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Brief Report

Spontaneous closure of right pulmonary artery-to-left atrium fistula: a case report

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Abstract Direct communication between the right pulmonary artery and the left atrium is rare. We report a case that was first detected in utero and confirmed by a two-dimensional echocardiographic study performed postnatally. The patient remained asymptomatic, and hence was managed conservatively. The fistula gradually decreased in size and closed spontaneously at 15 months of age. The rarity of the case is discussed.

Keywords: Right pulmonary artery; left atrial fistula; spontaneous closure

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Direct communication between the right pulmonary artery and the left atrium is a rare congenital anomaly. Such patients usually present with exertional dyspnoea, cyanosis, and clubbing or uncommonly present with complications of cerebral or systemic embolism. In the past, these patients underwent surgical closure of the fistula. More recently, the communication has been successfully closed using transcatheter techniques.

We present a case of a child who was detected to have a right pulmonary artery–left atrium fistula in utero and who subsequently had spontaneous closure of the fistula at 15 months of age. To the best of our knowledge, spontaneous closure of a right pulmonary artery–left atrium fistula has not been reported in the literature

Case report

The child was detected in utero to have a right pulmonary artery–left atrium fistula during fetal scan. At birth, he had persistent desaturation (SaO_2 88% on room air), and was therefore referred to confirm the diagnosis. Clinically, his vital parameters were normal. He had no cyanosis or clubbing and there was no audible murmur. His chest X-ray showed normal heart size and normal lung vascularity. The two-dimensional echocardiography and colour Doppler studies revealed normal segmental subset, small patent foramen ovale with a left-to-right shunt, intact interventricular septum, small closing ductus arteriosus, normally functioning valves, normal biventricular contractility, and a normal aortic arch. In addition, there was a small, low-pressure shunt seen entering the left atrium from the right pulmonary artery. The shunt at the site of its opening in the left atrium measured 3.5 mm and was significantly restricted (Fig 1a, video 1A).

As the child had no symptoms and the shunt was small and restrictive, conservative management was advised, and he was asked to follow-up at 3 months of age. At that time, the child remained asymptomatic with normal breathing, adequate feeding, and good weight gain. Repeat two-dimensional and colour Doppler studies showed spontaneous reduction in the size of the fistula, which measured 1.1 mm (Fig 1b, video 1B).

No intervention was thought necessary at this juncture, and the child was re-evaluated at 15 months of age. The child remained healthy with normal clinical examination. Repeat two-dimensional colour Doppler study showed complete spontaneous closure of the fistula (Fig 1c, video 1C).

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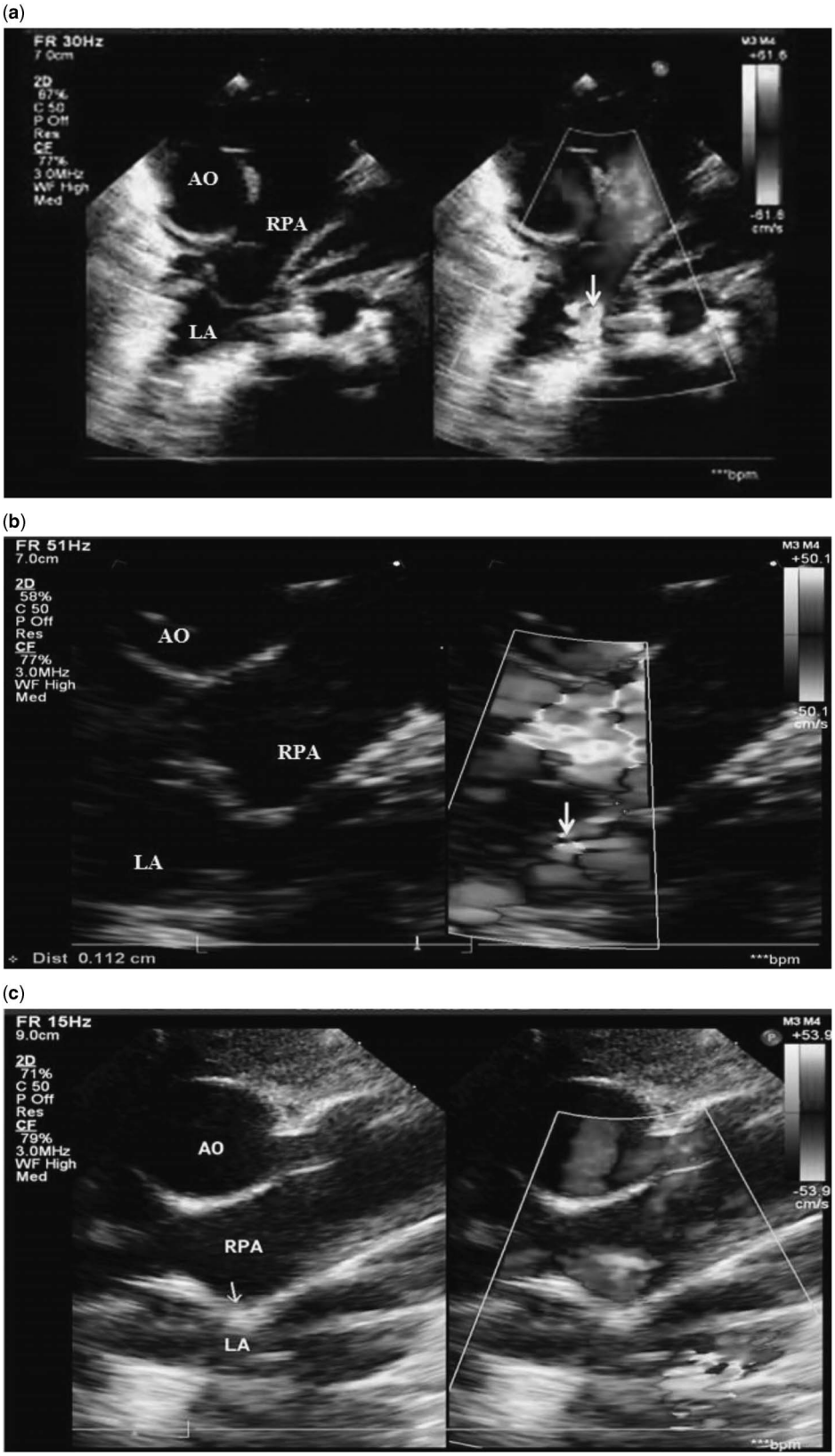

Figure 1.
*(**a**) Parasternal short-axis view at the level of the aorta (AO) showing a small, restrictive right pulmonary artery (RPA)-to-left atrium (LA) fistula (arrow) at 7 days of life. (**b**) Significant reduction in the size of the fistula (arrow) is seen at 3 months. (**c**) Shows complete closure of the fistula (arrow) at 18 months.*

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Discussion

Right pulmonary artery-to-left atrium fistula is a rare congenital anomaly initially reported by Friedlich et al[1] and surgically corrected for the first time by Blalock in 1950. Subsequently, more than 100 cases have been reported, managed either surgically[2] or by transcatheter techniques using coils or one of the devices from the Amplatzer family.[3] To the best of our knowledge, this is the first report of spontaneous closure of such a fistula.

This anomaly can present at different ages depending on the size of the fistula, which in turn determines the magnitude of the shunt. Those presenting late usually have good prognosis compared with those who present in infancy because they have a much smaller shunt.[4] The clinical presentation is due to right-to-left shunt and is characterised by shortness of breath, easy fatigability, cyanosis, and clubbing.[1] In the case of our patient, the shunt was minimal, and hence he remained completely asymptomatic. As the pulmonary artery directly communicates with the left atrium, some patients may present with paradoxical embolism or with cerebral abscesses.[5] Therefore, this anomaly should be considered in children who present with unexplained cerebral or systemic embolisation.

Right pulmonary artery-to-left atrium communication can mimic a pulmonary arteriovenous fistula in terms of its clinical presentation as both present with extracardiac right-to-left shunt in the presence of normal right ventricular pressure; two-dimensional echocardiography with colour Doppler and CT angiography can help differentiate the two.

On the basis of the presence or absence of the aneurysm and the drainage of the pulmonary veins, the right pulmonary artery-to-left atrium fistula has been classified angiographically into four types.[5] Type 1 is characterised by a normally branching right pulmonary artery with normal pulmonary venous return and a fistulous channel connecting the right pulmonary artery to the left atrium. In Type 2, the lower lobe branch of the right pulmonary artery communicates with an aneurysmally dilated right inferior pulmonary vein. In Type 3, all pulmonary veins drain into the aneurysmal pouch between the right pulmonary artery and the left atrium, whereas Type 4 has a very proximal right pulmonary arteriovenous fistula, which joins the left atrium via the right pulmonary veins. On the basis of this classification, our patient demonstrated a Type 1 fistula.

The diagnosis of right pulmonary artery–left atrium fistula is usually established using two-dimensional echocardiography and colour Doppler as in our patient. Contrast echocardiography is helpful in confirming the diagnosis in adults where colour flow imaging is suboptimal. A CT pulmonary angiogram is considered as the gold standard for establishing the diagnosis.[6] In our patient, the diagnosis was unequivocally confirmed in the neonatal period on two-dimensional echocardiography, the shunt was small, and no intervention was contemplated. Hence, we did not feel it was necessary to subject the child to CT pulmonary angiography as it involves exposure to radiation and injection of radiographic contrast.

Early repair is recommended in patients with right pulmonary artery–left atrium fistula to avoid complications, particularly systemic embolism, infective endocarditis, cerebral abscess, and rupture of aneurysmal communications. Surgical ligation of the fistula was the preferred treatment for this condition until recently. Of late, transcatheter coil occlusion and device closure have been performed successfully. As our patient had no symptoms and a small right-to-left shunt, it was decided not to intervene early. Fortunately, the fistula closed spontaneously and the child was saved from any form of intervention.

Although the exact mechanism of spontaneous closure remains unknown, it is likely that a fall in pulmonary vascular resistance in the postnatal period resulted in a significant drop in the pressure head with reduced flow through the restricted fistulous orifice. The flow eventually fell to a critical level, allowing the endothelium to grow and bridge the gap. The second mechanism that can be contemplated is that the duct-like tissue forming the fistulous tract constricted because of increased oxygenation in the postnatal period after the lungs became operational.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that all procedures mentioned in this case report, which did not involve human experimentation, are in accordance with the ethical standards of the Indian guidelines on human treatment.

Supplementary materials

For supplementary material/s referred to in this article, please visit http://dx.doi.org/10.1017/S1047951116000615

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