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Brief Report

Neonatal transcatheter closure of a large pulmonary arteriovenous fistula

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Abstract A congenital fistulous connection between the right pulmonary artery and the left atrium is a rare condition, resulting in early cyanosis and cardiac failure. These patients usually required urgent surgery. We present a neonate in whom we successfully closed such a large fistula via catheterization with an Amplatzer Duct Occluder, resulting in rapid clinical improvement, and obviating the need for surgical repair.

Keywords: Cardiac catheterization; congenital heart disease; intervention

CONGENITAL ARTERIOVENOUS FISTULAS ARE rare entities that are often associated with generalized disease, as in hereditary haemorrhagic telangiectasis, also known as the Rendu-Osler-Weber syndrome. The diagnosis is rarely made during infancy, and even more exceptionally in neonates. We present a neonate diagnosed with a large pulmonary arteriovenous fistula. This rare symptomatic lesion is usually treated surgically, with relatively small numbers reported thus far, with most undergoing surgical procedures.[1–4] We achieved successful transcatheter closure.

Case report

A newborn, delivered naturally at 37 weeks' gestation was admitted to our unit specializing in paediatric cardiology because of antenatal diagnosis of a pulmonary arteriovenous fistula. Cardiovascular examination revealed profound desaturation, 78% at rest, and a systolo-diastolic heart murmur. The initial echocardiographic examination revealed a large pulmonary arteriovenous fistula responsible for significant dilation of the left ventricle, which had a diastolic diameter of 27 millimetres, and exhibited systolic dysfunction. Cardiac catheterisation was performed under local anaesthesia. A 5 French introducer sheath was placed in the right femoral vein. Intravenous heparin, at 50 units per kilogram, was given after cannulation. Selective pulmonary angiograms confirmed the diagnosis, showing a direct communication from the right pulmonary artery to the left atrium. The left pulmonary arteriogram was normal. The injection in the right pulmonary artery showed the fistulous vessel arising from the posterior and inferior surface prior to the hilum, connecting to an aneurismal pouch, and entering the left atrium (Fig. 1). We advanced a right coronary catheter into the right pulmonary artery, and through the fistula into the left atrium. A stiff wire was then advanced inside the right coronary catheter, followed by a 6 French delivery sheath, which was placed over the wire in the left atrium. An 8/6 millimetres ductal occluder (Amplatzer, Minneapolis, United States of America) was loaded and delivered through the previously placed delivery sheath. Advancement and delivery of the device were easily performed. The time required for the procedure was 25 minutes. An angiogram subsequent to release of the occluder revealed the complete closure of the fistula, with disappearance of the aneurysmal pouch (Fig. 1). To avoid dislodgement of the device, and because angiograms in multiple projections did not reveal any pulmonary arterial obstruction, we avoided measurements of pressure to confirm the absence of pulmonary obstruction.

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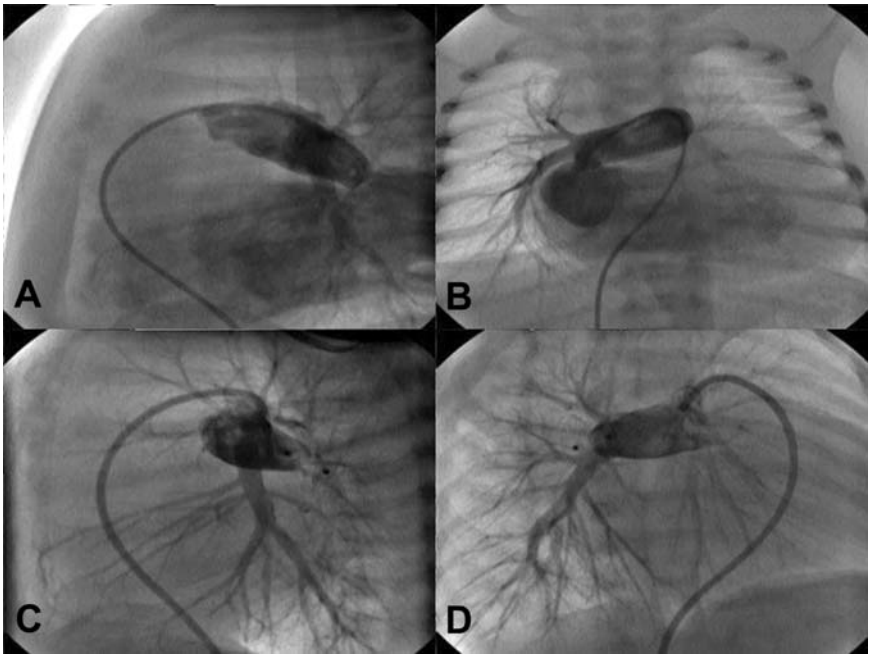

Figure 1.
Angiographic views before and after transcatheter closure. A and B: The fistulous vessel is arising from the postero-inferior side before the hilum by a neck connected to an aneurysmal pouch which connects to the left atrium (A: lateral, B: en face view). C and D: After transcatheter closure, the fistula is completely closed confirmed by the disappearance of the aneurysmal pouch and the absence of early left atrial opacification (C: lateral, B: 15 degree left anterior oblique). Note the absence of obstruction of the pulmonary arterial branches.

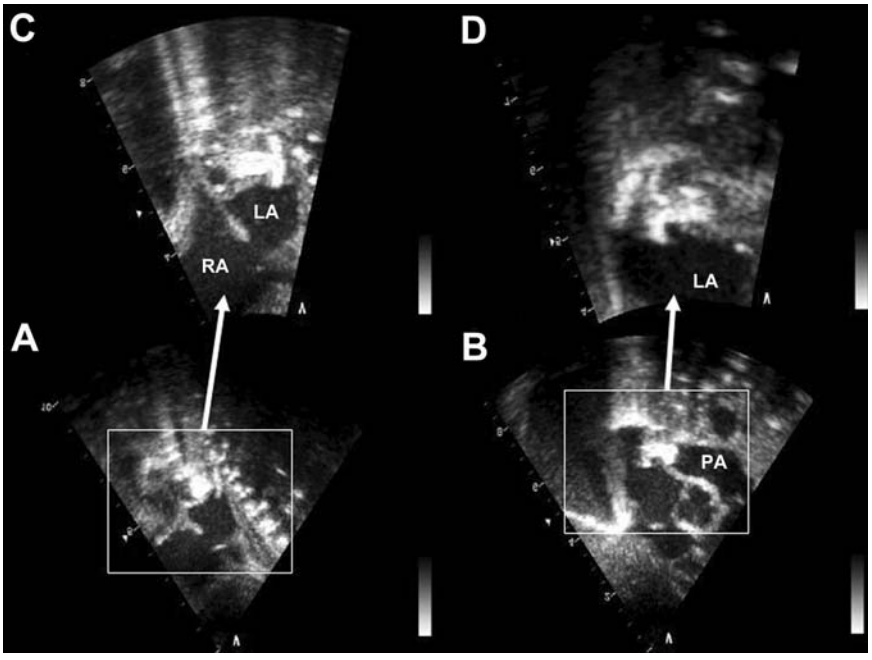

Figure 2.
Post-procedural echocardiographic views showing the device inside the fistula. Abbreviations: RA: right atrium; LA: left atrium; PA: pulmonary artery.

Concomitantly, we obtained evidence showing normalization of saturations. The post-procedural echocardiogram confirmed closure of the fistula, without any residual shunt, and showed reduction in left ventricular dilation, the diameter now measured at 23 millimetres (Fig. 2). The newborn was discharged home two days after the procedure. Six months after the procedure, he is in perfect condition, with normal echocardiographic findings.

Discussion

Acquired or congenital pulmonary arteriovenous fistulas are a rare but recognized cause of cyanosis. Proximal lesions are even more uncommon.[1–4] The majority of these fistulas arise from the right pulmonary artery. Most commonly, the opening of the fistula is in the posterior side of the artery, albeit that four anatomic subtypes have been described by Nelson

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et al.[5] The most common subtypes are a direct tubular connection to the left atrium, or a tubular vessel connecting to the right pulmonary vein, with an aneurysmal dilation of the vessel prior to its entry into the left atrium. They are thought to be either secondary to incomplete degeneration of the vascular septums between the arterial and venous plexuses at the level of the pulmonary bud,[6] or to a defect in the terminal capillary loops allowing dilation and formation of thin walled vascular sacs fed by a single artery, and drained by a single vein.[7] The timing of the onset of symptoms is primarily dependent on the percentage of blood that passes through the fistula. Extremely rarely, the fistula is large enough to cause profound cyanosis, tachypnea, and respiratory distress in the neonatal period. Electrocardiographic findings are left ventricular hypertrophy and left atrial enlargement. Echocardiography with color flow Doppler is usually diagnostic, showing dilated left cavities, and revealing the aneurysmal connection between the pulmonary artery and the left atrium or the pulmonary vein if present. Cardiac catheterization, when performed, confirms the diagnosis, showing the right-to-left shunt at atrial level, and demonstrating the dilated fistula vessel draining into the left atrium. A narrowing may be present along the course of the abnormal connection. The decision to intervene is usually made according to the amount of right-to-left shunt and the presence of cardiac failure. Surgical treatment has been the method of choice for most of reported cases, and was associated with significant morbidity, and mortality as high as one in five.[8–9] Transcatheter closure is an attractive alternative to surgery in symptomatic neonates. Various devices can be used depending on the size of the vessel and the presence of a narrowing. In our patient, we successfully inserted an Amplatzer Duct Occluder. Complete occlusion was obtained rapidly, with normalization of saturations revealed in the catheterization laboratory. Respiratory and cardiovascular symptoms rapidly disappeared and the patient was discharge home two days after the procedure. The transcatheter technique, therefore, can be a good option for neonates with haemodynamically significant direct connections between the pulmonary artery and the left atrium, and should be considered as an attractive alternative to surgery.

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