Congenital Aneurysm (Diverticulum) of the Right Atrium

Clinical Manifestations and Results of Operative Treatment

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SUMMARY
A patient is described in whom a large congenital aneurysm or diverticulum of the right atrium caused repeated attacks of supraventricular arrhythmia and a strikingly reduced cardiac output. The aneurysm, which contained a large thrombus, was demonstrated by preoperative angiographic examinations. At the time of operation, regular rhythm returned at the moment the aneurysm was transected. The patient is asymptomatic after operation, and has maintained sinus rhythm. A normal cardiac index was recorded at postoperative cardiac catheterization.

A young woman was recently referred to the National Heart Institute because of severe symptoms principally attributable to disorders of the cardiac rhythm. Clinical, hemodynamic, and angiographic examinations revealed a large aneurysm or diverticulum which originated from the right atrium. The manifestations of this unusual congenital cardiac malformation, the operative methods employed in its correction, and the results of postoperative evaluations are described in the report that follows.

Clinical Summary
N. H., a 23-year-old housewife, had been found to have an abnormal cardiac silhouette by x-ray when she was 9 years of age. Two siblings had died at birth with congenital heart disease of unknown type, but her parents and nine other siblings are well. At age 16 she first became symptomatic, and chest pain, fatigue, palpitations, and dyspnea necessitated hospitalization on many subsequent occasions. In 1959, right heart catheterization at another hospital revealed normal intracardiac pressures and no circulatory shunt. The positions of the catheter indicated striking enlargement of the right atrium, and the diagnosis of Ebstein's anomaly of the tricuspid valve was made. The patient's symptoms progressed in severity, and she had more frequent episodes of tachycardia, often accompanied by cyanosis, syncope, dyspnea, and precordial pain. Therapeutic doses of digitalis glycosides did not reduce the frequency of arrhythmias, but quinidine was sometimes helpful.

On examination the patient did not appear ill, and her development was normal. The heart was enlarged to percussion, but no precordial impulses could be felt. The heart sounds were extremely faint (inaudible to some examiners), and no murmurs were heard. The electrocardiogram (fig. 1) revealed normal sinus rhythm with large P waves and extremely low ventricular voltages. Roentgenograms of the chest (fig. 1) demonstrated marked enlargement of the cardiac silhouette, and in the lateral projection there was an unusual opacity of the retrosternal area.

Right heart catheterization revealed normal pressures in the right ventricle and pulmonary artery, and no intracardiac shunt. An intracardiac electrocardiogram recorded no right ventricular complexes proximal to the tricuspid valve, ruling out Ebstein's anomaly. The cardiac catheter could be passed anteriorly from the right atrium to the left border of the heart, where it appeared to lie against the left chest wall; in this position the catheter registered a normal atrial pressure, and the oxygen content of blood sampled from this site was identical to that of blood from the right atrium proper. Arterial indicator-dilution...
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The right atrial aneurysm constitutes the entire cardiac silhouette and the ECG is principally characterized by extremely low voltages in all leads.

Curves recorded after right atrial injection had marked prolongation of both ascending and descending limbs, but those following right ventricular and pulmonary arterial injection had normal contours. The cardiac index at rest was abnormally low, 1.89 L/min/m². A selective angiogram, exposed after right atrial injection, revealed a large chamber, which made up the entire cardiac silhouette, and in its left border a large filling defect was seen (fig. 2). In later films the left atrium and left ventricle appeared to be normal.

The patient was receiving therapeutic doses of digoxin and quinidine, but throughout the preoperative period, and particularly during the cardiac catheterization, she had repeated attacks of paroxysmal atrial tachycardia, atrial fibrillation, and atrial flutter. Each arrhythmia, however, reverted spontaneously to normal sinus rhythm after a variable period.

At operation, the heart was exposed through a bilateral anterior thoracotomy (fig. 3). After the pericardium was opened, the entire anterior surface of the heart was seen to be covered by a thin-walled, almost transparent sac, which was in continuity with the right atrium. No atrial appendage as such was apparent. The atrial wall on the right side was of normal consistency, and vena caval cannulae were introduced through it. With the institution of cardiopulmonary bypass the aneurysm emptied, and a firm mass could be felt in its apex (fig. 4). The aneurysm was
At operation the heart and great arteries were covered by the thin-walled aneurysm, and it was obvious that the insulating effect of the envelope of blood had rendered the heart sounds inaudible and had prevented the palpation of any precordial impulse. The body of the right atrium appeared normal in consistency, and caval cannulae were introduced through purse-string sutures in it.

The atrial incision was closed as shown, reconstituting an atrial chamber of approximately normal size and configuration.

Diagrammatic representation of the relations of the right atrial aneurysm to the cardiac chambers and great vessels. The right ventricle was compressed and indented by the aneurysm and the thrombus within it.
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6. The mass within the aneurysm was 5 cm in diameter. It consisted of organized thrombus. Histologic examination of the wall of the aneurysm revealed that it was composed of fibrous tissue and attenuated myocardial fibers with an endothelial lining.

The variations in cardiac rhythm recorded during the operative procedure were of unusual interest. As the pericardium was opened, atrial flutter with a varying block occurred, and when the aneurysm was manipulated, atrial fibrillation appeared. Thereafter, atrial fibrillation continued as the basic rhythm. The electrocardiogram recorded immediately before and after the aneurysm was amputated is reproduced in figure 7. A regular atrial rhythm appeared at the moment the sac was detached from the heart, and normal sinus rhythm reappeared within minutes.

Postoperatively the patient's course was uneventful, and no arrhythmia was ever noted. She returned to the Institute 6 months after operation for evaluation. She was asymptomatic and was engaging in all household and social activities without restriction. She had stopped taking digoxin and quinidine 1 week after returning home and had not noted palpitations. Examination revealed no cardiac abnormalities, and roentgenograms showed a striking reduction in the size of the heart. The electrocardiogram demonstrated sinus rhythm, normal voltages, and an anterior axis.

Cardiac catheterization revealed normal pressures in the pulmonary artery, right ventricle, and right atrium. The cardiac index at rest had risen to a normal level, 3.28 L/min/m². Indicator-dilution curves recorded after right atrial

and pulmonary arterial injections had similar normal contours. A selective angiocardiogram was carried out, and the appearance of the right atrium and ventricle is shown in figure 2.

Comment

It would appear that the atrial aneurysm in the patient described originated as an isolated congenital abnormality of the right atrial myocardium. The patient's cardiac silhouette had been enlarged since her childhood, and no intracardiac malformation which could have caused secondary atrial enlargement was evident either in the preoperative studies or at operation. Also, the aneurysm seemed to arise from the atrial appendage, since no appendage as such was present, and the myocardium of the body of the right atrium was normal.

True aneurysm of the right atrium has apparently not been previously described. Bailey excised a "diverticulum" of the right atrium in 1953, but the descriptions of the malformation in this patient indicate only symmetrical dilatation of an otherwise normal right atrium: the appendage was present, and the muscle making up the wall of the diverticulum had a normal structure. Pitts and Potts described the excision of a large saccular enlargement of the left atrium in 1954, and subsequently, there have been reports of 10 other left atrial aneurysms. In five of these patients, however, the lesion was actually an enlarged left atrial appendage that had herniated through a congenital pericardial defect, rather than a true aneurysm of the atrium.

The operative and pathological findings in the cases of true aneurysm of the left atrium were generally similar to those in the present patient. Thus, the aneurysms have been thin and translucent, with swirling blood visible within them, and the walls composed of fibrous tissue and attenuated muscle fibers. Most contained organized thrombus.

In all patients, including the present one, symptoms were primarily related to episodes of supraventricular tachycardia, and these were eliminated by excision of the aneurysm. It seems clear that the enormous mass of
atrial tissue was responsible for these arrhythmias, either by containing irritable ectopic foci or by providing extensive surfaces over which circus movements could occur. The immediate restoration of regular rhythm with transection of the aneurysm was clearly documented in the patient described (fig. 7).

The exercise intolerance and fatigue experienced by the present patient preoperatively are probably attributable to the reduced cardiac output demonstrated at preoperative catheterization. At operation the right ventricle was found to be severely compressed by the mass of the aneurysm, and abnormal right ventricular compliance was apparently the mechanism responsible for the low cardiac output, a physiological sequel similar to that seen in more common restrictive and constrictive diseases of the myocardium and pericardium.

References
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