

## RHEUMATIC HEART DISEASE IN THREE DECADES (1942-1971)

By Paul Szekely

Rheumatic heart disease has been known as a usually progressive disease the complications of which significantly affect morbidity and mortality. However, three more recent events have favourably influenced its course, namely the introduction of antistreptococcal prophylaxis, cardio-valvar surgery, and the natural decline in the severity of initial carditis. In this communication, the clinical course of rheumatic fever and rheumatic heart disease is outlined as has been observed at the Newcastle General Hospital, Newcastle upon Tyne over three decades between 1942 and 1971.

### MATERIAL

1303 patients with acute rheumatic fever and/or chronic rheumatic heart disease have been seen between 1942 and 1971. When first assessed, 5.5 per cent of the patients had acute rheumatic fever without cardiac involvement, 12 per cent had evidence of acute rheumatic carditis without pre-existent valve lesion, and 82.5 per cent showed already signs of an established valve lesion with or without evidence of active carditis. The long-term observations are based on 1099 patients who have been followed-up for 11033 patient-years, an average period of 10 years. At the final assessment 10 per cent of these patients had no cardiac involvement, and 90 per cent showed evidence of an established lesion of one or more valves, with predominant mitral valve disease in over 90 per cent of the cases.

### CLINICAL OBSERVATIONS

#### Initial Carditis

There has been a gradual decrease in the number of cases of established cardio-valvar lesion which in our experience reflects an important decline in the severity of acute carditis rather than in its incidence. There is indeed evidence that streptococcal virulence has diminished (Table I), and that the severity of acute carditis as judged by heart failure and/or cardiac enlargement and by the extent of residual valve lesion has declined (Table II).

#### Preventive Aspects

239 patients with initial carditis and/or established rheumatic valve disease have had uninterrupted antistreptococcal prophylaxis with oral penicillin over a period of 2800 patient-years, an average period of observation of 11.7 years.

In 91 patients who showed initially acute carditis without pre-existent valve lesion overt rheumatic fever recurred in 3.3 per cent of the cases. Deterioration of the cardiac condition as defined by the development of new murmurs, progressive cardiac enlargement, atrial fibrillation or heart failure took place in 12 per cent of the patients at the rate of 0.9 per cent per patient-year.

In a group of 148 patients who already had established valve lesion recurrence of overt rheumatic fever was observed in 2.7 per cent of the cases. Deterioration of the cardiac condition as judged by the same criteria took place in 38 per cent of the patients at a rate of 3.8 per cent per patient-year.

#### Heart Failure

369 patients (33 per cent) have developed heart failure at various stages during the follow-up period at a rate of 3.3 per cent patient-year.

78 per cent of the episodes of pulmonary oedema occurred in sinus rhythm. On the other hand, 68 per cent of the episodes of right heart failure developed in the presence of atrial fibrillation. Both pulmonary oedema and right heart failure were significantly less frequently observed during the past decade.

#### Atrial Fibrillation

136 patients (12.5 per cent) were already in atrial fibrillation when first seen. 256 patients (23.5 per cent) were in sinus rhythm when first seen and developed atrial fibrillation at a later stage. Many of these reverted to sinus rhythm after direct current shock or drug therapy which had been maintained for a variable length of time. 707 patients (64 per cent) have remained in sinus rhythm throughout the study.

#### Systemic Embolism and Anticoagulant Prophylaxis

131 patients (12 per cent) developed 164 systemic embolic episodes at a rate of 1.5 per cent per patient-year. 72 embolic episodes occurred in sinus rhythm giving an incidence of 0.8 per cent per patient-year. 92 episodes occurred in atrial fibrillation, an incidence of 4.6 per cent per patient-year.

Since the introduction of anticoagulants in 1960, 128 patients have been on continuous anticoagulant prophylaxis for 960 treatment-years. In this group, there were 8 embolic episodes in 8 patients (6 per cent). Thus, the incidence of systemic embolism in patients receiving anticoagulants was 0.9 per cent per patient-year: 0.4 per cent in sinus rhythm and 1 per cent in atrial fibrillation. During the same period of 11 years there have been 91 embolic episodes in a more or less comparable and not anticoagulated group of 366 patients followed for 2690 patient-years. Thus, the incidence of embolism was in this latter group 3.4 per cent per patient-year: 2.3 per cent in sinus rhythm and 8.5 per cent in atrial fibrillation.

#### Cardiac Surgery

280 patients (26.5 per cent) underwent closed mitral valvotomy. 28 patients had a second closed mitral valvotomy and one of these a third valvotomy.

The overall operative mortality rate was 4.3 per cent for first valvotomy and 11 per cent for second valvotomy, but since 1962 there has been no death in a consecutive series of 117 first valvotomies, and 2 deaths in 21 second valvotomies.

Of the 268 survivors of first valvotomies, 108 patients (40 per cent) deteriorated over a period of 2978 patient-years: heart failure occurred in 18 per cent, atrial fibrillation in 32 per cent, systemic embolism in 12 per cent, and late death in 9.5 per cent of the cases.

82 patients underwent open heart surgery, 41 had mitral valve replacement, 3 mitral valve repair, 23 aortic valve replacement, 14 double valve replacement and 1 triple valve replacement.

The overall operative mortality was 25 per cent, but in 1971 there was only 1 death among 20 valve replacement (5 per cent).

The 61 survivors have so far been followed for 185 patient-years. During this period systemic embolism occurred in 5 patients (8 per cent), 4 of whom were on anticoagulant treatment at the time. There were 4 late deaths (6.6 per cent): 2 from heart failure and 2 occurred suddenly outside hospital.

#### The Remote Effects of Pregnancy on the Course of Rheumatic Heart Disease

761 patients were closely followed in 1033 pregnancies. There were 11 cardiac deaths in pregnancy 6 of which were due to pulmonary oedema, and one non-cardiac death. No death occurred in a pregnant patient since 1961.

749 patients of this group have been subsequently followed-up for 8050 patient-years. During this period, heart failure developed in 54 patients (7.2 per cent), atrial fibrillation in 144 patients (21 per cent), and systemic embolism in 74 patients (10 per cent). 84 patients died during the follow-up period (11 per cent).

29 patients underwent closed mitral valvotomy during pregnancy, and 167 patients required cardiac surgery during the follow-up period.

TABLE I  
STREPTOCOCCAL INFECTION AND  
RHEUMATIC RECURRENCE  
NO ANTISTREPTOCOCCAL PROPHYLAXIS

Type of streptococcal infection	50 consecutive streptococcal infections before 1963			50 consecutive streptococcal infections after 1963		
	No.	Overt rheumatic recurrence		No.	Overt rheumatic recurrence	
		No.	Per cent		No.	Per cent
Symptomatic	31	8	26	28	3	10.5
Asymptomatic	19	4	21	22	0	0
TOTAL	50	12	24	50	3	6

TABLE II  
SEVERITY OF RHEUMATIC CARDITIS

Period of study	No. of patients with rheumatic fever	No. of patients with carditis	Cardiac enlargement with or without heart failure	Residual valve lesion
1948-1952	65	40 (62%)	10 (16%)	18 (28%)
1962-1966	75	50 (67%)	7 (9.5%)	9 (12%)
1967-1971	34	20 (59%)	2 (5.9%)	2 (5.9%)

TABLE III  
MORTALITY PATTERNS IN RHEUMATIC HEART DISEASE 1942 - 1971

Period of study	No. of deaths	Causes of death as percentages of all deaths							
		Heart failure	Embolism		At cardiac surgery	Sudden no exact cause established	Various		Unknown
			Systemic	Pulmonary			cardiac	non-cardiac	
1942 - 1951	71	51	8.5	2.8	0.0	11.2	7.0	5.5	14.0
1952 - 1961	64	58	4.8	0.0	20.0	12.2	3.0	6.0	6.0
1962 - 1971	51	24	10.0	0.0	44.0	4.0	6.0	4.0	8.0
TOTAL	186	42	7.5	1.3	19.2	9.60	5.4	5.4	9.6

### Mortality

186 patients (17 per cent) died including those who died at cardiac surgery.

The mortality pattern over three decades is shown in Table III.

### DISCUSSION

There has been a gradual decline in the incidence and especially in the severity of rheumatic fever and rheumatic heart disease during the past decade or so (Massell *et al.*, 1964; Scottish Health Service Council, 1967; Szekely, 1968). Bland reported already in 1960 that the previously common clinical manifestations of severe rheumatic fever such as pancarditis, heart failure and nodules had become only very occasional findings. Also the onset of the disease has been so insidious in recent years that it is very likely that some initial episodes of rheumatic carditis have escaped clinical recognition. Furthermore, it would appear that unlike in the past initial attacks have occurred more frequently after puberty (Biörck, 1955; Hall, 1961; Szekely and Farmer, 1964).

In addition to the natural decline in the severity of initial carditis, long-term antistreptococcal prophylaxis has favourably influenced the course of rheumatic fever. However, in view of certain limitations of secondary prophylaxis, more attention should now be paid to primary rheumatic fever prophylaxis by drugs or antistreptococcal vaccine. The feasibility of the latter procedure has been explored by Massell *et al.* (1969).

Closed mitral valvotomy has significantly improved cardiac function and has reduced morbidity and mortality. However, it is only a therapeutic procedure and progression of the disease continues at a rate peculiar to each patient (Lowther and Turner, 1963; Ellis and Harken, 1964). In our experience, 40 per cent of the patients who had a closed mitral valvotomy deteriorated over an average period of

observation of 11 years. Furthermore, mitral valvotomy did not reduce the incidence of systemic embolism and did not seem to delay the onset of atrial fibrillation. To what extent these observations also apply to valve replacement will become clear only after much longer follow-up studies.

Atrial fibrillation is the commonest significant cardiac dysrhythmia in association with rheumatic heart disease, although it has become less frequent in the past few years. The onset of atrial fibrillation is a medical emergency as it can initiate heart failure and systemic embolism. In many instances sinus rhythm can be restored and maintained with improvement in cardiac function and a lesser risk of potential complications (Szekely *et al.*, 1970).

Systemic embolism remains a serious complication in both medically and surgically treated cases. However, anticoagulant prophylaxis has reduced the incidence of recurrences and we believe also that of initial embolic episodes (Szekely, 1964).

The incidence of the main complications of rheumatic heart disease and the age at their onset were similar in parous women and in nulliparous women or men. We have no evidence that pregnancy influences unfavourably the long-term course of rheumatic heart disease.

The mortality pattern in rheumatic heart disease has changed over the past three decades. Heart failure has become a less frequent immediate cause of death. Cerebral embolism still remains an important cause of mortality, and we believe that the field of anticoagulant prophylaxis should be extended.

### SUMMARY AND CONCLUSIONS

Continuous antistreptococcal prophylaxis can arrest the progression of rheumatic heart disease in a number of cases, if started before the development of haemodynamically significant valve lesion.

Anticoagulant prophylaxis reduces the incidence of systemic embolism.

Cardio-valvar surgery improves cardiac function. However, closed mitral valvotomy does not delay the onset of atrial fibrillation and does not eliminate the danger of systemic embolism.

Pregnancy has no adverse effect on the long-term course of rheumatic heart disease.

The prevalence of rheumatic heart disease is decreasing, but a more striking feature is the decline in its severity.

Morbidity and mortality patterns underwent a considerable change during the past decade.

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