Tricuspid and Pulmonic Valve Echoes in Tricuspid and Pulmonary Atresia*

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An echocardiographic pattern of normal tricuspid valve motion in a patient with tricuspid atresia, and pulmonic valve motion in a patient with transposition of the great vessels and pulmonary atresia were documented. The recordings of the valve motion resulted in an initial erroneous diagnosis. Although M-mode echocardiography is very useful for the initial diagnostic evaluation of critically ill newborn infants with complex congenital heart diseases, on occasion recording of valvular echoes may result in conflicting clinical and pathologic correlation.

Tricuspid atresia and pulmonary atresia are relatively common cyanotic congenital heart diseases, and usually present with profound cyanosis in the neonatal period. Emergency cardiac catheterization study and immediate treatment are essential for infants with these cardiac anomalies. Rapid deterioration occurs as a result of decreased pulmonary blood flow as the ductus arteriosus constricts. Death invariably ensues. Echocardiography has been validated as a useful tool for initial screening in the acutely ill or cyanotic infant with cardiopulmonary diseases.1 The purpose of this report is to document a patient with tricuspid atresia and a patient with transposition of the great vessels and pulmonary atresia. Tricuspid valve and pulmonic valve echoes similar to normal motion were documented in the latter patient. The echocardiographic recording of the valve motion resulted in an initial erroneous diagnosis.

CASE REPORTS

CASE 1

A one-day-old, full term boy was admitted because of severe cyanosis at birth. Physical examination showed profound generalized cyanosis. The second heart sound was single and there was no murmur. Chest roentgenogram showed moderate enlargement of the right atrium and the superior vena cava and markedly diminished pulmonary markings. Electrocardiogram showed left axis deviation and decreased anterior forces.

An echocardiogram was obtained using a ultrasonoscope with a 3.5 MHz pediatric transducer (Smith-Kline Ekoline 20). A semilunar valve and an atrioventricular valve were identified. The atrioventricular valve had continuity with the great vessel (Fig 1). The left ventricle was moderately dilated. The interventricular septum was clearly identified.

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Figure 1. Echocardiogram of case 1. Mitral-aortic continuity is demonstrated. MV: mitral valve, Ao: Aorta.
and the small right ventricular chamber was noted. No other semilunar valve could be identified. With the transducer placed on the low left sternal border and tilted medially, a structure with a motion very similar to that of a normal tricuspid valve was identified. This anterior atrioventricular valve-like structure was clearly distinct from the mitral valve (Fig 2). Based on these echocardiographic findings, pulmonary valve atresia with intact ventricular septum with hypoplastic right ventricle was initially suspected.

Emergency cardiac catheterization studies confirmed the diagnosis of tricuspid atresia. Special attention was given to ensure that there was no patency of tricuspid valve by careful probing of the medial aspect of the right atrium and by right atrial cineangiography.

CASE 2

A one-day-old full term girl was admitted because of respiratory distress and intense generalized cyanosis which began shortly after birth. Physical examination showed severe cyanosis and moderate tachypnea. The second heart sounds were loud and single and a grade 2/6 crescendo-decrescendo systolic murmur was noted along the left sternal border. Chest roentgenogram showed mild cardiac enlargement and the pulmonary vasculature was normal. The electrocardiogram was within normal limits for the age of the patient.

Echocardiography identified two semilunar valves and two atrioventricular valves. The anterior atrioventricular valve was situated to the right of the posterior atrioventricular valve, which had continuity to the posterior great vessel (mitral valve: posterior and left; tricuspid valve: anterior and right; = ventricular d-loop) (Fig 3, 4). The anterior semilunar valve was located slightly anterior and to the right of the posterior semilunar valve. The tracing with two semilunar valves recorded simultaneously demonstrated an RC interval of 0.22 sec of the anterior semilunar valve, and 0.29 sec of the posterior valve (Fig 5). This suggested the diagnosis of d–transposition of the great vessels.

The infant underwent an emergency cardiac catheterization which confirmed the diagnosis of d–transposition of the great vessels with a ventricular septal defect. Balloon atrial septostomy was performed. Following the procedure, severe hypoxia continued and no obvious clinical improvement was noted. A second catheterization study showed evidence of

![Figure 2. Echocardiogram of case 1. This recording was obtained with the transducer placed in the 4th left intercostal space and tilted medially. An echo with motion very similar to that of a normal tricuspid valve is seen. TV: tricuspid valve.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21070/)

![Figure 3. Echocardiogram of case 2. Showing presence of mitral valve and ventricular septum. The left ventricle was smaller than normal. IVS: interventricular septum, LVPW: left ventricular posterior wall, MV: mitral valve.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21070/)
pulmonary atresia, transposition of the great vessels and a ventricular septal defect. Pulmonary blood flow was maintained through a patent ductus arteriosus. Shortly after the second catheterization and while the patient was being prepared for emergency systemic-to-pulmonary artery shunt procedure, sudden deterioration ensued and the infant expired.

Post-mortem examination confirmed the catheterization diagnosis. The left ventricle was moderately hypoplastic and the left ventricular outflow tract was atretic. The pulmonic valve was well developed with normal diameter of the valvular ring, and three apparent leaflets were seen, but no pulmonary aperture was evident. The main pulmonary artery was patent to the level of the pulmonic valve.

**DISCUSSION**

Echocardiography is an extremely important and essential diagnostic procedure for assessing critically ill or cyanotic infants and children. In many patients, echocardiography alone can allow an accurate anatomic diagnosis. However, in complex congenital cardiac anomalies, cardiac catheterization and angiography are necessary for final confirmation of the diagnosis especially prior to cardiac surgical procedures. With two-dimensional echocardiography more direct visualization of valve
motion, intracardiac anatomy and great vessel orientation can be appreciated. Hopefully two-dimensional echocardiography may provide accurate anatomic detail so that cardiac catheterization may be unnecessary in some instances.

M-mode echocardiographic findings of tricuspid atresia include an absent tricuspid valve echo, a small right ventricle and a large left ventricle. The mitral valve is easily recorded and a large aortic root which does not override the ventricular septum has been described. Absence of the tricuspid valve echo is characteristic. It should be noted that the tricuspid valve echo is relatively easy to record in normal infants. On pathologic examination, there is usually no connection between the right atrium and right ventricle, and no valvular material can be identified on either gross or microscopic examination. However, in some instances, the atresia may be of the membranous type, and there have been a few reports of patients with complete fusion of the valvular cusps. In those cases, it is conceivable that motion of atretic tricuspid portion of the right atrium can be visualized by M-mode echocardiography.

Pulmonary atresia with d-transposition of the great vessels is a rare congenital cardiac anomaly, comprising approximately 5 percent of neonates with transposition of the great vessels. The echocardiographic findings in this lesion have not been described in detail. However, one may expect to find absence of pulmonic valve echo and, possibly, some degree of hypoplasia of the left ventricle. However, in the presence of a normally moving semilunar valve echo with normal diameter of the great vessel at the valvular level, complete obstruction of the ventricular outflow tract or semilunar valvular atresia may not be suspected. In the second case report, pulmonary atresia was thought to be unlikely because of the recording of both semilunar valves. In this particular instance, motion of the well-developed pulmonic valvular leaflets was due to passive movement of the valve, since there was no antegrade flow across the pulmonic valve. In retrospect, the pulmonic valve echo was not quite normal in that multiple echoes were recorded during systole and that the separation of the valvular cusps was not appreciated.

These two patients are unusual and atypical. In general, failure to demonstrate a cardiac valvular structure by M-mode echocardiography indicates atresia of the valve unless it is due to technical factors. On the other hand, clear demonstration of valvular structure usually excludes atresia of that valve. However, there are exceptions to the rule, as demonstrated in our patients, and in one isolated case reported. Two-dimensional echocardiography is probably of greater value in such cases because it allows precise identification of the intracardiac spatial anatomy. However, some of the limitations of M-mode echocardiography may also be applicable to two-dimensional echocardiography.

REFERENCES