Two-Dimensional Echocardiographic Findings in Double Orifice Mitral Valve

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Between December 1981 and April 1984, five children ranging in age from 1 month to 5½ years examined by two-dimensional echocardiography appeared to have a double orifice mitral valve. The diagnosis was verified in one patient at surgery, one patient by angiography and one patient by necropsy. Associated malformations included mitral stenosis and regurgitation, coarctation of the aorta, ostium primum and secundum atrial septal defect, ventricular septal defect and hypoplastic left heart syndrome. Three varieties of double orifice mitral valve were observed: an incomplete bridge type (one patient), in which a small strand of tissue connected the anterior and posterior leaflets at the leaflet edge level; a complete bridge type (three patients), in which a fibrous bridge divided the atrioventricular orifice completely into equal or unequal parts and a hole type (one patient), in which an additional orifice with subvalvular apparatus occurred in the posterior commissure of the mitral valve. These three types could be distinguished by sweeping the transducer in cross-sectional view from the apex toward the base of the heart. Both orifices could be seen throughout the scan in the complete bridge type while in the incomplete bridge type the two orifices could be seen only at the level of the papillary muscles. In the hole type, the second orifice was seen at about midleaflet level. In all three types, the chordae surrounding each orifice attached to only one papillary muscle. Congenital mitral stenosis or regurgitation was evident in three patients. The type of the double orifice mitral valve did not predict the presence or severity of symptoms. (J Am Coll Cardiol 1985;6:383–7)

Methods

Patients. Between December 1981 and April 1984 two-dimensional echocardiograms were performed in five patients in whom double orifice mitral valve was recognized. The diagnosis of double orifice mitral valve was made retrospectively in two cases and prospectively in the other three cases. The patients’ ages ranged between 1 month and 5½ years. Doppler echocardiography was performed in three of the five patients. The diagnosis was verified by surgery in one patient, angiography in one patient and necropsy in another patient. In addition, the five patients had coarctation of the aorta (three patients), ventricular septal defect (one patient), mitral stenosis (two patients), mitral regurgitation...
Table 1. Patient Data in Five Cases

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (mo)</th>
<th>BSA (m²)</th>
<th>Diagnosis</th>
<th>Confirmation of DOMV</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>39</td>
<td>0.56</td>
<td>DOMV, CoA, MS, MR</td>
<td>Surgery</td>
</tr>
<tr>
<td>2</td>
<td>67</td>
<td>0.8</td>
<td>DOMV, BAV</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>20</td>
<td>0.5</td>
<td>DOMV, CoA, VSD muscular, ASD 1, MR</td>
<td>Angiography</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
<td>0.3</td>
<td>DOMV, CoA, MS, MR</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>0.3</td>
<td>0.24</td>
<td>DOMV, HLHS, ASD 2</td>
<td>Necropsy</td>
</tr>
</tbody>
</table>

ASD 1 = ostium primum atrial septal defect; ASD 2 = ostium secundum atrial septal defect; BAV = bicuspid aortic valve; BSA = body surface area; CoA = coarctation of aorta; DOMV = double orifice mitral valve; HLHS = hypoplastic left heart syndrome; MR = mitral regurgitation.

(three patients) and hypoplastic left heart syndrome (one patient) (Table 1).

Echocardiography. Echocardiography was performed using a Picker Echoview 80CI, a Diasonics CV 100 or an ATL Mark 600, with a 3.5 or 5.0 MHz transducer focused appropriately for the size of the patient. Images were recorded on ½ inch (1.25 cm) video cassette tape and were reviewed independently by three examiners. Parasternal long- and short-axis views and apical two and four chamber views were obtained in all patients and subxiphoid long- and short-axis views were recorded in three patients. The location of the orifices, the number and location of papillary muscles and the sites of attachment of the chordae of each orifice were determined from subxiphoid and parasternal short-axis views.

Angiographic diagnosis of double orifice mitral valve was made on the basis of visualization of two separate circular streams of unopacified blood entering the left ventricle during diastole.

Results

Two-dimensional echocardiography. The diagnosis of double orifice mitral valve was made prospectively in three patients. In retrospect, the double orifice mitral valve could be seen on the two-dimensional echocardiogram in the other two cases as well. The most useful views for demonstrating double orifice mitral valve were the parasternal or subxiphoid short-axis view and apical four chamber view. Long-axis views were not useful for diagnosing double orifice mitral valve or for distinguishing among the various types. In all five patients two normally located papillary muscles were identified. In Patient 3 with a hole type double orifice mitral valve, the axis of the smaller (anterior) orifice and subvalvular apparatus was almost orthogonal to the axis of the major (posterior) orifice. In contrast, the axis of the two orifices in the other types of double orifice mitral valve were nearly parallel. In each case the chordae of the lateral (anterior) orifice attached only to the anterolateral papillary muscle while the chordae of the medial (posterior) orifice attached to the posteromedial papillary muscle (Table 2).

Types. Sweeping the transducer in parasternal or subxiphoid short-axis view from the apex toward the base of the heart allowed differentiation among three types of double orifice mitral valve:

1. Complete bridge type (Fig. 1): Both orifices were visible from the leaflet edge all the way through the valve ring. Cuts at leaflet edge (Fig. 1C), at midleaflet (Fig. 1B)

Table 2. Two-Dimensional Echocardiographic and Doppler Results in Five Cases

<table>
<thead>
<tr>
<th>Case</th>
<th>Best Views</th>
<th>Orifice Size (diameter in cm)</th>
<th>Type</th>
<th>Doppler</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>Para short, sub short, apical 4</td>
<td>Med 0.8</td>
<td>Complete</td>
<td>MS, MR</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lat 0.8</td>
<td>bridge</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>Para short, apical 4</td>
<td>Med 0.7</td>
<td>Complete</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lat 0.8</td>
<td>bridge</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Para short, sub short, para long</td>
<td>Med 1.6</td>
<td>Hole</td>
<td>MR</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lat 0.3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Para short, sub short</td>
<td>Med 0.8</td>
<td>Incomplete</td>
<td>MR</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lat 0.8</td>
<td>bridge</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>Sub short</td>
<td>Med 0.2</td>
<td>Complete</td>
<td>—</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Lat 0.2</td>
<td>bridge</td>
<td></td>
</tr>
</tbody>
</table>

Apical 4 = apical four chamber; Lat = lateral; Med = medial; MR = mitral regurgitation; MS = mitral stenosis; para long = parasternal long axis; para short = parasternal short axis; sub short = subxiphoid short axis.
and at the valve ring (Fig. 1A) showed the double orifice mitral valve with the appearance of spectacles. Both orifices were circular, almost equal in size and well defined.

2. Incomplete bridge type (Fig. 2): A connection was seen between the anterior and posterior leaflet only at the leaflet edge (Fig. 2C) resulting in a double circle being seen only at this level. In more basal views (Fig. 2A and B), the mitral valve appeared normal again.

3. Hole type (Fig. 3): At the edge of the leaflets (Fig. 3C) only the medial orifice could be seen along with the anterolateral papillary muscle. At midleaflet level (Fig. 3B) the additional orifice in the lateral commissure became visible, with an axis almost orthogonal to the axis of the major orifice. At anulus level a cleft in the anterior leaflet of the mitral valve was demonstrated (Fig. 3A) but the minor orifice was no longer visible.

**Discussion**

**Echocardiographic appearance.** In our five patients we could clearly recognize three different types of double orifice mitral valve using two-dimensional echocardiography. A carefully performed sweep in parasternal or subxiphoid short-axis view from the base of the heart to the valve anulus showed the morphologic characteristics. Similar to Hartmann (3) and Cascos et al. (27), we found a complete bridge type, an incomplete bridge type and a hole type of double orifice mitral valve (Fig. 2 to 4). In the complete bridge type, a spectacles-like double orifice mitral valve could be imaged from the leaflet edge level through the mitral anulus. This characteristic appearance of the two circles could be seen only at the leaflet edge in the patient with an incomplete bridge type. Although the relative size of the orifices differed, the chordae of each orifice attached to only one papillary muscle creating a kind of double parachute valve. These two types may represent opposite ends of a spectrum in which a tissue bridge connects the mitral valve leaflets. However, reports of specimens give no hint that there are
Figure 2. Incomplete bridge type of double orifice mitral valve. Panel D shows the echocardiographic plane for panels A, B and C. The double orifice is visible only at leaflet edge level (C). Ao = aorta; I = inferior; MR = mitral ring; RV = right ventricle; S = superior; other abbreviations as in Figure 1.

Figure 3. Hole type of double orifice mitral valve. Panel D shows the echocardiographic plane for panels A, B and C. The additional orifice is demonstrated only at midleaflet level (B). The lateral orifice disappears further toward the apex and the anterolateral papillary muscle (ALPM) becomes visible (C). L-S = left and superior; R-I = right and inferior; other abbreviations as in Figures 1 and 2.

intermediary types, nor have we recognized any such cases echocardiographically.

In the patient with a hole type double orifice mitral valve, a secondary orifice in the lateral commissure of the mitral valve was visible only at midleaflet level and disappeared on scanning either toward the apex or base. The chordae from the anterior orifice inserted on the anterolateral papillary muscle, which appeared to be nearly perpendicular to the postero medial papillary muscle. No chordae of the major orifice of the mitral valve attached to the anterolateral papillary muscle, connecting instead to the postero medial papillary muscle. In this way two parachute orifices were again created.

Clinical correlation. The presence or severity of symptoms was not predicted by the type of double orifice mitral valve. Nonetheless, we suggest that patients with evidence of mitral stenosis or regurgitation undergo two-dimensional echocardiography to determine the anatomy of the mitral valve. In AV canal defects, the presence of a double orifice mitral valve may complicate the surgical repair (26). In Case 1 of our series (complete bridge type), a division of the tissue bridge was attempted, but severe regurgitation occurred as a result and a valve replacement was necessary. However, division of the tissue bridge may be more successful in patients with an incomplete bridge type.

M-mode echocardiographic examination was very insensitive for detecting double orifice mitral valve. However, the two-dimensional echocardiogram appears to be a useful noninvasive method for detecting and characterizing double orifice mitral valve.

Patient 3 has undergone surgical correction of cardiac defects with confirmation of the mitral valve configuration.

References