Current Practice

CHILD CARE IN GENERAL PRACTICE

Care of Children with Chronic Heart Disease

JOHN DUNCAN HAY,* M.A., M.D., F.R.C.P.

This article deals with established heart disease in children, which may necessitate care and supervision of the child by the family doctor for a prolonged period, often throughout childhood.

In such a case the practitioner's responsibilities include, firstly, helping the child's parents—and perhaps the child himself if he is old enough—to understand, to the extent that is considered wise, the nature of his heart condition and any limitations which it may impose on him. The parents must be helped to grasp the likely long-term prognosis and how it may be modified by treatment, medical and surgical. This has become especially important now that many parents have acquired from the radio and the popular press a little knowledge, often ill-digested, of the achievements of cardiac surgery. In many cases it is necessary to stress that a long period of observation may be advisable before full investigation or surgical treatment is undertaken. So that the views expressed to parents by consultant and practitioner may be uniform the closest co-operation between them is, of course, essential, and it is particularly important that the practitioner should receive a full report from the consultant as soon as possible after an out-patient consultation or after full investigation of the child in hospital.

Such a full understanding by parents, and sometimes also by the child, of the nature and implications of his heart condition assists the practitioner in the discharge of his second responsibility—namely, supervision of all aspects of the child's life.

His third responsibility is always to bear in mind, to watch for, and to treat or arrange promptly for the treatment of infections. Important examples are a sore throat in a child with rheumatic heart disease or bacterial endocarditis, or an exacerbation of rheumatic fever. He should also watch for a deterioration in the cardiac condition as indicated by increasing symptoms or the development of cardiac failure.

Formerly, family doctors were concerned in these ways more often with children suffering from rheumatic than congenital heart disease. The reverse is now the case, congenital heart lesions in schoolchildren being approximately six times as common as rheumatic lesions. Under 5 years of age congenital heart lesions are, of course, even commoner, as six or seven out of every 1,000 babies born alive have a malformation of the heart.

The child may be brought to the family doctor by his mother because of symptoms she has noticed, or a cardiac murmur may be heard during routine examination. In either case the practitioner's first concern must be early diagnosis. The nature of the lesion should be established as soon as possible and an estimate made of its severity. Prognosis can then be deduced and treatment and care planned without delay. In some children with an organic lesion this may be essential to a satisfactory outcome; others, in whom a murmur is found to be

functional, may immediately be encouraged to lead a normal life with full activity. Moreover, an early diagnosis may contribute considerably to the parents' peace of mind.

Diagnosis

The first symptoms of cardiac embarrassment may be dyspnoea on exertion, lack of energy, or lassitude; or a baby may fail to feed well and thrive. The grosser the lesion the earlier the symptoms. Severe dyspnoea always indicates grave myocardial insufficiency, and deep cyanosis of the lips with marked finger clubbing points to a large right-to-left intra-cardiac shunt. Occasionally, however, a severe lesion causes no symptoms until adolescence or early adult life, when their appearance may be followed by rapid deterioration in the patient's condition. Lack of symptoms alone should therefore not be regarded as indicating an insignificant lesion. Physical signs are often also difficult to interpret, especially in some forms of congenital heart disease; and functional murmurs sometimes resemble those of organic lesions. An early consultation with a consultant paediatrician or cardiologist is therefore advisable in most cases.

In some children a diagnosis may be established in the consulting-room by consideration of symptoms and physical signs with perhaps electrocardiography, phonocardiography, and radiology. This applies to most children with rheumatic mitral regurgitation or stenosis, aortic regurgitation, or evidence of previous pericarditis. In many children with congenital heart lesions, however, an accurate detailed diagnosis may be achieved only by means of cardiac catheterization, angiocardiography, or other investigation for which admission to hospital will be necessary. It may be desirable to undertake this immediately or in some patients, especially infants and young children who are symptomless, to postpone it, perhaps for years. A decision concerning this is based on our knowledge of the natural history of the lesion suggested by the initial examination and on estimation of its severity and of the need for early surgical treatment. The expectation of life in the untreated patient, the results of surgical treatment, and the mortality rate after operation all vary considerably with the nature of the malformation.

Care of the Child

With the diagnosis established as accurately as is considered desirable for the time being and a decision having been reached concerning the timing of surgical treatment, care of the child and supervision of his life at home and at school may be planned. The practitioner's main aim should be to encourage him to be as active as is compatible with his cardiac capacity, in the interests of both his physical and psychological well-being. The more severe his lesions, the more he is liable to a reduced exercise tolerance, but some children with severe lesions are capable of considerable exertion without undue dyspnoea.

* Professor of Child Health, University of Liverpool; Physician in Charge of Heart Clinic, Royal Liverpool Children's Hospital.

Exercise

Though the load imposed on a child's myocardium by an organic heart lesion will cause dyspnoea on less exertion than one might expect with an anatomically normal heart, exercise resulting in moderate breathlessness is in no way detrimental to his cardiac state, provided always that he is not at the time suffering from active rheumatism or other toxic or infective process, when exertion may be harmful. Exercise, especially in the fresh air, is indeed particularly beneficial for children with heart lesions in view of the importance of their maintaining good general health and high resistance to infection. The infant and young child may be safely allowed to limit his own activity to the desired extent—he will not voluntarily induce in himself more than moderate dyspnoea. The same applies to most older children, who should be encouraged to take part in physical exercises and organized games at school unless they cause marked dyspnoea, provided that the children and their teachers understand clearly that they are free to fall out if they wish. They should, however, be discouraged from taking part in competitive athletics unless their lesions are slight. Sudden death on exertion is extremely rare in children with heart disease, but in view of its occasional occurrence in cases of aortic stenosis a child with moderate or severe degrees of this lesion should not be permitted to take part in organized games or any strenuous activity while awaiting surgical treatment.

Schooling

Schooling must be arranged to suit the child's capacity for exercise and general rough-and-tumble. In all but the severely incapacitated a normal school should be tried if it seems at all likely that he will be able to take part in school activities sufficiently to avoid his feeling out of things. Travel to and from school may add considerably to the physical strain on a child, and transport by special bus may enable him to attend a school which would otherwise be beyond his physical capabilities. For the severely affected a school for physically handicapped children provides the more sheltered existence which they require, more constant medical supervision, and education to meet their particular needs. With suitable daily transport and an adequate home a child may attend daily, but for some a residential school is more satisfactory. For those who require frequent medical or nursing care a long-stay hospital with a hospital school may be preferable. For all school leavers with chronic heart disease, education authorities take special steps to ensure suitable employment.

Home Conditions

Satisfactory home conditions may contribute much to the care of a child with chronic heart disease. If he becomes breathless on mounting stairs or is so incapacitated that his mother or father have to carry him up, a flat high up in a tenement is clearly unsuitable. Overcrowding, inadequate ventilation, and perhaps damp will predispose him to infections, especially respiratory ones, which will be detrimental to his cardiac state. In these circumstances a practitioner is fully justified in pressing the local authority to provide better accommodation for the family.

Even more important than the physical surroundings are the competence of the mother, the support she receives from her husband, and the general attitude of all in the home to the child and his disability. On these factors largely depend the practitioner's ability to care for his patients efficiently and comprehensively at home.

General Measures

Reference has already been made to the importance of early and full discussion with the parents of all the implications of

their child's heart condition (Glaser *et al.*, 1964). Of special significance to them may be the practitioner's ability to relieve them of feelings of guilt in relation to its aetiology: these are not infrequent in parents of children with congenital or rheumatic heart disease (Maxwell and Gane, 1962). It is equally important to help especially the older child to achieve a satisfactory attitude to his limitations and, in doing so, to realize the unexpressed fears and doubts which thoughts of "heart disease" may be conjuring up in his mind. The avoidance of neurosis in a child with an organic heart lesion may be as important as its surgical treatment.

In the care of the child's physical condition the practitioner's first concern in most cases will be to maintain as high a standard of general health as possible, with a view to minimizing the incidence and severity of intercurrent infections. Adequate diet, sufficient sleep and fresh air, and a sensible daily routine in the home are of the greatest importance. Some children with heart disease tire easily and should rest for an hour or more after their midday meal. They should also be encouraged to rest during play when they wish. At the same time it should be recognized that children with heart disease do not differ from their fellows in becoming bored after walking for a time with their mothers and in demanding to be carried when they are still quite capable of further exertion themselves. In such cases the mother will frequently admit that the child plays vigorously with his friends for many hours without a break. It is natural that parents of children with heart disease should be apprehensive lest insistence on firmness and discipline may be bad for his cardiac state if it demands from him a physical effort or results in his crying. In some cases this may be so, as, for example, in a child with a cyanotic congenital heart lesion, in whom crying causes a serious increase in cyanosis; but the consequences of lack of discipline are in most cases far more serious, and the majority of children with heart disease derive only benefit from consistent, firm handling.

Reference has already been made to the amount of exercise that may be advisable. An invalid chair will enable a child who is limited by dyspnoea to 100 yards' walking on the flat to be out in the fresh air, to move further from home, and so to widen his psychological horizons in a way that would otherwise be impossible.

Iron-deficiency anaemia is not uncommon in children with both congenital and rheumatic heart disease. It must be promptly diagnosed and treated thoroughly with a preparation of iron.

The risks of infection may be considerably reduced by the regular administration of an antibiotic. In a child with rheumatic heart disease, streptococcal infections and therefore recurrences of rheumatic activity will be less likely if he regularly takes, orally, sulphadiazine (1 g. daily), or penicillin V (125 mg. twice daily); or if he is given a monthly intramuscular injection of 1.2 million units of the long-acting benzathine penicillin. The incidence of respiratory infections may also be reduced in a child whose congenital heart lesion renders him susceptible to them by similar continuous administration of penicillin or other antibiotic. The risks of bacterial endocarditis after dental extractions will also be less if the child with a cardiac lesion is given penicillin (1 million units of crystalline penicillin by injection) with the premedication, shortly before the extraction, followed by 125 mg. of penicillin V orally six-hourly for three days. If he has been taking penicillin prophylactically as described above his mouth will contain penicillin-resistant organisms and the antibiotic should be changed before the extraction to tetracycline or other antibiotic to which they have been shown by culture to be sensitive.

Treatment of Cardiac Failure

By all these measures the practitioner will assist the child with chronic heart disease to lead as full and active a life as

possible and will minimize any tendency there may be to deterioration in his heart condition. Investigation in hospital and surgical treatment, if indicated, will be arranged at the time appropriate to his type of lesion and its severity. This will apply to many children with congenital heart lesions, but only occasionally to those with rheumatic mitral stenosis. If surgical treatment is successful the practitioner will be able to ensure, firstly, that the child's parents really understand that his cardiac reserve has been improved or restored to normal, and, secondly, that they are carrying out the consultant's instruction to encourage their child in future to lead a more active or fully active life.

In many cases, however, when surgical treatment is not practicable, the cardiac reserve steadily falls, the capacity for exercise lessens, and heart failure eventually supervenes. It is then necessary, in consultation with the cardiologist or paediatrician concerned, to decide whether the child should receive treatment at home or in hospital. This will depend largely on the mother's capabilities and on the home conditions. If these are adequate and there are no additional factors which require investigation or treatment in hospital, such as infection or abnormal cardiac rhythm which may have precipitated the failure, then the earlier and milder manifestations of failure may be satisfactorily and safely treated at home; but if symptoms are not relieved in a few days he should be admitted to hospital and treatment continued there until the maximum response has been attained. If home treatment is then considered advisable it may subsequently be the practitioner's task, with the consultant's assistance, to treat the child for severer degrees of cardiac failure. Most infants in failure present a difficult therapeutic problem and should be treated in hospital, at any rate initially (Goldblatt, 1962). In all cases it must be remembered that failure may have been precipitated by anaemia, which should be treated by iron and, if necessary, blood transfusion, or by infection, which will require immediate treatment.

The first essential in treatment of heart failure is to reduce the demands on the heart. This is achieved by rest, posture, and, if necessary, sedation. Complete rest in bed is necessary, with the child propped up at an angle of 50–60°, the position of minimum cardiac output, and with a pillow under his knees to prevent his slipping down the bed. A baby is best strapped to a tilting board and turned from side to side to minimize pulmonary stasis. If the child is restless he should be given morphine sulphate intramuscularly (1 mg./10 lb (0.22 mg./kg.) body weight). This is particularly helpful in left-sided cardiac failure, when it also helps to relieve dyspnoea.

When dyspnoea or cyanosis are marked, 50% oxygen should be administered in a tent, provided that the child is not terrified by being enclosed; if he is more harm than good will probably result from this measure. A high humidity in the tent will prevent excessive loss of fluid in the breath, which is particularly dangerous in babies. Feeds should be small to avoid distension of the stomach and consequent respiratory difficulty. Babies may be exhausted by sucking and may require tube feeding, perhaps by continuous gastric drip through an indwelling tube.

Digitalis is of great value, its beneficial action being due to its slowing the heart rate and thereby allowing for adequate filling and emptying of the heart with each beat, and to its increasing the force of systolic contraction. Its diuretic effect is also helpful in oedematous patients. The most satisfactory preparation is digoxin, owing to its rapid action and wide margin between therapeutic and toxic doses. It may be given in tablet or liquid form (Lanoxin). The digitalizing dose is 0.04–0.06 mg./lb. (0.09–0.13 mg./kg.) body weight in 24 hours for infants and children under 2 years of age, and 0.02–0.04 mg./lb. (0.04–0.09 mg./kg.) for those over 2. Digitalization should be attempted first with the smaller dose, divided into

four equal doses and given orally to most patients. In urgent cases a first dose of one-quarter to one-half of the total digitalizing dose may be given intravenously and followed by intravenous or oral administration of subsequent doses as indicated by the patient's response. If relief of symptoms is inadequate after 18 hours' treatment with the smaller dose, and there are no signs of digitalis intoxication, the larger dose should be tried. A satisfactory maintenance dose is usually one-quarter to one-fifth of the digitalizing dose. Early symptoms of toxicity are anorexia, nausea, or vomiting, later headache and dizziness. The pulse rate may be unduly slow and there may be premature beats, sometimes regular and giving rise to "coupling." In infants an arrhythmia may be the first indication of overdosage. If toxicity is suspected the drug should be withdrawn. If symptoms or signs are marked the child must be admitted to hospital immediately for treatment under constant E.C.G. control. Potassium chloride is given orally, 1 g. in orange juice eight-hourly, or in the very severe case, intravenously, 40 mEq in 500 ml. of dextrose in water over a period of one hour but not exceeding a total dose of 1 mEq/lb. body weight. If the urinary output is inadequate potassium should not be given. If, in the child kept at home, the manifestations cease, the drug should be restarted in smaller doses in 24 to 48 hours.

When oedema does not respond to digoxin alone, a diuretic should be given and a low sodium diet considered. First, a mercurial diuretic is administered, mercaptopimerin sodium (Thiomerin Sodium) being the preparation of choice. It is given subcutaneously or intramuscularly. A test dose of 0.1 ml. is given to exclude hypersensitivity, as manifested by flushing, urticaria, or other allergic reaction. It is followed the next day by 0.125 to 0.25 ml. for an infant under 1 year old, 0.25 to 0.5 ml. between 1 and 5 years, and 0.5 to 1.0 ml. over 5 years of age. It should be given on alternate days for several doses and then less frequently in order to avoid electrolyte depletion, which may occur when the drug is administered for more than a few days. To counteract a tendency to hypochloaemic alkalosis, which decreases the diuretic action, ammonium chloride should be given orally in doses of $\frac{1}{4}$ to 1 g. three times a day before the injection of mercaptopimerin. Depletion of potassium, which predisposes to digitalis intoxication, may be avoided by maintaining a high intake of potassium in the form of either a quart (1.1 l.) of orange juice or 1 to 1.5 g. of potassium chloride daily. When the response to mercaptopimerin is poor, it may also be improved by aminophylline, given six-hourly in the form of a rectal suppository containing 12.5 mg. for infants under 6 months old and 25 mg. for older infants and children.

A satisfactory alternative diuretic in such cases is chlorothiazide (Saluric), a benzothiadiazine, which is given orally in a daily dose of 5–10 mg./lb. (11–22 mg./kg.) body weight, though it tends to cause more electrolyte depletion than mercaptopimerin.

If an adequate diuresis has not followed these measures, the carbonic-anhydrase inhibitor, acetazolamide (Diamox) may be given in addition, orally in a dose of 1 mg./lb. (2.2 mg./kg.) twice daily on alternate days.

The value of these diuretics may be considerably enhanced by a diet low in sodium, which should be achieved in all children with persistent oedema by not adding salt to food, using milk with a low sodium content (Lanolac), and avoiding heavily salted foods, such as bacon. Fluid restriction is unnecessary and in deeply cyanosed infants and children is dangerous.

REFERENCES

- Glaser, H. H., Harrison, G. S., and Lynn, D. B. (1964). *Pediatrics*, **33**, 367.
Goldblatt, E. (1962). *Lancet*, **2**, 212.
Maxwell, G. M., and Gane, S. (1962). *Amer. Heart J.*, **64**, 449.