

Transcatheter closure of a Right Pulmonary Artery to Left Atrial Fistula using the Amplatzer Muscular VSD Occluder Device

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A fistula formation between the right pulmonary artery (RPA) and left atrium (LA) is a very rare cyanotic congenital heart disease and the traditional treatment involves surgery. We presented the successful closure of the RPA to LA fistula using the Amplatzer Muscular VSD occluder in a child with cyanosis.

Case report

A 4-year-old girl was admitted with cyanosis and heart murmur. She had severe cyanosis, digital clubbing and effort intolerance (NYHA Class II-III). Cardiac MRI and MR Angiography showed a congenital fistula between the RPA and LA. Diagnostic cardiac catheterization was performed. The RPA showed dilatation and the inferior branch of the RPA, along with the right pulmonary veins via a large fistula, was draining to an aneurysmatic sac and then to the left atrium. A guide wire was advanced into the sac passing through the fistula via the RPA and the small ASD antegradely. An arteriovenous loop was created. The narrowest part of the fistula was 11 mm in balloon sizing. A 12-mm Amplatzer Muscular VSD occluder device was deployed at the narrowest site and the fistula was closed successfully (Figure 1) and control angiography demonstrated complete occlusion of the fistula. After the procedure, arterial oxygen saturation in room air increased from 63% to 97%. The patient was asymptomatic on follow-up evaluations at 3, 6, 12, 18 and 30 months with a mean oxygen saturation of 97%.

Discussion

A congenital RPA to LA fistula is a rare congenital heart disease that is typically associated with a normal cardiovascular system examination except for central cyanosis. There have been only a few cases reported in the literature where fistula closure using the Amplatzer Muscular VSD occluder device. In select cases, transcatheter closure appears to be a safe and effective alternative to surgical treatment.

Figure.1- A. Balloon sizing of the fistula, B. Pulmonary arteriogram in the posteroanterior projection shows complete occlusion of the fistula by the Amplatzer Muscular VSD occluder device

