Evaluation of Individuals with Prader-Willi Syndrome with GI Complaints

USA PRADER-WILLI SYNDROME ASSOCIATION Still hungry for a cure.

IPWSO INTERNATIONAL PRADER-WILLI SYNDROME ORGANISATION

Known or suspected binge, vomiting, lethargy, or abdominal pain/distension

Admit Patient for Observation

Is Patient stable and in no distress?

No

- Nothing by mouth, and place nasogastric tube. Urgent surgery consultation, Computed tomography of abdomen, Rectal exam
- Consider complete blood count, cortisol, metabolic profile, amylase, lipase, lactate, blood gas, coagulation studies, urine analysis

Evidence of gastric necrosis or perforation?

Yes

Emergent Surgery

There is no advantage to waiting for surgery.

No

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

Yes

- Recommend nothing by mouth and abdominal flat plate and upright
- Rectal exam for stool guaiac

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

Free Air?

Constipation

- Stool cleanout regimen
- Check for impaction either digitally or with fleet enema prior to starting oral cleanout
- Calorie restricted clear liquid diet
- Check thyroid, electrolytes
- Consider gastrointestinal 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

- Nothing by mouth
- Nasogastric tube for decompression
- Make surgery aware of Patient
- Reassess plain film in 6-8 hours unless condition worsens then urgent abdomen computed tomography
- Consider Gastrointestinal consultation, especially if guaiac positive stool

Neither

- Check for urinary tract infection, pancreatitis, cholelithiasis, etc.
- Abdominal 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.

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