Endocardial Fibrosis

Detection by Cardiac Pacing

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SUMMARY

Three patients with endocardial fibrosis who had their endocardial pacing thresholds measured are described. The threshold values were elevated to at least twice normal levels. By determining the endocardial pacing threshold value, a technique is available for detection of endocardial fibrosis during diagnostic cardiac catheterization.

Additional Indexing Words: Pacing threshold Myocardiopathy

Endocardial fibrosis occurs in diverse diseases. An infantile form appears to reflect an unexplained congenital derangement in which a thick layer of collagen and elastin lines the left ventricular cavity.1 Sporadic instances noted in the adolescent have also been presumed to be of congenital origin.2 A poorly understood cause of heart failure occurs frequently in Uganda and South Africa in association with extensive endocardial fibrosis.3 Endocardial fibrosis also occurs as one of the pathological changes of subendocardial myocardial infarction, and, in the right ventricle, in the carcinoid syndrome. Localized areas of endocardial fibrosis have been noted in association with sustained hypertension and with valvular stenosis and regurgitation.4 Despite the frequent occurrence of this abnormality in many types of heart disease, proof of endocardial fibrosis has not been possible except by autopsy, or rarely, by biopsy during cardiac surgery. The present study concerns a method by which endocardial fibrosis can be detected by endocardial pacing during cardiac catheterization.

The excitatory properties of myocardium have been recognized at least since the eighteenth century. Hyman,5 in 1932, first successfully stimulated the arrested heart of experimental animals. Electrical stimulation of the heart in man was initiated in 1952 by Zoll.6 Furman and Robinson7 introduced the rapidly advancing field of cardiac pacing with the first intracavitary cardiac pacing in 1958. Cardiac pacing has subsequently been used for many physiological studies.8 The normal values for endocardial stimulation thresholds have been well established in studies by Nash9 to be approximately 1.3 microjoules and by Davies and Sowton10 to be approximately 0.7 v for a 2-msec impulse.

The presence of endocardial fibrosis would be expected to increase the electrical energy necessary to stimulate myocardium with an endocardial electrode. This paper presents a test of this hypothesis in three patients, two of whom had anatomic confirmation of endocardial fibrosis.

Methods

The endocardial pacing threshold was determined during diagnostic cardiac catheterization.
in the left ventricle of patient 66-350 and in the right and left ventricles of patients 68-94 and 68-143. This was performed by using a bipolar pacing catheter* connected to a battery-powered pacemaker. The pacemaker provided square wave impulses with a duration of 2 msec and a controlled variable voltage.

Right ventricular pacing was accomplished via a right antecubital vein; for left ventricular pacing, the catheter was advanced into the ventricle from a brachial artery. Physical contact between the distal platinum electrode and the endocardium was assumed to exist when ventricular premature beats could be mechanically produced by advancing the catheter. The pacemaker rate was set slightly faster than the existing sinus nodal rate. The stimulus voltage was gradually increased. The level at which constant ventricular pacing occurred was taken as the threshold.

Threshold values were determined in several areas of each ventricle.

Similar studies have been conducted in the right ventricles of 50 patients and in the left ventricles of four patients without apparent cause for endocardial fibrosis. In none of these was more than 1.0 v required to achieve stimulation.

Report of Cases

Case 66-350

This 23-year-old, white male factory worker had cardiomegaly detected on a routine chest x-ray examination at age 15 years. Dyspnea began 3 days before admission. Six other members of his family have cardiomegaly and electrocardiographic evidence of left ventricular hypertrophy.

His blood pressure was 110/70 mm Hg, and his pulse rate was 78 beats per minute. Bilateral basal rates were present on the initial examination. He had marked cardiomegaly with a forceful left ventricular impulse. The second heart sounds were paradoxically split. Third and

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Figure 1

Right ventricular endocardium and adjacent myocardium of patient 68-94, × 10. The thick fibrous endocardium is evident, as indicated by the arrow.
fourth heart sounds were present; no murmurs were detected.

Electrocardiograms showed left ventricular hypertrophy with intraventricular conduction aberration. Chest x-rays showed a massive left ventricle.

Cardiac catheterization revealed only slight pulmonary hypertension (38/16 mm Hg), marked elevation of left ventricular end-diastolic pressure (26 mm Hg), and a low cardiac output of 1.72 L/min/m².

Left ventricular pacing was conducted eventfully. The threshold was measured in three locations. The values were 1.9, 5.3, and 6.7 v, at the lateral wall, apex, and septal wall of the left ventricle, respectively.

The patient had progressive congestive heart failure and expired 8 months after his initial admission. Postmortem examination performed by Dr. Clifford W. Atherton revealed a heart weight of 1,015 g. "The left ventricle varied in thickness from 3 cm at the base to 1 cm with marked thinning and scarring in the apical lateral left ventricular wall. . . . In the anterior interventricular wall there are irregular broad zones of fibrous scarring, subendocardial in location. The endocardium shows patchy areas of subendocardial fibrosis." Coronary arteries were normal.

Case 68-94

This 40-year-old white male architect had a heart murmur detected at age 7 years, but felt completely well until progressively severe exertional dyspnea and palpitations began 8 years ago.

He was an obese man with a blood pressure of 128/72 mm Hg and a pulse rate of 88 beats per minute. The jugular venous pressure was estimated to be 12 cm of blood. The left heart border was at the anterior axillary line. The pulmonic closure sound was soft. A grade IV/V, harsh systolic ejection murmur was audible over the pulmonic area.

The electrocardiogram revealed right ventricular and right atrial hypertrophy. Chest films showed right ventricular enlargement and prominence of the proximal pulmonary arteries.

Cardiac catheterization revealed right ventricular pressures of 113/19 and 24/19 mm Hg in the inflow and outflow portions, respectively. No lesions other than infundibular stenosis were found. Both right and left ventricular endocardial pacing thresholds were determined. They were 0.5 v at the apex and 0.7 v in an area on the lateral wall of the left ventricle. Right ventricular endocardial pacing thresholds were 2.0 v along the midportion of the right ventricular wall posteriorly. The floor of the right ventricular inflow tract had a pacing threshold of 2.7 v.

The patient underwent open heart surgery for repair of his infundibular stenosis. The right ventricular endocardium was carefully inspected at surgery and was noted to be opaque, white, and approximately 2 mm in thickness. This tissue was biopsied. The thick layer of endocardial fibrosis is shown in the biopsy section (fig. 1).

Case 68-143

This 2-year-old asymptomatic girl was found to have cardiomegaly on a routine examination and was admitted to the hospital for study. Her two sisters had died of heart disease at 3 months and 6 years of age. Each had had unexplained cardiomegaly with marked endocardial fibroelastosis of both ventricles at postmortem examination.

The patient's blood pressure was 90/60 mm Hg, and her heart rate was 120 beats per minute. No evidence of congestive failure was present. Her left ventricular impulse was diffuse in the left anterior axillary line. Heart tones were normal except for loud third and fourth sounds. A grade III/IV apical pansystolic murmur was heard.

Left ventricular hypertrophy was present electrocardiographically and roentgenographically.

Only an elevated left ventricular end-diastolic pressure of 18 mm Hg and an enlarged, poorly contractile, left ventricular cavity were found during routine catheterization and angiocardiography. A presumptive diagnosis of endocardial fibroelastosis was made.

Left ventricular pacing was accomplished in seven areas. The threshold value in one apical location was 0.8 v; the values ranged from 1.2 to 3.0 v in all other areas tested. Right ventricular pacing in three areas resulted in threshold values of 0.9, 1.2, and 1.5 v.

Anatomic confirmation has not been obtained.

Discussion

These three cases represent different causes of endocardial fibrosis. Patient 66-350 is presumed to have a congenital abnormality of the myocardium with associated endocardial fibrosis. The fibrosis in patient 68-94 may either be an integral part of his right ventricular structural anomaly or be a consequence of sustained right ventricular hypertension. Patient 68-143 is believed to have infantile endocardial fibroelastosis; the exact cause is not known. Regardless of the cause, elevated pacing thresholds were present in all three patients, two of whom had pathological confirmation of endocardial fibrosis.
This technique should permit more precise identification of endocardial fibroelastosis in infants and may provide supporting evidence of subendocardial myocardial infarction in adults. The ability to recognize fibrosis may, in addition, permit clearer distinction between the restrictive effects of the endocardial lining and the hemodynamic effects of associated valvular or other lesions.

Acknowledgment

We are indebted to Dr. Clifford W. Atherton, Pathologist, Rayburn Hospital, Ottawa, Illinois, who performed the autopsy, for providing us with the results.

We are grateful to Miss Susan Connors and Mrs. Linda Campbell for valuable technical assistance.

References


100 Years Ago

Concerning the Temperature in Acute Rheumatism

Abnormally mild cases are particularly common, or rather cases in which the temperature is either very slightly or, perhaps, not at all affected; although the local condition is not always correspondingly insignificant. Indeed, we cannot always tell why the fever should remain so trifling, or perhaps be altogether absent, when the joint affection is very severe; and cardiac complications are by no means excluded by the absence of fever. Cases with slight fever (not above 38.5° C.) (101.3° F.), or with only sub-febrile temperatures, constitute about one third of all the cases of acute rheumatism. All other deviations from the course described, comprehending more or less severe cases altogether, do not, at least in our country, amount to more than one sixth of the cases.—C. A. Wunderlich: On the Temperature in Diseases: A Manual of Medical Thermometry. London, The New Sydenham Society, 1871, p. 398.
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_Circulation_. 1968;38:1136-1139
doi: 10.1161/01.CIR.38.6.1136
_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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