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Hypothermia and Prader-Willi Syndrome – An Overview

By Barb Dorn, RN, BSN

Hypothermia can be a life threatening condition that some children and adults with Prader-Willi syndrome appear to be more susceptible to than the general population. This condition develops when the body temperature drops too low – to 95 degrees Fahrenheit or lower. Our body attempts to warm itself; but there are situations in which it is not able to do this successfully. When the body temperature drops too low, the heart, nervous system and other organs can't work normally. If left untreated, hypothermia can lead to heart and respiratory failure and in some cases even death.

Research and PWS

Over the years, there have been reports and research studies done on persons with PWS that have provided a mixture of results in supporting the premise that individuals with PWS have a defect in temperature regulation. We know that persons with PWS have abnormalities in their hypothalamus – the area of the brain which is also responsible for temperature regulation. As with many manifestations seen in persons with PWS, some individuals may be at greater risk of having problems while others may not. Wise et al. (1991) described 5 patients with PWS who experienced recurrent hyperthermia in infancy. On the basis of these patients and other reports of abnormal temperature regulation in PWS patients, particularly hypothermia with exposure to cold, they concluded that defects in temperature regulation may be a manifestation of hypothalamic dysfunction in PWS. Cassidy and McKillop (1991) concluded on the basis of surveys that clinically significant abnormal temperature control is not a common finding in this disorder. Similarly, Williams et al. (1994) concluded on the basis of surveys that the prevalence of febrile convulsions, fever-associated symptoms, and temperature less than 94 degrees Fahrenheit were not unique to PWS but can occur in any neuro-developmentally handicapped individual and do not necessarily reflect syndrome-specific hypothalamic abnormalities. DiMario et al (1994) found that patients with PWS had a detectable underlying autonomic dysfunction in their autonmic nervous system with a diminished parasympathetic nervous system activity. In 1999, Dr's James and Jeanne Hanchet from the Children's Institute presented case studies at the National PWSA | USA conference on 2 adults with PWS who were diagnosed and treated for hypothermia while in their care. DiMario and Burleson (2002) did not find differences in central regulation and cutaneous temperature flow in regulation in their patients with PWS compared to their control subjects. In 2013, Cherpes et al also shared findings at the National PWSA | USA conference on the "Effects of Atypical Antipsychotic Medications on Body Temperature in Patients with PWS." They did not find these medications to have significant impact on body temperature. However, there have been reports of persons with PWS of dying of hypothermia. More research is needed.

Factors Contributing to Hypothermia	**Symptoms of Hypothermia**
• Age – Infants and older persons are at greater risk • Exposure to cold temperatures • Inadequate heating in home or workplace – conditions do not have to be extreme. • Inappropriate dress – not wearing adequate warm clothing. • Falling in cold water • Medications – Opioids (pain medications), certain anti-hypertensive medications, anesthetics, some atypical antipsychotics and antidepressants. • Certain medical conditions – stroke, spinal cord injuries, hypothyroidism, severe arthritis, Parkinson's disease, dehydration, other nerve disorders.	• Early/Mild Hypothermia: • Shivering • Dizziness • Nausea • Increased breathing and heart rate • Slurred speech; difficulty talking • Slight confusion or lack of coordination • Fatigue • Cold, pale or blue gray skin • Moderate/Severe Hypothermia • Shivering - decreases and stops as progresses • Weak pulse, slow respirations • Increased confusion • Drowsiness – progresses to loss of consciousness • **Infants** – may appear limp, weak cry, cold, red skin

Prevention

Many children and adults with PWS may have factors that make them more susciptible to hypothermia. There are some steps that parents and caregivers can take to prevent and monitor for this condition.

Know the person with PWS' baseline temperature. Check the person's temperature daily for 2-4 weeks to determine his/her average temperature. In many cases it will be below 98.6 F – the average temperature.

When the outside temperature is low - make sure the person with PWS wears appropriate clothing and/or keep the person with PWS indoors or minimize time outdoors.

Keep the thermostat in the home at a reasonably warmer temperature. As the person with PWS ages, this temperature may need to be higher.

Don't let the person sleep in a cold room – this is especially true for infants and older adults with PWS.

Apply extra blankets, flannel sheets as needed for sleeping. **Dress in warm pajamas and/or blanket sleepers**.

What to Do if Severe Hypothermia is Suspected

Hypothermia is an emergency condition that can quickly lead to unconsciousness and death if left untreated. If you suspect that a child or adult with PWS has severe hypothermia, immediately WARM the person with blankets. If wearing wet clothes, remove and dress in warm clothing. Take to the nearest Emergency Department for evaluation. All cases must be investigated to rule out illness, brain tumor or other brain abnormality and/or medication reaction. In most cases, there is no specific treatment unless an illness, brain abnormality or medication have been found to be the cause. There is no medication or treatment that will raise the body's temperature other than those measures listed above. There have been documented cases of severe hypothermia in infants, children and adults with PWS. Awareness of this issues will allow parents and caregivers to take steps to monitor and prevent it and in severe cases – treat it.

References:

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