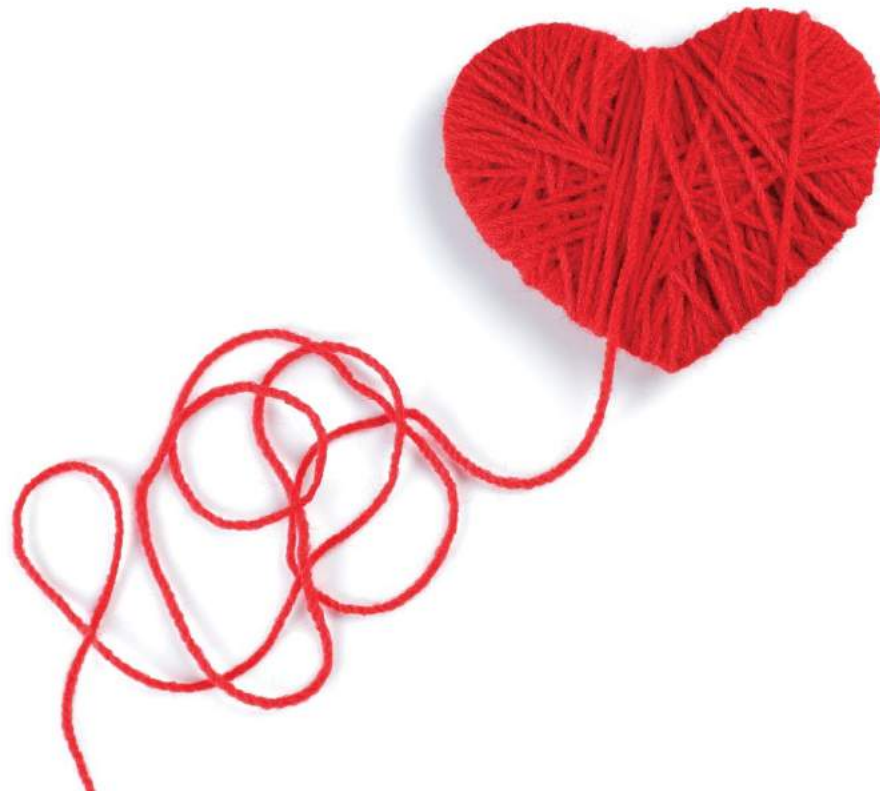


CARDIOLOGY  
2023

## BOX OF CHOCOLATES DAY

Mary Hoang, MSN, CPNP-AC/PC

2/25/2023



# OBJECTIVES

- Discuss different clinical presentations of critical congenital heart defects in pediatric.
- Recognize the uniqueness of pediatric congenital heart defects.

# SETTING

- Specialty Care Center
- Part of strip mall: Large grocery chain, Hallmark store, European wax center, Nothing Bundt Cake, and Chick-Fil-A.

# DRIVE TO CLINIC

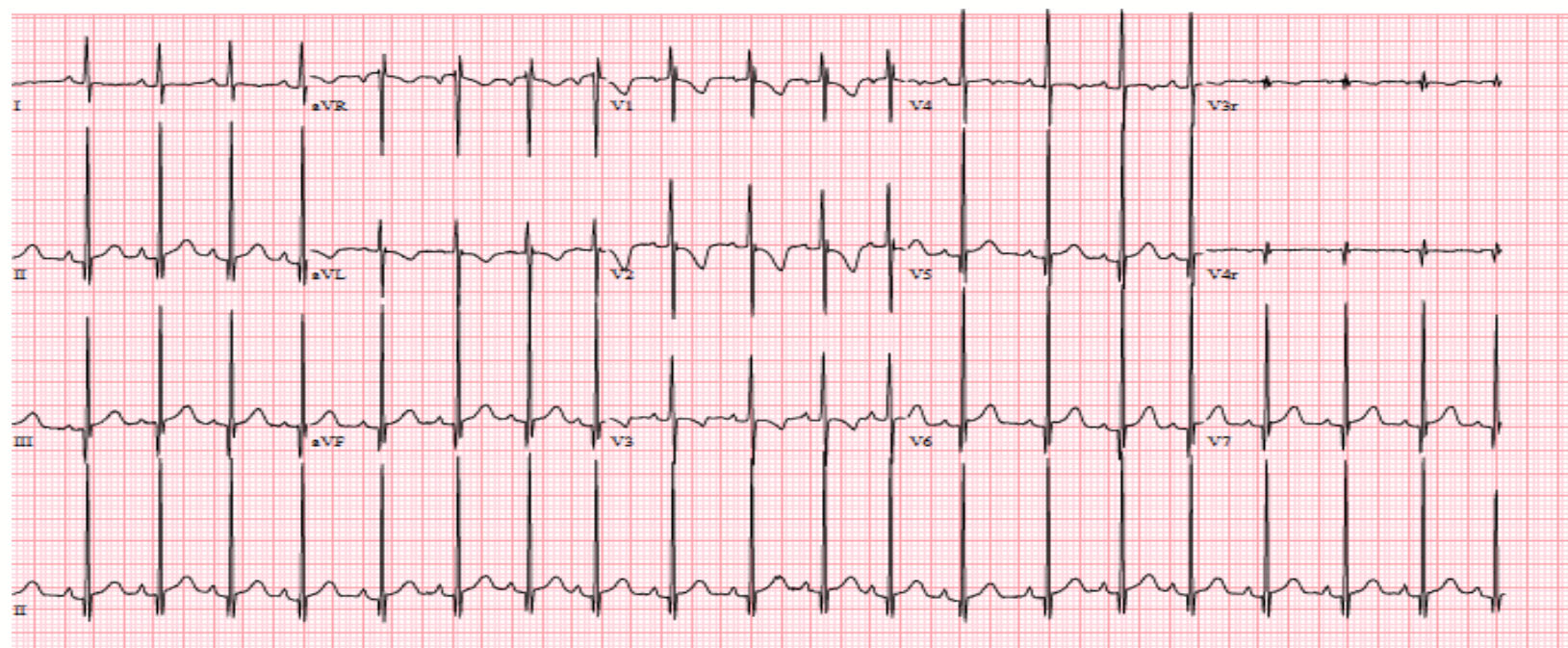


# EXAM ROOM



# REASON FOR REFERRAL: HEART MURMUR

- HPI: New heart murmur at recent 6 month well-visit. No other concerning symptoms.
- Family history: Noncontributory
- Exam: Healthy, well-developed, well-nourished. Non-dysmorphic. Cardiac: Normal rate and rhythm. Normal S1 and physiologically split S2. Grade II/VI medium-pitched systolic murmur to left sternal border and radiation to left upper sternal border. No click, rub, or gallops. Peripheral pulses are symmetrical and equal in caliber. Abdomen: Soft, non-distended, normal bowel sounds. The liver is not enlarged.



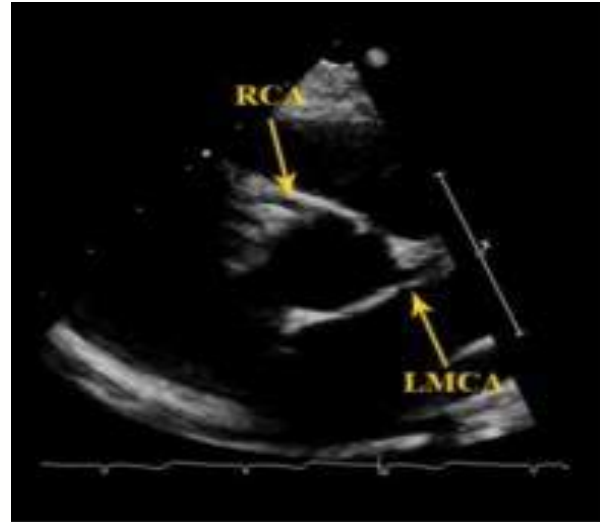
# I AM LOOKING AT THE ECHOCARDIOGRAM IMAGES AND...



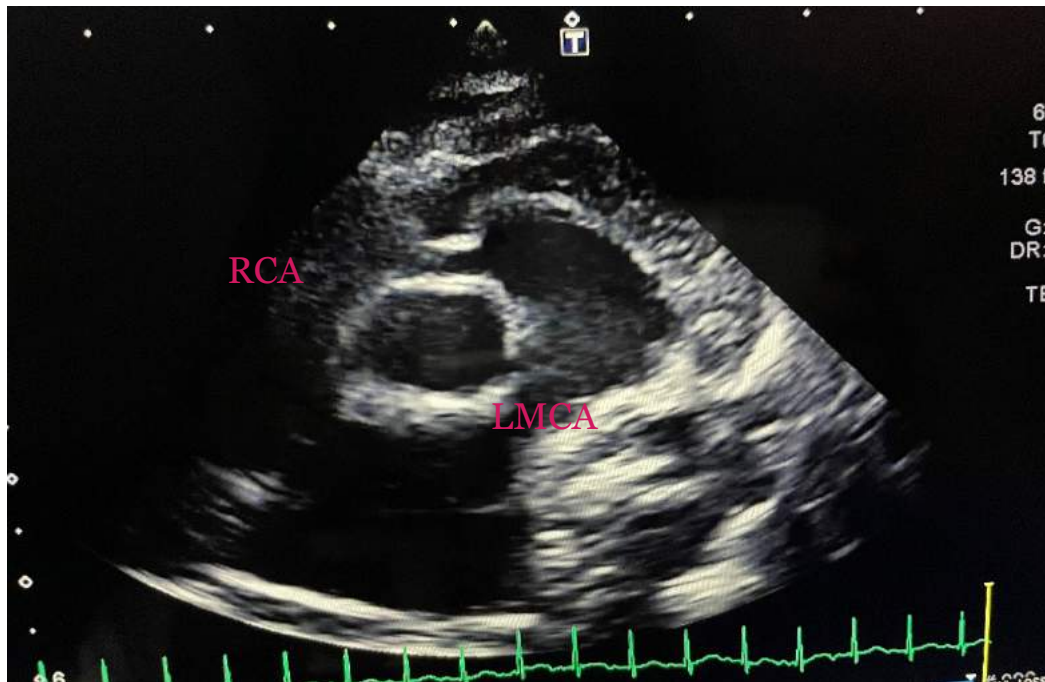




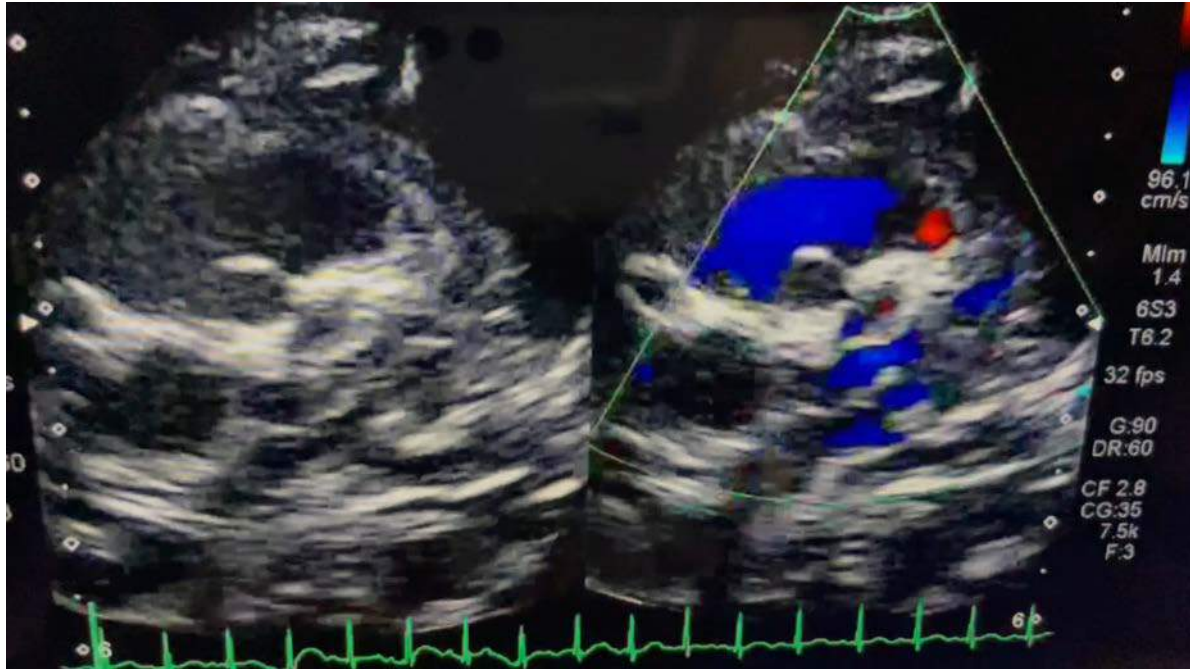
# JUST TO REVIEW



# NOT NORMAL



# IS IT REAL?



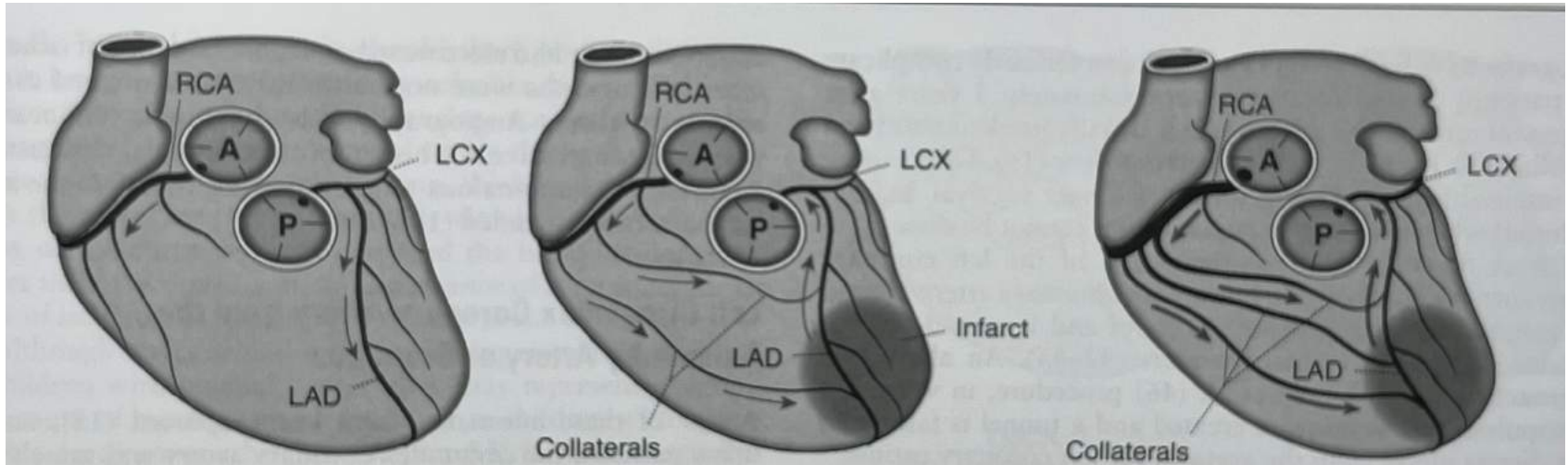
# ANOMALOUS RIGHT CORONARY ARTERY FROM THE PULMONARY ARTERY (ARCAPA)

- Anomalous right coronary artery from the pulmonary artery
- A very rare congenital heart defect.
- First description of ARCAPA was 1885 by John Brooks, Irish anatomist.
- First Case diagnosed by echocardiogram was 1985.
- First reported surgical intervention was 1972.
- Incidence in the general population is 0.002%.
- At TCH, there are 7 cases. This is the only one with no other congenital heart defect or genetic abnormality.

# PATHOPHYSIOLOGY

- The pulmonary and aortic pressures are similar during fetal life. Shortly after birth, the high pulmonary pressure causes antegrade flow of deoxygenated blood through the RCA.
- As pulmonary pressure decreases, blood then flows retrograde into the pulmonary artery and creates a “coronary steal” phenomena.
- The clinical presentation is related to the direction of blood flow in the coronary artery, extent of collateralization, and impact of oxygen delivery to the myocardium.
- If collateralization between the LCA and RCA is present and pulmonary artery pressure remains low, the LCA supplies oxygenated blood to the entire heart.
- Myocardial ischemia may be present even in absence of symptomatology.

# COMPARED TO ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY (ALCAPA)



Moss and Adams(2016)



# CLINICAL PRESENTATION OF ARCAPA

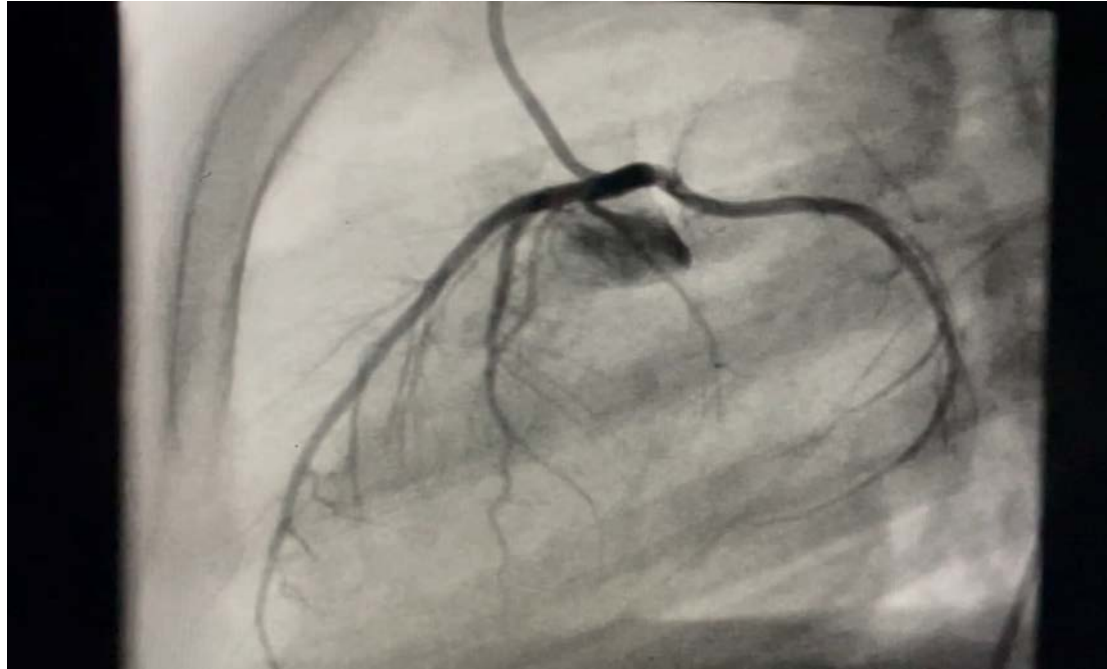
- Many remain asymptomatic during infancy.
- Neonatal arrhythmia, myocardial ischemia, and heart failure have been reported.
- Recent systemic review published in Annals of Thoracic Surgery by Guenther et al in 2020. From 1950 to 2019 using PubMed, Google Scholar, and Embase. 223 cases from 193 papers:
  - Murmur on exam: “systolic” or “continuous”
  - Bimodal distribution among symptomatic patients; neonatal and around 40-60 years.
  - Cardiac angiography was most common method for diagnosis.
  - Reimplantation of the RCA was method of repair.



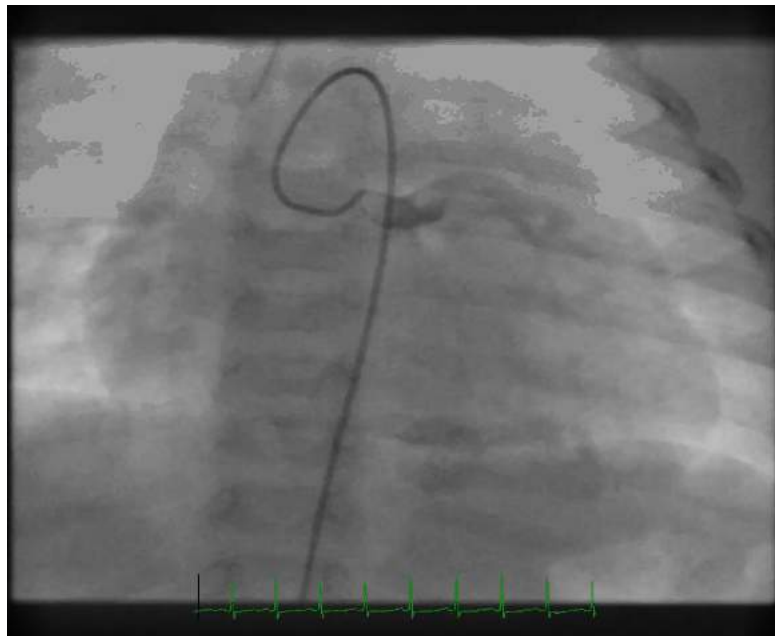
# NOW WHAT?

- Confirm diagnosis with reading physician.
- Discussed with receiving physician.
- She was admitted from clinic.
- Had cardiac catheterization to rule out coronary fistulae, per assigned surgeon.

# NORMAL



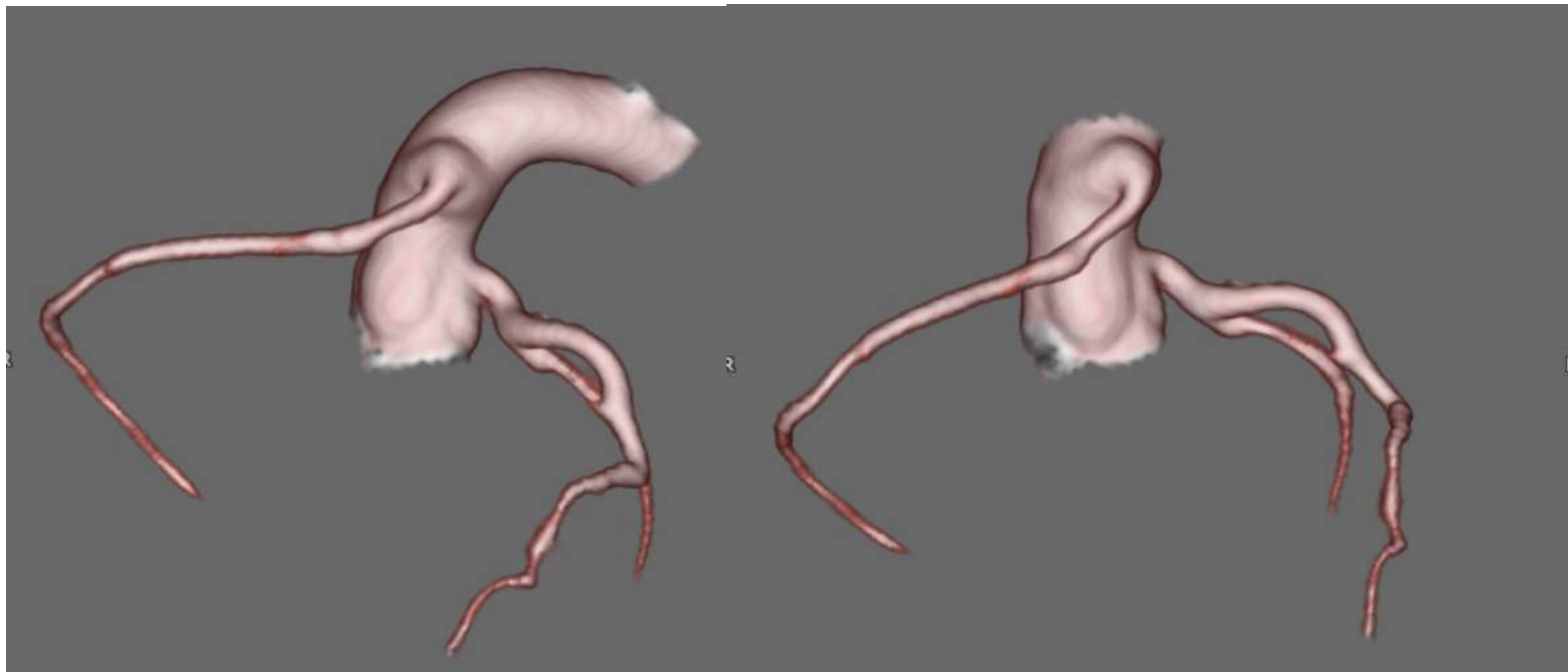
# MY PATIENT'S

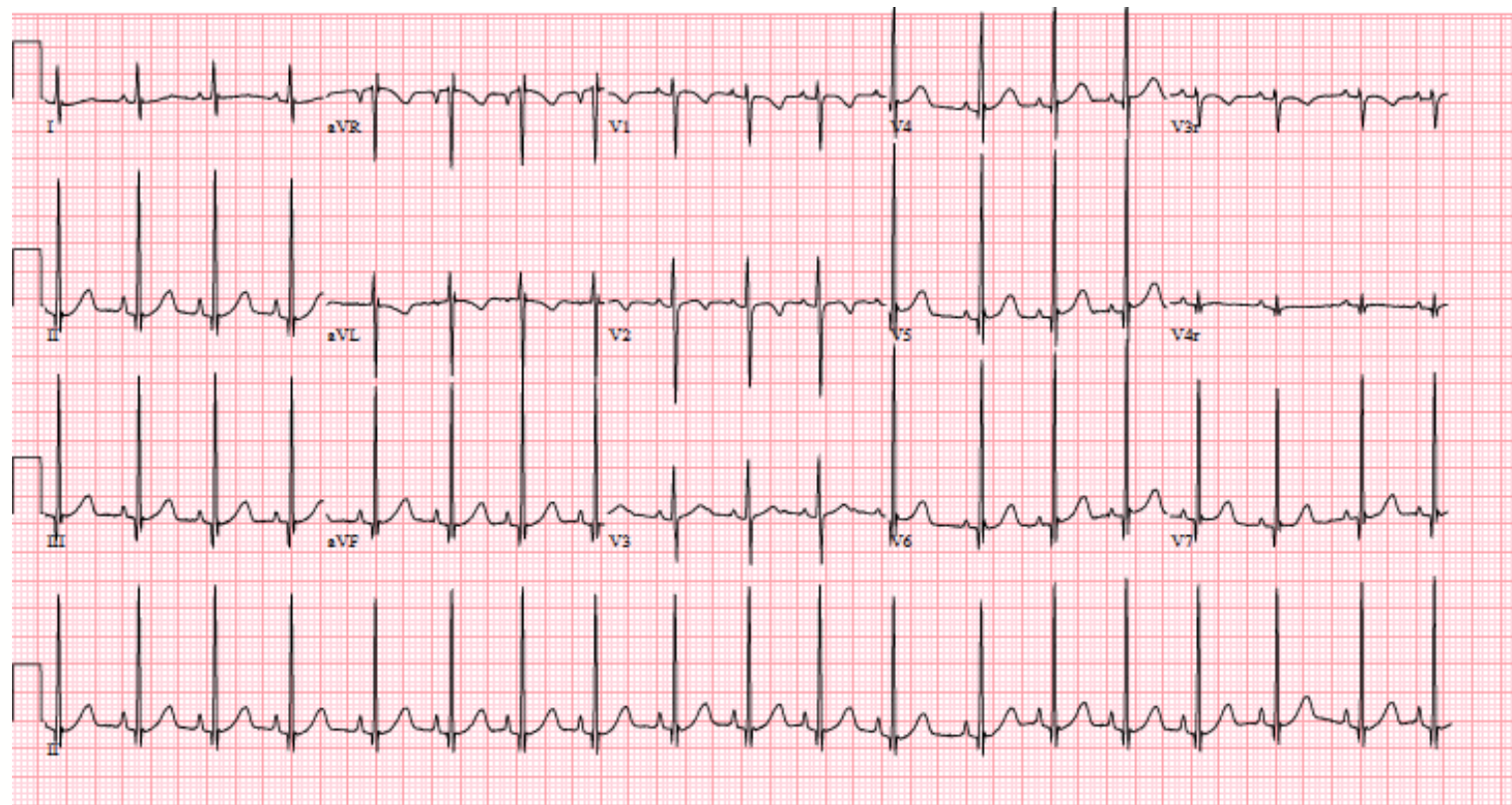


# MANAGEMENT

- Surgical repair upon diagnosis.
- Most are maintained on low dose aspirin.
- Late development of thrombus has been reported.
- Repair was done by re-implantation of the right coronary artery to the aorta.
- She did well and was discharged home on POD 7.
- Post-op meds: Lasix and aspirin

# POST-OPERATIVE CARDIAC CT 3 MONTHS LATER





# TAKE AWAY

- While rare, this unique case demonstrates need for clinicians to be aware of this anomaly.
- Early diagnosis can prevent sudden death and cardiac arrest.
- Not all heart murmurs are innocent.

