Echocardiography in Truncus Arteriosus

The Value of Pulmonic Valve Detection

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SUMMARY
Nine patients with a clinical presentation suggestive of truncus arteriosus were examined by echocardiography. Subsequent intracardiac studies including cardiac catheterization and angiocardiography divided this group into five patients with truncus arteriosus and four patients with other congenital heart diseases. In all patients with proven truncus arteriosus, it was possible to identify only one semilunar valve with a large truncal vessel overriding the ventricular septum. In none of these patients could a second semilunar valve be found.

This study indicates that echocardiography may provide a direct, yet noninvasive method to exclude the diagnosis of truncus arteriosus by finding the pulmonic valve. However, the diagnosis of truncus arteriosus is not tenable when based solely on the inability to demonstrate the pulmonic valve. The importance of the clinical application of this method is discussed.

Additional Indexing Words:
Ultrasound cardiography
Tetralogy of Fallot
Pulmonic valve
Hypoplastic right heart syndrome
D-Transposition of great vessels

TRUNCUS ARTERIOSUS is an uncommon congenital cardiac anomaly, but its differentiation from other anomalies characterized by left to right shunt at the great vessels and/or the ventricular level presents significant diagnostic problems, particularly in infants. Cardiac catheterization and angiocardiography may be necessary to exclude the possibility of this condition. Ultrasonic techniques have developed rapidly in recent months and have been shown to be of increasing value in supplementing clinical data in the pediatric age group with congenital heart disease. The purpose of this presentation is to describe our early experiences in the ultrasonic examination of patients suspected of having truncus arteriosus and to illustrate the value of pulmonic valve detection in the definitive exclusion of this diagnosis.

Methods and Materials
This study consists of nine patients all of whom were suspected of having truncus arteriosus on clinical grounds with either increased or decreased pulmonary blood flow. Following cardiac catheterization and angiocardiography, four were proven to have other cardiac anomalies and five indeed had truncus arteriosus. Group I contained those with proven truncus arteriosus (table 1). Patients in whom truncus arteriosus was excluded formed Group II (table 2). One patient was shown to have a ventricular septal defect, one had patent ductus arteriosus, and two had Tetralogy of Fallot.

Echocardiographic examinations were done on all patients. A commercially available ultrasonoscope (Picker) producing 1000 pulses/sec and a 2.0 mHz transducer (0.75 in diameter) were used. Recording was by slave oscilloscope and 35 mm oscilloscope record camera. In every patient, the mitral valve, tricuspid valve, aortic valve, and pulmonic valve were sought. The left ventricular diameter was determined and the left ventricular outflow tract was evaluated by slow transducer angulation from the mitral to the aortic valve.

The techniques and application of echocardiography in infants and children have been described. Previously published techniques for the identification of the pulmonic valve were used and can be summarized as follows. The transducer is placed in the left parasternal position usually one interspace above...
Table 1

**Group I: Truncus Arteriosus**

<table>
<thead>
<tr>
<th>Name</th>
<th>Age</th>
<th>Sex</th>
<th>Clinical diagnosis and angiocardiography</th>
<th>Diagnosis by cardiac catheterization and angiocardiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>R.S.</td>
<td>82-55-13</td>
<td>7 mos.</td>
<td>M</td>
<td>Truncus arteriosus type I</td>
</tr>
<tr>
<td>M.S.</td>
<td>59-97-16</td>
<td>7 yrs.</td>
<td>M</td>
<td>Truncus arteriosus type I</td>
</tr>
<tr>
<td>L.F.</td>
<td>75-91-23</td>
<td>6 yrs.</td>
<td>F</td>
<td>Truncus arteriosus type IV</td>
</tr>
<tr>
<td>B.M.</td>
<td>36-86-54</td>
<td>12 mos.</td>
<td>F</td>
<td>Truncus arteriosus type IV</td>
</tr>
<tr>
<td>R.C.</td>
<td>73-28-83</td>
<td>20 yrs.</td>
<td>F</td>
<td>Truncus arteriosus type IV</td>
</tr>
</tbody>
</table>

Table 2

**Group II: Truncus Arteriosus Suspects**

<table>
<thead>
<tr>
<th>Name</th>
<th>Age</th>
<th>Sex</th>
<th>Clinical diagnosis and angiocardiography</th>
<th>Diagnosis by cardiac catheterization and angiocardiography</th>
</tr>
</thead>
<tbody>
<tr>
<td>D.C.</td>
<td>82-07-53</td>
<td>6 wks.</td>
<td>M</td>
<td>Truncus arteriosus type IV, seizure disorder</td>
</tr>
<tr>
<td>W.C.</td>
<td>80-83-18</td>
<td>4½ mos.</td>
<td>M</td>
<td>Truncus arteriosus type IV</td>
</tr>
<tr>
<td>L.T.</td>
<td>77-64-54</td>
<td>5 mos.</td>
<td>M</td>
<td>Truncus arteriosus type II</td>
</tr>
<tr>
<td>B.U.</td>
<td>80-41-88</td>
<td>11 mos.</td>
<td>M</td>
<td>Truncus arteriosus type II</td>
</tr>
</tbody>
</table>

that used for aortic valve detection and a beam is angulated laterally toward the left shoulder. Alternatively, the transducer may be displaced still higher and then directed posteriorly and inferiorly. The pulmonic valve is recognized by its anterior position with respect to the aortic valve and the registration of the valve cusp movement (fig. 1). Pulmonary artery injections of Indocyanine green¹⁰ have validated its identity (fig. 2).

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**Figure 1**

**Echocardiogram of aortic and pulmonic valves in a 4-year-old normal child.** The pulmonic valve (PV) is shown on the right and the aortic valve (AV) is shown on the left. The movement of the valve cusps is demonstrated between the margins of the great vessels. Note the pulmonic valve is anterior to the aortic valve. RVO: Right ventricular outflow; LA: left atrium; PHONO: phonocardiogram; ECG: electrocardiogram.
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Results
In group I, with cardiac catheterization and angiographically proven truncus arteriosus, the ultrasonic studies showed a large aortic root which was identified in a position overriding the ventricular septum. Continuity between the ventricular septum and the anterior aortic margin was lost (fig. 3). The pulmonic valve could not be found in any of these patients. The atrioventricular valves appeared normal. In group II, those patients in whom truncus arteriosus was excluded by cardiac catheterization and angiography, the pulmonic valve was identified in each instance. As above, the atrioventricular valves appeared normal. The two patients who were proved to have Tetralogy of Fallot also showed evidence of aortic overriding often indistinguishable from the truncus arteriosus group.

Discussion
Despite its relative infrequency, the specific differentiation of truncus arteriosus from other congenital cardiac anomalies with similar clinical and hemodynamic abnormalities is important for appropriate medical and surgical management. It has been difficult to differentiate such anomalies as atypical Tetralogy of Fallot, large ventricular septal defect, patent ductus arteriosus, transposition of the great vessels with ventricular septal defect, and others with a clinical picture of aortic runoff from truncus arteriosus by standard clinical examination and other noninvasive methods.2,11 This study indicates that echocardiography may provide a direct, yet noninvasive method of excluding the diagnosis of truncus arteriosus. Recent advances in the examination of infants by echocardiographic techniques and the ability to identify the semilunar valves and their relationship to each other has been useful in the diagnosis of hypoplastic left and right heart syndrome,12-15 atrial septal defect,16-18 congenital mitral stenosis,19 and Tetralogy of Fallot in infants and children.20 Detection of the pulmonic valve is a relatively new examination technique. It has already been proved useful in the identification of D-transposition of great vessels.21 Our initial detection rate in

Figure 2
Identification of the pulmonic valve echoes by contrast injection. Indocyanine green was injected into the main pulmonary artery during cardiac catheterization. The cusp was identified by the dense contrast material filling it. INJ: injection signal; ECG: electrocardiogram; PCG: phonocardiogram, PV: pulmonic valve, AV: aortic valve; LA: left atrium; RA: right atrium.

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Figure 3

Echocardiograms from a normal child and a patient with truncus arteriosus. As the ultrasound beam scans from the left ventricle to the base of the heart, there is a continuity between the ventricular septum (VS) and the anterior aortic margin in a normal child (above). In truncus arteriosus (below), there is a discontinuity between the ventricular septum and the anterior aortic margin with a large aortic root and aortic overriding. AV: aortic valve; MV: mitral valve; LA: left atrium, PHONO: phonocardiogram; ECG: electrocardiogram.

the pediatric age group was 40% of patients undergoing routine studies. However, recent review of the last 100 infants and children studied shows that the detection rate is now 92%. It is somewhat more difficult to detect the pulmonic valve in the general adult population, but it is relatively easy in neonates and infants because the skeletal system is cartilagenous and there is no intervening lung tissue.

In the patient group with truncus arteriosus presented here, the aortic valve was easily detected, but the pulmonic valve could not be identified. The aortic root was enlarged and straddled the ventricular septum to a varying degree as described previously. The findings of aortic overriding and absence of pulmonic valve strongly suggest truncus arteriosus. In those patients with congenital heart disease other than truncus arteriosus, both semilunar valves were identified. Continuity between the ventricular septum and the anterior aortic margin was found in the patient with patent ductus arteriosus and the patient with ventricular septal defect, findings which also helped to rule out the possibility of truncus arteriosus in these patients.

A particularly difficult ultrasonic differential diagnosis is that between truncus arteriosus and Tetralogy of Fallot. The degree of aortic overriding offers no useful information and may be the same in both conditions. However, the detection of pulmonic valve cusp echoes is extremely useful in ruling
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out truncus arteriosus (fig. 4). We have been able to identify the pulmonic valve in Tetralogy of Fallot with a small pulmonary annulus. Theoretically, a small or distorted pulmonary annulus could present difficulties in recognition. However, this was not a problem in our small series. Present echocardiographic technique is unable to identify the number of valve cusps, and no statement can be made as to the cusp structure in a truncal vessel. The diagnosis of truncus arteriosus can easily be excluded if two semilunar valves and continuity between the ventricular septum and the anterior aortic margin can be demonstrated. However, the converse is not true, at least with our present techniques. Since we have 8% failure to detect the pulmonic valve in the general pediatric population, diagnosis of truncus arteriosus is not tenable when based solely on the inability to demonstrate the pulmonic valve in the presence of septal-aortic discontinuity.

We have not examined a patient with Tetralogy of Fallot with pulmonary atresia, but would expect that the echocardiographic picture would be identical to that found in truncus arteriosus.

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