Echocardiographic Findings of Congenital Absence of the Pulmonary Valve with Tetralogy of Fallot*

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The characteristic echocardiographic findings for congenital absence of the pulmonary valve with tetralogy of Fallot were described in three patients, whose diagnoses were confirmed by operation and/or autopsy. The abnormal linear echo, which was thought to be derived from the rudimentary pulmonary valve tissue, was recorded anteriorly to the dense echo from the subpulmonary muscle mass. The echocardiographic signs of right ventricular volume overload were associated with overriding of the aorta.

Congenital absence of the pulmonary valve is a rare anomaly. It is generally associated with other cardiac anomalies, the most common of which is tetralogy of Fallot. To date, approximately 150 proved cases have been reported in the literature. There is little echocardiographic description except for the report of one case by Weyman et al. Recently, we have seen three patients with congenital absence of the pulmonary valve with tetralogy of Fallot. We present their echocardiographic findings in this article.

**MATERIALS AND METHODS**

The diagnosis of three patients, ages 7, 11 and 13 years, was proven by angiography and operation. Two of the three were also confirmed by autopsy. The echocardiographic examinations were performed with an ultrasonic cardiograph Sanei Sokki model WM-09, using 2.25 MHz transducer of 16 mm diameter. The echo signals were recorded by strip chart recorder or polaroid camera.

**RESULTS**

The summarized data from echocardiography are shown in Table 1. Overriding of the aorta was identified in all three patients. The M-mode scan between the mitral and the pulmonary valve provided the most important diagnostic findings. As shown in Figure 1A (case 1), a dense and wide echo was found at the posterior wall of the outflow tract of the right ventricle between the mitral and the pulmo-

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FIGURE 1A. M-mode scan from the mitral valve to pulmonary artery. A linear echo (arrow) was recorded in the outflow tract of right ventricle anterior to the subpulmonary conal muscle.

An enlarged right ventricle and paradoxic motion of the interventricular septum were found in all patients (Table 1), which was consistent with the hemodynamic basis of associated pulmonary regurgitation.

DISCUSSION

The echocardiographic identification of overriding of the aorta has been reported in tetralogy of Fallot. Additional echocardiographic findings were seen in our three patients, which corresponds to anatomic and hemodynamic characteristics of the absence of the pulmonary valve. The outflow tract of the right ventricle and the pulmonary artery were easily and clearly recorded, although they have rarely been recorded in simple tetralogy of Fallot because of the lung over the underdeveloped pulmonary artery. This fact suggested the presence of a dilated blood pathway out of the right ventricle. Pulmonary regurgitation is known to be definitely associated with this particular type of anomaly be-

FIGURE 1B. Echogram during catheterization, recorded on the line in Figure 1A. A linear echo (arrow) was recorded between the catheter and the subpulmonary conal muscle. PC = subpulmonary conal muscle, PA = pulmonary artery, IVS = interventricular septum, RVO = right ventricular outflow tract, MV = mitral valve, LA = left atrium, C = catheter.

FIGURE 2. Outflow tract of the right ventricle and pulmonary artery shows the rudimentary pulmonary valve (arrow) and dilatation of the outflow tract of the right ventricle and pulmonary artery. AO = aorta, RV = right ventricle, PA = pulmonary artery, PC = subpulmonary conal muscle.

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cause of inadequate function of the valve. The volume overload of the right ventricle due to the regurgitation produces dilatation of its chamber, its outflow tract and the pulmonary artery, except the constricted pulmonic valve ring. So the rudimentary valve tissue can be easily recorded in front of the subpulmonary conal muscle and in the space of the dilated outflow tract. The combination of echocardiographic findings of overriding of the aorta and the volume overload of the right ventricle suggests an association of the congenital absence of the pulmonary valve. In such a case, the area from the outflow tract of the right ventricle to the pulmonary artery should be carefully examined to ascertain the abnormality of the pulmonary valve.

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