Aortosubpulmonary Fistula after Konno-Rastan Aortoventriculoplasty

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INTRODUCTION

The Konno-Rastan procedure is an anterior aortoventriculoplasty facilitating enlargement of the left ventricular outflow tract (LVOT) to accommodate a larger aortic valve prosthesis in patients with subvalvular, valvular, and/or supravalvular stenosis. We describe a case of fistulous formation between the aorta and right ventricular outflow tract (RVOT) due to degeneration and disintegration of the repair patch. Furthermore, we present multimodality imaging and detail the multidisciplinary management of this complex patient.

CASE PRESENTATION

A 48-year-old female patient presented with progressively worsening heart failure symptoms 2 years after a Konno-Rastan procedure. The patient's medical history was notable for an aortic valvulotomy at 4 years of age for congenital bicuspid valve with associated aortic stenosis. Subsequently, she redeveloped severe aortic valve stenosis and aneurysmal dilation of the ascending aorta, requiring an aortic valve replacement (19 mm, St. Jude Medical Trifecta aortic bioprosthesis, St. Jude Medical, St. Paul, MN) and supracoronary ascending aorta and hemiarch replacement at 36 years of age. Ten years later, she developed severe prosthetic stenosis and underwent a Konno-Rastan procedure to enlarge the aortic valve annulus and permit surgical implantation of a larger bioprosthetic valve (23 mm, Inspiris Resilia, Edwards, Irvine, CA). Bovine pericardium CardioCel (Admedus, Toowong, QLD, Australia) patches were utilized to reconstruct the LVOT, RVOT, and ascending aorta.

The transthoracic echocardiogram (TTE) revealed a normally functioning bioprosthetic valve in the aortic position with a mean gradient of 11 mm Hg and no prosthetic regurgitation. Two communications were noted. One large communication (~2 cm) with continuous and pulsatile flow was seen between the ascending aorta and an anterior expanding mass (Figure 1, Video 1), suspicious for a

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VIDEO HIGHLIGHTS

Video 1: Preoperative TTE images demonstrating post-Konno-Rastan CardioCel patch disintegration resulting in an anterior mass, suspicious for a pseudoaneurysm. Two-dimensional and color Doppler compare TTE image obtained from a modified parasternal long-axis window (third intercostal space) demonstrating a large ~2 cm communication with continuous, pulsatile, and mainly laminar flow through the anterior wall of the repaired ascending aorta (location of the CardioCel patch) into an echo free space (anterior aortic pseudoaneurysm cavity). The *orange arrow* identifies the site of patch disintegration. The *green x* marks the aortic pseudoaneurysm. The color image to the right demonstrates flow from the aorta into the pseudoaneurysm.

Video 2: Preoperative TTE images demonstrating an anterior aortic pseudoaneurysm. Two-dimensional TTE image in a parasternal short-axis window demonstrating the pseudoaneurysm (*green x*) compressing the distal RVOT (*orange arrow*).

Video 3: Preoperative TTE images demonstrating the dominant path of the left-to-right shunt. Two-dimensional and color Doppler compare images were obtained from a parasternal short-axis window demonstrating sequential communications from the aorta into an expansile pseudoaneurysm (*green x*) cavity and then into the distal RVOT (*orange arrow*).

Video 4: Intraoperative TEE modified bicaval view demonstrating the coronary sinus (CS) catheter (*orange arrow*) entering the CS. *RA*, Right atrium.

Video 5: Intraoperative TEE ascending aortic arch short-axis view demonstrating the pulmonary endovent catheter (*orange arrow*) in the main pulmonary artery (MPA). Note the balloon is inflated as these catheters are placed in the same fashion as a pulmonary artery catheter. *AA*, Ascending aorta.

Video 6: Intraoperative TEE image demonstrating the ascending aortic pseudoaneurysm. Transesophageal echocardiographic multiplanar reconstruction of the pseudoaneurysm (*green x*) arising from the disintegration of the aortic patch (*orange arrow*) compressing the RVOT.

Video 7: Intraoperative TEE (color) image demonstrating the ascending aortic pseudoaneurysm. Transesophageal echocardiographic color multiplanar reconstruction of the aortic pseudoaneurysm (*white x*) compressing the RVOT. The *orange arrow* identifies the location of the disintegrated CardioCel patch.

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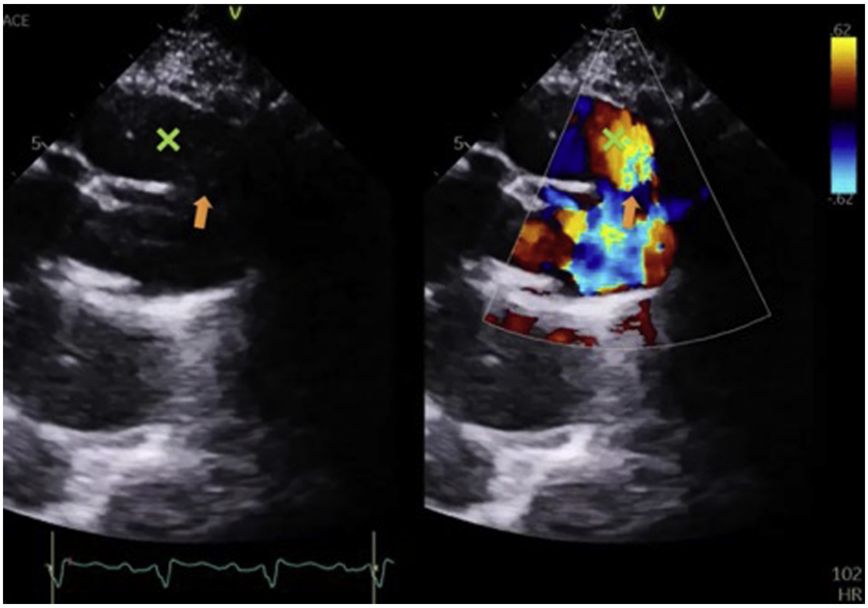

Figure 1 Preoperative TTE modified parasternal long-axis color compare image demonstrating post-Konno-Rastan CardioCel patch disintegration resulting in an anterior mass, suspicious for a pseudoaneurysm. The *orange arrow* identifies the site of patch disintegration. The *green x* marks the aortic pseudoaneurysm. The color image to the right demonstrates flow from the aorta into the pseudoaneurysm.

pseudoaneurysm. The pseudoaneurysm appeared to compress the RVOT (Figure 2, Video 2). A second communication with continuous flow was visualized between the pseudoaneurysm and the distal RVOT (Figure 3, Video 3). Contrast computed tomography (CT) confirmed a large (3.6 × 3.8 × 4.3 cm) anterior aortic pseudoaneurysm arising near the right coronary cusp with compression of the distal RVOT (Figure 4A). Consistent with the TTE findings, a fistulous communication was noted between the pseudoaneurysm and RVOT (Figure 4B). Right heart catheterization demonstrated elevated right-sided pressures (right ventricle, 70/30 mm Hg; pulmonary artery, 40/30 mm Hg) and a large left-to-right shunt (Qp:Qs, 2.1). The imaging and catheterization findings confirmed loss of integrity of the CardioCel patch. Furthermore, the CT showed the pseudoaneurysm, right ventricular free wall, and RVOT in immediate proximity to the mid and lower portions of the sternum, indicating inevitable injury to these structures during sternal reentry (Figure 5).

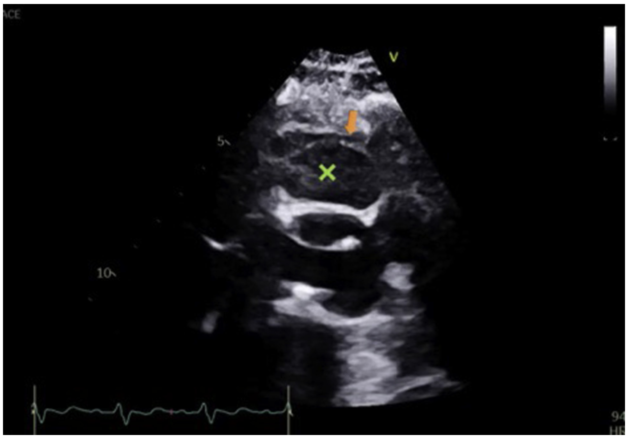

Figure 2 Preoperative TTE parasternal short-axis view demonstrating the anterior aortic pseudoaneurysm (*green x*) compressing the distal RVOT (*orange arrow*).

To avoid fatal hemorrhage on accidental injury to the adherent structures during sternotomy, peripheral cardiopulmonary bypass (CPB) and endoaortic occlussion with percutaneous retrograde cardioplegic arrest before completion of the sternotomy was planned. Following induction of anesthesia, a percutaneous retrograde cardioplegia catheter (ProPledge, Edwards Lifesciences) was introduced through the right internal jugular vein and positioned in the coronary sinus under transesophageal echocardiography (TEE) guidance (Figure 6, Video 4). The final position was confirmed by contrast fluoroscopy. Similarly, a pulmonary endovent catheter (EndoVent Catheter, Edwards Lifesciences) was placed in the main pulmonary artery under TEE guidance (Figure 7, Video 5).

After exposing the right axillary and both femoral arteries, a partial lower sternotomy was performed to define the planes and expose the right atrium and inferior vena cava. Cardiopulmonary bypass was established with right axillary and right femoral arterial inflow, and an inferior vena cava cannula was placed for venous return (later transitioned to bicaval cannulation after completion of sternotomy and dissection). A 32 mm endovascular occlusion catheter device (CODA balloon, Cook Incorporated, Bloomington, IN) was introduced via a 14F sheath in the left femoral artery. Using fluoroscopy, the CODA endoballoon was positioned in the ascending aorta graft proximal to the right brachiocephalic trunk. Cardiopulmonary bypass with systemic cooling was initiated. Upon ventricular fibrillation at 28°C, the endoballoon was inflated with diluted contrast and its position was confirmed on fluoroscopy (Figure 8). Following-occlusion of the ascending aorta, retrograde cardioplegia was delivered via the percutaneous cardioplegia catheter to arrest the heart. Sternotomy was completed after cardioplegic arrest thus preventing injury to the adherent structures. The endoballoon, well positioned in the

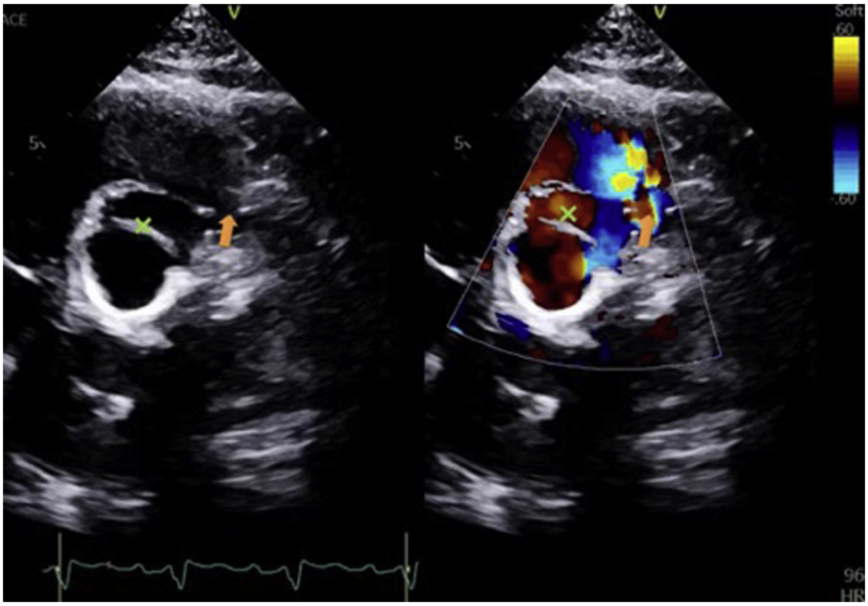

Figure 3 Preoperative TTE modified parasternal short-axis view demonstrating an anterior aortic pseudoaneurysm (*green x*) and a fistulous communication with the RVOT (*orange arrow*). Blood flows from the pseudoaneurysm into the RVOT, consistent with a left-to-right shunt.

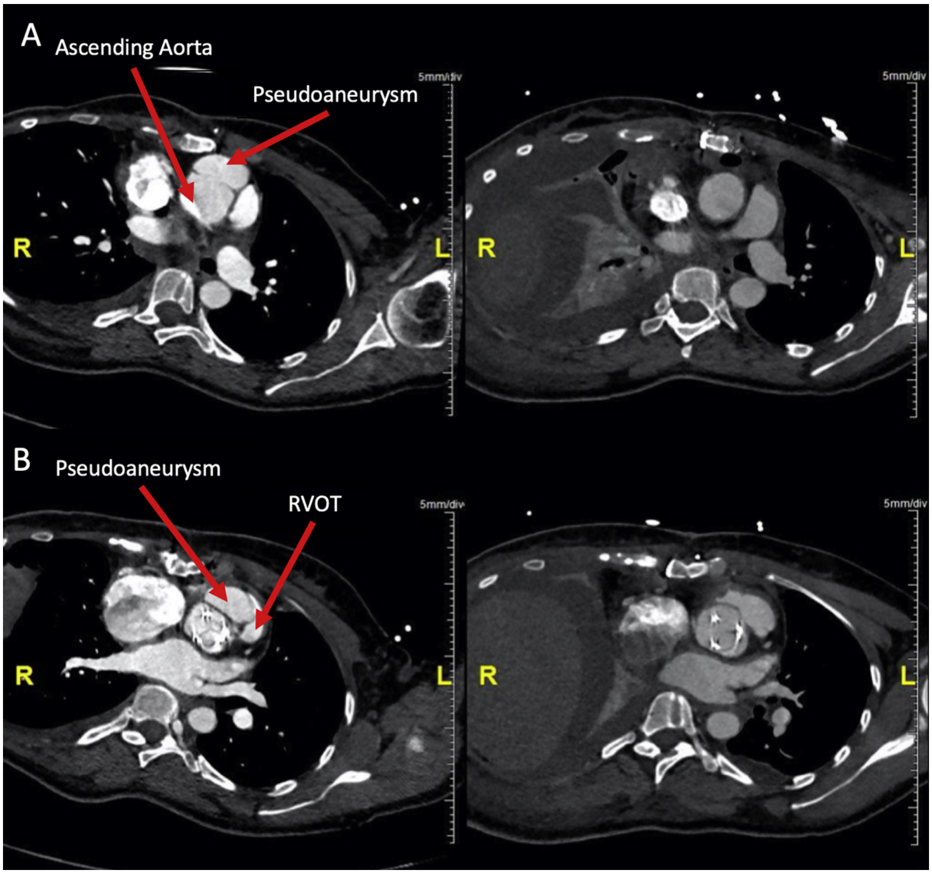

Figure 4 Computed tomography scan images before and after surgical correction. (**A**) Aortic pseudoaneurysm. Note the connection between the ascending aorta and the pseudoaneurysm. (**B**) Fistulous tract between the aortic pseudoaneurysm and RVOT.

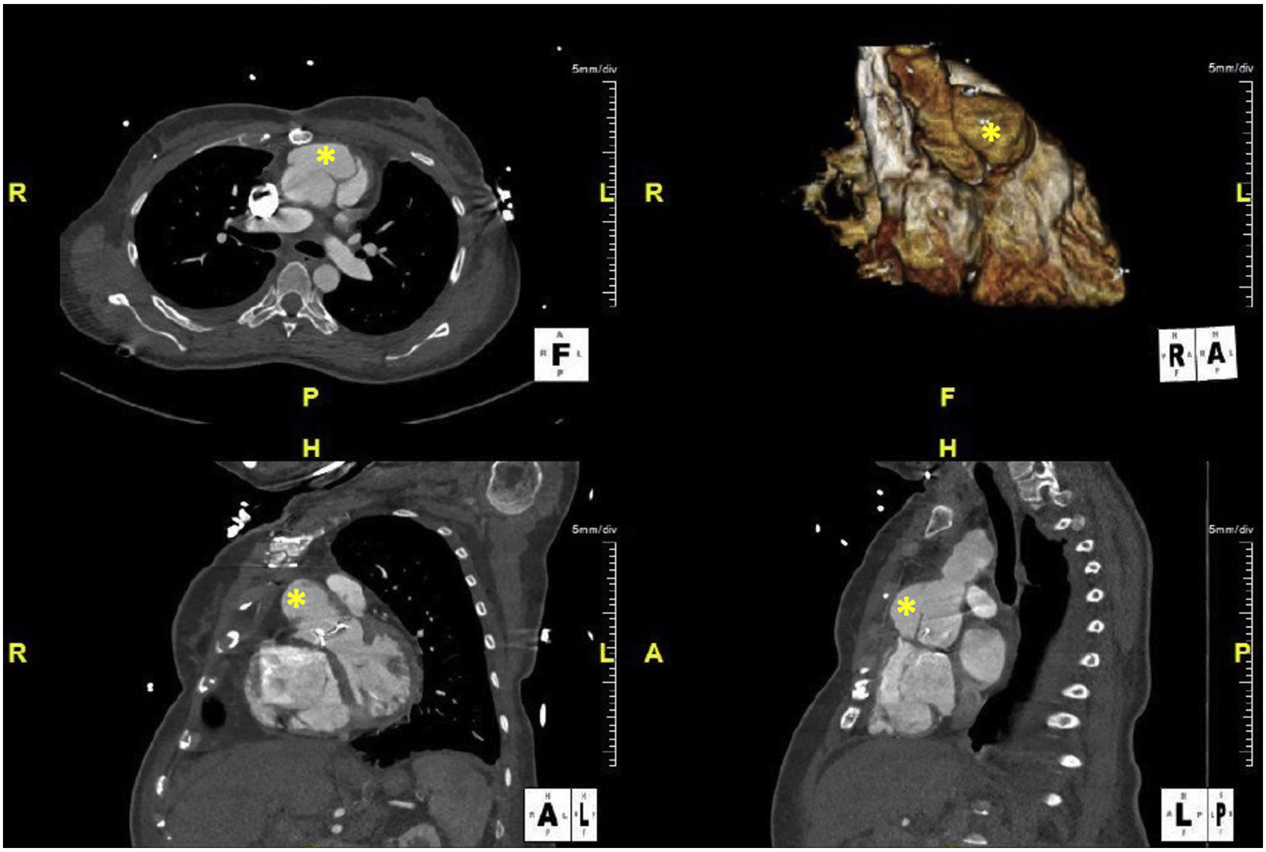

Figure 5 Multiplanar reconstruction CT demonstrating the pseudoaneurysm, right ventricle, and RVOT in close approximation to the mid and lower sternum. The pseudoaneurysm is marked with a *yellow asterisk*.

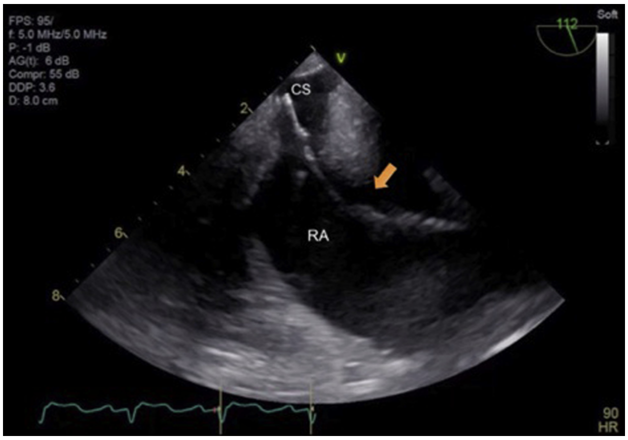

Figure 6 Intraoperative TEE modified bicaval view demonstrating the coronary sinus (CS) catheter (*orange arrow*) entering the CS. *RA*, Right atrium.

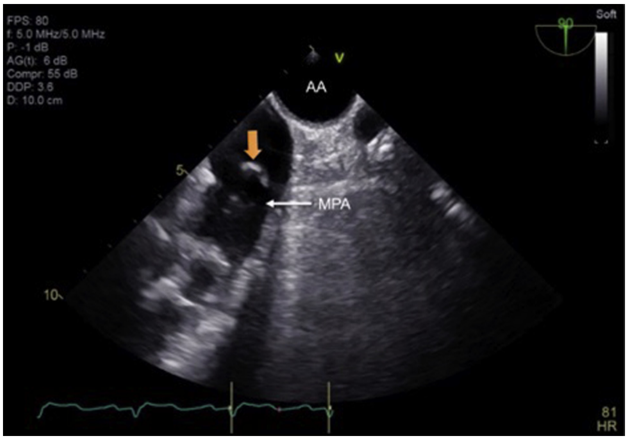

Figure 7 Intraoperative TEE ascending aortic arch short-axis view demonstrating the pulmonary endovent catheter (*orange arrow*) in the main pulmonary artery (MPA). Note the balloon is inflated as these catheters are placed in the same fashion as a pulmonary artery catheter. The endovent can be connected to the corresponding venting line from the CPB machine to aid in cardiac venting throughout the case. *AA*, Ascending aorta.

ascending graft, served as the aortic cross clamp throughout the operation.

The imaging findings were confirmed on intraoperative TEE (Figures 9 and 10, Videos 6 and 7) and surgical inspection (Figure 11). The CardioCel patch had lost its integrity above the prosthetic valve (Figure 11A; the patch covering the anterior ascending aorta has disintegrated) resulting in a pseudoaneurysm (Figure 12A). In the illustration, the pseudoaneurysm is demonstrated by the transparent bulge anterior to the ascending aorta and RVOT. This pseudoaneurysm developed a 7-8 mm large communication between the

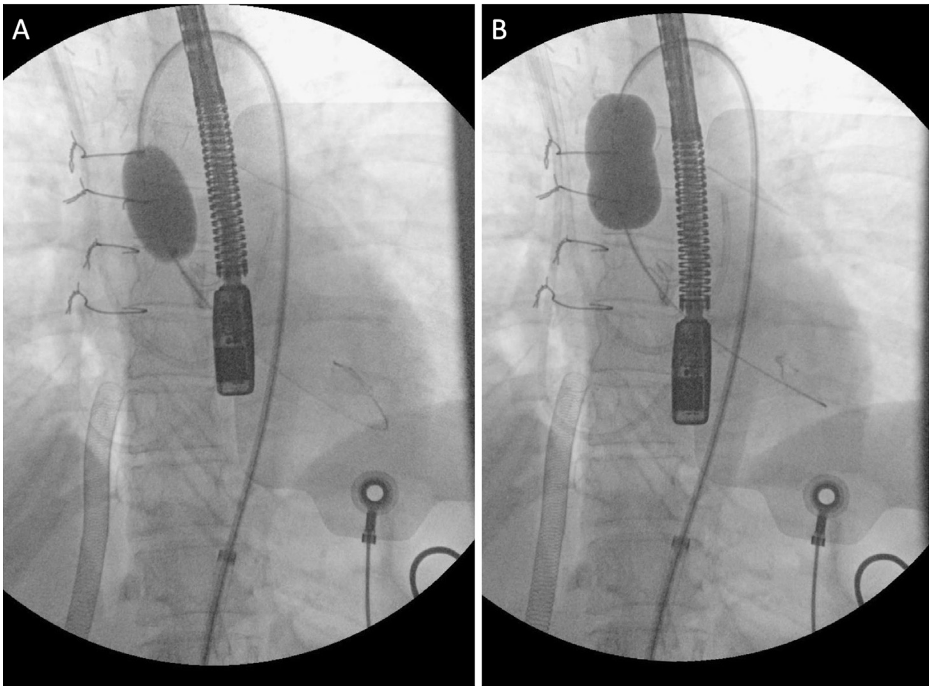

Figure 8 Contrast fluoroscopy with CODA endo balloon in the ascending aorta. **(A)** Partially inflated. The CODA balloon is inflated with contrast to allow localization of the balloon. **(B)** Fully inflated. Note that the inflated balloon on fluoroscopy demonstrates an indentation proximally as it extends partially into the pseudoaneurysm. This indentation signifies complete inflation of the balloon and occlusion of the ascending aorta.

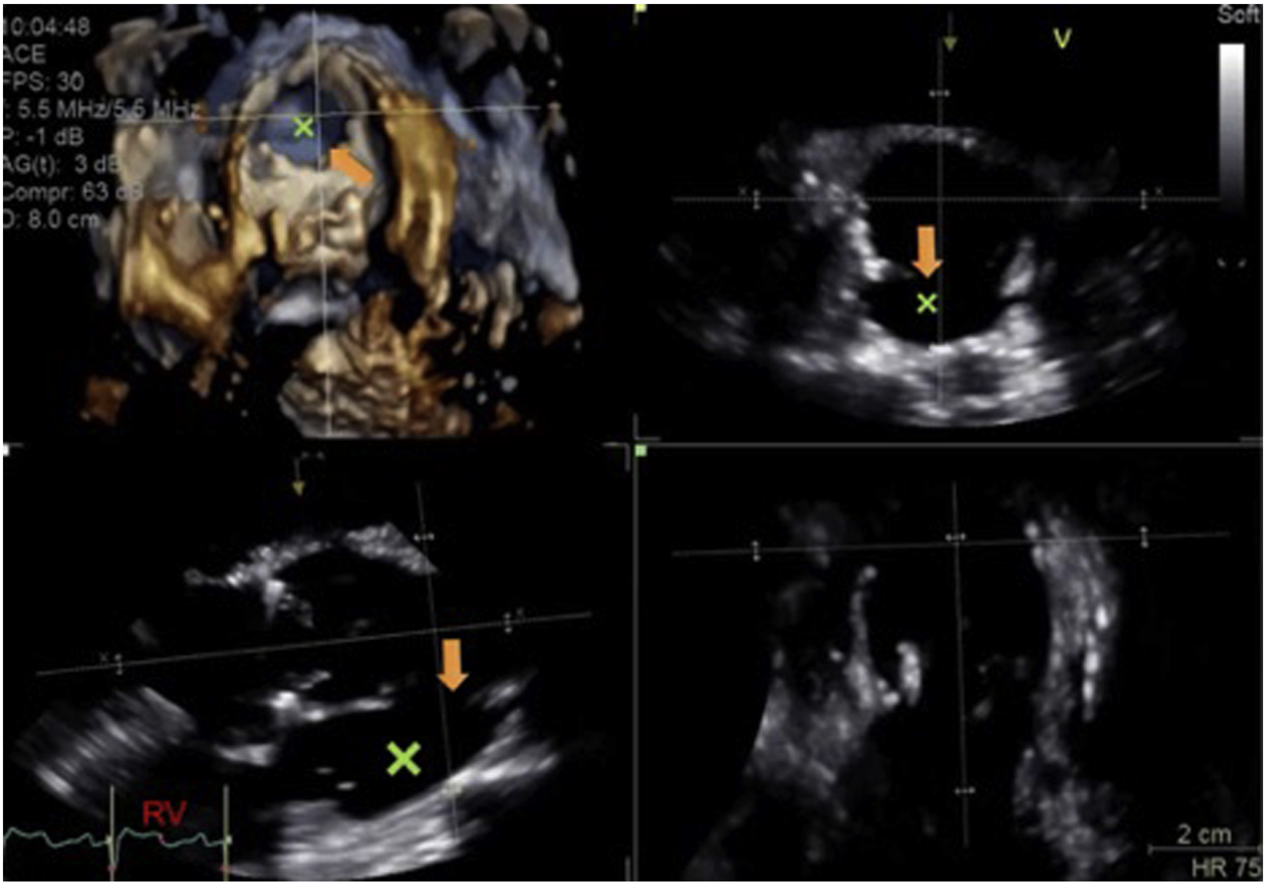

Figure 9 Intraoperative TEE imaging with three-dimensional multiplanar reconstruction demonstrating the ascending aortic pseudoaneurysm (*green x*) arising from the disintegration of the aortic patch (*orange arrow*).

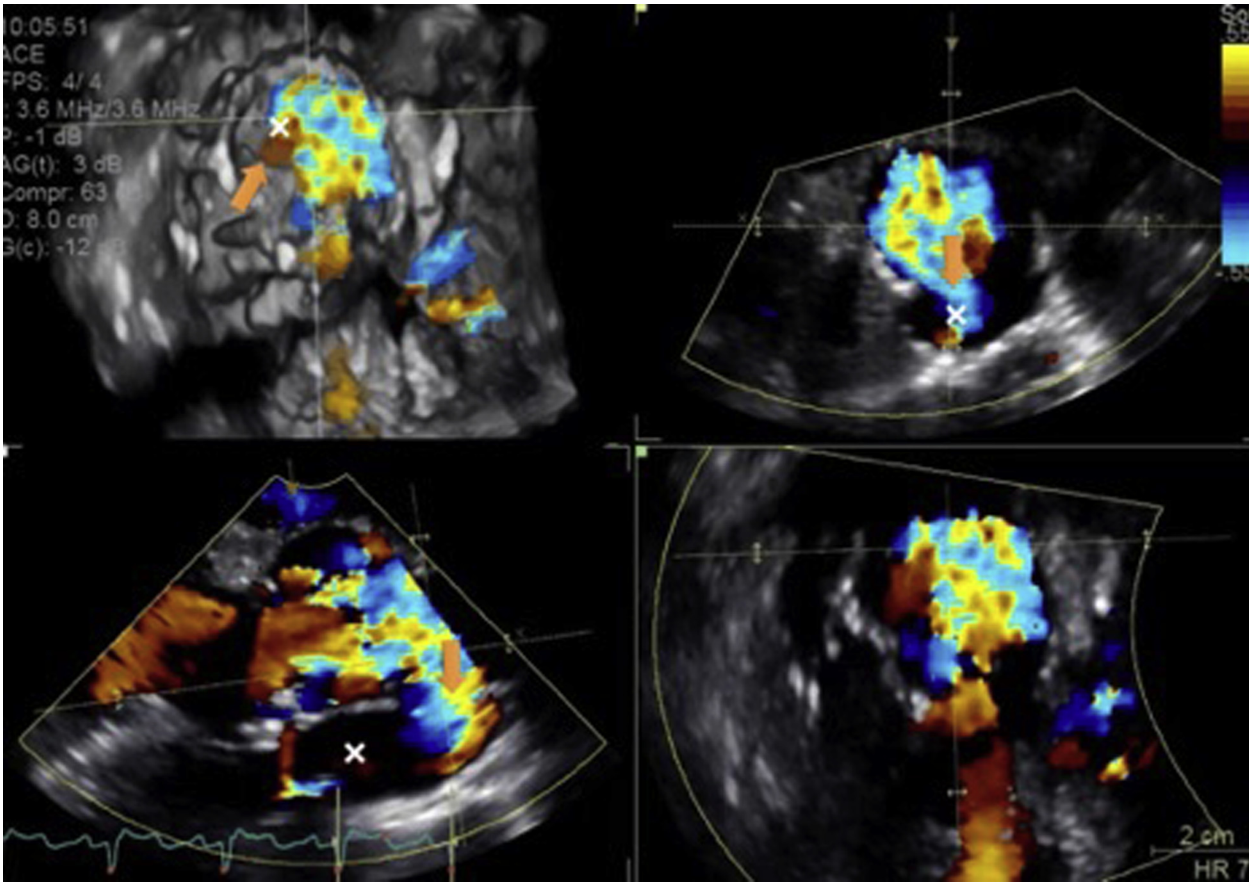

Figure 10 Intraoperative TEE color imaging with three-dimensional multiplanar reconstruction demonstrating the ascending aortic pseudoaneurysm (*white x*) arising from the disintegration of the aortic patch (*orange arrow*).

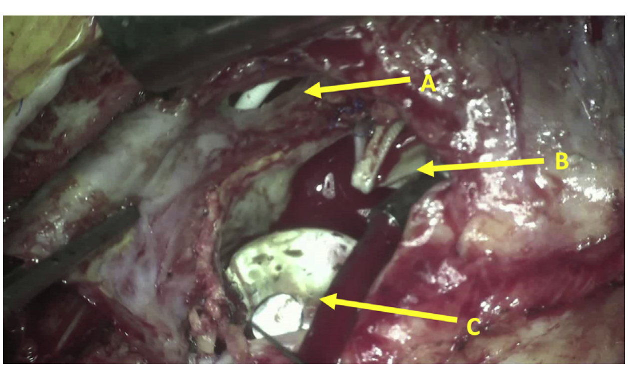

Figure 11 Surgical photograph after excision of the capsule of the aortic pseudoaneurysm and removal of the aortic patch. **(A)** Fistulous tract connecting the aortic pseudoaneurysm and the RVOT. **(B)** Bioprosthetic valve in the aortic position. **(C)** CODA endoballoon.

aorta and RVOT where the aortic and RVOT patches were compressed together. A small erosion was noted in the ventricular septal patch, resulting in a small ventricular septal defect close to the valve (Figure 12B) (This defect was not noticed on preoperative echocardiograms.) The prosthetic aortic valve appeared free of any pathology, with no signs of infection or dehiscence. After careful removal of the eroded patches, the ventricular septal, aortic, and RVOT defects were reconstructed using bovine pericardium (Edwards Lifesciences) without replacing the prosthetic aortic valve. The postoperative course was uneventful, and the patient was discharged home on postoperative day 7. Two months later, the patient presented with no heart failure symptoms. Transthoracic echocardiogram demonstrated preserved left ventricular systolic function with a normally functioning aortic valve prosthesis and no anastomotic leaks. Follow-up CT scan revealed a successful surgical repair (Figure 13).

DISCUSSION

The Konno procedure was first described by Soji Konno in a patient with congenital aortic stenosis and associated hypoplasia of the aortic valve.[1] A year later, Rastan and Koncz described a similar procedure.[2] The Konno-Rastan aortoventriculoplasty procedure entails a leftward longitudinal incision on the anterior ascending aorta, extended proximally into the right coronary sinus directed in between the right coronary artery and pulmonary valve. The RVOT is similarly incised anteriorly proximal to the pulmonic valve, and these two incisions are connected before further extending the incision across the aortic annulus into the upper portion of the interventricular septum. The location and size of the first septal artery are noted preoperatively to prevent damage to this artery. These incisions allow implantation of a larger prosthetic valve resulting in triangular defects in the ventricular septum, ascending aorta, and RVOT. These defects are reconstructed using bovine pericardium or synthetic patch material

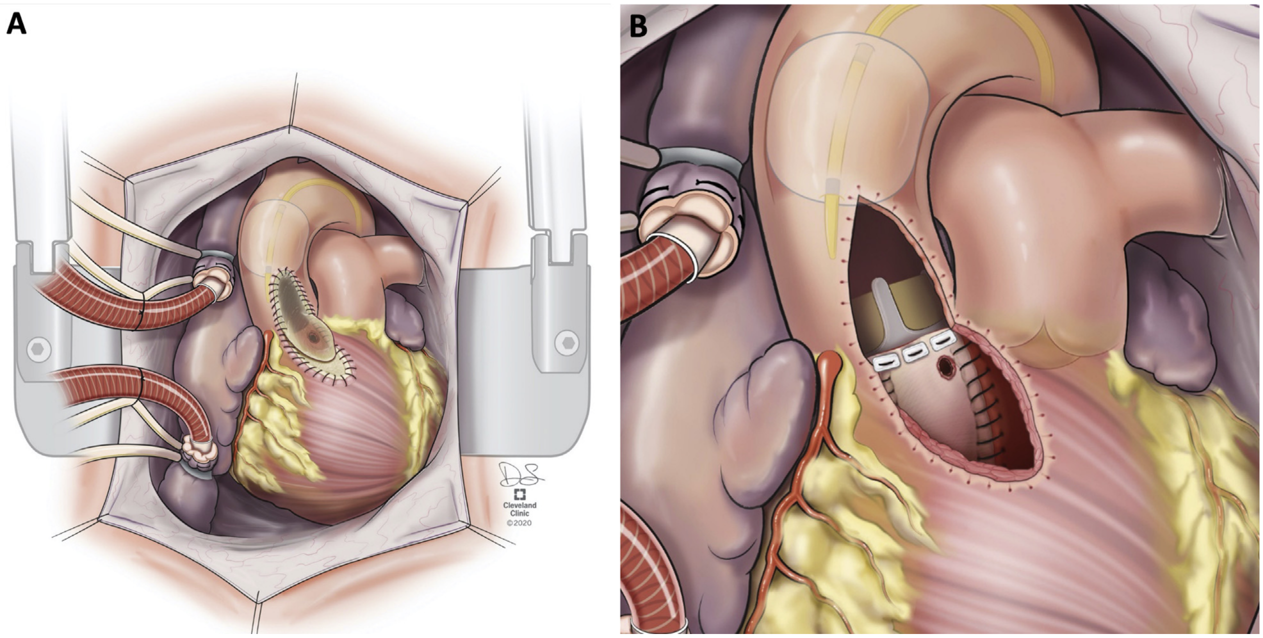

Figure 12 Schematic illustrations of the aortic pseudoaneurysm, fistulous tract between the pseudoaneurysm and the RVOT, and defect in the LVOT patch. The CODA balloon can be seen in the ascending aorta immediately distal to the pseudoaneurysm. The superior vena cava and inferior vena cava have been individually cannulated. **(A)** The anterior ascending aorta patch has disintegrated. Note the shadowed area inside the suture line above the aortic valve annulus. This disintegration formed a pseudoaneurysm, which is demonstrated as the transparent bulge anterior to the ascending aorta and RVOT. **(B)** With the pseudoaneurysm and external patch removed, two chambers of the heart can be seen. The superior aspect of the defect demonstrates the ascending aorta, while the inferior aspect of the defect demonstrates the RVOT. The suture line in the RVOT chamber is the ventricular septal patch, which separates the RVOT from the LVOT. A small erosion in the ventricular septal patch (LVOT) beneath the prosthetic aortic valve suture line is seen, resulting in a small ventricular septal defect. Illustrations courtesy of Dave Schumick.

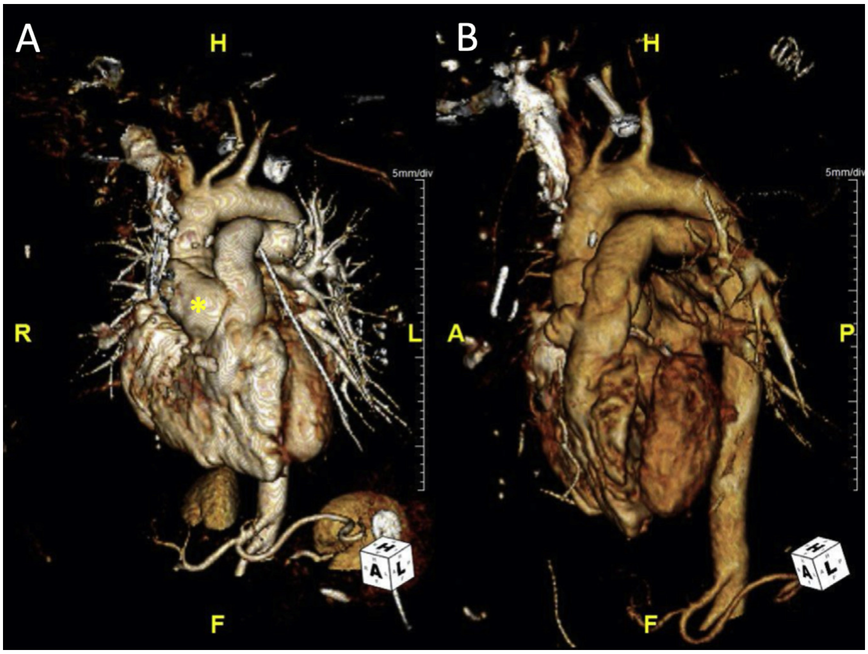

Figure 13 Three-dimensional CT scan rendering before **(A)** and after **(B)** surgery. Note the aortic pseudoaneurysm (marked with *yellow asterisk*). Following repair, the pseudoaneurysm has been surgically repaired.

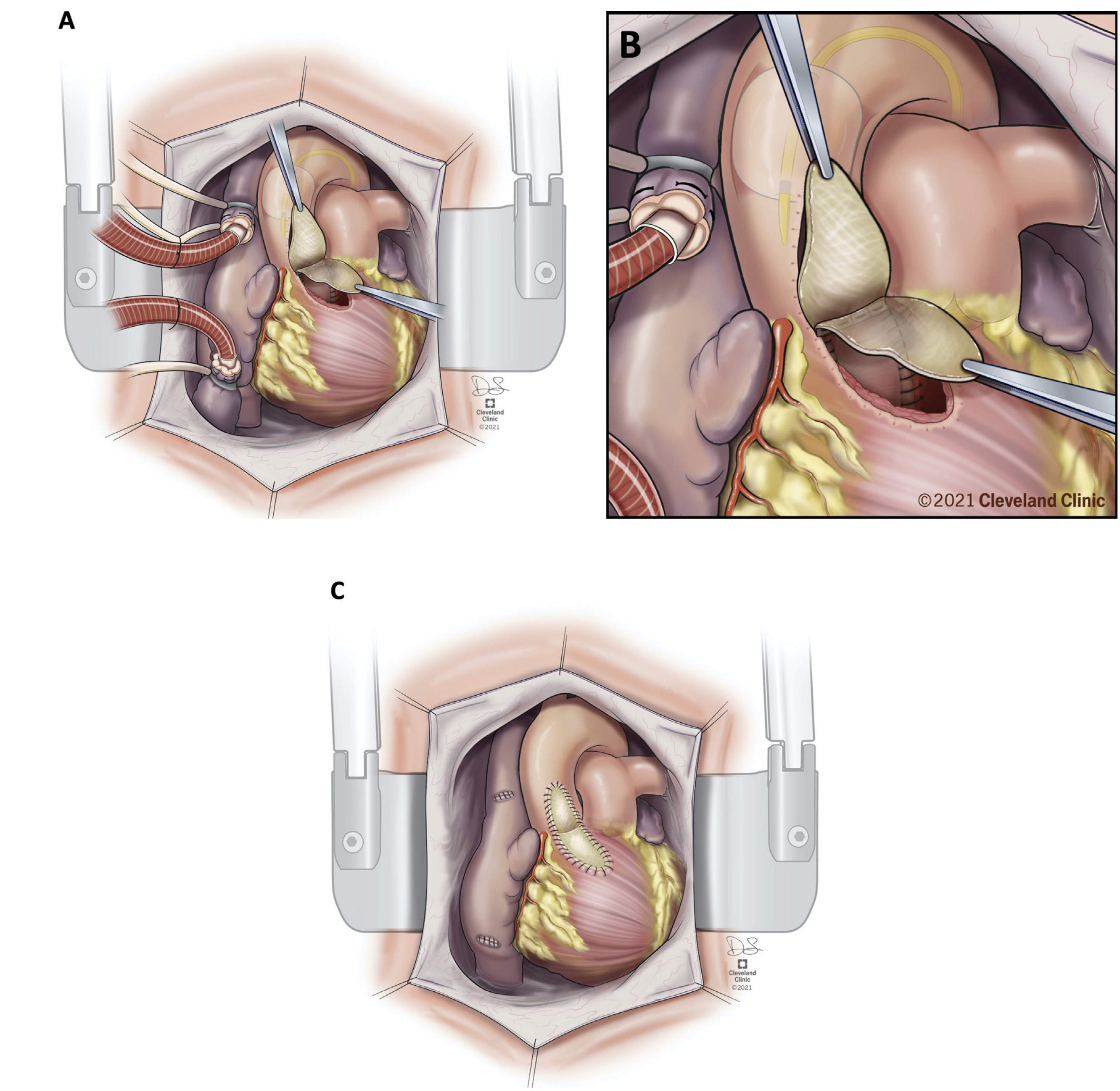

Figure 14 Illustration demonstrating the Konno-Rastan aortoventriculoplasty procedure. **(A)** There are three defects requiring reconstruction/closure: the anterior ascending aorta, the ventricular septum (LVOT), and the anterior RVOT. One patch is sutured to the ventricular septum (LVOT), prosthetic aortic valve sewing ring, and ascending aorta, while a separate patch is typically used to close the RVOT. Note the sutures interior to the RVOT patch demonstrating the ventricular septal patch portion. **(B)** Close-up image of the patches. **(C)** Completed procedure. Illustrations courtesy of Dave Schumick.

(Figure 14). A fusiform-fashioned single- or double-layer patch is sutured to the ventricular septum, the sewing ring of the prosthesis, and the ascending aorta to cover the corresponding defects, while the anterior portion of a double patch or a separate patch is used to refashion the RVOT.[3]

This case illustrates a serious early complication after Konno-Rastan procedure caused by degeneration and disintegration of the CardioCel patches used for the reconstruction. The case highlights the value of multimodality imaging to outline the pathology and a multidisciplinary team approach to an exceptionally high-risk reoperation.

Several publications testify to the safety and efficacy of the Konno-Rastan procedure.[4,5] Known complications associated with the procedure include prosthetic dysfunction, infective endocarditis, conduction blocks, congestive heart failure, and sudden death. Early degeneration of the CardioCel patch used to perform a Konno procedure has been noted only once previously, in a 12-year-old patient. The histology of the explanted patch demonstrated acute inflammation with destruction of collagen fibers and necrosis; however, no organism was identified.[6] CardioCel is prepared through a process in which bovine pericardium is decellularized and subjected to an anticalcification proprietary tissue engineering process (ADAPT TEP) to reduce the risk of calcification and degeneration. A low concentration of monomeric glutaraldehyde is used to promote strength and elasticity. Detoxification and storage are performed in glutaraldehyde-free solutions.[7,8] CardioCel has been used for the repair of congenital heart defects, aortic valve repair, and the Ozaki procedure. However, inflammation, fibrosis, and calcification persist, causing concern with its use, as documented in various reports.[9-11]

Dacron, bovine pericardium, and polyfluoroethylene are alternative patch materials used to reconstruct the surgical defects of the Konno-Rastan procedure. Synthetic patch materials are advantageous as they are resistant to degeneration, aneurysm formation, and rupture, but they are associated with more bleeding.[12]

The adherence of the pseudoaneurysm to the sternum in this fourth-time sternotomy presented a significant challenge to the team. A strategy previously described by us consisting of peripheral CPB with intra-aortic occlusion device (IntraClude, Edwards Lifesciences) and retrograde percutaneous cardioplegic arrest before completion of sternotomy was employed to prevent catastrophic bleeding and to minimize or avoid circulatory arrest time.[13] In this particular case, an endovascular occlusion balloon (CODA endoballoon) was inserted in place of the intra-aortic occlusion device (unavailable due to product recall) used previously. In comparison with the IntraClude device, the CODA endoballoon is unable to vent the left ventricle or deliver antegrade cardioplegia. Interestingly, both the devices have occlusion balloons with similar lengths (CODA 32 mm [3.7 cm] and IntraClude [3.75 cm]).[14,15] The preoperative CT scan was crucial in identifying a suitable landing zone for the CODA balloon in the ascending aorta graft proximal to the brachiocephalic artery. The estimated dimensions of the landing zone were of sufficient length and width to accommodate the inflated endoballoon device without occluding the coronary arteries proximally or the brachiocephalic artery distally. The inflated balloon on fluoroscopy demonstrated an indentation proximally as it extended partially into the pseudoaneurysm. This was an important clue to achieving complete occlusion of the ascending graft (Figure 10B). In contrast to previous cases, where the IntraClude catheter was withdrawn and replaced with a regular aortic cross clamp, the entire operation was carried out with the CODA balloon as a substitute for the aortic cross clamp.

A percutaneous retrograde cardioplegia catheter placed under TEE guidance was used to deliver cardioplegia. It is important to confirm the precise position and depth of the retrograde cardioplegia catheter within the coronary sinus. Demonstration of all the coronary sinus tributaries on contrast fluoroscopy after placement was crucial in confirming the adequate delivery of retrograde cardioplegia solution. The fistula between the aorta and pulmonary artery precluded delivery of antegrade cardioplegia and also necessitated a percutaneously placed pulmonary artery vent to decompress the pulmonary circulation on CPB.

CONCLUSION

Early CardioCel patch degeneration and disintegration after a Konno-Rastan procedure resulted in a complex pathology including a pseudoaneurysm, compression of the RVOT, an aorta to RVOT fistula, and a ventricular septal defect. Multimodality imaging, including echocardiography and CT, was crucial in diagnosing the pathology and planning the surgical repair. With careful preoperative planning, use of an endovascular aortic occlusion device, and peripheral CPB with percutaneous retrograde cardioplegic arrest, safe sternal reentry was possible to execute a successful repair without complications.

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SUPPLEMENTARY DATA

Supplementary data to this article can be found online at https://doi.org/10.1016/j.case.2021.07.009.

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