CASE REPORT

Transcatheter closure of direct right pulmonary artery to left atrial communication using vascular plug

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SUMMARY

A direct right pulmonary artery to left atrial fistula is a rare congenital condition in which patients usually present with isolated cyanosis in the absence of abnormal cardiovascular findings. We report the percutaneous closure of such a defect in a young woman who presented with severe cyanosis. This was achieved without making a conventional venovenous loop and was performed using an Amplatzer vascular plug. We believe, in patients with a suitable anatomy, closure with vascular plug without a venovenous loop should be the method of choice for treating this malady in view of the technical ease.

BACKGROUND

A fistulous communication between right pulmonary artery (RPA) and left atrium (LA) is a rare congenital anomaly, usually presenting in adolescence or adulthood. A normal cardiovascular examination in the presence of central cyanosis is the hallmark of this malady. The diagnosis is often arrived at by a combination of contrast echocardiogram with a CT angiogram, or with an invasive pulmonary angiogram. Since its first description in 1950,¹ open surgery was the therapy of choice; however, the last decade saw catheter-based techniques being used for treating a select group of patients.^{2–8} We report a case of successful device closure of congenital RPA to LA communication in the cardiac catheterisation laboratory using an Amplatzer vascular plug (AVP) without making a conventional venovenous loop.

CASE PRESENTATION

A 28-year-old woman presented with symptoms of gradually progressive effort intolerance for the past 5 years, which worsened after pregnancy and delivery 3 months prior. She reported bluish discolouration of her extremities since adolescence; the discolouration had never been evaluated. Clinical examination, including that of her cardiovascular system, was unremarkable except for central cyanosis (resting saturation: 77%) and grade II clubbing. ECG revealed sinus rhythm along with features of left atrial enlargement and left ventricular (LV) hypertrophy. Chest roentgenogram showed cardiomegaly of the LV contour with an abnormal right hilar silhouette (figure 1A). In view of the unexplained cyanosis, an echocardiogram was performed, which revealed severe LV systolic dysfunction (ejection fraction of 26%) with global

hypokinesia; however, there was no evidence of any intracardiac or arterial level shunt lesions.

INVESTIGATIONS

In order to elucidate the aetiology of cyanosis, contrast echocardiography was performed, using agitated saline, in which the bubbles appeared in the LA five beats after right heart opacification, suggesting a pulmonary arteriovenous (AV) fistula as the diagnosis (figure 1B, video 1). CT angiography was subsequently performed for anatomical delineation of the shunt, which conclusively proved the presence of a large fistulous communication between the RPA and the LA (figure 1C).

The exact aetiology of the ventricular systolic dysfunction was not clear, as this could not be explained by the pulmonary level shunt. The possibility of peripartum cardiomyopathy was considered, and hence the patient was started on heart failure medications including angiotensin receptor blockers, β -blockers and diuretics. As cyanosis was contributing to her symptoms and to avoid the complications associated with the pulmonary AV fistula,⁹ it was decided to percutaneously close this defect.

TREATMENT

After obtaining informed consent, the patient was taken up for device closure of the fistulous communication under local anaesthesia and conscious sedation. Percutaneous access to the right femoral artery and vein was obtained and the patient was adequately heparinised. Cardiac catheterisation disclosed a femoral artery saturation of 72%, normal pulmonary artery pressures and absence of cardiac level shunting (table 1). Pulmonary angiogram confirmed the CT findings of a large tubular communication (measuring 14 mm) from the lower branch of the RPA opening to an aneurysmal sac, which eventually drained into the LA (figure 1D, video 2). The presence of other fistulous communication on the contralateral side was excluded. In view of this peculiar anatomy, it was decided to close the long tubular portion of the communication using a vascular plug.

The right lower pulmonary artery was selectively entered using a Judkins right catheter. An 0.035/260 cm hydrophilic wire (Terumo) was then manipulated across the fistulous communication to enter into the aneurysmal sac; the wire was then changed to an 0.035/Amplatzer Super Stiff guidewire (Boston Scientific) inside the catheter. This was parked in the sac with a loop so as to provide the

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Figure 1 Chest X-ray showing cardiomegaly and abnormal right hilar shadow by the right pulmonary artery to left atrial fistula (A); bubble contrast echo showing opacification of the left-sided chambers (B). The fistula origin from right lower pulmonary artery (*) and exit into LA (arrow heads) are displayed in the three-dimensional reconstruction of CT (C) and invasive pulmonary angiogram (D). (LA, left atrium; LV, left ventricle; RA, right atrium; RLPA, right lower pulmonary artery; RV, right ventricle).

necessary support. The 7F femoral venous sheath was then changed to a 12F 80 cm long sheath (Cook Medical Inc, Indiana, USA), which was subsequently taken across the tubular portion of the fistula into the sac. An 18 mm AVP II (St Jude Medical, Minnesota, USA) was introduced through the 12F

sheath and positioned in the tubular segment. After confirming the position and stability of the device, the plug was released. An angiogram at this stage revealed closure of the main communication. However, a separate small feeder artery was seen, measuring 4 mm in diameter, arising proximal to the plugged

Video 1 Contrast echo using agitated saline showing the dense filling of the left heart with bubbles five beats after right heart opacification.

Table 1 The cardiac catheterisation data before and after the closure of the right pulmonary artery to left atrial fistula

	Before closure		After closure	
	Pressures (mm Hg)	Saturations (%)	Pressures (mm Hg)	Saturations (%)
Mixed venous	—	43		
Right atrium	5 Mean	44		
Right ventricle	26 Systolic, Ed 5	43		
Pulmonary artery	25/14/19	43	24/16/20	67
Pulmonary capillary wedge	14 Mean	—		
Left ventricle	96 Systolic, Ed 15	72		
Ascending aorta	98/56/70	72	104/64/76	96
Ed, end diastolic.

Video 2 Digital subtraction angiography using pigtail catheter in the right pulmonary artery showing the abnormal communication connecting to the left atrium.

channel and draining into the aneurysmal sac (figure 2A, asterisk). This was then coiled with a 018' 6–3 mm embolisation coil (Cook) using a microcatheter (figure 2B). The final angiogram showed complete closure of the RPA-LA fistula (figure 2C, video 3).

OUTCOME AND FOLLOW-UP

Immediately after the procedure, the femoral artery saturation improved to 96% and the clinical cyanosis disappeared. A bubble contrast echocardiography performed the next day confirmed the absence of any residual shunt (video 4). The patient was treated with oral anticoagulation for 6 months and is asymptomatic after 1 year of the procedure (figure 2D). There was partial recovery in LV function with ejection fraction improving to 45% in the echo performed after 1 year.

DISCUSSION

The fistulous connection between RPA and LA is a rare congenital pulmonary circulation anomaly with only about 65 cases reported so far.⁷ This is seen more frequently in males than in females, with a reported frequency of 3:1.⁹ Although a few patients have been diagnosed at infancy,² the majority are identified following delayed onset of cyanosis at adolescence or even adulthood. Symptoms at presentation include progressive cyanosis, effort intolerance, embolic manifestations, endarteritis, hyperviscosity symptoms and even fatal rupture.⁹ Diagnosis is made in the presence of central cyanosis with a normal cardiovascular system, although associated anomalies such as atrial septal defects may complicate the clinical picture. The embryological explanation for this condition is thought to be the persistence of an abnormal channel between the RPA and primitive pulmonary vein; the latter, when it gets incorporated into the LA, leads to the development of this malady.¹⁰

Figure 2 Angiographic image after deploying the Amplatzer vascular plug (arrow) showing a small feeder artery (*) draining into the distal sac (A). An embolisation coil (arrowhead) was used to close this feeder artery (B). Final angiogram showing complete exclusion of the fistulous communication (C). One year later, the patient is asymptomatic with X-ray showing reduction in cardiac size (D) (RLPA, right lower pulmonary artery).

Video 3 Postprocedure digital subtraction angiography showing the complete closure of the abnormal communication. The Amplatzer vascular plug and coils are both seen.

Surgery was the only treatment available for these patients until the first transcatheter coil closure was performed in 2000.² The surgical approach depends on the anatomy, with an off-pump ligation preferred whenever feasible, although alternative surgeries such as ligation and division, and excision and intracardiac repair, have been performed in a minority of patients, with excellent results.⁹ Recent advances in the catheter-based techniques brought this condition into the realm of treatment by interventionists. The first reported transcatheter closure was performed by Slack *et al*² in a sick neonate, using Gianturco coils. Subsequently, six more patients underwent device closure successfully, using either an Amplatzer septal occluder,^{4 8} a muscular ventricular septal defect occluder⁶ or a duct occluder.^{3 5 7} In all the patients except one,⁸ operators choose to use a venovenous loop to deliver the device.

In our patient, a venovenous loop was not required, as the aneurysm distal to the tubular communication provided adequate space for parking the stiff wire, which gave excellent support for the delivery catheter. Although use of a vascular plug was reported previously in treatment of pulmonary AV malformations,¹¹ this is the first time it was used in a patient with direct RPA to LA communication. We believe, whenever the device is being taken from a pulmonary arterial (PA) aspect, an AVP is preferred over a duct occluder. The latter, with its larger distal disc, runs the risk of embolisation as the PA

pressures are higher than those of the LA. Moreover, the tubular shape of the communication is ideally suited for closure with vascular plugs.

The odd finding in our patient was the presence of severe LV dysfunction at presentation. The reason for this remained unclear to us, as the intrapulmonary shunt per se could not explain this finding. Pulmonary AV malformations classically have been shown not to cause any haemodynamic burden on the heart, as the increase in flow through the fistula causes an equivalent reduction in flow through the remaining portion of the lungs.^{12 13} However, in one interesting case, a very large congenital pulmonary AV fistula caused heart failure in a neonate, who recovered after surgical correction.¹⁴ Our literature search did not reveal any reported case of pulmonary AV fistula with LV volume overload or ventricular dysfunction in adults. We considered the possibility of a coexistent peripartum cardiomyopathy causing LV dysfunction, as the patient was 3 months post partum when she came to us. Other possibilities were cardiomyopathy caused by long-standing cyanosis, or sub-clinical myocarditis. Although she showed partial improvement in ventricular function after the procedure, this may also be attributed either to the natural recovery process of the peripartum cardiomyopathy or to the beneficial effects of heart failure medications initiated by us. Although we did not perform an endomyocardial biopsy, if carried out, this would have probably shed light onto the aetiology of the ventricular dysfunction.

To conclude, transcatheter closure of a RPA to LA fistula is a feasible and safe alternative to surgery in selected patients. The symmetrical shape of the vascular plug is more advantageous and safer whenever the procedure is performed without a conventional venovenous loop.

Learning points

- ▶ A direct pulmonary artery to left atrial fistula should be suspected in patients presenting with isolated cyanosis in the absence of abnormal cardiovascular findings.
- ▶ Bubble contrast echocardiogram and contrast CT angiogram are the most useful investigations for confirming the diagnosis and for delineation of the anatomy of the fistula.
- ▶ In selected patients with a suitable anatomy, transcatheter closure of the pulmonary artery to left atrial fistula is feasible and is a safer alternative to open surgery.

Contributors VK and MJV were involved in the patient care and follow-up of the patient, and coauthored the work. OKG performed the procedure and carried out critical supervision of the manuscript writing.

Competing interests None declared.

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Video 4 Bubble contrast echo after the procedure, confirming the complete closure of the defect.

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