Congenital Right Ventricular Diverticulum Associated With a Ventricular Septal Defect: A Rare Echocardiographic Finding

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A neonate presented on the first day of life with tachypnea and poor feeding. The infant's initial echocardiogram demonstrated outpouching of the lateral wall of the right ventricle (RV) associated with a large ventricular septal defect (VSD). At 9 days of age he was diagnosed with osteogenesis imperfecta (OI). Despite treatment with digoxin, diuretics, and captopril he required hospitalization twice during his first 2 months of life for congestive heart failure (CHF). The VSD was closed at three and one-half months of age without resection of the diverticulum and CHF symptoms resolved. At 26 months of age he is doing well despite the residual RV diverticulum. Congenital cardiac diverticula are rare forms of cardiac malformations and their echo-Doppler features are herein discussed. (ECHOCARDIOGRAPHY, Volume 23, October 2006)

Congenital cardiac ventricular aneurysms or diverticula are rare. Fewer than 200 cases have been reported and only 18 of these involved the right ventricle.[1–3] Presentation varies from an incidental finding on chest x-ray to overt heart failure or sudden death from rupture. Herein, we report a case of a right ventricular diverticulum associated with a ventricular septal defect and osteogenesis imperfecta.

Case Report

A Caucasian male presented a few hours after birth with tachypnea and a cardiac murmur. The baby was 2.73 kg product of a 39 weeks uncomplicated gestation to a 23-year-old gravida 1 healthy mother. Evaluation within a few hours of birth revealed a heart rate of 160 beats/min, blood pressure of 68/53 (right arm) and 73/46 (left leg), and respiratory rate of 55. He was acyanotic and lungs were clear with symmetric chest excursion. Cardiac exam revealed diffuse precordial overactivity with normal S1, III/VI harsh systolic murmur at the mid left sternal border, single S2, ejection click and a III/VI diastolic rumble at the left lower sternal border. Femoral pulses were normal.

Electrocardiogram showed sinus rhythm with right axis deviation (axis +230) and complete right bundle branch block. Chest radiograph demonstrated cardiomegaly with a prominent right heart shadow and normal pulmonary vascularity. Echocardiogram revealed a large perimembranous ventricular septal defect, patent foramen ovale, patent ductus arteriosus, and an outpouching of the lateral wall of the right heart ventricle (Fig. 1). At 26 days of age the patient underwent cardiac catheterization. The right ventricular systolic pressure was equal to the aortic systolic pressure and the pulmonary to systemic flow ratio was 2:1. Angiography showed a large RV diverticulum that expanded with diastole and contracted with systole (Fig. 2).

He was treated with digoxin, furosemide, captopril, nasogastric feeding, and fluid restriction and was discharged from the hospital at 6 weeks of age. At 3.5 months of age he underwent surgical closure of the VSD. The RV diverticulum was not resected, since its contractile pattern was synchronous with body of the right ventricle and it contributed to a significant portion of the right ventricular stroke volume. The surgical course was uncomplicated and the patient was weaned from all medications by 11 months of age. Postoperative echocardiogram showed

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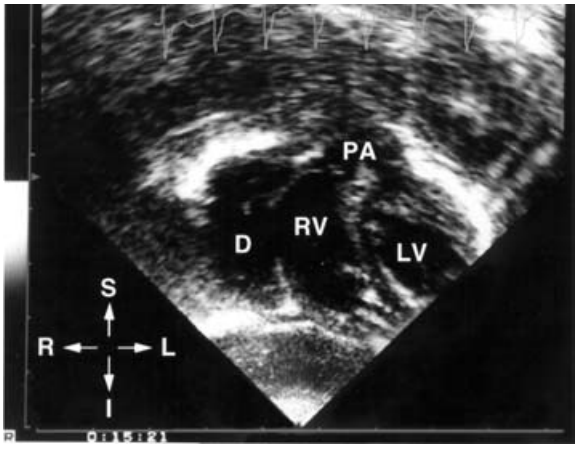

Figure 1. *Transverse subxyphoid echocardiographic diastolic image of the right ventricle (RV). The large anterolateral echo-free space in continuity with RV cavity is the diverticulum (D). Displayed are the right ventricle, right ventricular outflow tract, pulmonary artery (PA), and left ventricle (LV). Compass: I = inferior; L = left; R = right; S = superior.*

no residual ventricular shunt flow, normal contractility, and no valvular dysfunction. The RV diverticulum was unchanged. The patient has grown adequately and at the age of 36 months was asymptomatic from a cardiovascular standpoint.

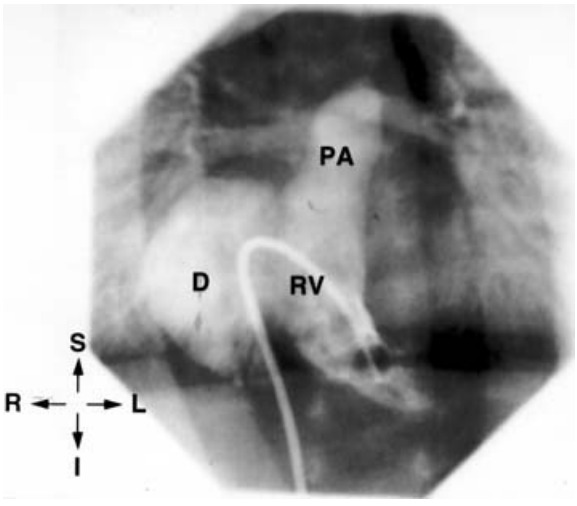

Figure 2. *Right ventricular (RV) cineangiocardiogram. Diastolic anterior-posterior projection corresponding to the echocardiographic image. The diverticulum (D) is large and fills with contrast. Contrast is also seen in the main pulmonary artery (PA) and its branches.*

Discussion

Pathogenesis

There has been much debate about the nomenclature of ventricular outpouching when diagnosed in infancy. A muscular defect with a narrow connection to the ventricle and associated mid-line defects as described by Cantrell et al.[4] have been termed a "diverticulum" while a fibrous, wide-based outpouching which resembles a ventricular aneurysm in adults has been termed "aneurysm." A recent report of 18 children distinguished aneurysms as akinetic or dyskinetic outpouching while diverticula had a normal or near-normal contractile pattern. In addition, aneurysms are usually isolated cardiac defects that often consist of only a fibrous layer with severe dyskinesis. Such aneurysms might be acquired by intrauterine myocardial infarction or myocarditis. The earliest diagnosis of a congenital aneurysm was made by fetal ultrasound at 24 weeks gestation, with subsequent intrauterine demise at 31 weeks.[5] Until more is known about the etiology of diverticula and aneurysms it seems reasonable to use the definition of Hamaoka et al.,[6] based on contractile pattern. Moreover, these authors classified left ventricular aneurysms and diverticula as annular subvalvar (subaortic and submitral) and apical. Right ventricular aneurysms were divided as outflow tract and apical (6). Thus, we have classified the defect described herein as a right ventricular outflow tract diverticulum.

Incidence

Congenital ventricular outpouchings are not common. Since 1940, fewer than 200 cases of left ventricular and only 18 or 20 cases of right ventricular outpouchings have been reported. This includes a number of cases discovered later in life, and in one instance at 62 years of age. Coexisting cardiac defects are listed in Table I. The most common associated lesions appear to be conotruncal malformations followed by isolated ventricular septal defects.[5]

Symptoms and Physical Findings

Early in life infants may present with tachycardia, various heart murmurs, poor feeding, and failure to thrive. Later in life, patients can present with dysrhythmias, chest pain, congestive heart failure, and syncope.[7] It is, however, unclear whether these symptoms result from the diverticulum or from the associated

Table I

Congenital Cardiac Defects Associated with Right Ventricular Diverticulum

Cardiac Defects	No. Cases Reported (Some Patients had Multiple Defects)
Atrial	
Atrial septal defect	4
Ventricular	
Ventricular septal defect	5
Valvular	
Aortic insufficiency	1
Pulmonary stenosis	2
Mitral insufficiency	1
Mitral valve prolapse	1
Conotruncal	
Truncus arteriosus	4
Double outlet right ventricle	2
Tetralogy of fallot	2

malformations. Since our patient was successfully weaned from all cardiac medications after ventricular septal defect closure, it appears that only this defect contributed significantly to the patient's congestive failure.

Diagnosis and Natural History

The diagnosis is often made by echocardiography or angiography. Both imaging modalities outline the contractile pattern of the outpouching and enable differentiation of an aneurysm from a diverticulum.

The natural history of right ventricular diverticulum is variable. Some cases are discovered incidentally at autopsy and others cause sudden death due to rupture or arrhythmias. Because the antemortem diagnosis is made in symptomatic patients, the number of patients with an asymptomatic diverticulum remains unknown. Treatment is supportive unless the diverticulum is causing symptoms.

Conclusions

Congenital right ventricular diverticula are rare congenital defects, which can present at any age with a variety of symptoms. Associated cardiac defects such as a VSD and conotruncal malformations may be more important determinants of the hemodynamics and prognosis than the diverticulum itself. In children, these defects can be easily detected noninvasively by echocardiography.

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