

RADY 403 Case Presentation

Pulmonary Embolus in a Patient with a Left-sided Duplicated IVC

Elizabeth O'Neil, MS4 | August 2019

Focused patient history and workup:

14-year-old male with a PMHx significant for a left clavicular fracture (surgically corrected 6/13/19) admitted from OSH to UNC PICU for confirmed saddle pulmonary embolus on outside chest CTA with concomitant hypoxemic respiratory failure.

- 6/13/19: Sustained left mid-shaft clavicular fracture with shortening and displacement after being tackled by a friend. Underwent open internal fixation with plate placement and initially did well post-op.
- Night of 8/6/19: Experienced chest pain, tachycardia, SOB, and increased WOB on RA.
- Original Labs at OSH: Lactate 2.6, INR 1.4, D-dimer in the 4,000s.
- Chest x-ray and chest CTA performed at OSH confirmed saddle PE.
- Received 70 mg loading dose of Lovenox followed by Heparin and then prepared for transfer to UNC.

Focused patient history and workup:

- During transfer to UNC: SpO2 dropped to low 80s. Placed on nonrebreather.
- Stat ECHO at UNC revealed a patent pulmonary artery suggesting the clot may have embolized into distal pulmonary vessels.
- Initial PICU evaluation at UNC: Patient critically ill with acute dyspnea and hypoxemic respiratory failure secondary to saddle pulmonary embolus.
- PVLs at UNC 8/6 showed no evidence of DVT.
- 8/7/19 left leg cool and mottled in appearance with decreased pulses. Repeat PVLs revealed an acute DVT.
- 8/7/19: CT Pelvic Venogram revealed a duplicated left-sided IVC with thrombus extending from the inferior aspect of the duplicated IVC through the left external iliac, left femoral veins, and into the proximal left greater saphenous vein.
- Hematology sent labs for initial thrombophilia workup.

List of imaging studies:

- XR Chest PA and Lateral 8/6/19 (**Performed at OSH**)
- Chest CTA 8/6/19 (**Performed at OSH**)
- XR Chest Portable 8/6/19
- Transthoracic Echocardiogram 8/6/19
- Bilateral PVLs 8/6/19
- Bilateral PVLs 8/7/19
- CT Pelvic Venogram Kid to Fem 8/7/19

List of imaging studies:

Let's focus on the 4 imaging studies outlined below

- XR Chest PA and Lateral 8/6/19 (**Performed at OSH**)
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Patient with chest pain, tachycardia, SOB, and increased WOB on RA. D-dimer at OSH in the 4,000s

Variant 2: Suspected pulmonary embolism. Intermediate probability with a positive D-dimer or high pretest probability.

Radiologic Procedure	Rating	Comments	RRL*
X-ray chest	9		⊕
CTA chest with IV contrast	9	This procedure should be optimized for pulmonary circulation.	⊕⊕⊕
CT chest with IV contrast	9	This procedure should be optimized for pulmonary circulation. This procedure may be an alternative to CTA, but both should not be performed.	⊕⊕⊕
Tc-99m V/Q scan lung	7	This procedure may be an alternative to CTA, but both should not be performed.	⊕⊕⊕
US duplex Doppler lower extremity	7	This procedure may be an initial study prior to CTA.	○
MRA chest without and with IV contrast	6		○
CTA chest with IV contrast with CT venography lower extremities	5		⊕⊕⊕
Arteriography pulmonary with right heart catheterization	3		⊕⊕⊕⊕
US echocardiography transthoracic resting	3		○
CT chest without IV contrast	2		⊕⊕⊕
CT chest without and with IV contrast	2		⊕⊕⊕
MRA chest without IV contrast	2	This procedure has limited sensitivity and may be indicated for rare situations or certain contraindications for a specific patient.	○
US echocardiography transesophageal	2		○
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

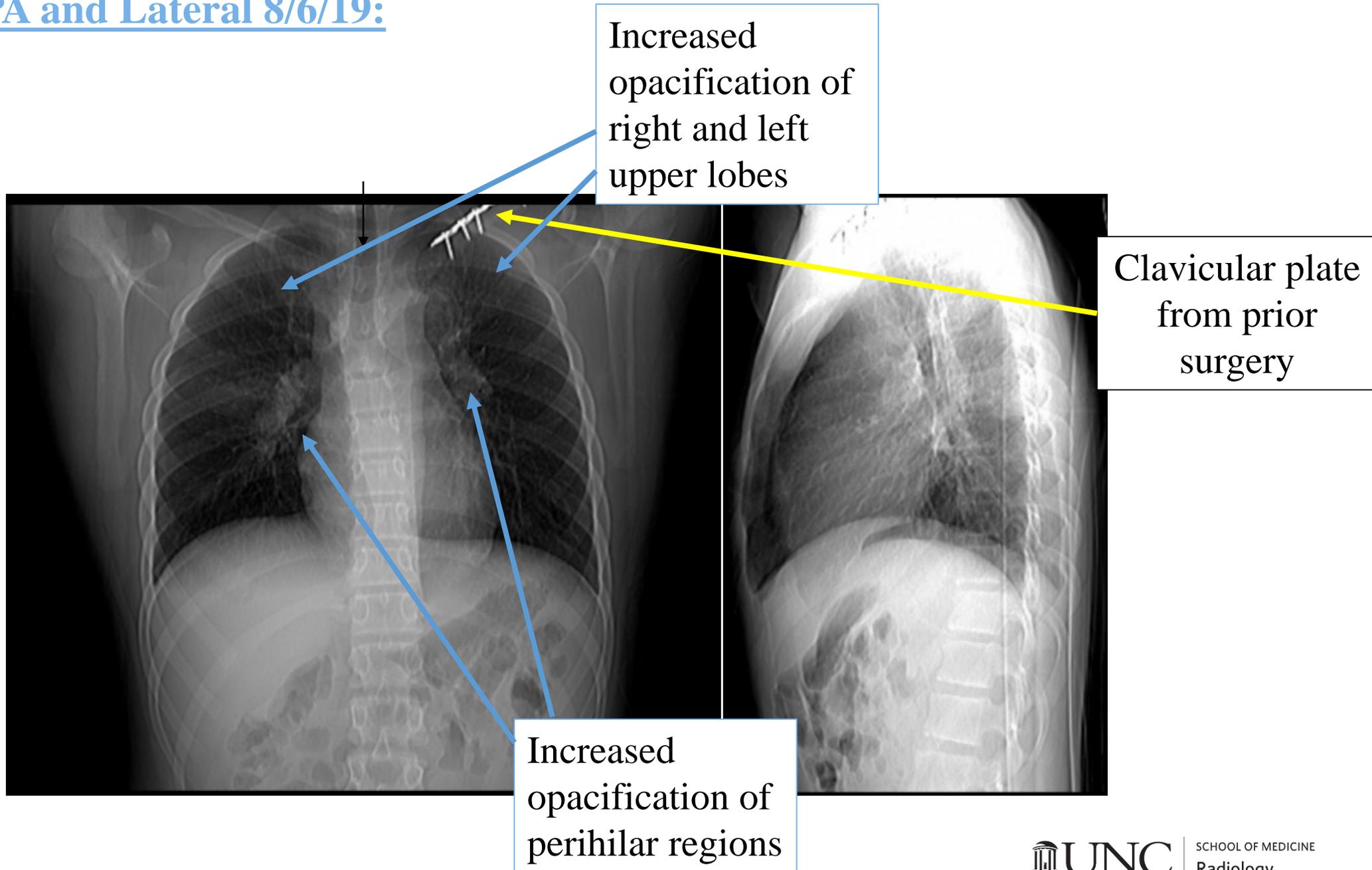
The patient's clinical presentation and positive d-dimer gave him a high pretest probability for a PE so a chest x-ray was performed, followed by a chest CTA².

The cost of a chest x-ray ranges from \$54-\$191, locally. Estimated national average cost = \$254³

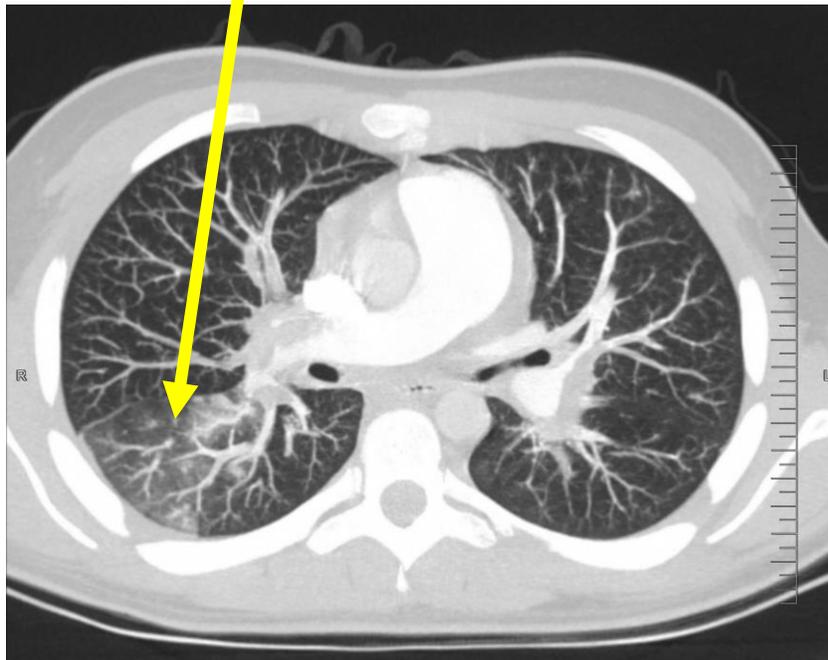
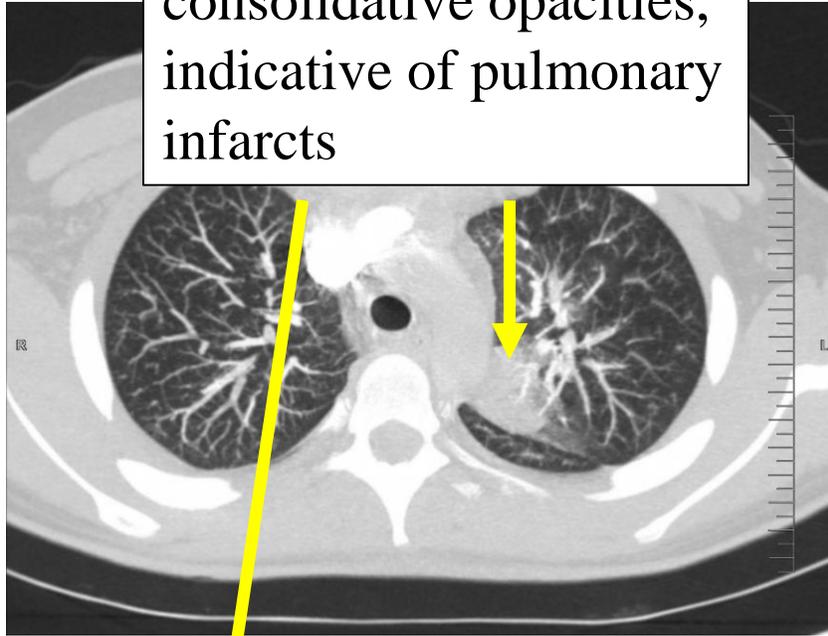
The cost of a CT angiogram ranges from \$326-\$799, locally. Estimated national average cost = \$896⁴

XR Chest PA and Lateral 8/6/19:

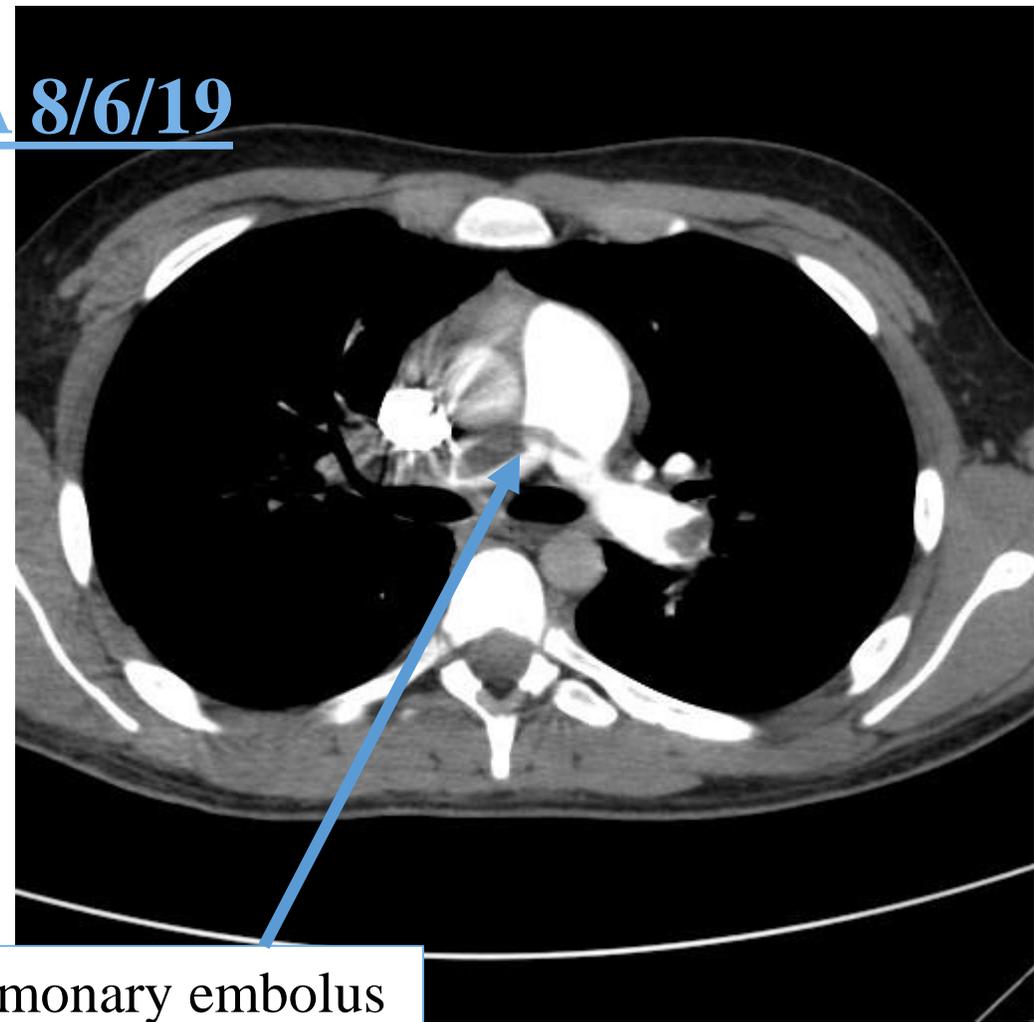
- Airway is midline and pt is centered
- No bone or soft tissue abnormalities
- Heart size is within a normal limit
- Lung parenchyma is well aerated
- Costophrenic angles are clear/no evidence of effusion



Wedge-shaped bilateral consolidative opacities, indicative of pulmonary infarcts

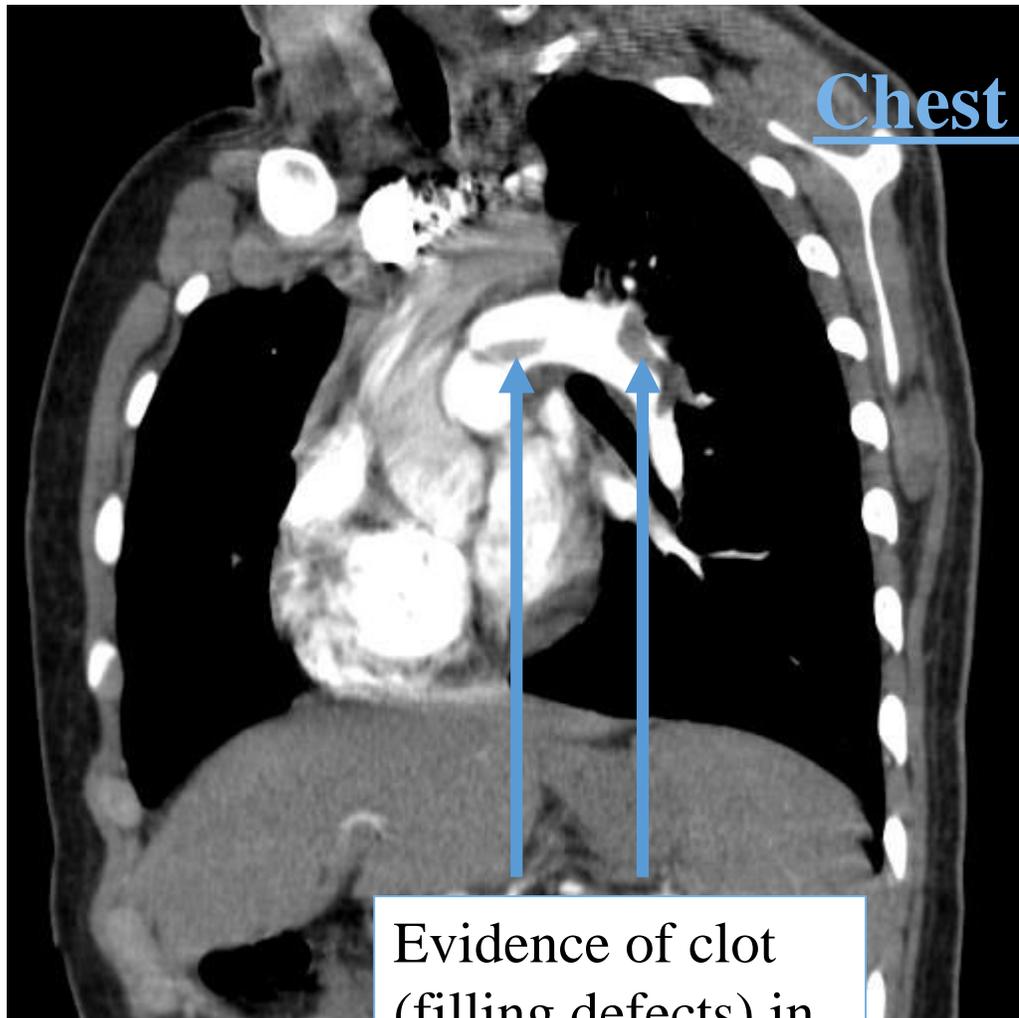


Chest CTA 8/6/19

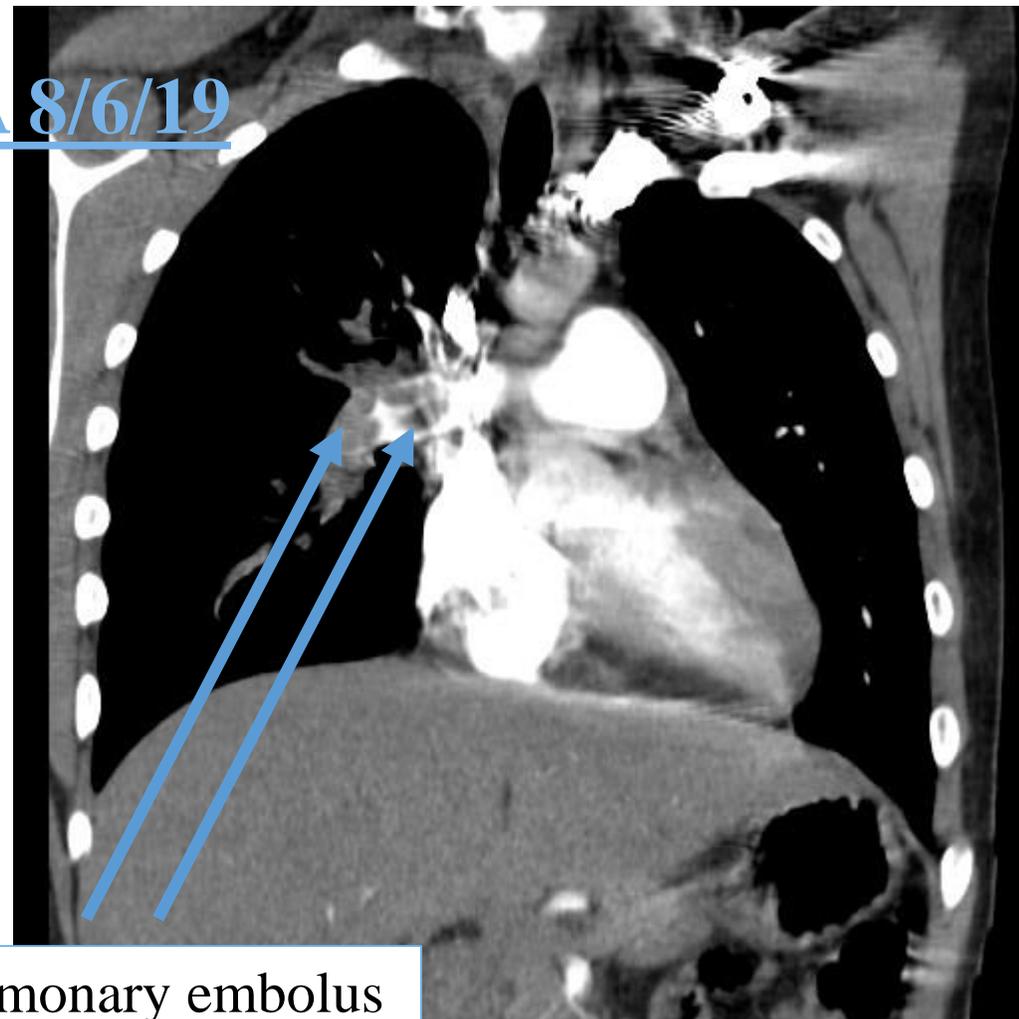


Pulmonary embolus (filling defects) extending into the right and left pulmonary arteries.

Chest CTA 8/6/19

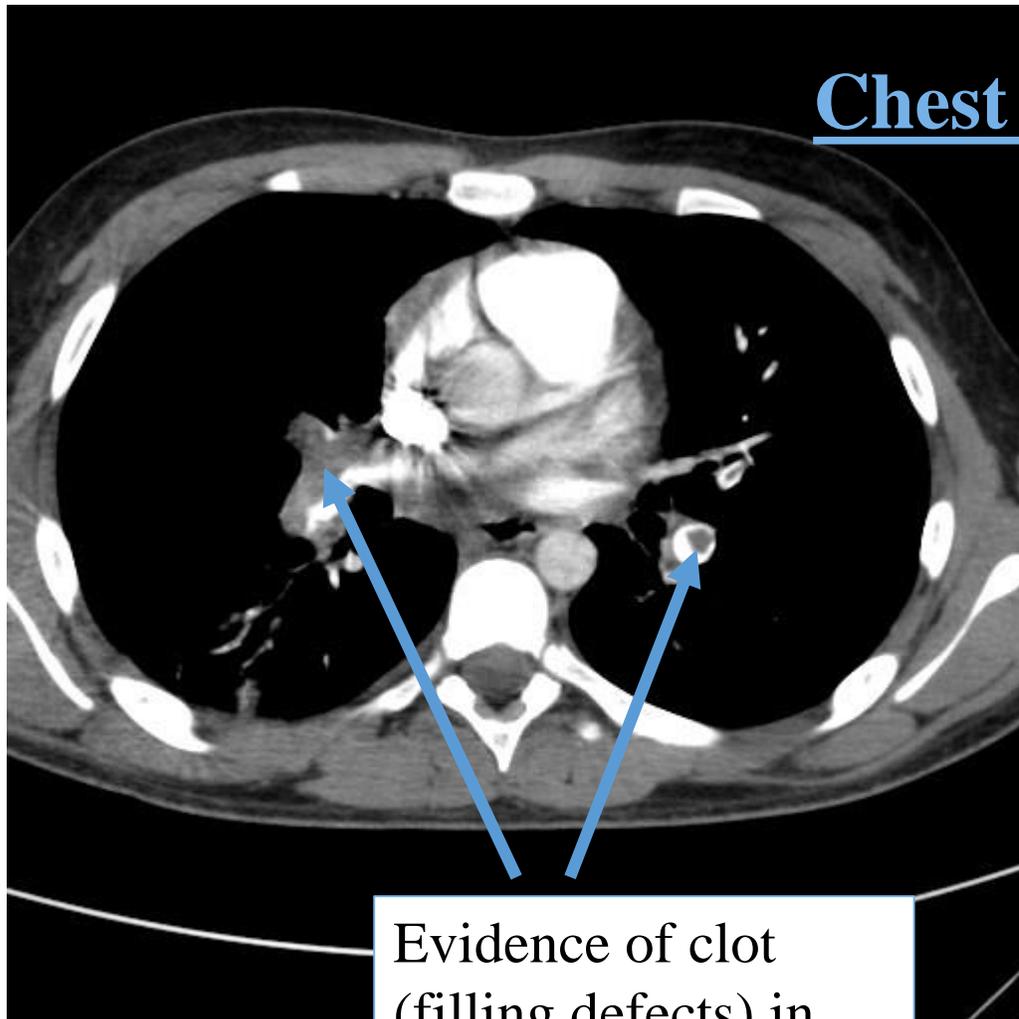


Evidence of clot (filling defects) in the main left pulmonary artery and its branches on LAO projection

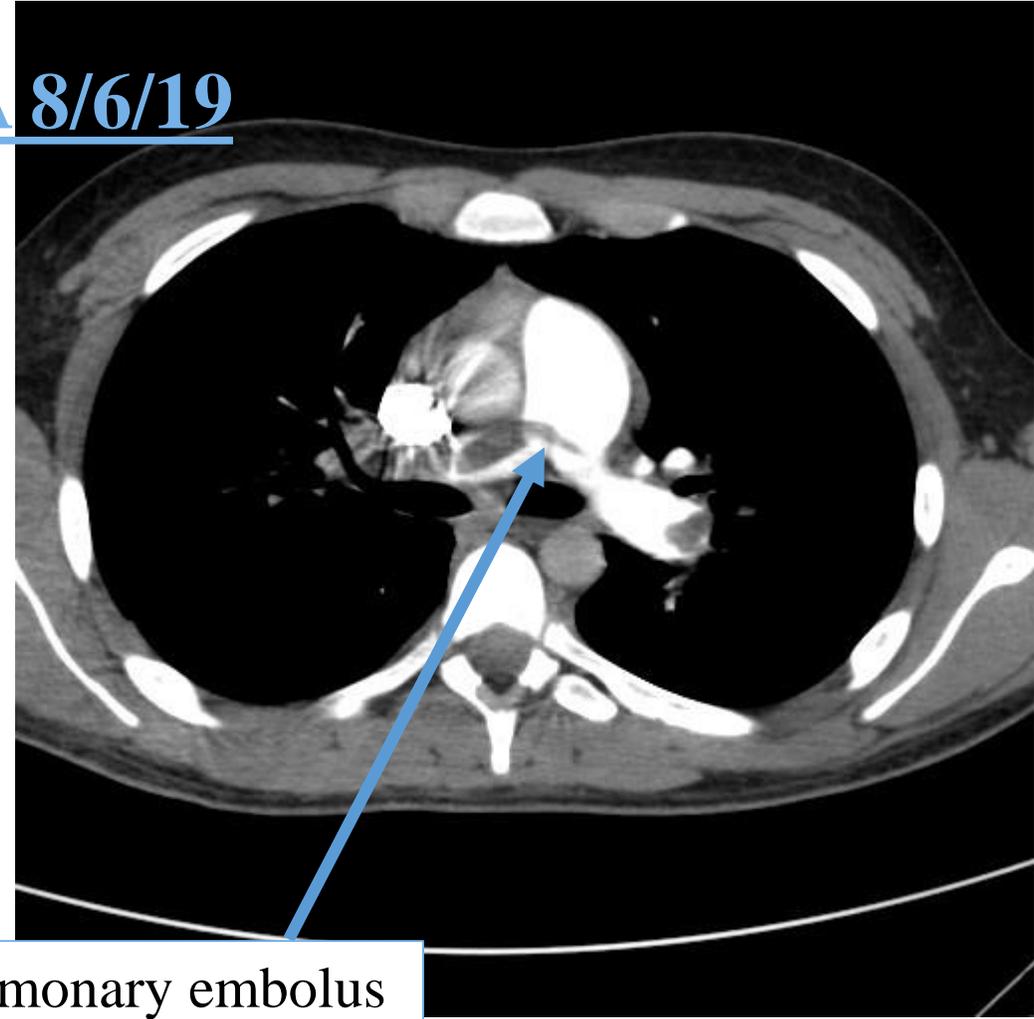


Pulmonary embolus (filling defects) in the main right pulmonary artery and its branches on coronal projection

Chest CTA 8/6/19



Evidence of clot (filling defects) in the right and left pulmonary artery branches on transverse projection

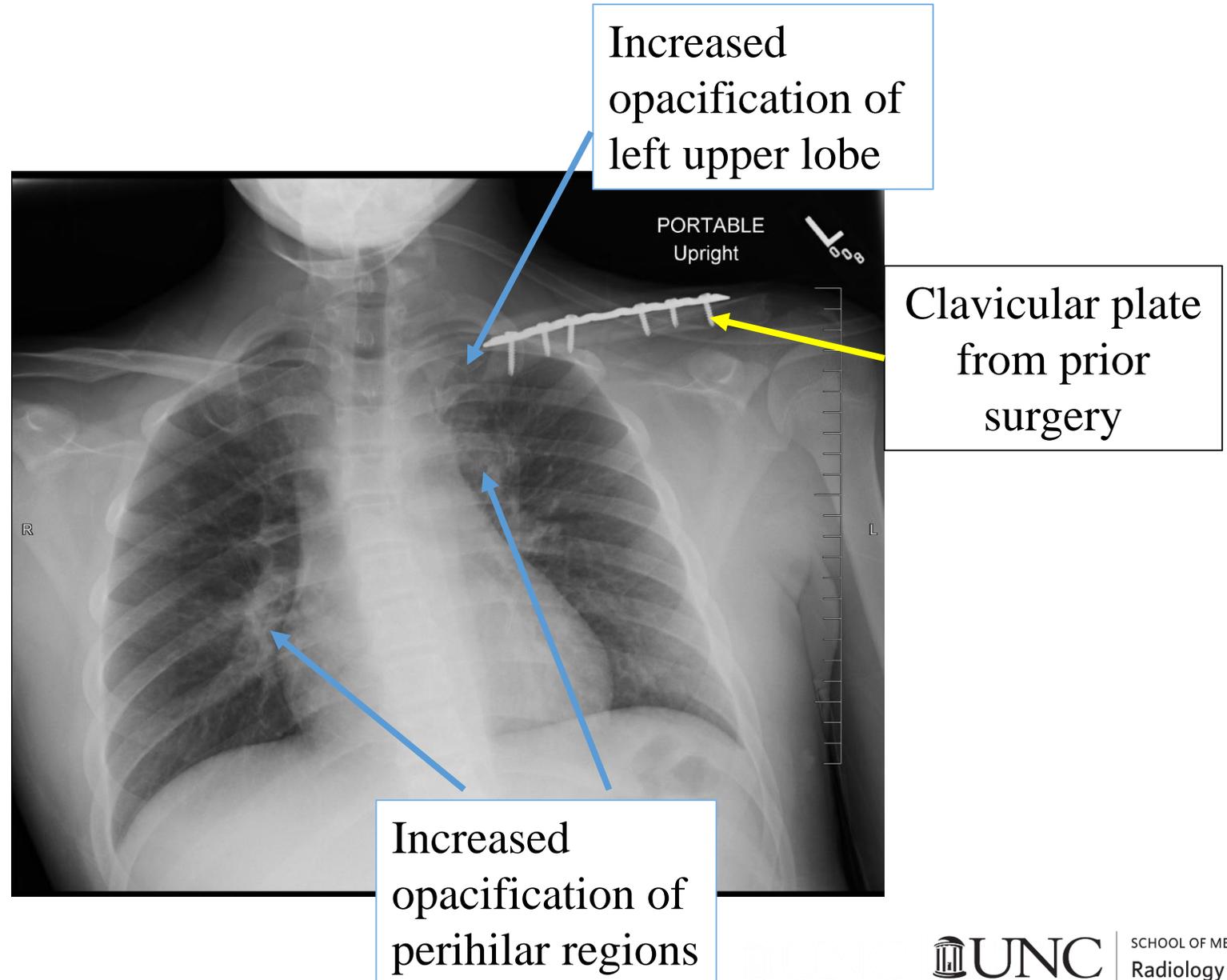


Pulmonary embolus (filling defect) that straddles both main pulmonary arteries is dubbed “saddle embolus”

XR Chest Portable AP 8/6/19:

Clinical Indication: 14-year-old male with hypoxemia and PE

- Airway is midline and pt is centered
- No bone or soft tissue abnormalities
- Heart size is within a normal limit
- Lung parenchyma is well aerated
- Costophrenic angles are clear/no evidence of effusion



8/7/19 left leg cool and mottled in appearance with decreased pulses

**American College of Radiology
ACR Appropriateness Criteria®
Sudden Onset of Cold, Painful Leg**

Variant 1: Sudden onset of cold, painful leg.

Radiologic Procedure	Rating	Comments	RRL*
Arteriography lower extremity	8	This procedure is the preferred option if clinical suspicion of acute arterial obstruction is intermediate to high.	☼ ☼ ☼
CTA lower extremity with IV contrast	7	Consider in place of arteriography if clinical suspicion of arterial obstruction is low and patient has a stable baseline eGFR ≥ 45 mL/min).	☼ ☼ ☼
MRA lower extremity without and with IV contrast	7	This procedure may help reduce total contrast dose. Consider prior to arteriography in patients with mild to moderate chronic kidney disease (GFR 30–89 mL/min).	0
MRA lower extremity without IV contrast	5	This procedure should be considered in patients with eGFR < 30 mL/min who are not yet on dialysis.	0
US duplex Doppler lower extremity	5	This procedure may be helpful for problem solving or targeted examinations (eg. bypass graft).	0
Rating Scale: 1,2,3 Usually not appropriate; 4,5,6 May be appropriate; 7,8,9 Usually appropriate			*Relative Radiation Level

Bilateral PVLs were performed, which revealed the presence of a left DVT. A CT Venogram was performed due to low suspicion for arterial involvement¹.

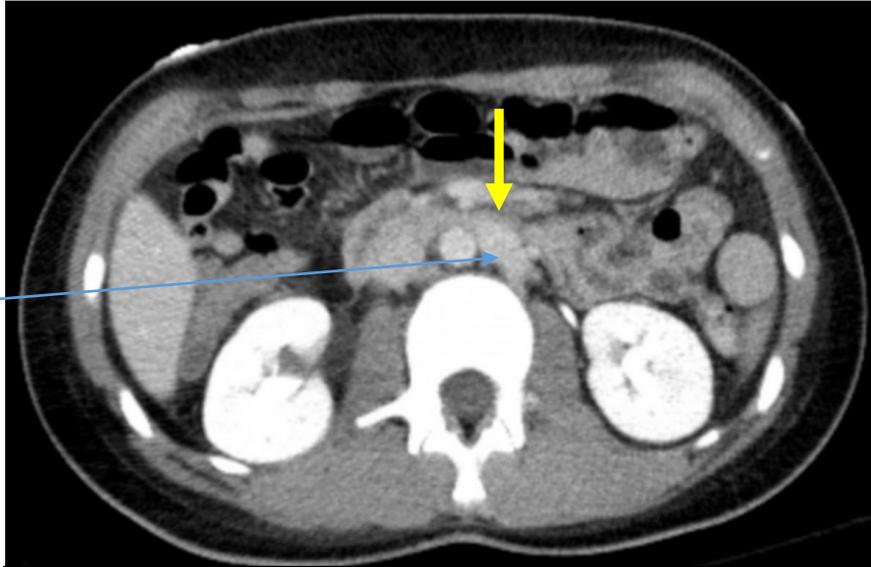
The cost of a bilateral venous doppler study ranges from \$217-518, locally¹⁰.

Estimated national average cost of CT venography is approximately \$2,583⁹.

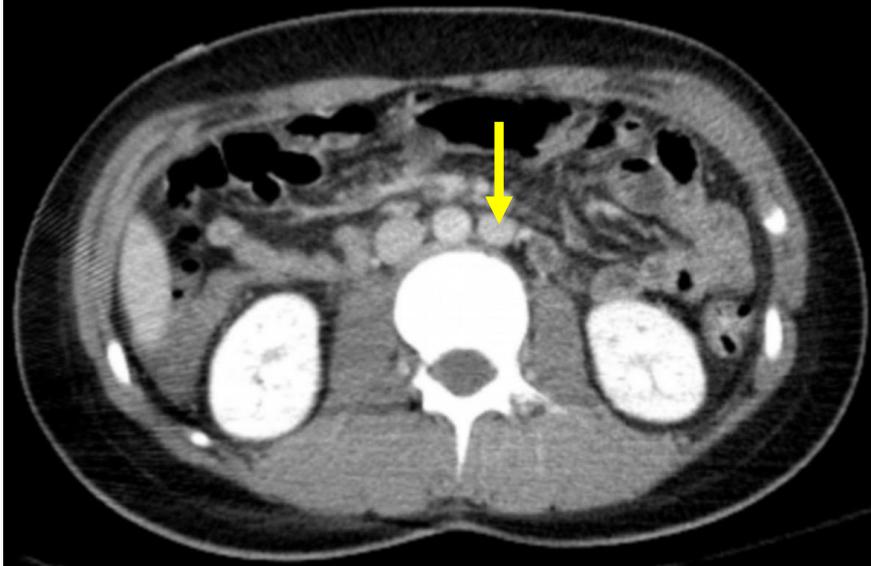
CT Pelvic Venogram Kid to Fem 8/7/19:

Clinical Indication: 14-year-old male with left common femoral vein occlusion

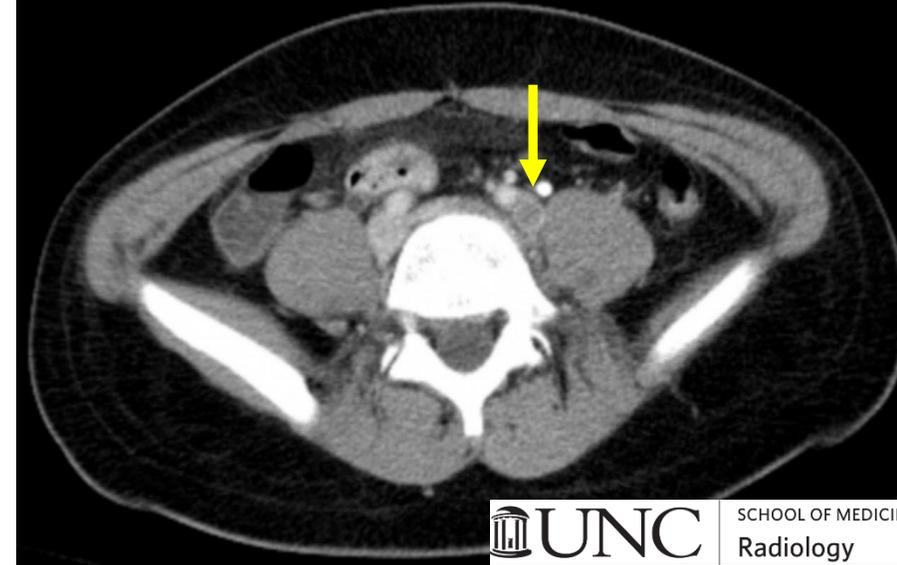
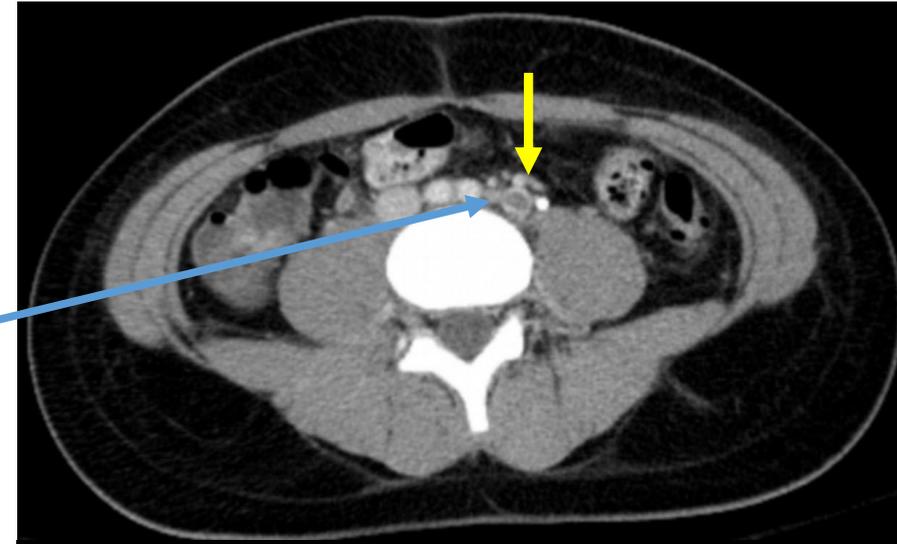
Left-sided duplicated IVC beginning to branch off from the left renal vein



Yellow arrows follow the course of the duplicated IVC



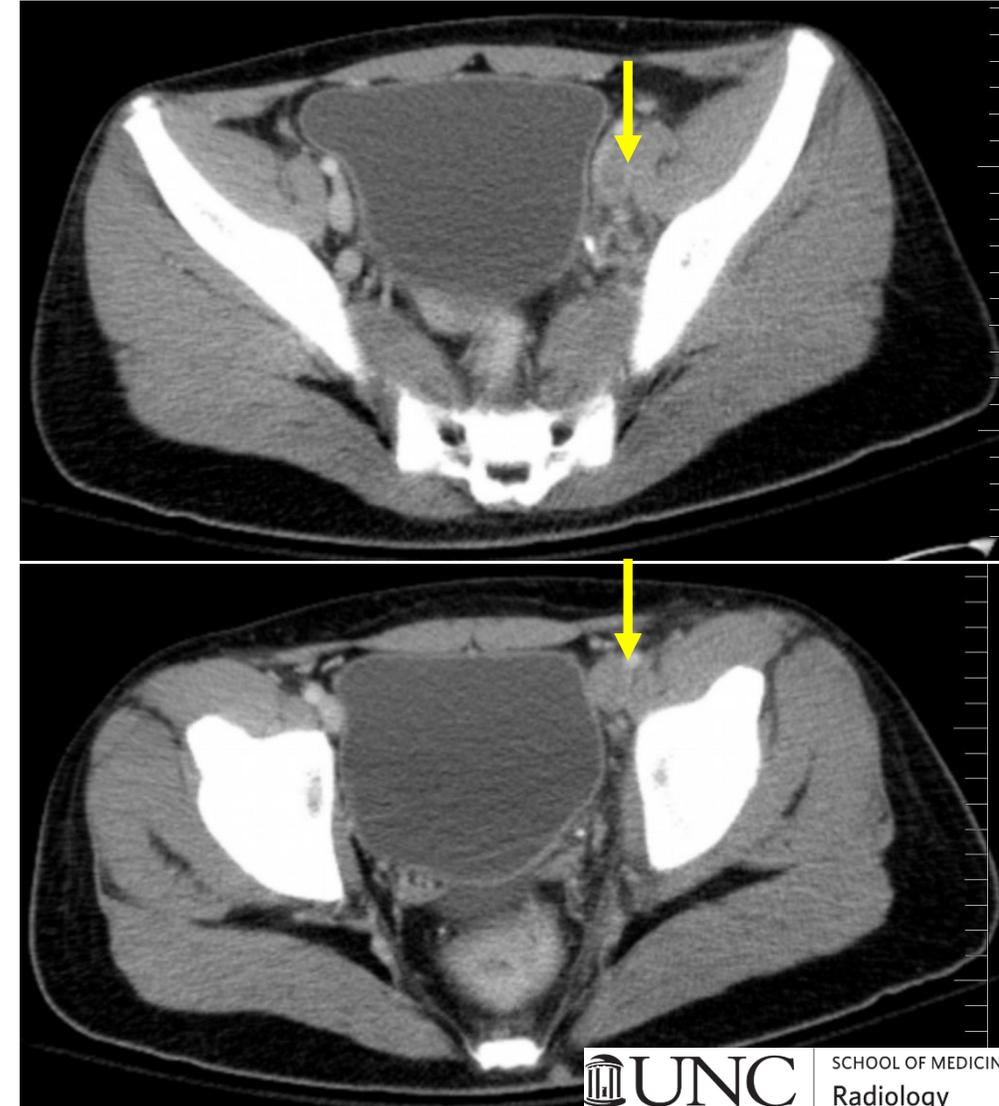
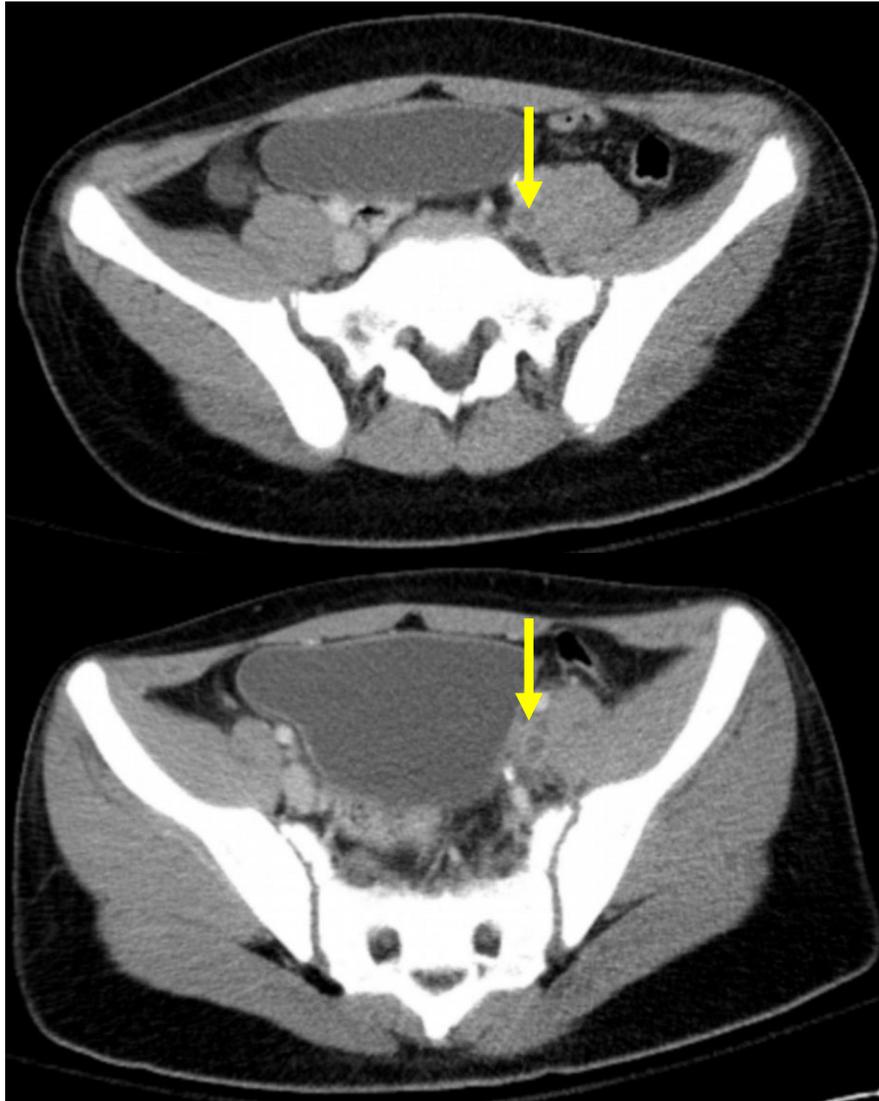
First evidence of thrombus formation in the left-sided duplicated IVC



CT Pelvic Venogram Kid to Fem 8/7/19 (Continued):

Clinical Indication: 14-year-old male with left common femoral vein occlusion

Yellow arrows follow the course of the duplicated IVC/thrombus



Risk of DVT and PE after Orthopedic Procedures in the Pediatric Population:

- Venous thromboembolic events (VTE) in pediatric patients is a rare condition which has not been well studied^{5,7}.
- Risk factors include: malignancy, infection, trauma, central venous catheter placement, operative procedures, and heritable prothrombotic disorders^{5,7}.
- A large (117,676 patients) retrospective prognostic study looked at the incidence of VTE in pediatric patients (<18 y.o.) who underwent elective orthopedic procedures⁵.
 - Incidence of VTE was found to be 0.0515%.
- Risk factors for VTE included admission type, obesity, diagnosis of metabolic conditions, increased age, and complications of surgical procedures and/or implanted devices ($p < .05$)⁵.

Inherited Thrombophilias:

- In pediatric patients with VTE, the most common inherited thrombophilias (ITs) are factor V Leiden, prothrombin G20210A mutation, antithrombin deficiency, and protein C and S deficiencies⁷.
- Identification of an IT during an acute VTE does not change management but may impact duration of therapy⁷.
 - Depends on the age of the patient and if event was or was not provoked
- In a pediatric patient with a VTE that is not central venous catheter related, it is recommended they undergo IT testing⁷.
 - Testing should include the conditions mentioned above as well as elevated homocysteine, elevated factor VIII, and antiphospholipid antibodies
 - Many non-DNA-based tests are affected by acute thrombosis and need to be confirmed with repeat testing if they are abnormal.

Duplicated IVC:

- Occurs when the left subcardinal and/or left supracardinal veins fail to regress in embryogenesis (occurs between weeks 6 and 8)^{6,8}.
- Thought to affect 0.2% - 3% of the population^{6,8}.
- Can be a risk factor for venous thromboembolism 2/2 retrograde stasis⁸.
- Over the past 100 years, there have been less than 10 reported cases involving duplicated IVC in association with venous thromboembolism⁸.
 - Age of first presentation of first thrombosis in most reports is less than 35 years of age.

Patient treatment or outcome:

- Patient's interdisciplinary care team did not think his DVT/PE was due to his duplicated left-side IVC.
- Per pediatric hematology/oncology: Patient likely had thrombus in iliac vein, which was not detected on PVLs 8/6. Thrombus likely enlarged on 8/7.
 - Patient was found to have mild heparin resistance in the setting of low antithrombin III levels.
- Patient became hemodynamically stable and tolerated room air in the PICU.
- Repeat PVLs 8/13 showed stability of DVT.
- Patient was transitioned from Heparin to Lovenox.
- Patient transitioned from PICU to floor.
- Patient will have a full IT workup outpatient.
- Inpatient IT Labs: No evidence of Factor V Leiden mutation or the Factor II gene mutation. Protein C and S levels were found to be normal.
 - Protein C and S levels can be elevated in the setting of heparin therapy.

Takeaway Points:

- Chest x-ray and CTA chest are appropriate studies for patients with a clinical presentation suggestive of PE and an elevated d-dimer.
- The risk of DVT and PE after elective orthopedic procedures is very low in the pediatric population but does occur.
- IT workup in the setting of an acute VTE does not change management but may impact duration of therapy.
- It is recommended that pediatric patients undergo IT workup if they have a VTE that is not central venous catheter related.
- Duplicated IVC occurs in 0.02% - 3% of the population and may be a risk factor for VTE.

References:

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