Clinical Study of Twenty-Three Cases of Ebstein's Anomaly of the Tricuspid Valve

By Gerold L. Schiebler, M.D., Paul Adams, Jr., M.D., Ray C. Anderson, Ph.D., M.D., Kurt Amplatz, M.D., and Richard G. Lester, M.D.

The clinical findings in 23 cases of Ebstein's anomaly are presented, together with a discussion of the diagnostic and surgical aspects of this defect. Attention is again drawn to the frequent occurrence of the Wolff-Parkinson-White electrocardiographic pattern in this anomaly.

In 1937 Yater and Shapiro reported a case of Ebstein's anomaly of the tricuspid valve from this hospital. No further cases were recognized by our group until 1951, when we identified case 18 in the present series on the basis of Taussig's description of this anomaly. Since then, we have come to recognize this defect with increasing frequency, and have now studied a total of 23 cases apart from the case of Yater and Shapiro. Eight of these have had postmortem studies. All patients were examined by the staff of the University of Minnesota Hospitals.

As originally described almost a century ago, the sine qua non of this valvular malformation is that a portion or all of the leaflets of the tricuspid valve are displaced downward from the anatomic annulus separating the right atrium and the right ventricle. This defect has become a well-recognized clinical entity. Our case material covers a wide spectrum of clinical symptomatology and anatomic variety, thereby allowing a fairly comprehensive treatment of the subject.

Incidence

The world literature now contains more than 140 cases of Ebstein's malformation, and the rapidity with which new cases are being reported indicates that this anomaly is not uncommon. Maude Abbott's review of 1,000 autopsied congenital heart lesions contained at least 1 case. Keith and co-workers suggested an incidence figure of 1:210,000 live births, and found this defect to constitute less than 1 per cent of all congenital heart defects.

Sex Ratio

Our series consists of 10 females and 13 males, supporting the current concept that there is an equal sex ratio in this entity.

Prenatal History

Information on this subject was available in 19 of the 23 cases. In 12 of these, the pregnancy was "uneventful" or "uncomplicated." The other 7 reported the following incidents during pregnancy: nausea and vomiting in the first trimester with transient bleeding in the sixth month (case 2), marked hypertension and premature labor preceded by hemorrhage (case 3), exposure to mumps during the seventh month of gestation (case 4), hyperemesis gravidarum during the entire gestational period (case 8), transient uterine bleeding in the third month (case 9), malaria during the sixth week (case 10), and exposure to measles during the third month with a "few red spots on the arms" (case 12). In the absence of control data, it is not possible to evaluate the etiologic importance of these incidents.

Family History

There have been no reports in the literature of multiple cases of Ebstein's anomaly in the same family. Grob et al. stated in their case report that "a cousin of the child's mother" had acyanotic congenital heart disease. Blount et al. and Bayer et al. reported cases that had a normal nonidentical twin. Blacket et al.

From the Departments of Pediatrics and Radiology, University of Minnesota Hospitals, Minneapolis, Minn.

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Table 1.—Symptoms in Patients Surviving the Neonatal Period (18 patients)

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Number</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dyspnea</td>
<td>18</td>
<td>None with orthostatic dyspnea²⁷</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>15</td>
<td>One of these (case 2) had a closed foramen ovale</td>
</tr>
<tr>
<td>Excessive fatigue</td>
<td>13</td>
<td></td>
</tr>
<tr>
<td>Neurologic symptoms</td>
<td>11</td>
<td>Syncope in 5 (2 with exertion, 2 with paroxysmal tachycardia, and 1 at rest); thromboembolic phenomena in 2; febrile convulsions in 3; &quot;dizzy spells&quot; in 1; transient visual loss in 1</td>
</tr>
<tr>
<td>Cardiac failure</td>
<td>9</td>
<td>Later or terminally in 6; with paroxysmal tachycardia in 2; with nephrosis in 1</td>
</tr>
<tr>
<td>Precordial or epigastric pain</td>
<td>7</td>
<td>With exertion or emotional stress in 4, and with paroxysmal tachycardia in 3</td>
</tr>
<tr>
<td>Paroxysms of tachycardia</td>
<td>7</td>
<td>Between paroxysms 4 had sinoatrial rhythm; 3 had the W-P-W pattern. Case 5 had paroxysm of tachycardia at catheterization only</td>
</tr>
<tr>
<td>Squatting</td>
<td>5</td>
<td>With exertion in 4; with paroxysmal tachycardia in 1</td>
</tr>
<tr>
<td>Headaches</td>
<td>5</td>
<td>All with emotion or exertion</td>
</tr>
<tr>
<td>Protracted cough</td>
<td>4</td>
<td>In 3 after a bout of exertion</td>
</tr>
<tr>
<td>Photophobia</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Failure to gain weight</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

reported a patient whose older brother was known to have heart disease and died suddenly at the age of 17 while playing tennis.

Detailed family information was available in 22 of our cases. There were no cases of congenital heart disease among the 44 parents and 53 siblings. One patient (case 7) had a maternal first cousin, once removed, who was shown to have a ventricular septal defect by cardiac catheterization. Otherwise, there were no known relatives with congenital heart disease. None of the cases was a member of twin pairs.

There appeared to be no significant relationship to birth order, birth weight (none was premature by weight), age of mother, or month of birth (though 14 were born in the 6 month period from September to February, 5 being in December). However, the series is so small that these features cannot be properly evaluated.

**Neonatal Symptoms and Signs**

Twelve of our cases had symptoms or signs of congenital heart disease during the neonatal period.

Seven of our 8 autopsied cases fell into this category. In 5, a murmur and cyanosis were noted at birth or shortly thereafter, 1 (case 1) dying in 26 hours of severe cardiac failure. Another showed only a murmur, and the last displayed cyanosis and "sluggishness" at birth and a paroxysm of supraventricular tachycardia at several weeks of age.

Five of our living patients (data available on the neonatal period in 13) had either a murmur or cyanosis or both during the neonatal period. In all, the neonatal cyanosis decreased or disappeared with the passage of time, but recurred later in life.

Numerous authors⁵⁻²⁶ have described cases with Ebstein's defect with neonatal difficulties. Five cases dying in the first few days of life have been reported.,⁺¹⁺⁴,⁺²¹,⁺²⁵ The cyanosis at birth and its gradual disappearance have been noted by Engle et al.²² and Kilby et al.¹³ This transient cyanosis may represent a right-to-left shunt through the foramen ovale secondary to normal high pulmonary bed resistance in the presence of a poorly functioning right ventricle. The cyanosis may then disappear or decrease with the normal drop in pulmonary resistance in early infancy. In addition, the normal decrease in hemoglobin level after birth may diminish the degree of cyanosis, so that the improvement may be more apparent than real. Unrecognized bouts of paroxysmal supraventricular tachycardia with spontaneous reversion may account for some cases of transient cyanosis.

**Symptoms after the Neonatal Period**

Twenty-two patients survived the first month of life. Four of these (ranging in age from 4 to 11 years, cases 9, 10, 11, and 15) are
currently asymptomatic. Of the remaining 18 patients, 11 had symptoms in the neonatal period; in the others the onset of symptomatology was vague and indefinite. In case 23 symptoms first developed at 30 years of age. As shown in table 1, dyspnea, cyanosis, and excessive fatigue were the most common symptoms.

**Physical Findings**

*Physique.* The values for height and weight were plotted on the Iowa grid, with serial measurements available in most of the children. Six patients were above the 55th percentile for both height and weight; 5 were below the 16th percentile; and 8 were between these limits (table 2). Thus, as a group, patients with this cardiac malformation show normal growth features. This is particularly true if cyanosis is absent, since none of the acyanotic patients was below the 16th percentile. Patients seen initially as adults had a normal weight for their age and height.

*Skin Color.* Seven patients have shown no cyanosis of the skin or mucous membranes at any time. One patient (case 2) showed cyanosis only terminally, 3 had cyanosis only with exertion, and 12 had cyanosis at rest. All living patients who had cyanosis in the neonatal period improved transiently, but now have cyanosis at rest or with exertion. As Engle et al.22 reported originally, cyanosis is not closely correlated with exercise tolerance. Thus, 2 of our severely cyanotic adults do manual labor without difficulty, while 1 of our acyanotic adults (case 23) has definitely decreased exercise tolerance.

Unusual facial colorations such as "violaceous hue,"17, 8, 12 "flushed,"17 "florid,"9, 28 and "red-cheeked"9, 29 have been described in patients with Ebstein’s anomaly. Similar observations have been made in our cases, often prior to the establishment of the correct diagnosis. In our 23 cases, 10 showed a peculiar facial coloration, which may have diagnostic significance somewhat akin to the "malar flush" of mitral stenosis.

Clubbing of the fingers was present in 6 patients, all cyanotic.

**Table 2.—Physical Findings in Ebstein’s Anomaly—Twenty-Three Cases**

<table>
<thead>
<tr>
<th>Finding</th>
<th>Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systolic murmur</td>
<td>23</td>
</tr>
<tr>
<td>P₂ equal or greater than A₂</td>
<td>23</td>
</tr>
<tr>
<td>Lungs clear to auscultation</td>
<td>23</td>
</tr>
<tr>
<td>Cardiomegaly</td>
<td>21</td>
</tr>
<tr>
<td>Normal physique</td>
<td>18</td>
</tr>
<tr>
<td>&quot;Triple&quot; or &quot;quadruple&quot; rhythm</td>
<td>18</td>
</tr>
<tr>
<td>Cyanosis</td>
<td>16 (13 at rest, 3 only with exertion)</td>
</tr>
<tr>
<td>Systolic thrill</td>
<td>15</td>
</tr>
<tr>
<td>Diffuse point of maximal impulse</td>
<td>13</td>
</tr>
<tr>
<td>Diastolic murmur</td>
<td>11</td>
</tr>
<tr>
<td>&quot;‘Split’&quot; first sound</td>
<td>11 (Some had a systolic ejection &quot;‘click’&quot;)</td>
</tr>
<tr>
<td>&quot;‘Metalliec’&quot; or &quot;‘clicking’&quot; heart tones</td>
<td>10</td>
</tr>
<tr>
<td>&quot;‘Facial erythema’&quot;</td>
<td>10</td>
</tr>
<tr>
<td>Cardiac failure</td>
<td>9</td>
</tr>
<tr>
<td>Normal chest contour</td>
<td>9</td>
</tr>
<tr>
<td>Dyspnea at rest</td>
<td>8 (6 terminal, 2 with paroxysms of tachycardia)</td>
</tr>
<tr>
<td>Localized point of maximal impulse</td>
<td>6</td>
</tr>
<tr>
<td>Clubbing</td>
<td>6</td>
</tr>
<tr>
<td>Left chest prominence</td>
<td>6</td>
</tr>
<tr>
<td>Bilateral chest prominence</td>
<td>5</td>
</tr>
<tr>
<td>Visible thrust of chest</td>
<td>5</td>
</tr>
<tr>
<td>Observed paroxysms of tachycardia</td>
<td>(2 at catheterization)</td>
</tr>
<tr>
<td>&quot;‘Fair’&quot; or &quot;‘weak’&quot; peripheral pulses</td>
<td>5</td>
</tr>
<tr>
<td>Pericardial friction rubs</td>
<td>3</td>
</tr>
</tbody>
</table>

*Chest Shape and Auscultation of the Lungs.* Information on chest shape is available in 21 of our 23 cases. Nine had a normal chest contour. Five were considered to have an increase in the anteroposterior diameter with bilateral chest prominence; 6 were reported to have left chest prominence; and 1, right chest prominence. Acyanotic patients invariably had a normal chest contour.

In no case, even when cardiac failure was present, were rales heard in the chest.
Peripheral Vessels. The peripheral vessels were usually described as normal, although in 5 of the cyanotic patients the pulses were noted to be "fair" or "weak." Blood pressures in the arms and legs were found to be within normal limits, except in 1 adult (case 20) whose systolic arm pressure was 160 mm. Hg. In measuring the blood pressure, the diastolic end point was sometimes difficult to ascertain, particularly if the patient was cyanotic or in heart failure. Similar observations have been made by Blacket et al.\(^9\) and Kilby et al.\(^{13}\)

In general, no specific observations were made regarding the neck veins. There was a definite presystolic pulsation of the neck vessels associated with tricuspid insufficiency in case 7, who was in severe congestive failure. However, pulsations of the liver were not noted. In 1 of our asymptomatic patients (case 9), a phlebogram showed evidence of tricuspid insufficiency.

Cardiac Findings. Five of the patients had a visible thrust of the precordium. The point of maximal impulse was described in 19 cases. In 13 it was "diffuse," but in the others it was "localized." These observations could not be correlated with heart size.

A systolic thrill was noted in 15 of the 23 cases at the lower left sternal border or at the "apex." In several cases the thrill was maximum over the displaced tricuspid valve as determined at cardiac catheterization. We agree with Blount et al.\(^3\) that the "apex" represents the location of the tricuspid valve in an enlarged heart with leftward displacement of the valve. No diastolic thrills were noted. The intensity of the systolic thrill was poorly correlated with heart size, but was more prominent in the presence of cardiac failure. During open heart surgery in 2 patients (cases 6 and 7), the thrill was felt over the huge right atrium during ventricular systole. Tricuspid valvular insufficiency was identified by direct observation.

The heart rate varied normally with age, except during episodes of paroxysmal tachycardia. In only 3 cases was marked sinus arrhythmia noted (cases 10, 19, and 23).

The heart sounds were variable. Soft and diminished heart sounds have been emphasized to be of diagnostic significance.\(^9,22\) We have found this feature only in a minority of cases, the heart sounds usually being of normal intensity. "Metallic" or "clicking" heart tones were described in 10 patients. The "triple" or "gallop" rhythm so frequently described in the literature was noted in 18 of our 23 cases, and in 6 of these the rhythm was recorded as "quadruple." The triple rhythm is produced by a low frequency early to mid-diastolic third heart sound or soft murmur, usually maximal along the lower left sternal border or the "apex." The "quadruple" rhythm is caused by an additional presystolic sound or murmur.

The first sound was noted to be "split" in 11 of the patients. Characteristically, the second portion of the split first sound was louder than the first portion, suggestive to some observers of a systolic ejection click. Also, the widely split first sound plus a second and third heart sound led to the occasional description of a "quadruple" rhythm. In children the second sound over the pulmonic area was usually described as "split" and louder than over the aortic area. In adults, the second sound over the pulmonic area was usually noted to be pure and equal in intensity to that over the aortic area. In no case was the second sound described as being diminished over the pulmonic area.

All our cases had a systolic murmur, although in 1 patient (case 2) it was not noted until 16 months of age. The maximal intensity of the murmur was always noted to be at the lower left sternal border, or from there to the "apex." Again, we think that the location of the murmur depends upon the location of the tricuspid valve. The majority of the murmurs were of grade II to III intensity, but varied from a barely audible grade I murmur to the grade V murmur. The intensity had no close relationship to heart size, but appeared to increase with cardiac failure. The quality of the murmur was also variable, only rarely being described as "coarse" or "scratchy."
As previously noted, 18 of our cases had diastolic sounds. Eleven of these represented a discrete murmur, varying from protodiastolic to presystolic in time, and variable also in intensity and quality. The usual location and greatest intensity were at the lower left sternal border or just to the left at the "apex." These diastolic sounds and murmurs were usually accentuated in the inspiratory phase of respiration, thus suggesting a tricuspid valve origin.

A to-and-fro murmur was described in 3 of our cases, as well as by others. One of these (case 17) is still living; in the other 2 patients (cases 4 and 7) postmortem examination showed pericardial effusion and fibrinous pericarditis. This combination of pericarditis and pericardial effusion, often bloody, has been noted by others, and has been attributed to extravasation of blood from the coronary veins because of increased venous pressure. On the basis of these observations, we believe that the to-and-fro murmurs in our cases were actually pericardial friction rubs.

Routine Laboratory Findings

The hemoglobin, erythrocyte count, and hematocrit were elevated in the cyanotic cases. Albuminuria was noted in individuals with cardiac failure and in the single patient in whom typical nephrosis was associated with Ebstein’s anomaly. Otherwise, routine laboratory studies were normal.

Roentgenographic Findings

We have found considerable variation in the cardiac contour on roentgenograms in patients with Ebstein’s anomaly (figs. 1 and 2). In 4 instances we have obtained roentgenograms within the first few days of life. In case 1 the heart was markedly enlarged at 5 hours of age, while in the others (cases 2, 3, and 12) the heart contour was within normal limits.

In the majority of our cases the cardiac contour could be loosely grouped into 2 types. One group showed moderate cardiomegaly and a contour simulating left ventricular enlargement in both the anteroposterior and left oblique views. Several of these were at first mistakenly diagnosed as mild aortic stenosis. The other group showed marked cardiomegaly and a boxlike contour. When this contour is present, it is highly diagnostic of Ebstein’s defect; it is due to an almost horizontal take-off of the left border of the heart near the aorta, sometimes extending virtually to the left chest border. Right atrial enlargement, leading to enlargement of the upper right border, may also contribute to this squarish appearance. The bulging of the upper left heart border is probably due primarily to the displacement of the infundibulum of the right ventricle by the markedly enlarged right atrium, and to a lesser degree by dilatation of the outflow tract of the right ventricle. This same contour may be encountered in patients with valvular pulmonic stenosis and cardiac failure, rheumatic heart disease, or total anomalous pulmonary venous connection to the right atrium.

The contours in some of our other cases resembled those seen in tetralogy of Fallot, tricuspid atresia, pulmonary atresia, or isolated pulmonary valvular stenosis.

In case 2 no definite cardiomegaly occurred prior to her death with nephrosis at 2\textfrac{1}{2} years. Individuals with this malformation having a normal-sized heart have been reported by several writers.

The pulmonary vascular markings were always normal or decreased. A close correlation exists between the degree of cyanosis and the decrease in the vascular markings. Even in cases with cardiac failure there was no increase in pulmonary vascular markings and no evidence of pleural effusion. The main pulmonary artery segment was often difficult to identify, and the aortic knob generally appeared small. However, both may be totally or partially obscured by the right-sided chamber enlargement. None of our cases showed roentgenographic evidence of left atrial enlargement, a very helpful sign in the exclusion of mitral valve lesions. Calcifications in the atrioventricular ring may appear both in Ebstein’s defect and in rheumatic heart disease.
Fig. 1 Top. Tracings of chest roentgenograms of autopsied cases to show cardiac contour and pulmonary vasculature. Numbers refer to case numbers.

Fig. 2 Bottom. Tracings of chest roentgenograms of living patients to show cardiac contour and pulmonary vasculature.
CLINICAL STUDY OF EBSTEIN’S ANOMALY OF TRICUSPID VALVE

Kymography showed the pulsations of the right side of the heart to be increased in some cases, and normal or decreased in others. They were not consistent enough to be of much diagnostic aid.

A more detailed report of the roentgenographic aspect of Ebstein’s anomaly is being published separately. 36

ANGIOCARDIOGRAPHY

Angiocardiography was performed here in cases 14 and 19 and elsewhere in case 6. Though our experience is limited, we agree with Kjellberg et al. 37 that peripheral angiocardiography is probably of very limited value, inasmuch as the contrast medium is so diluted in the large right atrium that anatomic details are difficult to visualize. Also, its potential danger to the patient is undoubtedly outweighs its diagnostic usefulness. On the other hand, selective angiocardiography performed from the right ventricle might be useful in delineating the anatomic features of the right ventricle, but we have not done such a procedure in any of our cases.

ELECTROCARDIOGRAPHIC FINDINGS

A. Without Wolff-Parkinson-White Pattern (Table 3)

Seventeen of our patients did not show the Wolff-Parkinson-White (W-P-W) pattern at any time, while 1 was variable in this respect. We have carried out a separate, detailed analysis of the electrocardiograms in these 18 cases (8 of whom had 2 or more tracings). All subsequent comparisons for children are stated in relation to Ziegler’s normal values.

The QRS axis was calculated on the basis of both area and voltage. The highly notched, slurred, and splintered QRS complexes make axis determination very difficult. The axis based on voltage is probably not meaningful, but that based on area may have greater significance. In our cases, the axis was usually right (including S1S2S3 patterns), but several cases showed either a normal or left axis.

The rhythm was usually sinoatrial, except for occasional atrioventricular nodal rhythm in case 19; and whenever sinoatrial bradycardia (under 60) was present, cases 8 and 13 developed atrioventricular dissociation. Only occasional nodal or ventricular premature beats were noted.

Six of the children showed a prolonged P-R interval. None of the children under 16 had a consistently normal QRS pattern during the course of the clinical observation; 2 children (cases 2 and 11) were initially normal and progressed to an incomplete right bundle-branch block. Another (case 9) progressed from an incomplete to a complete right bundle-branch block. Of the 5 patients over 16 years of age, 4 showed right bundle-branch block, and the other had a normal QRS with sinoatrial rhythm and complete right bundle-branch block when nodal rhythm supervened. The right bundle-branch block configuration was frequently atypical.

The uncorrected Q-T interval was normal in all but 3 patients. Case 1 had a prolonged Q-T for age and rate; cases 6 and 17 developed a prolonged Q-T interval as demonstrated by serial electrocardiograms. These latter 2 cases showed gradual prolongation of the P-R interval and the QRS duration, so that the prolonged Q-T may have been due to the long QRS.

Of the 18 cases, 14 showed maximum or higher than normal P waves, the peaking always occurring in V2 and V3 if present at all. There was prominent notching or slurring of the P wave in only 4 cases.

The duration of the P wave was increased in 3 adults and 5 children. Van Lingren and Baversfeld 39 postulated that the increase in the P-R interval was caused by an increase in the duration of the P wave, and that the P-R interval per se was not increased. Only 3 of our 6 cases with a prolonged P-R interval, however, had a P wave of abnormal duration.

The 4 children showing both an increase in height and duration of the P wave have all died, while the 2 adults with these features are living. The 4 patients who have normal P waves in all leads are still living and relatively asymptomatic. Those 3 patients whose only abnormality is an increased height of the P wave in the precordial leads are only mildly symptomatic. Patients showing abnormally
Table 3.—Electrocardiograms without W-P-W Pattern

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Voltage</th>
<th>Area</th>
<th>Rate</th>
<th>Rhythm</th>
<th>P-R interval (ms)</th>
<th>QRS lead II</th>
<th>Height of P in I, II, III</th>
<th>Max. duration of P waves in leads</th>
<th>Height of P in V1</th>
<th>V1 pattern</th>
<th>Highest R/S in V1</th>
<th>Intrinsicoid deflection of V1</th>
<th>V2 pattern</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>5 br.</td>
<td>+100</td>
<td>+110</td>
<td>140</td>
<td>Sinus</td>
<td>0.14</td>
<td>0.11</td>
<td>3.0 0.07 3.0</td>
<td>rsR'</td>
<td>8/4</td>
<td>0.08</td>
<td>RS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>1 mo.</td>
<td>+180</td>
<td>+170</td>
<td>300</td>
<td>SVT</td>
<td>—</td>
<td>0.04</td>
<td>— — —</td>
<td>rS</td>
<td>1.5/6</td>
<td>0.025</td>
<td>rs</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5 mo.</td>
<td>—150</td>
<td>+150</td>
<td>145</td>
<td>Sinus</td>
<td>0.12</td>
<td>0.07</td>
<td>3.5 0.07</td>
<td>3.0</td>
<td>rS</td>
<td>5/8</td>
<td>0.015</td>
<td>RS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 yr.</td>
<td>—110</td>
<td>+110</td>
<td>115</td>
<td>Sinus</td>
<td>0.14</td>
<td>0.09</td>
<td>4.0 0.07</td>
<td>4.0</td>
<td>rsR's'</td>
<td>1.5/1.8</td>
<td>0.04</td>
<td>qRS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>5 yr.</td>
<td>—150</td>
<td>—170</td>
<td>107</td>
<td>Sinus</td>
<td>0.17</td>
<td>0.11</td>
<td>2.5 0.10</td>
<td>4.0</td>
<td>rsR'</td>
<td>3.3/2.2</td>
<td>0.08</td>
<td>rsR's'</td>
<td></td>
</tr>
<tr>
<td>10 yr.</td>
<td>+180</td>
<td>+180</td>
<td>110</td>
<td>Sinus</td>
<td>0.22</td>
<td>0.13</td>
<td>4.5 0.09</td>
<td>3.5</td>
<td>r</td>
<td>3/0</td>
<td>0.09</td>
<td>rsR'S'</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>9 yr.</td>
<td>+100</td>
<td>+120</td>
<td>130</td>
<td>Sinus</td>
<td>0.16</td>
<td>0.10</td>
<td>3.5 0.08</td>
<td>6.0</td>
<td>rsR's'</td>
<td>2.5/1</td>
<td>0.06</td>
<td>RS</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>6 yr.</td>
<td>—45</td>
<td>—140</td>
<td>115</td>
<td>Sinus</td>
<td>0.20</td>
<td>0.12</td>
<td>5.0 0.09</td>
<td>4.0</td>
<td>rsR'</td>
<td>5/3.5</td>
<td>0.10</td>
<td>qRS</td>
<td></td>
</tr>
<tr>
<td>13 yr.</td>
<td>—100</td>
<td>—135</td>
<td>95</td>
<td>Sinus</td>
<td>0.22</td>
<td>0.18</td>
<td>7.5 0.11</td>
<td>5.0</td>
<td>rsR'</td>
<td>6/2.8</td>
<td>0.12</td>
<td>qRS</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>14 yr.</td>
<td>+120</td>
<td>+140</td>
<td>115</td>
<td>Sinus</td>
<td>0.20</td>
<td>0.15</td>
<td>4.5 0.12</td>
<td>5.0</td>
<td>rsR'</td>
<td>3.5/0.8</td>
<td>0.12</td>
<td>rsR'S'</td>
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<tr>
<td>8</td>
<td>13 yr.</td>
<td>+30</td>
<td>+105</td>
<td>70</td>
<td>Sinus or</td>
<td>0.18</td>
<td>0.13</td>
<td>3.0 0.10</td>
<td>4.0</td>
<td>rsR'</td>
<td>7/3</td>
<td>0.075</td>
<td>qRs</td>
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A-V dissociation

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Voltage</th>
<th>Area</th>
<th>Rate</th>
<th>Rhythm</th>
<th>P-R interval (ms)</th>
<th>QRS lead II</th>
<th>Height of P in I, II, III</th>
<th>Max. duration of P waves in leads</th>
<th>Height of P in V1</th>
<th>V1 pattern</th>
<th>Highest R/S in V1</th>
<th>Intrinsicoid deflection of V1</th>
<th>V2 pattern</th>
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<tbody>
<tr>
<td>9</td>
<td>11 mo.</td>
<td>+75</td>
<td>+100</td>
<td>120</td>
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<td>0.13</td>
<td>0.09</td>
<td>1.5 0.08</td>
<td>1.5</td>
<td>rsR'</td>
<td>7/2.5</td>
<td>0.06</td>
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<tr>
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<td>+85</td>
<td>85</td>
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<td>0.10</td>
<td>0.5 0.08</td>
<td>1.0</td>
<td>rsR's'</td>
<td>5/5</td>
<td>0.06</td>
<td>qRs</td>
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</tr>
<tr>
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<td>2 yr.</td>
<td>+110</td>
<td>+130</td>
<td>130</td>
<td>Sinus</td>
<td>0.16</td>
<td>0.08</td>
<td>2.5 0.08</td>
<td>3.0</td>
<td>qrs</td>
<td>4.5/3</td>
<td>0.035</td>
<td>qRs</td>
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<tr>
<td>6 yr.</td>
<td>+110</td>
<td>+120</td>
<td>105</td>
<td>Sinus</td>
<td>0.18</td>
<td>0.09</td>
<td>1.4 0.09</td>
<td>3.5</td>
<td>qrS</td>
<td>2.3/5.5</td>
<td>0.035</td>
<td>qRs</td>
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<tr>
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<td>8 yr.</td>
<td>—40</td>
<td>—80</td>
<td>65</td>
<td>Sinus or</td>
<td>0.15</td>
<td>0.13</td>
<td>1.0 0.09</td>
<td>0.5</td>
<td>r</td>
<td>3.4/8.5</td>
<td>0.02</td>
<td>qRs</td>
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A-V dissociation

<table>
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<tr>
<th>Case no.</th>
<th>Age</th>
<th>Voltage</th>
<th>Area</th>
<th>Rate</th>
<th>Rhythm</th>
<th>P-R interval (ms)</th>
<th>QRS lead II</th>
<th>Height of P in I, II, III</th>
<th>Max. duration of P waves in leads</th>
<th>Height of P in V1</th>
<th>V1 pattern</th>
<th>Highest R/S in V1</th>
<th>Intrinsicoid deflection of V1</th>
<th>V2 pattern</th>
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<tbody>
<tr>
<td>15</td>
<td>9 yr.</td>
<td>—20</td>
<td>—150</td>
<td>75</td>
<td>Sinus</td>
<td>0.15</td>
<td>0.13</td>
<td>1.0 0.10</td>
<td>1.5</td>
<td>rsR'</td>
<td>7/5</td>
<td>0.07</td>
<td>qRs</td>
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<tr>
<td>16</td>
<td>6 yr.</td>
<td>—40</td>
<td>—20</td>
<td>100</td>
<td>Sinus</td>
<td>0.12</td>
<td>0.10</td>
<td>3.0 0.09</td>
<td>3.5</td>
<td>r</td>
<td>0.4/4</td>
<td>0.02</td>
<td>qRs</td>
<td></td>
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<tr>
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<td>—35</td>
<td>90</td>
<td>Sinus</td>
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<td>0.10</td>
<td>4.5 0.09</td>
<td>4.0</td>
<td>rsr'</td>
<td>0.4/4.5</td>
<td>0.10</td>
<td>RsR's'</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>6 yr.</td>
<td>—60</td>
<td>—145</td>
<td>100</td>
<td>Sinus</td>
<td>0.19</td>
<td>0.12</td>
<td>2.5 0.09</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td></td>
</tr>
<tr>
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<td>70</td>
<td>Sinus</td>
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<td>0.20</td>
<td>2.5 0.09</td>
<td>2.5</td>
<td>rsR's'</td>
<td>4/2.8</td>
<td>0.017</td>
<td>qRs</td>
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<td></td>
</tr>
<tr>
<td>19</td>
<td>15 yr.</td>
<td>+110</td>
<td>+110</td>
<td>85</td>
<td>Noval</td>
<td>—</td>
<td>0.13</td>
<td>— —</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
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<td></td>
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<tr>
<td>20 yr.</td>
<td>+105</td>
<td>+110</td>
<td>90</td>
<td>Sinus</td>
<td>0.21</td>
<td>0.09</td>
<td>3.0 0.11</td>
<td>2.5</td>
<td>rsR's'</td>
<td>2/2</td>
<td>0.055</td>
<td>RsR'</td>
<td></td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>28 yr.</td>
<td>+130</td>
<td>+140</td>
<td>95</td>
<td>Sinus</td>
<td>0.17</td>
<td>0.13</td>
<td>3.5 0.11</td>
<td>5.0</td>
<td>rsR's'</td>
<td>8/4</td>
<td>0.055</td>
<td>Rs</td>
<td></td>
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<tr>
<td>21</td>
<td>35 yr.</td>
<td>—30</td>
<td>+20</td>
<td>80</td>
<td>Sinus</td>
<td>0.20</td>
<td>0.14</td>
<td>1.5 0.09</td>
<td>2.0</td>
<td>rsr'</td>
<td>4/4</td>
<td>0.10</td>
<td>qRs</td>
<td></td>
</tr>
<tr>
<td>22</td>
<td>36 yr.</td>
<td>+150</td>
<td>+170</td>
<td>75</td>
<td>Sinus</td>
<td>0.21</td>
<td>0.13</td>
<td>1.5 0.11</td>
<td>4.0</td>
<td>rsr'</td>
<td>4/3</td>
<td>0.11</td>
<td>qRs</td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>35 yr.</td>
<td>+60</td>
<td>+45</td>
<td>80</td>
<td>Sinus</td>
<td>0.18</td>
<td>0.12</td>
<td>0.5 0.10</td>
<td>1.0</td>
<td>rsr'</td>
<td>3/0.8</td>
<td>0.07</td>
<td>qRs</td>
<td></td>
</tr>
</tbody>
</table>

*Maximum instantaneous voltage and area of QRS were used for 2 independent determinations.

†Intrinsicoid deflection in V1 0.07 second on tracing taken at the age of 9.

high P waves in 8 or more of the standard electrocardiographic leads tended to have more severe symptoms or early death.

Notching or slurring of the QRS complexes was present in 1 or more leads of all patients. Lead I generally showed an rs pattern, lead II usually a qRs pattern, and lead III a QR or QRS pattern. Van Lingens's observation, that an R plus S voltage less than 7 mm. in the standard leads was of diagnostic value in this entity, was confirmed in 4 of the 5 adult cases, but was present in only 6 of the 13 children. That these complexes may change with time was shown by serial electrocardiograms in which there was often a progressive decrease in the voltage and an increase in the amount of notching or slurring as the child grew older.
In V1 the pattern was usually rS or occasionally qRS, both with wide splintering. The tallest R wave encountered in lead V1 in all tracings was 8 mm. This observation agrees well with that of Keith et al. who found that the tallest R wave in V1 was 7 mm. or less except in 1 case where it was 9 mm. A pattern of right ventricular hypertrophy was not seen in V1 in any patient. However, right ventricular hypertrophy in this malformation has been reported by others. In all but case 1 the ventricular activation time (intrinsicoid deflection) was 0.04 second or greater in V1. However, in 2 patients the progression from normal to prolonged time was noted during the course of our observations.

In V6, about one half the tracings showed no q wave, probably due to the displacement of the right ventricle by the large dilated right atrium. In these, a q wave could be seen in leads taken further to the left (V7 to V9). In only 1 (case 15) was the intrinsicoid deflection delayed in V6 when a q wave was present. No left ventricular hypertrophy patterns were noted.

The ST-segment changes were nonspecific and nondiagnostic. Slight depression or elevation in leads II, III, aVF, slight elevation in V1, and slight depression in V6 were occasionally seen.

The T waves were normal, except that in leads II, III and aVF, the incidence of diphasic or negative T waves appeared to be somewhat greater than might be expected. This was probably largely positional artefact, caused by the huge right atrium and an enlarged heart. Seven patients had diphasic T waves in V5 and 2 of these in V6, probably for the same reason. This view is supported by the finding of normal T waves in leads V7 to V9 where the QRS complex is initiated with a q wave.

In summary, our findings are very similar to those reported previously, the typical features being (1) incomplete or complete right bundle-branch block, with widely splintered, notched, and slurred QRS complexes, (2) increased height and, on occasion, duration of the P wave, (3) low voltage in the right pre-

cordial leads, with the tallest R wave in V1 not exceeding 8 mm., and (4) right axis deviation. Also encountered were a prolonged P-R interval, nonspecific ST-segment changes, bouts of supraventricular tachycardia, atrioventricular dissociation, premature beats, and a normal or left axis.

Sodi-Pallares has stated that the diagnosis of this cardiac anomaly must be strongly considered if the electrocardiogram shows a QR pattern and negative T waves from V1 to V4 or V6, particularly if found in a young person with cyanosis. In our review, we have found no case fitting these criteria. One of our patients (case 11) on a single tracing has a QR pattern all across the precordium with negative T waves from V1 to V4, but she is not cyanotic. Two others, with cyanosis or oxygen desaturation, had a QR pattern in leads V4R or V1, with a negative T wave, but this pattern did not extend to V2. Thus in our experience, this pattern is not of diagnostic usefulness.

Michel et al. proposed that the broad R' deflection of low amplitude in leads II and III, following a normally shaped R wave, represented the delayed activation of the atrialized portion of the right ventricle. They suggested that these be called P' waves because of their similarity to the P waves. Only 3 of our patients (cases 4, 7, and 17), show a pattern similar to this description. In 2 of these, serial tracings clearly demonstrate that this pattern develops with time and with increase in the notching and slurring of the QRS complexes. Even though Michel et al. found this in 2 of their cases, and in 6 of 9 cases they reviewed from the literature, in our experience this finding has little diagnostic significance.

B. With Wolff-Parkinson-White Pattern (Table 4)

We have found the W-P-W electrocardiographic pattern in 6 of our 23 cases, and believe that this finding deserves special emphasis. Sodi-Pallares first drew attention to this unique association, and interpreted the electrocardiogram of Yater and Shapiro as being a W-P-W pattern. We have reviewed
the original tracing in this case, reportedly the first electrocardiogram published in this anomaly, and agree with his interpretation.

Two general groups of W-P-W pattern have been described, those with right axis (called type A by Sodi-Pallares\textsuperscript{41}), and those with left axis (type B). The left axis in these cases was present by both voltage and area analysis. In type B the complexes simulate left bundle-branch block—with QS complexes usually present in leads II, III, and aVF, and upright ventricular complexes in the left preordial leads.

In 5 of our 6 cases, the W-P-W pattern was present on all occasions (all patients having at least 2 tracings), while in 1 (case 6) this pattern was seen only once. On other occasions there was a prolonged P-R interval and right bundle-branch block (fig. 3).

Sodi-Pallares\textsuperscript{41} reported 3 cases of Ebstein’s anomaly with type B W-P-W patterns. Single cases of W-P-W patterns in this anomaly have been published by Lev et al.,\textsuperscript{23} Götzsche and Falholt,\textsuperscript{18} Gotshalk et al.,\textsuperscript{20} Vogt,\textsuperscript{11} Hetjmaniec and Herrmann,\textsuperscript{43} Hernandez et al.,\textsuperscript{15} and Mahaim and van Nuenhuizen.\textsuperscript{26} The cases reported by Bruce, Yu, et al.\textsuperscript{44, 45} and Yim and Yu\textsuperscript{46} also appear to fall into this group.

In several of these cases the electrocardiogram showed normal rather than left axis, and Vogt’s\textsuperscript{31} case alternated her W-P-W pattern with a nodal rhythm on occasion. Kezdi and Wennemark\textsuperscript{34} have recorded a transient W-P-W pattern during the catheterization of the right ventricle in a patient with Ebstein’s defect.

The combination of type B W-P-W pattern and cyanotic congenital heart disease was considered to be diagnostic of Ebstein’s anomaly by Sodi-Pallares.\textsuperscript{41} However, Hetjmaniec and Herrmann\textsuperscript{43} recently described a case of transposition of the great vessels with W-P-W pattern. We have also seen an exceptional case of this type in which postmortem study showed complete transposition of the great vessels with patent foramen ovale. However, this electrocardiogram showed a normal axis.

On the other hand, the occurrence of type B W-P-W in a patient with cyanotic congenital heart disease and normal to decreased pulmonary vascular markings appears to be highly suggestive of Ebstein’s anomaly. Four of our 6 cases and that of Yater and Shapiro\textsuperscript{1} fall into this category. The other 2 remain acyanotic at ages 4 and 7, but may be expected to become cyanotic in the future. That this combination is not diagnostic is shown by Keith’s\textsuperscript{47} mention of a case of tricuspid atresia with the W-P-W pattern, and the postmortem study by de Oliveira et al.\textsuperscript{48} of a case
of tetralogy of Fallot with a variant of the type B pattern.

Three patients (6, 14, 18) of the 6 with W-P-W gave a history of recurrent attacks of tachycardia; in contrast, only 4 of the other 17 cases gave a similar history.

Tall peaked P waves ("P pulmonale") occurred in the right precordial leads in 5 of the 6 cases. There again appears to be a correlation between the height of the P wave and the clinical symptomatology; the tallest waves occurred in the 2 deceased patients, and the only normal P waves occurred in the lone asymptomatic patient (case 10) in this group.

The P wave was always positive in leads I, II, aV_l, aV_r, and V_2 to V_6, always negative in aV_R, and variable in the other leads.

All 6 cases showed type B W-P-W pattern, with tall R waves in lead I and deep S waves in lead III—up to 27 mm. The right precordial leads showed rS patterns with a depth up to 22 mm. There was decreasing height of R/S as increasing notching and slurring appeared with age. In the left precordial leads there was usually a high R wave, up to 34 mm. In the presence of type B W-P-W these observations cannot be interpreted as left ventricular hypertrophy or "strain."

Significantly, there were no normal tracings among the 70 or more electrocardiograms taken of our 23 patients.

Phonocardiography

Phonocardiograms were recorded in 12 of our 23 patients. In all cases the first sound was delayed (0.05 second or greater) after the onset of the QRS complex on the simultaneous electrocardiogram, as compared with the normal value of 0.02 to 0.04 second. The first sound was of normal intensity, though the second component of this sound was invariably louder than the first component, and was maximal at the apex or the lower left sternal border. It was not possible to measure the duration of the first sound inasmuch as the first sound regularly blended in with the succeeding medium frequency (100 to 600 cycles) systolic murmur. This murmur varied from a short decrescendo murmur in early and mid-
was loudest along the lower left sternal border or at the apex.

A discrete third heart sound or short mid-diastolic murmur was noted in 9 cases, usually best observed at the lower left sternal border. In another case (7) the area usually occupied by the third heart sound was filled by an early diastolic murmur. In the other 2 patients, both young children, neither a third heart sound nor a diastolic murmur was present. The third heart sound or mid-diastolic murmur was regularly of low frequency (90 to 150 cycles).

A presystolic extra sound was noted in 2 cases, and a presystolic murmur of varying type was present in 4 others. In the 1 case, in which we have serial phonocardiograms (case 18), there was a change from a presystolic extra sound to a definite presystolic murmur over a period of 6 years. In the other 5 cases, all children who are completely or relatively asymptomatic, no presystolic sound or murmur was present. This murmur or sound was regularly of medium frequency and maximal at the lower left sternal border or apex. A continuous systolic-diastolic vibration was noted in 3 cases (4, 6, 7), all of whom showed fibrinous pericarditis at postmortem examination.

In general our findings resemble those of Mayer, Nadas, and Ongley. They found the first sound to be delayed, of normal intensity, and maximal at the apex. The second sound was maximal at the left sternal border or apex in their 5 patients. The systolic murmur was described by them as being of moderate intensity, of medium frequency, and of decrescendo or crescendo-decrescendo configuration. A presystolic murmur of medium frequency and intensity was uniformly noted as well as a low-frequency mid-diastolic murmur at the left sternal border or apex in 2 cases.

Diminution or absence of the first heart sound noted by Vacea et al. was not seen on our phonocardiograms. Nor are our findings in agreement with those of Voill and Sterz, who labeled the second portion of the first sound as “4’’ to conform with Michel’s theory that delayed polarization of the atrialized ventricle manifests itself as a separate late deflection in the QRS complex (and termed P’ because of its resemblance to the original P wave).

**Cardiac Catheterization (Table 5)**

Seventeen of our 23 patients with Ebstein’s anomaly have undergone cardiac catheterization, several having had more than 1 study. There have been no deaths attributable to this procedure and no serious complications. Two pediatric cases (5, 14) developed bouts of supraventricular tachycardia during catheterization, but these were readily controlled by ocular pressure or digitalization.

Several authors have reported fatalities with this procedure, and the general impression has developed that this procedure is contraindicated or unwise in patients with this defect. Our catheterization studies were done under local anesthesia. Whether this technic or chance accounts for our lack of mortality is difficult to decide. Although we recognize that patients with this malformation undoubtedly represent a higher risk, we still think that cardiac catheterization is indicated if the diagnosis cannot definitely be established by other means.

In 6 patients the pulmonary artery was not entered. However, the primary purpose of catheterization was considered to be the establishment of a normal or slightly elevated right ventricular pressure and the documentation of the location of the tricuspid valve. Generally, the tricuspid valve was found to be 2 to 4 cm. to the left of the spine in the anteroposterior view. In 3 cases, the valve was located over the spine, but “lower and more anterior” than usual. This has also been noted by Mayer et al. and Kilby et al. Evidence of an enlarged right atrium was found consistently. In the cases in which the pulmonary artery was entered, the pulmonic valve was normally located. We have had no experience with the use of an intracavitary electrode at catheterization. This has been found to be of value by Sodi-Pallares, Hernandez et al., and Yim and Yu.
In addition, the pulmonary artery tracings resembled the right atrial or right ventricular tracing, an observation reported by several authors.\(^9\)\(^-\)\(^{27}\) In addition, the pulmonary artery pressure tracing often showed a small initial wave occurring after the P wave on the simultaneously recorded electrocardiogram. Blacket et al.\(^9\) and Blount et al.\(^7\) found similar tracings, and postulated that this early deflection represented initial pulmonary bed perfusion by the right atrium. Taken as a group, the pulmonary artery tracings on our patients showed great variation in contour.

Right ventricular pressures ranged from 11 to 40 mm. Hg systolic and -5 to 10 mm. Hg diastolic. In 4 of the 14 cases in which right ventricular end-diastolic pressure was recorded, the value was above 7 mm. Hg. Only 1 of our cases (8) had a right ventricular pressure above 30 mm. Hg, this individual having a systolic pressure of 40 mm. and a diastolic of -2 to 8 mm. Hg. This patient had a differential of 20 mm. Hg across the pulmonic valve, though at postmortem examination there was no evidence of anatomic stenosis. A similar differential across the pulmonic valve has been recorded by Kezdi and Wennemark.\(^{34}\) Several other writers\(^{15}\)\(^-\)\(^{28}\)\(^-\)\(^{37}\) have also reported cases with the systolic pressure in the right ventricle above 30 mm. Hg, but none had a differential pressure across the pulmonic valve greater than 12 mm. Hg.

### Table 5.—Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (yr.)</th>
<th>Pressure in mm. Hg (Systolic/early diastolic/late diastolic mean)</th>
<th>Oxygen content in volumes per 100 ml. (Van Slyke)</th>
<th>Systemic Artery</th>
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<td></td>
<td></td>
<td>PA RV RA</td>
<td>Capacity PA RV RA SVC IVC</td>
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</tr>
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<td>4</td>
<td>8*</td>
<td>22/10 22/6 (6)</td>
<td>20.6 — — 12.6 9.4 — 17.3 (88.5%)</td>
<td></td>
</tr>
<tr>
<td>10†</td>
<td></td>
<td>—                                                —                                             —</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>9</td>
<td>16/5 11/0</td>
<td>19.5 12.9 13.0 13.0 12.9 — 17.3 (88.5%)</td>
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</tr>
<tr>
<td>6</td>
<td>6‡</td>
<td>(26.5) 29/0</td>
<td>20.3 15.7 — 15.6 16.3 14.3 19.5 (94.3%)</td>
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</tr>
<tr>
<td>10§</td>
<td>22/6</td>
<td>22/1 21/5</td>
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</tr>
<tr>
<td>8</td>
<td>14</td>
<td>20/10 (12) 40/2/8 8/0 (2.5)</td>
<td>17.6 12.8 13.2 12.0 11.3 11.9 17.6 (98.3%)</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>2</td>
<td>11/4/5 15/-1/3 3.5/2.5</td>
<td>17.6 12.8 13.2 12.0 11.3 11.9 17.6 (98.3%)</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>4</td>
<td>27.5/15 (22.5) 27.5/1/4 8/1/5 (5)</td>
<td>18.9 13.1 12.6 11.0 11.1 — 18.5 (96.1%)</td>
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</tr>
<tr>
<td>11</td>
<td>4</td>
<td>19/10 21/0/2 10/5 (7)</td>
<td>17.7 — 13.3 12.6 12.9 13.5 17.5 (96.7%)</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>7</td>
<td>11/6 8/3 (5)</td>
<td>18.8 13.8 13.6 13.1 13.8 14.6 18.3 (95.9%)</td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>8</td>
<td>29/12 (15) 30/2/7 10/7</td>
<td>22.4 14.8 12.3 13.2 13.0 — 18.7 (82.0%)</td>
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</tr>
<tr>
<td>14</td>
<td>8</td>
<td>23/13 (16) 264/10 16/0 (14)</td>
<td>17.0 — 12.0 11.9 11.7 13.0 17.0 (98.3%)</td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>9</td>
<td>24/5/10 9/6/8 (8)</td>
<td>17.8 11.1 11.0 10.7 10.4 — 17.5 (97.2%)</td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>8</td>
<td>19/6 25/0/4 7/4</td>
<td>19.0 — 13.0 13.1 13.0 — 18.2 (94.2%)</td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>10</td>
<td>15/0/3 9/0</td>
<td>29.7 — 22.9 22.8 17.7 — —</td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>15</td>
<td>24/-1/2 8/4</td>
<td>26.1 21.1 — — 16.7 — 22.2 (83.7%)</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>18/9 (12)</td>
<td>18/0/6 13/3</td>
<td>27.9 — 18.4 16.8 14.7 16.0 20.5 (72.4%)</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>35</td>
<td>13.5/3/8 (8) 14/8 /10/0/3 (3)</td>
<td>21.0 16.0 16.4 16.1 13.4 — 20.2 (94.6%)</td>
<td></td>
</tr>
<tr>
<td>23</td>
<td>35</td>
<td>19/3 20/0/3 10/2 (2)</td>
<td>15.5 11.9 11.8 11.8 11.0 — 15.7 (99.2%)</td>
<td></td>
</tr>
</tbody>
</table>

* Catheterization at Columbia-Presbyterian Medical Center, New York, N. Y.
† Catheterization by Dr. Alfred R. Henderson, Asbury Park, N. J. No data available.
‡ Catheterization by Dr. James Campbell, Chairman, Department of Medicine, Presbyterian Hospital, Chicago, Ill.
§ Catheterization at Walter Reed Army Medical Center, Washington, D. C.
¶ Left ventricular sample.

The pulmonary artery pressure varied between 14 and 29 mm. Hg systolic to 3 to 15 mm. Hg diastolic. Infrequently, the pulmonary artery pressure tracing resembled the right atrial or right ventricular tracing, an observation reported by several authors.\(^9\)\(^-\)\(^{27}\) In addition, the pulmonary artery pressure tracing often showed a small initial wave occurring after the P wave on the simultaneously recorded electrocardiogram. Blacket et al.\(^9\) and Blount et al.\(^7\) found similar tracings, and postulated that this early deflection represented initial pulmonary bed perfusion by the right atrium. Taken as a group, the pulmonary artery tracings on our patients showed great variation in contour.
With 1 exception (case 9), the right atrial pressure was higher than normal, ranging from 7 to 21 mm. Hg systolic and from −2 to 7 mm. Hg diastolic.

Assessment of the physiologic competence of the tricuspid valve from pathologic material is difficult and often unreliable. A review of the pathologic material nevertheless suggested that stenosis was predominant in 2 cases (2, 3) and insufficiency predominant in 6. In 1 of the latter (case 5) there appeared to be evidence also of stenosis.

Physiologic observations tended to agree with postmortem interpretations. If we assume that one of the criteria of tricuspid stenosis is the occurrence of a right atrial diastolic pressure higher than the right ventricular end-diastolic pressure, then 4 of the catheterized patients had elements of stenosis.

Again, if we assume that the presence of an “s” wave (fig. 4) in the right atrial tracing may be considered a sign of tricuspid incompetence, then at least 10 of our patients would have a degree of tricuspid insufficiency. The “s” wave follows the QRS complex on the simultaneously recorded electrocardiogram and decreases the X′ descent in the normal atrial pattern. Shephard has described the occurrence of this wave in 3 cases of Ebstein’s anomaly as well as in adults with severe pulmonary valvular stenosis. Korner and Shillingford have noted the “s” wave in congestive heart failure, and we have noted it on occasion in cases of complete ostium primum.

In 14 of our cases, the right atrial tracing was suitable for interpretation, and all of these showed definite “s” waves. The height of the “s” wave varied from small 2 mm. deflections to waves large enough partially to obliterate the X′ descent and the V wave of the normal atrial pattern. In 4 cases, the A wave was the most prominent, and in the previously mentioned 10 cases the “s” wave was the larger.

Thus, the 4 patients with physiologic stenosis (5, 6, 11, 19) were also found to have concomitant evidence of physiologic incompetence. Six patients would then appear to show physiologic insufficiency alone, while the other 4 showed reasonably normal tricuspid function under the conditions of the study. It would seem logical that progressive cardiac enlargement, with the concomitant dilatation of the false tricuspid annulus, would enhance the development of valvular insufficiency. Ebstein’s defect thus presents a continuum of tricuspid function, from anatomic and physiologic stenosis to insufficiency, some cases having elements of both and other patients approaching normal.

In those catheterizations in which sufficient data were available for the calculation of physiologic values, 4 studies showed systemic arterial desaturation, and the 11 others were fully saturated (94 per cent or greater).

In the desaturated group, the average systemic saturation was 81.5 per cent. The average pulmonary blood flow was 2.5 L./M.2/min., and the average systemic blood flow 3.0 L./M.2/min.—indicating a small net right-to-left shunt. However, the actual calculated average right-to-left shunt was about 1 L./M.2/min., balanced in 3 patients by a simultaneous average left-to-right shunt of about 0.6 L./M.2/min.—both presumably at the atrial level. Thus, in our desaturated group, both the average systemic and pulmonary blood flow would appear to be less than the normal average of 4.0 L./M.2/min. As a consequence, the average total calculated pulmonary resis-
CLINICAL STUDY OF EBSTEIN'S ANOMALY OF TRICUSPID VALVE

ance in dynes/em./sec.\textsuperscript{5} was 2.2 times greater than normal for the surface area of these patients.

In the 11 patients who were fully saturated, the average saturation was 96.5 per cent. In contrast to the other group, the pulmonary and systemic flows approach normal, being about 3.5 L./M.\textsuperscript{2}/min. The average total calculated pulmonary resistance in this group is normal. One patient (21) would appear to have a left-to-right shunt at the atrial level.

Whether or not these physiologic differences are significant or represent errors inherent in sampling or calculations cannot be stated at the present time.

Differential Diagnosis

The following is limited to those clinical entities having normal or decreased pulmonary vasculature by roentgenograms.

1. Pulmonic Valve Stenosis, with or without Atrial Septal Defect

The young child with these malformations, particularly if in cardiac failure, may have a cardiac contour similar to some seen in Ebstein's defect. Diminished pulmonary vasculature and peaked P waves may also be seen in both anomalies. However, the harsh murmur of pulmonic stenosis in the primary pulmonary area (with the absence of a diastolic murmur), the systolic thrill in the same area and often in the suprasternal notch, and the marked right ventricular hypertrophy on the electrocardiogram help to identify this defect.

2. Myocarditis, Endocardial Fibroelastosis, and Familial Cardiomegaly

The cardiac contour, cardiomegaly with weak pulsations, distant heart sounds, increase in the P-R interval, diffuse ST segment and T wave changes, and the W-P-W pattern may be seen in these conditions and in Ebstein's anomaly.

In differentiating these lesions, left atrial enlargement on the roentgenogram should be of definite value, since it is often present in the above entities, and has not been seen in any of our series of Ebstein's anomaly. In addition, the pulmonary vascular markings are normal or even "full" in these diseases, even in the presence of a markedly enlarged heart; in contrast, in Ebstein's malformation, the very large heart is usually accompanied by decreased pulmonary vascular markings. Negative T waves in the left precordial leads would suggest endocardial fibroelastosis in the child, particularly if associated with cardiomegaly. The history may be of some aid in pointing to specific etiologic factors in myocarditis.

3. Tricuspid Atresia

Similarities between this malformation and Ebstein's anomaly include cardiac contour, peaked P waves, decreased pulmonary vasculature, cyanosis, and the electrocardiogram when the Ebstein's patient has a type B W-P-W pattern. This latter pattern would have left axis deviation, small or absent R waves in the right precordial leads, and tall R waves in the left precordial leads.

However, individuals with tricuspid atresia usually have left atrial enlargement, and ordinarily show a greater degree of cyanosis at an earlier age. The children with this malformation are usually dyspneic at rest, and show growth retardation.

4. Tetralogy of Fallot

This malformation may be confused with Ebstein's anomaly because cyanosis, decreased pulmonary vasculature, similar cardiac contour (when the Ebstein's has a normal or only slightly enlarged heart), syncope, squatting, peaked P waves, and right axis deviation may be found in individuals with either of these defects. Historically, those individuals with Ebstein's anomaly subjected to an extracardiac shunt procedure were diagnosed, as tetralogy of Fallot, trilogy of Fallot, or tricuspid atresia.\textsuperscript{22, 23, 25, 31, 59}

The cyanotic tetralogy, however, invariably has marked right ventricular hypertrophy with little delay in intraventricular conduction. The squatting and syncopal episodes are usually more frequent and severe, diastolic murmurs are rarely heard, and loud systolic murmurs are heard from the primary pulmonary area down the left sternal border.
5. Rheumatic Heart Disease

On rare occasions, in the older child or adult, the cardiac contour, the electrocardiogram, and the murmurs may be similar in Ebstein's anomaly and in rheumatic heart disease. This would be particularly so when the rheumatic lesion involves the tricuspid valve. This occurred in 1 of our adult cases who was diagnosed as having Ebstein's malformation because of "typical" clinical and catheterization findings. However, postmortem study showed a huge right atrium secondary to severe tricuspid stenosis. In addition, this case had calcific mitral and aortic stenosis. Of course, these entities can occur concomitantly, as happened in the unfortunate young man reported by Nöcker and Uibe.\(^\text{56}\)

Again, the presence of left atrial enlargement would suggest rheumatic heart disease. A history of acute inflammatory disease would be helpful, and the presence of typical mitral or aortic murmurs would aid the clinician.

6. Pericardial Effusion

A large globular cardiac contour, weak cardiac pulsations, distant heart sounds, absence of any murmur, low voltage, and diffuse ST-T changes may be found in both clinical entities.

However, absent or very weak cardiac pul-
sations, pulsus paradoxus on physical examination, and the clinical history on occasion, would suggest that the large cardiac contour is caused by a pericardial effusion.

7. Congenital Tricuspid Insufficiency

The clinical, roentgenographic, and electrocardiographic findings may be very similar, and differentiation would appear to require cardiac catheterization to demonstrate the critical displacement of the tricuspid valve in Ebstein's anomaly.

8. Aortic Stenosis

When the heart is only slightly enlarged, when a parasternal murmur of fair intensity is present, and particularly when type B W-P-W is interpreted as left bundle-branch block or left ventricular hypertrophy, then an Ebstein's anomaly may be erroneously diagnosed as "mild aortic stenosis." Misdiagnosis here is thus one of misinterpretation.

In addition, 1 of our patients was originally diagnosed as a "psychoneurotic" because of the multiplicity of her complaints and the paucity of her physical findings.

In most of the preceding conditions, it would be very unusual to hear a "triple" or quadruple" rhythm. Likewise, most of them would not have the marked right bundle-branch block and low voltages (particularly in the right precordial leads) so often seen in Ebstein's anomaly. The presence of a W-P-W pattern with congenital heart disease also should suggest Ebstein's anomaly, although the authors have infrequently seen this pattern in other congenital lesions.

Surgical Considerations

Prior to the accumulation of clinical knowledge that led to methods of detecting this defect, cases of Ebstein's anomaly were sometimes subjected to shunt procedures. The operation was of no benefit, and the correct diagnosis became evident at postmortem examination.

In the widely quoted case of Wright et al, temporary relief was gained by closing the incompetent foramen ovale. In marked tricuspid insufficiency, however, this palliative procedure would appear to have little usefulness. Three subsequent cases operated upon at the same center died during the induction of anesthesia or in the immediate postoperative period.

Recently a new surgical procedure for this anomaly was devised at our hospital. During total cardiopulmonary bypass with the helix reservoir pump oxygenator, a right atrial cardiectomy is made. The displaced leaflets are brought up to the true annulus by means of a plicating technic that excludes the thin wall of the atrialized portion of the right ventricle. Fenestrations in the leaflets are closed with silk sutures, and the atrial septal defect is closed. The tricuspid ring is reduced in diameter. The plicating sutures are so placed that they will interrupt the abnormal conduction pathways described by Lev et al in a case with W-P-W.

This special procedure was attempted in cases 6 and 7, but neither patient survived. The former patient was a semi-invalid because of his disabling paroxysms of tachycardia; the latter patient was in intractable cardiac failure even on a full medical program. Another, case 4, also in severe cardiac failure with thromboembolic manifestations, had cardiac arrest during the induction of anesthesia. Obviously, a trial on better-risk patients will be required before this operation can be evaluated properly.

It is of interest to note that at operation it
was not possible to tell whether the "atrial-ized ventricle" was contracting with the right atrium or with the "functional" right ventricle.

Pathologic Features

The mode of death in 3 of our cases has been noted. Cases 1, 3, and 5 had severe cardiac failure; case 2 died in cardiac failure shortly after the onset of the nephrotic syndrome; and case 8 died unexpectedly while sitting at his desk in school. Some of the postmortem specimens of the heart (figs. 5 to 9) demonstrate strikingly the typical abnormal positions of the tricuspid valve. The anterior leaflet is usually the largest and best developed leaflet and is attached to the true annulus along its medial aspect. The septal (medial) and posterior leaflets are displaced down from the true annulus, the posterior leaflet usually showing the greatest degree of deformity to the point of being absent entirely.

The valve tissue proper may be thick or thin, intact or with multiple perforations, or smooth or nodular. Leaflets may appear to be entirely functional, or they may be adherent to the endocardium ("tethered down"). The chordae tendineae vary from minute useless structures to those that approach normal. The papillary muscles similarly show wide variation in structure.

The right atrium is consistently enlarged, with dilatation and hypertrophy invariably present. On the other hand, the "atrialized" portion of the ventricle is always very thin, occasionally with thin aneurysmal areas. This latter feature was present in the 2 infants that died (cases 1, 3). The "functional" right ventricle may be rudimentary or very large, and its wall may be thin or hypertrophied or may show areas of each. The infundibulum usually shows hypertrophy. The size of the "functional" right ventricle varies more or less inversely with the "atrialized" right ventricle. Pechstein\textsuperscript{59} has attempted to classify this anomaly into 4 separate morphologic groups, utilizing valve positions and proportion of "atrialized" to "functional" right ventricle as the basis of his criteria.

All but 2 of our autopsied cases (2, 8) had an incompetent or fenestrated foramen ovale. Case 1 showed a patent ductus arteriosus, but this was of dubious significance inasmuch as death occurred shortly after birth. Only 1 other case (3), who died at 3½ months, had an associated cardiac abnormality—a ventricular septal defect. Although this was present, there was no evidence of increased pulmonary blood flow. Any blood shunting through this ventricular defect had 2 possible modes of exit: through the minute 3 mm. tricuspid orifice into the right ventricle, or through the foramen ovale to the left atrium. In view of the decreased vascularity on the roentgenograms, it is apparent that the small tricuspid aperture was effective in preventing excessive or even normal pulmonary blood flow.

The pulmonic, aortic, and mitral valves were uniformly normal, except for "slight nodularity" of the mitral valve in case 8. An attached thrombus was present in the apex of the left ventricle in case 5, and a nonattached thrombus practically filled the cavity of the small right ventricle in case 2. Fibrinous pericardi-
Atrophy of and defect.

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stein’s surfaces anomaly. of corrected atrial and right ventricular

ward al.11”), tricular of the left-sided atrioventricular valve, so-called “left-sided Ebstein’s anomaly.”

Other authors13, 61 have reported the ostium primum syndrome in conjunction with Ebstein’s anomaly, and 3 other cases5, 14, 62 have been described with an interventricular septal defect. In addition, Keith et al.5 had a case of Ebstein’s anomaly with pulmonary atresia and multiple atrial septal defects.

Chronic passive congestion, often with atrophy of the surrounding liver cells to the point of cardiac cirrhosis (case 6), was usually noted in our autopsied cases. One (case 4) showed an area of adenomatous hyperplasia of the liver in addition to large areas of central atrophy.

Four of our 8 cases showed pathologic changes in the brain. Case 2 showed an area of patchy demyelination in the frontal cortex, in addition to congestion and petechiae. Case 5 had congestion of all the vascular spaces and a large area of cystic degeneration in the floor of the fourth ventricle. Case 4 exhibited congestion of the vessels and areas of recent encephalomalacia in the left inferior parietal lobule and the inferior surface of the right occipital lobe, as well as slight calcification of the basal ganglia. Case 7 had a large cystic area of encephalomalacia and glial formation in the left cerebral hemisphere. The latter 2 patients had clinical evidence of cerebral emboli or thrombi.

Infarcts of the spleen and kidney were seen in case 7 and in the kidney in case 4.

None of the autopsied cases showed evidence of pulmonary tuberculosis, and such evidence is not present in the 15 living patients.

SUMMARY

Twenty-three cases of Ebstein’s malformation of the tricuspid valve are reviewed, including 8 with postmortem examinations. This anomaly occurs equally in males and females, and has been estimated to constitute less than 1 per cent of all congenital heart defects.

There was no evident etiologic relationship to various prenatal factors, birth order, maternal age, birth weight, or month of birth. Only 1 patient had a relative with known congenital heart disease.

Twelve of the 23 cases had neonatal symptoms or signs, generally cyanosis or a murmur. This neonatal cyanosis invariably diminished or disappeared with time, but recurred later in life in all cases. Symptoms after the neonatal period included dyspnea, cyanosis, excessive fatigue, central nervous system symptoms, cardiac failure, precordial or epigastric pain, paroxysms of tachycardia, and squatting.

The most common physical findings were a systolic murmur, a second sound which was always heard as well over the pulmonic area as over the aortic area, normal pulmonary breath sounds, cardiomegaly, normal growth features, a “triple” or “quadruple” rhythm, cyanosis, a systolic thrill, a diffuse area of maximal impulse, a diastolic murmur, a “split” first sound, “metallic” or “clicking” heart tones, cardiac failure, and an unusual “facial erythema.”

Roentgen studies regularly showed cardiomegaly, normal to decreased pulmonary vascular, and a normal-sized left atrium. Cardiac size and contour varied widely, though in certain cases the “box-like” configuration was diagnostic.

The most frequent electrocardiographic findings in the 18 patients with a normal sinus atrial rhythm were incomplete or complete right bundle-branch block (often with widely splintered or notched complexes), peaked and occasionally prolonged P waves, low voltage QRS complexes in the right precordial leads (none had an R wave in V1 exceeding 8 mm. in height), and right axis deviation. Five cases showed a constant type B Wolff-Parkinson-White (W-P-W) pattern, and another did so intermittently. The presence of a type B W-P-W pattern in a cyanotic child with congenital heart disease and normal to decreased
pulmonary vasculature should immediately suggest the possibility of Ebstein’s anomaly.

Phonoeardiography was performed in 12 patients. This is a valuable adjunct in elucidating the auscultatory findings.

A total of 20 right heart catheterizations were carried out in these 23 patients, without any mortality or serious complications. The principal value of cardiac catheterization is in documenting the location of the tricuspid valve, and in obtaining pressure tracings in the various vessels and chambers.

This defect may be confused with pulmonary valvular stenosis, myocarditis, endocardial fibroelastosis, tetralogy of Fallot, tricuspid atresia, rheumatic heart disease, pericardial effusion, congenital tricuspid insufficiency, and “mild aortic stenosis.”

Two of our patients underwent a new type of surgical procedure, but neither survived.

Physiologic data and studies of postmortem specimens indicate that Ebstein’s anomaly represents a continuum from predominant tricuspid stenosis to predominant tricuspid insufficiency, with some having elements of both, and others approaching normal.

ACKNOWLEDGMENT

We acknowledge gratefully the many persons who contributed clinical and pathologic data on the cases or aided in the preparation of the manuscript, particularly Robert A. Good, Ph.D., M.D., American Legion Memorial Heart Research Professor of Pediatrics, without whose generous aid and advice this manuscript could not have been completed.

SUMMARIO IN INTERLINGUA

Es presentate un revista de 23 casos del malformation de Ebstein del valvula tricuspidal. In 8, datos necroptic es includite. Ist anomalia occurre equalmente in masculos e in femininas. Il ha esseite estimate que illo constitue minus que 1 pro cento de omne congenite defectos cardiac.

Esseva notate nulle evidentia relation etiologic con varie factores prenatal, con le numero del fraternos senior, le etate del mater, le peso natal, o le mense del nascentia. Solmente 1 del patientes habeva un consanguineo con cognoscite morbo cardiac congenite.

Dece-duo del 23 habeva symptomos o signos neonatal, generalmente cyanosis o un murmure. Le cyanosis neonatal se diminuva o disparea invariablemente in le curso del tempore, sed in omne casos illo recurreva a un etate plus avaintiate. Le symptomas post le periodo neonatal includeva dyspnea, cyanosis, grados excessive de fatiga, symptomas del systema nervose central, disfallimento cardiace, dolores precordial o epigastic, paroxysmos de tachycardia, e statura pyenice.

Le plus commun constatationes physic esseva un murmure systolic, un secunde sono (semper audible supra le area pulmonic e supra le area aortic), normal sonos pulmone-respiratori, cardiomegalia, normal caracteristicas de crescentia, un “triple” o “quadrupe” rhythm, cyanosis, un fremito systolic, un area diffuse de impulso maximal, un murmure diastolic, un “finitite” prime sono, sonos cardiace “metallic,” disfallimento cardiae, e un inusual “erythema facial.”

Studios roentgenologic monstrava regularmente cardiomegalia, normal o reduceit vasculatura pulmonar, e dimensiones normal del atrio sinistre. Le dimensiones e le contorno del corde variava extensemente, ben que in certe casos le configuration “quadrate” esseva diagnostic.

Le plus frequente constatationes electrocardiographic in le 18 patientes con normal rhythm sino-atrial esseva bloco incomplete o complete de branca dextere (frequentemente con extense segmentation o indentation del complexos), punctate e a vices prolongate undas P, complexos QRS a basse voltage in le derivations dextero-precordial (un unda R in V₁ de plus que 8 mm de altor non occurreva in ume del casos), e deviation dextero-axial. In 5 casos, un configuration del typo B de Wolff-Parkinson-White esseva constante, in un sexte caso illo esseva presente intermittente. Le presentia de un configuration del typo B de Wolff-Parkinson-White in un puer cyanotic con congenite morbo cardiac et normal o reduceit vasculatura pulmonar debiera suggester immediatemente le possibilitate de anomalia de Ebstein.
Studios phonocardiographic esseva effectuate in 12 patientes. Esto es un ajuvente de valor in le elucidation del constatationes auscultatorii.

Un total de 20 catheterisationes dextero-cardiac esseva effectuate in iste 23 patientes. Occurrve nulla morte e null serie complicaciones in consequentia del catheterismo. Le principal valor de iste technica es que illo pote documentar le location del valvula tricuspid e que illo permite obtener registrationes del pression in le varie vasos e cameras.

Le defecto sub discussion pote esser confundite con stenosis del valvula pulmonar, myocardiitis, fibroelastosis endocardial, tetralogia de Fallot, atresia tricuspidal, rheumatic morbo cardiae, effusion pericardial, congenite insufficienția tricuspidal, et le "forma leve de stenosis aortic."

Due de nostre patientes esseva operate secundo un nove technica chirurgic. Supervișeva ni le un ni le altere.

Datos physiologic e studios de specimens necroptic indica que anomalia de Ebstein representant un continuo a predominante stenosis tricuspidal a predominante insufficienția tricuspidal. In certe cases, il ha elementos de ambas; altere cases approcha un stato normal.

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THE MODESTY OF A GREAT MIND

DAVID BREWSTER

Scottish physicist, 1781-1868

The modesty of Sir Isaac Newton, in reference to his great discoveries, was not
  founded on any indifference to the fame which they conferred, or upon any erroneous
  judgment of their importance to science. The whole of his life proves, that he knew
  his place as a philosopher, and was determined to assert and vindicate his rights. His
  modesty arose from the depth and extent of his knowledge, which showed him what a
  small portion of nature he had been able to examine, and how much remained to be
  explored in the same field in which he had himself labored. In the magnitude of the
  comparison he recognized his own littleness; and a short time before his death he
  uttered this memorable sentiment: “I do not know what I may appear to the world,
  but to myself I seem to have been only like a boy playing on the seashore, and diverting
  myself in now and then finding a smoother pebble or a prettier shell than ordinary,
  whilst the great ocean of truth lay all undiscovered before me.”—Memoirs of Sir Isaac
  Newton. From Great Companions. Readings on the Meaning and Conduct of Life
Clinical Study of Twenty-Three Cases of Ebstein's Anomaly of the Tricuspid Valve

GEROLD L. SCHIEBLER, PAUL ADAMS, JR., RAY C. ANDERSON, KURT AMPLATZ and RICHARD G. LESTER

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