

Medical Alert for Patients with Prader-Willi Syndrome

Information for Health Care Professionals in an Inpatient Setting

People with Prader-Willi syndrome (PWS) have unique health needs that require close attention. Contact PWSA | USA to speak to a physician, nurse, or other professional. Help is available 24/7. Call (941) 312-0400 or visit our website at www.pwsausa.org. This is a professional-to-professional service. To access the full [Medical Alerts Booklet](#), visit this link:

The following highlights some critical health concerns.

Cognitive Limitations: There are varying degrees of intellectual abilities. Due to communication challenges, self-reporting of health concerns may be difficult, inaccurate, or exaggerated. Many have guardians to assist with medical decisions.

Hyperphagia: Due to an abnormality in the hypothalamus, persons with PWS have an insatiable appetite. Many go to extreme measures to obtain food – begging, crying, stealing, and gorging. They tend to gain weight easily, and most must be on a reduced-calorie diet. ALL FOOD SOURCES MUST BE MONITORED AND CONTROLLED. One binge can result in death. Things you can do to help:

- DO NOT offer food without consulting parent or caregiver
- Place on a calorie restricted diet, discuss with parent or caregivers for calorie count
- NO access to ordering food
- Provide 3 healthy meals and 2 healthy snacks a day
- A private room is preferred to prevent access to a roommate's food
- Ensure no unauthorized access to food; food carts, other rooms, staff area, vending and coffee machines
- Those with a strong food drive or high behavioral needs, may require a bed alarm or 1:1 sitter
- Provide activities – watching TV, word search activities and other activities. Obtain an OT consultation.

Severe Gastric Illness: The person with PWS may present with abdominal distention, “not feeling well”, mild pain, diarrhea, and/or vomiting. **It is rare for a person with PWS to vomit.** This may indicate a gastric emergency. Some have experienced severe gastroparesis and ileus resulting in ischemia and/or a ruptured stomach. Follow the [GI Algorithm](#) or on the UpToDate website.

Altered Pain Response: Most have a very high pain threshold which may mask injury, infection, and illness. Many are unable to accurately localize pain.

Respiratory Concerns: Persons with PWS have increased risk for respiratory difficulties. Sleep apnea is common. Hypotonia and a weak cough put them at high risk for pneumonia and atelectasis.

Mental Health: Many people with PWS face challenges with behavior and co-existing mental health issues. Anxiety and emotional outbursts are common, especially with change and food-related issues. Work with family and caregivers for strategies that may help to decrease anxiety and behavior.

Swelling/Edema: Some individuals with PWS may have unrecognized edema or swelling.

Adverse Reactions to Anesthesia and Some Medications: Many people with PWS have unusual reactions to anesthesia and other medications. Drugs that cause sedation may have exaggerated responses. Antidiarrheal medications cause excessive slowing of intestines. Anti-diuretics have caused low serum sodium.

Body Temperature Abnormalities: Because of a hypothalamic malfunction, idiopathic hypo and hyperthermia have been reported. **Fever may be absent during serious illness.**

Skin Lesions, Picking and Bruises: Skin picking is common in persons with PWS. Findings are often mistaken as a sign of abuse. Persons hospitalized for surgery, cellulitis and/or wound infections require close monitoring and supervision. **For further details, please visit our website www.pwsausa.org to review the Medical Alerts Booklet or Medical Issues A-Z.**