

Sunrise Session: Optimizing Care and Outcomes in Complex CHD

Do not seek and you shall not find

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Case Presentation

- 5 m/o old male presents to outside hospital with poor feeding, cough, lethargy for ~3 days
- Recent mild viral URI
- Achieving milestones, weight 3%ile
- No relevant family history
- Flu/COVID-19 negative



Exam and CXR in ER

Exam:

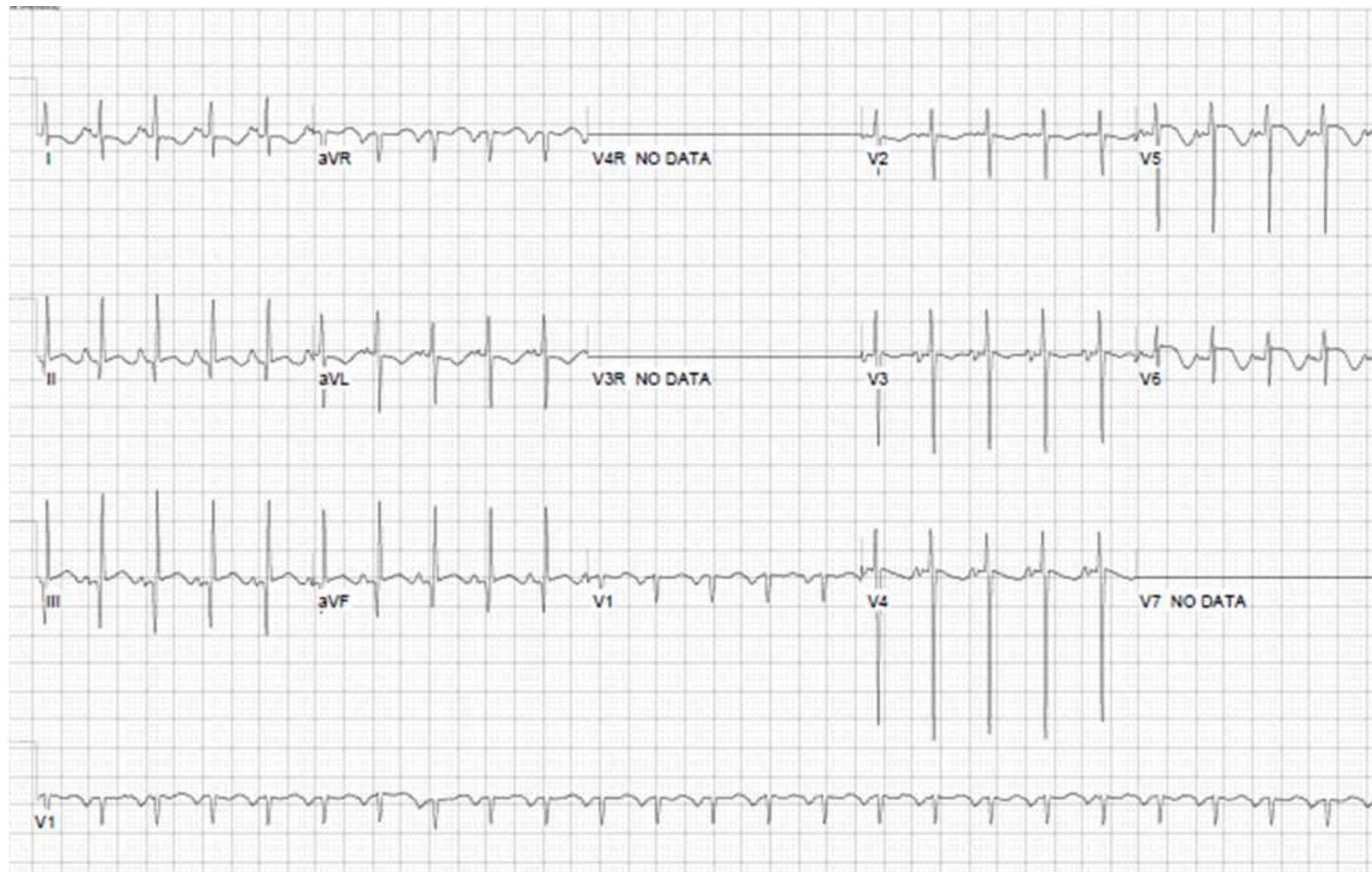
- Non-dysmorphic; uncomfortable
- HR 160, RR 68, BP 83/44, Pulse ox sats 98%
- Nasal flaring, tracheal tug, intercostal retractions
- III/VI systolic murmur at apex, + gallop present
- Liver 4 cm BCM, bowel sounds present, soft abd
- No rash, joint swelling, lymphadenopathy, mucosal/conjunctival changes



Key lab findings

- Unremarkable CBC
- Mildly abnormal INR (1.6) and AST/ALT
- Normal Na/K; creatinine and BUN mildly elevated
- Lactic acid 2.0
- C-reactive protein normal
- B-type natriuretic peptide 16,000 (upper limit 100)
- Troponin T 1.22 (upper limit 0.1)

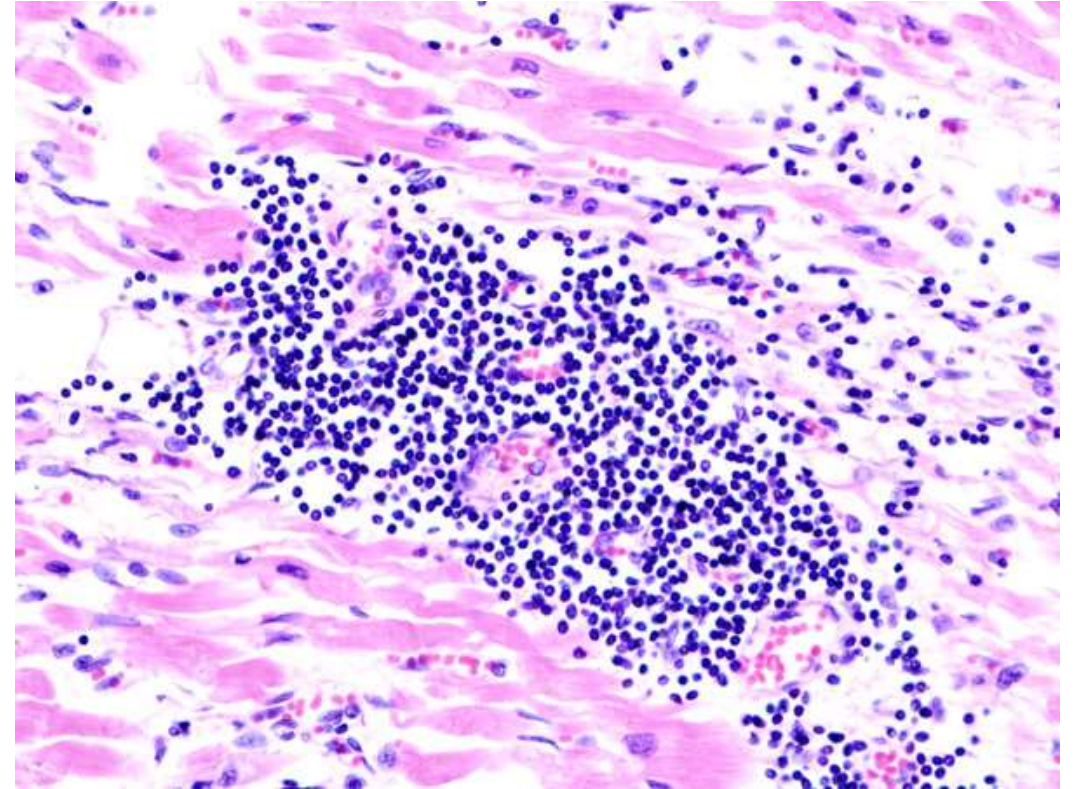




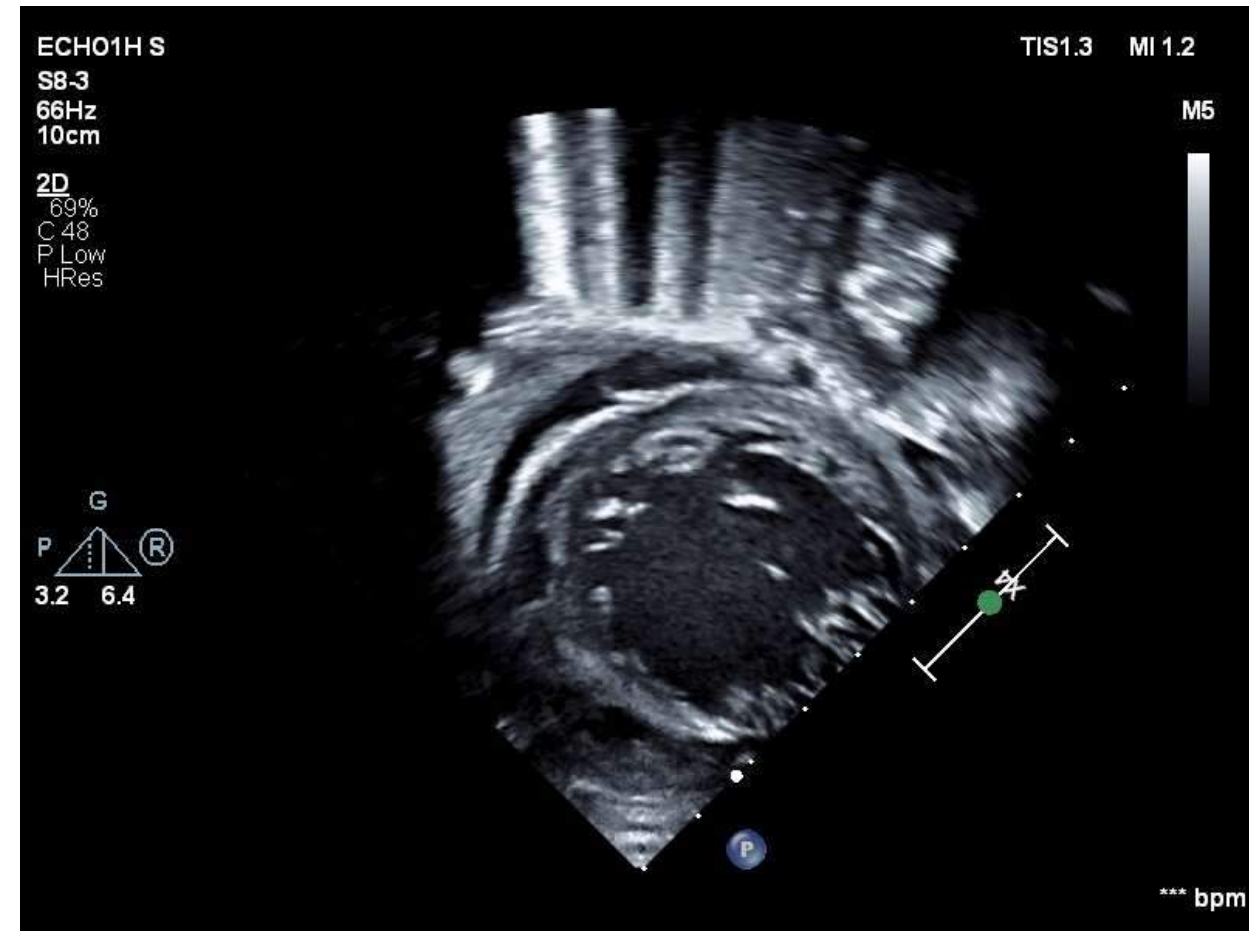
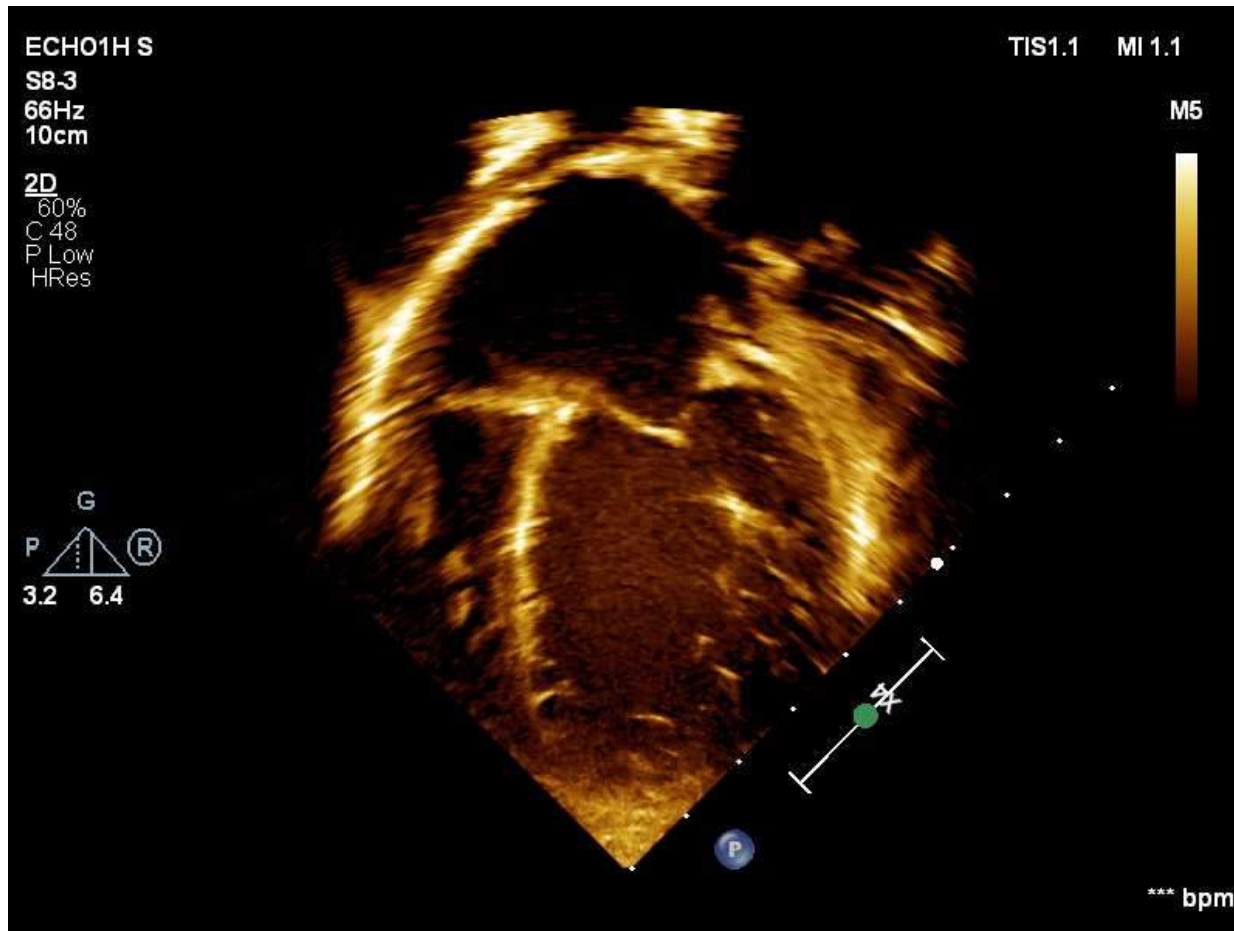
Comments: Significant ST-T wave changes in the lateral leads consistent with ventricular dysfunction versus cardiomyopathy. Possible lateral ischemia versus myocardial injury. Clinical correlation is strongly advised.

Partial Differential Diagnosis

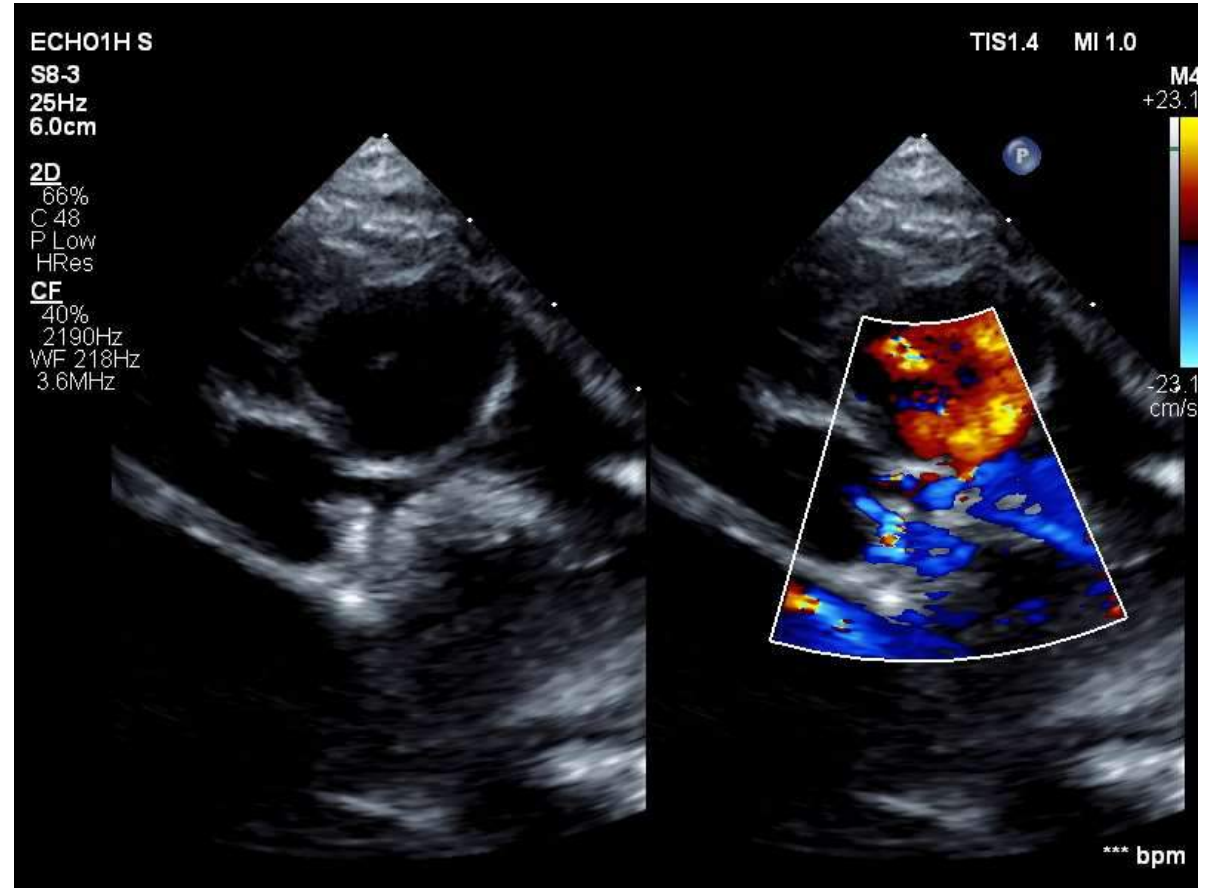
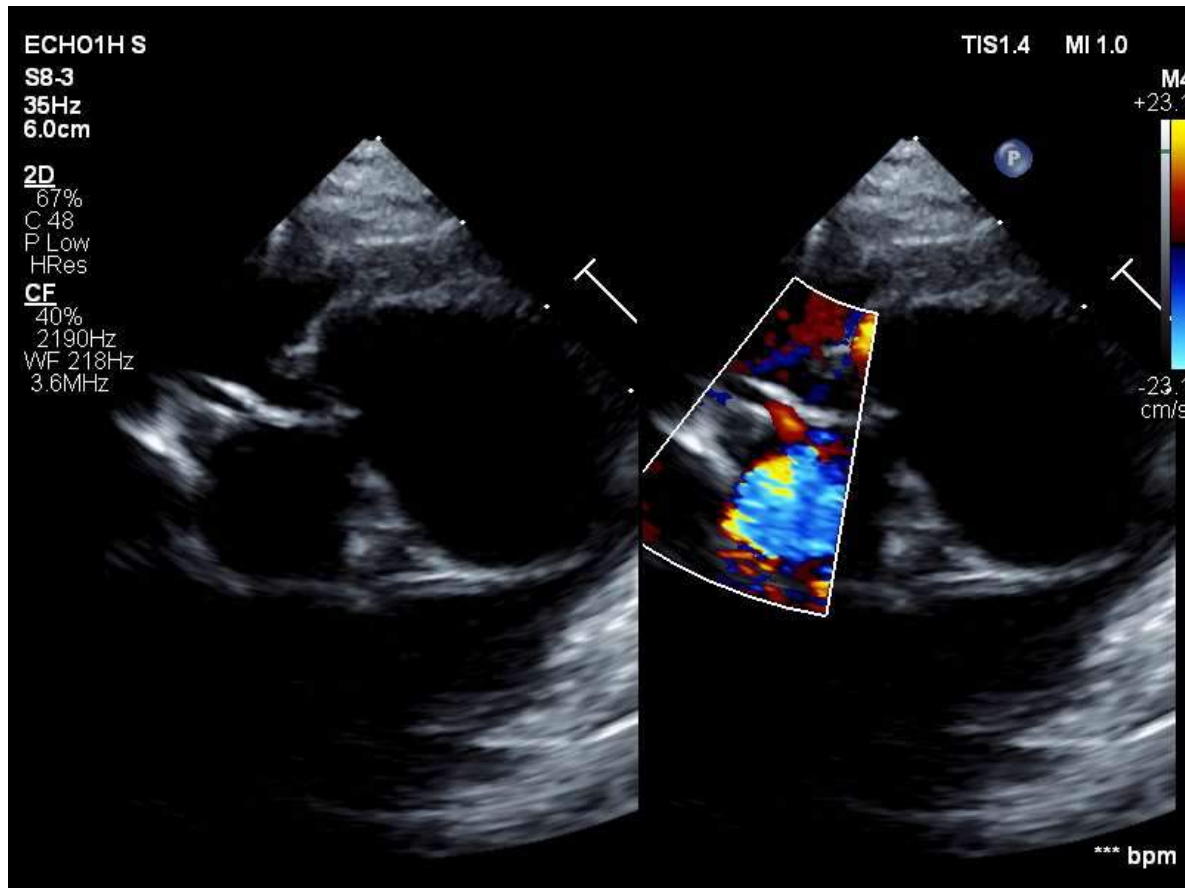
- Viral myocarditis
- Primary cardiomyopathy
- Myocardial ischemia
 - Congenital coronary anomaly (ALCAPA etc.)
 - Myocardial infarction (clot, dissection)
- Tachycardia-mediated cardiomyopathy
- Congenital heart disease (congenital MR, atypical Ebstein anomaly)



Echocardiogram



Echocardiogram



Echocardiogram: Summary

- Severe LV dysfunction and dilation, Severely dilated LA
 - LVEF 24%
 - LV end-diastolic volume 67 ml (z-score 6.8)
- “LV non-compaction”
- RV compressed by LV, moderately depressed RV function
 - RV hypertension, by TR jet ~40 mmHg plus RA v-wave
- LCA from aorta but more anterior/rightward than typical
- Bicommissural aortic valve (normal function), no coarctation
- Mild TR, Mild-moderate MR



Working diagnosis and next steps

- Presumed primary cardiomyopathy vs. myocarditis (viral studies –'ve)
- CICU: positive pressure ventilation, milrinone, low-dose dopamine/epi, diuretics
- Extubated PAD 6, weaned off epi/dopa, transferred to ward PAD 9
- Ward: Unable to wean off milrinone (feeding intolerance, tenuous fluid balance, intermittent tachycardia)
- Echocardiogram repeated 21 days after admission:
 - “Turbulent diastolic flow through the left coronary artery ostium with peak diastolic velocity of 160 cm/s. The anatomic and functional nature of the left coronary artery origin requires further investigation.”

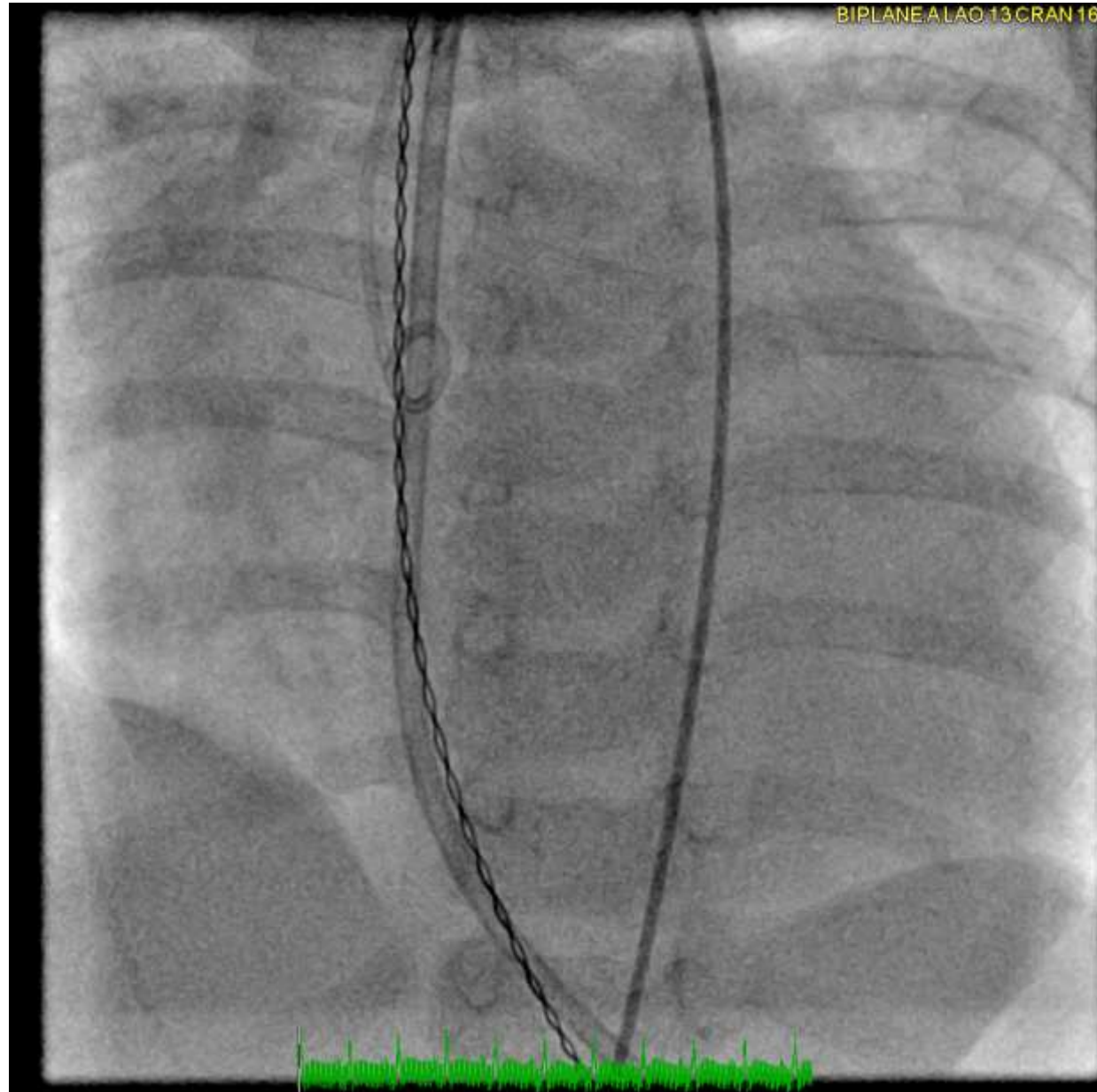


Cardiac CT

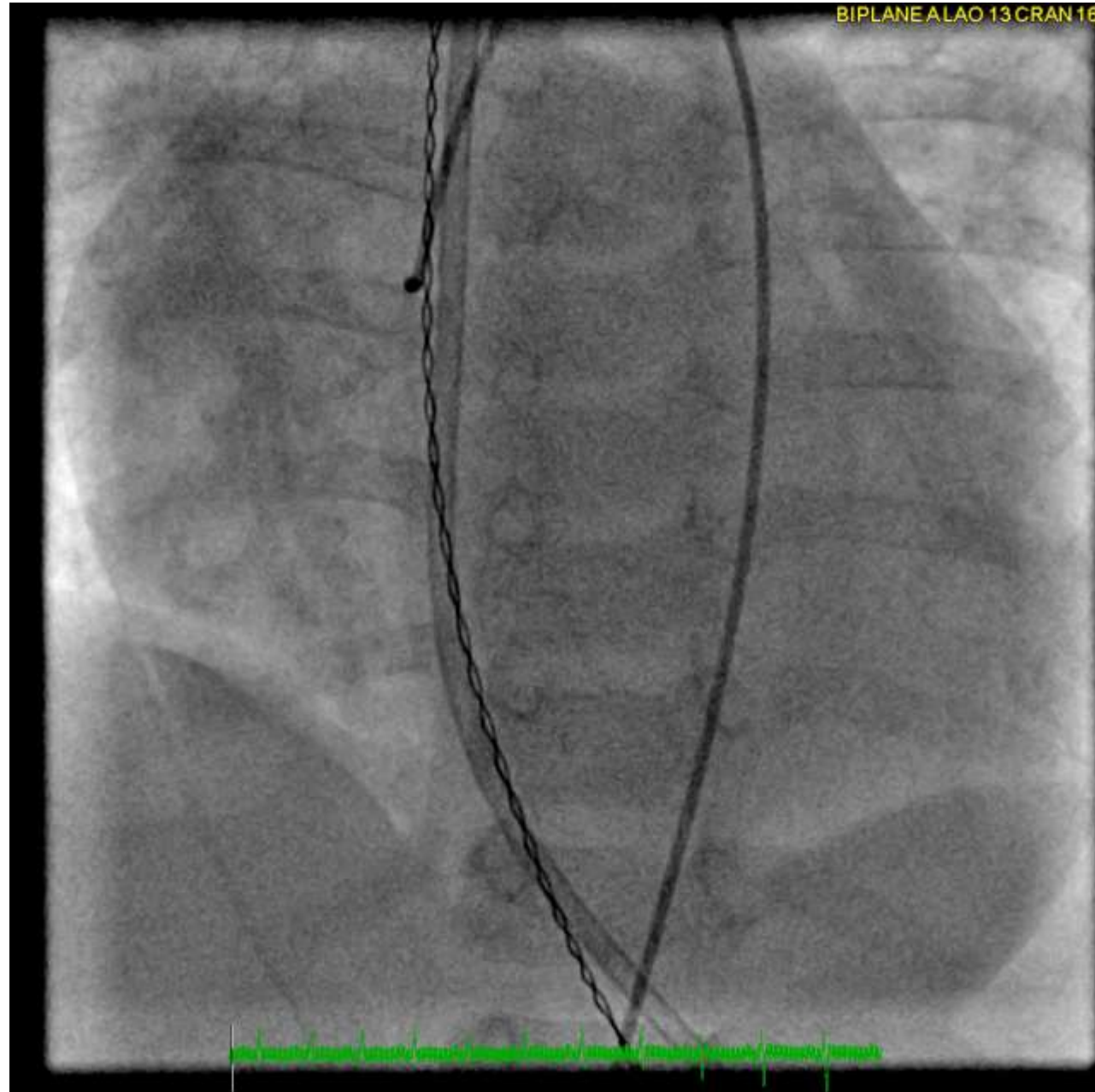


- Anatomy could not be clearly defined
- Concern for left coronary ostial stenosis

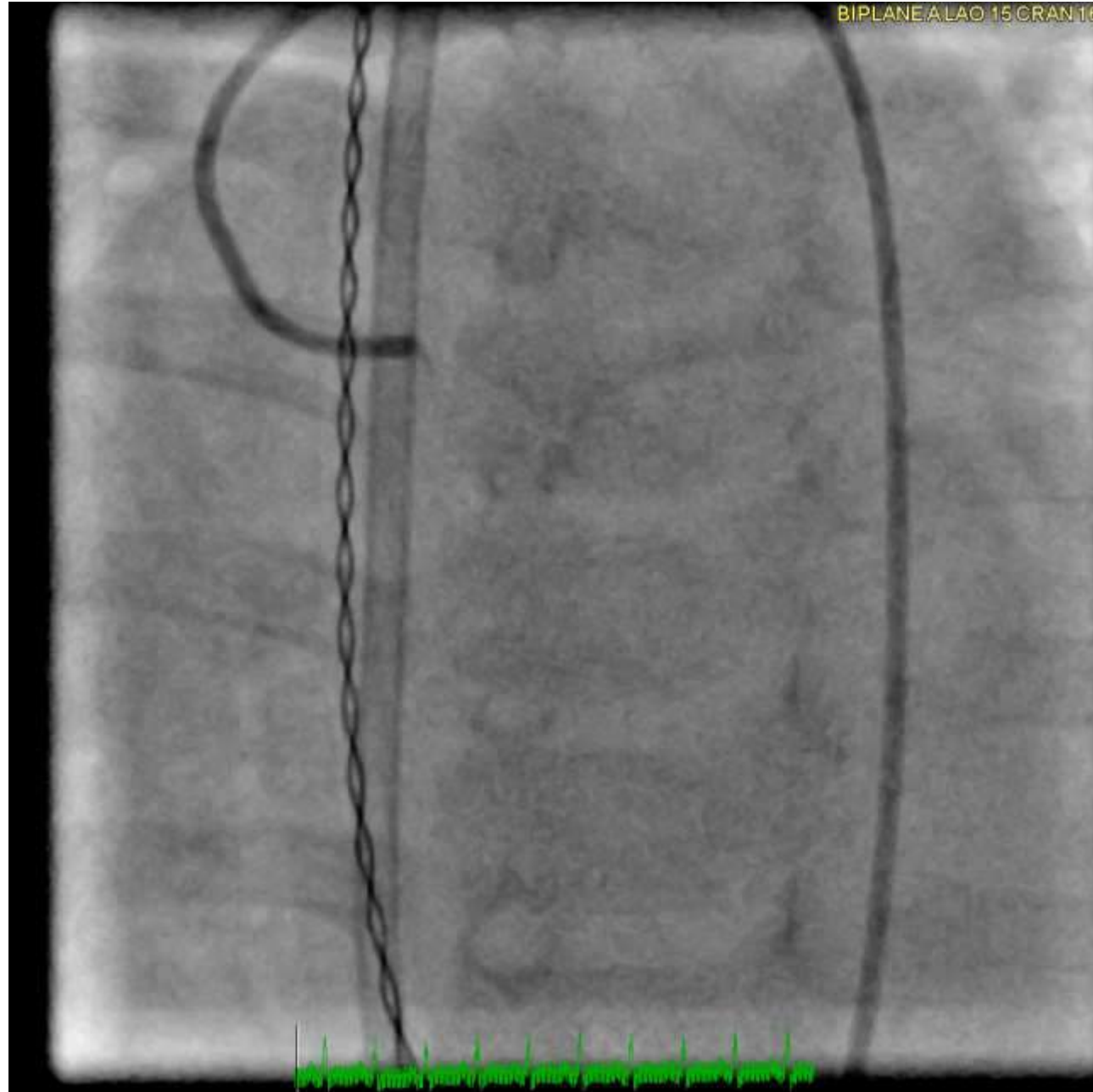
Angiography



Angiography

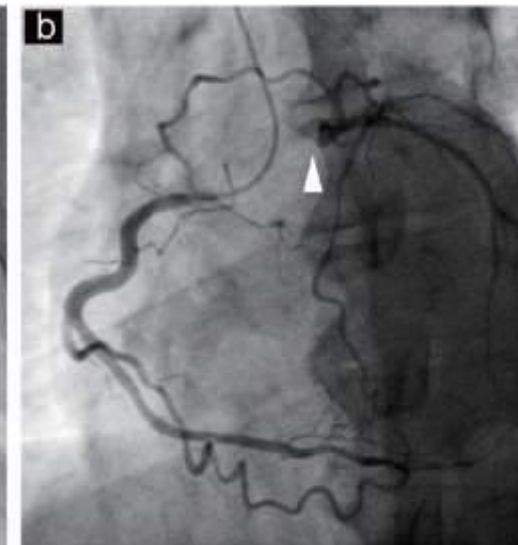
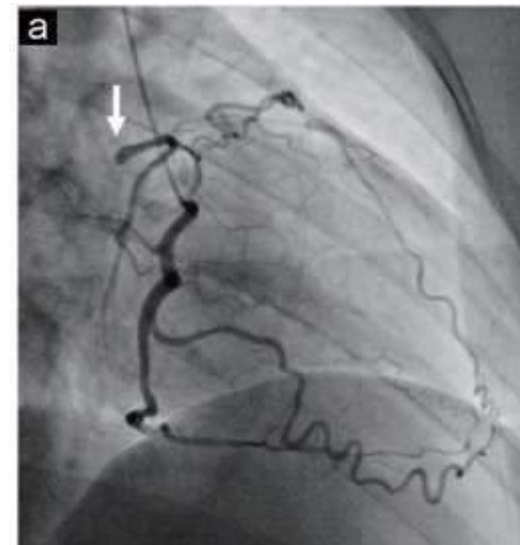


Angiography



Isolated congenital coronary ostial atresia/stenosis

- Rare congenital coronary anomaly
 - Much less common than ALCAPA (which occurs ~1:300,000 live births)
Karimi et al., World J for Ped Congen Heart Surg, 2015
- Largest case series has 5 patients
Hartz et al., JACC: Case Reports, 2021
- Has been described as a cause of neonatal mortality (autopsy)
Velumula et. al, Cureus, 2022
- Has also been reported in adults (>40 y/o) – CHF, sudden death
Horiuchi et al., J Med Cases, 2021



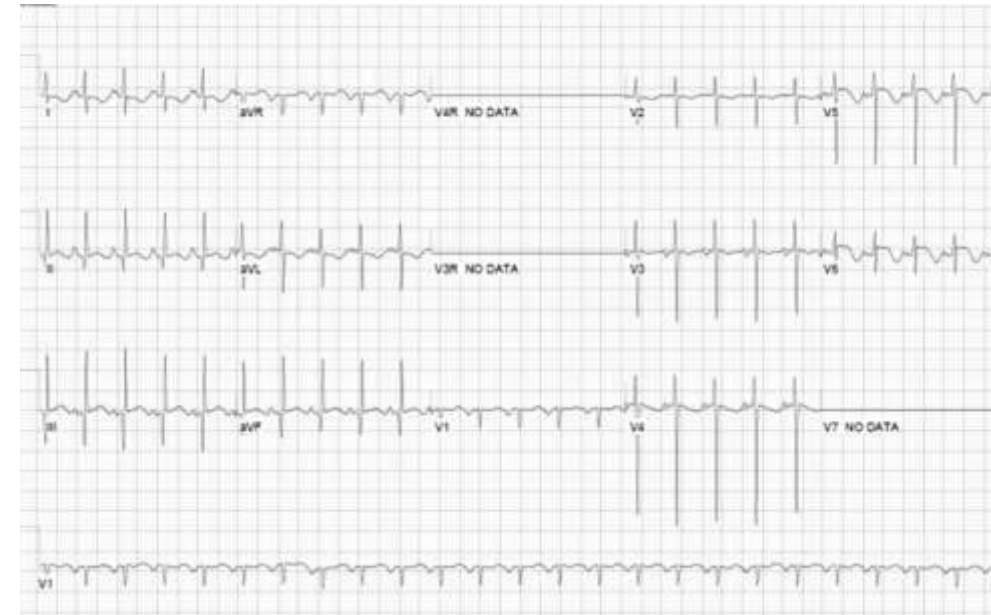
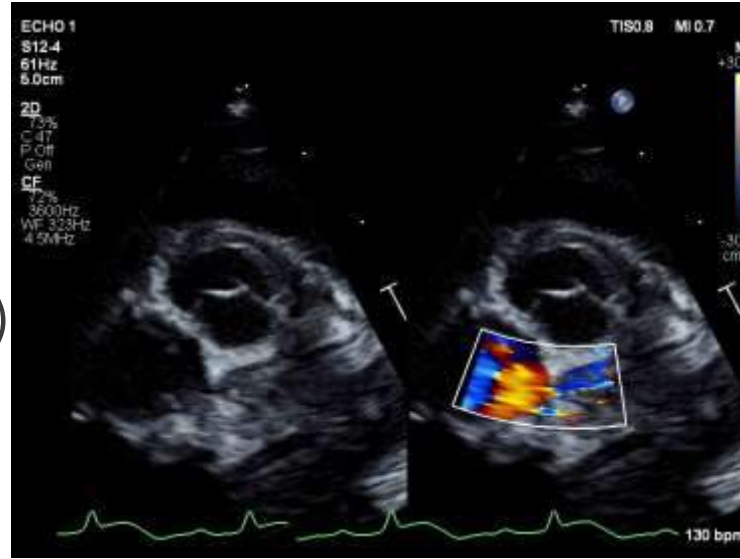
We made the diagnosis... eventually

- Extubated post-cath, debating surgical strategy
- 48 hrs post cath: acute deterioration with severe tachycardia, progressive metabolic acidosis, urgently intubated
- Cannulated to ECMO for persistent low output, lactic acidosis, hypoxemia
- Finally to OR
 - Ostium of the LCA was probe patent, and this was incised out into the bifurcation and then patched



Optimizing Care and Outcomes: What went wrong?

- “Common things being common...”
- False lead (recent viral illness)
- Alternative explanations (“LV non-compaction”)
- Inattentional blindness
 - Looking hard for **ALCAPA**...
- Silo effect
 - Limited secondary review of echo
- Inertia
 - We’ve ruled out a coronary issue... we have a plan...



Optimizing Care and Outcomes: Clinical Pearls

- Keep reversible causes of “DCM” on differential
 - Tachyarrhythmia
 - Coronary anomalies (ALCAPA and others)
- Respect the CXR and EKG
- Echo has imperfect sensitivity for coronary abnormalities
- Time matters when a coronary is compromised

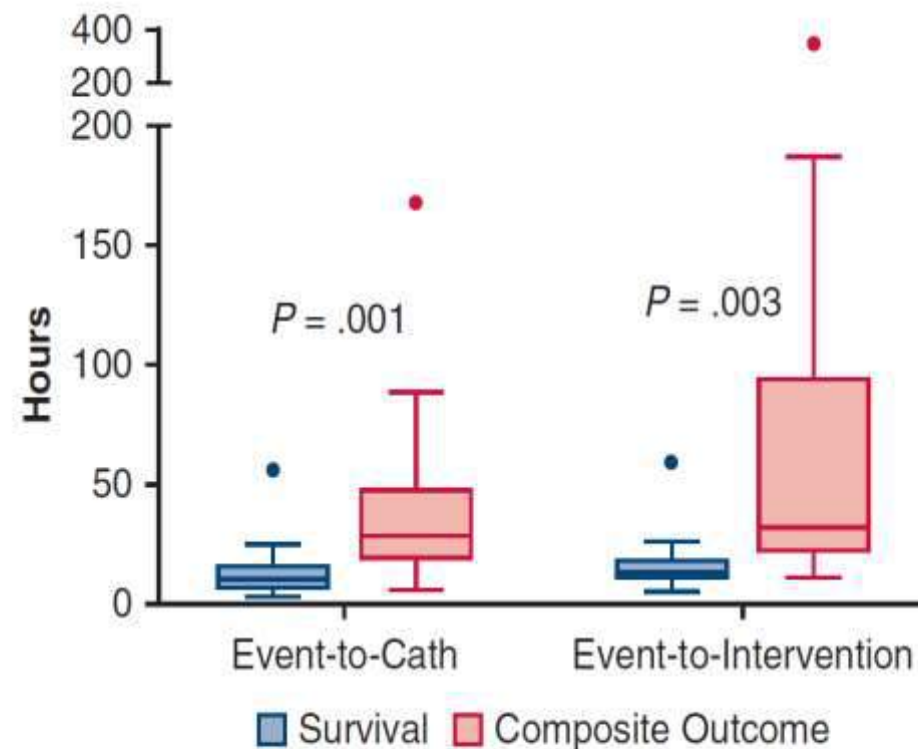


FIGURE 2. Box plot of time from sentinel event to cardiac catheterization and time from sentinel event to intervention, survivors versus those reaching composite outcome of death or listing for heart transplant.

Goldsmith *et al.*, JTCVS, 2019

Thank You!

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Boston Children's

Where the world comes for answers



What about our patient?

- Underwent LVAD placement a few days after coronary repair due to appearance of myocardium intraoperatively and no improvement in function
- LVAD could be removed about 2 months later after myocardial support and recovery
- Was listed for heart transplant, but eventually removed due to improved clinical status
- Most recent clinic echocardiogram... LV ejection fraction 63%