ILLUSTRATIVE ECHOCARDIOGRAM

Echocardiogram in Tetralogy of Fallot*

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This 20-year-old man was referred to the Mayo Clinic for corrective repair of tetralogy of Fallot. He had been cyanotic from birth. Tetralogy had been diagnosed when he was two years old, and a Blalock-Taussig anastomosis had been performed at six years of age because of severe cyanosis and limitation in exercise tolerance. After this operation, he grew and developed normally, and the cyanosis and exercise tolerance improved considerably. In the few months before the present examination, exercise tolerance had declined somewhat and cyanosis had increased.

CASE REPORT

At examination, the patient was 188 cm tall (74 inches) and weighed 70 kg (154 lb); his pulse rate was 80/min, and his blood pressure was 146/60 mm Hg. Moderate cyanosis and clubbing were present in all digits. The apex beat was felt in the anterior axillary line. A loud continuous murmur was heard over all of the precordium but was maximal in the right upper sternal area. The second heart sound was loud and single. A chest roentgenogram revealed cardiomegaly with increased pulmonary vascular markings. An electrocardiogram and a vectorcardiogram revealed right axis deviation, right bundle branch block, and right ventricular hypertrophy.

Echocardiography showed the following (Fig 1):

1. The right ventricular dimension (RVD), representing the vertical distance between the right ventricular epicardial echo and the right septal echo at end diastole, was increased to 2.6 cm (normal 0.5 to 2.1 cm). The RVD index (RVD/body surface area) was 1.34 cm/M² (normal value 0.5 to 1.1 cm/M²). The left ventricular internal dimension, representing the vertical distance between the left septal echo and the left ventricular endocardium at end diastole was 5.1 cm (normal 3.5 to 5.6 cm). The relatively large value in this patient was thought to be secondary to the functioning Blalock-Taussig anastomosis resulting in a relative volume overload of left ventricle.

2. The ventricular septal motion was normal. During ventricular systole, the left ventricular posterior wall (LPW) moved anteriorly while the septum (IVS) moved posteriorly; that is, both moved in opposite directions to each other with respect to the transducer (Fig 1). The mitral valve motion was normal. By medial and cephalad rotation of the transducer from the mitral valve position toward the aorta, the continuity between the anterior mitral leaflet (AML) and the posterior wall of the root of the aorta could be demonstrated (Fig 2 and 3). In normals, the AML and the posterior aortic wall are situated at the same depth, as are the interventricular septum and the anterior aortic wall (Fig 4 and 5). However, in this patient, the IVS and the anterior aortic wall were not at the same depth. In fact, the anterior aortic wall was anterior to the IVS and seemed to override it. The IVS echo seemed to end abruptly in its upper portion (Fig 2).

DISCUSSION

This echocardiographic feature (root of the aorta overlapping the ventricular septum) has been previously described in patients with tetralogy of Fallot,1 and a similar finding has also been noted in patients with double-outlet right ventricle.2 In the past, the differentiation between double-outlet right ventricle and tetralogy of Fallot has been based on mitral-semilunar valve discontinuity being demonstrated angiographically. Recently, Chesler et al2 have demonstrated this same feature echocardiographically, illustrating the application of this non-

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Figure 1. Echocardiogram revealing increased right ventricular dimension and normal ventricular septal motion. CW, chest wall; RV, right ventricle; IVS, interventricular septum; ECG, electrocardiogram; and LPW, left ventricular posterior wall.
invasive technique in delineating this important anatomic dissimilarity.

The echocardiographic feature of increased RVD index with normal ventricular septal motion is not specific, but is present in all conditions that lead to pressure overload of the right ventricle. Some of the conditions in which this echocardiographic feature has been obtained are pulmonary stenosis, pulmonary hypertension, large ventricular septal defect, tetralogy of Fallot, double-outlet right ventricle, transposition of the great arteries, and truncus arteriosus. In a cyanotic patient, tetralogy of Fallot can be tentatively diagnosed if the echocardiogram demonstrates an increased RVD index, normal ventricular septal motion, mitral-semilunar valve continuity, and an overriding aorta. We anticipate similar features in truncus arteriosus and are presently investigating this possibility.

Cardiac catheterization and selective right ventricular angiography in our patient confirmed the diagnosis of tetralogy of Fallot with severe infundibular pulmonic stenosis and a functioning right Blalock-Taussig anastomosis. The patient successfully underwent closure of the ventricular septal defect, relief of the pulmonic stenosis, and ligation of the shunt.

REFERENCES


