

RADY 401 Case Presentation

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Focused patient history and workup

- 5 hour old male born at 40w3d via uncomplicated SVD to G2P1011 mom with pregnancy complicated by fetal atrioventricular septal defect and dextrocardia on prenatal echo
- APGARS 8 and 9, no resuscitation required
- Vitals: T 37.2°C, HR 176, RR 60, BP 54/46 with MAP 50, SaO2 95%
- Physical exam with mild acrocyanosis and fixed split S2 with heart sounds primarily heard on right. Otherwise warm and well perfused with lungs CTAB and no respiratory distress. Pulses 2+ bilaterally. Abdominal exam benign. Neurologically intact.
- Normal karyotype and prenatal / postnatal microarray
- Admitted to NICU for further workup and management given inadequate visualization of dextrocardia and AVSD on prenatal echo

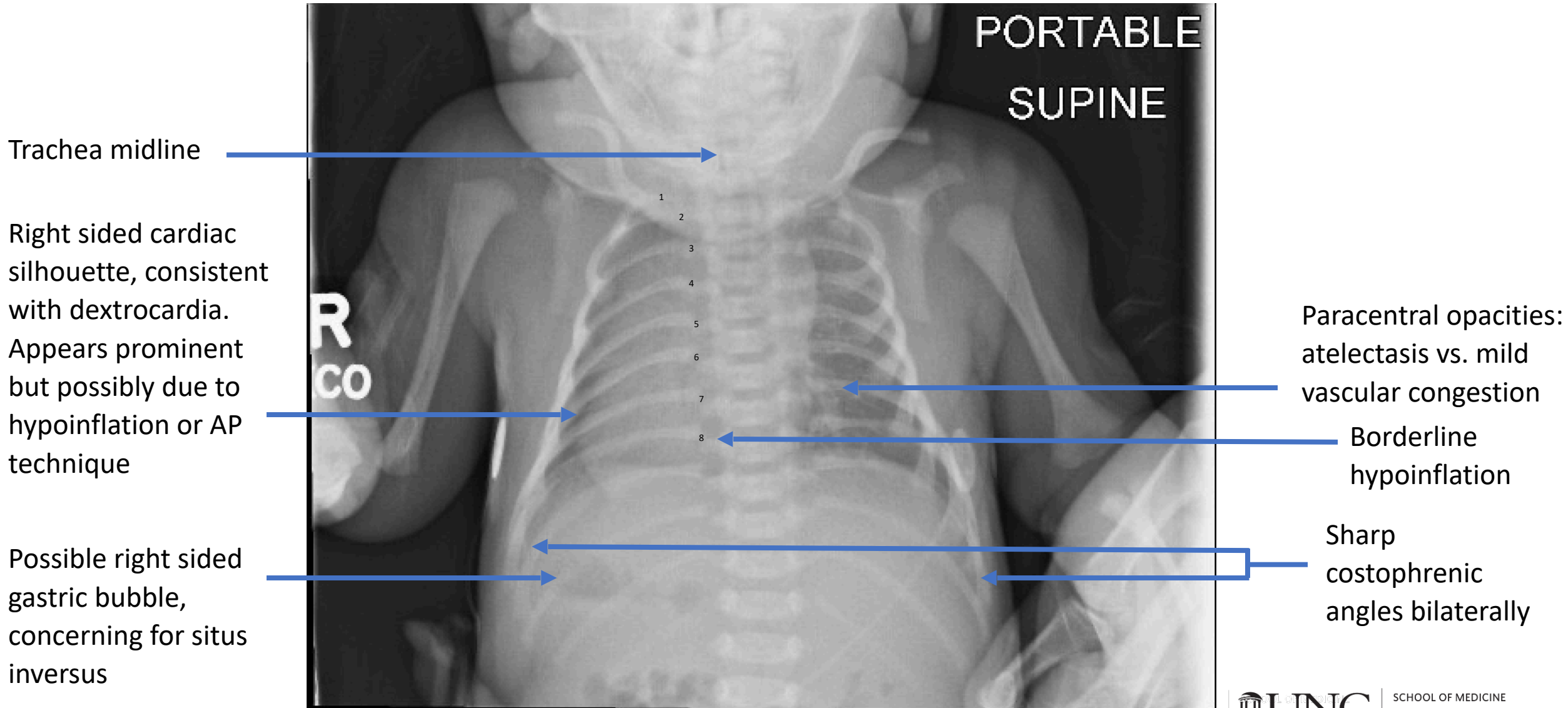
Dextrocardia

- Isolated dextrocardia (rare)
 - No associated vital organ abnormalities
- Dextrocardia with partial situs inversus
 - Some organs are on opposite side of body but usually function normally
- Dextrocardia with situs inversus totalis
 - All vital organs in chest and abdomen are on opposite side of the body and may be abnormally functioning
- Dextrocardia with heterotaxy
 - Some or all vital organs are misplaced and possibly absent or partially developed. Commonly associated with cardiac malformations and considerable morbidity and mortality
- Dextrocardia with Kartagener syndrome
- Associated congenital heart defects
 - Double outlet right ventricle, Endocardial cushion defect, Pulmonary stenosis, Tetralogy of Fallot, Transposition of the great arteries, Ventricular septal defect

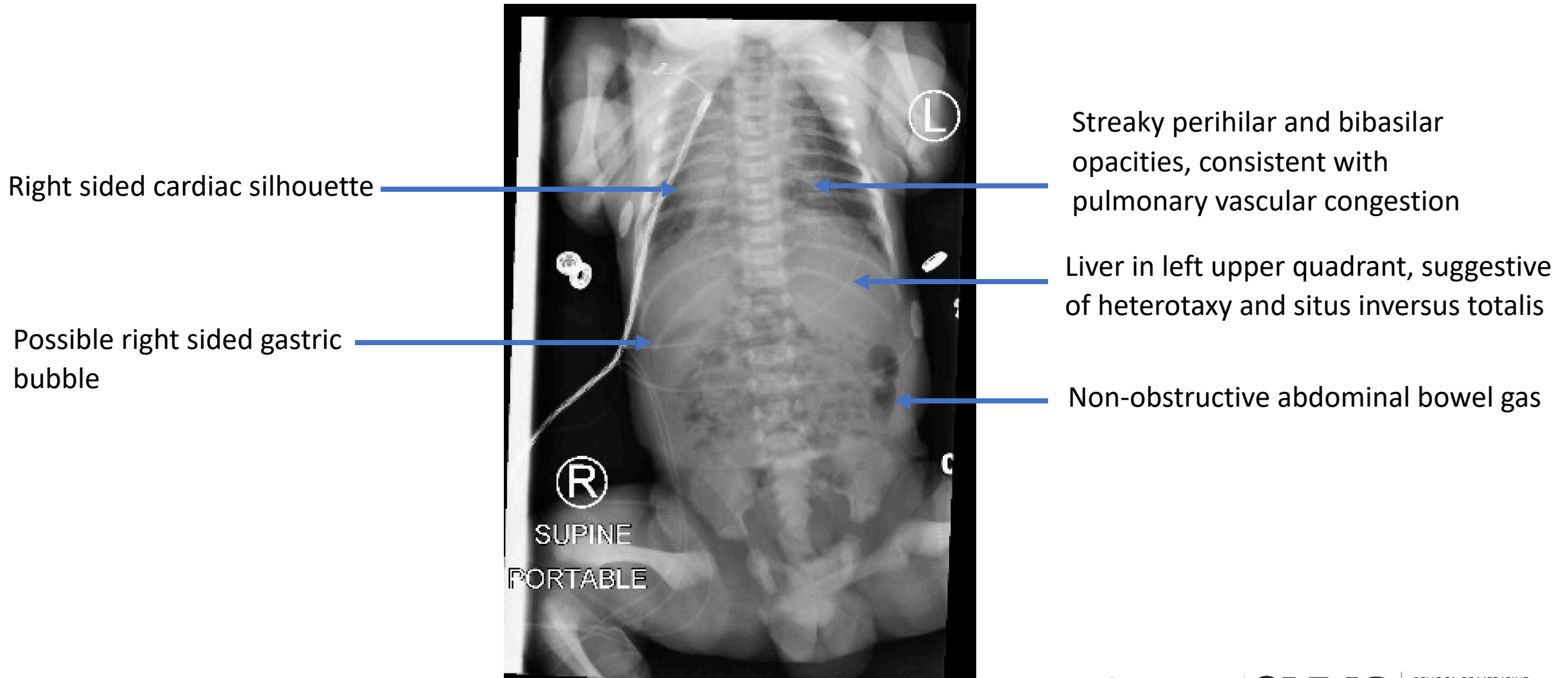
List of imaging studies

- Chest radiograph
- Abdominal radiograph
- Abdominal ultrasound
- Upper GI series

Imaging studies from PACS 1: AP supine portable CXR

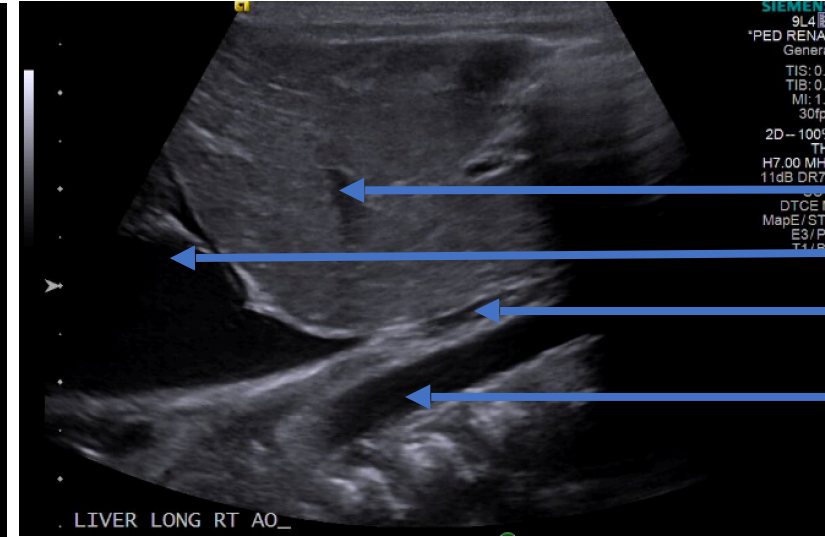
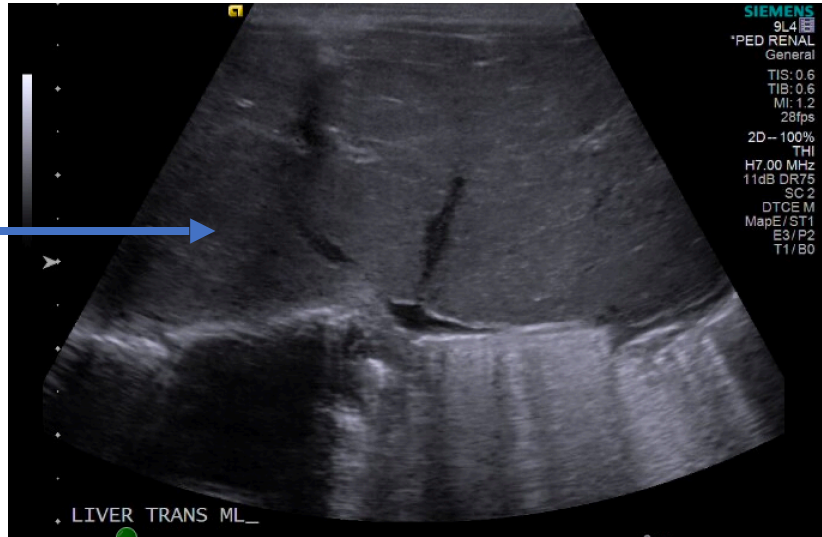


Imaging studies from PACS 2: AP supine portable AXR



Imaging studies from PACS 3: Abdominal ultrasound

Enlarged liver, mostly present in LUQ but crosses midline and present to a lesser extent in RUQ, more consistent with heterotaxy than situs inversus totalis



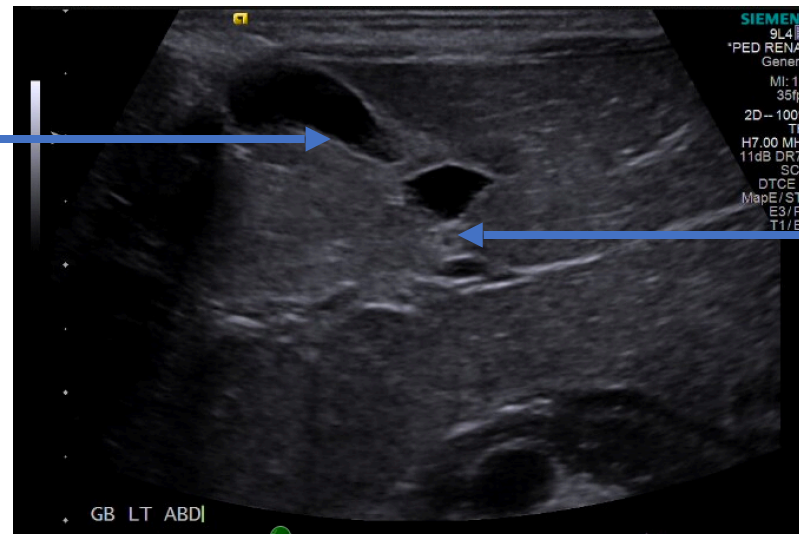
Hepatic veins

Right atrium

IVC

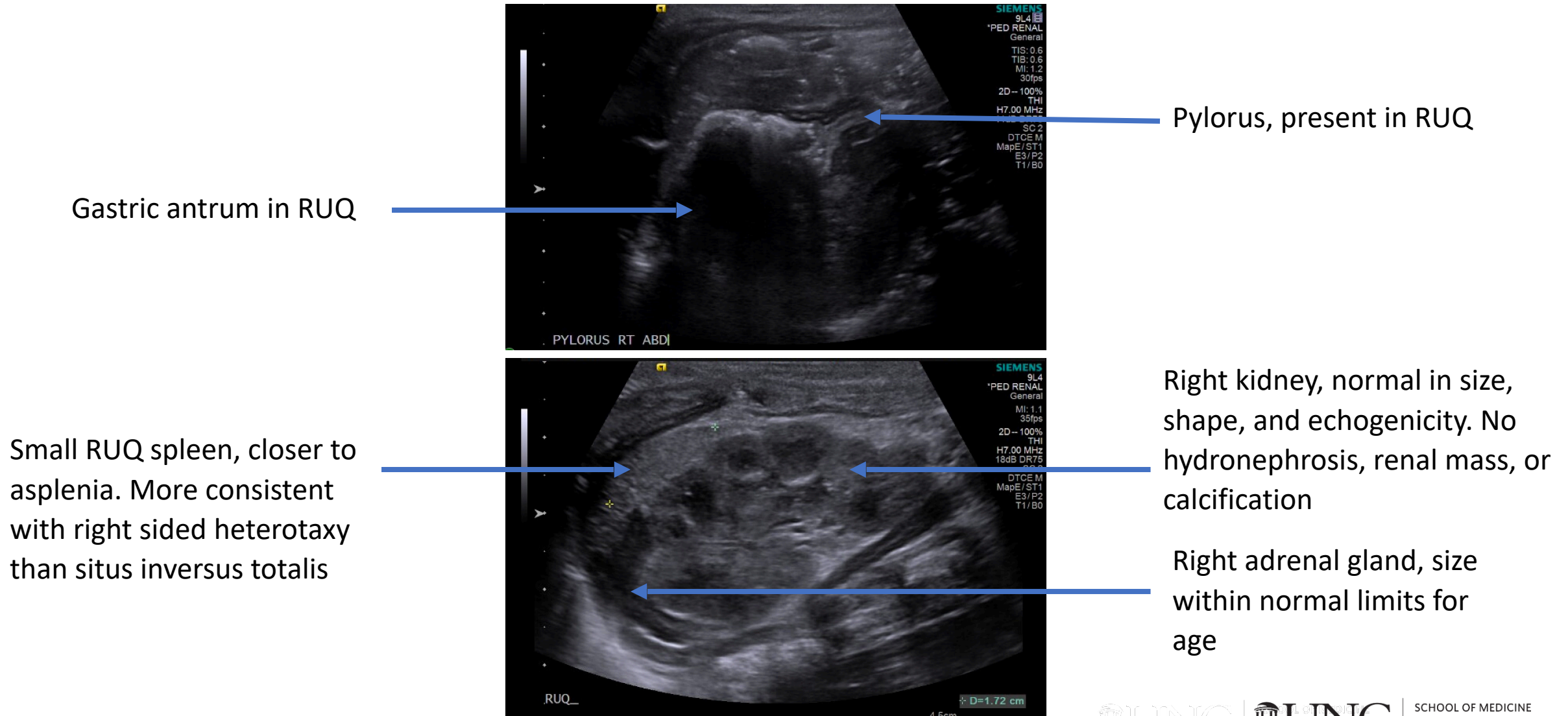
Aorta

Left sided gallbladder, no intraluminal gallstones or sludge



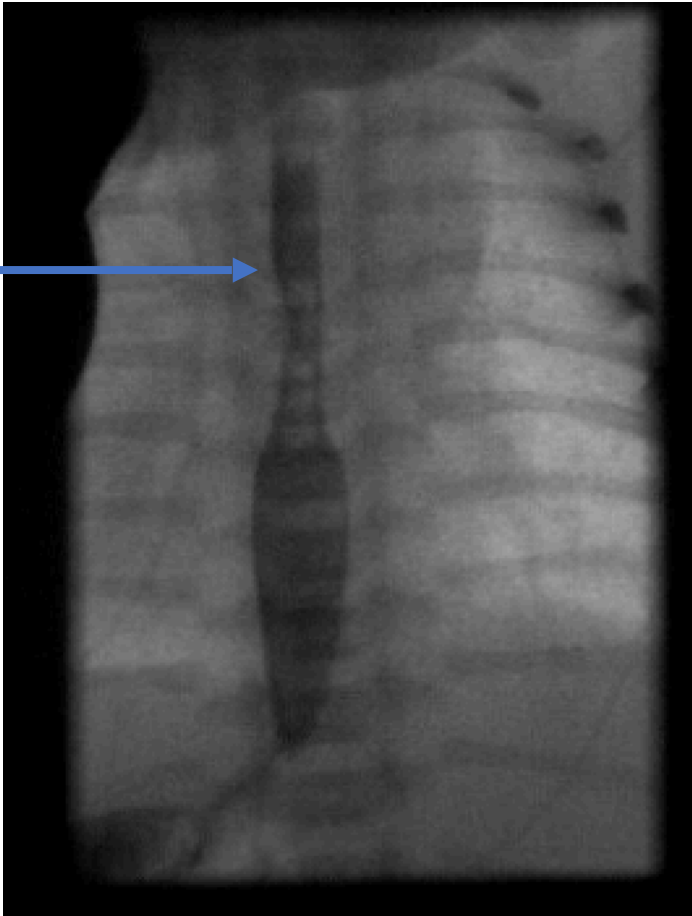
Common bile duct without dilation

Imaging studies from PACS 3: Abdominal ultrasound



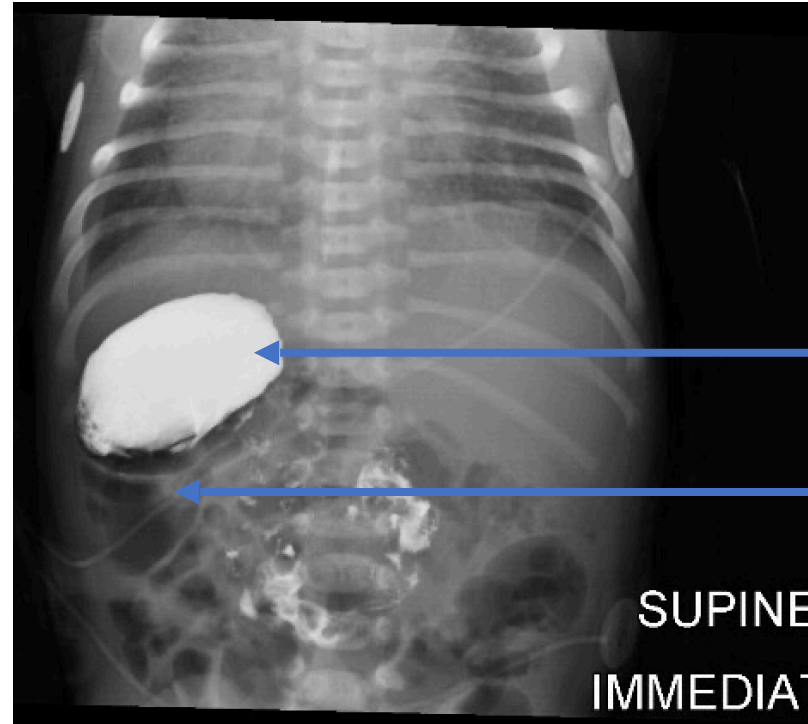
Imaging studies from PACS 4: Upper GI series with fluoroscopy and radiography

Esophagus with normal contour and peristalsis. No gastroesophageal reflux



Stomach in right hemiabdomen, empties readily

Duodenum and duodenojejunal junction in right hemiabdomen, consistent with malrotation. No evidence of obstruction or volvulus



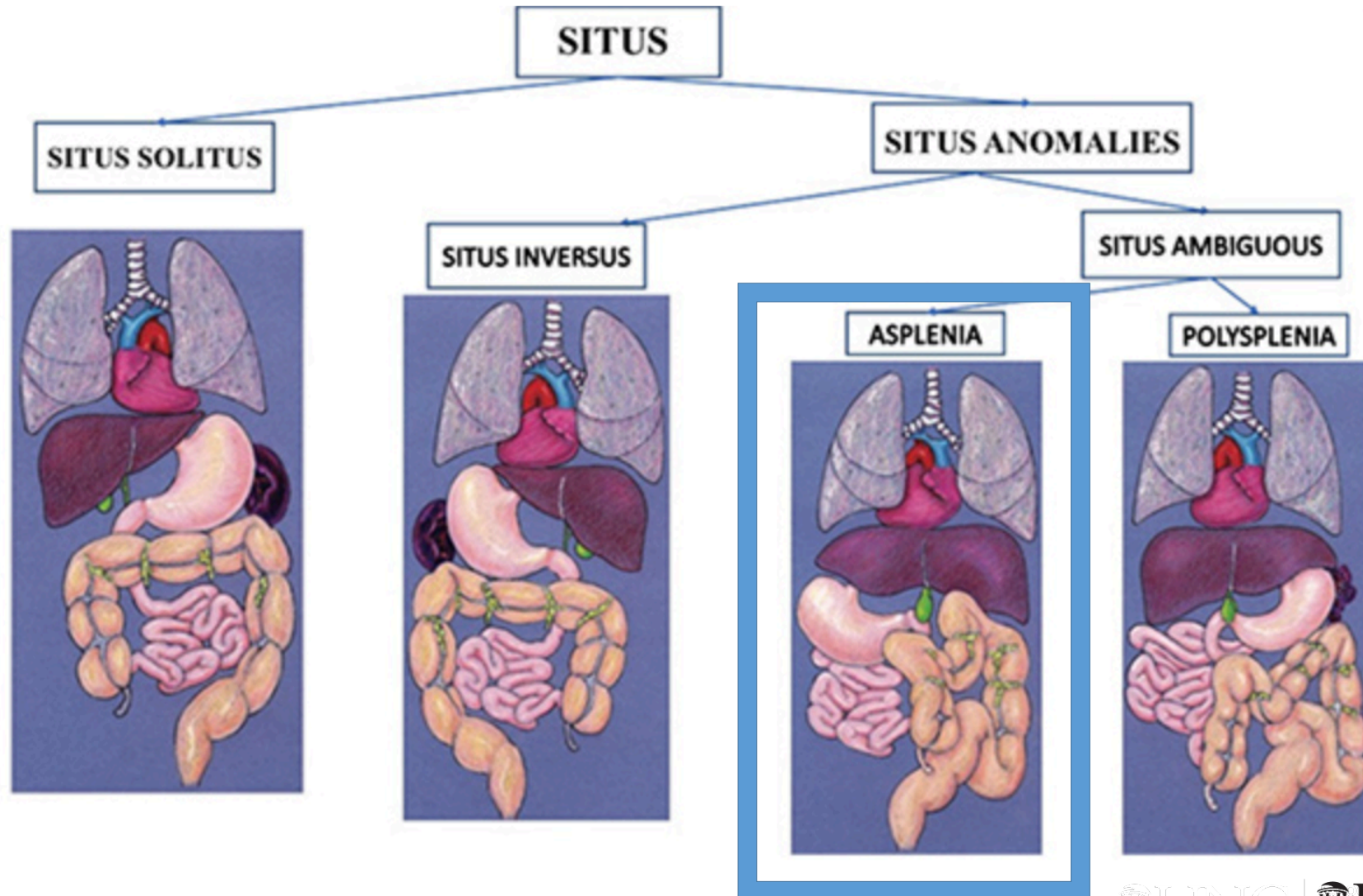
Patient treatment or outcome: Heterotaxy with malrotation

- Echo (not shown here): Hypoplastic LV and aorta, complete AV canal defect with common AV valve, TAPVR, double outlet RV, dilated RV. Normal RV and LV systolic function
- Diagnosed with heterotaxy with malrotation given dextrocardia with dextroversion, TAPVR, complete atrioventricular canal defect, right sided stomach, intestinal malrotation, RUQ spleen functionally asplenic, hepatomegaly with left sided liver, LUQ gallbladder
 - Complex congenital heart defects -> Lasix and staged cardiac reconstruction
 - Palliative pulmonary artery banding to decrease pulmonary circulation with ultimate goal of single ventricle circulation
 - TAPVR repair
 - Complete atrioventricular septal defect repair and pulmonary artery debanding (to provide normal 2 ventricle circulatory system)
 - Malrotation with risk of volvulus-> Ladd's procedure
 - Functional asplenia -> Amoxicillin prophylaxis
 - Severe GERD with feeding difficulty -> Temporary NG / PEG tube feeds
 - Dextrocardia of unknown etiology -> Pulmonology referral for workup of possible primary ciliary dyskinesia

Heterotaxy

- Heterotaxy: Abnormal arrangement of internal thoracic – abdominal organs across the left-right axis of the body. Commonly associated with cardiac malformations and considerable morbidity and mortality
 - Right atrial isomerism: duplication of right sided structures with absence of left sided structures (two right atria, hepatomegaly, asplenia)
 - Left atrial isomerism: duplication of left sided structures with absence of right sided structures (two left atria, discontinuity of IVC, polysplenia)
- Usually suspected prenatally with routine antenatal screening ultrasound and confirmed with fetal echocardiography
- Presentation at birth varied from asymptomatic to gravely ill with cyanosis depending on severity of cardiac malformations
- Management
 - Surgical cardiac reconstruction
 - Medical management to optimize cardiac function and pulmonary blood flow

Heterotaxy



Imaging discussion 1: Chest radiograph

Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
ICU patient, newly admitted, initial imaging	3084051	Radiography chest portable	<0.1 mSv ⊕	Not Assigned	Usually appropriate	●



Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Congenital coronary artery abnormality, TTE inadequate assessment of coronary morphology, next imaging study	3196447	US echocardiography transesophageal	0 mSv O	0 mSv [ped] O	Usually not appropriate	●
		Radiography chest	<0.1 mSv ⊕	<0.03 mSv [ped]..	Usually not appropriate	●
		MRA neck without IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate	●



- Appropriateness: unclear
- True indication: dextrocardia, confirmed congenital cardiac malformation on prenatal echo – initial postnatal imaging
- Sensitivity / specificity of chest radiograph for dextrocardia: no data
- Cost: ~\$55 (operating cost), cost to patient largely varied based on geographic location, imaging center, and insurance
- Radiation: <0.03 mSv

Imaging discussion 2: Abdominal radiograph

Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Vomiting, poor feeding, initial imaging Vomiting within the first 2 days after birth. Poor feeding or no passage of meconium. Initial imaging.	3191958	Radiography abdomen	0.1-1mSv ☼☼	0.03-0.3 mSv [ped]..	Usually appropriate	●
		US abdomen (UGI tract)	0 mSv O	0 mSv [ped] O	Usually not appropriate	●
		Fluoroscopy contrast enema	1-10 mSv ☼☼☼	3-10 mSv [ped]..	Usually not appropriate	●
		Fluoroscopy upper GI series	1-10 mSv ☼☼☼	0.3-3 mSv [ped]..	Usually not appropriate	●
		Nuclear medicine gastroesophageal reflux scan	Not Assigned	0.3-3 mSv [ped]..	Usually not appropriate	●



- Appropriateness: unclear
- True indication: dextrocardia with concern for visceral situs inversus, poor feeding without vomiting at 1 day of life
- Sensitivity / specificity of abdominal radiograph for situs inversus: no data
- Cost: ~\$55 (operating cost), cost to patient largely varied based on geographic location, imaging center, and insurance
- Radiation: 0.03-0.3 mSv

Imaging discussion 3: Abdominal ultrasound

Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Vomiting, poor feeding, initial imaging Vomiting within the first 2 days after birth. Poor feeding or no passage of meconium. Initial imaging.	3191958	Radiography abdomen	0.1-1mSv ☼☼	0.03-0.3 mSv [ped]..	Usually appropriate	●
		US abdomen (UGI tract)	0 mSv ○	0 mSv [ped] ○	Usually not appropriate	●
		Fluoroscopy contrast enema	1-10 mSv ☼☼☼	3-10 mSv [ped]..	Usually not appropriate	●
		Fluoroscopy upper GI series	1-10 mSv ☼☼☼	0.3-3 mSv [ped]..	Usually not appropriate	●
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Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Vomiting, bilious, malrotation suspected, initial imaging	3185451	Fluoroscopy upper GI series	1-10 mSv ☼☼☼	0.3-3 mSv [ped]..	Usually appropriate	●
		US abdomen (UGI tract)	0 mSv ○	0 mSv [ped] ○	May be appropriate	●
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- Appropriateness: unclear
- True indication: dextrocardia and situs inversus / heterotaxy with concern for malrotation. Poor feeding without emesis
- Pooled sensitivity 94% (82%-100%), pooled specificity 100% (55%-100%) for malrotation with or without midgut volvulus on abdominal ultrasound
- Cost: \$260 but varies based on location, imaging institution, and insurance
- Radiation: 0 mSv

Imaging discussion 4: Upper GI series

Scenario	Scenario Id	Procedure	Adult RRL	Peds RRL	Appropriateness Category	
Vomiting, bilious, malrotation suspected, initial imaging	3185451	Fluoroscopy upper GI series	1-10 mSv ⊗⊗⊗	0.3-3 mSv [ped]..	Usually appropriate	●
		US abdomen (UGI tract)	0 mSv ○	0 mSv [ped] ○	May be appropriate	●
		Radiography abdomen	0.1-1mSv ⊗⊗	0.03-0.3 mSv [ped]..	May be appropriate	●
		Fluoroscopy contrast enema	1-10 mSv ⊗⊗⊗	3-10 mSv [ped]..	Usually not appropriate	●
		Nuclear medicine gastroesophageal reflux scan	Not Assigned	0.3-3 mSv [ped]..	Usually not appropriate	●



- Appropriateness: likely appropriate
- True indication: dextrocardia and heterotaxy with concern for malrotation. Poor feeding without emesis
- Pooled sensitivity 91% (40%-100%), pooled specificity 94% (20%-100%) for malrotation on upper GI series
- Cost: \$600 but varies based on location, imaging institution, and insurance
- Radiation: 0.3-3 mSv

UNC Top Three (or Test Yourself or Wrap Up)

- Heterotaxy is the abnormal arrangement of internal thoracic – abdominal organs across the left-right axis of the body and is associated with significant morbidity and mortality due to associated cardiac malformations
- Heterotaxy is usually suspected prenatally with screening ultrasound and confirmed with fetal echocardiogram
- Management of heterotaxy depends on severity but commonly involves medical therapy and staged cardiac reconstruction

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