

# AMSER Case of the Month

## August 2021

13 yo female presents to the ED with acute abdominal pain with nausea and vomiting

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# Patient Presentation

- 13 year old female presented to the ED for acute abdominal pain with nausea and vomiting.
- ROS: negative
- PMHx: intestinal malrotation
- PSHx: status post Ladd procedure
- FamilyHx: none

# Pertinent Labs

- None

What Imaging Should We Order?

# Select the applicable ACR Appropriateness Criteria

**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  | ☼☼☼                      |

← This imaging modality was ordered by the ER physician

# Findings (unlabeled)



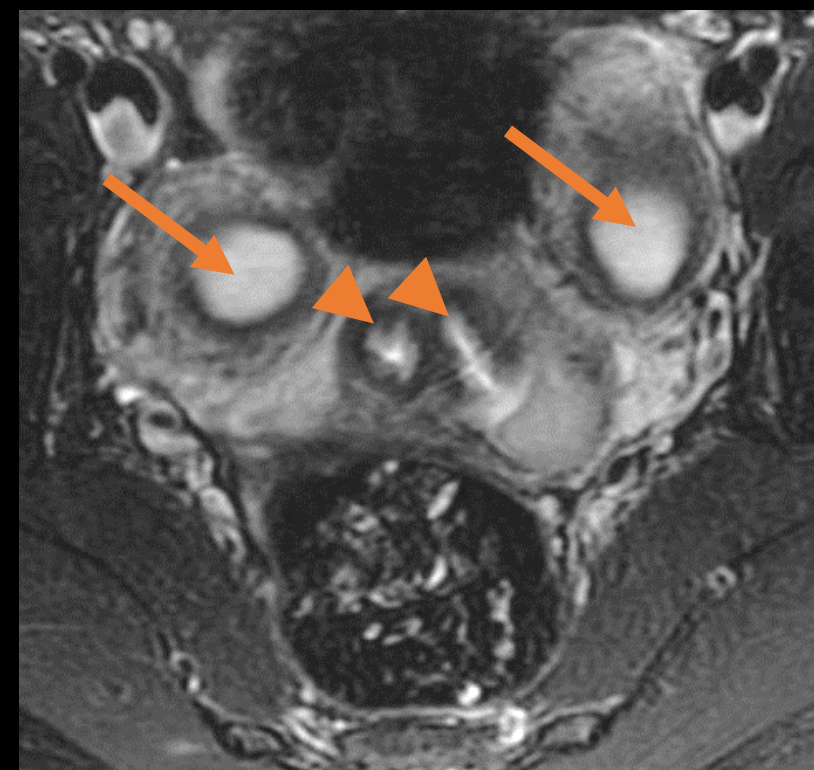
Axial CT of the Pelvis

CT abdomen pelvis demonstrated evidence of small bowel obstruction and incidental pelvic abnormality.

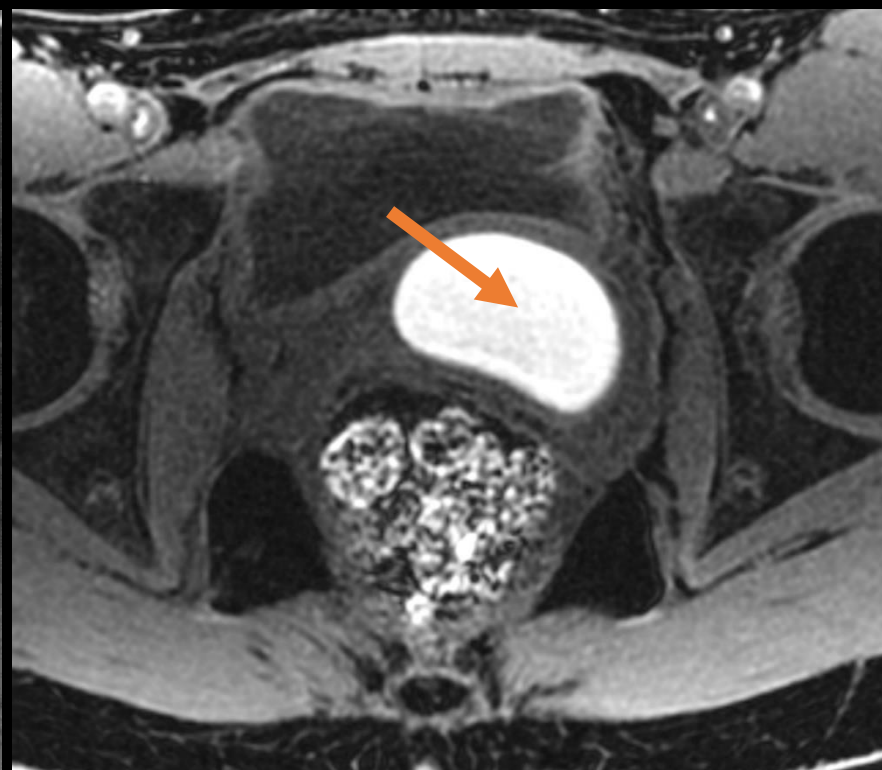
## Findings: (labeled)



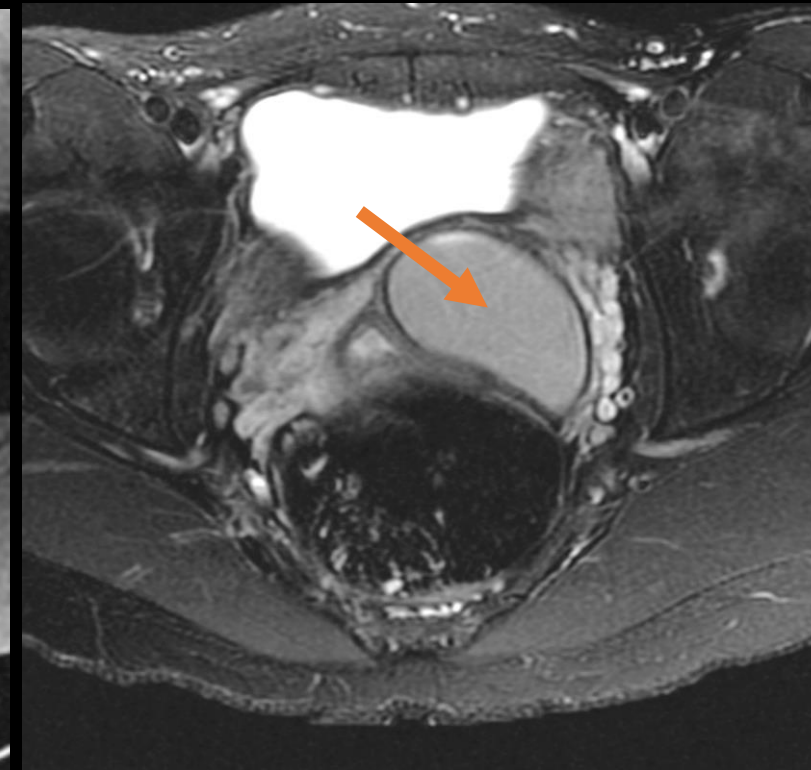
The arrows (orange) demonstrate uterus didelphys. Each arrow points to a horn of the uterus. For further evaluation and better characterization of the abnormality, the next appropriate imaging is an MRI.



A. Axial T2 oblique



B. Axial T1 image



C. Axial T2 image

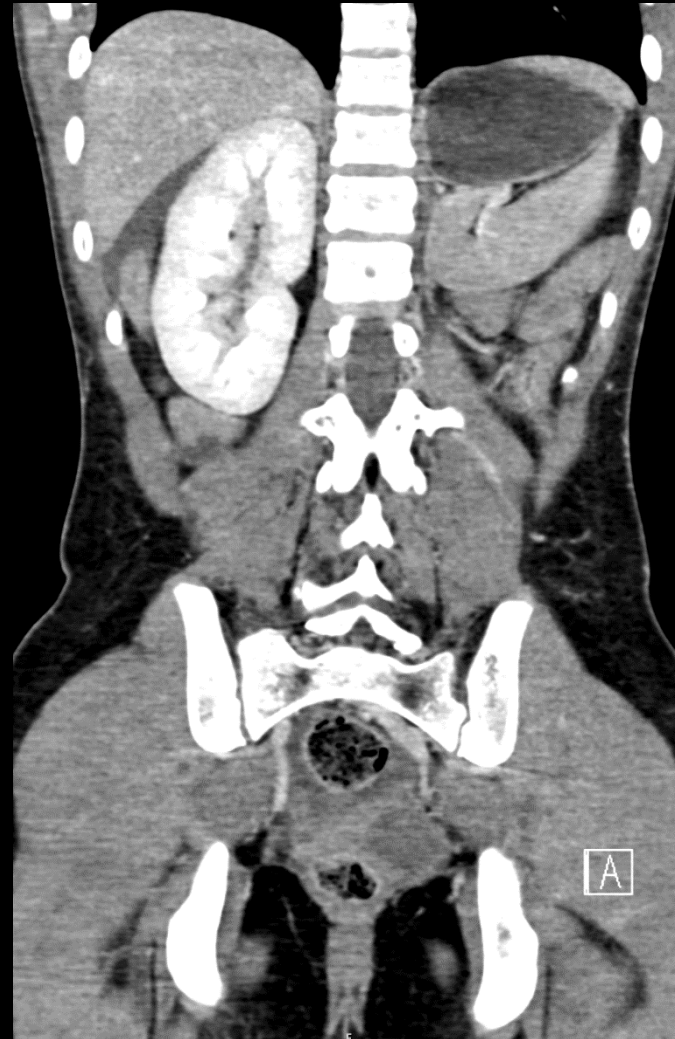
Image A demonstrates two uterine horns (arrows) with separate cervixes (arrowheads) (bicornis uterine didelphys). Images B and C shows an obstructed left hemi-vagina with T1 hyperintense/T2 intermediate fluid, consistent with hematocolpos.

What other organ should be evaluated in the disease process?



Answer: The kidneys!

A congenital anomaly of the uterus warrants further investigation of the urinary system, as Müllerian anomalies are highly associated with renal anomalies.



## Coronal CT Abdomen/Pelvis

Absent *left* kidney

## Final Dx:

OHVIRA (Obstructed Hemi-Vagina and Ipsilateral Renal Anomaly)

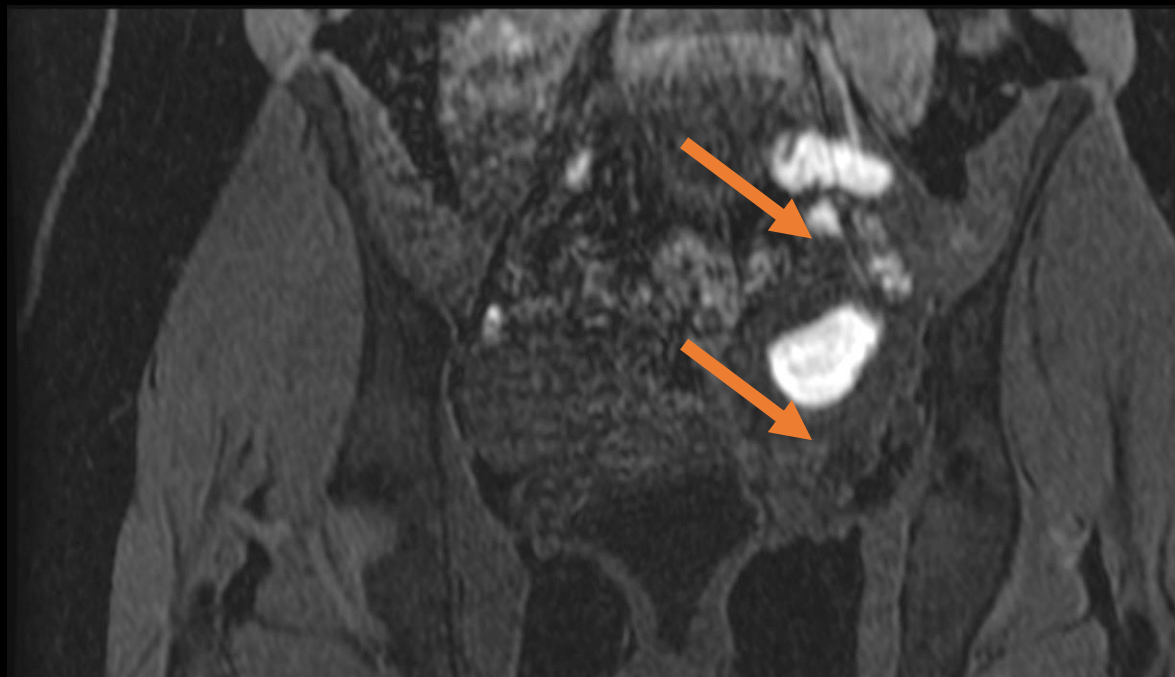
OHVIRA is characterized by a triad of uterine didelphys, unilateral renal agenesis, and unilateral hemi-vagina. It is a rare anomaly of the development of the Müllerian duct, and it typically presents after menarche. In this case, it was found incidentally during the patient's work up for acute abdominal pain.

# Pertinent companion case

14-year-old female presented with a very painful menstrual period with associated nausea and vomiting. NSAIDs and heating pads did not alleviate the pain. Diagnostic laparoscopy was performed and drainage of a left adnexal chocolate cyst along with aspiration of the left uterus. She is now status post resection of a left rudimentary horn and left fallopian tube/hemosalpinx, essentially giving her unicollis unicornuate uterus.

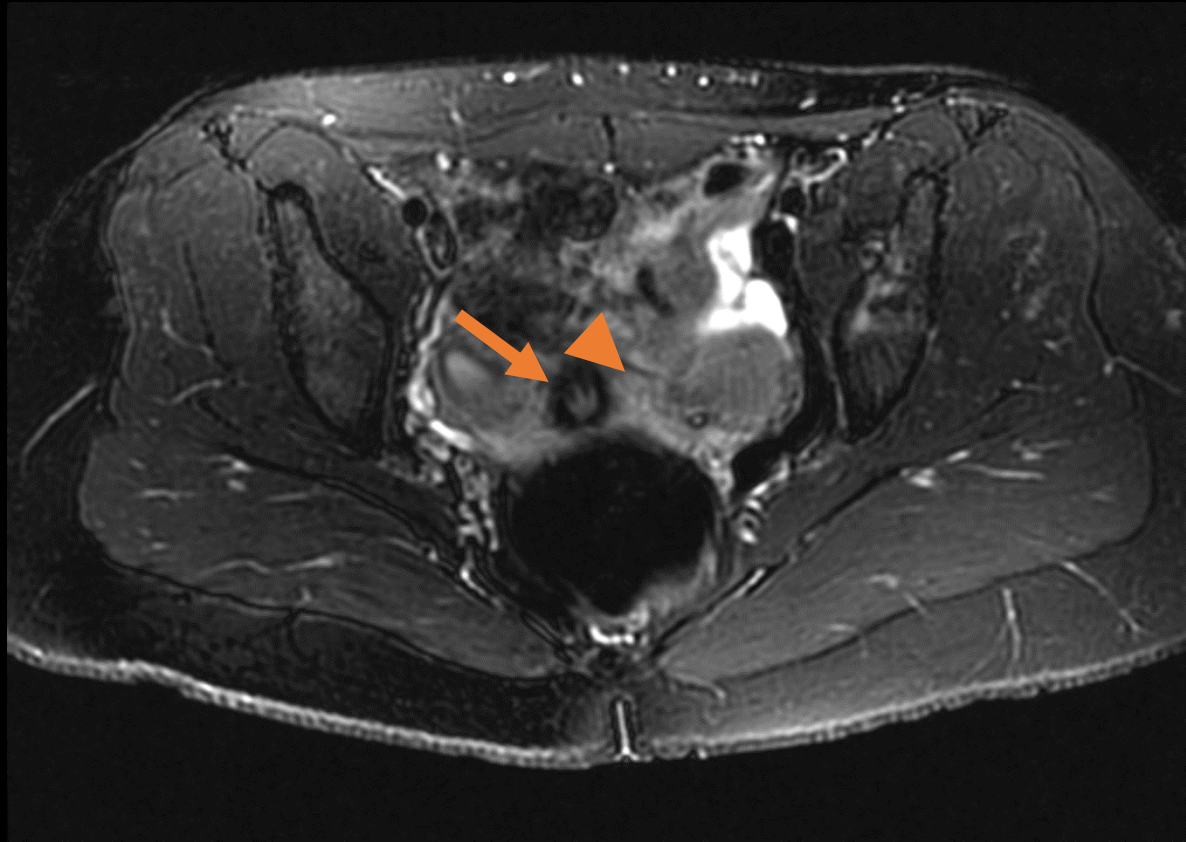


Coronal T2 image showing left renal agenesis.



Coronal oblique T1 pre-contrast showing left hematometra  
hemosalpinx

# Pertinent companion case (continued)



Ax T2 Fat Sat shows normal right cervix (arrow) and rudimentary left cervix (arrowhead).



Coronal oblique T2 shows obstructed rudimentary left horn (arrowhead) and cervix compared to the right horn (arrow) and cervix.

# Case Discussion

- Presenting symptoms of OHVIRA are non-specific, making it a difficult condition to diagnose.
- In evaluating OHVIRA, MRI provides excellent anatomic detail to allow accurate diagnosis without the use of invasive laparoscopic procedures.
- Delay in diagnosis can risk further complications, including pelvic infection and infertility.
- Although there is no definitive cure, surgical treatment includes a vaginoplasty.
- Management of potential complications includes drainage of blood from the hemivagina, antibiotics for infections, and surgery if necessary.
- Patients with OHVIRA should consider Cesarean section delivery to avoid unnecessary risk to the fetus during labor.

# References:

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2. Jha, S. & Abdi, S. Diverse Presentations of OHVIRA Syndrome: A Case Series. *Clin. Pediatr. (Phila.)* **60**, 6–8 (2021).
3. Bajaj, S. K., Misra, R., Thukral, B. B. & Gupta, R. OHVIRA: Uterus didelphys, blind hemivagina and ipsilateral renal agenesis: Advantage MRI. *J. Hum. Reprod. Sci.* **5**, 67–70 (2012).
4. Majmudar, A. & Cohen, H. L. “Lying-Down” Adrenal Sign: There Are Exceptions to the Rule Among Fetuses and Neonates. *J. Ultrasound Med.* **36**, 2599–2603 (2017).
5. American College of Radiology. ACR Appropriateness Criteria. Acute Nonlocalized Abdominal Pain. Revised 2018