PRADER-WILLI SYNDROME ASSOCIATION

Winter Warning on Hypothermia in PWS

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Due to a couple of crises recent situations involving significantly low body temperatures (hypothermia) in people with PWS, I thought it was time to remind parents and caregivers that there is a potential in PWS for this life-threatening problem. This is most likely going to happen in cold-weather states, and typically to someone middle-aged or older with the syndrome, but we recently had a report of a 17-year-old with severe hypothermia.

One big question not answered is how low can a person's temperature go without being life-threatening? There is not a clear answer but we've had a couple of situations a year where the person's temperature went down to 87 – 88°F. Dr. Jeannie Hanchett, along with the late Dr. Jim Hanchett wrote the first paper on hypothermia in PWS and they said that with PWS you have to look at other clinical signs such as: Is the person acting dizzy and/or disoriented? Unusually sleepy? Not eating or acting normally? Those with temperatures below 90°F have been known to go into an almost coma-like state.

Dr. Hanchett states that if a person with PWS has a temperature below 94°F the main advice is to WARM THAT PERSON UP! During cold weather if the person with PWS is known to have episodes of significantly low body temperature, use extra care to bundle the person up. If the temperature goes below 94°, put the person in warm place such as a bed with several blankets or an electric blanket. In some cases an emergency room visit or hospitalization is necessary. Even with hospitalizations, it is sometimes hard to stabilize the temperature.

"Several classes of medication can interfere with temperature regulation by inhibiting sympathetic outflow, shivering, and thermogenesis, including opioids, antihypertensives (beta adrenergic antagonists), anesthetics and atypical antipsychotics" (*Phenomenology of Malignant Hypothermia in PWS* by **Janice Forster, Francie McDougall, Jacqueline Durette and Linda M. Gourash). Hypothermia is listed as a potential side effect of some drugs, and this must be considered by the doctor. In a recent chart review study of PWS and atypical antipsychotics, a significant impact was not found on the mean body temperatures of pediatric and adult patients with PWS who were taking this type of medication at the time of admission to The Children's Institute. Temperatures less than or equal to 95° F on admission were seen in approximately 1.4% of the entire sample population. (Gregory Cherpes, Jennifer Padden Elliott, et al. - See 2013 scientific conference abstracts). If you are concerned about a case of hypothermia, please notify PWSA (USA) and we will send you more detailed articles on this topic.**