MINI-FOCUS ISSUE: CONGENITAL HEART DISEASE

INTERMEDIATE

CASE REPORT: CLINICAL CASE

Direct Percutaneous Transthoracic Cardiac Access for Recanalization of Longstanding Branch Pulmonary Artery Atresia

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ABSTRACT

We describe the use of direct percutaneous cardiac access to recanalize an atretic right pulmonary artery in an adolescent with complex congenital heart disease and right heart failure. This case highlights the problems associated with loss of central venous access and potential advantages of a direct cardiac approach to catheterization. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:180–6) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 17-year old girl presented with right heart failure (RHF) after surgical repair of pulmonary atresia with ventricular septal defect (PA-VSD) and discontinuous pulmonary arteries (PAs). As an infant, she underwent unifocalization of branch PAs, followed by PA plasty and right ventricle to pulmonary artery (RV-PA) conduit placement with VSD closure at age 3 years. At age 4 years, her right pulmonary artery (RPA) was noted to be atretic on a catheterization performed through the left internal jugular (IJ) vein, as both femoral veins and the right IJ were noted to be occluded. No attempt was made to recanalize the RPA during or following this procedure.

Over the next 13 years, the girl developed exercise intolerance. Cardiac magnetic resonance (CMR) was performed when she was 17 years of age, which demonstrated severe RV dysfunction (RV ejection fraction of 17%) and dilation (end-diastolic volume of 288 cm/m^2). A left IJ catheterization revealed suprasystemic RV pressure. The left PA had advanced PA hypertension (PAH). Attempts were made to recanalize a densely atretic RPA (**Figure 1**, Video 1), but the angles of approach made the intervention untenable. In an effort to relieve the suprasystemic RV pressures, the RV-PA conduit was stented to a diameter of 18 mm.

LEARNING OBJECTIVES

- To understand methods for DCA in order for complex cardiac interventions, particularly when central venous access is unavailable.
- To appreciate that return of parallel pulmonary circulation can significantly improve symptoms of right heart failure.
- To understand that longstanding atresia of proximal and distal pulmonary arteries can be successfully reversed, providing hemodynamic and clinical improvements.

Over the next 3 months, the patient developed overt signs of RHF with peripheral pitting edema, ascites, and massive hepatosplenomegaly. A computed tomographic angiography (CTA) revealed the course of the atretic RPA (Figures 1 and 2).

MEDICAL HISTORY

The patient was diagnosed as a newborn with pulmonary atresia and ventricular septal defect with discontinuous PAs; underwent placement of a right ventricle-to-pulmonary artery RV-PA conduit with VSD closure, right pulmonary atresia (RPA) atresia (2004) and pulmonary atresia hypertension (PAH) and left pulmonary atresia (LPA).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis for this patient was limited to causes of RV hypertension from left-sided PAH and RPA atresia.

Vital signs and physical examination. Weight was 65 kg; heart rate was 94 beats/min; blood pressure was 102/53 mm Hg; SpO$_2$ was 96%. The patient was an alert, pleasant young woman in no respiratory distress. She had a normal head and neck, and jugular venous distension. Her lung fields were clear. She had a prominent cardiac impulse at left sternal border with RV lift. On auscultation, normal S1, single S2 sounds. A harsh 3/6 systolic ejection murmur along the left sternal border radiating to the left lung. A 3/4 holodiastolic murmur along the left sternal border, a quiet right chest. The abdomen showed thin abdomen with firm hepatic margin palpated 6 cm below right costal margin; the spleen was palpable 4 cm below left costal margin. Moderate ascites with umbilical extroversion were present. Moderate lower extremity 3+ pitting edema with darkened discoloration on extensor surfaces was seen.

INVESTIGATIONS

CTA showed atretic RPA with calcified tract behind ascending aorta (Figure 1).

MANAGEMENT

A discussion with the cardiac surgeon made clear that surgical intervention or assistance with access would not be available. A consultation with the family took place to discuss transthoracic direct cardiac access (DCA) to attempt to recanalize the RPA. The patient and family consented to the procedure.

Surgery began by attaining left IJ and femoral artery access. A guidewire was placed in the left internal mammary artery (LIMA) to avoid puncture of the

ABBREVIATIONS AND ACRONYMS

CHD = congenital heart disease

CMR = cardiac magnetic resonance

CTA = computed tomographic angiography

CTO = chronic total occlusion

CV = central venous

DCA = direct cardiac access

IJ = internal jugular

LIMA = left internal mammary artery

PA = pulmonary artery

PAH = pulmonary arterial hypertension

PA-VSD = pulmonary atresia with ventricular septal defect

RHF = right heart failure

RPA = right pulmonary artery

RV = right ventricle

RVOT = right ventricular outflow tract

RV-PA = right ventricle to pulmonary artery

FIGURE 1 3-Dimensional Reconstruction of Computed Tomography Angiography
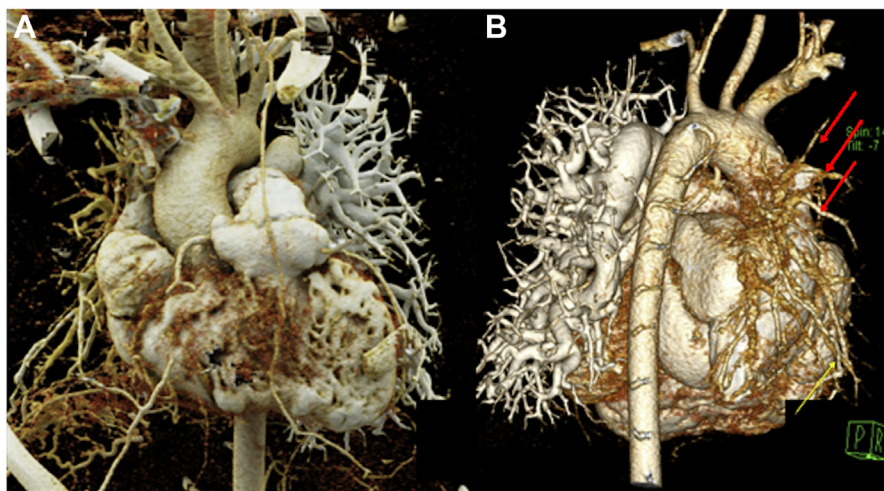

Computed tomography angiography demonstrates absent of right pulmonary arterial vasculature. The anteroposterior projection **(A)** demonstrates that the left internal mammary artery overlies the conduit. The posterior projection **(B)** demonstrates engorged left pulmonary artery branches, no right pulmonary arterial bed and hypoplastic right-sided pulmonary veins **(arrows)**.

FIGURE 2 Computed Tomography Angiography of Conduit and Atretic Right Pulmonary Artery
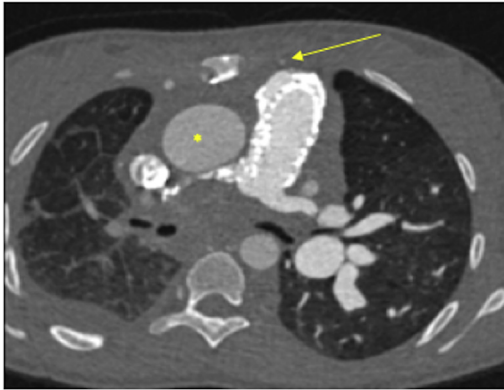
Computed tomography angiography image demonstrates an atretic right pulmonary artery which passes directly posterior to the ascending aorta **(*)**. The left internal mammary artery **(yellow arrow)** is directly anterior to the conduit.

LIMA with direct needle puncture of the right ventricular outflow tract (RVOT). Using the Seldinger technique, a short guidewire was passed from a 21-gauge access needle into the RVOT and then to the distal left PA (Videos 2 and 3). A 7-F short sheath was later upsized to an angled 8-F long sheath (Figure 3).

The RPA was carefully recanalized using chronic total occlusion (CTO) wires. Advancing a microcatheter over a CTO wire confirmed wire placement within the RPA, and contrast could be seen outlining the entire right lower lobe PA (Figure 4). The distal RPA pressure, after serial balloon angioplasty from 1.5- to 8-mm diameter was 22/10 mm Hg. The prehilar RPA was stented using premounted non-covered stents. Upon successful RPA recanalization and stenting, the long sheath was removed from the chest, leaving a guidewire in place. However, as pulsatile bleeding was noted at the access site, a sheath was then quickly placed back into the site, and an Amplatzer ductal occluder device (Abbott Medical, St. Paul, Minnesota) was placed in the access site with

FIGURE 3 Entry Into the Right Ventricle-to-Pulmonary Artery Conduit
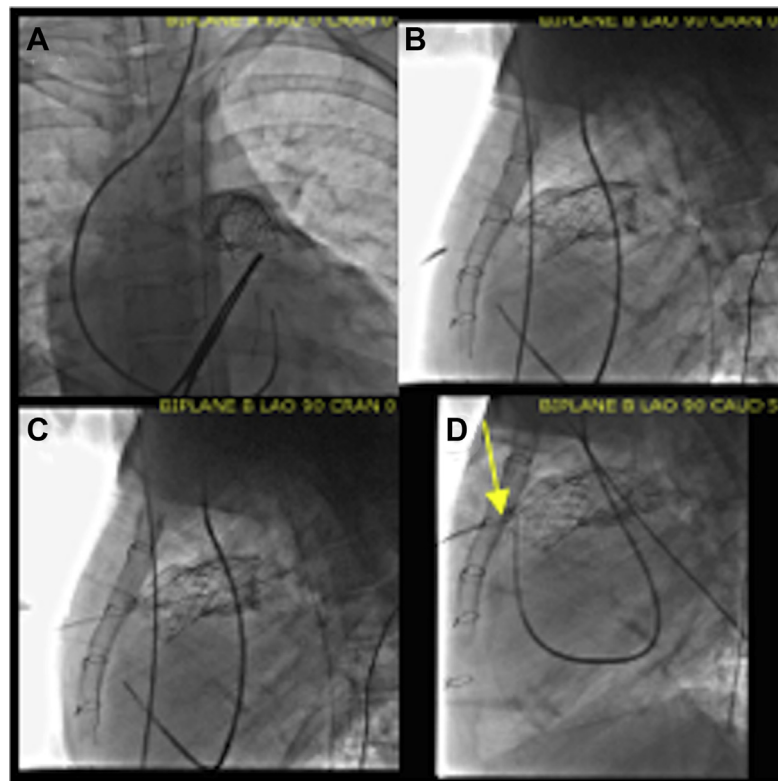
A hemostat helps identify the ideal focus of needle entry into the right ventricle-to-pulmonary artery (RV-PA) conduit **(A, B)**. Once the needle is passed into the RV-PA conduit, a guidewire is easily passed into the distal left pulmonary artery, allowing sheath placement **(C)**. Upon completion of the intervention, a ductal occlude device **(D)** is placed at the sheath entry into the RV-PA conduit to seal the tract **(yellow arrow)** to allow safe sheath removal.

FIGURE 4 Right Pulmonary Artery Recanalization and Stenting
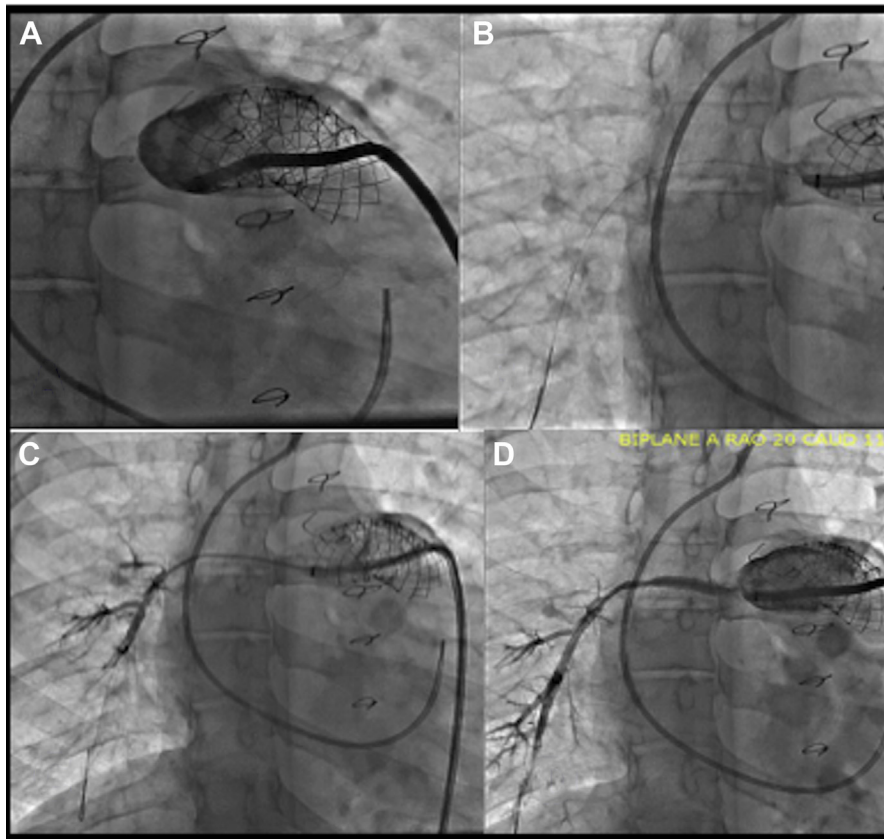
Angiograms demonstrate an atretic right pulmonary artery **(A)** which is recanalized using chronic total occlusion wires **(B)**. After recanalization, a hypoplastic but extensive pulmonary arterial vascular bed **(C)** is seen. After angioplasty with 4-, 5-, and 6-mm balloons, right pulmonary artery expands and exhibits antegrade flow **(D)**.

the proximal retention disk residing flush along the anterior surface of the RV outflow tract (**Figure 3D**, Video 4). The sheath and guidewire were removed; no bleeding was noted.

The patient was brought back for planned RPA stent redilation and further RPA rehabilitation 4 months later. At that time, her clinical symptoms of RHF had almost completely resolved. Her RV pressure was down to 75% systemic at repeat catheterization. The RPA stent was further dilated to 10-mm diameter (**Figure 5**). The right middle lobe PA branch was ultimately recanalized using the CTO wire technique and balloon angioplasty (**Figure 6**).

At the latest follow-up, the patient was clinically doing well and was attending college full time. Her RV function was moderately depressed, and her latest

hemodynamic catheterization reflected 60% systemic RV pressures. A lung perfusion scan following her second RPA intervention demonstrated 29% flow to the RPA (**Figure 7**). She has since undergone transcatheter pulmonary valve replacement to help reduce her RV volume, which eliminated her edema.

DISCUSSION

Previous publications have reported successful recanalization of central veins in this population (1,2). DCA to the left heart was initially described for diagnostic purposes in children and adults with aortic stenosis (3,4). Recently, direct left ventricular access has regained popularity to facilitate transcatheter aortic valve implantation (5). These procedures are

FIGURE 5 Subsequent Right Pulmonary Artery Rehabilitation
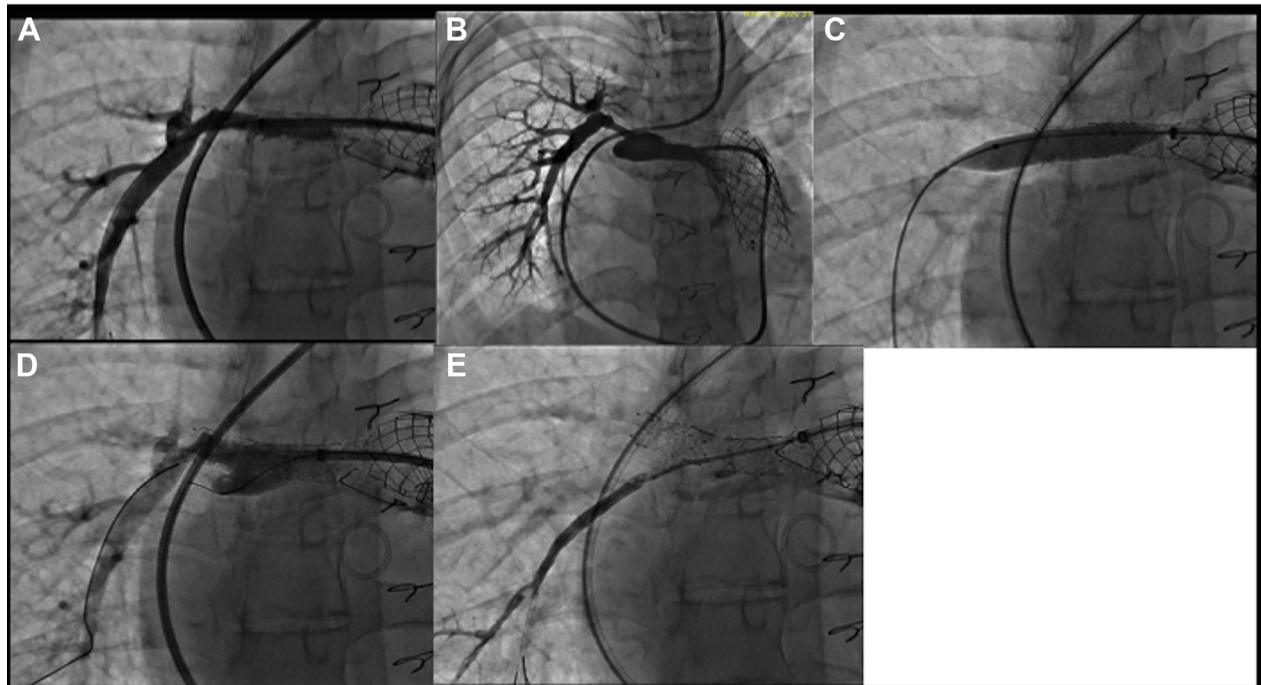
Angiography reveals a patent right pulmonary artery with preserved antegrade blood flow. Mild neointima within the stented region is noted **(A)**. Significant stenosis was noted **(B)** and ballooned **(C)**. An inferior pouch **(B)** represents origin of the right middle lobe pulmonary artery. This was carefully recanalized and ballooned **(D, E)**.

often facilitated by surgical access through the chest wall, allowing primary repair of the cardiac puncture site. However, in children with congenital heart disease (CHD), there are only rare reports of DCA, and those reports are limited again to left-sided diagnostic and interventional procedures (3,6,7).

Catheterization can be accomplished through alternate routes, including the transhepatic approach, although with risk of significant complications (8,9). Furthermore, alternate routes may not allow for optimal angles for certain interventions. In this patient, both the transhepatic approach and the left IJ approach failed to provide adequate counterpressure for CTO wires to the RPA. DCA was believed then to be an option of last resort. Surgical backup for DCA cases should be discussed prior to the procedure; in the present case, the surgical team believed the patient was not a candidate for advanced circulatory support, and therefore their support was minimal.

Interestingly, although use of DCA in the present case was deemed high risk, the ease with which access was gained was striking. Not only was sheath placement simple, but focusing a guide catheter into the area of interest thereafter was immediate. DCA enhanced our ability to successfully recanalize long-standing atresia. DCA for hemodynamic and interventional cardiac procedures has been previously described (3,4,6,7,10). In some cases, hemothorax has been described following sheath removal (6,7). Interestingly, in these other reports of DCA, the "exit strategy" following sheath removal from the chest is not described but obviously deserves consideration.

In the present case, the authors were cognizant of the fact that a range of strategies would need to be available once the sheath was removed at the end of the procedure. Once the bleeding around the guidewire was noted upon removing the sheath, a short sheath and a device to occlude the tract were quickly placed.

Providing an additional parallel vascular bed surface area is an important strategy which can at times be used to relieve RV hypertension (1,11). In this case, the presence of advanced vascular disease in the left lung made recanalization of the RPA even more

FIGURE 6 Final Angiograms and Transcatheter Valve Implantation
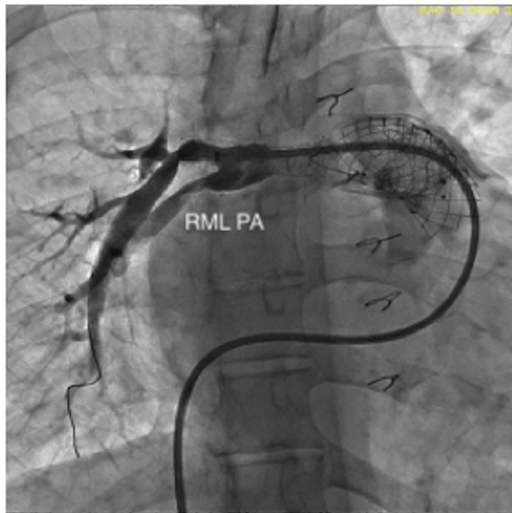
Placement of transcatheter pulmonary valve, and interval growth of the peripheral right pulmonary artery (PA) bed with no evidence of thrombosis or occlusion following recanalization. RML = right middle lobe.

FIGURE 7 Lung Perfusion Scan Following Right Pulmonary Artery Reintervention

A nuclear medicine lung perfusion scan demonstrates 29% flow to the right lung, with lack of flow to the right lung apex. LT= left; RT = right.

urgent. The hemodynamic consequences of recanalization include reduction in RV pressure and increase in cardiac output. In this case, distal RPA pressure was in fact low, and the resistance in that lung was much lower than the LPA, bringing the effective PVR much lower. This case also highlights the fact that recanalization and revascularization following many years of complete occlusion can be achieved and that such revascularization can result in meaningful redistribution of pulmonary blood flow. In this patient, at latest evaluation, there was 29% flow to the right lung. Hence, the interventionalist should be encouraged to consider recanalization of long-standing pulmonary artery atresia if RV hypertension or other indications are present.

FOLLOW-UP

At the last outpatient clinical encounter, this patient had improved dramatically. Her pulmonary valve has trace regurgitation, and her RV volume remains moderately dilated. Nevertheless, the patient attends college full time and reports sustained improvement after the series of interventions described above.

CONCLUSIONS

This case highlights the utility of a DCA approach for patients with diffuse CV occlusion and urgent right heart intervention. Care should be taken to note the local anatomy, and exit strategies should be carefully considered. Return of parallel pulmonary artery circulation can significantly reduce RV hypertension and can enhance cardiac output.

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KEY WORDS congenital heart disease, pulmonary artery, recanalization

APPENDIX For supplemental videos, please see the online version of this paper.