EDITORIAL

Paroxysmal Sympathetic Hyperactivity: It is Time to Use the New Diagnostic Criteria

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Paroxysmal sympathetic hyperactivity (PSH) is a term used to describe a syndrome of paroxysmal, episodic sympathetic hyperactivity occurring after any acquired brain injury secondary to varied etiologies.[1] This condition was first described by Wilder Penfield in a post-traumatic brain injury (TBI) patient. In 1929, Penfield described a case report of middle-aged woman with brain tumor causing seizures along with sympathetic hyperactivity in the form of hyperthermia, tachycardia, tachypnea, hypertension, excessive sweating, and posturing of the limbs.[2] Since the description, this condition has been referred by other names like sympathetic storms, hypothalamic dysregulation syndrome, paroxysmal autonomic instability with dystonia, diencephalic autonomic epilepsy, etc. The incidence of PSH ranges from 8 to 33% of adults and 13–14% of children following acquired brain injury.[3–6] Paroxysmal sympathetic hyperactivity may be underdiagnosed due to the absence of proper nomenclature and diagnostic criteria. A new diagnostic criterion using clinical features and diagnostic likelihood tool and a formal definition for this condition was proposed by a panel of experts in 2014.[5] The new diagnostic criterion named as the Paroxysmal Sympathetic Hyperactivity Assessment Measure (PSH-AM) combines 6 clinical features and 11 other diagnostic likelihood items to ascertain the probability of PSH in suspected patient groups. Paroxysmal sympathetic hyperactivity assessment measure tool was used in adult retrospective studies and has been validated in adult population. Paroxysmal sympathetic hyperactivity assessment measure clinical features scoring was modified and used in two pediatric studies.[6,7] Using the PSH-AM criteria in suspected patients over 3–4 days reduces the chances of misdiagnosis and improves the overall outcome in these patients.[5] Diagnosis of PSH is made by specific clinical features after excluding other close mimics.

The pathophysiology of PSH is complex and poorly understood. The main hypothesis behind PSH is the loss of cortical inhibition to the sympathetic centers in the diencephalon, brainstem, and spinal cord leading to exaggerated sympathetic responses for any external or internal stimuli. Maladaptive changes at the spinal cord leading to non-noxious stimuli being perceived as noxious stimuli and causing excitatory interneuronal activity are also proposed as the major reason for the excessive stimulation due to non-noxious stimuli.[1,4,6] Magnetic resonance imaging (MRI) changes affecting multiple brain regions typically diencephalon, midbrain, and periventricular white matter are more likely to cause PSH.[1,4,7] Other diagnostic possibilities like severe sepsis/sedation withdrawal should be ruled out by clinical and lab features.

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Evidence for specific pharmacological therapies in PSH is based on small case series. Management of PSH includes a combination of abortive and preventive medications to decrease the frequency and severity of episodes.[1,7–9] Abortive medications include sedatives like morphine, benzodiazepines, propofol, centrally acting B agonists (clonidine, dexmedetomidine), etc. Preventive medications include propranolol, gabapentin, bromocriptine, muscle relaxants (baclofen, dantrolene), etc. Avoidance of all noxious and non-noxious stimuli is the first step in these patients. Prevention of dehydration, contractures, heterotopic ossification, bed sores, and ulcers is also an important part of the overall management and rehabilitation. Paroxysmal sympathetic hyperactivity assessment measure episodes may be frequent during the initial days and may require frequent adjustments of doses and addition of drugs to control the severity of the episodes. Gradually, drugs and doses can be decreased as the number and severity of episodes decrease over time. The duration of symptoms/episodes is very much variable. The overall outcomes of PSH could be related to the severity of the underlying etiology/brain injury or the severity of PSH episodes itself.[1,6]

In this issue of the journal, Pallavi et al.[10] studied the incidence of PSH and its association with outcomes in children less than 12 years old requiring neurocritical care. The study was done in a tertiary care pediatric intensive care unit (PICU) over a period of 10 months. Roughly, a quarter (54 children) of the PICU admissions required neurocritical care and were included in the study. More than 1/3rd (37%) of the study subjects had infective meningoencephalitis as the diagnosis followed by peripheral nerve disorders (17%). Around 63% of the study subjects required mechanical ventilation. The authors used the criteria by Farias-Moeller et al.[11] to define PSH in children. Five

children fulfilled the four out of eight criteria for the diagnosis of PSH and 30 children who had 3 or less of the eight criteria were labeled as incomplete PSH. Four out of five children with PSH had infective meningoencephalitis, and one had Guillain–Barre syndrome. The mean duration of PSH was 26 days and the mean number of PSH episodes was 116 in these 5 children. Children with PSH had higher pediatric risk of mortality score (PRISM-3 SCORES), longer PICU stay, and ventilator requirements compared to children with incomplete PSH. Mortality was comparable in both groups.

The authors have studied about a relatively uncommon condition with limited pediatric literature. The literature regarding PSH has increased over the last 2 decades and is mostly adult literature. The study population had a mixture of neuro-critical patients rather than a specific group like TBI.

Few pediatric and adults[3,4,12] studies used 6–7 criteria (fever, tachycardia, tachypnea, hypertension, sweating, dystonia, or tonic posturing) for diagnosing patients to have PSH after ruling out other differential diagnosis. Farias-Moeller et al.[11] used 8 criteria (fever, tachycardia, tachypnea, hypertension, sweating, muscle rigidity, decreased level of consciousness, and pupillary dilatation) in critically ill children with meningoencephalitis and labeled affected children to have PSH if they satisfy 4 out of 8 criteria. Alofisan et al.[6] used PSH-AM criteria with modifications to the clinical feature score and looked at the incidence of PSH after severe TBI in children. The authors[10] used 4 out of 8 clinical features to diagnose PSH. The criteria encephalopathy and pupillary dilatation are not specific for PSH. Modifications of PSH-AM clinical variables for appropriate age groups along with the diagnostic likelihood tool would have been a better diagnostic option for PSH compared with 4 out of 8 clinical criteria.

Alofisan et al.[6] mentioned confounders for clinical features like the presence of culture-positive sepsis/sedation withdrawal/inotropes use/improper sedation, pain etc. The authors[10] should have given more information about culture-positive sepsis/pain and sedation management/sedation withdrawal/inotrope use in the study subjects considering the longer median duration of ventilation/length of stay (39 days) in the children with PSH.

The majority of the studies have looked at the neuroimaging findings[4,13–15] (site of involvement and specific features) and the association with the presence or absence of PSH, whereas there were no neuroimaging details mentioned by the authors. The authors' finding of longer duration of stay in the PSH group and comparable mortality among both the groups was similar to other studies.[6,11]

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