

TABLE 454-1 FUNCTIONAL CONSEQUENCES OF NORMAL ANS ACTIVATION

	Sympathetic	Parasympathetic
Heart rate	Increased	Decreased
Blood pressure	Increased	Mildly decreased
Bladder	Increased sphincter tone	Voiding (decreased tone)
Bowel motility	Decreased motility	Increased
Lung	Bronchodilation	Bronchoconstriction
Sweat glands	Sweating	—
Pupils	Dilation	Constriction
Adrenal glands	Catecholamine release	—
Sexual function	Ejaculation, orgasm	Erection
Lacrimal glands	—	Tearing
Parotid glands	—	Salivation

processing centers, or efferent limb of reflex arcs controlling autonomic responses. For example, a lesion of the medulla produced by a posterior fossa tumor can impair BP responses to postural changes and result in orthostatic hypotension (OH). OH can also be caused by lesions of the spinal cord or peripheral vasomotor nerve fibers (e.g., diabetic autonomic neuropathy). Lesions of the efferent limb cause the most consistent and severe OH. The site of reflex interruption is usually established by the clinical context in which the dysautonomia arises, combined with judicious use of ANS testing and neuroimaging studies. The presence or absence of CNS signs, association with sensory or motor polyneuropathy, medical illnesses, medication use, and family history are often important considerations. Some syndromes do not fit easily into any classification scheme.

SYMPTOMS OF AUTONOMIC DYSFUNCTION

Clinical manifestations can result from loss of function, overactivity, or dysregulation of autonomic circuits. Disorders of autonomic function should be considered in patients with unexplained OH, syncope, sleep dysfunction, altered sweating (hyperhidrosis or hypohidrosis), impotence, constipation or other gastrointestinal symptoms (bloating,

TABLE 454-2 CLASSIFICATION OF CLINICAL AUTONOMIC DISORDERS

I. Autonomic disorders with brain involvement	d. Cardiac arrhythmias
A. Associated with multisystem degeneration	e. Central sleep apnea
1. Multisystem degeneration: autonomic failure clinically prominent	f. Baroreflex failure
a. Multiple system atrophy (MSA)	g. Horner's syndrome
b. Parkinson's disease with autonomic failure	h. Vertebrobasilar and lateral medullary (Wallenberg's) syndromes
c. Diffuse Lewy body disease (some cases)	i. Brainstem encephalitis
2. Multisystem degeneration: autonomic failure clinically not usually prominent	II. Autonomic disorders with spinal cord involvement
a. Parkinson's disease	A. Traumatic quadriplegia
b. Other extrapyramidal disorders (inherited spinocerebellar atrophies, progressive supranuclear palsy, corticobasal degeneration, Machado-Joseph disease, fragile X syndrome [FXTAS])	B. Syringomyelia
B. Unassociated with multisystem degeneration (focal CNS disorders)	C. Subacute combined degeneration
1. Disorders mainly due to cerebral cortex involvement	D. Multiple sclerosis and neuromyelitis optica
a. Frontal cortex lesions causing urinary/bowel incontinence	E. Amyotrophic lateral sclerosis
b. Focal seizures (temporal lobe or anterior cingulate)	F. Tetanus
c. Cerebral infarction of the insula	G. Stiff-person syndrome
2. Disorders of the limbic and paralimbic circuits	H. Spinal cord tumors
a. Shapiro's syndrome (agenesis of corpus callosum, hyperhidrosis, hypothermia)	III. Autonomic neuropathies
b. Autonomic seizures	A. Acute/subacute autonomic neuropathies
c. Limbic encephalitis	1. Subacute autoimmune autonomic ganglionopathy (AAG)
3. Disorders of the hypothalamus	a. Subacute paraneoplastic autonomic neuropathy
a. Thiamine deficiency (Wernicke-Korsakoff syndrome)	b. Guillain-Barré syndrome
b. Diencephalic syndrome	c. Botulism
c. Neuroleptic malignant syndrome	d. Porphyria
d. Serotonin syndrome	e. Drug induced autonomic neuropathies-stimulants, drug withdrawal, vasoconstrictor, vasodilators, beta-receptor antagonists, beta-agonists
e. Fatal familial insomnia	f. Toxin-induced autonomic neuropathies
f. Antidiuretic hormone (ADH) syndromes (diabetes insipidus, inappropriate ADH secretion)	g. Subacute cholinergic neuropathy
g. Disturbances of temperature regulation (hyperthermia, hypothermia)	B. Chronic peripheral autonomic neuropathies
h. Disturbances of sexual function	1. Distal small fiber neuropathy
i. Disturbances of appetite	2. Combined sympathetic and parasympathetic failure
j. Disturbances of BP/HR and gastric function	a. Amyloid
k. Horner's syndrome	b. Diabetic autonomic neuropathy
4. Disorders of the brainstem and cerebellum	c. Autoimmune autonomic ganglionopathy (paraneoplastic and idiopathic)
a. Posterior fossa tumors	d. Sensory neuronopathy with autonomic failure
b. Syringobulbia and Arnold-Chiari malformation	e. Familial dysautonomia (Riley-Day syndrome)
c. Disorders of BP control (hypertension, hypotension)	f. Diabetic, uremic, or nutritional deficiency
	g. Dysautonomia of old age
	3. Disorders of reduced orthostatic intolerance: reflex syncope, POTS, associated with prolonged bed rest, associated with space flight, chronic fatigue

Abbreviations: BP, blood pressure; CNS, central nervous system; HR, heart rate; POTS, postural orthostatic tachycardia syndrome.