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Diagnosis and Management of Fontan Failure Secondary to Aortopulmonary Artery Fistula

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Abstract

Aortopulmonary artery fistula represents a rare anomalous communication between aorta and pulmonary artery. The treatment of these communications is well established and involves either surgical or percutaneous approach. We present a 15-year-old male with history of hypoplastic left ventricle, hypoplastic aorta and ventricular septal defect with Damus-Kaye-Stansel surgery in the past, who developed acute Fontan circuit failure secondary to the development of aortopulmonary fistula of unknown etiology. Fistula was successfully closed percutaneously, using Amplatzer duct occluder.

Key Words

Aortopulmonary artery fistula • Congenital heart disease • Duct occluder

Introduction

Aortopulmonary artery fistula (APF) represents an anomalous communication between aorta and pulmonary artery. It is a rare occurrence. There are two types of APFs described in the literature: congenital and acquired. Congenital APFs are extremely rare and have only been described in case reports [1]. On the other hand, acquired APFs are relatively more common and usually associated with pseudo-aneurysm that breaks into the pulmonary artery [2-4]. They may or may not be associated with trauma [5]. Treatment for these abnormal communications is well established and is either surgical or percutaneous [2, 6]. We present an interesting case of Fontan circuit failure due to development of aortopulmonary artery fistula in a 15-year-old male with history of complex congenital heart disease including hypoplastic left ventricle, hypoplastic aorta and ventricular septal defect (VSD) with Damus-Kaye-Stansel surgery in the past followed by Fontan palliation with fenestration and transcatheter fenestration closure years after primary surgery. He presented to emergency department with increasing abdominal pain in the setting of recent bicycle accident followed by dyspnea on exertion.

Case presentation

15-year-old male with history of complex congenital heart disease, status post Fontan palliation presented to an outside hospital (OSH) ER two weeks after a fall while riding his bike when flipped over the handle bars. Over the two-week period following the accident, patient developed increasing abdominal discomfort and dyspnea on exertion and finally pre-

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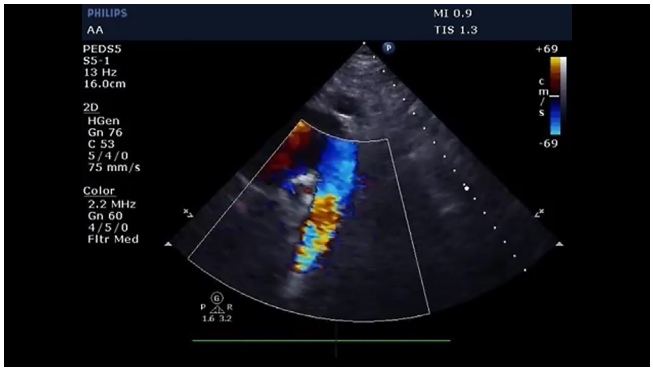
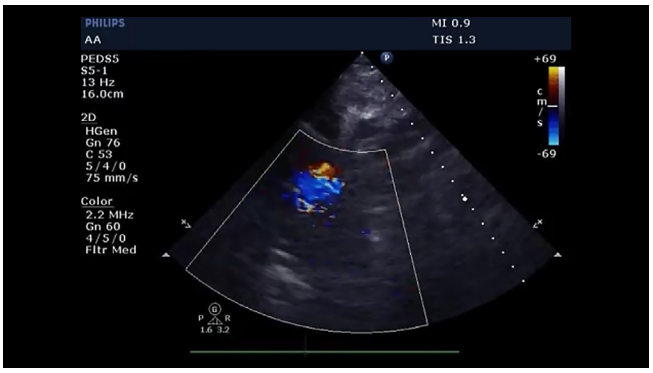
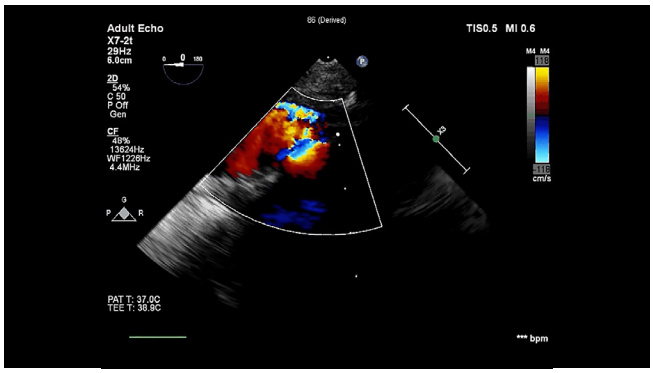


Video 1. The reversal of flow in descending aorta in diastole without aortic regurgitation. View supplemental video at https://doi.org/10.12945/j.jshd.2019.014.18.vid.01.



Video 2. Shunt-like flow coming from aorta to pulmonary artery (transthoracic echocardiogram). View supplemental video at https://doi.org/10.12945/j.jshd.2019.014.18.vid.02.



Video 3. APF between the posterior right aspect of the aorta and the pulmonary artery (transesophageal echocardiogram). View supplemental video at https://doi.org/10.12945/j.jshd.2019.014.18.vid.03.

sented to ER due to severe abdominal pain, mostly localized to right upper quadrant.

At the OSH ER abdominal CT was done which showed questionable injury to liver and spleen. At that moment it was decided to transfer the patient to our hospital for trauma evaluation. Following arrival to our hospital, abdominal CT was found to be reassuring. Upon further questioning it came to our knowledge that patient had multiple months of increased exercise intolerance including dyspnea on exertion which was occurring quicker into activities. Patient also reported orthopnea and was using an extra pillow to sleep during this time. Given the afore mentioned, chest X-ray was obtained and found to be concerning for bilateral pleural effusions.

Cardiology was consulted due to positive history of complex congenital heart disease. Transthoracic echocardiogram (TTE) was completed and showed reversal of flow in descending aorta in diastole without evidence of aortic regurgitation (Video 1). Further investigation on echocardiogram made us suspect that there is shunt-like flow coming from aorta to pulmonary artery (Video 2), but it was not clearly identified on TTE. It was determined that a diagnostic catheterization and transesophageal echocardiography (TEE) would be needed to identify the source.

The patient had increased oxygen requirements due to worsening pleural effusions and increasing ascites requiring transfer to the cardiac intensive care unit where bilateral thoracentesis was performed. Bilateral 8 Fr chest tubes were placed followed by almost complete resolution of his bilateral pleural effusions. However, prior to catheterization the patient had a persistent right sided pleural effusion causing desaturations while under anesthesia. Right chest tube (20 Fr) was placed which helped stabilize the patient but failed to adequately drain the effusion. The patient was taken to the catheterization lab where surgical drainage was performed under general anesthesia.

Once placed under anesthesia and the right sided pleural effusion drained, TEE was done. The TEE confirmed our suspicion of APF between the posterior right aspect of the aorta and the pulmonary artery (Video 3). It was determined that a percutaneous approach should be attempted with cardiothoracic surgery back up.

Right heart catheterization was performed using 7-Franch Berman angiographic catheter and revealed

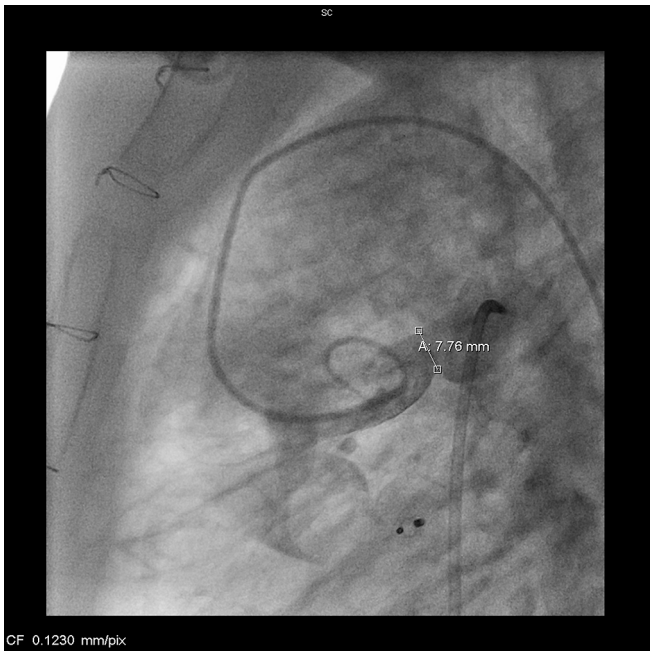
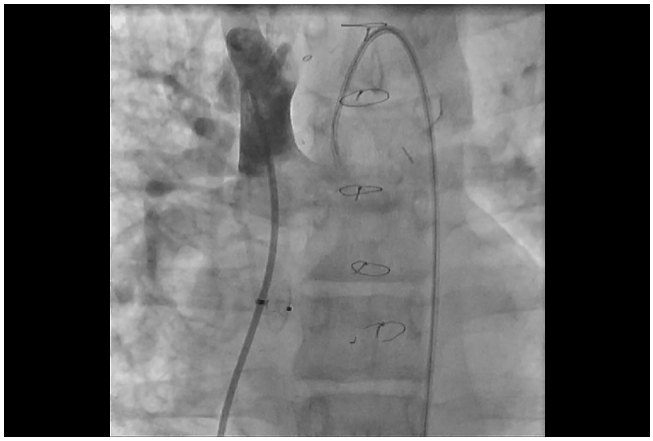
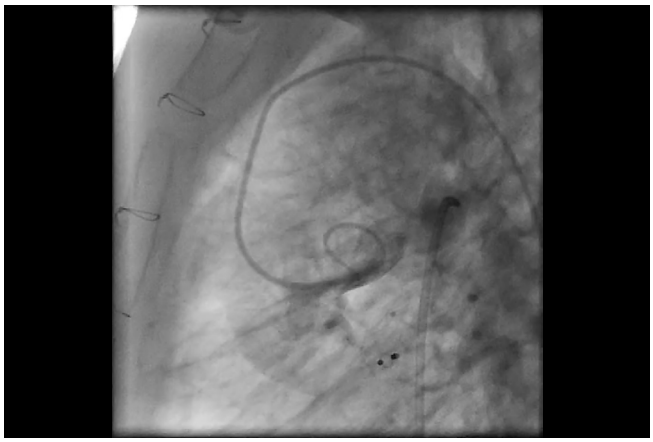


Figure 1. APF diameter measuring 7 mm.

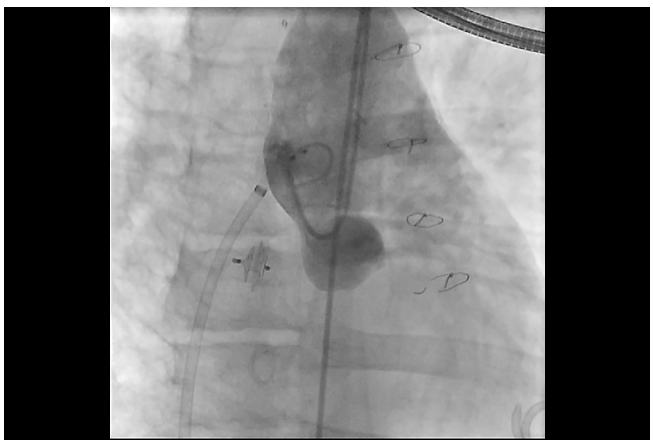
inferior vena cava, superior vena cava, Fontan baffle and pulmonary artery pressures to be all at a mean of 25 mm Hg implying increased pressure in Fontan circuit without obstruction. Pulmonary angiogram also showed contrast washout in pulmonary artery suggesting additional flow coming from high pressure setting (Video 4). Retrograde left heart catheterization performed with 6-French pigtail catheter revealed left ventricular pressure of 90/10 mmHg and no gradient to the ascending or descending aorta where pressure was 90/35 mmHg. An aortogram was performed in postero-anterior and lateral projections demonstrating competent aortic and neoaortic valve and normal coronary flow. Angiogram confirmed presence of aortopulmonary artery fistula (Video 5). The diameter of fistula was measured to be 7 mm (Figure 1) prior to advancing balloon sizing catheter. After creating arteriovenous loop, a balloon sizing catheter was advanced from the venous side and softly inflated across the fistula. The decision was made to close the fistula using Amplatzer duct occluder 10/8 mm in size. After successful positioning of the device and its release, a post-release aortogram confirmed good position and a trivial residual shunting through the device which was expected (Video 6). Post-release TEE also confirmed the device to be in a



Video 4. Contrast washout in pulmonary artery suggesting additional flow coming from high pressure setting (pulmonary angiogram). View supplemental video at https://doi.org/10.12945/j.jshd.2019.014.18.vid.04.



Video 5. Aortopulmonary artery fistula. View supplemental video at https://doi.org/10.12945/j.jshd.2019.014.18.vid.05.



Video 6. Device position with trivial residual shunting. View supplemental video at https://doi.org/10.12945/j.jshd.2019.014.18.vid.06.

good position. Following the closure of the fistula, the Fontan circuit pressure decreased from 25 mm Hg to 20 mm Hg of mean pressure.

The patient was extubated in catheterization lab and transferred back to the intensive care unit. Over the next couple of days, the patient improved and both chest tubes were removed. TTE prior to discharge showed a small shunt at the site of the fistula, but overall improved. Patient was discharged home with cardiology follow-up and twice daily Furosemide with plan to wean the medication over the next few weeks. Outpatient cardiology follow-up showed consistent improvement.

Discussion

Aortopulmonary artery fistula is a rare type of anomalous vascular communication. This unusual communication, reportedly, has been associated to aortic aneurysm or has appeared as a complication of aortic aneurysm rupturing into the pulmonary artery [2].

There are two types of APF: congenital and acquired. Congenital AFP are extremely rare but have been described in case reports in which patients also had an associated coronary artery fistula. The pathogenesis is not very well understood. [1]

Our case likely represents a case of acquired APF given that this is a patient with history of complex congenital heart disease who had multiple echocardiographic evaluations and catheterizations in the past not revealing the anomalous communication. However, the etiology is still not clear. It has been shown that aortopulmonary collateral vessels occur with the overall prevalence of 36% in the population of patients who have undergone bidirectional Glenn or Fontan procedure [7] , a category under which our patient falls.

Acquired APF in the pediatric and adult populations are shown to be associated with some level of defect in the wall of the great vessels including an intimal tear, pseudoaneurysm or aneurysm breaking into the pulmonary artery and resulting in direct communication between these great vessels [3, 4]. In pediatric populations, there have been multiple reports of iatrogenic APF formations after pulmonary angioplasty using different devices for percutaneous closure [8, 9]. In all cases, a mild intimal tear in the pulmonary artery or its branches was revealed using magnetic resonance imaging (MRI). Trauma is a significant risk factor for the development of mild to severe injuries of the vessel wall that can lead to fistula formation [4, 5]. Most vessel wall injuries, even minor ones, would likely be seen on MRI which was not done in this case. However, it is reasonable to say that his bicycle accident could have predisposed him to developing a minor tear in the wall of his great vessel. Although our patient did have a fall prior to presentation, his symptoms date back prior to the fall, therefore the etiology of his condition remains uncertain.

APF leads to significant left-to-right shunt and places a hemodynamic burden on the heart and lungs. Clinical presentation is consistent with variable level of congestive heart failure depending on the diameter of the anomalous vessel as well as the predisposing factors such as trauma. In case of significant left-to-right shunting from aorta to pulmonary artery, as described in our patient with APF where blood was shunted from aorta where systolic pressure was 90 mmHg to a much lower pressure in pulmonary artery leading to increased Fontan circuit pressures to 25 mm Hg and overcirculation of the pulmonary vascular bed with consequent dyspnea, pleural effusions and pulmonary edema. Additionally, increased Fontan circuit pressure leads to liver congestion, ascites and abdominal symptoms. In patients with Fontan palliation, in order for the circuit to function properly, low circuit pressures must be maintained. The increase in Fontan circuit pressure to 25 mm Hg in our patient contributed significantly to liver congestion and to ascites formation further on. It is well known that high venous pressures can compromise lymphatic circulation as well. Additionally, the extension of Glisson's capsule can also lead to right upper quadrant abdominal pain that our patient initially presented with.

The treatment of APF includes surgical repair or percutaneous closure of the anomalous vessel using different types of devices. Surgical treatment is the treatment of choice in the case of aortic pseudoaneurysm or aneurysm rupturing into the pulmonary artery and in case of severe trauma leading to the communication of the great vessels. Percutaneous embolization has become the treatment of choice for the occlusion of anomalous arteriovenous and

veno-venous connections frequently seen in patients with congenital heart disease [10]. Major advantages of transcatheter closure are the decreased need for reoperation leading to decreased postoperative morbidity and mortality [11]. APF occlusion using Amplatzer vascular occluder, in our patient, lead to a decrease in Fontan circuit pressures to a mean pressure of 20 mm Hg while the patient was still intubated and on positive pressure ventilation and it was expected for this pressure to decrease further with resolution of pleural effusions and ascites, as it was observed. Today, many devices are available for percutaneous closure of APF including coils, plugs, microspheres, glue and occlusion balloons and its safety and efficacy has been well established so far [6, 8, 10, 11].

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Conflict of Interest

The authors have no conflict of interest relevant to this publication.

Comment on this Article or Ask a Question

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