Surgical resection of severely calcified ascending arch in a pediatric congenital patient

Cathlyn K. Medina, BA, Berk Aykut, MD, Lauren E. Parker, BS, Lindsey Reynolds, BS, Stephen Miller, MD, Joseph Y. Cao, MD, Joseph W. Turek, MD, MBA,Douglas M. Overbey, MD, MPH, and Ziv Beckerman, MD.

From the Congenital Heart Surgery Research and Training Laboratory, School of Medicine, Division of Cardiothoracic Surgery, Department of Surgery, and Duke Children’s Pediatric and Congenital Heart Center, Duke University, Durham, NC.

This case report is approved under Pro00101549 (approved January 3, 2019) with waiver of individual patient consent. Received for publication Jan 11, 2024; revisions received March 7, 2024; accepted for publication March 17, 2024. Address for reprints: Cathlyn K. Medina, BA, School of Medicine, Duke University, 2301 Erwin Rd, DUMC 3474, Durham, NC 27710 (E-mail: cathlyn.medina@duke.edu).

CENTRAL MESSAGE
Reactive calcification causing near-total obstruction of the ascending aortic arch is an extremely rare presentation in a pediatric patient with congenital heart disease.

CASE DESCRIPTION
This case report is approved under Pro00101549 (approved January 3, 2019) with waiver of individual patient consent. Here, we present the case of a 15-year-old girl with hypoplastic left heart syndrome who had undergone 4 previous heart surgeries for single-ventricle palliation, most recently an intra-/extracardiac fenestrated Fontan procedure. The patient was 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. She subsequently underwent successful arch repair and Damus-Kaye Stansel (DKS) revision.

Mild calcification of the patient’s ascending neoaortic arch was first noted in October 2020 during diagnostic cardiac catheterization, 13 years after Fontan. At this time, the obstruction was not amenable to stenting given its proximity to the DKS, neoaortic valve, and arch branches. During routine catheterization in May 2023, elevated right ventricular end diastolic pressure, upper normal Fontan pressures and a 30 mm Hg gradient across the ascending aorta were noted, indicating need for surgical intervention. Computed tomography imaging demonstrated significant calcification of the ascending arch, measuring 4.4 × 1.7 × 1.9 cm (Figure 1, A).

PREOPERATIVE PLANNING
Given the findings from the catheterization, it was decided that the patient would have to undergo surgical arch repair and Fontan intervention. To aid in procedural decision making, a 3-dimensional model of the aorta was printed to assist with operative planning (Figure 1, B). With the procedure representing the patient’s fifth sternotomy, a hybrid approach was elected for staged management that consisted of stenting of the Fontan conduit in the catheterization lab followed by operative arch repair. If stenting was unsuccessful, the conduit would have been revised intraoperatively. Bypass cannulation strategy was carefully discussed given the unusual pattern of calcifications extending to the innominate artery as demonstrated by imaging.

CATHETERIZATION LAB
As the first step of the staged approach, balloon assessment was performed, showing a persistent waist at peak inflation across the Fontan conduit. A 5010 Palmaz (Cordis Corp)stent was subsequently positioned across the Fontan and deployed under fluoroscopy guidance. The stent was then serially dilated, and follow-up angiography demonstrated a well-positioned stent with improved Fontan baffle diameter.

CASE REPORT
OPERATIVE DETAILS
Following Fontan stenting, the patient underwent arch repair. Intraoperatively, right femoral vein cannulation was performed under fluoroscopy guidance to avoid Fontan stent dislodgement and the right axillary artery was exposed for bypass cannulation. A repeat median sternotomy was undertaken, and mediastinal dissection was performed with aortic no-touch technique to minimize risk of embolization. Following initiation of bypass, the patient was cooled to 25 °C, the ascending aorta was transected at the level of the DKS, and cardioplegia was administered down the DKS. The ascending aorta was near-totally obstructed, with only a minimal (3-4 mm) residual lumen (Figure 1, C). The base to proximal arch was resected en bloc (Figure 1, D), leaving an island of the arch vessels attached to the descending aorta. A 26-mm Gelweave (Terumo Aortic) graft was used to reconstruct the arch during a period of antegrade cerebral perfusion. The graft was next clamped proximally, arch vessels unsnared, and total body perfusion resumed through the graft. The DKS was reconstructed, and the proximal anastomosis between the DKS and aortic graft was completed. The patient separated from bypass without difficulty. Intraoperative echocardiogram demonstrated a patent arch and DKS with no neoaortic valve regurgitation. Pathologic evaluation of the resected arch revealed marked thickening and fibrosis of the aortic wall along with fibrous incorporation of the graft. The patient’s postoperative course was unremarkable aside for left vocal cord paresis. She was last seen in clinic on postoperative day 176 and continues to progress appropriately.

COMMENT
Although mild degrees of calcification of bovine jugular vein implants have been reported, the extent of calcifications in this patient represent an extremely rare presentation.1 On pathologic evaluation, the presence of a surrounding fibrotic aortic wall demonstrates that the calcification process was not limited to the foreign patch material. Cases of significant calcification of native vessels have only been described in adults with porcelain aortas or in pediatric patients with genetic conditions or aneurysms.2-4 As such, this case is among the first documented instances of severe calcifications of the aortic arch in a pediatric patient free of known genetic anomalies or vascular malformation.

Congenital heart disease with obstructive aortic lesions and prior arch interventions represents a complex surgical

![Figure 1](image_url)
problem that requires appropriate imaging along with careful preoperative planning to ensure safe surgical management and excellent outcomes.

**Conflict of Interest Statement**

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References


