Congenital Diverticulum of the Right Ventricle Associated with Coarctation of Aorta, Atrial and Ventricular Septal Defect and Ductus

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Congenital right ventricular muscular diverticula are extremely rare and are usually associated with other congenital cardiac anomalies, (in half of the cases tetralogy of Fallot). They functionally behave like an accessory ventricular chamber which contracts synchronously with the normal ventricles.

Less than 30 patients with a right ventricular diverticulum have been reported in literature. An apical right ventricular diverticulum occurs in patients with thoracolumbar defects or abnormalities of the cardiac position[1]. However, an antero-superior diverticulum is usually associated with other congenital cardiac defects, such as a ventricular septal defect, tetralogy of Fallot, double outlet right ventricle and pulmonary stenosis[2–9].

We report an 11-year-old boy with an antero-superior diverticulum of the right ventricle associated with a coarctation of aorta, ductus arteriosus, and atrial and ventricular septum defects. To the best of our knowledge, such an association has not been reported before.

Case Report

An 11-year-old boy underwent cardiac evaluation because of cyanosis. He had been asymptomatic until age 9, when he developed progressive exertional dyspnoea.

A grade III/VI systolic and diastolic murmur was heard along the left sternal border. The chest X-ray film showed cardiomegaly with slightly increased pulmonary vascularity. Electrocardiography showed right bundle branch block and biventricular enlargement.

A transthoracic two-dimensional echocardiogram revealed a large atrial and ventricular septal defect (Fig. 1) a coarctation of the aorta with a 64 mmHg pressure drop (Fig. 2), a patent ductus arteriosus and a contractile and trabeculated diverticulum arising from the base of the right ventricle, lateral to the tricuspid valve annulus (Figs 1 and 3). The shunting blood flow across the inter-ventricular septum was bi-directional and laminar, which means right and left pressures were similar.

Haemodynamic evaluation confirmed the echo/Doppler findings, the pulmonary and systemic pressures being at the same level. The patient is currently waiting for a cardiac and pulmonary transplant.

Discussion

A true congenital diverticulum is generally regarded as an accessory chamber. Its wall consists of normal myocardium that contracts synchronously with the ventricle and originates either from the apex or from the antero-superior wall. The narrow-based, finger-like apical diverticulum is usually associated with umbilical midline defects, defects of the lower sternum, absence of the anterior diaphragm, partial absence of the pericardium, and intracardiac defects[10]. The antero-superior right ventricular diverticulum is usually broad-based, and its mural morphology and pattern of trabeculation corresponds with that of the right ventricle[2,6,8]. In our patient, the diverticulum originated from the base of the right ventricle.

The clinical presentation in patients with an antero-superior diverticulum is commonly related to the associ-
ated defects. Only a few reported cases presented with a cardiac rhythm disorder\[9,11\] or with chest pain\[9-12\]. The electrocardiogram often shows an intraventricular conduction delay, which is more likely to occur peripherally than in the main right bundle.

Echocardiography can help to diagnose a left ventricular diverticulum, but is often of limited value for the diagnosis of diverticula of the right side\[13\]. In the present case, however, the diagnosis of a right ventricular diverticulum was made by echocardiography.

Figure 1. Apical four-chamber image demonstrating a large ventricular septal defect in the muscular portion, a large atrial septal defect and a diverticulum arising from the base of the right ventricle lateral to the tricuspid annulus (abbreviations: RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium; D, diverticulum).

Figure 2. Long axis plane through the aorta from the suprasternal position. Note the localized luminal reduction of the descending aorta.

Figure 3. Apical four-chamber view demonstrating a colour blood flow jet from the diverticulum during left ventricular systole.

References