Thorax, 1980, **35**, 641–643

Editorial

Sarcoid heart disease: a review and an appeal

Sarcoid heart disease is a neglected condition. Though it has long been recognised, clinicians have been reluctant to make the diagnosis. The case reporting in the United Kingdom is extremely patchy and it is difficult to think that it reflects the true incidence of the condition. Finally, there is much that we do not know about this disease and its prognosis.

Bernstein *et al*[1] first described sarcoid lesions of the heart in 1929 and 10 years later Gentzen[2] reported the first death from myocardial sarcoid. In subsequent years many reports of small numbers of cases and reviews of published series appeared, such as that of Porter.[3] Many of the patients were negroes in the southern United States and this fact still tends to be overemphasised.[4] However, most of the cases in the United Kingdom are white.

In the United Kingdom the sole report of a fatal case was the classic one of Forbes and Usher[5] in 1962, until from Cambridge, six necropsy cases were described in 1972.[6] This stimulated an enquiry throughout Britain, which resulted in the reporting of 50 cases,[7] 20 of them with necropsy confirmation. At the time this was much the largest published series. In Japan sarcoidosis is studied on a national basis, and in 1976 Matsui *et al*[8] made a most valuable contribution describing the clinicopathological analysis of 42 fatal cases. In 1977 Roberts *et al*[9] reported 35 necropsy cases and claimed to review the world publications (94 references) but their omission of any reference to the two large earlier series[7 8] invalidates some of their analyses and conclusions. An updated report on the United Kingdom series was given to the Eighth International Congress on Sarcoidosis and Other Granulomatous Diseases in Cardiff in 1978.[10] It comprised 128 cases with 57 deaths and 45 necropsies.

The collection of cases and their follow-up continues in the hope that we may in time build up a more complete picture of the epidemiology, natural history, and effects of treatment of this condition.

At the end of 1979 a cumulative total of 163 cases was on file. Only 11 of these patients were coloured. There are no available figures to indicate the percentage of coloured people in the population from which these cases originated. However, the large number of Caucasian cases is notable. The age at presentation with cardiac features ranged from 18 to 77 years (mean 44·4 yr) and there were roughly equal numbers of men and women. Seventy patients have died at an average age of 47 years. Death was sudden in 45, and in 26 no previous diagnosis of heart disease or sarcoid had been made. There were 53 necropsies.

Most of the patients in this series presented with cardiac features, as shown in the following table.

Presenting feature	*Number of cases*
Complete heart block	31
Lesser degree of heart block, including bundle branch block	56
Ventricular ectopic beats or ventricular tachycardia	64
Supraventricular tachycardia	35
Myocardial disease	34
Sudden death	26
Simulating myocardial infarction	10
Valve involvement	3
Pericardial involvement	2
Road traffic accident	3

This, of course, adds up to more than 163 because some cases present with more than one feature. Any possible bearing of the sarcoid on the three cases of road traffic accident is uncertain. Sarcoid heart disease should be suspected in any case of unusual cardiac disease for which there is no other explanation. The ultimate proof of the diagnosis is by necropsy and it is, therefore, with particular regret that we record a disappointing number and quality of necropsies in this series.[11]

In the living the diagnosis is made on the following grounds: (1) cardiac condition consistent with sarcoid heart disease; (2) clinical diagnosis of sarcoidosis; (3) histological confirmation of sarcoidosis—this should be sought wherever possible either on suspicion of the diagnosis or to confirm the clinical diagnosis. In the absence of any other biopsy material the Kveim test or a transbronchial biopsy can be of diagnostic value.

Subclinical cardiac involvement is probably very common in classical sarcoidosis. Silverman *et al*[12] report cardiac granulomata in 27% of patients

Address for reprint requests: Dr Hugh A Fleming, Regional Cardiac Unit, Papworth Hospital, Papworth Everard, Cambridge CB3 8RE.

dying of sarcoid. However, the heart may be massively involved while there is minimal involvement in other organs.[7] Endomyocardial biopsy has occasionally been positive and, of course, this is diagnostic, but a negative result is of no importance[11] because of the patchy distribution of the disease in the myocardium.

Geographical distribution

Although it is generally agreed that there are wide differences in the incidence of sarcoidosis in different areas and ethnic groups, accurate information about incidence (as opposed to prevalence) is available for only a few areas and groups.[13] In Britain the British Thoracic and Tuberculosis Association[14] compared the incidence in four areas. This varied from 0·21 per 10 000 among men in the most northerly area to 0·45 per 10 000 among women in the most southerly, with a generally lower incidence in the north.

The distribution of the centres through which cases of sarcoid heart disease have been reported to me is as follows: East Anglia 62, London 28, Oxford area 22, elsewhere 51, giving the total of 163.

No cases have been reported from the West country, even from the Devon Heart Block and Bradycardia Survey[15] and yet in the UK study of sarcoid[14] this area had the highest incidence. St George's Hospital, London with their long experience of heart block report no cases. Only 17 of the 163 cases reported came from north of Leicester, in spite of good communication with consultants in the north.

The 62 cases from East Anglia with a population of only two million is, therefore, remarkable. Three necropsy cases ranging in age from 25 to 41 years came from the City of Ely with a population of only 10 000.

If this discrepancy of distribution is real it could be of importance. In the meantime I assume that proximity to my known interest in Cambridge largely accounts for this distribution. I hope our efforts will enable us to compile a complete East Anglian register.

Prognosis and treatment

The prognosis in the individual case is extremely variable and it is difficult to know how actively to pursue treatment in the symptom-free case. Heart block, arrhythmia, and heart failure should obviously be treated in their own right, but when should steroids be given? The incidence of sudden death is very disturbing and clinicians having made the diagnosis of myocardial sarcoid cannot have any sense of security. There is recent evidence that sarcoid heart disease could have been involved in more than one fatal air crash. The matter is, therefore, of considerable importance.

Suspected cases should at least be investigated by 24-hour ECG tape monitoring, echocardiography, exercise testing, and probably by radionucleide myocardial imaging and estimations of angiotensin converting enzyme (ACE),[16] in an attempt to assess cardiac involvement and the activity of the sarcoidosis.

The collection of data about all cases of sarcoid heart disease appears to be the best way of assessing the importance of this condition and its natural history. It is to be hoped that other countries will follow the lead of Japan in establishing national registers for the collation of information.

In the United Kingdom there is no such national study but the present collection of cases continues, and it already provides a useful reservoir of clinical experience. Thus, if you have a proven or even a suspected case, please let me have details.

HUGH A FLEMING
Regional Cardiac Unit
Papworth Hospital
Cambridge

References

1 Bernstein M, Konzellman FW, Sidlick DM. Boeck's sarcoid: report of a case with visceral involvement. *Arch Intern Med* 1929; **44:**721–34.
2 Gentzen G. Über Riesenzellengranulome bei zwei Fällen von Endocardifibrose. *Beitr Pathol Anat* 1937; **98:**375–98.
3 Porter GH. Sarcoid heart disease. *N Engl J Med* 1960; **263:**1350–7.
4 Lorell B, Alderman EL, Mason JW. Cardiac sarcoidosis. Diagnosis with endomyocardial biopsy and treatment with corticosteroids. *Am J Cardiol* 1978; **42:**143–6.
5 Forbes G, Usher A. Fatal myocardial sarcoidosis. *Br Med J* 1962; **2:**771–3.
6 Ghosh P, Fleming HA, Gresham GA, Stovin PGI. Myocardial sarcoidosis. *Br Heart J* 1972; **34:**769–73.
7 Fleming HA. Sarcoid heart disease. *Br Heart J* 1974; **36:**54–68.
8 Matsui Y, Iwai K, Tachibana *et al.* Clinicopathological study of fatal myocardial sarcoidosis. *Ann N Y Acad Sci* 1976; **278:**455–69.
9 Roberts WC, McAllister HA, Ferrans VJ. Sarcoidosis of the heart. A clinicopathological study of 35 necropsy patients (group I) and a review of 78 previously described necropsy patients (group II). *Am J Med* 1977; **63:**86–108.

10 Fleming HA. Sarcoid heart disease. In: Williams WJ, Davies BH (eds). *Proceedings of the VIIIth International Conference on Sarcoidosis and other Granulomatous Diseases*. Cardiff: Alpha Omega Press, 1980:493–6.
11 Fleming HA. Sarcoid heart disease. *Br Heart J* 1980; **43**:366.
12 Silverman KJ, Hutchins GM, Bulkley BH. Cardiac sarcoid—a clinicopathological study of 84 unselected patients with systemic sarcoidosis. *Circulation* 1978; **58**:1204–11.
13 Mitchell DN, Scadding JG. Sarcoidosis. *Am Rev Respir Dis* 1974; **110**:774–802.
14 British Thoracic and Tuberculosis Association. Geographical variations in the incidence of sarcoidosis in Great Britain: a comparative study of four areas. *Tubercle* 1969; **50**:211–32.
15 Shaw DB, Kekwick CA. Potential candidates for pacemakers. *Br Heart J* 1978; **40**:99–105. Updated by Shaw 1980 personal communication.
16 Annotation. ACE in sarcoidosis. *Lancet* 1980; **1**: 804–5.