

RADY 401 Case Presentation: Extrarenal Wilms Tumor / Nephroblastoma

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Focused Patient History and Workup

- 10 y.o. male with a h/o asthma presents with
 - acute onset RLQ abdominal pain x1 day
 - 1x non-bloody, nonbilious emesis.
 - No fevers, constipation, diarrhea, urinary complaints, or testicular pain.
- Pertinent Physical Exam findings:
 - Abdominal exam shows normal bowel sounds, mildly distended but soft belly, and tenderness to palpation of RLQ w/o rebound or guarding. No hernias or masses palpated.
 - Both testicles descended appropriately.
- Labs:
 - Leukocytosis of 20.3 WBC with neutrophil predominance
 - Mild microcytic anemia : Hgb 11.8 (LLN), Hct 35 (LLN), MCV 70 (L), RDW 15.8 (H)
 - Unremarkable metabolic panel
 - Unremarkable urinalysis
 - C/F Appendicitis >>> MRI pelvis w/ and w/o IV contrast

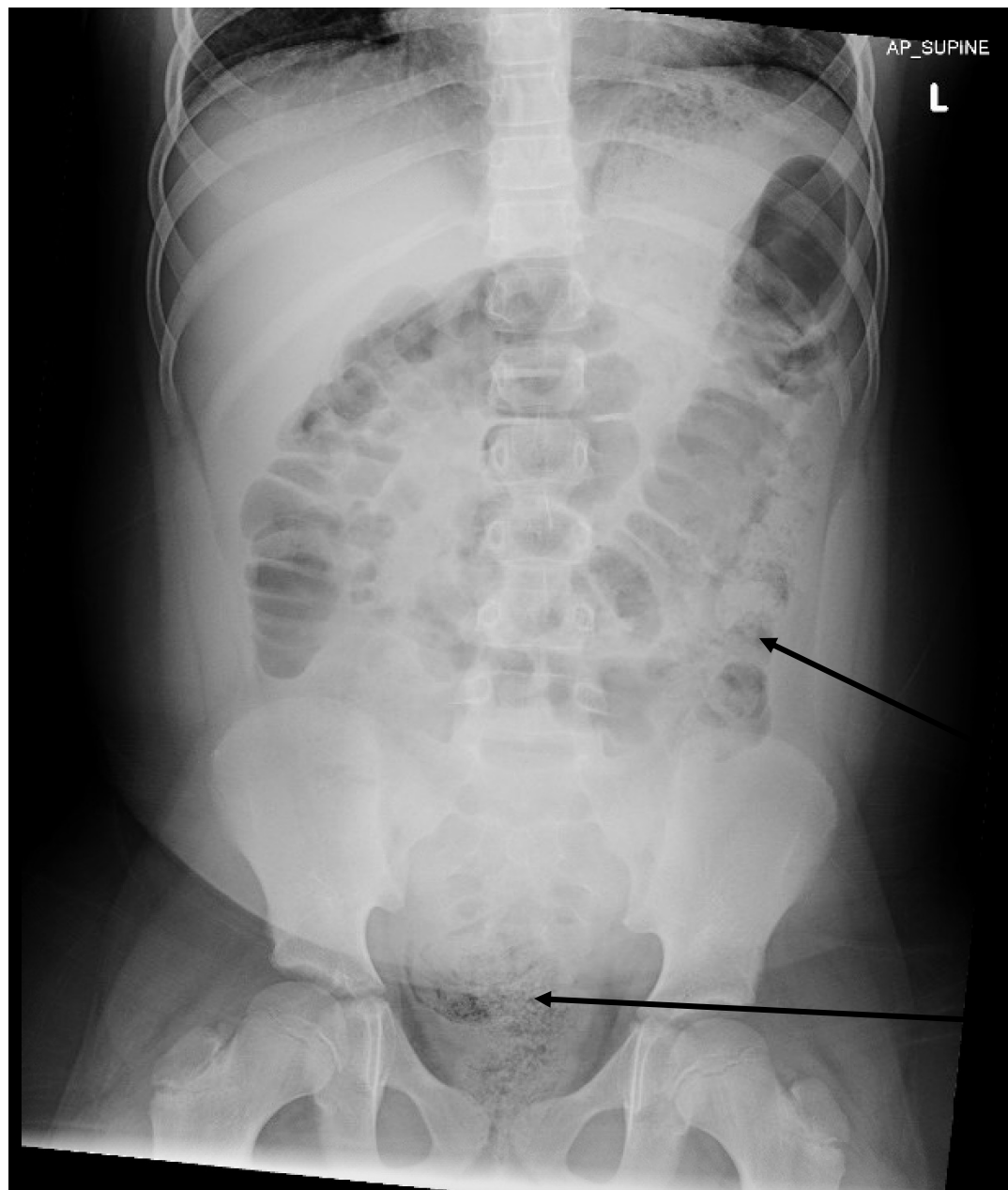
Differential Diagnosis for School-Aged Pediatric Vomiting/Abdominal Pain

- Gastroenteritis
- Appendicitis
- Diabetic Ketoacidosis
- Ovarian/Testicular Torsion
- Foreign Body Ingestion
- Volvulus
- Trauma
- Renal Dx/Stone/Pyelonephritis
- Biliary Dx

List of Imaging Studies

- ED Outside Hospital:
 - X-Ray Abdomen KUB
 - MRI Pelvis with and without IV contrast

X-Ray Abdomen KUB



Findings: *Large stool is demonstrated, throughout the colon and rectum. No dilated gas filled loops of bowel are demonstrated. No pneumatosis. No suspicious calcifications. Bones are unremarkable.*

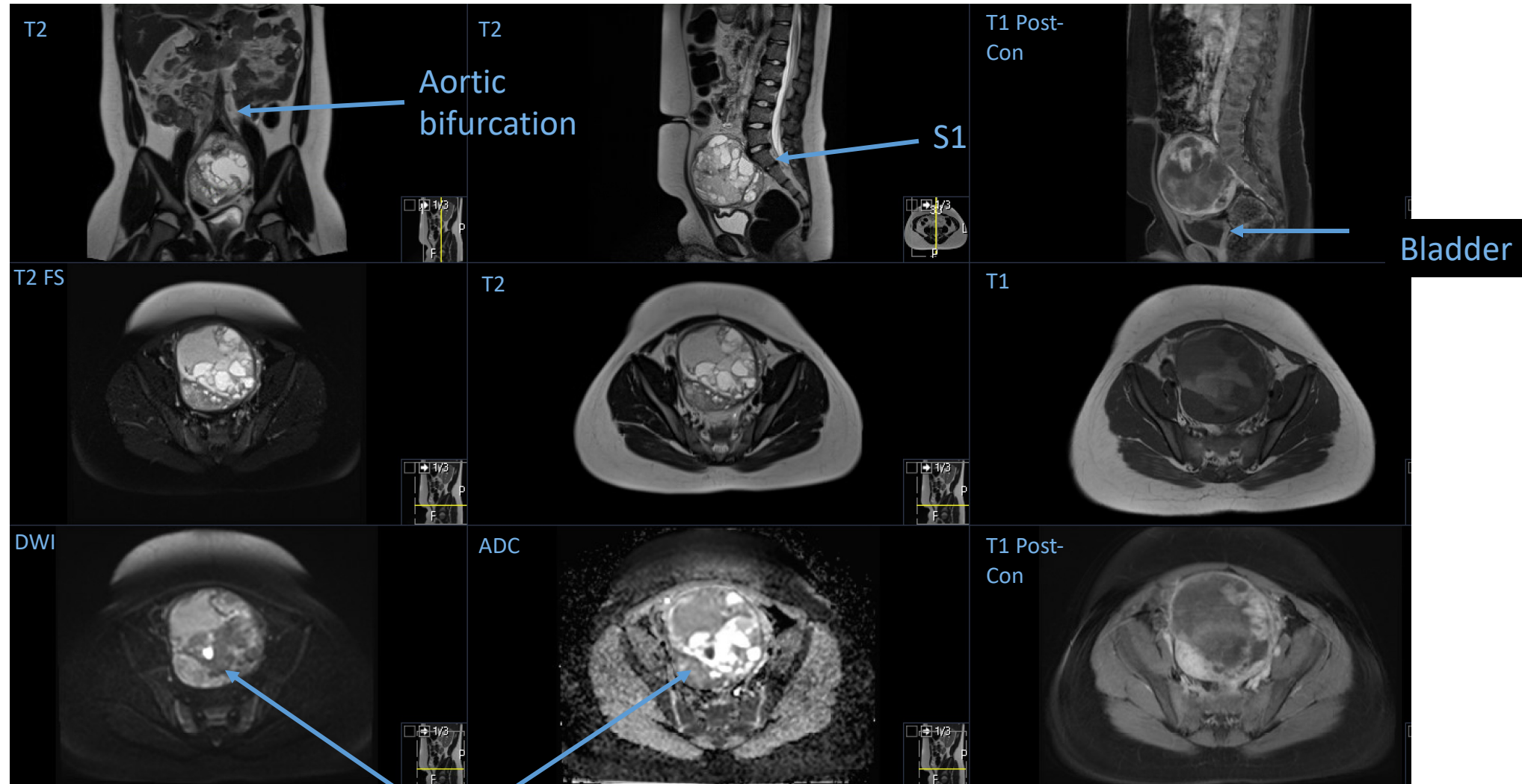
Impression: *Nonobstructive bowel gas pattern with a large colonic and rectal stool burden.*

Stool burden is recognizable on radiography as visible gas bubbles within semisolid/soft-tissue density. ⁽¹⁾

Stool burden in descending colon.

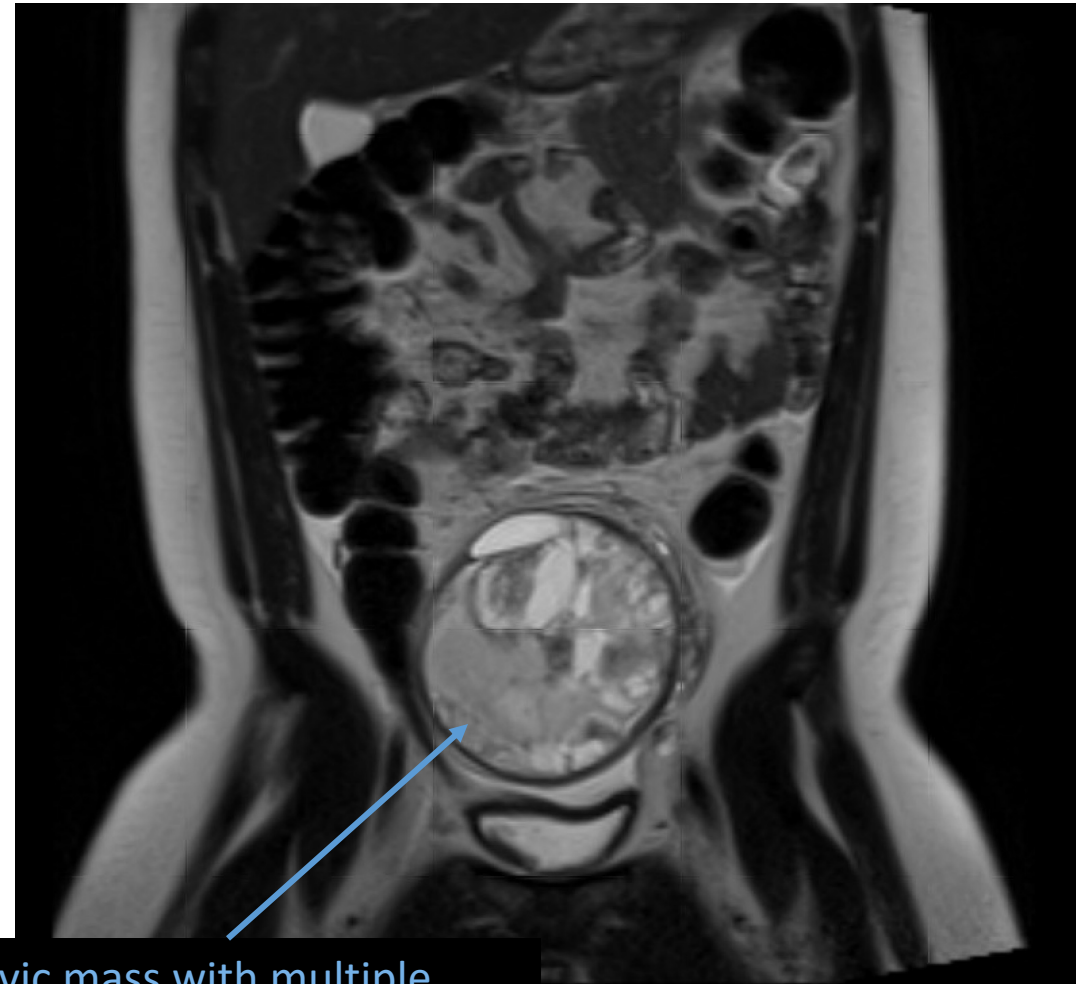
Stool burden in sigmoid colon and rectum.

MRI Pelvis with and without IV Contrast



Restricted diffusion.

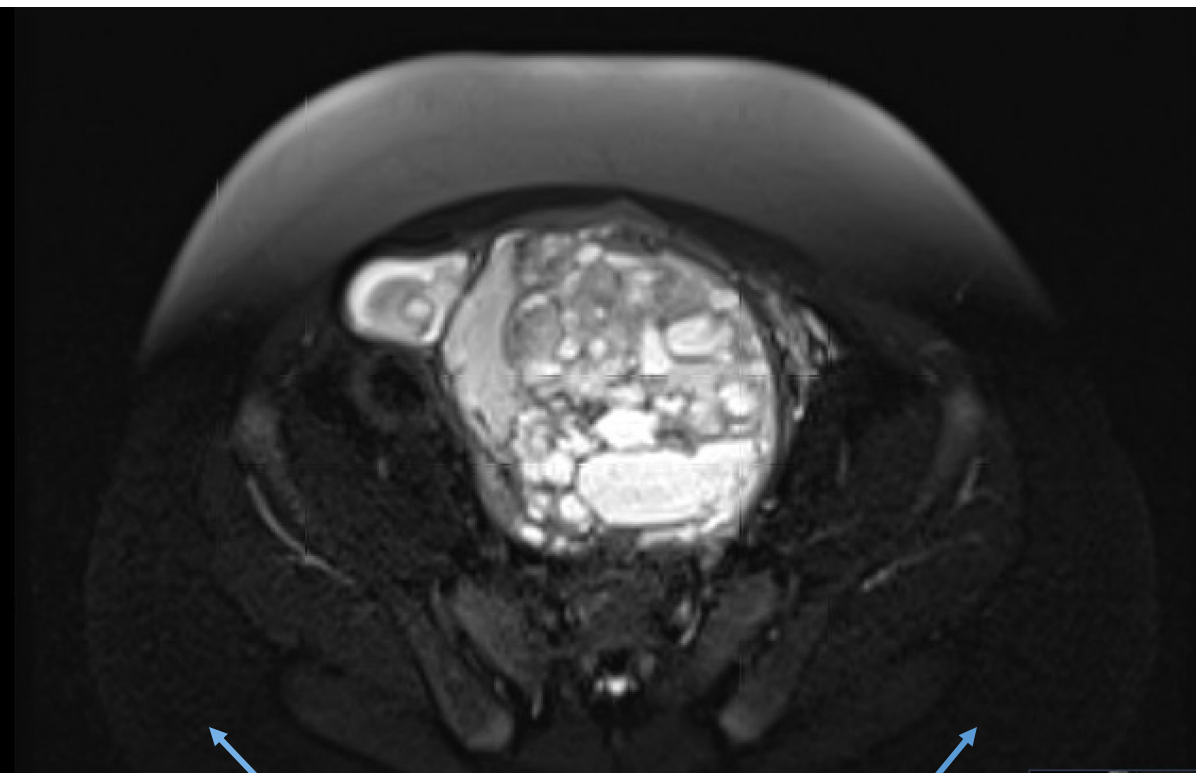
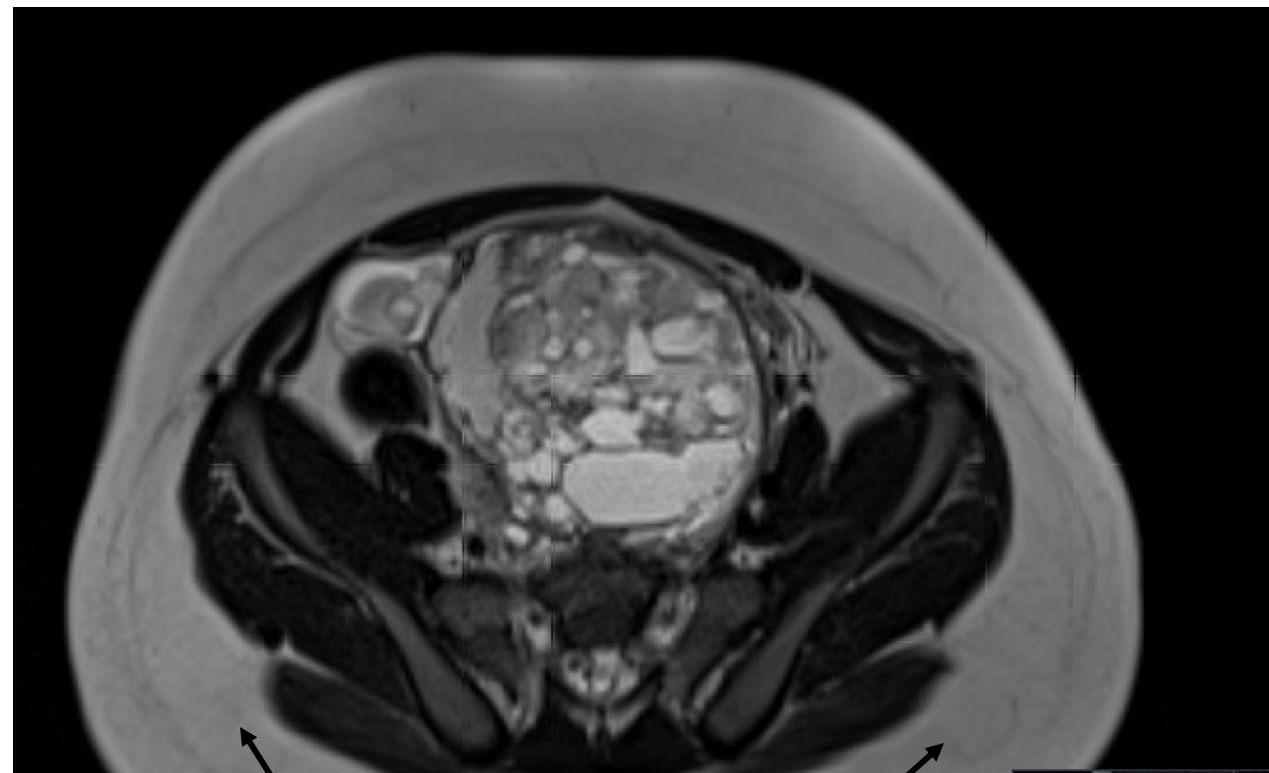
MRI T2 Coronal View



Large, heterogeneous pelvic mass with multiple internal septations with hyperintense cystic components and isointense solid components.

MRI T2 Axial View

MRI T2 Fat Saturated Axial

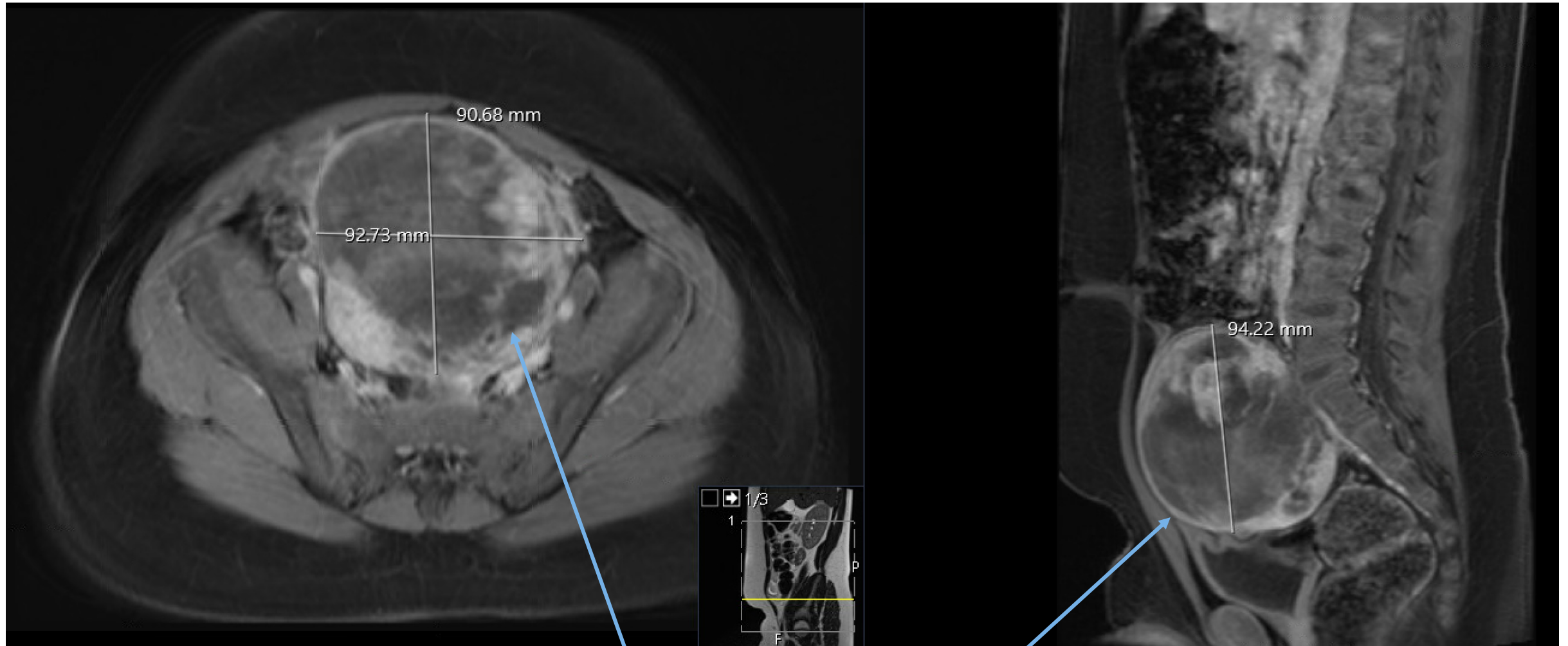


Hyperintense fat on T2.

Mass components unchanged.

Hypointense, suppressed fat on T2.

MRI T1 Post-Con Fat Saturated Axial and Sagittal Views



Large pelvic mass with peripheral enhancement and internal septation enhancement.

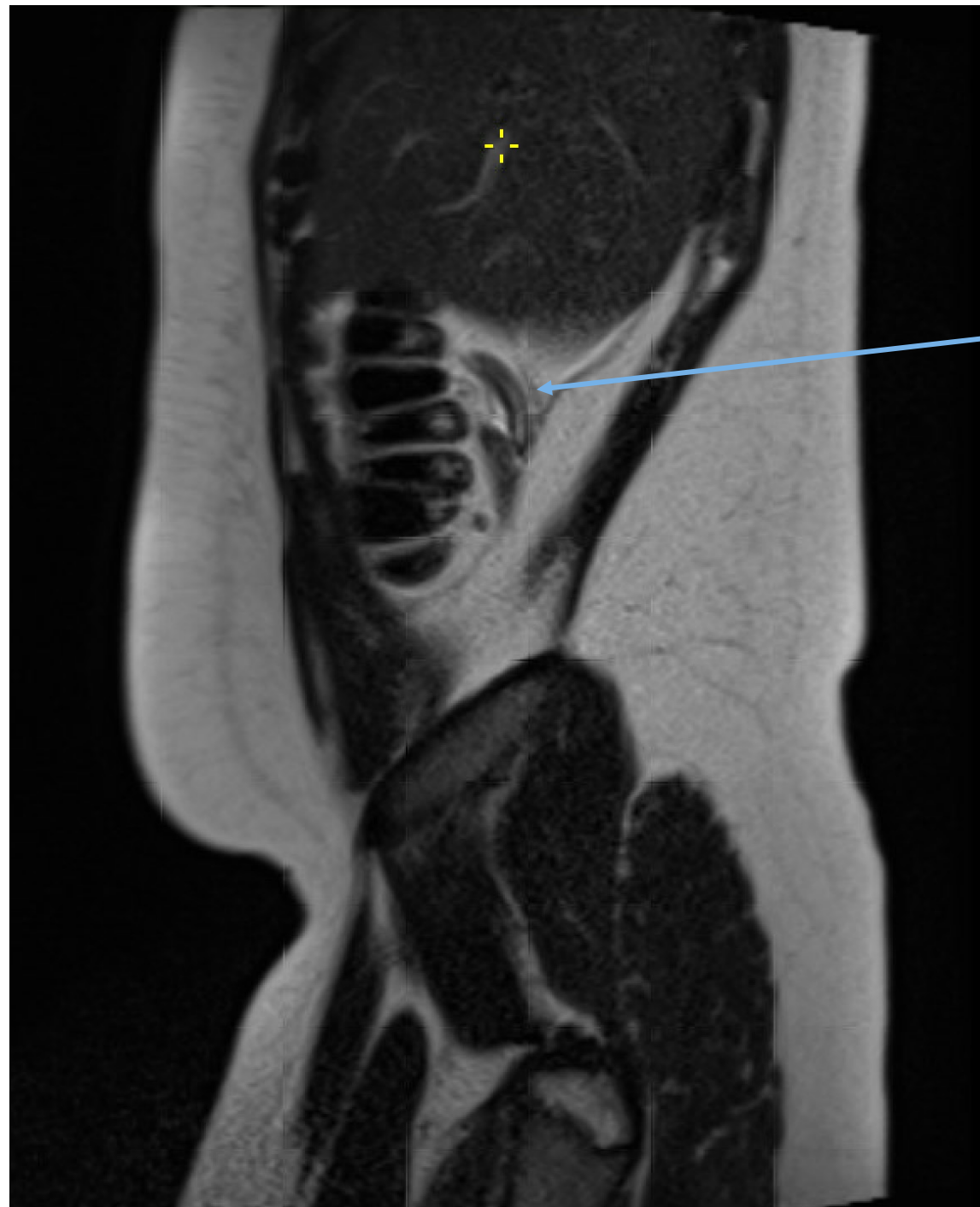


UNC

SCHOOL OF MEDICINE
Radiology

C/F Appendicitis?

MRI T2 Sagittal
View, right/middle
abdomen



Appendix Findings:
*Retrocecal
appendix is
visualized and is a
normal caliber.
There is small
amount of fluid
surrounding the
appendix.*

Final Impression, MRI Pelvis

1. *Large solid cystic pelvic mass* measuring up to 9.7 cm in the *central pelvis abutting the sacrum at S1*. *Differential* considerations would include *sacroccocygeal teratoma* although there are no definitive calcifications, *rhabdomyosarcoma*, or *extra-adrenal pheochromocytoma*.
2. *Mild RIGHT hydronephrosis*, likely external compression on the *RIGHT ureter* from the pelvic mass.
3. *Small pelvic and abdominal ascites*.

Patient treatment or outcome

- Emergent Exploratory Laparotomy and Tumor Resection
 - Large tumor originating from retroperitoneum and sacrum resected.
 - Tumor biopsied and sent to pathology for pediatric tumor work-up.
 - Evidence of pre-operative rupture with blood in abdomen and pelvis.
- Pathology:
 - *Triphasic Wilms tumor with additional heterologous elements (skeletal muscle and smooth muscle.) The amount of heterologous elements is not sufficient for use of the descriptive term "teratoid Wilms."*
- Referred to UNC Heme/Onc and tumor board review
 - CT Chest is recommended to evaluate for metastases.

ACR Appropriateness Criteria – Suspected Appendicitis-Child

American College of Radiology ACR Appropriateness Criteria® Suspected Appendicitis–Child

Variant 2:

Child. Suspected acute appendicitis, intermediate clinical risk. Initial imaging.

Procedure	Appropriateness Category	Relative Radiation Level
US abdomen RLQ	Usually Appropriate	○
US abdomen	Usually Appropriate	○
CT abdomen and pelvis with IV contrast	May Be Appropriate (Disagreement)	⚠⚠⚠⚠
CT abdomen and pelvis without IV contrast	May Be Appropriate (Disagreement)	⚠⚠⚠⚠
MRI abdomen and pelvis without and with IV contrast	May Be Appropriate (Disagreement)	○
MRI abdomen and pelvis without IV contrast	May Be Appropriate (Disagreement)	○

Pediatric EM Physician concerned for appendicitis opted for MRI over ultrasound due to patient's body habitus. BMI = 97th percentile for category.

ACR Appropriateness Criteria – MRI Abdominal and Pelvis with and without IV Contrast

Relative Radiation Level Designations

Relative Radiation Level*	Adult Effective Dose Estimate Range	Pediatric Effective Dose Estimate Range
○	0 mSv	0 mSv
☢	<0.1 mSv	<0.03 mSv
☢☢	0.1-1 mSv	0.03-0.3 mSv
☢☢☢	1-10 mSv	0.3-3 mSv
☢☢☢☢	10-30 mSv	3-10 mSv
☢☢☢☢☢	30-100 mSv	10-30 mSv

*RRL assignments for some of the examinations cannot be made, because the actual patient doses in these procedures vary as a function of a number of factors (eg, region of the body exposed to ionizing radiation, the imaging guidance that is used). The RRLs for these examinations are designated as “Varies”.

Radiation Dose = 0 mSv

Estimated Costs = \$106.93-369.59

CPT Code	Moderator	Status	Description	2021 Payment Rate	2022 Payment Rate	Percent Change in Payment Rate
72197		A	Mri pelvis w/o & w/dye	\$389.06	\$369.59	-5.0%
72197	26	A	Mri pelvis w/o & w/dye	\$107.82	\$106.93	-0.8%
72197	TC	A	Mri pelvis w/o & w/dye	\$281.24	\$262.66	-6.6%

- w/ IV Contrast: 94% sensitivity & 94% specificity
 - w/o IV Contrast: 86% sensitivity & 94% specificity

Classic Image Findings: Acute Appendicitis

- RUQ U/S:
 - Findings
 - Dilated appendix w/ diameter >6mm, noncompressible
 - Prominent appendiceal wall layers/wall thickening
 - Possible appendicolith with posterior acoustic shadowing
 - Periappendiceal fluid collection/fat ⁽⁴⁾
 - Cons
 - Operator dependent
 - Can be limited by obese body habitus, which is why managing physician ordered MRI for better visualization
 - Difficult to visualize through air or bowel ⁽⁵⁾
- MRI Pelvis & Abdomen w/ & w/o IV Contrast
 - Findings
 - Appendix lumen distension
 - Wall thickening
 - Periappendiceal free fluid
 - Cons
 - Time
 - Cost

Actual Dx: Extrarenal Wilms Tumor / Nephroblastoma

- Extrarenal manifestation is extremely rare and location can delay diagnosis due to lack of traditional presentation of palpable abdominal mass seen in toddler-aged children.⁽⁶⁾
- Renal Wilms Tumor Imaging:
 - 1ST Line: U/S = vascular tumor with hypoechoic areas of necrosis
 - Abdominal CT/MR = surgical excision mapping
 - CT Chest/Thorax = staging and metastases ⁽⁷⁾
 - This is the next step in this patient's treatment plan to assess for chest/lung metastases and determine next therapeutic steps.

Wrap Up

1. Imaging indications for appendicitis in pediatric patients aim to avoid excessive radiation exposure associated with CT.
 - a) Abdominal ultrasound
 - b) MRI abdomen and pelvis
2. Respect the limitations of ultrasound before ordering. Additionally, an equivocal or negative ultrasound study does not rule out clinically suspected appendicitis: further imaging is indicated.
3. It's not always appendicitis, but common things are common. Utilize ACR appropriateness criteria for clinical decision-making.

References

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