CRYPTOGENIC HEART DISEASE IN THE BANTU OF SOUTH AFRICA*

A meeting of the Association of Physicians of South Africa (M.A.S.A.) was held at Baragwanath Hospital, Johannesburg, on 9 July 1960.

Dr. V. H. Wilson, senior physician at Baragwanath Hospital, as Chairman of the meeting, welcomed Dr. M. M. Suzman, Dr. S. J. Fleishman, and 30 members of the Association of Physicians. He stated that up to 400 cases of cryptogenic heart disease (CHD) per year were admitted to Baragwanath Hospital, and added that this disease still defied explanation.

The Problem

Dr. K. J. Keeley (Baragwanath) presented the aetiological spectrum of heart disease as seen in the wards. Cryptogenic heart disease forms 37·5% of the total, outstripping chronic rheumatic heart disease (20·3%) and hypertensive heart disease (19·6%). Males constitute 64·0% of the cryptogenic cases.

Clinical Presentation

Dr. H. C. Seftel, of Baragwanath Hospital, outlined the clinical presentation of cryptogenic heart disease: The condition affects both sexes with a male predominance of 1·6 : 1. It occurs in all age groups, including children under 10 years, but the maximum age incidence for both sexes is between 30 and 50.

In respect of tribal origin, occupation, educational status, diet, alcohol and tobacco consumption, and the use of witch-doctor preparations, the patients with CHD appear to be essentially similar to the African population at large.

Clinically, the condition presents with heart failure, both left-sided and right-sided. The circulation is hypokinetic or normokinetic and sinus rhythm is usual. The blood pressure is often elevated on admission, but this is transient, returning to normal within a week or two. The heart is always enlarged, often markedly so. A heaving impulse is frequently felt over the left or right ventricle. Gallop rhythm is almost invariable. In many cases functional mitral or tricuspid systolic murmurs are heard. The great majority of cases show no clinical stigmata of malnutrition.

Electrocardiographically the majority of cases show increased voltage of the QRS complex and inverted T waves over the left ventricle.

With bed rest, digitalis, and diuretic therapy the signs of heart failure and gallop rhythm disappear and the systolic murmurs diminish in intensity. Cardiomegaly, however, usually persists and at this stage (about 1 - 2 weeks after admission) the patients are discharged from hospital. Our knowledge of the subsequent course is incomplete, no systematic follow-up study having yet been undertaken. The general view, based entirely on hospital experience, is that many, if not most, cases relapse and that with each relapse the condition becomes increasingly refractory to therapy.

There is, however, a group of patients with CHD whose course has been studied in some detail, and has been shown to be quite different from that described above. This group consists of patients in whom the onset of heart failure occurs in the postpartum period, or less often, in the last trimester of pregnancy. These peripartum cases are not uncommon, constituting about one third of all cases of CHD in females. They tend to be women of high parity and high child-bearing age, who have a higher-than-normal incidence of twin pregnancies.

These cases are clinically similar to the general run of cases with CHD except that in half the cases recovery ensues. This may occur within a few weeks, but usually requires between 3 and 12 months. Recovery may occur despite the cessation of digitalis and diuretic therapy and the resumption of normal physical activity while the heart is still enlarged. The other half pursue a chronic or relapsing course. The total mortality rate appears to be about 25%. Prognosis is significantly related to high age, high parity, and persistent lactation after the onset of heart failure. The condition may recover completely only to recur after a subsequent pregnancy.

Course of Illness

Dr. Keeley illustrated the possible course of the illness with selected cases. Death rarely results from the first attack of congestive failure; the number of patients who recover completely from the first attack is not known, because many subjects do not return after recovery from the congestive state. Usually there is a steady progression, punctuated by repeated admissions in congestive failure, until death ensues. By contrast, one patient was known to have had a complete clinical and radiological remission for at least 19 months after having been in congestive failure intermittently for 4½ years.

Radiology

Dr. N. Joffe, of Baragwanath Hospital, said that in patients who are X-rayed during an acute phase of the disease, be this the first episode of cardiac failure or a recurrence, the most constant radiological finding is the presence of cardiomegaly

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which varies from a slight increase in the transverse diameter of the heart to a gross degree of enlargement. Both the left and right sides of the heart are involved and, when the cardiomegaly is of a marked degree, the radiological appearances are not unlike those of a pericardial effusion. Although biventricular enlargement is the rule, predominance of the left ventricle is not uncommon. The aortic shadow shows no abnormality. Slight enlargement of the main pulmonary artery segment and pulmonary vascular engorgement are common findings while the patient is in cardiac failure; pulmonary oedema is rarely observed. Small unilateral or bilateral pleural effusions are frequently demonstrated, but large effusions are uncommon.

In the majority of cases fluoroscopic examination of the chest discloses a diminution in the amplitude of cardiac pulsations and, rarely, an almost complete absence of pulsations may be observed. In a fair proportion of cases, however, cardiac pulsations appear to be of normal amplitude. Pulsation of the aorta is commonly of poor amplitude.

The presence of associated primary lung disease is not common. Acute pulmonary infections and tuberculosis do not appear to be more frequent than in the average Bantu population; pulmonary infarcts are not infrequent in this disease and may be demonstrated radiologically by the presence of one or more pulmonary opacities which may be found in any situation in the lung fields. These opacities may be unilateral or bilateral in distribution and are occasionally accompanied by small pleural effusions or elevation of the corresponding dome of the diaphragm.

Recovery from the acute episode of cardiac failure is usually accompanied by radiological evidence of some reduction in heart size and an improvement in the amplitude of cardiac and aortic pulsations.

Pathology

Dr. C. Isaacson, of the South African Institute for Medical Research and Baragwanath Hospital, stated that this information had been obtained from 537 consecutive cases of heart failure in a total of 2,367 necropsies; the cases of heart failure thus formed 22·7% of all causes of death. Cryptogenic heart failure is the fourth commonest cause of cardiac failure in the Bantu in Johannesburg as seen at necropsy. It is probable that the incidence is even higher, since other cases of heart failure were observed, such as hypertensive heart failure in which the lesions present seemed inadequate to account for failure. The corrected sex ratio showed a male preponderence of 1·6:1.

Macroscopic examination showed that the heart was hypertrophied and dilated in all cases. The hypertrophy was usually generalized and involved all chambers equally, but in 3 cases the enlargement was more marked on the right side. In 30 hearts no other significant lesions were observed. In 48 hearts intraluminal thrombi were present. These varied from recent mural thrombi superimposed on an apparently intact endocardium and myocardium to dense organized thrombi forming fibrous plaques obliterating the spaces between the trabeculae carneae. In 2 hearts thick, fibrous plaques were present overlying the endocardium. From the microscopic examination it is believed that these represent old organized thrombi. In only 1 heart was the extent of the endocardial fibrous plaques comparable to that described in Uganda. No significant valvular lesions were seen in any case. In some hearts the coronary arteries showed mild atherosclerosis. In no case was significant diminution of the lumen present and the degree of atherosclerosis was no greater than that seen in hearts of patients dying from non-cardiac disease. Microscopic examination of the endocardium showed it to be normal in the majority of hearts with only occasional areas of non-specific diffuse and focal thickening. In some cases small foci of infiltration by lymphocytes, macrophages, and plasma cells were noted. In some hearts large bulky recent thrombi could be seen deposited on an apparently normal endocardium. In other areas the thrombi were composed of very small fibrin clots. The thrombi were most frequent between the interstices of the trabeculae carneae. The myocardium consistently showed hypertrophy of the muscle fibres which contained large 'stag-horn' nuclei. Sometimes the fibres showed a mild non-specific intracellular oedema with separation of myofibrils. No parasites or significant haemosiderin deposits could be demonstrated. In many hearts there was some degree of interstitial oedema between the muscle fibres associated sometimes with a deposition of a poorly-staining collagenous tissue. Small foci of fibrous scarring were sometimes observed with degeneration of muscle fibres. Such scarring was usually mild but was more prominent in 17 cases. Such areas of scarring did not imply coronary ischaemia, since the coronary vessels were normal. In no case did the small myocardial arteries and arterioles show evidence of fibrinoid necrosis or an arteritis. Examination of the other organs showed generalized venous congestion and in addition in 28 cases there was evidence of visceral infarction. The kidneys showed only the normal changes associated with increasing age.

The findings in the hearts of children dying of cryptogenic heart failure were essentially similar to those observed in adults. The endocardium of the left ventricle in these cases showed perhaps more fibrous thickening than in adults.

Points in Discussion

While the cases of CHD seen at the Baragwanath Hospital were mostly in urbanized Africans, Dr. J. V. O. Reid (Durban) and Dr. M. Nellen (Cape Town) stated that they had observed the condition in rural Africans as well. Dr. Reid had also encounted peripartum cases of CHD. His findings in these and in patients with CHD in general were similar to those of the Baragwanath Hospital workers.

Several speakers raised the problem of differentiating CHD from pericarditis. Dr. Wilson thought that if a heaving or thrusting impulse was felt over the praecordium, pericarditis was most unlikely. He stressed the danger of attempting to distinguish the two conditions by pericardial paracentesis. Dr. Reid agreed and advocated right atrial catheterization to assess the thickness of the atrial wall. He also stated that while pulsus paradoxus might occur in both conditions, a marked degree of paradoxus with disappearance of the pulse on inspiration favoured the diagnosis of pericarditis. Dr. Nellen stated that the finding of quadruple rhythm strongly suggested CHD. He had never found this rhythm in pericarditis.

Dr. M. M. Suzman (Johannesburg) raised the question of the relationship of CHD to beri-beri. Dr. Wilson reported that beri-beri was seen at the Baragwanath Hospital but, by contrast with CHD, was a rare disease. Patients with CHD did not have a hyperkinetic circulation, they were not influenced by thiamine and they were not usually rapidly reversible.

Dr. S. Fleishman (Johannesburg) asked whether cortisone had been used in CHD. Drs. Reid, Keeley, and Seftel had tried cortisone or its analogues but without success. Dr. Seftel mentioned that an attempt had been made to detect cardiac antibodies in the serum of cases of CHD by the agar-gel diffusion and tanned, red-cell techniques. The results were negative.

Dr. Suzman thought that CHD might have been diagnosed as hypertensive heart disease in the past.

Dr. L. Klugman (Johannesburg) suggested that the hypertrophy of the heart in CHD was due to the diastolic overload in a dilated heart. Dr. Isaacson considered that it had been satisfactorily demonstrated that the fibrous endocardial thickening seen in any heart failure was a mechanism to compensate for the increased tensile load placed on the myocardium of a dilated heart.

Dr. C. F. van der Merwe (Pretoria) asked what the autopsy findings were in cases of CHD in which recovery had ocurred and death was subsequently due to some unrelated cause. Dr. Seftel quoted a single case in which clinical, radiological and electrocardiographic recovery had occurred, the patient dying 2 months later from infective hepatitis. The heart still showed hypertrophy on postmortem examination.

Dr. Wilson, as Chairman, urged the meeting to accept that the cause of CHD was still unknown and warned that in attempting to find a stereotyped pathological explanation an important factor in myocardial dysfunction might be overlooked.