



# Complex Child E-Magazine

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## Autonomic Events and Crises: An Underdiagnosed Cause of Discomfort by Susan Agrawal

Many children with neurological or metabolic conditions have some degree of autonomic dysfunction, a topic that has been addressed in previous articles.<sup>1</sup> Certain other children have genetic forms of dysautonomia, called Hereditary Sensory and Autonomic Neuropathies (HSANs), that have similar symptoms. All struggle with similar issues ranging from mild to life-threatening symptoms, which may include everything from high blood pressure to low body temperature and motility problems of the gut. Table 1 lists common symptoms that develop in children with primary and secondary autonomic disorders.

Symptoms of dysautonomia, like those of many diseases, may be uncomfortable and painful for many children. But some of these children also experience autonomic events or crises, often on a daily basis, that can have a profound effect on their comfort and well-being. These crises may include nausea, vomiting, high blood pressure, fast heart rate, and other symptoms. The nature of these events and potential treatments for them is the subject of this article.

### **The Autonomic Crisis: Many Names for Similar Events**

Autonomic events and crises can go by many names, often to indicate the underlying cause. For example, individuals with traumatic brain injuries or strokes are often described as experiencing "Sympathetic Storms," a form of autonomic crisis in which the sympathetic nervous system is severely overactivated, usually in the period right after injury. Those with spinal cord injuries may experience Autonomic Dysreflexia, also known as Hyperreflexia, another form of autonomic crisis triggered by an interruption at the level of the spinal cord injury in the signals to and from the brain. For children with Familial Dysautonomia, a form of HSAN that involves crises more than other forms, the term Dysautonomic Crisis is often used. Other older terms include Hypothalamic-Midbrain Dysregulation Syndrome and Diencephalic Epilepsy.

Surprisingly enough, the autonomic crisis has not been described much in the medical literature, especially as it pertains to children with dysautonomia secondary to neurological or metabolic conditions. There is a great deal of literature specific to Autonomic Dysreflexia, and some on Sympathetic Storms after brain injury, but very

Table 1: Common Symptoms of Autonomic Disorders by System<sup>2</sup>

<b>System</b>	<b>Dysfunction</b>	<b>Symptom</b>
Vasomotor/cardiovascular	High blood pressure	Headache
	Low blood pressure	Dizziness, lightheadedness
	Heart rhythm disturbance	Fainting
	Circulatory problems	Cold hands and feet, blotching, blue fingers/toes
Gastrointestinal	Feeding problems	Poor suck, drooling, aspiration pneumonia
	Motility problems of the Esophagus	Difficulty swallowing
	Gastroesophageal reflux	Nausea
	Motility problems of the Stomach and Intestines	Recurrent vomiting, bloating, constipation, diarrhea
Ophthalmologic	No/few tears	Dry eye
	Nonreactive/sluggish pupils	Dark/light intolerance
	Different sized pupils	Nearsightedness
	Drooping eyelid(s)	Strabismus
Respiratory	Shallow or slow breathing	Blue tinge to skin, poor oxygen levels
	Apnea	Breath-holding spells
	Difficulty tolerating low oxygen levels	Fainting
	High CO <sub>2</sub> levels	
Temperature Regulation	Altered sweating	Too much or no sweating
	Altered body temperature	Decreased body temperature, dry skin, unexplained fevers
Neurologic	Altered pain perception	Sensory problems, decreased response to pain, self-mutilation
	Sleep problems	Insomnia
Urologic	Delayed bladder emptying	Bed-wetting
Psychological	Altered affect/mood	Poor social skills, inappropriate laughing/crying, tics
	Unusual emotional responses	Anxiety, phobias
	Learning/cognitive issues	Poor planning, poor school performance, learning disability, attention problems

little that describes events in children with neurological, metabolic, or genetic conditions that lead to autonomic dysregulation.

### **Features of Autonomic Crises and Events**

In an article on Familial Dysautonomia, the authors describe the typical symptoms of a crisis:

Dysautonomic crisis consists of a constellation of signs resembling a central sympathetic storm. Nausea is a consistent and often first feature that can escalate to retching or vomiting. In addition, hypertension [high blood pressure], tachycardia [fast heart rate], diffuse sweating, erythematous [red] skin blotching, and even negative personality change coexist. Sleep difficulties and major difficulties in oral coordination with difficulty swallowing saliva, and reluctance or inability to speak may also be present.<sup>3</sup>

The same article describes the symptoms of an individual child, including congestion, sweating, salivation, blotching, high blood pressure, fast heart rate, extreme irritability, increased respiratory rate, intermittent pulling at his left ear, and upward eye rolling.

Another article describes a few additional symptoms, including excess mucus production, excess gastric secretions, and mood changes, such as whining or withdrawal.<sup>4</sup> It also notes that many patients experience daily crises at about the same time each day, often in the morning upon awaking, likely due to their circadian rhythms.

A list of some of the most common symptoms that occur during autonomic events can be found in Table 2.

Table 2: Common Symptoms of Autonomic Events

- Nausea, retching, gagging, or vomiting
- Flushing or blotching of the face and extremities
- Fast heart rate
- High blood pressure
- Sweating
- Elevated body temperature
- Increased respiratory effort or decreased oxygen saturations
- Increased salivation, increased mucus production and congestion
- Increased gastric secretions
- Shaking or tremoring
- Changes in blood sugar
- Extreme irritability
- Whining, crying or screaming
- Withdrawal
- Difficulty speaking or swallowing
- Confusion

While the above description characterizes children with sympathetic nervous system overactivation, children with Autonomic Dysreflexia experience a combination of sympathetic and parasympathetic symptoms. Below the point of the spinal cord injury, they experience sympathetic symptoms (especially high blood pressure) in response to a trigger, often urinary fullness. These symptoms trigger a reflex to the brain, which responds by causing an overreaction of the parasympathetic nervous system (especially slowing the heart) above the point of injury. Other children with other forms of dysautonomia may also experience parasympathetic overreaction, leading to low blood pressure, a slow heart rate, low body temperature, and similar signs.

Autonomic events and crises can range from mild to very life-threatening. Typically, a mild event may include some nausea and flushing, and will resolve on its own without treatment. On the other end of the spectrum, some children, especially those with sympathetic storms or Autonomic Dysreflexia, may have extremely life-threatening crises that demand immediate intervention and are life-threatening emergencies. In these children, there is often so much activity of the nervous system that their vital signs become completely uncontrollable.

The exact mechanism for most forms of autonomic events is not fully understood at this time, with the exception of Autonomic Dysreflexia.

### **Autonomic Crisis Triggers**

One of the most important steps in treating autonomic events is to determine if there are specific triggers that set off the events or crises. Most triggers involve stresses on the body, and stressors such as infection, fever, overheating, and lack of sleep are common triggers for many children. Pain is also a big trigger for many children, especially the visceral or neuropathic pain that many children with autonomic dysfunction experience.

Some children, especially those with gastrointestinal or urological problems, may have crises or events that are related to dysfunction in those systems. A child who has visceral hyperalgesia or a super-sensitive gastrointestinal tract may trigger events every time she consumes food or formula, or when she is constipated. For children with spinal cord injuries, a very common trigger for Autonomic Dysreflexia is fullness of the bladder. Older girls may find menstruation to be a trigger event.

In some cases, even emotional disturbance may trigger a crisis. Getting very upset, angry, fearful, or excited may be enough to trigger autonomic events or crises.

Each child will have individual triggers, and these must be identified in order to fully treat the child.

## **Treatments for Autonomic Crises and Events**

The most important mechanism is prevention. The more events and crises that you can prevent, the less likely future events will occur. You can think of the progression of autonomic events as similar to a wind-up toy: the more it is wound up, the longer it will continue to play out. Each event that occurs winds up the nervous system a little bit more. If you keep winding up the nervous system with repeated events, it gets more and more wound up, without time to recover and return to baseline. Autonomic events then produce more frequent and more serious other autonomic events.

As such, the general recommendations for treatment are as follows:

- 1) Reduce triggers by treating underlying issues (such as gastrointestinal problems) and avoiding events (such as overheating) that provoke autonomic events.
- 2) When events do occur, intervene quickly and aggressively with rescue medications and comfort measures.
- 3) For frequent (daily or weekly) events, consider a preventative medication.

The primary strategy for prevention is to reduce all triggers. This will, of course, vary with the individual child. Common triggers such as pain, gastrointestinal issues, bladder issues, seizures, infections, overheating, and sleep problems should be treated as aggressively as possible by specialists.

Protocols should be put in place to reduce the impact of unavoidable triggers. For example, if fever is a frequent trigger, medications like Tylenol and Motrin should be administered to prevent autonomic events. During periods of stress, such as during an illness, the child should be treated aggressively with fluids and respiratory support to avoid stressing out the body as much as possible.

If an event does occur, it is important to recognize it early and treat it quickly and aggressively. Parameters should be put in place to guide treatment. For example, a common protocol is to administer oxygen to a child in crisis whenever his heart rate exceeds a certain level. Anti-nausea medications may need to be administered. Similarly, rescue medications may be given when comfort measures do not help or vital signs begin to spiral out of control.

Common treatments for children during a crisis include diazepam (Valium) and other similar drugs that cause sedation, or clonidine, an adrenergic agonist. Children with Autonomic Dysreflexia are usually treated with medications that reduce blood pressure. Dantrolene or morphine may be used for children with severe sympathetic storms. Other children may require specialized protocols related to their specific symptoms.

Children who continue to have frequent and uncomfortable events or crises may benefit from a preventative medication. In the past, the options were limited to alpha and beta

blockers and agonists like clonidine, propranolol, and labetalol. Most children, unfortunately, have complex dysautonomia affecting both the sympathetic and parasympathetic nervous systems, and while these medications may solve one problem (such as lowering blood pressure), they more likely than not will cause new problems. For example, an alpha blocker may help a child urinate with greater ease by relaxing the urinary sphincter, but it may also relax the sphincters in the gut, causing severe reflux and vomiting.

A recent case series has suggested using Pregabalin (Lyrica) as a preventative medication.<sup>5</sup> This medication appeared to reduce the number and severity of crises in children with Familial Dysautonomia significantly. Other similar medications, such as Gabapentin (Neurontin), may also be helpful.

### Find a Specialist

Mild autonomic issues are actually quite common and rarely require the attention of an autonomic specialist. On the other hand, severe autonomic disorders causing frequent or life-threatening autonomic events are rare, and very few doctors, including most pediatric neurologists and endocrinologists, have the knowledge and experience to treat them. If your child suffers from this level of autonomic dysregulation, it is important to consult with a specialist in autonomic disorders who can create a protocol for your child.

Autonomic issues are notoriously difficult to treat, but there are strategies for decreasing the number of events and reducing the discomfort associated with them. Hopefully, additional research in the future will lead to even more possible treatments.

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<sup>1</sup> See, for example,

- What on Earth is the Autonomic Nervous System? Dysautonomia and Autonomic Dysfunction
- Unusual Skin Symptoms in Autonomic Dysfunction: A Pictorial Essay
- Dysautonomia: A Minor Inconvenience Becomes a Devastating Disease

<sup>2</sup> Adapted from Felicia B. Axelrod, *et al.* Pediatric Autonomic Disorders. *Pediatrics* 2006;118(1):309-21.

<sup>3</sup> FB Axelrod, *et al.* Ictal SPECT during autonomic crisis in familial dysautonomia. *Neurology* 2000;55(1):122-5.

<sup>4</sup> Felicia B. Axelrod and Dena Berlin. Pregabalin: A New Approach to Treatment of the Dysautonomic Crisis. *Pediatrics* 2009;124(2):743-6.

<sup>5</sup> Felicia B. Axelrod and Dena Berlin. Pregabalin: A New Approach to Treatment of the Dysautonomic Crisis. *Pediatrics* 2009;124(2):743-6.