Cardiology in the Young

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Brief Report

Cite this article: Tomasulo CE, Ravishankar C, Natarajan S, Mascio CE, and Glatz AC (2021) Large aneurysms and pseudoaneurysms of surgically reconstructed right ventricular outflow tracts. *Cardiology in the Young* **31**: 1522–1524. doi: 10.1017/S1047951121001025

Received: 23 January 2021
Accepted: 20 February 2021
First published online: 17 March 2021

Keywords:
CHD; ventriculotomy; branch pulmonary artery stenosis; ventricular septal defect; tetralogy of Fallot

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Large aneurysms and pseudoaneurysms of surgically reconstructed right ventricular outflow tracts

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Abstract

Aneurysm and pseudoaneurysm development is a known, albeit uncommon, complication after right ventricular outflow tract surgical reconstruction. Large right ventricular outflow tract aneurysms and pseudoaneurysms have not been extensively described in recent literature and we report our experience with this unusual complication in five patients at our institution over the last 8 years. Although uncommon, this complication has potentially important clinical implications. Thus, clinicians should be aware of its potential, particularly in certain anatomic conditions.

The development of a large aneurysm or pseudoaneurysm can occur after right ventricular outflow tract surgical reconstruction. Multiple causes have been identified, including elevated right ventricular pressure,[1–3] residual ventricular septal defects,[4] right ventricular outflow tract or branch pulmonary artery obstruction,[1,2,4,5] and use of ventriculotomy and pericardial patch.[1,2,4] Progressive aneurysm dilation risks rupture, compression of adjacent structures, and thrombosis.[3,5,6] After the aneurysm is identified, patients often undergo surgical resection and repair, especially if additional surgical interventions are required. Large right ventricular outflow tract aneurysms and pseudoaneurysms have not been extensively described in recent literature, and we performed a retrospective chart review of five patients at our institution who presented with this rare complication over the last 8 years. This review focused on cardiac imaging findings and clinical course. Our institutional review board deemed this retrospective case series exempt from the need for ethics board review and approval.

Case series

Patient 1

A patient with a history of tetralogy of Fallot with pulmonary atresia and confluent pulmonary arteries initially underwent Blalock–Taussig shunt placement, followed by complete repair at 6 months of age with a transannular homograft patch and Dacron patch ventricular septal defect closure. She was suspected to have a large right ventricular outflow tract aneurysm by echocardiogram 5 months later and was noted to have poor weight gain, tachypnea, and desaturations with crying. Cardiac catheterisation performed 6 weeks later confirmed a right ventricular outflow tract aneurysm proximal to the pulmonary artery origin measuring 6.4 × 5.5 cm (Fig 1). She had mild right pulmonary artery stenosis and left pulmonary artery hypoplasia, greater than 2/3 systemic right ventricular pressure, with no residual shunts. She underwent surgical repair the same day with aneurysm resection, placement of a 22-mm pulmonary homograft right ventricle to pulmonary artery conduit, right pulmonary artery dilation, and left pulmonary artery plasty. The pathology was consistent with true aneurysm. She required catheterisation 7 days later for right pulmonary artery and left pulmonary artery stent placement and subsequent right pulmonary artery and left pulmonary artery balloon angioplasty another 8 months later. There was no aneurysm recurrence at 51 months' follow-up.

Patient 2

At 3 months of age, a patient with a history of VACTERL and tetralogy of Fallot with the left anterior descending artery from the right coronary artery underwent repair with right ventricular outflow tract muscle resection, Dacron patch ventricular septal defect closure, and a limited infundibular patch due to her coronary anatomy. Three months later, she was diagnosed with a right ventricular outflow tract aneurysm on echocardiogram. Cardiac catheterisation

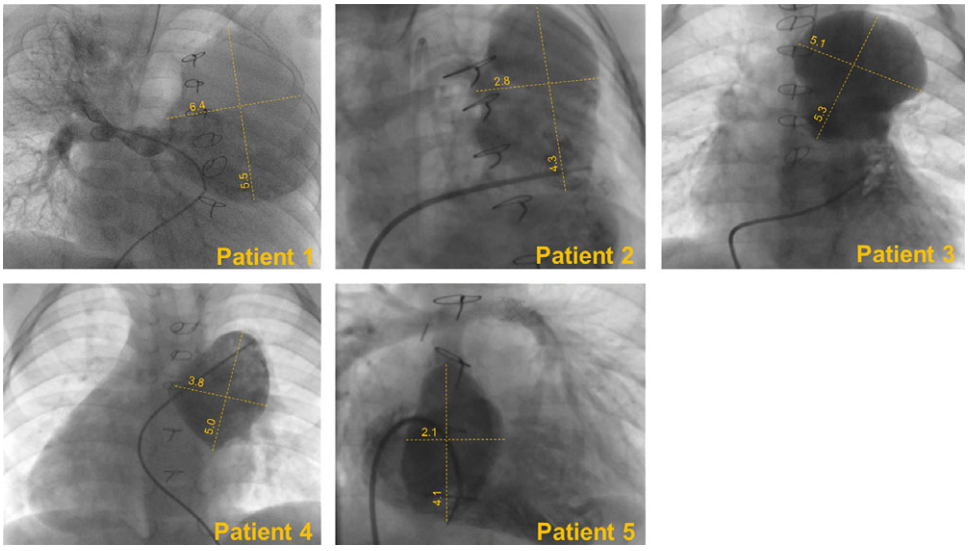

Figure 1. Angiographic still frames depicting the aneurysm or pseudoaneurysm in each patient, with measurements (in centimetres) noted.

demonstrated a large right ventricular outflow tract aneurysm at the right ventricle free wall measuring 4.3 × 2.8 cm (Fig 1). She had a small residual ventricular septal defect at the superior aspect of the patch, right ventricular outflow tract narrowing, and hypoplasia of the pulmonary valve, main pulmonary artery, and branch pulmonary arteries. Her right ventricular pressure was suprasystemic. She underwent surgical repair with aneurysm resection and placement of a 12-mm aortic homograft right ventricle to pulmonary artery conduit 7 days later. The pathology was consistent with true aneurysm. She required catheterisation the next day for right pulmonary artery stent placement. She underwent left pulmonary artery stent placement 3 months later and right pulmonary artery balloon angioplasty 3, 9, and 36 months later. There was no aneurysm recurrence at 61 months' follow-up.

Patient 3

At 2 months of age, a patient with a history of tetralogy of Fallot with severe pulmonary stenosis and major aortopulmonary collateral arteries underwent right ventricular outflow tract stent placement, with a second stent placed 1 month later and transannular patch repair without ventricular septal defect closure at 7 months of age. During cardiac catheterisation 12 months later, he was diagnosed with a large right ventricular outflow tract aneurysm at the main pulmonary artery measuring 5.3 × 5.1 cm (Fig 1). He had very small proximal branch pulmonary arteries, systemic right ventricular pressure, and net right-to-left shunt at the ventricular septal defect of 0.7:1. He then underwent surgical repair 3 days later with aneurysm resection, placement of a 16-mm aortic homograft right ventricle to pulmonary artery conduit and bilateral pulmonary arterioplasty. The ventricular septal defect was again left open. The pathology was consistent with true aneurysm. He required catheterisation 7 months later for right pulmonary artery and left pulmonary artery balloon angioplasty. There was no aneurysm recurrence at 31 months' follow-up, at which time he underwent Dacron patch ventricular septal defect closure, right ventricle to pulmonary artery conduit replacement, and left pulmonary arterioplasty.

Patient 4

A patient with a history of heterotaxy, dextrocardia, pulmonary atresia with right ventricle to aorta, malalignment ventricular septal defect, mildly hypoplastic left ventricle and bilateral superior vena cavae, segments {I,L,L}, was initially palliated with a Blalock–Taussig shunt, followed by a complete repair at 4 months of age with a Rastelli procedure with a 13-mm aortic homograft right ventricle to pulmonary artery conduit. He was diagnosed with a large right ventricular outflow tract aneurysm by echocardiogram 40 months later. Cardiac catheterisation demonstrated a 5.0 × 3.8 cm aneurysm arising below the conduit valve and laying to the left of the conduit (Fig 1). He had proximal right pulmonary artery stenosis, long-segment left pulmonary artery stenosis, systemic right ventricular pressure, and no residual shunts. He underwent surgical repair 37 days later with aneurysm resection, placement of a 17-mm aortic homograft right ventricle to pulmonary artery conduit, bilateral pulmonary arterioplasty, and suture ventricular septal defect closure. The pathology was consistent with pseudoaneurysm. He required catheterisation 54 days later for right pulmonary artery stent placement and left pulmonary artery balloon angioplasty. There was no aneurysm recurrence at 23 months' follow-up.

Patient 5

In the neonatal period, a patient with a history of transposition of the great arteries, ventricular septal defect, and aortic arch hypoplasia underwent an arterial switch operation with Lecompte manoeuvre, ventricular septal defect closure with Dacron patch, and augmentation of her hypoplastic aortic arch, followed by pulmonary arterioplasty and left pulmonary artery stent placement. She then underwent Dacron patch closure of a residual ventricular septal defect at 7 weeks of age. Two months later, cardiac catheterisation demonstrated a 4.1 × 2.1 cm right ventricular outflow tract aneurysm at the site of the right ventriculotomy patch (Fig 1). She had proximal right pulmonary artery stenosis, in-stent stenosis of the left pulmonary artery, and supravalvar narrowing of the main pulmonary artery. Her right ventricular pressure was greater than 2/3 systemic, with no residual shunts. The aneurysm remained stable in size on serial catheterisations and was closed with a homograft patch 7 years later, along with bilateral pulmonary arterioplasty. The pathology was consistent with pseudoaneurysm. She required catheterisation 3 months later for right pulmonary artery and left pulmonary artery stent placement. There was no aneurysm recurrence at 27 months' follow-up.

https://doi.org/10.1017/S1047951121001025 Published online by Cambridge University Press

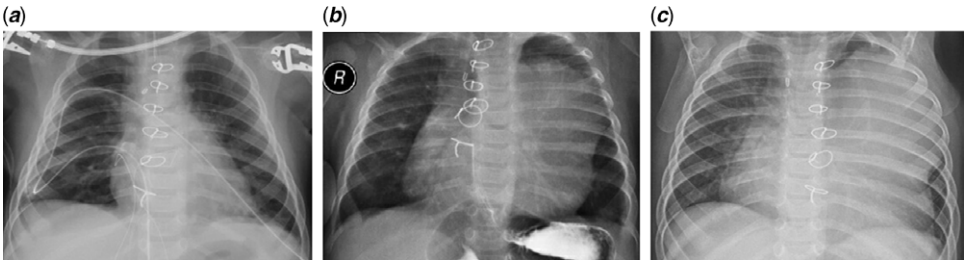

Figure 2. Serial chest radiographs from Patient 1 depicting aneurysm progression from ***(a)*** the day of tetralogy of Fallot repair, ***(b)*** 2 months later with an enlarging cardiac silhouette and opacity at the left superior aspect of the heart, and ***(c)*** just prior to aneurysm resection.

Discussion

These five cases demonstrate that large right ventricular outflow tract aneurysms/pseudoaneurysms can occur after right ventricular outflow tract surgical reconstruction. Patients should be monitored for this uncommon complication, especially during the first post-operative year. Careful imaging evaluation is important, since patients can often remain asymptomatic even when the aneurysm/pseudoaneurysm is large,[1] as seen in 4/5 patients in this series. In addition, it can be difficult to differentiate between true aneurysms and pseudoaneurysms clinically. While prior reports have shown pseudoaneurysms are more likely to increase in size rapidly and true aneurysms have a higher chance of stabilising,[2] Patient 5's pathology was consistent with a pseudoaneurysm, which remained stable in size until repair 7 years after diagnosis. Progression can be followed with chest radiographs (Fig 2), which may show widening of the cardiac silhouette or aneurysm/pseudoaneurysm calcification.[3] Echocardiography can be used to diagnose both true and pseudoaneurysms, though two patients in this series were not diagnosed until angiography was performed. Pseudoaneurysms may have a shorter neck and more often have rough margins in comparison to the smooth lining of a true aneurysm.[2] The neck of the pseudoaneurysm will also be narrower and colour flow can help demonstrate continuity of flow to the right ventricle.[3] Lack of myocardial or full thickness endothelial tissue in the pseudoaneurysm wall may increase the risk of rupture of the thin-walled structure, though this did not occur in any of our patients.[1]

This complication should be suspected when there are respiratory complaints, which Patient 1 presented with, as patients are often asymptomatic until the aneurysm/pseudoaneurysm is large enough to cause airway compression, leading to respiratory distress.[1] A chest radiograph may raise suspicion for an aneurysm/pseudoaneurysm, which can be confirmed in most patients by echocardiography. Usually, non-specific symptoms are discovered on routine follow-up, though patients may present with right ventricular failure or syncope.[1,6]

The location of the aneurysm/pseudoaneurysm can differ based on the type of surgical repair, as seen in this case series, and result from disruption or dehiscence of part of the surgically reconstructed outflow tract.[7] Use of ventriculotomy and pericardial patch,[1,2,4] residual obstruction of the right ventricular outflow tract or branch pulmonary arteries,[1,2,4,5] ventricular septal defects,[4] infection, and elevated right ventricular pressure causing excessive stress on the patch or suture line[1–3] have been reported as more common causes. At catheterisation, all patients in this series demonstrated >2/3 systemic right ventricular pressure and branch pulmonary artery stenoses, and three had residual ventricular septal defects.

After the aneurysm/pseudoaneurysm is identified, urgent surgery is usually indicated to resect the aneurysm/pseudoaneurysm and for additional surgical intervention for residual lesions. Four patients in this series had branch pulmonary artery stenoses addressed at the time of aneurysm/pseudoaneurysm repair, and one patient (Patient 2) required catheterisation the next day for right pulmonary artery stent angioplasty. All patients also required subsequent transcatheter bilateral branch pulmonary artery balloon and/or stent angioplasty within 8 months, but the most recent imaging demonstrated no aneurysm/pseudoaneurysm recurrence in any of our patients.

While no patient in this series had rupture, ongoing dilation is possible and, thus, prompt surgical revision should be considered in patients with this complication. Progressive aneurysm/pseudoaneurysm dilation risks rupture and compression of adjacent structures.[3,6] Patients should also be evaluated for related complications such as thrombus formation within the aneurysm, which has been reported and adds risk for embolisation to the pulmonary circulation. Early operative correction is especially prudent in these patients.[4,5] Close proximity or adherence of the aneurysm/pseudoaneurysm to the sternum may make re-operation challenging. Careful pre-procedure assessment with imaging is important to aid with surgical planning.[3]

This case series highlights an uncommon, yet striking, complication that can occur following surgical right ventricular outflow tract reconstruction with associated anatomic conditions which may place a patient at higher risk. Awareness of, and close clinical attention to, this potential complication is important.

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of interest. None.

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https://doi.org/10.1017/S1047951121001025 Published online by Cambridge University Press