

# Precision Diagnosis and Treatment of a Giant Pseudoaneurysm of the Right Ventricular Outflow Tract

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## ABSTRACT

Pseudoaneurysm (PSA) of the right ventricular outflow tract (RVOT) is an exceedingly rare adverse event after the surgical reconstruction of the RVOT for the treatment of congenital heart disease. We report an unusual giant PSA of RVOT in a 20-month-old child, who underwent correction of the tetralogy of Fallot. Her main symptoms were in the respiratory system, and chest X-ray also revealed the giant space-occupying lesion in the chest, which could've been misdiagnosed as a respiratory disease. After evaluation by the combination of echocardiography and cardiac computer tomography angiogram, the details of PSA were diagnosed, and surgical but not percutaneous intervention was selected. The exclusion of PSA successfully was performed by the femoral cannulation, exploratory through right ventriculotomy, closure of the defect using the Gore-Tex patch, and application of a retained drainage-tube inside the PSA.

## INTRODUCTION

Pseudoaneurysm (PSA) of the right ventricular outflow tract (RVOT) is a rare complication of cardiac surgery [Sykes 2017]. In considering the potential risk, intervention is necessary. However, the symptoms of PSA are different and lack specificity, and the diagnostic procedures, selection of intervention methods, and surgical approach still lack standard evaluation criteria. Herein, we report this case with the view to provide a reference for the accurate diagnosis and treatment of RVOT PSA.

## CASE REPORT

A 20-month-old child presented to us with a one-week history of wheezing, coughing, and shortness of breath. Past medical history was tetralogy of Fallot (TOF). At five months of age, she underwent complete ventricular septal

defect closure and enlargement of RVOT, using two pieces of the glutaraldehyde-treated autologous pericardium, measuring the size of 10×12mm and 10×20mm, respectively. The postoperative course was relatively simple, and postoperative thoracic drainage was reasonable, having no evidence of bleeding.

Chest X-ray revealed a giant space-occupying lesion in the mediastinum and left chest with the collapse of the upper lobe (Figure 1). On our examination, lung auscultation was notable for decreased sounds on the left, but clear respirations on the right without any wheezing or rhonchi. Percussion of her posterior chest revealed dullness halfway up her left lung. The remainder of this physical examination and initial laboratory workup was without significant abnormality.

In consideration of her past medical history, transthoracic echocardiography (TTE) was performed and revealed RVOT interruption and an echo-free space near it. Color Doppler flow mapping suggested communication between the RVOT and aneurysm and pulmonary artery blood flow velocity increased (Figures 2A and 2B).

Cardiac computer tomography angiogram (CTA) with three-dimensional reconstruction confirmed the diagnosis of the PSA, which arose from the distal RV-to-pulmonary artery (PA) anastomosis, measuring the size of 76×56×84mm with a

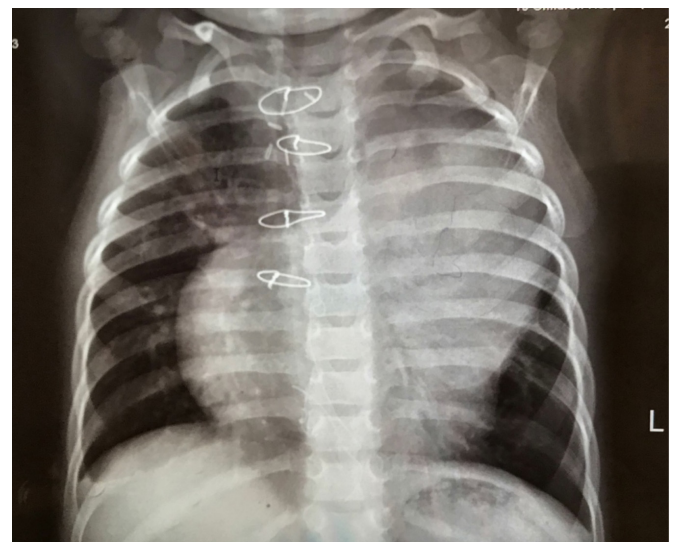


Figure 1. Chest radiograph demonstrating a giant space-occupying lesion in the mediastinum and left chest.

Received June 10, 2020; received in revised form June 24, 2020; accepted June 25, 2020.

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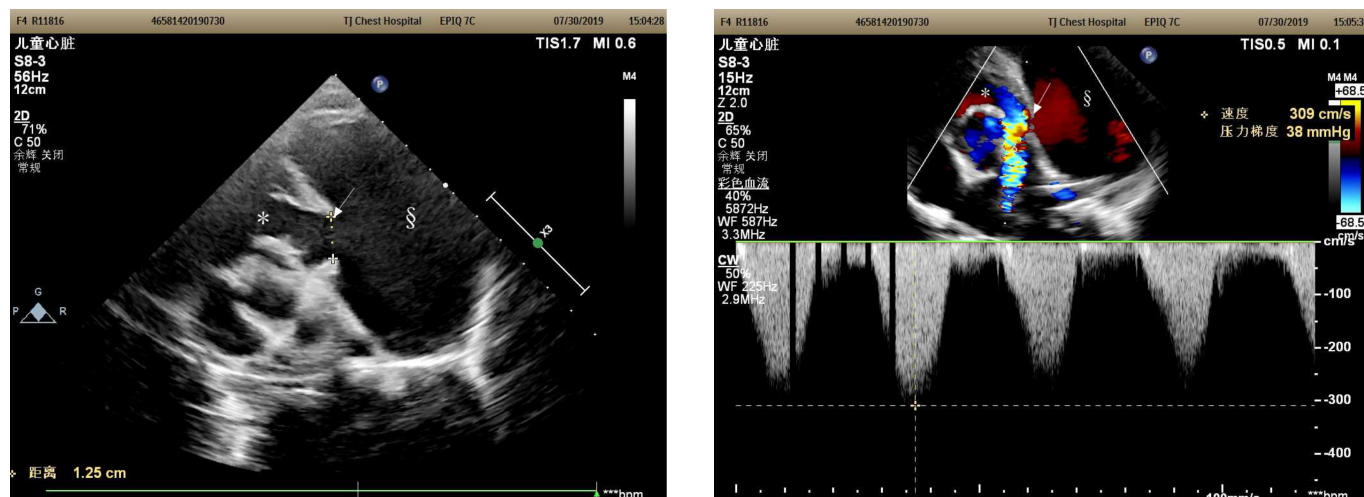


Figure 2. A, Transthoracic echocardiography indicating RVOT (asterisk) interruption (white arrow) and an echo-free space (section sign). B, Color Doppler flow mapping suggesting communication (white arrow) between the RVOT (asterisk) and aneurysm (section sign) (yin-yang or swirl sign) and pulmonary artery blood flow velocity increased.

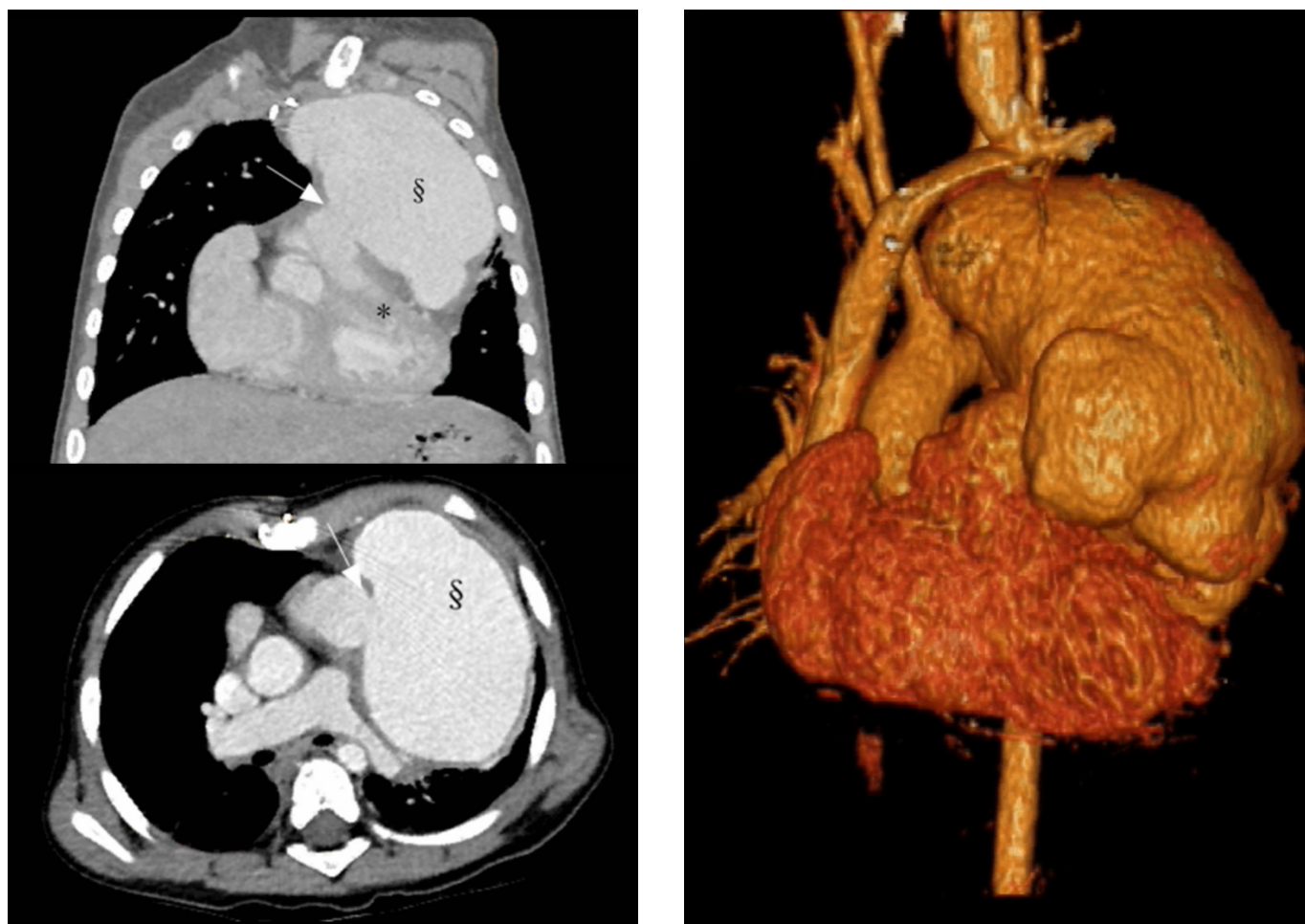


Figure 3. A, Cardiac CTA confirming giant PSA (section sign) arising from the distal RV-to-PA anastomosis with a 17mm fundus (white arrow), and compression atelectasis of upper lobe and the dorsal segment of the lower lobe of the left lung. B, 3D reconstruction of contrast-enhanced angio-CT demonstrating extensive giant PSA.

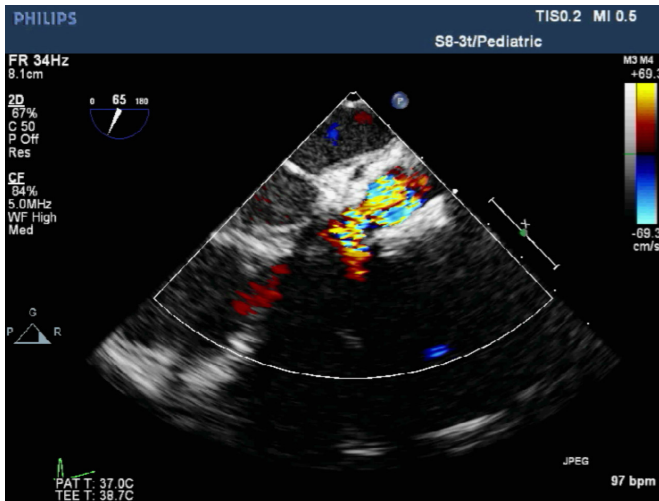


Figure 4. Transesophageal echocardiography further demonstrating the presence of bidirectional blood flow within the PSA (section sign) (yin-yang or swirl sign) and the size and location of PSA and defect (white arrow).

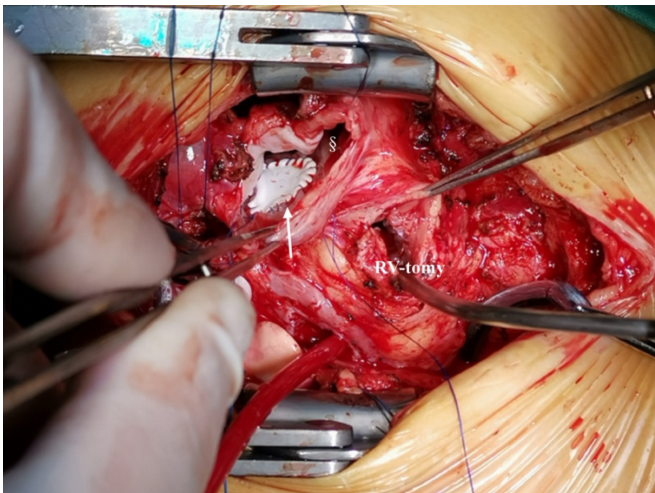


Figure 5. An approximately 17×18mm Gore-Tex patch (white arrow) was used to close the defect (section sign) of RVOT (section sign).

17mm fundus (Figures 3A and 3B). It resulted in compression atelectasis on the upper lobe and lower lobe dorsal segment of the left lung.

The child was taken to the operating room for surgical intervention. Transesophageal echocardiography (TEE) before incision further demonstrated the presence of bidirectional blood flow within the PSA (Figure 4), and the size and location of PSA and defect. A median sternotomy was performed with a standard oscillatory saw, and the adhesions carefully were dissected with cardiopulmonary bypass (CPB) via right femoral cannulation. A giant PSA arose from the RVOT as the echocardiography and CTA revealed, and it firmly adhered to the left lung. PSA exploratory was performed through the right ventriculotomy (Figure 5). The fundus of PSA or the defect of the RVOT measuring about

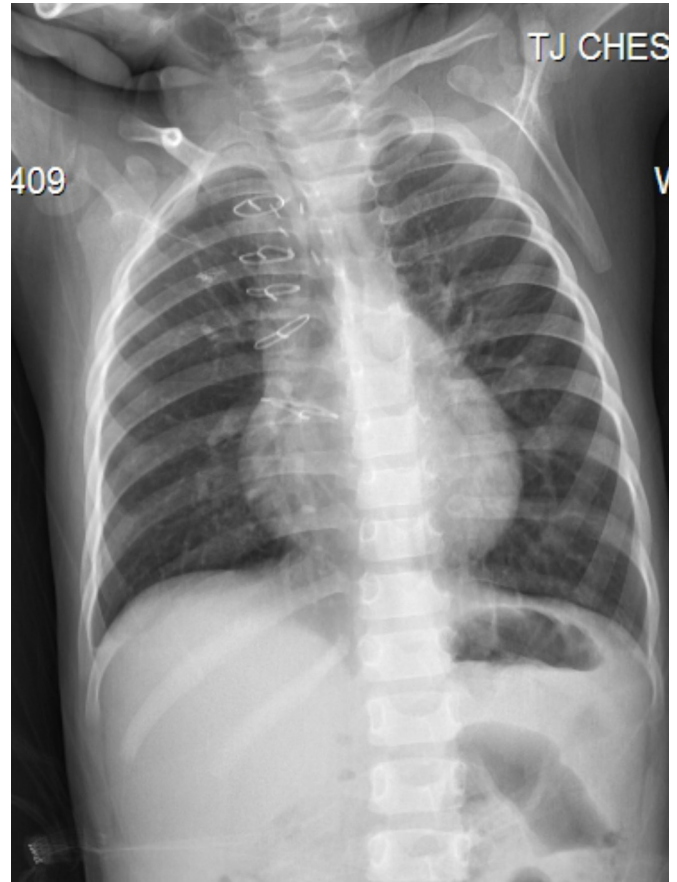


Figure 6. Chest X-ray at the three-month follow up.

15×16mm was finally confirmed. An approximately 17×18mm Gore-Tex (Gore, USA) patch was used to close the defect of RVOT (Figure 5), and a negative pressure drainage-tube was retained inside the PSA. Pathohistological findings confirmed a myocardium defect in the aneurysmal wall and fibrous hypertrophy of the epicardium, consistent with an RV PSA and past effusive pericarditis. The child recovered well and had no further complications at a three-month follow up (Figure 6).

## DISCUSSION

PSA of RVOT is an exceedingly rare adverse event after surgical reconstruction of the RVOT for the treatment of congenital heart disease by patch, synthetic conduit, or homograft placement, analogous to this case [Kaza 2009]. The true incidence of this condition and the risk factors for its development still are not evident. Michelle et al. [Sykes 2017] identified younger age, smaller size, and several other factors, both patient and surgical, that favored the development of a PSA, especially the underlying diagnosis of TOF because TOF is more likely to be associated with the significant branch or peripheral PA stenosis, and patients with tetralogy of Fallot therefore are more likely to have postoperative RV hypertension.



Although literature review documents that acute, dramatic, and life-threatening clinical presentation is infrequent, early intervention is usually recommended, including percutaneous [Lurz 2007; Herbert 2014] and surgical treatment.

The symptoms of PSA are varied, such as thromboembolism, compression of nearby structures, and infection [Sykes 2017]. However, these signs and symptoms are neither sensitive nor specific. Therefore, imaging modalities from echocardiography to CTA or MRI techniques usually are required to diagnose a PSA. TTE often is the first test performed, and color Doppler imaging can confirm the presence of bidirectional blood flow within the PSA, which is called the yin-yang or swirl sign [Lupattelli 2006], although it's low sensitivity. TEE may allow improved detection of PSA diagnosis. Unlike echocardiography, cardiac CT offers high spatial resolution, tissue characterization, and improved visualization of segments that may be difficult to see on echocardiography [Sykes 2017]. Thus, in suspected cases, the combination of TTE, TEE, and cardiac CTA can diagnose the location, size, and precise anatomical relations of PSA. Certainly, surgical and pathohistological evaluation of tissue layers remains the gold standard for diagnosis.

In our case, as outlined above, these procedures proved vital toward accurate diagnosis because her main symptoms were in the respiratory system, and chest X-ray also revealed the giant space-occupying lesion in the chest, which could've been misdiagnosed as a respiratory disease. Intervention was imperative. Percutaneous treatment may have been possible to close the PSA and repair RVOT. Still, it would not have quickly and effectively solved the problems of the compression on the left lung by the giant PSA, the impact on the development of the child, and subsequent infections from the absorption of thrombus. As such, surgical intervention was

selected. During the operation, the CPB technique with femoral cannulation to prevent PSA rupture undergoing median sternotomy and dissecting the adhesions, PSA exploratory through right ventriculotomy and application of a retained negative pressure drainage-tube inside the PSA, which can relieve the left lung compression caused by the PSA effusion after the operation and promote the reexpansion of the left lung, were critical points.

## ACKNOWLEDGEMENT

This work was supported by Science and Technology Funding of Tianjin Chest Hospital [grant numbers 2018XKZ14].

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