A man aged 57 years, with a strong holosystolic murmur throughout the precordium and a concomitant thrill, was admitted to the cardiology department for progressively deteriorating dyspnoea.

The two-dimensional echocardiogram, in the subxiphoid position, long axis view, showed a perimembranous defect in the form of a break in the interventricular septum, below the aorta and posterior to the septal tricuspid valve leaflet (Figure 1). Colour Doppler is the method of choice for demonstrating aplastic sections of the interventricular septum. The apical four-chamber view (Figure 2) and the subxiphoid long axis view (Figure 3) showed a ventricular septal defect, depicting the turbulent flow as a mosaic of colours passing through the perimembranous shunt from the left ventricle to the right during systole.

The patient had previously undergone magnetic resonance angiography of the thoracic aorta, in which a three-dimensional phase contrast series had shown a variation in the arterial origins from the aortic arch, with a common origin of the left common carotid and the innominate artery (Figure 4).

Ventricular septal defect is the most common congenital heart disease, comprising 20% of all congenital heart diseases. The defects are classified as subcristal (perimembranous and muscular) and supracristal (above the supraventricular crista). Perimembranous defects are those most often seen on echocardiography (90%). Ventricular septal defect occurs either as an isolated congenital anomaly or in combination with other cardiac anomalies, such as pulmonary artery stenosis, transposition of the great arteries, pulmonary artery atresia, and common atioventricular shunt. There is no report in the available literature of the coexistence of a large ventricular septal defect with this specific anatomical variation of the aortic arch.

References

Figure 1. Two-dimensional echocardiogram, subxiphoid position, long axis view, showing a perimembranous defect (D), in the form of a break in the interventricular septum below the aorta. LV – left ventricle; AV – aortic valve; RV – right ventricle.

Figure 2. Colour Doppler, apical four-chamber view, showing blood flow (arrows) from the left ventricle (LV) to the right ventricle (RV) via the perimembranous shunt. la – left atrium; ra – right atrium.

Figure 3. Colour Doppler, subxiphoid position, long-axis view, showing turbulent blood flow (arrows) as a mosaic of colours passing through the defect from the left ventricle (LV) to the right ventricle (RV). AO – aorta; IVS – interventricular septum.

Figure 4. Magnetic resonance angiography showing the common origin (arrow) of the left common carotid and innominate arteries.