

# Rapunzel Syndrome

Aashini Patel

7/21/2020

RAD 4003 Pedi Elective

Dr. Susan Greenfield



# Clinical History

7 yo female who presented with episodic bilious vomiting, abdominal pain for 1 week

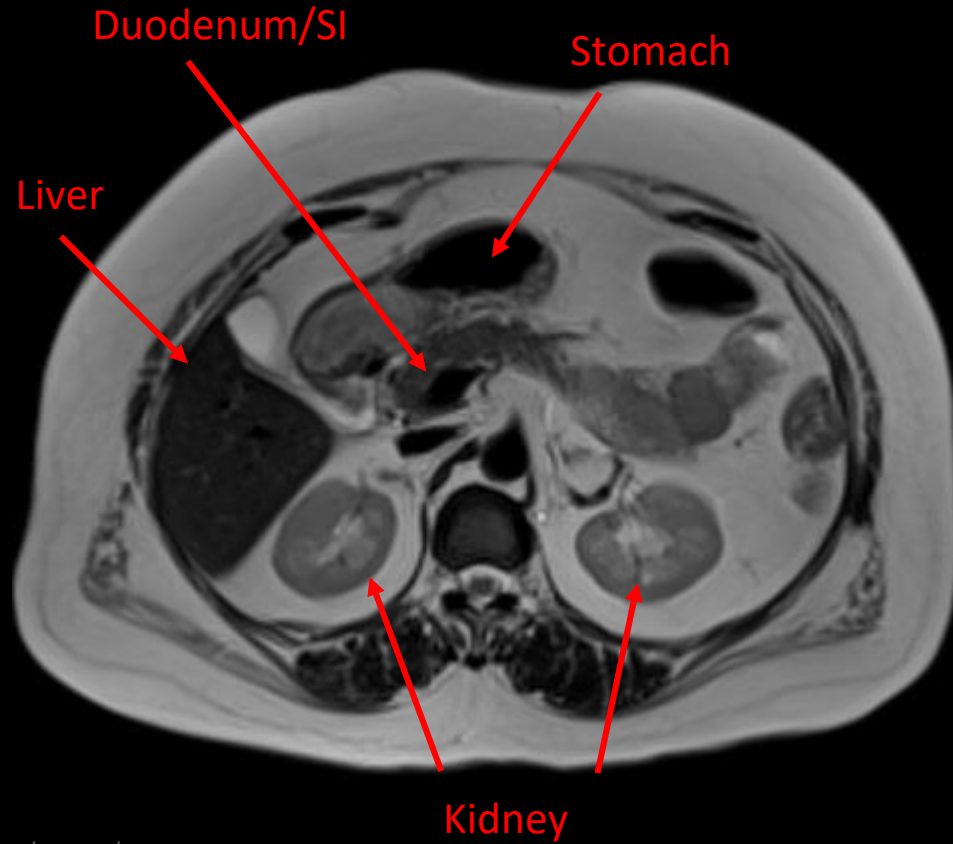
- **Associated symptoms:** severe pain that radiates to her back, weight loss, decreased PO intake and urinary output
- **Hx of hair pulling** since 2yo and other nervous habits/ hx of being bullied and constipation
- **PMHx:** nocturnal enuresis
- **Family Hx:** Mother -OCD, anxiety
  
- **Physical Exam:** tender to palpation in the epigastric region. Palpable **rubbery mass** in the epigastric region/RUQ about 1 cm, hair thinning

# Clinical History-Labs

- Glucose: 64
- **Lipase: 2255** uptrending to 7000s during inpt stay over 3 days
- Tbili: 1.4
- Normal electrolytes, creatine, AST/ALT,/Alk phos, GGT
- UA: ketones, leukocyte esterase, WBC
  
- Initial U/S: mild right hydronephrosis and gallbladder sludge, normal pancreas
  
- Pt was treated for acute pancreatitis and continued to vomit during the course of her stay

# Normal Images

T2 MRI



<https://radiopaedia.org/cases/normal-upper-abdominal-mri?lang=us>

AP X-ray

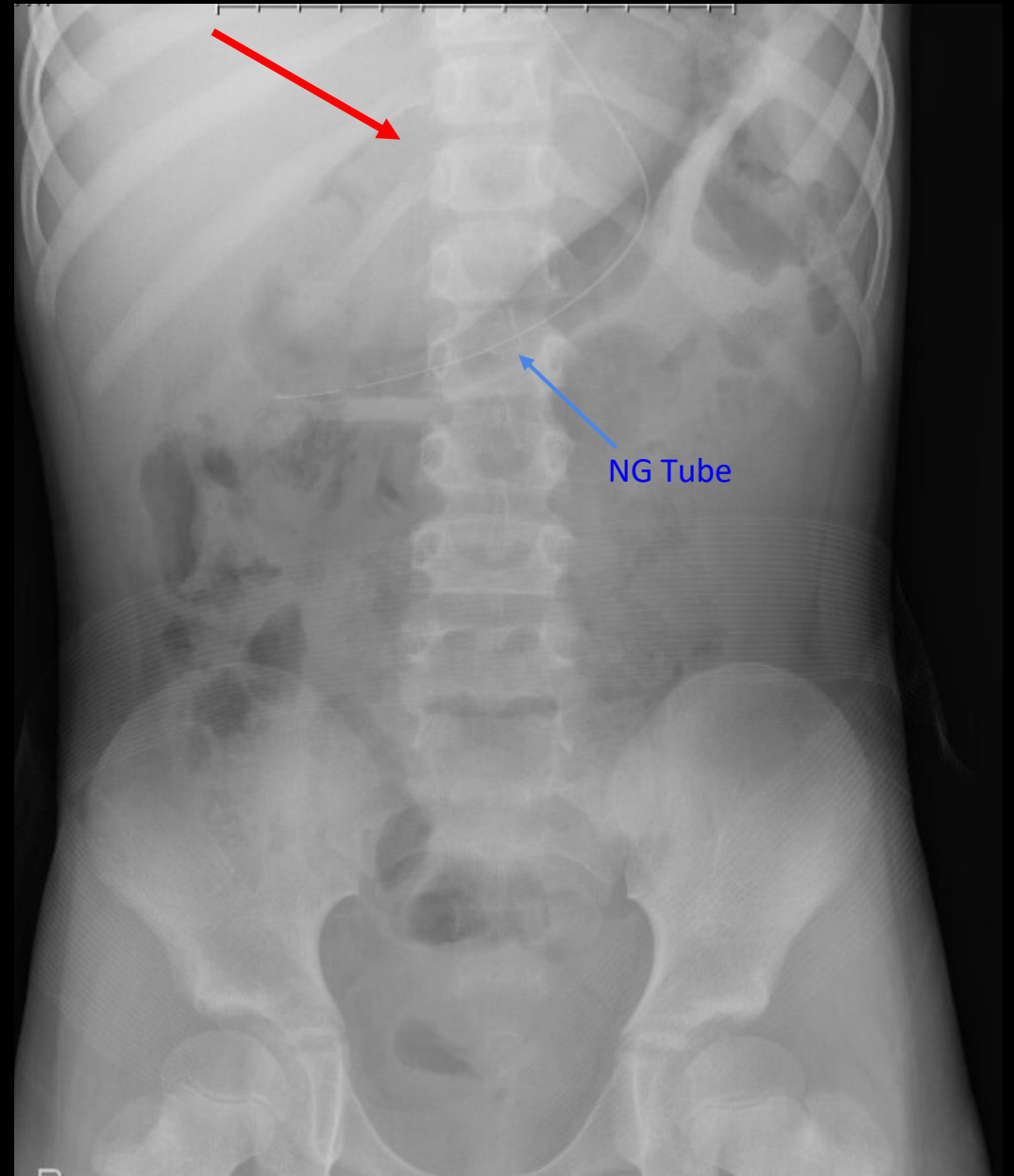


Normal air gas pattern

<https://radiopaedia.org/articles/paediatric-abdomen-ap-supine-view?lang=us>

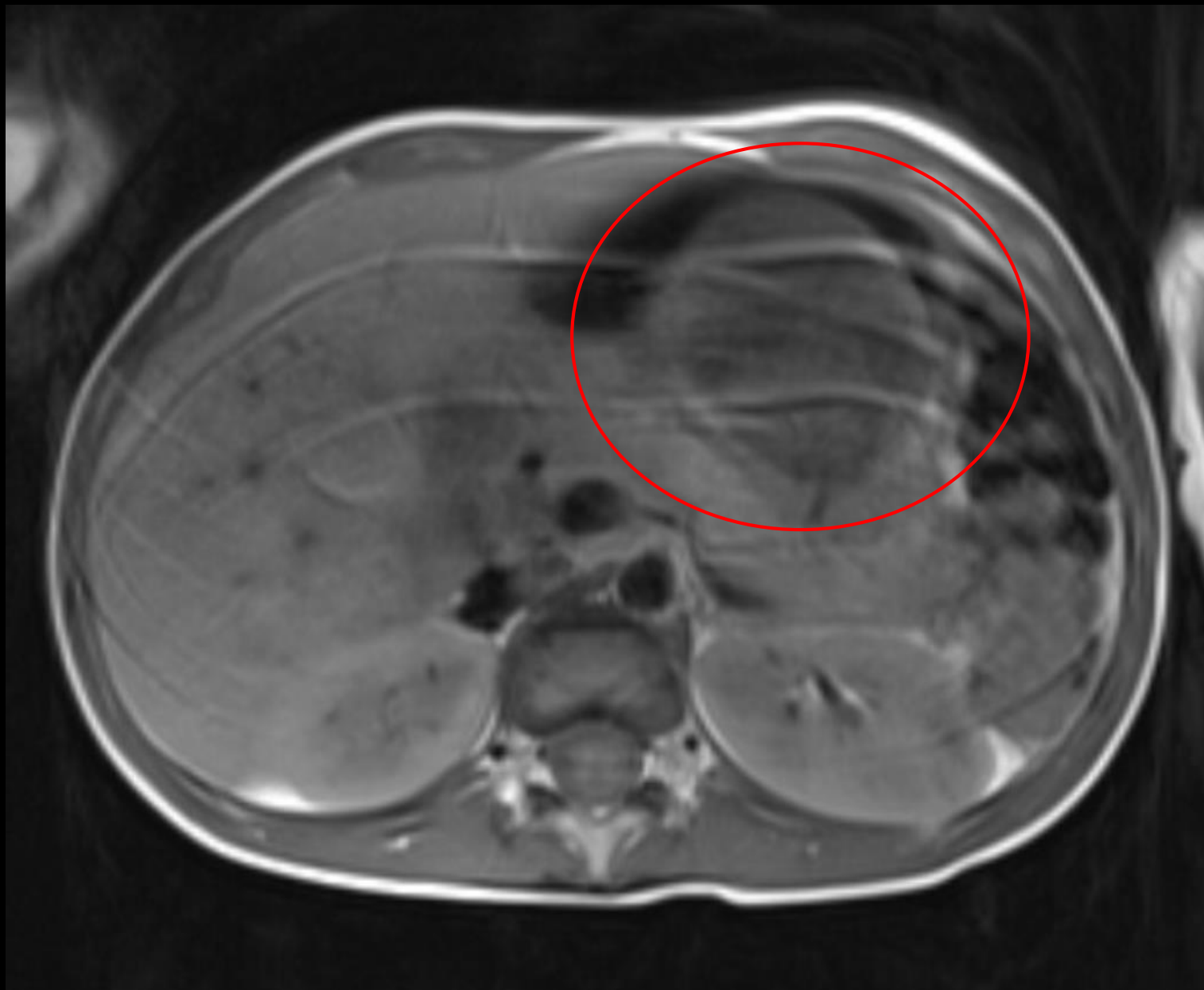
6/29: Abdomen 1 view X-ray

Incidental finding of a **dense ovoid material** in the stomach with attempt of NG tube placement



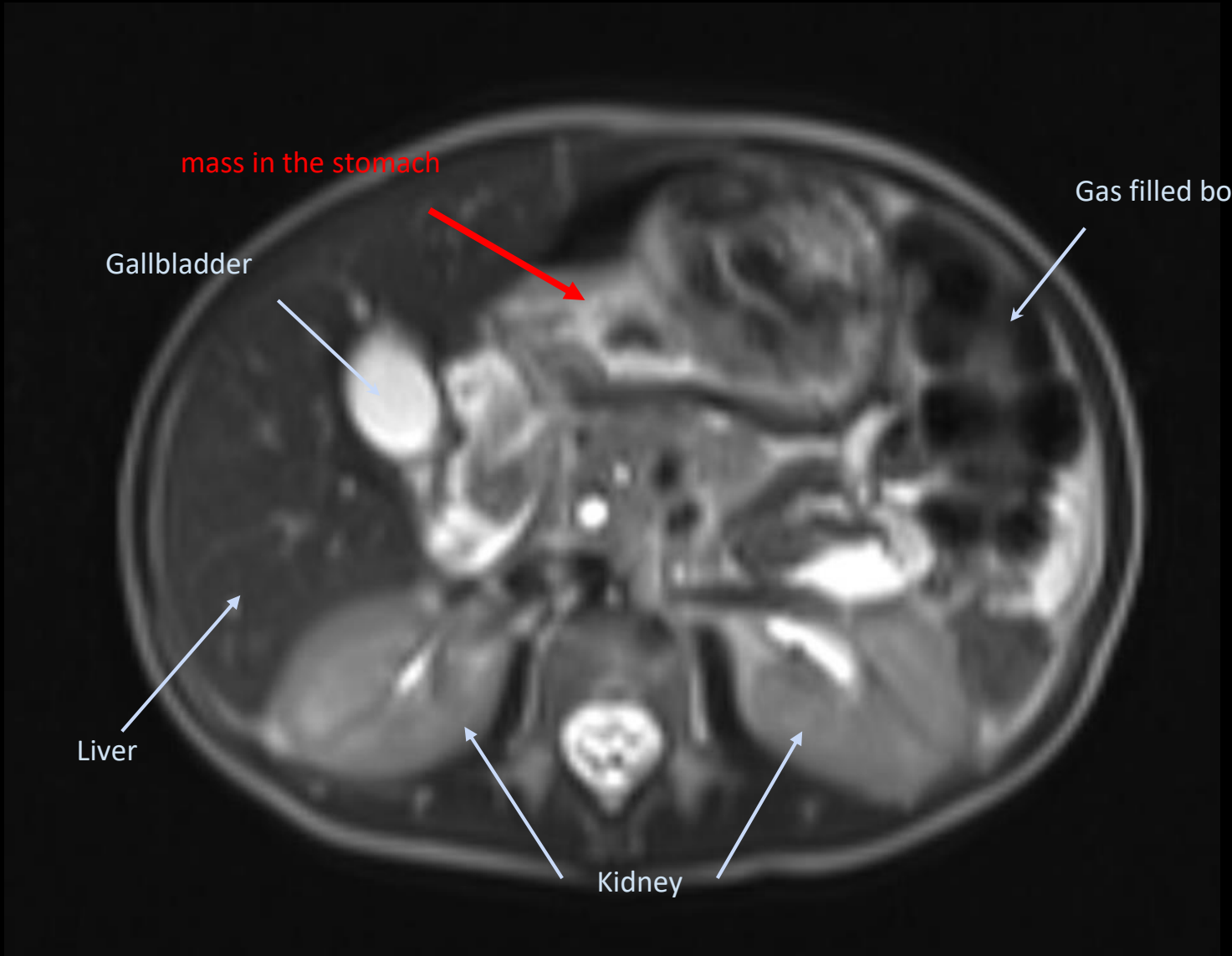
6/29- MRI w/o contrast

T1 Axial



6/29- MRI w/o contrast

T2 Axial view



6/29- Axial T2 MRI

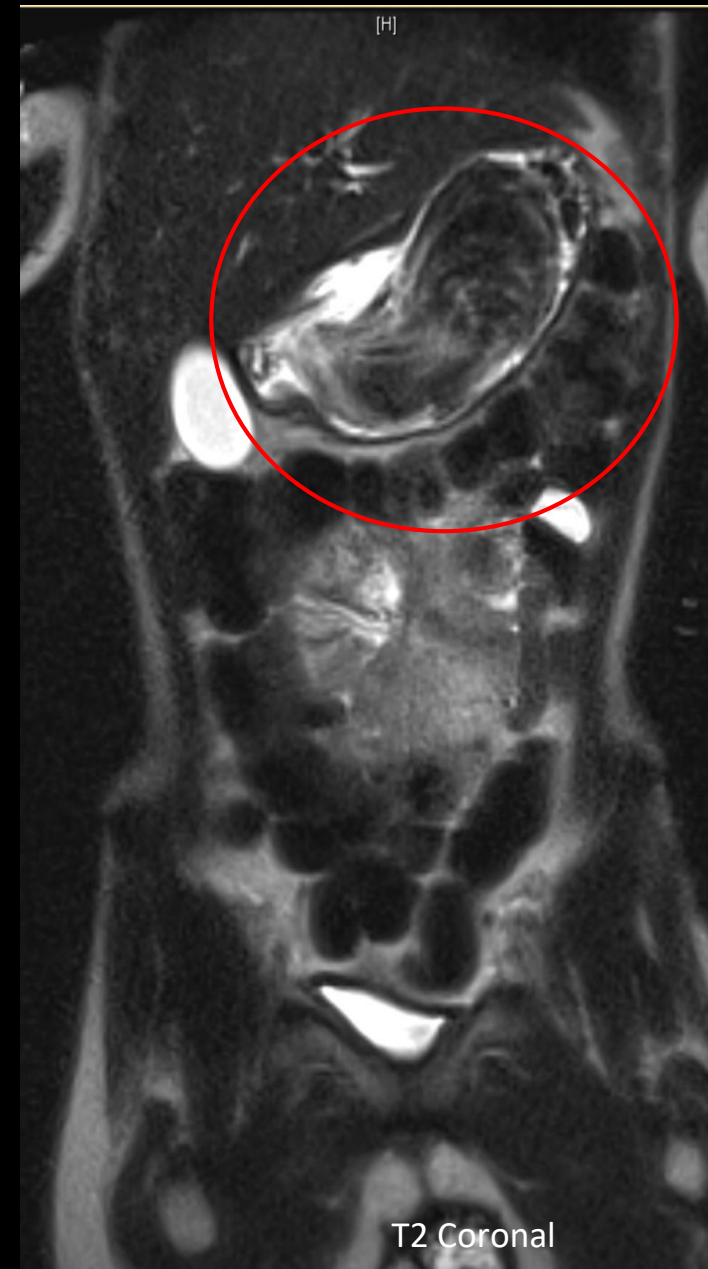




6/29- MRI w/o  
contrast



T2 Sagittal



T2 Coronal

# Key Imaging Findings

Pt with n/v and epigastric pain and a palpable rubbery mass being managed for acute pancreatitis

- U/S showed some peripancreatic fluid but no apparent mass
- Abdominal XR showed a dense ovoid material in the stomach with the attempted placement of NG tube
- MRI showed a tubular structure that is T2 hypointense mass found occupying the stomach and extending into the first part of the duodenum

# Differential Diagnosis

- **Bezoar-trichobezoar, lactobezoar, phytobezoar**
  - Presents with abd pain, bowel obstruction/constipation, hx of anxiety or psychiatric illnesses
- **Gastric carcinoma**
  - Rare
  - weight loss, n/v, abdominal pain, anemia
- **Intramural mass**
  - GI stromal tumor, metastasis, lymphoma
    - anemia, wt loss, n/v, abdominal pain, can be asymptomatic
- **Post-prandial food**

# Final Diagnosis

- **Trichobezoar** that fully occupied the **stomach** and extended to the **duodenum** due to **trichotillomania** or nervous habit
- **Rapunzel syndrome**: trichobezoar that extends into the small intestine

# Discussion-Trichobezoar

- Caused by ingesting hair with an underlying psychiatric disorder-trichotillomania/ trichophagia
  - Hair pulling seen in 1-4% of population of that 5-18% ingest
- Hair can not pass peristalsis→ mass forms combining with food→ obstructed at the pylorus over time
- Seen in female children 6-10 years old
- GI symptoms present when the bezoar is more advanced in size
- Increased morbidity compared to phytobezoar
  - Phytobezoar-vegetable/ fiber matter
    - most common
    - tx: medically managed

# Discussion

- **Symptoms**: halitosis, abdominal pain, n/v, weight loss, early satiety, can also be asymptomatic/incidental findings on imaging
  - palpable mass in LUQ
- **Complication**: pancreatitis (rare), constipation, gastric ulcers, obstruction
- **Dx**: abdominal radiograph +/- barium, non contrast CT, confirmed with EGD to get samples (Gold standard)
- **Management**: endoscopic removal, surgery
  - if palpable/ complicated bezoar: surgery
  - Common to reoccur
    - Need behavior modification/therapy

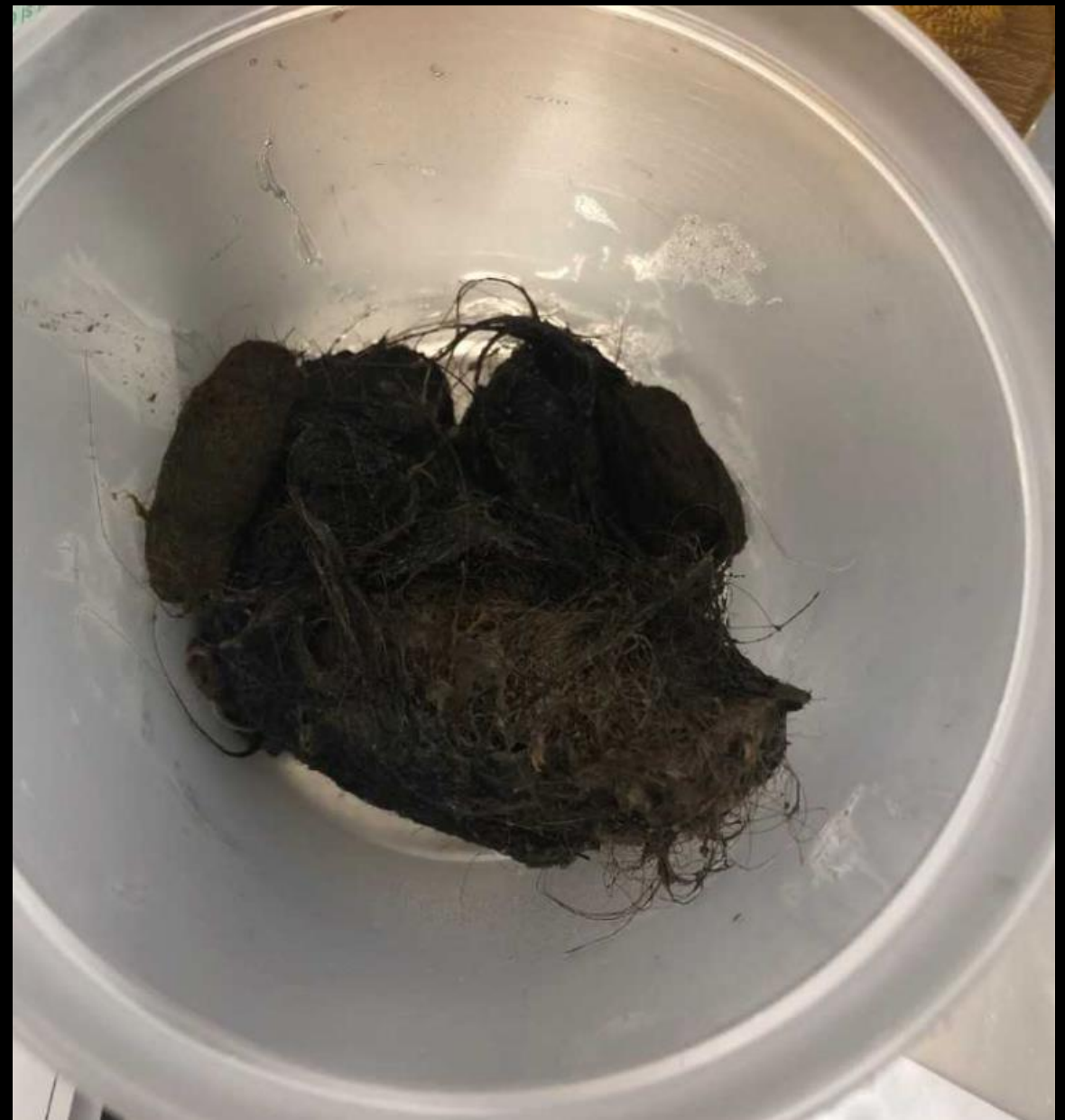
# Discussion-Imaging

- Only 18% of bezoars are seen on **X-ray**
- **U/S**: determine gastric bezoar 25% of the time
- **CT scan** show floating mass at the air-fluid level
  - correlates 97% with surgical findings
- **MRI**-not as useful
  - harder to distinguish bezoar with surrounding tissue/structure

# Treatment

Pt was taken to surgery

- Ex-lap, gastrotomy with removal of bezoar, EGD to remove remaining bezoar in the duodenum
- Bezoar occupied the entire stomach and extended into the first part of the duodenum
- Multiple ulcer in the stomach and the duodenum
- Recommended Habit reversal therapy





# ACR appropriateness Criteria-Palpable mass

- There was not a ACR criteria for bezoar.
- Following ACR criteria based on pt's presentation of abdominal pain, palpable mass, and acute pancreatitis

Revised 2019

American College of Radiology  
ACR Appropriateness Criteria®  
Palpable Abdominal Mass-Suspected Neoplasm

**Variant 1:** Palpable abdominal mass. Suspected intra-abdominal neoplasm. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen with IV contrast	Usually Appropriate	⊕⊕⊕
US abdomen	Usually Appropriate	○
MRI abdomen without and with IV contrast	May Be Appropriate	○
CT abdomen without IV contrast	May Be Appropriate	⊕⊕⊕
MRI abdomen without IV contrast	May Be Appropriate	○
CT abdomen without and with IV contrast	Usually Not Appropriate	⊕⊕⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕⊕
Radiography abdomen	Usually Not Appropriate	⊕⊕
Fluoroscopy contrast enema	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy upper GI series	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	⊕⊕⊕

# ACR Criteria-Acute non-localized abdominal pain

**Variant 4:**

**Acute nonlocalized abdominal pain. Not otherwise specified. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
CT abdomen and pelvis with IV contrast	Usually Appropriate	⊕⊕⊕
CT abdomen and pelvis without IV contrast	Usually Appropriate	⊕⊕⊕
MRI abdomen and pelvis without and with IV contrast	Usually Appropriate	○
US abdomen	May Be Appropriate	○
MRI abdomen and pelvis without IV contrast	May Be Appropriate	○
CT abdomen and pelvis without and with IV contrast	May Be Appropriate	⊕⊕⊕⊕
Radiography abdomen	May Be Appropriate	⊕⊕
FDG-PET/CT skull base to mid-thigh	Usually Not Appropriate	⊕⊕⊕⊕
WBC scan abdomen and pelvis	Usually Not Appropriate	⊕⊕⊕⊕
Nuclear medicine scan gallbladder	Usually Not Appropriate	⊕⊕
Fluoroscopy upper GI series with small bowel follow-through	Usually Not Appropriate	⊕⊕⊕
Fluoroscopy contrast enema	Usually Not Appropriate	⊕⊕⊕

# ACR Criteria-Acute Pancreatitis

**Variant 1:**

**Suspected acute pancreatitis. First-time presentation. Epigastric pain and increased amylase and lipase. Less than 48 to 72 hours after symptom onset. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
US abdomen	Usually Appropriate	0
CT abdomen and pelvis with IV contrast	May Be Appropriate	☼☼☼
MRI abdomen without and with IV contrast with MRCP	May Be Appropriate	0
MRI abdomen without IV contrast with MRCP	May Be Appropriate	0
US duplex Doppler abdomen	May Be Appropriate	0
CT abdomen and pelvis without and with IV contrast	Usually Not Appropriate	☼☼☼☼
CT abdomen and pelvis without IV contrast	Usually Not Appropriate	☼☼☼
US abdomen with IV contrast	Usually Not Appropriate	0

- The imaging modality of **US, Abdominal, Radiograph, and MRI without contrast** were appropriate.
- CT without contrast might have been better to visualize the mass

# Cost of Imaging-MHH Inpt

- Cost of bezoar imaging:

- Abdomen 1 view X-ray (1): \$670
- Abd MRI w/o contrast (1): \$4,610

>TOTAL: \$5,280

- Total cost of imaging during inpatient stay:

- Abdomen 2 view X-ray (1): \$771
- Abdomen 1 view X-ray (5):  $\$670 \times 5 = \$3,350$
- US Abdomen limited (3):  $\$1,493 \times 3 = \$4,479$
- Abd MRI w/o contrast: (1): \$4,610
- Chest X-ray 1 view (1): \$683

>TOTAL: \$13,893

# Take Home Points

- Pt is symptomatic when the bezoar is quite extensive
- Consider trichobezoar on the differential in female children with abdominal mass/pain, GI symptoms, underlying psychiatric illnesses
- Best diagnosed with non contrast CT and confirmed with EGD + sampling
- Acute pancreatitis is a rare complication of bezoar
- Treatment is surgery severe, advanced bezoar

# References

- Aybar, Ahmet, and Anca M. Safta. "Endoscopic Removal of a Gastric Trichobezoar in a Pediatric Patient." *Gastrointestinal Endoscopy* 74.2 (2011): 435–437. Web.
- Mathai, John et al. "Rapunzel Syndrome: a Diagnosis Overlooked." *Acta Pædiatrica* 96.1 (2007): 135–137. Web.
- Barrows, Amy et al. "Trichobezoars Detected and Treated Based on Plain Radiography." *Military medicine* 180.10 (2015): e1136–e1138. Web.
- Gorter, R.R et al. "Management of Trichobezoar: Case Report and Literature Review." *Pediatric surgery international* 26.5 (2010): 457–463. Web.
- Gonuguntla, Veena, and Divya-Devi Joshi. "Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar." *Clinical medicine & research* vol. 7,3 (2009): 99-102. doi:10.3121/cm.r.2009.822
- Subbiah, Vivek et al. "Gastric adenocarcinoma in children and adolescents." *Pediatric blood & cancer* vol. 57,3 (2011): 524-7. doi:10.1002/pbc.23051
- Kaemmer, D A et al. "The Gist of literature on pediatric GIST: review of clinical presentation." *Journal of pediatric hematology/oncology* vol. 31,2 (2009): 108-12. doi:10.1097/MPH.0b013e3181923cd8
- [https://www.uptodate.com/contents/gastric-bezoars?search=trichobezoar&source=search\\_result&selectedTitle=1~5&usage\\_type=default&display\\_rank=1#H15](https://www.uptodate.com/contents/gastric-bezoars?search=trichobezoar&source=search_result&selectedTitle=1~5&usage_type=default&display_rank=1#H15)
- <https://acsearch.acr.org/list>



Questions?