A Surgical Approach to Ebstein's Disease


The natural history of Ebstein's disease is variable. Usually progressive disability results in death by the end of the second decade.1 Of 48 deaths reviewed by Mayer et al.,2 40 per cent died before the age of 10, with an additional 30 per cent in the second decade. Contrasted to these premature deaths are the six patients referred to that survived beyond the sixth decade of life and the many asymptomatic children.

Dyspnea, cyanosis, congestive heart failure, and arrhythmias are the cardinal manifestations of the disease and account for the majority of deaths. The management of symptomatic patients has been unsatisfactory until recently. Response to antieongestive and antiarrhythmic drugs is variable and of little value to the patient who is hypoxic from a right-to-left shunt and diminished pulmonary blood flow. Experience with such a patient treated surgically constitutes the basis of this report.

Case History

R.R., a 7-year-old boy was first noted to be cyanotic 6 hours after birth. The cyanosis persisted along with retarded physical development. At 10 months of age hemoptysis occurred. He began to walk at 1 year, at which time dyspnea was first noted.

Cardiac catheterization was performed at 3 1/2 years of age at the Children's Medical Center, Boston, Massachusetts. The data, table 1, indicated a right-to-left shunt at the atrial level with right atrial pressures exceeding pressures in the left atrium. A gradient between the pulmonary artery and right ventricle indicated mild infundibular stenosis. The tricuspid valve was abnormally displaced toward the apex and the body of the right ventricle was extremely small. The position of the tricuspid valve and the small right ventricle, associated with a right-to-left shunt at the atrial level, was considered diagnostic for Ebstein's disease. No therapy was recommended.

During the 3 1/2 years since catheterization his symptoms worsened. At age 5 hemoptysis recurred. Cyanosis deepened and dyspnea with effort became more severe. The year prior to admission he was hospitalized on two occasions for phlebitis because of polycythaemia. Owing to a progressive deterioration in his clinical course, he was referred to the National Naval Medical Center for surgical consideration.

Physical findings revealed a small, 37-pound boy with marked generalized cyanosis and clubbing. Peripheral pulses were normal. Blood pressure was 108/58 in the right arm. A prominent A wave was seen in the jugular venous pulse. Precordial palpation indicated normal activity. At the lower left sternal border there was a presystolic crescendo murmur that varied with respiration and ended in an accentuated first sound (fig. 1). A fourth sound was heard best at the fourth left intercostal space. The second sound was single and of normal intensity. There was no evidence of congestive heart failure.

Chest x-ray (fig. 2) revealed slight cardiac enlargement with prominence of the right atrial border. The peripheral pulmonary vasculature was diminished. A "lacy" pattern in the lung fields suggested increase in bronchial collateral vessels. The electrocardiogram (fig. 3) revealed right axis deviation, marked right atrial hypertrophy, and normal precordial leads. The hemoglobin was 19.8 Gm. per cent, with a hematocrit level of 70 volumes per cent.

The clinical findings confirmed the previous diagnosis of Ebstein's disease. Because of progressive deterioration it was decided to bypass partially the right heart with a superior vena cava to right pulmonary artery anastomosis.

On August 16, 1962, through a right thoracotomy, an anastomosis of the end of the right pulmonary artery to the side of the superior vena cava was performed. The size of the anastomosis was limited by a 7- to 8-mm. pulmonary artery. A ligature was placed around the superior vena cava at its junction with the right atrium, and acute occlusion elevated caval pressure to 30 mm. Hg.
SURGICAL APPROACH TO EBSTEIN'S DISEASE

producing facial plethora. After release, gradual constriction to an estimated internal opening of 1 mm. was carried out. At this point the superior vena caval pressure was 10 mm. Hg. No significant facial congestion or cyanosis developed. It was anticipated that the point of constriction would thrombose over a period of time because of intimal damage at that site with low pressure flow.

Postoperatively, the child was kept in Fowler's position. There was a marked decrease in cyanosis without evidence of venous congestion in the head or neck. He had an uneventful recovery and was discharged on the tenth postoperative day.

Since surgery he has shown marked improvement. His cyanosis has diminished and he now attends school full-time without undue fatigue. Follow-up examination on September 21, 1962, revealed mild generalized cyanosis. Resting arterial saturation averaged 81 per cent by Beckman analysis. The presystolic murmur at the lower left sternal border is now barely audible. X-ray (fig. 2) and electrocardiogram (fig. 3) show little change except for slight diminution in the amplitude of the P waves in the latter. Venous angiography (fig. 4) shows adequate filling of the right pulmonary artery from the superior vena cava with no dye entering the right atrium.

Discussion

Impaired right heart function in Ebstein's disease is related to the "atrialization" of the right ventricle by the downward displacement of the tricuspid valve. In this anomaly the right ventricle is divided into a proximal "atrial" and a distal "ventricular" chamber.

Table 1

<table>
<thead>
<tr>
<th>Site</th>
<th>Oxygen saturation</th>
<th>Pressure mm. Hg</th>
<th>Flows L./min./M.²</th>
</tr>
</thead>
<tbody>
<tr>
<td>SVC</td>
<td>45.9</td>
<td>a = 15 (6)</td>
<td>Pulmonary 1.2</td>
</tr>
<tr>
<td>RA</td>
<td>47.9</td>
<td>a = 12 (5.5)</td>
<td>Systemic 2.0</td>
</tr>
<tr>
<td>RV</td>
<td>47.6</td>
<td>30-40/-</td>
<td></td>
</tr>
<tr>
<td>PA</td>
<td>47.6</td>
<td>9/3</td>
<td></td>
</tr>
<tr>
<td>PV</td>
<td>100</td>
<td></td>
<td></td>
</tr>
<tr>
<td>LA</td>
<td>70.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>LV</td>
<td>70.3</td>
<td>90-95/0-5</td>
<td></td>
</tr>
<tr>
<td>FA</td>
<td>70.8</td>
<td>90/55 (70)</td>
<td></td>
</tr>
</tbody>
</table>

SVC, superior vena cava; RA, right atrium; RV, right ventricle; PA, pulmonary artery; PV, pulmonary vein; LA, left atrium; LV, left ventricle; FA, femoral artery.

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Figure 1

Phonocardiogram showing presystolic murmur (PSM) ending in loud first sound (S₁).

Figure 2

Chest x-ray (top, before operation; bottom, after operation) showing no change in size or contour.
Electrical depolarization of these two chambers occurs simultaneously. During right atrial contraction the atrialized portion of the right ventricle is in diastole. Forward blood flow into the right ventricle is thus, in part, taken up by the proximal chamber reducing the effectiveness of atrial systole. To compensate, right atrial stroke volume is increased and dilatation ensues.

On the other hand, the distal right ventricular chamber is diminutive as a result of downward displacement of the tricuspid valve. The small volume and lack of distensibility of the ventricular chamber act to obstruct right atrial emptying accounting for high right atrial pressures. Lack of significant gradients across the tricuspid valve and the large A wave in the right ventricular pressure curves indicate absence of obstruction at the valvular level.

Dilatation and increased pressure within the right atrium prevent normal closure of the foramen ovale. An atrial right-to-left shunt develops and pulmonary blood flow diminishes, resulting in the clinical appearance of cyanosis and exertional dyspnea. If the opening in the atrial septum is not adequate to allow sufficient decompression of the right atrium, further dilatation ensues and congestive heart failure develops. Marked dilatation of the right atrium and the frequent occurrence of the Wolff-Parkinson-White syndrome predispose to supraventricular arrhythmias.

In the past there has been no good surgical procedure for Ebstein's disease.3-5 Glenn in 1958 suggested partial circulatory bypass of the right side of the heart by anastomosing the superior vena cava to the right pulmonary
artery. Ample animal experimentation supported the feasibility of this approach. In 1959 Gasul et al. reported the first patient with Ebstein's disease in whom a successful superior vena cava-right pulmonary artery anastomosis was performed with marked improvement in symptomatology and arterial oxygen saturation. Subsequently, he reported three additional patients similarly treated, with one death. McCredie et al. reported subjective improvement in a 45-year-old woman with Ebstein's disease, in whom a superior vena cava to right pulmonary artery anastomosis was performed, and recommended further trial in severely cyanotic cases for whom no other safe procedure is available.

Partial bypass of the right heart is particularly suited to the impaired function encountered in Ebstein's disease. A superior vena cava to right pulmonary artery anastomosis reduces venous return to the right atrium by 30 to 40 per cent and in so doing reduces right atrial volume and pressure. Reduction in right atrial pressure will also diminish the volume of the right-to-left shunt which, in part, accounts for the improvement in arterial oxygen saturation. Finally, pulmonary blood flow is improved without increasing the volume work of the left ventricle or subjecting the pulmonary vessels to systemic arterial pressures. The latter undesirable conditions are encountered with various systemic-pulmonary shunts.

The patient presented emphasizes the benefit from partial right heart bypass in Ebstein's disease. It is our belief that right heart bypass with superior vena cava to right pulmonary artery anastomosis is the surgical procedure of choice in the treatment of children and adults severely limited with Ebstein's disease.

Summary

Ebstein's disease in a severely cyanotic 7-year-old boy was successfully treated by anastomosing the superior vena cava to the right pulmonary artery.

A discussion of impaired right heart function in Ebstein's disease is presented emphasizing the inefficiency of right atrial contraction as a consequence of the "atrialization" of the right ventricle.

It is our belief that right heart bypass with a superior vena cava to right pulmonary artery anastomosis is the surgical procedure of choice in patients severely ill from Ebstein's disease.

References

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