Complete Transposition of the Atrioventricular Valves Associated With Left Ventricular Apical Hypoplasia

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A 22-year-old woman presented with chest discomfort. ECG showed atrial fibrillation, right axis deviation and T-wave abnormality. Transthoracic echocardiogram and MRI revealed that the 2 atrioventricular valves were transposed completely with mild to moderate regurgitation. However, the atrioventricular connection was concordant with normal atrial arrangement (Figure 1 and Movies I and II in the online-only Data Supplement). The left atrium was connected to the left ventricle by the tricuspid valve (Figure 2 and Movies III and IV in the online-only Data Supplement), whereas the right atrium was connected to the right ventricle through the mitral valve (Figure 3 and Movie V in the online-only Data Supplement). The atrial and ventricular septum was intact. In addition, the left ventricular apex was hypoplastic with patent ductus arteriosus.

It is commonly accepted that the mitral valve is always connected to the morphological left ventricle, and the tricuspid valve is invariably connected to the morphological right ventricle. However, our findings disagreed with this opinion, and complete transposition of the atrioventricular valves may represent a special congenital anomaly. Further research is required to reevaluate the embryological development of the atrioventricular valves.

Disclosures
None.

Figure 1. A, Transthoracic echocardiogram (in the 4-chamber view) showed that the left ventricular apex was hypoplastic, and mild to moderate regurgitation presented in the 2 atrioventricular valves. B, MRI confirmed the findings of echocardiogram. In addition, fat replacement was identified in the wall of the left ventricular apex.
Figure 2. In the short-axis views, transthoracic echocardiogram (A) and MRI (B) revealed that the left atrium was connected to the left ventricle by the tricuspid valve.

Figure 3. Transthoracic echocardiogram showed that only 2 leaflets were recognized in the right ventricle, and the dotted line was added to demonstrate the orifice of right atrioventricular valve (A, diastole; B, systole).
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