Calming the Storm: Dysautonomia for the Pediatrician

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Dysautonomia is a potentially life-threatening syndrome seen in many different types of brain injuries. It involves paroxysmal sympathetic hyperactivity and typically includes a constellation of symptoms, including: tachycardia, tachypnea, hyperthermia, hypertension, diaphoresis, hypertonia, and/or decerebrate or decorticate posturing. It is a clinical diagnosis of exclusion. A multimodal treatment approach is necessary including environmental modifications along with pharmacotherapy. Early management can help prevent comorbidities including secondary brain injury while also improving patient outcomes. This discussion serves as an overview of dysautonomia with a focus on management in the pediatric population including an example of a clinical algorithm and a review of the commonly used medications.

Curr Probl Pediatr Adolesc Health Care 2017;47:145-150

Introduction

Disruption in sympathetic regulation after a brain injury from different causes (including traumatic as well as non-traumatic brain injury such as brain tumor, infectious encephalitis, autoimmune encephalitis, metabolic disorders, and hypoxic-ischemic injuries) is common. It can present with diverse signs of sympathetic dysregulation and motor hyperactivity. This phenomenon has been recognized since at least 1954.[1,2] However, this syndrome still does not have a well-established name and is known by myriad of different terms (over 30)[1]: central autonomic dysfunction, hypothalamic–midbrain dysregulation syndrome, autonomic or sympathetic storming, paroxysmal autonomic instability with dystonia, paroxysmal sympathetic hyperactivity, and dysautonomia—the most commonly used in our experience. Recently, a consensus group identified "paroxysmal sympathetic hyperactivity" as the preferred terminology.[1] However, given its seeming commonality and for the purpose of consistency throughout this article, we will use the term dysautonomia as a surrogate for paroxysmal sympathetic hyperactivity (PSH).

Dysautonomia is a clinical diagnosis of exclusion.

Dysautonomia presents with paroxysmal sympathetic and muscle hyperactivity.[3] There is a host of possible symptoms and signs including increased temperature, increased respiratory rate, increased heart rate, increased blood pressure, diaphoresis, agitation, posturing (decerebrate or decorticate), and hypertonia (spasticity, dystonia, and/or rigidity). These individual symptoms and signs may be brief and only last a few seconds or may persist without intervention. There have been multiple different diagnostic criteria reported.[1,3] In previous work, it has been defined as simultaneous presentation of five or more of the following symptoms: tachycardia, tachypnea, hyperthermia, hypertension, diaphoresis, dystonia, and decerebrate or decorticate posturing[4–9]; some further defined a duration of at least 1 cycle per day for at least 3 days.[4,10] Recently, Baguley et al. on behalf of the Consensus Working Group published an article in 2014 listing 11 clinical items relevant to the diagnosis along with a "symptom severity index" that was combined into a diagnostic tool, the Paroxysmal Sympathetic Hyperactivity—Assessment Measure (PSH-AM).[1] The authors noted that initial data is encouraging but formal validation is not yet complete.[1] This diagnostic tool was developed for adults but adapted to pediatric patients as reported in an article by Pozzi et al.[11] Currently, there is not one set of diagnostic criteria that is widely accepted for clinical practice. It is, however, widely held that dysautonomia is a clinical diagnosis of exclusion and early recognition and treatment of dysautonomia can prevent secondary brain injury.[1]

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Curr Probl Pediatr Adolesc Health Care 2017;47:145-150
1538-5442/$ - see front matter

http://dx.doi.org/10.1016/j.cppeds.2017.06.009

Pathophysiology

As with many aspects of brain injury medicine, the pathophysiology of dysautonomia is not fully clear and is still being studied. At the core, there is excessive activation of the sympathetic system. This sympathetic hyperactivity leads to end organ activation which includes the adrenal glands which in turn release catecholamines. The central and peripheral release of catecholamines leads to the clinical manifestations that we are used to seeing including hyperthermia, hypertension, tachypnea, diaphoresis, and the hypertonic conditions (spasticity, rigidity, and dystonia). What is not fully clear is how injuries to the brain cause the excessive activation of the sympathetic system. Two popular theories are the disconnection theory and the excitatory:inhibitory ratio model.[5,6,12–17] The theories center around the diencephalon (thalamus and hypothalamus), which are thought to be the central excitatory foci during the paroxysmal episodes.[17,18] In the disconnection theory, it is thought that disruption of pathways between the cortex and hypothalamus start the cascade.[5,6,13–16] In the excitatory:inhibitory ratio model, there is thought to be excessive excitatory output due to damage to central inhibitory structures.[17] Clouding the picture further is the question of whether there is some level of cortical control or if there are spinal cord processes that contribute.

Most commonly recognized symptoms of dysautonomia are hyperthermia, hypertension, tachypnea ,diaphoresis, spasticity, rigidity and dystonias.

Dysautonomia is often triggered by a noxious stimulus. There are numerous possible triggers, but some of the more common triggers include pain, a distended bladder, inability to pass stool, abdominal distention, mucous plug, increased respiratory secretions, infection, pressure ulcers, skin wounds, seizure, IV site irritation, passive stretching, tight clothing, and cold tube feeds. The clinical picture can get more confusing when you realize that the same diagnoses on the differential, such as infection and seizure, can occur along with or even trigger dysautonomia.

The most common noxious triggers of dysautonomia include distended bladder, constipation, skin wounds, seizure, tight clothing, and cold tube feeds.

Epidemiology

The onset of dysautonomia in patients with a new brain injury can vary from one day after injury up to 60 days after injury.[10,18] The duration is equally variable, lasting anywhere from days to years.[3,6,18,19] Data on the incidence is variable but a few studies in adult patients with severe brain injuries report a prevalence of 8–10%.[1,4,5,20] Pediatric data is more sparse but the prevalence has been reported at 13–14%.[18,21] It is more common in severe brain injuries.[21,22] Overall, dysautonomia is more common in hypoxic and anoxic brain injuries compared to traumatic brain injuries.[3,21]

Clinical Manifestations

As stated above there are numerous possible symptoms and signs including increased temperature, increased respiratory rate, increased heart rate, increased blood pressure, diaphoresis, agitation, posturing, hypertonia including spasticity, dystonia, and/or rigidity. Vital signs should be analyzed according to a patient's baseline number if available and to age-matched norms. Ongoing symptoms may lead to or be associated with other comorbidities including hyperthermia, weight loss, heterotopic ossification (extra bone formation), cardiac damage, and immune suppression.[3,23–26]

Diagnostic Workup

There are no labs or tests that will confirm the diagnosis of dysautonomia. It is a clinical diagnosis and should be a diagnosis of exclusion. Other diagnoses in the broad differential include but are not limited to: pain, dehydration, an infectious process such as pneumonia or UTI or osteomyelitis, sepsis, seizure, drug fever, rhabdomyolysis, neuroleptic malignant syndrome, narcotic withdrawal, deep vein thrombosis, and pulmonary embolism. The timing of symptom onset to a noxious stimulus, the possible improvement of symptoms after correction of the noxious stimulus and the combination of symptoms occurring together can help clue providers into a diagnosis of dysautonomia. It is important to note, however, that medications may mask some of the symptoms, which can make diagnosis more challenging. As mentioned above, a diagnostic tool, PSH-AM, for adults and an adapted