Peripheral Autonomic Nervous System

Fundamentals

The autonomic nervous system, working in concert with the endocrine system (see above) and various nuclei in the brainstem, regulates vital functions that are necessary for the maintenance of the internal environment (homeostasis), including respiration, circulation, metabolism, body temperature, water balance, digestion, secretion, and reproductive function. The designation "autonomic" is derived from the fact that these functions are controlled by unconscious (involuntary) mechanisms, as discussed above.

As already mentioned, the hypothalamus is the main regulatory center for the entire peripheral autonomic system. It exercises its control over many bodily functions partly through nerve impulses and partly through hormonal pathways, by means of the hypothalamic-pituitary system (see above and standard works on endocrinology, physiology, and anatomy).

The efferent arm of the autonomic nervous system is composed of two complementary systems, the **sympathetic** nervous system and the **parasympathetic** nervous system, whose effects are generally antagonistic to each other. The efferent fibers of both systems mainly innervate the smooth muscle of the viscera, blood vessels, and glands and are thus commonly called *visceral efferent (visceromotor) fibers*, to distinguish them from the sensory *visceral afferent fibers*. The latter, unlike the visceral efferent fibers, are not divided into two systems.

General scheme of the sympathetic and parasympathetic nervous systems. The final efferent pathway of both the sympathetic and the parasympathetic nervous systems consists of two neurons in series (Fig. 6.**14**). The cell body of the **first (preganglionic) neuron** lies within the central nervous system, while that of the **second (postganglionic) neuron** is found in a peripheral ganglion.

The first neurons of the sympathetic nervous system lie in the thoracic and lumbar segments of the spinal cord (intermediolateral cell column, T1-L2); for this reason, the sympathetic nervous system is sometimes called the **thoracolumbar system**. Some of the first neurons of the parasympathetic nervous system are found in the nuclei of cranial nerves III, VII, IX, and X (see below), while the remainder are found in the lateral horns of the sacral segments of the spinal cord (pelvic parasympathetic system, S2-S4). Thus, the parasympathetic nervous system is sometimes called the **craniosacral system.**

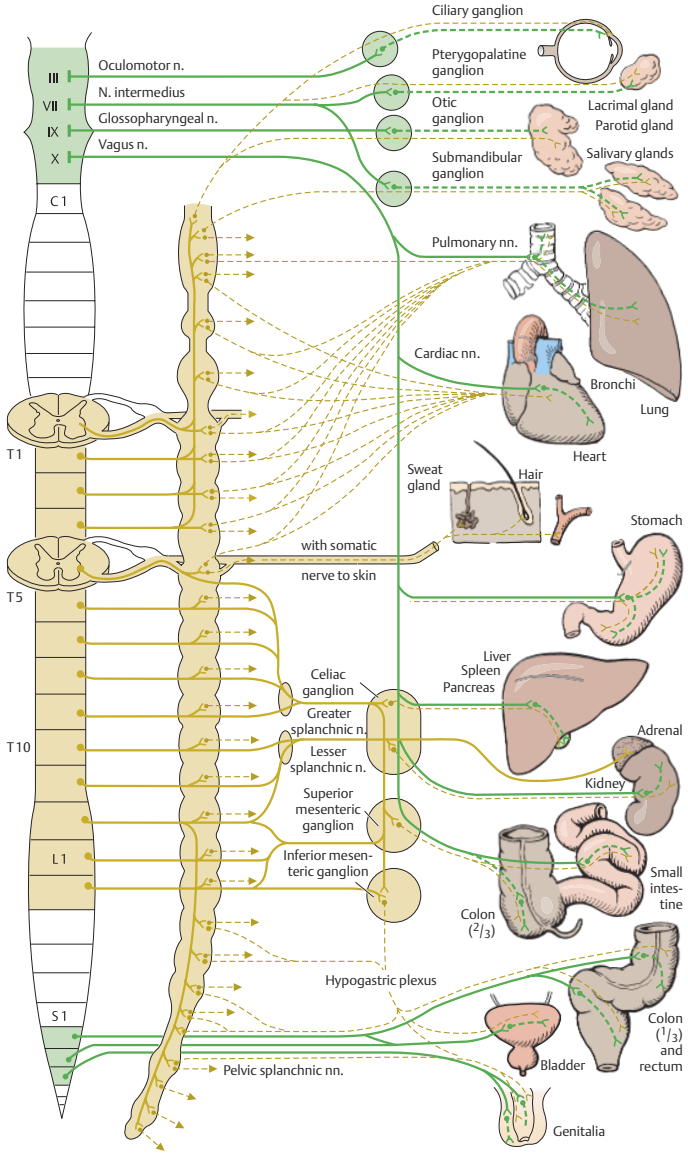


Fig. 6.**14 The sympathetic and parasympathetic nervous system** (schematic diagram). Yellow: sympathetic. Green: parasympathetic.

The second neurons of the sympathetic nervous system are arranged in prevertebral and paravertebral chains of ganglia (the sympathetic chains), while those of the parasympathetic nervous system generally lie in the walls of the innervated organs (intramural ganglia). The first neurons of both systems use acetylcholine as their neurotransmitter. The second neurons of the parasympathetic nervous system also use acetylcholine as their neurotransmitter (a further alternative name for the parasympathetic nervous system is, therefore, the **cholinergic system**). The neurotransmitter of the postganglionic sympathetic neurons, however, is norepinephrine (**adrenergic system**). The sweat glands are an exception to this rule: the second sympathetic neuron innervating them is cholinergic, like a second neuron in the parasympathetic nervous system.

Hypothalamic control of the sympathetic and parasympathetic nervous systems. Stimulation of the **rostral** hypothalamus induces **increased parasympathetic (trophotropic) activity**, including reduction of the cardiac minute volume, hypotonia, slowing of the heartbeat, reduction of the respiratory volume, lowering of the basal metabolic rate, vasodilatation, sweating, salivation, contraction of the bladder, reduced secretion of epinephrine, increased peristalsis, and pupillary constriction. Stimulation of the **caudal** hypothalamus, on the other hand, induces **increased sympathetic (ergotropic) activity**, including a rise in blood pressure, acceleration of the heartbeat, increased blood supply to the skeletal muscle and lungs, vasoconstriction in blood depots such as the capillary bed of the digestive tract, decreased blood supply to the abdominal viscera, increased respiratory volume, a rise in the blood glucose level, inhibition of peristalsis, urinary retention, increased secretion of epinephrine, widening of the palpebral fissure, and pupillary dilatation. A mass reaction thus occurs in the entire body, directed toward physical exertion and therefore enabling the whole organism to deal optimally with situations of attack and stress. While the sympathetic, ergotropic reaction is directed toward physical exertion, the parasympathetic, trophotropic reaction is directed toward rest and recovery. Despite these general principles, however, the distinction between parasympathetic and sympathetic activity is not always clear-cut.

Neural connections of the hypothalamus to the peripheral autonomic nervous system. The hypothalamus exerts its regulating and controlling functions over the sympathetic and parasympathetic nervous systems by means of descending pathways including the *medial forebrain bundle* (Fig. 6.**9**), the *mamillotegmental tract*, and the *dorsal longitudinal fasciculus* (of Schütz) (Fig. 6.**10**).

These three fiber pathways connect the hypothalamus to the *descending midbrain reticular system*, which, in turn, carries the central impulses to the various components of the parasympathetic and sympathetic nervous systems.

Sympathetic Nervous System

The sympathetic nervous system innervates the smooth musculature of the blood vessels, abdominal viscera, bladder, rectum, hair follicles, and pupils, as well as the cardiac muscle, the sweat glands, and the lacrimal, salivatory, and digestive glands. The smooth musculature of the abdominal viscera, bladder, rectum, and digestive glands is inhibited, while that of all other target organs is stimulated to contract.

The caliber of the body's arteries is mainly regulated by the sympathetic nervous system. Increased sympathetic activity leads to vasoconstriction, and decreased sympathetic activity to vasodilatation.

Anatomy. The origin of the preganglionic fibers from thoracic segments T1 through T12 and from the first two lumbar segments is shown in Fig. 6.**14**. Some of the preganglionic fibers terminate on second neurons in the right and left sympathetic chains (only the left sympathetic chain is depicted in the figure). The remainder pass through the sympathetic chain without a synapse and terminate on a second neuron in a prevertebral ganglion. In either case, the postganglionic fiber of the second neuron transmits the sympathetic impulses onward to the target organ.

Sympathetic chain. As shown in Fig. 6.**15**, the preganglionic fibers emerge from neurons in the lateral horn of the spinal cord (intermediolateral cell column) and then join the axons of the somatic motor neurons to exit from the spinal cord in the anterior root. At the level of the spinal ganglion, the autonomic fibers separate from the somatic fibers once again and enter the sympathetic chain by way of the *white ramus communicans*, which is white because its fibers are myelinated. Some preganglionic fibers already terminate on the second neuron in the pathway at the same segmental level, but others travel one or more levels up or down the sympathetic chain before making a synapse onto their second neuron. Yet other fibers traverse the sympathetic chain without making a synapse and then terminate on a second neuron in a prevertebral ganglion. In all cases, the unmyelinated postganglionic fibers leave the sympathetic chain in the *gray ramus communicans*, which rejoins the spinal nerve at the same segmental level, so that its fibers travel to the corresponding cutaneous dermatome. In the skin, the autonomic fibers innervate the cutaneous vessels, the piloerector muscles, and the sweat glands.

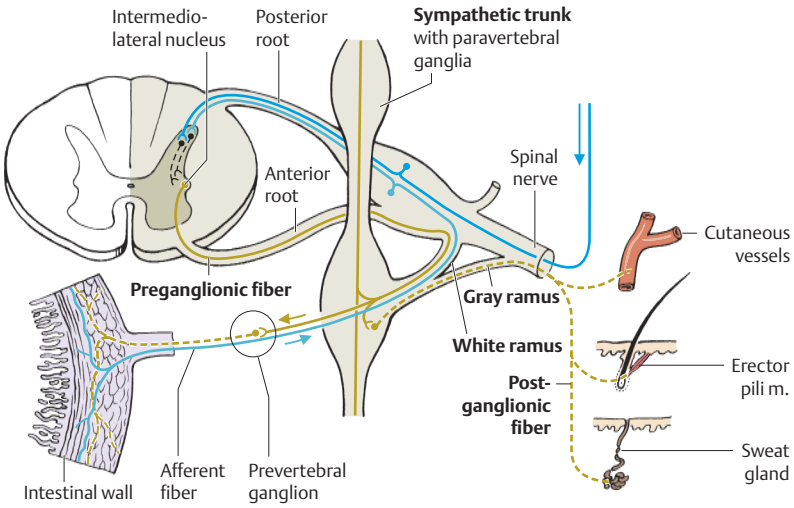


Fig. 6.**15 The sympathetic trunk and the preganglionic and postganglionic sympathetic fibers** (schematic diagram)

Sympathetic innervation of the head and neck. As mentioned above, some postganglionic fibers reach their targets in the periphery by way of the segmental spinal nerves, but others do so by traveling along the blood vessels and their branches, particularly in the head and neck. The cervical spinal cord contains no sympathetic nuclei; thus, the sympathetic innervation of the head and neck is derived from the intermediolateral cell column of the upper four or five thoracic segments. Postganglionic fibers from these segments ascend in the sympathetic chain, and terminate in three ganglia at its rostral end: the *superior cervical ganglion*, the *middle cervical ganglion*, and the *cervicothoracic (stellate) ganglion*. These ganglia are the sites of the synaptic relay onto the second neurons, which emit the postganglionic fibers. Some of these fibers travel with the spinal nerves to the cervical cutaneous dermatomes. Other, unmyelinated fibers from the superior cervical ganglion form the *external carotid plexus*, which accompanies the external carotid artery and its branches to the head and the face, innervating the sweat glands, the smooth muscles of the hair follicles, and the blood vessels. Yet other fibers accompany the internal carotid artery as the *internal carotid plexus*, which innervates the eye (dilator pupillae muscle, orbitalis muscle, and tarsal muscle) as well as the lacrimal and salivary glands (Figs. 4.**27** and 4.**28** [p. 158 f.] and 6.**14**).

Sympathetic innervation of heart and lungs. Postganglionic fibers from the cervical and upper four or five thoracic ganglia run in the *cardiac nerves* to the *cardiac plexus*, which innervates the heart. *Pulmonary nerves* innervate the bronchi and lungs (Fig. 6.**14**).

Sympathetic innervation of the abdominal and pelvic organs. Preganglionic fibers arise in thoracic segments T5 through T12 and travel, by way of the *greater* and *lesser splanchnic nerves*, to the unpaired prevertebral ganglia (the *celiac*, *superior mesenteric*, and *inferior mesenteric ganglia*), which are located along the aorta at the levels of origin of the correspondingly named aortic branches. Within these ganglia, the splanchnic fibers make synapses onto the second sympathetic neurons, which, in turn, emit the postganglionic fibers for the abdominal and pelvic viscera. In contrast to the parasympathetic fibers, the sympathetic postganglionic fibers are very long and form various plexuses before reaching their target organs (Fig. 6.**14**).

Adrenal medulla. The adrenal medulla occupies a special position in the sympathetic nervous system. It is analogous to a sympathetic ganglion, in that it is directly innervated by preganglionic fibers. These fibers form synapses onto modified second neurons within the adrenal medulla, which, rather than possessing an axon, secrete epinephrine and norepinephrine into the bloodstream (Fig. 6.**14**). Sympathetic activation induces the adrenal medulla to secrete epinephrine and norepinephrine, which then exert sympathetic effects in the periphery. This is particularly important under conditions of stress.

Clinical Symptoms of Sympathetic Lesions

Horner syndrome. As mentioned in Chapter 4 (p. 157 ff.), lesions affecting the ciliospinal center, the cervical sympathetic chain (cervicothoracic ganglion), or the autonomic plexuses along the blood vessels of the head and neck cause ipsilateral Horner syndrome. This consists of the clinical triad of a constricted pupil/**miosis** (due to loss of contraction of the dilator pupillae muscle), a hanging eyelid/**ptosis** (due to loss of contraction of the tarsal muscle), and an inwardly sunken globe/**enophthalmos** (due to loss of contraction of the orbitalis muscle). There is also loss of sweating (**anhidrosis**) and **vasodilatation** (due to loss of the vasoconstrictive effect of the sympathetic nerves) on the ipsilateral half of the face, which therefore appears dry and reddened.

Causes of Horner syndrome. Interruption of the sympathetic pathway to the head and neck at any point can cause Horner syndrome. One common cause is a bronchial carcinoma at the apex of the lung (**Pancoast tumor**) impinging on

the cervical sympathetic chain. Such tumors may present with Horner syndrome before becoming otherwise symptomatic.

Dissection of the internal carotid artery is another important cause of Horner syndrome. When the intima of the artery is torn, blood enters the vessel wall and the lumen is narrowed or occluded; rupture of the artery with pseudoaneurysm formation is rare. Carotid dissection has many possible etiologies; dissection may be *traumatic* or due to an *intrinsic abnormality* of the tissue of the vessel wall, e. g., fibromuscular dysplasia, which predisposes to the development of an intimal tear. In most cases, however, the etiology of carotid dissection cannot be determined.

The pathogenesis of sympathetic dysfunction in carotid dissection is not yet fully understood. According to one current hypothesis, *compression* of the sympathetic nerve branches by an intramural hematoma leads to nerve injury and dysfunction. According to another hypothesis, *ischemia* of the sympathetic nerve branches is the major cause of their dysfunction, as these nerve branches are supplied by small perforating branches of the internal carotid artery, which can be displaced or occluded by the dissection. Neither hypothesis is fully satisfactory.

Horner syndrome also arises as a result of brainstem lesions affecting the central sympathetic pathway, as in Wallenberg syndrome (p. 226 ff.)

Vasomotor phenomena in sympathetic dysfunction. The vasodilatation that follows a sympathetic lesion can be exploited therapeutically: *sympathectomy* is sometimes performed to increase regional blood flow, e. g., in Raynaud disease.

The vasodilatation due to a sympathetic lesion is also evident after interruption of the splanchnic nerves, which leads to a large increase of intravascular volume in the blood vessels of the bowel, i.e., to pooling of blood in the splanchnic area, with the risk of internal hemorrhage.

Parasympathetic Nervous System

In contrast to the sympathetic nervous system, the parasympathetic nervous system does not evoke any systemic responses, but instead produces its effects in individual, circumscribed areas, as reflected in the fact that its second (postganglionic) neurons lie near their target organs. Furthermore, acetylcholine, which is released as a neurotransmitter at the parasympathetic nerve terminals, is rapidly broken down by cholinesterases, and its effect is thus relatively short-lived.

The preganglionic fibers of the parasympathetic nervous system are long (unlike the short preganglionic fibers of the sympathetic nervous system).

They emerge from nuclei in the brainstem and sacral spinal cord (S2, S3, S4) (Fig. 6.**14**).

Cranial Portion of the Parasympathetic Nervous System

Parasympathetic innervation of the head. The cell bodies of the preganglionic neurons lie in various *brainstem nuclei*, and their axons are found in *cranial nerves III*, *VII*, *IX*, *and X*. (The anatomy and course of these nerves was described in Chapter 4.) The preganglionic fibers travel to a number of ganglia that lie very close to their respective end organs (the *ciliary*, *pterygopalatine*, *submandibular*, and *otic ganglia*). These ganglia are relay stations in which the preganglionic fibers form synapses onto the second (postganglionic) neurons. The parasympathetic postganglionic fibers in the head are short, as they have only a short distance to travel before they reach their end organs. Like the sympathetic postganglionic fibers, they innervate smooth muscle, sweat glands, and lacrimal and salivary glands (Fig. 6.**14**). The smooth muscle of the blood vessel walls receives no parasympathetic innervation.

Parasympathetic innervation of the thoracic and abdominal organs. The parasympathetic portion of the *vagus nerve* (Fig. 4.**49**, p. 199) originates in the *dorsal nucleus of the vagus nerve* and carries preganglionic fibers for the innervation of the heart, lungs, and abdominal viscera down to the distal third of the transverse colon (Fig. 6.**14**). The second (postganglionic) neurons are found in autonomic plexuses located immediately adjacent to their end organs, or else within the bowel wall (myenteric plexus of Auerbach, submucosal plexus of Meissner).

Sacral Portion of the Parasympathetic Nervous System

Parasympathetic innervation of the pelvic organs and genitalia. The sacral portion of the parasympathetic nervous system carries impulses in the *pelvic splanchnic nerves* and the *superior and inferior hypogastric (pelvic) plexuses* to ganglia in the muscular wall of the colon (from the distal third of the transverse colon onward), rectum, bladder, and genitalia (Fig. 6.**14**). In the pelvic area, the parasympathetic nervous system is responsible for the emptying of the rectum and bladder. It also brings about penile erection, while sympathetic fibers are responsible for ejaculation, which occurs through contractions of the ductus deferens and the seminal vesicles.

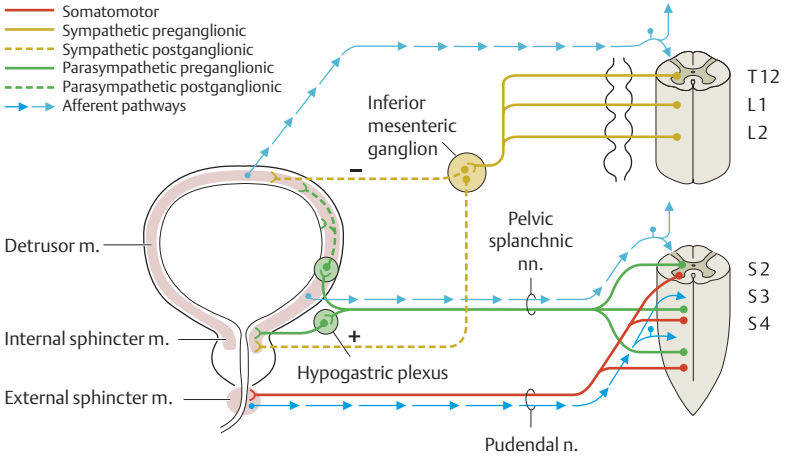


Fig. 6.**16 Innervation of the bladder**

Autonomic Innervation and Functional Disturbances of Individual Organs

The sympathetic and parasympathetic innervation of individual organs is summarized in Table 6.**2**. The innervation of the pelvic organs will be discussed in greater detail in the following sections, because the function of these organs is commonly impaired in disturbances of the autonomic nervous system. Bladder dysfunction is the most important problem of this type.

Innervation of the Bladder

Parasympathetic innervation. The motor innervation of the urinary bladder is mostly parasympathetic. The pelvic splanchnic nerves, derived from segments S2, S3, and S4, travel to parasympathetic ganglia in the bladder wall and to the smooth muscle of the internal urethral sphincter (Figs. 6.**14** and 6.**16**). Parasympathetic stimulation induces contraction of the smooth detrusor muscle of the bladder wall and simultaneous relaxation of the internal urethral sphincter. Micturition results.

Sympathetic innervation. The sympathetic fibers innervating the bladder are derived from neurons in the intermediolateral cell column of the lower thoracic and upper lumbar spinal cord (segments T12, L1, and L2). These fibers

Tabelle 6.2 The Sympathetic and Parasympathetic Nervous System

	Sympathetic			*Parasympathetic*		
Organ	***Preganglionic neuron***	***Postganglionic neuron***	***Activity***	***Preganglionic neuron***	***Postganglionic neuron***	***Activity***
Eye	T1–T2	Superior cervical ganglion	Mydriasis	Edinger–Westphal nucleus (accessory oculomotor nucleus)	Ciliary ganglion	Miosis, contraction of the ciliary muscle (accommodation)
Lacrimal, sublingual, and submandibular glands	T1–T2	Superior cervical ganglion	Vasoconstriction Secretion (viscous)	Superior salivatory nucleus	Pterygopalatine ganglion	Lacrimation, salivation (watery), vasodilation
Parotid gland	T1–T2	Superior cervical ganglion	Vasoconstriction Secretion	Inferior salivatory nucleus	Otic ganglion	Salivation
Heart	T1–T4 (T5)	Superior, middle, and inferior cervical ganglia and upper thoracic ganglia	Acceleration Dilation of coronary arteries	Dorsal nucleus of the vagus nerve	Cardiac plexus	Bradycardia, constriction of coronary arteries
Small intestine and ascending colon	T6–T10	Celiac ganglion, superior mesenteric ganglion	Inhibition of peristalsis and secretion	Dorsal nucleus of the vagus nerve	Myenteric plexus (of Auerbach), submucosal plexus (of Meissner)	Peristalsis, secretion, vasodilation
Pancreas	T6–T10	Celiac ganglion	—	Dorsal nucleus of the vagus nerve	Periarterial plexus	Secretion

	Sympathetic			*Parasympathetic*		
Descending colon and rectum	L1–L2	Inferior mesenteric ganglion, hypogastric ganglion	Inhibition of peristalsis and secretion	S2–S4	Myenteric plexus (of Auerbach), submucosal plexus (of Meissner)	Secretion, peristalsis, evacuation
Kidney Bladder	L1–L2	Celiac ganglion, renal and hypogastric plexuses	Activation of internal sphincter muscle, vasoconstriction	S2–S4	Hypogastric plexus (vesical plexus)	Relaxation of the internal sphincter muscle, contraction of the detrusor muscle, vasodilation
Adrenal gland	T11–L1	Adrenal cells	Secretion (norepinephrine, epinephrine)	—	—	—
Male genitalia	L1–L2 (pelvic splanchnic nerves)	Superior and inferior hypogastric plexuses (pelvic plexus)	Ejaculation Vasoconstriction	S2–S4	Hypogastric plexus (pelvic plexus)	Erection, vasodilation, secretion
Skin of head and neck	T2–T4	Superior and middle cervical ganglia	Vasoconstriction Sweating Piloerection	—	—	—
Arms	T3–T6	Inferior cervical ganglion and upper thoracic ganglia		—	—	—
Legs	T10–L2	Lower lumbar and upper sacral ganglia		—	—	—

travel through the caudal portion of the sympathetic chain and the inferior splanchnic nerves to the inferior mesenteric ganglion. Postganglionic sympathetic fibers then travel, by way of the inferior hypogastric plexus, to the bladder wall (tunica muscularis) and to the smooth muscle of the internal urethral sphincter (Fig. 6.**14** and 6.**16**).

Sensory innervation. Afferent fibers originate in nociceptors and proprioceptors of the bladder wall, which respond to stretch. As the bladder fills, there is a reflexive increase in muscle tone in the bladder wall and internal sphincter, which is mediated by the sacral segments (S2-S4) and the pelvic splanchnic nerves Increasing tension on the bladder wall is consciously perceived, as some of the afferent impulses travel centrally, by way of the posterior columns, to the so-called pontine micturition center, which lies in the reticular formation near the locus ceruleus. From the micturition center, impulses travel onward to the paracentral lobule on the medial surface of the cerebral hemispheres, and to other brain areas.

Regulation of Bladder Function: Continence and Micturition

The bladder performs its two major functions, the **continent storage of urine** and **periodic**, **complete emptying**, as follows.

Urinary continence is achieved by *activation of the internal and external urethral sphincters*, and, in women, mainly by activation of the *muscles of the pelvic floor*. Sympathetic efferent fibers from T11-L2 activate alpha-receptors of the internal sphincter and are also thought to inhibit the detrusor muscle by a mechanism that has not yet been determined. The external urethral sphincter is a striated muscle that, like the muscles of the pelvic floor, receives its somatic innervation through efferent fibers of the pudendal nerve (S2-S4, see above).

As the bladder is filled and the tension on the bladder wall increases, involuntary reflex contraction of the detrusor muscle is effectively countered by activation of the external sphincter by the sacral somatic motor neurons. At the same time, lumbar sympathetic activation induces closure of the internal sphincter as well as relaxation of the detrusor muscle.

Micturition. The most important stimulus for micturition is *stretching of the bladder wall*, which excites visceral sensory afferent neurons, induces the urge to void, and, with the cooperation of higher nervous centers, leads to *contraction of the detrusor muscle*. This hollow muscle receives its parasympathetic innervation from the sacral spinal cord by way of the pelvic nerve. Bladder emptying is further promoted by somatic, voluntarily controlled *abdominal*

pressing and by *simultaneous relaxation of the internal and external urethral sphincters*.

At a supraspinal level, micturition is controlled by the *pontine micturition center*, which projects descending efferent fibers in the medial and lateral reticulospinal tracts to coordinate the simultaneous relaxation of the internal and external sphincters and contraction of the detrusor muscle. The neurotransmitter glutamate may play a role in this pathway. The pontine micturition center is anatomically poorly characterized. It can be inhibited through afferent fibers from higher centers, including the frontal cortex, cingulate gyrus, paracentral lobule, and basal ganglia.

Bladder Dysfunction

As discussed in the last section, the regulation of continence and micturition requires the perfect functional cooperation of numerous anatomical structures, some of which are very distant from others. Lesions at many different sites in the central or peripheral nervous system can have far-ranging deleterious effects on bladder function.

Bladder dysfunction may be due to structural/anatomical lesions of the bladder or urethra (**bladder dysfunction of urological origin**: vesical tumors, infravesical obstruction by urethral stricture or prostatic hypertrophy), or it may be due to a lesion of the neural structures innervating the bladder (**neurogenic bladder dysfunction**). The responsible neural lesion may lie in the peripheral nerve pathways, the autonomic plexuses, the spinal cord, or higher centers.

Impairment of supraspinal control mechanisms frequently causes bladder dysfunction in patients with multiple sclerosis, for example. Disturbances of the interaction between the pontine micturition center and other, higher centers that modulate it play an important role in the types of neurogenic bladder dysfunction seen in neurodegenerative diseases, including Parkinson disease.

Neurogenic Bladder Dysfunction

Typical manifestations of neurogenic bladder dysfunction include *urinary frequency and urgency*, *incontinence*, *difficult and incomplete bladder emptying*, and *recurrent urinary tract infections*.

The first step toward the successful treatment of neurogenic bladder dysfunction is a correct clinical diagnosis. Various aspects of urinary function must be taken into account, including the answers to the following questions: When and how frequently is the bladder emptied? Is it emptied completely? Is the urge to void normal, diminished, or abnormally severe (urinary urgency)? Has a urinary tract infection been ruled out? Is the patient continent?

Detrusor instability and detrusor hyperreflexia are characterized by premature detrusor contractions during the vesical filling phase. The term "instability" refers to a lack of the normal inhibition of detrusor contraction; the term "hyperreflexia" implies that a neurological disease is causing the bladder emptying disorder. Thus, clinical entities such as uninhibited neurogenic bladder, automatic bladder, and motor instability of the bladder all belong within the etiological category of detrusor hyperreflexia. In such cases, *the lesion lies above the sacral spinal cord* and impairs the function of suprasacral inhibitory projections to the detrusor muscle. The major symptom of isolated detrusor hyperreflexia is **imperative urinary urgency with urge incontinence and low residual volume**. The more common causes are multiple sclerosis, cerebrovascular diseases, normal pressure hydrocephalus, Parkinson disease, spinal cord trauma, and trauma or tumor affecting the frontal lobes of the brain.

Detrusorsphincter dyssynergia is defined as involuntary detrusor contraction without relaxation of the external urethral sphincter. The lesion lies *between the sacral spinal cord and the pontine micturition center*. The major symptom is **imperative urinary urgency with incomplete emptying of the bladder**. Detrusor-sphincter dyssynergia causes complications (in particular, ascending urinary tract infections) more frequently in men than in women, because women have a lower bladder outlet resistance than men. The more common causes are multiple sclerosis, cervical myelopathy, spinal tumors, vascular malformations, and trauma. This entity should be distinguished from the rare *functional obstruction of the bladder neck*, a disorder of unknown etiology that is associated with increased residual volume and can impair renal function.

Detrusor areflexia results from deficient afferent or efferent innervation of the detrusor muscle. Afferent and efferent disturbances hardly ever occur in isolation, presumably because both afferent and efferent impulses travel through the pelvic parasympathetic nerves and the sacral spinal segments, so that any lesion impairing one type of impulses necessarily impairs the other. The clinical manifestations of detrusor areflexia are **reduced urge to void**, **inability to initiate micturition**, **and overflow incontinence** with an increased bladder volume (up to 2000 ml). *The lesion lies within the sacral spinal cord or the peripheral nerves that enter and emerge from it*. Causes include tumors involving the conus medullaris and/or cauda equina, lumbar spinal stenosis and disk herniation, polyradiculitis (including Guillain-Barré syndrome), diabetic or alcoholic polyneuropathy, tabes dorsalis, pelvic surgery and radiation therapy, myelodysplasia, and tethered cord syndrome.

Detrusor areflexia due to sacral spinal cord dysfunction is found in 20-30% of patients with multiple sclerosis. Most of these patients have markedly ele-

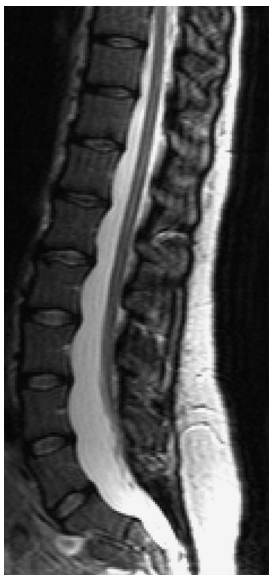
vated residual volumes because the attempt to urinate is further thwarted by lack of relaxation of the external urethral sphincter.

Case Presentation 3: *Tethered Cord Syndrome*

This previously healthy 27-year-old nurse complained to her family physician of difficulty urinating. She had trouble initiating the flow of urine, needed to strain to urinate, and felt that her bladder was still full afterward. At other times, she passed small amounts of urine involuntarily. Finally, she had also had a single episode of stool incontinence. She was very worried and embarrassed, was afraid to leave the house, and had stopped going to work. She denied having pain or any history of trauma.

Neurological examination revealed hypesthesia in the sacral dermatomes (saddle hypesthesia), normal strength in the lower limbs, and markedly diminished sphincter tone. An MRI scan was ordered to rule out a mass compressing the conus medullaris or cauda equina (Fig. 6.**17**). This study revealed a developmental anomaly in the lumbosacral spinal canal, in which the conus medullaris lay at an abnormally low level (tethered cord syndrome).

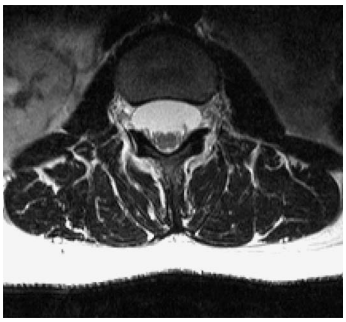
In this disorder, the conus, because it lies immediately under the dorsal dura mater and adheres to it, cannot ascend normally to the L1–2 level over the course of development. The resulting neurological deficits may not arise until later in life, and their pathogenesis remains incompletely understood. Because of her progressive neurological deficits, the patient presented here was treated neurosurgically, with an operative detachment of the conus from the dura mater. Her deficits resolved completely thereafter.



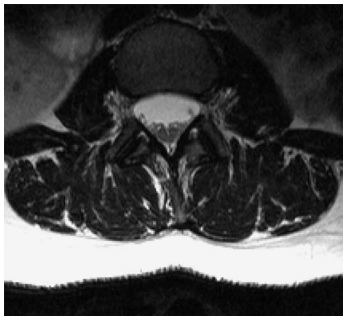
a

Fig. 6.**17 Tethered cord syndrome. a** The sagittal T2-weighted image shows an enlarged lumbar spinal canal with the conus medullaris lying at an abnormally low position (L4) immediately underlying the dorsal dura mater. In this case, there were no associated anomalies such as a dermal sinus, lipoma, or meningomyelocele. **b**, **c** The T2-weighted axial sections through the spinal canal at T12 (**b**) and L2 (**c**) reveal spinal cord at both levels. Even at the L2 level, the cord has a greater diameter than the cauda equina. It adheres to the dorsal dura mater.

Figs. 6.**17 b, c** ▷



b



c

Genuine stress incontinence is said to be present when detrusor function is normal and stress incontinence is due solely to deficient activation of the external urethral sphincter. Genuine stress incontinence, the most common type of bladder emptying disorder in women, occurs mainly after hysterectomy and in multiparous women with uterine prolapse. Its incidence rises with age. It also occurs as a manifestation of various neurogenic bladder emptying disorders, including detrusor hyperreflexia and detrusor-sphincter dyssynergia.

Nonneurogenic Bladder Dysfunction

Infravesical obstruction usually occurs in men, often as the result of benign prostatic hyperplasia, and manifests itself clinically with urinary urgency, pollakiuria, nocturia, urinary retention, and overflow incontinence.

Dysfunction of the external urethral sphincter, preventing adequate relaxation of the sphincter muscle, has been found to be a common cause of obstructive bladder emptying disturbances in young women. It is characterized by myotoniform discharges in the EMG. Electromyographic study is necessary to distinguish this disorder from two important alternative diagnoses in young women with bladder emptying disturbances, namely, multiple sclerosis and psychogenic bladder dysfunction.

Enuresis is defined as bedwetting, by day or night, in individuals over the age of 4 years, in the absence of any demonstrable causative lesion. Enuresis is thus, by definition, not a neurogenic disturbance. The important differential diagnoses include *organic* neurological and urological causes of bedwetting, including epilepsy, spina bifida occulta, and malformations of the urogenital tract. A 24-hour EEG recording is indicated in some cases.

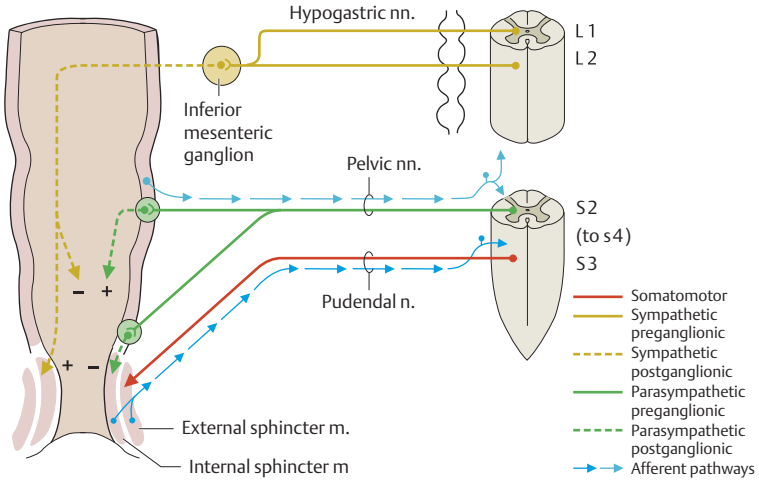


Fig. 6.**18 Innervation of the rectum**

Innervation of the Rectum

Emptying of the rectum is analogous to emptying of the bladder in many respects (Fig. 6.**18**).

Filling of the rectum activates stretch receptors in the rectal wall, which transmit impulses by way of the inferior hypogastric plexus to segments S2 through S4 of the sacral spinal cord. Afferent impulses then ascend the spinal cord to higher control centers, which are probably located in the pontine reticular formation and the cerebral cortex.

Rectal peristalsis is induced by parasympathetic activation from segments S2 through S4, which also induces relaxation of the internal sphincter. The sympathetic nervous system inhibits peristalsis. The external sphincter consists of striated muscle and is under voluntary control.

Rectal emptying is mainly accomplished voluntarily by abdominal pressing.

Rectal Emptying Disorders

Fecal retention. Transection of the spinal cord above the lumbosacral centers for defecation leads to fecal retention. Interruption of the afferent arm of the reflex pathway for defecation deprives higher centers of information about the filling state of the rectum, while interruption of descending motor fibers im-

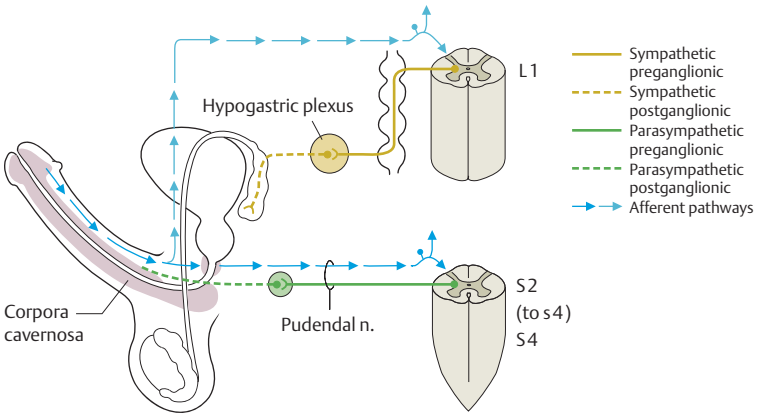


Fig. 6.**19 Innervation of the male genitalia** (erection and ejaculation)

pairs voluntary abdominal pressing. Sphincter closure is often inadequate because of spastic weakness.

Fecal incontinence. Lesions of the sacral spinal cord (S2-S4) abolish the anal reflex and produce fecal incontinence. If the stool is watery, involuntary loss of stool occurs.

Innervation of the Male Genitalia

Efferent sympathetic fibers from the upper lumbar spinal cord travel by way of a periarterial nervous plexus (the hypogastric plexus) to the seminal vesicles, prostate, and ductus deferentes. Stimulation of the plexus causes ejaculation (Fig. 6.**19**).

Parasympathetic fibers from segments S2 through S4 travel through the pelvic splanchnic nerves (the nervi erigentes) to the corpora cavernosa. Parasympathetically induced vasodilatation in the corpora cavernosa brings about penile erection (Fig. 6.**19**). The urethral sphincter and the ischiocavernosus and bulbospongiosus muscles are innervated by the pudendal nerve.

Genital function is ultimately under the control of hypothalamic centers, which exert their effects partly through neural connections (reticulospinal fibers) and partly by humoral means (hormones).

Genital Dysfunction

Spinal cord transection at a thoracic level causes impotence. Reflex priapism may occur, and occasional ejaculation is also possible. Paraplegia has been reported to be associated with testicular atrophy.

Lesions of the sacral spinal cord from S2 to S4 also cause impotence. In these cases, neither erection nor ejaculation is possible.

Visceral and Referred Pain

Afferent autonomic fibers participate in a large number of autonomic regulatory circuits. Most of the impulses traveling in these fibers do *not* rise to consciousness.

Visceral pain. The individual *can*, however, consciously perceive the filling state of the hollow viscera, which is reported to the central nervous system through afferent autonomic fibers arising from pressure or stretch receptors in the visceral wall. Overfilling of a hollow viscus is perceived as pain. Moreover, irritation of the wall of a viscus can cause reflex spasm of smooth muscle, which also gives rise to pain (biliary colic due to gallstones, renal colic due to kidney stones). Visceral inflammation or ischemia is also painful, e. g., angina pectoris.

Pain originating in the internal organs is diffuse and poorly localizable. Furthermore, the patient may report feeling pain not in the organ itself but in a related zone of the body surface (these are the zones of Head, cf. Fig. 6.**20**).

Referred pain. The cell bodies of the afferent autonomic fibers, like those of the somatic afferent fibers, are located in the spinal ganglia. The autonomic fibers enter the spinal cord through the posterior root together with the somatic afferent fibers from the myotome and dermatome of each segmental level. Thus, each individual segment of the posterior horn receives converging afferent input, both from the internal organs and from the related myotome and dermatome. Activation from either set of afferent fibers (visceral or somatic) is transmitted centrally by the same fibers of the lateral spinothalamic tract (Fig. 6.**21**). It is therefore understandable that pain arising in a particular viscus is sometimes felt elsewhere, namely, in the dermatome or myotome represented by the same spinal segment. This phenomenon is called referred pain. It may be accompanied by a certain degree of hypersensitivity to somatosensory stimulation in the dermatome to which pain is referred. The abdominal wall may also become rigid. The exact mechanism by which referred pain arises has not yet been conclusively explained, though there are a number of hypotheses.

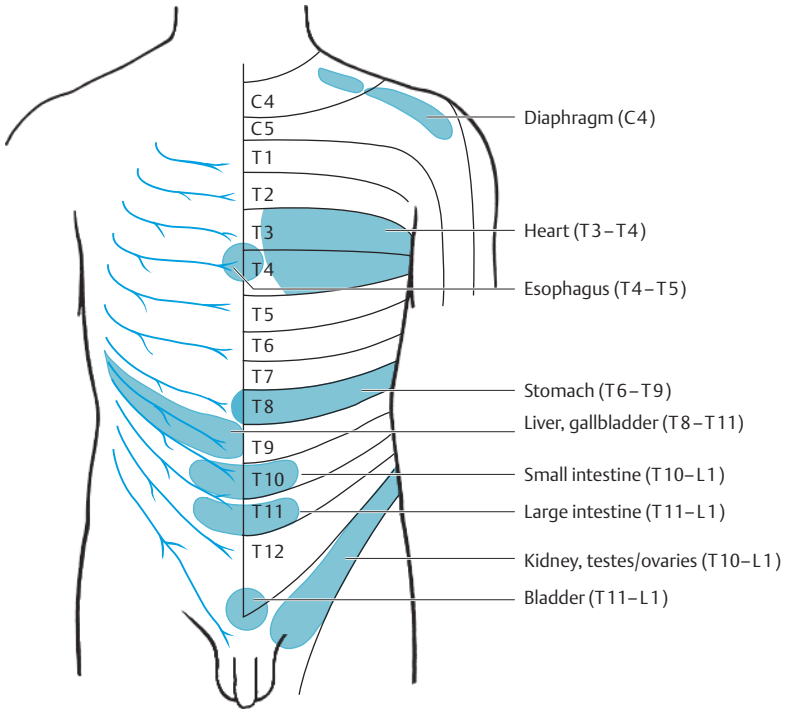


Fig. 6.**20 The zones of Head**

Pain of cardiac origin, for example, is often referred elsewhere. The upper thoracic segments on the left side receive somatic afferent fibers from the left side of the chest and the left arm, as well as visceral afferent fibers from the heart. Cardiac disease, particularly ischemia, often produces pain in one of these dermatomes (angina pectoris). The particular zones to which pain is referred from the individual internal organs are very important in physical diagnosis and are called the zones of Head (Fig. 6.**20**). It is also the case, however, that impulses arising from the skin can be projected (referred) to the internal organs. Clearly, the somatic afferent fibers are interconnected with visceral reflex arcs within the spinal cord. This may explain how therapeutic measures at the body surface (such as the application of warmth or heat, compresses, rubbing, etc.) often relieve pain arising from the autonomically innervated viscera.

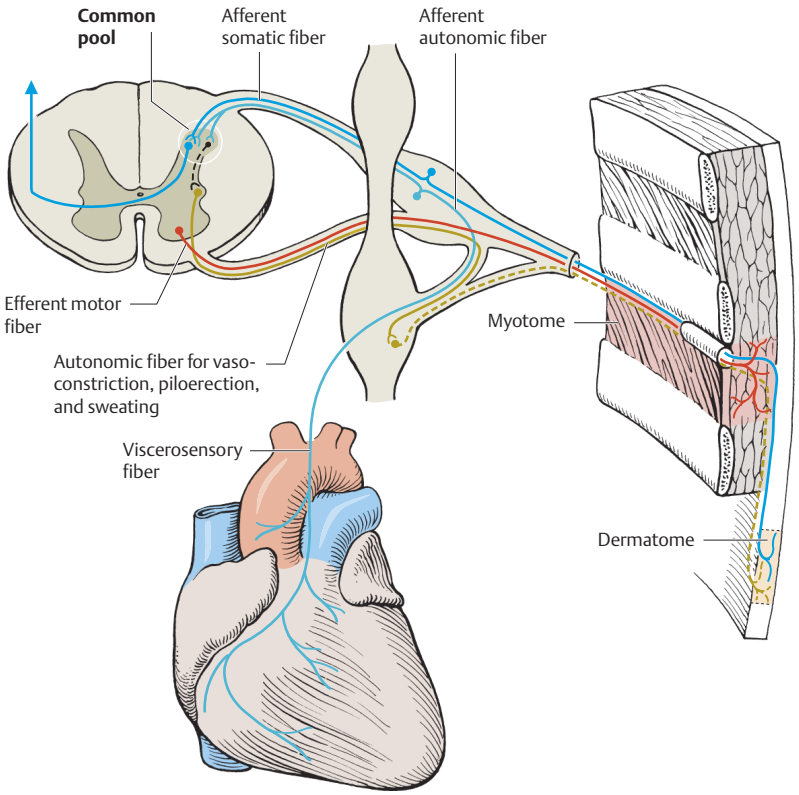


Fig. 6.**21** **The viscerocutaneous reflex arc** with myotome, dermatome, and enterotome. Viscerosensory and somatosensory impulses converge at the level of the posterior horn onto a common neuron, which transmits further impulses centrally along a single common pathway. Thus, afferent signals from the internal organs can be "misinterpreted" as having arisen in the corresponding cutaneous or muscular areas (dermatome or myotome). This is the mechanism of referred pain.