

Complex Repair of a Calcified Aortic Arch in an Adolescent Single Ventricle Patient

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Case Presentation

Past Surgical History

15yo girl with HLHS s/p four previous open-heart surgeries for single ventricle palliation, most recently an intra/extracardiac fenestrated Fontan procedure.

Presentation

Found to have Fontan baffle narrowing along with extensive coral-reef-like reactive calcifications of the bovine jugular vein graft used for reconstruction of the ascending aorta and aortic arch.

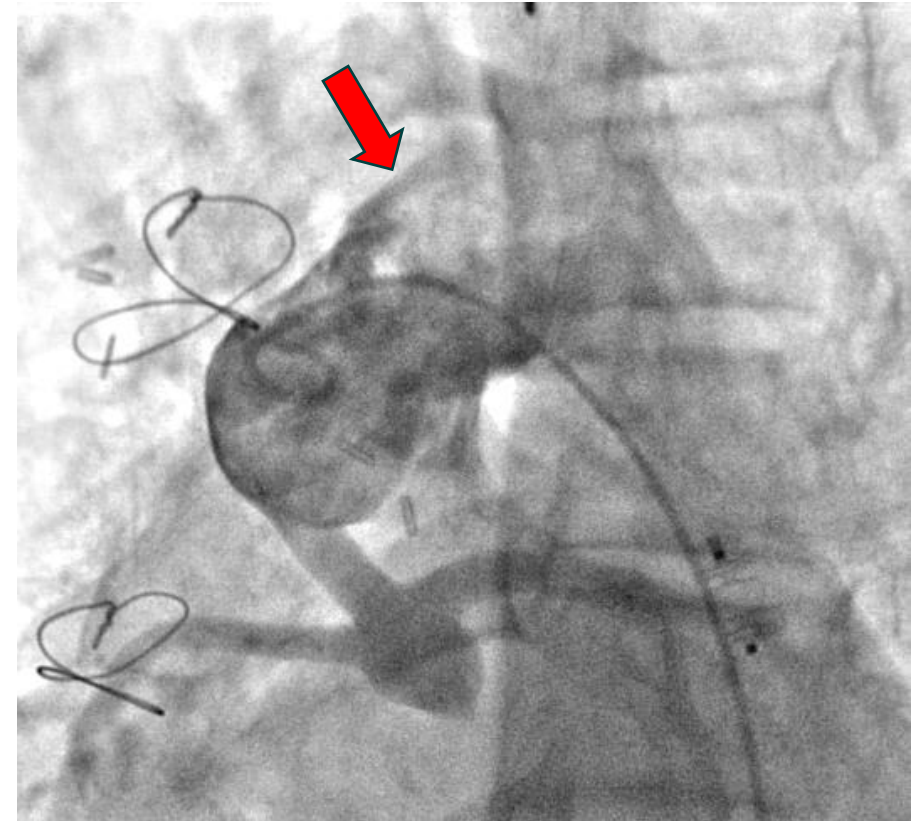
Management

Subsequently underwent successful arch repair, DKS revision and Fontan baffle stenting.

Case Presentation

Diagnostic catheterization findings in October 2020

Severe calcification of the patient's ascending neo-aortic arch. Obstruction not amenable to stenting given its proximity to the DKS anastomosis and neo-aortic valve.



Case Presentation

Routine catheterization findings in May 2023

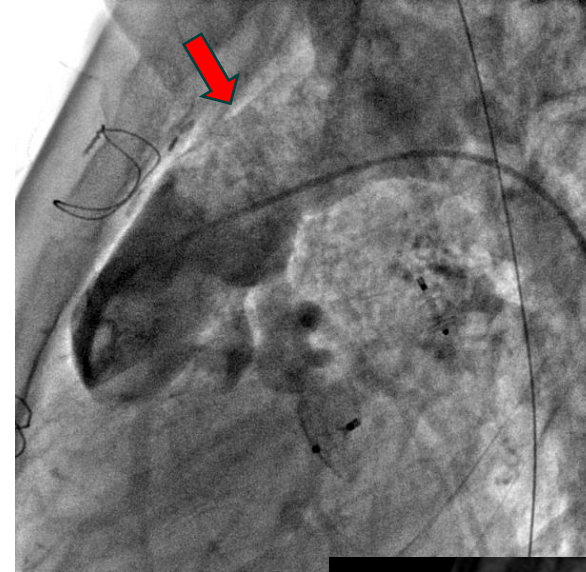
Elevated RVEDP, upper normal Fontan pressures and a 30mmHg gradient across the ascending aorta.

CT imaging findings

4.4 x 1.7 x 1.9cm calcification of the ascending arch.

Multidisciplinary conference recommendation

Recommendation for hybrid approach including stenting of the Fontan baffle followed by hemi arch replacement.



Step 1: Fontan baffle stenting

Balloon assessment

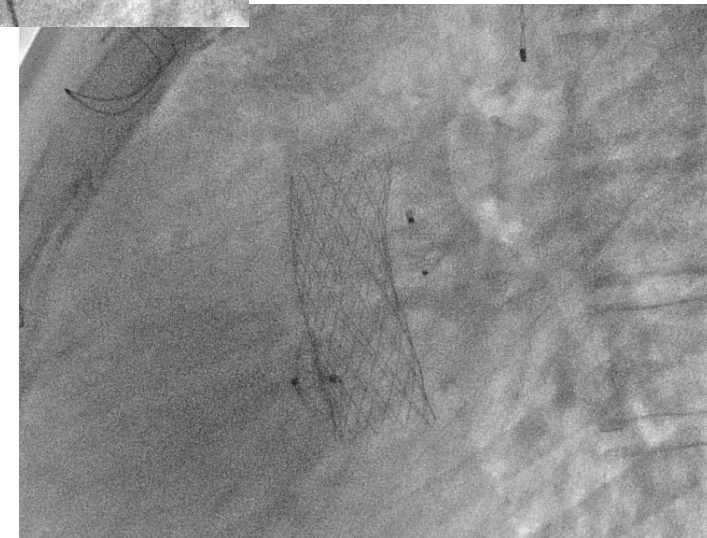
16mm x 4cm Z-Med II balloon showing a persistent waist at peak inflation.

Stenting

Positioning of a Palmaz 5010 stent across the Fontan.

Stent dilation

Serial dilation of the stent with follow-up angiography demonstrating a well-positioned stent and improved Fontan baffle diameter.



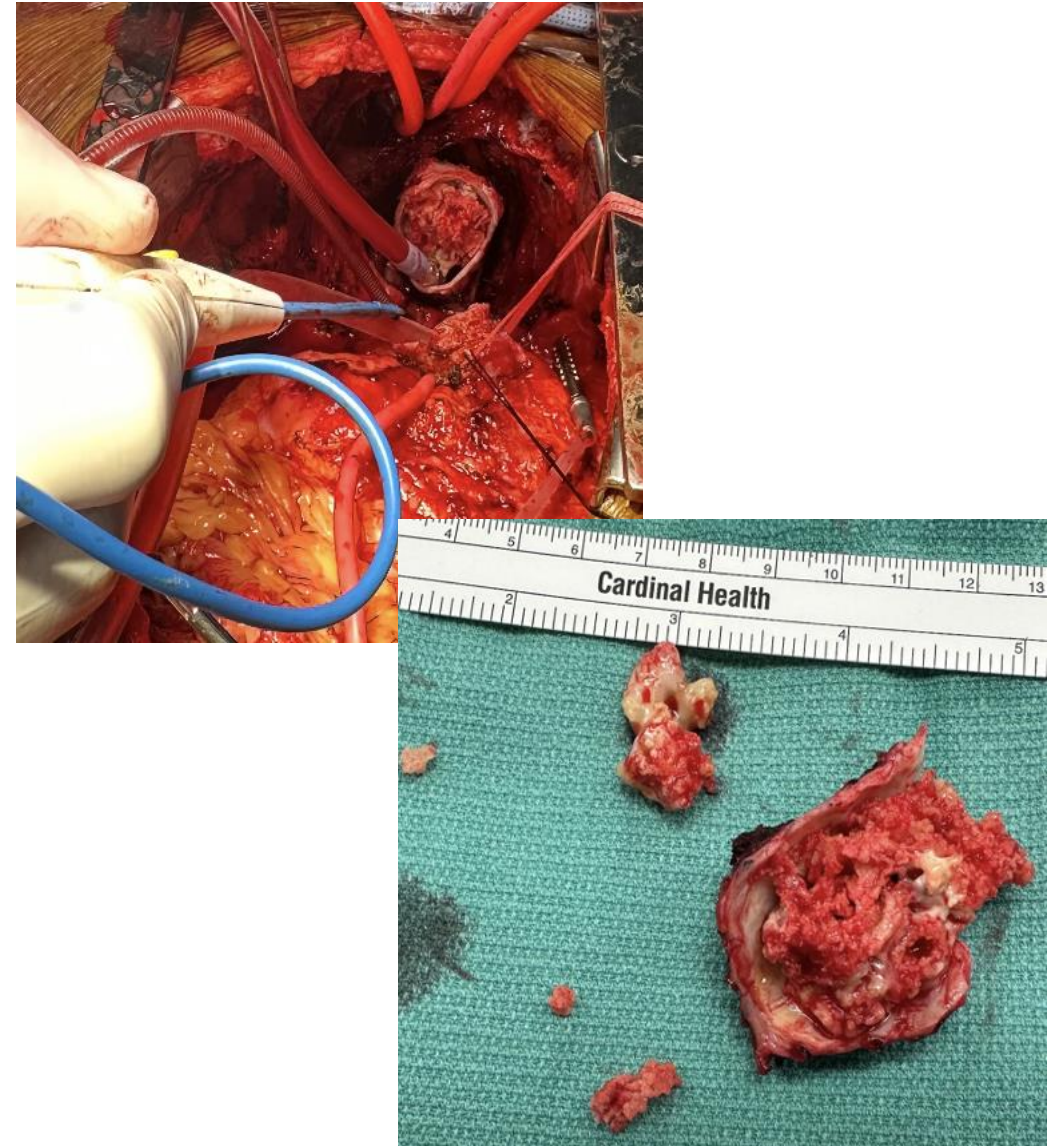
Preoperative Planning

Virtual and printed 3D models of the aorta to assist with operative planning.



Step 2: Operative Repair

- Fifth median sternotomy with aortic no-touch technique to minimize risk of embolization.
- Right axillary artery cannulation and femoral vein-Fontan baffle cannulation for venous drainage.
- Near-total obstruction of the ascending aorta with a minimal (3-4mm) residual lumen.
- En bloc resection of the base of the arch and ascending aorta.
- Anastomosis of a 26mm Gelweave graft to the distal aorta and arch vessels.



Postoperative Outcome

- The procedure was performed successfully with avoidance of any neurologic event.
- The patient's post-operative course was notable for a video swallow study revealing aspiration with thin liquids.
- She was last seen in outpatient clinic on post-operative day 50 where she was continuing her work with speech therapy and progressing appropriately.

Comment

- While mild degrees of calcification of bovine jugular vein implants have been reported, the extent of calcifications in this patient represent a previously undescribed presentation.
- The hybrid approach allowed for minimization of repeat dissection through amassed adhesions, decreasing the risk for intraoperative complications.
- This patient presented the additional technical challenge of a calcified ascending arch, prohibiting the standard approach for bypass and aortic cross-clamping.
- Taken together, aortic pathology in patients with congenital heart disease and prior arch intervention represent a complex surgical problem and require careful preoperative planning.
- A hybrid approach for optimization and minimization of surgical repair is recommended when feasible. This approach can mitigate the otherwise prohibitive surgical risk entailed with reoperation and repair of a calcified arch and increase procedural success.

Thank you