# AMSER Case of the Month February 2022

HPI: 34w2d infant with incidental finding of a pulmonary mass

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#### Patient Presentation

• HPI: Pt is a late preterm male (34 wks gestation) delivered via a scheduled C-section who was admitted to the Pediatric Cardiac ICU (PCICU) with prenatally suspected cyanotic congenital heart disease (ductal dependent), postnatally confirmed as tricuspid atresia with intact ventricular septum. Since birth the patient has been given prostaglandins (PGE) to maintain ductal patency until corrective surgery, caffeine loading and non-invasive respiratory support. On DOL 5, the patient started to have down trending saturations into the 60s and a low PaO2.

• PMHx: Tricuspid atresia and hypoplastic right ventricle



#### Patient Presentation

- Birth Hx: Scheduled C-section. Apgars 4 and 8 at birth. Placed on CPAP due to subcostal retractions and grunting. Transferred to PCICU, PGE initiated.
- Maternal Hx: 28yo G5P3114 with gestational diabetes controlled with insulin. Serologies negative.
- Physical Exam:
  - Vitals: HR 107, BP 91/52, spO2 79% on BiPAP (24%FiO2, Rate 40)
  - Bradycardia, 2/6 systolic ejection murmur. No nasal flaring or retractions, but occasional apneas. Normal breath sounds.

## Pertinent Labs

- WBC 6.3K, Hb 12
- Lactate 1.9
- ABG:
  - pH = 7.3
  - pO2 = **32.9**
  - pCO2 = 39.2
  - HCO = 19.2



# What Imaging Should We Order?



## Select the applicable ACR Appropriateness Criteria

Variant 3:	Intensive care unit patient with clinically worsening condition. Initial imaging.
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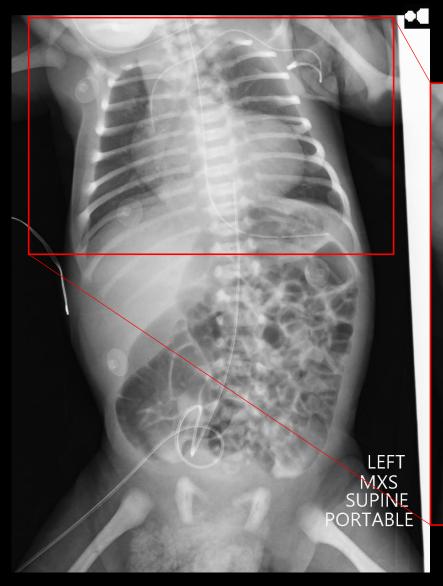
Procedure	Appropriateness Category	Relative Radiation Level	
Radiography chest portable	Usually Appropriate	<b>⊕</b>	
US chest	May Be Appropriate (Disagreement)	0	

This imaging modality was ordered by the PCICU

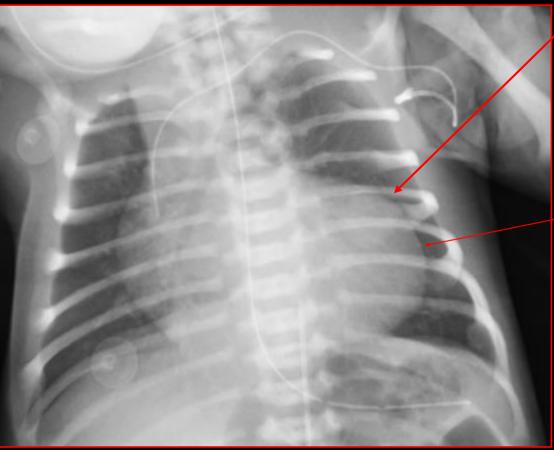


# Findings (unlabeled)





# Findings: (labeled)

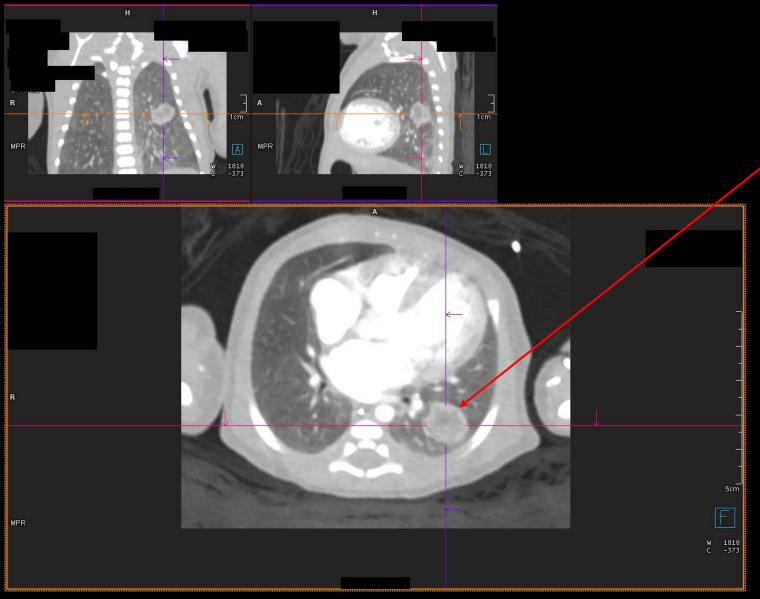


Rounded density projecting over the left heart border

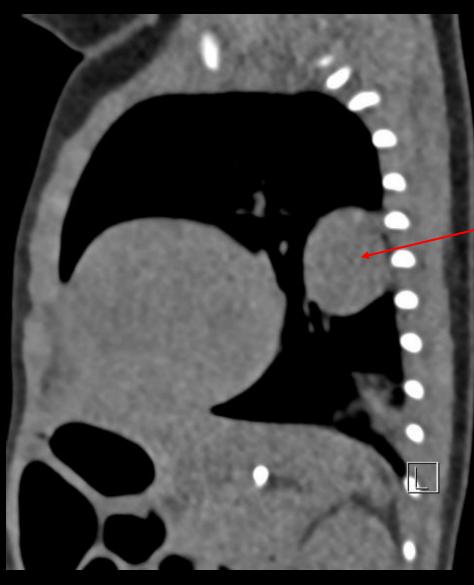
Cardiomegaly



## Cardiac Chest CT w/ IV Contrast



Large, non-calcified rim enhancing hypodense 1.3 x 1.0 x 1.2 cm lesion in the left lower lobe (LLL)



## At 1 month old

Circumscribed homogeneous soft tissue mass in LLL now measuring 2.5 x 1.6 x 2.3 cm w/o evidence of internal fat or calcifications

Scattered bilateral foci of subcutaneous emphysema likely benign cystic pneumatosis



## At 2 month old

Enhancing solid mass in posterior left hemithorax now 3.1 x 2.1 x 3.1 cm. Low central attenuation likely reflecting central necrosis or fibrosis component

Extensive subcutaneous emphysema



Left lower lobectomy and immunohistochemical stain for GLUT-1 confirmed the final diagnosis

# Final Diagnosis: Pulmonary Infantile Hemangioma



#### Case Discussion

• Pulmonary Infantile Hemangioma: Rare benign vascular tumor manifesting most commonly in the skin or liver. Characterized by a phase of rapid growth within the first few months of life, followed by a variable involution phase over the next several months to years.

#### • Risk factors:

 Prematurity, low birth weight, multiple gestations, advanced maternal age, chorionic villous sampling

#### • Classification:

- Infantile hemangioma: Positive for GLUT-1
- Congenital hemangioma: Negative for GLUT-1



#### Case Discussion

#### Radiographic features:

- Solid diffusely and avidly enhancing mass with sharply defined borders
- Cystic, multiple enhancing or endobronchial masses may also be seen
- Enlargement of pulmonary artery or vein due to vascular shunting

#### Histopathology differs based on stage:

- Rapidly proliferating/high-flow stage: Marked epithelioid endothelial cells accompanied by pericytes with numerous mitotic activity in both
- Involuting stage: Mitotic activity decreases. Endothelial cells appear more attenuated
- End-stage: Ghost vessels in a fibroadipose background



#### Case Discussion

- Clinical features:
  - Often asymptomatic
  - Respiratory distress dyspnea, stridor, cough, especially if endotracheal/endobronchial
- Management options: (Currently no FDA approved agents) Several cases reported high response to oral propranolol, a primary medical treatment. Laser treatment is especially for airway hemangiomas. No role for sclerotherapy.

Modality	Efficacy
Pharmacological therapies (glucocorticoids, propranolol, interferon alpha 2a or 2b, vinca alkaloids)	+++
Surgical excision/resection	
Lasers (FPDL, Nd-YAG, Diode)	+
Direct puncture sclerotherapy	

FPDL = Flashlamp pulsed dye laser



#### References:

- 1. Emiralioğlu, N. *et al.* Successful treatment of pulmonary hemangioma with propranolol. *Pediatr Pulmonol* **49**, 829–833 (2014).
- 2. Galdeano, F. *et al.* Multiple pulmonary infantile hemangiomas responsive to oral propranolol. *Pediatr Dermatol* **38**, 226–228 (2021).
- 3. Lichtenberger, J. P. *et al.* Primary Lung Tumors in Children: Radiologic-Pathologic Correlation From the Radiologic Pathology Archives. *RadioGraphics* **38**, 2151–2172 (2018).
- 4. Pandya, R. & Tummala, V. Giant infantile pulmonary hemangioma. *Pediatr Radiol* **40 Suppl 1**, S63-67 (2010).
- 5. Siaghani, P. J., Chavez, C., Anselmo, D. M. & Shane, L. Pulmonary infantile hemangioma presenting as a mass in a premature male infant: a case report focusing on pathological features. *Pediatr Dev Pathol* **18**, 66–70 (2015).

