

MEDICAL ALERT

Important Considerations For Routine Or Emergency Treatment

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. Carefully monitor respiratory function.

Water intoxication has occurred in relation to use of certain medications with antidiuretic effects as well as from excess fluid intake alone.

High pain threshold

Lack of pain 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. Infants in particular may be at risk for respiratory failure due to illness. Laboratory evaluation of respiratory function may be necessary.

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 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.

Body temperature abnormalities

Idiopathic hyper- and hypothermia have been reported. Hyperthermia may occur during minor illness and in pro-

cedures 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.

Edema

Individuals with PWS may have unrecognized edema. Careful clinical evaluation and diuretics may be necessary.

PWSA (USA) BACKUP PHONE NUMBERS

In the event of a serious medical emergency when immediate advice is needed from a medical expert, physicians and ER nurses may call our 24 hour crisis line at 1-800-926-4797 .

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 for Developmental Disorders, created by NIH in cooperation with the University of Maryland and the University of Miami. Prompt action is essential for tissue preservation. Families are advised to contact the closest Brain and Tissue Bank directly: 1-800-847-1539 (Maryland) or 1-800-59-BRAIN (Florida).