THE DIAGNOSIS AND TREATMENT OF AORTIC STENOSIS COMPLICATED BY HEART BLOCK

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The association between calcific aortic and mitral heart disease and atrio-ventricular block is well recognized especially since Yater and Cornell (1935) demonstrated the spread of calcification from the valve into the nearby conduction tissue. Recently Sellers *et al.* (1963) have pointed out the risk of damage to the same area during insertion of a prosthetic aortic valve. On the other hand Mahaim (1963) has reported relief of atrio-ventricular block following surgical correction of aortic stenosis.

In atrio-ventricular dissociation, the large stroke volume associated with a slow heart rate makes unusual demands on a normal left ventricle, which are increased by even mild obstruction to left ventricular output. This report concerns patients with heart block and aortic stenosis to show the special difficulties in the diagnosis and management of this lesion, and the effects of varying pace-maker rates in the presence of aortic valve disease.

These cases form part of a series of 100 patients with heart block treated with artificial pace-makers (Harris *et al.*, 1965).

CASE REPORTS

Patient 1. W.C., a married man aged 58 years, was first admitted to hospital in November 1961 after repeated Stokes-Adams attacks over the preceding 10 days. He gave a history of rheumatic fever at the age of 8; a murmur was first heard at the age of 36. He was free of symptoms until he was 56 years old, when he developed slight exertional dyspnoea without angina. At the time of admission he was in complete heart block with a rate of 28 a minute and blood pressure of 100/60 mm. Hg, and was unaware of his slow rate. The carotid pulses were slow rising, the apex beat was displaced to the left, and the cardiac impulse was sustained. On auscultation there was a moderately loud aortic systolic murmur (grade 3/6) and a soft early aortic diastolic murmur (grade 2/4). The second heart sound was single. The jugular venous pressure was not raised, but frequent cannon waves were observed. The electrocardiogram showed varying patterns: complete block with right bundle-branch block pattern idio-ventricular rhythm, 2:1 block with right bundle-branch block, and sinus rhythm with right bundle-branch block.

In view of recurrent Stokes-Adams attacks he was paced for 3 days by an endocardial electrode catheter (C.50 No. 5 U.S. Catheter and Instrument Co.) inserted via the right external jugular vein. The severity of the aortic valve disease was assessed by right and left heart catheterization and angiocardiology. There was a systolic gradient of 75 mm. Hg across the aortic valve with a cardiac output of 3.4 l./min. Biplane Schönander angiocardiology showed fixed calcified aortic valve cusps and a small regurgitant jet. It was decided that when his clinical condition had improved aortic valvotomy should be carried out and at the same operation an abdominal-epicardial system of artificial pacing inserted. He remained free from Stokes-Adams attacks and the electrode catheter was removed in the fourth week following its insertion. This was

followed by the onset of a persistent pyrexia and blood cultures grew a penicillin-sensitive *Staphylococcus pyogenes*. The septicæmia was controlled by 40 million units of penicillin intravenously, daily for 9 days. During this period he developed a left homonymous hemianopia which was considered to be due to a cerebral embolus. Three weeks later he developed severe central chest pain which persisted for several hours. The associated electrocardiographic abnormalities were difficult to interpret due to the presence of complete right bundle-branch block, but there was S-T elevation in V1 and a few days later a Q wave appeared in V3 and the electrical axis swung to the left. He developed congestive cardiac failure and died in a comatose state 6 days later.

Necropsy. The heart weighed 720 g. with hypertrophy mainly of the left ventricular wall to an average thickness of 2.4 cm. There was severe stenosis of the aortic valve from thickening and immobility of a bicuspid aortic valve with extensive calcification; a few small platelet thrombi were present on the valve cusps, but no evidence of active bacterial endocarditis. There was also an aneurysm of the anterior sinus of Valsalva. A recent extensive infarct measuring $10 \times 8 \times 1$ cm. involved the anterior two-thirds of the inter-ventricular septum and the anterior wall of the left ventricle. Radiographs of the heart and coronary injection studies revealed a dominant right coronary artery with a calcified embolus in its posterior descending branch; this branch had supplied an area of the myocardium in which an old scar was present. There was confluent atheroma in the proximal 3 cm. of the anterior descending branch of the left coronary artery with the lumen reduced to one-quarter of normal at its origin. There was no thrombus or embolus at this site. The circumflex branch of the left coronary artery had confluent atheroma in the proximal 1 cm., with no significant narrowing. Old infarcts were present in the spleen, kidneys, and brain.

Patient 2. K.O., a married woman aged 65 years, was first seen in July 1958 because of recurrent Stokes-Adams attacks over the preceding 5 years. The frequency of attacks had increased to 50 to 60 a day shortly before admission. Ephedrine, prednisone, and atropine therapy had been ineffective in controlling or decreasing the frequency of the Stokes-Adams attacks. At the time of admission she was in complete heart block, with a heart rate of 38 a minute, and despite this slow rate, a blood pressure of only 130/60 mm. Hg. The jugular venous pressure was not raised, but 'a' waves at a rate of 114 a minute were present. The apex beat was impalpable, which may have been related to presence of emphysema. She was examined by several competent observers, but no abnormality was observed in the form of the carotid arterial pulse. A systolic ejection murmur (grade 3/6), maximal in the aortic area, was conducted into the neck and to the mitral area. The second heart sound was soft. The electrocardiogram showed, at various times, sinus rhythm with complete right bundle-branch block, complete heart block and right bundle-branch block with continuing atrial activity at a rate of 112 a minute, and 2:1 A-V block with right bundle-branch block which masked the left ventricular hypertrophy subsequently found at necropsy.

The patient was found to have a large number of symptomless gall-stones, and cholecystectomy was carried out in the hope that it might relieve the frequency of her Stokes-Adams attacks; an endocardial electrode catheter was in position in the right ventricle during the operation for emergency pacing. The attacks ceased for 3 weeks and then recurred with the same severity as before. Subsequently, the frequency of the attacks declined and she was seen at regular intervals as an out-patient over the next 2 years during which time she averaged 1 Stokes-Adams attack a month.

The attacks then increased again and in January 1961 a pacemaker was inserted subcutaneously in the epigastrium and electrodes were attached to the surface of the heart. Unfortunately 2 days after operation she had a prolonged period of asystole due to detachment of the epicardial electrodes which were replaced at a second operation. Although satisfactory pacing was re-established, the patient remained comatose and died 24 hours later.

Necropsy. The heart weighed 360 g. and showed some hypertrophy of the left ventricle. The heart muscle and coronary vessels were normal. There was fairly severe aortic stenosis due to fusion of the right and left coronary cusps along the whole length of the commissure between them. The valve cusps were thickened, calcified, and very rigid. The mitral, pulmonary, and tricuspid valves were normal. The lungs showed evidence of widespread diffuse emphysema. There was extensive thrombosis of the pelvic veins and several emboli were present in the medium-sized pulmonary arteries.

Patient 3. M.G., a married man, aged 41, was first seen in November 1963 when he complained of nocturnal dyspnoea for 3 weeks. For 20 years, he had noted moderate limitation of exercise by dyspnoea, and shortly before admission he had developed congestive cardiac failure and severe limitation of exercise tolerance. At the time of admission to hospital he was in complete heart block with a heart rate of 30 to 40 a minute and a blood pressure of 105/55 mm. Hg. The carotid pulses were slow rising; the left ventricle felt enlarged and hypertrophied. The jugular venous pressure was raised 3 cm. with 'a' waves at a rate of 115 a

TABLE
RESULTS OF RIGHT AND LEFT HEART CATHETERIZATION IN PATIENTS W.C. AND M.G.

Patient	Heart rate/min.	Pressure (mm. Hg)					Cardiac output (l./min.) (Fick)	Systolic ejection time (sec.)	Aortic valve area (cm. ²)	Percentage increase in cardiac output	Stroke volume
		Pulmonary artery	Mean pulmonary capillary	Left ventricular cavity	Central aorta	Systolic gradient across aortic valve					
W.C.	94	10/4 (6)	3	150/0	75/50	75	3.4	0.36	0.35	—	36
M.G.											
Idio-ventricular rhythm	37		22	215/25	120/50	95	—	0.48	—	—	—
	41	72/25 (40)	20	175/25	105/55	70	4.4	0.4	0.5	—	107
Paced											
	42	—	25	190/20	100/45	90	4.4	0.4	0.41	—	104
	88	—	22	165/15-20	95/70	60	6.2	0.38	0.63	40	70
	107	—	20	145/12	95/70	50	5.0	0.28	0.5	27	52

minute, with occasional cannon waves. On auscultation there was an aortic systolic murmur grade 5/6 conducted into the neck and down to the mitral area. There was a soft early diastolic murmur grade 1-2/4, maximal at the left sternal edge. The second heart sound was single.

Electrocardiograms confirmed the left ventricular hypertrophy and showed complete heart block with an atrial rate of 110 a minute, a ventricular rate of 30 a minute, and coupled beats due to ventricular ectopics. Sympathomimetic amines had proved ineffective in increasing the heart rate before admission.

As a preliminary to right and left heart catheterization a C.50 No. 5 electrode catheter was passed from a left antecubital vein, and the tip was wedged in the apex of the right ventricle. As a precaution the patient was given penicillin throughout the period of intravenous electrode pacing.

During the early phase of left heart catheterization a brief episode of ventricular fibrillation occurred which reverted with a short period of external cardiac massage.

Three to five minutes at each pacing rate were allowed before intravascular pressures and the cardiac output were measured (Table). Valve calcification was not seen on screening with an image intensifier. The operative findings were calcific aortic stenosis with fusion of the posterior cusp with the right and left coronary cusps. Aortic valvotomy with débridement of the cusps was carried out (C.D.) under profound hypothermia with circulatory arrest on January 21, 1964. Following a period of rewarming, an abdominal pacemaker with epicardial electrodes was inserted. His post-operative course was entirely uneventful. He has been seen as an out-patient at 3-monthly intervals and he is asymptomatic and leading a normal life. He still has some physical signs of aortic stenosis with slight incompetence, but these are less than those found before operation. The pacemaker rate is fixed at 74 a minute.

DISCUSSION

Diagnosis. The difficulty in diagnosis is illustrated by the second case (K.O.) where undiagnosed aortic stenosis was found at necropsy. A systolic murmur is commonly heard in uncomplicated heart block, due to the ejection of a large stroke volume through a normal aortic valve. Furthermore, the duration of aortic ejection is increased in proportion to this increase in stroke volume and the pulse feels sustained. The third patient (M.G.) doubled his stroke volume when his heart rate fell from 107 to 41 a minute. The ejection time lengthened from 0.28 to 0.4 sec. It is, therefore, necessary to be aware of the possibility of aortic valve disease and not to discount the systolic murmur as arising from increased stroke volume alone. The systemic pressure in all 3 patients was relatively low at slow idio-ventricular rates. This is an unusual feature of heart block alone when the greatly increased stroke volume is associated with a big pulse pressure (Harris *et al.*, 1965). With further

FIG. 1.—Case 3 (M.G.). Simultaneous electrocardiogram (lead II) and the left ventricular (LV) pressure tracing during pacing. With a P-R interval of 0.2 sec. augmentation of left ventricular pressure occurs. Paper speed 50 mm./sec.; time marker 0.04 sec.

FIG. 2.—Case 3 (M.G.). Simultaneous electrocardiogram (lead II), left ventricular (LV) pressure tracing, and phonocardiogram recorded at the left sternal edge during pacing (S). The ejection systolic murmur increases in intensity when atrial contraction augments left ventricular filling. Paper speed 100 mm./sec.; time marker 0.04 sec.

experience it may be possible to decide, by external carotid pulse recording (Robinson, 1963), that the ejection time is longer in patients with heart block and aortic stenosis than in heart block alone.

The presence of left ventricular failure at an unexpected stage of the disease may alert one to the diagnosis of aortic valve disease in addition to heart block. The presence of emphysema and right bundle-branch block are factors that may obscure the diagnosis.

Bacterial Endocarditis. Although mild aortic valve disease may have little hæmodynamic significance, and though it can be argued that any obstruction that has not caused easily-recognized signs or symptoms with a slow heart rate is very unlikely to do so after treatment is directed at an increase in rate, the diagnosis is not solely academic if treatment by an artificial pacemaker is contemplated. The first patient developed bacterial endocarditis and died of it before it was realized that it was unsafe to use the intravenous electrode catheter, with an external pacemaker, in the presence of valve lesions, except as a short-term measure with antibiotic prophylaxis. The pacemaking system we favour at present uses a unit implanted in the axilla connected to an electrode catheter passed to the right ventricle by way of an external jugular vein (Harris *et al.*, 1965). Bacterial endocarditis has occurred in 4 patients out of 53 paced with an electrode catheter. However, the electrode catheter had been exteriorized in the neck and attached to an external pacemaker. Every patient develops a local area of sepsis around the electrode catheter in the neck, and this was almost certainly the source of the bacterial endocarditis. Of these 4 patients, 3 had valve lesions and 1 had an extensive myocardial infarction. It now seems probable that endocarditis does not

occur if the endocardial electrode and the attached pacemaker is buried in the axilla. When intravenous electrode pacing is contraindicated, the pacemaker is inserted in the anterior abdominal wall and connected to epicardial electrodes; there is no risk of endocarditis with this system and it is the method of choice when cardiac surgery is to be carried out.

Hæmodynamic Data. There have been reports of catheter and cardiac output studies on patients with uncomplicated heart block at various heart rates from several centres (Hudson, 1962; Piemme *et al.*, 1963; Sowton, 1964; Segel *et al.*, 1964). It was usually found that each subject had an optimal heart rate, after which the cardiac output did not continue to rise, and might fall. This optimum was found to lie between 84 and 100 a minute by Piemme *et al.* (1963) and 60 to 80 by Sowton (1964).

It is of interest that our third patient M.G. was able to increase his cardiac output by 40 per cent as a result of pacing at a rate of 88 a minute. At a rate of 107 a minute the output was lower, but this may not be significant. The left ventricular end-diastolic pressure fell strikingly as a result of pacing. The calculation of the aortic valve area by the Gorlin formula (Gorlin and Gorlin, 1951) was remarkably constant at different heart rates and corresponded very well to that found at operation confirming the accuracy of the method. The heart size, as judged by standard chest radiography, did not change during an acute study with varying heart rate, but after 4 months had decreased slightly.

The value of atrial contraction in augmenting ventricular systole has been pointed out by several authors (Gesell, 1916; Wiggers and Katz, 1922; Braunwald and Frahm, 1961; Judge, Wilson, and Siegel, 1964), and its final value has been questioned by others (Burchell, 1964), but it is well illustrated in Fig. 1, where a well-timed atrial contraction considerably increased the ventricular pressure and the intensity of the ejection systolic murmur (Fig. 2).

The patient also illustrates the difficulty in diagnosing aortic valve calcification. Despite heavy calcification confirmed at operation, and careful scrutiny, no calcium was seen on the image intensifier screen.

SUMMARY

Three patients with aortic stenosis and heart block are described. The diagnosis of aortic stenosis may be difficult in the presence of heart block, but may be suspected by a prolonged ejection time on the external carotid trace, and an abnormally low systemic arterial pressure for a slow ventricular rate. The electrocardiographic features of left ventricular hypertrophy may be obscured by the presence of right bundle-branch block which is commonly present in complete heart block. Clinical evidence of left ventricular hypertrophy may also be obscured when emphysema is present.

Bacterial endocarditis occurred in one patient when pacing was carried out with an endocardial electrode catheter and an external pacemaker without prophylactic penicillin.

Aortic valvotomy and long-term pacing by an epicardial-abdominal system of pacing has been successfully carried out with complete relief of symptoms.

The value of atrial contraction in augmenting ventricular systole has been confirmed.

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