REFRESHER COURSE FOR GENERAL PRACTITIONERS

RHEUMATIC FEVER

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Since the war there has been, in most parts of the country, a striking decrease in the incidence and severity of acute rheumatism. While the cause of this change is not clear, it has led to rather greater difficulty in the diagnosis of the disease, as the classical severe attacks are comparatively rarely seen. It is important that we should be as accurate as possible in making a diagnosis of acute rheumatism in a child or young adult, since this usually means a long and protracted treatment if the utmost is to be done to minimize the risk of permanent cardiac damage. Unfortunately we have no specific test to aid us, and we are forced to rely on a careful assessment of the history and clinical findings.

History

In about half the attacks or relapses of acute rheumatism there is a more or less clear history of a sore throat or pharyngitis ten days to three weeks before the onset of the disease. This sore throat is due to infection with the haemolytic streptococcus, and may be so mild that it produces little more than 24 hours' malaise; on the other hand, it may last some days and may be associated with considerable toxæmia and, perhaps, a rash—when it will be diagnosed as scarlet fever. The severity of the antecedent throat infection seems to have no bearing on whether or not the patient will subsequently develop acute rheumatism. In the past some clinicians have referred to "rheumatic" sore throats. It cannot be stressed too strongly that the sore throat is not in itself a manifestation of acute rheumatism, it does not occur at the same time as the arthritis or other rheumatic features, and there is nothing to distinguish the sore throat that is followed by rheumatic fever from the one in which convalescence is rapid and uncomplicated.

On the other hand, a history of a sore throat during the preceding weeks offers strong support to a diagnosis of rheumatic fever. Further help in reaching a correct diagnosis may be obtained by remembering that the vast majority of first attacks of this disease occur between the ages of 5 and 15. Thus we should hesitate to make the diagnosis in a child below the age of 5 or in an adult with no previous history of the disease. Once a child has had an attack of acute rheumatism or chorea there is a greatly increased liability to further attacks. Thus if we have knowledge of a previous attack any suspicions about the present illness are considerably strengthened.

Clinical Findings

The pain of rheumatic fever is due to an arthritis. There is no convincing evidence that this disease ever gives rise to muscle or limb pain in any other way. "Muscular rheumatism," if it occurs at all, is no part of the symptomatology of rheumatic fever. From this it follows that the pain will be situated in a joint. Further, in many cases careful observation will show that the affected joint is swollen and tender. As is well known, the arthritis, even without treatment, is evanescent and "flits about" from joint to joint, each of which is involved for three to four days at the most.

Despite the name, fever may not be very pronounced (see Fig. 1) and the "rheumatic hyperpyrexia" described by earlier physicians is now rarely seen. Whether this is

FIG. 1.—Temperature chart of a child developing a fatal rheumatic relapse ten days after a haemolytic streptococcal sore throat.

due to the almost invariable use of salicylates early in the course of the disease, to the fact that other diseases associated with high fever were once confused with rheumatic fever, or whether there has been a change in the character of the disease is impossible to say. Perhaps the commonest mistake made is to regard the aches and pains which are so common a feature of almost any acute infection as evidence of rheumatic fever, especially in cases in which no obvious cause for the fever can be found. This likelihood is increased by anxiety on the part of the doctor not to miss the onset of rheumatic fever.

Carditis and Other Rheumatic Manifestations

Help in the diagnosis can be obtained from a consideration of the other manifestations of acute rheumatism, which are, of course, carditis, chorea, nodules, and rash.

Carditis may reveal its presence by evidence of enlargement of the heart—in the acute phase this usually gives rise to a diffuse apex beat, maximal beyond the mid-clavicular line; by an apical systolic murmur which may be associated with an apical diastolic murmur (the so-called Carey Coombs murmur) or perhaps an aortic diastolic murmur; and by a pericardial rub. Difficulty is caused in some cases by an innocent or "functional" systolic murmur. This usually differs from the systolic murmur met with in carditis in that it is loudest towards the base, and is, of course, not associated with other murmurs or with any sign of cardiac enlargement.

Chorea very rarely occurs at the same time as the acute arthritis and so is of little help in diagnosis in the acute stage, although a history of chorea previously is of exactly the same significance as a previous history of rheumatic fever.

Subcutaneous nodules usually appear late in the attack, commonly some weeks after the onset of the arthritis. Their main clinical importance is not so much in assisting diagnosis as in giving us sure evidence that the rheumatic pro-

cess is still active. They may be found over most subcutaneous bony or tendinous surfaces, and are particularly to be sought for on the bony points around the elbow, on the finger knuckles, over the tendons in front of the wrist (Fig. 2), on the back of the skull, and over the prominences around the knee.

FIG. 2.—Rheumatic subcutaneous nodules along the palmaris longus tendon.

The rheumatic rash is comparatively rarely seen. It is known as erythema marginatum or annulare, and these names fully describe its character. It starts as a patch of erythema which may just be palpable and spreads outwards, and as it does so the central skin which was first affected returns to normal. Thus there is formed a ring or margin of erythema which gradually extends until it finally fades away. In its fully developed phase the irregular rings of erythema are quite characteristic (Fig. 3). It

FIG. 3.—Erythema marginatum (annulare) in a child with acute rheumatism.

occurs usually in the acute phase of rheumatic fever, but comes and goes and may continue to do so for a long time. When this peculiar rash is found it is highly suggestive, if not actually diagnostic, of rheumatic fever, and its persistence should be regarded as strong evidence that the pathological process is still active.

Differential Diagnosis

Difficulties in diagnosis also arise from confusion with acute osteomyelitis. In this condition, however, the fever is generally higher, and careful examination will usually show that the pain and swelling is greatest not over the joint but at the end of one of the bones. Further, it is rare for more than one site to be involved in osteomyelitis, and, of course, the manifestation remains localized to the original site.

Occasionally rheumatic fever presents with abdominal pain, often more or less localized to one or other iliac fossa. The mechanism of this is far from clear; it may be due to an acute arthritis of the hip, but some workers think that it may represent an actual involvement of the peritoneum by the rheumatic process and regard it as analogous to the pleurisy that is rarely associated with rheumatic fever. Such cases with abdominal pain as the first symptom are particularly difficult and are often subjected to laparotomy, on the suspicion of acute appendicitis, before the true nature of the condition is disclosed by the subsequent development of a fleeting polyarthritis or signs of carditis.

Salicylate Therapy

Since we are ignorant of the actual cause of rheumatic fever we have no specific curative agent. The whole aim of treatment must be directed to the prevention of carditis or, if this develops, to doing all we can to reduce to a minimum the permanent cardiac damage.

Salicylates in full dosage will relieve the arthritis and abolish the fever within 24 to 48 hours. In fact in cases in which the diagnosis of an acute arthritis is in doubt they may be used as a diagnostic test. However, it is essential to remember that despite this symptomatic cure the all-important carditis may progress steadily. This is not to say that salicylates should not be used—they should be—but we must take care not to be lulled into a sense of false security by their effect.

There is no proved virtue in any of the special salicylate preparations so commonly recommended. Sodium salicylate or acetylsalicylic acid are still probably the preparations of choice. It will be remembered that it has long been taught that sodium salicylate should be prescribed only with the same, or even double, the dose of sodium bicarbonate, and it has been claimed that this reduces the toxic effects of the drug. It has now been shown, however, that the only effect of the alkali is to increase the excretion of the drug and that therefore in order to obtain a satisfactory effect it is necessary to prescribe larger amounts of salicylate than if given alone. Thus nothing is in fact gained. Some years ago it was claimed that even the carditis could be controlled if salicylates were given intravenously. It is now generally agreed that this is not correct, and because the danger of intravenous administration is great it is practically never justifiable.

For a child of 10 years of age 20 gr. (1.3 g.) of sodium salicylate or of aspirin should be given every four hours and continued until toxic effects appear. Some degree of deafness and tinnitus is practically unavoidable and may be disregarded. More serious toxic effects are vomiting, drowsiness, and deep respirations. If any of these develop the drug should be stopped at once for 24 hours and then prescribed in a smaller dose. The most serious complication of salicylate therapy is the development of a haemorrhagic state due to hypoprothrombinaemia. If any haemorrhages or purpura occur the drug should, of course, be stopped and the patient immediately given vitamin K (inj. menaphthone, 20 mg. intravenously), or better still a transfusion of 500 ml. of fresh blood.

Management of the Carditis

However, as has been said, all that can be hoped for from salicylates is relief from pain and fever. The carditis can be avoided or helped only by absolute rest under opti-

most conditions until we are as sure as we can be that the rheumatic process is at an end. It has long been recognized that rheumatic fever has a striking social incidence, being much rarer in the wealthy members of the community than in those not so well off. While the actual factor at fault has not been determined, it is fairly clear that the predisposition to rheumatic fever is connected with the patient's social environment. Therefore it is reasonable and desirable to do what we can to remove the child from this undesirable environment. This usually means admission to hospital. Further, despite the name "acute rheumatism," it is now well known that the disease is in fact anything but acute and commonly runs a course of many weeks or months. If a patient is to be kept at rest in bed for such a long time he needs occupation and diversion. The best form of occupation for a child of school age is some form of schooling and education. Thus arises the need for hospital schools for the treatment of children with rheumatic fever. In such schools they can rest in bed for as long as may be necessary and can have their minds occupied. Provided that the hospital school has adequate nursing and medical staff there is everything to be gained by admitting the child as soon as the diagnosis is made. Patients with rheumatic fever should receive a full well-balanced diet as soon as they can take it.

The Return to Normal Activity

As has already been stressed, the arthritis and fever are evanescent and provide no real indication of the progress of the attack so far as the carditis is concerned. The presence of rash, nodules, or chorea makes it clear that the disease process is still active, as of course does the development of any further evidence of cardiac damage. However, these are not common, and on this all-important problem we have to be guided by a consideration of various non-specific tests. The most valuable of these are the pulse rate, the weight, and the erythrocyte sedimentation rate.

The pulse is a very valuable guide to activity in most infections. However, the unreliability of the pulse in nervous or excitable patients is well recognized. For this reason we should try to take the pulse rate in rheumatic fever when the patient is asleep. A careful record of the sleeping pulse rate should always be kept. Normally this is about 10 beats a minute slower than the rate when the child is awake, but during the active phase of the disease it is raised and about the same as the waking pulse rate. As the disease subsides it falls.

A normal healthy child gains weight fairly steadily, especially when convalescent from an acute illness. For this reason a careful record of the weight every week is of great value, as a steady gain indicates that the illness is probably over.

The erythrocyte sedimentation rate is perhaps the most delicate index of a persistent infection that we have, and no child with rheumatic fever should be allowed out of bed until the sedimentation rate has been normal for a month. This is necessary because some cases of the disease exhibit what has been called a "polycyclic" course, in which the activity waxes and wanes so that a single normal reading cannot be accepted as proof that all danger is past.

After a long period of rest in bed it is necessary to return the child to normal activity by easy stages of rehabilitation. A method that has proved of value has been to wait till the sedimentation rate and sleeping pulse rate have been normal for a month and there is a steady gain in weight. The patient is then given graduated exercises while remaining in bed. Particular attention is paid to the feet in an attempt to prevent the development of flat-feet when the child first gets up. If after a month there is no evidence of relapse, more elaborate graduated exercises are performed with the child out of bed for the exercises. After a further month the patient is allowed up for half the day, and then, a month later, he is up and about the ward all day. At this stage, provided there is no gross cardiac enlargement,

the child is encouraged to lead a full normal life with games and exercises, and on discharge from the hospital school returns to a normal school life.

It cannot be stressed too strongly that the child with chronic rheumatic heart disease in the shape of some slight cardiac enlargement and a mitral lesion and with no active rheumatic infection needs no special treatment or restriction. If there is gross cardiac enlargement or any genuine restriction of activities by dyspnoea school should be permitted but not games or drill. The presence of an aortic lesion usually indicates more severe damage, but here, too, it is probably safe to allow the child to lead a normal school life provided there is no pronounced cardiac enlargement. This normal school life is extremely important from two aspects. First, it is essential that we do not occasion a cardiac neurosis by over-restricting a child with a minimal and, at this stage, insignificant cardiac lesion. Secondly, the child with rheumatic heart disease will ultimately have to compete with other children for employment. It is necessary for them to seek relatively sheltered and sedentary employment, not so much because of the immediate ill effects of a laborious occupation, but because ultimately they will suffer as a result of their heart disease. Provided the child has adequate schooling it is usually fairly easy to avoid a laborious occupation on leaving school, but it is extremely difficult for a doctor to obtain sedentary employment at 35, the age at which the patient with chronic rheumatic heart disease is likely to begin to develop symptoms. For these reasons as full an education as possible and advice concerning suitable employment are very important for these children.

To sum up the management of the child with rheumatic fever we may say, complete rest in bed for as long as there is any suggestion of persistent rheumatic infection, followed by gradual return to normal life. But, provided there is no evidence of active rheumatic infection, and in the absence of gross cardiac enlargement or genuine symptoms of early cardiac failure, the physical activities of the child with chronic rheumatic heart disease need not be restricted.

Prophylaxis

Great hopes were raised by the preliminary reports of the value of cortisone and A.C.T.H. in the treatment of rheumatic fever. There is no doubt that these agents relieve the fever and arthritis as promptly as salicylates, but whether they exert any beneficial effect on the carditis is far from clear. It is certain that the disease may, and often does, show an acute exacerbation or relapse when the drug is stopped. Hench has very aptly compared the effect of these drugs in rheumatic fever to clothing a fireman in the middle of a fire with an asbestos suit: it may prevent him from being burnt, but it will not put out the fire.

Finally, can anything be done to reduce the well-known tendency of patients who have once had rheumatic fever to have another attack? Attempts have been made to do this by preventing the antecedent haemolytic streptococcal sore throat by the continuous administration of sulphonamides or penicillin. While these measures have met with some success, they cannot be recommended for general use. Sulphonamides carry with them the risk of various toxic effects, and very large doses of penicillin are needed by mouth, while repeated injections are clearly impracticable. Our best method of attempting to deal with this problem would seem to be to do what we can to improve the environmental factors, perhaps paying particular attention to the patient's diet, and by regular routine clinical examinations in order to recognize any mild relapse as soon as possible.

In the event of any relapse of acute rheumatism, in any form, it is most important that complete rest should be reimposed and the relapse treated as if it were an initial attack.

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