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## Autonomic Dysfunction

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## Introduction

The autonomic nervous system (ANS) includes all regions implicated in the control of “autonomic,” unconscious, and involuntary functions in the context of total body homeostasis. In general, the wide range of physiologic functions are ultimately necessary for human survival, and allow us to interact with the external environment in a wide range of conditions. Together with the slow-acting, long-lived effects of the endocrine system, the ANS exerts its fast-acting, short-lived effects on the following functions, among other:

- Perfusion of all the body with blood through heart rate and blood pressure control,
- Homoeothermic function through sweating control and shivering,
- Processing of nutrients through control and coordination of different parts of the gut and glands,
- Urinary motility, and
- Pupil movement, focusing and lacrimation.

Dysfunction of one or more subdivisions of the ANS, when accompanying other diseases, is linked to a worse prognosis of the latter. In some circumstances or when severe, dysfunction of ANS itself results in symptoms and disability, which may, in turn, prompt treatment.

## Etiology

Autonomic dysfunction can be caused by a myriad of the following factors, and more than one can concur even in the same patient.

- Degenerative neurologic diseases, such as Parkinson’s disease, multiple-system atrophy, and pure autonomic failure presenting or suffering from Orthostatic Hypotension (OH) and fixed heart rate responses, etc.
- An immune-mediated attack, either idiopathic or paraneoplastic, can be a source of focal or generalized autonomic dysfunction, as in Guillain-Barre syndrome, or in autoimmune autonomic ganglionopathy
- Metabolic and toxic damage, as in diabetes mellitus, amyloidosis and Fabry disease
- Genetic disorders, like in dopamine-beta-hydroxylase deficiency and familial dysautonomia
- Abnormal reflex responses in carotid sinus hypersensitivity, vasovagal syncope, and other neuro-mediated syncopes; and probably in Postural Tachycardia Syndrome (PoTS)
- Abnormal sweating responses, as in generalized or focal hyperhidrosis, related to an excessive activation

- Traumatic or tumoral spinal cord injuries at different levels and presenting with the so-called autonomic dysreflexia
- Pharmacologic effects of several drugs that interfere with normal autonomic function and provoke symptoms, for example, alpha- and beta-blocker-triggered OH

## Epidemiology

Autonomic dysfunction, taken as a whole, is not infrequent. The most common autonomic dysfunction occurs in the cardiovascular control sphere and consists of an abnormal vasovagal response that leads to syncope. Other common manifestations are related to Postural Tachycardia Syndrome (POTS), or changes seen with Parkinson disease and other parkinsonisms. Urinary incontinence, related to other areas of autonomic control, as observed in multiple sclerosis and other nervous system disorders, is unspecific but not rare. There are symptoms of autonomic disturbance in some situations where they are second in importance but help in diagnosis, for example, the facial vasomotor and ocular symptoms in trigeminal autonomic headaches.

## Pathophysiology

Depending on what the area of control of the ANS is affected, the pathophysiology will follow. It is not unlikely to find closely anatomically-related areas of the ANS malfunctioning, and neither it is to find anatomically remote isolated dysfunction. In the cardiovascular system, three pathophysiological syndromes are the most typically related to a chronic dysfunction of the ANS. These are Postural orthostatic tachycardia syndrome, orthostatic hypotension and supine hypertension, and reflex cardiovascular syndromes. Hyperhidrosis and hypohidrosis are the main features in temperature control. Fixed mydriasis and miosis, also known as, Adie's pupil and Horner syndrome in the eye.

## History and Physical

A detailed history of symptoms in each area should guide us to a list of possible diseases with autonomic dysfunction. Place special emphasis on cardiovascular, urinary, and sudomotor symptoms, together with accompanying neurologic and non-neurologic manifestations, along with a thorough examination. A key feature of autonomic dysfunction, either orthostatic syncope or presyncope should guide us to a suspicion of cardiovascular autonomic dysfunction, the latter with dizziness, giddiness, blurred or tunnel vision, headache or neckache (coat-hanger pain), nausea, or fatigue. The alternating sweating intensity in different areas of the body, most typically a length dependent distal hypohidrosis with palms and soles preserved, may be a clue to a sudomotor autonomic lesion. Urgency and incontinence tend to relate to a neurogenic bladder more than retention, but the latter should be addressed in a general exam.

## Evaluation

Testing of the most disturbing or salient features of autonomic dysfunction is often necessary to confirm a diagnosis and give objective evidence to support pharmacologic treatment.

Tilt table test with usual cardiovascular stimuli, for example, Valsalva maneuver, hyperventilation, or cold pressor are among some of the more common types of techniques utilized. This is especially true in the case the bedside blood pressure and heart rate measurements (while supine and after three minutes standing, the orthostatic test) are not informative and the suspicion of an abnormal cardiovascular autonomic function is high. Sympathetic skin responses can add objective information, and thermoregulatory sweat test might be a good option if the symptoms of altered sweating predominate. Vesical ultrasonography and urodynamic studies help figure out what the problem looks like and what the best pharmacologic approach is for urinary dysfunction. Other tests are not as useful in changing the management but may play a role in support of an explanation for the patient's symptoms, or as adjunctive for syndromic diagnosis, as it is the case with blocking eyedrop tests and pupillometry for pupillary abnormalities. On the other hand, positive testing results for autoantibodies, for example, antibodies against alpha-3-acetylcholine receptor, help support immunosuppressive therapy.

## Treatment / Management

Treatment, when indicated, can be organized into three levels. The physiopathologic, symptomatic level is the most frequently used and must address first the most bothersome symptoms while keeping in mind the whole picture. They consist of a wide range of measures from the non-pharmacologic, supportive hydration, salt intake, dietary changes to the alfa-agonist midodrine and mineralocorticoid fludrocortisone for orthostatic hypotension, octreotide for postprandial hypotension, powder desiccants for hyperhidrosis patients. Physical measures like exercise, tailored physiotherapy, pressure stockings can help in some cardiovascular autonomic dysfunctions, and sunglasses for mydriatic pupils. The pathophysiologic aspect can be addressed at least in the case of immune-mediated disorders through immunotherapy, for example, steroids and other immune suppressor drugs, intravenous immunoglobulins, and plasma exchange. Last but not least, etiology must always be addressed and can be as important to treat the malignancy found in paraneoplastic, autoimmune autonomic neuropathy as it is to taper or withdraw the drugs interfering with the ANS in drug-induced, symptomatic OH.

## Pearls and Other Issues

Autonomic dysfunction is a very prevalent health problem that remains underdiagnosed, undertreated, and underappreciated across healthcare systems. In most cases, patients tend to have a late diagnosis, when their symptoms are chronic and long-standing. The lack of acknowledgment from the general population and the medical community, a diagnostic testing set in the development, and a finite set of extensively tested and validated drugs to offer, among other causes, make the field one of the most promising for research.

Autonomic dysreflexia is a severe, potentially life-threatening condition affecting the cardiovascular system of patients exposed to a variety of at-risk exposures and risk factors -- ranging from painful/noxious stimuli to more subtle conditions (e.g. bowel or bladder obstructions). Clinicians should remain cognizant depending on the specific patient and clinical constellation of symptoms. The latter condition can lead to myocardial ischemia, brain hemorrhage, seizures, and even death.

## Questions

To access free multiple choice questions on this topic, [click here](#).

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