



Prader-Willi Syndrome Association (USA)

MEDICAL ALERT

Important Considerations For Routine Or Emergency Treatment

Medical professionals can contact PWSA(USA) to obtain more information and put you in touch with a specialist as needed.

Anesthesia, medication reactions

People with PWS may have unusual reactions to standard dosages of medications and anesthetic agents. Use extreme caution in giving medications that may cause sedation: prolonged and exaggerated responses have been reported.

Anesthesia - <http://www.pwsausa.org/research/anesthesia.htm>

Adverse reactions to some medications

People with PWS may have unusual reactions to standard dosages of medications. Use extreme caution in giving medications that may cause sedation: prolonged and exaggerated responses have been reported. Water intoxication has occurred in relation to use of certain medications with antidiuretic effects, as well as from excess fluid intake alone.

Water intoxication - http://www.pwsausa.org/support/water_intoxication_alert.htm

High pain threshold

Lack of typical pain signals is common and may mask the presence of infection or injury. Someone with PWS may not complain of pain until infection is severe or may have difficulty localizing pain. Parent/caregiver reports of subtle changes in condition or behavior should be investigated for medical cause.

Respiratory concerns

Individuals with PWS may be at increased risk for respiratory difficulties. Hypotonia, weak chest muscles, and sleep apnea are among possible complicating factors. Anyone with significant snoring, regardless of age, should have a medical evaluation to look for obstructive sleep apnea.

Recommendations for Evaluation of Breathing Abnormalities Associated with Sleep in Prader Willi Syndrome - <http://www.pwsausa.org/syndrome/RecEvalSleepApnea.htm>

Respiratory - <http://www.pwsausa.org/syndrome/respiratoryproblems.htm>

Lack of vomiting

Vomiting rarely occurs in those with PWS. Emetics may be ineffective, and repeated doses may cause toxicity. This characteristic is of particular concern in light of hyperphagia and the possible ingestion of uncooked, spoiled, or otherwise unhealthful food items. The presence of vomiting may signal a life-threatening illness.

Severe gastric illness

Abdominal distention or bloating, pain, and/or vomiting may be signs of life-threatening gastric inflammation or necrosis, more common in PWS than in the general population. Rather than localized pain, there may be a general feeling of unwellness. If an individual with PWS has these symptoms, close observation is needed. An X-ray and an endoscopy with biopsy may be necessary to determine degree of the problem and possible need for emergency surgery.

Acute Gastrointestinal - http://www.pwsausa.org/syndrome/acute_gastrointestinal_episodes.htm

Medical Alert Stomach - http://www.pwsausa.org/syndrome/medical_alert_Stomach.htm

Another consideration is gastroparesis, a weakness of the stomach. This is a condition that is common with PWS and can be more life threatening than in a typical situation. A child with Prader-Willi syndrome when diagnosed with Gastroparesis may need hospitalization. Eating while the stomach is distended with gastroparesis can be very dangerous. For more information go to: <http://www.gicare.com/pated/ecdgs45.htm>

Body temperature abnormalities

Idiopathic hyper- and hypothermia have been reported. Hyperthermia may occur during minor illness and in procedures requiring anesthesia. Fever may be absent despite serious infection.

Skin lesions and bruises

Because of a habit that is common in PWS, open sores caused by skin picking may be apparent. Individuals with PWS also tend to bruise easily. Appearance of such wounds and bruises may wrongly lead to suspicion of physical abuse.

Hyperphagia (excessive appetite)

Insatiable appetite may lead to life-threatening weight gain, which can be very rapid and occur even on a low-calorie diet. Individuals with PWS must be supervised at all times in all settings where food is accessible. Those who have normal weight have achieved this because of strict external control of their diet and food intake.

Surgical and Orthopedic Concerns

In view of the increasing number of infants and children with PWS undergoing sleep assessments prior to growth hormone treatment and the potential rise in surgical procedures (e.g., tonsillectomy) requiring intubation and anesthesia, it will be important to alert the medical team about complications. These complications may include trauma to the airway, oropharynx, or lungs due to possible anatomic and physiologic differences seen in PWS such as a narrow airway, underdevelopment of the larynx and trachea, hypotonia, edema, and scoliosis.'

Musculoskeletal manifestations, including scoliosis, hip dysplasia, fractured bones and lower limb alignment abnormalities, are described in the orthopedic literature. However, care of this patient population from the orthopedic surgeon's perspective is complicated by other clinical manifestations of PWS.

Prader-Willi Syndrome: Clinical Concerns for the Orthopedic Surgeon -

<http://www.pwsausa.org/syndrome/Orthopedic.htm>

IN THE EVENT OF DEATH

Reporting of Deaths

The Prader-Willi Syndrome Association (USA) has created a research database of reported deaths of individuals with PWS. Although most premature deaths are attributable to morbid obesity, cases unrelated to obesity have recently been noted, leading PWSA to begin a formal investigation of causes of death.

PWSA also provides bereavement support to families who have lost children with PWS. In the event of death of someone with PWS, please contact PWSA (USA): 1-800-926-4797.

Organ Donation for Research

When a child or adult with PWS dies, the family may wish to consider donation of organs for research. PWSA (USA) has established a procedure for such donations to the Brain and Tissue Banks.

<http://www.pwsausa.org/research/B&TBank.htm> for more information.