Congenital Aneurysmal Defect of the Membranous Portion of the Ventricular Septum Associated with Heart Block, Ventricular Flutter, Adams-Stokes Syndrome and Death

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A case of congenital aneurysmal defect of the ventricular septum is reported which appears to be unique in that this lesion was the only significant finding at autopsy. The patient had evidence of auriculoventricular block for a period of 26 years, at first partial and then complete. She succumbed at the age of 47 to a series of Adams-Stokes attacks which were demonstrated to be set off by paroxysmal ventricular flutter followed by ventricular tachycardia and ventricular standstill. Pertinent literature is briefly reviewed.

The following case is considered worthy of recording since it appears to be the only one which we can find on record where an aneurysmal defect of the ventricular septum was the sole significant anatomic lesion found at autopsy. It demonstrates that ventricular flutter, unsuspected without cardiographic evidence, may precipitate the Adams-Stokes type of syncope.

Previous Literature

Aneurysm of the Ventricular Septum. Because of its presumed lack of clinical significance, little has been written on the subject of aneurysm of the ventricular septum. Abbott, in Osler’s Modern Medicine,1 described the pathologic condition briefly and reviewed the original studies of Mall.2 In 1938 Lev and Saphir3 published two cases and reviewed 70 cases which had been reported in the literature. Since this review we find only one further case reported, that of Castoldi4 in 1942.

Adams-Stokes Syndrome. The Morgagni-Adams-Stokes syndrome as originally described referred to the association of syncope, epileptiform convulsions and marked slowing of the heart action. Parkinson and co-workers5 in 1941 reviewed all reported cases of the Adams-Stokes syndrome with electrocardiographic tracings and found that only 55 per cent of reported attacks were associated with ventricular arrhythmias. Parkinson defined the Adams-Stokes disease as the “name applicable to patients with heart block who suffer from recurrent attacks of loss of consciousness due to ventricular standstill, ventricular tachycardia, ventricular fibrillation or a combination of these.” Schnur,6 in 1948 reiterated these views and presented a case of ventricular fibrillation, tachycardia and asystole imposed upon complete heart block. Pastor and Worrilow7 have recently reviewed the electrocardiographic patterns in the Adams-Stokes syndrome and found 20 cases on record in which both ventricular arrhythmias and ventricular standstill had been shown to occur during the Adams-Stokes syncope; in eight of these cases the arrhythmias were in association with complete A-V block.

Congenital Heart Block. Heart block of congenital origin as reported in the literature has been reviewed by Yater and associates,8 with a study of 44 acceptable cases of which a ventricular septal defect was found to be present in 26. Faessler9 reported eight cases of the Adams-Stokes syndrome associated with congenital heart disease, of which six were diagnosed as having ventricular septal defects. Essentially all types of congenital defects have been reported as associated with heart block.10

Ventricular Flutter versus Fibrillation. In 1925

From the Cardiac Department, Winchester Hospital and the Cardiac Clinics and Laboratory of the Massachusetts General Hospital, Boston.
Sir Thomas Lewis stated that the nature of the disturbances included under the term ventricular fibrillation was undefined. He believed that varying grades of ventricular arrhythmias occurred, as in the auricles, ranging from flutter, to impure flutter and grossly impure flutter or fibrillation. Fastier and Smirk described the condition of ventricular flutter, observed in experimental cats and dogs, as showing characteristic regular undulatory waves on the electrocardiogram which are quite in contrast with the disorganized movement, usually of smaller amplitude, seen in ventricular fibrillation.

CASE HISTORY

S. B., aged 47. This patient was said to have been in normal health, except for diphtheria in childhood, until 1921, when, at the age of 21, she was delivered of her first and only child. A review of her hospital record indicated that following a normal labor and delivery she had had five fainting spells followed by a convulsion, associated with irregularity of the pulse which at times was not palpable. By the patient's own statement, her pulse subsequent to this was usually slow, about 40 per minute, but she was generally well and led a normal life. In 1936 a careful examination showed a pulse rate of 44, a blood pressure of 136/80, with heart sounds of good quality. An electrocardiogram showed first degree A-V block with a ventricular rate of 42 and a P-R interval of 0.24 of a second; the tracing was otherwise not remarkable.

In March 1947, at the age of 47, the patient was seen in consultation by one of us (R. J. C.). For the previous four or five years she had complained of "dizzy sensations," distinct increase in dyspnea and brief bouts of low substernal pressure with radiation to the left arm brought on by excitement or exercise. Examination showed a pulse rate of 48, regular and full. The blood pressure was 165/100. The heart showed a left border of dullness 2 cm. outside the midclavicular line. The heart sounds were forceful in character with an accentuated pulmonary second sound which was greater than the aortic second sound. There was a grade 2 systolic murmur, without a thrill, heard best between the apex and the left sternal border. The lungs were clear and there was no evidence of congestive failure. Fluorescopic examination showed a slight increase in the transverse diameter of the heart with the appearance of slight left ventricular hypertrophy. An electrocardiogram showed a regular ventricular rate at 42 with the P-R interval increased to 0.26 of a second, but was otherwise not remarkable. A urinalysis was normal. A Hinton test was negative. Photoelectric determination of hemoglobin was 13.5 Gm. The presumptive diagnosis at that time was congenital heart disease with a ventricular septal defect and associated heart block with superimposed coronary artery disease.

On the morning of June 15, 1947, the patient was admitted as an emergency case to the Winchester Hospital because of an attack of generalized convulsions and unconsciousness of 10 minutes duration. Directly after admission a recurrent attack was observed. The pulse could not be obtained and no heart sounds were heard. One-half cc. of 1:1000 adrenaline solution was given intramuscularly. Another attack occurred after a half hour, during which an electrocardiogram was obtained. This showed ventricular flutter at a rate of 220 in the portion taken during the acute attack. This was followed by complete A-V block with an auricular rate of 130 and a ventricular rate of 42, also variable ectopic ventricular beats. The basic pattern had not otherwise changed significantly from that found in March.

When it became apparent that the true situation was one of Adams-Stokes episodes associated with ventricular flutter, adrenaline was omitted and the patient was started on quinidine sulfate by mouth and atropine sulfate, 0.6 mg. by hypodermic injection. During the afternoon she continued to have convulsions at approximately hourly intervals; frequently clusters of two or three attacks covered a period of 15 to 20 minutes. On several occasions, between attacks, the patient complained of severe substernal oppression with radiation to the left arm wherefore morphine sulfate 10.6 mg. by hypodermic was given. She was maintained in an oxygen tent. The patient was seen in consultation by one of us (P.D.W.) that afternoon. Quinidine sulfate was stepped up in dosage from 0.2 Gm. orally to 0.4 Gm. of lactate every two hours intramuscularly because of vomiting. During the night the patient's attacks decreased slightly in frequency but the following day at 12:30 P.M. there was a more severe seizure in which she expired. During her period of hospitalization the patient had some 30 convulsive seizures. In the course of 24 hours she received 3.5 Gm. of quinidine.

Numerous electrocardiographic observations were made which showed periods of ventricular flutter lasting up to three minutes, frequently followed by periods of ventricular tachycardia lasting two or three seconds, followed by periods of ventricular standstill with P waves decreasing in frequency. The longest period of measured standstill was 75 seconds. Following the ventricular standstill the patient resumed complete heart block with the ventricular rate speeded to about 60 at first then dropping into the 40's. Figure 1 shows a recording of one of the more severe episodes.
The clinical diagnosis prior to autopsy was acute myocardial infarction imposed upon coronary artery disease, with possible congenital heart block.

Pathologic Examination

An autopsy was performed by Doctors Donald McKay and Donald A. Nickerson. Significant findings were limited to the heart, which weighed 350 Gm. The pericardium was smooth and glistening. The epicardium was smooth. The coronary arteries were thin walled and patent throughout. The right auricle appeared dilated. The myocardium of the left ventricle was firm and red brown, showing no evidence of fibrosis or infarction; it measured 1.5 cm. in thickness. The right ventricular myocardium was slightly softer but otherwise not remarkable; the wall of the right ventricle measured 0.4 cm. in thickness. The membranous portion of the interventricular septum was paper thin and herniated into the right ventricle, forming a pouch with its mouth on the left ventricular side (fig. 2). The mouth of the pouch measured 1.8 cm. in diameter. The membrane was fused with the medial cusp of the tricuspid valve. The pouch, when ballooned out with the finger, almost completely filled the orifice of the tricuspid valve. The remainder of the valves showed thin delicate leaflets with no evidence of inflammation. The chordae tendineae were normal. Apart from the interventricular aneurysmal defect there were no congenital malformations of the heart or great vessels. The aorta was thin walled, elastic and lined by a smooth velvety intima. Microscopic sections taken from the right and left ventricular musculature showed no recognizable abnormality of

Fig. 1. Continuous electrocardiographic tracing, lead II, taken during a severe convulsive episode. A. Onset with ventricular “flutter” at rate of 220. B. Widening of the beam caused by severe convulsing and necessity of holding the patient. C. Shift to ventricular tachycardia at slower rate again followed by a brief reversion to “flutter.” D. Appearance of ventricular standstill with only P waves in evidence, with progressive slowing of auricular activity. E. Resumption of ventricular activity (escape) with sinus bradycardia followed by high grade auriculoventricular dissociation. Note the disappearance of wide beam shadow as convulsive movements cease. Time intervals equal 0.04 second.
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muscle fibers or blood vessels. There was a moderate degree of passive congestion in the lungs, liver, kidneys and spleen. No specific cause of death was found anatomically.

Subsequently more detailed study of the heart was carried out in the Pathological Laboratory of the Massachusetts General Hospital by Doctors David Freiman and Robert Scully. Block sections were made of the entire aneurysm. These sections failed to show evidence of anything which looked convincingly like the Bundle of His. The pouch was composed of relatively acellular collagenous tissue. No evidence was found of mural endocarditis.

DISCUSSION

The diagnosis of aneurysmal defect of the ventricular septum cannot be made clinically. In this case historical evidence pointed strongly to the existence of heart block since at least the age of 21 years probably with Adams-Stokes syncope occurring following childbirth. A respite of 26 years without further occurrence is remarkable. The "dizzy spells" which this woman had experienced for about four years were doubtless a result of her heart block. The history pointed strongly toward coronary insufficiency with mild angina pectoris in a woman with early hypertension, yet no coronary disease was found at autopsy. One might speculate as to whether her heart block could have produced a relative coronary insufficiency giving rise to dyspnea and substernal distress. She had diphtheria in childhood. It has been demonstrated that residual heart block from diphtheria occurs in only very rare individuals.13 Barring such an assumption, the most likely cause of block at this age would be a congenital lesion, either of the septum or of the conduction fibers themselves. The heart murmur in this patient was not typically that of a ventricular septal defect either in location or intensity. No other congenital lesion was suggested. A tentative diagnosis of a ventricular septal defect with an atypical murmur was made, but such is doubtless an improbable assumption because of the absolute rarity of this condition.

In previously quoted cases6-7 with a similar sequence of arrhythmias, quinidine was quite ineffectual in therapy, as it proved to be in this instance. The use of atropine in similar cases has been suggested to be of value.14,15 Lately procaine intravenously has been advocated in ventricular tachycardia and fibrillation, both for prophylaxis and for emergency use on the operating table.16 Should another case similar to the one here reported be encountered, a trial of Pronestyl (procaine amide hydrochloride) would seem worth while.17

SUMMARY

A case history is presented of a woman with heart block of at least 26 years duration, who died at the age of 47 with Adams-Stokes attacks precipitated by paroxysms of ventricular flutter followed by ventricular tachycardia, ventricular standstill and complete auriculo-ventricular dissociation. At postmortem exami-
nation the only significant abnormality was the presence of a large aneurysmal defect of the ventricular septum.

ACKNOWLEDGMENT

We are indebted to Dr. Ernest MacDougall of Wilmington, Massachusetts, who referred this patient to us.

REFERENCES

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Circulation. 1952;5:725-729
doi: 10.1161/01.CIR.5.5.725

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/5/5/725

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