A British Medical Association Lecture ON RHEUMATISM IN CHILDREN.

DELIVERED BEFORE THE DUNDEE BRANCH,
FEBRUARY 5TH, 1929,

BY
GEO. A. ALLAN, M.D., F.R.F.P.S., M.R.C.P.,
VISITING PHYSICIAN AND LECTURER IN CLINICAL MEDICINE,
WESTERN INFIRMARY, GLASGOW.

THE reawakening of interest in the subject of rheumatism has been a striking feature of medical history in this country during the past five years. The investigations that had previously been undertaken with characteristic vigour by the American Heart Association were no doubt a factor in the process, but since 1923, when Poynton (who has been an enthusiastic worker on this subject for over a quarter of a century) published a plea for the prevention of rheumatic heart disease, this matter has been constantly before the profession.

The special committee of the British Medical Association appointed to inquire into this subject can also claim some credit for the renewed interest, and in its published reports has helped not a little to clarify the situation and to point the way to further methods of advance. The chief medical officer of the Board of Education in England has repeatedly emphasized the importance of the subject in his annual reports. The Medical Research Council, the Section of Diseases of Children of the British Medical Association, the Society of Medical Officers of Health, the Child Welfare Section of the Sanitary Institute Congress, and the Society of School Medical Officers of Scotland, in addition to numerous Divisions and Branches of the British Medical Association, have all had the matter under discussion. The experiments in after-care carried out by the Invalid Children's Aid Association in their heart homes, and by others in different parts of the country, notably in Birmingham, have demonstrated what can be done in the direction of limiting the devastation of this serious disease.

When one looks up the subject of acute rheumatism or rheumatic fever in some of the older books, description is found of an affection of the joints, which are red, swollen, and painful, exquisitely tender so that even a touch on the bed is feared, usually associated with fever, sweating, and malaise, and with, as a possible complication, heart disease. The description when applied to the adult may be correct, but when transferred to the child is apt to leave a very erroneous impression. The more the disease is studied in the child, the more it is realized that rheumatism is a general infection, of which after all the joint condition is in essence a minor manifestation. The latter is eminently curable, leaves no deformity, and were the disease so limited one would not be so much concerned with its high incidence among the population of this country. In the child the joints seldom occupy the prominent place that they do in the picture of the adult disease, while the heart less rarely escapes from the effect of the general infection.

There may be different explanations offered as to the special susceptibility of the heart in the child, but comparatively recent investigations by Gross and his collaborators in America into the vascularity of the heart afford a possible clue to the problem. It is now generally accepted that rheumatism in the heart is brought to it by the coronary circulation, and not by the blood passing through its cavities. In the child, and for a variable period in adult life, there is a blood supply which can be demonstrated right into the cusps of the various valves. This vascularity tends to disappear first from the arterial valves, and later from the auriculo-ventricular valves, the last cusp to lose its blood supply being the aortic cusp of the mitral valve. It is very suggestive in this connexion that the mitral valve is more frequently involved than the aortic valve, and the cusp more severely damaged as a rule over a series of cases will be found to be the aortic cusp

of the mitral. Another point that must be clearly realized at the outset is that rheumatism in its first attack is essentially a disease of childhood. The highest peak of incidence varies slightly, according to different observers, from the seventh to the tenth year of life, the vast majority of primary attacks of rheumatism taking place before the age of 15. When once rheumatism has affected the heart, particularly the valves, it is found that vessels persist in the valves for a much longer period than in healthy hearts, so that in hearts once damaged, even to a slight extent, subsequent attacks of rheumatism are more prone to produce further damage, even in adult life, than in patients who had not had an attack in childhood. If any good, therefore, is to be done towards lessening the prevalence of heart disease, steps must be taken to circumvent the infection in the child. If, with our present knowledge, it is impossible to prevent rheumatism by the study of the disease in the child, it may still be possible to prevent relapses and to lessen the number of cardiac cripples. The aim of anyone interested in this subject must therefore be (1) to determine as far as possible the conditions which produce or predispose to the infection, and (2) to acquire a knowledge of the earliest manifestations which may indicate the presence of the disease. With our present knowledge of the natural history of rheumatism this may be an ideal difficult to attain; this should not hinder the attempt but rather stimulate to further effort.

PREDISPOSING OR EXCITING FACTORS.

Much uncertainty still exists as to the factors which predispose to rheumatic infection. In inquiring into the family history of cases of children suffering from rheumatism, a history is obtained of parents, brothers, or sisters who have been similarly affected in about as great a proportion as is found in the case of tuberculosis, so that the question of heredity in some degree cannot be lightly dismissed. Many believe, and with considerable ground for their opinion, that some children are born with a special liability to acquire rheumatic infection. That children have been born actually suffering from rheumatic endocarditis has been demonstrated, though so rarely as to be hardly worthy of serious consideration in this connexion. Some have gone so far as to suggest that alteration in the balance between the various ductless glands, notably the thyroids, adrenals, and pituitary, has been the basis of the tendency which has been inherited; and there is certainly in many rheumatic children a nervousness and a tendency to tachycardia that make one hesitate before dismissing lightly such a possibility. Such questions, however, have comparatively little practical application, except in so far as helping one generation may be a means of lessening the incidence of rheumatism in the next.

No one, I think, doubts that environment plays an important part in the incidence of this disease, and while statistical evidence as regards dampness is rather contradictory, the spot maps of certain towns in England where a careful inquiry into this question has been carried out are very difficult to explain on any other basis than that dampness in dwellings is a potent factor in predisposing to the disease. Poynton is confident that the rheumatic child fares badly in such surroundings, and that there is a tendency for the rheumatism in such circumstances to become intractable. He states, "I have seen too many examples to be disturbed by any statistics on this point." In addition to the dampness of dwellings, dampness of clothing and footwear should also be considered. These certainly have a direct influence in promoting catarrhal affections, particularly of the nasopharynx. Further, social status apparently plays a part, though the exact factor therein is not obvious. The elementary council or board school provides a much greater percentage of cases than the average boarding or private school.

Sources of Infection.

With regard to the actual infection, little more need be said than that the disease seems undoubtedly to be due to infection of some kind, but that there is still no uniformity of opinion as to whether the micrococcus of Poynton and Payne, or some other organism or symbiosis of organisms, is the exciting factor. Apart from knowledge of the actual

organism, attempts have been made to determine its site of entry into the body, and the one which gains most support is the tonsil. The teeth have also come under suspicion, but with less ground, while some believe that the intestine may harbour the organism after its entry. The results of enucleation of the tonsils have been more or less unsatisfactory. Some of the latest investigations along this line show that relapses are as frequent in the operated as in the non-operated cases. Miller's figures, however, seem to show that removal of the tonsils lessens the liability to rheumatic joint relapses, and that patients who do relapse afterwards usually manifest the infection in the form of chorea. That enucleation of the tonsils is not a preventive against relapses cannot, of course, be used as an argument against their being the site of primary infection, because it is well recognized that once the infection gains entry to internal organs its eradication is a much more difficult problem.

While the last word has not been written on any of these factors, experiments have been carried out over a sufficiently long period, both in this country and in America, to warrant the view that improvement in environmental conditions can affect for the better the prospects of the rheumatic subject.

EARLY MANIFESTATIONS.

Cardiac Rheumatism.

Apart from the comparatively few cases of congenital heart disease, there is no common affection of the heart in childhood except rheumatism, and it should be recognized that the heart may be the first part to show signs of the infection. Mitral stenosis is usually accepted as a typically rheumatic manifestation, and yet in a series of hospital cases with this lesion over a period of ten years 25 per cent. of the males and 31 per cent. of the females gave no history of antecedent rheumatic manifestation. In many of these cases the heart had been quietly damaged without other sign of the disease, for all patients could not have forgotten other symptoms of rheumatism.

We frequently speak of endocarditis, or of pericarditis, but it may be accepted that in neither case does the myocardium escape. The effect which should be sought in this connexion is enlargement of the heart as made out by percussion. We are too apt to ascribe all changes in the size of the heart to pressure effect, but the dilatation is really more dependent on the extent of muscle damage than on the degree of valvular lesion. Likewise in pericarditis with effusion the increase in dullness, which is often ascribed to the fluid, is in many cases due chiefly to the dilated heart. Dilatation of the heart in a child subject to even suspicious rheumatic manifestations would warrant the assumption that the heart was already in the grip of the rheumatic poison. Dilatation of the heart is really the most reliable early sign in a child, as characteristic lesions which would allow one to diagnose valvular involvement, such as mitral stenosis or even aortic incompetence, do not necessarily present themselves early in the disease. Mitral stenosis particularly may take some years before it develops characteristic physical signs. The presence of a systolic murmur at the apex is perhaps one of the most difficult to assess. In the absence of any dilatation of the heart it can usually be neglected if it is the only sign, but in association with dilatation of the heart it is presumptive evidence that the mitral valve is also affected. An aortic systolic murmur is much less common as an early indication of heart involvement, and is rarely present without a diastolic murmur of aortic incompetence as well. The heart rate, in the absence of auriculo-ventricular block, is usually accelerated, but taken as a solitary sign is not of much value in determining implication of the heart, as so many other causes in the young child can produce the same increased frequency. This sign, however, cannot be neglected, and in the absence of any other cause for acceleration, should be reckoned a suspicious feature. Disorder of rhythm in the child is not common. Auricular fibrillation is rare under 15 years of age; heart-block, however, may occur during the acute stage and may be a transient feature. The presence of extra-systoles, generally infrequent in childhood, would suggest some toxæmia.

Mackenzie indicated that the respiratory type of sinus irregularity could be considered as a sign that the heart had escaped. This view is not now generally accepted. Sinus irregularity is not common with a rapid heart action, and consequently its reappearance may be expected if the heart rate becomes slow. This is probably its only significance, as it has been found in association with such a definite muscle lesion as heart-block.

I have purposely placed cardiac rheumatism first on the list, not only on account of its very great intrinsic importance, but because in the child the heart very rarely completely escapes, and also to emphasize the fact that it may be the first part to show signs of the disease.

According to a recently published statistical inquiry covering 500 children in a New York cardiac clinic, the heart was the first part to be affected in 6 per cent. of the patients. Furthermore, some figures which have recently come to light in connexion with disordered action of the heart—irritable heart—suggest that rheumatic infection may lead to this condition in the absence of valvular disease. Thus Parkinson found disordered action of the heart in a higher percentage of cases in men with a rheumatic history without valvular disease than in those with no rheumatic history.

Any of the above signs have a much greater significance when found in association with other typical rheumatic manifestations, and we may now proceed to consider some of these.

Polyarthritis.

Articular rheumatism is the commonest condition, or at least the commonest to be recognized, antecedent to cardiac infection. In the New York series it occurred in 66 per cent. and was the earliest manifestation in 40 per cent. In many cases, of course, there were relapses from one to five times. The diagnosis of these cases presents little difficulty.

Growing Pains.

This is the group to which I would like specially to direct attention. That it is in most cases in the nature of a true rheumatic manifestation seems undoubted. In the New York series it occurred in 67 per cent. of the cases and was the first manifestation in 27 per cent. It should be noted that in 10 per cent. carditis followed without any other polyarthritis or chorea. Until growing pains are considered as worthy of medical treatment, both by the doctor and the parent, many cases will be allowed to drift into the state of cardiac cripples.

Tonsillitis.

It is difficult to present valuable figures for this condition in its relation to heart disease. That tonsillitis is a frequent accompaniment of other definite rheumatic features, that diseased tonsils are more frequent in rheumatic than in other subjects, and that attacks of tonsillitis often accompany or usher in relapses, is undoubted. It was the first manifestation in 14 per cent. of the New York series. Unfortunately the removal of tonsils may also be immediately followed by rheumatic symptoms.

Chorea.

Apart from the chorea which was a feature of lethargic encephalitis at one period of the epidemic, chorea must be considered as rheumatic in origin. It may be the only rheumatic precursor of carditis, and was so in 21 per cent. of the New York series, in which it occurred in 43 per cent. of the cases. It is as frequently followed by severe carditis as is polyarthritis, and its early recognition is important. The fidgety, restless, nervous child requires very special care at school, and it would be an interesting study to determine how much the strain of school examinations is a determining factor in the production of chorea in susceptible children.

Nodules and Other Signs.

I place great stress on the presence of nodules as an aid to diagnosis and treatment. Figures are unreliable, as the nodules must be looked for regularly if their frequency

is to be recognized. Sometimes transient, coming in crops, they are neither painful nor tender, and so may be overlooked. They have a structure similar to that of the submiliary nodules in the heart.

Anaemia, pleurisy, and pyrexia of unknown origin, while also suggestive of tuberculosis, may be early evidence of rheumatic infection.

Clinical Syndromes.

Generally speaking, cases of rheumatic heart disease may be grouped along certain broad lines, as suggested by Poynton. Perhaps the commonest group is associated with polyarthritis, tonsillitis, or growing pains. The second group is that associated with chorea, with or without some of the other manifestations. In either of those the heart may be quietly damaged, and the full effect of the lesion may only be manifest after one or more relapses. In another group, more often also associated with polyarthritis, the heart is the seat of pancarditis, the endocardium, myocardium, and pericardium being involved, and the patient may be very seriously ill from the onset. In still another group without other rheumatic manifestations the heart may be quietly damaged.

Relapse.

This is a well-known feature, and as succeeding attacks rarely fail to increase the cardiac damage, attention to them, or, if possible, their prevention, may do much to lessen the cardiac effects. In the New York series 70 to 85 per cent. of the patients relapsed, the largest figure being found in those in whom the initial attack occurred in very early life. The relapse may be within a year, or delayed for several years. In half the cases in this series the first relapse occurred within one year; in two-thirds within two years; in four-fifths within three years, and in the remainder up to ten years. The susceptibility to infection decreased after the age of 12.

TREATMENT AND SUPERVISION.

In treatment of the rheumatic infection in childhood time and patience are all-important factors, and it is to inability to secure these from all the parties concerned that many of the failures must be ascribed.

The salicylates, while potent in relieving symptoms and fever in joint cases, seem to fail when the heart or brain is involved. Not only so, but some would argue that the rapid amelioration of symptoms in joint cases induces a false sense of security, so that sufficient time is not allowed for the heart to recover its tone. When the heart is actually the seat of infection salicylates seem to be of little value, and relapses of fever are about as frequent with salicylates as without them.

Rest in bed should be demanded till all signs of activity have passed, and for some time thereafter. Fever and pain are definite and easily appreciated indications for rest, but, in their absence, nodules or tachycardia are equally significant. When it is decided to allow the patient to get up the heart rate should be carefully watched, and any undue acceleration accepted as an indication for more rest. Later, gradually increasing exercise, with the same precaution, is of value. Iron and cod-liver oil help to restore vigour to the cardiac muscle, and should be continued for some time. Experience shows that for many of the cases prolonged convalescence, under good hygienic conditions, with fresh air and sunshine, is necessary.

The problem with which we are faced in trying to deal with rheumatic heart disease hinges on the difficulty of securing lengthy treatment. The public (medical and lay) have been educated to face prolonged treatment in tuberculosis, but not yet in rheumatism. Heart disease kills annually as many as tuberculosis, and, like it, the rheumatic type kills early. Looked at as an economic problem rheumatism deserves as much consideration as tuberculosis, but, of course, the question of isolation does not arise in the same way, and so a good argument is lost.

For the well-to-do the doctor and parents can, if alive to the need, secure the necessary treatment; but rheumatic heart disease is more common in a grade of society that

cannot readily face such a course. The hospitals obviously cannot at present provide beds for the lengthy residence that is very often required. The hospital convalescent homes are suitable neither in size nor arrangement to deal with those cases. The British Medical Association subcommittee put forward a plea for the establishment of special residential hospital schools or homes, and the Medical Research Council, in no uncertain words, argues on similar lines: "The plain need for organized after-care offers an urgent but soluble problem of preventive medicine, too long neglected, and every motive of humanity and wise economy should impel the community to provide the after-care as rapidly as possible."

All cases, of course, are not suitable for such homes—for instance, advanced or acute cases. But there are the children who have just passed through an attack of arthritis, chorea, or carditis, or very mild or even potential cases, or quiescent cases, for whom the strain of ordinary school life is too great. The Birmingham people, as a result of the earlier years' working, have had the accommodation doubled, and all reports are favourable as to the value to the patients. Until such places are available, and even afterwards, much remains to be done. All rheumatic children need special and regular supervision, and it seems to me that only by co-operation between practitioners, ante-natal clinic workers, school medical officers, and hospitals, will this be efficient, as first manifestations may be observed by any one of them.

The Rheumatism Clinic.

My own feeling is that any scheme on the lines adopted at Paddington Green under Dr. Reginald Miller serves a useful purpose. Attached to the hospital is a supervisory centre where all known rheumatic children are asked to attend. A temporary scheme of notification is adopted so that the medical officer of health can also give his help. Patients attending are carefully recorded and registered. Any line of treatment considered necessary is recommended only, and is carried out by the family doctor or the hospital, as is considered best. Children who would benefit by country residence are recommended to the care of the Invalid Children's Aid Association.

Another very useful function of such a centre is that of education. The parents are provided with a card giving general instructions on the care of rheumatic children. Similar instructions have been given in other parts of the country through the school medical officers whenever a case presented itself at the routine examination. Education of school nurses and teachers has also an important place in any scheme, as these can often detect early cases of chorea or other features of ill health before the doctor is called in.

The essential feature of any such scheme is that there must be co-operation between the various bodies concerned. No patient should be dismissed from hospital without information being sent to the practitioner or rheumatic centre, and, if fit for school, to the school medical officer. Similarly, cases seen at school should be notified to the practitioner or centre for supervision, while there should be ready access to hospital for patients requiring special care. The function of the centre would naturally be that of co-ordinating and supervising.

To make any impression on the incidence of rheumatic heart disease the problem must be faced from the aspect of rheumatism in childhood. At that stage alone is there any hope. The treatment of auricular fibrillation is often an eminently satisfactory piece of work, but, after all, it is only a patched-up job. To prevent relapses in a child, or, better still, to prevent rheumatism, is to save lives.

Even without a complete knowledge of all the facts of the natural history of this disease much good can be done now by: (1) Education of all in touch with child life—parents, teachers, nurses, doctors—in the early manifestations, the best plan to combat them, and the risk of neglect. (2) Prolonged care, by the doctor, of the child who has been attacked until all signs of activity have ceased. (3) Prolonged after-care in suitable surroundings for those who are recovering. (4) Constant supervision to detect the early signs of ill health, or of relapses.