Evaluation of Individuals with Prader-Willi Syndrome with New GI Complaints

Known or suspected binge, unusual vomiting, loss of appetite, 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

Is Patient stable and in no distress?

- NO
  - Evidence of free air, gastric necrosis or perforation?
    - YES
      - Emergency 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
  - YES
    - Constipation
      - Stool cleanout regimen
      - Check for impaction either digitally or with fleet enema prior to starting oral cleanout
      - Calorie restricted clear liquid test
      - Check TFTs, CMP, phos, mag
      - 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 or surgical consultation should be considered.

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

- NO
  - Gastric distension or constipation / impaction / obstruction?
    - YES
      - Recommendations for NPO and KUB flat plate and upright
    - NO

- Neither
  - Check for UTI, pancreatitis, cholelithiasis, 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.