Periodic Spontaneous Hypothermia Syndrome (PSHS): Looking Beyond Shapiro Syndrome

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Periodic Spontaneous Hypothermia Syndrome (PSHS) is an extremely rare condition, only around 70 cases has been reported since initially it was identified in 1969 as Shapiro syndrome, characterized by a triad of: episodic hypothermia, hyperhidrosis and agenesis of corpus callosum. Recently with the finding of episodic hypothermia and hyperhidrosis seen with COVID-19 cases it has been postulated that PSHS could be a variant of Shapiro syndrome.

Introduction

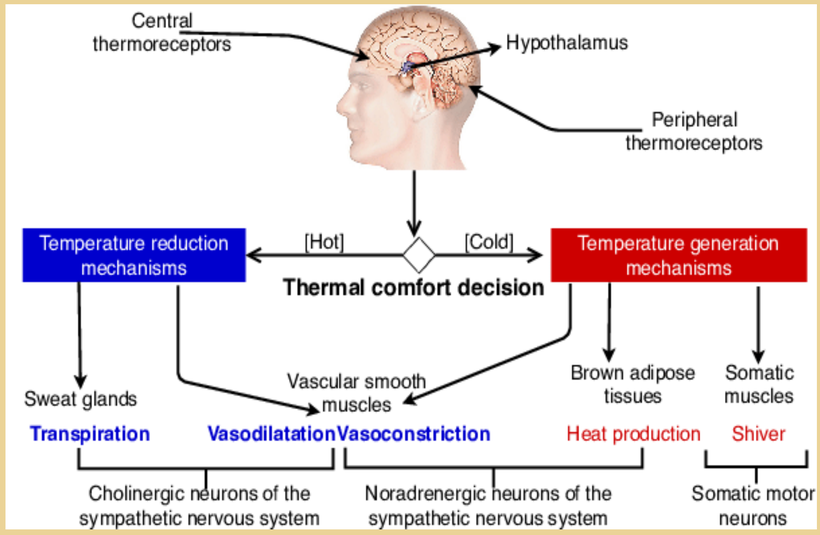
Hypothermia is defined as core body temperature of <95°F or <35 °C. PSHS is a rare condition. Only around 70 cases has been reported since initially it was identified in 1969 as Shapiro syndrome.

Shapiro syndrome is characterized by episodic hypothermia, hyperhidrosis and agenesis of corpus callosum. Some form of congenital or acquired thermal regulation dysfunction of hypothalamic or perioptic area is suspected as the cause of thermo-dysregulation. Many acquired causes are considered as likely causes including neurotransmitter disequilibrium especially of dopaminergic system, inflammatory process, autoimmune disorders and epilepsy to name a few.

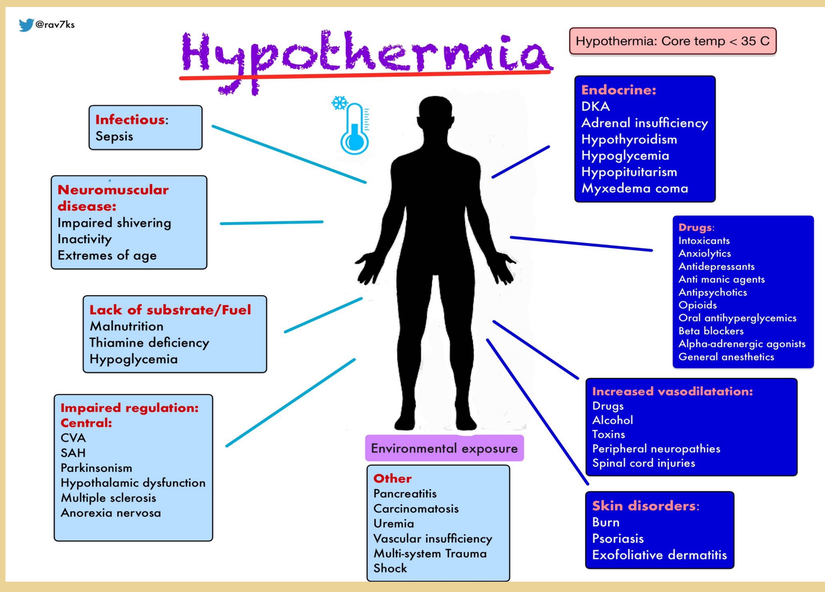
We present 2 psychiatric patients with PSHS, both had history of COVID-19, who were on antipsychotics for many years. Anti-psychotics, anticonvulsants and at times serotonergic medications all can worsen these hypothermic episodes. In absence of any of these medications management of psychiatric patients with PSHS is much more challenging then other PSHS patients.

There has been several reports of PSHS and episodic hyperhidrosis and hypothermia after the recovery from COVID-19. The patient may experience episodes of profound hypothermia, triggered by minimal exertion.

Thermoregulation Mechanism



Causes of Hypothermia



Cases of PSHS

Case Report I: JW is a 52 years old, black male on disability, living in AFC for over 12 years. History of recurrent episodes of hypothermia, pituitary adenoma, high lipids, HTN, intellectual disability and schizophrenia, presented with altered mental status, with core body temp of 89.9°F (32.1 °C). Labs were normal except: Glucose 59, ALP 144. Brain imaging unremarkable. On lithium, clozapine, quetiapine and bromocriptine at home. History of numerous inpatient psychiatric hospitalizations. Psychiatrically stable since last psych admission in 2007. This was patient's 6th presentation with PSHS to this hospital since Sept 2019. History of COVID infection in Feb 2021.

Case Report II: VB is 62 years old, black female, on lifelong disability, living in AFC for last 20 years. Brought to the ED by police after she was found wondering in Walmart late at night. She has type 2 diabetes, CKD stage III, schizoaffective disorder and intellectual disability. Presented with core body temp of 90.3 °F (32.4° C). Currently only taking Klonopin 0.5 mg QHS, all antipsychotics have been discontinued since Dec 2022 after she started developing PSHS. Patient has presented with PSHS, 4 times to this hospital since Feb 2021. Labs normal except U/A: LE, Nitrite and Bacteria +ve. Brain imaging unremarkable for preoptic or hypothalamic lesion. History of COVID outbreak in AFC, she was sick but refused COVID test.

References

1. Blondin NA. Diagnosis and management of periodic hypothermia. Neurol Clin Pract. 2014 Feb;4(1):26-33.
2. Attout H et al. Spontaneous Periodic Hypothermia in the Elderly: A Rare or Under-Recognised Syndrome. Eur J Case Rep Intern Med.2020 Sep 28;7(12)
3. Ren L et al. A new perspective of hypothalamic disease: Shapiro's syndrome. Front Neurol. 2022 Jul 29;13:9.
4. Lamotte G, Benarroch EE, Coon EA. Paroxysmal hypothermia and hyperhidrosis with exacerbation after COVID-19 Infection. Clin Auton Res. 2021 Apr;31(2):327-329.
5. Tansey EA, Johnson CD. Recent advances in thermoregulation. Adv Physiol Educ. 2015 Sep;39(3):139-48.

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- Kizito Nkurikiyeyezu PhD. An efficient thermal comfort delivery in workplaces. Conference Paper · March 2019 (Figure 1)
- Dr. Ravi Singh.Cuses of Hypothermia. Academic Hospitalist and APD @SinaiBmoreIMRes, Clinical reasoning, Simulation and POCUS enthusiast - https://twitter.com/rav7ks (Figure 2)

Scan for full abstrct

Conclusion

Both of our patients were black adults with intellectual disabilities and chronic psychotic illness, on multiple antipsychotics for many years. By this presentation we hope that we can enhance the understanding of PSHS to help treat patients with PSHS in general medical units in a better way.