Ebstein’s Anomaly
Presentation of Ten Cases

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Ten cases of Ebstein’s anomaly are presented and the literature is reviewed. Certain diagnostic features of the clinical profile, in particular the auscultatory findings, are emphasized.

Since first described by Ebstein in 1866,1 approximately 80 cases of this anomaly have been reported.2-4 Its differentiation from forms of congenital heart disease amenable to surgery is of importance and in the majority of cases may be accomplished by clinical means.

The 10 cases studied at the Children’s Medical Center, Boston, ranged in age up to 25 years. There were 4 males and 6 females. Postmortem material was available in 4, cardiac catheterization had been performed in 4, and angiocardiography in 2. One cardiac catheterization was performed at the Maine General Hospital, Portland, Me.

Results

History and Physical Examination

Symptomatology was evaluated in 9 patients. Slightly increased fatigability and mild dyspnea on exertion were present in all patients over 1 year of age. Severe dyspnea occurred only with the onset of congestive failure. Cyanosis was observed in the neonatal period in 6 cases; 2 of these subsequently became cyanotic only with crying or activity. Cyanosis appeared after infancy in 2, and 1 patient at the age of 25 years had never been cyanotic. Three patients developed congestive failure in the neonatal period, at 2 years, and at 8 years of age, respectively. Paroxysmal atrial tachycardia occurred in 1 case. Squatting was observed infrequently in 2 patients and in only 1 was an anoxic spell with loss of consciousness described.

Examination of these patients revealed normal growth in the majority. Five were mildly cyanotic but only 2 presented clubbing. The cardiac impulse was not forceful and extended diffusely from sternum to apex. A systolic thrill was palpable between xiphoid and apex in 6 patients. On auscultation the outstanding findings, noted in 8, were a triple or quadruple rhythm, a diminished pulmonary second sound, and systolic and diastolic murmurs of medium frequency, heard maximally at the lower left sternal border and apex. The systolic murmur was frequently described as “scratchy” or coarse.

Phonocardiogram

Sound tracings available on 5 patients showed remarkable similarity (fig. 1). The first heart sound was delayed and normal in intensity, maximal at the apex. The second heart sound was maximal at either the lower left sternal border or apex in all cases, and was narrow or unsplit in 3. All presented a third sound, and in 3 a fourth sound was observed as well.

A systolic murmur of moderate intensity and of medium frequency and decrescendo or crescendo-decrescendo in configuration was recorded in all tracings. Also uniformly recorded was a presystolic murmur of medium frequency and intensity. A very low frequency mid-diastolic murmur was present in 2. These murmurs were of greatest intensity at the left lower sternal border or apex.

Electrocardiogram

Electrocardiographic studies were done in 9 cases. Tall, peaked or broad P waves over 2.5 mm. in leads II, V1, and V2 were present in 8 patients and in many they measured over 4 mm. in height. Atrioventricular conduction was prolonged in 5 tracings.

Right bundle-branch block was a constant finding; in 4 the QRS complex measured 0.12
second or more; in the remainder the block was incomplete. Ventricular potentials of small amplitude in the right precordial leads were observed in all who were beyond early infancy. Paroxysmal atrial tachycardia was documented by electrocardiogram in 1 patient; frequent ventricular ectopic beats were seen in another.

The evolution of the characteristic electrocardiographic pattern is observed in figure 2. The first electrocardiogram taken at 3 days of age showed complete reversal of the adult R/S progression from V₄R to V₆ with an R in V₁ of 20 mm. It was within normal limits for age. In the second tracings at 3½ years of age there had appeared prolonged atioventricular conduction with a P-R interval of 0.20 at a rate of 105 per minute, P-pulmonale with a P in lead II of 5 mm., incomplete right bundle-branch block, and potentials of low amplitude in the right precordial leads.

**Radiologic Examination**

Marked cardiomegaly involving the right side of the heart was noted by roentgenologic examination in all patients (fig. 3). The contour was frequently described as "globular." A narrow base with a small aorta was typical; the pulmonary artery was poorly visualized in 7 patients and the pulmonary vascular markings were decreased in all except 1. Diminution in the amplitude of cardiac pulsation was specifically noted in 3 cases.

**Angiocardiography**

Two patients were studied by angiocardiography (figs. 4 and 5). The dye entered a huge atrial chamber where it was greatly diluted and from which it emptied slowly. In one patient the pulmonary artery filled poorly; in the other it appeared normal. A right-to-left shunt was suggested by simultaneous opacification of the aorta and pulmonary artery in both cases; in one the shunt was demonstrated at the atrial level.

Figure 5 was taken in a child who had been in chronic congestive failure and who died shortly after the procedure. He was found at postmortem examination to have a markedly dilated distal as well as proximal right ventricular chamber.

**Cardiac Catheterization**

This procedure was performed in 4 patients; it was not completely satisfactory in any instance, but yielded suggestive information in 3 (table 1). In each the catheter coiled in a large atrium (fig. 6). In patient S. P. a predominant "a" wave with a peak of 2.5 to 5 mm. Hg was obtained in this chamber. The pulmonary artery could not be entered; the right ventricular systolic pressure was within normal limits. Unsatisfaction of left atrial blood suggested a right-to-left shunt at this level. Peripheral arterial oxygen saturation was 85 per cent.
In patient R. B. it was impossible to proceed to the right ventricle because of the appearance of the ectopic beats when the tricuspid valve area was approached. This region appeared to lie farther to the left than normal. Peripheral arterial saturation was 95 per cent. A rise in oxygen saturation at the atrial level suggested a small left-to-right shunt. In patient J. L. the catheter entered an area with a right ventricular pressure curve at a “lower” site than normal. Systolic pressure in the right ventricle was not elevated and there was no gradient across the pulmonary valve. Peripheral arterial saturation was 90 per cent.

Course

Six patients are doing well and have minimal symptoms. One has had frequent episodes of paroxysmal atrial tachycardia eventually controlled with digitalis.

Four patients have died. The first, a stillborn, died presumably as a result of a prolapsed cord in a difficult breech delivery. The second child, L. C., a 9-day-old infant, died in severe congestive failure, and at postmortem examination was found to have a hypoplastic aorta and coarctation as well as Ebstein’s anomaly. The third patient, L. H., was a 2-year-old boy, cyanotic since birth, who had been treated for congestive failure on admission, with good response. He developed marked bradycardia and coma during angiocardiography and died 36 hours later. The fourth patient, C. C., an 8-year-old girl, had been in congestive failure for several months and was rapidly deteriorating. She had been cyanotic since $4\frac{1}{2}$ years of age. Cardiac arrest occurred during cardiac catheterization and she failed to recover.
EBSTEIN'S ANOMALY

Fig. 3. Representative posteroanterior roentgenograms of patient C.M. (left) top, at 3 days of age and bottom, at 3 years of age.

Fig. 4. S.P., age 8 days. Angiocardiogram in left lateral position. The large chamber of the right atrium and proximal right ventricle was opacified from the inferior vena cava. The pulmonary artery and aorta opacified simultaneously.

Postmortem Material

Autopsies were performed in 4 cases. In each instance the weight of the heart was 2 to 3 times normal. The right atrium was markedly dilated in all, with hypertrophied muscle bundles lining the wall of the atrium and its appendage. These muscles bundles were prominent in the atrial wall of the stillborn.

The foramen ovale was patent in every case. In 1 patient a small ventricular septal defect was present as well.

The right ventricle was divided into proximal and distal portions by the misplaced valve leaflets. In the stillborn (fig. 7) these were distinctive, the proximal portion forming a thin-walled pocket measuring 1.2 cm. by 1.6 cm., the distal or outflow portion being of normal appearance and thickness. No histologic defect in the cellular structure of the proximal ventricular myocardium was observed.

In patient L. C., who had a complicated lesion, the divisions of the right ventricle were
Table 1.—Findings on Cardiac Catheterization in Three Cases of Ebstein's Anomaly of the Tricuspid Valve

<table>
<thead>
<tr>
<th>Patient and age</th>
<th>O₂ content volume per cent</th>
<th>O₂ capacity</th>
<th>Pressure, mm. Hg</th>
<th>Flow/M.²</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>SVC</td>
<td>RA</td>
<td>RV</td>
<td>PA</td>
</tr>
<tr>
<td>S. P., 8 days...</td>
<td>13.5</td>
<td>14.7</td>
<td>14.7</td>
<td>—</td>
</tr>
<tr>
<td>R. B., 4 yrs.....</td>
<td>12.7</td>
<td>14.5</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>J. O., 25 yrs.....</td>
<td>9.8</td>
<td>11.3</td>
<td>12.1</td>
<td>12.5</td>
</tr>
</tbody>
</table>

* M = Mean pressure

not so well differentiated; the distal portion was somewhat dilated and hypertrophied and measured 4 mm. in thickness. The dilated proximal portion was variable in thickness, averaging 2 mm.

In patient L. H., who had been in congestive failure, both the proximal and distal divisions were dilated and thin walled and the pulmonary valve appeared thickened (fig. 8). Patient C. C., in whom congestive failure had also been present, had a paper-thin, 12 by 15 cm. proximal portion and a dilated distal division measuring 9 by 6 cm. (fig. 9). In this case the pulmonary valve appeared to be dilated.

The abnormal valve cusps varied in detail from one patient to another. In the stillborn the anterior cusp was normal in structure and in its origin from the valve ring. The posterior and medial cusps, arising 1 cm. below the ring from the wall of the right ventricle and its septum, were adherent to the wall and to sparse, thin chordae tendineae. The endocardium showed no evidence of scarring. In patient L. C., the picture was similar, with a normal anterior leaflet, and with small posterior and septal leaflets arising from the right ventricular wall midway between valve ring and apex. No chordae or papillary muscles were attached to these leaflets. In patient L. H., the anteromedial, medial, and a small part of the posterior leaflet arose from the ring; the anterolateral and most of the posterior cusps, from the wall of the right ventricle 1 to 3 cm. below the valve ring. The displaced cusps were almost completely adherent to the wall and there were no well-formed papillary muscles in this area. In patient C. C., the anterior leaflet arose as a large membranous curtain, 8 by 6 cm. in measurement, from the valve ring; the left border was attached to the ventricular wall, the right border was free. The septal leaflet was normal. The posterior leaflet arose from the ventricular wall, was poorly developed, and was bound down to the wall.

In each instance, passive congestion of the liver was noted.

Patient L. C. had additional malformations of the great vessels: a hypoplastic aorta with a coarctation 1 cm. in length proximal to a patent ductus arteriosus. Patient C. C. had a probe-patent ductus arteriosus.

Discussion

Clinical Findings

The symptomatology of Ebstein's anomaly is generally mild. A number of patients have been asymptomatic, in particular those who survived to an advanced age. Cyanosis, persistent or intermittent, was observed in about four fifths of the cases; in one third it was present from birth. The most common
history was one of cyanosis appearing some time after infancy, intermittent with cold or exertion at first, and gradually becoming more marked.

Slight to moderate dyspnea and fatigability are almost invariably present, appearing usually after the period of early childhood. Cardiac arrhythmias are extremely frequent; 5 patients had documented paroxysmal atrial tachycardia,\textsuperscript{19, 29, 37, 46} 1 had atrial fibrillation,\textsuperscript{33} and another had atrial flutter.\textsuperscript{41} Wolff-Parkinson-White syndrome was described in 1 case.\textsuperscript{30} About one third of all patients experienced bouts of palpitation. Squatting, anoxic spells, and precordial pain are uncommon.

At least 12 cases have been in congestive failure. In 5 patients it occurred in the first decade, in 5 others in the second decade. Its occurrence is probably a poor prognostic sign. Only 2 were alive at the time of reporting; one was a 29-year-old man living 5 years after his first bout of failure.\textsuperscript{32} In the remainder there were, at least as an associated cause of death, pulmonary tuberculosis in 2,\textsuperscript{2, 9} myocardial infarction in 2,\textsuperscript{19, 35} erysipelas in 1, embolism in 1,\textsuperscript{5} and fatal outcome of diagnostic procedures in 2.

Almost without exception these patients were normal in growth. Clubbing of the fingers has been noted in about one third. Precordial chest deformities were infrequent and the cardiac impulse was commonly diffuse and quiet. In the majority, a systolic thrill was palpable between xiphoid and apex.

Auscultatory findings of wide variety have been described in the literature. The findings in our series have been so uniform as to be characteristic: a triple or quadruple rhythm; a second sound maximal in intensity at the lower left sternal border or apex; a medium frequency, moderate intensity systolic murmur decrescendo or crescendo-decrescendo in contour, and a late diastolic or presystolic murmur of moderate intensity, both maximal at the xiphoid or apex. These characteristic findings may not be present, however, for 6 patients,\textsuperscript{16, 18, 27, 29, 47} including one of our own, had no murmurs at all.

It has frequently been commented that clinical signs of tricuspid insufficiency are rarely seen. A pulsating liver was noted in none of our cases and pulsating neck veins only in a patient in failure. It has been suggested that clinical signs of insufficiency may be lacking.
because of the great distensibility of the combined chamber above the tricuspid valve, which allows regurgitation of a considerable volume of blood without transmission of a visible pressure wave to the systemic veins.43

The hugely dilated and hypertrophied right atrium is reflected in the characteristic tall P waves of the electrocardiogram, which with right bundle-branch block and low amplitude of right ventricular potentials have been almost constant findings in all the cases. Prolonged atrioventricular conduction is common. The characteristic pattern may not be present in early infancy,44 in our patient also this pattern developed over a 2-year period. Abnormally tall right ventricular potentials have been reported in 2 cases, case C of Kjellberg et al.,45 and case III of Brown et al.,47 so that the diagnosis of Ebstein's anomaly cannot be entirely dismissed in the presence of tall right ventricular potentials.

The radiologic picture characteristically is one of marked cardiomegaly, a contour consistent with right-sided enlargement, a narrow base, diminished pulmonary vascular markings, and a poorly delineated pulmonary artery. Fluoroscopically, pulsation of the right border is frequently weak although a few observers have described increased presystolic pulsations. The contour of the heart mimics that of pericardial effusion and the amplitude of pulsation in each instance is diminished. The similarity of the roentgenologic picture to that of severe pulmonic stenosis with intact ventricular septum has been pointed out.21

Angiocardiography has been performed in 23 cases. Ours appears to be the only reported fatality due to the procedure. Characteristically, the dye opacifies and is greatly diluted in a huge chamber representing right atrium and proximal right ventricle. This chamber shows delayed emptying and obscures to a large extent the remainder of the cardiac silhouette. The pulmonary artery and its branches are poorly opacified. Several authors have demonstrated a distinct small outflow portion of the right ventricle, filling later, and separate from the large chamber. Evidence of a right-to-left shunt has been observed in about one half of the cases by early opacification of the aorta or by observation of the dye shunting to the left atrium. The information obtained from the procedure does not appear to be pathognomonic and does not contribute greatly to the diagnosis.

Cardiac catheterization (table 2) has been reported in 38 patients and was performed in 4 of our own. Seven of the cases were proved by autopsy. There is a marked tendency toward the development of arrhythmias, and the procedure carries with it a definite hazard: 3 patients have died during or shortly after the procedure.26, 29 A fourth23 died 21 days later with emboli that produced gangrene in both lower extremities and thrombi in branches of
the left coronary artery. Thrombi were present in the left atrium and left ventricle, both of which had been explored with the catheter. In addition, Campbell in an editorial states that both he and Wood have lost 1 patient in 5 or 6 who were catheterized. We think, however, that in questionable cases more pertinent information may be gained from catheterization than from angiocardiography.

The malformation results in a reduction in size of the functioning right ventricle. Pulmonary flow may be diminished, and with a rise in pressure in the atrial chamber a right-to-left shunt may occur at the atrial level.

Typically the catheter coils in the huge right atrial chamber. The tricuspid valve appears to lie to the left of its usual position and the right ventricle may be entered with difficulty. Arrhythmias frequently occur as the catheter approaches the valve area.

A number of observations may be made from the reported data. Peripheral arterial unsaturation is usual. Only 9 cases with arterial saturation over 90 per cent have been reported. Post-mortem examination revealed no interatrial communication in 2 cyanotic patients, and it has been suggested that cyanosis may be of a peripheral type in some instances because of a low cardiac output.

Ten cases revealed at the atrial level a rise in oxygen saturation of sufficient degree to suggest a left-to-right shunt of small magnitude. Right atrial mean pressures are usually moderately elevated. The pressure contour has been described in a number of cases: in 8 a predominant V wave was observed suggesting that tricuspid insufficiency is indeed present in many. One patient, in whom a "v" wave of 14 mm. was recorded, died in congestive failure and was found to have a greatly dilated distal right ventricular chamber. In 7 cases an "a" wave was predominant. Transmission of this atrial wave to the right ventricle and pulmonary artery has been described and it has been suggested that the contraction of the atrial chamber may supplement the right ventricular func-

**Fig. 9.** C.C., age 8 years. A. Exterior of the heart illustrating large right atrium, RA. RV, right ventricle. B. Interior of same heart. The right atrium is opened and the tricuspid valve, TV, is viewed from above. The large membranous anterior cusp arises from the valve ring.
tion of promoting pulmonary flow in some instances. Of the catheterized cases that have been autopsied, the few without defects in the atrial septum had the highest peak pressures in the right atrium.

Closure of the atrial defect was performed in 1 case\textsuperscript{48} with symptomatic improvement. This procedure would not be expected to be satisfactory, however, in patients with marked tricuspid regurgitation.

Right ventricular systolic pressure has not been significantly elevated in any case. The abnormal systolic ejection contour is delayed in onset and in reaching a peak.\textsuperscript{48} Several authors have reported elevated end-diastolic pressures, which in some instances may represent the transmitted atrial wave.

The pulmonary artery was entered in 27 cases. In none was there a significant gradient across the pulmonary valve.

\begin{table}
\centering
\caption{Summary of Catheterization Data in Thirty-Eight Reported Cases of Ebstein's Anomaly}
\begin{tabular}{|l|c|c|c|c|c|}
\hline
Author and ref. no. & Pressure in mm. Hg & & & & \\
 & R. A. & R.V. & P.A. & Peripheral arterial O\textsubscript{2} saturation & Suggestive L-R shunt at atrial level & Autopsy evidence of atrial communication \\
\hline
Engle\textsuperscript{21} & 8/1 & 21/7 & * & 66 & + & F.O. \\
Brown\textsuperscript{18} & 4 & & 10 & & & \\
Brown\textsuperscript{18} & 14 & & & & & \\
Brown\textsuperscript{18} & 11 & & & & & \\
Blacket\textsuperscript{22} & 4 & & 8 & 8 & 8 & \\
Kjellberg\textsuperscript{43} & v wave & 14/5 & 13/7 & 81 & & \\
Kjellberg\textsuperscript{43} & M-10 v wave 14 mm. & 36/6 & 24/7 & 90 & & \\
Kjellberg\textsuperscript{43} & M-13 v wave 13 mm. & 23/8 & 24/8 & 94 & & \\
Kjellberg\textsuperscript{43} & M-10 v wave 15 mm. & 33/9 & 27/17 & 85 & & \\
Tourniaret\textsuperscript{20} & 15 & & 15 & 20 & & \\
Medd\textsuperscript{40} & 8/4 & & 18/4 & & & \\
Van Lingen\textsuperscript{40} & 7/4 & & 22/4 & 22/10 & 87 & + \\
Van Lingen\textsuperscript{40} & & & 22/8 & 22/10 & 69 & + \\
Getzsch\textsuperscript{34} & 7/2 & & 18/2 & 14/8 & 87 & + \\
Getzsch\textsuperscript{34} & 8/2 & & 27/2 & 16/4 & 89 & \\
Getzsch\textsuperscript{34} & 8/1 v wave & 25/1 & * & 89 & & \\
Getzsch\textsuperscript{34} & 15/3 v wave & 22/4 & 17/10 & 93 & & \\
Getzsch\textsuperscript{34} & 2 & & 15/0 & 15/8 & 96 & \\
Getzsch\textsuperscript{34} & 3 & & 24/2 & 16/6 & 90 & + \\
Broadbent\textsuperscript{32} & 10/4 & 13/4 & 19/4 & 20/10 & 89 & + \\
Broadbent\textsuperscript{32} & 11/4 & 13/4 & 18/3 & 18/10 & 62 & F.O. \\
Broadbent\textsuperscript{32} & 25/13 v wave & 34/10 & 33/13 & 34/22 & 84 & \\
Kerwin\textsuperscript{15} & 10/10 & 32/6 & & 86 & & None \\
Henderson\textsuperscript{32} & 7 & & 9 & 14 & 91 & \\
Goodwin\textsuperscript{24} & 2.5 & & & & & \\
Grob\textsuperscript{31} & 10/0 & 30/0 & 30/15 & & & \\
Baker\textsuperscript{23} & 10 & & * & * & 75 & A.S.D. \\
Baker\textsuperscript{23} & not given & & * & * & 93 & A.S.D. \\
Reynolds\textsuperscript{31} & 11-17 & & * & * & 79 & + \\
Bayer\textsuperscript{41} & 10/0 v wave & 20/0 & 26/0 & 26/10 & 80 & + \\
Bayer\textsuperscript{41} & 7/3 a wave & 10/3 & 18/0 & 15/9 & 78 & + \\
Bayer\textsuperscript{41} & 4/0 a wave & 7/0 & 15/0 & 15/7 & 79 & \\
Schaede\textsuperscript{26} & not given & & & & & \\
Schaede\textsuperscript{26} & not given & & & & & \\
Blount\textsuperscript{48} & 7/2 a wave & 24/0/4 & 19/9 & 90.9 & & \\
Blount\textsuperscript{48} & 12/5 a wave & 24/4/11 & 20/9 & 76.6 & & \\
Blount\textsuperscript{48} & 10/4 a wave & 29/3/13 & 22/10 & 95.4 & & \\
Blount\textsuperscript{48} & 10/5 a wave & 29/0/9 & 22/9 & 96 & & \\
\hline
\end{tabular}
\end{table}

M, mean pressure;* not entered; F.O., foramen ovale; A.S.D., atrial septal defect.

† When two pressures are listed, the first is that of the proximal right ventricle, the second that of the distal chamber.
Ebstein’s anomaly may be compatible with considerable longevity, as evidenced by 2 patients who survived to the eighth decade\textsuperscript{27,47} and 4 who were in their sixth and seventh decades.\textsuperscript{2,6,18,48} However, of 48 patients who have died, 40 per cent died before the age of 10 years and an additional 30 per cent by the age of 20.

Ten patients died apparently as a direct result of a diagnostic or operative procedure. Four died as a result of cardiac catheterization and 1 following angiocardiography. In 5 published cases operative procedures were undertaken. Presumptive diagnoses in these cases were tetralogy of Fallot and pulmonic stenosis. Shunt procedures in 3\textsuperscript{21,31} exploratory thoracotomy 1,\textsuperscript{23} and induction of anesthesia in 1\textsuperscript{32} resulted in death. Because of these experiences such procedures are undertaken with some concern, and a diagnosis made by clinical means becomes of more than academic interest.

Sudden death occurred in 7 patients;\textsuperscript{1,8,13,17,21,26,49} cardiac arrhythmias may have been the cause.

Central nervous system complications have been described: cerebral embolism\textsuperscript{14} was the supposed cause of death in 1; 2 patients died with brain abscesses,\textsuperscript{7,28} and another with meningitis.\textsuperscript{7}

Tuberculosis was a common cause of death in the earlier cases.\textsuperscript{1,3,8,9,12} Two patients died with myocardial infarction,\textsuperscript{19,35} and 2 in congestive failure without other associated cause of death.\textsuperscript{11,43}

\textbf{Pathology}

We have gathered 48 descriptions of post-mortem specimens, including our own. The hearts were well over normal weight and pathologic change was generally limited to the right side.

The primary abnormality appeared to lie in the development of the tricuspid valve, the papillary muscles, and the chordae tendineae. The valve leaflets were abnormal in origin and structure, arising in whole or in part from the wall of the ventricle below the valve ring. In structure they varied from small nubbins to large membranous curtains that fused so as to allow communication only through small fenestras. They were occasionally plastered down to the wall of the right ventricle to form an obviously insufficient valve. Cusps in some instances were completely absent. Most commonly the posterior and medial leaflets were grossly deformed but the anterior leaflet was frequently involved as well. In the region of the abnormal cusp, the papillary muscles and chordae were invariably poorly formed, sparse, vestigial, or absent.

The right ventricle was divided into 2 parts by the abnormal valve. The proximal portion was a thin-walled, dilated chamber that had taken on the characteristics of the atrium. The right atrium was hugely dilated and frequently hypertrophied. No specific histologic defect has been noted in the myocardium of the proximal right ventricle. The distal portion was frequently limited to the outflow chamber. Usually its wall was of normal thickness. In 6 instances\textsuperscript{33,29,41} including 3 of our own, this distal chamber was found to be dilated and the pulmonary valve dilated or thickened; in 3 of these congestive failure was clinically well documented.

A patent foramen ovale was observed in 29 cases, an atrial septal defect in 6. Thus, the mechanism for a right-to-left shunt was present in a large number.

Associated cardiovascular abnormalities have not been commonly reported: a probe-patent ductus arteriosus was found in 1 of our cases and a hypoplastic aorta with fetal coarctation in another.

\textbf{Differential Diagnosis}

The differential diagnosis is of considerable importance, since no established surgical procedure is available in Ebstein’s anomaly and exploratory thoracotomy is probably unusually hazardous.

Tetralogy of Fallot may be differentiated by a clinical course in which squatting, syncopal spells, dyspnea, and stunted growth are more prominent. Diastolic murmurs are uncommon; the electrocardiogram shows less block and more evidence of right ventricular hypertrophy; cardiomegaly is uncommon and a coeur en sabot rather than a globular contour is typical.

Tricuspid atresia usually is accompanied by
earlier and more marked symptoms, particularly cyanosis, dyspnea, poor growth, and limited activity. The electrocardiogram also shows large P waves but the considerable right intraventricular block is not usually present; left ventricular dominance or hypertrophy is the rule. The cardiac contour is also quite different in that it has a straight right border or it mimics dextrocardia or coeur en sabot.

Severe pulmonic stenosis with intact ventricular septum, particularly when a right-to-left shunt occurs at the atrial level, represents the most difficult differential diagnosis. The similarity in radiologic appearance has been noted. The typical stenotic, well-transmitted systolic murmur at the pulmonic area accompanied by a thrill, absent diastolic murmur, the observation of presystolic pulsation in neck veins or liver, coupled with definite right ventricular hypertrophy by electrocardiogram usually identifies the malformation. On occasion cardiac catheterization may be the only means of differentiation; to date, elevated right ventricular systolic pressure and a significant gradient across the pulmonary valve have not been described in Ebstein’s syndrome.

The auscultatory findings suggest complete transposition of the pulmonary veins. Marked right ventricular hypertrophy by electrocardiogram and engorged pulmonary vascular markings by x-ray differentiate this condition. Blount has pointed out the similarity of auscultatory findings in adult patients to those of rheumatic heart disease.45

Summary

Ten cases of Ebstein’s anomaly of the tricuspid valve have been presented. The findings in these cases have been correlated with those reported in the literature to emphasize certain clinical features that are of diagnostic value. 1. Symptomatology is usually mild with cyanosis, dyspnea, fatigability, and commonly a history of bouts of palpitation. 2. The physical findings are characterized by normal growth, frequent cyanosis and infrequent clubbing; a quiet cardiac impulse and a systolic thrill between xiphoid and apex; a triple or quadruple rhythm, a second sound diminished at the pulmonary area, and a combination of systolic and diastolic murmurs maximal at the lower left sternal border or apex. 3. Phonocardiographic studies confirm the presence of a triple or quadruple rhythm, show a delayed first sound of normal intensity, demonstrate a systolic murmur of moderate intensity and medium frequency, and a presystolic murmur at the lower left sternal border and apex. Less constant is a mid-diastolic murmur in this area. 4. The characteristic electrocardiogram has tall P waves, frequently prolonged atrioventricular conduction, considerable right bundle-branch block, and right ventricular potentials of low amplitude. 5. By radiologic examination marked cardiomegaly is seen, a contour consistent with right-sided enlargement, a narrow base, diminished pulmonary vascular markings, and poorly delineated pulmonary artery. 6. Angiocardiography reveals a huge right atrial chamber with delayed emptying, poorly opacified pulmonary radicles, and frequent evidence of a right-to-left shunt. 7. Cardiac catheterization demonstrates a large atrium, displacement of the tricuspid valve to the left, moderately elevated right atrial pressure, normal right ventricular systolic pressure and absence of a significant gradient across the pulmonary valve. Peripheral arterial oxygen unsaturation is usual.

The importance of clinical recognition of this entity is stressed in view of the definite hazard involved in undertaking cardiac catheterization and surgical procedures in these patients.

Summario in Interlingua

Es presentate dece casos de anomalía de Ebstein del valvula tricuspid. Le constataiones in iste casos es correlationate con le datos reportate in le litteratura pro signalar emphaticamente certe aspectos clinic que es de valor diagnostic.

1. Le symptomatologia es usalmente leve, con cyanosis, dyspnea, fatigabilitate, e comunmente un historia de episodios de palpitação. 2. Le constatationes physic include characteristicamente un normal crescentia, frequentia de cyanosis, e infrequentia de digitos hippocratic; un quiete impulso cardiac e un fremito systolic inter xiphoide e apice; un rhythmo triple o quadruple, un secunde sono
diminuit in area pulmonar, e un combination de murmures systolice e diastolice que es maximal al margine sternal sinistro-inferior o al apice. 3. Studios phonocardiographic confirma le presentia de un triple o quaduple rhythm. Illos mostravera un retarde prime sono de intensitade normal, un murmur systolico de intensitade moderate e frequentia intermediari, e un murmur presystolico al margine sternal sinistro-inferior e al apice. Un murmur medio-diastolico in iste area es minus constante. 4. Le electrocardiogramma typic es characterisate per alte undas F, frequente per prolongate conduction atrioventricular, per considerabile grados de bloco de branca dextere, e per potentiales dextero-ventricular de basse amplitude. 5. Le examine radiologic revela marcate grados de cardiomegalia, un contorno compatible con allargamento al latere dextere, un base de largor reducite, diminuite marcas pulmo-vascular, e un delination pauc clar del arteria pulmonar. 6. Le examine angiocardiographic revela un enorme camera dextero-atrial con retardo de vacuum, imperfecte opacification del radicules pulmonar, e frequentemente signos de un derivation dextero-sinistre. 7. Catheterisation cardiae demonstra un grande atrio, displaciamiento del valvula tricuspe verso le latere sinistre, elevation moderate del pression dextero-atrial, normal nivellos del pression systolico dextero-ventricular, e absentia de un gradient significative trans le valvula pulmonar. Non-saturation oxygenic in le arterias peripherie es le regula.

Le importancia del recognition clinic de iste entitate es subliniate in vista del definite hasardo representate per le execution de catheterisation cardiae o de operationes chirurgic in casos de iste genere.

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REFERENCES

18 Walton, K., and Spencer, A. G.: Ebstein's
Ebstein’s Anomaly: Presentation of Ten Cases
FLORENCE E. MAYER, ALEXANDER S. NADAS and PATRICK A. ONGLEY

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