STUDIES IN JUVENILE RHEUMATISM

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The purpose of this review is to summarize the results of four years' special study in juvenile rheumatism among Cardiff school children. It will be convenient to describe our experiences under three headings—(1) the rheumatism clinic, (2) the hospital, and (3) the after-care of rheumatic children.

The rheumatism clinic.

The clinic was inaugurated in 1927 and originally met once a fortnight. As the work expanded, in order to deal promptly with new cases and to keep in touch with all rheumatic children, weekly sessions were held during 1929. Later still, further sessions were found to be necessary, and between 1st January and 30th June, 1931, 42 sessions were held, an average of 7 a month. There was a total of 693 attendances at the rheumatism clinic during this period of

TABLE 1.
INCIDENCE OF RHEUMATIC HEART DISEASE.

Area	Estimated population, 1929	Total cases	Cases per 1,000 of population	Estimated present school population	School cases per 1,000 of present school population
Cardiff	224,200	206	0.91	35,367	5.82
Bath	69,240	15	0.21	7,500	1.28
Bristol	391,320	754	1.90	54,673	7.72
Gloucestershire ..	329,346	89	0.27	53,501	1.03
Somerset	406,560	158	0.39	42,804	2.17
Wiltshire (excluding Swindon)	246,930	73	0.30	33,160	} 1.5
Swindon	62,020	13	0.20	10,238	

six months, 178 being new cases. On 30th June, 1931, there were 601 children remaining on the rheumatism register, a rate of 16.9 per 1,000 of the school population, or rather more than $1\frac{1}{2}$ per cent. Of these, 206 were found to be suffering from heart disease, giving a figure of almost 6 per 1,000 for rheumatic heart disease amongst the present Cardiff school population. It is interesting to compare this figure with the rates of incidence recently given by Savage¹ in the interim report on the Gloucestershire, Somerset and Wiltshire inquiry (Table 1).

Cases are referred to the rheumatism clinic from school inspections and clinics, by private practitioners, public health nurses, teachers and school attendance officers. At each attendance a child receives a complete medical examination, special attention being paid to the heart. Frequency of re-examination in individual cases depends, of course, on the activity of the rheumatic process, but an effort is made to see every rheumatic child at least once in six months. Apart from the discovery of the early signs of rheumatic heart disease with a view to providing suitable hospital treatment, the main function of the clinic is educational. Parents receive instruction on the importance of juvenile rheumatism in the production of heart disease and the special care and management required by rheumatic children. Leaflets setting out the more essential points in this connection are also distributed. In order to be able to devote sufficient time to each case the numbers attending each session are restricted to 18 (5 or 6 being new cases), as we have not found it possible to deal adequately with more than this number at any one session of

TABLE 2.
TYPES OF ONSET.

Origin	Cases ascertained during 1927		Cases ascertained during 1928-31		Total	
	No.	%	No.	%	No.	%
Rheumatic pains	79	37.4	230	55.4	309	49.3
Chorea	43	20.3	124	29.8	167	26.6
Rheumatic fever	55	26.0	46	11.0	101	16.1
Other definite symptoms ..	11	5.2	0	0.0	11	1.7
Insidious	23	10.9	15	3.6	38	6.0
Total	211	—	415	—	626	—

approximately three hours' duration. In the case of children not confined to bed, admission to hospital is always arranged through the clinic. Children who are too ill to attend the clinic are occasionally referred by their medical attendants, and these are seen at home by arrangement with the doctor, and, if considered suitable for admission, are conveyed to hospital by one of the City Council's ambulances.

The type of onset, the season and age at which it developed, and the ultimate condition of the heart in 626 children referred to the clinic during the last four years are set out in Tables 2-5. The findings concerning some 200 children included in this number and previously reported on² are given separately in Tables 2 and 5 for comparative purposes.

Types of onset.—The decline during recent years in the proportion of cases commencing with rheumatic fever which has been commented on by Gray Hill³ and the relative increase in the milder types of onset are shown in Table 2. The significance of this tendency will be referred to later. It is interesting to find that 49 per cent. of 626 first attacks of the disease in Cardiff school children took the form of subacute articular rheumatism, as compared with 48 per cent. of 1,700 first attacks in London children⁴.

Seasonal incidence.—It will be seen in Table 3 that the majority of cases (66·3 per cent.) were found to commence during the cold wet months of winter and spring. If the cases beginning with chorea are excluded this tendency is accentuated. When the seasonal onset of chorea alone is investigated it is found that there is a greater likelihood of this manifestation occurring at the end of a school term, particularly in adolescent girls.

TABLE 3.
SEASON OF ONSET.

	No.	%
Spring	125	22·7
Summer	107	19·4
Autumn	78	14·1
Winter	240	43·6
	550	—
Uncertain	76	—
Total	626	—

Age of onset.—It will be noted in Table 4 that in the great majority of instances in school children (73·7 per cent.) juvenile rheumatism commences between the ages of 5 and 10 years. When these figures are plotted out, the resultant curve closely approximates to that obtained for 1,700 rheumatic children recently reported on⁴ by the School Medical Officer to the London County Council. The disproportionately low figure for cases beginning at the age of 14 years is, of course, artificial, being due to the intervention of the school-leaving age.

TABLE 4.
AGE OF ONSET.

Age in years	No. of cases
2	4
3	14
4	36
5	70
6	67
7	72
8	84
9	75
10	65
11	42
12	38
13	20
14	1
	588
Uncertain	38
Total	626

Incidence of heart disease.—The first column in Table 5 shows the condition of the heart in 214 rheumatic children ascertained before any special provision was made in Cardiff for rheumatic children. The second column shows the present condition of the heart in 412 children who had been dealt with during the three years the rheumatism supervisory scheme has been in operation.

Although the working of the clinic during the last three years has led to a fuller ascertainment of the earlier and milder forms of the disease in which cardiac involvement is less commonly encountered, it is fair to conclude that the lessened incidence of permanent heart disease amongst rheumatic children ascertained subsequent to 1927 is, in some measure, due to the establishment of the scheme which ensures the regular medical supervision of all rheumatic children and provides a special hospital for the treatment of early cases of carditis.

TABLE 5.
INCIDENCE OF HEART DISEASE.

	Cases ascertained during 1927		Cases ascertained during 1928-31		Total	
	No.	%	No.	%	No.	%
Normal	41	19.1	196	47.5	237	37.8
Cardiac abnormalities not amounting to permanent disease	44	20.5	120	9.1	164	26.2
Mitral regurgitation ..	101	60.2	69	23.3	170	35.9
Mitral stenosis	23		22		45	
Aortic regurgitation ..	3		2		5	
Aortic stenosis	0		1		1	
Aortic and mitral regurgitation	2		1		3	
Mitral regurgitation and pulmonary stenosis ..	0		1		1	
	214	—	412	—	626	—

Social status.—In order to assess the significance, if any, of general environmental conditions in the ætiology of juvenile rheumatism, an attempt was made to grade 600 rheumatic children from the social standpoint, the following arbitrary standards being used :—

Grade A. Children from well-to-do homes.

Grade B. Those whose parents have a struggle to make ends meet, but who were not suffering from the effects of privation.

Grade C. Those who are destitute, semidestitute, or in receipt of public or charitable assistance.

The numbers in each group are shown in Table 6.

That poverty or destitution is not a potent factor in the causation of juvenile rheumatism is suggested by the fact that almost 75 per cent. of the children examined did not come within this category.

Damp.—The significance from the ætiological standpoint of damp in houses was also further investigated. Amongst 600 rheumatic children the houses were stated to be free of dampness in 61·1 per cent. of cases. In a control group of non-rheumatic children investigated in 1927 the percentage of houses not damp was 62·2 per cent. Clearly this factor is of little moment in the causation of juvenile rheumatism.

Family incidence.—The incidence of juvenile rheumatism in other members of the same family was also recorded. The result was rather surprising. In 125 out of 279 separate family histories investigated, one or more parents of rheumatic children had themselves suffered during childhood from some form of acute rheumatism—a percentage of 44·7. In all, 138 parents were affected. In investigating this factor, histories of rheumatic pains contracted in adult life as the result of war service, strain, or exposure of any kind were disregarded. Further, 385 rheumatic children were found to have 122 brothers or sisters affected with juvenile rheumatism. It would seem, then, that family relationships play a bigger part in the causation of juvenile rheumatism than has hitherto been realized.

TABLE 6.
SOCIAL GRADE.

Social grade	No.	%
A	113	18·8
B	336	56·0
C	151	25·1
Total	600	—

Infectivity.—It was not possible to arrive at any definite conclusion with regard to the infectivity of juvenile rheumatism. The theory of the infective origin of this disease is attractive and has much to recommend it. The marked seasonal prevalence, the predominance of cases during the latter half of the first decade of life, and the tendency for other cases to occur in a family where one child is already affected, are suggestive features in this connection; but in the light of the facts ascertained from a scrutiny of over 600 case histories it must be admitted that, if the disease is communicable, its infectivity is of a low grade when compared with the exanthemata. If juvenile rheumatism is an infectious disease it would appear to be akin epidemiologically to the virus infections of the central nervous system, such as poliomyelitis and encephalitis lethargica. During the last four years many striking instances of multiple cases of juvenile rheumatism occurring contemporaneously in the same house have come to light, but on the other hand multiple cases have failed to occur in hundreds of other cases living under similar home conditions. The only conclusion that seems warrantable on the present evidence is that, while case-to-case infection in juvenile rheumatism is possible, it is neither the usual nor even a common mode of origin. The possibility of carrier spread and of

TABLE 7.
COLOUR OF HAIR AND EYES.

Hair	No.	%	Eyes	No.	%
Fair	111	34.6	Blue	145	45.1
Medium fair	107	33.3	Grey	49	15.2
Red	16	5.0	Hazel	20	6.2
Medium dark	39	12.1	Brown	107	33.3
Dark	48	14.9			
Total	321	—	Total	321	—

infection by sub-clinical types cannot, however, be ignored, and may be worthy of investigation when the bacteriological relationships of the disease are more clearly defined.

Influence of scarlatina.—The influence of scarlet fever in predisposing to juvenile rheumatism has received careful attention. Amongst 626 rheumatic children, 95 were found to have suffered from scarlet fever, and of these only 8 developed it after the first rheumatic manifestation. In 37 cases (almost 6 per cent. of the total rheumatic children examined) the onset of juvenile rheumatism was directly post-scarlatinal. Of these 11 were found to have permanent valvular disease and most of these had occurred before special hospital provision was made locally for rheumatic children. Of the 26 post-scarlatinal cases ascertained between 1928—1931 only three developed permanent heart disease. Our experience has been that when rheumatism develops after an attack of scarlet fever it is less likely to be followed by grave cardiac sequelæ than when the disease develops independently.

Colouring.—The colour of hair and eyes was recorded in 321 rheumatic children, and the results show that about two-thirds of the children were of the fair type and had blue or grey eyes. The figures are given in Table 7.

TABLE 8.
INCIDENCE OF DENTAL CARIES.

	Rheumatic group		Non-rheumatic group	
	No.	%	No.	%
Sound teeth	135	48.5	122	40.6
1 carious	40	14.3	36	12.0
2 „	48	17.2	45	15.0
3 „	28	10.0	37	12.3
4 „	13	4.7	31	10.3
5 „	5	1.8	7	2.3
Over 5 carious	9	3.2	22	7.3
Total	278	—	300	—

No distinctive type of complexion or facies was recognized, apart from this definite preponderance of fair children with blue or grey eyes. Chorea seemed to be the rheumatic manifestation exhibited by many of the dark-haired children.

Dental caries.—The incidence of dental caries amongst 278 rheumatic children was also investigated, and the findings as compared with those in a group of non-rheumatic children are set out in Table 8.

It will be seen that the incidence of carious teeth was slightly higher in the non-rheumatic group. It appears that dental caries may safely be discounted as a factor of ætiological significance in juvenile rheumatism.

The hospital.

That some special form of institutional provision for rheumatic children was required in Cardiff has for years been apparent from the records of the School Medical Service. A special survey of Cardiff rheumatic children carried out in 1927² showed that in spite of the facilities then existing in general hospitals and convalescent homes, 129 children were suffering from permanent heart disease, the consequence of untreated or insufficiently treated juvenile rheumatism. How unsatisfactory the domiciliary treatment of juvenile rheumatism can be even in the homes of the well-to-do, is well known by all who have to deal with this disease in clinics and out-patient departments. The immediate needs of a child suffering from any form of acute or subacute rheumatism may be summed up in a few words—physical and mental rest until the disease subsides. Even if rest in bed can be ensured in a working-class home, it is at best a make-shift. The mother, busy with household tasks and the care of the other children, is frequently unable to provide the amount of supervision and care required by the ailing rheumatic child. In the days before the City Council had provided special hospital accommodation for these cases, it was not unusual for children to continue attending the clinic with active rheumatism for nine and twelve months at a stretch, and in these circumstances it will be realized that eventual cardiac involvement was the rule. Since the Lord Pontypridd Hospital was opened on 8th April, 1929, conditions have completely changed, for it is found that the most intractable of rheumatic manifestations speedily respond to a comparatively short period of in-patient treatment. Our aim in selecting cases for hospital has been to admit only those who are in a sufficiently early stage of heart disease to respond to treatment. Any rheumatic child presenting the signs of early carditis as below described is considered a suitable case for admission. In addition, all acute forms of juvenile rheumatism (rheumatic fever, chorea, and severe rheumatic pains), even though the heart may appear normal on clinical examination, are regarded as potential cases of heart disease and so qualify for hospital treatment. In subacute cases a period of observation at the clinic is sometimes necessary before the question of hospital treatment can be decided. It occasionally happens that a child has a permanent valvular condition present on admission to hospital. These are usually cases suffering from acute manifestations (e.g., chorea, arthritis or severe pains) and the determining factor in their

admission has often been the knowledge that the patient cannot secure in the unsatisfactory environment of his own home the rest and care required to carry him safely through an acute attack. Again, a child found to have early carditis when seen at the clinic may develop the signs of valvular disease while awaiting admission to hospital. This is more prone to happen in the winter and spring when the demand on the hospital accommodation is heaviest. Starting with 16 beds, the accommodation has been increased to 25, but the experience gained during two winters indicates that even this is insufficient for our present needs. In a city like Cardiff it is probable that one bed for every 1,000 of school population (approximately 35 beds) would not be an excessive provision for this type of case.

In the period April 8th, 1929, to July 31st, 1931, there were 249 admissions to hospital. Of these, 10 children were transferred to other Corporation hospitals (9 cases of communicable disease and one case of the post-encephalitic syndrome which had been admitted in error as chorea). 17 children were for various reasons withdrawn by their parents after short periods, leaving 222 cases treated to a conclusion in the wards for one or other form of juvenile rheumatism. All children admitted were of school age. The form of rheumatism present on admission to hospital in these cases was as follows :

Chorea	118 cases
Rheumatic pains	111 ..
Rheumatic fever	8 ..
Early carditis without definite symptoms	12 ..
Total →	249 cases

One child died in hospital after almost five months' hospital treatment. She was a girl aged 11 who was admitted with severe chorea of three months' standing and an active carditis causing mitral regurgitation. She remained completely uninfluenced by all treatment while in hospital and had two attacks of cerebral embolism, the second causing a left-sided hemiplegia. The clinical course of this case corresponded closely to the variety of sub-infective endocarditis described by Horder and others under the title of endocarditis lenta. One other advanced case of mitral stenosis and acute chorea, admitted after tonsillectomy at the urgent request of the parents, developed rapid failure of compensation and died three days after transfer to a general hospital. The remaining 220 cases were discharged free of active disease.

In the great majority of cases some abnormality of the heart was detectable on admission to hospital. In more than half of the cases this took the form of early carditis, but reference to Table 9 shows that no less than 47 of the 222 cases treated to a conclusion were thought to be suffering from permanent valvular disease on admission. Although cases with established heart disease are not, generally speaking, considered suitable for this type of institution, their admission has in several instances unexpectedly justified itself, for with prolonged rest and treatment definite murmurs have disappeared and the heart has returned to normal functional capacity. Even with increasing experience it is not always possible to decide, on clinical examination, whether

certain lesions of the heart, of comparatively recent origin, are likely to respond to treatment or not. In these cases the practice is to give the child the benefit of the doubt, and accordingly seven children, classified on admission as having established valvular conditions, completely lost all signs of heart disease after a prolonged period of in-patient treatment, while four others were regarded on discharge as not suffering from valvular disease, although the heart sounds had not quite regained their normal clearness.

Cardiac complications.—The most constant physical sign of early rheumatic carditis in our experience has been enlargement of the area of cardiac dullness, coupled with an alteration in the character of the first sound of the heart. The alteration in the great majority of cases takes the forms of blurring or a want of clearness, and this is usually accompanied by an accentuation of the second sound. Sometimes the first sound is definitely roughened and prolonged, or again, the muscular element in it may be lacking, rendering it valvular in character. In extreme cases the first sound may be entirely inaudible, and at a later stage in the disease a soft systolic murmur localized to the mitral area makes its appearance. It is probable that all these signs express myocardial rather than endocardial mischief, and with the exception of the absence of the first sound and the localized murmur it is the rule for these abnormalities to disappear with prolonged rest in bed. If the condition is ignored, however, the development of permanent valvular disease is almost invariable. In the case of localized murmurs or absence of the first sound, complete recovery is not impossible, but a more usual result is some degree of hypertrophy of the cardiac musculature with a slighter degree of impurity in the character of the sounds. When endocardial damage sufficient to evoke the classical signs of disease exists, it is, of course, the exception for the heart to return to normal or for any improvement other than increased hypertrophy of the left ventricle to take place, but the existence of a systolic murmur at the mitral area which is conducted around to the interscapular region is not conclusive proof that the valvular defect is permanent. A very few of these cases do respond to treatment, and if it is known with certainty that such a conducted murmur is of recent origin it is always worth while trying the effect of rest in hospital before deciding that the physical signs connote irretrievable damage to the valve tissues. A change in the quality of the sounds of the heart, or in the adventitious sounds accompanying or replacing them, does not readily occur when the condition is one of long standing and the disease is quiescent, but the clearness of the sounds and the character of the bruits expressing early carditis do change within relatively short periods. With these changes detectable on auscultation there will be present the other invariable sign of active carditis, namely, an alteration in the size of the area of cardiac dullness. Another constant sign of active carditis is a change in the rate of the pulse. In assessing the activity of rheumatic disease of the heart, pulse, rather than temperature readings, should receive primary consideration. In addition to the usual twice daily record, which should preferably be graphed, it is desirable, in the case of children, to chart the rate of the sleeping pulse, and if a heart, not the seat of an established valvular condition, gives readings persistently

over 90 during sleep, the case should be very closely watched for other signs of rheumatic carditis. Persistent frequency of the pulse may be the earliest sign of active carditis. It is often the only sign for perhaps several weeks on end. A fluctuating pulse, especially with evening readings reaching 100, is another suspicious circumstance, but in emotional children and in the absence of a frequent sleeping pulse, too much importance should not be attached to this. Irregularity in rhythm may or may not connote active carditis. It is much more frequently found in established valvular disease. It is a safe rule to insist on absolute recumbency, even during meals, wherever the sleeping pulse of a rheumatic child who has not an established valvular leak exceeds 90 on three consecutive evenings. It cannot be too strongly emphasized that hearts can become progressively damaged from rheumatism without any appreciable pyrexia, and, indeed, without any obvious constitutional disturbance. Thus a scrutiny of the charts of 216 consecutive cases showed that the pulse readings alone were elevated in 92, the temperature alone in 24, and both in 37.

One of the most unexpected of our experiences has been the comparative rareness of long continued pyrexia in the acute and sub-acute cases of the disease treated in the wards. It is, of course, common for the temperature to be elevated when a joint is swollen, but defervescence is generally very speedy, a matter of a few days at most. No doubt the complete immobilization of patients which the hospital régime prescribes helps to produce this effect, but it is difficult to avoid the conclusion that juvenile rheumatism is to-day a much less acute disease than it was formerly. It should, however, be pointed out that this milder symptomatology of present-day rheumatism is not in any way analogous with the mildness of modern scarlet fever and smallpox, for cardiac involvement is an integral part of juvenile rheumatism and there is no evidence that the prevalent insidious forms of the disease are less likely to lead to heart mischief. Indeed, the converse may be true, for with the less severe manifestations now encountered there is less disposition on the part of parents to seek medical advice at a sufficiently early stage of the disease to render preventive treatment practicable.

The aim in treatment is to safeguard the heart during the acute and sub-acute phases of the disease, and, so far as is possible, by the provision of rest and careful management, to prevent the development of permanent cardiac damage. Broadly speaking, this object is attained by the insistence on absolute rest in bed during the acuter stages, followed by a very gradual return to normal activities when the disease has become quiescent. It is essential to keep the child completely at rest during the active phases of rheumatism. During the first fortnight after admission to hospital the child is closely observed and questioned for signs and symptoms. He is kept lying flat in bed and is allowed to do nothing for himself, and is fed by a nurse. To ensure freedom from emotional upsets visits from relatives are not allowed during the first week, at the end of which time the child has usually grown accustomed to the ritual and the personnel of the hospital. At the end of the fortnight the extent and degree of activity will have been gauged. As long as there are any signs of

active disease the patient is kept lying flat and usually for the whole of this time he is not allowed to feed himself. The subsequent management of the cases depends largely on the development of signs and symptoms. In a case speedily responding to rest the patient may be allowed to sit up in bed for 20 minutes each day a week or so after the commencement of self-feeding. The effect of each concession on the pulse rate, the quality of the heart sounds and the area of cardiac dullness is carefully noted, and if no ill effects follow, increments of 20 minutes are added to the sitting-up period every two or three days until the patient is sitting up for three hours each day. About a week later he is allowed to get up and sit about the ward, and subsequently proceeds to walk about the grounds, first on the flat and later without restrictions. This is an account of the straightforward case pursuing an uneventful course towards recovery, but this régime is liable to interruptions. Elevation or fluctuations in the pulse or temperature readings, alteration in the character of the heart sounds or in the area of cardiac dullness, return of twitchings or pains, increase in pallor, the development of nose bleeding, or the appearance of purpuric patches or urticarial rashes, e.g., erythema marginatum, may determine a withholding of further concessions or a return to complete recumbency. Persistent frequency or fluctuations of the pulse rate is always taken as an indication for complete immobilization in bed. Sometimes in spite of this treatment the heart condition steadily progresses until a systolic murmur makes its appearance. It is surprising how satisfactory may be the general condition of the child while this change is taking place. There may be a complete absence of pains or movements, but usually the colour is poor and there is, in addition, the instability of the pulse rate and possibly of the temperature. If a murmur does evolve, it is, at first, audible only at the mitral area, but in the course of a few weeks it becomes more distinct locally and is now heard in the axilla and still later in the inter-scapular area. Enlargement in the area of cardiac dullness accompanies the development of these physical signs, but the process of progressive cardiac involvement may stop at any intermediate stage, leaving most commonly a murmur localized to the mitral area which persists after the disease has become quiescent. Sometimes this local bruit, after remaining stationary for many weeks, may clear up and the heart may be again free of murmurs. More usually in this type of case some softening of the first sound is left together with a degree of cardiac enlargement. In the earlier phases of rheumatic carditis signs of pericardial involvement have been constantly looked for, but so far have not been encountered. Precordial pains are, however, not uncommon in this stage and may express a slight degree of pericardial inflammation. As shown by Table 9, the mitral valve has, in the great majority of cases, been the seat of endocardial involvement.

In the treatment of these rheumatic conditions little use is made of drugs. Salicylates are used for the relief of pain chiefly. Aspirin has been tried with the hope of cutting short relapses, but no striking results have yet been obtained. Precipitated calcium in lactic acid has been given to the severer cases of chorea with a preparation containing the Vitamins A and D, but the cases so treated

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did not appear to lose their movements any more rapidly than those who were treated merely by rest. Iron tonics are used regularly for combating the anæmia associated with the disease, and various proprietary preparations containing cod-liver oil are given to children with subnormal nutrition. No modification in the diet of these children is considered necessary. An ample and nourishing diet is given in all cases, solids being withheld only when the temperature exceeds 100° F.

The value of open-air in the treatment of juvenile rheumatism has been amply demonstrated since the inception of this work. During the summer months as many as possible of the beds are wheeled out on the lawn, and on fine days the children lie exposed to the sunlight wearing a minimum of clothing. For the whole of the summer two shelters in the grounds are utilized

TABLE 9.
EFFECT OF HOSPITAL TREATMENT ON CONDITION OF HEART.

Condition of the heart					On admission		On discharge	
					No.	%	No.	%
Myocardial conditions					116	52.2	30	13.5
Blurred first sound					100		29	
Valvular first sound					14		0	
Absent first sound					2		1	
Endocardial conditions					77	34.6	57	25.6
Localized murmur					30		19	
Mitral regurgitation					35		22	
Aortic regurgitation					0		1	
Mitral and aortic regurgitation					1		1	
Mitral stenosis					10		13	
Mitral regurgitation and pulmonary stenosis ..					1		1	
Normal hearts					29	13.0	135	60.8
Total					222	—	222	—

as open-air wards and accommodate six children with a nurse who sleeps there at night. The children in the shelters improved much more rapidly in general health, became deeply pigmented and seemed to lose their rheumatic manifestations more quickly than those sleeping indoors. It was observed that the children who had been treated in the open-air shelters seemed less liable to relapses after discharge and the pigmentation of the skin acquired by these children persisted for many months after they had left hospital. It is to be regretted that the construction of the hospital (which is an adapted private residence) does not permit of the application of open-air treatment on a much wider scale.

The results obtained from hospital treatment so far as the heart was concerned in the first 222 cases are set out in Table 9.

It will be observed that 116 definitely myocardial conditions on admission were reduced to 30, that 77 cases regarded as endocardial on admission were reduced to 57 on discharge, and that the figure for normal hearts (29) on admission was raised to 135 on discharge. Up to the present the great majority of the cases treated in hospital are remaining well. In dealing with a disease like rheumatism relapses must be expected, and it is this tendency which makes the periodical medical examination of rheumatic children so essential a feature of any scheme attempting to deal with the problem. So far the indications are that the children discharged from hospital continue to keep well, recurrences being milder in character and more fleeting in duration than in the case of children who have not undergone in-patient treatment. The educational value of the hospital treatment undoubtedly plays a large part in producing this effect, as it has been noted again and again that even quite young children absorb sufficient of the spirit of the hospital régime to render subsequent domiciliary treatment a very much simpler matter from the parents' point of view.

Complications other than cardiac.—Having regard to the speedy subsidence of acute symptoms which complete immobilization effects, it is not surprising that complications other than cardiac arising during hospital treatment should be very few. Amongst 222 cases of acute and sub-acute rheumatism, whose combined treatment in hospital exceeded 15,000 days, the following complications were observed :—

Pharyngitis and tonsillitis	..	..	..	..	24
Periodontal abscess	..	..	..	..	2
Otitis and otorrhœa	..	..	..	..	4
Urticaria	..	..	..	..	5
Purpura	..	..	..	..	4
Herpes zoster	..	..	..	..	3
Rhinitis	..	..	..	..	3
Conjunctivitis	..	..	..	..	3
Epistaxis	..	..	..	..	6
Bronchitis	..	..	..	..	1

The occurrence of throat inflammations and their effect on the rheumatic process has been closely watched. During treatment 19 children developed a total of 24 attacks of either tonsillitis or pharyngitis. In 10 instances the attack of tonsillitis or pharyngitis preceded a relapse by periods varying from one to four weeks. In the remaining cases the throat inflammation seemed to produce no effect. It was noticed that in 12 other cases who did not complain of sore throat, but who had sudden rises in temperature lasting for 24 hours, relapses followed within a similar period, generally two or three weeks after the elevation in temperature. The relapses in all cases involved the heart and were never choreic in nature. The significance of these sharp attacks followed by an intermediate period of quiescence and then a relapse may be, as suggested by the writings of Schlesinger⁵, Sheldon⁶ and Collis⁷, the presence in the throat during the period of initial fever of the hæmolytic streptococcus which leads to an allergic response in tissues sensitized by previous infection. No bacteriological investigations have been so far carried out in Cardiff in this connection.

It was observed amongst the children in hospital that the tendency towards an attack of tonsillitis seemed to be quite independent of the state or size of the tonsils, and in three-fourths of the children developing attacks while under treatment the tonsils were either normal or had been completely removed by previous tonsillectomy. The results obtained in 1927, when a group of rheumatic children was compared as to the state of their tonsils with a control group of non-rheumatic children, have been confirmed by subsequent observation (see Table 10).

TABLE 10.
STATE OF TONSILS.

Tonsils	Rheumatic children		1927 controls	
	No.	%	No.	%
Normal	276	46.9	54	46.3
Slightly enlarged	131	22.2	31	26.7
Moderately enlarged	68	11.5	13	11.2
Enlarged	45	7.6	10	8.6
Removed	68	11.5	8	6.9
Total	588	—	116	—

Allowing for the disproportion in the size of the two groups, it is obvious that there is no definite relationship between the state of the tonsils and the development of juvenile rheumatism.

Mere frequency of sore throats seems also to possess no ætiological significance (see Table 11).

TABLE 11.
INCIDENCE OF SORE THROATS.

Sore throat	Rheumatic children		Non-rheumatic controls (1927)	
	No.	%	No.	%
Never	407	65.8	82	70.6
Very rare	37	5.9	6	5.0
Occasional	98	15.8	15	12.8
Frequent	76	12.2	13	11.2
Total	618	—	116	—

It is possible that the importance of throat conditions in this disease may be found to depend more upon the bacteriological findings during pyrexial periods than upon alterations in the size, or frequency of inflammation, of the tonsils.

The experience of the last three years has also confirmed the view expressed in 1928* that tonsillectomy does not prevent the onset of juvenile rheumatism, for of 407 rheumatic children examined since then, 51 had had their tonsils

removed and of these no less than 39 (76 per cent.) were operated upon before the first sign of rheumatism manifested itself. Having regard to the frequency with which an acute attack of chorea is precipitated by tonsillectomy and the questionable influence of the operation in the prevention of other forms of rheumatism, it is clear that the indiscriminate removal of tonsils in rheumatic children should be condemned. Striking instances of how an injudicious tonsillectomy can light up a quiescent cardiac condition with dire results have been given by Coburn* in his treatise on the rheumatic state recently published.

The after-care of rheumatic children.

Children discharged from hospital rest quietly at home for one month and are then re-examined at the clinic. In the majority of cases they are then found fit to return to school, but in some cases a further period of rest at home is necessary before return to school. With some children it may be necessary even to resume school more gradually, the child attending the afternoon session only, and resting in bed in the mornings. As a general rule children who have had acute attacks do not return to school for at least three months after the onset, and, on an average, two months of this period is spent in hospital. There are, of course, instances which require much longer periods of rest than this, and a few children with insidious forms of carditis have been in hospital for periods of six or eight months at a time. Owing to the pressure on the accommodation at the hospital, cases have undoubtedly to be sent home too soon after getting up and, although in the majority of cases the mothers can be trusted to supervise the completion of convalescence, there have been instances where children have relapsed after longer or shorter intervals of home management. Out of the total admissions up to July 31, 1931, 18 were re-admissions, 16 children undergoing two periods of hospital treatment and one child three. The latter had two attacks of severe chorea and one of rheumatic fever in eighteen months, but was finally discharged with a normal heart. In most of the cases re-admitted the home conditions had been unfavourable and the parental management unsatisfactory.

Generally speaking, the cases which have passed through the hospital have remained well and any recurrences occurring have been slight and of brief duration. Co-operation with the school attendance department has now been developed to such an extent that rheumatic children not excluded from school are readily excused from attending on particularly wet days, especially if they would be obliged to walk a long distance in order to do so. The willing co-operation of head teachers and school attendance officers in making special arrangements in respect of the attendance of rheumatic school children is deserving of mention.

An experiment was begun at Easter, 1930, of sending to an open-air school a certain number of rheumatic children, who, although free of rheumatic manifestations when discharged from hospital, were still of subnormal physique. The children selected were those who had escaped the graver cardiac sequelæ and were of the type who seemed likely to benefit from the open-air conditions

and the regular and balanced diet available at the open-air school. It need hardly be mentioned that the unmodified school régime cannot be followed by the rheumatic child, but with adequate precautions as to exposure to the weather and restriction of exercise and recreation it is clear that selected rheumatic children can make considerable progress at an open-air school. The factor of continuous medical supervision which is available at this type of school has special advantages in the case of the rheumatic child, for recurrences can be detected at an early stage and appropriate advice and treatment prescribed.

While there can be no question as to the value of special hospital treatment in bringing about a speedy subsidence of the acute or sub-acute manifestations of juvenile rheumatism, it is, of course, far too early yet to express any opinion as to its effect in lessening the incidence of heart disease amongst the general population. All that can be said at present is that the hospital treatment of the earlier phases of rheumatic carditis, coupled with the regular medical supervision of rheumatic children, gives results which are encouraging enough to hold out the hope that a disease which has been estimated by some to be responsible for 25,000 deaths every year in England and Wales may yet prove to be largely, if not entirely, preventible.

Summary.

(1) A four years' survey of juvenile rheumatism in Cardiff showed that 16.9 per 1,000 of the school population are affected, of whom 5.82 were suffering from rheumatic heart disease.

(2) Further evidence of the relative decrease in the prevalence of rheumatic fever at the expense of the milder and more insidious forms of juvenile rheumatism was obtained.

(3) Among 550 case histories investigated, the onset of the disease occurred during the winter or spring months in 475 (66.3 per cent.).

(4) Among 588 rheumatic children 433 (73.7 per cent.) developed the first rheumatic manifestation between the age of 5 and 10 years.

(5) Among 626 rheumatic children examined 225 (35.9 per cent.) were found to be suffering from permanent disease of the heart.

(6) An investigation of the social conditions under which 600 rheumatic children lived showed that almost 75 per cent. of the children were not suffering from the effects of poverty or privation.

(7) Further support for the view that dampness of houses is a factor of no aetiological significance in juvenile rheumatism was obtained.

(8) The parents of rheumatic children were found to have themselves suffered from juvenile rheumatism in 44.7 per cent. of 279 separate family histories investigated, and 385 rheumatic children were found to have 122 brothers or sisters affected with the disease.

(9) Although family relationships play some part in the causation of the disease no conclusive evidence as to the infectivity of juvenile rheumatism came to light.

(10) In 6 per cent. of 626 cases the onset of the disease was directly post-scarlatinal but the post-scarlatinal cases were found to be less likely to develop cardiac sequelæ than those developing the disease independent of scarlet fever.

(11) No distinctive type of rheumatic child was recognized, but a majority of the children were fair-haired and had either blue or grey eyes.

(12) The incidence of dental caries was found to be no higher in rheumatic than in non-rheumatic children.

(13) A clinical description of the earlier phases of rheumatic carditis is given, and the methods and results of hospital treatment in 222 cases outlined.

(14) The significance of throat conditions in juvenile rheumatism is discussed. Figures are given to support the contention that the state of the tonsils is of little importance in the causation of this disease.

(15) Further evidence is adduced as to the limitations of tonsillectomy in the treatment of juvenile rheumatism.

(16) The effect of open-air treatment in the hospital treatment and after-care of rheumatic children is described.

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