Echocardiographic Estimation of Ventricular Hypoplasia in Complete Atrioventricular Canal

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SUMMARY Echocardiograms from 10 patients with complete atrioventricular canal (CAVC) were compared with autopsy specimens to determine the capabilities of echocardiography in identifying patients with ventricular hypoplasia. On the basis of echocardiographic ventricular size, patients could be divided into three groups: 1) "balanced" CAVC patients had both increased right and left ventricular end-diastolic dimensions (RVED and LVED) and an increased RVED/LVED ratio; 2) "dominant left ventricular" CAVC patients had an increased LVED and small or normal RVED and a diminished RVED/LVED ratio; 3) "dominant right ventricular" CAVC patients had an increased RVED, but small LVED, and increased RVED/LVED. There was complete agreement between echocardiographic ventricular dominance and pathologic findings. This study demonstrates that echocardiography may be valuable in assessing ventricular dominance in the presence of CAVC.

PULMONARY VASCULAR obstructive disease develops rapidly in infants with complete atrioventricular canal (CAVC). Unsatisfactory results from palliative pulmonary artery banding in CAVC and advances in surgical technique make primary repair the procedure of choice.

Bharati and Lev divided heart specimens from patients with CAVC into three pathological groups, based on the size of the ventricular chambers. The balanced form of CAVC had a large, hypertrophied right ventricle (RV) and a normal or enlarged and hypertrophied left ventricle (LV). The dominant right ventricular (DRV) form of CAVC had a large, hypertrophied RV with a smaller-than-normal LV, while the dominant left ventricular (DLV) form had a large and hypertrophied LV with a RV that was smaller than normal. These patterns of ventricular dominance in living patients with CAVC have been confirmed by recent angiographic studies.

A dominant left or right ventricle may make such a heart inoperable by present surgical techniques. In this study we tried to determine if these patterns of ventricular dominance could be identified echographically. A specific echographic pattern would permit the cardiologist and surgeon to assess the possibility of ventricular hypoplasia at the time of cardiac catheterization or surgery.

Patients

The echocardiograms and heart specimens of 10 patients with CAVC were evaluated. The patients died at an age from 2 months to 2.5 years (mean 1 year). Five patients died postoperatively, while the other patients died from various causes, such as pneumonia, septicemia, congestive heart failure, or upper airway obstruction. Eight patients had Down's syndrome. Three patients had an additional patent ductus arteriosus, but none had pulmonic stenosis documented by cardiac catheterization.

The echograms of 20 normal infants, age 1–18 months, were controls for the patient group.

Methods

Echocardiograms were obtained with a Hoffrel 101C ultrasonoscope interfaced with an Irex Contintrace Recorder. All patients underwent echographic examination on several occasions. The echogram performed nearest to the time of death was used for comparison with the pathologic specimens. The echocardiograms were performed at a mean age of 11 months, an average of 30 days before death.

The left ventricular end-diastolic dimension (LVED) was measured at the Q wave of the simultaneous ECG, from the left septal surface to the left ventricular endocardium. The measurement was made at the level of the mitral valve. Because the mitral valve was abnormal, both leaflets were often present at the LVED. The right ventricular end-diastolic dimension (RVED) was likewise measured from the right septal surface to the right ventricular epicardium. Each was measured to the nearest millimeter and compared with normal values described by Meyer and Gutgesell. Dimensions within 2 standard deviations about the mean normal value were considered normal.

A ratio of RVED to LVED (RVED/LVED) was calculated for patients with CAVC and compared with a group of normal controls. The absolute RVED and LVED values for this normal group of children was similar to that reported by Meyer and Gutgesell. We believed that the ratio might better reflect ventricular hypoplasia than any other dimension. The control group consisted of 20 children age 1–18 months (mean 7 months) and weight 3–10.5 kg (mean 6.7 kg).

Autopsy specimens were evaluated by three examiners without knowledge of the echographic findings. The anatomy of the atrioventricular valve and its relationship and attachment to the interventricular...
septum was assessed. The size of the LV and RV was measured and the RV was evaluated for the presence of a sinus, trabeculated portion, and conus. We compared the size of LV to the RV for each specimen. The significance of differences between patient and control groups was performed by $t$ test, and $p$ values $< 0.05$ were considered significant.

Results

Echocardiograms

In all patients the diagnosis of CAVC was made echographically on the basis of reported ultrasonic features. All patients had continuity of the mitral and tricuspid valves through a relatively deficient interventricular septum, abnormal configuration of the anterior leaflet of the mitral valve, and narrowing of the left ventricular outflow tract (fig. 1). In three patients with hypoplasia of the RV the tricuspid valve could not be distinctly visualized within the right ventricular cavity, but the mitral valve did traverse the interventricular septum in these instances (fig. 2). All patients had fragmentation of the septal echogram, except one child in whom the LV was hypoplastic (fig. 3).

On the basis of echographic ventricular dimensions (table 1), patients with CAVC could be separated into three groups. Group 1 consisted of six patients in whom the RVED was enlarged and the LVED normal or slightly enlarged. The mean RVED/LVED ratio...
Figure 3. The echocardiogram demonstrates features of the dominant right ventricular form of complete atrioventricular canal. The ratio of the right ventricle (RV) to left ventricle (LV) was 1.5. The mitral valve was fragmentary, and we did not observe septal fragmentation.

was 0.68, and larger than the normal ratio of 0.5 ($p < 0.005$). These patients were designated as having a balanced ventricular form of CAVC. The three children in group 2 had a large LVED and normal to small RVED with a RVED/LVED (0.29) that was significantly smaller than normal and other children with CAVC. Group 2 children were designated as the dominant left ventricular form of CAVC. Although only one patient was present in group 3, the LVED was small, the RVED very large, and the RVED/LVED much greater than normal (1.5 vs 0.5). This child was considered to have the DRV form of CAVC. There was no overlap in the patients in the three echographic groups (fig. 4).

Pathology

All 10 patients had CAVC as evident by a single atrioventricular orifice guarded by anterior, posterior and two lateral valve leaflets. Five patients died after attempted surgical correction; the others had no surgery. Examination of autopsy specimens revealed that the six patients in echographic group 1 had a large RV and a normal to slightly enlarged LV. These

<table>
<thead>
<tr>
<th>Pt no.</th>
<th>Age at echo</th>
<th>Wt at echo (kg)</th>
<th>RVED</th>
<th>LVED</th>
<th>RVED/LVED Ratio</th>
<th>Type of CAVC</th>
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<tbody>
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<td></td>
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<tr>
<td>1</td>
<td>4 mos</td>
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<td>1.2*</td>
<td>2</td>
<td>0.6</td>
<td>B</td>
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<tr>
<td>2</td>
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<td>1.4*</td>
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<tr>
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<td>9½ mos</td>
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<td>1.8*</td>
<td>2.7*</td>
<td>0.66</td>
<td>B</td>
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<tr>
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<tr>
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<tr>
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<td>B</td>
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<tr>
<td>Mean</td>
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<td>1.5*</td>
<td>2.2</td>
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<td>13 mos</td>
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<td>10</td>
<td>10 mos</td>
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<td>2.1*</td>
<td>1.4†</td>
<td>1.5</td>
<td>DRV</td>
</tr>
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</table>

*More than 97th percentile.
†Less than 3rd percentile.

Abbreviations: B = balanced form of complete atrioventricular canal; CAVC = complete atrioventricular canal; DLV = dominant left ventricular type of complete atrioventricular canal; DRV = dominant right ventricular type of complete atrioventricular canal; ECHO = echocardiogram; LVED = left ventricular end-diastolic dimension; RVED = right ventricular end-diastolic dimension.
Figure 4. The ratio of the right ventricular end-diastolic dimension/ left ventricular end-diastolic dimension (RVED/LVED) is shown for normal children and patients with atrioventricular canal. There is complete separation of the three types of canal by the RVED/LVED measurement.

Figure 5. The pathologic specimen is from a patient with the balanced form of complete atrioventricular canal. The right ventricle (RV) and left ventricle (LV) are large and normally developed. The large defect is guarded by a common atrioventricular valve (CAVV). PA = pulmonary artery; LA = left atrium.
specimens were the balanced form of CAVC as suggested by echocardiography (fig. 5). The three specimens which belonged to echographic group 2 had a large LV and diminutive RV sinus and were consistent with the DLV form of CAVC. The RV was primarily represented by the conus (fig. 6). The patient in echographic group 3 had a hypoplastic left ventricular chamber as suggested by ultrasound (fig. 7).

**Discussion**

Most patients with CAVC have an enlarged RV with a normal or large LV. These patients represent the balanced form of CAVC. Goor et al.\(^6\) described four cardiac specimens in which the common atrioventricular valve was exclusively related to the LV and the right margin of the atrioventricular valve was opposite to the interventricular septum. Bharati and Lev\(^7\) similarly reported five such specimens and described this as the DLV type of CAVC. In the same report three specimens with a hypoplastic LV were described and called the DRV form of CAVC.

Clinical recognition of these forms of CAVC is important because it may influence operability, surgical technique and prognosis. Ventricular hypoplasia usually has a very poor prognosis, making diagnosis before surgery imperative. Recent reports have described the presence of ventricular hypoplasia as indicated by measuring ventricular volumes during biplane cineangiography.\(^6,8\)

Echographic features of CAVC have been well described,\(^6,8,9\) but no one has attempted to delineate the features of ventricular hypoplasia by echocardiography. There appears to be rather clear separation of the three types of CAVC by calculating the RVED/LVED ratio. The RVED/LVED ratio in the child with the DLV form was less than 0.36, much smaller than other patients with CAVC and most normal children. Although there was only one patient who had the DRV form, the RVED/LVED was much greater than other children with CAVC.

The measurement of absolute right ventricular chamber size may be particularly difficult in this disorder, because the ultrasound beam traverses the RV obliquely. A comparison of the RV to LV by measurement of RVED/LVED nonetheless allows preliminary diagnosis of ventricular hypoplasia.

The surgical mortality in the groups with hypoplasia of the RV or LV was 100%. In our institution, in patients who have the balanced form of CAVC, the survival rate is greater than 75% at the time of surgical repair. We suggest that when hypoplasia of the RV or LV is suggested by the RVED/LVED ratio, the ventricular dimensions should be delineated at the time of cardiac catheteriza-

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**FIGURE 6.** This pathologic specimen represents the dominant left ventricular form of complete atrioventricular canal. The sinus portion of the right ventricle (RV) was hypoplastic and the RV was represented by a conus. Abbreviations as in figure 5.
The heart depicts the dominant right ventricular form of complete atrioventricular canal. The left ventricle (LV) is normally formed but hypoplastic. The right ventricle (RV) is large. The large defect guarded by a common atrioventricular valve (CAVV) is demonstrated in the left panel. Ao = aorta; other abbreviations as in figure 5.

References
Echocardiographic estimation of ventricular hypoplasia in complete atrioventricular canal.
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