BRITISH MEDICAL JOURNAL

LONDON: SATURDAY, JULY 20th, 1935

ACUTE RHEUMATISM AND ITS RESULTS*

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Acute rheumatism is one of the most important diseases of this country—because it is common, because it causes serious cardiac damage and produces reduced efficiency, ill-health, or chronic invalidism in a considerable proportion of its victims, and because it shortens many lives. Its history is comparatively brief: Shakespeare makes no use of the word rheumatism in its modern sense, and it was only in the year of his death that acute arthritis was first called rheumatism by a Frenchman, Gwilliam de Baillou.

Sydenham's Descriptions

The first accurate clinical description was written by Sydenham at the end of the seventeenth century. He said: "The disease comes on at any time, but especially in the autumn, and chiefly seizes those that are in the flower of their age . . . it begins with shivering and shaking and presently heat, restlessness and thirst, and other symptoms which accompany a fever. After a day or two, and sometimes sooner, the patient is troubled with a violent pain sometimes in this, sometimes in that joint, in the wrists and shoulders, but most commonly in the knees. It now and then changes places and seizes elsewhere, leaving some redness and swelling in the part last possessed."

He described chorea too, but does not appear to have associated the two conditions. The picture of chorea is very good: "This disease is a kind of convulsion which seizes children of both sexes from the tenth to the fourteenth year. It manifests itself by a halting or unsteadiness of one of the legs, which the patient draws after him like an idiot. If the hand of the same side be applied to the breast, or any other part of the body, the child cannot keep it a moment in the same posture, but it will be drawn into a different one by a kind of convulsion, notwithstanding all his efforts to the contrary. Before a child who hath this disorder can get a glass or a cup to his mouth he useth abundance of odd gestures, for he does not bring it in a straight line thereto, but his hand being drawn sideways by the spasm, he moves it backwards and forwards till, at length the glass accidentally coming nearer his lips, he throws the liquor hastily into his mouth, and swallows it greedily as if he meant to divert the spectators."

Towards the Modern Conception

But it is not until the year 1816 that we first find anything resembling the modern conception of the pathology of acute rheumatism. For whereas Sydenham,

Haygarth, Cullen, and all the earlier writers had regarded rheumatism as essentially a disease of articular structures, Balfour took a wider and more modern view. According to him the seat of the disease was in the cellular membrane, and he wrote: "The cellular membrane abounds everywhere in the body . . . all the blood vessels receive a coat from it, from the aorta where it emerges from the heart to the minutest capillary that enters a tendon." Similar ideas were held by Charles Scudamore. His *Treatise on Nature and Cure of Gout and Rheumatism* (1819) constitutes the disease as "a peculiar species of inflammation, affecting parts which have a fibrous texture, and most frequently synovial membrane. . . ." He realized, as did Balfour, the remarkable predilection of rheumatism for the synovial membranes of the body, and gave as examples of this fact instances of inflammation of the dura mater and of the pericardium. "The pericardium," he stated, "also sometimes becomes the seat of similar morbid action which I conceive as secondary and induced by the inflammation which is affecting some part of the tendinous structures of the heart."

A French physician, Lasègue, made a statement which is so graphic that it ought to be preserved: "Rheumatism is a disease which licks the joints, the pleura, and the meninges, but it bites the heart." The greatest names associated with the development of our knowledge of acute rheumatism and its results are English. In 1843 Sir Thomas Watson pointed out that children as early as 3 or 4 years of age might develop carditis, and that the earlier they were attacked the more severe was the resultant damage. All the earlier writers thought in terms of endocarditis or pericarditis, and it was not until 1876 and later that the essential damage to the myocardium was recognized. This was the result of the work of Sturges, Barlow, Cheadle, Lees, and Poynton.

So we see that whereas early in the nineteenth century acute rheumatism was considered merely a disease of joints, with the occasional involvement of heart, brain, or lungs by a kind of metastatic spread, and that in the occasional cardiac involvement pericarditis was common, endocarditis rare, and myocarditis undreamed of, by the end of the century a very marked change in medical knowledge had caused acute rheumatism to be regarded as a protean disease commonly associated with serious results inasmuch as a characteristic type of inflammation involved blood vessels throughout the body but especially in the heart and synovial membranes. Moreover, it was recognized that rheumatism was essentially a disease of childhood, and that while the involvement of joints and subcutaneous tissues was of small importance, yet in every case some cardiac damage occurred.

* Abstract of the Ingleby Lectures delivered at the University of Birmingham on June 13th and 20th, 1935.

Aetiology

Many and various have been the theories propounded to explain acute rheumatism, for the problems involved are complex and none is yet completely solved. Cold and damp were first suggested as causal factors, but chemical abnormalities such as an excess of lactic acid or uric acid have been suggested, and indeed were responsible for the alkaline treatment of the condition. J. R. Mitchell suggested that it was a trophic disorder, and T. J. MacLagan thought it was allied to malaria, and on this assumption sought and found a remedy (salicin) in the bark of the willow which grew where rheumatism was most common.

The bacteriology of the disease began with Mantle's discovery of a diplococcus in the blood of a rheumatic child in 1886, and in the opening years of the century Poynton and Payne made a series of most careful and consistent investigations which resulted in a diplococcus being incriminated as the causal organism. Subsequent observers failed to repeat their findings, and very detailed serological and immunological studies have had no far-reaching results.

In 1895 Sir Arthur Newsholme, in the Milroy Lectures, considered the subject from the statistical standpoint, and came to some remarkable conclusions. That the disease is a specific disorder due to an infecting micro-organism; that the disease follows an initial sore throat, tends to relapse, and predisposes to subsequent attacks. That the infectivity of any case is very low, probably because the organismal cause is concentrated about the joints, and that the infecting agent is probably a saprophyte which can become a parasite.

It has been suggested that the lesions of rheumatism are not due to any specific organism, and are not the result of a toxæmia, but are really allergic manifestations due to a peculiar tissue reaction in certain individuals. The known facts and the finding of absolutely characteristic tissue changes are against such a theory. The latest suggestion is that of Schlesinger, Signy, and Amies, who believe that they have recovered a virus from cases of rheumatism in children, and although it is too early to pronounce definitely upon this suggestion it fits the known facts very well indeed.

Rheumatism is a disease of temperate climates and is rare in the Tropics; it is essentially a disease of childhood, and appears chiefly in the winter and spring, and in epidemics. In wards of hospitals small epidemics and epidemic recurrences follow streptococcal sore throats, but there are epidemic years in which the disease is at least four times as common as in the relatively free years. Girls are slightly more susceptible than boys, and there is a well-marked family incidence, but the most important known factor is environment, for acute rheumatism is a disease of the relatively poor. Still found that 13.1 per cent. of his hospital patients had rheumatism, while only 0.7 per cent. of his private patients suffered from it. The disease is really extraordinarily rare in our big public schools.

Symptoms

The diagnosis of rheumatism is not usually as easily made in the child as in the adult, for the onset is less acute, and joint involvement is seldom so marked in the young. The rheumatic child may be anaemic, listless, and easily tired, may lose weight or more often fail to gain weight normally. Pains of a rather vague type are common in muscles or joints, are made worse by exercise or wet weather, and are relieved by warmth, sunshine, or a holiday. Epigastric discomfort is common, and is often associated with repeated sore throat. In other cases the child becomes unnaturally jumpy and fidgety, is emotional and incapable of sustained attention. Insomnia,

night terrors, enuresis, or tics may appear. Such children react badly to the stresses of school, and may become frankly choreic. One of the earliest signs of chorea is irregularity of respiratory movement, which usually antedates the characteristic quick and irregular contortions of the face or limbs. It is important to recognize that such movements are not repetitions like a true tic.

But the first definite sign of rheumatism in the child may be a frank cardiac abnormality, and only too often when first seen the patient has a seriously damaged heart. As a rule some vague illness has existed for some time, but has not been sufficiently striking to demand attention. Irregular fever with sweating and some degree of malnutrition are commonly associated with early carditis. One of the first signs of cardiac rheumatism is a quickened pulse, the normal juvenile variation in the pulse rate disappears, and the pulse becomes unnaturally accelerated, especially during sleep. The heart sounds change in character—the first sound at the apex becomes soft, blurred, and accompanied by a systolic murmur. The second sound at the base may be intensified or split.

Change in the size of the heart in the early stages of carditis is a result of pericardial effusion, and is associated, too, with alteration in the outline of the heart, blurring of both heart sounds in all areas, an almost impalpable cardiac impulse and some degree of pulsus paradoxus. As the heart adapts itself to myocardial damage or valvular defects, enlargement of a permanent nature may appear, and the degree of this increase will give a useful clue to the prognosis, for the larger the heart the more serious is the damage. Acute dilatation of the heart, as a result of acute rheumatism or any other infection, if it ever occurs, must be exceedingly rare, for years of careful observation and frequent measurement, with the help of *x*-ray examinations, have never shown me a single example of the condition.

Electrocardiographic abnormalities are often found early in rheumatic carditis, but are seldom gross in really early cases and may completely disappear. Such are abnormalities of the QRS, flattening of T, and minor degrees of heart-block—lengthening of P-R. Gross change in any of the complexes, or the onset of auricular fibrillation always implies a most serious carditis and a poor outlook, but auricular flutter is sometimes found in children who do well.

Perhaps the most characteristic single sign in acute rheumatism is the rheumatic nodule. This is a subcutaneous lump which appears singly or in groups about the elbows, knees, metacarpo-phalangeal joints, wrists, and ankles. They may be found in great numbers or may be very scanty; sometimes they appear in the scalp about the occiput and occasionally they are found along the iliac crests; frequently they can be felt as peculiar small, slightly tender thickenings in the lumbar muscles. The lumps may appear or disappear with astonishing rapidity, although usually they remain for some weeks.

Results of Acute Rheumatism

Rheumatic inflammation, wherever it appears, produces a characteristic microscopic lesion—the submiliary focus. It is common in heart muscle, pericardium, endocardium, synovial, and subcutaneous tissues, and is found in the adventitia and media of arteries in all parts of the body. The typical focus shows four distinct points in its structure:

1. A fibrinous matrix.
2. Certain large multinucleated cells.
3. Plasma cells or fibroblasts.
4. An aggregation of leucocytes—darkly staining mononuclear cells.

At first the nodule shows an aggregation of lymphocytes and plasma cells, later the characteristic multinucleated cells appear, and finally the lesion is replaced by a patch of fibrous tissue. These foci are always accompanied by more or less marked changes in the parenchyma of the tissue invaded, and produce a considerable degree of vascular proliferation and hyperaemia. In the more chronic forms of acute rheumatism—especially that deadly yet mild type of the disease which gradually leads to mitral stenosis, the degree of fibrosis of the myocardium may be truly astonishing. It is of the greatest importance to realize that even in the mildest case the myocardium is always involved, the endocardium may escape; when it is damaged, with consequent valvular defect, the lesion is preceded by changes in the valve leaflets, and the valves are not directly infected from the blood in the chambers of the heart. This statement is borne out by the common knowledge that the mitral valve is most frequently, and the aortic valve quite commonly, damaged, while the tricuspid and pulmonary valves are relatively seldom attacked. It is now held that damage is the result of both stresses and vascularity: the relatively avascular pulmonary valves, which sustain comparatively little stress, escape; while the vascular and heavily stressed mitral valve is most seriously and most often affected.

Acute rheumatism is common: Sir George Newman has calculated that 0.8 per cent. of all elementary school children have rheumatic cardiac disease—50,000 cases in England and Wales. Carey Coombes showed that about 40 per cent. of all heart disease was rheumatic, that the average rheumatic cardiac patient lived to 28½ years, and that only 7 per cent. of such patients attained the age of 50. He also showed that nearly half the children who acquire rheumatism before the age of 5 years develop serious cardiac damage, while only a quarter of those who begin their rheumatism after 10 years become cardiac patients.

Treatment

The first step, then, must be the education of civic authorities so that health may be ensured by well-built houses in sunny smokeless cities, which are not overcrowded and contain spacious suitable playgrounds. The second step should be the education of parents in the wise nurture of children, so that their resistance to disease may be high and good health may be preserved. This implies wise feeding, proper clothing, and adequate changing and drying of wet clothing and boots, as well as the early and careful attention to minor infections, sore throats, colds, and damaged teeth. When acute rheumatism, in any of its varied manifestations, does occur it must be recognized early and treated promptly and thoroughly.

To-day there are usually three great essentials in treatment. Rest is the greatest of these, and should be complete in the early stages and prolonged if carditis supervenes. Rest implies that the child is carefully nursed nearly flat in bed, fed, and so attended that the patient learns to make no effort or unnecessary movement. In severe carditis it may be necessary to continue this rest for months, and in any case the return to activity should be carefully graduated and watched to see what effect effort has on pulse rate and cardiac action. By means of really expert nursing the most severe carditis may be prevented from causing serious permanent damage. The second essential is the removal of foci of infection especially common about the mouth and nose; for infected teeth and tonsils, whether they be the portal of entry of rheumatism or not, depress the general health of children. It may not be easy to decide when a tonsil is infected, but it is certainly abnormal if the glands at the angle of

the jaw are enlarged, and such tonsils ought to be removed as soon as is possible, having regard to the general condition of the patient, because experience has shown that children with carditis who have had a complete tonsillectomy, and in whom the glands have subsequently disappeared, are definitely less likely to develop serious cardiac defects than those in whom the tonsils remain or have been imperfectly removed, or the glandular enlargement persists. Tonsillectomy after rheumatic infection has taken place will not prevent recrudescences, but it does tend to protect the heart from the more serious consequences of such recrudescences. Every measure calculated to improve the general condition of health should be employed. The child who puts on weight usually does well.

It has become the fashion to decry or at least belittle the use of salicylates, but there is no group of drugs which has so much influence over the rheumatic infection, nor is there any reliable evidence that any untoward effects follow large doses over a prolonged period. The natural course of acute rheumatism has recently been studied in America by four physicians, who watched a series of over a hundred children with the disease. These children were not treated with any anti-rheumatic measures. But the history of the condition, especially after 1876, with the introduction of salicin and the almost universal employment of salicylates in treatment, is far stronger evidence than that afforded by this brief study. To a child of 10 years it is quite possible to give 10 grains of sodium salicylate every hour, or 240 grains in each day, without any discomfort or even anorexia. To reach this dosage it is necessary to have the child at rest, to allow considerable amounts of fluid with the food, to maintain a milky diet, to ensure a regular action of the bowels, and also to give twice as much of the bicarbonate as of the salicylate of sodium. As children easily tire of any medicine it is useful to give two prescriptions with different flavourings to alternate every two or three hours, and to change the flavouring every three or four days.

Salicylates certainly abolish the pyrexia which results from rheumatism. They also remove pain from the affected joints; but they do more than this, for they produce a restoration to their normal size and contour and a re-absorption of effusions be they peri- or intra-articular. If, then, salicylates can produce such effects upon the rheumatic process in the joints, why should not the same result occur in other tissues damaged in like manner? Given in adequate doses, salicylate does influence the carditis of acute rheumatism and does modify the course and prognosis of this most serious disease. But every known measure likely to benefit the patient and raise resistance is important, and none can be neglected: tonics, vitamins, and above all a prolonged sea visit, are advisable—the latter, alas! often very difficult to arrange. And when all has been done one can only hope that the damage left by the infection will be small, and the resultant incapacity relatively slight.

Let us hope that the future has greater things in store for those susceptible to rheumatism.

V. de Lavergne and H. Accoyer (*Rev. d'Hyg. et de Méd. Préval.*, June, 1935, p. 421) state that during the period August, 1928, to April, 1934, twenty-seven cases of paratyphoid B occurred in a French battalion, twenty-six of which developed in epidemic form, while one was sporadic. Of the twenty-seven patients twenty-two had been inoculated or reinoculated, three had not been inoculated, and two had been inoculated in the incubation period, and therefore might be regarded as not inoculated. A severe attack occurred in four soldiers, three of whom had been inoculated and one not. All recovered. The cause of the infection was found to be contamination of the water supply.