# Endomyocardial fibrosis in a Zambian

M. N. Lowenthal M.B., Ch.B. (Rand.), M.R.C.P. (Edin.) Physician, General Hospital, Ndola, Zambia

J. Fine, M.D. (Glas.), D.P.H.
Pathologist, Central Hospital, Kitwe, Zambia

The purpose of this communication is to report a proven case of endomyocardial fibrosis (E.M.F.) in a Zambian African.

Most of Zambia is situated on a high plateau 4,000 feet or more above sea level, and the country enjoys a temperate climate with sharp differentiation between the rain season (November-April) and the dry season (May-October).

Parry (1964, 1965) points out that endomyocardial fibrosis (E.M.F.) has been extensively reported from the hot, wet parts of Africa, but not from the drier or more temperate parts at the northern and southern extremetics of the continent. Brockington et al. (1967) in reviewing the cases of E.M.F. that have been reported in Europeans resident in tropical Africa, state that the disease is one of humid tropical zones in Africa and South America. Cases have also been reported from Ceylon and Malaya (Nagaratnam and Dissanayake, 1959; La'Brooy 1957) and South India and Brazil (W.H.O. Chronicle 1967). Davidson and Ross (1966) briefly described a case from the Ndola General Hospital who at autopsy, was found to have a "thin, fibrotic film covering the endocardium of the left ventricle". and in whom, microscopically, the endocardium showed fibrosis. Davidson subsequently, (1967), stated that this case was one of E.M.F.

#### CASE REPORT

A male (village Ndobela, chief Tungati, boma Luwingu) aged about 25, was admitted to the General Hospital, Ndola, on 22nd November, 1966. He stated that his abdomen had been swollen for the four months prior to his admission. A similar episode had occurred in 1964 for which he was admitted to hospital and from which he had recovered after treatment.

On physical examination the patient was a young man in gross congestive cardiac failure. Temperature, 98°F.; heart rate, 72 per minute; blood pressure, 120/90. The face was hyper-pigmented suggesting alcoholism, but this was denied by the patient and by his relatives. The jugular venous pressure was very high and the veins were very distended and hardly pulsated. The pulse was of small volume. The precordium was quiet but the heart was obviously enlarged to palpation. The first and second heart sounds were normal and added sounds were not audible. Murmurs of mitral and of tricuspid regurgitation were present. Ascites was very marked, but ankle oedema was only slight. After paracentesis the spleen was palpable and the liver enlarged and coarsely nodular. Crepitations were present throughout the lung fields.

# INVESTIGATIONS

Haemoglobin, 10 gms./100 ml.; total serum proteins, 7.1 gms./100 ml.; albumin, 2.6 gms./100 ml.; globulin 4.5 gms./100 ml.; serum bilirubin, 2.1 mgs.% (conjugated 0.9 mgs.); alkaline phosphatase 5 King Armstrong units, thymol turbidity 4 units. Kahn test negative. Urine test for protein ±, glucose, nil. X-ray

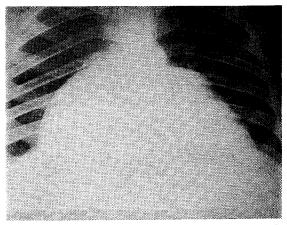


Fig. 1. Chest X-ray showing cardiac enlargement and prominence of right atrium and pulmonary conus.

of the chest (Fig. 1), showed a very lrage cardiac shadow with the right atrium and the pulmonary conus especially prominent. The intravenous injection of carbon dioxide (Phillips et al. 1961) demonstrated the absence of excess

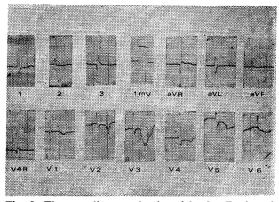


Fig. 2. Electrocardiogram showing right bundle branch block and T wave changes.

pericardial fluid or of pericardial thickening. An electrocardiogram (Fig. 2) showed right bundle branch block and T wave changes, and in lead III a prominent Q wave was present. Heaf test was positive (blister). Ascitic fluid protein was 3.1 gms./100ml.

## **PROGRESS**

He was treated with bed rest and digoxin, frusemide (Lasix), mersalyl, multivitamin tablets, isoniazid and thiacetazone (Thiazina) and potassium chloride, but did not improve.

The entire hospital course was a stormy one, with the patient frequently complaining of headache and joint pains. Bouts of shivering occurred from time to time and on several occasions the temperature spiked to 101°F. On the 23rd day after admission a pleural rub was noted. The following day the cardiac signs were found to have altered to a remarkable degree. The pulse had a water hammer quality and blood pressure was 125/50, heart rate was 56 per minute, and over the whole precordium a loud to-fro murmur was present, in addition to a pericardial scratch at the left sternal edge and at the apex. Corrigan's sign was present in the arteries of the neck and pulsation in the finger pulps was detected. The temperature at this time was 99°F. The lung fields were clear. The following day the patient died.

#### POST-MORTEM EXAMINATION

This examination was performed by M.N.L. 19 hours after death. Following this the heart was sent first to J. F. in Kitwe and then to Prof. M. S. R. Hutt of Makerere University College, Kampala.

Heart (macroscopic): The heart weighed 615 grammes. On the right side there was gross enlargement of the right atrium which contained ante-mortem thrombus and which showed patchy endocardial thickening with an area of calcification in the wall. The tricuspid ring was dilated and easily admitted four fingers (Fig. 3).

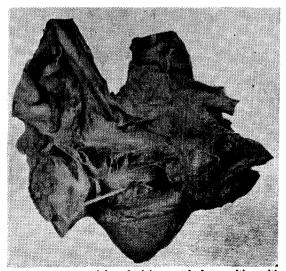


Fig. 3. Right atrial and right ventricular cavities with dilated tricuspid ring.

The tricuspid valves were normal although the chordae tendineae were pulled down and incorporated into a thick endocardial scar which obliterated the apex of the right ventricle (Fig. 4). The scarring was extensive but

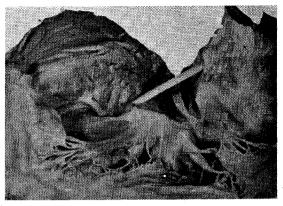


Fig. 4. Right ventricular cavity showing chordae tendineae incorporated into endocardial scar which is obliterating apex of right ventricle.

did not extend deeply into the underlying myocardium. However, the papillary muscles and trabeculae carneae were partly replaced by the fibrous tissue. The outflow tract of the right ventricle was slightly dilated. The pulmonary valve was normal. The mitral valve ring was slightly dilated but the valve cusps were normal and the aortic valves were normal. There was a small patch of fibrosis, approximately 1 cm. across, on the inflow tract of the left ventricle just above the apex. The coronary arteries were normal. The thoracic and abdominal aorta were free from atheroma. The pericardial cavity was obliterated.

Heart (microscopic): Sections of the right ventricle showed a thick collagenised scar with some fibrin on the surface. The scar tissue surrounded papillary muscles which were undergoing atrophy. In the deeper areas of the scar there was a typical zone of vascularization with a few lymphocytes and histiocytes in the looser connective tissue. Some strands of fibrous tissue extended into the inner portion of the myocardium. The deeper myocardium showed no abnormalities. The endocardium of the right atrium was fibrous and thickened. The right atrial myocardium showed fibrosis and degeneration of muscle fibres. The pericardium showed fibrous thickening and round-celled infiltration.

A vast quantity of ascitic fluid was present. The liver showed a coarse nodular cirrhosis. The spleen was greatly enlarged. The kidneys were congested but otherwise normal as were the adrenals, uretes and bladder. The right pleural cavity contained approximately 250 mls. of clear straw-coloured fluid. The bronchi contained an excess of frothy fluid and bilateral pulmonary oedema was present.

## DISCUSSION

Precise residential details during the patient's lifetime are not known—the district of his birth, Luwingu, is at an altitude of 4.650 feet and is situated at latitude 10°30 minutes south, longitude 29°30 minutes east. Ndola's altitude is 4,126 feet and is situated at latitude 13° south, longitude 28° 40 minutes east. In both areas the mean annual rainfall is 46 inches falling during November to April, the other months being rain free. Mean annual temperature for both areas is 65°-70° F.

The clinical course in this patient was indicative of a continuing active process. The autopsy findings were those of "classical" right ventricular E.M.F. (Williams et al. 1954, Davies and Ball 1955, Abrahams 1962). We believe that cases of E.M.F. will in future prove to be not uncommon in Zambia and neighbouring countries. despite absence of the disease in previously reported series from Rhodesia (Gelfand 1957, Baldachin 1963) and South Africa (Schrire 1960, Schwartz et al. 1958). Recently Dukes and Gelfand (1967) have described the case of a Malawian resident in Rhodesia in whom aneurysmal enlargement of the right atrium occurred in association with mitral incompetence. These authors suggest that this case may be one of E.M.F. This diagnosis is strongly suspected in at least four patients currently attending the cardiac clinic at the Ndola General Hospital. The present lack of facilities prevents proof of this diagnosis.

# **SUMMARY**

A case of endomyocardial fibrosis proved at postmortem is described in a young Zambian African male. This is probably the first well-documented case report of the condition from Southern Africa. It is believed that the condition will not prove to be a rarity locally.

# **ACKNOWLEDGEMENTS**

We are grateful to Mrs. J. Storrs, radiographer, Ndola General Hospital, for photographic assistance. Climatic data was kindly provided by the Meteorological Office, Ndola Airport. The Permanent Secretary, Ministry of Health, Zambia, kindly gave permission to publish. Our thanks are due to Professor M. S. R. Hutt for his opinion on this heart, and to Dr. Arthur Hollman, consultant cardiologist, and Dr. David Wayne, both of University College Hospital, London, for their assistance.

#### REFERENCES

 Abrahams, D. G. (1962) Quart. J. Med. N.S., 31, 1. Endomyocardial Fibrosis of the Right Ventricle.

- Baldachin, B. J. (1963) Centr. Afr. J. Med., 9, 463.
   Cardiovascular Disease in the African.
- 3. Ball, J. D., Williams, A. W. and Davies, J. N. P. (1954) Lancet, 1, 1049 Endomyocardial Fibrosis.
- Brockington, I. F., Olsen, E. G. J., and Goodwin, J. F. (1967) Lancet 1, 583. Endomyocardial Fibrosis in Europeans Resident in Tropical Africa.
- Davies, J. N. P. and Ball, J. D. (1955) Brit. Heart, J. 17, 337. The Pathology of Endomyocardial Fibrosis in Uganda.
- Davidson, J. C., and Ross, D. M. (1966) Centr. Afr. J. Med., 12, 138. The African Cardiomyopathies
- 7. Davidson, J. C. (1967) Personal communication.
- Dukes, D. C. and Gelfand, M. (1967) Centr. Afr. J. Med., 13, 203. Aneurysmal Enlargement of the Right Atrium.
- Gelfand, M. (1957) The Sick African, p. 467 3rd ed. Juta, Cape Town.
- La'Brooy, E. B. (1957) Proc. Alumni Ass. Malaya,
   303. (Title not known and paper not read).
- Nagaratnam, N., and Dissanayake, R. V. P. (1959)
   Brit. Heart J., 21, 167. Endomyocardial Fibrosis in the Ceylonese.
- 12. Parry, E. H. O. (1964) Cardiomyopathies. Ciba Foundation Symposium, p. 324. London.
- 13. Parry, E. H. O. (1965) Brit. med. J., 1, 921. Endomyocardial Thickening.
- Phillips, J. H., Burch, G. E., and Hellinger, R. (1961)
   Amer. Heart J., 61, 748. Use of Intracardiac Carbon Dioxide in Diagnosis of Pericardial Disease.
- Schrire, V. (1960) Amer. Heart J. 59. 835. The Racial Incidence of Heart Disease at Groote Schuur Hospital, Cape Town. Part III. The less common forms of heart disease.
- Schwartz, M. B., Schamroth, L. Seftel, H. C. (1958) Med. Proc. 4, 275.
- 17. W. H. O. Chronicle (1967) 21 407, The Cardiomyopathies.