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What on Earth is the Autonomic Nervous System? Dysautonomia and Autonomic Dysfunction

Most people have never heard of the Autonomic Nervous System, even though it controls most of the fundamental functions within the body. And when something goes wrong with the Autonomic Nervous System, most people have no idea how devastating that can be, and the wide range of symptoms that can occur.

This edition focuses on Dysautonomia and Autonomic Issues, a subject that affects a wide range of children who are medically complex, but is neither well understood nor discussed as often as it should be by the medical community. This article will attempt to provide a basic overview of how the Autonomic Nervous System works and the types of primary and secondary conditions that can affect Autonomic Nervous System function, causing Dysautonomia or Autonomic Dysfunction.

Overview of the Autonomic Nervous System

Your nervous system consists of the Central Nervous System (brain and spinal cord) and the Peripheral Nervous System, which includes all of the nerves in the body apart from the brain and spinal cord.[1] The Peripheral Nervous System, in turn, is divided into two divisions: the Somatic Nervous System and the Autonomic Nervous System. While the Somatic Nervous System controls the skeletal muscles and can be powered volitionally by the body to jump or lift a hand, the Autonomic Nervous System regulates the glands and muscles on the inside of the body, and governs the body processes that work automatically.

The Autonomic Nervous System is responsible for the unconscious body processes that we simply *expect* to work, such as regulating our body temperature, managing how much blood is pumped to the brain and around the body, and keeping the heart rate and blood pressure in check. It is the Autonomic Nervous System that makes you sweat when you are hot, constricts your pupils when it is sunny, or increases your heart rate when you are in danger.

The Autonomic Nervous System transmits messages via nerves throughout the body using hormones or neurotransmitters. It has three separate parts, the Enteric Nervous System in the gastrointestinal tract which is not well understood, the Parasympathetic Nervous System that regulates “vegetative” activities like eating and digesting, and the

Sympathetic Nervous System, which in tandem with the adrenal glands controls the "fight or flight" responses.

The Autonomic Nervous System works by transmitting messages from nerve terminals to receptors in glands or muscles. For example, to dilate your eyes in response to darkness, a chemical message is sent down the nerve to the eye that then activates a receptor in the eye. This then triggers the eye to dilate appropriately.

In the Parasympathetic Nervous System, neurotransmitters, specifically Acetylcholine, are the chemical messengers that transmit messages to body systems. Acetylcholine triggers receptors that cause processes such as increasing bladder contractions, constricting the pupils, increasing salivation, decreasing the pulse rate, or increasing motility in the gut.

Another neurotransmitter, Norepinephrine, along with the hormone Epinephrine, transmits most messages within the Sympathetic Nervous System. Norepinephrine and Epinephrine trigger effects such as developing goosebumps, dilating the pupils, responding to hot or cold temperatures, constricting blood vessels, slowing the gut, and increasing the heart rate during stress.

Organ or Function[2]	Sympathetic Nervous System Action	Parasympathetic Nervous System Action
Heart Rate	increased	decreased
Skin	constricts	--
Bronchioles (lung)	dilates	constricts
GI muscle	relaxes	contracts
GI sphincters	contracts	relaxes
Bladder walls	relaxes	contracts
Bladder sphincters	contracts	relaxes
Male genitalia	ejaculation	erection
Eye (pupils)	dilates	constricts
Sweat glands	increased	--

In general, the Parasympathetic and Sympathetic Nervous Systems work in opposite ways. For example, the Parasympathetic Nervous System causes the eyes to constrict, while the Sympathetic Nervous System causes them to dilate. While the Parasympathetic Nervous System slows the pulse rate, the Sympathetic Nervous System speeds it up.

When Things Go Wrong: Dysautonomias and Autonomic Dysfunction

Sometimes the Autonomic Nervous System does not work properly. This typically results from either too much activity or not enough activity of any part of the Autonomic Nervous System. While this can be a sudden and temporary event, for many children it is a long term, chronic problem.

There are two basic categories of Autonomic Dysfunction or Dysautonomia. The first category is Primary Dysautonomia, containing genetic disorders and idiopathic conditions. The second type of Autonomic Dysfunction is Secondary Dysautonomia, meaning symptoms result as a side effect or complication of another underlying disease, disorder, or the aging process.

Rare types of Primary Dysautonomias called Hereditary Sensory and Autonomic Neuropathies (HSAN) are caused by genetic disorders. They are characterized by impaired pain sensation and temperature regulation, with many additional symptoms depending on the child and form of the disease. Some are progressive and extremely life-threatening, while others, while still very serious, can be managed. These HSANs include:[3]

- HSAN I: Hereditary sensory radicular neuropathy
- HSAN II: Congenital sensory neuropathy (CSN)
- HSAN III: Familial dysautonomia (FD)/Riley Day
- HSAN IV: Congenital insensitivity to pain with anhidrosis (CIPA)
- HSAN V: Congenital insensitivity to pain with partial anhidrosis
- Congenital autonomic dysfunction with universal pain loss (CAD)
- Progressive panneuropathy

Other Primary Dysautonomias include mild disorders such as Postural Orthostatic Tachycardia Syndrome (POTS), Neurocardiogenic Syncope (NCS), Neurally Mediated Hypotension (NMH), and Vasovagal Syncope. Most of these are characterized by dizziness or fainting and while sometimes difficult to manage, they are not life-threatening. Other more serious forms of Dysautonomia include Autoimmune Autonomic Failure, Pure Autonomic Failure, and Multiple System Atrophy (Shy-Drager Syndrome), among others. Many of these only appear later in life and range from chronic diseases to progressive, fatal disorders.

Secondary Autonomic Dysfunction or Dysautonomia includes any autonomic problems that result as a side effect or complication of another disease. This category encompasses a wide range of conditions, including autonomic symptoms resulting from genetic disorders like Menkes disease, many different forms of mitochondrial disorders, diabetes, and neurological disorders such as cerebral palsy, encephalopathy, and Parkinson's disease. Symptoms may range from isolated and mild (such as heat intolerance), to complicated and life-threatening (such as alterations in heart rate, blood pressure and other vital functions).

An Example of Autonomic Dysfunction in a Child with Cerebral Palsy Secondary to Hypoxic Ischemic Encephalopathy

My daughter suffered a severe injury to her brain and organs due to massive blood loss from an umbilical cord rupture at birth. She was diagnosed with Hypoxic Ischemic

Encephalopathy. Around her third birthday, she became ill with an unusual progression of conditions, including an enterovirus, pancreatitis, and sepsis, and began having increased symptoms of Autonomic Dysfunction. It is unknown whether her Autonomic Dysfunction is due entirely to the severe injury to her brain, or whether her illnesses triggered some sort of autoimmune response, worsening the ability of her Autonomic Nervous System to function appropriately.

These are her Autonomic Symptoms:

- ❑ Does not regulate temperature appropriately; is typically hypothermic with body temperatures as low as 90F; can become hyperthermic (feverish) when exposed to hot temperatures
- ❑ Does not sweat when hot
- ❑ Does not shiver when cold
- ❑ Does not get goosebumps when she is cold (gets them when she is hot instead)
- ❑ Eyes are always dilated and do not respond appropriately to light
- ❑ Typically has high blood pressure
- ❑ Typically is tachycardic (fast heart rate)
- ❑ Has circulation problems, including cold, mottled, purple extremities and no peripheral pulses
- ❑ Develops red blotches, patches on skin, and flushing, especially when stressed
- ❑ Does not sense temperature on skin well
- ❑ Does not sense pain on skin well
- ❑ Neuropathic and visceral pain throughout body
- ❑ Sphincters in bladder are too tight, while muscles are too floppy, requiring cathing
- ❑ Sphincters and muscles in GI tract do not work appropriately, causing motility problems
- ❑ Nerves in GI tract are hypersensitive, causing pain, vomiting, and impaired motility, and requiring parenteral nutrition
- ❑ Bone Marrow production is impaired during stress (drops platelets and hemoglobin)

In my daughter's case, her Autonomic Dysfunction, which is consider moderate in nature, requires multiple daily interventions. These include catheterizing her bladder, use of parenteral (IV) nutrition, constant monitoring of vital signs, extensive pain medication regimen, and lifestyle modifications to address temperature regulation problems, eye dilation, and similar problems. While not all of her issues can be solely attributed to Autonomic Dysfunction, they tend to be exacerbated by her Autonomic Nervous System's inability to work correctly.

An Under-Diagnosed Condition

Autonomic Dysfunction, and especially autonomic issues secondary to neurological conditions like mitochondrial disease and cerebral palsy, is not commonly diagnosed or treated. Even top pediatric neurologists may have little to no experience diagnosing and treating these disorders. While treatments are limited and focus primarily on lifestyle

modifications and supportive measures, in some cases significant medical intervention may be helpful and lifesaving.

If your child is significantly limited by his or her autonomic symptoms, it may be appropriate to contact a specialist in Autonomic Dysfunction. Specialty clinics for children are available at New York University (NYU) Medical Center and the Mayo Clinic.

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[1] For more detailed information on this subject presented in an easy to understand format, see the excellent NDRF handbook at http://www.ndrf.org/NDRFHandbook.htm

[2] Chart adapted from Linda S. Costanzo. *Physiology* (Philadelphia: WB Saunders Company, 1998). 45.

[3] See http://www.med.nyu.edu/pediatrics/fd/hsan/index.html