Large pseudoaneurysm following right ventricle to pulmonary artery homograft placement in an infant

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ABSTRACT
Pseudoaneurysm (PSA) is a known but rare complication of the right ventricle to pulmonary artery (RV-PA) conduits. The patient’s clinical presentation can be variable ranging from asymptomatic to potential rupture. We describe an unusual case of a massive PSA in an infant who underwent RV-PA pulmonary homograft placement after relief of right ventricular outflow tract obstruction.

Keywords: Homograft conduit, pseudoaneurysm, right ventricular outflow

INTRODUCTION
Right ventricle to pulmonary artery (RV-PA) conduits are commonly used for repair in various complex congenital heart diseases. There are several complications related to these conduits, the most common being conduit stenosis or regurgitation necessitating conduit replacement. A very rare complication is the formation of pseudoaneurysm (PSA), especially along the suture sites. This could be a potentially life-threatening condition requiring urgent surgery for replacement of the conduit. Our case is unique as we describe an infant <5 kg who presented with atypical symptoms and had an incidental discovery of a large PSA.

CASE REPORT
Patient “FT” is a 7-month-old girl with a history of d-transposition of great vessels with side-by-side vessels and a single coronary artery arising anteriorly and coarctation of the aorta. She initially underwent an arterial switch and coarctation repair with augmentation of the aortic arch in the newborn period. Three months later, she presented with severely elevated right ventricular pressures and was found to have hypoplasia of the neopulmonary valve with subvalvular obstruction. She underwent infundibular resection with the placement of a 9 mm pulmonary homograft conduit from RV to PA with right PA angioplasty. The native connection to the pulmonary arteries was left open.

At a routine follow-up visit four months later, the patient’s mother reported feeding intolerance with solids only, with frequent gag initiation. The patient was also noted to have mild tachypnea and fussiness when she was supine and constantly wanted to be held in upright position. On physical examination, her oxygen saturation was 98% on room air. There was a Grade II/VI to-and-fro murmur heard best at the left upper sternal border. The chest radiograph revealed widening of the anterior mediastinum in the region of the PA [Figure 1]. Transthoracic echocardiography showed normal biventricular function and a large fluid collection anterior to the RV [Figure 2]. A subsequent cardiac magnetic resonance imaging (MRI) demonstrated a large PSA in the anterior mediastinum related to the RV-PA conduit measuring 5.5 cm × 3.3 cm × 7.1 cm in transverse, anteroposterior,
and superoinferior dimensions, respectively [Figure 3]. The neck of the aneurysm measured 1.5 cm × 2.5 cm. The ascending aorta was displaced posteriorly secondary to its mass effect. She underwent open heart surgery the next day. Cardiopulmonary bypass was achieved through cervical incision. The right common carotid artery and internal jugular vein were heparinized and cannulated. Once the patient was cooled to 22 degrees, sternotomy was done. During her previous surgery, a Gore-Tex membrane was placed which basically served as the anterior portion of the capsule of the PSA. This membrane was removed which revealed a massive PSA extending from the base of diaphragm to the innominate vein and included the entire homograft. The native connection from the RV to the pulmonary arteries was still patent. There was stenosis of the origin of the right and left pulmonary arteries. The entire capsule of PSA was excised and replaced with a 15 mm pulmonary homograft. The patient did well thereafter and was discharged on postoperative day 8 with a normal echocardiogram on the day of discharge.

DISCUSSION

The large PSA that we report is a rare complication of right ventricular outflow tract (RVOT) conduits. Its pathogenesis is not clear. In our case, the PSA might have occurred as a result of elevated right sided pressures secondary to the branch PA stenosis. Elevated right ventricular pressure can lead to mechanical strain in the suture line and cause PSA. A small dehiscence in the reconstructed RVOT leads to leakage of blood in the surrounding tissue causing a hematoma into the potential space lined by pericardium. Other contributing factors such as cardiac muscle degradation, suture material, and technique have also been implicated.[1]

These PSAs can be asymptomatic or can present with symptoms related to compression of surrounding mediastinal structures, thrombosis, or infection.[2] The tachypnea was attributed to the massive PSA compressing the trachea, especially in supine position. The gagging sensation noted with solid foods in our patient was attributed to compression of the esophagus. Rarely, PSAs can present with syncopal attacks.[3] Progressive dilation and rupture of PSA are a recognized outcome making it imperative for prompt diagnosis and treatment.[4] Our patient developed a massive PSA involving the entire homograft conduit. In the setting of intolerance to solids, tachypnea, and significant echocardiogram findings, a PSA should be considered. The chest x-ray often shows widening of the right mediastinum. It is important to perform advanced imaging, such as an MRI, to clearly define the size and anatomy and help plan surgical approach.[5]

The options for repair of a PSA include endovascular device placement versus surgical replacement. While there are case reports of endovascular closure followed by percutaneous pulmonary valve implantation, this strategy was mostly utilized in adult patients with little distortion of the existing conduit.[6,7] Cardiac catheterization followed with device occlusion with Amplatzer device has been used as a successful option in an infant as young as 5 months of age.[8] In our patient, the large stalk combined with massive aneurysm size and distortion of previously placed conduit necessitated surgical approach. Care must be taken during sternotomy to avoid entering the large aneurysm inadvertently.[9] Preoperative advanced imaging with MRI or computed tomography scan to fully delineate the location of the PSA helps plan for the route of bypass for repair.

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Conflicts of interest
There are no conflicts of interest.
REFERENCES

1. Pillai SK, Reddy HP, Kulkarni S, Murthy KS, Cherian KM. Pseudoaneurysm of homograft placed in right ventricular outflow tract. Ann Thorac Surg 2004;78:1068-70.

2. Lee Y, Lee JT, Cho JY, Kim GJ. Pseudoaneurysm of surgically reconstructed right ventricular outflow tract complicated by superior vena cava syndrome. Korean J Thorac Cardiovasc Surg 2014;47:541-4.

3. Calabro R, Santoro G, Piscacane C, Pacileo G, Russo MG, Vosa C, et al. Repeat syncopal attacks due to postsurgical right ventricular pseudoaneurysm. Ann Thorac Surg 1999;68:252-4.

4. Vlodaver Z, Coe JI, Edwards JE. True and false left ventricular aneurysms. Propensity for the latter to rupture. Circulation 1975;51:567-72.

5. Shashidharan S, Wells WJ, Dettterich J, Wong J, Kumar SR. Giant pseudoaneurysm of reconstructed right ventricular outflow tract. Ann Thorac Surg 2015;100:734.

6. Lurz P, Taylor A, Bonhoeffer P. Percutaneous treatment of a giant right ventricular outflow tract pseudo-aneurysm and severe pulmonary regurgitation. Eur Heart J 2007;28:2086.

7. Dryžek P, Góreczny S, Moszura T, Stefańczyk L, Szymczyk K, Sysa A, et al. Right ventricular outflow tract giant pseudoaneurysm: Percutaneous approach and complications. Kardiol Pol 2013;71:1076-8.

8. Herbert C, Ikemba C, Nugent A. Device closure of a pseudoaneurysm of the right ventricular outflow tract in an infant with right ventricle-to-pulmonary artery homograft. Catheter Cardiovasc Interv 2014;83:587-90.

9. Kirshbom PM, Myung RJ, Simsic JM, Kramer ZB, Leong T, Kogon BE, et al. One thousand repeat sternotomies for congenital cardiac surgery: Risk factors for reentry injury. Ann Thorac Surg 2009;88:158-61.