however. In the initial stages of treatment it is essential to estimate the blood electrolytes, particularly chloride and potassium, at least weekly. There is a tendency to hyperchloraemia, so ammonium chloride should not be used as an adjuvant to mercurial diuretics while resins are being given. Special care is necessary when there is any evidence of renal damage. Treatment should always be started in hospital but can be maintained after discharge.

In some of our cases we have noticed that although mercurial diuretics produced little response before resin treatment, a good diuresis followed subsequent injections. Some patients who improve very little with resin treatment alone may improve when this is combined with a low sodium diet. The best combination of sodium intake, mercurial diuretics and resin treatment still seems to be a matter of trial and error in refractory cases of heart failure and it must be admitted that although the cation exchange resins

are a valuable new weapon in the control of sodium metabolism in heart failure our use of them is still rather tentative and exploratory.

In covering so broad a field as the medical management of rheumatic heart disease it is not possible to discuss details comprehensively, but I have tried to place some emphasis upon three aspects of the approach to management; first, upon the paramount importance of regular moderate exercise in maintaining general fitness, especially in the earlier stages of the disease; next, upon the need for constant vigilance in the anticipation and early recognition of complications; and finally upon the necessity to search for and treat causes of deterioration not directly connected with the heart disease. It will be obvious that in such an approach constant alert supervision by the family doctor is far more likely to ensure success than occasional survey by a consultant.

DIAGNOSIS AND TREATMENT OF RHEUMATIC FEVER WITH SPECIAL REFERENCE TO EARLY CARDITIS

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Diagnosis of Rheumatic Fever

The diagnosis of rheumatic fever is seldom wrongly made when a migratory polyarthritis and fever follow a sore throat. Occasionally it may be difficult to distinguish between rheumatic fever and other conditions, notably Still's disease, osteomyelitis, Henoch-Schonlein purpura, leukaemia and poliomyelitis. In Still's disease there may be involvement of several joints at the onset and the joint changes may be of such short duration as to simulate rheumatic fever. Furthermore, pericarditis may occur; friction may easily be mistaken for heart murmurs and effusion for cardiac dilatation. Often in early cases of Still's disease a characteristic rash consisting of discrete pink macules is present which enables the correct diagnosis to be made. Furthermore there is frequently a persistent high swinging temperature and generalized glandular enlargement. Rarely the joint involvement in rheumatic fever may be most marked in the hands, and these changes may persist for days or even weeks, suggesting a diagnosis

of Still's disease. In long-standing rheumatic fever also, there may be nodule formation over the knuckles simulating the fusiform fingers of rheumatoid arthritis; rarely ulnar deviation may result as a residual deformity (Jaccoud's syndrome).

The diagnosis between rheumatic fever and osteomyelitis may be particularly difficult. The presenting symptom in osteomyelitis may be pain in the joints above and below the site of the lesion; there may be pain in the shoulder and elbow from osteomyelitis of the humerus. On the other hand, in rheumatic fever there may be severe involvement of one joint only, often a large one such as the hip, and the signs may persist in that joint alone for several days; the differential diagnosis is then between rheumatic fever and suppurative arthritis or osteomyelitis. X-rays are of no value in diagnosis at this stage as bone changes do not occur until later. Neither is the total or differential white blood count, for there may be little or no leukocytosis in the first 48 hours of osteomyelitis and there is frequently a high white count in rheu-

FIG. 1.—Erythema marginatum.

matic fever. In cases where there is any doubt, full penicillin therapy and immobilization should be instituted forthwith and the opinion of an orthopaedic surgeon sought.

Henoch-Schonlein purpura may cause joint changes indistinguishable from those of rheumatic fever, but the diagnosis can usually be made on the appearance of the characteristic maculopapular rash with central petechiae, most marked below the knees and over the buttocks. Petechiae may, however, occur in rheumatic fever. Leukaemia may cause joint pain which, together with fever, tachycardia and anaemia, may lead to a wrong diagnosis of rheumatic fever. Furthermore, there may be cardiac enlargement and systolic and diastolic murmurs due to anaemia. Often the correct diagnosis is revealed by the blood picture, but if the disease happens to be in the aleukaemic phase, marrow puncture is necessary. Malignant disease, e.g. neuroblastoma, with secondaries in the bone may cause similar difficulty. In poliomyelitis there may be severe limb pains maximal in the joints and the case may be diagnosed as rheumatic. Not until paralysis develops is the true character of the disease revealed.

Two rheumatic stigmata that are particularly helpful in diagnosis are erythema marginatum and nodules. Erythema marginatum (Fig. 1) may develop at any time from the onset, and its appearance makes a doubtful diagnosis of rheumatic fever certain. The rash may occur anywhere on the trunk and limbs, rarely on the face. It is fleeting and may vary in extent and distribution from hour to hour. Nodules do not usually de-

velop until six to eight weeks after the onset of rheumatic activity. They are not helpful, therefore, in the diagnosis of early cases, but their appearance later is undoubted evidence of recent activity. They occur most often over the olecranon processes where they may be preceded by thickening of the subcutaneous tissue (pre-nodular thickening) for a few days or a week. Nodules may rarely occur in Still's disease but may be distinguished from those of rheumatic fever by their larger size and longer duration, as well as histologically.

An increased antistreptolysin o titre is often mentioned as an important point in the diagnosis of rheumatic fever. Although helpful it is not diagnostic as it is raised after 80 per cent. of Group A streptococcal infections. A raised antistreptolysin o titre soon after the onset of suspected rheumatic fever means that there has been a recent streptococcal infection, a point in favour of the diagnosis. In some cases of rheumatic fever, about 20 per cent., the antistreptolysin o titre is not raised.

The blood sedimentation rate is usually high at the onset and then gradually falls over a period of several weeks. Occasionally, however, it may return to normal after two or three weeks. A normal blood sedimentation rate, therefore, soon after the onset of suspected rheumatic fever does not preclude this diagnosis.

Occasionally there may be no acute joint pain at the onset but merely vague pains in the legs or in the arms and legs. Such limb pains are not uncommon in normal children but when they are

FIG. 2.—Disappearance of loud mitral systolic murmur over a period of six weeks. (L.F—low frequency; H.F—high frequency.)

accompanied by other symptoms such as poor appetite, fatigue and loss of weight the possibility of active rheumatism must be considered. In many such cases a diagnosis of innocent 'growing pains' is wrongly made, the patients are allowed to continue leading normal school lives and, when seen some weeks or months later, have well-marked heart lesions. In many young children who already have severe cardiac involvement when admitted to hospital, there is no history of previous acute joint pains, only transitory discomfort in the limbs and symptoms of general ill health.

The Diagnosis of Carditis

Having made the diagnosis of rheumatic fever, the question of heart involvement arises and must be considered from the following points of view. Are there or are there not any abnormal signs in the heart? If there are, are they due to rheumatic or to congenital heart disease? If the former, are they related to the present attack or to previous ones?

The possibility of associated congenital heart lesions must always be borne in mind lest abnormal heart signs be mistakenly attributed to rheumatism. Congenital lesions which may cause difficulty are atrial septal defect, ventricular septal defect, patent ductus arteriosus and coarctation of the aorta. In atrial septal defect there may be an abnormal apex beat, a noticeable systolic murmur and sometimes a basal diastolic murmur. The systolic murmur of ventricular septal defect may be well heard towards the apex, simulating a mitral lesion; there may also be a mitral diastolic murmur. The murmur of patent ductus arteriosus may sound like that of rheumatic aortic disease, and a mitral diastolic murmur may be heard at the apex. In coarctation an aortic diastolic murmur may be present and again there may be a mitral diastolic murmur. At times it may be necessary to perform cardiac catheterization to confirm the presence or absence of congenital lesions.

If it is thought that abnormal heart signs are due to rheumatism, it must be decided whether they are due to the present attack or to previous ones. The absence of a history of a previous attack, though helpful, is not conclusive, for there may have been unnoticed or unrecorded ones. We believe that a diagnosis of active carditis, i.e. carditis related to the present attack, can only be made under the following circumstances:

1. When murmurs either develop or disappear under observation or show a striking change in intensity.
2. When a significant change in heart size occurs (more than 1 cm.).
3. In the presence of pericarditis or heart failure.

The Diagnosis of Recent Carditis on Auscultation

The diagnosis of recent and active carditis can most often be made on auscultation; on the development, disappearance or change in intensity of organic murmurs. Organic murmurs—mitral or aortic diastolic murmurs or systolic murmurs maximal at the apex and filling systole—may develop within a short time of the onset of rheumatic fever, certainly within 24 hours, though occasionally not for several days or weeks. Their subsequent behaviour then varies from case to case; in some they may persist unchanged throughout the period of observation in hospital, in others they may become louder or softer and they may even disappear altogether. The diastolic murmurs of recent carditis are soft, short, localized and difficult to hear. The mitral diastolic murmur is heard best at the apex with the child lying on the left side, using light pressure with the bell of the stethoscope. The basal diastolic murmur is best heard to the left of the lower end of the sternum with the child supine, on expiration, and using firm pressure with the diaphragm. Murmurs that are already present when the patients are admitted

FIG. 3.—Marked diminution in intensity of basal diastolic murmur five days after admission. Further diminution 15 days later.

may later show a significant change in intensity; only then can they be related with certainty to the present attack. Thus soft aortic and mitral diastolic murmurs may increase in intensity or loud murmurs become softer and even disappear. Sometimes very loud murmurs may prove to be due to the present attack as in the following two cases. A girl, aged nine, was admitted within three weeks of her first known attack. She had then a Grade III mitral systolic murmur with enlargement and paradoxical pulsation of the left auricle. Six weeks later she had no significant murmurs and screening showed no abnormality (Fig. 2). A boy aged eight, admitted within a few days of his first known attack, had a very loud basal diastolic murmur with a marked thrill. Five days later the murmur was much softer and the thrill had disappeared (Fig. 3).

In most cases the diagnosis of active carditis can be made on the auscultatory findings; in a few it is made on other criteria—pericarditis, a significant change in heart size, or heart failure. Pericarditis occurs in about 15 per cent. of cases of rheumatic fever. Pericardial friction is usually easy to diagnose, but may be mistaken for the murmurs of aortic disease; the electrocardiogram may be normal in the absence of effusion. Pericardial effusion may be suspected clinically when a rise in temperature and venous pressure occur, together with abnormal signs at the lung bases; abnormal cardiac dullness and altered heart signs appear later (Thomas, Besterman and Hollman, 1953). The earliest radiological evidence of effusion is straightening of the left border; the

effects of changes in posture are of little value (Besterman and Thomas, 1953).

Cardiac enlargement is often quoted as an early sign of rheumatic carditis, but in our experience is absent in the majority of recent cases. In those with longstanding activity or who have had previous attacks, however, some enlargement may be present on admission and subsequent changes in heart size may indicate further activity. In interpreting films for changes in heart size the following factors must be borne in mind: the height of the diaphragms, the heart rate and the stage of the cardiac cycle—systole or diastole; an increase or decrease of 1 cm. or more is thought to be significant. Sudden increase in heart size is rare save in cases of established valve lesions and failure; many examples of so-called acute dilatation are due to pericardial effusion.

Signs of failure in children with rheumatic heart lesions indicate active carditis.

Tachycardia has often been quoted as an important sign of active carditis. We have found, however, that there is nothing remarkable about the pulse rate in the majority of recent cases unless the attacks are particularly severe. The pulse rate is seldom raised out of proportion to the temperature and it returns to normal with the temperature; this applies to both the day and sleeping pulse rates. Not infrequently there may be a sinus bradycardia.

Electrocardiographic changes are few and infrequent. The PR interval was prolonged in only 35 out of 479 (7 per cent.) cases of chorea and rheumatic fever with carditis seen here over a

period of four years. In the majority of these 35 patients the PR was prolonged for only the first month after admission and most of them had well-marked heart lesions. The QTc (QT interval corrected for heart rate; $QTc = \frac{QT}{\sqrt{RR}}$) has recently been stressed as a useful sign of active carditis (Taran, 1947; Abrahams, 1949). In a recent series of 58 cases with carditis seen in this Unit, the QTc was raised in only 13. In 27 there was certain clinical evidence of active carditis; the QTc was abnormal in nine of them (25 per cent.). The remaining 18 had carditis but there was no certain evidence that the carditis was active; four (18 per cent.) had a raised QTc. The measurement of the QTc is difficult except for someone practised in it and errors are easily made. T wave changes are rare except as a sequel to pericarditis.

Treatment

Bed rest. Patients should be confined to bed for three to four weeks after all signs of rheumatic activity have disappeared; they should have had normal weight increments, no anaemia and normal blood sedimentation rates. An exception may be made in the case of girls over the age of puberty who may have raised blood sedimentation rates—20 to 30 mm. per hour Westergren—long after all other signs of activity have disappeared. If otherwise well they should be allowed up, when not infrequently the sedimentation rates fall to lower levels. If there has been slight carditis—soft diastolic murmurs and no cardiac enlargement—10 to 12 weeks in bed is probably wise, even though signs of activity may have disappeared much earlier. With more severe degrees of carditis, often associated with continued activity, longer periods of rest are necessary.

Drugs. Salicylates are useful in that they lower the temperature and ameliorate joint swellings and pain, but we have no evidence that they influence carditis. They should be given in the maximum dosage that can be tolerated; mild tinnitus is not an indication to reduce the dose. Cortisone and ACTH are similar in their effects to salicylates; there is yet no indication that they are any more beneficial to the heart. Both drugs should be given for periods of not less than six weeks and preferably for 12. Their side effects are well known; both may cause sodium retention. Relapses not infrequently occur on stopping treatment, but usually subside spontaneously. A suitable scheme of dosage for cortisone is 300 mgm. for one day, 200 mgm. for four days, 100 mgm. for the remainder of the first three weeks, 75 mgm. for two weeks and 50 mgm. thereafter; and for ACTH, 80 mgm. for four days, 60 mgm.

for three days, 40 mgm. for two weeks, 30 mgm. for two weeks and 20 mgm. thereafter.

The *anaemia* of active rheumatism does not respond to iron or other specific therapy, but the possibility of an associated iron deficiency anaemia must not be overlooked.

Streptococcal infection. Overt clinical tonsillitis should be promptly treated with intramuscular penicillin. Ideally, and in hospital, throat and nose swabs should be taken weekly and positive streptococcal infections treated with penicillin. There is now a good deal of evidence that prophylaxis against streptococcal infection is important (American Heart Association Statement, 1953). Penicillin is the drug of choice but is expensive, and sulphonamides in the dose of 1 gm. daily are effective. Agranulocytosis is uncommon. Should it occur, prophylaxis should be changed to oral penicillin. Tonsillectomy is indicated only when there is chronic tonsillar infection; it should be carried out under penicillin cover.

Heart failure is uncommon except in cases with marked valvular lesions and cardiac enlargement. Its treatment then differs in no way from that of failure in established heart disease; digitalis, mersalyl and low salt diet. Sodium retention, despite 0.6 gm. per day sodium diets, occurs not infrequently in patients with cardiac enlargement who are being treated with ACTH or cortisone. It usually responds to mersalyl and there is seldom need to abandon hormone therapy. So-called heart failure in early cases—manifested by a rise in venous pressure, enlargement of the liver and increase in respiration rate—is usually due to pericardial effusion; there is little or no response to digitalis, in contrast to the good response seen in cases with more longstanding disease who have well-marked valve lesions and cardiac enlargement. It may be difficult sometimes to distinguish between the signs of failure and those of effusion, but in failure the temperature and blood sedimentation rate usually fall whereas in effusion they rise; and signs at the lung bases, particularly at the left and straightening of the left border of the cardiac silhouette, are common early on in effusion. No specific therapy is indicated for pericardial effusion, though when there is marked fever and tachypnoea salicylates may be effective in reducing both, often without any change in size of the effusion. There is no point in aspirating the pericardial sac unless the effusion is massive. Moreover, paracentesis is frequently unsuccessful as the fluid so quickly clots and loculates. Aspiration of an accompanying pleural effusion, if large, may be of benefit however.

After care. At the time of discharge from hospital advice is given on any limitations of physical activity that may be necessary, on the need

for prompt treatment of sore throats or recurrences of limb or joint pains and on the importance of penicillin cover for dental extractions if heart lesions persist. If sulphonamide prophylaxis has been started in hospital it should be continued for at least another two years. Patients should attend after-care clinics at regular intervals. Our own are seen at the third, sixth and twelfth month after discharge and then once each year. These examinations are of value for the following reasons. First, to establish as far as possible whether there has been any fresh rheumatic activity; the occurrence of sore throats or limb and joint pains is noted; the haemoglobin, blood sedimentation rate and weight are recorded and a search is made for nodules. If there is any question of present activity the patients are re-admitted for assessment and treatment.

Secondly, to ensure an adequate check on any change in the cardiac status they may have occurred; at follow-up clinical and radiological signs in the heart may be either more or less marked than they were in hospital and further advice on physical activity must be given. Thirdly, to re-emphasize to the patients or their parents the continued need for sulphonamide prophylaxis and prompt treatment of sore throats or limb or joint pains should they occur.

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MITRAL STENOSIS: SELECTION OF CASES FOR MITRAL VALVOTOMY

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When it became apparent that the stenosed mitral valve could be treated by surgery, clinicians immediately were faced with the problem of deciding which patients would benefit by operation. The more obvious indications were predicted from the abnormal anatomy and physiology. Others were arrived at in time by the expedient of trial and error. A number of important points are still *sub judice*.

It was soon evident that not every person with mitral stenosis was suitable for operation. Some of the earlier cases were failures partly because of the newness of the technique of operating inside the heart and partly because of clinical features which today would have contraindicated operation. In each of the first four cases, all fatal, reported by Bailey and his colleagues (1950), one or more of the following features were present: A very large heart, mitral incompetence, advanced cardiac failure, gross left atrial enlargement and bronchiectasis. The unsuitability of each of these findings will be referred to later.

The broad principles for selection laid down by the earlier workers in this field (Bailey, *et al.*,

1950; Harken, *et al.*, 1950; Baker, *et al.*, 1950) were applied to our first patients (Bedford, *et al.*, 1953). With experience, criteria were modified slightly, mainly towards including patients with features which heretofore would have been regarded as unfavourable or frank contraindications. The current basis for selection, influenced to some extent by discussion with others interested in the subject, but mainly by our observation and experience, is in close accord with the views expressed recently by Baker and his associates (1952). The term 'mitral valvotomy' refers to splitting of the mitral valve commissures by finger or knife; it is synonymous with 'valvulotomy' and 'commissurotomy' used by other writers.

Symptoms

The main indication for mitral valvotomy is breathlessness attributable to mitral stenosis. This fact needs emphasis, for patients with mitral stenosis may be breathless from other causes such as severe associated aortic valve disease or chronic lung disease.