ECRONICON OPEN ACCESS

EC PAEDIATRICS
Case Report

# Syndrome of Paroxysmal Sympathetic Hyperactivity as a Variant of the Complicated Course of the Postoperative Period in a 9-Month-Old Child with Neurosurgical Pathology. Clinical Observation

**Savvina IA[1,2,3]*, Gurskaya VI[1], Blinov SA[3,4] and Alexandrovich Yu S[4]**

[1]*Almazov National Medical Research Center, St. Petersburg, Russia*

[2]*North-West State Medical University Named After I.I. Mechnikov, St. Petersburg, Russia*

[3]*Children's Region Clinical Hospital, St. Petersburg, Russia*

[4]*St. Petersburg State Pediatric Medical University, St. Petersburg, Russia*

***Corresponding Author:** Savvina IA, Almazov National Medical Research Center, St. Petersburg, Russia.

**Received:** October 29, 2024; **Published:** November 13, 2024

## Abstract

The paper describes a clinical observation with a complicated course of the postoperative period in the form of the development of paroxysmal sympathetic hyperactivity (PSH) syndrome in the infant after acute cerebral injury caused by neurosurgical intervention to remove the formation of the oculomotor nerve on the left. The issues of diagnosis and intensive therapy of PSH syndrome are considered.

***Keywords:*** *Neurosurgery; Paediatric Patients; Acute Cerebral Injury; Paroxysmal Sympathetic Hyperactivity Syndrome*

## Introduction

Acute cerebral injury in infants and children after neurosurgical intervention for intracranial neoplasms of the brain is often the cause of severe neurological deficiency caused by hemorrhage into a tumor, the development of cerebral vasospasm, the formation of hydrocephalus, infectious complications and paroxysmal sympathetic hyperactivity (PSH) syndrome [1-6]. The purpose of the publication of this clinical observation is to demonstrate the PSHA syndrome in a child a younger age group after acute cerebral injury caused by neurosurgical trauma due to the removal of a tumor of the left oculomotor nerve. A feature of this clinical observation is early childhood, acute cerebral injury with the development of secondary complications in the form of cerebral vasospasm and hemispheric ischemic stroke, severe course of PSHA syndrome, multiple cerebroprotective therapy [7,8].

## Clinical Observation

Patient A., 9 months old, was admitted to the Children's Region Clinical Hospital with the main clinical diagnosis: volumetric formation of the left oculomotor nerve (schwannoma?). Hemangioma of the right parietal region. Upon admission, left-sided mydriasis, left-sided exophthalmos and ptosis were noted. It is known from the anamnesis of the disease that for the 1st time parents paid attention to the different size of the pupils 3 days before the moment of hospitalization, during the examination period. It is known from the anamnesis of life that the child was full-term, grew and developed according to age, on the 22nd day of life the child underwent surgery for herniation on the right for a pinched inguinal hernia, and later the period of life proceeded without peculiarities.

A neuroimaging examination on the first day of hospital stay, performed using contrast-enhanced MRI, revealed a voluminous formation extending into the cavity of the left orbit and intracranially into the middle cranial fossa (Figure 1). A formation with smooth, clear contours with dimensions of 2.8 x 1.3 x 2.0 cm, moderately heterogeneous MR-structures in central departments, isointensive on all types of weighting. The formation is located along the oculomotor nerve. It has a mass effect on the left optic nerve intraorbitally, pushing it medially, without distinct signs of germination, with signs of edema of the left optic nerve in the form of an uneven increase in the MR signal from it in the adjacent sections, has a mass effect on the anterior lower-medial sections of the left temporal lobe, pushing it laterally, without distinct signs of germination, without signs of perifocal edema.

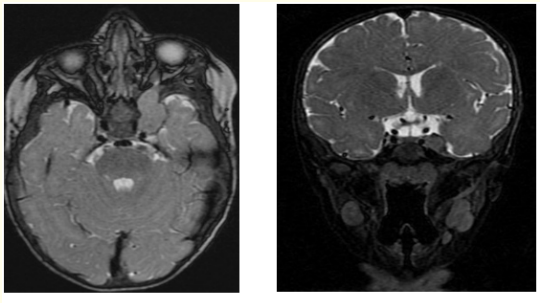


***Figure 1A and 1B:*** *MRI of the brain of patient A., 9months old. Illustration of volumetric formation on a series of tomograms.*

Surgical intervention was performed - subtotal removal of the tumor of the left oculomotor nerve. The duration of the operation was 5 hours. The intraoperative period was smooth. The hemodynamic profile remained stable, laboratory data remained within the reference values, and no centrogenic reactions were registered at the main stage. In the postoperative period, drug-induced sleep was continued for 7 hours in order to complete hemostasis in the postoperative area, as well as to prevent complications against the background of hemodynamic rearrangements in the surgical area. After 7 hours, the patient was extubated against the background of spontaneous breathing, satisfactory muscle tone and stable blood gas parameters. 10 minutes after extubation, there was a sharp deterioration in the condition in the form of generalized convulsive activity, depression of consciousness up to 8 points on the Glasgo Coma Scale, and a decrease in saturation. In this connection, an emergency reintubation was performed, followed by the resumption of neurovegetative stabilization: fentanyl, benzodiazepines, propofol.

In a control study of MRI of the brain with contrast enhancement, there was a picture of partial removal of the intracranial component of the volumetric formation of the left oculomotor nerve; subaponeurotic fibroma of the right parietal region, ischemic stroke in the left internal carotid artery (Figure 2).

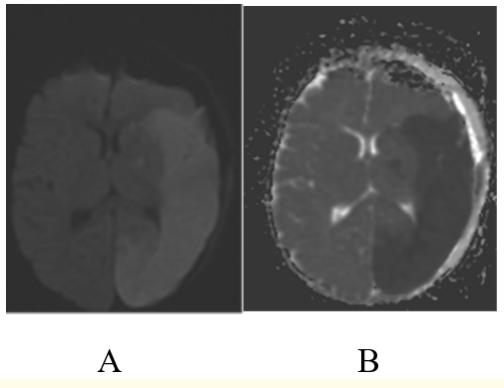


***Figure 2A and 2B:*** *Magnetic resonance imaging of the brain of patient A. Ischemic stroke in the left middle cerebral artery with a predominant lesion of the temporo-occipital region in the cortical-subcortical sections.*

With MR angiography, the picture of anterior trifurcation of the left internal carotid artery. Lack of blood flow along the left ICA and left CMA, decreased blood flow along the left CMA. Depletion of peripheral blood flow in all basins is most likely due to vasospasm (Figure 3).

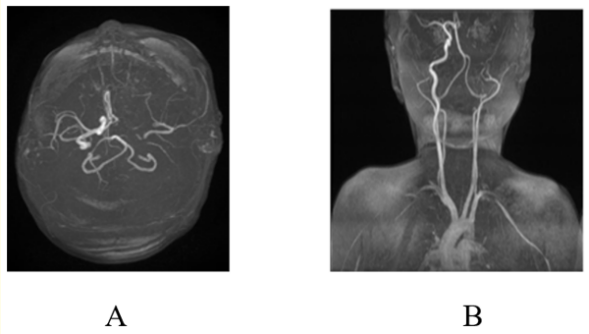


***Figure 3A and 3B:*** *MR angiography of the cerebral vessels of patient A. The left ICA and the left CMA are not visualized. The blood flow along the left posterior cerebral artery is sharply reduced, threadlike. The left ACA is traced in the A1 segment in fragments. Anterior trifurcation of the right ICA is determined.*

Transcranial duplex scanning (TCDS) data of arteries and veins - blood flow asymmetry in the cerebral hemispheres is visualized, 50% less in the left hemisphere, blood flow in the left hemisphere is reduced; RI index is not changed: SMA - on the right, systole - 70 cm/s, diastole - 21 cm/s, on the left - systole - 32 cm/s, diastole - 13 cm/s, index - RI - 0.54-,69; PMA - on the right, systole - 42 cm/s, diastole - 16 cm/s, on the left, systole - 25 cm/s, diastole - 12 cm/s, index - RI - 0.55-,62; ZMA - on the right, systole - 65 cm/s, diastole 21 cm/s, index RI - 0.57-,69, left, systole - 27 cm/s, diastole - 12 cm/s, index RI - 0.57-,69.

Duplex scanning of intracranial sections of brachiocephalic arteries - extracranial: vascular sections of the carotid department, duplex scanning, asymmetry - asymmetric in ICA, 50% smaller on the left: ICA on the right - 106 cm/s, on the left - 50 cm/s, vessel diameter: on the right - 3 mm, on the left 2.8 mm, vascular tone - not changed, venous outflow - not obstructed, compression tests - not carried out because of the critical condition of the child.

Vertebral-basilar basin: vertebral arteries d - 66 cm/s, sufficient blood flow, vascular tone - not changed, venous outflow - not obstructed, diameter of the right vertebral artery, mm - 2.7 mm, smooth stroke, left vertebral artery - 42 cm/s, 30% less than on the right, sufficient blood flow, tone: - not changed, venous outflow: - not obstructed, diameter of the left vertebral artery, mm - 2.8 mm, smooth stroke, blood flow a. Bas1lar1s - visualized through the fontanel - failed, head rotation tests - not performed because of the critical condition of the child.

With a duplex of the vessels of the neck, asymmetry of blood flow along the ICA was revealed, 2 times less on the left, most likely against the background of vasospasm; asymmetry of blood flow through the vertebral arteries, 30% less on the left (Figure 4).

Taking into account the revealed vasospasm along the cerebral arteries, therapy was modified with the appointment of a selective calcium channel blocker of class II, a derivative of dihydropyridine - nimodipine 15 - 30 mg/kg/h, the addition of cerebroprotective therapy - Inosine + Nicotinamide + Riboflavin + Succinic Acid, as well as correction of doses of neurovegetative stabilization [11-13].

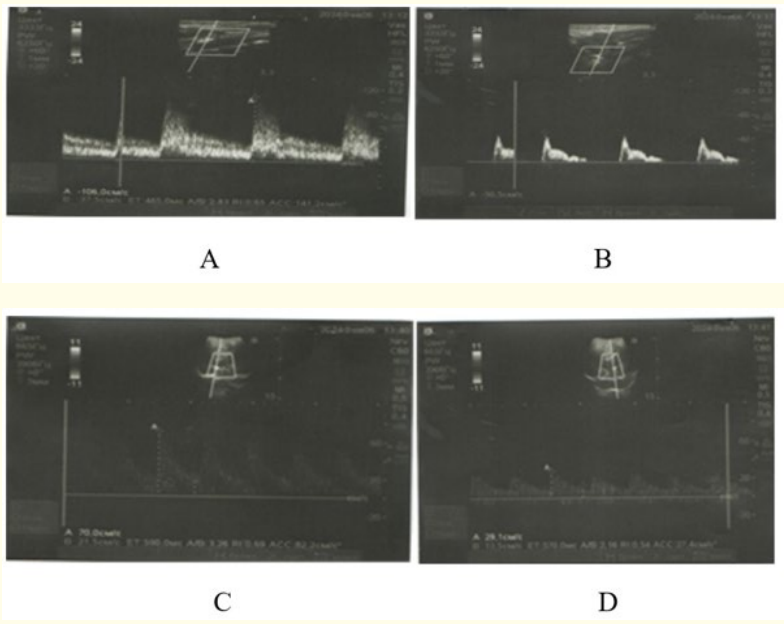


***Figure 4A-4D:*** *Transcranial duplex scanning of arteries and veins. A - right internal carotid artery; B - left internal carotid artery; C - right cerebral middle artery; D - left cerebral middle artery. The asymmetry of blood flow in the cerebral hemispheres is visualized, 50% less in the left hemisphere, blood flow in the left hemisphere is reduced.*

Further, on the 5th postoperative day, a control MRI examination of the brain was performed, according to the results of which the acute ischemic stroke was determined in the left ICA; left CPA; right CMA (pole of the frontal lobe). Dislocation of the median structures, descending tentorial wedge on the left. Signs of occlusive subcompensated hydrocephalus with dilation of the right lateral ventricle (Figure 5).

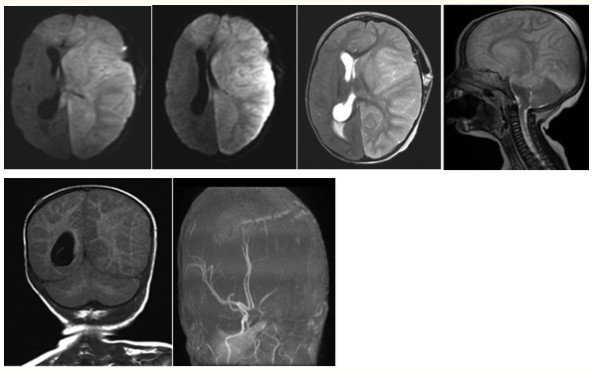

***Figure 5:*** *MRI of the brain of patient A. An extensive ischemic stroke in the left frontal and parietal lobes, the pole region of the right frontal lobe. The stem structures are shifted to the right and down, partially compressed. A descending tentorial wedge on the left. Occlusive subcompensated hydrocephalus with dilation of the right lateral ventricle. Decreased blood flow along the left ICA and left CMA, left CMA.*

Against the background of manifestations of unstable hemodynamics, revealed dislocation of median structures, as well as signs of the brain structures insertion, an emergency operation was performed - decompressive craniotomy of the left temporal, parietal and partially left frontal bones. In the postoperative period, hemodynamic correction was performed using inotropic and vasopressor support with dopamine and norepinephrine, anticonvulsant therapy was also performed, decongestant therapy and deep neurovegetative stabilization were continued.

On the 11th day after the first operation without vasopressor support of hemodynamics, body hyperthermia indicators up to 39.9°C with isothermia were recorded, high systolic blood pressure values were also recorded, this condition was accompanied by pronounced tachycardia up to 160 beats per minute, when evaluating the vegetative Kerdo index - a sharply positive value (up to 69). Against the background of hyperthermia of central origin, stable arterial hypertension, marbling of the skin, inconstancy of thermoregulation were observed. These clinical manifestations were extremely difficult to control: minimal positive reaction to antipyretics, physical cooling methods, jet injection of sedatives. This pathological condition was regarded as a manifestation of severe paroxysmal sympathetic hyperactivity (PSH) syndrome. In this connection, the doses for NSAIDs were adjusted up to the maximum [10-12].

When registering the EEG under condition of pharmacological neurovegetative stabilization, an interhemispheric asymmetry of background activity was noted in the form of gross suppression of bioelectric activities in the left hemisphere; low-amplitude, very slow delta activity was recorded in the right hemisphere. Electrographic cyclic organization of physiological states “wakefulness/sleep” - on the right, physiological phenomena of sleep (sleep spindles); on the left, none. No epileptiform activity or epileptic seizures were recorded (Figure 6).

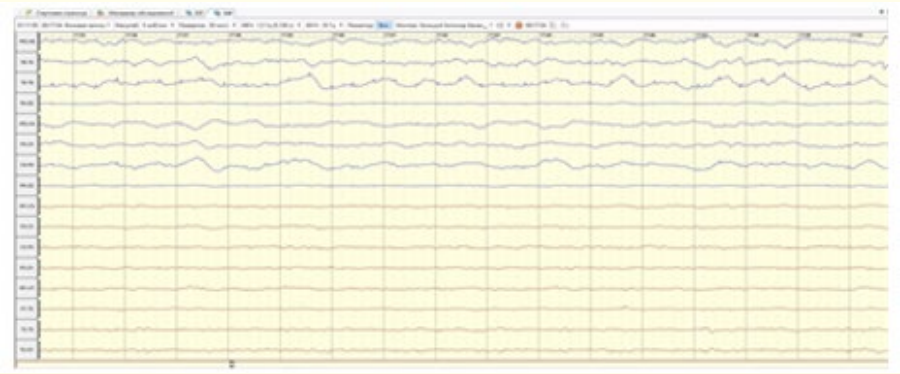

***Figure 6:*** *EEG under condition of neurovegetative stabilization fentanyl+propofol. Interhemispheric asymmetry of background activity, electrographic cyclic organization of physiological states "sleep/wakefulness" - on the right, physiological phenomena of sleep (sleep spindles); on the left, none.*

According to the study of somatosensory evoked potentials (SSEP), symmetrical signs of slowing down of conduction at the ponto-mesencephalic level are recorded from two sides. Cortical responses are not recorded in the study of visual VP (SEP) from both sides. According to acoustic stem evoked potential data, symmetrical signs of slowing down of conduction at the ponto-mesencephalic level are recorded from two sides.

In view of the obtained EEG and EP data on bioelectrical activity of the brain, indicating a pronounced functional violation of the unified system, neurovegetative stabilization was continued, with subsequent monitoring of the electroencephalogram.

On the 21st postoperative day of stay in the ICU, the patient's condition with negative dynamics in the form of the appearance of abundant liquid intestinal contents of the "melena" type, clinical data in the form of an increase in tachycardia, a moderate decrease in saturation, the appearance of oxygen dependence indicated accompanied anemia on the background of bleeding in the gastrointestinal tract. A decrease in hemoglobin and hematocrit was confirmed in the laboratory. Esophagoduodenoscopy was performed urgently, followed by clipping of the duodenal bulb ulcer, from which bleeding was detected (Figure 7).

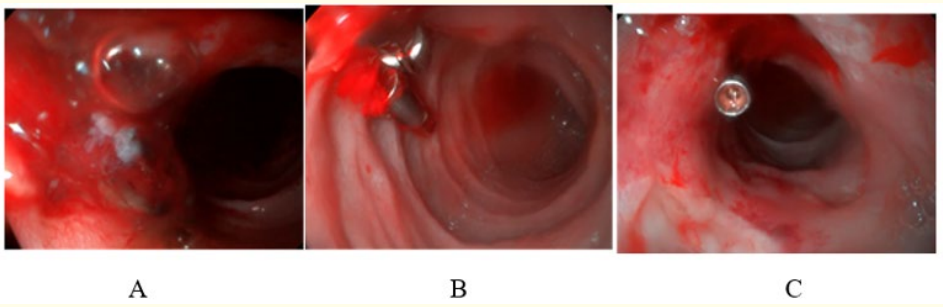


***Figure 7:*** *Esophagoduodenoscopy with clipping of the ulcer of the duodenum bulb in patient A.*

The hemoglobin level against the background of developed gastrointestinal tract bleeding reached extremely low values - 63 g/l, in connection with which transfusion of erythrocyte suspension and transfusion of freshly frozen plasma was performed. In the following days, the patient's condition remained severe, stable, and the laboratory data corresponded to the reference values.

On the 22nd day after surgery, the control of cerebral vascular TCD was performed, where significant asymmetry of blood flow was still preserved (Figure 8A and 8B).

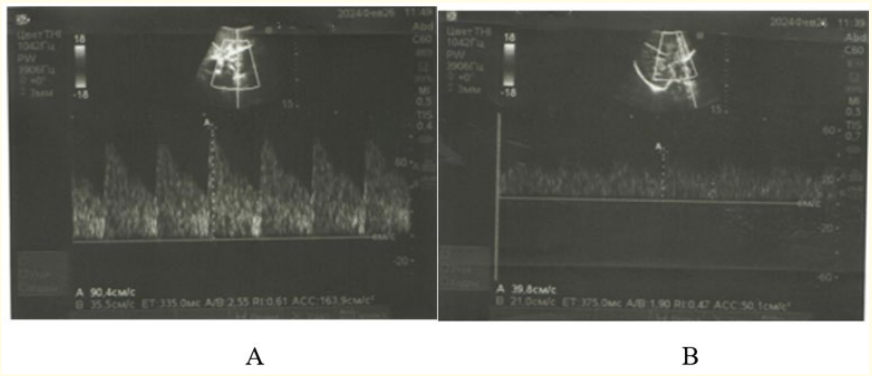


***Figure 8A and 8B:*** *Transcranial duplex scanning of arteries and veins. A - right CMA; B - left CMA. Absence of vascularization of the left hemisphere of the brain; improvement of vascularization of the right hemisphere of the brain in dynamics.*

Also, on the 22nd day, the patient was removed to the diagnostic window. With a gradual sequential decrease in doses of NSAIDs, manifestations of PSH increased in the form of increasing tachycardia up to 150-170 beats per minute, an increase in arterial hypertension, the appearance of bright hyperemia of the cheeks and upper half of the trunk, a rise in body temperature to 38.9°C without effect on the administration of antipyretics, also a decrease in doses of NSAIDS led to resynchronization with a ventilator, episodes of decreased saturation, at the same time, attempts to switch to independent breathing in CPAP mode are unsuccessful.

Thus, on day 22, PSHA syndrome remained the dominant pathological condition in the severity of the patient's condition, and therefore the alpha2-adrenoagonist clonidine was added to the structure of the pharmacological neurovegetative stabilization. Clinical manifestations of PSH were with some regression, however, when recording EEG against the background of clonidine without fentanyl, benzodiazepines and propofol, an interhemispheric asymmetry of background activity continued to be recorded in the form of gross suppression of bioelectrical activity in the left hemisphere, while no epileptiform activity was recorded (Figure 9).

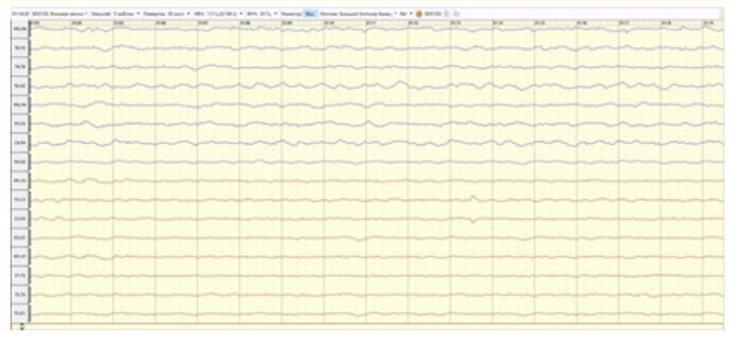

***Figure 9:*** *Electroencephalogram of patient A. under condition of clonidine 0,4 mcg/kg/h. Interhemispheric asymmetry of background activity (gross suppression of bioelectrical activity in the left hemisphere; in the right hemisphere, a drop in amplitude parameters by 35%), "sleep/wakefulness" - on the right, physiological sleep phenomena (sleep spindles); on the left, there are no.*

Over the next 3 days, therapy with NSAIDs, fentanyl, propofol and clonidine, were continued. Then the gradual cessation of the administration of fentanyl and propofol was performed, clonidine was left for another 3 days.

On the 31st postoperative day, extubation and self-respiration were performed. The clinical manifestations of PSH were moderate.

On the 36th day, the patient was transferred to the specialized department for further observation and rehabilitation measures. During the observation period, there was a gradual restoration of consciousness to moderate deafness (Figure 10). Rehabilitation measures were initiated in the acute period of brain damage and included proper positioning and laying to prevent spasticity and trophic disorders, constant passive exercises to prevent contractures in the joints. During EEG registration, the interhemispheric asymmetry of background activity continues to be recorded in the form of gross suppression of bioelectrical activity in the left hemisphere.

***Figure 10:*** *Patient A. on the 36th day after surgery to remove a tumor of the left oculomotor nerve.*

## Discussion

In the presented clinical example, we observed a variant of the development of impaired consciousness with manifestations of PSH, due to the consequence of cerebral brain damage as a result of neurosurgical intervention and the development of cerebral vasospasm. The acute period of cerebral insufficiency was characterized by signs of pathological organization of brain function with depression of consciousness and signs of PSH.

It should be noted that paroxysmal hyperactivity of the sympathetic nervous system in response to brain damage is one of the manifestations of a stress reaction. The pediatric paroxysmal sympathetic hyperactivity scoring reference is represented in the table according to Pozzi M., *et al*. 2014 [10], but there are no data concerning newborns and infants.

| | 0 | 1 | 2 | 3 |
|---|---|---|---|---|
| Heart rate | 1-4 years: < 110<br>5-15 years: < 100 | 1-4 years: 110-124<br>5-15 years: 100-119 | 1-4 years: 125-139<br>5-15 years: 120-139 | 1-4 years: ≥140<br>5-15 years: ≥ 140 |
| Respiratory rate | 1-4 years: < 30<br>5-15 years: < 25 | 1-4 years: 30-34<br>5-15 years: 25-29 | 1-4 years: 35-39<br>5-15 years: 30-34 | 1-4 years: ≥ 40<br>5-15 years: ≥ 35 |
| Systolic blood pressure | 1-4 years: < 100<br>5-15 years: < 120 | 1-4 years: 100-109<br>5-15 years: 120-129 | 1-4 years: 110-119<br>5-15 years: 130-139 | 1-4 years: ≥ 120<br>5-15 years: ≥ 140 |
| Diastolic blood pressure | 1-4 years: < 65<br>5-15 years: < 75 | 1-4 years: 65-72<br>5-15 years: 75-82 | 1-4 years: 73-79<br>5-15 years: 83-89 | 1-4 years: ≥ 80<br>5-15 years: ≥ 90 |
| Temperature | < 37°C | 37-37.9°C | 38-38.9°C | ≥39°C |
| Sweating | Normal | Increased sweating | Localized diaphoresis | Generalized diaphoresis |
| Muscle tone increase | Absent | Mild increase | Neat increase | Generalized spasticity<br>or opisthotonus |

***Table:*** *Pediatric paroxysmal sympathetic hyperactivity scoring reference [10].*

In our opinion, an important point is the timely correction of the signs of PSH by creating a "pharmacological dominant", as well as relieving pathological afferentation of pain using analgesics and/or, if possible, eliminating the cause of pain. The determination of the severity of PSH syndrome also underlies the assessment of the adequacy of the treatment of acute cerebral injury, namely, the method of neurovegetative stabilization. When assessing the PSH syndrome in children with acute cerebral insufficiency, it should be remembered that the regulation of cardiac activity is due to a combination, on the one hand, the insufficiency of mature sympathetic regulatory mechanisms, and on the other hand, the insufficiently pronounced influence of the parasympathetic division of the autonomic nervous system. In this case, the criterion of adequacy was the results of dynamic observation of clinical, laboratory and instrumental research methods.

## Conclusion

This clinical observation demonstrates the important of the key principles of intensive care of acute cerebral damage - neurovegetative stabilization, and understanding the neuroanatomical basis of paroxysmal sympathetic hyperactivity for the successful treatment of PSH.

## Bibliography

1. Baguley IJ., *et al*. "Paroxysmal sympathetic hyperactivity after acquired brain injury: consensus on conceptual definition, nomenclature, and diagnostic criteria". *Journal of Neurotrauma* 31.17 (2014): 1515-1520.

2. Meyfroidt G., *et al*. "Paroxysmal sympathetic hyperactivity: the storm after acute brain injury". *Lancet Neurology* 16.9 (2017): 721-729.

3. Podell JE., *et al*. "Paroxysmal sympathetic hyperactivity after traumatic brain injury: a case-control study". *Critical Care Medicine* 49.10 (2021): e989-e1000.

4. Godoy DA., *et al*. "Paroxysmal sympathetic hyperactivity: An entity to keep in mind". *Medicina Intensiva* 43.1 (2019): 35-43.

5. Thomas A and Greenwald BD. "Paroxysmal sympathetic hyperactivity and clinical considerations for patients with acquired brain injuries: a narrative review". *American Journal of Physical Medicine and Rehabilitation* 98.1 (2019): 65-72.

6. Louraoui SM., *et al*. "Paroxysmal sympathetic hyperactivity after traumatic brain injury: What is important to know?" *Cureus* 14.5 (2022): e24693.

7. van Eijck MM., *et al*. "The use of the PSH-AM in patients with diffuse axonal injury and autonomic dysregulation: A cohort study and review". *Journal of Critical Care* 49 (2019): 110-117.

8. Hinson HE., *et al*. "Neuroanatomical basis of paroxysmal sympathetic hyperactivity: a diffusion tensor imaging analysis". *Brain Injury* 29.4 (2015): 455-461.

9. Pozzi M., *et al*. "Clinical scales for paroxysmal sympathetic hyperactivity in pediatric patients". *Journal of Neurotrauma* 31.22 (2014): 1897-1898.

10. Rabinstein AA and Benarroch EE. "Treatment of paroxysmal sympathetic hyperactivity". *Current Treatment Options in Neurology* 10.2 (2008): 151-157.

11. Savvina IA., *et al*. "Cerebral vasospasm: Mechanisms, Pathomorphology, Diagnostics, Treatment". In: Cerebrovascular Diseases - elucidating Key Principles/ Ed. by Dr. Patricia Bozzetto Ambrosi. IntechOpen, London (2022): 138.

12. Savvina IA and Petrova AO. "The treatment of acute stroke/ischemic stroke". Ed. by Pratap Sanchetee. London: IntechOpen (2021): 85-102.

13. Savvina IA., *et al*. "Management of pediatric severe traumatic brain injury: what are news?" *EC Anaesthesia* 6.10 (2020): 1-14.

**Volume 13 Issue 12 December 2024**
**©All rights reserved by Savvina IA., *et al.***