

Variations in Manifestations of Rheumatic Fever in Relation to Climate*

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THE statement can frequently be found in textbooks and monographs that rheumatic fever is ubiquitous. This generalization (which is often attributed to Hirsch¹) can, however, scarcely be accepted today. Aside from the interest which has always been aroused by the seasonal incidence and familial occurrence of rheumatic fever, as well as the yearly variations in the severity of the disease, there has recently been some attempt to study, more accurately than has been possible before, the geographical distribution of the disease. The matter is one of importance not only in relation to rheumatic fever but in connection with other diseases. It is now well recognized, for instance, that infections such as scarlet fever and diphtheria, so common in the temperate zones, are almost unknown in some tropical countries. The geographical distribution of pernicious anemia, also, is very irregular. It is said to be almost unknown in China and Japan (Mills²).

My interest in possible climatological differences in the symptomatology

of rheumatic fever was awakened, almost ten years ago, on coming to Baltimore from New York. Arthritis was often the predominant symptom of acute rheumatic fever in the adult in New York, and the one for which the patient called the physician or entered the hospital. Combined with the severe arthritis, or sometimes without severe arthritis, there was not infrequently observed acute, severe, and sometimes fatal endocarditis, pericarditis, myocarditis, pleurisy and pneumonia.

In Baltimore, on the other hand, the acute severe arthritis, so familiar in New York, was rarely encountered in the wards of the hospital. In general, though the disease seemed common in Baltimore, it presented a somewhat different clinical picture, appearing more insidious, less outspoken in its arthritic manifestations, not so fulminant in its severer forms, but suggesting usually a chronic or relapsing progressive disease of the heart. In order to obtain more definite information on these points, the case histories of patients with rheumatic fever treated during the last five years in the adult medical wards of the Johns Hopkins Hospital have been analysed.

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It is difficult, as Newsholme³, who was interested in this question, found many years ago, to obtain accurate information regarding the regional distribution of rheumatic fever throughout the world. The statistics collected from hospitals by Faulkner and White,⁴ by Harrison and Levine,⁵ and by Seegal and Seegal⁶ indicate that rheumatic fever is commoner and more severe in the colder portions of the temperate zones than in the warmer portions. Seegal and Seegal also found that the incidence of rheumatic fever was greater from 1916 to 1918 than from 1918 to 1925. Excellent as these statistics are, they are based on diagnoses made in a variety of hospitals, and one cannot help but wonder upon what criteria the diagnoses were made. Do these statistics represent only those cases of rheumatic fever presenting arthritis, or do they include all cases of rheumatic fever? It is usually assumed that practically all cases of mitral stenosis are instances of rheumatic fever in the active, quiescent or healed stage of the disease. It is therefore very important in investigating the geographical distribution of rheumatic fever to know the regional distribution of mitral stenosis. Meleney and Kellers⁷ state that though rheumatic arthritis is rare in China, mitral stenosis is common. Harrison and Levine found mitral stenosis frequent in Boston, St. Louis and Baltimore, much less frequent in Galveston, Richmond and Oklahoma, and rare in New Orleans. Wood, Jones and Kimbrough⁸ find rheumatic fever and rheumatic heart disease about half as common in Virginia as in Massachusetts. Clarke⁹

believes that rheumatic fever is almost unknown in natives in the tropics, defining the tropics as an area lying between 23° 28' North and South. He states that he did not see a single case of rheumatic heart disease among 150,000 hospital cases in Perak, Malay States. Coburn¹⁰ quotes Getz as stating that in the last 4 years only three unquestioned cases of rheumatic pancarditis have been recognized at autopsy at the Hospital of Santo Tomas in Panama. According to Coburn, rheumatic fever is extremely rare in Porto Rico. In about 500 autopsies studied by Dr. Lambert and Dr. Pappenheimer no gross or microscopic lesions of rheumatic fever were found. In a later series, however, two autopsies showing rheumatic pancarditis have been recorded.

The information, therefore, that is obtainable through published statistics and from such important personal surveys as that made by Coburn, goes to show that rheumatic fever, in all its forms, is exceedingly rare in the tropics, though rheumatic heart disease is not unknown in some tropical countries; whereas all the manifestations of rheumatic fever are common in the colder portions of the North and South temperate zones, where rheumatic arthritis is particularly prominent. In the intermediate and warmer regions of the temperate zones, rheumatic fever is certainly not recognized with as great frequency as it is in the colder regions. The geographical distribution, according to Coburn, corresponds to that of scarlet fever.

In analysing the cases at the Johns Hopkins Hospital, rheumatic fever has

been considered as a generalized disease, and consequently there have been included in this category all instances of chorea, of mitral stenosis, of rheumatic pancarditis and of rheumatic arthritis, whether they have been observed in the active, quiescent or possibly healed stages of the disease. In the majority of the cases there was some definite evidence of activity while the patient was under observation in the hospital wards. It may be seen from Table I that rheumatic fever is quite common in Baltimore, for 1.37 per cent of all patients admitted to the adult medical wards suffer from rheumatic fever. The cases of rheumatic fever, moreover, form a fair proportion of all autopsies, at least 1.66 per cent. The figures for

TABLE I
ADMISSION OF RHEUMATIC FEVER TO
ADULT MEDICAL WARDS

Five Year Period—Sept. 1st, 1925 to Oct. 1st, 1930.

Total Admissions	= 10,385
Rheumatic Fever	= 142
Admission Rate	= 1.37%
Total Autopsies 1908-1929	= 8,164
Total Rheumatic Carditis	= 146
Per Cent Rheumatic Carditis	= 1.6

the admission rate are a little higher than those given by Faulkner and White for the Peter Bent Brigham Hospital in Boston (Table II).

Since 36 of the 146 autopsies showed rheumatic heart disease in acute or subacute form, it may be inferred that the disease may occur in Baltimore as an acute and severe infection. The total mortality for the 142 cases was 16.2 per cent. Table III shows the age incidence at the

TABLE II
REGIONAL DISTRIBUTION OF RHEUMATIC FEVER
(Modified from Faulkner and White)

PLACE	Av. Yearly Med. Admis.	Rheumat. F. Chorea	Per Cent
Johannesburg, S. Africa	2,906	169	5.8
Glasgow Royal Infirmary	2,655	126	4.74
Mt. Sinai, N. Y.	1,641	58	3.6
Royal Prince H., Australia	1,966	58	2.9
London Hospital	10,273	274	2.7
Univ. Hosp., Iowa	1,537	38	2.4
P.B.B. Hosp., Boston	2,480	31	1.3
J. H. Hosp., Baltimore	1,723	23	1.37
Univ. Hosp., Omaha	760	5	0.7
Barnes Hosp., St. Louis	1,358	65	0.47
Charity Hosp., New Orleans	5,349	28	0.4
Univ. Hosp., Atlanta, Ga.	2,500	2	0.08

TABLE III
142 CASES OF RHEUMATIC FEVER

Age	Total Cases	Auricular Fibrillation	Heart Block	Bacterial Endocarditis	Deaths
0-20	49	3	9	3	13
21-30	40	4	3	6	7
31-40	33	12	6	2	8
41-50	14	11	1		3
51+	6	6		1	2
Total	142	35	19	12	23
Per Cent		25.3	13.4	8.5	16.2

time of observation, the occurrence of auricular fibrillation, the incidence of prolonged A-V time and the number of cases complicated by bacterial endocarditis. It is interesting to note that the proportion of patients with auricular fibrillation increases with the increase in age, and that bacterial endocarditis usually occurs in the younger individuals. The percentage of 1st, 2nd and 3rd degree heart block is small, but this is no doubt due to the fact that single electrocardiographic records were made in many instances. The figures show that delay in A-V conduction is also commoner in the younger than in the older patients, associated in all probability with the greater activity of the rheumatic process in the younger patients.

Males and females were almost equally affected; many more instances of rheumatic fever occurred in the white than in the colored race (Table IV). A careful analysis of the histories of these 142 cases and the condition on admission to the hospital discloses some interesting facts. Table V records the frequency with which various manifestations of rheumatic

fever occurred during the life of these patients before they were seen in the hospital. The past history shows that in 15 there was no history of any illness, simulating rheumatic fever, and in 27, or 19+ per cent, there was no history of any rheumatic manifestation other than tonsillitis. There was a history of some form of arthritis without a history of cardiac disease in 37 patients, or in only 26+ per cent. On the other hand, cardiac disease is found to be remarkably common. In 14 cases there was a history of cardiac disease alone, and in 73 patients, or over 50 per cent, a history of cardiac disease either alone or in combination with some other manifestation of rheumatic fever. It is thus obvious that even in the histories of these patients cardiac disease is an important feature.

When one analyses the condition of the patients on admission to the hospital, the importance of cardiac disease becomes even more impressive (Table VI). Of the 142 patients, only 6 were admitted with arthritis alone and one with chorea alone. Of the entire number, 58, or only about 40 per cent, suffered with arthritis on admission, or during their stay in the hospital; while 135, or over 95 per cent, were admitted with cardiac disease or showed evidence of cardiac disease while in the hospital.

TABLE IV
SEX AND COLOR
142 CASES OF RHEUMATIC FEVER

Male	73	Female	69
White	59	White	55
Colored	14	Colored	14

TABLE V
HISTORY OF PREVIOUS MANIFESTATIONS OF RHEUMATIC FEVER—
142 CASES

No arth., ton., chorea, card.....	15	Cardiac alone	14
Tonsillitis alone	12	Arth. and cardiac	43
Chorea alone	5	Chorea and cardiac	5
Arthritis alone	34	Arth., chorea and cardiac	11
Arthritis and chorea	3	Total cardiac	73
Total arthritis, no cardiac	37		

TABLE VI
RHEUMATIC FEVER—142 CASES, CONDITION ON ADMISSION

Diagnosis	No.	Diagnosis	No.
Arthritis alone	6	Cardiac disease alone	80
Chorea alone	1	Arthritis and cardiac disease.....	49
Arthritis and chorea	0	Chorea and cardiac disease	3
Total arthritis	58	Arthritis, chorea and cardiac disease..	3
		Total cardiac disease	135

The observations upon these patients in hospital show quite definitely that many of them, though they gave no history of cardiac disease and though they were ignorant of the fact that they had cardiac disease, had had, nevertheless, cardiac disease probably for some years.

The figures thus emphasize the fact that rheumatic fever, as we see it, is essentially a disease of the heart, which may be preceded or accompanied by arthritis, often mild in character, by chorea, by tonsillitis, or occasionally by pleurisy, pneumonia, subcutaneous nodules and skin eruptions. During the acute stages the disease may be very severe or even fatal. Acute pericarditis occurred in 9 cases, acute pleurisy in 2; subcutaneous fibroid nodules were found in only 3 cases. Though many of these patients have died during the healed stage of the disease from the effects of the cardiac lesions or from such complications as bacterial endocarditis, at least 9 of the 22 autopsies showed that death was associated with some form of acute rheumatic carditis. The analyses which Dr. Thayer¹¹ has made of the fatal cases of acute and subacute rheumatic fever show that fatalities during the acute and subacute stages of the disease are not very rare at the Johns Hopkins Hospital.

Table VII shows the forms of heart disease observed in the 135 cases. Al-

most all of these patients presented the signs of disease of the mitral valve. A comparatively large number also showed the signs of aortic insufficiency. In three cases the signs were those of aortic insufficiency alone, though it seems probable that mitral disease also existed in these cases. In a few instances myocarditis or chronic adhesive pericarditis was present without the signs of mitral disease.

TABLE VII
FORMS OF CARDIAC LESIONS
IN 142 CASES OF RHEUMATIC FEVER

Mitral stenosis and insufficiency	76
Mitral sten. and aortic insuff.	50
Aortic insufficiency	3
Myocarditis	3
Acute pericarditis	3
Total	135

DISCUSSION

In reviewing these histories of patients, many of whom I have studied in the wards, the fact becomes quite clear that rheumatic fever is practically as common in the hospitals of Baltimore as in the hospitals of Boston. The character of the disease is not precisely the same, for arthritis, in the severe form, is certainly not common in Baltimore, and arthritis, even in mild degree, occurs in only a moderate proportion of cases. Cardiac disease, on the other hand, is extremely common and has been present in 95 per cent of the cases that we have studied. Patients are not often

seen in the florid stage of the disease, though severe acute rheumatic pancarditis is by no means unknown. As compared with the incidence of cardiac disease in other series of cases of rheumatic fever, the figures at the Johns Hopkins Hospital are rather high. Mackie¹² states that serious cardiac disease occurred, irrespective of age, in 68.3 per cent of his series of 393 cases of rheumatic fever, and that between the ages of 10 and 15 approximately 78.2 per cent of all cases presented evidence of cardiac disease during the first attack. Poynton¹³ found cardiac disease present in 70 per cent of 500 rheumatic children whom he examined. These figures are a high average for those found in the literature.¹⁴ At the risk of redundancy, then, it may be repeated that rheumatic fever, as seen in Baltimore, is essentially rheumatic carditis often of insidious onset, with comparatively mild acute exacerbations, but progressing none the less to a chronic deforming endocarditis with involvement of the myocardium and often of the pericardium, and resulting eventually in chronic invalidism and death. The carditis may be preceded or accompanied by attacks of arthritis, usually mild in character or by chorea.

The predominance of carditis in rheumatic fever and the insignificance of arthritis as a feature of the disease has been observed elsewhere. Meleney and Kellers have called attention to the fact that mitral stenosis occurs only a little less frequently in Peiping, China, than at St. Bartholomew's Hospital in London, and yet rheumatic arthritis is almost unknown in Peiping or in North China. Coffen¹⁵ points out that mitral disease is encountered in Ore-

gon with a frequency entirely disproportionate to the number of cases of rheumatic arthritis, and Houston¹⁶ in describing 88 cases of rheumatic fever which occurred during a period of four years amongst 115,213 general admissions to the Charity Hospital in New Orleans, emphasized the mild character of arthritic symptoms and the frequency with which cardiac disease occurred (51.1 per cent).

It seems possible, therefore, that rheumatic fever might be detected more frequently in the Southern States and in semi-tropical countries if the disease were regarded as one primarily of the heart, and if it were thoroughly appreciated that arthritis is an insignificant feature; an episode which may attract little attention or may be entirely absent.

CONCLUSIONS

The available statistics concerning the geographical distribution of rheumatic fever indicate that the disease is very rare or almost unknown in the tropics, and much less commonly observed in the warmer portions of the mid-temperate zones than in the colder portions.

In some regions where rheumatic arthritis is said to be rare, mitral stenosis is quite frequently observed.

At the Johns Hopkins Hospital in Baltimore the admission rate to the adult medical wards for rheumatic fever, in all its forms and in all its stages, over a period of five years was 1.37 per cent. The autopsy rate for rheumatic heart disease over a period of 21 years was 1.66 per cent. The disease, therefore, is comparatively common.

An analysis of 142 cases of rheumatic fever studied during this period

showed that symptoms or signs of cardiac disease appeared in the past histories of 50 per cent of the cases.

Cardiac disease was present on admission to the hospital or was detected during observation in hospital in 95 per cent of these cases. Only 6 of the entire 142 cases presented symptoms and signs of arthritis alone, but in 77+ per cent arthritis, often of mild degree, occurred at some time during the illness for which they were admitted to the hospital.

Rheumatic fever, as it is seen at the Johns Hopkins Hospital, is essentially

a disease of the heart, frequently preceded or accompanied by arthritis, often of mild degree, or by chorea. Severe acute arthritis is rarely seen, but acute pericarditis and pleurisy are not very infrequent.

It is suggested that the infrequency of severe acute arthritis, the great frequency of carditis and the comparative insidiousness of the disease during the acute and subacute stages, renders the clinical picture of rheumatic fever somewhat different in Baltimore from that generally described for more northern sections of the United States.

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