Ultrasound Cardiography in Single Ventricle and the Hypoplastic Left and Right Heart Syndromes


SUMMARY
Seven cases of single ventricle, six cases of hypoplasia of the right heart (tricuspid atresia), and one case of hypoplasia of the left heart were investigated with reflected ultrasound. In hypoplasia of the right heart there was abnormal anterior movement of the mitral valve; in hypoplasia of the left heart, abnormal anterior movement of the tricuspid valve, and in single ventricle, similar movement of the anterior component of a single valve. In one case of single ventricle, two separate valves were identified at different depths. In none of the cases could a ventricular septum separating two ventricular cavities be identified. Ultrasound cardiography may be particularly useful as a non-invasive preliminary investigation of infants with congenital heart disease. Failure to demonstrate a ventricular septum combined with the presence of a single atrioventricular valve moving abnormally far anteriorly may be an expression of marked hypoplasia of the left or right ventricles or an anatomically single ventricle with one valve.

Additional Indexing Words:
Mitral atresia  Tricuspid atresia  Atrioventricular valves

ULTRASOUND cardiography has proved to be a useful tool in the diagnosis of mitral valve disease,1–5 pericardial effusion,6–8 and hypertrophic obstructive cardiomyopathy.9–11 In addition, following recent clarification of the source of intracardiac echo patterns, identification of the cardiac chambers,10 the ventricular septum, and the aortic root is now possible, and left and right ventricular size,12 left atrial volume, and left ventricular stroke output have been estimated.13,14 Fewer studies have been performed with ultrasound in the field of congenital heart disease. Rapid motion of the mitral and tricuspid valves has been recognized as the result of the high flow in cases of atrial septal defect, ventricular septal defect, patent ductus arteriosus, and ostium primum defect, while a slow mitral downslope has been described in cases of membranous subaortic stenosis.15

Since it is now possible to identify echoes originating from the walls of the left and right ventricles, the ventricular septum,12 and occasionally the tricuspid valve,16 the potential for investigating more complex forms of congenital heart disease is apparent. During the course of our investigation of congenital heart disease by ultrasound cardiography it was noticed that cases of single ventricle and the hypoplastic left and right heart syndromes exhibited similar abnormalities of valve movement. The purpose of this paper is to describe these findings and to discuss the diagnostic value of ultrasound in children with congenital heart disease.
Methods

This paper is based on a study of six patients with tricuspid atresia, seven with single (common) ventricle, and one with the hypoplastic left heart syndrome. All the patients were fully investigated by cardiac catheterization and angiography. Particular attention was paid to the cineangiographic identification of the atrioventricular valves and their range of movement within the ventricular cavities.

The six patients with tricuspid atresia varied in age from 7 months to 11 years. All were cyanosed and their arterial oxygen saturations ranged from 24% to 80%. In the five cases in which the great vessels were normally related, angiography demonstrated a large left ventricle communicating through a narrow ventricular septal defect with the infundibular portion of the right ventricle; the inflow portion of the right ventricle was characteristically absent. In the remaining case, the great vessels were transposed, and a small right ventricular cavity was identified. In five cases the body type was situs solitus, and in one case there was situs inversus with mirror-image dextrocardia.

The seven patients with single ventricle varied in age from 30 months to 24 years. All were cyanosed, and their arterial oxygen saturations ranged from 56 to 80%. Single ventricle was diagnosed largely on the basis of angiographic evidence when a ventricular chamber received inflow of blood from a common atrioventricular valve (six cases) or from two separate atrioventricular valves (one case). A rudimentary chamber was identified in every case. In three cases the great vessels were transposed and non-inverted (D-transposition); in two, the great vessels were transposed and inverted (L-transposition) and in the remaining two these vessels were normally related. Pulmonary stenosis was present in all cases except in one of the cases with normally related great vessels. Two patients had abdominal heterotaxia, one with asplenia and levocardia, the other with polysplenia and marked dextroversion.

The patient with the hypoplastic left heart syndrome died at the age of 4 days following cardiac catheterization and angiography on the third day of life. Autopsy revealed polysplenia, dextroversion, and gross hypoplasia of the left atrium, left ventricle, mitral and aortic valves, and ascending aorta. The right ventricle was markedly enlarged and formed most of the cardiac bulk. The pulmonary artery was enlarged and supplied the descending aorta via a patent ductus arteriosus; pre-ductal coarctation of the aorta was also present.

Ultrasound examinations were performed on patients in the supine position. A commercially available ultrasonic apparatus transmitting bursts of 2.25 mHz ultrasound vibrations 1,000 times/
cavity of the right ventricle lies between the right septal echo and the endocardial surface of the right ventricle as an echo-free space 1 to 3 cm wide. An enlarged right ventricular cavity in a patient with double-outlet right ventricle and pulmonary stenosis is illustrated in figure 2. Posterior to the mitral valve echo, structures identified are usually the left atrial wall which moves posteriorly during ventricular systole and occasionally the left ventricular wall which moves anteriorly during systole (figs. 1 and 2).

**Results**

All the patients exhibited the same anatomic abnormality of the chambers ultrasonically. In none could a ventricular septum or two ventricular cavities be identified. In addition, abnormalities of valvular movement were evident.

In all six patients with tricuspid atresia, the anterior leaflet of the mitral valve exhibited a degree of movement toward the endocardium of the anterior ventricular wall in excess of normal. The distance between the point of maximum diastolic opening of the anterior leaflet (E point) and the endocardial surface of the anterior ventricular wall was usually 1 cm or less. The E point was thus found at a depth of approximately 2 to 3 cm from the anterior chest echoes depending on the thickness of the ventricular wall (fig. 3).

Of the seven patients with single ventricle, six exhibited findings like those observed ultrasonically in the patients with tricuspid atresia, namely a single leaflet moving to within a centimeter of the endocardial surface of the anterior ventricular wall (fig. 4A). Cineangiography in these cases showed a large sail-like anterior leaflet opening to the full extent of the ventricular chamber. In the remaining patient with single ventricle, two leaflets were identified ultrasonically; the anterior leaflet moved in close apposition to the anterior ventricular wall like a tricuspid valve, and the other leaflet lay posteriorly in the position of a normal mitral valve (fig. 4B). The leaflets in the latter case moved synchronously like the anterior leaflets of two separate valves rather than paradoxically like the anterior and posterior leaflets of a single valve. A ventricular septum separating the two leaflets was not identified by ultrasound or angiography.

The neonate with the hypoplastic left heart syndrome exhibited similar findings. A ventricular septum could not be identified and a
single atrioventricular valve moving into diastolic apposition with the anterior ventricular wall was evident (fig. 5).

**Discussion**

By definition, a single ventricle is one which receives both the tricuspid and mitral valves or a common atrioventricular valve. In most cases of single ventricle the tricuspid and mitral valve rings are separate, but because no ventricular septum is present, there is continuity between the basal aspects of the septal and anterior mitral leaflets. In other cases of single ventricle, particularly when it is associated with a common atrium as in the polysplenic or asplenic syndromes, a single
large atrioventricular valve ring with one large anterior and a smaller posterior leaflet may be present. From a functional point of view, both these types of valvular anomalies would account for the ultrasonic and cineangiographic findings in six of our seven cases in which one large anteriorly moving leaflet was found (figs. 4A, 5, and 6A). Less commonly in single ventricle the mitral and tricuspid valves are completely separated by muscular tissue. This type of arrangement would explain the findings in our one case in which two separate valves were identified at different depths (fig. 4B). The rudimentary chamber varied in size, but even when fairly large, it did not occupy a position in front of the anteriorly moving valve leaflet in the plane of the transducer beam.

In the hypoplastic right heart syndrome with a small or absent inflow portion of the right ventricle, ultrasound yielded similar findings to those observed in single ventricle with one atrioventricular valve inasmuch as the anterior mitral leaflet moved more anterior than normal (fig. 6B). This is exemplified in our cases of tricuspid atresia in which the left ventricle communicating with a basal outlet chamber formed by the infundibulum of the right ventricle. Pulmonary atresia, type 1, with a slitlike right ventricular cavity may be expected to yield similar ultrasonic abnormalities.

The hypoplastic left heart syndrome is classically associated with a minute, posteriorly situated left ventricle. The mitral or aortic valves, or both, may be atretic or markedly hypoplastic and stenotic. The enlarged right ventricle, with a single, anteriorly moving tricuspid valve, therefore, functions as a single ventricle supplying all, or most of the systemic circuit through an enlarged pulmonary artery and a patent ductus arteriosus (fig. 6C).
Hence this syndrome produces ultrasonic findings similar to those in single ventricle and the hypoplastic left heart syndrome.

While ultrasound cardiography does not provide absolute information as to the nature of a congenital malformation used in conjunction with other diagnostic procedures, it may be of considerable assistance. Visualization of the ventricular septum by angiography is in our experience not always easy and it is frequently difficult to distinguish between corrected transposition of the great vessels with two ventricles on the one hand and single ventricle with transposition of the great vessels and inverted infundibulum on the other. Ultrasonic demonstration of a ventricular septum, two ventricles, and a normally positioned mitral valve would militate against a diagnosis of single ventricle or the hypoplastic left and right heart syndromes. This feature may be of particular assistance in the neonate in whom these diagnostic problems arise. One of our patients was 3 days old, and the advantage of using a non-invasive bedside technic is apparent. From the point of view of therapy of single ventricle in the future, these cases in which two distinctly separate atrioventricular valve rings and leaflets are demonstrated may be more amenable to corrective surgery than the situation in which there is a common atrioventricular valve.

Addendum

Since submission of this paper we have studied an additional four cases: One case of tricuspid atresia and two cases of single ventricle showed a single anteriorly moving A-V valve. The remaining case was a single ventricle with two A-V valves. A ventricular septum was not identified in any of these patients.

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