RHEUMATIC FEVER IN MALTESE CHILDREN

E. A. CACHIA
M.B.CH.B. (MANCH.), D.C.H. (LOND.)

H. M. LENICKER
M.D., D.C.H. (LOND.)

Pediatric Department,
St. Luke's Hospital

The incidence of acute rheumatism in Maltese children has not been studied so far. In 1956 Professor J. E. Debono was of the opinion that the disease had become more frequent since the end of the last war. Our impression is that the disease has become even more common.

TABLE I
Showing number of children treated for acute rheumatism in one of the children's wards

1957	2
1958	3
1959	7
1960	6
1961	6
1962	5
1963	11
1964	6
1965	11
1966	13

During the years 1957 to 1966 inclusive 70 children were treated for rheumatic fever at one of the two Children's Wards at St. Luke's Hospital. It is reckoned that a similar number were treated at the other Children's Ward during the same period. At the same time an unknown number of children over 9 years of age were treated in the Medical Wards (since 1966 patients under the age of 12 years have been admitted to the Children's Wards) and an unknown number were treated at home and in private hospitals. Were all the figures known they would make up a considerable number. Rheumatic fever occurs too frequently to permit complacency. Bland and Jones (1951) in their study of 1000 rheumatic patients followed since childhood found signs of heart disease in 80%. They also concluded that 25% of rheumatic patients die within 30 years of the onset of the illness.

Initial attacks of rheumatic fever and subsequent recurrence are causally related to preceding infection by any type of Group A. haemolytic streptococci. This has now been accepted on clinical, serological, epidemiological and prophylactic evidence. Studies of rheumatic fever in closed communities have shown a fair constant attack rate of 3% following untreated epidemic streptococcal infections.

The symptoms and signs of rheumatic fever vary greatly. The clinical findings are determined by the sites of involvement, the severity of the lesions, the time of appearance during the course of the illness, and the stage at which the patient is first examined (Markowitz and Kuttner 1965). Yet it is very important that the physician be reasonably certain of the diagnosis. Treatment is prolonged

and expensive. The patient is confined to bed for several weeks or months and diagnosis may produce acute or chronic emotional tension in the parents or patient (Baum 1963).

Unfortunately there is no single diagnostic symptom or sign and no specific laboratory test for rheumatic fever. In 1944 the late T. Duckett Jones published criteria to guide the physician in making the diagnosis. These were subsequently revised by the American Heart Association. It is worthwhile repeating them here:

Major Criteria

Carditis
Polyarthritis
Chorea
Subcutaneous nodules
Erythema marginatum.

Minor Criteria

Fever
Arthralgia
Prolonged P-R interval in E.C.G.
Increased E.S.R. or C-reactive protein
Preceeding group A streptococcal infection

Previous rheumatic fever or inactive rheumatic heart disease.

The Co-operative Clinical Trial (1955) of the Medical Research Council and American Heart Association acepted two major manifestations or one major and two minor manifestations as justifying the diagnosis. Difficulties in diagnosis, however, may still occur because of the limitations of Jones's criteria and the great variability of the disease. A mild attack of rheumatic fever or a child presenting with abdominal pain may not satisfy the criteria. One of the commonest sources of difficulty is when the patient presents with polyarthritis, fever and a raised E.S.R. because this group of findings is common to several other diseases. Other manifestations, not included in the criteria, but which provide additional evidence of rheumatic fever are pallor, loss of weight, easy fatiguability, erythema nodosum, pre-cordial pain and epistaxis.

During the years 1962 to 1966 inclusive 66 children were referred to the Children's Ward for rheumatic fever. Following a period of observation and investigation 42 children were found to be actually suffering from the disease. Seventeen children were found to have short term fever or non-specific arthritis which cleared up within 3 days of admission. Of the remain ing seven patients, two had rheumatoid arthritis, one was found to have an appendix abscess, one had a neuroblastoma with metastases at the lower ends of the femora, one had acute lymphoblastic leukaemia, one had typhoid fever and the remaining one had undulant fever. Two patients were originally referred for fever of unknown origin and two for acute abdominal pain and these were subsequently found to be suffering from rheumatic fever. This made up our total of 46 patients. Thirty seven had an initial attack and 9 had a recurrence.

We have made a study of the 46 children who were treated for rheumatic fever. Rheumatic fever has long been associated with poor living conditions and substandard housing. We have confirmed this. Affected children came mainly from large families, the average number of children in each family being 5.4. The average number of rooms per family was 3.6. Regarding the father's occupations 12 belonged to class V, 8 to class IV, 15 to class III and 3 to class II. The occupations of the remaining 8 fathers had not been recorded. The ages of the patients ranged from 3 to 11 years with an average of 7.3 years. Fourteen of the patients had a previous history of a sore throat or tonsillitis and 2 patients had furunculosis. In those patients who had no such history we assumed that there was a mild or subclinical infection.

Four of the children had had their tonsils removed before they had their initial attack of rheumatic fever.

Carditis occurred in 40 of the children. A significant heart murmur was found in 33. X-ray chest for heart was taken of the 22 patients and the heart was enlarged in 14 of them. One child had pericarditis with effusion. Congestive heart failure occurred in 8 patients. Five of the latter had a history of rheumatic fever. Polyarthritis as evidenced by pain and clinical signs involving two or more joints occurred in 28. Chorea affected 5 girls and

one boy. One girl developed hemi-chorea while she was being treated for rheumatic carditis. In three of the children chorea was the sole manifestation of the disease. Subcutaneous nodules were not noted in any of the children but erythema marginatum was observed in one child.

The most common minor manifestation met with was a raised E.S.R. which was found in 41. Fever occurred in 38. Arthralgia was complained of by 4 children who did not have polyarthritis. More frequent use of the electrocardiogram in the Children's Ward has only been possible during the last two years. In all 13 of the patients had an E.C.G. A prolonged P-R interval was found in one patient. This may possibly be due to the fact that the electrocardiograms were taken several days after admission. Of the other manifestations, pallor was almost universal during the acute stage.

Comment

Rheumatic fever can be a very serious and crippling disease. It is evident that the disease is far from rare in Malta and is probably on the increase. In order that the incidence of rheumatic fever and the prevalence of rheumatic heart disease in the Maltese Islands be known the disease should be made notifiable. This has been the practice for some years in selected areas of Great Britain notably Sheffield and Bristol. The criteria for notification being:

1. Rheumatic pains or arthritis accompanied by a rise in temperature.
2. Rheumatic chorea.
3. Rheumatic carditis.
4. Valvular heart disease of rheumatic origin (Jameson and Parkinson 1963). Notification and registration of the disease has also been strongly recommended by the W.H.O. expert committee on Rheumatic Fever 1966 as the first step in any programme directed at the prevention of rheumatic fever. For a start, known cases of rheumatic fever should be notified. The next step would be to detect previously unknown cases through examination of special population groups such as school children and expectant mothers.

If group A streptococcal infection could be eradicated rheumatic fever would disappear.. The prevention of acute rheumatism is possible today only by prevention or treatment of group A streptococcal infections (W.H.O. Report 1966). Initial attacks which might possibly be prevented still occur because many children do not receive medical attention for antecedent streptococcal infection either through ignorance and poverty, or through the misguided advice of neighbours and persons selling medicines over the counter. Unfortunately a popular belief shared by many is that tonsillectomy prevents rheumatic fever but, even in this small series, 4 children developed acute rheumatism in spite of previous tonsillectomy.

The diagnosis of streptococcal pharyngitis or tonsillitis cannot be made unless the physician has access to a bacteriological service which is available every day including Sundays, holidays and festas. Many cases of pharyngitis and tonsillitis — with or without exudate — are non-bacterial in aetology and do not lead to rheumatic fever. Patients with such infections do not benefit from antibiotic therapy. A culture of the throat would confirm or disprove streptococcal infection. Throat cultures from patients with acute streptococcal infections are positive for Beta-haemolytic streptococci in over 95% of cases (Markowitz and Kuttner 1965).

The drug of choice in the treatment of streptococcal infections is penicillin. As yet no group A streptococcus has been confirmed as penicillin resistant. Unfortunately it often happens that the drug is only given for two or three days until symptoms have ceased to worry the parents. For penicillin to be effective in eliminating the streptococcus it must be given for seven to ten days. An alternative and convenient way has been recommended where crystalline penicillin G., procain penicillin and dibenzyl penicillin 1,200,000 I.U. are given in a single injection. Sulphonamides even though they suppress the symptoms and signs of acute streptococcal tonsillitis have been found

to be ineffective in the prevention of rheumatic fever presumably because they are unable to eliminate the streptococcus from the upper respiratory tract (W.H.O. Report 1966). Broad spectrum antibiotics are less efficient than penicillin and much more expensive. Erythromycin has been recommended for use in those allergic to penicillin.

Recurrent attacks of rheumatic fever in known rheumatic patients can be prevented by continuous prophylactic administration of penicillin, sulphonamides or broad spectrum antibiotics. This prophylactic treatment is also recommended for those patients with inactive rheumatic heart disease and in those with "probable rheumatic Fever" not meeting Jones's criteria, e.g. those with "pure" chorea. In the present state of knowledge prophylactic treatment is recommended for several years. Phenoxymethyl penicillin 100 to 125 mg orally may be given twice daily. When the parents are unreliable it is better to give dibenzyl penicillin 1,200,000 i.u. intramuscularly every 4 weeks to children and every three weeks to adolescents. If sulphonamides are used in prophylaxis sulphadiazine for example, 0.5 G. a day may be given to children and 1 G. daily to adolescents.

Summary

Rheumatic fever is still one of our great paediatric problems and is probably on the increase. A study of 46 children who had acute rheumatism has been presented. The disease should be made notifiable. Facilities for the isolation of the streptococcus should be made available to all physicians. Rheumatic fever and its recurrences can be prevented by adequate chemotherapy.

References

BAUM, D. (1963). Pediat. Clin. N. Amer., *10*, 899.

BLAND, E.F. and JONES, T.D. (1951). Circulation, *4*, 836.

DEBONO, J.E. (1956). "The Rheumatic Child", A survey of Child Welfare Problems, by the "Save the Children Fund". (Malta, Gov. Muscat).

JAMESON and PARKINSON (1966). A synopsis of Hygiene, 12th Edition, J & A. Churchill, London.

MARKOWITZ and KUTTNER (1965). Rheumatic Fever diagnosis and prevention. W.B. Saunders, London.

World Health Organisation (1966). Prevention of Rheumatic Fever, Technical Report Series, No. 342. Geneva.