Ebstein’s Anomaly

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Four cases of Ebstein’s anomaly are described, revealing that the clinical features permit diagnosis in the majority of instances and that cardiac catheterization affords precise confirmation of the diagnosis. The occurrence of this anomaly in acyanotic adults is pointed out, and the similarity to acquired valvular rheumatic heart disease is stressed.

The congenital cardiac defect known as Ebstein’s anomaly has been recognized as a pathologic entity for 90 years. The first 80 years of this period constituted a relatively dormant era characterized by sporadic descriptions of the lesion at postmortem examination and by intimation that clinical recognition of this anomaly was not possible. Knowledge regarding Ebstein’s anomaly has accumulated rapidly following the introduction of cardiac catheterization. A rapidly expanding body of information has accumulated in the literature in the past 10 years that has totally dispelled the myth of the diagnostic impregnability of Ebstein’s anomaly. This recent experience has permitted the establishment of a definitive pattern of diagnostic clinical features. These features, when applied in the younger age patients, should lead promptly to the recognition of this anomaly.

The less well described occurrence of Ebstein’s anomaly is in adults without cyanosis. This type of patient with Ebstein’s anomaly constitutes a more formidable diagnostic challenge than does the cyanotic child in view of the possible confusion with acquired cardiac lesions. A description of the congenital malformation first described by Ebstein in 1866 is considered to be pertinent at this time. Although the details in the individual case have varied somewhat, in general this malformation consists of an abnormality of the leaflets and origin of leaflets of the tricuspid valve. This consists of a fusion of the leaflets, particularly the septal and posterior leaflets, into a membranous structure extending into the cavity of the right ventricle and separating the right ventricle into a proximal and distal chamber. The proximal portion, which consists of the sinus of the right ventricle, is continuous with the right atrium, while the distal portion, composed of the outflow tract of the right ventricle, functions as the right ventricle. The anterior leaflet of the valve is frequently normal; however, the septal and posterior leaflets are deformed and may be completely fused with the endocardium of the ventricle and not attached to the annulus fibrosus.

A defect in the atrial septum is almost invariably present and may consist of an anatomically patent foramen ovale or true atrial septal defect. The thin ventricular wall of the atrialized portion of the right ventricle has been considered as an integral part of the malformation. However, this view is not definitely established, and Edwards believes that the thinning may be the result of the altered dynamics that exist secondary to the deformity of the valve rather than a primary abiotrophy of the muscle tissue. The distal or functional portion of the right ventricle may reveal hypertrophy of the musculature. An excellent drawing of this anomaly is included in the article on Ebstein’s anomaly by Engle and associates.

The purpose of this communication is to present clinical and hemodynamic data from 4 patients with Ebstein’s anomaly. Two patients represent the classical picture of the lesion in childhood. The remaining 2 patients were acyanotic adults presenting clinical features suggesting acquired heart disease. Emphasis is placed on the elements aiding in the establishment of a diagnosis in this latter group.
CASE REPORTS

Case 1. E. K. was admitted to the Pediatrics Service at Colorado General Hospital on June 7, 1949, at the age of 2 weeks because of cyanosis. The baby was the product of a normal pregnancy and an uneventful delivery and was a nonidentical twin. The fraternal twin was found to be normal in all respects. The patient was noted to be cyanotic at 5 days of age.

Physical examination revealed a well developed, well nourished baby weighing 6 lb. 14 oz., who was questionably cyanotic. The heart size was normal by palpation, and no thrills were felt. A sinus rhythm with a rate of 150 beats per minute was found. A grade I systolic murmur was heard in the third and fourth intercostal spaces at the left sternal border. The remainder of the examination was normal except for the presence of a supernumerary nipple on the right side of the abdomen.

The electrocardiogram (fig. 1) demonstrated sinus rhythm, right axis deviation, and a vertical electric position. The QRS interval was normal in duration, being 0.07 second. An R wave 11 mm. in amplitude was present in lead V2 with an S wave of 2 mm. The intrinsicoid deflection time was 0.02 second in this lead. The tracing was interpreted as suggestive of right ventricular hypertrophy on the basis of the R-S ratio.

Roentgenologic examination (fig. 2) revealed a normal vascularity of the lung fields. There was a conspicuous concavity in the area of the main pulmonary artery segment. The right atrium was considerably enlarged, dominating the over-all cardiac silhouette. The ventricular mass appeared normal in size.

A definitive cardiac diagnosis was not established at that time and the patient was discharged. Shortly after discharge from this hospital, cyanosis was again observed by the parents and the baby was rehospitalized elsewhere for a 4-week period. On returning home, the baby gained weight very slowly and was retarded in development, lagging behind the twin both physically and mentally. Episodes of severe dyspnea associated with deep cyanosis occurred on 5 occasions during the first year of life. Profuse sweating of the head and neck was observed repeatedly throughout infancy. The child began to walk at 15 months of age and by that time showed only minimal limitation of exercise tolerance. No squatting was observed at any time.

The patient was first seen by the Cardiology Service when 25 months of age. The physical examination at that time showed no cyanosis. The heart was enlarged by palpation to the anterior axillary line in the left fifth intercostal space. A systolic thrill was palpable along the lower left sternal border. A systolic murmur was audible over the entire precordium and back, with maximum intensity at the lower left sternal border. The murmur was grade IV in intensity and blowing in quality. A protodiastolic sound was audible at the base of the heart producing a gallop rhythm. The lower border of the liver was palpable 2 cm. below the right costal margin.

The electrocardiogram revealed a distinct change from the previous tracing in that the QRS interval had widened to 0.12 second with a resulting pattern of right bundle-branch block. The P waves had increased in amplitude in V2.

Fluoroscopic examination revealed a decreased vascularity of the lung fields, a normal size of the right and left pulmonary arteries, and a concavity in the region of the main pulmonary artery. The right atrium was considerably enlarged. The ventricular area was also enlarged with no specific configuration (fig. 2).
The child continued to be active, keeping up with the twin in all but very vigorous activities. Her weight and height remained less than the twin. Repeated examinations up to the age of 5½ years revealed a varying location and intensity of the cardiac murmurs. At one period, a loud to-and-fro murmur was audible along the left sternal border. The electrocardiographic and roentgenologic patterns remained essentially unchanged during this period (figs. 1 and 2). These clinical features suggested the presence of Ebstein's anomaly.

Cardiac catheterization was carried out when the patient was 5½ years of age (table 1). Sinus tachycardia was maintained throughout the study except for the occurrence of short runs of ventricular premature syostoles on several occasions when the catheter tip lay in the right ventricle. There was no evidence of a shunt. The calculated cardiac index was within normal limits. Normal arterial oxygen saturation was present both at rest and following exercise. The right ventricular systolic pressure was normal at 24 mm. Hg. The right atrial pressure tracing...
showed a moderately prominent a wave. A slight pressure gradient of 5 mm Hg was present between the right ventricle and pulmonary artery. There was moderate difficulty in introducing the catheter into the right ventricle. The point at which an atrial form of pressure wave changed to a ventricular form was observed to lie well toward the left border of the cardiac silhouette.

The catheterization data in conjunction with the clinical features were considered to establish the diagnosis of Ebstein’s anomaly.

Case 2. T. R. was an 8-year-old white boy seen for the first time on May 8, 1953. He was the product of a normal pregnancy and delivery, and his growth and development were normal. The parents first noted cyanosis when he was 2 years of age. Moderate exertional dyspnea also appeared at 2 years. He was never able to keep up with other children but was able to walk half a mile at a slow pace. A doctor was first consulted when the patient was 4 years of age and the mother was informed that “something was wrong with his heart.”

Physical examination showed the patient to be a well developed, well nourished boy with cyanosis of the lips and nailbeds. There was a slight bulging of the precordium, and the heart was slightly enlarged by percussion. A systolic thrill was palpable along the lower left sternal border. Auscultation revealed a harsh systolic murmur of grade IV intensity over the entire precordium with maximum intensity in the fourth and fifth left intercostal spaces para-sternally. The second heart sound in the left second intercostal space was normal in intensity and pure in quality.

The electrocardiogram (fig. 3) demonstrated complete right bundle-branch block with very tall P waves in the right precordial leads, suggesting right atrial enlargement. Roentgenologic examination (fig. 4A) demonstrated a slightly decreased vascular pattern of the lung fields with small, quiet right and left pulmonary arteries. The aorta appeared normal but the main pulmonary artery was inconspicuous. The superior vena cava was seen as a prominent shadow at the base of the heart on the right. The right atrium was moderately enlarged with a considerable cephalic element to the enlargement. The right ventricular area was moderately enlarged with a fullness high along the left cardiac border. Oblique views showed that the left ventricle and left atrium were normal in size and configuration. The over-all clinical picture suggested Ebstein’s anomaly.

Cardiac catheterization was performed on June 22, 1953 (table 1). The right ventricle and pulmonary artery were intubated only with difficulty, the catheter repeatedly coiling in the large right atrium. The point at which pressure contours changed from atrial to ventricular in form was observed to be far to the left of the midline, well within the usual region of the body of the right ventricle. A rather sharp angulation of the catheter course occurred on advancing the catheter through the cavity of the right ventricle and into the pulmonary artery. A paroxysmal supraventricular tachycardia with a rate of 180 beats per minute occurred during the procedure; 4 minutes after the manipulation of the catheter was stopped, it reverted to a sinus rhythm. There was no evidence of left-to-right shunt at the atrial levels. A significant right-to-left shunt was present with an estimated volume of 2.0 L. per minute. The right atrial pressure tracing initially showed prominent a waves averaging 12 mm Hg in amplitude (fig. 5). During the supraventricular tachycardia, high pressure waves were recorded that were considered to represent tricuspid insufficiency. Following reestablishment of sinus rhythm, a pressure pattern of mild tricuspid insufficiency was evidenced by a pressure plateau throughout the period of ventricular systole. The right ventricular pressure tracing showed a prominent pressure wave occurring in late ventricular diastole with the same configuration and time characteristics as the a wave recorded in the right atrium. The systolic pressure in the right ventricle was normal, averaging 24 mm Hg. A mild pressure gradient of 4 mm Hg occurred between the right ventricle and pulmonary artery. The pulmonary arterial pressure wave demonstrated a variable contour. The pressure curve illustrated in figure 5, recorded in the main pulmonary artery, reveals a small

![Figure 3: Right precordial leads in case 2 (top) and case 4 (bottom) demonstrating prominent P waves and complete right bundle-branch block with low amplitude R waves.](http://circ.ahajournals.org/)

**FIG. 3.** Right precordial leads in case 2 (top) and case 4 (bottom) demonstrating prominent P waves and complete right bundle-branch block with low amplitude R waves.
Fig. 4. Cases 2(A), 3(B) and 4(C). All 3 films demonstrate a prominent right atrium, a narrow pedicle with an inconspicuous main pulmonary artery, and a globular configuration to the enlarged heart.

Fig. 5. Case 2. The right atrial pressure tracings show a prominent a-wave initially, a high wave of tricuspid insufficiency during tachycardia, and a mild pattern of residual insufficiency immediately following the tachycardia. A pressure wave occurring at the time of right atrial contraction is present in the pulmonary arterial and right ventricular tracings.

late diastolic wave coincident in timing with the a wave in the right atrium.

Venous angiography was carried out on June 24, 1953, without incident. The study demonstrated the presence of a large right atrium in which contrast media lingered throughout the entire series. Early filling of the left atrium was evident with subsequent good filling of the left ventricle and aorta. A suggestion of a small deformed right ventricle was present. Opacification of the main pulmonary artery segment and lung fields was faint (fig. 6).

The clinical pattern and the diagnostic studies were considered clearly to confirm the diagnosis of Ebstein's anomaly.

Case 3. L. B., a 21-year-old white man, was seen at Colorado General Hospital for the first time on November 23, 1955, for evaluation of a heart murmur. This murmur was first detected during early childhood. The patient grew and developed normally and was able to keep up with companions in
all activities. In school he engaged in team athletics. For the past 4 years he had worked as a truck driver and loader without difficulty. He noticed exertional dyspnea only on prolonged strenuous activity. In October 1955, the patient consulted a physician because of a respiratory tract infection. A cardiac murmur was heard and the patient referred to this service for further evaluation.

Physical examination revealed a well developed, vigorous young man. A mild violaceous hue was present over the malar eminences. A presystolic venous pulsation was noted in the neck when the patient was recumbent. Palpation over the precordium revealed a diffuse cardiac thrill in the left fifth intercostal space between the midclavicular and anterior axillary lines. A faint systolic thrill was palpable at the apex. Auscultation revealed a grade IV, blowing systolic murmur at the apex that was transmitted into the left axilla. A low pitched, middiastolic murmur of grade II intensity was heard at the apex. The first heart sound in this area was accentuated. The second sound heard in the pulmonary area was diminished in intensity.

The electrocardiogram (fig. 7) showed complete right bundle-branch block. Tall, wide P waves were present in leads II, aVF, and in the right precordial leads, indicating right atrial enlargement. Roentgenologic examination revealed a decreased vascularity in the peripheral lung fields (fig. 4B). The right and left pulmonary arteries were small in size and quiet in activity. The aorta was small and the main pulmonary artery was so inconspicuous as to prevent identification in the frontal view. The right atrium was markedly enlarged, projecting far to the right and superiorly. The right ventricular area appeared considerably enlarged, filling the greater portion of the retrosternal space. A conspicuous fullness high along the left cardiac border tended to produce a globular configuration of the greatly enlarged over-all cardiac silhouette. The left heart chambers were normal.

A diagnosis of Ebstein's anomaly was made on the basis of the clinical features. Cardiac catheter-
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Fig. 8. Case 3. The position of the tricuspid valve as determined by a change in the pressure wave from ventricular to atrial in type is shown to lie considerably to the left of the midline.

Fig. 9. Case 3. The right atrial tracing, R.A., shows an augmented a-wave. The right ventricular tracing, R.V., demonstrates a broadened and delayed right ventricular contraction wave. The pressure tracing from the main pulmonary artery shows a prominent late diastolic pressure wave.

zation was performed for evaluation of the hemodynamic status. Marked enlargement of the right atrium was demonstrated by the large diameter loop formed by the catheter as it repeatedly coiled in this chamber. The right ventricle and pulmonary artery were intubated with difficulty, the catheter being somewhat angulated when its tip lay in the pulmonary artery. The point at which right ventricular pressure pulses changed to a right atrial pressure pattern was observed well to the left of the midline (fig. 8). No shunts were demonstrable at rest. A decrease in brachial arterial oxygen saturation to 89.2 per cent following exercise was interpreted as indicating a small right-to-left shunt.

The pressure tracings from the right heart chambers are shown in figure 9. In the right atrium the a wave was 10 mm. Hg in amplitude at rest and 13 mm. Hg on exercise. The right ventricular pressure tracing demonstrated a high amplitude end-diastolic pressure wave resulting from right atrial contraction. The systolic pressure in the right ventricle was within normal limits and did not rise on exercise. A pressure gradient of 7 mm. Hg at rest and 12 mm. Hg on exercise was present at the pulmonic valve level. The pulmonary arterial pressure tracing showed a prominent pressure wave in late diastole that was considered to reflect right atrial contraction.

A sinus rhythm was maintained throughout the major portion of the study. Following exercise, after blood samples had been obtained and pressures determined, atrial flutter began with varying 2:1 to 4:1 block. The ventricular rate averaged 132 beats per minute. The procedure was terminated and the patient was put at bed rest. The flutter changed to atrial fibrillation at the end of 1 hour, with a ventricular rate of 86. Spontaneous conversion to a normal sinus rhythm occurred 2 hours after the onset of the arrhythmia. These data clearly confirmed the diagnosis of Ebstein’s anomaly.

Case 4. Mrs. M. P., a 35-year-old white woman, was seen for the first time at Colorado General Hospital on September 2, 1954, for an evaluation of cardiac murmurs. A cardiac murmur and cardiac enlargement had first been detected 8 years previously. Since that time mild fatigue and dyspnea were noted on severe exertion. Repeated episodes of rapid heart action had occurred in the past 6 years, with rates of approximately 200 beats per minute and abrupt onset and termination. Recurrent throat infections occurred since childhood.

Physical examination revealed a well developed, well nourished woman with flushing of the malar eminences of the face. In the left lateral decubitus position a short, presystolic thrill was felt at the apex. A localized shock-like heave was palpable at the lower left sternal border. The heart was enlarged to the anterior axillary line on percussion. Auscultation revealed reduplication of the first sound in the
right second intercostal space and a grade II systolic murmur in this area. The second sound in the left second intercostal space was reduplicated and was of greater intensity than the second sound in the aortic area. The first sound at the cardiac apex was accentuated, producing a staccato quality. A second heart sound of normal intensity was present at the apex and was followed by a faint third heart sound. A low pitched, diastolic murmur with presystolic accentuation was heard immediately after the third heart sound. A grade II, blowing systolic murmur was present at the lower left sternal border. This murmur increased in intensity during inspiration.

Electrocardiography revealed a prolonged QRS interval of 0.12 second with a pattern of complete right bundle-branch block (fig. 3). High amplitude broadened P waves were present in the right preordial leads.

Roentgenologic examination (fig. 4C) showed the pulmonary vascularity to be normal. The right and left pulmonary arteries and the main pulmonary artery were normal in size and in amplitude of pulsation, while the aorta was small and quiet. The over-all heart size was moderately increased owing to the enlargement of the right heart chamber.

Cardiac catheterization was carried out without incident on October 19, 1954 (table 1). At the time of cardiac catheterization, the diagnosis was rheumatic heart disease with mitral and tricuspid valve involvement. Right heart chamber samples were not obtained, and the catheter position on withdrawal from the right ventricle to the right atrium was not specifically observed. Full arterial oxygen saturation was present at rest. Following exercise, moderate unsaturation to 84.7 per cent occurred. The cardiac output was significantly reduced at rest, and rose only moderately on exercise. The pressure tracing recorded in the right atrium showed prominent α waves with an amplitude of 10 mm. Hg (figs. 10 and 11). The right ventricular pressure tracing demonstrated a prominent late diastolic pressure wave resulting from right atrial contraction. A mild pressure gradient of 5 mm. Hg was present between the right atrium and right ventricle in early diastole, suggesting some degree of tricuspid stenosis. The peripheral pulmonary arterial pressure tracing showed a prominent secondary pressure wave in late diastole that corresponded in timing with the α wave of the right atrium. A systolic pressure gradient of 8 mm. Hg at rest, and 11 mm. Hg on exercise was present between the right ventricle and pulmonary artery. During exercise, prior to drawback of the catheter into the ventricle, the pressure in the pulmonary artery rose as high as 45/15 mm. Hg. Normal pulmonary arterial wedged pressure levels were recorded both at rest and with exercise.

These data obtained by cardiac catheterization failed to substantiate the diagnosis of rheumatic heart disease with mitral valve impairment. The possibility of acquired tricuspid and pulmonic valve stenosis associated with a carcinoid tumor was considered. However, a subsequent review of the entire clinical and hemodynamic pattern led to the somewhat delayed realization that all the criteria for a diagnosis of Ebstein's anomaly were fulfilled.

Discussion

Symptomatology. The symptoms associated with Ebstein's anomaly are, for the most part, nonspecific and afford little aid in establishing a diagnosis. The symptoms occurring in children with the cyanotic form of this anomaly include such general features as inadequate development and weight gain, excessive perspiration of the head and neck, and limitation of exercise tolerance. The mild degree of these symptoms with cyanosis often occurring later in childhood is of minor diagnostic value in that the most common lesion producing cyanosis in children, the tetralogy of Fallot, tends to be excluded. The finding in case 2 that a cyanotic child could walk half a mile, did not squat, and had no episodes of paroxysmal dyspnea, suggested the presence of anomalies other than the tetralogy of Fallot.

The symptoms in noncyanotic patients, particularly those in the age groups where acquired valvular heart disease is common, are almost totally devoid of diagnostic significance. Relatively good exercise tolerance in the presence of considerable cardiomegaly, as in case 3, might be considered as a possible diagnostic aid.

The occurrence of paroxysmal tachycardia constitutes the single valuable feature in the symptomatology of Ebstein's anomaly. This symptom was a salient feature in case 4 in this series and was helpful in the establishment of the diagnosis.

The natural course of patients with this anomaly varies widely, from the occurrence of death in infancy to the maintenance of good health until late in life. Congestive failure may occur terminally, as was the case in the initial patient reported by Ebstein. Death presumably due to systemic emboli may occur as a result of embolization through a defect in the atrial septum. Sudden and unexpected death is a common terminal event in patients with this lesion. The mechanism responsible for this event has been con-
sidered to be related to the paroxysmal arrhythmias characteristic of Ebstein’s anomaly.

**Physical Examination.** The information derived by physical examination may more often be misleading than helpful in the recognition of Ebstein’s anomaly. The cyanotic child constitutes a somewhat less confusing problem on examination than does the acyanotic adult, inasmuch as the diagnostic possibilities are usually limited to congenital lesions. The murmurs accompanying this lesion in the child afford the least important diagnostic information. Location, intensity, and timing of murmurs vary widely, not only from patient to patient, but at times in the same patient on serial examinations, as demonstrated in case 1.

The critical feature of the physical examination consists in the recognition that right ventricular hypertrophy is not present. This is suggested by the absence of a lift of a hypertrophied right ventricle along the lower left sternal border, and by the absence of signs of pulmonic stenosis or of pulmonary hypertension, the 2 common causes of right ventricular hypertrophy. Cyanosis in the absence of right ventricular hypertrophy should immediately suggest the possibility of Ebstein’s anomaly. The physical findings in the adult with Ebstein’s anomaly rarely alert the examiner to the correct diagnosis. Mild cyanosis, if present, is readily attributable to the peripheral cyanosis associated with rheumatic tricuspid valvular disease. If less common cardiac diseases are considered, the mild cyanosis may even suggest the flushing associated with carcinoid heart disease. Venous pulsations in the neck, as seen in case 3, are easily assigned to acquired tricuspid valve disease. The cardiac murmurs, present in cases 3 and 4 in this series and in many of the adults described in other series, closely mimic the murmurs of rheumatic mitral and tricuspid valve disease. The murmurs in patients with Ebstein’s anomaly undoubtedly arise primarily as a result of stenosis and insufficiency of the deformed tricuspid valve. The displacement of this valve to the left explains the frequent apical location of these murmurs. The similarity of the auscultatory features of rheumatic mitral disease and Ebstein’s anomaly is strikingly illustrated by the finding of an accentuated first heart sound and an opening snap at the apex in case 4. These characteristics of the heart sounds that have classically been limited to mitral valve disease can apparently arise from the tricuspid valve in Ebstein’s anomaly.12, 24

This bewildering overlapping of signs in Ebstein’s anomaly and rheumatic valvular disease permits the suggestion that a significant number of adults with this congenital anomaly are being erroneously considered examples of rheumatic heart disease.

**Electrocardiography.** The electrocardiogram offers valuable diagnostic aid in patients with Ebstein’s anomaly. A classic feature that occurs in a high percentage of cases is complete right bundle-branch block with low amplitude R waves in the right precordial leads.6 This form of right bundle-branch block is usually readily distinguished from that associated with severe right ventricular hypertrophy, where tall R waves are present in the right precordial leads. The characteristic configuration of the QRS complex is demonstrated in cases 2, 3, and 4 (figs. 3 and 7), and in case 1 after the age of 2 weeks (fig. 1).

A different configuration of the QRS complex occurs infrequently in patients with Ebstein’s anomaly. The most common variant described is incomplete right bundle-branch block.6, 8, 11, 12, 24-26 Complete right bundle-branch block with very tall R waves in the right precordial leads has been demonstrated in 1 case by Kjellberg and associates.12 This very unusual pattern suggesting severe right ventricular hypertrophy was present despite a right ventricular systolic pressure of only 33 mm. Hg. The presence of free tricuspid insufficiency in this patient was considered an important element leading to the development of right ventricular hypertrophy. A right ventricular hypertrophy pattern with a normal QRS interval represents an additional unusual variation from the classic electrocardiogram. Evidence of right ventricular hypertrophy with a normal QRS duration is present in the right precordial leads in case 3 of Brown and associates.26 A normal QRS duration is demonstrated in case 2 in the present series at 2 weeks of age (fig. 1) with an R-S ratio suggesting right ventricular hypertro-
phy. Normal ventricular conduction time with no evidence of right ventricular hypertrophy was found in case 1 in the series reported by Broadbent and associates. Further unusual patterns include incomplete bundle-branch block of indeterminate type as seen in case 4 of Brown, and complete left bundle-branch block as described by Adams and Hudson.

The occurrence of these variant forms of QRS complexes permits a reconstruction of the development of the classic pattern of complete right bundle-branch block. The finding of normal conduction and intermediate degrees of incomplete block suggests that the conduction impairment is not an intrinsic element of the anomaly caused by a defect in the conduction system itself, but is a result of changes in structure of the right ventricle due to the physiologic stress imposed by the tricuspid valve deformity. The serial tracings from case 1 (fig. 1) offer conclusive evidence that intraventricular conduction may be normal shortly after birth and be progressively lengthened with increasing age. It is postulated that the right bundle-branch block arises as a result of the dilatation and thinning of the atrialized proximal portion of the right ventricle. Normal conduction or partial degrees of right bundle-branch block may be present at an early age or in milder forms of the anomaly where this portion of the right ventricle is less markedly dilated.

Right ventricular hypertrophy is a disconcerting feature that considerably diminishes the diagnostic value of the electrocardiogram when present. Hypertrophy of the right ventricle has seldom been described at postmortem examination. However, hypertrophy of the distal portion of the right ventricle was described by Brown and associates in 2 patients, 1 of whom demonstrated electrocardiographic evidence of right ventricular hypertrophy. Inasmuch as a normal right ventricular pressure and a normal or decreased right ventricular output is the invariable physiologic pattern in this anomaly, it is difficult to account for this occasional right ventricular hypertrophy. A possible explanation is that the functioning portion of the right ventricle constitutes only a fraction of the total right ventricular mass and therefore is subject to a relative increase in work load in relation to a normal output. Evidence of severe right ventricular hypertrophy must be ascribed to other types of physiologic stress such as the marked tricuspid insufficiency present in the patient described by Kjellberg.

The occasional finding of left bundle-branch block is considered a manifestation of an associated cardiac lesion. Coronary atherosclerosis may well have been responsible for this phenomenon in the elderly patient described by Adams and Hudson.

The P-wave abnormalities are an equally consistent feature of the electrocardiogram in Ebstein's anomaly. Tall, broadened P waves characteristically occur in precordial leads overlying the right atrium and in the limb leads, reflecting the potential developed in this chamber. The increased amplitude of these P waves is clear evidence of hypertrophy of the right atrium. The broadened duration of these P waves frequently results in prolongation of the PR interval. The broadening and slurring of the P waves can be interpreted as reflecting dilatation of this chamber with consequent delay in conduction.

An additional electrocardiographic feature of Ebstein's anomaly is the occurrence of supraventricular arrhythmias. These may consist of paroxysmal atrial or nodal tachycardias and occasionally atrial flutter or fibrillation (case 3).

Roentgenography. Fluoroscopic examination of the patient with Ebstein's anomaly offers the most specific diagnostic evidence that can be obtained by the simpler methods of evaluation. A characteristic pattern is present that frequently will permit the establishment of a definitive diagnosis. Enlargement of the right atrium is the dominant radiologic feature in Ebstein's anomaly. The first film in the series, shown in figure 2, shows a very prominent right atrium in a 2-week-old infant. This prominence of the right atrium in case 1 progressed with increasing age. The right atrium enlarges anteriorly, encroaching on the retrosternal area, and also develops in a cephalad direction, elevating the point of juncture with the ascending aorta. The right ventricle is also...
enlarged and shows a rounded configuration with low amplitude pulsations. Bulging of the right ventricular outflow tract frequently produces a prominence high along the left cardiac border, a finding demonstrated in all patients in this series (figs. 2 and 4). The protrusion of the distal portion of the deformed right ventricle that is responsible for this roentgenologic configuration is well illustrated by Edwards.

The pulmonary arterial pattern is also characteristic on fluoroscopy. The main pulmonary artery is small and in conjunction with the dilated right ventricle immediately below may be scarcely detectable in the frontal view (fig. 4). The right and left pulmonary arteries are usually small and show small amplitude pulsations. The vascularity of the lung fields may appear normal or decreased, depending on the magnitude of the right-to-left shunt. The aorta is typically small and inactive.

The over-all pattern produced by these features is an enlarged, quiet heart with a globular configuration and a narrow pedicle usually accompanied by decreased vascularity of the lung fields. This pattern is generally distinctive from that of the tetralogy of Fallot or of isolated valvular pulmonic stenosis, 2 congenital lesions often confused with Ebstein's anomaly. The differential diagnosis in adults must include consideration of acquired tricuspid valve disease and pericardial effusion.

Angiocardiography has proved a useful diagnostic aid in this anomaly. The usual elements noted following injection of contrast material are a voluminous right atrium that remains opacified for a prolonged period, prompt filling of the left atrium through a patent foramen ovale in the cyanotic patient, a deformed right ventricular chamber situated well toward the left heart border, and poor visualization of the main and peripheral pulmonary arteries.

Cardiac Catheterization. Ebstein's anomaly has been established as a clinical entity largely as a result of information derived by cardiac catheterization. This diagnostic procedure continues to be the most effective means of confirming the presence of this anomaly. The emphasis on serious complications accompanying cardiac catheterization in the initial reports has not been substantiated in the more recent reports of larger numbers of patients. The arrhythmias occurring in cases 2 and 3 in this series were no more significant than similar arrhythmias occurring during cardiac catheterization in patients with other forms of congenital cardiac anomalies.

The principal diagnostic features on cardiac catheterization consist of a greatly enlarged right atrium, displacement of the tricuspid valve to the left, normal right ventricular, pulmonary arterial and pulmonary arterial wedge pressure, and absence of an arteriovenous shunt. These elements in a cyanotic patient are conclusively diagnostic. In an acyanotic adult considered to have rheumatic valvular disease, the hemodynamic pattern should reveal the diagnostic error. However, cardiac catheterization will not be diagnostic of Ebstein's anomaly unless the clinician includes this congenital defect in the precathe-terization differential diagnosis.

The specific details of the pressure phenomena recorded in the right heart chambers and pulmonary arterial system are of minor diagnostic value. They are of considerable value, however, in leading to an understanding of the altered hemodynamics associated with this unusual form of congenital heart disease. The result of contraction of the enlarged right atrium influences the pressure patterns obtained at all levels. An elevated mean pressure with a prominent a wave is the usual pattern recorded in the right atrium. This accentuated a wave reflecting hypertrophy of the right atrium can be considered to result from an impedance to normal emptying of the right atrium due to the small volume of the distal portion of the right ventricle, and to possible minor degrees of tricuspid stenosis. A pressure pattern of tricuspid insufficiency may occur less commonly in the right atrium. Free tricuspid insufficiency is demonstrated in the right atrial pressure tracing presented by Götzsche and Falholt, and by Kjellberg and associates. The series of right atrial pressure curves in figure 5 show the development of tricuspid insufficiency during tachycardia with
Fig. 10. Case 4. The right atrial pressure tracing shows an accentuated a wave. The pulmonary arterial tracing shows a bizarre, bifid wave with the secondary peak occurring at the time of right atrial contraction.

Fig. 11. Case 4. A continuous pressure tracing shows a systolic pressure gradient at the pulmonic valve level and a mild early diastolic pressure gradient at the tricuspid valve level.

a mild residual insufficiency following the arrhythmia.

The right ventricular pressure curves are characterized by normal systolic, normal early diastolic, and high end-diastolic levels. The high late diastolic pressure wave can be identified by its temporal relations and magnitude as resulting from right atrial contraction. This transmission of the pressure wave attending right atrial contraction to the right ventricle is illustrated in figures 5 and 10, and in the pressure tracings presented by Medd, Matthews, and Thursfield. It is reasonable to assume that this forceful end-diastolic filling is a significant aid in effecting maximum filling of the small distal portion of the right ventricle, and thereby assures maximum output from this chamber. A mild pressure gradient is present between the right atrium and right ventricle in early diastole in the pressure trac-
ing in case 4 (fig. 11) indicating a mild degree of tricuspid stenosis. The form of the right ventricular ejection wave itself is abnormal with widening, delay in onset, and delay in reaching a peak. This may reflect the abnormal electric activation of the right ventricle.

A systolic pressure gradient is frequently present between the right ventricle and pulmonary artery (fig. 11). This evidence of mild obstruction to right ventricular outflow was noted in all the present cases and in many of the cases previously reported.\textsuperscript{11,12} Postmortem studies of the heart in Ebstein's anomaly seldom reveal actual pulmonic valve stenosis. This obstruction, therefore, is probably a manifestation of a hypoplastic pulmonic valve ring and pulmonary artery. The occurrence of pulmonic hypertension during exercise in case 4, with a normal pulmonary arterial wedge pressure is a further suggestion that the pulmonary arterial system is decreased in capacity.

The pulmonary arterial pressure tracings demonstrate an unusual series of pressure waves that vary considerably in configuration in different regions of the pulmonary arterial system. It is possible to identify a pressure wave with temporal relationships similar or identical to those of the augmented a wave in the right atrium (figs. 9–11). Similar right atrial pressure activity was described by Black et co-workers\textsuperscript{6} in the pulmonary arterial pressure tracing of a patient with Ebstein's anomaly. The presence of right atrial pressure activity superimposed on pulmonary arterial tracings\textsuperscript{31} and even systemic arterial tracings\textsuperscript{32} has been noted in other forms of cardiac disease. A comparison of the pressure levels in the pulmonary artery and in the right atrium at the time of right atrial contraction in end-diastole shows that the right atrial pressure may equal or exceed that in the pulmonary artery at that phase of the cardiac cycle. This pressure relationship suggests that right atrial contraction may affect pulmonary arterial filling and contribute in some degree to the propulsion of blood into the pulmonary arterial system.\textsuperscript{8} This phenomenon would assign to the right atrium the role of an accessory right ventricle.

\textit{Therapy.} A satisfactory surgical procedure has not as yet been developed for correction of the cardiac defect in Ebstein's anomaly. This situation is readily understandable in view of the nature of the basic abnormality, consisting as it does of an inadequate propulsive force generated by the deformed right ventricle. Direct manipulation of the deformed tricuspid valve, even with the ample open operative period afforded by present techniques, would not appear to be a fruitful approach to the correction of this anomaly.

Two types of palliative surgical procedures have been performed in patients with Ebstein's anomaly. The first of these is the Blalock-Taussig anastomosis. This procedure theoretically offers the possibility of palliation in patients with right-to-left shunts of large volume through an atrial communication. This method thus utilizes the left ventricle as a partial substitute for the ineffective right ventricle to achieve a more normal pulmonary blood flow. Despite the theoretical benefit to be derived from a Blalock-Taussig anastomosis, its application in patients with Ebstein's anomaly has been unrewarding. Operative fatality has occurred in all instances, regardless of whether the correct diagnosis was established preoperatively,\textsuperscript{8} or whether the erroneous diagnosis of a tetralogy of Fallot\textsuperscript{11,26} was made.

A second type of palliative surgical procedure consists of closure of the communication between the 2 atria. This has been accomplished in a 25-year-old man with Ebstein's anomaly, reported by Wright and associates,\textsuperscript{10} with relief of cyanosis and improvement in exercise tolerance. As the authors point out, if nothing else, the procedure results in the elimination of the right-to-left shunt with its attendant hazard of thromboembolic disease from paradoxical emboli or from the secondary polycythemia. In addition to this benefit, the closure of this outlet to right atrial outflow may result in an increased right atrial pressure and consequent enhancement of the role of the right atrium as an accessory right ventricle. This possible benefit from an increased right atrial pressure might, however, be achieved only at the price of hastening the onset of con-
gestive failure. Operative fatality associated with closure of an atrial defect has been mentioned in 2 patients with Ebstein’s anomaly since this initial successful case of Wright.

Operative intervention has not been recommended in any of the 4 patients reported here.

**Summary**

Four cases of Ebstein’s anomaly are presented that demonstrate the characteristic clinical and hemodynamic features of this congenital cardiac defect. Two of these patients were acyanotic adults who presented auscultatory findings closely resembling those of acquired mitral and tricuspid valve disease. It is suggested that a significant number of patients with Ebstein’s anomaly are mistakenly considered to have rheumatic valvular heart disease.

**Summario in Interlingua**

Es presentate quatro casos del anomalía de Ebstein que exhibi le characteristic aspectos clinic e hemodynamic de iste congenite defecto cardiac. Duo del patientes eseva adultos acyanotic qui presentava constatations auscultatori multo simile a illos de acquirir morbo de valvula mitral e tricuspid. Es stipulate que un numero significative de patientes con anomalía de Ebstein es erroneemente considerate como affigite de rheumatic morbo de valvula cardiac.

**REFERENCES**


Biosynthetically labeled C¹⁴-digitoxin was administered intravenously in multiple doses to 3 terminal patients. The various tissue samples at autopsy were assayed for both the unchanged drug and its metabolic products. The myocardium does not have any special affinity for the cardiac glycoside in comparison to other organs. On a tissue-weight basis, the kidney, gallbladder contents, and entire intestines have the highest concentration of unchanged digitoxin, whereas the spleen, jejunal contents, and gallbladder contents have the highest concentration of metabolic products. On a whole organ basis, the liver has the largest amount of both digitoxin and its metabolites. The hypothetic scheme of the possible course of events based on the results is as follows: After its intravenous administration, there is a rapid initial removal from the vascular system as evidenced by the disappearance of approximately 60 per cent of drug from the blood stream within 15 min. after its administration. During this period, as well as after the digitoxin blood level has reached equilibrium, some of the drug is metabolized by the liver and both the glycoside and its metabolic products enter the gastrointestinal tract via the biliary route. A major portion of the metabolites and some of the unchanged digitoxin are then reabsorbed by the small intestine and enter the enterohepatic cycle. With this passage, small amounts of the metabolic products and lesser amounts of the unchanged drug are continuously removed from the vascular system by the kidney. This accounts for the greater excretion of the drug and metabolites by way of the kidneys than by way of the feces.

Aviado
Ebstein's Anomaly
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Circulation. 1957;15:210-224
doi: 10.1161/01.CIR.15.2.210
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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