Left Ventricular Anomalies
Associated with Ebstein’s Malformation of the Tricuspid Valve

ALI A. MONIBI, M.D., WILLIAM H. NECHEES, M.D., CORA C. LENOX, M.D.,
SANG C. PARK, M.D., ROBERT A. MATHEWS, M.D., AND J. R. ZUBERBUHLER, M.D.

SUMMARY Seventeen patients with Ebstein’s malformation of the tricuspid valve have been evaluated. The majority of patients (11/17) presented in the newborn period with cyanosis and cardiomegaly. The remainder (6/17) were referred because of cardiomegaly on chest X-ray and/or a heart murmur heard on routine examination. Three patients have died; one in the newborn period and two suddenly at the age of 11 and 15 years. Thirteen patients have undergone right and left heart cardiac catheterization within the last seven years. Twelve of these 13 patients (92%) had angiographic left ventricular contraction abnormalities. Five patients also had mitral valve prolapse associated with left ventricular dysfunction. Since many patients with Ebstein’s anomaly may have significant left ventricular abnormalities, careful and systematic evaluation of the left ventricle is warranted.

EBSTEIN’S MALFORMATION of the tricuspid valve (EBS) is most commonly associated with a patent foramen ovale or fossa ovalis type of atrial septal defect. Other associated lesions such as pulmonic stenosis, pulmonary atresia, ventricular septal defect, and rarely transposition of the great arteries or tetralogy of Fallot have also been described. In contrast, abnormalities of the left side of the heart have not been well documented. Over the last few years, some unusual left ventricular (LV) abnormalities have been noted on angiography in patients with EBS seen at Children’s Hospital of Pittsburgh. This report will describe the findings in these patients.

Materials and Methods

Over the 7½ year period from January 1970 through June 1977, 17 patients with EBS were seen at Children’s Hospital of Pittsburgh: 9 males and 8 females. Thirteen patients had cardiac catheterization during this study period while the remaining four patients had cardiac catheterization prior to this study. All patients who underwent catheterization during this period of time had evaluation of both left and right heart hemodynamics and cineangiography.

The criteria for inclusion of patients in this study were the availability of adequate cardiac catheterization and cineangiographic data and also current clinical information. In each case the diagnosis of EBS was established by cineangiography.

Three patients died during this study period. The ages of the surviving patients ranged from 10 months to 22 years (mean age 10.8 years). In all patients the clinical records as well as previous catheterization data and cineangiograms were reviewed. Autopsy specimens were available in two of the three patients who died.

Results

Clinical Data

A majority of the patients (11/17) presented in the newborn period with cyanosis and cardiomegaly. Three patients between 1 and 12 months of age and three over one year of age were referred because of cardiomegaly on chest roentgenogram and/or a heart murmur heard on routine examination.

Of the 14 patients currently being followed, nine have no symptoms and five have mild exercise intolerance. Nine patients are acyanotic, three have mild to moderate cyanosis and two have severe cyanosis (table 1). Three patients who are currently 10, 11, and 19 months of age presented as newborns. They each had clearing of their cyanosis during the newborn period and are currently doing well without symptoms or cyanosis. Two of four patients between 6 and 10

From the Pediatric Cardiology Division of Children’s Hospital of Pittsburgh and the Department of Pediatrics of the University of Pittsburgh School of Medicine, Pittsburgh, Pennsylvania.

Supported in part by a grant from the Beaver County (PA) Heart Association.

This investigation was done during the tenure of Dr. Neches as an American Heart Association Teaching Scholar.

Address for reprints: William H. Neches, M.D., Cardiology Division, Children’s Hospital of Pittsburgh, 125 De Soto Street, Pittsburgh, Pennsylvania 15213.

Received July 7, 1977; revision accepted October 3, 1977.
years of age have mild exercise intolerance. Three of these four have mild cyanosis and one had severe cyanosis. Four patients over 15 years of age are mildly symptomatic (one with severe cyanosis and two without cyanosis).

Three of the 17 patients have a history of supraventricular tachycardia (SVT) without evidence of Wolff-Parkinson-White syndrome (WPW) on electrocardiogram. One patient with WPW, Type B, who had no history of SVT died suddenly after minor trauma. Two patients have sinus rhythm with occasional sinus arrest and junctional escape beats. One patient developed complete A-V block after tricuspid valvuloplasty and plication of the atrialized portion of the right ventricle.

Cardiac Catheterization Data

Analysis of oxygen saturation data obtained at cardiac catheterization showed systemic arterial oxygen saturations over 90% in 11 of the 14 patients who were beyond the newborn period (table 2). Two patients had oxygen saturation between 80 and 89% and one had an oxygen saturation less than 80%.

Current hematocrit values in three patients are less than 40%. Eight have hematocrits of 40–44% which could be considered in the normal range but are unusual for the population of patients seen in this center. Five patients have hematocrits of 45% or more. Thus, 13 of 16 patients have either high normal or elevated hematocrits despite the absence of cyanosis at rest in the majority of patients.

Right ventricular systolic pressure was normal (20–25 mm Hg) in 16 patients. One patient had valvular pulmonic stenosis and an elevated right ventricular pressure of 45 mm Hg. Mean right atrial pressure was less than 5 mm Hg in 12 patients and between 5 and 8 mm Hg in five patients.

Thirteen of the 17 patients had LV catheterization and cineangiography. Left ventricular pressures (systolic and end-diastolic) were normal in all patients. However, a number of patients had abnormalities of the LV seen on cineangiography. These consisted of abnormality of contour and/or contractility with or without associated

<table>
<thead>
<tr>
<th>Current age</th>
<th>N</th>
<th>Symptoms</th>
<th>Cyanosis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>None</td>
<td>Mild</td>
</tr>
<tr>
<td>&lt;1 yr</td>
<td>2</td>
<td>2</td>
<td>—</td>
</tr>
<tr>
<td>1-5 yr</td>
<td>1</td>
<td>1</td>
<td>—</td>
</tr>
<tr>
<td>6-10 yr</td>
<td>4</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>11-15 yr</td>
<td>4</td>
<td>4</td>
<td>—</td>
</tr>
<tr>
<td>&gt;15 yr</td>
<td>3</td>
<td>—</td>
<td>3</td>
</tr>
<tr>
<td>Total*</td>
<td>14</td>
<td>9</td>
<td>5</td>
</tr>
</tbody>
</table>

*3 additional patients died: 1 newborn, 1 11-yr-old, and 1 15-yr-old. The two older patients had mild cyanosis and symptoms.

TABLE 1. Ebstein's Malformation—Current Status

Figure 1. Left ventricular cineangiogram in systole (A), diastole (B) and a composite tracing of the systolic-diastolic configuration (C). There is asymmetry of ventricular septal and lateral wall contraction and the overall contour is irregular. Systolic prolapse of the mitral valve is seen (arrow).
prolapsing mitral valve. Nine of the 13 patients had abnormal contour of the LV with an unusually elongated and distorted long axis and irregularity of the septal and lateral walls (figs. 1 and 2). All nine patients with abnormal contour of the LV also had abnormal contraction. Twelve of the 13 patients had an abnormal LV contraction pattern. Three of these had a left ventricle that was both dyskinetic and hypokinetic. The other nine patients had either hypokinetic or dyskinetic ventricular contraction. Five of the 13 patients had moderate to severe prolapse of the mitral valve and each had an abnormally functioning LV with hypokinesia and/or dyskinesia (figs. 1 and 2).

Echocardiographic Data

Nine of the patients in this series have had echocardiography performed. All patients have characteristic findings previously described in Ebstein's anomaly of the tricuspid valve.44 One patient had evidence of left ventricular dysfunction with diminished left ventricular posterior wall motion. No patient with cineangiocardiographic evidence of a prolapsing mitral valve had an echocardiogram diagnostic of mitral valve prolapse.

Autopsy Data

Three patients died during the study period. A newborn infant died 12 hours after cardiac catheterization with severe metabolic acidosis and cardiovascular collapse. Postmortem examination was not permitted. An 11-year-old black male

![Figure 2. Left ventricular cineangiographic diagram in systole (A), diastole (B) and a composite tracing of the ventricular contraction pattern (C). The long axis of the left ventricle is angulated and there is irregularity of lateral wall and septum. Asymmetry of the left ventricular contraction pattern is seen. Systolic prolapse of the mitral valve is present (arrow).](image-url)
died suddenly two hours after minor head trauma. He was known to have Type B WPW but had no history of SVT. Cineangiocardiology had shown a hypokinetic and dyskinetic LV with an abnormal contour. At autopsy the left ventricle was grossly abnormal. The interventricular septum was deformed and bulged into the left ventricle. The mitral valve apparatus appeared normal. No cause of death was found. These findings were also seen in the third patient, a 15-year-old boy with only minimal cyanosis and exercise intolerance, who died suddenly. In addition to the septal deformity, this latter patient also had an irregularly distorted lateral LV wall. In both patients there was a moderately severe degree of downward displacement of the tricuspid valve into the right ventricle. Microscopic examination of many different areas of myocardium of both patients did not reveal any histologic abnormality.

Discussion

Abnormalities of the left side of the heart in association with Ebstein's anomaly of the tricuspid valve have not been well documented. Mitral valve prolapse has been described in patients with various forms of underlying cardiac disease, including Marfan's syndrome, atrial septal defect, hypertrophic cardiomyopathy, ischemic and rheumatic heart disease.7-12 Roberts et al.19 recently reported prolapse of the mitral valve in two patients with Ebstein's anomaly. However, no abnormality of LV function or structure has previously been described in patients with this disorder.

In all probability, a major reason for the absence of previous reports of abnormalities of the left side of the heart in EBS was that these anomalies are not evident by clinical examination and thus no attempts were made to study the left ventricle. In recent years catheterization of the left side of the heart with selective LV cineangiography has been routine in most patients studied at Children's Hospital of Pittsburgh. As a result, abnormalities such as those presented in this series have been discovered even in the absence of clinical signs. On retrospective clinical evaluation of the patients with EBS in this series, no clinical signs of left ventricular dysfunction were evident. The echocardiogram did not prove helpful in detecting the left ventricular or mitral valve abnormalities that had been seen on angiocardiography. Also, right ventricular cineangiocardiology did not adequately demonstrate the LV dysfunction or mitral valve abnormality on levophase of cineangiograms.

The data from this series have demonstrated that a significant number of patients with EBS have some degree of abnormality of the left side of the heart consisting of a prolapsing mitral valve and/or LV dysfunction. Since these abnormalities were not detected by clinical means or diagnostic noninvasive studies such as echocardiography, selective left ventricular angiography is essential in the evaluation of patients with this disorder. The cause of the angiocardiographic LV abnormalities seen in this series is unknown. These abnormalities may represent a form of generalized myocardial disorder although this hypothesis is not supported by any histologic evidence of pathology. Further evaluation of these unusual left ventricular abnormalities may, in the future, provide additional insight into the problems of arrhythmia and sudden death that have previously been described in patients with EBS.

References

Left ventricular anomalies associated with Ebstein's malformation of the tricuspid valve.
A A Monibi, W H Neches, C C Lenox, S C Park, R A Mathews and J R Zuberbuhler

Circulation. 1978;57:303-306
doi: 10.1161/01.CIR.57.2.303
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1978 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/57/2/303