Right Ventricular Dependent Coronary Circulation in Pulmonary Atresia with Intact Ventricular Septum

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INTRODUCTION

- Pulmonary atresia with intact ventricular septum (PA/IVS) causes complete obstruction to right ventricular outflow thus an alternate source of pulmonary blood flow is required for survival
- PA/IVS accounts for 1-3% of all congenital cardiac defects
- Pathogenesis is unknown and is not associated with other genetic syndromes like other cardiac malformations
- **PA/IVS has a wide range of phenotypic cardiac defects such as degree of RV and tricuspid hypoplasia or presence of coronary artery malformations**
- PA/IVS causes RVOT obstruction and blood can only exit the RV via TR or through the RV to the coronary artery circulation (fistula/sinusoids)
- These patients are ductal dependent and rely on a patent ductus as the sole source of pulmonary blood flow

CASE DESCRIPTION

- 36wk M with cyanosis and desaturation to 52% at 24HOL. Post natal diagnosis of PA/IVS made and patient intubated and started on PGE

- TTE on DOL1 showed TV z score of -4.7 and large PDA (4mm throughout)
- Small/hypoplastic RV
- RCA unable to be imaged on TTE
- Network of vessels across RV with low grade antegrade/retrograde flow suggestive of sinusoids

- **Cardiac MRI obtained on DOL 2 showing: Absent RCA ostium w/ RV sinusoids and RV-DCC**
- PGE continued as pt had tortuous ductus and anatomy was too high risk for PDA stent

- Listed Status 1A for OHT,, transplanted at 2 months of age without complication

LEARNING OBJECTIVES

There is no standardized approach for correction/palliation of PA/IVS which makes recognition and initial evaluation crucial as some variants with RVDCC are uniformly fatal

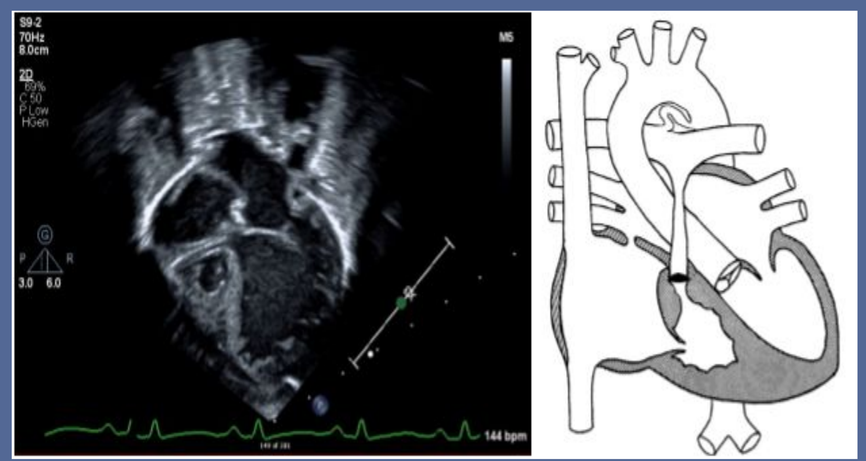

Figure 1- echocardiographic and pictorial 4-chamber representation of PA/IVS showing small plate like tricuspid valve and RV hypoplasia

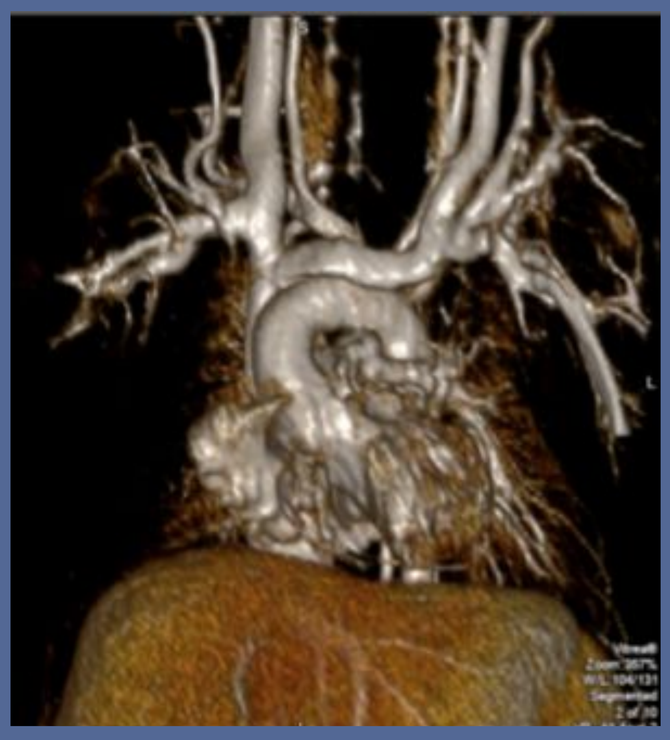

Figure 2- High-resolution 4D music and 4D flow cardiac MRI imaging using volume rendered reconstructions showing RV hypoplasia and tortuous PDA.

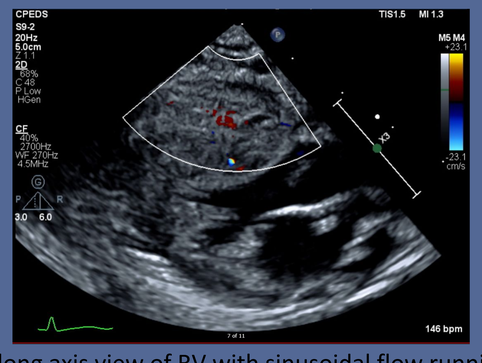

Figure 3- long axis view of RV with sinusoidal flow running across

DISCUSSION

- The obstruction of the RVOT from PA/IVS leads to RV hypoplasia and high pressures in the RV
  - Blood must escape from the RV via TR or via sinusoids
- **Sinusoids can fistulize with the coronary arteries causing stenosis, atresia, and RVDCC.**
  - **The coronary blood flow will now become RV-dependent**
- It is critical to identify RVDCC as any RV decompression may lead to coronary artery steal with resultant myocardial ischemia
- **This case of PA/IVS with right coronary ostial atresia and RVDCC necessitated OHT for survival**

- Initial stabilization begins with PGE
- Next, there are 4 important parts of the TTE evaluation
  - RV size
  - TV z-score
  - assessment of mixing via PFO/ASD and PDA
  - Coronary artery evaluation
- Significant dysplasia leads to higher RV pressures and increased likelihood of RV sinusoids and coronary artery abnormalities

- If coronaries are unable to be visualized on TTE further assessment is needed with catheterization
  - This determines if coronary anomalies and RV-DCC are present which will change the treatment course
- cMRI was used in this case as the infant was not a candidate for PDA stenting given its tortuous course

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