

# RADY 403 Case Presentation: Vascular Ring

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May 2022

# Focused patient history and workup

- 8 week old female with a pmh of Down Syndrome presented to a routine medical genetics office appointment with choking, spitting up, and regurgitation of food after most feeds
- Vitals:
  - Temp 36.3 °C (97.3 °F) (Temporal)
  - Ht 55 cm (21.65")
  - Wt 4415 g (9 lb 11.7 oz)
  - HC 37 cm (14.57")
  - BMI 14.59 kg/m<sup>2</sup>
  - BSA 0.26 m<sup>2</sup>
- Labs:
  - TSH 1.964
  - CBC: WBC 4.6, HGB 12.1, Plt 223, Absolute Lymphs 1.8

# List of imaging studies

- Barium swallow 12/21/2021
- Echocardiogram 2/23/2022
- CTA Chest 5/10/2022

# Barium Swallow

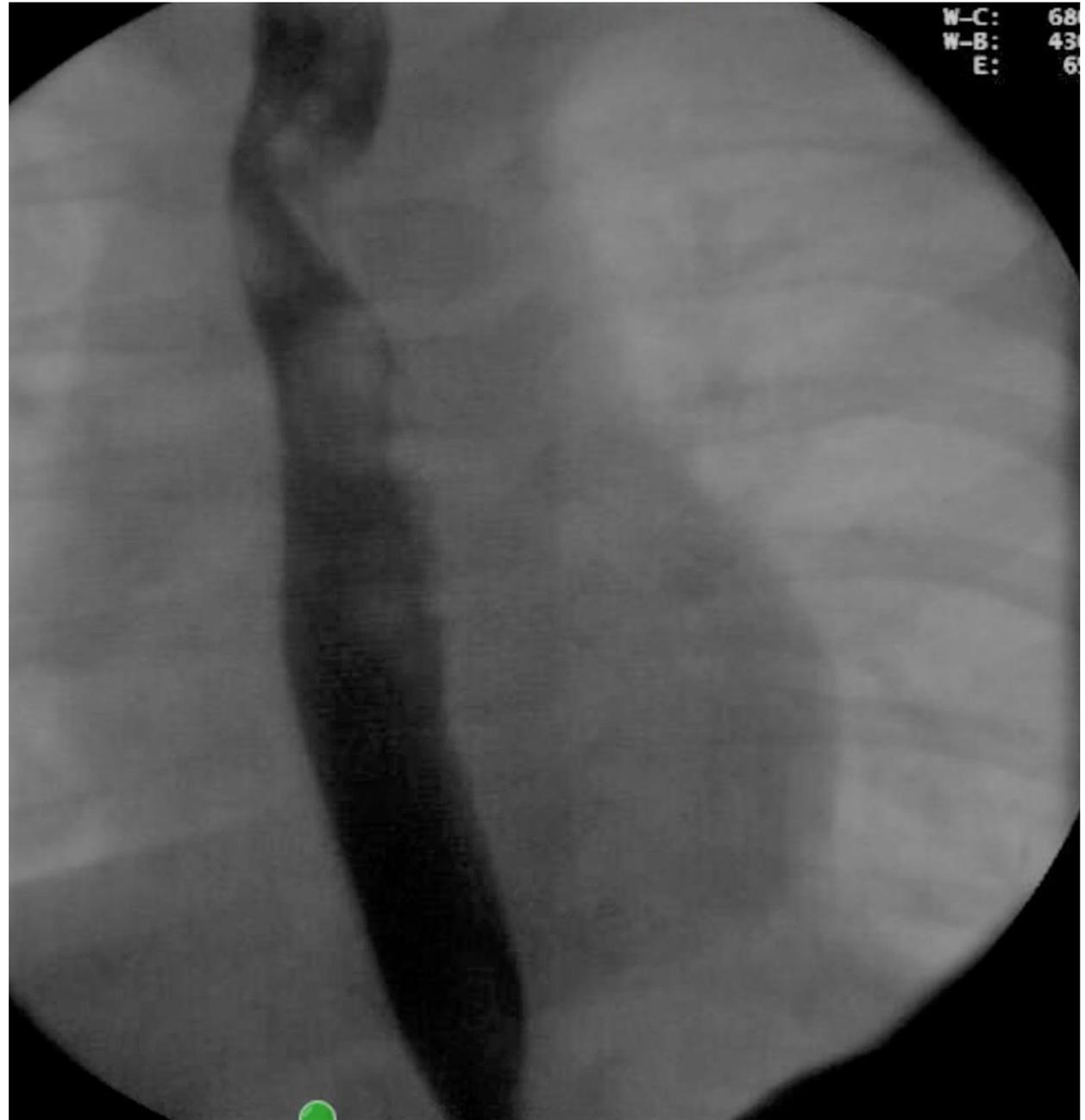


# Barium Swallow

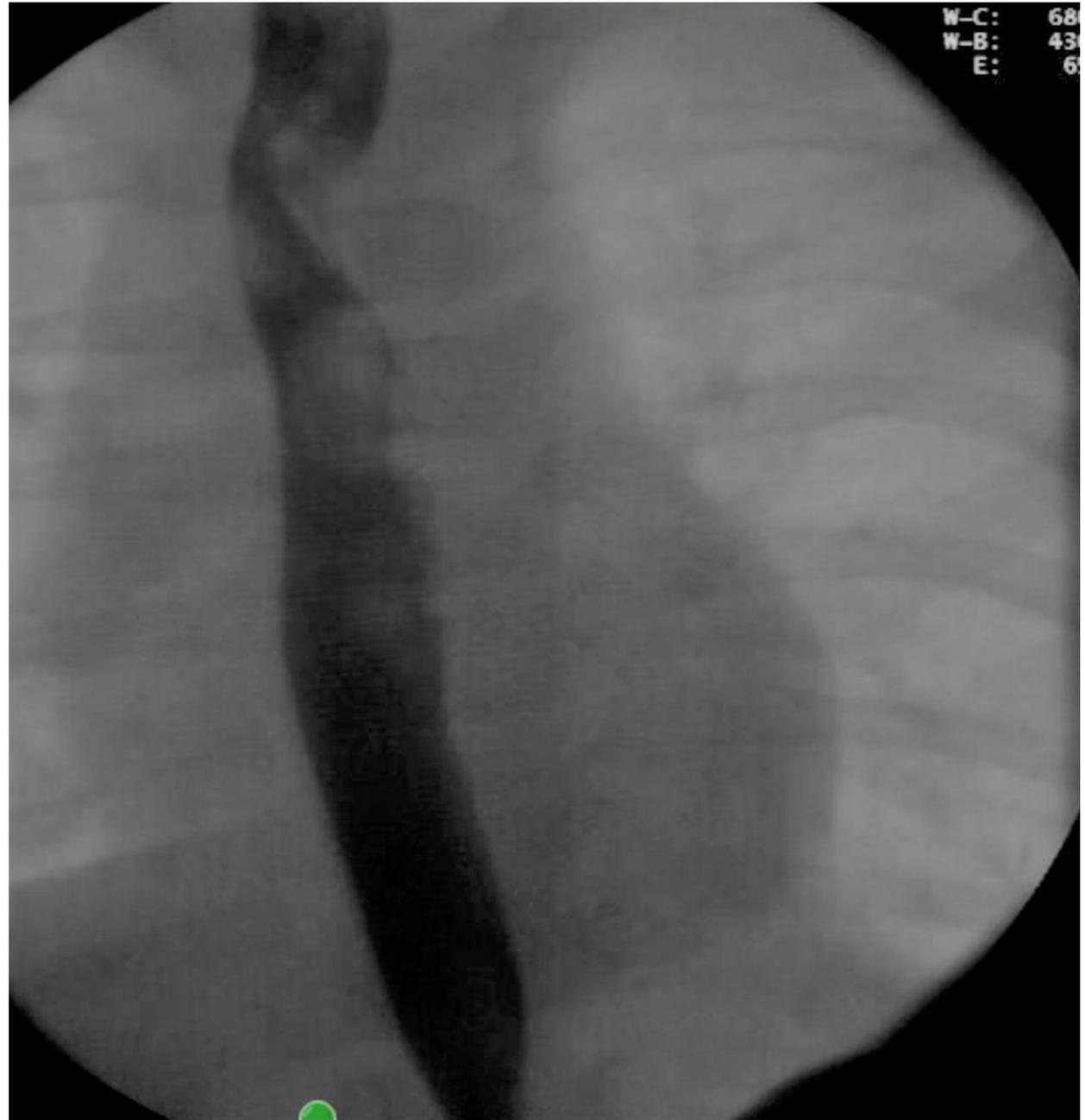
- Esophagram lateral projection shows **posterior esophageal compression** at the level of the second posterior rib



# Barium Swallow



# Barium Swallow



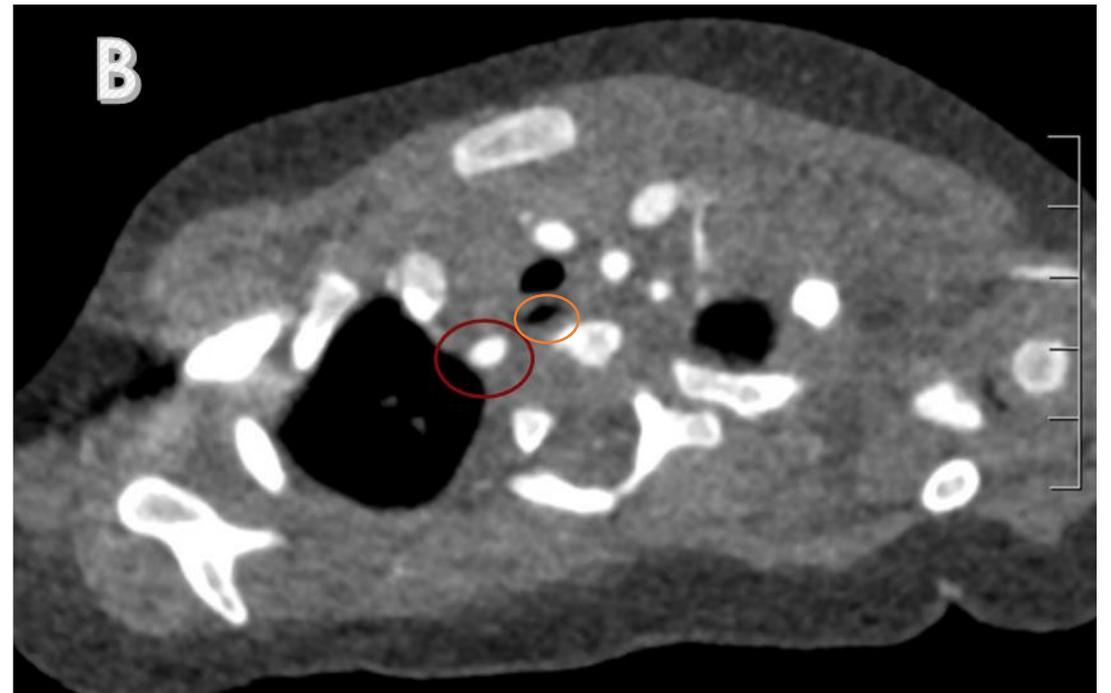
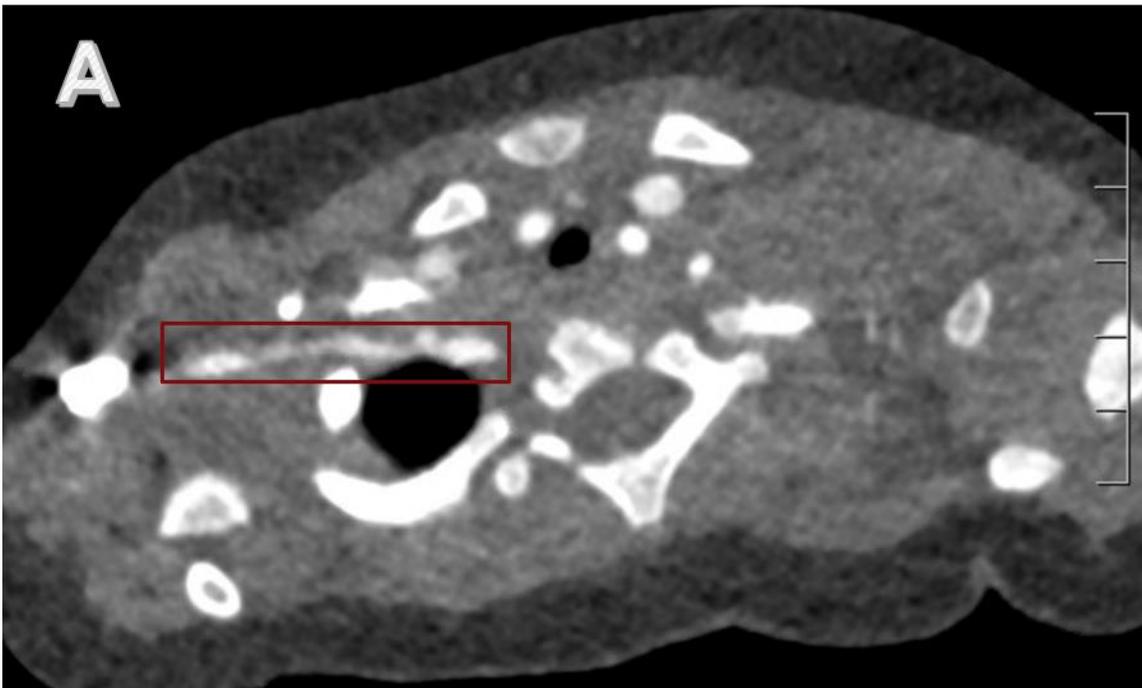
# Barium Swallow

- Esophagram anterior projection demonstrates **left-sided esophageal compression** at the level of the 2<sup>nd</sup> posterior rib



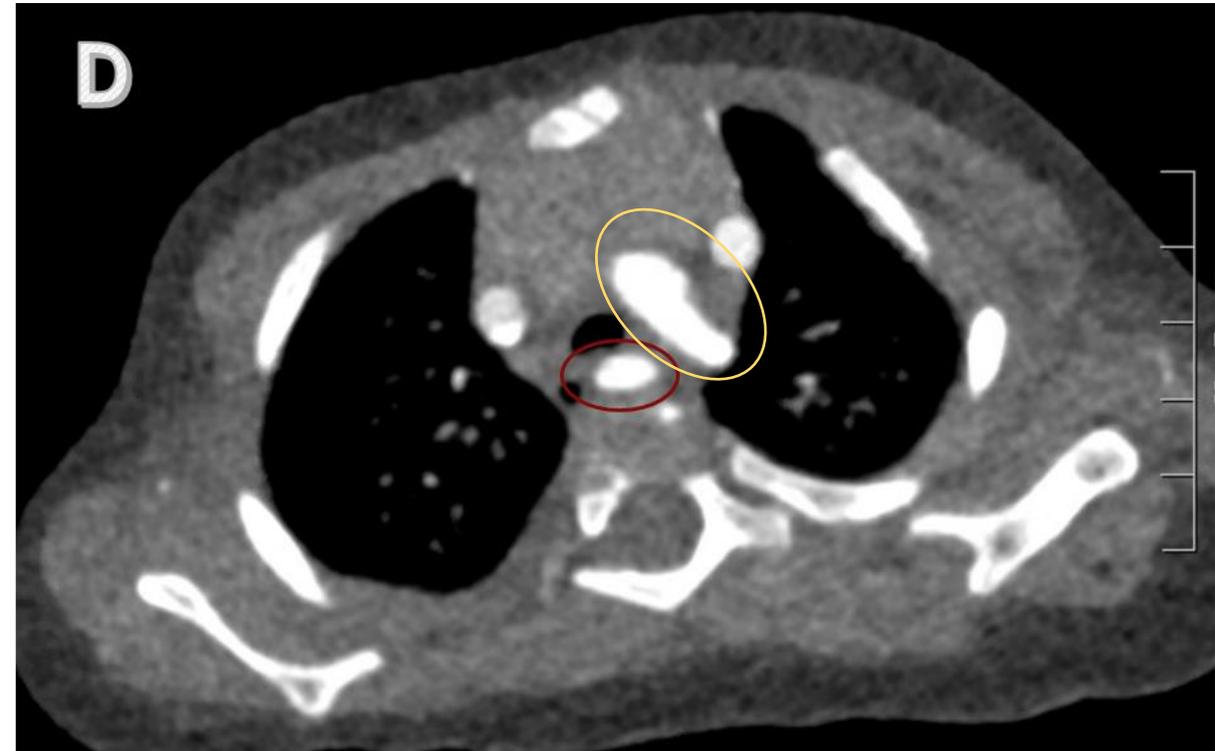
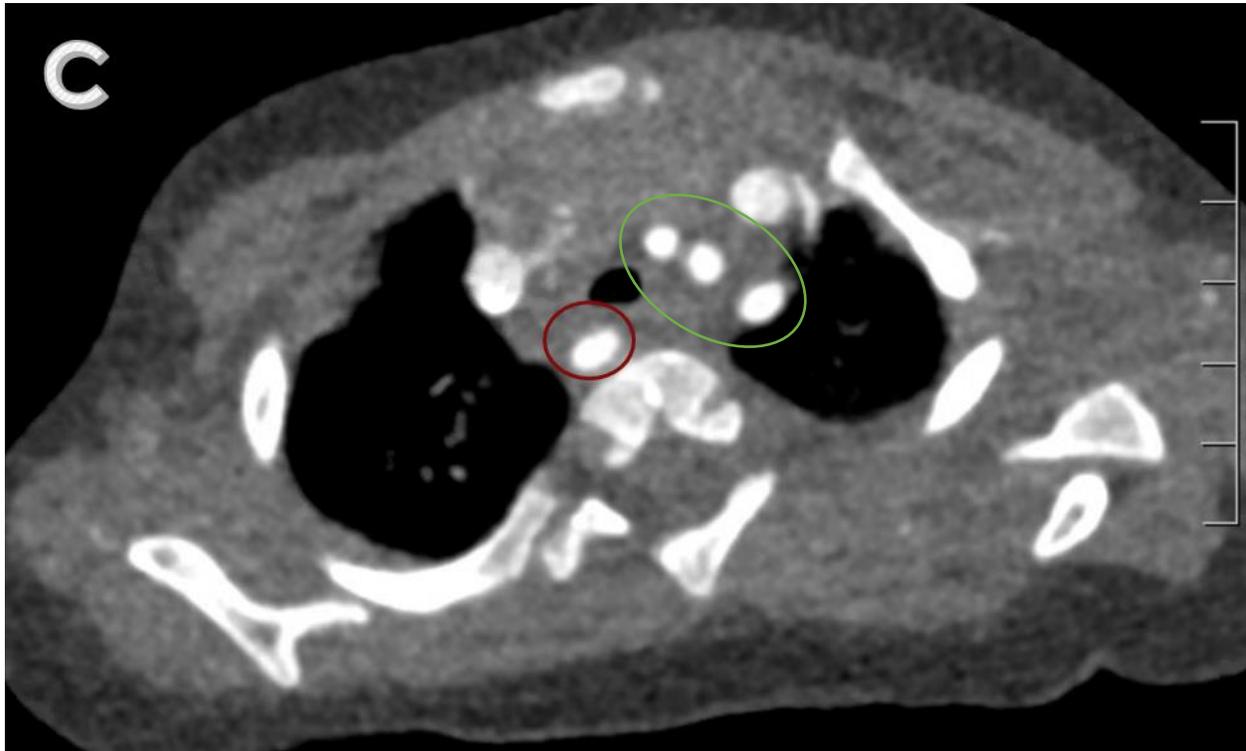
## Axial CTA Chest

- A-B: Aberrant right **subclavian** coursing inferiorly from the right axilla to the mediastinum. Note the posterior location relative to the **esophagus** in (B).



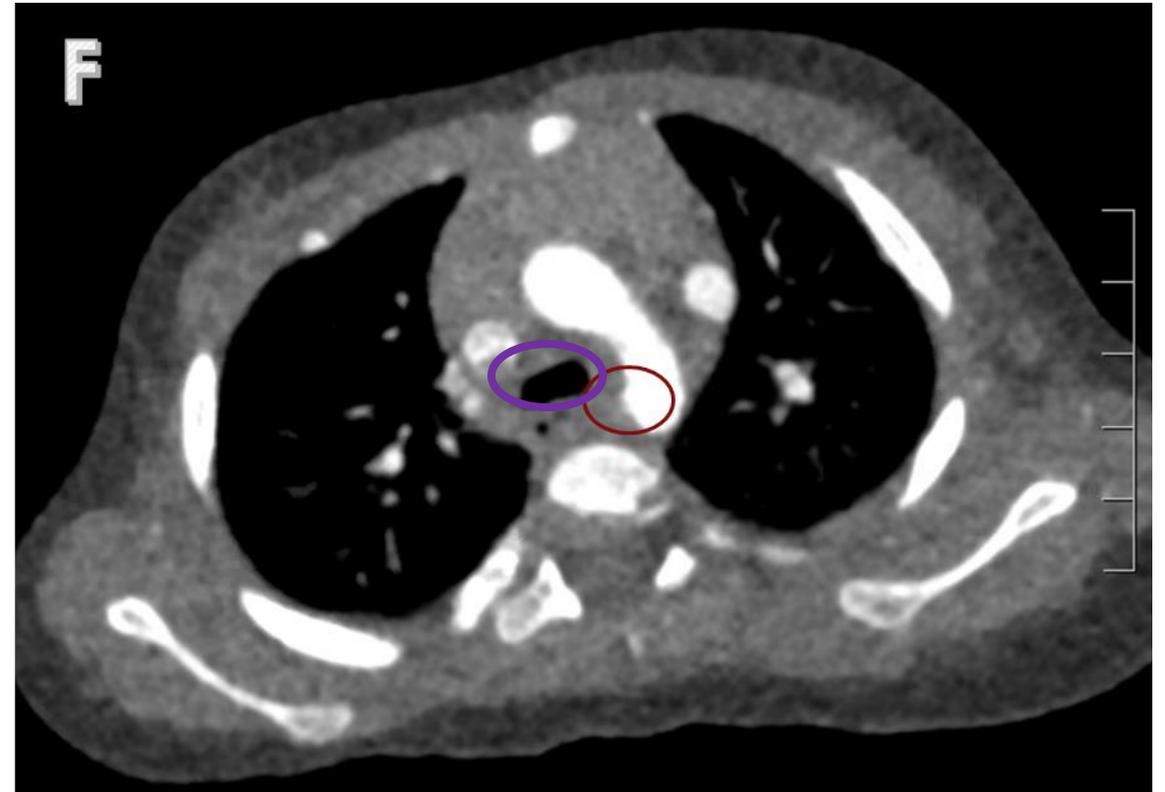
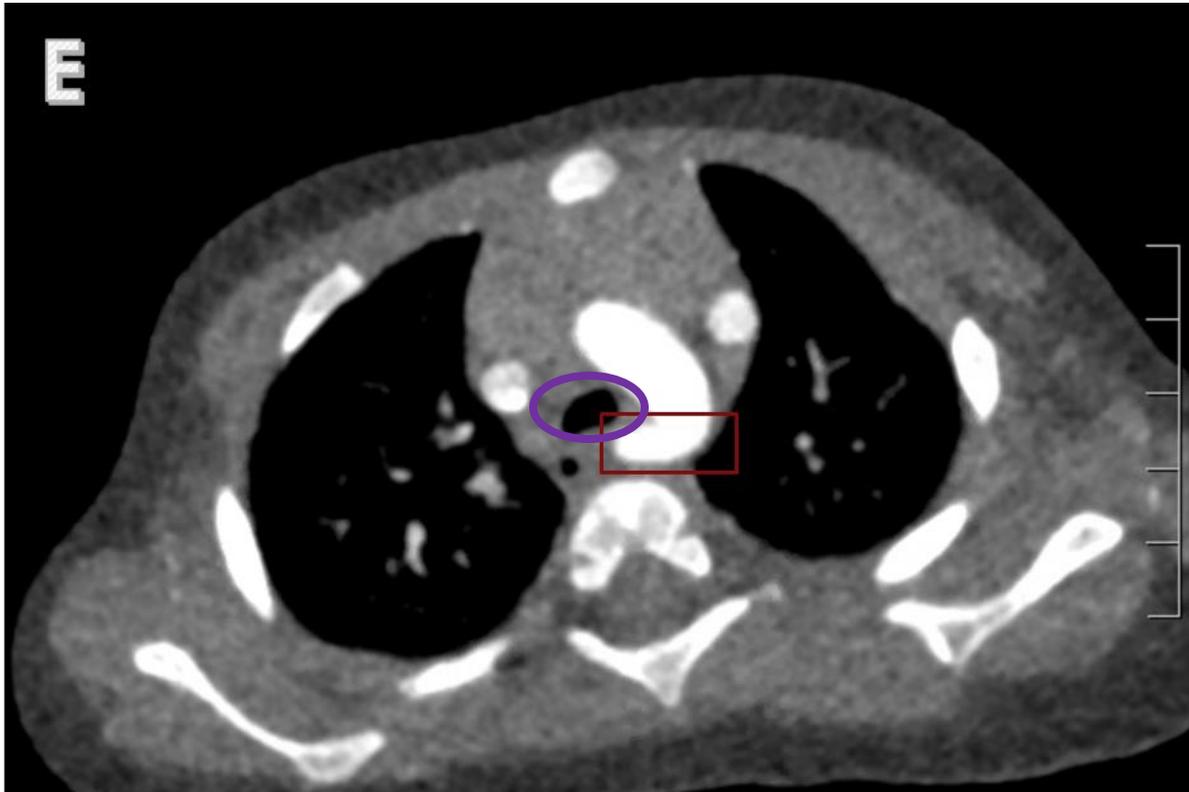
## Axial CTA Chest

- C-D: Aberrant right **subclavian** continues to track inferiorly in the mediastinum from the right to the left side. Note the left sided location of three aortic **branches** in (C) and the **arch** in (D).



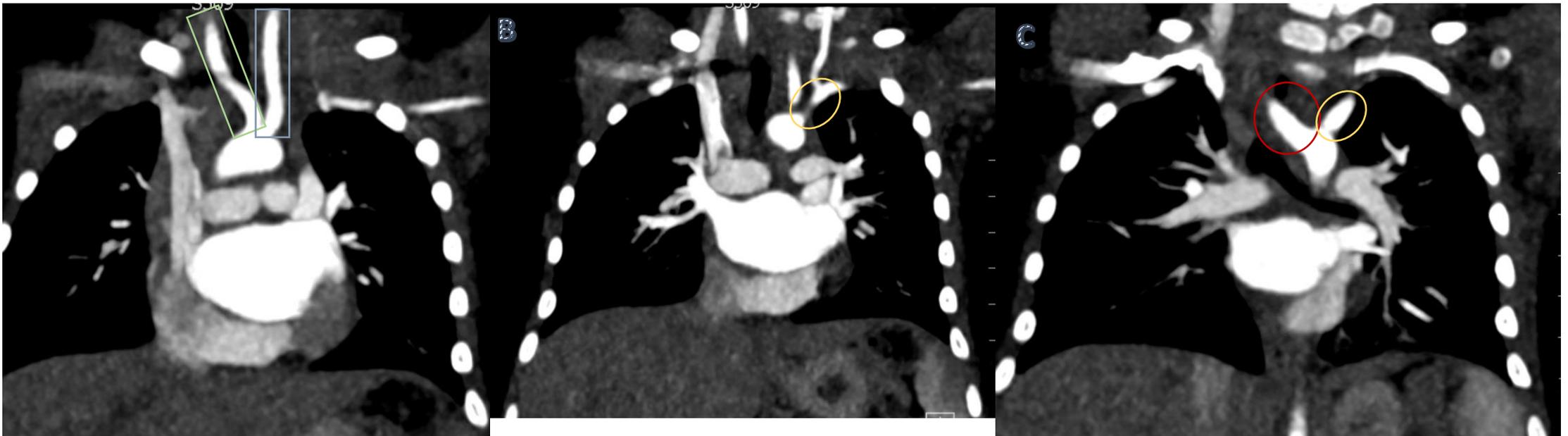
## Axial CTA Chest

- E-F: The aberrant right **subclavian** joins the left sided descending aorta in the left mediastinum. Note the abnormal flattening, or pancaking, of the **trachea**.



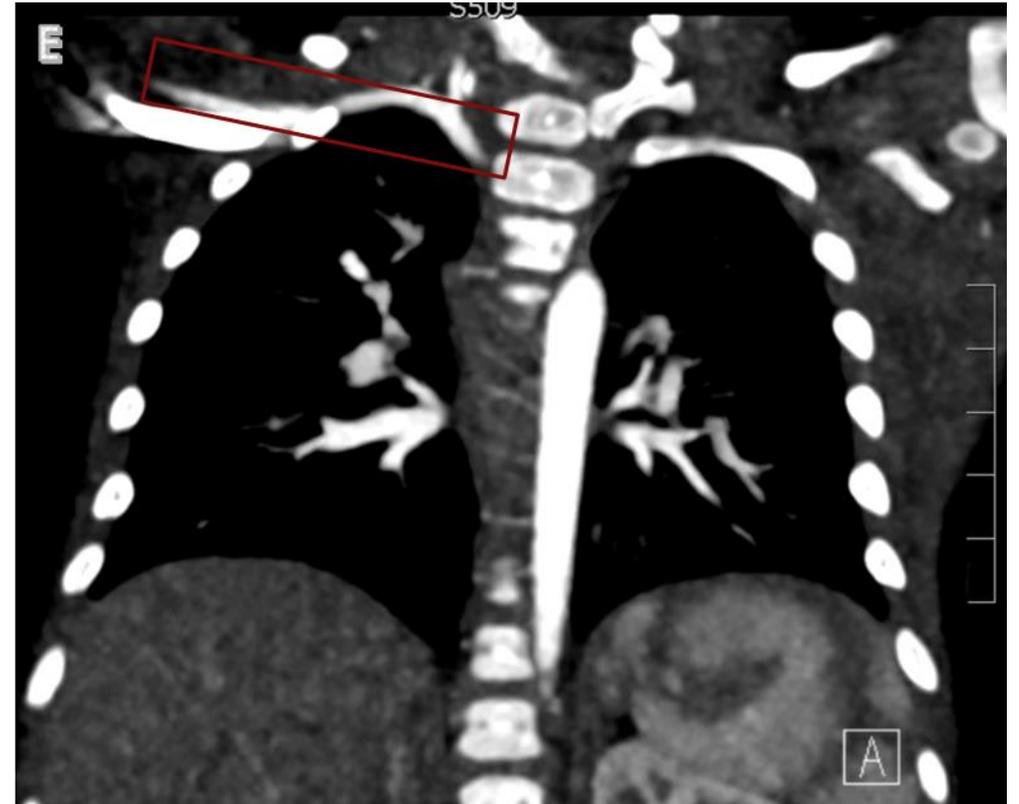
# Coronal CTA Chest

- A-C: Can see the more anterior **right** and **left** common carotid arteries in (A). Note the normal **left subclavian** in (B) and (C) with the origin of the aberrant right **subclavian** coming into view distally from the descending aorta in (C).



# Coronal CTA Chest

- D-E: Moving posteriorly in the chest the **aberrant right subclavian** can be seen exiting the mediastinum in (D), and entering the axilla in (E)

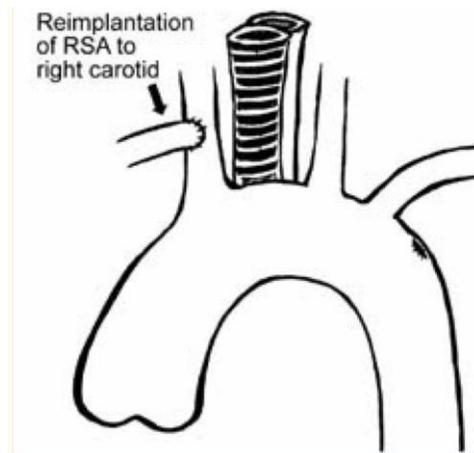


## Patient treatment or outcome

- Barium swallow demonstrated posterior esophageal compression on the left lateral view and left sided esophageal compression on the AP
  - Consistent with right aberrant subclavian
- Echocardiogram consistent with right aberrant subclavian, also incidentally found duplicated left sided SVC
- CTA of the chest performed for management planning confirmed diagnosis of left aortic arch with right aberrant subclavian and duplicated SVC
- The patient is currently pending treatment

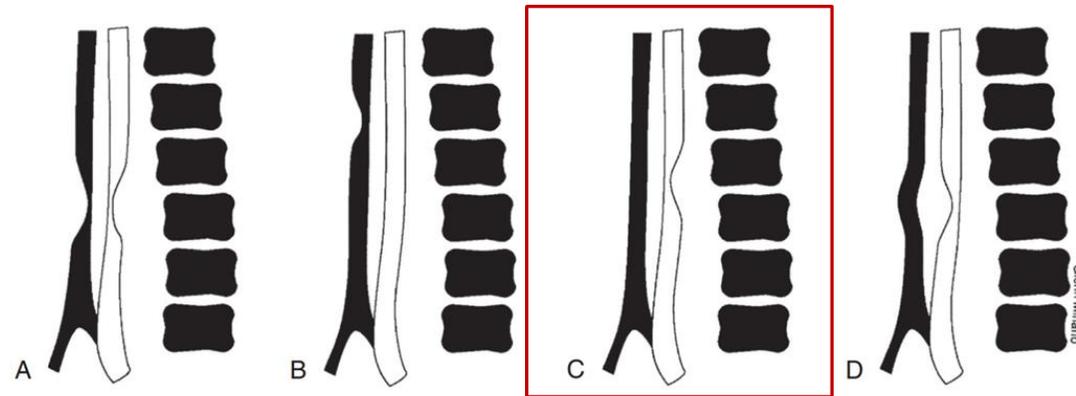
# Patient treatment or outcome

- General treatment for symptomatic right aberrant subclavian is surgical
  - The approach is individualized, but a common approach is division of the aberrant subclavian with anastomosis to the carotid (known as subclavian carotid transposition)
- Aneurysmal dilation is common at the origin of the aberrant artery and requires prophylactic surgery due to risk of aortic dissection



# Vascular Rings General Information

- Vascular rings with compression of the trachea and esophagus comprise 1-3% of all congenital cardiac anomalies
- Divided into two categories
  - Complete rings: Double arch, Right arch/left ligamentum arteriosum
  - Partial rings: Pulmonary artery sling(aberrant left pulmonary artery), innominate artery compression, **left arch with aberrant right subclavian**



A. Double aortic arch

B. Innominate artery compression

C. Left arch with aberrant right subclavian

D. Aberrant left pulmonary artery

*NB: The black structure is the trachea and the white structure is the esophagus.*

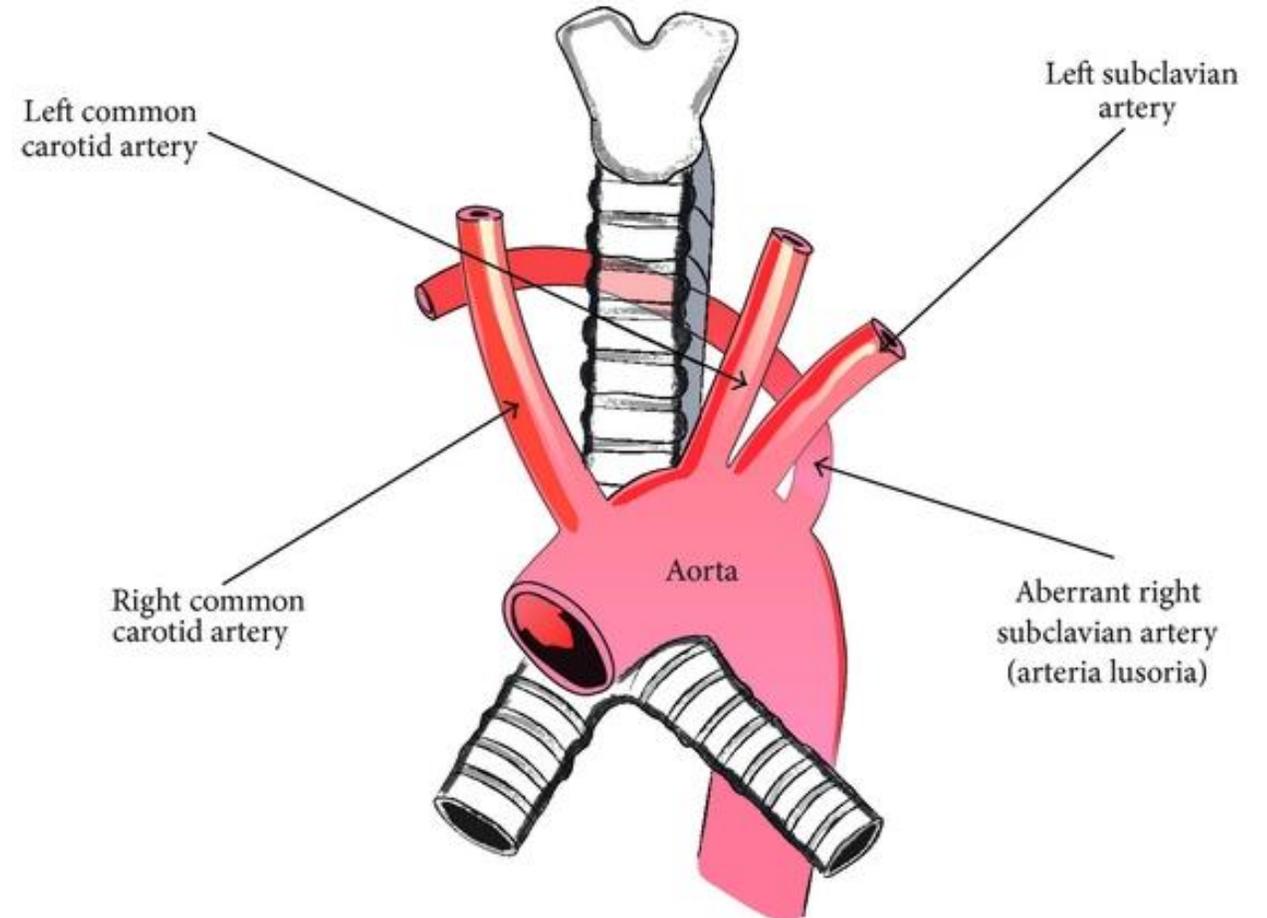
# Vascular Rings General Information

- Most common vascular rings
  - Right aortic arch and an aberrant left subclavian artery and left-sided ductus arteriosus = 30-65% of cases
  - Double aortic arch = 30-45% of cases
  - Innominate artery compression syndrome = 3-20% of cases
  - Aberrant left subclavian = 3-7% of cases
  - Pulmonary artery sling = <5% of cases
  - Aberrant right subclavian = <5% of cases
- NB: Aberrant right subclavian is the most common embryologic abnormality of the aortic arch, but is very uncommon with accompanying left sided aortic arch

# Aberrant Right Subclavian With Left Aortic Arch

- Has an incidence as low as 0.07-0.2% in individuals with left sided aortic arch
- Symptomatic left arch with aberrant subclavian most commonly causes posterior esophageal compression WITHOUT tracheal compression
  - Can cause feeding difficulty called dysphagia lusoria
  - No issues with breathing

(Note! can be fooled thinking classic presentation of posterior esophageal course because 15% between trachea and esophagus and 5% anterior to the trachea rather than posterior)



# Aberrant Right Subclavian Artery Associations

- Associated anatomic variations:
  - Truncus bicaroticus (common origin of carotid arteries) 19.2% of the time
  - Proximal aberrant subclavian aneurysm ( Kommerell diverticulum) 14.9% of the time
  - Aneurysm of the distal aberrant subclavian 12.8% of the time
  - Right sided aortic arch 9.2% of the time
- Some studies suggest an incidence of up to 19-36% in children with down syndrome
  - Also suggested higher incidence in other trisomies such as 18

# ACR Appropriateness Criteria

- In pediatric patients, especially infants, differentiating oropharyngeal from retrosternal dysphagia is extremely challenging
- Patients with unexplained dysphagia should have a biphasic esophagram to look at structures from the oral cavity to the gastric cardia
  - Lesions of the esophagus or gastric cardia can cause referred dysphagia sensation to the pharynx

## **Variant 2: Unexplained oropharyngeal dysphagia. Initial imaging.**

Procedure	Appropriateness Category	Relative Radiation Level
Fluoroscopy biphasic esophagram	Usually Appropriate	☼☼☼
Fluoroscopy barium swallow modified	May Be Appropriate	☼☼☼
Fluoroscopy single contrast esophagram	May Be Appropriate	☼☼☼
Fluoroscopy pharynx dynamic and static imaging	May Be Appropriate (Disagreement)	☼☼☼
Esophageal transit nuclear medicine scan	May Be Appropriate	☼☼☼
CT neck and chest without IV contrast	Usually Not Appropriate	☼☼☼☼
CT neck and chest with IV contrast	Usually Not Appropriate	☼☼☼☼
CT neck and chest without and with IV contrast	Usually Not Appropriate	☼☼☼☼

## **Variant 3: Retrosternal dysphagia in immunocompetent patients. Initial imaging.**

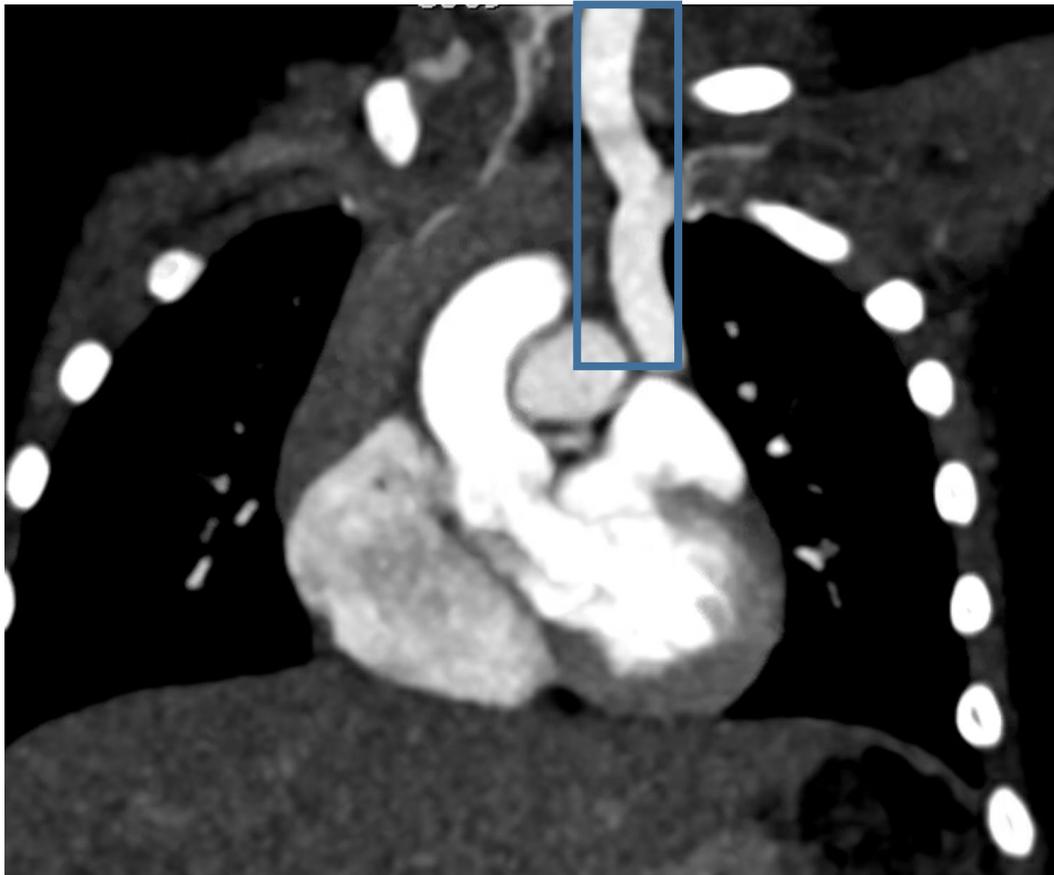
Procedure	Appropriateness Category	Relative Radiation Level
Fluoroscopy biphasic esophagram	Usually Appropriate	☼☼☼
Fluoroscopy single contrast esophagram	May Be Appropriate	☼☼☼
Esophageal transit nuclear medicine scan	May Be Appropriate	☼☼☼
Fluoroscopy barium swallow modified	May Be Appropriate	☼☼☼
CT neck and chest without IV contrast	Usually Not Appropriate	☼☼☼☼
CT neck and chest with IV contrast	Usually Not Appropriate	☼☼☼☼
CT neck and chest without and with IV contrast	Usually Not Appropriate	☼☼☼☼
Fluoroscopy pharynx dynamic and static imaging	Usually Not Appropriate	☼☼☼

## UNC Top Three

- Pediatric dysphagia should prompt consideration of a vascular ring
- Biphasic esophagram to look at structures from the oral cavity to the gastric cardia should be performed in unexplained dysphagia per the ACR appropriateness criteria
- Right aberrant subclavian is a specific prevalent ring that should be looked for in patients with Down Syndrome

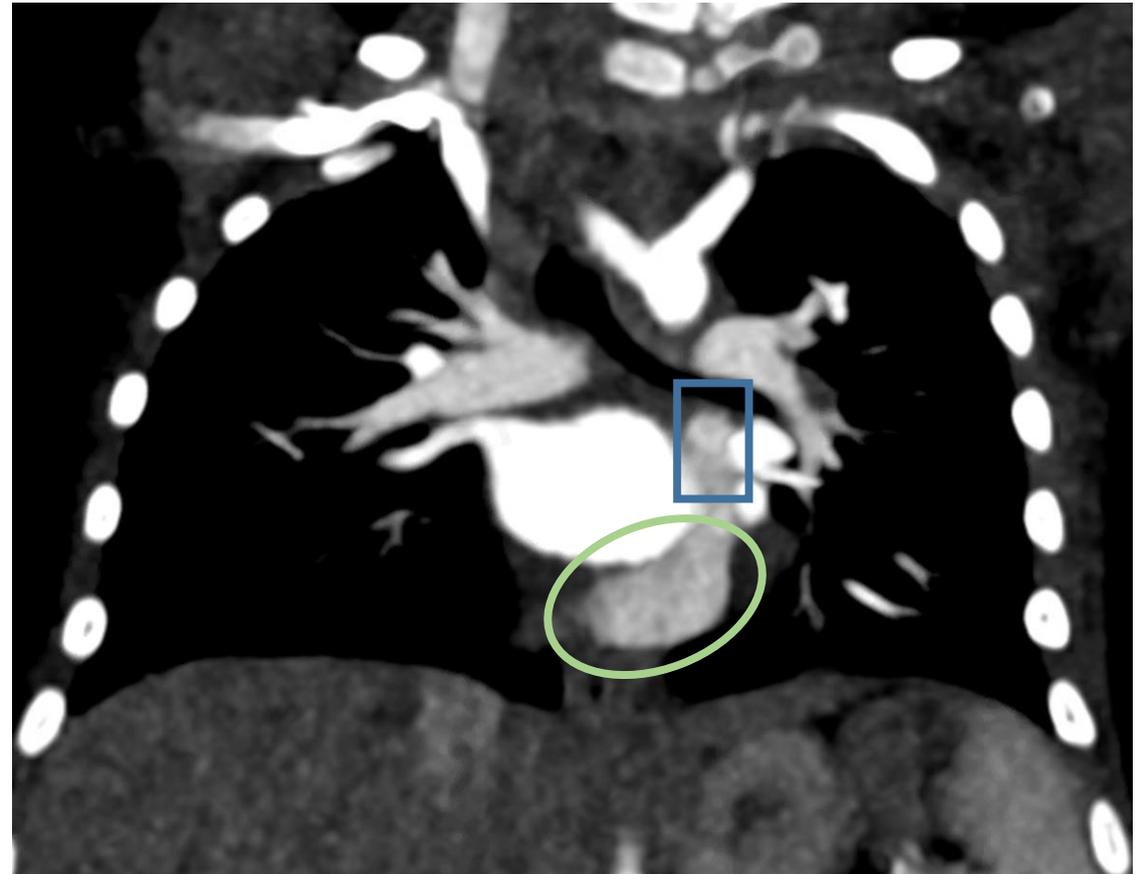
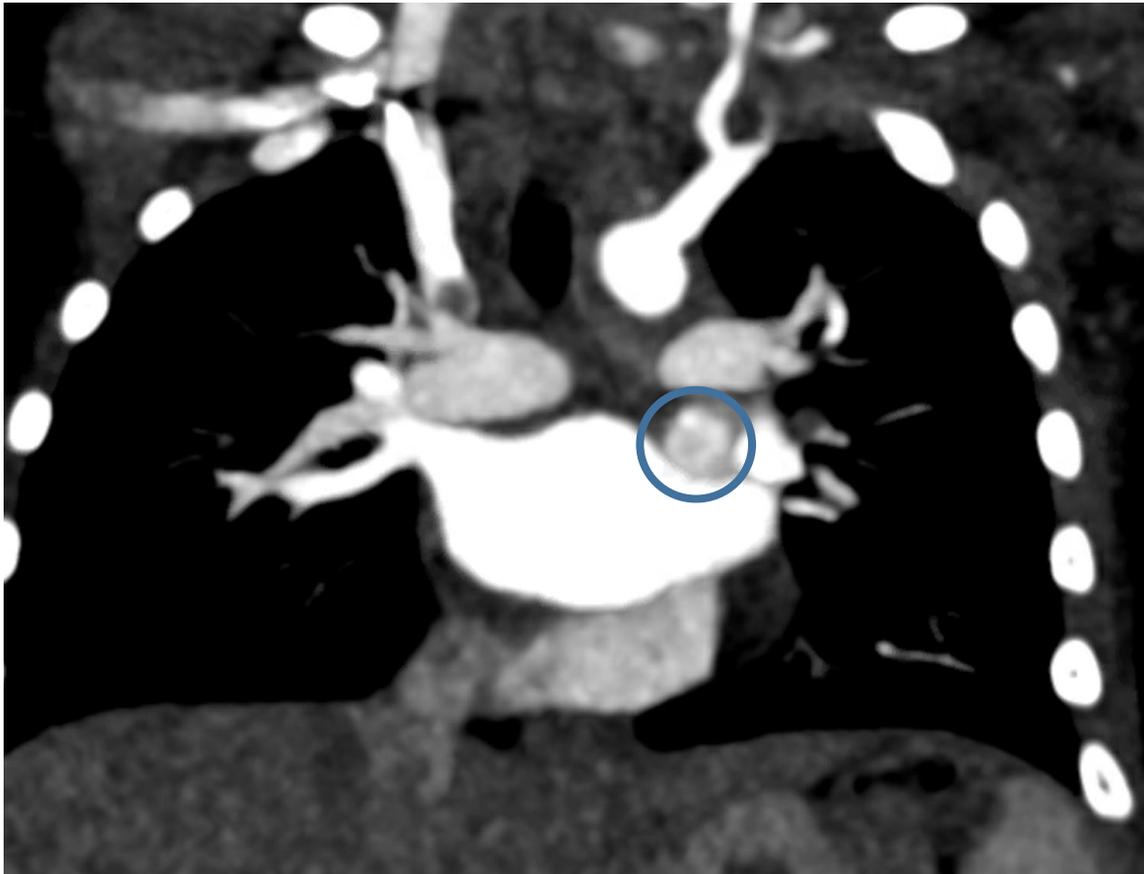
## Additional Finding – Duplicated Left SVC

- Duplicated **SVC** is seen coursing down the left side of the mediastinum rather than returning to the right heart via a normal left brachiocephalic vein



## Additional Finding – Duplicated Left SVC

- Moving from anterior to more posterior CT slices, the duplicated SVC drains into the coronary sinus, which is dilated as result



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