

## Organisation and the Rheumatic Child

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RHEUMATISM in childhood has been known since the early nineteenth century, but it is only within recent years that the numerical extent of the disease has been ascertained and its significance appreciated. The position which now confronts us can be summarised as follows: From the statistics of 1920, 500,000 of the school-children of the country are estimated to suffer from rheumatism, 10,000 of the children of school age in London are known to be rheumatic, and the vital statistics of 1924 showed that the deaths from heart disease exceeded those from either tuberculosis or cancer.

This incidence of disease in the community has two consequences. The first are the recurring periods of ill-health and loss of education to the child. This is a feature difficult to realise and it is something of a shock to read in a recent report (1) that a quarter of the chronic ill-health of school-children of the country is estimated to be due to rheumatism. The second effect is the adult carditis resulting from the disease. The extent of this is indicated by the estimation that 60 per cent of rheumatic children ultimately develop heart disease. These facts represent one of the many baffling problems confronting medicine today. It is a problem, however, which is fortunately attracting considerable attention, and the past year has seen the publication of two authoritative reports concerning it and one containing suggestions with regard to its prevention. It is the purpose of this paper to attempt to summarise the present position as regards our knowledge of the disease, and to examine the grounds upon which hope of control and of understanding may fairly be based.

(1) Annual Report of the London County Council for 1925, Public Health Pub.

### Three Main Facts.

If the disease be considered as a whole, three main facts become evident. The first is a marked lack of definition in the clinical outline. Only a short time ago, acute and subacute rheumatic fever, chorea, tonsillitis, and carditis were held to cover the manifestations of rheumatism in childhood. As these are well-defined conditions, the features of the disease appeared to be clear and easily recognisable. But within recent

years, it has been argued that many forms of debility, anaemia, vague muscular pains, dyspepsias, and general irritability should also be considered as of rheumatic origin. Furthermore, there is a good deal of evidence which goes to show that carditis can result equally from these as from the more definite forms of the disease, although this is a point upon which unequivocal proof is still needed.

The, second fact is that the greater significance of rheumatism in childhood does not lie in the typical manifestations to which the name is given, and from which the child can be seen to be suffering, but in a single feature, of these—i.e., heart disease. This, in any but its later stages, is not to be detected by the general public. Yet, as is the case with all diseases of children, it is on parents as part of the public that we must rely for the making of the contact between patient and physician. The peculiar situation, therefore, arises that the significance of the disease is in a region inaccessible to the observation of patient or mother, and without relation to the severity of the symptoms appreciable by either.

Thirdly, we have no certain knowledge of the actual cause and nature of the disease. Two views are current. By the early observers, rheumatism, or more strictly, those manifestations of it that had been identified, was looked upon as evidence of a particular constitution of the body. Somewhat later, a specific infection seemed the more probable explanation. It is on this hypothesis, put forward and developed by F. J. Poynton and A. Paine, (2) that nearly all modern conceptions have been based.

(2) *Researches in Rheumatism*, London, 1913.

#### Infection or Diatheses?

To the practical aspect of the question the choice of hypothesis is crucial. If it be granted that a given disease be the result, or the result mainly, of a specific micro-organism, then effective measures against the disease must wait upon the successful isolation of the organism, or the practical application of immunological processes yet to be discovered. In the period before this has occurred the lines of work most likely to be fruitful will be those directed towards the modification of conditions within and around the child that favour infection with this organism. If, on the other hand, the diathesis theory be assumed, other lines of approach to the problem come into view.

In the disease under review during the past two years certain evidence has been adduced to suggest the value once more of considering the constitutional or diathesis (3) hypothesis. According to this view, the outbreak of the disease would depend rather upon constitutional modifications of the bodily habit than an "infection" with a specific organism.

The importance of this point cannot be exaggerated. If the constitutional view be conceded, the emphasis of study, and ultimately of treatment, is thereby shifted. The dependence of treatment upon identification of the organism becomes greatly diminished, and the inquiry shifts to points concerning the endocrine and biochemical make-up of the individual. For the designing of work either with regard to the study of the disease or to the devising of means for its control, the choice of hypothesis is of fundamental importance.

(3) Llewellyn J. Llewellyn: Proc. Roy. Soc., Med. Jan. 1926. P.1. 4 Brit. Med. Jour., May 7th, 1927.

### Bearing on the Study of the Disease

Hitherto, all inquiry into the nature of the disease has been conducted by separate investigations specifically designed and carried out alongside of the ordinary clinical work. That is to say, while nearly all workers in this field have been keenly interested in the problems of the disease and have contributed much of great value to our knowledge of its clinical course and manifestations, when a specific aspect has come to be investigated, a separate effort has been needed to examine and report on the material available. Two examples of this kind have recently been provided by the reports of the B.M.A. and M.R.U. Committees. Just so long as the infective theory of the disease exclusively dominates our conception of it this system is inevitable. Bacteriological and statistical work have no bearing upon the treatment of the individual patient at the time concerned until their goal has been reached.

If, however, the alternative hypothesis be taken for examination, a different type of attack is possible. Here the nature of the disease is conceived as within the constitution of the patient himself, as has been brought out recently by Hurst. (4) According to this view, the whole individual is the subject of inquiry and this inquiry can only be carried out by observation of the individual, whereas by an infection theory the subject of inquiry has to be isolated from material supplied by the patient. It is therefore necessary, if this hypothesis be held, to devise methods of study which shall concern themselves with the life—history of the patient, his biochemical and endocrine processes, his response to specific rheumatic infections, and to circumstances of his daily life. Considered from this point of view, no attempt has yet been made to carry out exhaustive research, although, as will be seen later, some tentative beginnings are already in being. The obstacle to all work of this kind is the nature of the contact at present existing between child and physician. It has been abundantly proved that acute rheumatism shows its main prevalence among children of the hospital classes. The contact of the profession with these children is through the work of the general practitioner and the hospital out-patient and in-patient departments. All these are heavily burdened with work, so that, on the one hand, it is only

the child who is really ill—that is to say, who is suffering an exacerbation of his condition—who is brought for treatment. On the other hand, it is only possible by the undertaking of special and arduous work to correlate the records of these children. There is, therefore, in the system at present in force no facility for the continuous study of children from the constitutional point of view, and if this is to be undertaken some other machinery must be devised.

### Bearing on Prevention and Control.

It is in a sense true in this, as in all diseases, that the problem of prevention and control is dependent upon the discovery of the cause or causes of the disease. But it is perhaps worth remembering, in the face of the difficulty of the problem, that the elimination of a disease can be achieved Without an exact knowledge of its causation, as in the case of leprosy and typhus in Europe, and that the discovery of the Klebs-Löffler bacillus has not been followed by the disappearance of diphtheria.

In rheumatism it is clear that there are two aspects to the problem, each of which requires separate consideration. The one, that to which attention is increasingly being directed is the care of the heart of the already rheumatic child. The other is the attempt to control the incidence of the disease itself, and to achieve means for the prevention of its appearance in children who can be expected to become rheumatic. These problems, though intimately related, and common in origin, need to be kept very carefully distinct. All chances of success depend upon their separate consideration, and it is the failure to do so that determines the ineffectiveness of so many of the proposals at present current.

It is obvious that any scheme, either for the care of children already rheumatic, or for the study of the disease, must be based upon a definite hypothesis of the nature of the disease. Up to 1922, as has already been pointed out, work upon rheumatism has been conceived from the infective point of view and the results have been, on the whole, disappointing. In America a new step was taken with the formation of the Association for the Relief and Prevention of Heart Disease. This was intended primarily for adults, and embraced all forms of heart disease, but in 1915, W. St. Lawrence (5) adapted these methods to the study of the effect of rheumatism in childhood, and the first children's cardiac clinic was opened at St. Luke's Hospital, New York. The aim of this clinic was the continuous treatment and observation, upon a classified system, of rheumatic children, with special reference to their heart condition.

In this country nothing of the kind was attempted until 1923. In this year, interested by St. Lawrence's work, Prof. Leonard Findlay and the present writer organised a clinic at the Royal Hospital for Sick Children, Glasgow, for the continuous supervision of rheumatic

children attending that hospital. (6) As a result of this work certain facts became clear. It appeared that the manifestations of rheumatism in childhood, usually covered by such terms as the acute and convalescent stages, needed the addition of a third term for a complete description—that of the quiescent stage. Under this head is considered the period between attacks, when the disease smolders, but is not apparent. Upon the conception of this quiescent stage the whole work of a supervisory clinic is based.

#### A New Basis of Classification.

The second point of note arising out of the clinic is that of the importance of the basis of classification of these children. Hitherto, all classification of rheumatic children, whether explicit in records, or implicit in the mind of the observer has been based upon the clinical form of the disease—erg, rheumatic fever, chorea, tonsillitis. It has, however, been agreed that the significance of the disease does not lie here, but in its tendency to cardiac involvement. These terms, therefore, offer no help to the observer in his attempt to protect the future of the child. The use of them, moreover, necessitates a somewhat lengthy and detailed note on the cardiac condition being added to each case, and renders very difficult any survey of a number of cases.

(5) Hospital Service Series, May, 1920, pp. 151—181.

(6) Brit. Med. Jour., 1926, 1., 817 and 820.

For the working of this clinic the emphasis was shifted, and a classification was devised upon the basis of the children's cardiac history. Since the clinical findings by different observers in any given cardiac case are not always necessarily identical, an endeavour was made to arrange the classes so the discrepancies arising from this source should be reduced to a minimum. The classification finally adopted was as follows :—

Class P. . . Potential heart cases—that is, cases with definite rheumatic history who have never had cardiac involvement and are now well.

Class A. . . Cases with a history of definite rheumatism with cardiac involvement, which has now disappeared.

Class B. . . Cases with a definite rheumatic history and present signs of cardiac involvement, but no symptoms referable to the heart.

Class C. . . Children with a definite rheumatic history and present signs and symptoms of cardiac involvement, but no disability for ordinary life.

Class D.1. . . Cases with a definite rheumatic history, present signs and symptoms of cardiac involvement, and definite disability for ordinary life.

Class D.2 . . . Cases without a rheumatic history, but showing signs of a cardiac lesion, and with some definite disability for ordinary life.

Miscellaneous. . . Cardiac irregularities, effort syndrome and cutaneous eruptions supposed to be of rheumatic origin.

From a practical point of view such a classification has the following advantages. When the original classification has been made, subsequent notes are reduced to a minimum. "Mild Class B.," for instance, can describe all the essential points about a case. Whether this lesion is the result of an acute rheumatic fever, of growing pains, or of tonsillitis, though of interest, from the point of view of immediate treatment prognosis or recommendations to the mother, is of comparatively little significance. It also makes possible a rapid survey of a large number of cases without elaborate cross-indexing. By collection of those children whose classes have been changed during the year, immediate information is given regarding percentage improvement or deterioration in cardiac condition of the children. Further, any class of child can be quickly and easily isolated for reference, examination, or experimental therapeutics.

It will be noted that this classification embraces only children already the subject of a definite rheumatic condition. This was deliberate, because the original clinic was devised to study the possibilities of prevention and control of heart disease in children who were already rheumatic. In other words, to keep Class P. children from moving into any other class, and to move as many of those in the other categories as possible to Class A.

#### An Attack Upon the Rheumatic Constitution.

Another fact stands out clearly as the result of the work of this clinic—viz., that while work of this kind is very useful and effective within its own limits, if an attempt is to be made to achieve the prevention of rheumatism, a still earlier attack must be made upon the disease. But since all signs of rheumatism in children, other than those of acute rheumatic fever, chorea, and tonsillitis are ill-defined and susceptible of varying interpretation, the formation of a clinic upon this basis will involve the assumption of the existence of a rheumatic constitution. That is to say, children of rheumatic stock and those children who are subject to all forms of vague and indefinite rheumatism will also need to be included. Not only must the basis of the clinic be thus widened, but the treatment of

these children must follow different lines and be directed as much towards raising the child's general resistance, as to combating his definitely rheumatic tendencies.

These are as yet hypothetical conceptions and need wide testing before their acceptance. One attempt to do so is already in existence in the form of a "children's cardiac clinic" at the St. Marylebone General Dispensary, which has been in operation since 1926. At this clinic children, whether definitely or possibly rheumatic, are received from the age of 2 years, and supervised until puberty, when they join one of the adult clinics of the dispensary. The modifications introduced into the classification are as follows:—

1. Class P. becomes subdivided into——

P.1 Children with history and complaints suggestive of rheumatism, no cardiac involvement, and no disability for ordinary life.

P.2 Children with history and complaints suggestive of rheumatism, no cardiac involvement, and some disability for ordinary life.

P.3 As in original classification.

2. Class B. becomes subdivided into——

B.1 As in original classification.

B.2 Children with a history suggestive of rheumatism present signs of cardiac involvement, but no symptoms referable to the heart.

The standardisation of the classification with that already in use elsewhere allows both accurate comparison of results, and full development of preventive measures in either.

Since the publication of the report of the Glasgow Clinic, the Science Committee of the British Medical Association have sat for a further consideration of the problem and have issued a second report (7) on the Prevention and Control of Juvenile Rheumatic Infection. In this communication the existence of the quiescent stage is accepted, and the principle of continuous medical supervision emphatically recommended. The details of the one centre to which reference is made in the report are, however, so unsubstantial that it is impossible to discover upon what lines it is organized. What class of child is accepted, and how classified, and upon what lines the suggestions of treatment are made. The

actual proposals of the report also suffer from the same vagueness; and this is unfortunate, because in the medical supervision of rheumatic children we have not only a method of treatment but also an instrument Which can be used both for the examination of the effects of treatment and to the study of the disease itself. At present treatment of the disease and inquiry into its nature are far too apt to be kept in watertight compartments. What is needed is a method by which the natural history of the disease, the factors influencing or retarding its development, the effect of attempts to modify the constitutional habit, can be provided in a largo number of cases in such a form as to be readily and easily manipulated and studied. It should be possible to achieve this and simultaneously to provide abundant material (or aetiological and therapeutic research.

(7) Brit. Med. Jour., April 16th, 1927.

It is clear that work of this kind opens new avenues for correlated research, while it demands some modification in our attitude to systems for control of disease. Such methods, by providing material for investigation upon any desired lines, and by demonstrating the life-history of the disease, will exactly fill the obvious gap in our present system. It is true that there may be difficulties in associating these principles with the work of a voluntary hospital, but they are, I am confident, not insuperable. On the other hand, by work of this kind a weapon can be forged for the study of the constitutional hypothesis of rheumatism, which, it is contended, is the next and most hopeful step to be taken towards an understanding of the disease.