LATE-PRESENTING SINGLE VENTRICLE WITH SEVERE BILATERAL PA STENOSES AND HYPOPLASTIC RPA: PRE-OPERATIVE PA REHABILITATION WITH PDA STENTING AND BALLOON DILATION

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History and physical:

An infant with severe cyanosis was referred from a neighboring country at 8 months of age, weighing 5.3kg.

Imaging:

Diagnosis by echocardiography: pulmonary atresia with intact ventricular septum (PAIVS), hypoplastic right ventricle, pulmonary arteries from the PDA, bilateral stenoses of proximal pulmonary arteries. The right pulmonary artery (RPA) was very small, with minimal flow. Destination: single ventricle tract.

Indication for intervention:

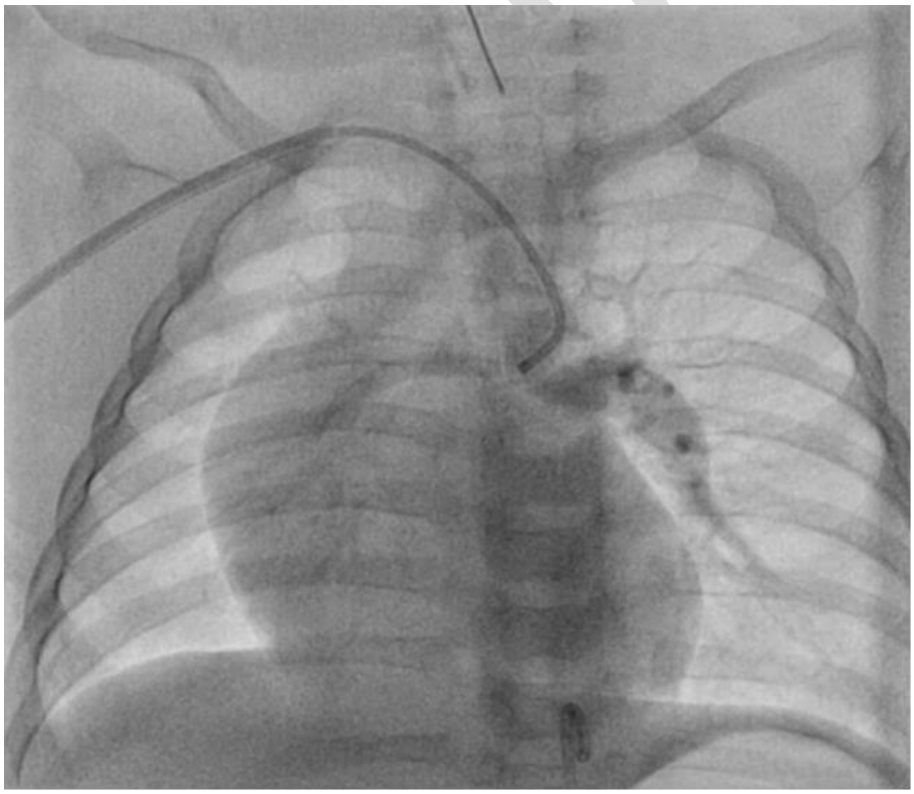
1. To improve cyanosis (SpO2 65%) with PDA stenting and balloon dilation of the pulmonary arteries.
2. To rehabilitate the pulmonary arteries in preparation for Glenn and Fontan operations.
3. To improve restrictive patent foramen ovale (PFO) with balloon atrial septostomy.

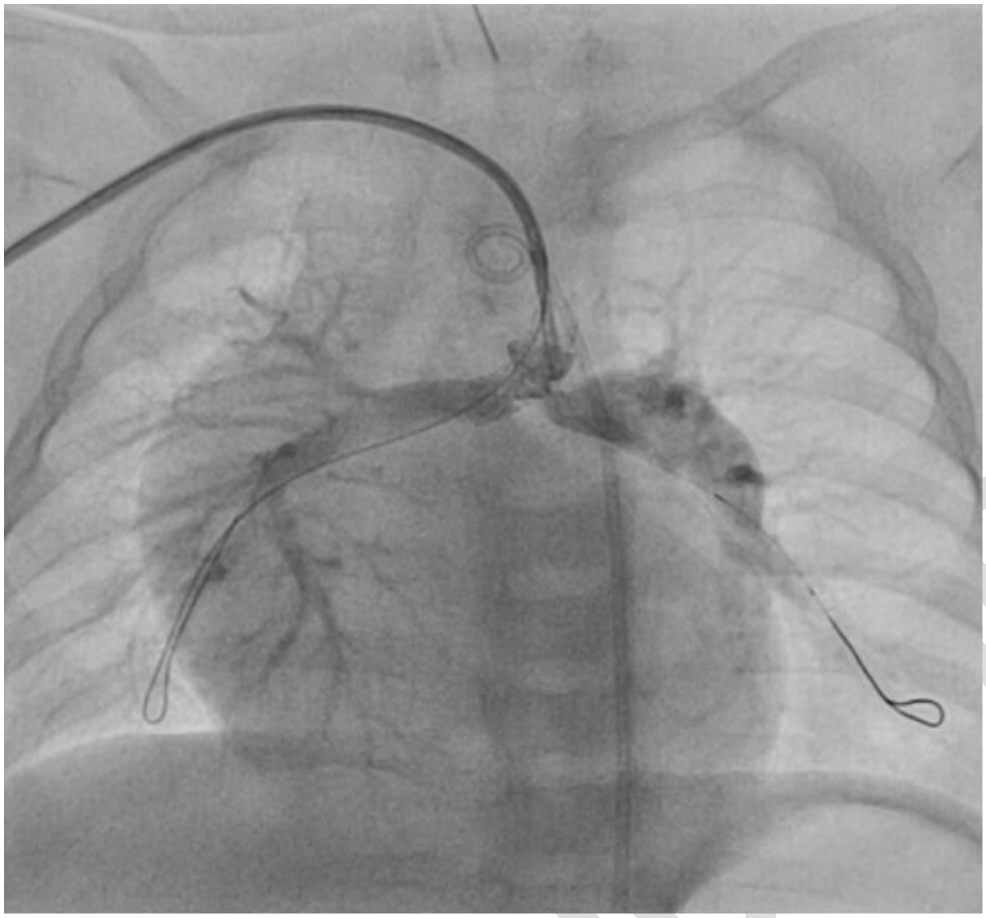
Intervention:

1. The right axillary artery was cannulated using a 5/4 French slender sheath for access to the PDA. A 4 French JR diagnostic catheter, maneuvered into the PDA, revealed the following anatomy:
    - The PDA originated from the underbelly of the aortic arch and was short and straight.
    - Both pulmonary arteries originated from the PDA.
    - The RPA was hypoplastic (3mm in diameter), almost occluded and hardly visible due to very tight stenosis at its origin from the PDA.

- The left pulmonary artery (LPA) had moderate-to-severe stenosis at its origin but adequately sized distally with good lung arborization.
- There was no main pulmonary artery.

2. To salvage the RPA for future Glenn shunt insertion, a balanced middle weight (BMW) 0.014' coronary wire was navigated into RPA & anchored in a distal branch. The 4 French JR catheter was exchanged with a 5 French JR guiding catheter for stent delivery. An Onyx 4.5 mmm x 15 mm stent was implanted into the RPA with good positioning.
3. Following stent delivery, there was no flow into the LPA due to jailing of the severely stenosed LPA origin. The BMW wire was then navigated into the LPA across the stent struts. Balloon dilatation of struts was performed with 3.5 x 15 mm Trek coronary dilatation catheter.
4. Final angiogram showed good flow into the RPA and LPA.
5. Lastly, balloon atrial septostomy was performed with a Z-5 Septostomy catheter.
6. Post-procedure, the patient had mild overshunting and was kept ventilated for 48 hours. Heparin was infused for 72 hours. Oral aspirin and clopidogrel was started after that. Pre-discharge echocardiography showed patent PDA stent, good flow into both PA branches, and distal RPA visualized.





Learning points of the procedure:

1. Ductal-dependent pulmonary circulation has a high incidence of branch PA stenosis at the site of PDA connection – mostly unilateral branch stenosis but bilateral in this patient.
2. PDA origin from the arch underbelly requires non-conventional arterial access from the right axillary artery.
3. Jailing of the LPA following stent deployment requires immediate rescue to prevent occlusion. Decision was made in this case for balloon dilation of the struts instead of LPA stenting.