CHAPTER 8

Lesions of the Spinal Cord

I. Overview: Lesions of the Spinal Cord (Figure 8-1)

- may be classified according to the area of origin or the area affected.

A. Lower motor neuron (LMN) lesions

B. Upper motor neuron (UMN) lesions

C. Sensory pathway lesions

D. Peripheral nervous system (PNS) lesions

E. Combined upper and lower motor neuron lesions

F. Combined motor and sensory lesions

G. Herniations of the intervertebral disk

II. Lower Motor Neuron Lesions (Figure 8-2A)

- result from damage to motor neurons of the ventral horns or motor neurons of the cranial nerve nuclei.
- result from interruption of the final common pathway connecting the neuron via its axon with the muscle fibers it innervates (the motor unit).

A. Neurologic deficits resulting from LMN lesions

1. **Flaccid paralysis**
2. **Muscle atrophy (amyotrophy)**
3. **Hypotonia**
4. **Areflexia**
   - consists of loss of muscle stretch reflexes (MSRs) (knee and ankle jerks) and loss of superficial reflexes (abdominal and cremasteric reflexes).
5. **Fasciculations (visible muscle twitches)**
6. **Fibrillations** (seen only on electromyogram)

B. Diseases of LMNs (see Figure 8-2A)

1. **Poliomyelitis**
   - is an acute inflammatory viral infection affecting the LMNs; it is caused by an enterovirus.
   - results in a flaccid paralysis.

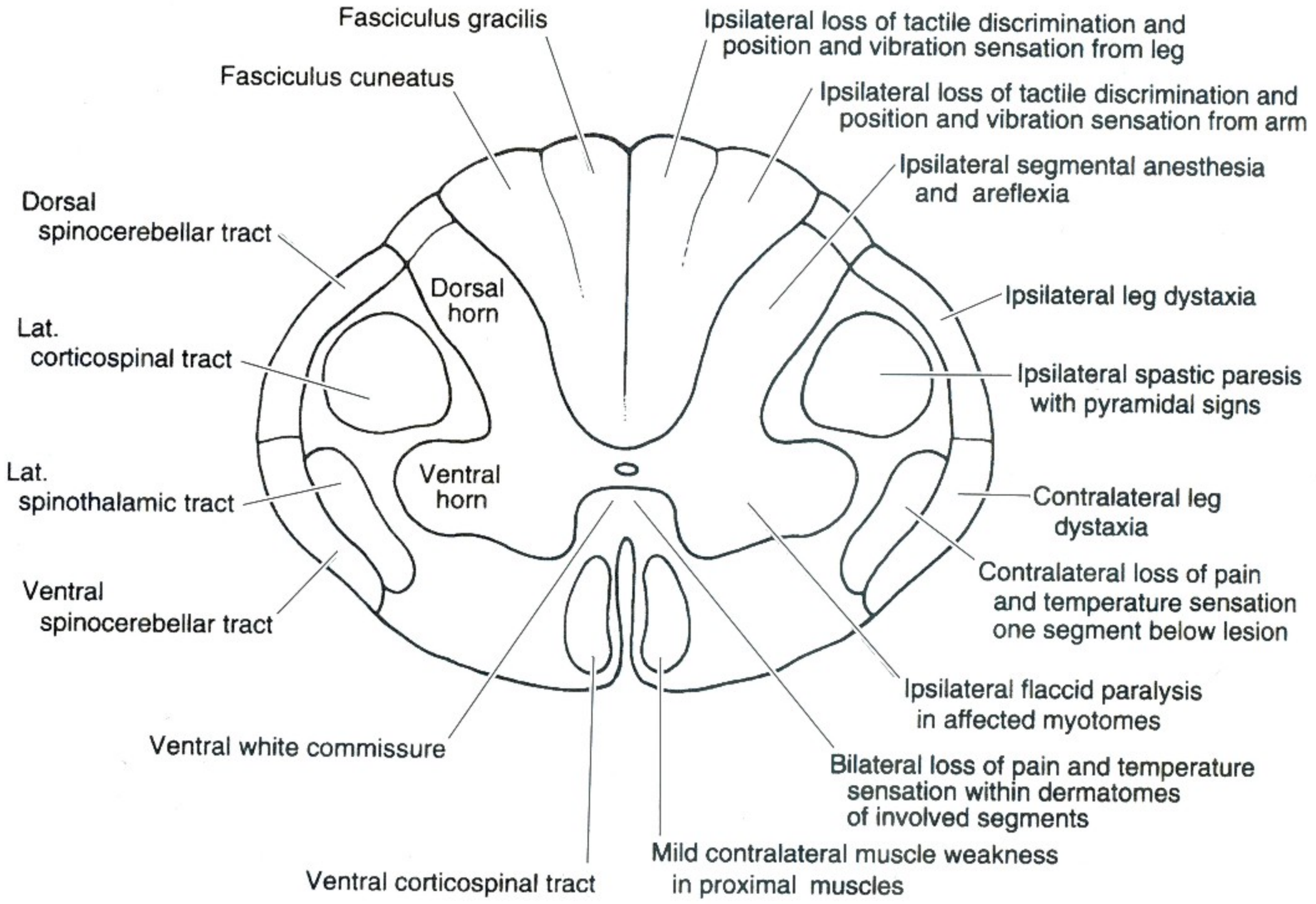


Figure 8-1. Transverse section of the cervical spinal cord. Clinically important pathways are shown on the left side; clinical deficits resulting from the interruption of these pathways are shown on the right side. Destructive lesions of the dorsal horns result in anesthesia and areflexia, and destructive lesions of the ventral horns result in LMN lesions and areflexia. Destruction of the ventral white commissure interrupts the central transmission of pain and temperature impulses bilaterally via the lateral spinothalamic tracts.

2. **Progressive infantile muscular atrophy (Werdnig-Hoffmann disease)**
   - is a heredofamilial degenerative disease of infants that affects LMNs.
3. **Kugelberg-Welander disease** (juvenile hereditary LMN disease)
   - appears at 3 to 20 years of age.
   - affects the large girdle muscles first and then the distal muscles.

III. Upper Motor Neuron Lesions

- result from damage to cortical neurons that give rise to corticospinal and corticobulbar tracts.
- are lesions of the corticospinal and corticobulbar tracts, called **pyramidal tract lesions**.
- may occur at all levels of the neuraxis from the cerebral cortex to the spinal cord.
- When rostral to the pyramidal decussation of the caudal medulla, they result in deficits below the lesion, on the contralateral side.
- When caudal to the pyramidal decussation, they result in deficits below the lesion, on the ipsilateral side.

A. Lateral corticospinal tract lesion

- results in the following ipsilateral motor deficits found below the lesion:
  1. **Spastic hemiparesis with muscle weakness**
  2. **Hyperreflexia (exaggerated muscle stretch reflexes)**
  3. **Clasp-knife spasticity**
     - When a joint is moved briskly, resistance occurs initially and then fades (like the opening of a pocketknife blade).

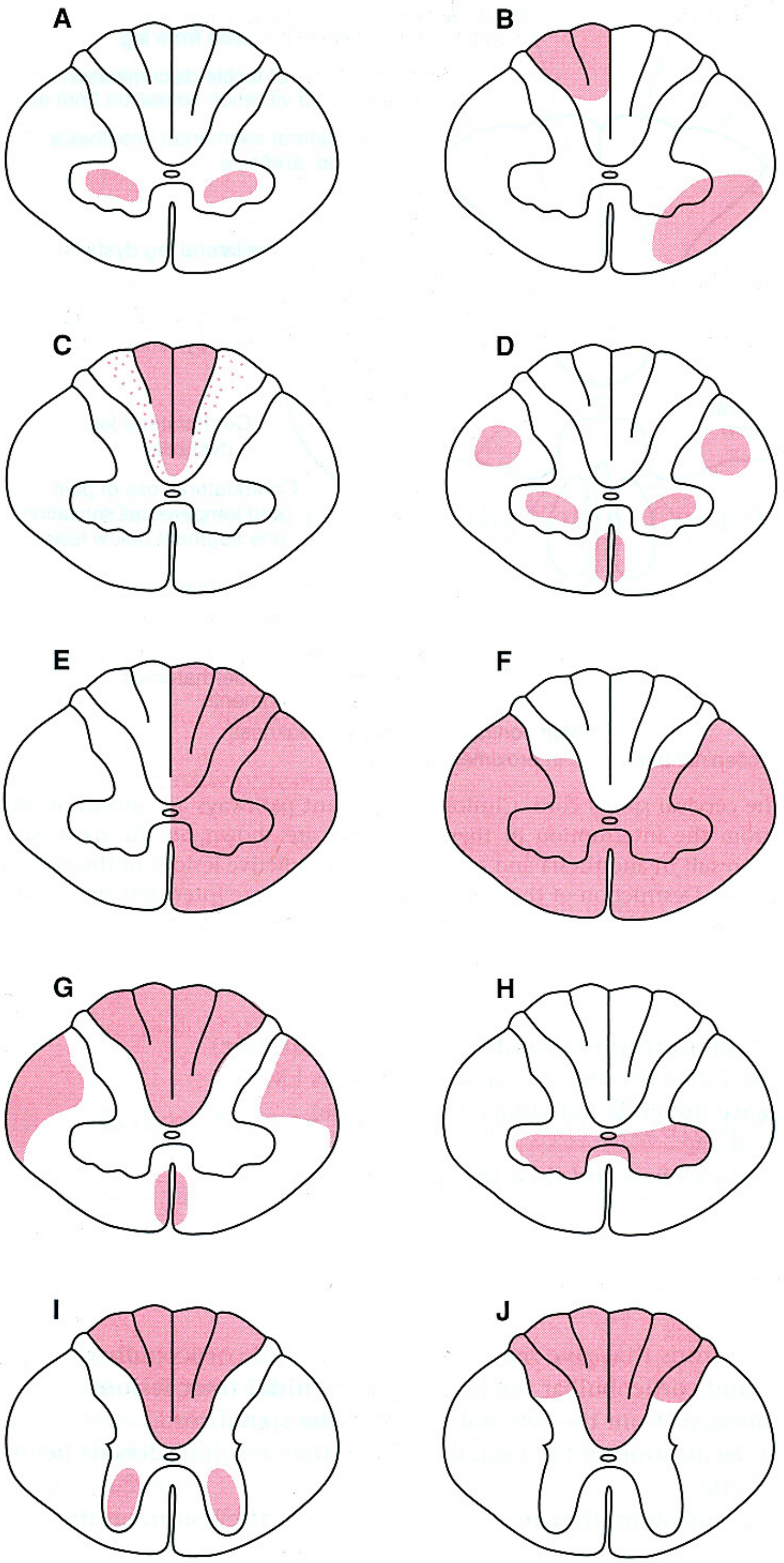


Figure 8-2. Lesions of the spinal cord: **(A)** poliomyelitis and progressive infantile muscular atrophy (Werdnig-Hoffmann disease); **(B)** multiple sclerosis; **(C)** dorsal column disease (tabes dorsalis); **(D)** amyotrophic lateral sclerosis; **(E)** hemisection of the spinal cord (Brown-Séquard syndrome); **(F)** complete ventral spinal artery occlusion of the spinal cord; **(G)** subacute combined degeneration (vitamin B_{12} neuropathy); *(H)* syringomyelia; **(I)** Charcot-Marie-Tooth disease (hereditary motor–sensory neuropathy type 1); **(J)** complete dorsal spinal artery occlusion. (Modified from Fix JD: *High-Yield Neuroanatomy,* 3rd ed. Philadelphia, Lippincott Williams & Wilkins, 2005, p 70.)

4. **Loss of superficial (abdominal and cremasteric) reflexes**
5. **Clonus**
   - consists of rhythmic contractions of muscles in response to sudden, passive movements (wrist, patellar, or ankle clonus).
6. **Babinski sign**
   - consists of plantar reflex response that is extensor (dorsiflexion of big toe).

B. Ventral corticospinal tract lesion

- results in **mild contralateral motor deficit**. Ventral corticospinal tract fibers decussate at spinal levels in the ventral white commissure.

C. Hereditary spastic paraplegia or diplegia

- is caused by bilateral degeneration of the corticospinal tracts.
- results in gradual development of spastic weakness of the legs with increased difficulty in walking.

IV. Sensory Pathway Lesions

A. Dorsal column syndrome (see Figure 8-2C)

- includes the fasciculi gracilis (T6–S5) and cuneatus (C2–T6) and the dorsal roots.
- is seen in subacute combined degeneration (vitamin B_{12} neuropathy).
- is seen in neurosyphilis as **tabes dorsalis** and in nonsyphilitic sensory neuropathies.
- results in the following **ipsilateral sensory deficits** found below the lesion:
  1. **Loss of tactile discrimination**
  2. **Loss of position (joint) and vibratory sensation**
  3. **Stereoanesthesia** (astereognosis)
  4. **Sensory (dorsal column) dystaxia**
  5. **Paresthesias and pain** (dorsal root irritation)
  6. **Hyporeflexia or areflexia** (dorsal root deafferentation)
  7. **Urinary incontinence, constipation, and impotence** (dorsal root deafferentation)
  8. **Romberg sign** (sensory dystaxia) (standing patient is more unsteady with eyes closed)

B. Lateral spinothalamic tract lesion

- results in contralateral loss of pain and temperature sensation one segment below the level of the lesion.

C. Ventral spinothalamic tract lesion

- results in contralateral loss of light (crude) touch sensation three or four segments below the level of the lesion.
- does not appreciably reduce touch sensation if the dorsal columns are intact.

D. Dorsal spinocerebellar tract lesion

- results in ipsilateral leg dystaxia; patient has difficulty performing the heel-to-shin test.

E. Ventral spinocerebellar tract lesion

- results in contralateral leg dystaxia; patient has difficulty performing the heel-to-shin test.

V. Peripheral Nervous System Lesions

- may be sensory, motor, or combined.
- affect spinal roots, dorsal root ganglia, and peripheral nerves.

A. Herpes zoster (shingles)

- is a common **viral infection** of the nervous system.
- consists of an acute inflammatory reaction in the dorsal root or cranial nerve ganglia.
- is usually limited to the territory of one dermatome; the most common sites are **T5 to T10.**
- causes irritation of dorsal root ganglion cells, resulting in pain, itching, and burning sensations in the involved dermatomes.
- produces the characteristic vesicular eruption in the affected dermatome.

B. Acute idiopathic polyneuritis (Guillain-Barré syndrome)

- is also called **postinfectious polyneuritis**.
- usually follows an infectious illness.
- results from a cell-mediated immunologic reaction directed at peripheral nerves.
- affects primarily motor fibers and causes segmental demyelination and wallerian degeneration.
- produces **LMN symptoms** (muscle weakness, flaccid paralysis, and areflexia).
- results in symmetric paralysis that begins in the lower extremities and ascends to involve the trunk and upper extremities; the facial nerve frequently is involved bilaterally.
- elevates cerebrospinal fluid (CSF) protein; however, the CSF cell count remains normal (**albuminocytologic dissociation**).

VI. Combined Upper and Lower Motor Neuron Lesions

A. Characteristics

- are muscle weakness and wasting without sensory deficits.

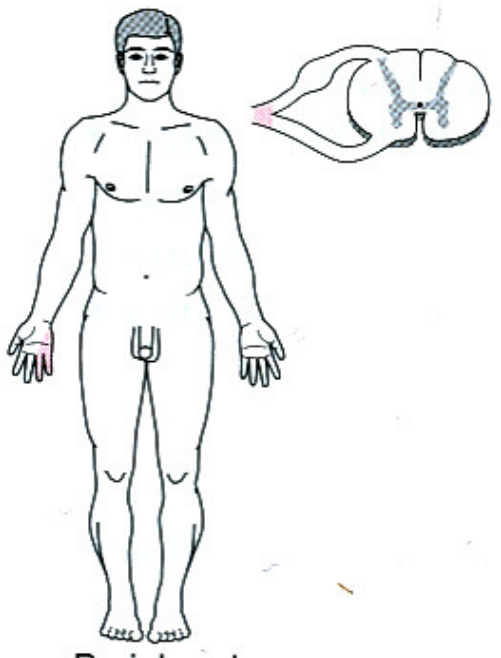
B. Prototypic disease—amyotrophic lateral sclerosis (AML) (see Figure 8-2D)

- is also called **Lou Gehrig disease**, motor neuron disease, or motor system disease.
- usually occurs in persons 50 to 70 years of age.
- affects twice as many men as women.
- involves both LMNs and UMNs; either component may dominate the clinical picture.
- progressive (spinal) muscular atrophy or progressive bulbar palsy refers to an LMN component.
- pseudobulbar palsy or primary lateral sclerosis refers to a UMN component.

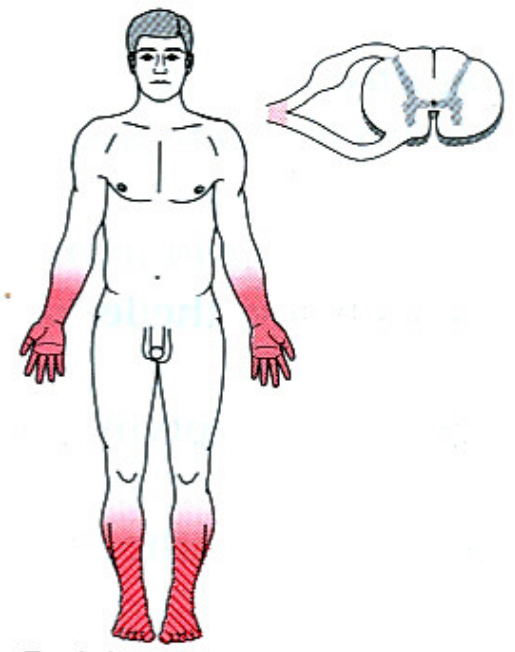
VII. Combined Motor and Sensory Lesions

A. Spinal cord hemisection (Brown-Séquard syndrome) (Figure 8-3; see Figures 8-1 and 8-2E)

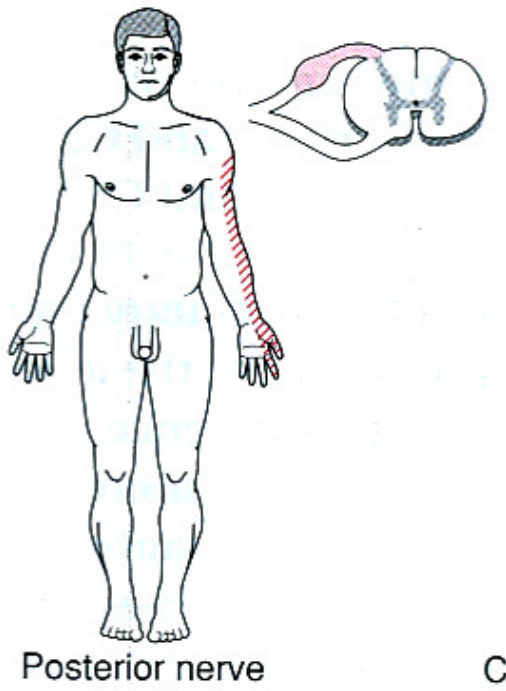
1. **Dorsal column transection**
   - results in ipsilateral loss of tactile discrimination, form perception, and position and vibration sensation below the lesion.
2. **Lateral spinothalamic tract transection**
   - results in contralateral loss of pain and temperature sensation, starting one segment below the lesion.
3. **Ventral spinothalamic tract transection**
   - results in contralateral loss of crude touch sensation starting three or four segments below the lesion.
4. **Dorsal spinocerebellar tract transection**
   - results in ipsilateral leg dystaxia.
5. **Ventral spinocerebellar tract transection**
   - results in contralateral leg dystaxia.
6. **Hypothalamospinal tract transection rostral to T2**
   - results in Horner syndrome.
7. **Lateral corticospinal tract transection**
   - results in ipsilateral spastic paresis below the UMN lesion with the Babinski sign.
8. **Ventral corticospinal tract transection**
   - results in minor contralateral muscle weakness below the lesion.
9. **Ventral horn destruction**
   - results in ipsilateral flaccid paralysis of somatic muscles (LMN lesion).
10. **Dorsal horn destruction**
    - results in ipsilateral dermatomic anesthesia and areflexia.



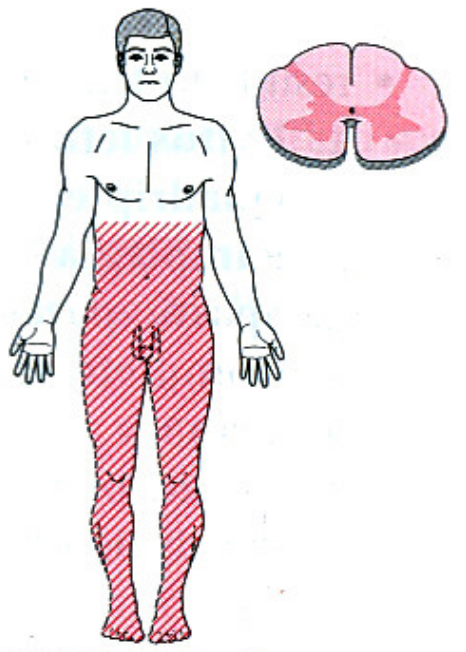
Peripheral nerve
(mononeuropathy lesion)



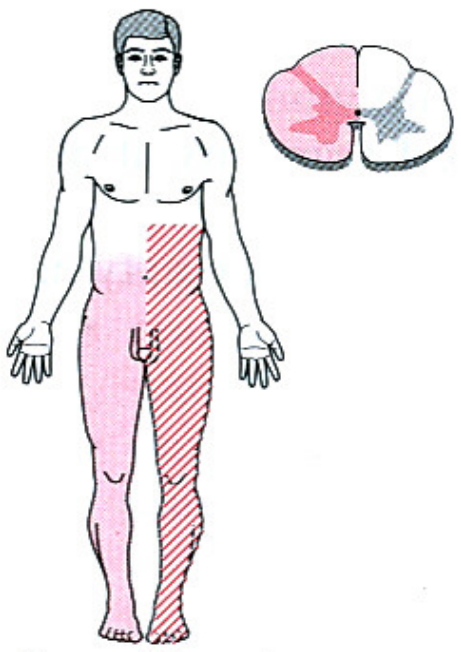
Peripheral nerve
(polyneuropathy lesion)



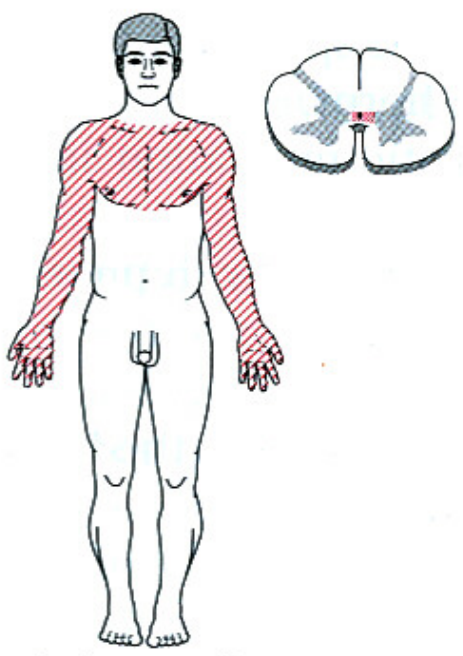
Posterior nerve
root (C6) lesion



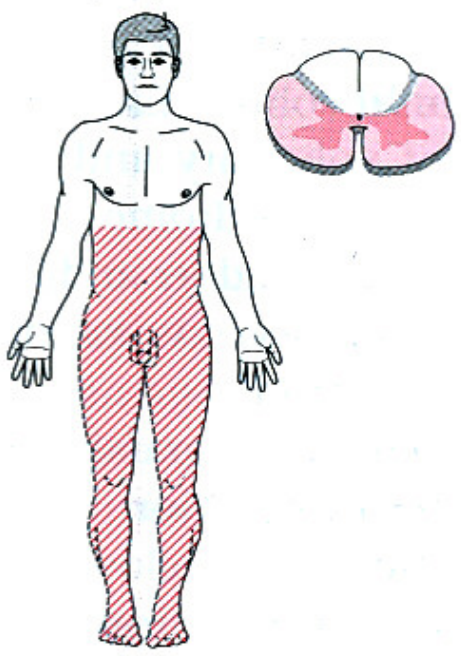
Complete transection
of the spinal cord



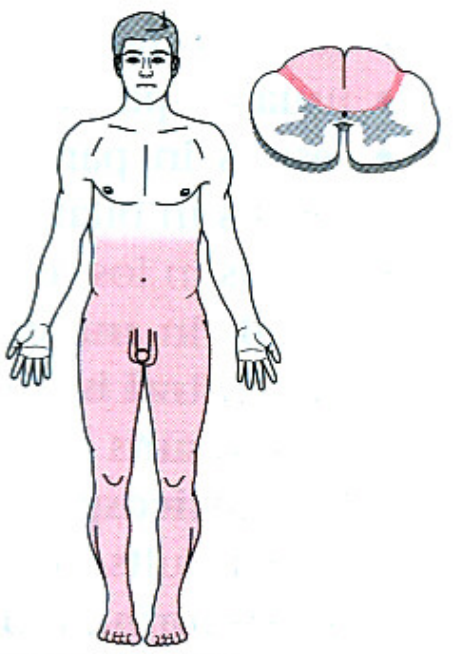
Brown-Séquard
syndrome



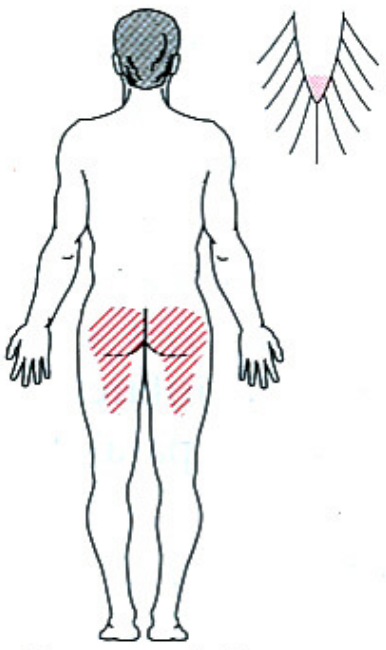
Syringomyelic
syndrome



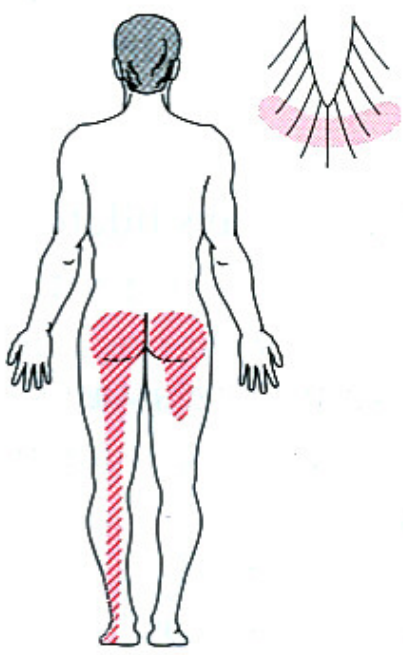
Anterior cord syndrome
(anterior spinal artery syndrome)



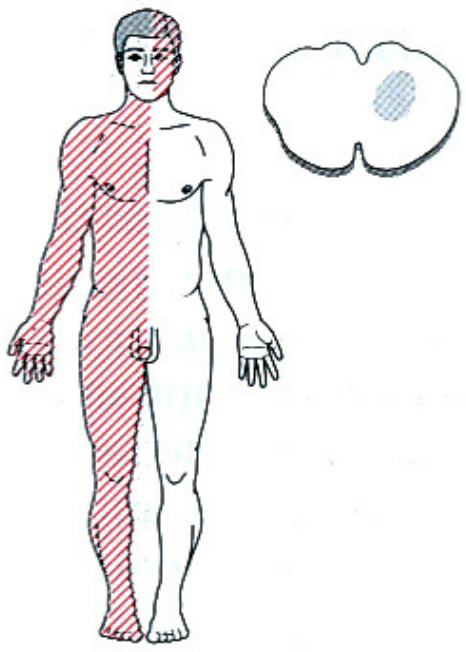
Posterior column
syndrome



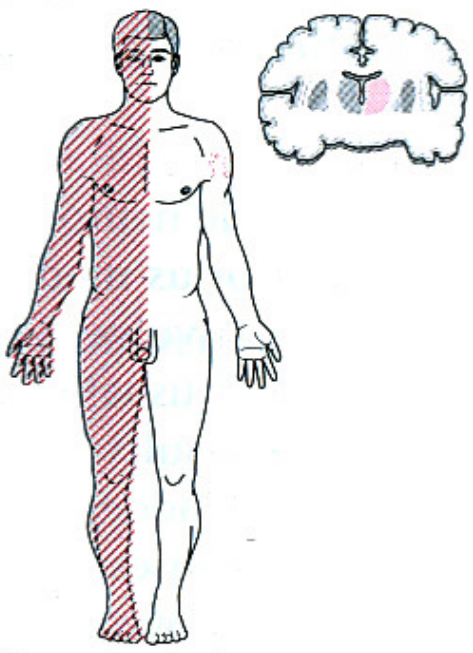
Conus medullaris
lesion



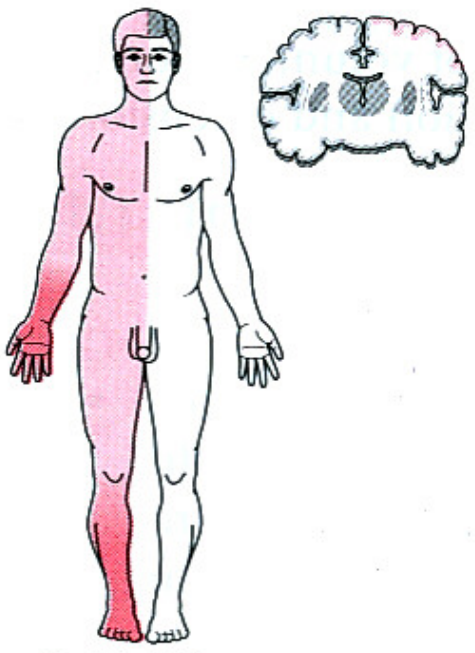
Cauda equina
lesion



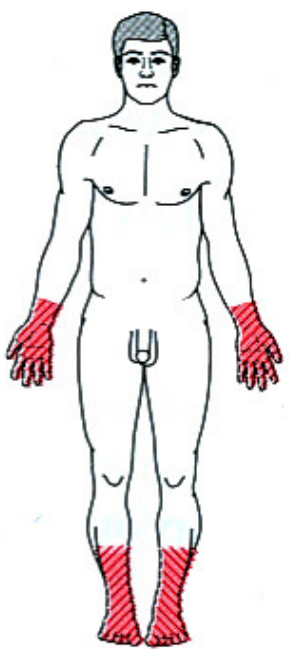
Medulla oblongata
lesion



Upper brainstem
or thoracic syndrome



Parietal lobe
syndrome



Hysteria

Loss of pain sense

Loss of tactile or position sense

Figure 8-3. Localization of sensory disorders.

B. Complete transection of the spinal cord

- results in the following conditions:
  1. **Exitus lethalis** between C1 and C3
  2. **Quadriplegia** between C4 and C5
  3. **Paraplegia** below T1
  4. **Spastic paralysis** of all voluntary movements below the lesion
  5. **Complete anesthesia** below the lesion
  6. **Urinary and fecal incontinence**, although reflex emptying may occur
  7. **Anhidrosis and loss of vasomotor tone**
  8. **Paralysis of volitional and automatic breathing** if the transection is above C5 (the phrenic nucleus is found at C3–C5)

C. Ventral (anterior) spinal artery occlusion (see Figure 8-2F)

- causes infarction of the ventral two-thirds of the spinal cord.
- usually spares the dorsal columns and dorsal horns.
- results in paralysis of voluntary and automatic respiration in cervical segments; it also results in bilateral Horner syndrome.
- results in loss of voluntary bladder and bowel control, with preservation of reflex emptying.
- results in anhidrosis and loss of vasomotor tone.

1. **Ventral horn destruction**
   - results in complete flaccid paralysis and areflexia at the level of the lesion.
2. **Corticospinal tract transection**
   - results in a spastic paresis below the lesion.
3. **Spinothalamic tract transection**
   - results in loss of pain and temperature sensations, starting one segment below the lesion.
4. **Dorsal spinocerebellar tract and ventral spinocerebellar tract transection**
   - results in cerebellar incoordination, which is masked by LMN and UMN paralysis.

D. Conus medullaris and epiconus syndromes

- include neurologic deficits and signs that are most always bilateral.

1. **Conus medullaris syndrome**
   - involves segments S3 to Co.
   - is usually caused by small intramedullary tumor metastases or hemorrhagic infarcts.
   - results in destruction of the sacral parasympathetic nucleus, which causes paralytic bladder, fecal incontinence, and impotence.
   - causes perianogenital sensory loss in dermatomes S3 to Co (saddle anesthesia).
   - shows an absence of motor deficits in the lower limbs.
2. **Epiconus syndrome**
   - involves segments L4 to S2.
   - results in reflex functioning of the bladder and rectum but loss of voluntary control.
   - is characterized by considerable **motor disability** (external rotation and extension of the thigh are most affected).
   - affects the ventral horns and long tracts.
   - is associated with absent Achilles tendon reflex.

E. Cauda equina syndrome

- classically involves spinal roots L3 to Co.
- produces neurologic deficits similar to those seen in conus or epiconus lesions.
- results in signs that frequently predominate on one side.
- may result from intervertebral disk herniation.
- commonly results in severe spontaneous radicular pain.

F. Filum terminale (tethered cord) syndrome

- results from a thickened, shortened filum terminale that adheres to the sacrum and causes traction on the conus medullaris.
- results in sphincter dysfunction, gait disorders, and deformities of the feet.

G. Subacute combined degeneration (vitamin B_{12} neuropathy) (see Figure 8-2G)

- is a spinal cord disease associated with pernicious anemia.
- consists of demyelination of dorsal columns, resulting in loss of vibration and position sensation.
- consists of demyelination of spinocerebellar tracts, resulting in arm and leg dystaxia.
- consists of demyelination of corticospinal tracts resulting in spastic paresis (UMN signs).

H. Friedreich hereditary ataxia (see Figure 8-2G)

- is the most common hereditary ataxia with autosomal recessive inheritance.
- results in spinal cord pathology and spinal cord symptoms that are similar to subacute combined degeneration with dorsal column, spinocerebellar, and corticospinal tract involvement.
- cerebellar involvement (Purkinje cells and dentate nucleus) is frequent with progressive ataxia.
- commonly leads to cardiomyopathy, pes cavus, and kyphoscoliosis.

I. Syringomyelia (see Figures 8-2H and 8-3)

- is a central cavitation of the cervical spinal cord of unknown etiology.
- results in destruction of the ventral white commissure and interruption of decussating spinothalamic fibers, causing bilateral loss of pain and temperature sensation.
- can result in extension of the syrinx into the ventral horn, causing an LMN lesion with muscle wasting and hyporeflexia. Atrophy of lumbricals and interosseous muscles of the hand is a common finding.
- can result in extension of the syrinx into the lateral funiculus, affecting the lateral corticospinal tract and resulting in spastic paresis (a UMN lesion).
- can result in caudal extension of the syrinx into the lateral horn at T1 or lateral extension into the lateral funiculus (interruption of descending autonomic pathways), resulting in Horner syndrome.

J. Multiple sclerosis (see Figure 8-2B)

- is the most common form of demyelinating disease.
- has asymmetric lesions and may affect all tracts of the spinal cord white matter. Spinal cord lesions occur most frequently in the cervical segments.

K. Charcot-Marie-Tooth disease (hereditary motor–sensory neuropathy type I) (see Figure 8-2I)

- is also called peroneal muscular atrophy.
- is the most common inherited neuropathy.
- affects the posterior columns, resulting in a loss of conscious proprioception.
- affects the anterior horn motor neurons, resulting in muscle weakness (atrophy).

VIII. Intervertebral Disk Herniation

A. Overview: intervertebral disk herniation

- consists of prolapse or herniation of the **nucleus pulposus** through the defective **annulus fibrosus** into the vertebral canal. The nucleus pulposus impinges on spinal roots, resulting in root pain (radiculopathy) or muscle weakness.
- may compress the spinal cord with a large central protrusion (above VL1).
- is recognized as the major cause of severe and chronic low back and leg pain.
- appears in 90% of cases at the L4–L5 or L5–S1 interspaces; usually a single nerve root is compressed, but several may be involved at the L5–S1 interspace (cauda equina).
- appears in 10% of cases in the cervical region, usually at the C5–C6 or C6–C7 interspaces.
- is characterized by **spinal root symptoms**, which include paresthesias, pain, sensory loss, hyporeflexia, and muscle weakness.

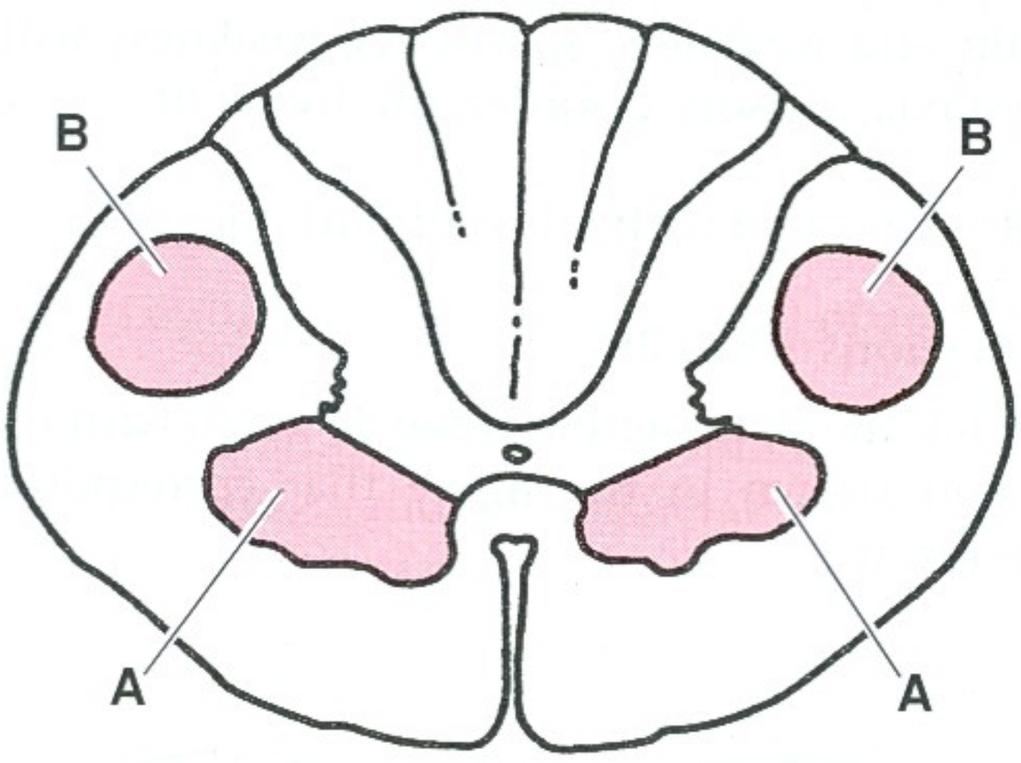
B. Cervical spondylosis with myelopathy

- is the most commonly observed myelopathy.
- consists of spinal cord or spinal cord root compression by calcified disk material extruded into the spinal vertebral canal.
- presents as painful stiff neck, arm pain and weakness, and spastic leg weakness with dystaxia; sensory disorders are frequent.

REVIEW TEST

Questions 1 to 3

Questions 1 to 3 relate to the figure.



Neuropathologic examination of the spinal cord reveals two lesions labeled A and B. Lesion A is restricted to five segments.

1. The result of lesion A is best described as

(A) bilateral arm dystaxia with dysdiadochokinesia
(B) spastic paresis of the legs
(C) flaccid paralysis of the upper extremities
(D) loss of pain and temperature sensation below the lesion
(E) urinary and fecal incontinence

2. The result of lesion B is best described as

(A) dyssynergia of movements affecting both arms and legs
(B) flaccid paralysis of the upper extremities
(C) impaired two-point tactile discrimination in both arms
(D) spastic paresis affecting primarily the muscles distal to the knee joint
(E) bilateral apallesthesia

3. Lesions A and B result from

(A) an intramedullary tumor
(B) an extramedullary tumor
(C) thrombosis of a spinal artery
(D) multiple sclerosis
(E) amyotrophic lateral sclerosis

4. Neurologic examination reveals an extensor plantar reflex on the left side, hyperreflexia on the left side, a loss of pain and temperature sensation on the right side, and ptosis and miosis on the left side. A lesion that causes this constellation of deficits would most likely be found in the

(A) paracentral lobule, left side
(B) crus cerebri, right side
(C) dorsolateral medulla, left side
(D) cervical spinal cord
(E) lumbar spinal cord

5. A 50-year-old woman complains of clumsiness in her hands while working in the kitchen: she recently burned her hands on the stove without experiencing any pain. Neurologic examination reveals bilateral weakness of the shoulder girdles, arms, and hands, as well as a loss of pain and temperature sensation covering the shoulder and upper extremity in a cape-like distribution. Severe atrophy is present in the intrinsic muscles of the hands. The most likely diagnosis is

(A) amyotrophic lateral sclerosis
(B) subacute combined degeneration
(C) Werdnig-Hoffmann disease
(D) syringomyelia
(E) tabes dorsalis

6. A 50-year-old man has a 2-year history of progressive muscle weakness in all extremities, with severe muscle atrophy and reduced MSRs in both legs. In his arms, the muscle atrophy is less pronounced and the MSRs are exaggerated. Which of the following types of neuronal degeneration would postmortem examination most likely show?

(A) Loss of Purkinje cells
(B) Loss of neurons from the globus pallidus
(C) Loss of neurons from the paracentral lobule and from the anterior horns of the spinal cord
(D) Demyelination of axons in the posterior and lateral columns
(E) Demyelination of axons in the posterior limb of the internal capsule

7. Transection of the spinothalamic tract results in

(A) Loss of pain and temperature sensation
(B) Complete flaccid paralysis
(C) Spastic paresis
(D) Cerebellar incoordination
(E) Areflexia

8. Which of the following is a characteristic of Lou Gehrig disease?

(A) Loss of tactile discrimination
(B) Loss of vibratory sensation
(C) Dorsal root irritation
(D) Progressive bulbar palsy
(E) Stereoanesthesia

9. Clasp-knife spasticity results from a lesion in the

(A) Ventral corticospinal tract
(B) Ventral spinothalamic tract
(C) Lateral corticospinal tract
(D) Dorsal spinocerebellar tract
(E) Lateral spinothalamic tract

10. Which of the following syndromes is associated with an absent Achilles tendon reflex?

(A) Filum terminale
(B) Cauda equina
(C) Conus medullaris
(D) Epicomus
(E) Syringomyelia

11. An example of a peripheral nervous system lesion is

(A) Guillain-Barré syndrome
(B) Charcot-Marie-Tooth disease
(C) Friedreich ataxia
(D) Lou Gehrig disease
(E) Brown-Séquard syndrome

12. A patient has the ability to stand with open eyes but falls with closed eyes. A lesion of which pathway is likely responsible for this symptom?

(A) Ventral spinothalamic tract
(B) Dorsal spinocerebellar tract
(C) Lateral spinothalamic tract
(D) Ventral spinocerebellar tract
(E) Dorsal column syndrome

Questions 13 to 18

The response options for items 13 to 18 are the same. Select one answer for each item in the set.

(A) Amyotrophic lateral sclerosis
(B) Cauda equina syndrome
(C) Cervical spondylosis
(D) Friedreich ataxia
(E) Guillain-Barré syndrome
(F) Multiple sclerosis
(G) Subacute combined degeneration
(H) Tabes dorsalis
(I) Werdnig-Hoffmann disease

Match each statement below with the syndrome that corresponds best to it.

13. A pure lower motor neuron disease

14. Elevated CSF protein with a normal CSF cell count

15. Characterized by asymmetric lesions found in the white matter of cervical segments

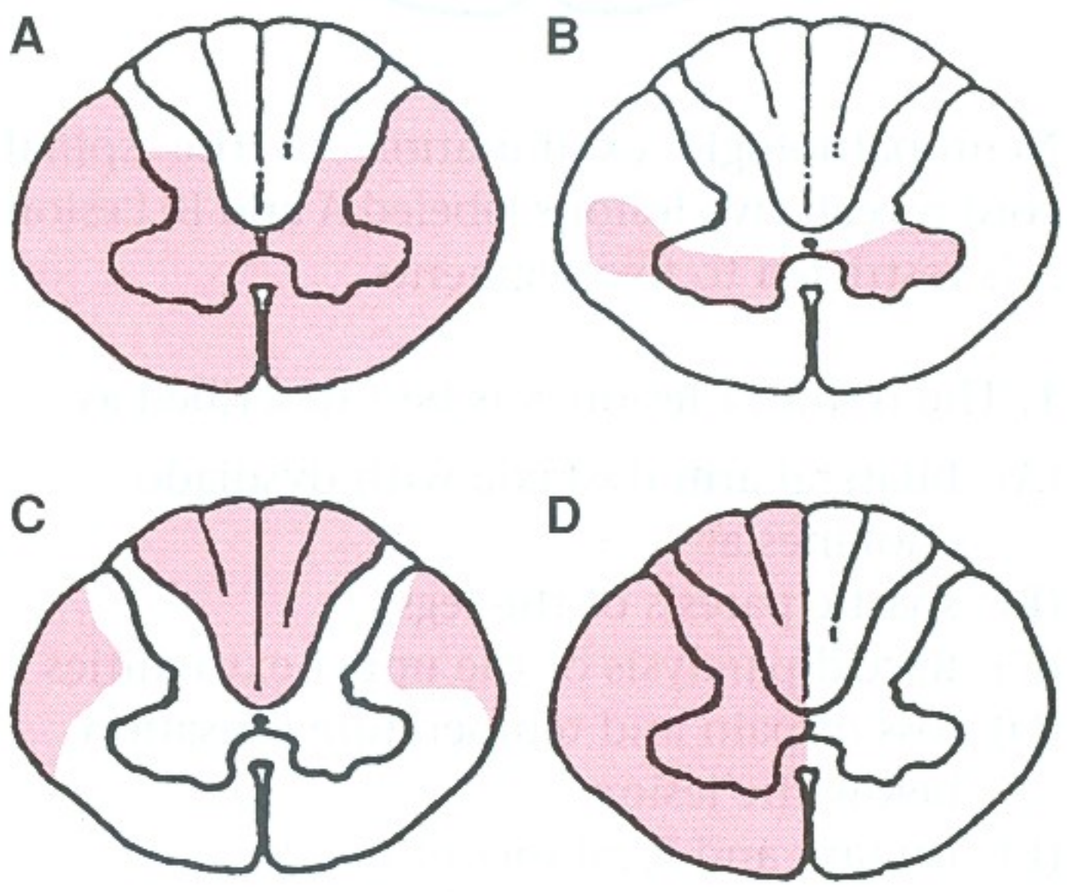
16. May result from intervertebral disk herniation

17. Symptoms include a painful stiff neck, arm pain and weakness, spastic leg weakness with dystaxia; sensory disorders are frequent

18. Associated with a loss of Purkinje cells

Questions 19 to 26

Match the statement in items 19 to 26 with the lesion shown in the figure that corresponds best to it.



19. Neurologic manifestation of vitamin B_{12} deficiency

20. Lesion due to vascular occlusion

21. Loss of vibration sensation on the right side; loss of pain and temperature sensation on the left side

22. Bilateral loss of pain and temperature sensation in the legs

23. Bilateral loss of pain and temperature sensation in the hands; muscle atrophy in both hands; spastic paresis on the right side only

24. Urinary incontinence and quadriplegia

25. No muscle atrophy or fasciculations

26. Demyelinating disease

ANSWERS AND EXPLANATIONS

1–C. Lesion A involves degeneration of the ventral horns bilaterally at midcervical levels, resulting in flaccid paralysis in the upper extremities.

2–D. Lesion B involves degeneration of the lateral corticospinal tracts bilaterally, resulting in spastic paresis of the lower extremities and primarily affecting the muscles distal to the knee. Spastic paresis of the upper extremities is masked by flaccid paralysis resulting from lesion A. Apallesthesia is the inability to perceive a vibrating tuning fork.

3–E. Lesions A and B are the result of amyotrophic lateral sclerosis, a pure motor disease.

4–D. A lesion of the cervical spinal cord could result in ipsilateral Horner syndrome, ipsilateral spastic paresis, and contralateral loss of pain and temperature sensation. Horner syndrome is always manifested on the ipsilateral side. This lesion produces a classic Brown-Séquard syndrome.

5–D. Syringomyelia is a cavitation of the spinal cord most commonly seen in the cervicothoracic segments. This condition results in bilateral loss of pain and temperature sensation in a cape-like distribution as well as wasting of the intrinsic muscles of the hands. Amyotrophic lateral sclerosis is a pure motor syndrome; subacute combined degeneration includes both sensory and motor deficits; Werdnig-Hoffmann disease is a pure motor disease; and tabes dorsalis is a pure sensory syndrome (neurosyphilis).

6–C. Amyotrophic lateral sclerosis affects both the upper and lower motor neurons. It is also referred to as motor systems disease. A loss of Purkinje cells as seen in cerebellar cortical atrophy (cerebello-olivary atrophy) results in cerebellar signs. Cell loss in the globus pallidus and putamen is seen in Wilson disease (hepatolenticular degeneration). Demyelination of axons in the posterior and lateral columns is seen in subacute combined degeneration. Demyelination of axons in the posterior limb of the internal capsule results in contralateral spastic hemiparesis.

7–A. Transection of the spinothalamic tract results in loss of pain and temperature sensations, starting one segment below the lesion. Ventral horn destruction results in complete flaccid paralysis and areflexia at the level of the lesion. Corticospinal tract transection results in spastic paresis below the lesion. Dorsal spinocerebellar tract and ventral spinocerebellar tract transection results in cerebellar incoordination.

8–D. Progressive bulbar palsy is a lower motor neuron component of amyotrophic lateral sclerosis, or Lou Gehrig disease. Disease characteristics are muscle weakness and wasting without sensory deficits. Loss of tactile discrimination, loss of vibratory sensation, stereoanesthesia, and dorsal root irritation are all sensory deficits found in dorsal column syndrome.

9–C. Clasp-knife spasticity is an ipsilateral motor deficit found below a lesion of the lateral corticospinal tract. It is characterized by initial but fading resistance of a briskly moved joint.

10–D. Epicomus syndrome involves segments L4 to S2 and results in loss of voluntary control of the bladder and rectum, motor disability, and an absent Achilles tendon reflex.

11-A. Acute idiopathic polyneuritis, or Guillain-Barré syndrome, is a peripheral nervous system lesion. It typically follows an infectious illness and results from a cell-mediated immunologic reaction.

12-E. Dorsal column syndrome results in a sensory deficit known as sensory dystaxia, or Romberg sign. Patients are Romberg positive when they are able to stand with the eyes open but fall with the eyes closed.

13–I. Werdnig-Hoffmann disease is a heredofamilial degenerative disease of infants that affects only LMNs.

14–E. Guillain-Barré syndrome is characterized by elevated CSF protein with normal CSF cell count (albuminocytologic dissociation).

15–F. Multiple sclerosis is characterized by asymmetric lesions frequently found in the white matter of cervical segments.

16–B. The cauda equina syndrome frequently results from intervertebral disk herniation; severe spontaneous radicular pain is common.

17–C. Cervical spondylosis is the most commonly observed myelopathy. Its symptoms include a painful stiff neck, arm pain and weakness, and spastic leg weakness with dystaxia; sensory disorders are frequent.

18–D. Friedreich ataxia is the most common hereditary ataxia with autosomal recessive inheritance. Dorsal columns, spinocerebellar tracts, and the corticospinal tracts show demyelination. Friedreich ataxia results in a loss of Purkinje cells in the cerebellar cortex and a loss of neurons in the dentate nucleus.

19–C. A neurologic manifestation of vitamin B_{12} deficiency is subacute combined degeneration. There is no involvement of LMNs.

20–A. Lesion A shows the territory of infarction resulting from occlusion of the ventral (anterior) spinal artery.

21–D. A spinal cord hemisection (Brown-Séquard syndrome) on the right side results in a loss of vibration sensation on the right side and a loss of pain and temperature sensation on the left side (dissociated sensory loss).

22–A. Total occlusion of the ventral spinal artery, involving five cervical segments, results in infarction of the ventral two-thirds of the spinal cord and interrupts both lateral spinothalamic tracts. The patient would have a loss of pain and temperature sensation caudal to the lesion.

23–B. Lesion B shows a cervical syringomyelic lesion involving the ventral white commissure, both ventral horns, and the right corticospinal tract. The patient would have a bilateral loss of pain and temperature sensation in the hands, muscle wasting in both hands, and a spastic paresis on the right side.

24–A. In lesion A, both lateral and ventral funiculi have been infarcted by arterial occlusion. Bilateral destruction of the lateral corticospinal tracts at upper cervical levels results in quadriplegia (spastic paresis in upper and lower extremities). Bilateral destruction of the ventrolateral quadrants results in urinary and fecal incontinence.

25–C. In lesion C, subacute combined degeneration, there is no involvement of LMNs, hence no flaccid paralysis, muscle atrophy, or fasciculations.

26–C. In lesion C, subacute combined degeneration, there is symmetric degeneration of the white matter, both in the dorsal columns (fasciculi gracilis) and in the lateral funiculi (corticospinal tracts). In this degenerative disease, both the myelin sheaths and the axis cylinders are involved. Subacute combined degeneration is classified under nutritional diseases (in this case a vitamin B_{12} neuropathy). In true demyelinative diseases (e.g., multiple sclerosis), the myelin sheaths are involved but the axis cylinders and nerve cells are relatively spared.

CHAPTER 14

Lesions of the Brainstem

I. Introduction: Lesions of the Brainstem

- are most frequently syndromes of arterial occlusion or circulatory insufficiency that involve the vertebrobasilar system.

II. Vascular Lesions of the Medulla

- result from occlusion of the vertebral artery or its branches (i.e., the anterior and posterior spinal arteries and the posterior inferior cerebellar artery [PICA]).

A. Medial medullary syndrome (Figure 14-1A)

- results from occlusion of the anterior spinal artery.
- includes the following affected **structures** and resultant **deficits**:
  1. **Corticospinal tract**
     - contralateral hemiparesis of the trunk and extremities
  2. **Medial lemniscus**
     - contralateral loss of proprioception, discriminative tactile sensation, and vibration sensation from the trunk and extremities
  3. **Hypoglossal nerve roots (intra-axial fibers)**
     - ipsilateral flaccid paralysis of the tongue

B. Lateral medullary syndrome (PICA syndrome) (see Figure 14-1B)

- is also called Wallenberg syndrome.
- results from occlusion of the vertebral artery or one of its medullary branches (e.g., PICA).
- includes the following affected **structures** and resultant **deficits**:
  1. **Vestibular nuclei (medial and inferior)**
     - nystagmus, nausea, vomiting, and vertigo
  2. **Inferior cerebellar peduncle**
     - ipsilateral cerebellar signs (dystaxia, dysmetria, dysdiadochokinesia)
  3. **Nucleus ambiguus of cranial nerve (CN) IX, CN X, and CN XI (somatic visceral efferent [SVE])**
     - ipsilateral laryngeal, pharyngeal, and palatal paralysis (loss of the gag reflex [efferent limb], dysarthria, dysphagia, and dysphonia [hoarseness])
  4. **Glossopharyngeal nerve roots (intra-axial fibers)**
     - loss of the gag reflex (afferent limb)
  5. **Vagal nerve roots (intra-axial fibers)**
     - neurologic deficits same as those seen in lesion of the nucleus ambiguus
  6. **Spinothalamic tracts**
     - contralateral loss of pain and temperature sensation from the trunk and extremities

CHAPTER 13

Cranial Nerves

I. Introduction: Cranial Nerves

- are the 12 pairs of nerves that arise from the brain and supply the structures of the head and neck (Figures 13-1 through 13-3; see Figures 1-1 and 1-7).

II. Olfactory Nerve (CN I) (see Chapter 20 I and Appendix)

A. General characteristics of CN I

- is a special visceral afferent (**SVA**) nerve that mediates the **sense of smell** (olfaction).
- consists of unmyelinated axons of bipolar neurons located in the nasal mucosa, the olfactory epithelium.
- enters the skull via the foramina of the cribriform plate of the ethmoid bone.
- projects directly to the telencephalon.
- synapses with mitral and tufted cells found in the olfactory bulb, an outgrowth of the telencephalon.
- is the only cranial nerve that projects directly to the forebrain.

B. Clinical correlation: CN I damage

- results in **anosmia**, loss of olfactory sensation (e.g., ethmoid bone fracture).

III. Optic Nerve (CN II) (see Figures 1-2, 17-2, and 17-4; see Chapter 17 III B)

A. General characteristics of CN II

- is a special somatic afferent (**SSA**) nerve that subserves **vision** and **pupillary light reflexes** (the afferent limb).
- consists of axons of neurons located in the ganglion cell layer of the retina.
- enters the skull via the optic canal of the sphenoid bone.
- has axons that continue via the optic chiasm and optic tracts to the lateral geniculate body, a thalamic relay nucleus that projects to the visual cortex (area 17) of the occipital lobe.
- is **not a true peripheral nerve** but a tract of the diencephalon.
- contains fibers from the nasal retina that decussate in the optic chiasm.
- contains fibers from the temporal retina that continue ipsilaterally through the optic chiasm.
- contains axons that are myelinated by oligodendrocytes.
- is invested by the dura and pia–arachnoid membranes and lies within the subarachnoid space.

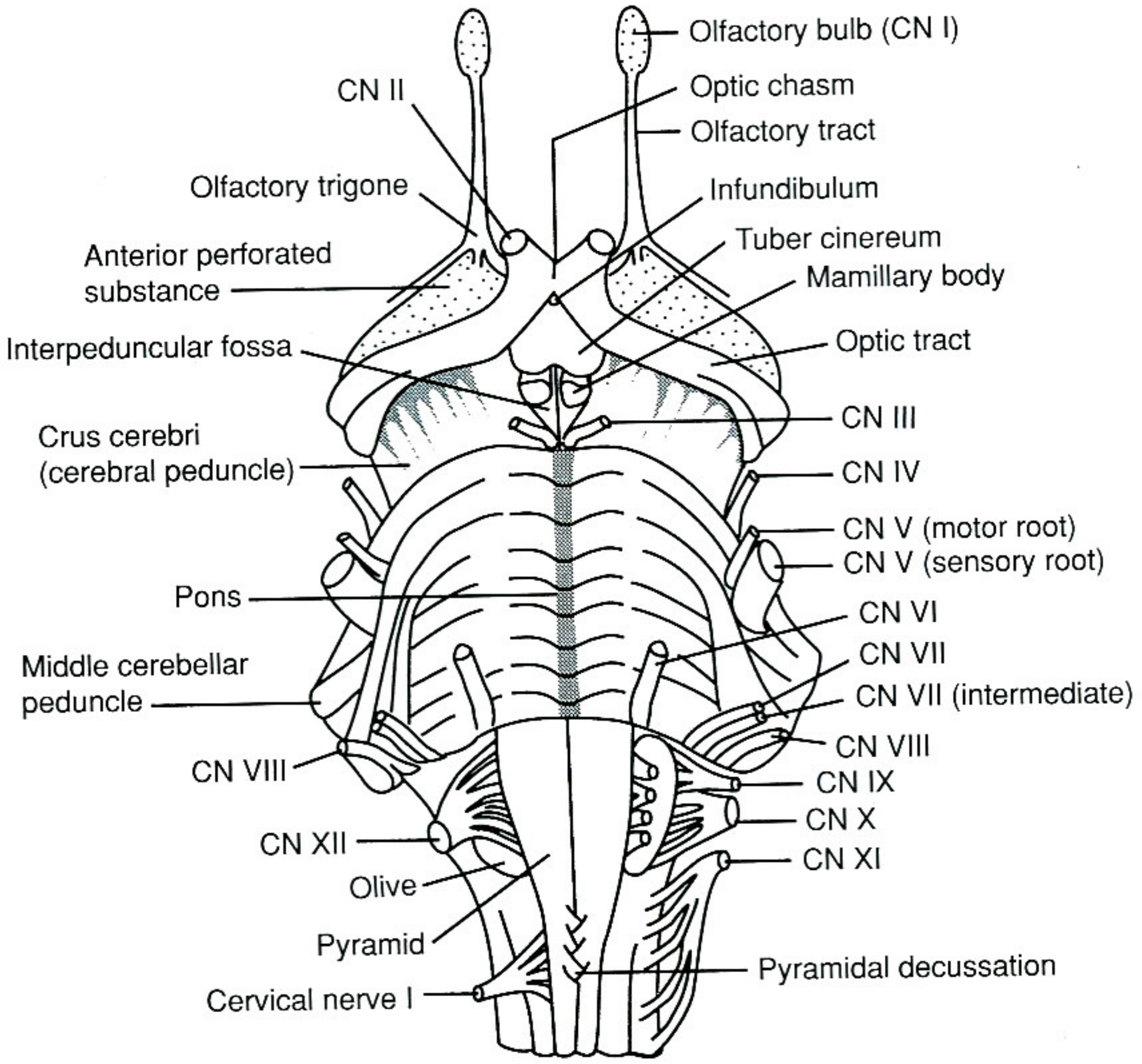


Figure 13-1. The base of the brain with attached cranial nerves (*CN*). (Reprinted with permission from Truex RC, Kellner CE: *Detailed Atlas of the Head and Neck*. New York, Oxford University Press, 1958, p 34.)

B. Clinical correlations: CN II

- When it is transected, **ipsilateral blindness** and **loss of direct pupillary light reflex** result; regeneration of the optic nerve does not occur.
- When it is subjected to increased intracranial pressure (e.g., tumor), **papilledema**, a choked optic disk results.
- When it is constricted, **optic atrophy** (i.e., axonal degeneration) results.

IV. Oculomotor Nerve (CN III) (see Figures 1-1, 1-7, and 13-3; Chapter 17)

A. General characteristics of CN III

- contains general somatic efferent (**GSE**) and general visceral efferent (**GVE**) fibers.
- is a pure motor nerve that **moves the eye**, **constricts the pupil**, **accommodates**, and **converges**.
- exits the brainstem from the interpeduncular fossa of the midbrain, passes through the lateral wall of the cavernous sinus, and enters the orbit via the superior orbital fissure.

1. **GSE component**
   - arises from the oculomotor nucleus of the midbrain.
   - innervates four extraocular muscles and the levator palpebrae muscle. (Remember the mnemonic: **SIN**, **S**uperior muscles are **IN**torters of the globe.)

   a. **Medial rectus muscle**
      - adducts the eye.
      - with its opposite partner, converges the eyes.

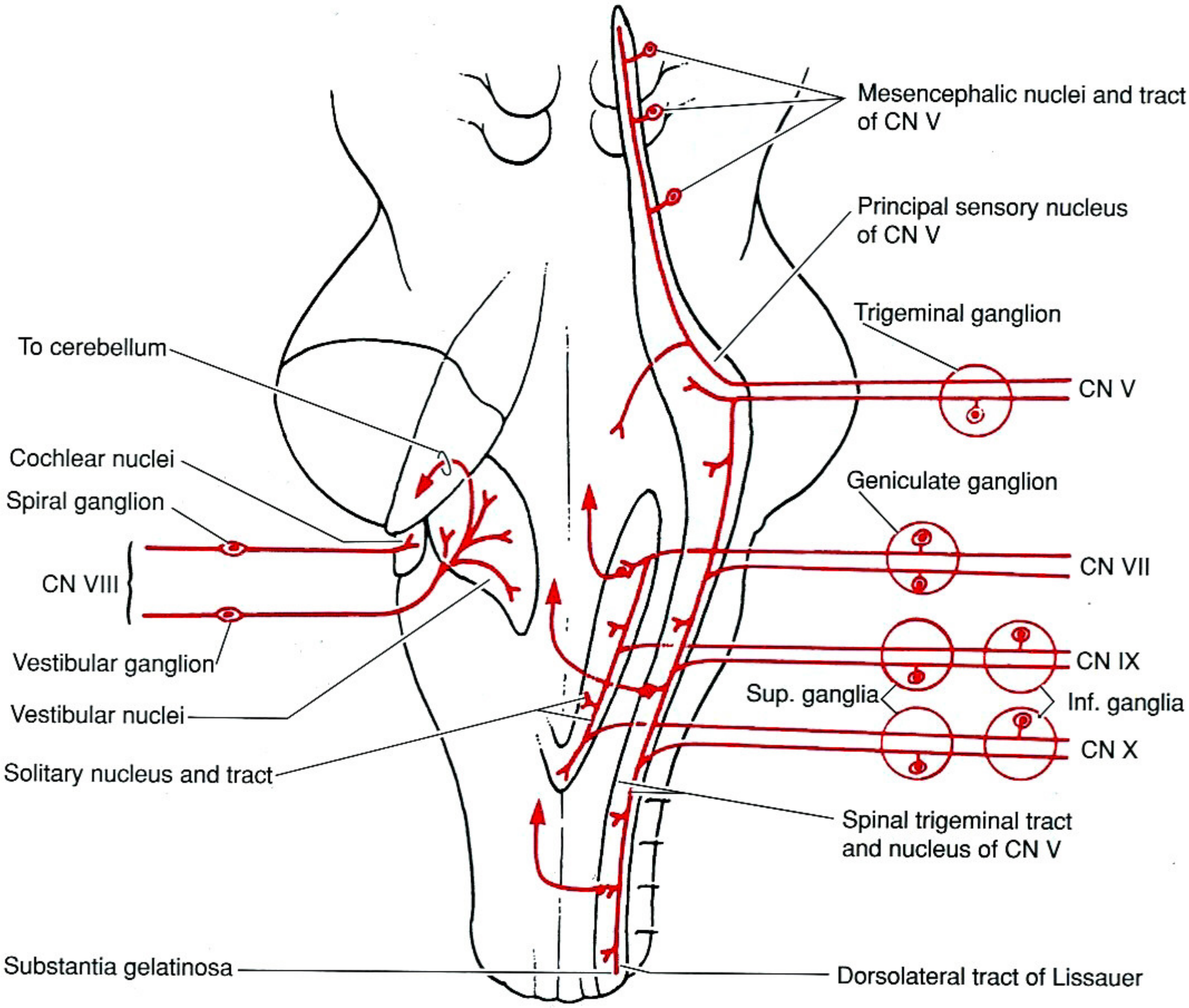


Figure 13-2. Location of the sensory cranial nerve nuclei within the brainstem. Phantom view of the brainstem from the dorsal aspect. The spinal trigeminal tract and nucleus extend into the cervical cord (C3). Three sensory areas are prominent: the special somatic afferent (SSA) area, including the cochlear and vestibular nuclei of CN VIII; the combined general visceral afferent (GVA) and SVA column, the solitary nucleus of CN VII, CN IX, and CN X; and the GSA column, including the spinal trigeminal, principal, sensory, and mesencephalic nuclei of CN V, CN VII, CN IX, and CN X. (Modified with permission from Noback CR, Demarest RJ: *The Human Nervous System*. Baltimore, Williams & Wilkins, 1991, p 222.)

b. Superior rectus muscle
- elevates, intorts, and adducts the eye.

c. Inferior rectus muscle
- depresses, extorts, and adducts the eye.

d. Inferior oblique muscle
- elevates, extorts, and abducts the eye.

e. Levator palpebrae muscle
- elevates the upper lid.

2. GVE component

a. Composition
- consists of preganglionic parasympathetic fibers.

b. Pathway
- arises from the Edinger-Westphal nucleus (accessory oculomotor nucleus) of the midbrain.

(1) Edinger-Westphal nucleus
- projects to the ciliary ganglion of the orbit via CN III.

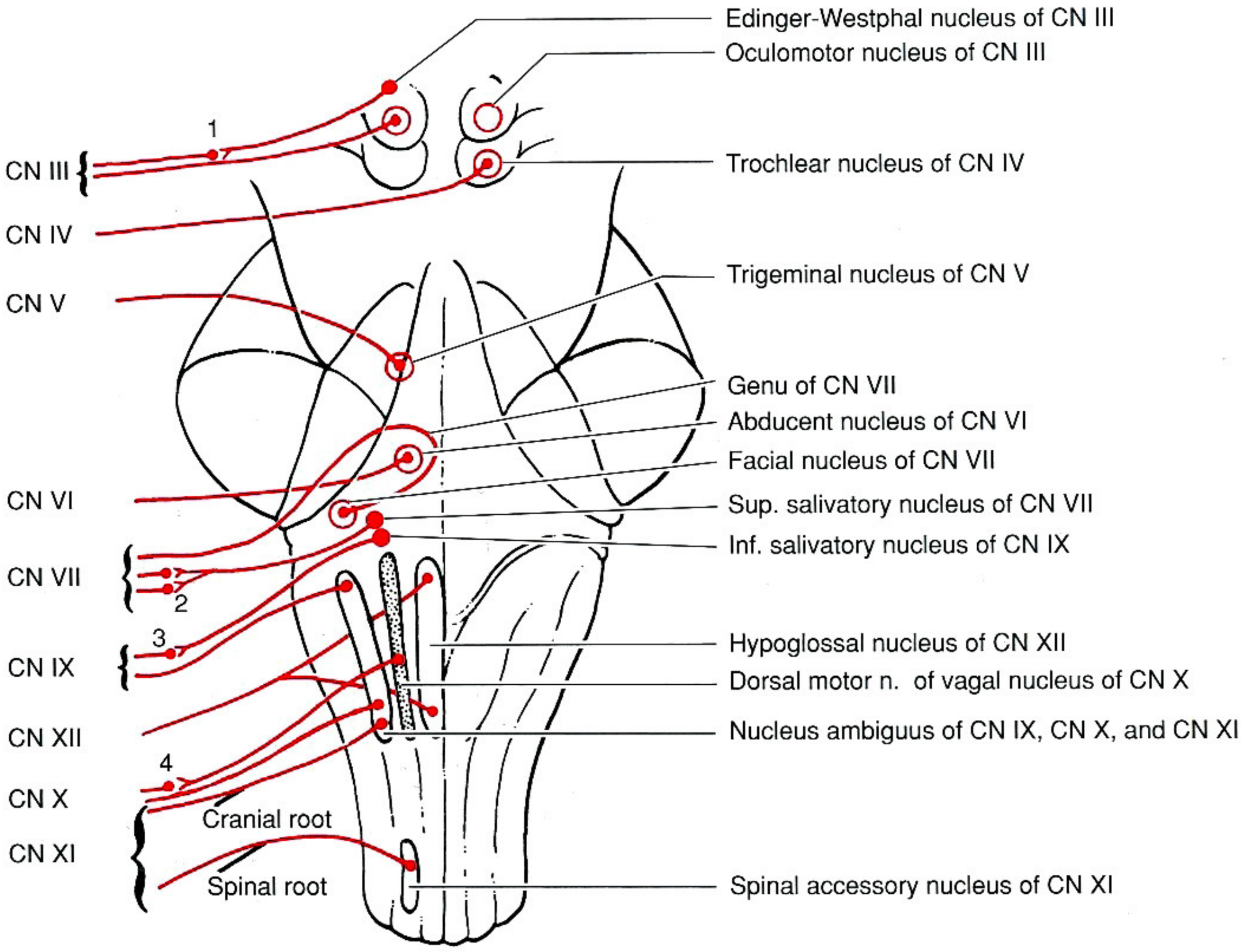


Figure 13-3. Location of motor cranial nerve nuclei within the brainstem. Three functional cell columns are visible from medial to lateral; the GSE column of CN III, CN IV, CN VI, and CN XII; the GVE column of CN III, CN VII, CN IX, and CN X; and the SVE column of CN V, CN VII, CN IX, CN X, and CN XI. Parasympathetic ganglia are indicated as *1* = ciliary ganglion; *2* = pterygopalatine and submandibular ganglia; *3* = otic ganglion; and *4* = terminal (intramural) ganglia. (Modified with permission from Noback CR, Demarest RJ: *The Human Nervous System*. Baltimore, Williams & Wilkins, 1991, p 223.)

(2) Ciliary ganglion

- projects postganglionic parasympathetic fibers to the sphincter muscle of the iris (miosis) and to the ciliary muscle (accommodation).

B. Clinical correlations: CN III

1. Oculomotor paralysis

- is seen frequently with **transtentorial herniation** (subdural or epidural hematoma).
- results in **diplopia** (double vision) when the patient looks in the direction of the paretic muscle.
- Denervation of the levator palpebrae muscle results in **ptosis** (drooping of the upper eyelid).
- Denervation of the extraocular muscles causes the affected eye to **look down and out** because the action of the lateral rectus and superior oblique muscles is unopposed. The superior oblique and lateral rectus muscles are innervated by CN IV and CN VI.
- Interruption of parasympathetic innervation results in a **dilated and fixed pupil** and **paralysis of accommodation (cycloplegia)**.

2. Other conditions associated with CN III impairment

a. Transtentorial (uncal) herniation

- Increased supratentorial pressure (tumor) forces the hippocampal uncus through the tentorial notch and compresses the oculomotor nerve. Pupilloconstrictor fibers are affected first, resulting in a dilated and fixed pupil; somatic efferent fibers are affected later, resulting in an external strabismus (exotropia).

b. **Aneurysms (carotid and posterior communicating arteries)**
   - frequently compress the oculomotor nerve within the cavernous sinus or the interpeduncular cistern.
   - usually affect the peripheral pupilloconstrictor fibers first, as in uncal herniation.

c. **Diabetes mellitus (diabetic oculomotor palsy)**
   - frequently affects the oculomotor nerve, damaging the central fibers and sparing the pupilloconstrictor fibers.

V. Trochlear nerve (CN IV) (see Figures 1-7 and 13-3)

A. General characteristics of CN IV

- is a pure **GSE** nerve that **innervates the superior oblique muscle**, which **depresses, intorts**, and **abducts** the eye.
- arises from the contralateral trochlear nucleus of the midbrain.
- decussates within the midbrain and exits the brainstem on its dorsal surface, caudal to the inferior colliculus.
- encircles the midbrain in the subarachnoid space, passes through the lateral wall of the cavernous sinus, and enters the orbit via the superior orbital fissure.

B. Clinical correlations: CN IV paralysis (Figure 13-4)

- results in the following conditions:
  1. **Extorsion of the eye and weakness of downward gaze**
  2. **Vertical diplopia**, which increases when looking down
  3. **Head tilting**, to compensate for extorsion

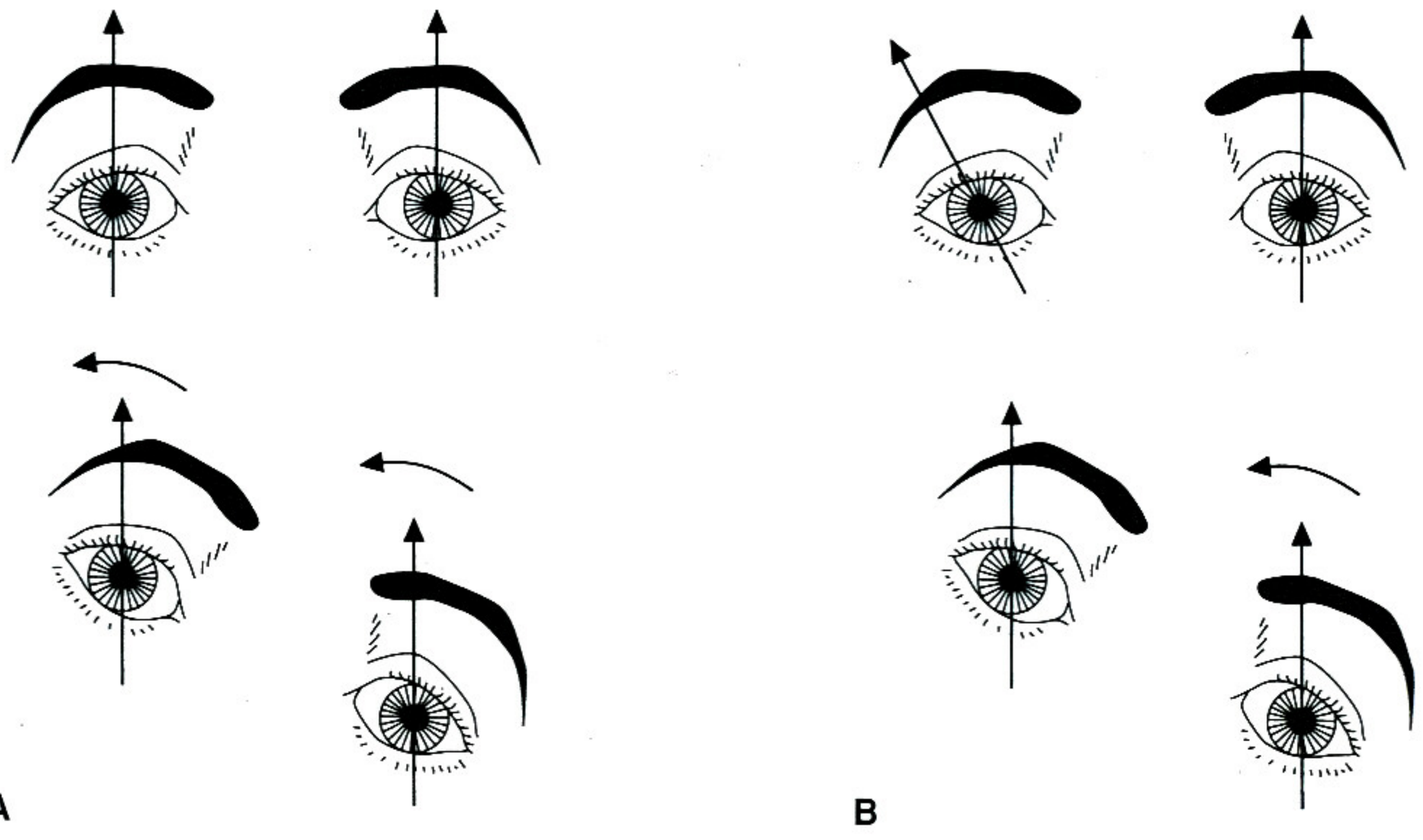


Figure 13-4. Paralysis of the right superior oblique muscle. (A) A pair of eyes with normal extorsion and intorsion movements. Tilting the chin to the right side results in compensatory intorsion of the left eye and extorsion of the right eye. (B) Paralysis of the right superior oblique muscle results in extorsion of the right eye, causing diplopia. Tilting the chin to the right side results in compensatory intorsion of the left eye, thus permitting binocular alignment.

VI. Trigeminal Nerve (CN V) (see Figures 1-1, 1-7, 10-4, 13-2, and 13-3; see Chapter 10)

A. General characteristics of CN V

- contains general somatic afferent (**GSA**) and special visceral efferent (**SVE**) fibers.
- innervates the **muscles of mastication** and mediates **general sensation** from the face, eye, and nasal and oral cavities.
- is the nerve of the first pharyngeal arch (mandibular).
- exits the brainstem from the pons.
- contains first-order sensory neurons in the trigeminal ganglion and in the mesencephalic nucleus.
- contains motor neurons in the motor trigeminal nucleus of the rostral pons.
- has three divisions: **ophthalmic** (CN V-1), **maxillary** (CN VI-2), and **mandibular** (CN V-3) (see Figures 10-1 and 10-2; see Chapter 10 I A 1–3).
  1. **GSA component** (see Figure 10-1)
     - provides **sensory innervation** to the face, mucous membranes of the nasal and oral cavities and frontal sinus, teeth, hard palate, soft palate, and deep structures of the head (proprioception from muscles and the temporomandibular joint).
     - innervates the dura of the anterior and middle cranial fossae.
     - innervates the external ear with CN VII, CN IX, and CN X.
  2. **SVE component**
     - innervates the **muscles of mastication** (temporalis, masseter, lateral and medial pterygoids), the **tensores tympani** and **veli palatini**, the **mylohyoid**, and the **anterior belly of the digastric muscles**.

B. Clinical correlations: lesions of CN V

- result in the following conditions:
  1. **Loss of general sensation** from the face and mucous membranes of the oral and nasal cavities
  2. **Loss of the corneal reflex** (afferent limb, CN V-1)
  3. **Flaccid paralysis of the muscles of mastication**
  4. **Deviation of the jaw to the weak side** due to the unopposed action of the opposite lateral pterygoid muscle
  5. **Paralysis of the tensor tympani**, leading to hypacusis (partial deafness to low-pitched sounds)

VII. Abducent Nerve (CN VI) (see Figures 1-1, 1-7, and 13-3)

A. General characteristics of CN VI

- is a pure **GSE** nerve that innervates the lateral rectus muscle, which **abducts the eye**.
- arises from the abducent nucleus of the caudal pons.
- exits the brainstem from the inferior pontine sulcus.
- passes through the Dorello canal and cavernous sinus and enters the orbit via the superior orbital fissure.

B. Clinical correlations: CN VI paralysis

- is the most common isolated muscle palsy.
- results in the following conditions:
  1. **Convergent strabismus** (**esotropia**), with inability to abduct the eye because of the unopposed action of the medial rectus muscle
  2. **Horizontal diplopia**, with maximum separation of the double images when looking toward the paretic lateral rectus muscle

VIII. Facial Nerve (CN VII) (see Figures 1-1, 1-7, 13-2, 13-3, and 13-5)

A. General characteristics of CN VII

- contains **GSA**, **SVA**, **SVE**, and **GVE** fibers.
- mediates **facial movements**, **taste**, **salivation**, and **lacrimation**.
- is the nerve of the second pharyngeal arch (hyoid).
- includes the **facial nerve proper** (motor division), which contains the SVE fibers that innervate the muscles of facial expression.
- includes the **intermediate nerve** (sensory division), which contains GSA, SVA, and GVE fibers. All first-order sensory neurons are found in the geniculate ganglion within the temporal bone.
- exits the brainstem in the cerebellopontine (CP) angle.
- enters the internal auditory meatus and facial canal.
- exits the facial canal and skull via the **stylomastoid foramen**.

1. **GSA component**
   - has cell bodies in the geniculate ganglion.
   - innervates the **posterior surface of the external ear** via the posterior auricular branch of the facial nerve.
   - projects centrally to the spinal trigeminal tract and nucleus.
2. **SVA component**
   - has cell bodies in the geniculate ganglion.
   - projects centrally to the solitary tract and nucleus.
   - innervates the **taste buds** from the anterior two-thirds of the tongue via:

   a. **Intermediate nerve**

   b. **Chorda tympani** (Figure 13-6)
      - is located in the tympanic cavity medial to the tympanic membrane and lateral to the malleus.
      - contains **SVA** and **GVA** fibers.

   c. **Lingual nerve** (a branch of CN V-3)

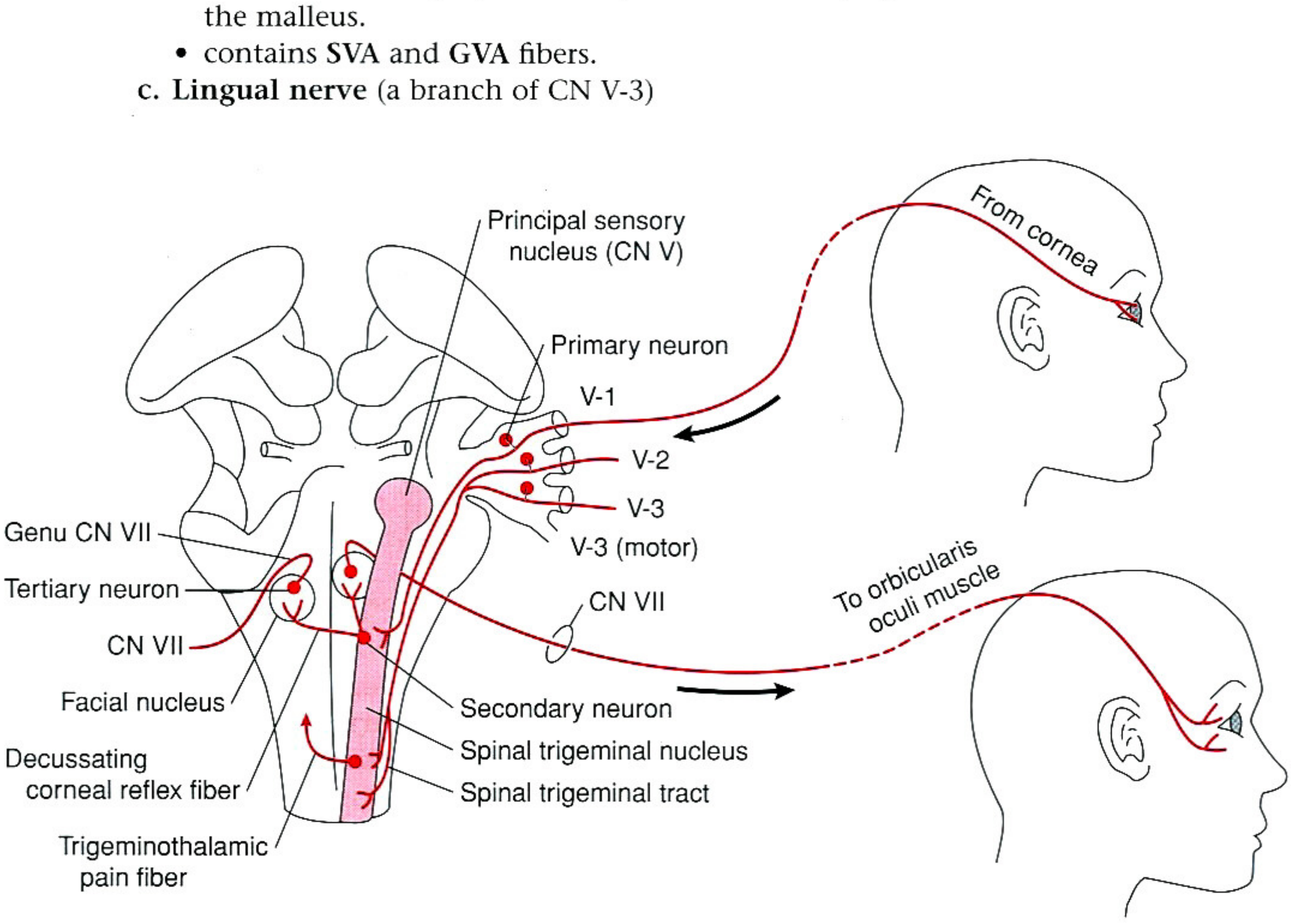


Figure 13-5. The corneal reflex pathway showing the three neurons and decussation. This reflex is consensual, like the pupillary light reflex. Second-order pain neurons are found in the caudal division of the spinal trigeminal nucleus. Second-order corneal reflex neurons are found at more rostral levels. (Reprinted with permission from Fix JD: *High-Yield Neuroanatomy,* 3rd ed. Philadelphia: Lippincott Williams & Wilkins, 2005, p 93.)

3. **GVA component**
   - has cell bodies in the geniculate ganglion.
   - innervates the soft palate and adjacent pharyngeal wall.
   - has no clinical relevance.
4. **GVE component**
   - is a parasympathetic component that innervates the **lacrimal**, **submandibular**, and **sublingual glands**.
   - contains preganglionic neurons in the superior salivatory nucleus of the caudal pons.

   a. Lacrimal pathway (see Figure 13-6)
   - begins in the superior salivatory nucleus, which projects via the intermediate nerve, the greater petrosal nerve, and the nerve of the pterygoid canal to the pterygopalatine ganglion.
   - continues as the postganglionic neurons of the pterygopalatine ganglion project through the inferior orbital fissure and via the zygomatic nerve (a branch of CN V-2) and the lacrimal nerve (a branch of CN V-1) to innervate the lacrimal gland.

   b. Submandibular pathway (see Figure 13-6)
   - begins in the superior salivatory nucleus, which projects via the intermediate nerve and chorda tympani to the submandibular ganglion.
   - continues as the postganglionic neurons of the submandibular ganglion, which project to and innervate the submandibular and sublingual glands.

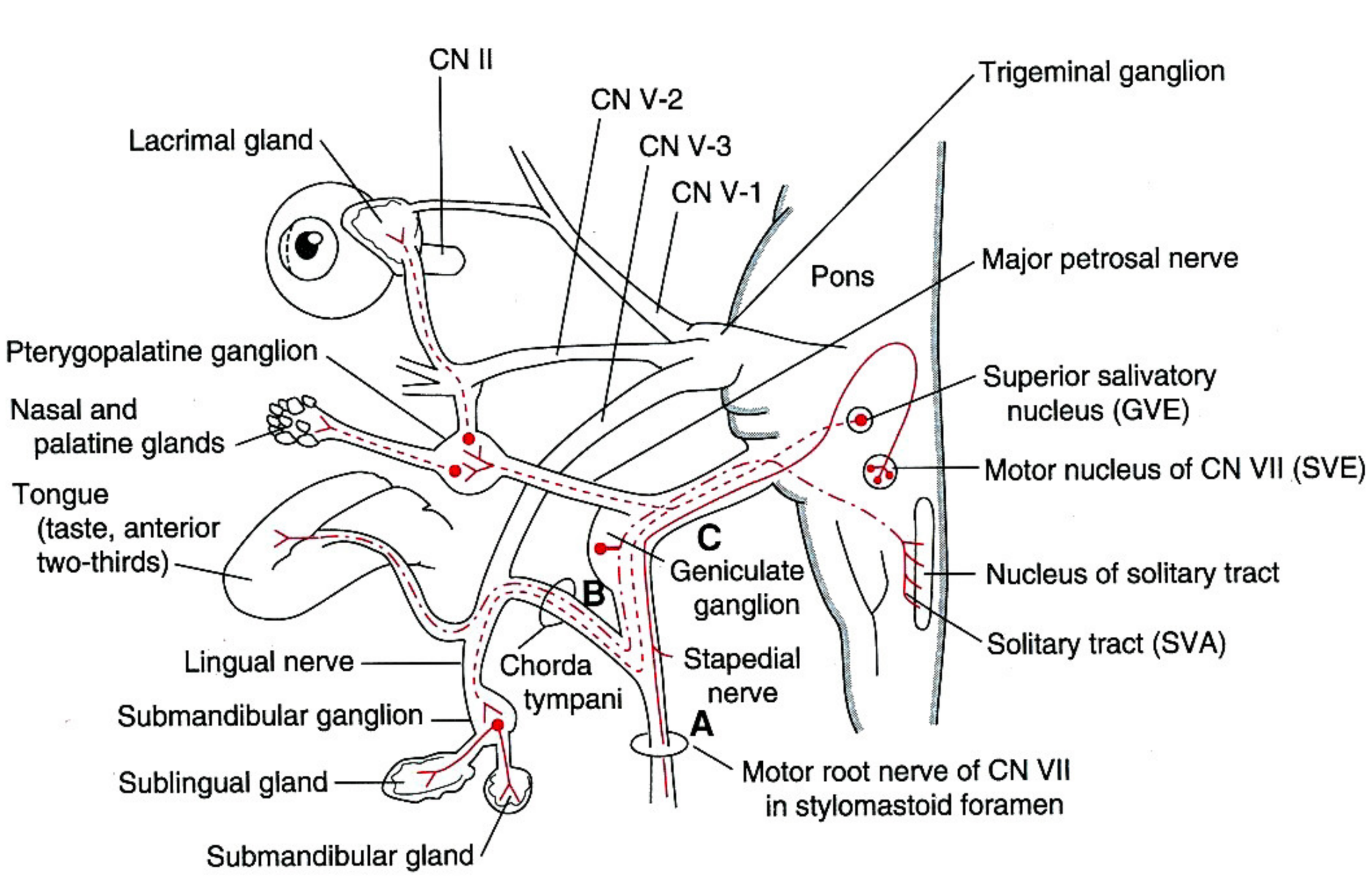


Figure 13-6. Functional components of the facial nerve (CN VII). The intermediate nerve is the sensory and visceromotor division of the seventh nerve. **A**, **B**, and **C** indicate three lesions of the nerve. Lesion **A** is at the stylomastoid foramen and spares lacrimation, nasal and palatine secretion, taste to the anterior two-thirds of the tongue, salivation, and the stapedial reflex; the patient has a lower motor neuron lesion involving the muscles of facial expression. Lesion **B** is between the geniculate ganglion and the chords tympani and spares lacrimation and secretion from the nasal palatine glands. Lesion *C* is proximal to the geniculate ganglion and is total. *GVE* = general visceral efferent; *SVA* = special visceral afferent; *SVE* = special visceral efferent.

5. **SVE component**
   - arises from the facial nucleus of the caudal pons and exits the brainstem in the CP angle.
   - enters the internal auditory meatus, traverses the facial canal, sends a branch to the stapedius muscle of the middle ear, and exits the skull via the stylomastoid foramen.
   - innervates the **muscles of facial expression**, the **stylohyoid muscle**, the **posterior belly of the digastric muscle**, and the **stapedius muscle**.

B. Clinical correlations: lesions of CN VII (see Figure 13-6)

- result in the following conditions:

1. **Flaccid paralysis** of the muscles of facial expression (upper and lower face)
2. **Loss of the corneal (blink) reflex** (efferent limb), which may lead to corneal ulceration (keratitis paralytica)
3. **Loss of taste** (ageusia) from the anterior two-thirds of the tongue
4. **Hyperacusis** (increased acuity to sounds), due to stapedius paralysis
5. **Bell palsy** (see Figure 9-11)
   - is caused by trauma to the nerve within the facial canal.
   - is a lower motor neuron (LMN) lesion with paralysis of all muscles of facial expression.
6. **Bell phenomenon**
   - is seen in Bell palsy.
   - occurs when trying to close the eyes—the affected eye looks up and out.
7. **Central facial palsy** (supranuclear palsy)
   - results from transection of corticobulbar fibers in the internal capsule.
   - results in contralateral facial weakness below the orbit.
   - is an upper motor neuron (UMN) lesion affecting the muscles of the lower face.
8. **Crocodile tears syndrome** (lacrimation during eating)
   - is caused by a facial nerve lesion proximal to the geniculate ganglion. Regenerating preganglionic salivatory fibers are misdirected to the pterygopalatine ganglion, which projects to the lacrimal gland.
9. **Möbius syndrome** (congenital oculofacial paralysis)
   - consists of a congenital facial diplegia (CN VII) and a convergent strabismus (CN VI).

IX. Vestibulocochlear Nerve (CN VIII) (see Figures 1-1, 1-7, and 13-2)

- **maintains balance** and **mediates hearing.**
- consists of two functional divisions: the **vestibular nerve** and the **cochlear nerve.**
- is a pure **SSA** nerve.
- exits the brainstem at the CP angle.
- enters the internal auditory meatus and is confined to the temporal bone.

A. Vestibular nerve (see Chapters 11 II C 6 and 12 I)

1. **General characteristics of the vestibular nerve**
   - plays a role in **equilibrium** and **balance.**
   - is associated functionally with the cerebellum (flocculonodular lobe).
   - regulates **compensatory eye movements.**
   - has first-order sensory bipolar neurons in the vestibular ganglion of the internal auditory meatus.
   - projects peripheral processes to the hair cells of the cristae ampullares of the semicircular ducts and to hair cells of the utricular and saccular maculae.
   - projects central processes to the four vestibular nuclei of the brainstem and to the flocculonodular lobe of the cerebellum.
   - conducts efferent fibers to hair cells from the brainstem.
2. **Clinical correlation: lesions of the vestibular nerve**
   - result in **disequilibrium**, **vertigo**, and **nystagmus.**

B. Cochlear nerve (see Chapter 11 III C)

1. **General characteristics of the cochlear nerve**
   - serves **audition** (hearing).
   - has first-order sensory bipolar neurons in the spiral (cochlear) ganglion of the modiolus of the cochlea, within the temporal bone.
   - projects peripheral processes to the hair cells of the organ of Corti.
   - projects central processes to the dorsal and ventral cochlear nuclei of the brainstem.
   - conducts efferent fibers to the hair cells from the brainstem.
2. **Clinical correlations: lesions of the cochlear nerve** (see Chapter 11 V B 2)
   - result in **hearing loss** (sensorineural deafness) (destructive lesions).
   - cause **tinnitus** (irritative lesions).

X. Glossopharyngeal Nerve (CN IX) (see Figures 1-1, 1-7, 13-2, and 13-3)

A. General characteristics of CN IX

- contains **GSA**, **GVA** (general visceral afferent), **SVA**, **SVE**, and **GVE** components.
- mediates **taste** (gustation), **salivation**, and (with CN X and CN XII) **swallowing**.
- mediates **input from the carotid sinus**, which contains baroreceptors that monitor arterial blood pressure.
- mediates **input from the carotid body**, which contains chemoreceptors that monitor the carbon dioxide and oxygen concentration of the blood.
- is the nerve of the third pharyngeal arch.
- is predominantly a sensory nerve.
- exits the brainstem (medulla) from the postolivary sulcus with CN X and CN XI.
- exits the skull via the jugular foramen with CN X and CN XI.

1. **GSA component**
   - innervates **part of the external ear** and the **external auditory meatus** via the auricular branch of the vagus nerve.
   - has cell bodies in the superior ganglion.
   - projects its central processes to the spinal trigeminal tract and nucleus.
2. **GVA component**
   - innervates **structures derived from the endoderm** (e.g., pharynx [foregut]).
   - innervates the **mucous membranes of the posterior third of the tongue**, **tonsil**, **upper pharynx** (soft palate), **tympanic cavity**, and **auditory tube**.
   - innervates the **carotid sinus** (baroreceptors) and the **carotid body** (chemoreceptors) via the sinus nerve.
   - has cell bodies in the inferior (petrosal) ganglion.
   - is the afferent limb of the gag reflex and the carotid sinus reflex.
3. **SVA component**
   - innervates the **taste buds** of the posterior third of the tongue.
   - has cell bodies in the inferior (petrosal) ganglion.
   - projects its central processes to the solitary tract and nucleus.
   - is the most important nerve for taste sensation.
4. **SVE component**
   - innervates the **stylopharyngeus muscle**.
   - arises from the nucleus ambiguus of the lateral medulla.
5. **GVE component**
   - is a parasympathetic component that innervates the **parotid gland**.
   - consists of preganglionic neurons in the inferior salivatory nucleus of the medulla that project, via the tympanic nerve and via the lesser petrosal nerve, through the innominate canal to the otic ganglion; postganglionic fibers from the otic ganglion project to the parotid gland via the auriculotemporal nerve (CN V-3).

B. Clinical correlations: lesions of CN IX

1. **Loss of the gag (pharyngeal) reflex** (interruption of afferent limb)
2. **Loss of the carotid sinus reflex** (interruption of the sinus nerve)
3. **Loss of taste from the posterior third of the tongue**
4. **Glossopharyngeal neuralgia**

XI. Vagal Nerve (CN X) (see Figures 1-1, 1-7, 13-2, and 13-3)

A. General characteristics of CN X

- contains **GSA**, **GVA**, **SVA**, **SVE**, and **GVE** components.
- mediates **phonation**, **swallowing** (with CN IX and CN XII), **elevation of the palate**, and **taste**.
- innervates **viscera of the neck**, **thorax**, and **abdomen**.
- is the nerve of the fourth and sixth branchial arches.
- exits the brainstem (medulla) from the postolivary sulcus.
- exits the skull via the jugular foramen with CN IX and CN XI.

1. **GSA component**
   - innervates the **infratentorial dura** (with C2 and C3) **posterior surface of the external ear**, **external auditory meatus**, and **tympanic membrane**.
   - has cell bodies in the superior (jugular) ganglion.
   - projects its central processes to the spinal trigeminal tract and nucleus.
2. **GVA component**
   - innervates the **mucous membranes** of the **pharynx**, **larynx**, **esophagus**, **trachea**, and **thoracic and abdominal viscera** (to the left colic flexure).
   - has cell bodies in the inferior (nodose) ganglion.
   - projects its central processes to the solitary tract and nucleus.
3. **SVA component**
   - innervates the **taste buds** in the **epiglottis**.
   - has cell bodies in the inferior (nodose) ganglion.
   - projects its central processes to the solitary tract and nucleus.
4. **SVE component**
   - innervates the **pharyngeal arch muscles of the larynx and pharynx**, **striated muscle of the upper esophagus**, **muscle of the uvula**, and **levator veli palatini** and **palatoglossus muscles**.
   - receives SVE input from the cranial division of the spinal accessory nerve (CN XI).
   - arises from the nucleus ambiguus in the lateral medulla.
   - provides the efferent limb of the gag reflex.
5. **GVE component** (see Figure 18-2)
   - innervates the **viscera of the neck** and the **thoracic and abdominal cavities** as far as the left colic flexure.
   - consists of preganglionic parasympathetic neurons in the dorsal motor nucleus of the medulla, which project to the intramural ganglia of the visceral organs.
   - consists of preganglionic parasympathetic neurons in the **nucleus ambiguus** of the medulla, which project to the intramural ganglia of the heart.

B. Clinical correlations: lesions of CN X (Figure 13-7)

- result in the following conditions:

1. **Ipsilateral paralysis** of the soft palate, pharynx, and larynx leading to **dysphonia** (hoarseness), **dyspnea**, **dysarthria**, and **dysphagia**
2. **Loss of the gag (palatal) reflex** (efferent limb)
3. **Anesthesia of the pharynx and larynx**, leading to unilateral loss of the cough reflex
4. **Aortic aneurysms and tumors of the neck and thorax**
   - frequently compress the vagal nerve.

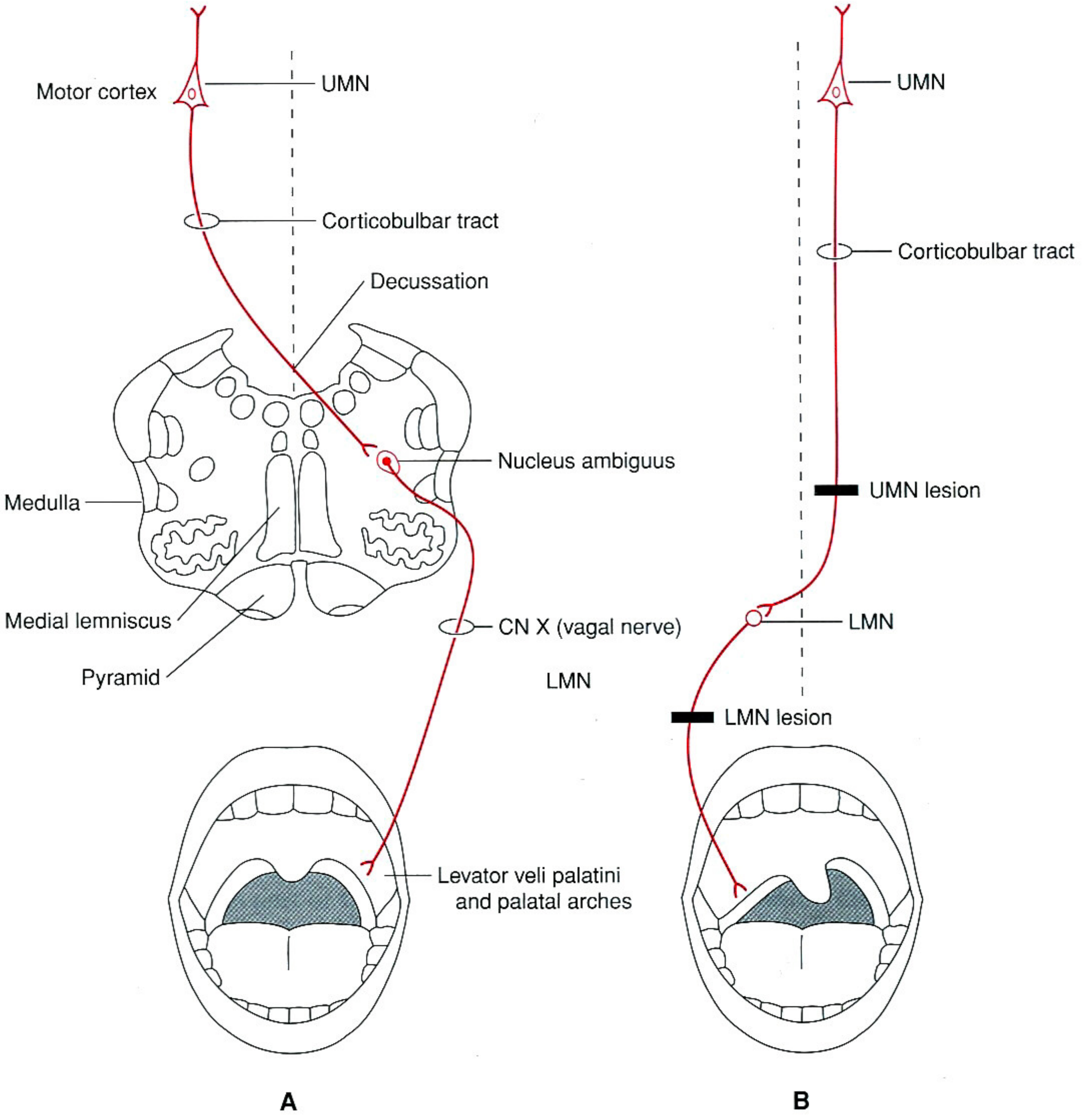


Figure 13-7. Innervation of the palatal arches and uvula. Sensory innervation is mediated by the glossopharyngeal nerve (CN IX). Motor innervation of the palatal arches and uvula is mediated by the vagus nerve (CN X). **(A)** A normal palate and uvula in a person who is saying "ah." **(B)** A patient with an upper motor neuron (*UMN*) lesion (*left*) and a lower motor neuron (*LMN*) lesion (*right*). When this patient says "ah," the palatal arches sag. The uvula deviates toward the intact (left) side. (Modified with permission from DeMyer WE: *Technique of the Neurological Examination; A Programmed Text,* 4th ed. New York, McGraw-Hill, 1994, 6–9, p. 191.)

XII. Accessory Nerve (CN XI) (Figure 13-8; see Figures 1-1, 1-7, and 13-3)

A. General characteristics of CN XI

- contains the **SVE** component.
- mediates **head and shoulder movement** and innervates **laryngeal muscles**.
- includes the following divisions:
  1. **Cranial division**
     - arises from the **nucleus ambiguus** of the medulla.
     - exits the medulla from the postolivary sulcus and joins the vagal nerve (CN X).

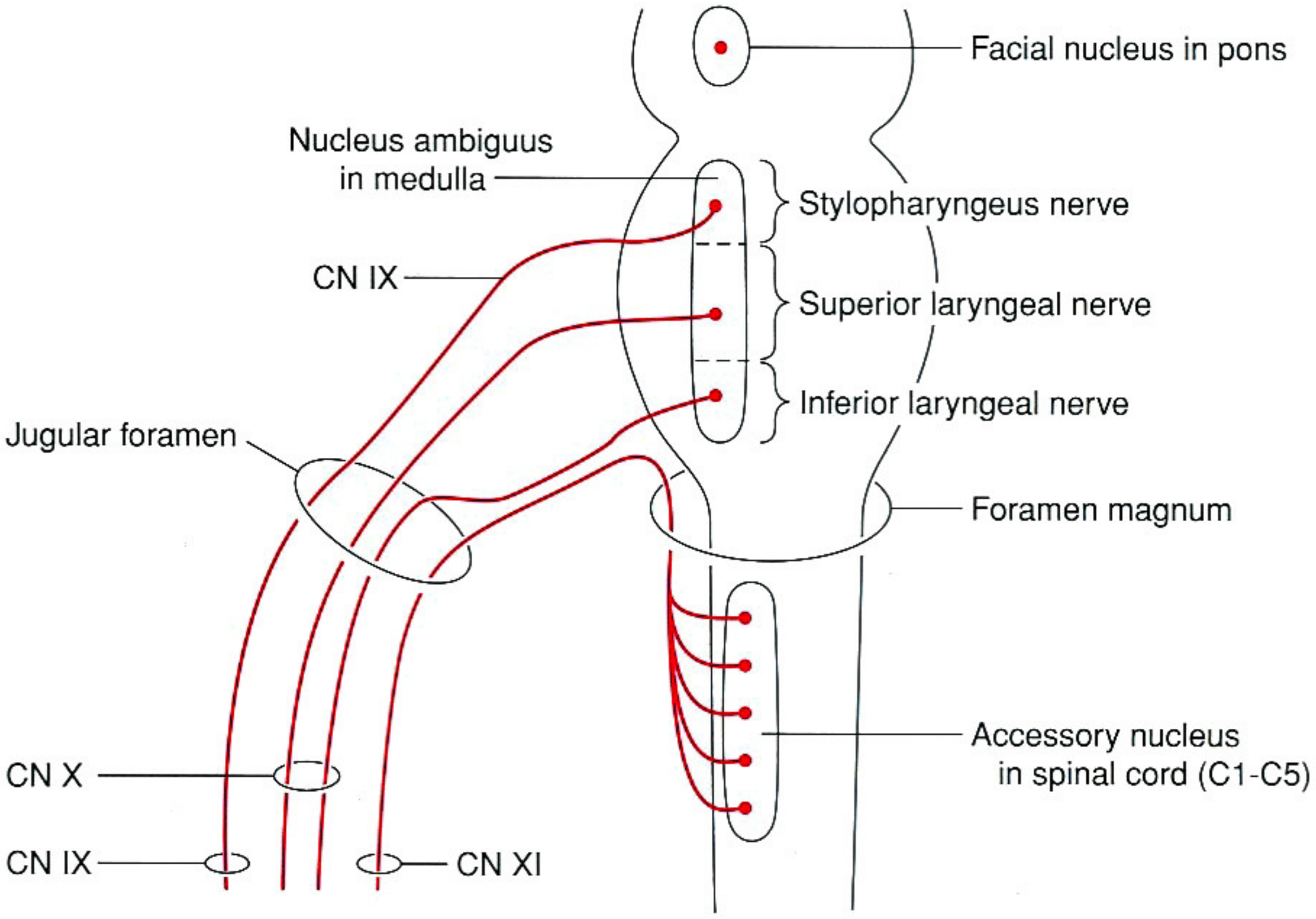


Figure 13-8. The cranial and spinal divisions of the accessory nerve (CN IX). The cranial division hitchhikes a ride with the accessory nerve, then joins the vagal nerve to become the inferior (recurrent) laryngeal nerve. The recurrent laryngeal nerve innervates the intrinsic muscles of the larynx, except for the cricothyroid muscle. The spinal division innervates the trapezoid and sternocleidomastoid muscles. Three nerves pass through the jugular foramen (glomus jugulare tumor). (Reprinted with permission from Fix JD: *High-Yield Neuroanatomy,* 3rd ed. Philadelphia: Lippincott Williams & Wilkins, 2005, p 101.)

- exits the skull via the jugular foramen with CN IX and CN X.
- innervates the **intrinsic muscles of the larynx** via the inferior (recurrent) laryngeal nerve, with the exception of the cricothyroid muscle.

2. **Spinal division**
   - arises from the ventral horn of cervical segments C1 to C6.
   - Spinal roots exit the spinal cord laterally between the ventral and dorsal spinal roots, ascend through the foramen magnum, and exit the skull via the jugular foramen.
   - innervates the **sternocleidomastoid** (with C2) and **trapezius muscles** (with C3 and C4).

B. Clinical correlations: lesions of CN XI

- result in the following conditions:

1. **Paralysis of the sternocleidomastoid muscle**
   - results in difficulty in turning the head to the side opposite the lesion.
2. **Paralysis of the trapezius muscle**
   - results in a shoulder droop.
   - results in the inability to shrug the ipsilateral shoulder.
3. **Paralysis of the larynx** occurs if the cranial root is involved.

XIII. Hypoglossal Nerve (CN XII) (see Figures 1-1, 1-7, and 13-3)

A. General characteristics of CN XII

- mediates **tongue movement**.
- is a pure **GSE** nerve.
- arises from the hypoglossal nucleus of the medulla.

- exits the medulla in the preolivary sulcus.
- exits the skull via the hypoglossal canal.
- innervates **intrinsic and extrinsic muscles of the tongue.**
- has three extrinsic muscles: the genioglossus, styloglossus, and hyoglossus.

B. Clinical correlations: CN XII (Figure 13-9)

- When it is transected, **hemiparalysis of the tongue** results.
- When it is protruded, the tongue points toward the weak side due to the unopposed action of the opposite genioglossus muscle.

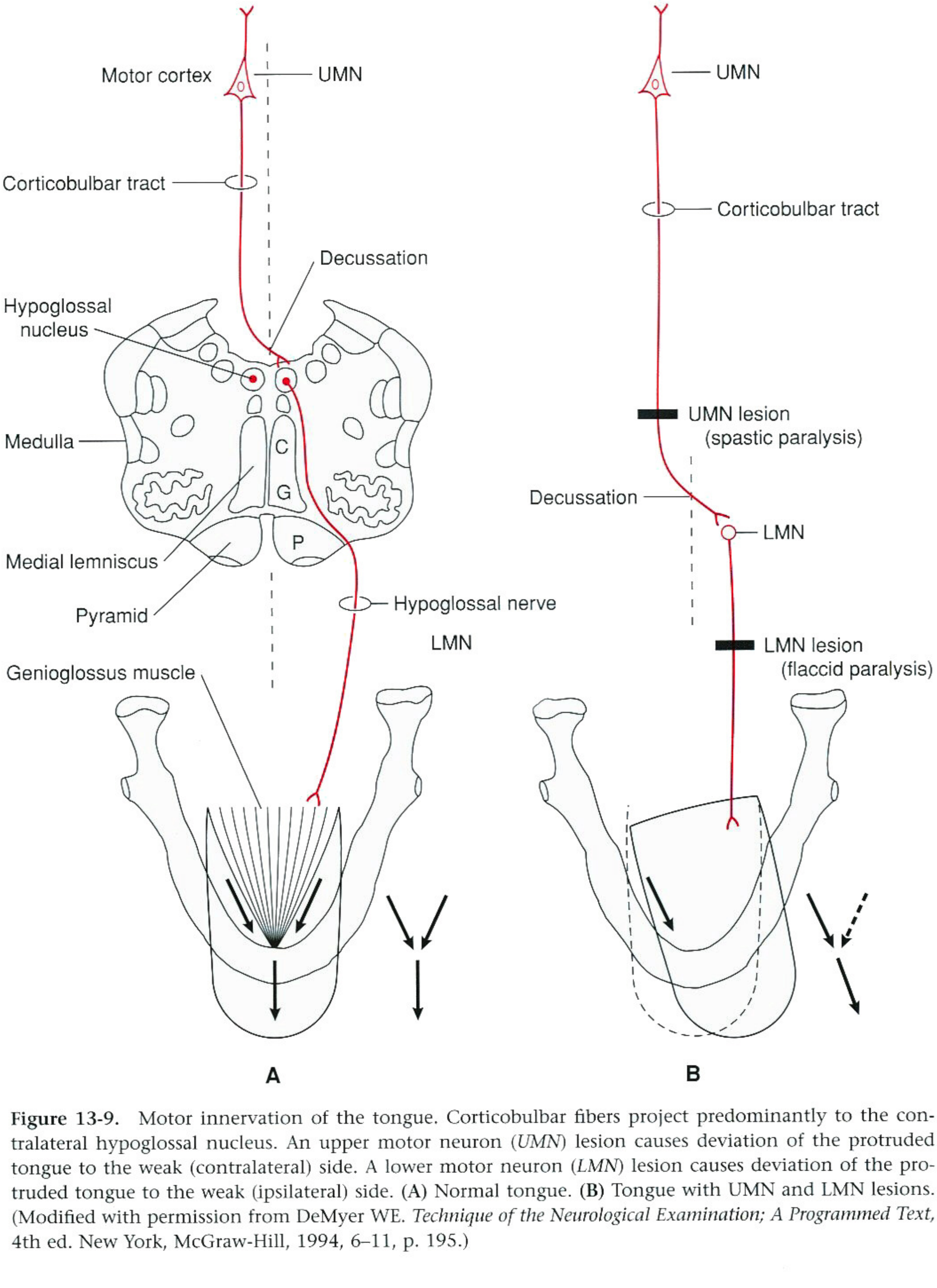


Figure 13-9. Motor innervation of the tongue. Corticobulbar fibers project predominantly to the contralateral hypoglossal nucleus. An upper motor neuron (*UMN*) lesion causes deviation of the protruded tongue to the weak (contralateral) side. A lower motor neuron (*LMN*) lesion causes deviation of the protruded tongue to the weak (ipsilateral) side. (**A**) Normal tongue. (**B**) Tongue with UMN and LMN lesions. (Modified with permission from DeMyer WE. *Technique of the Neurological Examination; A Programmed Text,* 4th ed. New York, McGraw-Hill, 1994, 6–11, p. 195.)

REVIEW TEST

1. A 50-year-old family physician has vertical diplopia; the man feels unsure when descending stairs. He can eliminate the double vision by tilting his chin toward the paretic side. Which of the following extraocular muscles is responsible for the ocular malalignment?

(A) Superior rectus
(B) Inferior rectus
(C) Inferior oblique
(D) Lateral rectus
(E) Superior oblique

2. Anosmia results from damage to which cranial nerve?

(A) CN I
(B) CN II
(C) CN III
(D) CN IV
(E) CN V

3. A 50-year old retired army major complained of severe pain in the ear and throat. Pain was episodic and triggered by swallowing, chewing, coughing, and laughing. Symptoms included loss of gag (pharyngeal) reflex; analgesia and anesthesia in the region of the tonsils; and dysphagia. A lesion of which cranial nerve would produce these neurologic deficits?

(A) Facial nerve
(B) Glossopharyngeal nerve
(C) Hypoglossal nerve
(D) Trigeminal nerve
(E) Vagal nerve

4. Which cranial nerve's fibers are myelinated by oligodendrocytes?

(A) CN I
(B) CN II
(C) CN III
(D) CN VII
(E) CN X

5. A 25-year-old woman's a neck was injured in an automobile accident. At examination, she reports difficulty in turning her head away from the side of her neck that was injured. She also has a visible shoulder droop. Which nerve was likely damaged?

(A) CN VIII
(B) CN IX
(C) CN X
(D) CN XI
(E) CN XII

Questions 6 to 10

The response options for items 6 to 10 are the same. Select one answer for each item in the set.

(A) Glossopharyngeal nerve
(B) Accessory nerve
(C) Trigeminal nerve
(D) Facial nerve
(E) Vagal nerve

Match each description with the appropriate nerve.

6. Innervates the parotid gland

7. Is the efferent limb of the corneal reflex

8. Is the efferent limb of the gag reflex

9. Innervates the infratentorial dura

10. Is a pure motor nerve

Questions 11 to 16

The response options for items 11 to 16 are the same. Select one answer for each item in the set.

(A) Foramen jugular
(B) Innominate canal
(C) Foramen magnum
(D) Foramen ovale
(E) Foramen rotundum
(F) Foramen spinosum
(G) Foramen stylomastoideum
(H) Superior orbital fissure

Match the anatomic structure(s) below with the foramen or fissure through which it passes.

11. A branch of the maxillary artery

12. The nerve that innervates the buccinator muscle

13. The nerve that innervates the skin of the upper lip

14. CN IX, CN X, and CN XI

15. The nerve that projects to the otic ganglion

16. Four cranial nerves traverse this orifice

ANSWERS AND EXPLANATIONS

1–E. The superior oblique muscle depresses, abducts, and intorts the eye. Paralysis of this muscle results in extorsion and weakness of downward gaze. Head tilting compensates for extorsion.

2–A. Anosmia, a loss of olfactory sensation, results from damage to the olfactory nerve, or CN I.

3–B. Glossopharyngeal neuralgia has the following neurologic deficits: Excruciating, paroxysmal pain that comes from the tonsillar area and radiates into the ear; loss of taste sensation from the posterior third of the tongue; and loss of palatal and gag reflexes. Potential causes of glossopharyngeal impairment include fractures of the skull base, thrombosis of the sigmoid sinus, tumors, and aneurysms of the posterior fossa. Pain may be triggered by a blood vessel pressing on the nonmyelinated root of the glossopharyngeal nerve, and relocation of the vessel may alleviate symptoms. Treatment is with carbamazepine and other antiepileptic drugs.

4–B. The fibers of the optic nerve (CN II) are myelinated by oligodendrocytes. This is an important distinction from the other cranial nerves, whose fibers are myelinated by Schwann cells, because the optic nerve is considered a tract of the central nervous system and thus incapable of regeneration.

5–D. The accessory nerve (CN XI) mediates head and shoulder movement and innervates laryngeal muscles. Lesions result in paralysis of the sternocleidomastoid muscle, making it difficult to turn the head to the side opposite the lesion, and paralysis of the trapezius muscle, resulting in a shoulder droop and inability to shrug the shoulder on the side of the lesion.

6–A. The glossopharyngeal nerve (CN IX) innervates the parotid gland via the tympanic and lesser petrosal nerves, the otic ganglion, and the auriculotemporal nerve.

7–D. The facial nerve (CN VII) provides the efferent limb of the corneal reflex (orbicularis oculi muscle).

8–E. The vagal nerve (CN X) provides the efferent limb of the gag reflex (muscles of the soft palate). The glossopharyngeal nerve provides the afferent limb of the gag reflex.

9–E. The vagal nerve (CN X) innervates, via the recurrent meningeal ramus, the infratentorial dura (the dura of the posterior cranial fossa).

10–B. The accessory nerve (CN XI) is a pure SVE motor nerve. The cranial division innervates, via the recurrent laryngeal nerve, the intrinsic muscles of the larynx; the spinal division innervates, via motor branches, the sternocleidomastoid muscle and upper parts of the trapezius muscle.

11–F. The middle meningeal artery, a branch of the maxillary artery, traverses the foramen spinosum.

12–G. The facial nerve (CN VII) exits the base of the skull via the stylomastoid foramen; CN VII innervates the muscles of facial expression, including the buccinator muscle.

13–E. The maxillary nerve (CN V-2) exits the skull via the foramen rotundum.

14–A. CN IX, CN X, and CN XI exit the posterior cranial fossa via the jugular foramen.

15–B. The lesser petrosal nerve of CN IX passes through the innominate canal to synapse with postganglionic neurons of the otic ganglion. The innominate canal lies between the foramen ovale and the foramen spinosum.

16–H. CN III, CN IV, CN VI, and CN V-1 pass through the superior orbital fissure.

CHAPTER 14

Lesions of the Brainstem

I. Introduction: Lesions of the Brainstem

- are most frequently syndromes of arterial occlusion or circulatory insufficiency that involve the vertebrobasilar system.

II. Vascular Lesions of the Medulla

- result from occlusion of the vertebral artery or its branches (i.e., the anterior and posterior spinal arteries and the posterior inferior cerebellar artery [PICA]).

A. Medial medullary syndrome (Figure 14-1A)

- results from occlusion of the anterior spinal artery.
- includes the following affected **structures** and resultant **deficits**:
  1. **Corticospinal tract**
     - contralateral hemiparesis of the trunk and extremities
  2. **Medial lemniscus**
     - contralateral loss of proprioception, discriminative tactile sensation, and vibration sensation from the trunk and extremities
  3. **Hypoglossal nerve roots (intra-axial fibers)**
     - ipsilateral flaccid paralysis of the tongue

B. Lateral medullary syndrome (PICA syndrome) (see Figure 14-1B)

- is also called Wallenberg syndrome.
- results from occlusion of the vertebral artery or one of its medullary branches (e.g., PICA).
- includes the following affected **structures** and resultant **deficits**:
  1. **Vestibular nuclei (medial and inferior)**
     - nystagmus, nausea, vomiting, and vertigo
  2. **Inferior cerebellar peduncle**
     - ipsilateral cerebellar signs (dystaxia, dysmetria, dysdiadochokinesia)
  3. **Nucleus ambiguus of cranial nerve (CN) IX, CN X, and CN XI (somatic visceral efferent [SVE])**
     - ipsilateral laryngeal, pharyngeal, and palatal paralysis (loss of the gag reflex [efferent limb], dysarthria, dysphagia, and dysphonia [hoarseness])
  4. **Glossopharyngeal nerve roots (intra-axial fibers)**
     - loss of the gag reflex (afferent limb)
  5. **Vagal nerve roots (intra-axial fibers)**
     - neurologic deficits same as those seen in lesion of the nucleus ambiguus
  6. **Spinothalamic tracts**
     - contralateral loss of pain and temperature sensation from the trunk and extremities

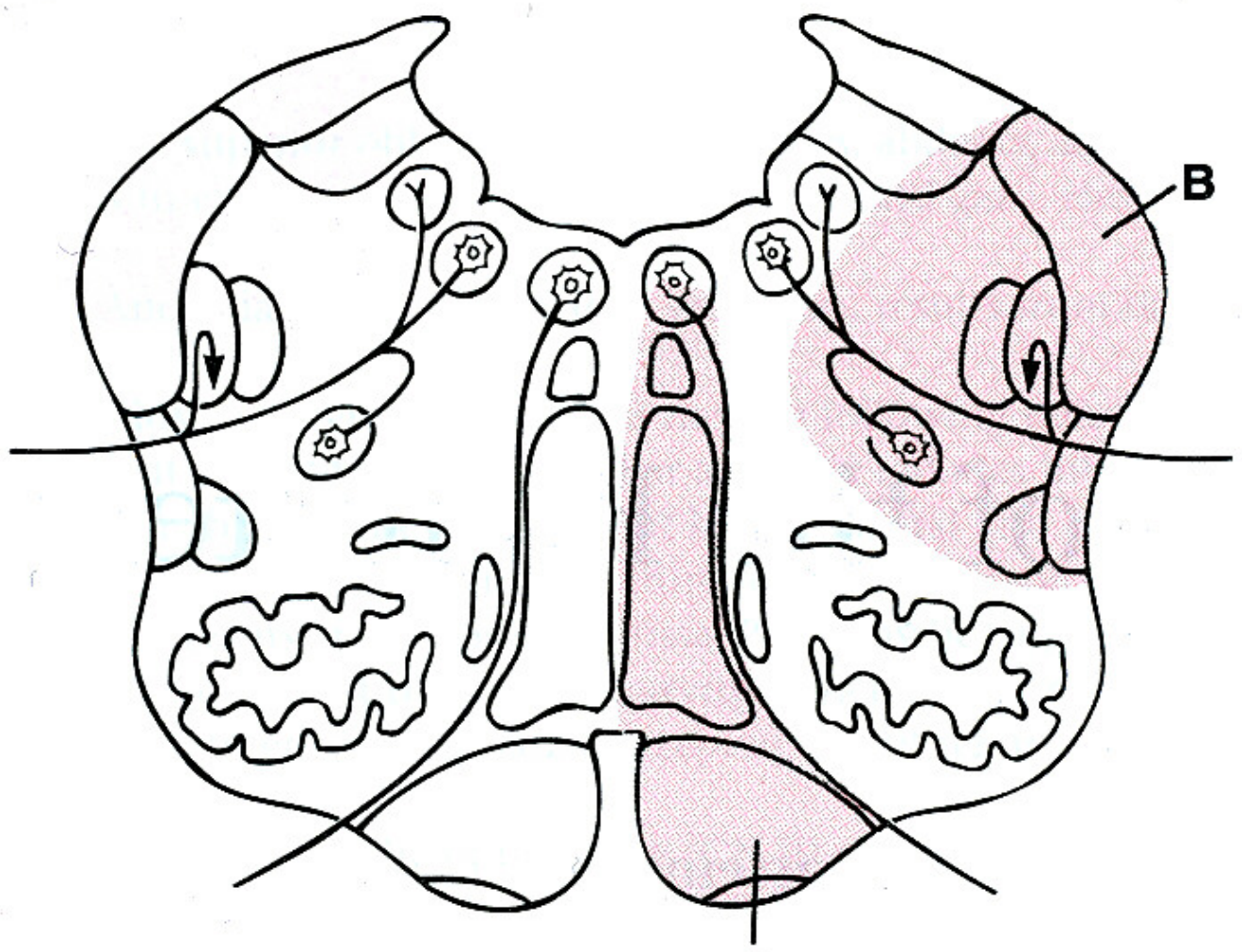


Figure 14-1. Vascular lesions of the caudal medulla at the level of the hypoglossal nucleus of CN XII and the dorsal motor nucleus of CN X. **(A)** Medial medullary syndrome (anterior spinal artery). **(B)** Lateral medullary syndrome (PICA syndrome).

7. **Spinal trigeminal nucleus and tract**
   - ipsilateral loss of pain and temperature sensation from the face
8. **Descending sympathetic tract**
   - ipsilateral Horner syndrome (ptosis, miosis, hemianhidrosis, vasodilation, and apparent enophthalmos)

III. Vascular Lesions of the Pons

- result from occlusion of the basilar artery or its branches (the anterior inferior cerebellar artery [AICA], transverse pontine arteries, and superior cerebellar artery).

A. Medial inferior pontine syndrome (Figure 14-2A)

- results from occlusion of the paramedian branches of the basilar artery.

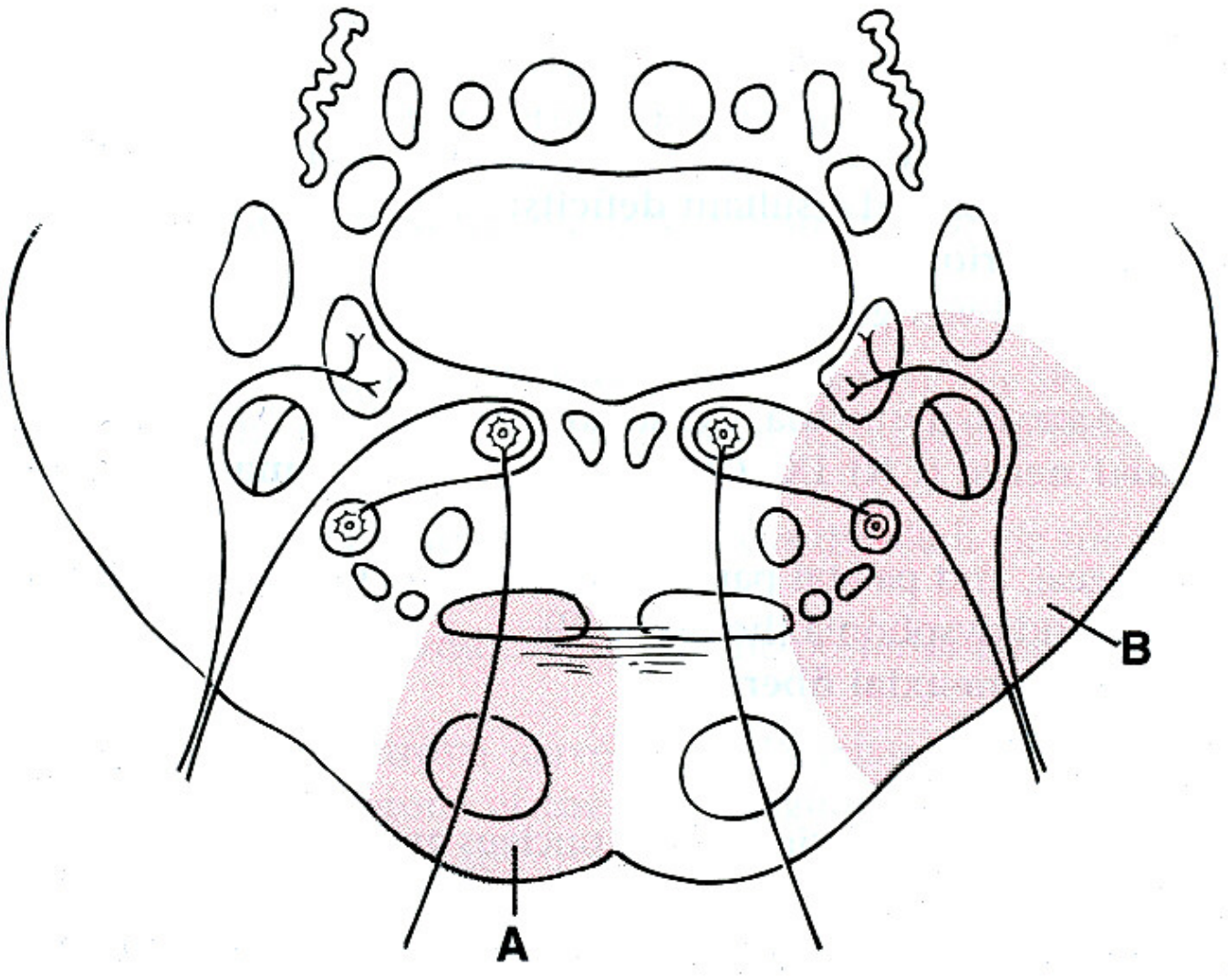


Figure 14-2. Vascular lesions of the caudal pons at the level of the abducent nucleus of CN VI and the facial nucleus of CN VII. **(A)** Medial inferior pontine syndrome. **(B)** Lateral inferior pontine syndrome (AICA syndrome).

- includes the following affected **structures** and resultant **deficits**:
  1. **Abducent nerve roots (intra-axial fibers)**
     - ipsilateral lateral rectus paralysis
  2. **Corticobulbar tracts**
     - contralateral weakness of the lower face
  3. **Corticospinal tracts**
     - contralateral hemiparesis of the trunk and extremities
  4. **Base of the pons (middle cerebellar peduncle)**
     - ipsilateral limb and gait ataxia
  5. **Medial lemniscus**
     - contralateral loss of proprioception, discriminative tactile sensation, and vibration sensation from the trunk and extremities

B. Lateral inferior pontine syndrome (AICA syndrome) (see Figure 14-2B)

- results from occlusion of a long circumferential branch of the basilar artery, AICA.
- includes the following affected **structures** and resultant **deficits**:
  1. **Facial nucleus and intra-axial nerve fibers**
     - ipsilateral facial nerve paralysis
     - loss of taste from the anterior two-thirds of the tongue
     - loss of the corneal and stapedial reflexes
  2. **Cochlear nuclei and intra-axial nerve fibers**
     - unilateral central nerve deafness
  3. **Vestibular nuclei and intra-axial nerve fibers**
     - nystagmus, nausea, vomiting, and vertigo
  4. **Spinal trigeminal nucleus and tract**
     - ipsilateral loss of pain and temperature sensation from the face
  5. **Middle and inferior cerebellar peduncles**
     - ipsilateral limb and gait dystaxia
  6. **Spinothalamic tracts**
     - contralateral loss of pain and temperature sensation from the trunk and extremities
  7. **Descending sympathetic tract**
     - ipsilateral Horner syndrome (ptosis, miosis, hemianhidrosis, vasodilation, and apparent enophthalmos)

C. Lateral midpontine syndrome

- results from occlusion of a short circumferential branch of the basilar artery.
- includes the following affected **structures** and resultant **deficits**:
  1. **Trigeminal nuclei and nerve root (motor and principal sensory nuclei)**
     - complete ipsilateral trigeminal paralysis, including:
       a. **Paralysis of the muscles of mastication**
       b. **Jaw deviation to the paretic side** (due to unopposed action of the intact lateral pterygoid muscle)
       c. **Facial hemianesthesia** (pain, temperature, touch, and proprioception)
       d. **Loss of the corneal reflex** (afferent limb of CN V-1)
  2. **Middle cerebellar peduncle (base of the pons)**
     - ipsilateral limb and gait dystaxia

D. Lateral superior pontine syndrome

- results from occlusion of a long circumferential branch of the basilar artery, the **superior cerebellar artery**.
- includes the following affected **structures** and resultant **deficits**:
  1. **Superior and middle cerebellar peduncles**
     - ipsilateral limb and trunk dystaxia
  2. **Dentate nucleus**
     - signs similar to those seen with damage to the superior cerebellar peduncle (dystaxia, dysmetria, and intention tremor)

3. **Spinothalamic and trigeminothalamic tracts**
   - contralateral loss of pain and temperature sensation from the trunk, extremities, and face
4. **Descending sympathetic tract**
   - ipsilateral Horner syndrome (ptosis, miosis, hemihidrosis, and apparent enophthalmos)
5. **Medial lemniscus (lateral division [gracilis])**
   - contralateral loss of proprioception, discriminative tactile sensation, and vibration sensation from the trunk and lower extremity

E. Locked-in syndrome (pseudocoma)

- results from infarction of the base of the superior pons; infarcted structures include the corticobulbar and corticospinal tracts, resulting in quadriplegia and paralysis of the lower cranial nerves.
- also may result from **central pontine myelinolysis.**
- Communication occurs only by blinking or moving the eyes vertically.

IV. Lesions of the Midbrain

- result from vascular occlusion of the mesencephalic branches of the posterior cerebral artery.
- may result from aneurysms of the posterior circle of Willis.
- may result from tumors of the pineal region.
- may result from hydrocephalus.

A. Dorsal midbrain (Parinaud) syndrome (Figure 14-3A)

- is frequently the result of a **pinealoma** or **germinoma** of the pineal region.
- includes the following affected **structures** and resultant **deficits**:
  1. **Superior colliculus and pretectal area**
     - paralysis of upward and downward gaze, pupillary disturbances, and absence of convergence
  2. **Cerebral aqueduct**
     - noncommunicating hydrocephalus (as a result of compression from a pineal tumor)

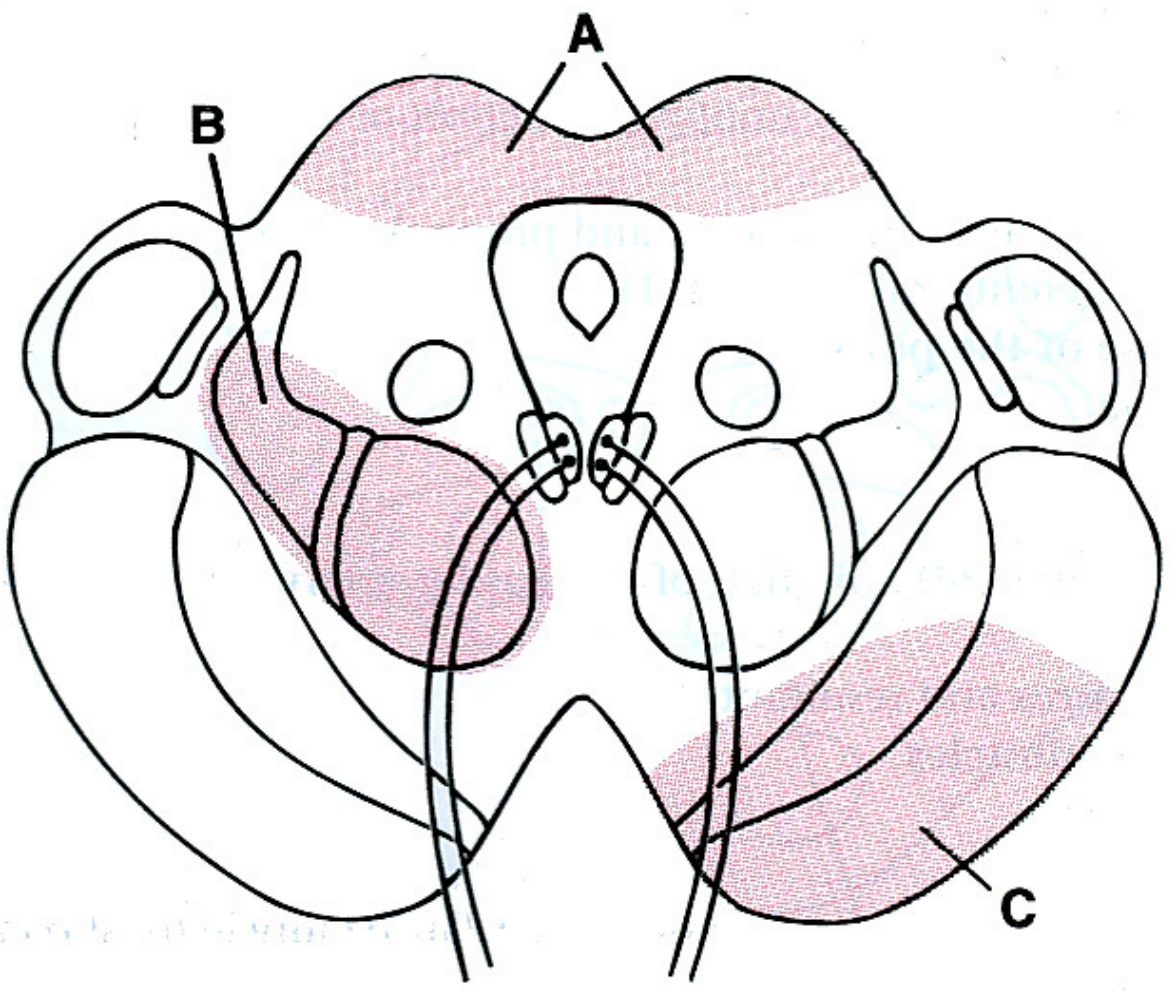


Figure 14-3. Lesions of the rostral midbrain at the level of the superior colliculus and oculomotor nucleus of CN III. **(A)** Dorsal midbrain (Parinaud) syndrome. **(B)** Paramedian midbrain (Benedikt) syndrome. **(C)** Medial midbrain (Weber) syndrome.

B. Paramedian midbrain (Benedikt) syndrome (see Figure 14-3B)

- results from occlusion or hemorrhage of the paramedian midbrain branches of the posterior cerebral artery.
- includes the following affected **structures** and resultant **deficits**:
  1. **Oculomotor nerve roots (intra-axial fibers)**
     - complete **ipsilateral oculomotor nerve paralysis**
     - **eye abduction and depression** because of the unopposed action of the lateral rectus (CN VI) and the superior oblique (CN IV) muscles
     - severe **ptosis** (paralysis of the levator palpebrae muscle)
     - ipsilateral **fixed and dilated pupil** (complete internal ophthalmoplegia)
  2. **Red nucleus and dentatorubrothalamic tract**
     - contralateral cerebellar dystaxia with intention tremor
  3. **Medial lemniscus**
     - contralateral loss of proprioception, discriminative tactile sensation, and vibration sensation from trunk and extremities

C. Medial midbrain (Weber) syndrome (see Figure 14-3C)

- results from occlusion of midbrain branches of the posterior cerebral artery and aneurysms of the circle of Willis.
- includes the following **structures** and resultant **deficits**:
  1. **Oculomotor nerve roots (intra-axial fibers)** (see IV B 1)
  2. **Corticobulbar tracts**
     - contralateral weakness of the lower face (CN VII), tongue (CN XII), and palate (CN X)
  3. **Corticospinal tracts**
     - contralateral hemiparesis of the trunk and extremities

V. Acoustic Neuroma (Schwannoma) (Figure 14-4)

- is a benign tumor of the Schwann cells affecting the vestibulocochlear nerve (CN VIII).
- is a posterior fossa tumor of the internal auditory meatus and the cerebellopontine (CP) angle.
- frequently compresses the facial nerve (CN VII), which accompanies CN VIII in the CP angle and internal auditory meatus.
- may impinge on the pons and affect the spinal trigeminal tract (CN V).
- includes the following affected **structures** and resultant **deficits**:

A. Cochlear nerve of CN VIII

- unilateral nerve deafness and tinnitus

B. Vestibular nerve of CN VIII

- vertigo, nystagmus, nausea, vomiting, and unsteadiness of gait

C. Facial nerve (CN VII)

- facial weakness and loss of corneal reflex (efferent limb)

D. Spinal trigeminal tract (CN V)

- paresthesias and anesthesia of ipsilateral face
- loss of the corneal reflex (afferent limb)

E. Abducent nerve (CN VI) (in advanced cases with large tumors)

- diplopia

F. Corticospinal tract [in advanced cases with large tumors]

- contralateral spastic paresis

VI. Internuclear Ophthalmoplegia (INO)

- is also known as medial longitudinal fasciculus (MLF) syndrome, which results from a lesion of the MLF. Lesions occur in the dorsomedial pontine tegmentum and may affect one or both MLFs.
- is a frequent sign of multiple sclerosis.
- results in medial rectus palsy on attempted lateral gaze and monocular nystagmus in the abducting eye with normal convergence.
- Lesions of the abducent nucleus of CN VI result in all MLF signs and a lateral rectus paralysis with internal strabismus.

VII. Jugular Foramen (Vernet) Syndrome

- affects CN IX, CN X, and CN XI.
- includes the following affected **structures** and resultant **deficits**:

A. Glossopharyngeal nerve (CN IX)

- loss of the gag reflex (afferent limb)
- loss of taste sensation in the posterior third of the tongue
- unilateral loss of the carotid sinus reflex

B. Vagal nerve (CN X)

- laryngeal paralysis with dysarthria, dysphagia, and dysphonia (hoarseness)
- palatal paralysis with loss of the gag reflex (efferent limb)

C. Accessory nerve (CN XI)

- weakness of the sternocleidomastoid and upper trapezius muscles (the shoulder droops)

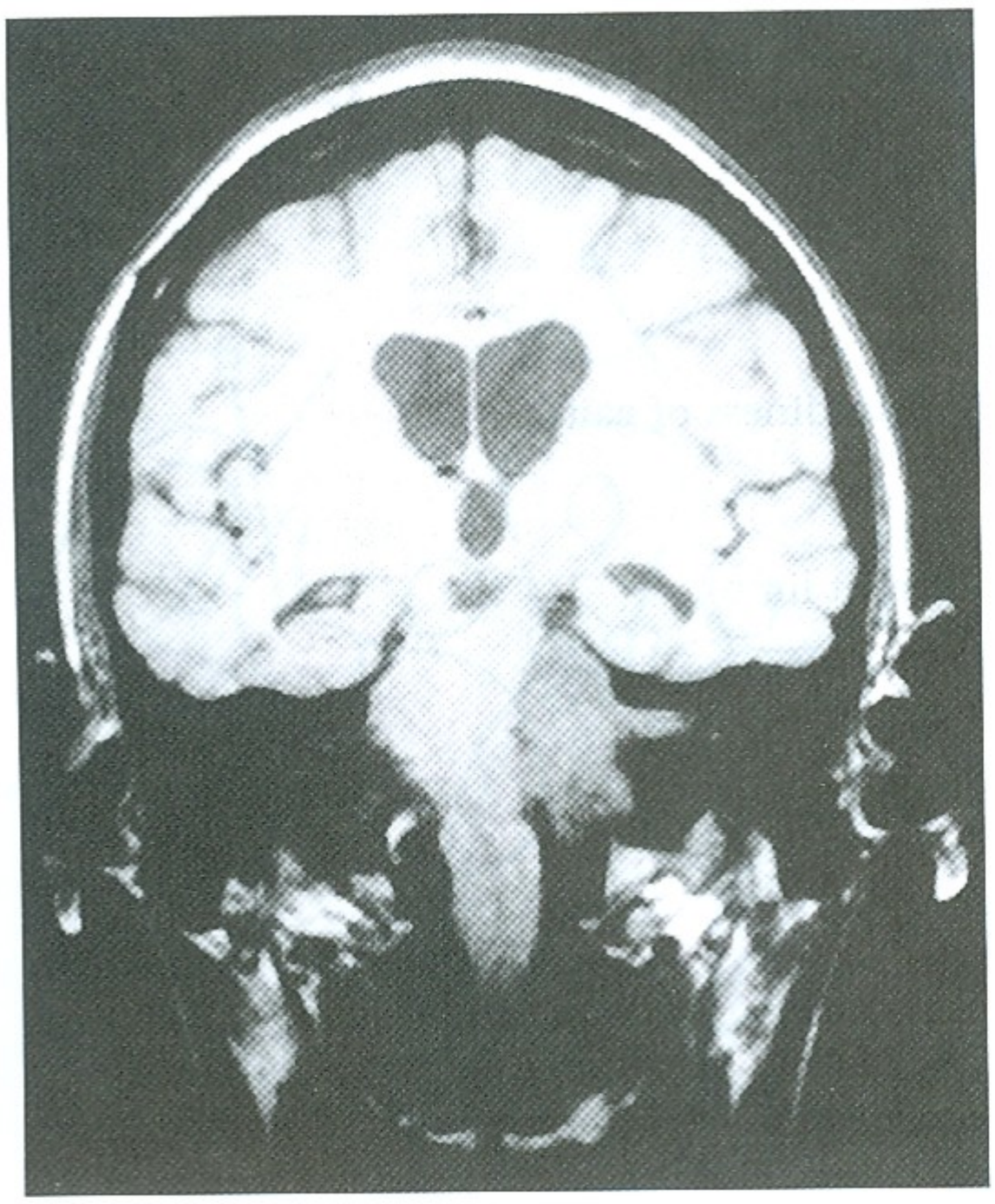


Figure 14-4. Magnetic resonance image of an acoustic neuroma. This coronal section shows dilation of the ventricles. The vestibulocochlear nerve is visible in the left internal auditory meatus. The tumor indents the lateral pons. Cranial nerve palsies include CN V, VII, and VIII. Symptoms include unilateral deafness, facial anesthesia and weakness, and an absent coronal reflex. This is a T_1-weighted image. (Reprinted with permission from Fix JD: High-Yield Neuroanatomy, 3rd ed. Philadelphia, Lippincott Williams & Wilkins, 2005, p 108.)

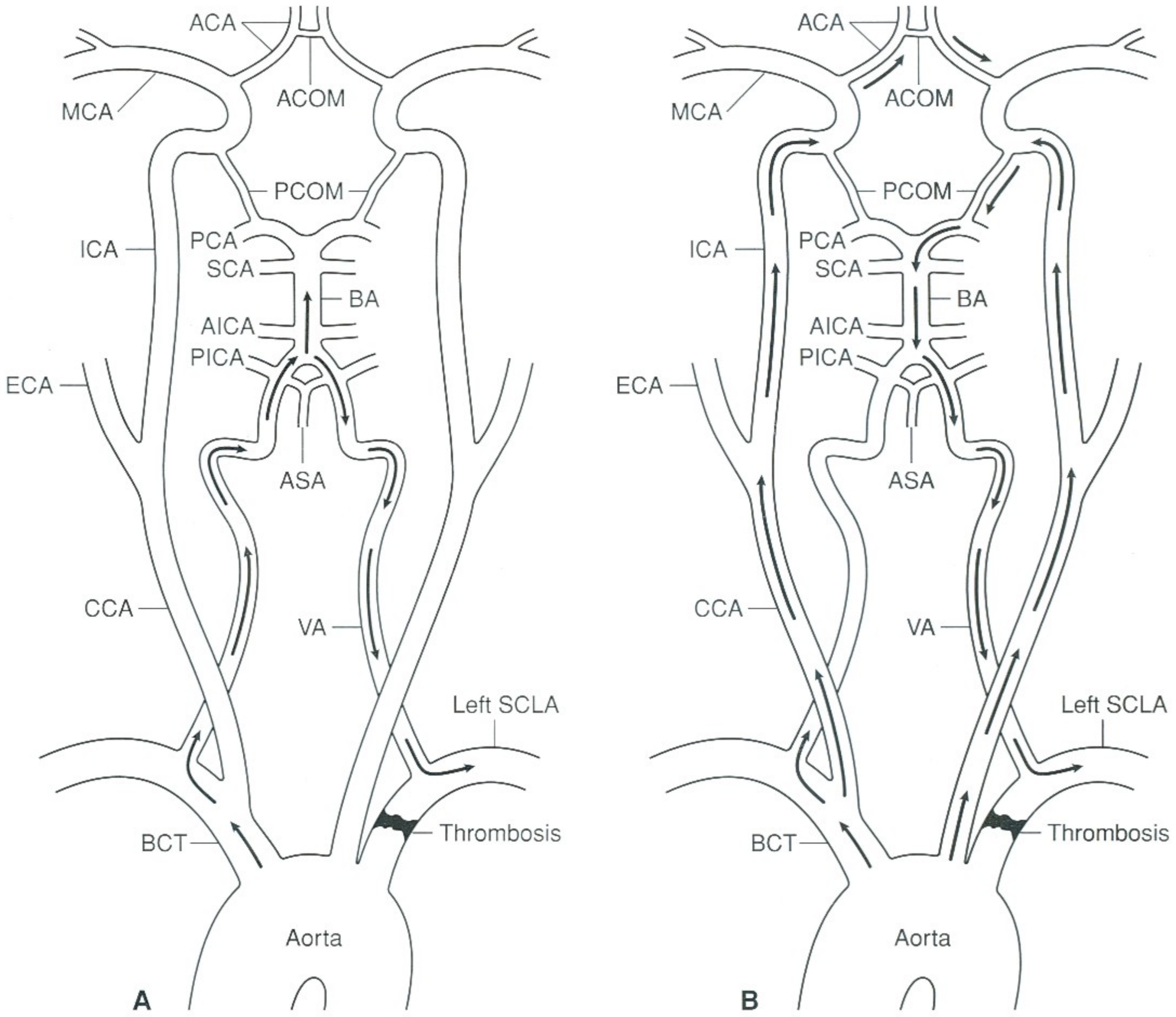


Figure 14-5. Anatomy of subclavian steal syndrome. Thrombosis of the proximal part of the subclavian artery on the left side results in retrograde blood flow through the ipsilateral vertebral artery and into the left subclavian artery. Blood can be shunted from the right vertebral artery and down the left vertebral artery (**A**). Blood may also reach the left vertebral artery via the carotid circulation (**B**). *ACA* = anterior cerebral artery; *ACOM* = anterior communicating artery; *AICA* = anterior inferior cerebellar artery; *ASA* = anterior spinal artery; *BA* = basilar artery; *BCT* = brachiocephalic trunk; *CCA* = common carotid artery; *ECA* = external carotid artery; *ICA* = internal carotid artery; *MCA* = middle cerebral artery; *PCA* = posterior cerebral artery; *PCOM* = posterior communicating artery; *PICA* = posterior inferior cerebellar artery; *SCA* = superior cerebellar artery; *SCLA* = subclavian artery; *VA* = vertebral artery. (Reprinted with permission from Fix JD: *High-Yield Neuroanatomy,* 3rd ed. Philadelphia: Lippincott Williams & Wilkins, 2005, p 109.)

VIII. Subclavian Steal Syndrome (Figure 14-5)

- results from thrombosis of the left subclavian artery proximal to the vertebral artery. Blood is shunted retrograde down the vertebral artery and into the left subclavian artery.
- leads to the following clinical signs: transient weakness and claudication of the left arm on exercise and vertebrobasilar insufficiency (vertigo, dizziness).

REVIEW TEST

1. During a gang fight, a 16-year-old male is shot with a 22-caliber short bullet in the occiput. Computed tomography (CT) shows that the bullet is lodged in the left medullary pyramid. The most prominent neurologic deficit is

(A) Apallesthesia, right side
(B) Exaggerated muscle stretch reflexes, left side
(C) Plantar reflex extensor, right side
(D) Fasciculations, right side
(E) Hyperreflexia, left side

2. A 70-year-old retired army colonel has right-sided hemiparesis. Which of the following signs best localizes the lesion to the brainstem?

(A) Loss of kinesthetic and pallesthetic sensation, right side
(B) Lower facial weakness (numbness), right side
(C) Exaggerated muscle stretch reflexes, right side
(D) Tonic deviation of eyes to the right
(E) Lateral strabismus

3. A 10-year-old boy has right arm and leg dystaxia, nystagmus, hoarseness, along with miosis and ptosis on the right. Bronchoscopy reveals a paretic vocal cord on the right. The lesion site responsible is most likely the

(A) right dorsal motor nucleus of CN X
(B) left red nucleus
(C) dorsolateral medulla
(D) dorsolateral pons
(E) internal capsule

4. Neurologic examination reveals miosis, ptosis, hemianhidrosis, left side; laryngeal and palatal paralysis, left side; facial anesthesia, left side; and loss of pain and temperature sensation from the trunk and extremities, right side. The lesion is in the

(A) caudal medulla, ventral median zone, right side
(B) rostral medulla, lateral zone, left side
(C) rostral pontine base, left side
(D) caudal pontine tegmentum, lateral zone, right side
(E) rostral pontine tegmentum, dorsal median zone, left side

5. Neurologic examination reveals severe ptosis, eye looks down and out, right side; fixed, dilated pupil, right side; spastic hemiparesis, left side; and lower facial weakness, left side. The lesion is in the

(A) caudal pontine tegmentum, dorsal median zone, left side
(B) rostral pontine tegmentum, dorsal lateral zone, right side
(C) pontine isthmus, dorsal lateral tegmentum, left side
(D) rostral midbrain, medial basis pedunculi, right side
(E) rostral midbrain, medial tegmentum, left side

6. Neurologic examination reveals sixth nerve palsy, right side; facial weakness, left side; hemiparesis, left side; and limb and gait dystaxia, right side. The lesion is in the

(A) caudal pontine tegmentum, lateral zone, right side
(B) caudal pontine tegmentum, dorsal median zone, left side
(C) caudal medulla, ventral median zone, right side
(D) rostral pontine tegmentum, lateral zone, left side
(E) caudal pontine base, median zone, right side

7. Neurologic examination reveals paralysis of upward and downward gaze, absence of convergence, and absence of pupillary reaction to light. The lesion is in the

(A) rostral midbrain tectum
(B) caudal midbrain tectum
(C) rostral pontine tegmentum
(D) caudal pontine tegmentum
(E) caudal midbrain tegmentum

8. Neurologic examination reveals bilateral medial rectus paresis on attempted lateral gaze, monocular horizontal nystagmus in the abducting eye, and unimpaired convergence. The lesion is in the

(A) midpontine tegmentum, dorsomedial zones, bilateral
(B) rostral midbrain tectum
(C) caudal midbrain tectum
(D) caudal pontine base
(E) rostral midbrain, bases pedunculorum

9. Neurologic examination reveals ptosis, miosis, and hemianhidrosis, left side; loss of vibration sensation in the right leg; loss of pain and temperature sensation from the trunk, extremities,

and face, right side; and severe dystaxia and intention tremor, left arm. The lesion is in the

(A) rostral midbrain tegmentum, right side
(B) rostral pontine tegmentum, dorsal medial zone, left side
(C) pontine isthmus, dorsal lateral zone, left side
(D) rostral medulla, lateral zone, left side
(E) caudal medulla, lateral zone, right side

10. Neurologic examination reveals weakness of the pterygoid and masseter muscles, left side; corneal reflex absent, left side; and facial hemianesthesia, left side. The lesion is in the

(A) midpontine tegmentum, lateral zone, left side
(B) midpontine base, medial zone, left side
(C) caudal pontine tegmentum, lateral zone, left side
(D) caudal pontine tegmentum, dorsal medial zone, left side
(E) foramen ovale, left side

11. Neurologic examination reveals loss of the stapedial reflex, loss of the corneal reflex, inability to purse the lips, and loss of taste sensation on the apex of the tongue. The lesion is in the

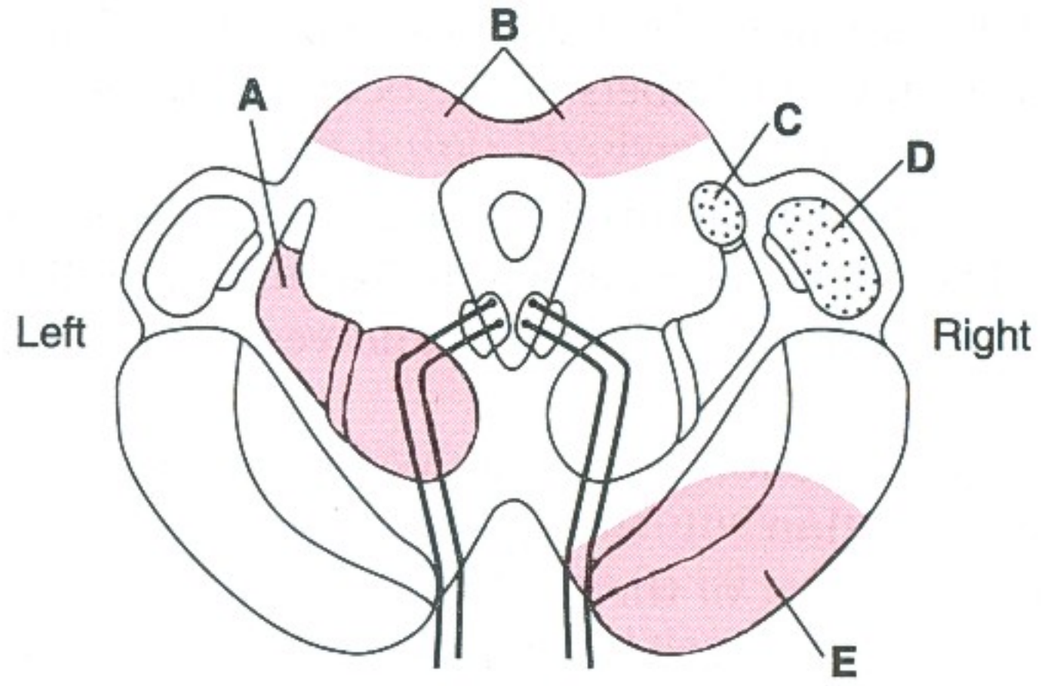
(A) stylomastoid foramen
(B) basis pedunculi of the midbrain
(C) rostral lateral pontine tegmentum
(D) caudal lateral pontine tegmentum
(E) rostral medulla

12. Paramedian infarction of the base of the pons involves which of the following structures?

(A) Trapezoid body
(B) Descending trigeminal tract
(C) Rubrospinal tract
(D) Pyramidal tract
(E) Ventral spinocerebellar tract

Questions 13 to 20

Match the description in items 13 to 20 with the appropriate lettered structure shown in the figure.



13. Paralysis of upward gaze

14. Loss of pain and temperature on the left side of the body

15. Deviation of the tongue to the left side and the uvula to the right side

16. Intention tremor on the right side

17. Complete third nerve palsy on the right side

18. Loss of vibration sensation in the right extremities

19. Babinski sign on the left side

20. Lesion leads to terminal axonal degeneration in the right transverse gyrus of Heschl

ANSWERS AND EXPLANATIONS

1–C. The bullet transected the left medullary pyramid, which contains the uncrossed corticospinal tract. This UMN lesion has produced a right contralateral spastic paresis with all pyramidal signs.

2–E. Lateral strabismus (exotropia) is seen in midbrain lesions (e.g., Weber syndrome) that transect intra-axial fibers of the oculomotor nerve. The intact lateral rectus pulls the globe laterally.

3–C. The lateral medullary syndrome is also called PICA syndrome. The dorsolateral medulla contains the nucleus ambiguus (larynx), hypothalamospinal tract (Horner syndrome), inferior cerebellar peduncle (dystaxia), and vestibular nuclei (nystagmus).

4–B. The lesion is a classic Wallenberg syndrome (PICA syndrome) of the lateral medullary zone. Interruption of the descending sympathetic tract produces ipsilateral Horner syndrome. Involvement of the nucleus ambiguus or its exiting intra-axial fibers accounts for LMN paralysis of the larynx and soft palate. The ipsilateral facial anesthesia is due to interruption of the spinal trigeminal tract; the contralateral loss of pain and temperature sensation from the trunk and extremities is due to transection of the spinothalamic tracts. The combination of ipsilateral and contralateral sensory loss is called alternating hemianesthesia. Singultus (hiccup) is frequently seen in this syndrome and is thought to result from irritation of the reticulophrenic pathway.

5–D. This constellation of deficits constitutes Weber syndrome, which affects the basis pedunculi and the exiting intra-axial oculomotor fibers. Severe ptosis (compare mild ptosis of Horner syndrome), the abducted and depressed eyeball, and the internal ophthalmoplegia (fixed, dilated pupil) are third nerve signs. The contralateral hemiparesis results from interruption of the corticospinal tracts; lower facial weakness is due to interruption of the corticobulbar tracts. The combination of ipsilateral and contralateral motor deficits is called alternating hemiplegia.

The corticospinal tract is closely related to three cranial nerves (CN III, CN VI, and CN XII); third nerve signs put the lesion in the midbrain, sixth nerve signs put the lesion in the pons, and twelfth nerve signs put the lesion in the medulla. With the exception of the trochlear nerve, all cranial nerves have ipsilateral signs. Transection of the corticospinal tract rostral to the decussation results in a contralateral spastic hemiparesis. The trochlear nucleus, an exception, gives rise to intra-axial axons that cross the midline and exit just caudal to the frenulum of the superior medullary velum. A lesion of the trochlear nucleus results in a contralateral superior oblique palsy.

6–E. These signs point to the base of the pons (medial inferior pontine syndrome) on the right side and include involvement of the exiting intra-axial abducent fibers that pass through the uncrossed corticospinal fibers; this results in an ipsilateral lateral rectus paralysis (LMN lesion) and contralateral hemiparesis. Contralateral facial weakness results from damage to the corticobulbar fibers prior to their decussation. Involvement of the transverse pontine fibers destined for the middle cerebellar peduncle results in cerebellar signs. Again, the involved cranial nerve and pyramidal tract indicate where the lesion must be to account for the deficits. An ipsilateral sixth nerve paralysis and crossed hemiplegia is called the Millard-Gubler syndrome.

7–A. These deficits indicate the Parinaud syndrome, dorsal midbrain syndrome. This condition frequently is the result of a tumor in the pineal region (e.g., germinoma or pinealoma); a pinealoma compresses the superior colliculus and the underlying accessory oculomotor nuclei that are responsible for upward and downward vertical conjugate gaze. Patients usually have pupillary disturbances and absence of convergence.

8–A. The MLF is located in the dorsomedial midpontine tegmentum. MLF syndrome is frequently seen in multiple sclerosis and less often in vascular lesions. Another pontine lesion results in one-and-a-half syndrome; it includes the MLF syndrome and a lesion of the abducent nucleus (CN VI). See Chapter 17:.

9–C. These deficits correspond to a lesion in the dorsolateral zone of the pontine isthmus, lateral superior pontine syndrome. Interruption of the descending sympathetic pathway to the ciliospinal center of Budge (T1–T2) results in Horner syndrome (always ipsilateral). Involvement of the lateral aspect (includes the leg fibers) of the medial lemniscus results in a loss of vibration sensation and other dorsal column modalities. Damage to the trigeminothalamic and spinothalamic tracts at this level results in contralateral hemianesthesia of the face and body. Infarction of the superior cerebellar peduncle leads to severe cerebellar dystaxia on the same side.

10–A. These signs indicate the lateral midpontine syndrome. This lesion involves the motor and principal trigeminal nuclei and the intra-axial root fibers of the trigeminal nerve as it passes through the base of the pons. All signs are ipsilateral and refer to CN V. The afferent limb of the corneal reflex has been interrupted. This syndrome results from occlusion of the trigeminal artery, a short circumferential branch of the basilar artery.

11–D. These signs constitute the lateral inferior pontine syndrome (AICA syndrome). The neurologic findings are all signs of a lesion involving the facial nerve (CN VII). The facial nerve nucleus and intra-axial fibers are found in the caudal lateral pontine tegmentum. A lesion of the stylomastoid foramen would not include the absence of the stapedial reflex or the loss of taste sensation from the anterior two-thirds of the tongue. The stapedial nerve and the chorda tympani exit the facial canal proximal to the stylomastoid foramen.

12–D. The base of the pons includes the corticospinal (pyramidal), corticobulbar, and corticopontine tracts, pontine nuclei, and transverse pontine fibers. At caudal levels, intra-axial abducent fibers of CN VI pass through the lateral pyramidal fascicles.

13–B. Paralysis of upward gaze results from compression of the mesencephalic tectum by a tumor in the pineal region; this is called Parinaud syndrome.

14–C. Loss of pain and temperature on the left side of the body is due to a lesion on the right side of the lateral spinothalamic tract.

15–E. Deviation of the tongue to the left side results from transection of the right corticobulbar fibers (CN XII) in the medial aspect of the crus cerebri. Deviation of the uvula to the right side results from transection of the right corticobulbar fibers (CN X) in the medial aspect of the crus cerebri.

16–A. Transection of the left dentatothalamic tract results in an intention tremor on the right side. The dentatothalamic tract decussates in the caudal midbrain, below the level of this lesion.

17–E. Complete third nerve palsy on the right side results from transection of the oculomotor nerve fibers as they pass through the right side of the crus cerebri.

18–A. A loss of vibration sensation in the right extremities results from destruction of the left medial lemniscus.

19–E. A Babinski sign on the left side results from transection of the corticospinal tract within the middle three-fifths of the crus cerebri.

20–D. Destruction of the right medial geniculate body results in terminal axonal degeneration of the auditory radiation in the right transverse gyrus of Heschl.