Ostium primum defects with cleft mitral valve

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Ostium primum defects are common; by 1955, 37 operated cases had been reported by Kirklin, Daugherty, Burchell, and Wood. The two main problems in the surgical treatment of primum defects have been the avoidance of air embolism and heart block. The aim of this paper is to discuss the technical details found to be important in this respect.

MATERIAL

The material is summarized in Table I. During a five-year period I have performed open heart surgery in 525 patients. Of these, 150 patients had atrial septal defects of the secundum type operated under hypothermia with two deaths, and 17 patients had the primum type, four of whom died after surgery.

The two youngest patients were 3½ and 4 years old; both died, one from heart block, the other from brain damage following deep hypothermia. The two oldest patients were 39 and 46 years and both died, one from block, the other from circulatory insufficiency.

The heart was significantly enlarged in five patients. In three of these, including the two with an abnormal venous return, the pulmonary artery pressure at rest was above 70 mm. Hg. The most important diagnostic sign was a left axis deviation in the electrocardiogram (Toscano-Barbosa, Brandenburg, and Burchell, 1956), which was found in 14 of the 17 patients. At surgery a cleft mitral valve was found in all patients. Only in one was this cleft mitral valve proved to be completely competent. Additional secundum defects were found in three patients (cases 11, 13, and 17), a total abnormal venous return in one woman 46 years old (case 8; arterial oxygen saturation 82%, pulmonary artery saturation 84%), and an abnormal venous return of the right pulmonary veins in a 27-year-old woman (case 11; arterial oxygen saturation 82%). Two patients had heart block and died. A further two patients died; one 4-year-old child died from brain damage following

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Sex</th>
<th>Age (yrs)</th>
<th>Heart Size (ml/m²)</th>
<th>Shunt Left to Right in % of Pulmonary Flow</th>
<th>Systolic Pulmonary Artery Pressure (mm.Hg)</th>
<th>Perfusion Time (min.)</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>18</td>
<td>530</td>
<td>66</td>
<td>34</td>
<td>37</td>
<td>Deep hypothermia; epileptic fits; cleft mitral valve overlooked, sutured at re-operation 2 years later</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>4</td>
<td>460</td>
<td>66</td>
<td>34</td>
<td>41</td>
<td>Deep hypothermia; died after 1 month from brain damage</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>3-5</td>
<td>800</td>
<td>65</td>
<td>23</td>
<td>38</td>
<td>Deep hypothermia; died after 1 day with heart block</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>17</td>
<td>450</td>
<td>60</td>
<td>22</td>
<td>41</td>
<td>Deep hypothermia; Good result</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>10</td>
<td>530</td>
<td>50</td>
<td>47</td>
<td>53</td>
<td>Good result</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>28</td>
<td>400</td>
<td>65</td>
<td>24</td>
<td>81</td>
<td>Good result</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>23</td>
<td>460</td>
<td>65</td>
<td>24</td>
<td>81</td>
<td>Also total abnormal venous return with cyanosis, arterial O₂=82%; Died after 6 days with heart block</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>46</td>
<td>620</td>
<td>72</td>
<td>127</td>
<td></td>
<td>Good result</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>9</td>
<td>470</td>
<td>50</td>
<td>35</td>
<td>76</td>
<td>Good result</td>
</tr>
<tr>
<td>10</td>
<td>M</td>
<td>8</td>
<td>420</td>
<td>50</td>
<td>22</td>
<td>80</td>
<td>Also 2 secundum defects + abnormal return of right pulmonary veins with cyanosis, arterial O₂=82%; good result</td>
</tr>
<tr>
<td>11</td>
<td>M</td>
<td>27</td>
<td>700</td>
<td>73</td>
<td>85</td>
<td></td>
<td>Good result</td>
</tr>
<tr>
<td>12</td>
<td>M</td>
<td>17</td>
<td>840</td>
<td>70</td>
<td>18</td>
<td>78</td>
<td>Also 2 secundum defects; died after 1 day in circulatory insufficiency</td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>39</td>
<td>580</td>
<td>75</td>
<td>18</td>
<td></td>
<td>Good result</td>
</tr>
<tr>
<td>14</td>
<td>M</td>
<td>6</td>
<td>450</td>
<td>60</td>
<td>22</td>
<td>45</td>
<td>Good result</td>
</tr>
<tr>
<td>15</td>
<td>F</td>
<td>11</td>
<td>435</td>
<td>70</td>
<td>30</td>
<td>53</td>
<td>Good result</td>
</tr>
<tr>
<td>16</td>
<td>F</td>
<td>35</td>
<td>700</td>
<td>70</td>
<td>22</td>
<td>62</td>
<td>Also secundum defect; good result</td>
</tr>
<tr>
<td>17</td>
<td>F</td>
<td>20</td>
<td>450</td>
<td>83</td>
<td>26</td>
<td>52</td>
<td>Good result</td>
</tr>
</tbody>
</table>

Perfusion time corresponds to time for total circulatory arrest in cases 1, 2, 3, and 4

Thorax (1965), 20, 405.
deep hypothermia and one 39-year-old woman died from circulatory insufficiency.

**DISCUSSION**

**PREVENTION OF AIR EMBOLISM** In opening the right atrium during bypass, in the presence of an atrial defect and competent mitral valves, there is always a risk of air embolism in the beating heart. A primum defect with an audible apical systolic murmur in the left axilla may be associated with competent mitral valves; a fact that I have observed at angiocardiography by injecting contrast medium into the left ventricle. For this reason at operation the competence of the mitral valves is always first estimated by palpation from the right auricular appendage before cannulation. In many patients a definite systolic regurgitant jet cannot be palpated in the mitral area and the aorta is therefore occluded for a few minutes when the right atrium is opened and the mitral leaflet is rendered incompetent by tension on a suture placed through the two distal corners of the mitral cleft. The aortic clamp is then immediately removed and coronary perfusion is continued. A row of isolated 4-0 silk sutures is placed at 2 mm. intervals through the edges of the cleft mitral leaflets. It is important to observe that sometimes these edges are turned inwards under the ventricular side of the leaflets. By turning the edges out and taking small bites with the suture, the leaflet area will be made larger. If the leaflet edge is held inwards by a chorda tendinea this may be divided, but this is rarely necessary. If a cleft mitral leaflet is competent it is not advisable to divide any chordae tendineae before the suture of the cleft. Sometimes some chordae close to the interventricular ridge may be divided to allow

**FIG. 1.** The sutures are placed so as to avoid block in ostium primum defects. From the cleft to the level of the coronary sinus all sutures are isolated through the base of the mitral valve and placed parallel to its insertion. The suture in the cleft has one branch on one side and one on the other side of the cleft going up through the prosthesis. With this technique the bundle of His will never be in danger.
better movement of the mitral valve. The sutures are continued to the distal corner of the cleft, care being taken to avoid causing mitral stenosis.

Once the mitral cleft has been closed and found to be competent, the aorta is cross-clamped and the heart packed in crushed ice. The dacron prosthesis is then sutured in place. When the heart fibrillates after a few minutes the aortic clamp is removed and the heart with coronary perfusion is kept fibrillating with the ice to prevent air embolism. The pressure is increased in the lungs and the heart massaged to evacuate air from the the left side before the last stitch in the prosthesis is drawn taut. A puncture hole in the highest point of the ascending aorta is made before the heart is defibrillated for evacuation of any trapped air bubble.

PREVENTION OF HEART BLOCK  Heart block is best prevented by placing the sutures for fixation of the prosthesis in such a way that the bundle of His is never crossed. The first suture is placed at the base of the mitral valve before the cleft is closed by one end introduced from the ventricular side of the mitral valve, the other end of the suture being introduced in a similar fashion on the other side of the cleft. Then a row of interrupted sutures is placed through the mitral valve parallel and close to its base and through the prosthesis from the cleft to the level of the coronary sinus (Fig. 1). Thereafter there is no further danger of producing a heart block, and a continuous suture can be used eventually with some isolated mattress sutures through the base of the tricuspid valve on the other side of the cleft (Fig. 2).
CONCLUSION

From a surgical point of view it seems to be of no importance to differentiate between the partial and the total form of common atrioventricular canal as the operative technique applied is identical. The ostium primum defect and the cleft mitral valve are operated upon but not the tricuspid valve. There is usually too little tissue left in the septal tricuspid leaflet for a plastic procedure even though the valve is competent. The policy not to accept patients with cyanosis and pulmonary hypertension (DuShane, Weidman, Brandenburg, and Kirklin, 1960) has not been followed. In this series both patients with cyanosis were complicated with an abnormal venous return and in one the operation was successful (Figs 3 and 4). Only angiocardioigraphy from the pulmonary artery could verify the suspicion of an abnormal venous return. During the first period only the tricuspid leaflet was used for fixation of the prosthesis at the valvular level, and this technique is still advocated by Gerbode, Kerth, Sabar, Selzer, and Osborn (1964). Sutures in the mitral valve for fixation of the prosthesis were avoided for fear of

FIG. 3. Case 8. The heart in a 46-year-old woman having an ostium primum defect (P.D.) with a cleft mitral valve as well as total abnormal venous return (T.A.V.R.) with drainage into the right atrium (R.A.) through a small secundum defect. The arterial oxygen saturation was 82% and each time during surgery that the secundum defect was closed by the fingertip on palpation the blood pressure decreased.

FIG. 4A. Case 11. The heart in a 27-year-old woman with an ostium primum defect (P.D.) with a cleft mitral valve, a secundum defect (S.D.) and a right abnormal venous return (R.A.V.R.). The arterial oxygen saturation was 82%.

FIG. 4B. The operative procedure in which the secundum defect was joined to the primum defect after closure of the cleft in the mitral valve. A large patch of dacron was sutured around the right abnormal venous return and the secundum and primum defects. The patient made an uneventful recovery.
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distortion and insufficiency of the mitral valve. When the sutures were placed through the base of the tricuspid leaflet the bundle of His had to be crossed. In one patient a stitch caused a block that disappeared on withdrawal of the needle, but two patients got permanent heart block. The mitral valve is so thickened in these patients that it will not be distorted. By placing all sutures from the cleft to the coronary sinus at the base of the mitral valve, the bundle of His will never be in danger (see Fig. 1).

SUMMARY

The experience of surgical treatment in 17 patients with primum defects and cleft mitral valve is reported. The technical steps for the prevention of air embolism and heart block are discussed.

REFERENCES


Ostium Primum Defects with Cleft Mitral Valve

Viking Olov Björk

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