EMERGENCIES IN GENERAL PRACTICE

ACUTE RHEUMATIC FEVER

BY

STANLEY GRAHAM, M.D., P.R.F.P.S., F.R.C.P.Ed.

Professor of Child Health, University of Glasgow

Rheumatic fever belongs to the group of disorders now classed as collagen diseases in which there is damage to collagen fibres and connective-tissue structures. In the last few decades there have been several slow and subtle changes in the manifestations and incidence of this disease. Although it may present the familiar picture of high fever, flitting joint pains, and prostration, it is now more often a condition with an insidious onset, unassociated with fever, and in at least 10% of cases with no history of joint involvement. Because of this variability and because most cases have their onset in childhood, the use of Miller's term "juvenile rheumatism" in preference to acute rheumatic fever has much to commend it.

Various writers have drawn attention to the fact that the incidence of acute rheumatic fever goes parallel with that of scarlet fever, and there has been a steady decline in the incidence of both diseases which began about the turn of the century. Since 1935, however, the fall has been accentuated, and it is a moot point whether or not the introduction of sulphonamide therapy into general use has played a part. In 1928 the deaths recorded in England and Wales from rheumatic fever were 1,500, in 1938 1,135, and in 1948 446. As morbidity goes parallel with mortality, these figures will serve as an indication of the extent of the diminution in the incidence of the disease. Rheumatism and its treatment and prevention, however, still remain important public health problems.

There is general agreement that rheumatism is related in some way to infection, usually in the throat, with Group A beta-haemolytic streptococci. By no means all subjects infected with this particular strain of streptococci suffer from rheumatism, but for some reason as yet unknown a subject may become "sensitized" to it: he is then a rheumatic subject and prone to recurrent attacks.

Clinical Findings

Rheumatism is very rare in children under 3 years of age and uncommon even in those under 5 years. The acute fulminating type of case is still seen occasionally and can be an emergency in general practice, but the more usual story is of a sore throat with perhaps some constitutional disturbance which is followed by a latent period of 14 to 21 days; then, with malaise, fever, and anorexia, vague joint pains make their appearance. The joints are painful on movement; there will be some slight swelling but seldom redness. The flitting and fleeting nature of this joint mischief is often marked. It responds dramatically to adequate dosage of salicylate but will also disappear without treatment.

Heart.—Carditis is the only crippling manifestation of rheumatism. The lesions may vary from mild enlargement and a mitral systolic murmur to gross enlargement with obvious involvement of both mitral and aortic valves. Presumably in every case of acute rheumatism there will be some myocardial involvement whether it can be detected or not, and the congestive failure seen in the acute phase of the disease is due to myocardial involvement rather than to valvular damage. The fully developed clinical picture of mitral stenosis with or without auricular fibrillation is rarely encountered before the fourteenth year. Pericarditis is undoubtedly the most serious of the cardiac lesions and is often

associated with other evidence of the massiveness of the attack, such as subcutaneous nodules, fever, cyanosis, and prostration. At the base of the left lung in rheumatic pericarditis there will often be found an area of dullness and intensely tubular breath sounds, the true nature of which must be recognized, as it may sometimes lead to a mistaken diagnosis of lobar pneumonia. Precordial pain, which is often regarded as a constant feature, is frequently absent in children.

Subcutaneous Nodules.—The appearance of subcutaneous nodules on the elbows, knees, malleoli, scalp, and spines of the scapulae is usually regarded as pathognomonic of the rheumatic infection, but it must be remembered that they can also be found in rheumatoid arthritis. Histologically they are roughly similar to the Aschoff nodes in the heart muscle and as a rule indicate a severe infection with severe cardiac involvement. The possibility of their persistence after all other signs of activity have disappeared seems unlikely.

Skin Manifestations.—The characteristic rash in rheumatism is erythema marginatum. It is a ringed eruption changing daily in outline, with very slightly raised margins and clear centres, and is seen mainly on the trunk and thighs. Once it has appeared it is likely to persist off and on throughout the active phase and even after all other signs of activity have disappeared. It does not signify a severe attack; indeed, one's impression is just the opposite. Erythema nodosum should not be looked upon as evidence of rheumatism. This skin lesion may appear as an allergic phenomenon in association with various infections, notably streptococcal, meningococcal, and tuberculous. When erythema nodosum is associated with rheumatism it can be regarded as evidence of the streptococcal infection rather than of rheumatism. With rare exceptions the remaining cases will be due to primary tuberculosis and should be regarded as such until proved otherwise. It should also be remembered that a rash indistinguishable from erythema nodosum may also result from the administration of sulphathiazole, probably as a result of sensitization to the drug.

Chorea.—This deserves special mention, because of all the manifestations of rheumatism it is the one most likely to occur alone and is least likely to lead to serious heart lesions. The subject, usually female, may have several attacks of chorea without apparent damage to the heart, and indeed without any other signs of rheumatism. Nevertheless all chorea should be regarded as rheumatic in aetiology. Habit spasm, in which the same movement is repeated over and over again, is readily confused with chorea and its differentiation is often a matter of great difficulty.

Differential Diagnosis

The diagnosis of acute rheumatism must be made on the history and clinical findings alone; in some subacute cases it may be necessary to delay making a definite diagnosis until the early course of the disease can be observed.

Abdominal pain during the early and acute phase of the rheumatic process may arouse suspicion of an acute abdominal condition, or an acutely tender and swollen joint may be confused with haematogenous osteitis, but in each case the history and a careful physical examination of the patient should serve to establish the diagnosis.

Anaphylactoid purpura (Henoch-Schönlein type) with its joint involvement bears a superficial resemblance to rheumatism. It is a form of primary purpura and is characterized by other manifestations such as abdominal pain, haematuria, and, of course, purpura, which should serve to distinguish it from rheumatism. The heart is not affected in anaphylactoid purpura.

Rheumatoid arthritis (Still's disease) may have an onset indistinguishable from that of rheumatism. Although they have many signs in common (arthritis, nodules, pericarditis), the course and prognosis are quite different. In rheumatoid arthritis the lesions tend to be symmetrical and the smaller joints, especially the interphalangeal ones, are more often affected so that the fingers are swollen at the base and taper towards the tip. The arthritis is not relieved by salicylates in the same way as it is in rheumatism, although it is customary for some improvement to occur. Splenomegaly and enlarged lymph nodes are found in about one-third of the cases. Furthermore, rheumatoid arthritis does not seem to be associated with infection by the Group A beta-haemolytic streptococcus as is rheumatism.

Also polyarteritis nodosa, disseminated lupus erythematosus, and occasionally subacute bacterial endocarditis may at times simulate rheumatism when heart and joint involvement are associated with pyrexia.

Treatment

Modern hormone therapy notwithstanding, salicylate, either as the sodium salt or as acetylsalicylic acid, remains the safest and most satisfactory drug in the routine treatment of rheumatism. In a recently published controlled study of the value of cortisone, A.C.T.H., and aspirin* no evidence was obtained that any of the three agents resulted in a termination of the disease, and some of the patients in each group developed fresh manifestations during the period of treatment. In the acute fulminating case and in pericarditis, however, the use of cortisone or A.C.T.H. is indicated and indeed may be life-saving. Such treatment should be carried out only under strict supervision and preferably in hospital. "Rebound" phenomena are likely to occur and may prove as difficult to control as the original manifestations, and for these salicylate will be found more valuable than the hormones.

There is no generally accepted dosage of salicylates: 1 gr. per lb. (0.14 g. per kg.) of body weight a day has been suggested, but some subjects tolerate the drug poorly and caution is required. To a child of 6 to 8 years 10 gr. (0.65 g.) of sodium salicylate and an equal amount of sodium bicarbonate every four hours would be a moderate dosage. The alkali diminishes the risk of salicylate poisoning by increasing its rate of excretion. It can be argued that smaller doses might be given without alkali. This is true, and if high blood levels of salicylate (40 mg. per 100 ml.) are desired the alkali may be omitted, but blood estimations should be carried out. Such a procedure is suitable only in hospital.

Salicylate poisoning is not uncommon. Vomiting is an early sign, although some authorities put this down to "gastric irritation," which is said to be relieved by the addition of the alkali. Tinnitus as a warning sign of intolerance or excessive dosage is valuable but is often, especially in children, either absent or unrecognized. Later, headache, hyperpnoea, and mental confusion may occur. The hyperpnoea results from the direct action of the salicylate on the centres in the medulla and is said to lead to a respiratory alkalosis. If this occurs the salicylate should of course be omitted and sodium bicarbonate given to increase the rate of excretion of the drug. For safety, it is sufficient to recommend the omission of one or two doses of the medicine if vomiting is troublesome or if there is any increase in the respiratory rate. Some children appear to tolerate acetylsalicylic acid better than sodium salicylate, and in such cases a change should be made. The dosage is approximately the same as for salicylate with alkali—namely, 10 gr. (0.65 g.) four times a day for the 6-8-year-old child and progressively larger doses as age advances. Alkali of course cannot be added. Calcium aspirin is pleasanter to take and is more soluble, but is expensive.

Modern conceptions of the aetiology of rheumatism suggest that at the onset of an attack a throat swab should be cultured for the presence of specific streptococci, and if found

a five-day course of penicillin should be given. Tonsillectomy merely because the patient is a rheumatic subject cannot be justified. Relapses occur in tonsillectomized children as often as in non-tonsillectomized children.

General Care

Nothing so far has been said about the general care and rest in bed. These are important. The duration of rest in bed for the average case is about 8 to 12 weeks, during the first four of which salicylate is given in diminishing doses. Often, however, relapses occur and the period of rest must be correspondingly lengthened. The sedimentation rate, the sleeping pulse rate, the size of the heart, and the general well-being of the patient must all be considered in assessing the amount of bed rest necessary. The convalescence will be long, with gradually increasing activity over several months. Even late in convalescence strenuous exercise causing undue fatigue should not be allowed. Hard-and-fast rules cannot be laid down and each case must be considered on its merits. On the other hand, one must avoid creating an atmosphere of invalidism, which in the long run may produce in children serious psychological problems. Residual murmurs in patients seen by the doctor for the first time need very careful consideration, and in those unfortunate subjects in whom there is severe and permanent damage to both mitral and aortic valves the decision to confine the patient to bed should not be lightly taken.

Prevention

Recent work has indicated that by the prevention of streptococcal throat infections over long periods the number of relapses and recurrences can be greatly reduced. Short courses of aspirin every two to three months were tried, but the results were not striking. The use of one of the sulphonamides in doses of 0.5 g. once or twice daily has been more successful, and the results obtained during the last war in the United States Navy were impressive. Because this form of therapy is not entirely without danger it has been replaced by penicillin: a daily oral dose of 300,000 units, given under fasting conditions to ensure absorption, has been recommended. Whether this form of prevention should be used in every case of rheumatism is arguable. Many subjects have only one attack of rheumatism (monocyclic) and in them prophylaxis may not be necessary, but in those with frequent attacks (polycyclic) and in those in whom the mischief seems to smoulder its use is probably justified. The duration of the prophylactic treatment has not yet been determined, but in view of the seasonal incidence of streptococcal infection and rheumatism it should be given during the late winter and spring months for several years if it is to be effective. Recurrences are most frequent within five years of the initial attack. The only other method of prevention is for the subject to live in an area where streptococcal infections are seldom encountered, a measure that is seldom practical.

Prognosis

Generalizations about the prognosis are not very helpful when it comes to assessing the individual case: there are too many variables. The prognosis is dependent on the degree of damage to the heart, which may not be apparent for years after the rheumatic process is apparently at an end. There are in fact two waves of mortality; the first is at its height during the first two years after the initial infection, gradually disappearing during the next three years. These early fatalities result from the overwhelming nature of the rheumatic infection. The second wave occurs in adult life, when the late effects of the rheumatic process cause serious valvular damage.

It has been estimated that 50% of patients recover from their initial attack without apparent heart damage, but 50% of these will show signs of rheumatic heart disease in later life.

Next article on Emergencies in General Practice.—
"The Acute Abdomen" (Part I), by Mr. R. Milnes Walker.

*British Medical Journal, 1955, 1, 555.