

Phenomenology of Malignant Hypothermia in PWS

Janice Forster MD¹, Francie McDougall RN², Jacqueline Durette APRN, BC² and Linda Gourash, MD¹

¹Pittsburgh Partnership, Pittsburgh PA; ²Latham Centers, Brewster MA

Background: Persons with PWS have difficulty maintaining core body temperature when ambient temperature changes. In addition to faulty homeostatic mechanisms in the hypothalamus, there is reduced transmission of temperature sensation from the skin; impaired judgment that interferes with selection of appropriate clothing to match the weather; and cognitive inflexibility that interferes with compliance for seasonal changes in clothing. A few individuals have had episodes of sustained (malignant) hypothermia <92°F, severe enough to be life threatening, and these individuals appear to be susceptible to experiencing subsequent episodes.

Methods: Literature review and presentation of recent cases of hypothermia in PWS informs the discussion of precipitating factors, phenomenology of symptoms, morbidity, methods of successful intervention and recommended strategies for prevention.

Results: Middle aged persons with PWS appear to be more susceptible to malignant hypothermia. They present with medical, cognitive and behavioral symptoms; hypothermia <92°F is usually noted on exam. Infection or sepsis, coagulopathy, adrenal insufficiency, and hypothyroidism are often suspected. Bradycardia and cardiac arrhythmia can occur. Laboratory findings include pancytopenia, hyponatremia, elevated blood urea nitrogen and transaminases, all of which normalize when body temperature returns to baseline. In two cases, external warming was episodically successful but not sustained, resulting in recurrent office visits, emergency room visits and multiple medical admissions for investigation and treatment of hypothermia. Subspecialty consultation included neurology, endocrine, hematology, and cardiology. Symptoms remitted when medications known to be associated with hypothermia were discontinued (Atenolol, Abilify).

Discussion: Thermoregulation in typical persons involves afferent sensing, central control and efferent responses. Afferent sensors for cold are located in skin and are triggered by below threshold ambient temperatures. Efferent responses to cold are mediated by cognitively directed behavior and somatosensory mechanisms involving increased sympathetic outflow causing subcutaneous vasoconstriction (alpha 1 adrenergic receptors), increased skeletal muscle tone (shivering) and increased metabolic activity (adrenal secretion of catecholamines; thyroid release of thyroxine). In typical individuals, core temperature is modulated within 0.36 °F through these mechanisms. These mechanisms are not effective in PWS. In a previous study (Hanchett, 1999), persons with PWS were determined to decrease their body temperature by 4-7 °F when challenged by ambient temperatures below 66 °F despite adequate dress and vigorous exercise; all returned to their baseline when ambient temperature normalized, however. In addition to the psychiatric and medical complications of malignant hypothermia, decreased metabolism of medications can also occur contributing to an increased severity of symptoms. Several classes of medication interfere with temperature regulation by inhibiting sympathetic outflow, shivering, and thermogenesis, including opioids, antihypertensives (beta adrenergic antagonists), anesthetics and atypical antipsychotics.

Conclusions: Malignant hypothermia is a life threatening problem seen in PWS. Investigation of an underlying medical cause is essential considering iatrogenic etiologies. All individuals with PWS are at risk for mild hypothermia because of impaired peripheral somatosensory and central thermoregulation, poor judgment and cognitive inflexibility. Aging individuals with PWS are at greater risk for malignant hypothermia. Physicians, families and residential providers must learn about the risks of hypothermia in PWS. Recommendations include greater supervision and behavioral reinforcement for wearing appropriate clothing, early detection by monitoring vital signs in cold weather, and ongoing monitoring of body temperature in those who have already experienced an episode of malignant hypothermia.

Bibliography:

Hanchett JE, Hanchett JM: Hypothermia in PWS, presented at the scientific meeting of the PWSA.USA in San Diego in 1999, and published in the Gathered View in 2008.

Diaz M and Becker D: Thermoregulation: Physiological and Clinical considerations during sedation and anesthesia, *Anesth Prog* 57:25-33, 2010.

Presented at the 25th Annual PWSA (USA) Scientific Meeting in Orlando, FL

November 11, 2011