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CASE REPORT

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Descending aorta to right atrial fistula: Transcatheter embolization of a very rare anomaly with coils

Stefanos Despotopoulos[1] | **Sotiria Apostolopoulou**[1] | **George Vagenakis**[1] | **Meletios Kanakis**[2] | **George Samanidis**[3] | **Panagiotis Zachos**[4] | **Anastasios Chatziantoniou**[1] | **John Papagiannis**[1] | **Spyridon Rammos**[1] | **Alexandros Tsoutsinos**[1]

[1]Department of Pediatric Cardiology and Adult Congenital Heart Disease, Onassis Cardiac Surgery Center, Athens, Greece

[2]Department of Pediatric and Adult Congenital Heart Surgery, Onassis Cardiac Surgery Center, Athens, Greece

[3]1st Department of Cardiac Surgery, Onassis Cardiac Surgery Center, Athens, Greece

[4]Pediatric Cardiology & Adult Congenital Heart Disease Department, General Hospital of Karditsa, Karditsa, Greece

Correspondence
Alexandros Tsoutsinos, Department of Pediatric Cardiology and Adult Congenital Heart Disease, Onassis Cardiac Surgery Center, Athens, Greece.
Email: tsutsi@otenet.gr

Key Clinical Message

Descending aorta to right atrial (RA) fistula is a rare and distinct clinical entity mimicking patent ductus arteriosus (PDA) and it may lead to rapid development of pulmonary vascular disease. Correct diagnosis and treatment, especially in the presence of other congenital heart defects, is very important. Interventional management is the treatment of choice.

Abstract

We present a case report of a trisomy 21 infant with atrial and ventricular septal defects and small patent ductus arteriosus (PDA) complicated by the presence of descending aorta to right atrial (RA) fistula with large left to right shunt leading to rapid increase in pulmonary vascular resistance. Transcatheter occlusion of the fistula followed by closure of the PDA with Nit-Occlud coil systems led to decreased pulmonary pressure and resistance permitting successful surgical repair of the patient's intracardiac defects with good outcome over 3 years of follow-up. Descending aorta to RA fistula is a rare and distinct clinical entity mimicking PDA and its correct diagnosis and treatment, especially in the presence of other congenital heart defects, is very important as it may lead to rapid development of pulmonary vascular disease.

KEYWORDS

aorta to right atrial fistula, cardiac surgery, coil, congenital heart disease, transcatheter occlusion

1 | INTRODUCTION

Communications between the aorta and the right and left atria are uncommon anomalies and usually involve the ascending aorta either in the form of congenital coronary cameral fistulae[1] or acquired connections associated with aortic dissection[2] and endocarditis. Fistulous malformation from the descending aorta is very rare and has only

been described in three previous reports, connecting to the right atrium,[3] left atrium[4] or the superior vena cava, azygos vein and the innominate vein.[5] In all cases it was isolated, mimicked the clinical picture of patent ductus arteriosus (PDA) or other aorto-cardiac fistulae and it induced large left to right shunt which was abolished surgically. The fistula was successfully occluded by transcatheter intervention.

2 | CASE REPORT

A 7-month-old 5.6 kg girl with trisomy 21 presented to our hospital with tachypnea and failure to thrive. On physical examination, she had a continuous murmur auscultated best over the right precordium and the left back and hepatomegaly 2 cm below the left coastal margin. On echocardiography, she had a moderate 10 mm secundum atrial septal defect, a small 4 mm high muscular ventricular septal defect, a small PDA, severe right ventricular dilation and high-velocity continuous flow entering the RA with 70 mmHg gradient. An abnormal tortuous vessel con depicted originating from the descending aorta below the isthmus, while Doppler interrogation of the tricuspid regurgitation and the flow through the ventricular septal defect indicated systemic pulmonary artery pressures. The patient underwent cardiac catheterization after written informed consent was obtained from her parents to delineate the anatomy and hemodynamic impact of the vessel as her intracardiac defects did not account for the degree of her failure to thrive and pulmonary hypertension.

On catheterization, she had systemic systolic pulmonary artery pressure at 101 mmHg with pulmonary to systemic flow ratio (Qp/Qs) 3.1, calculated pulmonary vascular resistance (PVR) 7.3 Wood units and pulmonary to systemic vascular resistance ratio 0.24. On administration of 100 FiO_2 Qp/Qs increased and dropped her PVR to 3.9 Wood units. Angiography depicted the atrial septal defect, the small muscular ventricular septal defect and PDA and a continuous flow vessel with multiple stenotic and dilated parts originating from the descending aorta just below the isthmus and, after a long tortuous course, entering the right atrium (Figure 1). After entering the fistula from the right atrial opening, a 5×4 mm Nit-Occlud (PFM, Cologne, Germany) coil system (nitinol based spiral device with double disk pyramid configuration designed for interventional PDA occlusion) was used in order to achieve a complete occlusion without complications (Figure 2A). The PDA was not addressed at this session as it appeared small and not hemodynamically significant on angiography and the patient had received >7 mL/kg of contrast during the procedure.

After the intervention, the patient improved significantly with resolution of the tachypnea and hepatomegaly and satisfactory weight gain. Repeat catheterization 4 months later at weight 7.5 kg, showed complete occlusion of the fistula, systolic pulmonary artery pressure 2/3 systemic at 71 mmHg with Qp/Qs 5.1, PVR 2.8 Wood units and pulmonary to systemic vascular resistance ratio 0.12. Closure of the PDA with another 5×4 mm Nit-Occlud coil system (Figure 2B) did not change hemodynamics at all. Due to the elevated, albeit improved, pulmonary artery pressures, the presence of trisomy 21, which may accelerate pulmonary vascular disease, interventional closure of her septal defect was performed with a benign postintervention course, despite the patient's small size.

Pulmonary artery waveform showed a significant decrease in pressure after the closure compared to this before the closure (Figure 3). Six months later, the patient underwent a midline sternotomy and septal defects were successfully closed. Postoperative course was uneventful. She remains well with no cardiovascular symptoms and normal weight gain over a 5-year of follow-up, while echocardiography demonstrates normal biventricular size and function and mild tricuspid regurgitation without pulmonary hypertension.

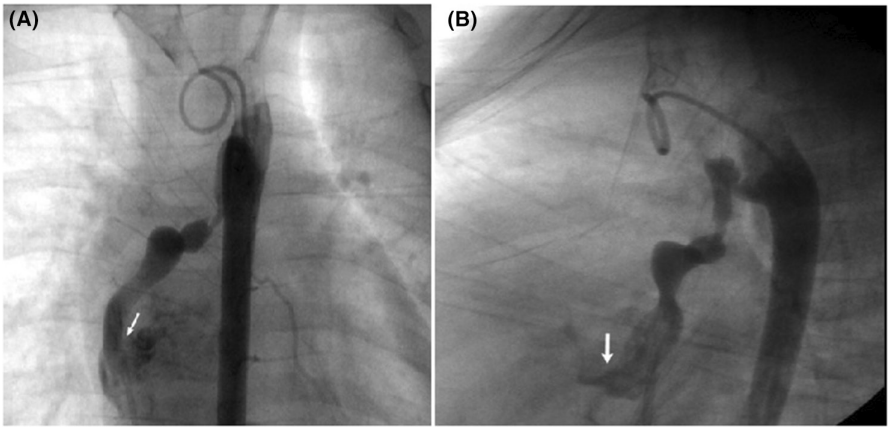

FIGURE 1 Frontal (A) and lateral (B) projection of descending aortogram depicting a very tortuous vessel with many stenotic and dilated portions originating just below the isthmus and entering the right atrium (arrow).

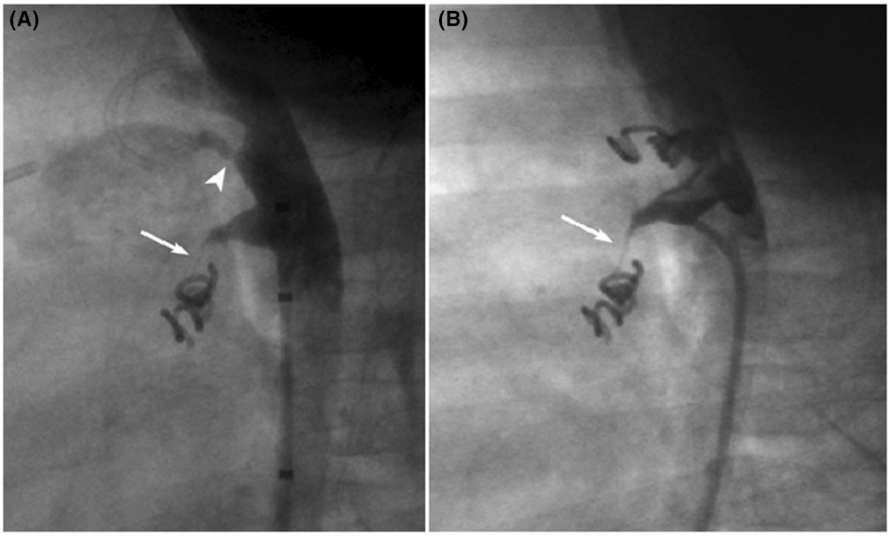

FIGURE 2 (A) Lateral projection of descending aortogram 4 months after the fistula closure showing complete occlusion of the fistula by the Nit-Occlud coil system with very narrow tract (arrow) from the ampulla of the fistula to the coil. The arrowhead points to the patent dusctus arteriosus. (B) Lateral projection of descending aortogram after the patent ductus arteriosus closure depicts the Nit-Occlud coil system in the occluded fistula with the narrow tract leading to it and placed Nit-Occlud coil system above it occluding the patent ductus arteriosus.

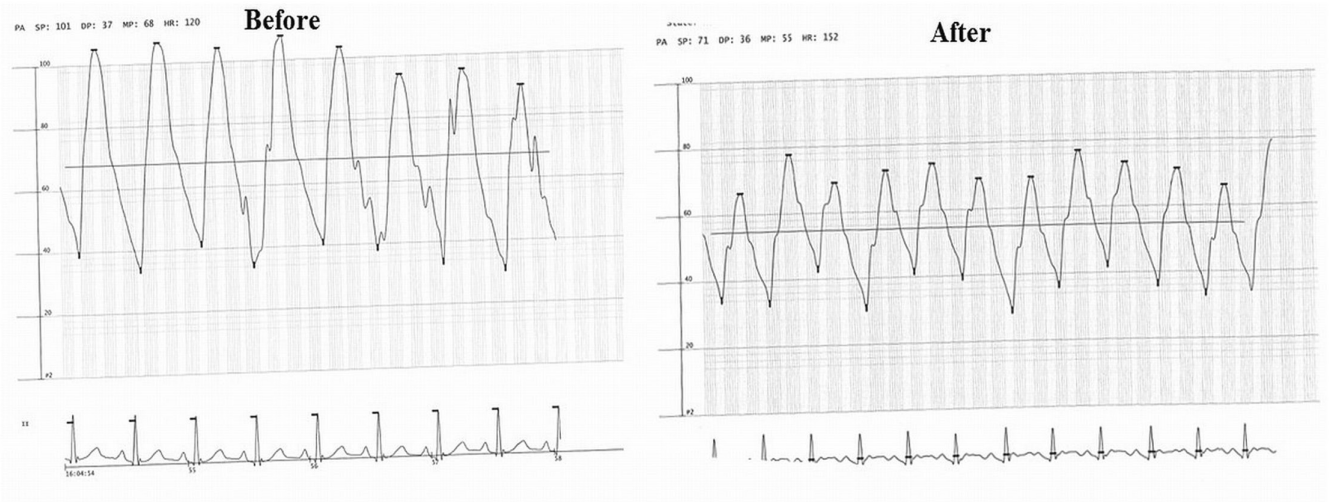

FIGURE 3 Pulmonary artery waveform before and after intervention depicting a significant drop in the pressure.

3 | DISCUSSION

Arteriovenous fistulas of the heart are relatively rare and may involve one of the coronary arteries, an intercostal artery or an internal mammary artery. They may be associated with continuous machinery murmurs, mimicking PDA. Some of these fistulas may join the aorta to the right or left atrium. Most previous reports refer to acquired ascending aorta to cameral fistulae in conjunction with aortic dissection,[1,2,6] surgery,[3–5] infection for example, endocarditis[7–12] and foreign bodies such as ASD septal occluder,[13–15] or pacemaker wire.[16] Furthermore, aorto-atrial fistulas could be associated with thoracic trauma,[17–21] or heart transplantation.[22] Congenital aorta to atrial fistulous communications is very rare.

Danilowicz et al.[23] reported a congenital fistulous tract between the right aortic sinus of Valsalva and the right atrium in a newborn, who presented with congestive heart failure on the first day of life and was addressed surgically at 5 days of age.

Moreover, Rosenberg et al.[24] reported four cases of unusual congenital aortic-right atrial communications, in asymptomatic patients with an atypical continuous murmur on examination. Three of these fistulas were surgically corrected.

In addition, Soler reported a case of congenital arteriovenous fistula from the descending aorta draining separately to the superior vena cava, the azygos vein, and the innominate vein. Clinically, the case simulated a PDA.

A case of congenital fistula between the descending thoracic aorta and the left atrium was reported by Nihoyiannopoulos et al.[25] and was surgically ligated.

Finally, Elwatidy et al.[26] reported a congenital descending aorta to right atrial channel, similar to our patient's fistula, not associated with other congenital abnormalities, which initially was misdiagnosed as a PDA and was eventually closed surgically.

The embryologic origin of such arterio-cardiac chamber communications is unclear. Most probably it is due to an abnormality during the simultaneous embryologic development of the respiratory system and the heart tube formation. On the one hand, the first is formed by the primitive foregut and their common vascular system, called the splanchnic plexus, which drains into the cardinal and umbilicovitelline venous systems. As the lung buds develop, the anterior portion of the splanchnic plexus differentiates into the primitive pulmonary vascular plexus.[27,28] On the other hand, the second one is formed by the dorsal mesocardium, which originates from part of the splanchnic mesoderm, connecting the primitive heart to the lung mesenchyme. In other words, an anomaly of the sixth aortic arch may be associated with abnormal regression of the cardinal veins or sinus venosus and contribute to the development of that type of fistula. A high index of suspicion for this lesion is advocated in patients with unusual location of continuous murmurs and congestive heart failure or pulmonary hypertension out of proportion for the size of their intracardiac shunts.

Our patient is a quite rare case of descending aorta to right atrial fistula in a trisomy 21 patient with atrial and ventricular septal defects leading to rapid increase in pulmonary vascular resistance. The fistula was successfully occluded following transcatheter intervention.

Diagnosis and treatment of these fistulae becomes very important as they usually produce very increased left to right shunt with ventricular dilation[26] and may lead to rapid increase of PVR, especially in a patient with trisomy 21, as in our case. Echocardiography usually leads to the diagnosis, but cardiac catheterization plays an essential role in the diagnosis and management of this defect as it permits complete hemodynamic evaluation, precise anatomic delineation and, if possible, interventional closure.

4 | CONCLUSION

In summary, this is the first literature report of a descending aorta to right atrial fistula in conjunction with atrial and ventricular septal defects and PDA in a trisomy 21 infant, which caused rapid increase of pulmonary vascular resistance and was successfully occluded by transcatheter placement of Nit-Occlud coil system.

AUTHOR CONTRIBUTIONS

Stefanos Despotopoulos: Writing – original draft. **Sotiria Apostolopoulou:** Investigation. **George Vagenakis:** Investigation; writing – original draft. **Meletios Kanakis:** Investigation; methodology; supervision; visualization; writing – original draft. **George Samanidis:** Conceptualization; investigation; validation; visualization; writing – original draft. **Panagiotis Zachos:** Investigation. **Anastasios Chatziantoniou:** Investigation. **John Papagiannis:** Investigation; writing – original draft. **Spyridon Rammos:** Investigation; methodology; supervision; writing – original draft. **Alexandros Tsoutsinos:** Writing – original draft.

FUNDING INFORMATION

None.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author.

CONSENT

Written informed consent was obtained from the parent of children to publish this report in accordance with the journal's patient consent policy.

ORCID

Meletios Kanakis https://orcid.org/0000-0002-9708-0523
George Samanidis https://orcid.org/0000-0001-6355-1932

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