International Resource Limitations Lead to Delayed Presentation of Rare Single Ventricle Physiology

Mary Breige O'Donnell-Smith, MD, Bonnie Brooks, MD, Koyelle Papneja, MD
Pediatric Cardiology, Department of Pediatrics
UCLA Mattel Children's Hospital, Los Angeles, CA

Background

- There is known association of Interrupted Aortic Arch (IAA) with Double Inlet Left Ventricle (DILV), Double Outlet Right Ventricle (DORV) and Transposition of the Great Arteries (TGA) (Marek, et al., 2021; Wernovsky, et al., 1995). It is exceedingly rare for three of these lesions to occur together.
- A late presentation of this constellation of lesions after the neonatal period, in the setting of ductal dependency with IAA, is even more uncommon.

The Case

- A 5-month-old male was born full-term in Kenya and was diagnosed with CHD at 1 week of life. With no available surgical option in his home country, his father moved him to the United States.
- Transthoracic and transesophageal echocardiography showed:
  - {S,L,L}, situs solitus, levocardia
  - DILV with superior-inferior L-looped ventricles
  - DORV with L-malposed great vessels arising primarily from the rudimentary left-sided RV
  - Hypoplastic aortic valve, root, and ascending aorta with IAA Type A
  - Patent ductus arteriosus
  - There was trivial left sided AVVR and normal LV systolic function.
- By catheterization, Qp:Qs was 2.4:1, the LVEDP was 10 mmHg, transpulmonary gradient was 35 mmHg, and PVRi 4.02 WUi.
- A Norwood procedure with a 4-mm Blalock-Taussig-Thomas shunt, Damus-Kaye-Stansel, atrial septectomy, and IAA repair was performed.
- LV dysfunction necessitated VA-ECMO for postoperative cardiogenic shock. He was decannulated on POD 2 and underwent delayed sternal closure on POD 5.
- He remained in the CTICU for 14 days and discharged home with good weight gain on oral diuresis and afterload reduction.

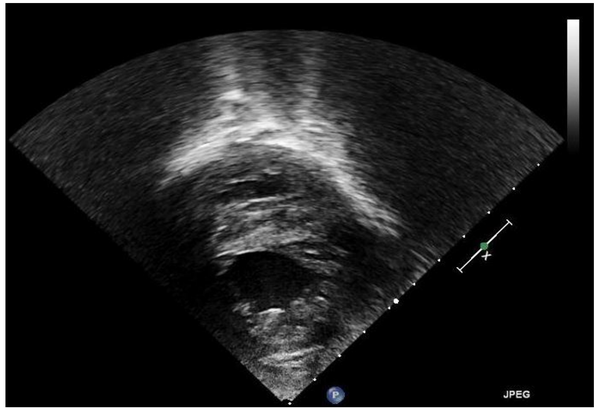

Figure 1: Subcostal short axis view of superior-inferior L-looped ventricles

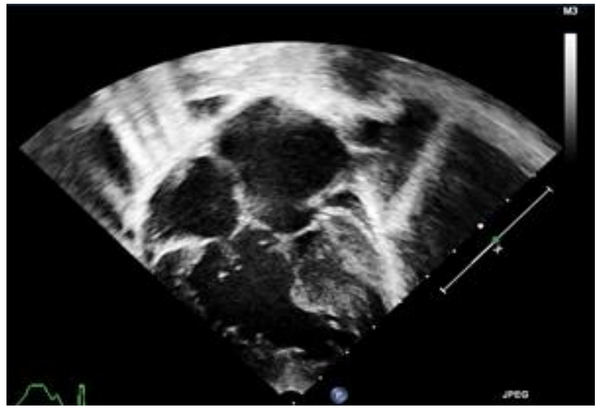

Figure 2: 2D apical 4 chamber view of DILV

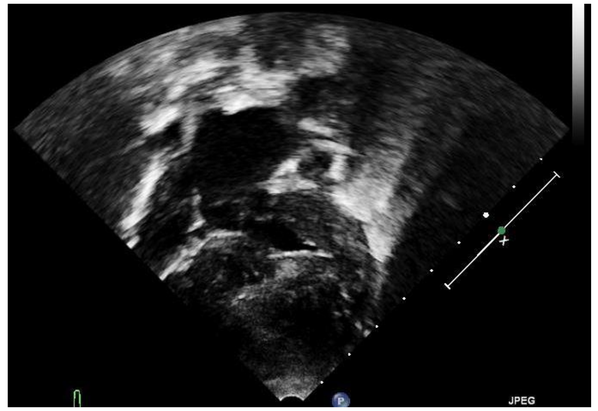

Figure 3: 2D apical 2 chamber view of the pulmonary artery arising from the rudimentary left-sided RV

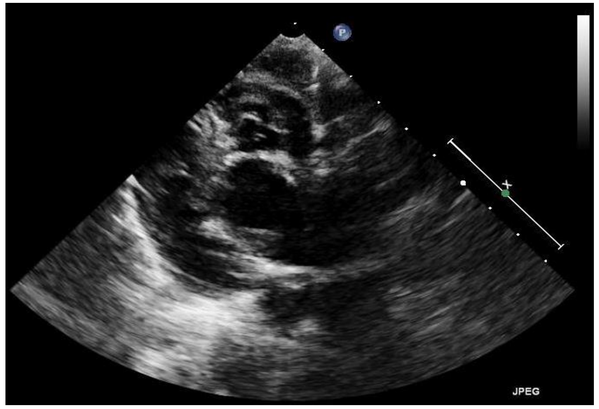

Figure 4: 2D parasternal short axis view of the hypoplastic aortic valve anterior to the pulmonary valve

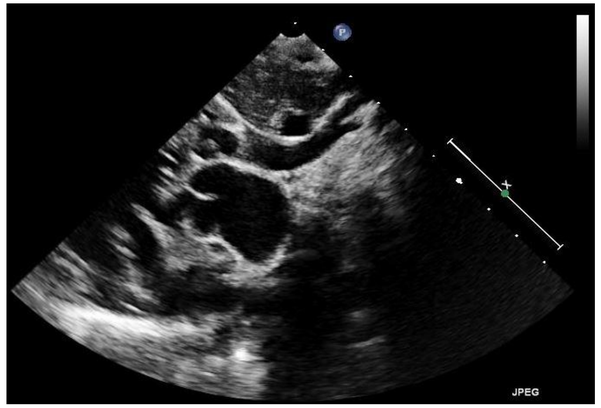

Figure 5: 2D high parasternal view showing parallel great vessels and a hypoplastic aortic valve, root, and ascending aorta

Conclusion

- This is a rare case of late-presenting, unrepaired single ventricle CHD with IAA and ductal dependent systemic circulation.
- Successful stage 1 surgical palliation was performed on another continent at 5 months of life.
- This case highlights the variability of congenital cardiac care on an international level.