

MEDICAL ALERT

Health Care Providers in an Inpatient Setting

PLEASE READ THE FOLLOW IMPORTANT HEALTH INFORMATION:

It has come to our attention that you are caring for a person who has Prader-Willi syndrome (PWS). Persons with PWS have some unique health needs that require close attention. The following are just some of these more critical concerns that you, as his/her health care provider need to monitor and keep in mind. NOTE: The health issues and dietary needs of an infant with PWS are very different. For more information, visit our website at www.pwsausa.org or call toll free: 1-800-926-4797.

COGNITIVE LIMITATIONS: Approximately 90% of persons with PWS have cognitive limitations. Many have guardians who assist with medical decisions. Simple, understandable language needs to be used to educate and obtain informed consent.

HYPERPHAGIA – Because of an abnormality in the hypothalamus, persons with PWS have an insatiable appetite – the feeling of fullness never reaches their brain. Many will go to extreme measures to obtain food – begging, crying, stealing and gorging. Because of hormone deficiencies, they can gain weight on half the calories of others their age. All must be on a calorie restricted diet. ALL FOOD SOURCES MUST BE MONITORED AND CONTROLLED. PERSONS WITH PWS HAVE HAD BINGE EATING EPISODES RESULTING IN STOMACH RUPTURE. This has occurred while in a hospital setting.

Place patient on a calorie restricted diet. Have a nutritional consultation. Providing small more frequent meals 5-6 times a day is often helpful.

If possible, the patient with PWS should be in a private room. If they have a roommate, he/she may steal their food. Ask the roommate to remove food items that may be visible..

Make sure there is constant monitoring of food carts that are delivering food to patients. They can

quickly take a tray while staff is not looking or delivering a tray.

Promptly remove food trays that are collected. Watch for food items that may be left out. They will eat from any plates where food is remaining.

Limit access to the nursing unit kitchen. Keep a supply of diet Jell-O, diet soda and other low calorie food items if additional food may be needed for them.

Limit the quantity of fluids consumed. They can and will drink enormous amounts of diet soda, diet Kool Aid ... and suffer from water intoxication. Most do not drink or care for water.

Make one staff person responsible for giving this patient ALL food. They are clever at getting different people to get them food. Keep lines of communication open with family members, care providers and visitors to keep informed of food the person has already consumed.

Check weight daily. Believe what you see and tighten your vigilance!

If the patient goes off of the unit – they must have 1:1 supervision. They can often get money to access vending machines, gift shops and cafeterias.

Keep break room doors closed – especially if food is present.

SEVERE GASTRIC Distention with Ischemia- This is a life threatening situation that may also result in the rupturing of the stomach. The person with PWS may present with abdominal distention, pain and/or vomiting but may only complain of mild abdominal discomfort.. Because of an altered pain response and the rarity of vomiting seen in these individuals, the presence of pain and/or vomiting must be evaluated closely. This gastric inflammation with necrosis has most often been seen in persons with PWS who have had a recent

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binge episode and whose weight is under control. Many persons also have gastroparesis, which can become dangerous if overeating occurs.

ADVERSE REACTIONS TO ANESTHESIA AND SOME MEDICATIONS: People with PWS have unusual reactions to standard doses of medications and anesthetic agents. Use extreme caution in giving medication that may cause sedation. Prolonged exaggerated responses including respiratory arrest have been reported. Psychotropic medications must always be started at very low doses. Medications that have anti-diuretic effects may cause water intoxication.

Monitor postoperative patients very closely. You may want to keep a pulse oximetry monitor in place for 24 hours.

Be very conservative in administering pain medications.

Closely monitor all liquids the patient is getting – both intravenously and orally.

ALTERED PAIN RESPONSE: Persons with PWS have a very high pain threshold which may mask injury, infection and illness. Do not use pain as a primary means of diagnosis. Some may not complain of pain until the infection or injury has become very severe. Use extreme caution when prescribing and administering pain or other narcotic medications.

RESPIRATORY CONCERNs: Individuals with PWS are often at risk for respiratory difficulties. Sleep apnea is common. Hypotonia, especially in the

trunk area put them at high risk for developing pneumonia and atelectasis. Obese patients who have been chronically hypoxic may not tolerate fully corrective use of Oxygen and are likely to start retaining CO₂

LACK OF VOMITING: Vomiting rarely occurs in persons with PWS. If it occurs, severe gastric illness must be ruled out. In cases of poisoning and ingestion of spoiled or contaminated food, emetics are often ineffective. Toxicity can occur if repeated doses of emetics are attempted.

BODY TEMPERATURE ABNORMALITIES: Because of hypothalamic malfunction, idiopathic hypo and hyperthermia have been reported. Hyperthermia may occur during minor illness and in procedures requiring anesthesia. Fever may be absent in cases of severe infection.

SKIN LESIONS – PICKING: It is common to see open sores caused by skin picking. Bruising is also seen. Appearance of such wounds and bruises may wrongly lead to suspicion of abuse. Persons hospitalized for cellulitis and/or wound infections require close monitoring and supervision. Extra measures to cover incisions, IV sites and/or wounds should be used. Steps to keep hands occupied are often helpful.

If you have any questions or concerns, please contact PWSA (USA). We have physicians and other health care professionals who are willing to provide consultative services. This service is professional to professional only. Phone numbers of professionals will not be shared with family members or care provider staff. Call toll free: 1-800-926-4797 for this service to be arranged.