus is frequently present. Matutinal vertigo (vertigo occurring on arising in the morning) may be peripheral or central. Other peripheral causes include peripheral vestibulopathy (extramedullary), vestibular neuronitis, acute labyrinthitis (associated with tinnitus and hearing loss), Ménière's disease (vestibular type), and vertigo secondary to middle ear disease or viral infections.

Posttraumatic vertigo and unsteadiness may be due to injury to the vestibular nuclei and cerebellum.[149] The incidence of vertigo after head trauma has been reported as 24% and 78%.[149–151] Centrally produced vertigo can also result from vascular causes (transient ischemic attacks mainly of the vertebrobasilar artery) and is mostly of prolonged duration. Other central causes are labyrinthine stroke, Wallenberg's syndrome, multiple sclerosis, cere-bellopontine angle tumor (causing hearing loss more frequently than vertigo), and vestibular epilepsy.

Nystagmus is an eye movement disorder consisting of a slow drift, which is quickly corrected by saccades to return to the desired focus. Nystagmus may be induced by central or peripheral vestibular lesions. The eyes drift parallel to the plane in which the diseased canal lies. Nystagmus may be horizontal or vertical.[152] A history of dysequilibrium (ataxia and unsteady gait) may be observed with vestibular nerve injury.

Evaluation

Clinical evaluation of the *cochlear nerve function* employs the following hearing tests[148]:

- The auditory screening test is performed by the examiner whispering into the ear or presenting a ticking watch at a distance of 60 cm, while the opposite ear is masked by gently occluding the external ear or pressing on the tragus. Sound should be perceived by a person of normal hearing.
- Pure tone audiometry (Figure 7-28) measures the hearing sensitivity to selected pure tone frequencies at calibrated sound pressure levels. The audiogram records the results graphically. It easily demonstrates low frequency or high frequency hearing loss and so may suggest the type of hearing deficit.
- The normal intensity of sound pressure reception is between 10 and 25 decibels (dB). The dB is a unit for expressing the difference between a given sound pressure level and a standard reference pressure of 0.0002 dyne/cm^2.

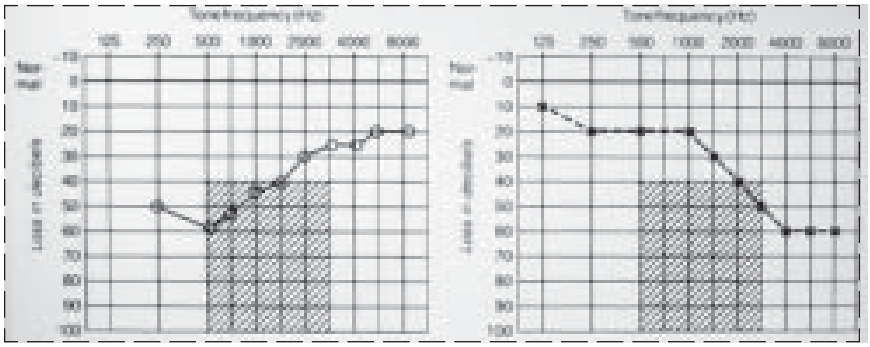

Fig. 7-28. Audiogram. Left: Middle ear (conduction) deafness. Right: perception (nerve) deafness. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 161.

- The unit dB is used not only to describe sound-pressure level (SPL) but also to measure hearing sensitivity. The standard reference for hearing sensitivity is the amount of energy needed for a person of normal hearing to perceive that particular sound (the hearing threshold level—HTL). Threshold is defined as the level at which 50% correct responses are elicited. Although the frequencies perceived by the human ear can range from 20 to 20,000 Hz, the critical area is 500 to 3,000 Hz.
- Tympanometry (acoustic impedance—immittance evaluation) assesses the integrity and performance of the peripheral auditory system. The test uses a probe tip with three holes. One hole is for a probe tone at approximately 220 Hz; one is for cavity air pressure control; and one is for a microphone to record the sound pressure level in the ear canal. The probe tip is sealed into the external auditory meatus. A sound properly adjusted to the mass and stiffness of the system and the acoustical resistance of the ear is introduced, and the reflection from the tympanic membrane is measured.[147]

  Tympanometry provides an extremely sensitive measure of middle ear function. The normal middle ear absorbs and transmits energy easily, causing the tympanic membrane to have a low acoustic impedance. If the tympanic membrane is scarred or fluid is located in the middle ear, impedance is increased (static compliance is decreased).
- The stapedial reflex test may be performed if tympanometry is normal. Monaural stimulation causes bilateral contraction of stapedius muscles. Absent stapedial reflex strongly suggests a retrocochlear CN VIII lesion.
- A speech discrimination or word discrimination test evaluates reception of 50 phonetically balanced words at approximately 30 to 40 dB above the patient's speech reception threshold. Findings offer clues to the suspected severity of the handicap.
- The alternate binaural loudness balance test shows abnormal recruitment, an abnormal growth in the loudness of sound when intensity is increased above threshold levels. Sound is presented to both ears. The intensity to one side is held constant, while the other side is varied until the patient perceives the two as equal. Inequality of delivered sound intensities is identified. Abnormal recruitment suggests hearing loss due to cochlear disease.
- The test for hearing scotomas determines whether all frequencies can be heard. Testing is done with audiometry.
- An examination is made for sensory aphasia, or word deafness. This is an ability to hear but not comprehend words. Sensory aphasia is associated with lesions of the posterior portion of the superior temporal gyrus of the dominant cerebral hemisphere.
- Weber's test is performed by placing a tuning fork against the midline vertex of the scalp. Normally, no lateralization of sound to either ear is reported. If the sound is referred to the ear with hearing impairment, loss of hearing is due to impaired conduction in the external middle ear. If the sound is referred to the unimpaired hearing ear, loss of hearing is attributed to impaired function of the auditory nerve or cochlea.
- Rinne's test compares air and bone conduction. A vibrating tuning fork is placed first on the mastoid bone process, and then held with its tip beside the auricle. A patient with normal hearing or mild nerve deaf-

ness will report that it is louder when near the auricle. A patient with middle ear (conductive) deafness will report the tuning fork to be louder when placed on the mastoid bone. In severe nerve deafness no sound is heard in either position. Low frequency hearing impairment is mostly conductive hearing loss.

- Bine's test is conducted by placing a tuning fork (with pitch C, or 256 vibrations/s) on the vertex with one ear occluded. Normally the closed ear hears sound best by bone conduction. If no sound is heard in the closed ear, nerve deafness is suspected.
- An otoscopic examination examines the external canal to detect or rule out foreign bodies, congenital malformation, abnormal condition of the tympanic membrane, presence of wax, or evidence of infection. If a polyp is observed, care must be taken not to touch it because it may bleed disastrously.

Evaluation of vestibular nerve function includes inquiry about general symptoms such as history of diaphoresis, tachycardia, nausea, vomiting, and low blood pressure, such as may occur with labyrinthine disease. The following tests are performed:

- Nystagmus is observed and described as to type, direction of slow and fast components, and the head position in which it occurs. It may represent a disturbance in the reflex control of the ocular muscles, which is mainly a function of the semicircular canals. Other forms of nystagmus occur in central cerebellar and cerebral diseases.[152]
- In a positional test the patient is required to look to the left and then is assisted to recline quickly so that the head lies over the edge of the couch 45° below horizontal. The patient remains in this position with the eyes turned to the left for 30 seconds. Observation is made for possible nystagmus (peripheral type) and the patient is asked for complaints of vertigo. The patient then sits up with the head turned to the left for half a minute and is continuously observed. After any vertigo or nystagmus has ceased (in about a minute), the procedure is repeated with the head and eyes turned to the other side. Nystagmus and vertigo will normally occur after a latent period of 10 to 20 seconds. No latent period is observed with central lesion or habituation. With fatigue, signs and symptoms lessen after repetition of provocative maneuvers. With peripheral lesions, nystagmus and accompanying vertigo are significantly more severe, and often signs of brainstem involvement are seen.
- During caloric testing, cold water (to 10°C) is instilled into the right ear with the subject seated and head tilted backward 60°. Nausea responses normally occur, and there is horizontal nystagmus with a *slow* component to the right, pass-pointing to the left, and falling to the right; with warm water, the *quick* component is observed to the right. Complete interruption of vestibular nerve function is characterized by absence of the reaction to external irrigation; partial interruption of vestibular nerve function produces a diminished response. In the Hallpike caloric test, approximately 250 cm^3 of water at 30°C is applied to the external auditory canal for 40 seconds. The application is then repeated with water at 44°C. The normal response is nystagmus of 90 to 140 seconds duration.
- The Romberg test, which evaluates the ability to stand with feet together and eyes closed, is performed also with eyes open and the patient is observed for unsteadiness. Unilateral ataxia is almost always an indicator of focal posterior fossa abnormality (infarct, demyelination, abscess, or tumor).
- Posturography[153] is a quantification of the Romberg test. A recently developed dynamic posture platform is used to isolate the vestibular system for the purposes of the test. A movable visual field is provided with a special platform to remove visual and proprioceptive cues that otherwise would assist in maintaining posture. The patient's ability to maintain posture is evaluated. This procedure has not been properly validated, and further research is needed before scores can be adequately integrated.
- The examiner tests for presence of pass-pointing, which is the unidirectional drifting of outstretched fingers. Pass-pointing is a definite indication of tonic imbalance in the vestibular system.
- The examiner should test blood count, electrolytes, glucose, and thyroid function, and

should also check for hypercholesterolemia or increased triglycerides.

Electrodiagnosis

An electrical test can be performed to study the amount of galvanic current (mA) necessary to produce nystagmus, pass-pointing, and inclination of the head when current is passed between two saline pads placed one over each ear. The comparative effect of placing the cathode on the right and then on the left ear is determined.

In addition, electrodiagnostic evaluation includes ENG, electrocochleography, and brainstem auditory evoked potential (BAEP). BAEP is also called brainstem evoked response or auditory brainstem response (ABR).[154,155]

ENG offers an objective means of evaluating the optokinetic reflex, smooth pursuit, and saccades.[79] It can assist also in the diagnosis and interpretation of nystagmus by providing analysis of superficially recorded corneal-retinal potentials during gaze, positional changes, and simultaneous caloric testing. Bitemporal recording records only the sum of the movement of the two eyes. This simplifies the procedure but is unable to detect asymmetries between the two eyes, as may occur in internuclear ophthalmoplegia. A significant result is a difference of greater than 20% to 25% between the average slow component velocity (SCV) resulting from thermal stimulation of one ear compared to the SCV resulting from thermal stimulation of the other ear. This finding is an indication of hypofunction in one peripheral vestibular system (canal paresis).[79] Another benefit of ENG is that it can determine the presence of nystagmus through the eyelid in patients with ptosis or an inability to cooperate. The incidence of spontaneous nystagmus observed with ENG after head injury is reported as 23%.[154]

Electrocochleography measures electrophysiological activity that originates within the cochlea or the auditory nerve. This test studies the compound action potential of the auditory nerve. The recording electrodes must make contact with the promontory of the cochlea at the transtympanic membrane.

During BAEP studies, five main distinct wave forms representing different areas of the auditory pathways have been documented (Figure 7-29).[155] These important wave forms are believed to represent the following origins:

- wave I, the auditory nerve;
- wave II, the CN VIII or cochlear nuclei;
- wave III, the superior olive;
- wave IV, the lateral lemniscus; and
- wave V, the inferior colliculus.

It must be remembered that exact localization of the origin of these wave forms has not been determined at this time.

Cochlear nerve involvement in concussive states after closed head trauma may be objectified by BAEPs. Abnormal BAEPs were reported in 27.3%[156] of patients in postconcussive states, as manifested by unilateral or bilateral increase of interpeak latencies. These changes are often reversible.[156,157] Some conditions in which BAEPs are useful are listed below.

- In multiple sclerosis the most common abnormalities are an increase in waves I–V interpeak latencies and a reduction of amplitude of wave V.
- Acoustic neuroma may be associated with the absence of all waves, or the interpeak latencies of early waves I–III may be increased.

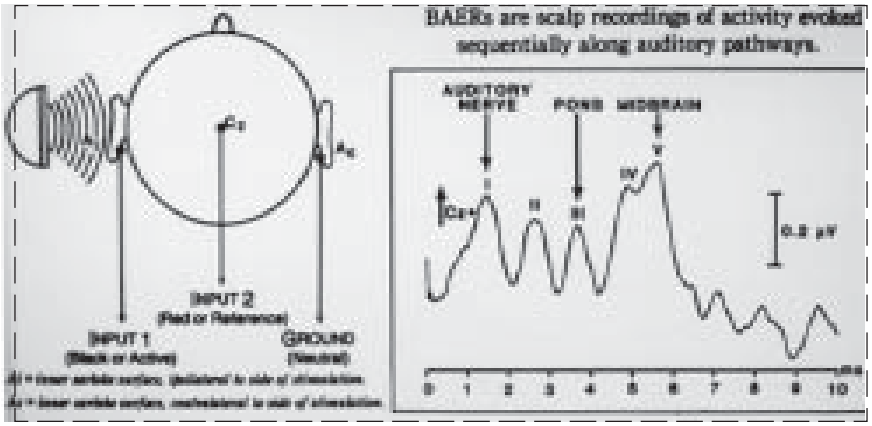

Fig. 7-29. Brain stem auditory evoked potential. Electrode placement and evoked response (schematic). Reprinted with permission from TECA Corp. *Brainstem Auditory Evoked Response (BAER).* In: TECA Applications Bulletin, No. 1001. Pleasantville, NY: TECA Corp: 1981.

- Brainstem tumors may show the absence of all waves after wave III. Multilevel lesions may abolish all waves except wave I.
- BAEPs may be used for intraoperative monitoring during brainstem surgical procedures.[14–16]
- In Sudden Infant Death Syndrome (SIDS), BAEPs may possibly be useful for detection of susceptibility.
- BAEPs can be used to assess brain death and coma, along with clinical and EEG assessment.

Procedure of BAEP Studies

Stimulations are monaural clicks presented to the ear under examination. Click polarity may be rarefaction or condensation. Duration of stimulus is 100 microsecond and the rate is 10/s. Intensity should be 60 to 80 dB above the patient's hearing threshold. The contralateral ear is masked by white noise of at least 40 dB above the patient's hearing threshold to prevent the stimulus from affecting the contralateral ear.

Recording electrodes are placed on the ear lobe and a reference electrode is placed on position C_Z (International 10–20, see Figure 7-7). Parameters and procedures are (*a*) sensitivity, 2.5 μV/division; (*b*) sweep speed, 10 ms/division; (*c*) filters: low frequency cut-off 100 to 200 Hz, high frequency cut-off 2 to 3 kHz; and (*d*) averaging number of responses is 1,000 to 4,000. Measurements taken are absolute peak latencies, interpeak latencies, peak-to-peak amplitudes (amplitudes are measured from peak to the following trough), relative peak-to-peak amplitudes, and inter-ear variations.

Normal values, with 10/s stimulation

- Latency to peak of wave, in milliseconds:
  wave I 1.7 ± 0.15
  wave II 2.8 ± 0.17
  wave III 3.9 ± 0.19
  wave IV 5.1 ± 0.24
  wave V 5.7 ± 0.25
  wave VI 7.3 ± 0.29
- Interwave latencies (interpeak), in milliseconds:
  I–III 2.1 ± 0.15
  I–V 4.0 ± 0.23
  III–V 1.9 ± 0.18
- Amplitudes peak to peak, in μV:
  wave I 0.28 ± 0.15
  wave III 0.23 ± 0.14
  wave IV 0.40 ± 0.13
  wave IV/V 0.47 ± 0.16 (highest peak)
  wave V 0.43 ± 0.16

The most significant abnormality is interwave separations (I–III or I–V), which are seen with cerebellopontine angle tumors.

The prognosis for improvement in auditory function is significantly greater for conductive than for sensorineural hearing loss. Patients with different types of presbycusis have varying degrees of success from the use of hearing aids. Patients with metabolic presbycusis have a significantly better prognosis than those with neural, sensory, or mechanical forms of presbycusis. The fact that hearing impairment is the most prevalent chronic physical disability certainly suggests limited possibilities of recovery.[148]

Recovery for patients with central vestibular damage is less complete than for patients with peripheral lesions. A study[143] of 321 patients with mild or moderate head injuries showed that 60% to 70% of patients with central vestibular dysfunction had persisting symptoms 5 years after the injury. Improvement in vestibular function is sometimes due to habituation of appropriate oculovestibular reflexes.

Prognosis for the recovery of CN VIII deficit secondary to ototoxic drugs is markedly improved if patients seek a physician's help at the earliest sign of tinnitus. Discontinuing or modifying the ototoxic medication may often preserve hearing.

Management

Cochlear Dysfunction

Tinnitus can be very disabling, as is often seen in patients who have had excessive noise exposure, head injury, or ototoxic medication. Medical management employs vasodilators, large doses of vitamin A, xylocaine, and carbamazepine. Biofeedback for tinnitus may be effective in some cases. Tinnitus maskers can be attached to the ear and a band of white noise introduced. The patient controls the intensity of the masking sound, which most likely occurs at the frequency of 2,000 Hz or higher. Another approach is external electrical stimulation. Electrodes are placed over the mastoid bone and different frequencies (which generate different size waves) are applied for progressively longer periods. Results have not been fully studied yet.[157,158]

The management of patients with hearing impairment varies with the etiology and severity of the impairment.[159]

Conductive hearing loss: Surgical or medical intervention or a combination of the two may be successful in treating conductive hearing loss. Pro-

cedures include repair and reconstruction of the tympanic membrane (myringoplasty), replacement of ossicles, plastic surgery on the outer ear, and early removal of CN VIII tumor.

Sensorineural hearing loss: Contrary to what is commonly believed, hearing aids can often improve sensorineural hearing loss, especially when caused by head injury. Audiological rehabilitation is necessary, in addition to using a hearing aid.

Cochlear implants: For selected patients with profound sensorineural hearing loss cochlear implants are a therapeutic option. Electrical stimulation of the auditory nerve affords some awareness of environmental noise signals, but not necessarily recognizable speech. Intensive audiological rehabilitation is necessary after the implants have been provided.

Hearing aids: Hearing aids are the major rehabilitation tool. They should be considered for all patients with irreversible forms of hearing impairment. Their function is to improve the delivery of sound to the ear. Modern electronic types allow speech to be presented at a comfortable level. The components of hearing aids are a microphone for converting sound energy to electrical energy, an amplifier, and a receiver (the earphone) that converts the amplified signal back to acoustical energy. Hearing aids are available in different types. Some are worn behind the ear or at other places on the body, and some are worn completely inserted in the ear. Digital hearing aids have been developed recently and represent a significant advance for persons with hearing deficits. These instruments amplify primarily the spoken word, not all signals indiscriminately.

Other assistive devices: Adaptive devices for the telephone may be extremely helpful in allowing the patient with a hearing impairment to maintain communication with friends and family. Also helpful are captioned television and home appliances that signal with light instead of sound.

Speech reading: This is the skill of observing the lips of a person speaking and using other clues as well to increase the speed and effectiveness of recognizing meaning, if not actually to "read" the words said. Some persons have significant difficulty learning this skill while others learn it quickly.[160] Speech readers require assistance to sharpen their visual perception and to become sensitive to nuances of language.

Vestibular Dysfunction

Vertigo. Positions that provoke dizziness or vertigo should be avoided. Medications used are mainly antihistamines, such as meclizine (Antivert 25–100 mg daily in divided doses), cyclizine (50 mg 1 to 2 times daily), or anticholinergics. Especially useful is the scopolomine transdermal patch, which is attached every 1 to 3 days. Other medications used are antiemetics, such as promethazine (25–50 mg/d); or tranquilizers, such as diazepam (5–10 mg one to three times a day), or oxazepam (10–60 mg/d), or halopredol (0.5–1 mg one to two times per day), or combined preparations.

Positional vertigo. Treatment for positional vertigo emphasizes the postural control underlying stability. Exercises include strengthening on isokinetic equipment, and strategies that address muscular dyscoordination. Such strategies employ EMG biofeedback, functional electrical stimulation, and facilitation techniques that use manual cues to improve motor performance. Exercises are mainly focused on improving balance by concentrating on visual and somatosensory surface inputs (eg, by walking on uneven surfaces). Patients with positional vertigo seem to have an increased sensitivity to visual motion cues, but these patients lose balance if the visual cues are not interpreted correctly. The patient is taught to maintain balance in progressively more difficult movement tasks.[161,162]

Gaze stabilization. Exercises to improve eye–head coordination for stabilizing gaze during head movements employ visual tracking tasks, first with the head stationary, and then with progressive head movement. The exercises are designed to improve visual modulation of the vestibuloocular reflex.[162]

Management

General rehabilitative measures for managing auditory dysfunction include the following:

Education and training: Teaching of sign language is especially appropriate for children and young adults. Teaching speech to children who were born deaf or became deaf at an early age is an awesome task. Proprioceptive, tactile, and visual cues must be used in the absence of auditory feedback. For vertigo, medications that suppress vestibular function can at times be detrimental because such medications also suppress central nervous system function. Patients are taught to avoid positions that provoke vertigo. This deliberate control of posture, however, must be considered carefully because it might delay recovery. The purposes of exercises are to habituate to dizziness, to improve eye–head coordination, and to retrain sensory and motor components of postural control. Treatment of diz-

ziness is based on repeated exposure of the patient to provocative positions or movements (5–10 times/d) in order to habituate to dizziness. Exercises are performed twice a day in the hospital and the patient continues these activities when discharged to home.

Emotional support: It is essential that the patient is assisted to achieve emotional acceptance of the new status imposed by the impairment.

Preventive measures: These extremely important measures include (*a*) protection from exposure to excessive noise by wearing earplugs in the firing range, in industrial environments, and in other places where noise exposure is a problem; (*b*) careful monitoring when ototoxic drugs have been administered; and (*c*) prevention of prenatal rubella.

Referrals to self-help groups: Persons with similar problems can often be of significant help to a patient who has difficulty adjusting to an impairment.

CRANIAL NERVE IX: GLOSSOPHARYNGEAL

The glossopharyngeal nerve is mixed. Branches contain various somatic and visceral afferents and efferents. The anatomy is shown in Figures 7-30 and 7-31.

Anatomy and Function

The motor branchial efferent fibers originate in the nucleus ambiguus, pass through the superior and petrous ganglia (within or distal to the jugular foramen), then leave the skull via the jugular foramen together with CN X and the bulbar fibers of CN XI. CN IX motor fibers are anterolateral to CN X, innervate the stylopharyngeus muscle (a pharyngeal elevator and the sole striated muscle supplied by CN IX), and then penetrate the pharyngeal constrictor muscles at the base of the tongue. The pharyngeal constrictors are supplied by CN IX and X.

The parasympathetic-visceral efferent fibers originate in the inferior salivatory nucleus. Preganglionic fibers reach the otic ganglion via the tympanic branch of CN IX and via the lesser superficial petrosal nerve. Postganglionic fibers supply the parotid gland via the auriculotemporal nerve (a branch of CN V) and stimulate secretion of saliva.

The sensory-somatic afferent fibers have their primary neurons in the petrous ganglion. The petrous ganglion receives sensory fibers (Jacobson's nerve)[141] from the area between the ear and mastoid cells, and from the lining of the tympanic membrane and the eustachian tube. The sensory fibers terminate eventually in the nucleus of the spinal tract of CN V.[163]

The sensory-visceral afferent fibers have their primary neurons in the superior or petrous ganglion, which receives fibers arising from unipolar cells. The sensory-visceral afferent fibers convey sensation from the posterior third of the tongue, epiglottis, posterior and lateral pharynx (including pain fibers), posterior part of the soft palate,[164] tonsillar region, uvula, and the eustachian tube. Centrally they terminate in the tractus solitarius.

The sensory-visceral-gustatory afferent fibers have their primary neurons in the petrous ganglion and terminate centrally in the nucleus solitarius.

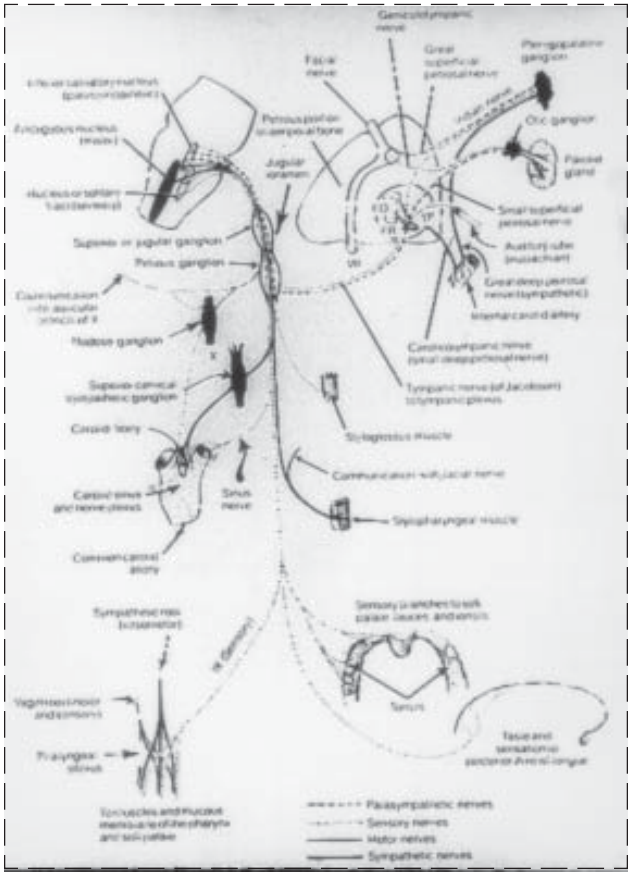

Fig. 7-30. The glossopharyngeal nerve. FO: Fenestra ovalis (oval window); FR: Fenestra rotunda (round window); TP: Tympanic plexus. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy*. 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 168.

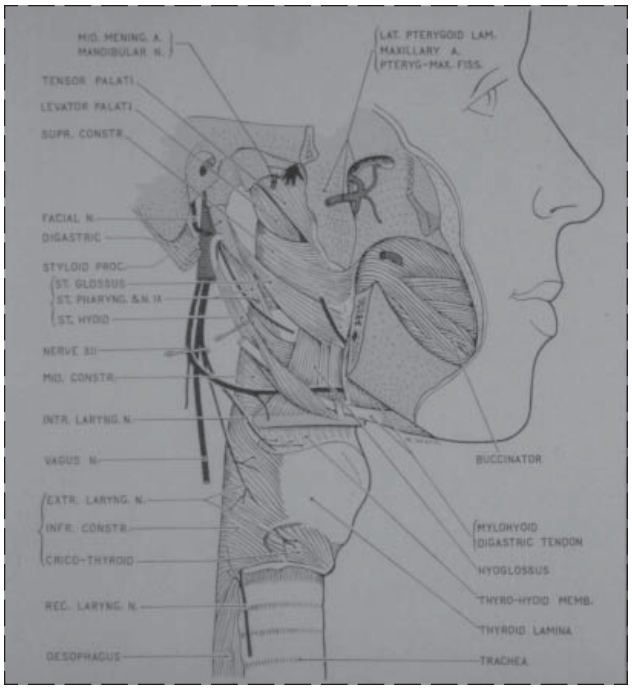

Fig. 7-31. Muscles and structures relevant to swallowing, lateral view. Reprinted with permission from Anderson J. *Grant's Atlas of Anatomy.* 7th ed. Baltimore, Md: Williams & Wilkins; 1978.

The petrous ganglion receives fibers conveying taste from the posterior third of the tongue.

The sensory-visceral, chemoreceptor and baroreceptor afferent fibers have their primary neurons in the petrous ganglion, which receives impulses conveyed through the sinus nerve fibers from the special chemoreceptors and baroreceptors in the carotid body and carotid sinus, respectively. The sensory-visceral chemoreceptor and baroreceptor afferent fibers are concerned with reflex control of respiration, blood pressure, and heart rate. Centrally, these fibers terminate in the nucleus solitarius.

The central connections from the nucleus ambiguus travel via the corticobulbar tract. They have reflex connections from the extrapyramidal and tectobulbar tracts as well as from the nucleus tractus solitarius. The inferior salivatory nucleus receives cortical impulses via the dorsal longitudinal tract and via reflex connections from the nucleus or the tractus solitarius. The sensory fibers are connected with the cortex via the medial lemnisci and thalamus, and reflexly with the salivatory nucleus, the nucleus ambiguus, and the motor nucleus of CN VII.

Injuries and Lesions

The most common cause of injury or lesion is trauma, especially head injury due to blunt trauma. Isolated injuries of CN IX are rare because CN IX, X, XI, and XII are in close anatomical relationship in the posterior cranial fossa, the nucleus ambiguus, and the jugular foramen. As noted previously, upper cervical lesions may also affect the last four cranial nerves and CN VI (the latter because of its long course through the base of the skull).

Penetrating gunshot wounds are the most common cause of traumatic injury to the last four cranial nerves.[165] Missile wounds or skull fractures at the jugular foramen are likely to damage CN IX and XI without involvement of CN X (Vernet's syndrome).

Nontraumatic lesions may be due to syringobulbia, tumors (meningioma,[166] chordoma,[167] glomus jugularis tumor[168]), multiple sclerosis, and Guillain-Barré syndrome (in almost all fatal and in many nonfatal cases, CN IX and X are affected). At times, even cervical lymphadenopathy, infections of the mastoid, and basal meningitis can cause CN IX lesions and glossopharyngeal neuralgia. The incidence of glossopharyngeal neuralgia as compared with trigeminal neuralgia is 1:100.[169,170]

Syndromes involving the glossopharyngeal nerve are:

- Bonnier's syndrome (CN VIII, IX, and X) is caused by a lesion at the lateral vestibular nucleus (Deiter's) and adjacent pathways. It is manifested by paroxysmal vertigo; symptoms and signs of CN IX, X, and occasionally, III and V lesions; contralateral hemiplegia; occasional somnolence; apprehension tachycardia; and weakness.
- Vernet's syndrome (jugular foramen syndrome, CN IX, X, XI) usually results from basilar skull fracture. It presents with paralysis of muscles innervated by ipsilateral glossopharyngeal, vagal, and accessory nerves. The *Vernet's Rideau* phenomenon, which normally has constriction of the posterior pharyngeal wall when saying "ah," is absent in CN IX lesions.
- Glossopharyngeal neuralgia (tic douloureux of the glossopharyngeal nerve) presents with painful attacks similar to trigeminal neuralgia, but in the glossopharyngeal nerve distribution.
- Reichert's syndrome is an incomplete "neuralgia" affecting the tympanic branch

(Jacobson's nerve) of the glossopharyngeal nerve. It may be relieved by intracranial section of the glossopharyngeal nerve. Pain is limited to the ear and eustachian tube.

The symptoms and signs of glossopharyngeal nerve lesions are:

- Loss of gag reflex.
- Slight dysphagia.
- Loss of taste in posterior third of tongue.
- Deviation of uvula to the uninvolved side.
- Loss of sensation in the pharynx, tonsils, fauces, back of tongue.
- Loss of constriction of the posterior pharyngeal wall when saying "ah."
- Increased salivation, involvement of the tympanic plexus in middle ear lesions.
- Possible nystagmus of the uvula in central inflammatory and respiratory lesions (rare).
- Tachycardia, probably from disturbance of the carotid sinus.
- Sharp, knife-like pains in the territory of the glossopharyngeal nerve (region of tonsillar fossa, pharynx, or base of tongue) in glossopharyngeal neuralgia; these are spontaneous or triggered by swallowing, yawning, clearing the throat, or talking.

Evaluation

Clinical evaluation begins with an observation of the palatal arch at rest and on vocalization. The affected site of the palatal arch may be somewhat lower at rest, causing a mild dysphagia. However, with vocalization the palate usually elevates symmetrically.[171]

Sensation of the posterior third of the tongue, the soft palate, tonsillary regions, and pharyngeal wall must be tested. Reflex function is tested by eliciting the pharyngeal (gag) reflex. The posterior pharyngeal wall or base of tongue is stimulated. The expected response is constriction and elevation of the pharyngeal musculature and retraction of the tongue. The afferent arc is carried by the glossopharyngeal nerve; the efferent arc by CN IX and X.

To test the palatal reflex, the soft palate is stimulated and elevation of the soft palate and ipsilateral deviation of the uvula is observed. Again, the afferent arc is carried by the glossopharyngeal nerve; the efferent arc by CN IX and X.

The sensory component of the carotid sinus reflex is tested by pressing over the carotid sinus. This normally produces slowing of heart rate and fall of blood pressure. The reflex may be deficient in lesions of CN IX or X.

The swallowing reflex is triggered by food or saliva at the base of the anterior faucial pillars. The afferent arc is carried by CN IX, X, and XI to the swallowing center in the medullary reticular formation located within the brainstem.[172] The efferent arc is carried by motor fibers of CN IX and X. CN VII, V, and XII may add components to this reflex. The reflex consists of three major phases: (1) oral, (2) pharyngeal, and (3) esophageal.

Swallowing during the three phases is preferably observed through a modified barium swallow (Figure 7-32). A barium swallow with cineesophagram (recorded on videotape) is modified by using three boluses of different consistencies (liquid, paste, and solid) to permit assessment of the oropharyngeal phases.

The *oral phase* is voluntary and accomplished by the CN VII innervated orbicularis oris and buccinator (lip seal); by CN V innervated muscles of mastication; and by CN XII innervated intrinsic and extrinsic muscles of the tongue.

Fig. 7-32. Schematic presentation of swallowing of a radio-opacified liquid bolus during fluoroscopy. First two pictures (A and B) represent the oral phase; the second two (C and D) represent the pharyngeal phase; these are followed by a picture of the esophageal phase (E). Reprinted with permission from Logemann T. *Evaluation and Treatment of Swallowing Disorders.* Austin, Tex: Pro-Ed; 1983.

The *pharyngeal phase* is short (0.5 second) and mainly involuntary. It is most critical for possibly life-threatening aspiration. In this phase, appropriate placement of the bolus is assured by preventing back flow (velar-nasopharyngeal seal, tongue-palate seal, lip closure seal) and inappropriate forward flow (tracheal seal). There is reflex inhibition of breathing. The CN V innervated tensor veli palatini assists the CN IX and X innervated levator veli palatini in creating the velar seal.

The CN V innervated anterior belly of the digastric, the CN VII innervated posterior belly, and the stylohyoid muscle (CN VII) assist the CN XII innervated hyoglossus and genioglossus muscles in laryngeal displacement. CN IX and X provide innervation to the aryepiglottis muscle (epiglottic tilt), to the lateral cricoarytenoid and thyroarytenoid (glottic seal), to the stylopharyngeus, and to the three pharyngeal constrictors to provide pharyngeal compression. The CN XII innervated styloglossus and hyoglossus assist in this function. Pharyngoesophageal relaxation is innervated by CN IX and X (cricopharyngeal inhibition). The *esophageal phase* is involuntary and is accomplished by CN X innervated muscles.[173]

The quantity of salivary secretion from the parotid gland can be measured. It may be absent, decreased, or occasionally increased, with glossopharyngeal lesions. Assessment for glossopharyngeal neuralgia requires checking the heart rate. Bradycardia or even asystole may cause syncopal episodes during neuralgic attacks, possibly due to stimulation of the nucleus solitarius and dorsal motor nucleus of the vagus by impulses originating in glossopharyngeal afferents.[171]

Electrodiagnosis and Imaging

Electrodiagnostic evaluation consists of standard EMG testing of the palate, evaluating pharyngeal constrictors supplied by CN IX and X. See the discussion of CN X below. Imaging by video fluoroscopy is the examination of choice for determining swallowing deficits.

Prognosis

The prognosis for recovery from swallowing problems secondary to stroke, closed head injury, neurosurgical procedures, or poliomyelitis is quite good. The prognosis is poor for swallowing problems secondary to degenerative disease such as Parkinson's, amyotrophic lateral sclerosis, multiple sclerosis, muscular dystrophy, and dermatomyositis. Prognosis for recovery from glossopharyngeal neuralgia is good if the patient responds to anticonvulsants, or if pathology can be identified during surgery and corrected. The stylopharyngeus is difficult to assess. Most other objective tests also examine CN X (see below).

Management

Any patient found to aspirate more than 10% of every bolus regardless of consistency of food should not receive oral feedings. Fluoroscopy examination (modified barium swallow) will assist the physician and speech therapist in deciding whether treatment should be directed toward appropriate adaptive behaviors, or whether exercises should be carried out to improve appropriate motor controls as prerequisites to normal swallowing. Therapeutic techniques may be applied during the fluoroscopic study to evaluate whether these would in fact reduce aspiration. Management also applies to CN X lesions.[173] Available techniques[174] of therapy are as follows:

- Exercises to improve motor control.
- Range of lip motion exercises, to improve labial closures.
- Tongue resistance exercises.
- Exercises to hold a cohesive bolus.
- Bolus propulsion exercises.
- Range of tongue motion exercises, to improve oral transit.
- Possible use of palatal reshaping prothesis to improve oral manipulation of food and speed of oral transit.
- Therapy to stimulate the swallowing reflex with a small, long handled, iced laryngeal mirror.
- Exercises to increase adduction of vocal folds to improve airway protection while swallowing.
- Special maneuvers to improve swallowing:
  - Close airway before and during the swallow (eg, supraglottic swallow); prolong laryngeal elevation and cricopharyngeal opening (Mendelsohn maneuver).
  - Postural changes to redirect bolus (eg, head rotated to damaged side for unilateral pharyngeal paresis); widen vallecula and place epiglottis in protection position (eg, head down "chin tuck" for delayed pharyngeal reflex and weak pharyngeal peristalsis); facilitate oral transit (eg, head back).[175]

  - Food and liquid consistency modifications are recommended after evaluation (clinical and/or with modified barium swallow fluoroscopy study) according to items patient can swallow without aspiration.
- Dilation of cricopharyngeus sphincter. Progressively larger bougies are gently passed through the sphincter gradually stretching the muscle. However, dilation is usually temporary, and eventually a myotomy will have to be performed.
- Teflon injection into damaged vocal folds, to improve closure and airway protection during swallowing .[176]
- Nasogastric feeding.

If no improvement is noted in 3 to 6 months, surgery, such as cricopharyngeal myotomy, may have to be considered.[177] If the impairment cannot be altered significantly, rehabilitation efforts should be aimed at reducing the effect of the impairment by providing alternate means of safe eating and drinking, possibly by modifying the diet[178,179] or swallowing patterns. Eventually a permanent gastrostomy may be necessary.

Medical management for glossopharyngeal neuralgia is the same as for tic douloureux (trigeminal nerve), using anticonvulsants (phenytoin, carbamazepine). Suboccipital craniectomy with exploration of the glossopharyngeal nerve is indicated in patients failing to respond to anticonvulsants. If a compressing blood vessel is found and mobilized, pain will stop. If no structural deficit can be identified, the nerve should be sectioned.[92]

CRANIAL NERVE X: VAGUS

The vagus nerve is mixed, containing somatic and visceral afferents and somatic and visceral efferents. See Figures 7-33 through 7-36.

Anatomy and Function

The branchial efferent vagal fibers derive from the nucleus ambiguus, which also contributes motor rootlets to the glossopharyngeal nerve and the internal ramus of the accessory nerve. These three cranial nerves leave the skull through the jugular foramen. The vagus nerve fibers supply all the striated muscles of the soft palate, pharynx, and larynx, except for the tensor veli palatini supplied by CN V and the stylopharyngeus supplied by CN IX. Other motor fibers of the vagus nerve, together with fibers of the accessory nerve, join outside the skull, and through the recurrent laryngeal nerve supply all of the intrinsic muscles of the larynx except for the cricothyroid. The left recurrent laryngeal nerve arises as a branch in the thorax and recurs by looping around the aortic arch; on the right side it arises in the root of the neck looping around the right subclavian artery to recur. Both recurrent nerves then ascend between the trachea and esophagus to reach the laryngeal muscles. The cricothyroid muscle as well as the inferior constrictor muscle of the pharynx are innervated by the external branch of the superior laryngeal nerve. The internal branch of this nerve gives sensory supply to the larynx.

The parasympathetic-visceral efferent preganglionic fibers arise in the dorsal motor nucleus of the vagus and terminate on ganglia close to the thoracic and abdominal viscera. The postganglionic fibers supply the smooth muscles and glandular structures of the pharynx, larynx, and viscera.

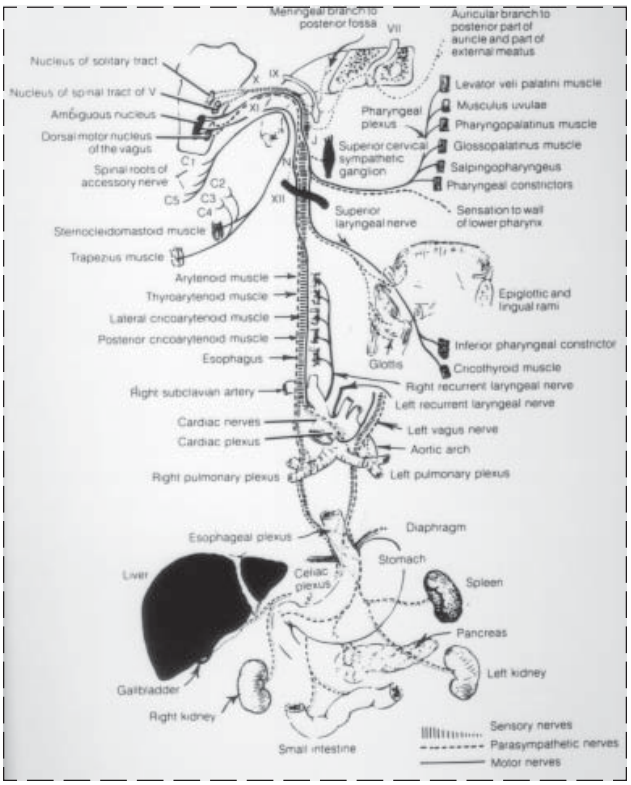

Fig. 7-33. The vagus nerve. N: Nodose (inferior) ganglion; J: Jugular (superior) ganglion. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy*. 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 171.

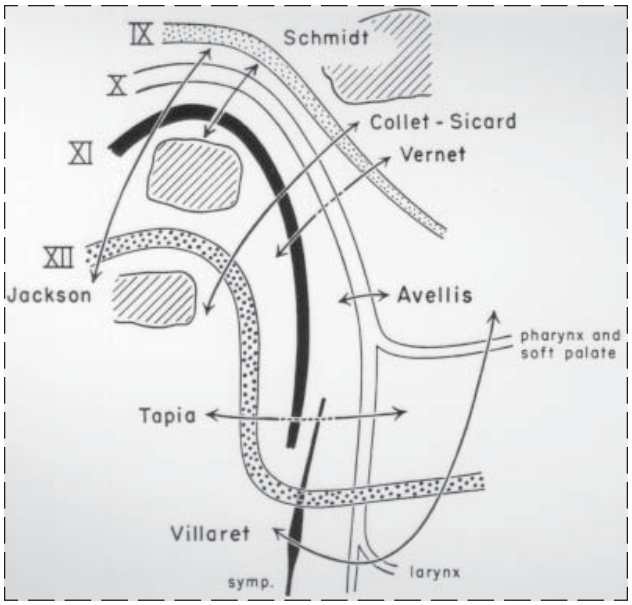

Fig. 7-34. Combinations of nerves affected in various syndromes. The broken lines over certain nerves indicate that they may or may not be affected. Reprinted with permission from Vernet M. *J Laryng Otologol*. 1918; 33:354.

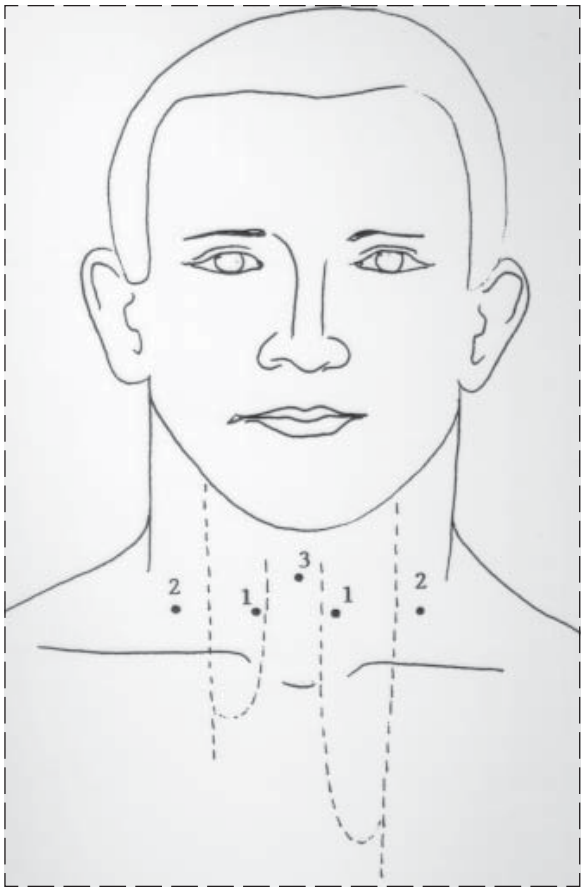

Fig. 7-35. Sites of electrode placement for paratracheal stimulation of the recurrent laryngeal nerve. 1) Recurrent laryngeal nerve; 2) Vagus nerve; 3) Thyroid cartilage. Drawing: Courtesy of artist: Dr. Anna Bettendorf, University of Virginia.

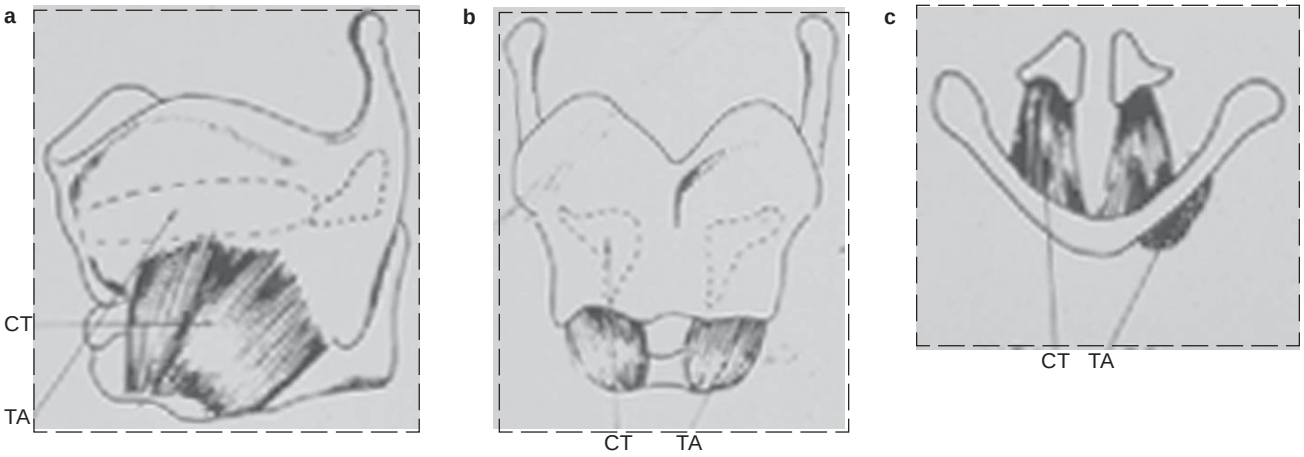

Fig. 7-36. (**a**) Lateral, (**b**) anterior, and (**c**) superior projection of the larynx and EMG needle insertion sites. CT: Cricothyroid muscle. TA: Thyroarytenoid muscle. Reprinted with permission from Simpson D, Sternman D. Vocal Cord Paralysis: Clinical and Electrophysiologic Features. *Muscle Nerve.* 1993;16:953**.**

Stimulation of the vagus nerve produces bradycardia, while vagus nerve paralysis results in tachycardia. CN X fibers also vasoconstrict the coronary arteries. There is inhibition of suprarenal secretions and stimulation of gastrointestinal peristalsis and gastric, hepatic, and pancreatic glandular activity. In general, the vagus nerve is the largest and most important parasympathetic nerve in the body, and also the longest and most widely distributed cranial nerve.

The somatic afferent fibers have their primary neurons in the jugular ganglion subserving fibers carrying sensation from the posterior portion of the external acoustic meatus, the adjacent part of the tympanic membrane, and a small area in the posterior aspect of the pinna. The fibers eventually terminate on the trigeminal nucleus. Afferent fibers carrying pain sensation from the dura mater of the posterior cranial fossa relay in the jugular ganglion and terminate in the spinal tract of the trigeminus and its nucleus. Fibers conveying sensory information from structures above the vocal cords travel in the internal laryngeal nerves; those below the vocal folds travel in the recurrent laryngeal nerves.

The visceral afferent fibers have their primary neurons in the ganglion nodosum carrying sensation from the lower pharynx, larynx, trachea, esophagus, and the thoracic and abdominal viscera. The fibers collect centrally as the fasciculus solitarius and terminate on its nucleus. Fibers that carry gustatory sensation from the epiglottis and the arytenoids also relay through the ganglion nodosum and terminate in the nucleus solitarius.[171] The central connections for the vagus nerve are as described under CN IX above.

The muscles supplied by the vagus nerve are

- Levator veli palatini: occludes the nasal passages in swallowing.
- Musculus uvulae: retracts uvula backward, helping to block off nasal passages.
- Palatoglossus: narrows the fauces in swallowing by elevating posterior tongue and depressing soft palate.
- Palatopharyngeus: approximates the pharyngopalatine arches and closes posterior nares and faucial orifice.
- Salpingopharyngeus: raises upper and lateral portions of pharynx.
- Superior, middle, and inferior constrictors of the pharynx: Together, these flatten and contract the pharynx in swallowing. The three constrictors help to force the food into the esophagus and initiate peristaltic waves. They also assist in production and modulation of speech.

The muscles supplied by the recurrent laryngeal nerve (branch of the vagus nerve) are

- Posterior cricoarytenoids: chief abductors that separate the vocal cords.
- Lateral cricoarytenoids: chief adductors.
- Thyroarytenoids: shorten and relax vocal cords (also adduct).
- Arytenoid (unpaired): closes the opening of the glottis, especially the posterior aspect. The arytenoid has two parts. The oblique arytenoid acts as a sphincter of the upper larynx, and the transverse arytenoid closes the posterior pad of the glottis.

The muscles supplied by the superior laryngeal nerve (external branch) are the cricothyroids, the chief tensors of the vocal chords. The extrinsic muscles of the larynx (functionally a part of the voice apparatus) include also those supplied by CN XII and by upper cervical nerves that insert on the hyoid bone.

Reflexes with vagus nerve participation are gag, swallowing, cough, sucking, hiccup, yawning, carotid sinus, and Hering-Brewer. The Hering-Brewer reflex is an overstretching of receptors in the visceral pleura and the tracheobronchial tree, causing inhibition of respiration to prevent overdistension.

Injuries and Lesions

The most common cause of injury is trauma, blunt and penetrating. Such traumas most frequently involve the posterior fossa. Injuries and other lesions also frequently involve CN IX, XI, and XII (Collet-Sicard syndrome). See Figure 7-34. The incidence of dysphagia following closed head trauma is reported as 26% to 27%.[178,179]

Injuries to CN VI, IX, X, XI, and XII may also be concomitants of high cervical lesions.[1,2] Neck and thorax injuries and lesions can affect the superior laryngeal and the recurrent laryngeal nerves. Vocal cord paralysis is a relatively common disorder. Adour[180] reported superior laryngeal nerve paralysis to have a higher incidence than facial palsy. Yamada et al[181] investigated 564 patients with recurrent laryngeal nerve paralysis in a 10-year period (1971–1980). The largest group of cases were

idiopathic (230 cases), 68 were caused by thyroid surgery, and 63 were due to endotracheal intubation. The last group had the best recovery rate.

There are reports from World War I of penetrating injury to the vagus nerve caused by shrapnel.[182] In 1909, an injury to the recurrent laryngeal nerve following a gunshot wound to the neck was successfully repaired.[183]

Surgical trauma is the most common cause of vocal cord paralysis.[184,185] The recurrent laryngeal nerve is especially vulnerable during thyroidectomies.[186] The incidence is reported as 0.3% to 13.2% depending on the experience of the surgeon.[187] Purposeful sacrifice of the recurrent laryngeal nerve during cancer surgery because of involvement in the tumor is no longer advocated by many surgeons if the vocal cord function is normal.[188]

Nontraumatic lesions are due to vascular insults (lateral medullary or Wallenberg's syndrome), motor neuron disease, tumors, syringobulbia, inflammatory disease, Guillain-Barré syndrome, carcinomatous meningitis, metastases, sarcoidosis, nasopharyngeal diphtheria, and chronic lead poisoning. Abductor paralysis in children is frequently associated with Arnold-Chiari malformation; other congenital malformations; birth trauma; or are idiopathic, possibly of viral origin. These nontraumatic lesions are mainly reported in the cranium. Lesions affecting the vagus nerve in the neck or thorax are mainly tumors, aneurysms of the internal carotid artery, enlarged lymph nodes, or the result of surgical trauma. The sites of lesions of CN IX, X, XI, and XII are shown in Figure 7-34. Syndromes involving the vagus nerve are

- Jugular foramen syndrome of Vernet, involving CN IX, X, XI;
- Schmidt's syndrome, involving CN X, XI;
- Hughlings-Jackson syndrome, involving CN X, XI, and XII;
- Collet-Sicard syndrome, involving CN IX, X, XI, and XII; and
- Avellis's syndrome, involving CN X and XI.

See also the discussion of syndromes for CN IX above.

Symptoms and Signs

The symptoms and signs of CN X malfunctions are motor disturbances within the pharynx and larynx; sensory disturbances within the pharynx and larynx; and autonomic disturbances involving heart rate, glandular dysfunction, and gastrointestinal functions. These symptoms and signs of CN X lesions are summarized below.

Motor Disturbances

- Aphonia (paralysis of vocal cords or hysteria).
- Dysphonia (impairment of voice, often with unilateral lesions producing nasal speech, hoarseness, weak voice).
- Position change of vocal cord, observed on examination (can be asymptomatic).
- Dysphagia (difficulty in swallowing) is caused by damage to the pharyngeal branches of the vagus resulting in paralysis of the palatini muscles and the pharyngeal constrictors. Dysphagia may be profound, especially for liquids, which tend to escape through the nasal cavity.
- Defective elevation of the palate on phonation and movement of the uvula toward the unaffected side (clinical symptoms may be absent).
- Unilateral vocal cord paresis; except for some snoring at night or minor nasal speech, the ipsilateral vocal cord will assume the cadaveric position (midway between adduction and abduction). There is little if any dyspnea.
- Bilateral vocal cord paresis; respiration may be severely affected.
- Stridor, especially in children.
- Vocal cord immobility (observed during laryngoscopy) can be caused by cricoarytenoid joint fixation, interarytenoid scarring, recurrent laryngeal nerve paralysis, or laryngeal synkinesis. To establish definitely the cause of immobility, laryngeal EMG must be performed.
- Esophageal, cardiac, or pyloric spasm not due to local causes or possibly of viral origin.
- Loss of gag reflex (paralysis of soft palate).
- Anesthesia of larynx, hoarseness, and voice fatigue (as secondary to unilateral superior laryngeal nerve paralysis, which is rare and usually traumatic).

Sensory Disturbances

- Pain or paresthesias of the pharynx, larynx, and external auditory meatus. In vagus or superior laryngeal neuralgia (tic douloureux), pain may be severe and knife-like.

- Anesthesia of the lower pharynx and larynx, as occurs in complete vagus nerve lesions.
- Cough, a constant symptom of vagus irritation.
- Dyspnea or pseudoasthma as may occur if the reflex vagal control of respiration is interrupted.
- Salivary hypersecretion as may occur with irritative lesions.
- Hypersecretion of acidic gastric fluids, which may cause gastric ulceration. Selective vagotomy may be necessary for recurring ulcers.

Parasympathetic (vegetative) disturbances

- Bradycardia occurs with irritative lesions.
- Tachycardia occurs with palsy of the vagus nerve.
- Gustatory sweating occurs after vagus nerve damage, possibly due to aberrant regeneration (see glossopharyngeal nerve).
- Intermittent diarrhea (lasting 1 to 2 days and recurring every 1 to 2 weeks) due to under activity of vagus (diabetes).
- Possible esophageal dysfunction, gastric dilation, and nocturnal diarrhea.

Evaluation

Clinical examinations should begin with observation of the soft palate and uvula at rest and in phonation. The palate should elevate symmetrically with no deviation of the uvula.[189] The character of the voice, breathing, and cough are evaluated. During laryngoscopy the laryngeal movements can be observed. Reflex testing should include the gag reflex (CN IX and X), the occulocardiac reflex (pressure on the orbit should cause cardiac slowing), the carotid sinus reflex (pressure on carotid sinus to produce cardiac slowing), sinus arrhythmia, and pulse retesting for variations during deep inhalations followed by slow deep exhalation.

Electrodiagnosis

One kind of electrodiagnostic testing consists of nerve conduction studies of the recurrent laryngeal nerve.[190] Because of the relative inaccessibility of the structures and the unreliability of distance measurements, the method has not found wide acceptance and is rarely performed. The procedure is described below. EMG[191–195] (see Figures 7-35 and 7-36) of the laryngeal muscles is carried out according to standard procedures.

Procedure for Recurrent Laryngeal Nerve Conduction Study

Stimulation: A needle stimulating electrode (5 cm) is inserted 2.5 cm below the most prominent point of the cricoid arch paratracheally. The anode is placed in the subcutaneous tissue of the jugular fossa. A second stimulus is applied at the same level as the first stimulus, but at the posterior edge of the sternocleidomastoid muscle. The depth of the cathode is adjusted to the lowest threshold that will produce an action potential. Again, the anode is placed in the subcutaneous tissue.

Recording: Electrodes inserted through a laryngoscope are placed in the thyroarytenoid muscle. Distance from point of stimulation averages 10 cm on the right and 21.5 cm on the left.

Normal values:

- Distal latency: 2.1 ± 0.05 millisecond (average distance 5.9 ± 0.05 cm)
- Conduction velocity: 60–70 m/s
- Amplitude: 6.8 ± 0.7 mV
- Duration: 6.6 ± 0.5 millisecond

The *cricothyroid muscle* is tested by inserting a small (approximately 2 cm) electrode approximately 1 cm from the midline over the lower aspect of the thyroid cartilage, to a depth of a few millimeters. The patient is instructed to phonate a low pitched vowel ("e" as in English "sea") and gradually raise the pitch. Increasingly more muscle activity is identified. To reduce discomfort, the skin may be infiltrated with 1% lidocaine (1 cm^3) with 1:100,000 epinephrine before insertion of needle.

The *thyroarytenoid muscle* can be reached by inserting a 7.5 cm needle through the cricothyroid membrane 0.5 cm laterally from midline. In males, the needle should then be angled 45° superiorly, while in females the angle should be approximately 30°. The needle is inserted to a depth of approximately 2.0 cm. The patient is required to say "ee" repeatedly; as the recording needle tip approaches the fibers of the thyroarytenoid or adjacent muscles (lateral cricoarytenoids), action potentials are detected. Observation is as with conventional EMG; at rest it should be silent even though in intrinsic laryngeal muscles there are often baseline normal action potentials observed. These potentials are diphasic or triphasic, of 3 to 6 millisecond duration and 100 to 300 μV in amplitude. Further activity summates into an interference pattern. Examination can be performed in most cases without difficulty, especially in the absence of laryngospasm, he-

matoma, or other complications. However, a laryngoscope, endotracheal tube, Ambu bag, and other resuscitative equipment should be available.

The most common conditions for which EMG is performed are unilateral recurrent laryngeal palsy, superior laryngeal palsy, recurrent laryngeal palsy, spastic dysphonia, chronic hoarseness, immobile vocal cords, incomplete recovery of cricoarytenoid joint fixation, or idiopathic vocal cord paralysis.

A recent report by Simpson and colleagues[192] discussed electromyographic findings in 52 vocal cords of 44 patients with idiopathic vocal cord paralysis. The most common complaints of these patients were hoarseness and shortness of breath (with bilateral involvement). Not as common were other symptoms suggesting laryngeal incompetence, such as loss of high pitch phonation. In 67.4% of vocal cords, evidence of acute or chronic denervation was shown (55% in the thyroarytenoids, 10% in the cricothyroids, and 2% involving both of these muscles); 33% were normal. Another study[194] by the same investigators reports on 48 patients (60 vocal cords) with laryngeal disorders due to different conditions. In 53% of vocal cords there was cranial nerve involvement (28% including the thyroarytenoid, 10% the cricothyroid, and 15% both). Patients with spastic dysphonia showed no spontaneous activity; action potential parameters were of normal values, but firing patterns were altered. There were some patients who showed normal position and activation of vocal cords during the laryngoscopic examination, but EMG detected some evidence of denervation. Conversely, there were patients whose laryngoscopic examinations were suggestive of cranial nerve lesions, but EMG changes were not observed. Involvement of the cricothyroid alone was mostly observed in professional singers.

Posterior criocarytenoid muscles can also be examined as described by Hiroto and colleagues.[193] These tests may be performed with a curved needle electrode or a hooked-wire electrode.

Since the criocarytenoid muscles are supplied by the recurrent laryngeal nerve, electromyography adds little to the diagnosis of a lower motor neuron process and is, therefore, only confirmatory. For kinesiological electrodiagnostic study, however, additional wire electrode placement may be of interest for observing the functional capacities and synergistic movements during swallowing and phonation. This may be especially applicable to the study of spastic dysphonias.

The superior constrictor muscles of the pharynx[196,197] and the cricopharyngeal muscle can be studied effectively with special bipolar suction electrodes.

Quantitative vagus parasympathetic fiber testing can be accomplished by assessment of sinus arrhythmia with an electrocardiograph, EMG, or other computer-assisted analog integrator. Normal vagus parasympathetic innervation of the heart mediates slowing of heart rate with exhalation, which follows an increasing heart rate during inhalation. Vagus denervation abolishes sinus arrhythmia, although exact reflex pathways are not known. During the test, 5 to 6 breaths per minute are taken and heart rate is recorded for 1 to 2 minutes. The maximal-to-minimal heart rate per respiratory cycle is determined, and the mean maximal-to-minimal rate variation is calculated. Test results are influenced by patient's body position, respiratory rate, age, and other factors such as changes in intrathoracic and intraabdominal pressures (Valsalva maneuver) or drugs (propanolol, atropine as autonomic blocking agents).[198]

Esophageal manometry can be used to identify disruptions in the peristaltic wave through the pharynx and esophagus. With the help of fluoroscopy, upper and lower esophageal sphincter disorders may be identified.

Prognosis

The prognosis of acute complete bilateral lesions of the vagus nerve is unfavorable without immediate emergency measures. Unilateral recurrent laryngeal nerve injury, which may simply cause hoarseness, is usually transient. Bilateral recurrent laryngeal palsies (postthyroidectomy, polyneuropathy, or carcinoma) may produce severe airway limitation, often necessitating a tracheostomy. Swallowing difficulties, especially after stroke and head injury, may be severe but often improve with time. Spastic dysphonia is quite resistant to treatment, and prognosis for recovery is poor. Unilateral recurrent laryngeal nerve sections have been advocated by Dedo and Izbedski.[199] Idiopathic paralysis (viral infection) usually recovers spontaneously within 6 months. The majority of patients showed long term improvement with only minor hoarseness as a complication.

Management

Management of dysphagia is described in the discussion of CN IX. Unilateral paralysis of laryngeal muscles if bothersome to the patient can be treated with speech therapy for better voice production. Bilateral paralysis usually needs tracheostomy or intralaryngeal surgery, such as arytenoidectomy or lateral fixation of the arytenoids.

Recurrent laryngeal nerve anastomosis, as well as regeneration without surgery, carries a significant risk for aberrant regeneration (synkinesis), which may cause spasticity during phonation (dysphonia), airway difficulty, or aspiration. It is reported[200] as a frequent problem following immediate repair. The reason for this aberrant regeneration is believed to be the composition of the recurrent laryngeal nerve. It consists of nerve fibers to the abductors, which are inspiratory muscles, and fibers that supply the adductors, which produce phonation. Therefore all options must be carefully considered. Crumley et al[201] describes the following choices: in unilateral vocal cord paralysis, an attempt is made to medialize the involved cord; while in bilateral paralysis one of the better vocal cords must be lateralized to open the airway.

Other treatment options include (1) recurrent laryngeal nerve neurorrhaphy; (2) ansa hypoglossi recurrent laryngeal nerve anastomosis; (3) neuromuscular pedicle transfer (a small piece of hypoglossal innervated omohyoid muscle); and (4) Isshiki thyroplasty (laryngoplasty).

Placement of a silastic block in a subperichondrial plane medializes the vocal cords.[202] Nonsurgical invasive procedures are gelfoam paste, phonagel, or Teflon vocal cord injections. Gelfoam is best used as a temporary measure when recovery is expected. Teflon paste is permanent, for use when recovery is not expected. Another option is to observe and wait.

Spastic dysphonia may be treated with section of the unilateral recurrent laryngeal nerve block or botulin toxin injections. Another treatment option is the electrolarynx, an artificial larynx that may assist in communication if no voice can be produced. The device introduces sound through the soft tissue of the neck, and the tongue and lips articulate this sound into intelligible speech. Instruction by a speech pathologist is mandatory in order to accomplish appropriate sound placement, timing, and articulation. If the vocal cords and the larynx are partially destroyed by a malignancy, laryngectomy followed by insertion of a voice prosthesis may be necessary. The most popular technique is the Singer and Blom technique.[202–204] Tracheoesophageal speech with a voice prosthesis offers the most acceptable communication, and importantly, avoidance of aspiration.[205]

Vagal or superior laryngeal neuralgia is treated with medications such as carbamazepine or other anticonvulsants. If there is no relief, suboccipital craniectomy with decompression should be considered. If no lesion can be identified, sectioning of vagal and glossopharyngeal nerves is recommended.[206] Deficits caused by involvement of parasympathetic fibers need medical management.

CRANIAL NERVE XI: ACCESSORY

The accessory nerve is all motor. The anatomy is given in Figure 7-37.

Anatomy and Function

The branchial efferent fibers have two nuclear origins. The cranial component of the accessory nerve arises from the caudal part of the nucleus ambiguus and is closely related to the radicles of the vagus nerve. The accessory and vagus nerves together supply the musculature of the pharynx and larynx. The spinal component arises in the anterior horn of the upper cervical spinal cord (C1–C5), enters the skull through the foramen magnum, and is somatotopically arranged. That is, C1–C2 predominantly innervate the sternocleidomastoid muscle and C3–C4 the trapezius. The cranial and spinal roots leave the skull through the jugular foramen together with CN IX and X, and then again separate. The external ramus penetrates and supplies the sternocleidomastoid muscle, then crosses the posterior cervical triangle and supplies the trapezius muscle. The internal ramus joins the vagus nerve to supply the pharynx and larynx. The lower part of the trapezius is said to have some innervation by the third and fourth cervical roots through the cervical plexus. However, these are mainly proprioceptive fibers. Muscles innervated by the accessory nerve are represented cortically between the elbow and the trunk in the homunculus of the precentral gyrus.

Central connections are mediated through fibers of the corticobulbar tract, which travel from the cortex, accompany the corticospinal tract to the midbrain, and eventually terminate in the brainstem as well as in the voluntary motor nucleus ambiguus. There are reflex connections with the tectospinal and vestibulbospinal tracts for postural reflexes. Corticobulbar fibers to the trapezius are crossed, and therefore lesions cause contralateral deficit. Corticobulbar fibers to the sternocleidomastoid are uncrossed, or more likely, are doubly decussated. Therefore, lesions cause ipsilateral deficit.[207,208] In the brainstem the fibers to the trapezius are located

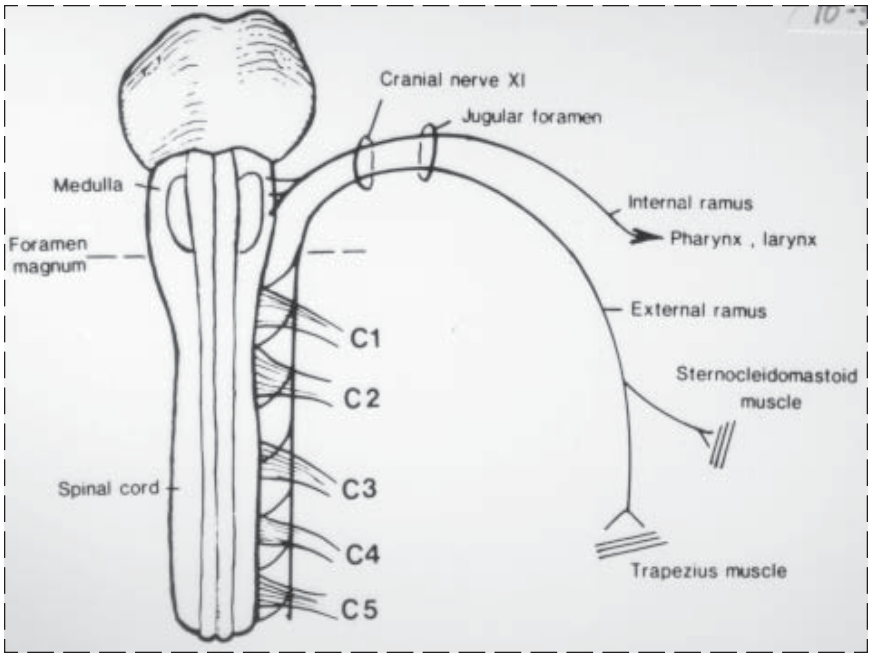

Fig. 7-37. The accessory nerve. Reprinted, with permission from Brazis P, Masdeu J, Biller JL. *Localization in Clinical Neurology*. Boston, Mass: Little, Brown; 1990: 251.

ventrally, and the fibers to the sternocleidomastoid are located in the brainstem tegmentum.[110] A ventral pontine lesion can therefore present with supranuclear paresis of the trapezius but not of the sternocleidomastoid muscle.

Injuries and Lesions

The most common injuries and lesions are the result of trauma such as head injury, especially basal skull fracture and posttraumatic syrinx following spinal cord injury. Neck injuries[209,210] may cause compression or percussion of the accessory nerve in the posterior cervical triangle (football injury),[211] where the accessory nerve is located quite superficially.

Other causes may be surgical trauma such as removal of lymph nodes, biopsy, or more commonly, radical neck dissections. Cannulation of the internal jugular vein[212] may be followed by weakness or paralysis of the trapezius muscle, the sternocleidomastoid, or both.

Nontraumatic lesions are mainly tumor, motor neuron disease, meningitis or other infections, spontaneous accessory nerve lesion[213] or other central lesions.[214,215] The causes and expressions of lesions according to location are given in Tables 7-5 and 7-6. Syndromes involving CN XI are as follows:

Vernet's syndrome is manifested by ipsilateral dysphagia (CN IX, X); diminished gag reflex (CN IX, X); palatal droop (CN IX); ipsilateral vocal cord paralysis (CN X); and weakness and atrophy of the sternocleidomastoid and trapezius muscles (CN XI).

Collet-Sicard syndrome[216] presents with ipsilateral paralysis of the trapezius and sternocleidomastoid (CN XI); paralysis of vocal cord (CN X); paralysis of the pharynx (CN IX); hemiparalysis of the tongue (CN XII); loss of taste from the posterior third of the tongue (CN IX); hemianesthesia of the palate, pharynx, and larynx (CN IX, X).

Symptoms and Signs

The symptoms or signs of supranuclear lesions of CN XI are ipsilateral sternocleidomastoid weakness, contralateral trapezius weakness, and hemiplegia. The symptoms or signs of lesions in the nuclear area are ipsilateral weakness of the sternocleidomastoid and trapezius muscles. Infranuclear lesions present with nuclear and CN IX, X, and XII deficits. Peripheral accessory nerve lesions show normal sternocleidomastoid muscle function, but weakness of the trapezius muscle, pain, atrophy, and drooping of shoulder. Other peripheral lesions may cause torticollis.

Evaluation

The clinical examination consists of strength evaluation of the sternocleidomastoid by applying resistance to head turning, opposite to the side of the muscle tested. Resistance is given also to head tilting. Resistance applied to head flexion tests the

TABLE 7-5

CAUSES AND EXPRESSION OF LESIONS BY LOCATION (CRANIAL NERVE XI)

	Traumatic Lesions	Nontraumatic Lesions	Symptoms/Signs
Supranuclear	Gunshot wounds, other trauma	Tumor	Contralateral hemiplegia including contralateral trapezius, and possible ipsilateral sternocleidomastoid weakness
Nuclear	Posttraumatic syrinx	Tumor, motor neuron disease	May have associated medullary or upper cervical cord dysfunction
Foramen Magnum	Skull fracture, other trauma	Neoplasm, meningitis	May also involve CN IX, X, and XII. Ipsilateral findings, dysphonia, dysphagia, loss of taste posttongue, vocal cord paralysis, tongue paresis, and atrophy
Juglar Foramen	Basal skull fracture	Tumor, infections	May also involve CN IX and X (Vernet's syndrome). Findings are ipsilateral dysphonia and dysphagia (paralysis of ipsilateral vocal cord), and loss of taste and sensation from posterior tongue
Retropharyngeal Space	Trauma	Intramedullary lesions	May also involve CN IX, X and XII. Collet-Sicard syndrome findings are ipsilateral.
Neck	Compression or percussion injury (football)[1,2] Shoulder dislocation[3]	Internal jugular cannulation in posterior triangle, radiation[4] therapy, adenopathy, postsurgery	Ipsilateral findings trapezius and/or sternocleidomastoid muscle weakness.

CN: cranial nerve

(1)Markey K, Di Benedetto M, Curl WW. Upper trunk brachial plexopathy: The stinger syndrome. *Am J Sports Med.* 1993;21:650–656. (2) Di Benedetto M, Markey K. Electrodiagnostic localization of traumatic upper trunk brachial plexopathy. *Arch Phys Med Rehabil.* 1984;65:15–17. (3) Mitchell SW. *Injuries of the Nerves and Their Consequences.* Philadelphia, Pa: JB Lippincott; 1872; 335. (4) Hoffman JC. Permanent paralysis of the accessory nerve after cannulation of the internal jugular vein. *Anesthesiology*. 1983;58:583.

TABLE 7-6

TRAPEZIUS AND STERNOCLEIDOMASTOID MUSCLE INVOLVEMENT

Lesion	Trapezius	Sternocleidomastoid
Upper motor neuron lesion,ipsilateral to sternocleidomastoid above oculomotor complex	Contralateral	Ipsilateral
Ventral brain stem lesion (supranuclear fibers to trapezius) or a lower cervical cord lesion (somatotopic arrangement)	Ipsilateral	
Lower brain stem tegmentum lesion or upper cervical accessory roots		Ipsilateral
Contralateral brainstem lesion or ipsilateral high cervical cord lesion (Peripheral accessory nerve lesion before the nerve divides into muscular branches.)	Ipsilateral	Ipsilateral

two sternocleidomastoid muscles simultaneously. Reflex can be elicited by tapping the insertion of the sternocleidomastoid muscle.

The strength of the trapezius muscle is tested by observing the position of the shoulders in lateral abduction of the arm from approximately 100° to 180° with the arm internally rotated and hand pronated.[217] The levator scapula and the rhomboids also elevate the shoulder. Therefore, the observer must be certain that the trapezius is isolated before judging normal. The position of the scapula is observed; the trapezius elevates, rotates, and retracts the scapula and abducts the arm above the horizontal. Scapular winging indicates a lesion of CN XI, especially the fibers to the middle trapezius. The scapula is more prominent at rest, with the superior angle moving away from the midline.

Electrodiagnosis

Electrodiagnostic evaluation consists of nerve conduction studies[208] of the accessory nerve and EMG of the sternocleidomastoid and trapezius muscles. Conduction studies are summarized below. EMG evaluates insertional activity, activity at rest, minimal and maximal contraction, and recruitment.

Procedure of Nerve Conduction Studies of CN XI

1. *Stimulation* in the posterior cervical triangle.
2. Upper trapezius (scapular elevator). The active electrode is placed on the upper trapezius muscle 10 cm inferior and lateral to the point of stimulation. The reference electrode is placed over the acromion.[211,218]
   Normal values:
   - Latency: 2.3 ± 0.3 millisecond
   - Amplitude: 1.5 ± 0.5 mV
3. Middle trapezius (scapular adductor). The active electrode is placed halfway between the midpoint of the ipsilateral scapular spine and the thoracic spine. The reference electrode is placed on the nearest spinous process.[219]
   Normal values:
   - Latency: 3.0 ± 0.2 millisecond
   - Amplitude: 2.5 ± 1.0 mV
4. Lower trapezius (scapular depressor). The active electrode is placed 2 finger breadths (4–5 cm) from the spinal column on the level of the inferior angle of the scapula.[220] The reference electrode is placed on the nearest spinous process.
   Normal values:
   - Latency: 4.6 ± 0.3 millisecond
   - Amplitude: 1.3 ± 0.9 mV

Imaging and Prognosis

Imaging studies such as high resolution CT through the jugular foramen and MRI are helpful.

The prognosis of CN XI lesions is generally favorable, but considerable time may be required for recovery. Central lesions may recover spontaneously especially if the lesion is nonprogressive.

Management

Range-of-motion exercises should be performed to prevent contractures. Resistive exercises promote strengthening. Preventive measures should be applied when possible. For example, properly fitted shoulder pads and neck rolls for players of American football have been shown to reduce occurrence of stingers (burns).[211]

CRANIAL NERVE XII: HYPOGLOSSAL

The hypoglossal nerve is all motor and is depicted in Figures 7-38 and 7-39.

Anatomy and Function

The somatic efferent fibers arise from the hypoglossal nucleus, which consists of a longitudinal cell column in the medulla. Approximately 10 to 15 rootlets emerge from the ventrolateral sulcus of the medulla along the lateral border of the pyramid. These rootlets are gathered into two bundles and pass through the dura mater and the hypoglossal canal of the skull. The two bundles then combine and descend toward the angle of the mandible, passing near the internal carotid artery and the internal jugular vein. A descending ramus is given off and joins the ansa hypoglossi. The hypoglossal nerve then supplies the intrinsic muscles of the tongue and also the hypoglossus, styloglossus, genioglossus, and geniohyoid muscles. It supplies all extrinsic muscles of the tongue except the palatoglossus, which is supplied by CN X. The intrinsic

muscles of the tongue alter the shape of the tongue, and the extrinsic muscles alter its shape and position. The hypoglossal nucleus contains some spindle afferents.

The central connections are fibers controlling tongue movement that originates from the lower portion of the precentral gyrus near the sylvian fissure. Corticobulbar tract fibers accompany the corticospinal tract and terminate in the brainstem on the motor hypoglossal nucleus. Fibers may be crossed or uncrossed. There are reflex connections with the cortex, extrapyramidal, and tectospinal tracts, and with sensory nuclei of CN V, IX, and X. Corticobulbar control of the genioglossus muscles is contralateral. Most of the other tongue muscles appear to have bilateral supranuclear control.[221]

Injuries and Lesions

Blunt trauma in head injury causing basilar skull fractures may affect CN XII in isolation or in combination with other cranial nerves. However, blunt trauma is not a common cause for CN XII injury. More common are penetrating missile or gunshot wounds in the submandibular region, reported as common causes of hypoglossal nerve injury during the U.S. Civil War (1861–1865),[222] World War I,[223] and in a recent study from Los Angeles County (California) Hospital.[224] Injury of CN XII can occur in the neck or near the nerve entry into the tongue. This is a common site of injury during surgery, especially injury involving endarterectomies. Dislocation or fracture of the upper cervical vertebrae may involve CN XII.

Nontraumatic lesions include vascular lesions, especially in the internal capsule; lacunar strokes; or tumors compressing the corticobulbar fibers to

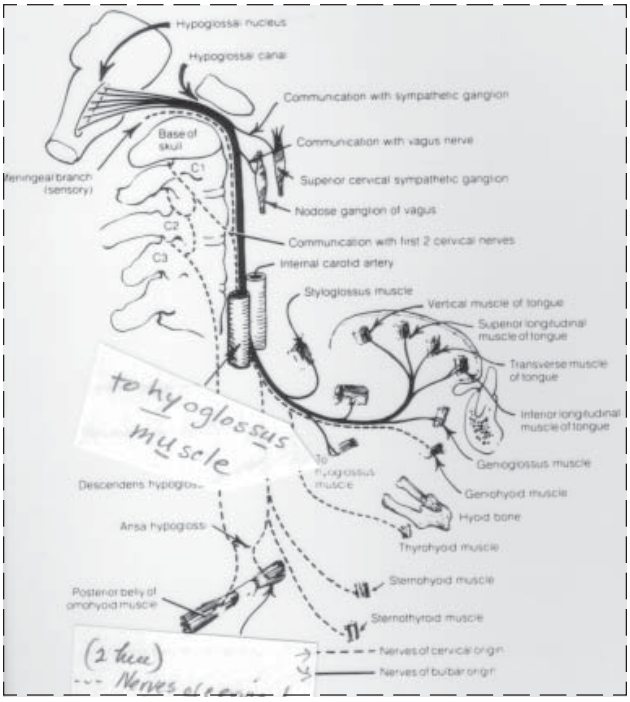

Fig. 7-38. The hypoglossal nerve. Reprinted with permission from deGroot J, Chusid JG. *Correlative Neuroanatomy.* 20th ed. Norwalk, Conn: Appleton & Lange; 1988: 175.

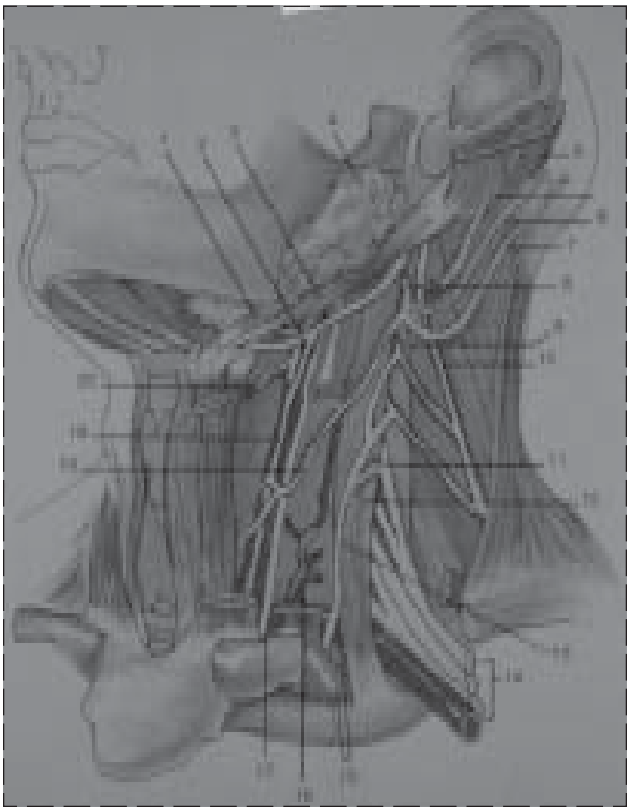

Fig. 7-39. Root of the neck showing location of the hypoglossal nerve (CN XII). 1: Stylohyoid muscle. 2: Cranial nerve XII. 3: Digastric muscle. 4: Parotid gland. 5: Sternocleidomastoid muscle. 6: Greater auricular nerve. 7: Lesser occipital nerve. 8: Ventral ramus C2. 9: Ventral ramus CC3. 10: Cranial nerve XI. 11: Ventral ramus C5. 12: Anterior scalene muscle. 13: Phrenic nerve. 14: Brachial plexus. 15: Subclavian artery and vein. 16: Thyrocervical trunk. 17: Cranial nerve X. 18: Inferior root ansa cervicalis. 19: Superior root ansa cervicalis. 20: Superior thyroid artery. Reprinted with permission from Maves MD. Surgical Anatomy of the Head and Neck. In: Byron E, Bailey J, eds. *Head & Neck Surgery: Otolaryngology*. Philadelphia, Pa: JB Lippincott; 1993: 20.

the hypoglossal nuclei in the nuclear and intramedullary area. Metastatic lesions invading the skull base may cause hypoglossal nerve lesions. Extracranial neurofibromas[225] or multiple sclerosis and syringobulbia may produce hypoglossal nerve lesions. Other nontraumatic causes are amyotrophic lateral sclerosis, poliomyelitis, and Dejerine's anterior bulbar syndrome. This is a rare syndrome that affects the hypoglossal nerve in its intramedullary course and is caused by occlusion of the anterior spinal artery. Still other nontraumatic lesions in the neck are carotid aneurysm and lesions of the parotid and other salivary glands, and the base of the tongue, with subsequent neck radiation.[226] Diseases such as myasthenia gravis, tuberculosis, and cerebral syphilis also must be considered. Common toxins that can interfere with hypoglossal nerve function are lead, alcohol, arsenic, and carbon monoxide. Certain tongue movement disorders may be induced by drugs or may be a form of seizure that injures the hypoglossal nerve.[227] Continuous lingual myoclonus may occur after head injury.[228]

Syndromes: Signs and Symptoms

Syndromes involving CN XII include Dejerine's anterior bulbar syndrome and Collet-Sicard syndrome.

Dejerine's anterior bulbar syndrome presents with ipsilateral paresis, atrophy, and fibrillations of the tongue. The protruded tongue deviates toward the side of the lesion. There also presents contralateral hemiplegia (pyramid involvement) with sparing of face, contralateral loss of position, and vibratory sense (medial lemniscus).

Collet-Sicard syndrome is manifested by paralysis of the trapezius and sternocleidomastoid muscles (CN XI); unilateral paralysis of vocal cord (CN X) and pharynx (CN IX); hemiparalysis of the tongue (CN XII); loss of taste on the posterior third of tongue (CN IX); and hemianesthesia of palate, pharynx and larynx (CN IX and X).[229]

Other syndromes involving lower cranial nerves are Jackson's syndrome (CN X, XI, and XII); Tapia's syndrome (CN X and XII); and Villaret's syndrome (CN IX, X, XI, and XII).

Signs and symptoms of CN XII lesions are secondary to weakness of the tongue. Specific observable signs and symptoms of CN XII lesions are listed below.

- Weakness and atrophy of the tongue (unilateral or bilateral depending on the lesion).
- Fibrillation of tongue (only in lower motor neuron lesions).
- Protruded tongue deviating to side of lesion (action of unopposed contralateral genioglossus muscle) in a lower motor neuron lesion.
- Nonprotruded tongue pointing to normal side (voluntary contraction, unopposed action of styloglossus).[230]
- Protruded tongue deviating to opposite side of lesion in an upper motor neuron lesion.
- Possible respiratory difficulties (in bilateral lesions, atrophic tongue slips back to throat).
- Dysarthria (difficulty with speech articulation, especially difficulty in pronouncing dental consonants). See Table 7-7.
- Possible Horner's syndrome (if cervical sympathetic trunk is interrupted in the region where the hypoglossal nerve emerges from the skull).
- Possible oral-buccal-lingual dyskinesia, athetosis, palatal myoclonus, tremor.

Evaluation

Clinical examination consists of observation of the tongue at rest, looking for fasciculations (motor neuron disease), atrophy, or furrowing. The tongue is then observed during movement within the mouth for intrinsic tongue muscle activity, and protruding from the mouth for extrinsic tongue muscle activity.

The strength of tongue muscles is assessed by providing resistance to the protruded tongue. Quickness of tongue movement is evaluated as well as the articulation of speech and tongue movements during eating. The paralyzed tongue has difficulty moving bolus toward the pharynx and from side-to-side in the preparatory phase of swallowing. Breathing must also be observed, to rule out obstruction of the pharynx with a paralyzed tongue.

Electrodiagnosis

Electrodiagnostic evaluation can be performed by nerve conduction studies[231] and EMG. The procedure for nerve conduction is as follows.

Stimulation of the hypoglossal nerve is conducted anterior and inferior to the corner of the mandible (one third the distance from the angle of the jaw to

TABLE 7-7

CRANIAL NERVE LESION AS CAUSE OF DYSARTHRIA

Involved CN	Laryngeal (Phonatory)	Velopharyngeal	Oral
XII			
Flaccid	Normal	Normal	Unilateral or bilateral atrophic weak tongue, fasciculations, decreased range-of-motion of tongue, drooling, imprecision of vowels and lingual consonants
X			
Flaccid	Hoarseness, breathiness, low volume, diplophoniae	Hypernasally nasal mission, if lesion is above pharyngeal branch	Normal
VII			
Flaccid	Normal	Normal	Unilateral or bilateral weak orbicularis oris, reduced range-of-motion in lips and cheek, imprecision of vowels and labial consonants
V			
Flaccid	Normal	Normal	Mandibular muscles weakness, loss of tongue sensation with lingual nerve involvement, imprecision of vowels and consonants
Multiple CN			
Flaccid	Breathiness, reduced volume, inhalatory, stridor, monopitch	Hypernasally, nasal emission	Imprecision of vowel and consonants
Bilateral Upper Motor Neuron			
Spastic	Hyperadduction of vocal folds, strained, strangled voice, hoarseness, low pitch, monopitch	Incompleteness of palatopharyngeal closure, hypernasality	Slowness, weakness of tongue and mimetic muscles, imprecision of consonants

CN: cranial nerve
Reprinted with permission from Aronson AE. Motor speech signs of neurologic disease. In: Darby JK Jr. *Speech Evaluation in Medicine.* New York: Grune & Stratton; 1981: 159–180.

the mental protuberance along the base of the mandible and 1 cm medial).

Recording is carried out with superficial electrodes (a bar embedded and placed on top of the tongue).

Normal values:

- Latency: 2.2 ± 0.4 millisecond
- Amplitude: 3.8 ± 1.6 mV.

EMG of tongue is carried out through direct insertion of electrodes into the tongue or into the mylohyoid muscle, through the skin beneath the chin. The examiner looks for possible evidence of denervation, action potential parameter abnormalities, reduced recruitment, or myoclonus.

Imaging and Prognosis

Imaging the skull base with CT, MRI, or both, is helpful. The prognosis for recovery is better for upper motor neuron lesions than for lower motor neuron lesions.

Management

Management of hypoglossal nerve lesions is assisted by speech therapy, using strengthening ex-

ercises for the tongue to facilitate speech and improve the oral phase of swallowing. See the discussion of CN IX for therapeutic interventions to improve the oral phase of swallowing.

Severe involvement makes tube feeding or gastrostomy necessary. Tracheostomy or a surgical procedure may be required to promote efficient breathing.

CONCLUSION

In this chapter, each of the 12 cranial nerves has been considered in detailed account as a single anatomical and functional entity. The signs and symptoms of deficits following peripheral and central injuries and lesions have been described and evaluated. Syndromes were tabulated and briefly discussed. Injuries and lesions of the head and neck frequently display conjoint nerve involvement due to the close proximity or origin of central tracts and nuclei of termination and the close anatomical intracranial and peripheral course of one nerve path with others. Such injuries and lesions then present a complexity of deficits, each attributable to the specific nerve (or nucleus or tract) involved. Keeping in mind that branches of nerves attach onto branches of other nerves and may course over several nerves from sensory origin to motor destination, such vulnerability and subsequent inclusion is far more frequent in injuries than single nerve involvement. Therefore, within the discussion of each cranial nerve as an entity, pertinent multiple involvement has been discussed.

In addition to more routine methods of examination and diagnosis, the use of the most current tools of electrodiagnosis and imaging have been emphasized. Techniques and procedures have been tabulated concisely. These sophisticated techniques offer advantages for use in surgical procedures in neuromonitoring as precautionary measures.

Lastly, prognosis for the various conditions and syndromes is integral to and supports the discussion of management and rehabilitative measures.

REFERENCES

1. Maclean JG, Taylor A. Combined lower cranial nerve injury: Complication of upper cervical or basal skull fracture. *J R Coll Surg Edinb.* 1991;36:188–189.

2. Hammer AJ. Lower cranial nerve palsies: Potentially lethal in association with upper cervical fracture dislocation. *Clin Orthop.* 1991;266:64–69.

3. Edmund J, Godtfredsen E. Unilateral optic atrophy following head injury. *Acta Ophthalmol.* 1963;41:693.

4. Roberts AH. *Severe Accidental Head Injury.* New York: Macmillan; 1979.

5. Russell WR. Injury to cranial nerves including the optic nerves and chiasma. In: Brock S, ed. *Injuries of the Skull, Brain, and Spinal Cord.* 2nd ed. Baltimore, Md: Williams & Wilkins; 1943:104.

6. Costanzo RN, Becker DFP. Sense of smell and taste disorders in head injury and neurosurgery patients. In: Meiselman HL, Rivlin RS, eds. *Clinical Measurement of Taste and Smell.* New York: Macmillan; 1986: 565–578.

7. Kitchins JL, Groff DB, Nagaraj HS, Fallat ME: Basilar skull fractures in childhood with cranial nerve involvement. *J Pediatr Surg.* 1991;26(8):992–994.

8. Friedman AP, Merritt HH. Damage to cranial nerves resulting from head injury. *Bull Los Angeles Neurol Soc.* 1944;9:135–139.

9. Hughes B. The results of injury to special parts of the brain and skull: The cranial nerves. In: Rowbotham GR, ed. *Acute Injuries of the Head.* Edinburgh, Scotland: Livingston; 1964: 410.

10. Dobson JE, Newell MJ, Shepherd JP: Trends in maxillofacial injuries in wartime (1914–1986). *Br J Oral Maxillofac Surg.* 1989;27:441–450.

11. Keane JR, Baloh RW. Post-traumatic cranial neuropathies. *Neurol Clin.* 1992;10(4):853.

12. Jacobi G, Ritz A, Emrich R. Cranial nerve damage after pediatric head trauma: A long term follow up study of 741 cases. *Acta Paediatr Hung.* 1986;27:173–187.

13. Harner SG, Danbe JR, Beatly CW, Ebersvold MJ. Intraoperative monitoring of the facial nerve. *Laryngoscope.* 1988;98:209–212.

14. Moller AR. Neuromonitoring in operations in the skull base. *KEIO J Med.* 1991;40(3):151–159.

15. Nelson KR, Phillips LH. Neurophysiologic monitoring during surgery of peripheral and cranial nerves and in selective dorsal rhizotomy. *Semin Neurol.* 1990;10(2):141–149.

16. Ruth RA. Neurophysiologic intraoperative monitoring. In: Bailey BJ, ed. *Head and Neck Surgery: Otolaryngology.* Philadelphia, Pa: JB Lippincott; 1993:1505–1516.

17. Sekhar LN, Lanzino G, Sen CN, Pomonis S. Reconstruction of the third through sixth cranial nerves during cavernous sinus surgery. *J Neurosurg.* 1992;76:935–943.

18. Ransom DT, Dinapoli RP, Richardson RL. Cranial nerve lesions due to base of the skull metastases in prostate carcinoma. *Cancer.* 1990;65(3):586–589.

19. Sumner D. Post traumatic anosmia. *Brain.* 1964;87:107–120.

20. Jennett B, Teasdale G, eds. *Management of Head Injuries.* Philadelphia, Pa: FA Davis; 1981: 272–278.

21. Meacy J. *Pediatric Head Injuries.* Springfield, Ill: Charles C Thomas; 1968: 88–136.

22. Bakay L. Olfactory meningioma, the missed diagnosis. *JAMA.* 1984;251:53–55.

23. Doty RL, Bartoshuk LM, Snow JB. Causes of olfactory and gustatory disorders. In: Getchell TV, Doty RL, Bartoshuk LM, Snow JB Jr, eds. *Smell and Taste in Health and Disease.* New York: Raven Press; 1991: 451.

24. Jones BP, Butter NM, Moskowitz HR, Montgomery K. Olfactory and gustatory capacities of alcoholic Korsakoff patients. *Neuropsychologia.* 1988;16:323.

25. Moberg PF, Pearlson GD, Speedie LJ, Lipsey JR, Strauss ME, Folstein SE. Olfactory recognition: Differential impairments in early and late Huntington's and Alzheimer's diseases. *J Clin Exp Neuro Psych.* 1987;9:650–664.

26. Warner MD, Peabody CA, Flattery JJ, et al. Olfactory deficits and Alzheimer's disease. *Biol Psychiatry.* 1986;21:116.

27. Schatz, NJ, Smith JL. Nontumor causes of the Foster Kennedy Syndrome. *J Neurosurg.* 1948;40:245.

28. Doty RL, Riklan M, Deems DA, Reynolds C, Stellar S. The olfactory and cognitive deficits of Parkinson's disease: Evidence for independence. *Ann Neurol.* 1989;25:166–171.

29. Schiffman SS. Age related damages in taste and smell and their possible causes. In: Meiselman HL, Rivlin RS, eds. *Clinical Measurement of Taste and Smell.* New York: Macmillan; 1986: 326–342.

30. Deems DA, Doty RL, Settle RG, et al. Smell and taste disorders: A study of 750 patients from the University of Pennsylvania smell and taste center. *Arch Otolaryngol Head Neck Surg.* 1991;117:519–528.

31. Schweitzer VG. Osteolytic sinusitis and pneumomediastinum: Deceptive otolaryngologic complications of cocaine abuse. *Laryngoscope.* 1986;96:206–210.

32. Levin HS, High WM, Elsenberg HM. Impairment of olfactory recognition after closed head injury. *Brain.* 1985;108:579–591.

33. Caruso V, Hagan J, Manning H. Quantitative olfactometry in measurement of post traumatic hyposmia. *Arch Otolaryngol.* 1968;90:500.

34. Cain WS, Gent JF, Goodspeed RB, Leonard G. Evaluation of olfactory dysfunction in the Connecticut chemosensory clinical research center. *Laryngoscope*. 1988;98:83–88.

35. Costanzo RM, Zasler ND. Head trauma. In: Getchell TV, Doty RL, Bartoshuk LM, Snow JB Jr, eds. *Smell and Taste in Health and Disease.* New York: Raven Press; 1991: 715.

36. Kobal G, Hummel T. Olfactory evoked potentials in humans. In: Getchell TV, Doty RL, Bartoshuk LM, Snow JB Jr, eds. *Smell and Taste in Health and Disease.* New York: Raven Press; 1991: 255–275.

37. Kobal G. *Elektrophysiologische Untersuchungen Des Menschlichen Geruchsinns.* Stuttgart: Thieme; 1981.

38. Zasler ND, Costanzo RM, Heywood PG. Neuroimaging correlates of olfactory dysfunction after traumatic brain injury. *Arch Phys Med Rehabil.* 1990;71:814.

39. Costanzo RM. Neural-regeneration and functional reconnection following olfactory nerve transsection in hamster. *Brain Res.* 1985;361:258–266.

40. American Medical Association. *Guides to the Evaluation of Permanent Impairment.* Chicago, Ill: AMA; 1988.

41. Dimond SJ, Farrington L, Johnson P. Differing emotional response from right and left hemisphere. *Nature.* 1976;261:690–692.

42. Shokunbi T, Agbeja A. Ocular complications of head injuries in children. *Childs Nerv Syst.* 1991;7:147–149.

43. LaGrange F. Fractures of the orbit and injuries to the eye in war. *Adv Ophthal Plast Reconstr Surg.* 1986;5:169–176.

44. LaGrange F. Fractures of the orbit with preservation of the eyeball. *Adv Ophthal Plast Reconstr Surg.* 1987;6:145–192.

45. Newman NM, Di Loreto DA, Ho JT, Klein JC, Birnbaum NS. Bilateral optic neuropathy and osteolytic sinusitis: Complications of cocaine abuse. *JAMA.* 1988;259:72–74.

46. Harriman DG, Garland H. The pathology of Adie's syndrome. *Brain.* 1968;91:401–418.

47. Zihl J, Von Cramon D. Restitution of visual function in patients with cerebral blindness. *J Neurol Neurosurg Psychiatry.* 1979;42:312–322.

48. Ross Russell RW, Bharucha N. Visual localization in patients with occipital infarction. *J Neurol Neurosurg Psychiatry.* 1984;47:153–158.

49. Spector RH, Glaser JS, David NJ, Vining DQ. Occipital lobe infarctions: Perimetry and computed tomography. *Neurology.* 1981;31:1098–1106.

50. Hennekes R. Clinical electroretinography. *Fortschr Ophthalmol*. 1989;86:146–150.

51. Chiappa KH. Pattern-shift visual evoked potentials. In: Chiappa KH, ed. *Evoked Potentials in Clinical Medicine.* 2nd ed. New York: Raven Press; 1989: 37–155.

52. Hughes B. Indirect injury of the optic nerves and chiasma. *Bull Johns Hopkins Hosp.* 1962;111:98–126.

53. Geisler FH, Dorsey FC, Coleman WP. Recovery of motor function after spinal cord injury: A randomized, placebo controlled trial with GM-1 ganglioside. *N Engl J Med.* 1991;324:1829–1838.

54. Spooner JW, Sakala SM, Baloh RW. Effect of aging on eye tracking. *Arch Neurol.* 1980;37:57–59.

55. Nadeau SE, Trobe JD. Pupil sparing in ocularmotor palsy: A brief review. *Ann Neurol.* 1983;13:143.

56. Warren M. A hierarchical model for evaluation and treatment of visual perceptual dysfunction in adult aquired brain injury. *Am J Occup Ther.* 1993;47:42–65.

57. Russell WR. Injury to cranial nerves and optic chiasm. In: Brock S, ed. *Injuries of the Brain and Spinal Cord and Their Coverings.* 4th ed. Baltimore, Md: Williams & Wilkins; 1960.

58. Kontroupas S, Meyerhoff NL. Surgical treatment of orbital floor fractures. *Arch Otolaryngol.* 1982;108:184–186.

59. DiNubile MJ. Septic thrombosis of the cavernous sinus. *Arch Neurol.* 1988;45:567.

60. Donaldson D, Rosenberg NL. Infarction of abducens nerve fasicle as cause of isolated sixth nerve palsy related to hypertension. *Neurology.* 1988;38:1654.

61. Carter S, Sciarra D, Merritt HH. The course of multiple sclerosis as determined by autopsy proven cases. *Res Publ Assoc Res Nerv Ment Dis*. 1950;28:471.

62. Spector RH, Smith JL, Chavis PS. Charcot-Marie-Tooth disease mimicking ocular myasthenia gravis. *Ann Ophthalmol.* 1978;10:1033–1038.

63. Hyams SW. Oculomotor palsy following dental anesthesia. *Arch Ophthalmol.* 1976;94:1281.

64. Masdeu J, Brazis PW. The localization of lesions in the ocularmotor system. In: Brazis PW, Masdeu JC, Biller J, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 140–143.

65. Yaskin JC, Kornblum K. Neurologic aspects of petrositis. *Arch Neurol Psychiat.* 1937;37:307–333.

66. Keane JR, Zaias B, Itabashi HH. Levator sparing oculomotor nerve palsy caused by a solitary midbrain metastasis. *Arch Neurol.* 1984;41:210–212.

67. Conway VH, Rozdilsky B, Schneider RJ, Sundaram M. Isolated bilateral complete ptosis. *Can J Ophthalmol.* 1983;18:37–40.

68. Growden JH, Winkler GF, Wray SH. Midbrain ptosis. A case with clinico-pathologic correlation. *Arch Neurol.* 1974;30:179.

69. Leigh RJ, Zee DS. *The Neurology of Eye Movement.* 2nd ed. Philadelphia, Pa: FA Davis; 1991: 111.

70. Smith JL, David NJ, Klindworth G. Skew deviation. *Neurology.* 1964;14:96.

71. Neger RE. The evaluation of diplopia in head trauma. *J Head Trauma Rehabil.* 1989;4:27–34.

72. Warren M. Identification of visual scanning deficits in adults after cerebrovascular accident. *Am J Occup Ther.* 1990;44:391–399.

73. VonNoorden GK. *Burian-VonNoorden's Binocular Vision in Ocular Motility.* 2nd ed. St. Louis, Mo: Mosby; 1980.

74. Thompson BM, Corbett JJ, Kline LB, Thompson HS. Pseudo Horner's syndrome. *Arch Neurol.* 1982;39:108–111.

75. Sydnor CF, Seaber JH, Buckley EG. Traumatic superior oblique palsies. *Ophthalmology (Rochester).* 1982;89: 134–138.

76. Frank JW, Kushner BJ, France TD. Paradoxic pupillary phenomena: A review of patients with pupillary constriction to darkness. *Arch Ophthalmol.* 1988;106:1564–1566.

77. Bouska MJ, Kauffman MA, Marcus S. Disorders of the visual perceptual system. In: Umphred DA, ed. *Neurological Rehabilitation.* Vol 4. St. Louis, Mo: Mosby; 1990.

78. Cytowic R, Stump DA, Larned DC. Closed head trauma: Somatic, ophthalmic and cognitive impairment in nonhospitalized patients. In: Whitaker HA, ed. *Neuropsychological Studies of Nonfocal Brain Damage.* New York: Springer-Verlag; 1990.

79. Troost BT. Neuro-otology. In: Bradley WG, Daroff RB, Feinchel GM, Marsden CD, eds. *Neurology and Clin Practice.* Vol 1. Woburn, Mass: Butterworth-Heineman; 1991: 655.

80. Berrol S. Cranial nerve dysfunction. In: *Physical Medicine and Rehabilitation*: *State of the art reviews.* 1989;3:89.

81. Zasler ND, McClintic N. Functional orthoptic evaluation after traumatic brain injury. Presented at the Postgraduate Course on Rehabilitation of the Brain Injury in Adult and Child; June 1991; Williamsburg, Va.

82. Trobe JD, Acosta PC, Krischer JP, Trick GL. Confrontation visual field techniques in the detection of anterior visual pathway lesions. *Ann Neurol.* 1981;10:28–34.

83. Silver WL, Finger TE. The trigeminal system. In: Getchell TV, Doty RL, Bartoshuk LM, Snow JB Jr, eds. *Smell and Taste in Health and Disease.* New York: Raven Press; 1991: 97.

84. Greenward HS Jr, Keeney AH, Shannon GM. A review of 128 patients with orbital fracture. *Am J Ophthalmol* 1974;78:655.

85. Yadav YR, Khosla VK. Isolated 5th-10th cranial nerve palsy in closed head trauma. *Clin Neurol Neurosurg.* 1991;93:61–63.

86. Gonzalez Revilla A. Differential diagnosis of tumors at the cerebello-pontine recess. *Bull Johns Hopkins Hosp.* 1989,83:187–212.

87. Blackburn CW, Bramley PA. Lingual nerve damage associated with the removal of lower 3rd molars. *Br Dent J.* 1989;167:103–107.

88. Rushton JG, Olafson RA. Trigeminal neuralgia associated with multiple sclerosis. Report of 35 cases. *Arch Neurol.* 1965;13:383–386.

89. Jannetta PJ, Rand RW. Arterial compression of trigeminal nerve at the pons in patients with trigeminal neuralgias. *J Neurosurg.* 1967;26(Suppl):159.

90. Ashworth B, Tait GBW. Trigeminal neuropathy in connective tissue disease. *Neurology*. 1971;21:609–614.

91. Caselli RJ, Hunder GG, Whisnant JP. Neurologic disease in biopsy-proven giant cell (temporal) arteritis. *Neurology.* 1988;38:352–359.

92. Brazis PW. The localization of lesions effecting cranial nerve V. In: Brazis PW, Masdeu JC, Biller JL, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 197.

93. Loeser JD. Cranial neuralgias. In: Bonica JJ, ed. *The Management of Pain*. 1990: 676–686.

94. Lance JW, Anthony M. Neck-tongue syndrome on sudden turning of the head. *J Neurol Neurosurg Psychiatry.* 1980;43:97–101.

95. Campbell WW. The numb cheek syndrome: A sign of infraorbital neuropathy. *Neurology.* 1986;36:421.

96. Mountcastle VB, Talbot WH, Darian-Smith I, Kornhuber HH. Neural basis the sense of flutter-vibration. *Science.* 1967;155:597–600.

97. Dellon AL. The moving two-point discrimination test. Clinical evaluation of the quickly adapting fibre/receptor system. *J Hand Surg.* 1978;3:474–481.

98. Ongerboer DE, Visser BW, Goor C. Jaw reflexes and masseter electro-myograms in mesencephalic and pontine lesions. An electrodiagnostic study. *J Neurol Neurosurg Psychiatry.* 1976;39:90–92.

99. Godoux E, Desmedt GE. Exteroceptive supression and motor control of the masseter and temporalis muscles in normal man. *Brain Res.* 1975;85:447–458.

100. Ongerboer DE, Visser BW, Goor C. Cutaneous silent period in masseter muscles: A clinical and electrodiagnostic evaluation. *J Neurol Neurosurg Psychiatry.* 1976;39:674–679.

101. Ongerboer DE, Visser BW, Cruccu G, Manfredi M, Koelman JH. Effects of brainstem lesions on the masseter inhibitory reflex. Functional mechanism of reflex pathways. *Brain.* 1990;113:781–792.

102. Cruccu G, Pauletti G, Agostino R, Berardelli A, Manfredi M. Masseter inhibitory reflex in movement disorders. Huntington's chorea, Parkinson's disease, dystonia, and unilateral masticatory spasm. *Electroencephalogr Clin Neurophysiol.* 1991;81:24–30.

103. Hopf HC, Ellrich J, Hundermer H. *The Pterygoid Reflex in Man and its Clinical Application Muscle and Nerve.* 1992;15:1278–1283.

104. Kimura J, Powers JM, Van Allen MW. Reflex response of orbicularis oculi muscle to supraorbital nerve stimulation: Study in normal subjects and in peripheral facial paresis. *Arch Neurol.* 1969;21:193–199.

105. Kimura J. Conduction abnormalities of the facial and trigeminal nerves in polyneuropathy. *Muscle Nerve.* 1982;5:139–144.

106. Kimura J, Bodensteiner J, Yamada T. Electrically elicited blink reflex in normal neonates. *Arch Neurol.* 1977;34:246–249.

107. Accornero N, Berardelli A, Cruccu G, Bini G, Manfredi M. Corneal reflex elicited by electrical stimulation of human cornea. *Neurology*. 1980;30:782–785.

108. Di Benedetto M. Trigeminal sensory nerve conduction. Unpublished data.

109. Stohr M, Petruch F. Somatosensory evoked potentials following stimulation of the trigeminal nerve in man. *J Neurol.* 1979;220:95–98.

110. Dillingham TR, Spellman NT, Belandres PV. Trigeminal motor nerve conduction: Deep temporal and mylohyoid nerves. *Muscle Nerve.* 1993;16:1115.

111. Jannetta PJ. Microsurgical approach to the trigeminal nerve for tic douloureux. *Prog Neurosurg*. 1976;7:180–200.

112. Roberts AM, Person P. Etiology and treatment of idiopathic trigeminal and atypical facial neuralgias. *Oral Surg.* 1979;48:298–308.

113. Brazis PW. The localization of lesions affecting cranial nerve VII (the facial nerve). In: Brazis PW, Masdeu JC, Biller JL, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 206–207.

114. Bartoshuk LM, Kveton JF, Karrer T. Taste. In: Bailey BJ, ed. *Head and Neck Surgery: Otolaryngology.* Philadelphia, Pa: JB Lippincott; 1993: 520–529.

115. Jenny AB, Saper CB. Organization of the facial nucleus and cortico-facial projection in the monkey: A reconsideration of the upper motor neuron facial palsy. *Neurology.* 1987;37:930–939.

116. Borod JC, Koff E, Lorch MP, Nicholas M, Welkowitz J. Emotional and nonemotional facial behaviour in patients with unilateral brain damage. *J Neurol Neurosurg Psychiatry.* 1988;51:826–832.

117. Sumner D. Post traumatic ageusia. *Brain.* 1967;90:187–202.

118. Costanzo RM, Zasler ND. Epidemiology and pathophysiology of olfactory and gustatory dysfunction in head trauma. *J Head Trauma Rehabil.* 1992;7:15–24.

119. Schechter PJ, Henkin RI. Abnormalities of taste and smell after head trauma. *J Neurol Neurosurg Psychiatry*. 1974;37:802–810.

120. Fisch U. Facial paralysis in fractures of the petrous bone. *Laryngoscope.* 1974;84:2141–2154.

121. Miller H. Facial paralysis. *Br Med Bull.* 1967;3:815.

122. Park HW, Watkins AL. Facial paralysis: Analysis of 500 cases. *Arch Phys Med.* 1949;30:749–762.

123. Karnes WE. Diseases of the seventh cranial nerve. In: Dyck PJ, Thomas PK, Lambert EH, eds. *Peripheral Neuropathy.* Philadelphia, Pa: WB Saunders; 1975: 570–603.

124. Brown JW. Physiology and phylogenesis of emotional expression. *Brain Res.* 1967;5:1–14.

125. Hanson MR, Sweeney PJ. Lower cranial neuropathies. In: Bradley WG, Daroff RB, Feinchel GM, Marsden CD, eds. *Neurology and Clinical Practice.* Vol 1. Woburn, Mass: Butterworth-Heineman; 1991: 220.

126. Frank ME, Smith EV. Electrogustometry. In: Getchell TV, Doty RL, Bartoshuk LM, Snow JB Jr, eds. *Smell and Taste in Health and Disease.* New York: Raven Press; 1991: 503–514.

127. Plattig KH. Gustatory evoked brain potentials in humans. In: Getchell TV, Doty RL, Bartoshuk LM, Snow JB, eds. *Smell and Taste in Health and Disease*. New York: Raven Press; 1991;14:227–286.

128. Dumitru D, Walsh NE, Porter LD. Electrophysiologic evaluation of the facial nerve in Bell's palsy. A review. *Am J Phys Med Rehabil.* 1988;137–143.

129. Mitz M, Prakash AS, Melvin J, Piering W. Motor nerve conduction indicators in uremic neuropathy. *Arch Phys Med Rehabil.* 1980;61:45–48.

130. Jannetta PJ, Abbasy M, Maroon JC, Ramos FM, Albin MS. Etiology and definitive microsurgical treatment of hemifacial spasm. *J Neurosurg.* 1977;47:321–328.

131. Granger C. Prognosis in Bell's palsy. *Arch Phys Med Rehabil.* 1976;61:45–48.

132. Diamant H, Kirkstrand T, Wiberg A. Prognosis of idiopathic Bell's palsy. *Arch Otolaryngol.* 1972;95:431.

133. Drachman DA. Bell's palsy: A neurological point of view. *Arch Otolaryngol.* 1969;89:173.

134. El-Ebiary HM. Prognosis in Bell's palsy. *Rheumatol Phys Med.* 1971;11:111.

135. Langworth EP, Taverner D. The prognosis in facial palsy. *Brain*. 1963;86:465–480.

136. Schaumburg HH. Diseases of the peripheral nervous system. In: Wyngaarden JB, Smith LH, eds. *Cecil Text-book of Medicine.* Philadelphia, Pa: WB Saunders; 1988: 2267.

137. Crumley R, Robinson L. Traumatic facial nerve injury. In: Gates GG, ed. *Current Therapy. Head and Neck Surgery*. Philadelphia, Pa: BC Decker; 1990: 97–101.

138. Pitty LF, Tator CH. Hypoglossal-facial nerve anastomosis for facial nerve palsy following surgery for cerebellopontine angle tumors. *J Neurosurg.* 1992;77:724–731.

139. Wilson-Pawels L, Akesson EJ, Stewart PA. Cranial nerves. In: *Anatomy and Clinical Comments.* Hamilton, Ontario, Canada: BC Decker; 1988: 104.

140. Sakai CS, Mateer CA. Otological and audiological sequalae of closed head trauma. *Semin Hearing.* 1984;5:157–173.

141. Barber HO. Positional nystagmus especially after head injury. *Laryngoscope.* 1964;74:891–944.

142. Barber HO. Head injury: Audiological and vestibular findings. *Ann Otol Rhinol Laryngol.* 1969;78:239–252.

143. Berman J, Fredrickson J. Vertigo after head injury: A 5-year follow up. *J Otolaryngol.* 1978;7:237.

144. Healy GB. Hearing loss and vertigo secondary to head injury. *N Engl J Med.* 1982;306:1029.

145. Clark K, Rees TS. Post traumatic endolymphatic hydrops. *Arch Otolaryngol* 1983;103:1009–1012.

146. Vernon J, Northern JL, eds. *Tinnitus. Hearing Disorders.* Boston: Little, Brown; 1984.

147. Clark J, Yanick P. *Tinnitus and its Management.* Springfield, Ill: Charles C Thomas; 1984.

148. Schein JD, Miller MH. Diagnosis and rehabilitation of auditory disorders. In: Kottke FJ, Lehmann JF, eds. *Krusen's Handbook of Physical Medicine and Rehabilitation.* Philadelphia, Pa: WB Saunders; 1990: 953.

149. Tuohimaa P. Vestibular disturbances after acute mild head injury. *Acta Otolaryngol.* 1979;359(Suppl):1–67.

150. Windle WF, Groat RA, Fox CA. Experimental structural alterations in the brain during and after concussion. *Surg Gynecol Obstet.* 1944;79:561–572.

151. Griffiths MV. The incidence of auditory and vestibular concussion following minor head injury. *J Laryngol Otol.* 1979;9:253–265.

152. Masdeu JC, Brazis PW. The localization of lesions in the oculomotor system. In: Brazis PW, Masdeu JC, Biller J, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 172.

153. Troost BT. Neuro-otology. In: Bradley WG, Daroff RB, Feinchel GM, Marsden CD, eds. *Neurology and Clinical Practice.* Vol 1. Woburn, Mass: Butterworth-Heineman; 1991: 657.

154. Chiappa KH. Brainstem auditory evoked potentials. Methodology. In: Chiappa KH, ed. *Evoked Potentials in Clinical Medicine,* 2nd ed. New York: Raven Press; 1989: 174–220.

155. Bergamasco BP, Gilli BC, Ferrero P, et al. Brainstem auditory evoked potentials in postconcussion syndromes. *Ital J Neurol Sci.* 1986;4:291–297.

156. Montgomery A, Penton GW, McClelland RJ. Delayed brainstem conduction time in post concussional syndrome. *Lancet.* 1984;1:1011.

157. Shulman A. External electrical stimulation in tinnitus control. *Am J Otolaryngol.* 1985;6:110–115.

158. Karnick PP, Kirtane MV, Wagh SP, et al. Otoneurologic problems in head injuries and their management. *Int Surg.* 1975;60:466.

159. Berger KW. *Speech Reading Principles and Methods.* Baltimore, Md: National Education Press; 1972.

160. Shumway-Cook A, Horak F. Rehabilitation strategies for patients with vestibular deficits. *Neurol Clin.* 1990;8:441.

161. Igarashi M, Ishixawa M, Yamane HI. Physical exercise and balance compensation after total ablation of vestibular organs. *Prog Brain Res.* 1988;76:395–401.

162. Herdman SJ. Treatment of vestibular disorders in traumatically brain injured patients. *J Head Trauma Rehabil.* 1990;5:63.

163. Hunt JR. Geniculate neuralgia: A further contribution to the sensory system of the facial nerve and its neuralgic conditions. *Arch Neurol Psychiatry.* 1937;37:253–285.

164. Kanagasuntheram R, Wong WC, Chan HL. Some observations on the innervation of the human nasopharynx. *J Anat.* 1969;104:361.

165. Mohanty SK, Barrios M, Fishbone H, Khatib R. Irreversible injury of cranial nerve IX–XII (Collet-Sicard Syndrome). *J Neurosurg.* 1973;30:86–88.

166. Russell JR, Buct PC. Meningiomas of the posterior fossa. *Surg Gynecol Obstet.* 1953;96:183.

167. Demir R, Steegmann AT. Intracranial chordoma. *Neurology*. 1959;9:514.

168. Henson RA, Crawford JV, Cavanagh JB. Tumours of the glomus jugulare. *J Neurol Neurosurg Psychiatry.* 1953;16:127–138.

169. Rushton JG, Stevens JC, Miller RH. Glossopharyngeal (vagoglossopharyngeal) neuralgia. *Arch Neurol.* 1981;38:201.

170. Laha RK, Jannetta PJ. Glossopharyngeal neuralgia. *J Neurosurg.* 1977;47:316.

171. Brazis PW. The localization of lesions affecting cranial nerves IX and X (the glossopharyngeal and vagus nerves). In: Brazis PW, Masdeu JC, Biller JL, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 240–248.

172. Sumi T. Role of the pontine reticular formation in the neural organization of deglutition. *Japanese J Physiol.* 1972;22:295–314.

173. Siebens AA. Rehabilitation for swallowing impairment. In: Kottke FJ, Lehmann JE, eds. *Handbook of Physical Medicine and Rehabilitation*, 4th ed. Philadelphia, Pa: WB Saunders; 1990: 765–776.

174. Logemann J. *Evaluation and Treatment of Swallowing Disorders.* San Diego, Calif: Coll-Hill Press; 1983: 129–215.

175. Shanahan TK, Logemann JA, Rademaker AW, Palkoski BR, Kahrilas PJ. Chin-down posture effect on aspiration in dysphagic patients. *Arch Phys Med Rehabil.* 1993;74:736–739.

176. Sessions D, Zill R, Schwartz J. Deglutition after conservation surgery for cancer of the larynx and hyperpharynx. *Otolaryngol Head Neck Surg.* 1979;87:779–796.

177. Mitchell R, Armanini G. Cricopharyngeal myotomy: Treatment of dysphagia. *Ann Surg.* 1975;181:262–266.

178. Cherney LR, Halper AS. Recovery of oral nutrition after head injury in adults. *J Head Trauma Rehabil.* 1989;4:42.

179. Winstein CJ. Neurogenic dysphagia: Frequency, progression and outcome in adults following head injury. *Phys Ther.* 1983;12:1992.

180. Adour KK. Diagnosis and management of facial paralysis. *N Engl J Med.* 1982;307:348–351.

181. Yamada M, Hirano M, Ohkubo H. Recurrent laryngeal nerve paralysis: A 10-year review of 564 patients. *Auris Nasus Larynx*. 1983;10(Suppl):1–15.

182. Collet FJ. Surg un nouveau syndrome paralytique pharyngolarynge par blessure de guerre. *Lyon Med.* 1916;124:121–129.

183. Horsely J. Suture of the recurrent laryngeal nerve with reports of a case. *Trans South Surg Gynecol Assoc.* 1909;22:161.

184. Woodson GE, Miller RH. The timing of surgical intervention in vocal cord paralysis. *Otolaryngol Head Neck Surg.* 1981;89:264.

185. Holinger LD, Holinger PC, Holinger PH. Etiology of bilateral abductor vocal cord paralysis. *Ann Otol.* 1976;85:428.

186. Blackburn G, Salmon LF. Cord movements after thyroidectomy. *Br J Surg.* 1960;48:371.

187. Clark OH. Total thyroidectomy: The treatment of choice for patients with differentiated thyroid cancer. *Ann Surg.* 1982;196:361–370.

188. Clark OH. *Endocrine Surgery of the Thyroid and Parathyroid Glands.* St. Louis, Mo: Mosby; 1985.

189. Palmar ED. Disorders of the cricopharyngeus muscle: A review. *Gastroenterology*. 1976;71:510.

190. Peytz F, Rasmussen H, Buchtal F. Conduction time and velocity in human recurrent laryngeal nerve. *Dan Med Bull*. 1965;12:125–127.

191. Blair RL. Laryngeal electromyography: Techniques and application. *Otolaryngol Clin North Am*. 1978;11:2.

192. Simpson DM, Sternman D, Graves-Wright J, Sanders I. Vocal cord paralysis. Clinical and electrophysiologic features. *Muscle Nerve.* 1993;16:952–957.

193. Hiroto I, Hirano M, Tomita H. Electromyographic investigation of human vocal cord paralysis. *Am Otol Rhinol Laryngol.* 1968;77:246–304.

194. Simpson DM, Kaufmann H, Sanders I, Wolfe DE. Laryngeal dystonia in multiple system atrophy. *Muscle Nerve.* 1992;15:1213–1215.

195. Rodriguez A, Myers B, Ford C. Laryngeal electromyography in the diagnosis of laryngeal nerve injury. *Arch Phys Med Rehabil.* 1990;71:587–590.

196. Tanaka E, Palmer J, Siebens A. Bipolar suction electrodes for pharyngeal electromyography. *Dysphagia.* 1986;1:39–40.

197. Palmer JP, Holloway AM, Tanaka E. Detecting lower motor neuron dysfunction of the pharynx and larynx with electromyography. *Arch Phys Med Rehabil.* 1991;72:214–219.

198. Eckberg DL. Parasympathetic cardiovascular control in human disease. A critical review of methods and results. *Am J Physiol.* 1980;234:581–593.

199. Dedo HH, Izdebski K. Intermediate results of 306 recurrent laryngeal nerve sections for spastic dysphonia. *Laryngoscope.* 1983;93:9–15.

200. Crumley L. Repair of the recurrent laryngeal nerve. *Otolaryngol Clin North Am.* 1990;23:553–563.

201. Crumley L, Izdebski K, McMicken B. Nerve transfer versus teflon injection for vocal cord paralysis: A comparison. *Laryngoscope.* 1988;98:1200–1204.

202. Isshiki N, Morita H, Okamura H, et al. Thyroplasty, a new phono-surgical technique. *Acta Otolaryngol (Stockh).* 1974;78:451.

203. Johns ME, Cantrell RW. Voice restoration of the total laryngectomy patient: The Singer-Blom technique. *Otolaryngol Head Neck Surg.* 1981;89:82–86.

204. Johns ME. "How do I do it"—Head and neck. A targeted problem and its solution. The Panje button. *Laryngoscope.* 1982;92:204.

205. Singer MI. Voice rehabilitation after laryngectomy. In: Bailey BJ, ed. *Head and Neck Surgery: Otolaryngology.* Philadelphia, Pa: JB Lippincott; 1993: 1361–1372.

206. Kunc Z. Treatment of essential neuralgia of the IXth nerve by selective tractotomy. *J Neurosurg.* 1965;23:494.

207. Geschwind N. Nature of the decussated innervation of the sternocleidomastoid muscle. *Ann Neurol.* 1981;10:495.

208. DeToledo J, Smith DB, Kramer RE, Rosenbaum T, Stanulis R. Cortical innervation of the sternocleidomastoid in humans: Clinical and Neurophysiological Observations. *Ann Neurol.* 1989;26:171.

209. Patterson WR. Inferior dislocation of the distal end of the clavicle. *J Bone Joint Surg.* 1967;49A:1184.

210. Logigian EL. McInnes JM, Berger R, Busis NA, Lehrich JR: Stretch induced spinal accessory nerve palsy. *Muscle Nerve.* 1988;11:146–150.

211. Markey K, Di Benedetto M, Curl WW. Upper trunk brachial plexopathy: The stinger syndrome. *Am J Sports Med.* 1993;21:650–656.

212. Hoffman JC. Permanent paralysis of the accessory nerve after cannulation of the internal jugular vein. *Anesthesiology.* 1983;58:583.

213. Eisen A, Bertrand G. Isolated accessory nerve palsy of spontaneous origin. *Arch Neurol.* 1972;27:496–502.

214. Manon E, Espaillat R, Ruff RL. Dissociated weakness of sternocleidomastoid and trapezius muscles with lesions in the CNS. *Neurology.* 1989;38:796.

215. Mastaglia FL, Knezevic W, Thompson PD. Weakness of head turning in hemiplegia. *J Neurol Neurosurg Psychiatry.* 1986;49:195–197.

216. Mohanty SK, Barrios M, Fishbone H, Khatib R. Irreversible injury of cranial nerve IX–XII (Collet-Sicard Syndrome). *Neurosurgery.* 1973;38:86–88.

217. Anderson R, Flowers RS. Free grafts of the spinal accessory nerve during radical neck dissection. *Am J Surg.* 1969;118:796–799.

218. Di Benedetto M, Markey K. Electrodiagnostic localization of traumatic upper trunk brachial plexopathy. *Arch Phys Med Rehabil.* 1984;65:15–17.

219. Green RF, Brien M. Accessory nerve latency to the middla and lower trapezius. *Arch Phys Med Rehabil.* 1985;66:23–24.

220. Shankar K, Means KM. Accessory nerve conduction in neck dissection subjects. *Arch Phys Med Rehabil.* 1990;71:403–405.

221. Brodal A. *Neurological Anatomy in Relation to Clinical Medicine.* 3rd ed. New York: Oxford University Press; 1981: 453–459.

222. Mitchell SW. *Injuries of the Nerves and Their Consequences.* Philadelphia, Pa: JB Lippincott. 1872: 335.

223. Alrich EM, Baker GS. Injuries of the hypoglossal nerve. Report of ten cases. *Mil Surg.* 1948;103:20–25.

224. Keane JR, Baloh RW. Post-traumatic cranial neuropathies. *Neurologic Clin.* 1992;10:864.

225. Jgnelzi RJ, Bucy PC. Intracranial hypoglossal neurofibroma. *J Neurosurg.* 1967;26:352–356.

226. Berger PS, Bataini JP. Radiation induced cranial nerve palsy. *Cancer.* 1977;40:152–155.

227. Jabbari B, Coker SB. Paroxysmal rhythmic lingual movements and chronic epilepsy. *Neurology.* 1981;31: 1364–1367.

228. Troupin AS, Kamm RF. Lingual myoclonus: Case report and review. *Dis Nerve Syst.* 1974;35:378.

229. Brazis PW. The localization of lesions affecting cranial nerve XII (the hypoglossal nerve). In: Brazis PW, Masdeu JC, Biller J, eds. *Localization in Clinical Neurology*. 2nd ed. Boston, Mass: Little, Brown; 1990: 263.

230. Riggs JE. Distinguishing between extrinsic and intrinsic tongue muscle weakness in unilateral hypoglossal palsy. *Neurology.* 1984;34:1367.

231. Redmond MD, Di Benedetto M. Hypoglossal nerve conduction. *Muscle Nerve.* 1988;11:447–452.