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Surgery

Surgical Repair of Severe Supravalvular Aortic Stenosis with Bicuspid Aortic Valve and Single Coronary Artery in a 5-Year-Old Child

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Abstract | **Case Report**

Background: Supravalvular aortic stenosis (SVAS) is a rare form of left ventricular outflow tract obstruction in children. Its coexistence with a bicuspid aortic valve (BAV) and a single coronary artery is uncommon and presents significant surgical challenges. ***Case Summary:*** We report a 5-year-10-month-old child with severe SVAS associated with BAV, post-stenotic aortic dilatation, and a single right coronary artery giving rise to the left coronary artery. Multidetector CT angiography confirmed the anomalous coronary anatomy. The patient underwent median sternotomy, supravalvular aortoplasty (Brom's procedure), single-patch augmentation of the non-coronary sinus and ascending aorta, and PDA ligation. Postoperative echocardiography demonstrated a peak gradient of 2–4 mmHg across the left ventricular outflow tract with no residual aortic regurgitation. The patient had an uneventful recovery. ***Conclusion:*** Single-patch supravalvular aortoplasty with PDA ligation is an effective and safe surgical strategy in children with SVAS, BAV, and anomalous coronary anatomy, providing relief of obstruction while preserving valve and coronary function.

Keywords: Supravalvular aortic stenosis; Bicuspid aortic valve; Single coronary artery; Pediatric cardiac surgery; Brom's procedure.

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INTRODUCTION

Supravalvular aortic stenosis (SVAS) is the least common form of congenital left ventricular outflow tract obstruction. It may occur as part of Williams–Beuren syndrome or as an isolated lesion and is often associated with other cardiovascular anomalies, including bicuspid aortic valves and coronary artery anomalies.

The combination of SVAS, BAV, and a single coronary artery is rare, making surgical management challenging due to the risk of coronary compromise during aortic reconstruction. Various surgical techniques, including single-patch, two-patch, and three-patch reconstructions, have been described. We present a case of a 5-year-old child with this rare combination, managed successfully with Brom's supravalvular aortoplasty, single-patch augmentation, and PDA ligation.

Patient History

The patient is a 5-year-10-month-old child, full-term spontaneous vaginal delivery (FTSVD), birth weight 3.7 kg, with an uneventful perinatal history. A murmur was noted at birth; echocardiography revealed moderate aortic stenosis and a patent ductus arteriosus (PDA).

The child remained asymptomatic on regular follow-up under the care of HSA. Recent imaging demonstrated severe SVAS with post-stenotic aortic dilatation and a bicuspid aortic valve. Multidetector CT angiography revealed a single right coronary artery giving rise to the left anterior descending artery.

During a multidisciplinary team discussion in September 2025, the patient was identified as high-risk for surgery due to the coronary anatomy. The surgical plan included supravalvular aortoplasty and PDA ligation.

Operative Findings and Procedure

With the patient supine under general anesthesia, a right radial arterial line and right internal jugular venous line were placed. A median sternotomy was performed, a large thymus removed, and the pericardium opened. The heart was moderately enlarged with normal pulmonary and systemic venous drainage. The ductus arteriosus was patent. Pronounced

constriction of the ascending aorta at the sinotubular junction was noted; the pulmonary arteries were normal.

Following heparinization, the aorta and right atrium were cannulated and cardiopulmonary bypass (CPB) established. The patient was cooled, and the aorta cross-clamped. Half-dose del Nido cardioplegia was delivered via the aortic root, with subsequent cardioplegia via the coronary ostia.

Transection of the ascending aorta revealed severe circumferential stenosis at the sinotubular junction with a thick fibrotic ring. A bicuspid aortic valve was identified with fusion of the left and non-coronary cusps, restriction of the right cusp, and a single right coronary artery supplying the left system. Commissurotomies and shaving of thickened leaflet edges were performed, allowing passage of an 11 mm Hegar dilator.

The non-coronary sinus was augmented with a shield-shaped Cardiocel patch secured with 5/0 polypropylene, extending into the ascending aorta. The ascending aorta was reconstructed and anastomosed to the sinotubular junction. PDA ligation was performed.

The patient was rewarmed, de-aired, and weaned off CPB successfully with milrinone and adrenaline support. CPB time was 71 minutes and cross-clamp time 46 minutes. Postoperative echocardiography showed a peak left ventricular outflow tract gradient of 2–4 mmHg with no residual aortic regurgitation and preserved left ventricular function. The chest was closed in layers with placement of temporary epicardial pacing wires.

Postoperative Course

The patient tolerated the procedure well. Hemodynamics remained stable postoperatively, and no complications were noted. The patient was transferred to the cardiac ICU in stable condition. Early follow-up echocardiography confirmed satisfactory relief of obstruction and preserved valve and ventricular function.

DISCUSSION

This case illustrates the rare association of SVAS, bicuspid aortic valve, and single coronary artery in a pediatric patient. The anomalous coronary anatomy posed significant surgical risk, requiring careful planning to avoid compromise during aortic reconstruction. Brom's single-patch supravalvular aortoplasty allowed effective relief of obstruction while preserving valve and coronary function.

The surgical outcome demonstrates that even in complex anatomy, careful intraoperative assessment, patch augmentation, and valvuloplasty can achieve excellent short-term results. Long-term follow-up is essential to monitor valve function and detect potential recurrence of obstruction.

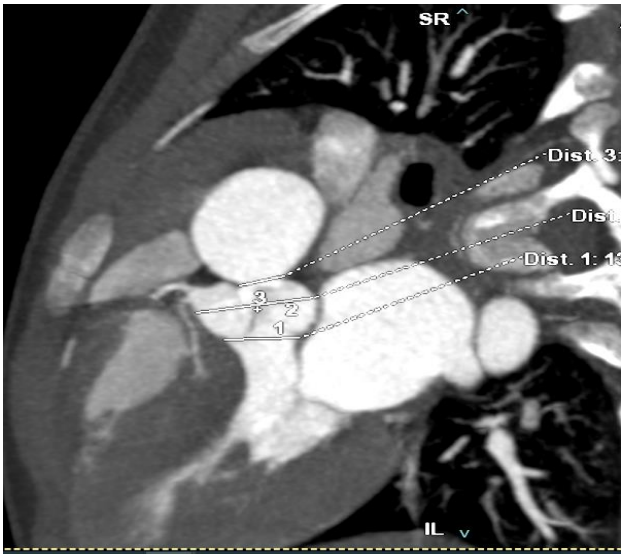
CONCLUSION

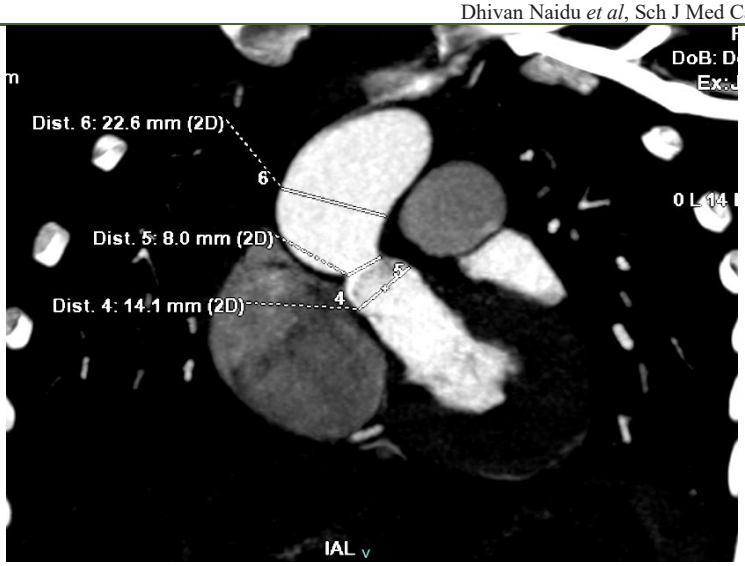
Supravalvular aortic stenosis associated with bicuspid aortic valve and single coronary artery is rare but can be safely managed surgically in children. Single-patch augmentation of the non-coronary sinus with Brom's aortoplasty and PDA ligation provides effective relief of obstruction, preserves aortic valve function, and avoids coronary compromise.

Operative Details:

- CPB time: 71 minutes
- Cross-clamp time: 46 minutes
- Valve competence: No residual regurgitation

Figure Legends

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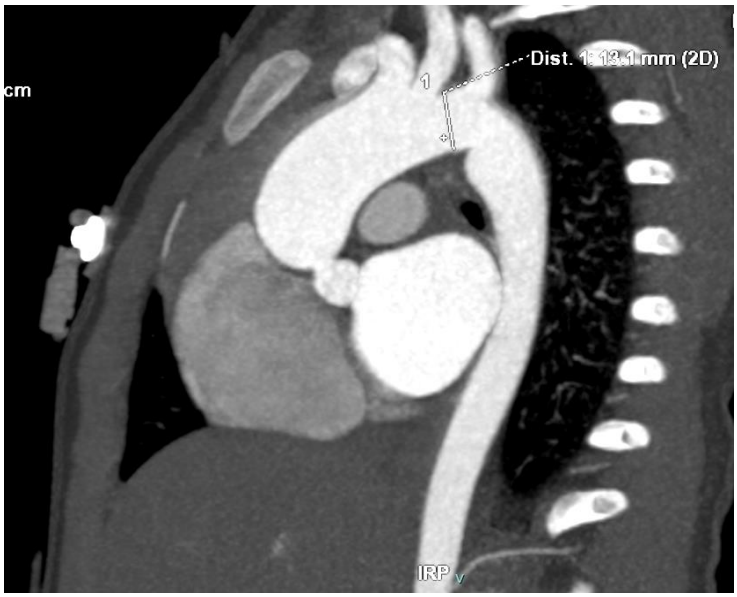
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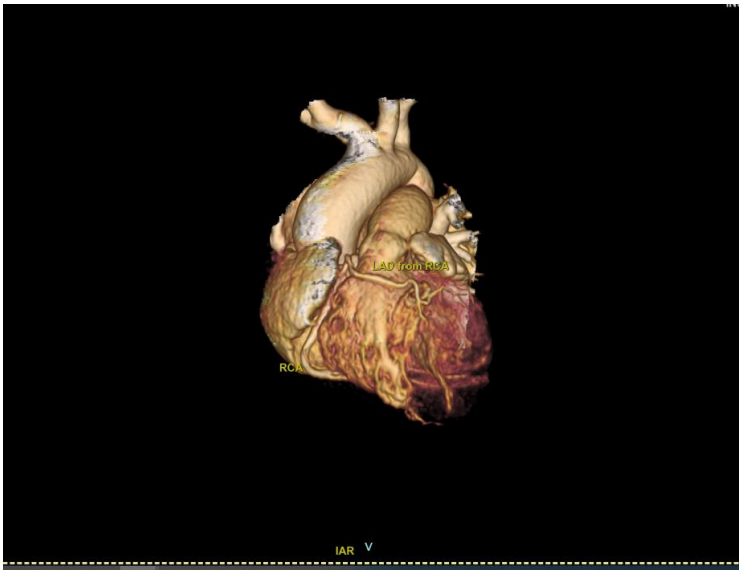
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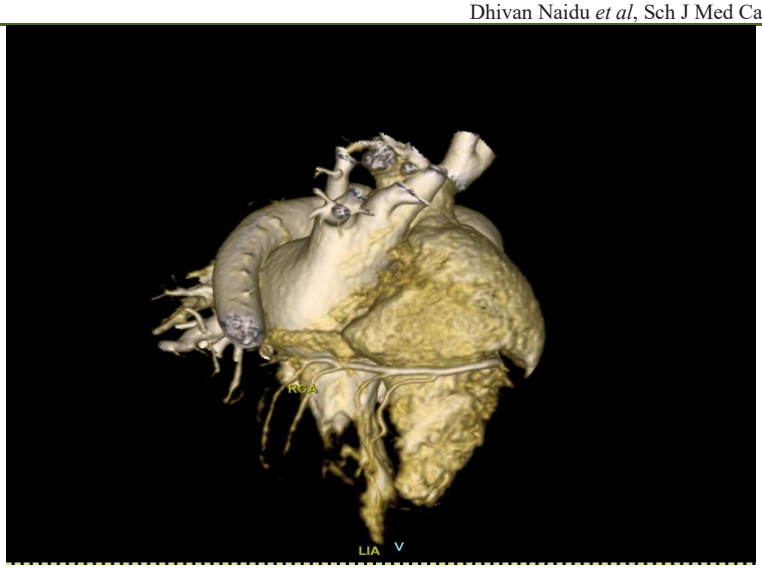
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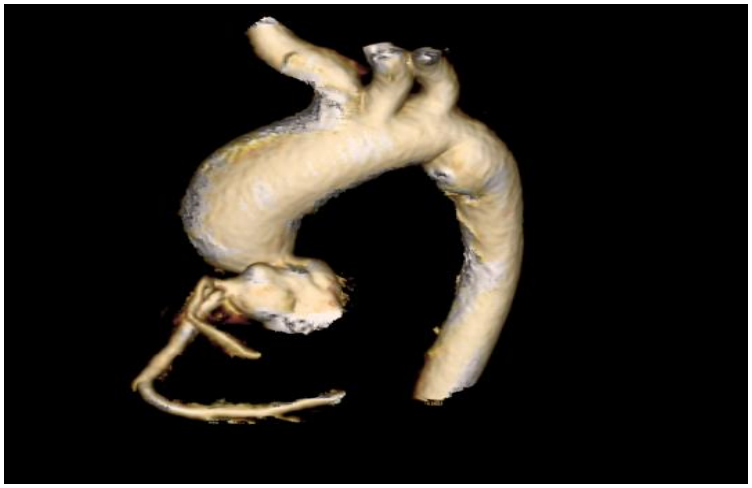

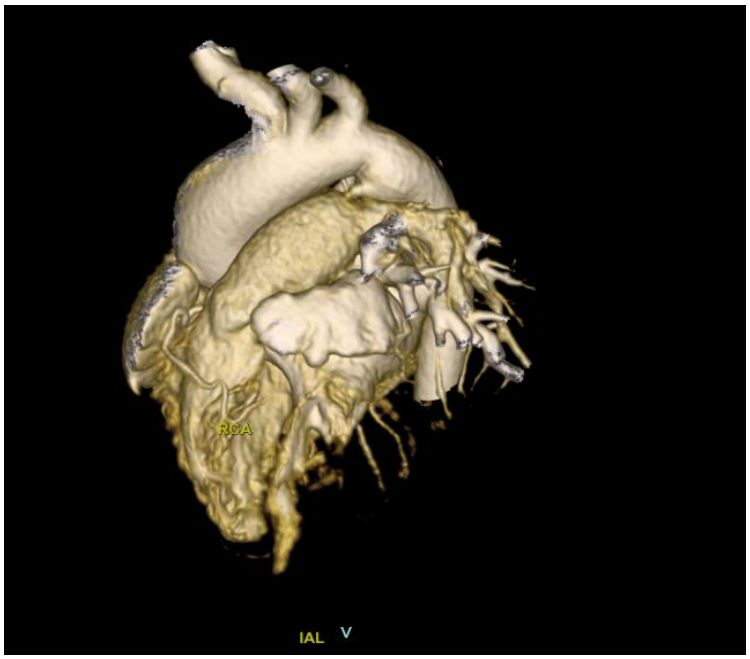
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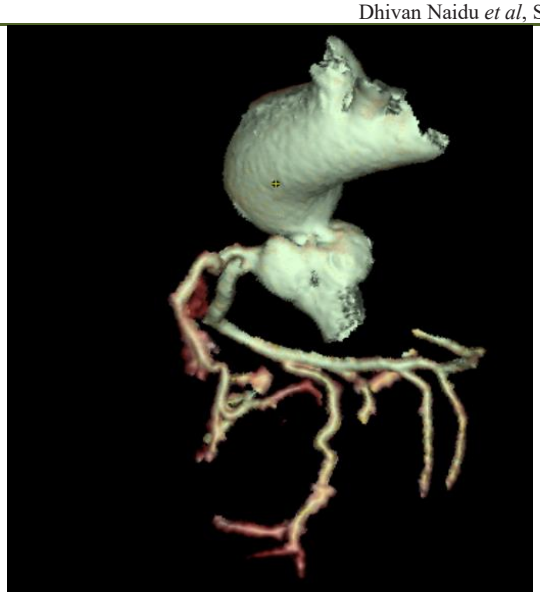
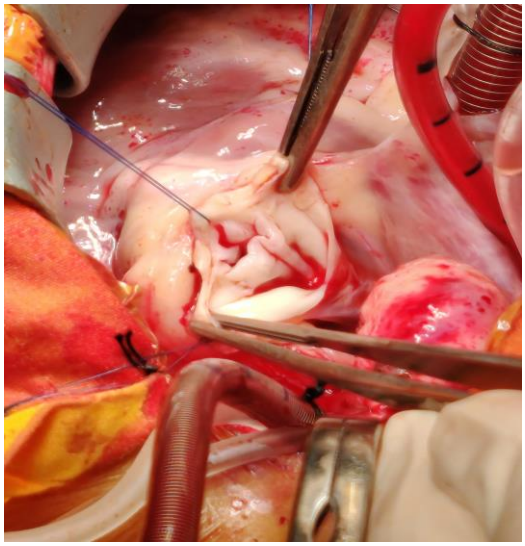


Operative images



Declarations

Conflict of Interest: The authors declare no conflicts of interest.

Funding: None.

Ethical Approval: This study was reviewed by the National Heart Institute, Institut Jantung Negara Research Ethics Committee (IJNREC) and was waived from full ethical review. Approval to proceed was granted.

Informed Consent: Written informed consent was obtained from the patient's parents or legal guardians for publication of this case report.

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