Value of an Electrode Catheter in Diagnosis of Ebstein’s Disease

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Various features of Ebstein’s disease, an unusual congenital malformation of the tricuspid valve, are discussed briefly. Cardiac catheterization with the use of an electrode catheter in such a case is described. The pathophysiologic changes as illustrated by simultaneous intracardiac potentials and pressures are discussed.

EBSTEIN’S disease is a congenital malformation and displacement of the tricuspid valve, in which the 3 leaflets of the valve are displaced distally and attached directly to the ventricular wall.1-3 Frequently individual leaflets cannot be identified and are represented only by remnants of a basket-like arrangement of fibers and cords. The deformed valve divides the right ventricle into 2 chambers—a distal one in the region of the right ventricular outflow tract with walls of normal thickness and a proximal grossly dilated chamber with markedly thin walls upon which the tricuspid valve or its representatives are adherent. This proximal chamber is continuous with a dilated and generally hypertrophied right atrium. In most cases an interatrial septal defect or patent foramen ovale is present.

A clinical picture has evolved so that one may suspect the diagnosis with a high degree of accuracy during life.1, 3-7 On occasions, however, the picture may be confused with acquired tricuspid or mitral valvular disease, pulmonic stenosis with interatrial septal defect, or pericardial effusion. Indeed, operations have been performed inadvertently in the past in patients with Ebstein’s disease with dire results.1, 4, 8, 10

On routine cardiac catheterization the diagnostic points reported include the presence of a large dilated right atrium; and 2 areas of different pressures to the left of the spine, presumably from the right ventricle, with the proximal low “ventricular” pressure being indistinguishable from right atrium.3, 6, 11, 12 The sole factor in attributing pressures to be right “ventricular,” however, was the location of the catheter in relation to the spine. Normal pressures in the pulmonary circuit and frequently a shunt at the atrial level are additional findings.

The utility of an electrode catheter was first suggested when Sodi-Pallares and associates18, 14 described their method of obtaining monophasic ventricular potentials by the use of an exploring electrode pressed against certain points of the interventricular septum in a case of Ebstein’s disease. Based upon the observations of Sodi-Pallares, Hernandez and his associates15 reported in 3 cases of Ebstein’s disease right ventricular intracavity electrocardiographic patterns in the portion of the right ventricle near the tricuspid valve where low pressure was recorded. Pathologic studies support the thesis of identifying ventricular musculature in a zone where “atrial” pressures are obtained. With simultaneous recording of intracardiac electric potentials and pressures in a clinically suspected case, the location of the catheter within the heart was clearly identified, and 2 right ventricular chambers with different pressure patterns were demonstrated. Emslie-Smith and co-workers16 also suggested using the electrode catheter to define anatomic lesions in congenital heart disease, with particular reference to the localization of pulmonic stenosis.

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This study was supported in part by a grant-in-aid from the National Heart Institute, by a grant from the American Heart Association partially supported by the Cayuga County New York Heart Chapter, and by the Hochstetter Fund and Ernest L. Woodward Fund.
CASE REPORT

J. K. #338710, a Negro woman, was first seen at age 24 at the University of Rochester Medical Center in January 1952 because of abdominal pain. The pain was thought to be due to pelvic inflammatory disease. Surgery was deferred because of findings relative to the heart.

She gave a history of heart disease since childhood with limitation of activity, but there was no suggestion of rheumatic fever. She reported a recent increase in shortness of breath on exertion, and repeated “heart attacks” in the past. These were characterized by apprehension, palpitations, frequent fainting, dusky nails, but not chest pain.

She had been hospitalized elsewhere for 1 year previously for a spontaneous abortion. Two years before an appendectomy and removal of a left ovarian cyst were performed.

On examination she appeared apprehensive but not acutely ill. The blood pressure was 100/70 mm. Hg. There was no evidence of clubbing of the fingers nor of cyanosis. The lungs were clear. The maximum cardiac impulse was seen diffusely in the fifth and sixth intercostal spaces at the left anterior axillary line. A soft systolic thrill was felt along the left sternal border, extending to the apex. A short, harsh systolic murmur of grade IV intensity was heard loudest along the left sternal border in the third and fourth interspaces and was transmitted over the entire precordium. A third heart sound was present in the same area as the systolic murmur. Diastolic murmurs could not be definitely identified. The abdomen was tender to deep palpation in the lower quadrants. The liver was not palpable.

Routine laboratory data showed a negative serologic test for syphilis and the absence of polycythemia.

The electrocardiogram showed a sinus rhythm with a rate of 72, P-R interval of 0.18 second, a QRS of 0.12 second, and was interpreted as showing an intraventricular conduction defect (probably right bundle-branch block), abnormal peaked P waves, and inverted T waves over V_5-6 (fig. 1). The chest film showed a generally enlarged heart of globular shape with clear lung fields (fig. 2).

A phonoangiogram confirmed the systolic murmur and third heart sound, and also suggested the presence of a short presystolic murmur along the left sternal border at the third and fourth intercostal spaces. Fluoroscopy on the horizontal table again showed generalized heart enlargement without hilar pulsation. It appeared that the right ventricle was particularly enlarged, although there was a wide swing of the esophagus on barium swallow. A cineangiogram was technically poor because of a spontaneous Valsalva maneuver and because of dilution defects in the large heart.

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**Fig. 1.** The routine 12-lead electrocardiogram shows peaked P waves and prolonged intraventricular conduction resembling right bundle-branch block. The long leads of II and V_1 are representative tracings during a bout of supraventricular tachycardia.

**Fig. 2.** Chest film.
However, it was thought to be suggestive of an interatrial septal defect. The arterial oxygen saturation was 88.6 per cent at rest.

The patient was discharged after a stay of 10 days. She was advised to visit the cardiac clinic but was not seen again until June 1954, when she returned with a supraventricular tachycardia at a rate of 160. In the preceding 2 years she had experienced “heart attacks” repeatedly, 9 in the month before admission. Examination was unchanged except for a definite presystolic murmur heard along the left sternal border at the third and fourth intercostal spaces. The arrhythmias stopped spontaneously. On this admission the diagnosis of Ebstein’s disease was suggested.

Cardiac catheterization was performed on January 25, 1955, and an electrode catheter was used. Arrhythmias were not encountered. Simultaneous recordings of intracardiac pressures and potentials were obtained while the tip of the catheter was withdrawn from the pulmonary artery, across the pulmonic valve, through the right ventricle, across the tricuspid valve, and into the right atrium. Repeated observations demonstrated a pressure change occurring near the lower portion of the right ventricular outflow tract from ventricular to atrial, although the simultaneous intracardiac electrocardiogram remained ventricular in form. In this position, the tip of the catheter was clearly far beyond the left side of the spine. When the tip of the catheter was slowly withdrawn across the spine, no significant change in the contour of the pressure pattern was observed although the intracardiac electrocardiogram suddenly changed to an atrial type.

The catheterization thus demonstrated a proximal ventricular chamber with ventricular potentials but with atrial pressures. In addition, the blood gas analysis demonstrated a predominantly left-to-right shunt at the atrial level. A co-existing right-to-left shunt was suggested by the degree of arterial oxygen desaturation demonstrated earlier. The pressure-potential tracings are depicted separately (fig. 3) and the location of the tip of the catheter is illustrated in figure 4.

Since first being seen, this patient has had over 30 visits to the hospital, usually in the emergency department, and generally for bouts of palpitation. The attacks were similar to those described with an occasional report of dusky nails and fainting. Signs of heart failure had not been observed. Many attacks had been documented electrocardiographically to be bouts of supraventricular tachycardia, usually stopping in a short period of time. On several occasions intravenous quinidine in 0.4-Gm. doses given slowly over several hours had been required. Various regimens for prophylaxis had been tried, quinidine appearing to be most effective. There seemed to be no reliable program because of lapses of taking the medications on the patient’s part.

Throughout the follow-up period physical examinations, chest films, and electrocardiograms revealed no progressive changes.

D I S C U S S I O N

The diagnosis of Ebstein’s disease in this case was suspected on the basis of documented bouts of paroxysmal supraventricular tachycardia, the configuration of the heart, and the pattern of the electrocardiogram, and it was confirmed by cardiac catheterization. Many authorities state that Ebstein’s disease as a clinical syndrome may be diagnosed or at least suspected with a high degree of accuracy during life.\(^1\)\(^,\)\(^2\)\(^,\)\(^3\)\(^,\)\(^4\)\(^,\)\(^5\)\(^,\)\(^6\) Their findings are summarized below.

Symptoms are remarkably mild in relation to the heart size. Cyanosis is usually mild or absent at rest, and becomes more evident with exercise or during bouts of tachycardia. When present, cyanosis usually occurs in later life. Exercise tolerance is unusually good, although the majority of patients complain of some degree of fatigability. The incidence of paroxysmal palpitations is strikingly frequent.

On examination clubbing of the fingers, polycythemia, and cyanosis at rest are uncommon. The heart is grossly enlarged. Despite the size, the heart is unusually quiet.
Fig. 3. These tracings illustrate the precordial electrocardiograms (top), intracardiac (intraeavity) electrocardiograms (middle), and pressure patterns (bottom) taken simultaneously at different sites during cardiac catheterization. The large deflections of the pressure tracings are artifacts. A. Right pulmonary artery. The intracardiac potential shows a deep inverted P wave, an rSr' complex and an inverted T wave. The pulmonary pressure is 33/11 with a mean of 15 mm. Hg. B. Right ventricular outflow tract. The intracardiac potential is characterized by a small upright P wave followed by a small R wave and a deep S wave. The T wave is deeply inverted. The right ventricular pressure is 33/5 mm. Hg. Note that the initial upstroke of the ventricular pressure begins after the R wave in both precordial and intracardiac electrocardiograms. C. Below the right ventricular outflow tract, adjacent to the left border of the heart. The right ventricular intracardiac potential continues to be similar to those of the right ventricular outflow tract (B) except for a deeper S wave and a less deeply inverted T wave. The pressure pattern in this area is distinctive in that the initial upstroke begins after the P wave but before the QRS complex. In this respect it resembles an atrial pressure pattern.

D. ‘‘Mid right ventricle.’’ The intracardiac potential continues to show right ventricular activity, resembling those recorded at position B. Despite this, the pressure is atrial in configuration. The magnitude of the pressure in this area and in location (C) is definitely lower than that in the right ventricular outflow tract (B). It is almost identical to that obtained in the right atrium proper (F). E. Drawing back across the spine, the usual location of the tricuspid valve, where the intracardiac potential changes from right ventricle to right atrium. This potential change is clearly shown by the sudden appearance of a large diphasic P wave followed by an rSr' complex. However, there is no change in the contour or magnitude of the pressure. F. High right atrium, along the right cardiac border. The intracardiac potential shows a large negative P wave and a tiny QRS complex. This atrial pressure is elevated and shows a distinct ‘‘a’’ wave.

Signs of right ventricular hypertrophy such as a parasternal heave or lift are absent. A moderately loud systolic murmur is present to the left of the sternum and may be transmitted over the entire precordium. A common finding is the presence of a triple rhythm near the apex. Frequently in the same area a diastolic murmur may be heard. Wood\textsuperscript{5}
has stated that a superficial diastolic scratch may be a pathognomonic sign.

Radiographic examination shows a large globular heart shadow resembling pericardial effusion, with a prominent right border, small pedicle, and pulmonary artery segments, and clear lung fields. Distinctive features in the electrocardiogram include tall peaked P waves and prolonged intraventricular conduction usually of a right bundle-branch block type. Occasionally the QRS complexes are bizarre. The P-R interval may be long. The frequent bouts of paroxysmal tachycardia are supraventricular in origin.

Many workers have stressed the point that surgery, particularly a shunting procedure, is contraindicated in this condition and there are several reports describing death following surgery. Caution has been urged in the catheterization of patients with Ebstein's disease because of the danger of arrhythmias. Campbell stated that cardiac catheterization should not be carried out in these patients unless there are special reasons for it. On the other hand, several workers have not encountered serious arrhythmias during catheterization in a large group of patients. However, it should be stressed that we cannot dismiss lightly the hazard of this procedure in patients with Ebstein's disease. If this procedure is performed, it requires constant monitoring of the electrocardiogram. The presence of supraventricular or ventricular tachycardia should be an indication for discontinuing the procedure.

As mentioned previously, the distinctive features on catheterization that have been described include a grossly dilated right atrium in which the catheter may coil, and elevated atrial pressure. Displacement of the tricuspid valve has been assumed by the demonstration of 2 areas of different pressures well to the left of the midline. In a given case, however, a variety of abnormal findings may be encountered that may be difficult to interpret. A pressure gradient across the pulmonic valve may suggest pulmonic stenosis. A gradient across the tricuspid valve suggesting tricuspid stenosis is also described. On the other hand, a pattern simulating tricuspid insufficiency may be present. In some cases pressure impulses from atrial contraction may be transmitted and superimposed on those from the right ventricle or pulmonary artery.

Although the clinical syndrome of Ebstein's disease may be apparent during life without cardiac catheterization, this procedure remains the most useful tool for obtaining a definitive diagnosis, especially in a confusing case where the question of surgical intervention may be raised. Hecht and Kossmann and his co-workers have shown that intracardiac potentials are distinctive and representative for various positions within the heart. With the electrode catheter in the present case, simultaneous pressures and potentials provided a graphic representation of the structural abnormalities. This is demonstrated by a distinct change in pressure from ventricular to atrial just below the right ventricular outflow tract. Atrial pressures persist in this zone despite simultaneous demonstration of intracardiac potentials of ventricular form. This point where pressures suddenly change is most likely that area where the displaced tricuspid valve divides the right ventricle proper into 2 chambers. Slow withdrawal of the catheter reveals a second point lower in the right heart, near the usual point.
sition of the tricuspid valve, where the intracardiac potential suddenly changes from ventricular form to that of atrial, but where there is no concomitant alteration in pressure. This is further evidence for the "atrialization" of the proximal right ventricle.

SUMMARY

A case is reported that possesses all the clinical features of Ebstein's disease. Cardiac catheterization, with the use of an electrode catheter to record simultaneous potentials and pressures, gave graphic representation of the anatomic changes. It is suggested that the diagnosis of Ebstein's disease can be made with a greater degree of certainty by use of the electrode catheter than by the usual means heretofore reported.

SUMMARIO IN INTERLINGUA

Es reportate un caso exhibente omne le aspectos clinic de morbo de Ebstein. Catheterisation cardiac, per medio de un catheter electrodic que permetteva le registration simultanea de potenciales e pressiones, resultava in un representation graphic del alterationes anatomic. Es postulate que le diagnosto de morbo de Ebstein potte esser establite con un plus alte grado de certitude per usar le catheter electrodic que per le medios communemente reportate in le passato.

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Circulation. 1958;17:543-548
doi: 10.1161/01.CIR.17.4.543

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:
http://circ.ahajournals.org/content/17/4/543

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