PRADER-WILLI SYNDROME MEDICAL ALERTS

(800) 926-4797 • (941) 312-0400
www.pwsausa.org

SAVING AND TRANSFORMING LIVES!

REVISED 2015
PRADER-WILLI SYNDROME

Prader-Willi syndrome (PWS) is a complex neurobehavioral genetic disorder resulting from abnormality on the 15th chromosome. It occurs in males and females equally and in all races. Prevalence estimates range from 1:12,000 to 1:15,000. Incidence in newborns is unknown.

PWS typically causes low muscle tone, short stature if not treated with growth hormone, cognitive deficits, incomplete sexual development, problem behaviors, and a chronic feeling of hunger that, coupled with a metabolism that utilizes drastically fewer calories than normal, can lead to excessive eating and life-threatening obesity.

It is felt to be a multistage disorder with decreased fetal movement prenatally and low birth weight. Infants have failure to thrive due to feeding problems and hypotonia. Toddlers have increased weight gain, then hyperphagia and obesity as they get older, if calories are not restricted. Most of the medical problems in Prader-Willi syndrome are related to the obesity, hypotonia, and hypothalamic dysfunction.

Some of the other factors that may cause difficulties include adverse reactions to medications, high pain tolerance, gastrointestinal and respiratory issues, lack of vomiting, and unstable temperature. Adrenal insufficiency may also occur.

Severe medical complications can develop rapidly in individuals with PWS.

Members of the Clinical Advisory Board are available for consultation with physicians through the Prader-Willi Syndrome Association (USA).
Prader-Willi Syndrome
Medical Alerts
by
Medical Specialists in
Prader-Willi Syndrome
Mission
Prader-Willi Syndrome Association (USA) is an organization of families and professionals working together to raise awareness, offer support, provide education and advocacy, and promote and fund research to enhance the quality of life of those affected by Prader-Willi syndrome.

Table of Contents
Foreward.........................................................inside front cover
MEDICAL ALERT Important Considerations ......................... 5
Anesthesia Reactions...................................................... 5
Adverse Reactions to Some Medications ......................... 6
Water Intoxication.......................................................... 6
High Pain Threshold.......................................................... 6
Central Adrenal Insufficiency in Individuals with
Prader-Willi Syndrome..................................................... 7
Body Temperature Abnormalities................................. 7
Skin Lesions and Bruises.................................................. 8
Hyperphagia (Excessive Appetite)................................. 8
Lack of Vomiting............................................................ 8
Swallowing and Choking.................................................. 9
Table of Contents, continued...

Respiratory Concerns ................................................................. 9
  Recommendations for Evaluation of Breathing
    Abnormalities Associated with Sleep in Prader-Willi Syndrome ......................................................... 10
  Growth Hormone Treatment Consensus Statements.... 15

In-Patient Considerations ..................................................... 15
  Anesthesia............................................................................. 15
  Surgical and Orthopedic Concerns ..................................... 18
  Postoperative Monitoring.................................................... 19

Severe Gastric Intestinal Concerns........................................ 22
  Lack of Vomiting................................................................... 22
  Severe Gastric Illness............................................................... 22
  Constipation.......................................................................... 23
  Risk of Stomach Necrosis and
    Rupture / Warning Signs.................................................... 25-26

IN THE EVENT OF DEATH............................................................... 26
  Reporting of Deaths............................................................... 27
  Organ Donation for Research............................................... 27

Life-Saving GI Evaluation Algorithm Chart .......................
  .................................................................Foldout page in back of publication
MEDICAL ALERT
Important Considerations for Routine or Emergency Treatment

Obesity and its related complications is the major cause of morbidity and mortality in Prader-Willi syndrome. Keeping the individual at a healthy weight will minimize these complications. There are problems unique to Prader-Willi syndrome and they will be discussed in this Alert.

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

Anesthesia Reactions
People with PWS may have unusual reactions to standard dosages of anesthetic agents. Use caution in giving anesthesia. See page 15 or go to the website at http://www.pwsausa.org and view the Medical section for articles on Anesthesia.

• Anesthesia and Prader-Willi Syndrome: James Loker, M.D., Laurence Rosenfield, M.D.
• Anesthesia Concerns for Patients with PWS: Winthrop University
Adverse Reactions to Some Medications
People with PWS may have unusual reactions to standard dosages of medications. Use extreme caution in giving medications, especially those that may cause sedation; prolonged and exaggerated responses have been reported.

Water Intoxication
Water intoxication has occurred in relation to use of certain medications with antidiuretic effects, as well as from excess fluid intake alone. Anti diarrheal medications may cause severe colonic distension, necrosis and rupture and should be avoided. http://www.pwsausa.org and view Medical section under Water Intoxication.

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. Any complaint of pain by a person with PWS should be taken seriously.
Central Adrenal Insufficiency in Individuals with Prader-Willi Syndrome

Individuals with Prader-Willi syndrome may be at risk for central adrenal insufficiency (CAI). The presence or absence of CAI cannot be determined by ONLY measuring an 8 a.m. cortisol level - the individual must be tested while stressed (e.g., with febrile illness) or using a stimulation test.

http://www.pwsausa.org and view Medical section under Adrenal Insufficiency.

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. All individuals with PWS are at risk for mild hypothermia because of impaired peripheral somatosensory and central thermoregulation, poor judgment and cognitive inflexibility. Malignant hypothermia is a life-threatening problem occasionally seen in PWS.

http://www.pwsausa.org and view Medical section under Temperature.
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 erroneously lead to suspicion of physical abuse. These lesions can cause serious life-threatening infections. There are possible effective treatments for picking.

http://www.pwsausa.org and view Medical section under Skin Picking.

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.

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 and may warrant immediate treatment.
Swallowing and Choking
Persons with PWS are highly likely to have an undetected swallowing problem that places them at risk for asphyxiation of a food bolus (choking), and they require a specific type of swallowing evaluation. A clinical or bedside evaluation is not sufficient to detect dysphagia in this population. For entire article go to: http://www.pwsausa.org and view Medical section under Choking/Swallowing.

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.

Other problems that can cause respiratory difficulties in the young can be chronic stomach reflux and aspiration. Although the lack of vomiting is felt to be prominent in PWS, reflux has been documented and should be investigated in young children with chronic respiratory problems. Individuals with obstructive apnea are at more risk for reflux as well.

Respiratory Problems in Prader-Willi Syndrome:
James Loker, M.D. http://www.pwsausa.org and view Medical section under Respiratory.
Recommendations for Evaluation of Breathing Abnormalities Associated with Sleep in Prader-Willi Syndrome

PWSA (USA) Clinical Advisory Board Consensus Statement - 12/2003

Problems with sleep and sleep disordered breathing have been long known to affect individuals with Prader-Willi syndrome (PWS). The problems have been frequently diagnosed as sleep apnea (obstructive [OSA], central or mixed) or hypoventilation with hypoxia. Disturbances in sleep architecture (delayed sleep onset, frequent arousals and increased time of wakefulness after sleep onset) are also frequently common. Although prior studies have shown that many patients with PWS have relatively mild abnormalities in ventilation during sleep, it has been known for some time that certain individuals may experience severe obstructive events that may be unpredictable.

Factors that seem to increase the risk of sleep disordered breathing include young age, severe hypotonia, narrow airway, morbid obesity and prior respiratory problems requiring intervention such as respiratory failure, reactive airway disease and hypoventilation with hypoxia. Due to a few recent fatalities reported in individuals with PWS who were on growth hormone therapy (GH), some physicians have also added this as an additional risk.
One possibility (that is currently unproven) is that GH could increase the growth of lymphoid tissue in the airway thus worsening already existing hypoventilation or OSA. Nonetheless, it must be emphasized that there is currently no definitive data demonstrating that GH causes or worsens sleep disordered breathing. However, to address this new concern, as well as the historically well documented increased risk of sleep-related breathing abnormalities in PWS, the Clinical Advisory Board of the PWSA (USA) makes the following recommendations:

1. **A sleep study or a polysomnogram** that includes measurement of oxygen saturation and carbon dioxide for evaluation of hypoventilation, upper airway obstruction, obstructive sleep apnea and central apnea should be contemplated for all individuals with Prader-Willi syndrome. These studies should include sleep staging and be evaluated by experts with sufficient expertise for the age of the patient being studied.

2. **Risk factors that should be considered to expedite the scheduling of a sleep study should include:**
   - Severe obesity - weight over 200% of ideal body weight (IBW).
   - History of chronic respiratory infections or reactive airway disease (asthma).
• History of snoring, sleep apnea or frequent awakenings from sleep.
• History of excessive daytime sleepiness, especially if this is getting worse.
• Before major surgery including tonsillectomy and adenoidectomy.
• Prior to sedation for procedures, imaging scans and dental work.
• Prior to starting growth hormone or if currently receiving growth hormone therapy.

Additional sleep studies should be considered if patients have the onset of one of these risk factors, especially a sudden increase in weight or change in exercise tolerance. **If a patient is being treated with growth hormone, it is not necessary to stop the growth hormone before obtaining a sleep study unless there has been a new onset of significant respiratory problems.**

Any abnormalities in sleep studies should be discussed with the ordering physician and a pulmonary specialist knowledgeable about treating sleep disturbances to ensure that a detailed plan for treatment and management is made. Referral to a pediatric or adult pulmonologist with experience in treating sleep apnea is strongly encouraged for management of the respiratory care.
In addition to a calorically restricted diet to ensure weight loss or maintenance of an appropriate weight, a management plan may include modalities such as:

- Supplemental oxygen
- Continuous positive airway pressure (CPAP) or BiPAP
- Oxygen should be used with care as some individuals may have hypoxemia as their only ventilatory drive and oxygen therapy may actually worsen their breathing at night.
- Behavior training is sometimes needed to gain acceptance of CPAP or BiPAP.
- Medications to treat behavior may be required to ensure adherence to the treatment plan.

If sleep studies are abnormal in the morbidly obese child or adult (IBW > 200%) the primary problem of weight should be addressed with an intensive intervention - specifically, an increase in exercise and dietary restriction. Both are far preferable to surgical interventions of all kinds. Techniques for achieving this are available from clinics and centers that provide care for PWS and from the national parent support organization [PWSA (USA)]. Behavioral problems interfering with diet and exercise may need to be addressed simultaneously by persons experienced with PWS.
If airway related surgery is considered, the treating surgeon and anesthesiologist should be knowledgeable about the unique pre- and postoperative problems found in individuals affected by Prader-Willi syndrome.

Tracheostomy surgery and management presents unique problems for people with PWS and should be avoided in all but the most extreme cases. Tracheostomy is typically not warranted in the compromised, morbidly obese individual because the fundamental defect is virtually always hypoventilation, not obstruction. Self-endangerment and injury to the site are common in individuals with PWS who have tracheostomies placed.

At this time there is no direct evidence of a causative link between growth hormone and the respiratory problems seen in PWS. Growth hormone has been shown to have many beneficial effects in most individuals with PWS including improvement in the respiratory system. Decisions in the management of abnormal sleep studies should include a risk/benefit ratio of growth hormone therapy. It may be reassuring for the family and the treating physician to obtain a sleep study prior to the initiation of growth hormone therapy and after 6-8 weeks of therapy to assess the difference that growth hormone therapy may make. A followup study after one year of treatment with growth hormone may also be indicated.
Growth Hormone Treatment and Prader-Willi Syndrome

PWSA (USA) Clinical Advisory Board Consensus Statement - 6/2009
PWSA International Consensus Statement 2013

Both statements are found at http://www.pwsausa.org and view Medical section under Growth Hormone.

In-Patient Considerations

Anesthesia and Prader-Willi Syndrome

In individuals with Prader-Willi syndrome there are health issues that can alter the course of anesthesia. The majority of problems does not come from general anesthesia but from conscious sedation if it is not well monitored.

Obesity

• Consideration for obstructive apnea, pulmonary hypertension, diabetes, and right heart failure should be addressed.

Narcotics

• Individuals may have an exaggerated response to narcotics and the lowest possible dose to achieve the desired state of anesthesia should be used.
Pulmonary embolism
• Individuals with PWS are at risk for pulmonary embolism. DVT prophylaxis should be considered in all obese individuals.

High pain threshold
• Unexplained tachypnea or tachycardia may be only indication of pain.

Individuals with PWS may not respond to pain in the same manner as others. While this may be helpful in postoperative management, it may also mask underlying problems. Pain is the body’s way of alerting us to problems. After surgery, pain that is out of proportion to the procedure may alert the physician that something else is wrong. Since pain may not be present, other possible signs of underlying problems should be monitored.

Temperature instability
• Leads to hypo- or hyperthermia. There is no known predisposition to malignant hyperthermia, but depolarizing muscle relaxants should be avoided if possible.

Thick saliva
• Can complicate airway management especially during conscious sedation and increase the risk of caries.
Food seeking behaviors
• Assume individual has eaten unless verified by caregiver.

Hypotonia
• May complicate ability to cough effectively and clear airways

Skin picking
• May complicate healing of IV sites and incisional wounds. Restraints or gloves may be necessary to protect wounds during healing.

Difficult access
• Obesity and poor muscle tone may complicate line placement. Obesity may also complicate ability to intubate.

Behavior problems
• Individuals are more prone to emotional outbursts, obsessive-compulsive behaviors, and psychosis. Psychotropic medications may affect anesthesia.

Hypothyroidism
• Risk of hypothyroidism is 20-30% and may be unknown prior to surgery.

Growth hormone efficiency
• All individuals should be considered to be GH deficient.
Central adrenal insufficiency

• Several studies have shown CAI in individuals with PWS. Stress dose of cortisol may be indicated unless measurement of stress cortisol levels has been done.

• Outpatient procedures and general sedation may be especially problematic. Care must be taken during procedures done in out of hospital settings that proper equipment for resuscitation is immediately available and consideration for doing these in the OR should be discussed. Procedures where more than light sedation is used may warrant an overnight observation.

To view this article in its entirety, see: http://www.pwsausa.org and view Medical section for articles on Anesthesia.

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. [http://www.pwsausa.org](http://www.pwsausa.org) and view Medical section under Orthopedic Issues.

**Postoperative Monitoring of Patients with Prader-Willi Syndrome**

Patients with PWS are known to have increased morbidity after surgery due to:

- Abnormal physiological response to hypercapnia and hypoxia
- Hypotonia
- Narrow oropharyngeal space
- High incidence of central, obstructive and mixed apnea
- Thick secretions
- Obesity
• Increased incidence of scoliosis with decreased pulmonary function
• Prolonged exaggerated response to sedatives
• Increased risk for aspiration
• Decreased pain sensation
• Possible challenges with compliance to pre- and postoperative treatment procedures due to:
  – Extreme food seeking behavior and hyperphagia due to hypothalamic dysfunction.
  – High incidence of gastroparesis and slow motility of the intestinal tract.
  – Extreme skin picking which may interfere with wound healing.
  – Altered temperature regulation – fever may be absent in the presence of infection. There does not seem to be a higher incidence of malignant hyperthermia.
  – The possibility of central adrenal insufficiency.

**RECOMMENDATIONS:**
• Patients with PWS who undergo deep sedation and general anesthesia should be recovered overnight in a monitored unit.
• Infants and children may require intensive care monitoring.
• Continuous monitoring of pulse-oximetry for 24 hours postoperative with attention to airway and breathing.

• A conservative approach to pain management and use of narcotic agents.

• Full assessment of return of GI motility prior to initiation of intake by mouth because of the predisposition to ileus after surgery.

• Scheduling procedure as early in the day as possible to prevent prolonged time period where food seeking could take place.

• Direct supervision (1:1) to prevent foraging postoperatively.

• Monitor for picking at wounds and/or incisions. May require additional dressings and other barriers to prevent access to wound.

• Close observation of wound for signs of infection.

• Utilization of respiratory therapy interventions to prevent atelectasis and/or postoperative lung infection.

• Due to the hypotonia and obesity, individuals with PWS are at risk for deep venous thrombi (DVT) and pulmonary embolism. Patients should be under the guidelines for DVT prophylaxis.

http://www.pwsausa.org and view Medical section under Postoperative Monitoring of Patients with Prader-Willi Syndrome.
Severe Gastric Intestinal Concerns

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 and may warrant immediate treatment.

Severe Gastric Illness
Gastric problems are very common in PWS with the majority of individuals having some degree of decreased motility and gastroparesis. Abdominal distension or bloating, pain and/or vomiting may be signs of life-threatening gastric dilation, inflammation or necrosis. Rather than localized pain, there may be a general feeling of unwellness. Any individual with PWS with these symptoms needs immediate medical attention. An X-ray, CT scan or ultrasound can help with the diagnosis and confirm if there is gastric necrosis and/or perforation. If distension is noted, these individuals need close monitoring, made NPO and may need decompression with an NG tube.
Gastric necrosis or perforation is a medical emergency requiring exploratory laparotomy or emergent surgery. Individuals with PWS may not have tenderness, rigidity or rebound normally associated with an acute abdomen.

In addition to gastric distension, colonic impaction may also be present and need to be addressed. Stomach pain can also be due to gallstones or pancreatitis. An ultrasound, chemistry analysis of the blood and CT of the abdomen will help with the diagnosis.

**Constipation in Individuals with Prader-Willi Syndrome**

James Loker, M.D., Pediatric Cardiologist
Ann Scheimann, M.D., M.B.A., Gastroenterologist
PWSA (USA) Clinical Advisory Board Members

Constipation is a common problem in individuals with Prader-Willi syndrome (PWS). It takes longer for food to move through the GI system in Prader-Willi syndrome*. This slower passage of food can lead to serious issues similar to the ones seen related to the stomach. Outpatient methods used to clear constipation in non-PWS patients may be ineffective due to poor fluid intake and hypotonia. Inpatient regimens frequently use large volumes of fluid which
may cause problems. Reliance on these methods may lead to life-threatening conditions such as necrosis and perforation of the colon and subsequent sepsis. Due to decreased muscle tone and altered pain response, individuals with PWS may not have the same clinical exam that a non-PWS patient would have. A heavier reliance on imaging may be necessary. Individuals with PWS may be at higher risk for impaction. Rectal examination and enema may be required in addition to oral cleanout regimen. This may also be problematic in some leading to rectal picking.

Patients with PWS having constipation and receiving repeated regimens of oral PEG (polyethylene glycol) solution for bowel cleansing should be monitored closely for abdominal distention and retention. Failure of standard constipation protocols to clear the stool in a timely manner, especially in the face of increasing abdominal distension, vomiting, decreased appetite, stoppage of food consumption and/or abdominal pain warrants surgical or GI consultation. Emergent surgical or colonoscopic intervention may be necessary.

Prader-Willi Syndrome (USA) ALERT!
Risk of Stomach Necrosis and Rupture
Possibly Related to Chronic Gastroparesis
A Cause of Death from Sepsis, Gastric Necrosis or Blood Loss

Signs and symptoms of stomach necrosis and rupture:

- **Vomiting**- Any vomiting is very unusual in Prader-Willi syndrome
- **Loss of appetite**- (ominous sign)
- **Lethargy**
- **Complaints of pain, usually non-specific**- Pain sensation is abnormal in Prader-Willi syndrome due to high pain threshold; rarely complain of pain
- **Pain** is often poorly localized
- **Peritoneal signs** may be absent
- **Abdominal/stomach bloating and gastric dilation**
- **Fever** may or may not be present
- **Temperature regulation** is altered in Prader-Willi syndrome
- **Guaiac positive stools** (chronic gastritis)

An algorithm for ER evaluation of an individual with PWS and abdominal complaints is on a foldout page in the back of this publication.
These Signs should raise suspicion of STOMACH NECROSIS/RUPTURE as a possible diagnosis which can be LIFE-THREATENING!

History may include:
- History of binge eating within the week
- Hyperphagia and binge eating are characteristic of Prader-Willi syndrome, regardless of whether obese or slim
- Frequently occurs after holiday, or social occasion with less supervision of intake
- History of gastroparesis- Common in Prader-Willi syndrome, though often undiagnosed
- Often slim or history of significant obesity followed by weight loss- May leave the stomach wall thinned

http://www.pwsausa.org and view Medical section under Gastric/Intestinal.

IN THE EVENT OF DEATH
In the case of a death or impending death, please call PWSA (USA) immediately at 1-800-926-4797 for support and advice.
**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 been noted. PWSA (USA) has a formal investigation of causes of death.

PWSA (USA) also provides bereavement support to families who have lost children with PWS. Please call PWSA (USA) to report a death and also so the family can receive support.

**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.

For donating brain tissue, contact the Brain and Tissue Bank at the University of Maryland, (800) 847-1539.

[http://www.pwsausa.org](http://www.pwsausa.org) and view Medical section under “In the Event of Death”.
Evaluation of Individuals with Prader-Willi Syndrome with GI Complaints

- Known or suspected binge, vomiting, lethargy, or abdominal pain/distension
  - Admit Patient for Observation

  - NPO, and place NG. Urgent surgery consultation, CT abdomen
  - Rectal exam
  - Consider CBC, cortisol, CMP, amylase, lipase, lactate, ABG, PT/PTT, UA, BG

- NO
- Is Patient stable and in no distress?

- YES
- Recommend NPO and KUB flat plate and upright
- Rectal exam for stool guaiac

- Is there evidence of gastric distension or constipation/impaction/obstruction?

  - Free Air?

  - Evidence of gastric necrosis or perforation?

    - YES
    - Emergent Surgery
      - There is no advantage to waiting for surgery.

      - Admit to ICU for observation
      - Surgical consultation
      - Check plain film every 6–8 hours unless examination changes
      - Assess for other etiologies

    - NO

      - Constipation
        - Stool cleanout regimen
        - Check for impaction either digitally or with fleet enema prior to starting oral cleanout
        - Calorie restricted clear liquid diet
        - Check TFEs, electrolytes
        - Consider GI or surgical consultation, especially if stool guaiac positive
        - Reassess plain film in 12–24 hours
        - Increasing distension or failure to clear stool may indicate impaction or obstruction. Further GI consultation should be considered

      - Gastric distension
        - NPO
        - NG for decompression
        - Make surgery aware of Patient
        - Reassess plain film in 6–8 hours unless condition worsens then urgent CT
        - Consider GI consultation, especially if guaiac positive stool

      - Neither
        - Check for UTI, pancreatitis, choledolithiasis, etc.
        - RUQ ultrasound
        - If patient vomiting consider esophageal obstruction or toxic ingestion

- NOTE:
  - Individuals with Prader-Willi syndrome have decreased muscle tone and pain response so clinical evaluation for acute abdomen is altered similar to a patient on steroids.

---

**Prader-Willi Syndrome Association**

*Still hungry for a cure.*

*Credit: James Loker, M.D., Pediatric Cardiologist • Ann Scheimann, M.D., M.B.A., Gastroenterologist*  
**PWSA (USA) Clinical Advisory Board Members**

www.pwsausa.org
Important Notes
PWSA (USA) gratefully acknowledges the printing of these booklets was made possible by a gift from: Dr. James and Carolyn Loker in honor of their daughter Anna.