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CASE REPORT
Structural intervention

Management of an asymptomatic pulmonary arteriovenous fistula diagnosed prenatally by detachable balloon embolization: a case report

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Background Pulmonary arteriovenous fistulas (PAVFs) are abnormal vascular malformations that connect pulmonary arteries and pulmonary veins, resulting in a right-to-left shunt. This anatomical shunt leads to a decrease in arterial oxygen saturation. Patients with PAVFs are usually treated with embolization or surgical procedures.

Case summary A 15-month-old child was diagnosed with PAVF *in utero* at 36 weeks' gestational age. The patient had no history of dyspnoea, repeated haemoptysis, or pneumonia, and there was no relevant family history. A chest computed tomography (CT) scan showed a bilobed opacity located in the inferior lobe of the right lung, and an echocardiogram detected a small atrial septal defect with positive contrast echocardiography. The patient underwent successful embolization using a detachable balloon catheter due to the relatively large diameter of the fistula. After the surgery, a significant decrease in flow was observed in the fistula, and the contrast is no longer visible.

Discussion Here, we report a case of a paediatric patient with congenital asymptomatic PAVF, diagnosed prenatally, and successfully treated with detachable balloon embolization. The case illustrates the detachable occlusion balloons appear to be an effective tool for embolization of PAVF in children, especially in cases involving large feeding vessels.

Keywords Pulmonary arteriovenous fistulas • Foetal echocardiogram • Contrast echocardiography • Embolization • Detachable occlusion balloons • Paediatric • Case report

ESC curriculum 2.2 Echocardiography • 7.5 Cardiac surgery

Learning points

- Pulmonary arteriovenous fistulas (PAVFs) can be diagnosed early by foetal echocardiogram.
- Patients with PAVFs are usually asymptomatic; however, the complications of PAVFs are severe. Treatment decisions should be made on a case-by-case basis for asymptomatic patients, especially for children.
- The detachable occlusion balloons seem to be an effective tool for embolotherapy of PAVF in children, particularly in cases with large feeding vessels.

Introduction

Pulmonary arteriovenous fistulas (PAVFs) are a rare condition that leads to hypoxaemia as deoxygenated blood bypasses the pulmonary arteries and enters the pulmonary veins. Pulmonary arteriovenous fistulas can be congenital or acquired, and patients may have a history of hereditary haemorrhagic telangiectasia (HHT) caused by endoglin (HHT1) and/or activin receptor-like kinase (ALK1) (HHT2) mutations.[1]

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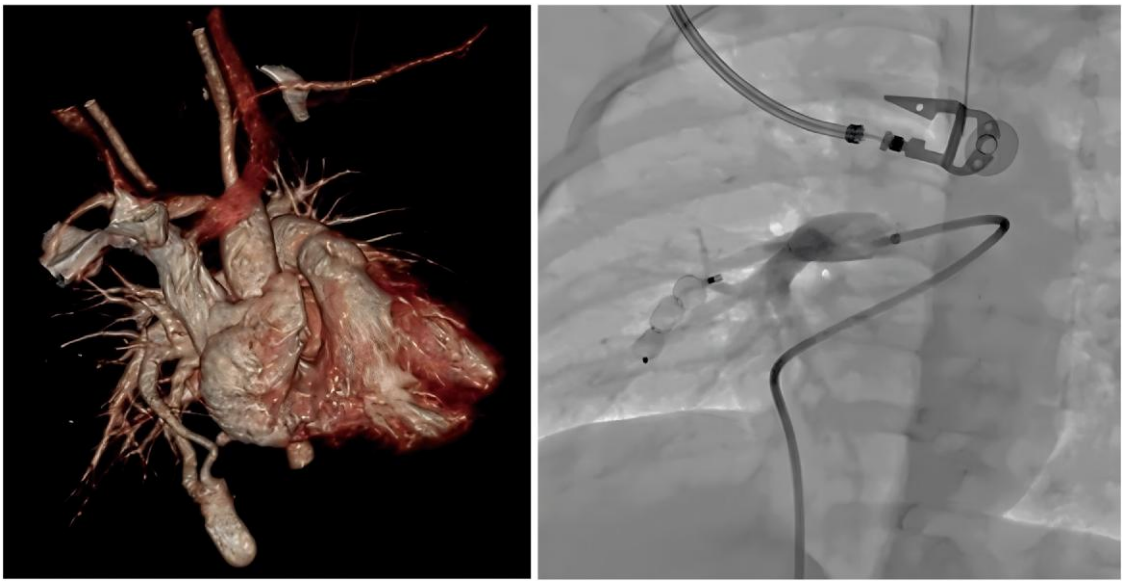
In the early stages, most individuals with PAVF do not exhibit obvious symptoms. However, patients may experience dyspnoea, recurrent haemoptysis, and even pulmonary or cerebrovascular embolism, which can be potentially life threatening.[1,2] Clinically, PAVFs are usually classified into focal PAVFs and diffuse PAVFs. Focal PAVFs are generally treated with embolization using coils, detachable occlusion balloons, or vascular plug.[3] However, embolization by coil or vascular plug has limitations, particularly in treating large paediatric PAVFs. High incidence of recanalization and air embolism have been reported with coil embolization,[4] and the vessels in paediatric patients are often too small to accommodate the vascular sheath required for large vascular plug device. Kaneko *et al.*[5] reported that embolization with detachable balloons is an effective procedure for treating bilateral multiple PAVF in adults. Here, we present the case of a paediatric patient with large focal PAVFs who was diagnosed prenatally and remained asymptomatic during the first year of follow-up after birth. The patient successfully recovered following embolization with a detachable balloon catheter.

Summary figure



Case presentation

A 15-month-old child presented to our cardiac centre for follow-up of PAVF, which had been diagnosed *in utero* at 36 weeks' gestational age (*Figure 1*). Following birth, the child's oxygen saturations were satisfactory, and the child was placed under regular follow-up care every 3 months. The child had no history of dyspnoea, recurrent haemoptysis, or pneumonia, and there was no relevant family history. A general physical examination revealed no significant findings, and arterial blood gas analysis showed no signs of hypoxaemia. Chest radiography revealed a bilobed opacity in the inferior lobe of the right lung (*Figure 2A*). Chest computed tomography (CT) and contrast-enhanced CT identified a 1.0 × 1.74 cm lesion in the right lower lung lobe (*Figure 2B*). Volume-rendered images from CT pulmonary angiography confirmed a PAVF connecting the right lower pulmonary arteries and right lower pulmonary veins (*Figure 2C* and *D*). Transthoracic echocardiography showed a small atrial septal defect with no other abnormalities. Additionally, contrast echocardiography demonstrated the presence of contrast in both the left atrium and right atrium within one cardiac cycle after injecting 10 mL of saline into a peripheral vein while simultaneously imaging both atria (*Figure 3A* and *B*; Supplementary material online, *Video S1*).

We further confirmed the diagnosis of PAVF through pulmonary artery catheterization and angiography, conducted via right transfemoral venous access. To prevent pericatheter thrombosis, heparin (100 U/kg) was administered. Selective right pulmonary angiography and superselective catheterization of the feeding artery, using a 5F multipurpose guiding catheter, revealed a large, high-flow fistula with rapid opacification of the draining vein (*Figure 4A*; Supplementary material online, *Video S3*). The feeding artery's diameter was measured at 5.3 mm. Given the relatively large size of the fistula and the patient's young age, we decided to proceed with embolization using an Amplatzer detachable balloon catheter (see Supplementary material online, *Videos S4–S6*). These balloons, made of soft, flexible materials, expand when inflated, allowing for controlled occlusion of the blood vessel. A stainless steel microscrew is welded to the platinum/iridium marker bands, enabling attachment to the stainless steel delivery cable. The delivery system allows the operator to confirm the proper position and stability of the device before detachment. After preloading the device and adequately flushing the loader to remove air, the balloons were advanced along the guiding catheter to the distal part of the feeding artery. The catheter was then retracted, allowing the balloons to expand and occlude the fistula. Following deployment, a marked reduction in flow was observed in the fistula (*Figure 4C* and *D*), and contrast was no longer visible in the left heart chambers on contrast echocardiography (*Figure 3C* and *D*; Supplementary material online, *Video S2*).

After the procedure, the patient was completely asymptomatic and was transferred to the general ward within a few days. Four weeks later, the patient underwent an outpatient follow-up. A contrast echocardiography revealed no contrast in the left atrium within 20 cardiac cycles after 10 mL of saline was injected into a peripheral vein.

Discussion

Pulmonary arteriovenous fistula is a rare anomaly, with an incidence of 2–3 per 100 000 individuals.[6] Most PAVFs are congenital, resulting from

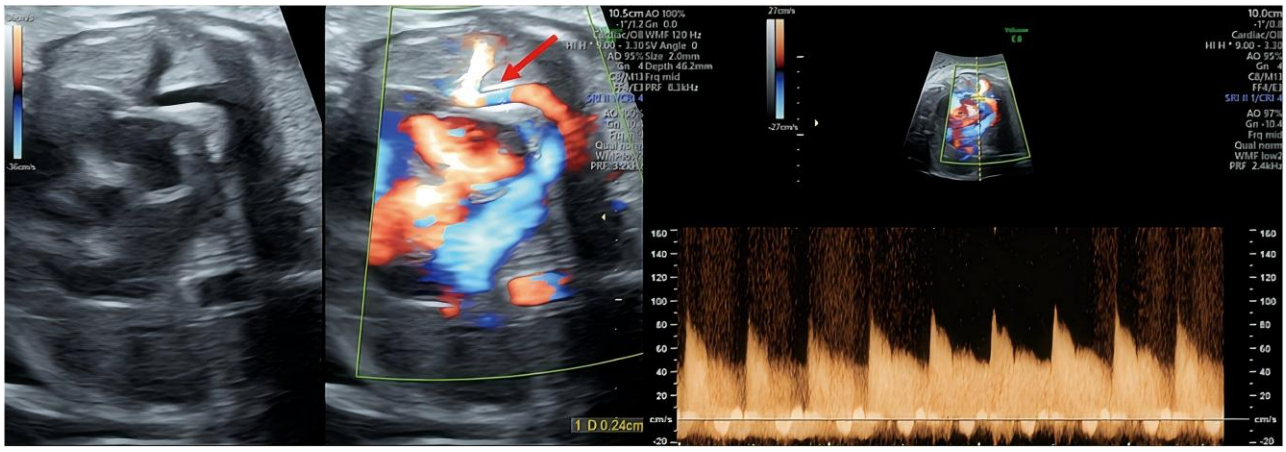


Figure 1 Foetal echocardiography showing high-velocity flow and distal constriction of pulmonary arteriovenous fistula (red arrows).

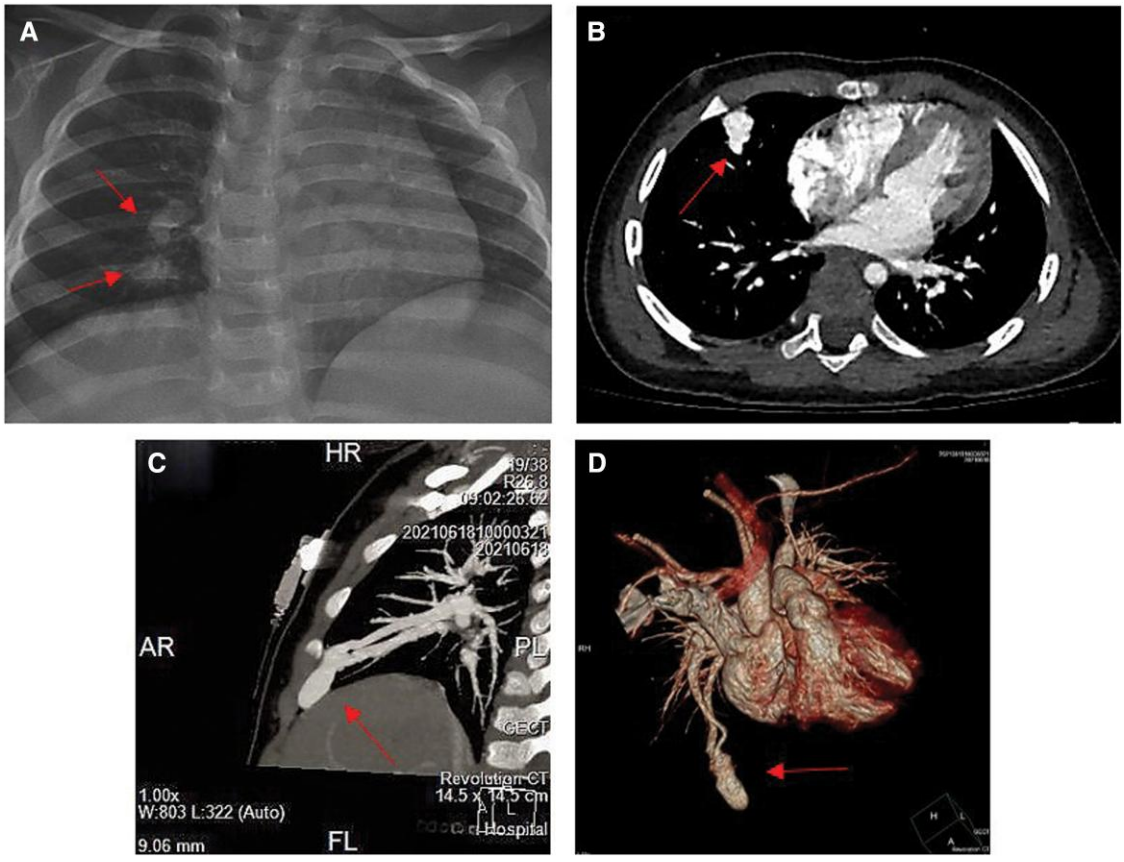


Figure 2 Chest X-ray and computed tomography before embolization. (*A*) Chest X-ray showing a bilobed opacity in the right lung. (*B*) Contrast-enhanced computed tomography depicting images of lesion. (*C*, *D*) A volume-rendered reformatting image of the computed tomography pulmonary angiography showing the pulmonary arteriovenous fistula with feeding vessels. Red arrows showing the pulmonary arteriovenous fistula.

the failure of differentiation of the embryonic vascular plexus.[7] Approximately 70% of cases are associated with HHT.[1] In this case, however, the patient did not meet any of the diagnostic criteria for HHT. Routine diagnosis of PAVF is challenging. Patients presenting with cyanosis, dyspnoea, and decreased oxygen saturation should be considered for this diagnosis. Prenatal diagnosis of PAVFs is rarely described in the literature.[8] What is particularly noteworthy in this case is that the diagnosis was made through foetal echocardiography during

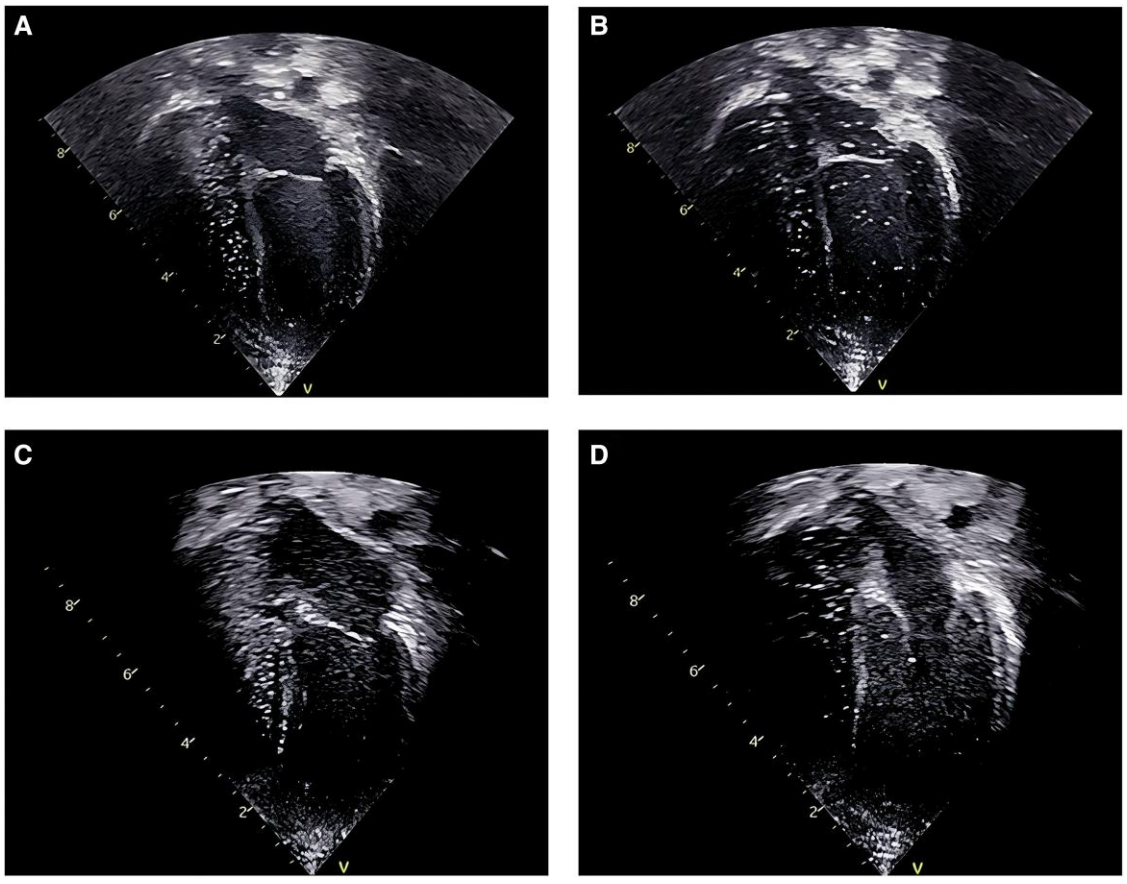


Figure 3 Contrast echocardiography of the patient. (*A, B*) Echocardiography of the patient before embolization with the injection of 10 ml of saline into a peripheral vein; (*C, D*), contrast echocardiography of the patient after embolization.

prenatal care and later confirmed by contrast-enhanced CT, contrast echocardiography, and selective pulmonary angiography after birth. In general, a prominent pulmonary artery and vein, along with Doppler echocardiographic visualization of the origin of pulmonary venous flow, can aid in accurate prenatal diagnosis. Approximately 98% of PAVF patients exhibit abnormalities on chest radiographs. The typical radiographic appearance of PAVFs is a mass of uniform density, often lobulated but sharply defined, typically located in the lower lobes.[9] Additionally, contrast-enhanced CT and contrast echocardiography are valuable diagnostic tools for evaluating the vascular anatomy of PAVFs.[1]

Another interesting aspect of this case is that the child remained asymptomatic, despite the presence of a significant right-to-left shunt due to the PAVF. Asymptomatic patients are not uncommon in such cases; Swanson *et al.*[10] from the Mayo Clinic reported a morbidity rate of 26%–33% and a mortality rate of 8%–16% in untreated patients with asymptomatic PAVFs. Common complications of PAVFs include cerebral and systemic emboli, cerebral abscesses, fatal rupture of the fistula, and heart failure. The incidence of central nervous system (CNS) complications associated with PAVFs includes migraines, transient ischaemic attacks, strokes, abscesses, and seizures.[11,12] Compared to surgery, endovascular embolization is the preferred treatment due to its lower complication rate and shorter hospital stays. International guidelines for the management of pulmonary PAVFs in patients with HHT recommend that treatment decisions for asymptomatic children be made on a case-by-case basis. The selection of PAVFs for embolization typically depends on the diameter of the feeding artery, with a threshold of 3 mm or greater, although an artery as small as 2 mm may also be considered appropriate for embolization.[13]

In this case, the diameter of the PAVF increased from 2.4 to 5.3 mm during the first year after birth. Given the progressive enlargement of the PAVF, interventional therapy was deemed necessary. Treatment of significant PAVFs typically involves embolization using coils, detachable occlusion balloons, or vascular plugs. These methods occlude the feeding arteries with minimal invasion. In 1977, Porstmann first reported the use of handmade steel coils for PAVF embolization.[14] Coil embolization is generally recommended for PAVFs with diameters <5 mm. The procedure involves localizing the PAVF through angiography, selectively catheterizing the feeding artery, and deploying the coils, which should assume their natural shape and adhere to the vessel wall. However, complications such as coil migration and residual shunt can occur, especially if an oversized coil is used. Vascular plugs may be used for larger PAVFs, but their use is limited in paediatric patients, particularly infants, due to the small size of their blood vessels, which cannot accommodate the vascular sheath required for the plugs. Another option for embolization is detachable occlusion balloons. In this method, a balloon catheter is inserted and positioned at the neck of the PAVF after localization by angiography. The balloon is then inflated with radiopaque contrast material, and angiography is repeated to confirm vessel occlusion. The balloon can be detached once occlusion is verified, and its ability to be deflated and repositioned if necessary is an added advantage.[1] In this case, the

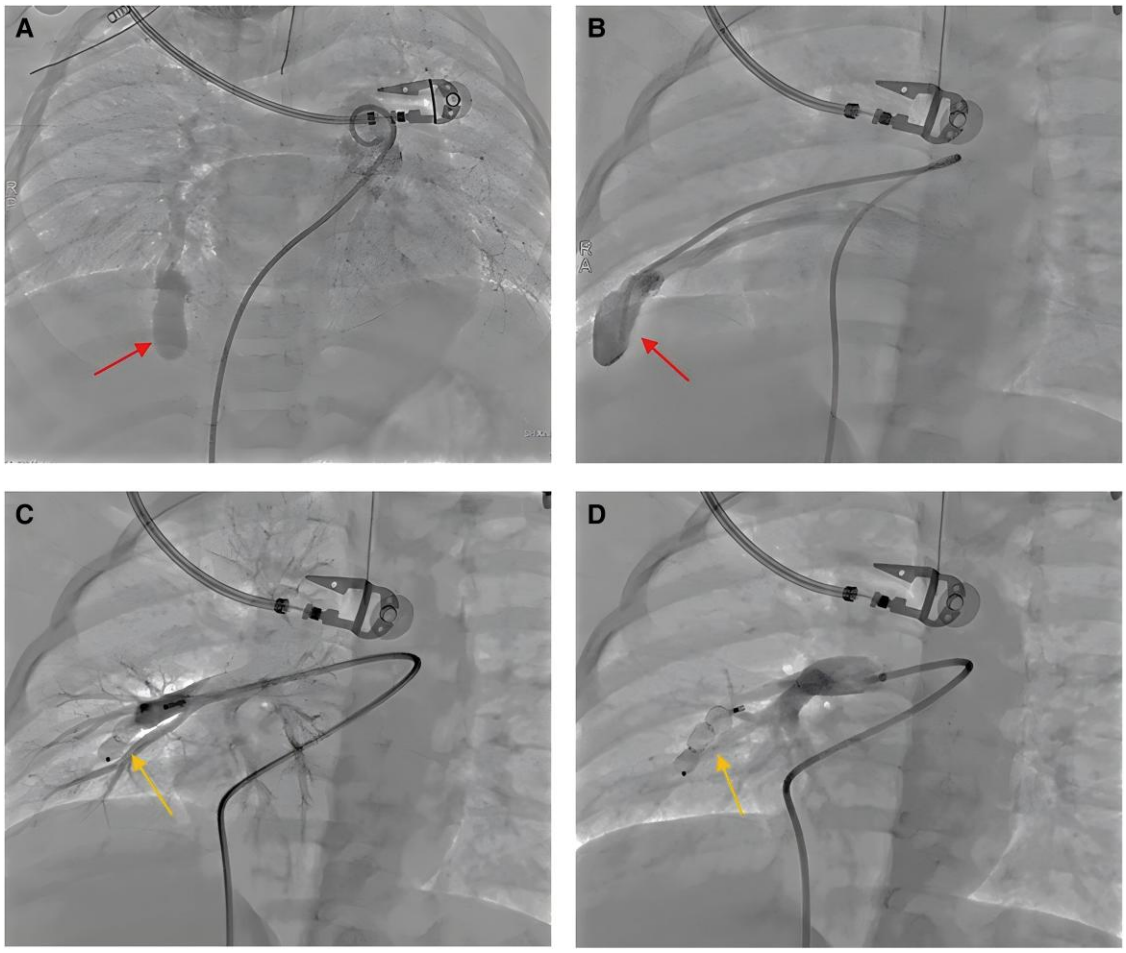


Figure 4 Cardiac catheterization of the patient. (*A*) Pulmonary catheterization. (*B*) Superselective catheterization of the fistula-feeding artery shows the location of the pulmonary arteriovenous fistula (red arrows). (*C*, *D*) Left anterior oblique view demonstrating the correct positioning of the detachable balloon (yellow arrows). A test injection reveals complete thrombosis of the feeding artery following superselective catheterization of the fistula-feeding artery and selective right pulmonary catheterization.

feeding arteries of the PAVF were successfully occluded using detachable occlusion balloons.

In summary, detachable occlusion balloons seem to be an effective tool for embolization of PAVFs in children, particularly in cases involving large feeding vessels. However, information on the use of detachable occlusion balloons in children and long-term results should be provided by future investigations.

Lead author biography

Sun Chen is the chief of Pediatric Cardiology Centre at Xinhua Hospital, Affiliated to Shanghai Jiao Tong University School of Medicine in Shanghai, China. His main clinical interest is congenital heart disease. He has an active research interest in paediatric cardiology.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

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Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.

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