Congenitally Abnormal Aortic Valve causing Coronary Obstruction and Cardiac Arrest in Infancy

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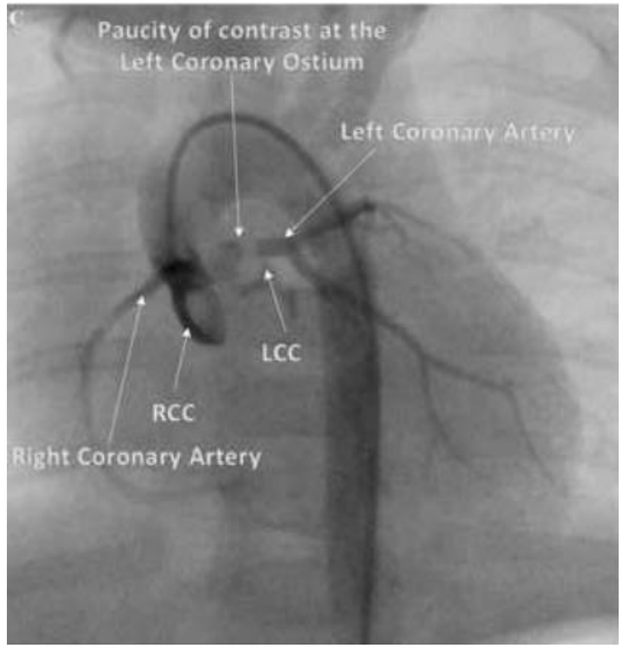
Advocate Children's Hospital
Tomorrow starts today.

Background

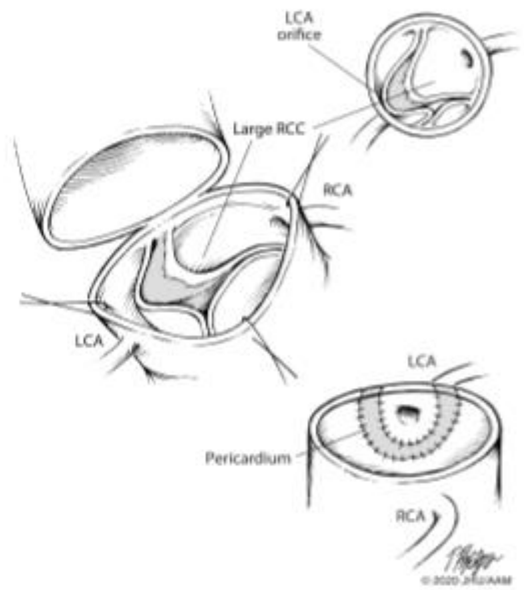
- Many instances of congenitally small coronary cusps causing ipsilateral coronary ostium occlusion are documented in the literature.
- Detection and treatment of an asymmetric, large aortic cusp causing obstruction of a coronary ostium in a symptomatic infant remains unreported.
- We present the diagnosis and emergent surgical intervention of a symptomatic two-month-old infant with a large, asymmetric right coronary cusp (RCC) occluding the left coronary ostium.

Case Presentation

- A two-month old female was reported unresponsive while being removed from a car seat, requiring road-side cardiopulmonary resuscitation for ventricular fibrillation.
- Following two subsequent episodes of ventricular arrhythmia while in the intensive care unit requiring CPR and defibrillation, an aortic root angiogram demonstrated a notable lack of contrast in the left coronary artery.
- Exploratory cardiac surgery demonstrated a relatively large RCC overriding a smaller left coronary cusp causing obstruction of the left coronary artery (LCA) ostium.
- A left coronary artery button was used to relocate the ostium more cephalad to overcome the overriding cusp and the left coronary sinus was reconstructed with autologous pericardium.



Aortic root angiogram showing paucity of blood flow at the left coronary artery origin. LCC, Left Coronary Cusp; RCC, Right Coronary Cusp



Operative Findings and Repair: Large right coronary cusp (RCC) overriding the left coronary cusp, thereby obstructing the left coronary ostium. The left coronary ostium was relocated more cephalad and the sinus of Valsalva was reconstructed using autologous pericardium. LCA, Left coronary artery; RCA, Right coronary artery.

Discussion

- Previous case studies describe treatment of aortic cusp attachments causing ipsilateral coronary obstruction in older patients.
- These cases either had less severe and emergent presentation, were identified and corrected at an older age, or utilized more invasive surgical procedures to conduct an operative repair.
- Due to our experience with coronary button translocation in the arterial switch procedure, we surmised that cephalad transplantation of the coronary button and subsequent reconstruction of the sinus of Valsalva would sufficiently restore left coronary blood flow.
- This approach preserved the aortic valve and the integrity of the right side of the heart.

Conclusion

- Creating a coronary button to relocate the LCA ostium allowed for successful repair while preserving native valve function and coronary button translocation should be considered an advantageous surgical technique for coronary ostial obstruction.

Follow Up

- Ten days following surgery, the patient was discharged home. She remains asymptomatic with normal cardiac function and absence of aortic valve dysfunction five months later