THE CHILD HANDICAPPED BY CARDIAC CONDITIONS*

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The cardiac causes of crippling in childhood fall into two main categories—congenital cardiovascular lesions and rheumatic carditis. There are, of course, other examples—often of great medical interest—but they are comparatively rare, and I need not mention them.

There is a general impression that over the past fifteen years or so congenital deformities of the heart and large vessels have become more common, while rheumatic carditis has diminished in frequency. It is difficult to obtain reliable evidence. In regard to congenital malformations, the great advances made during recent years in the surgical treatment of these deformities, and in anaesthesia for these patients, have focused the attention both of doctors and of parents upon this type of disability, and the hospital services for the detailed diagnosis and treatment of these cases are under increasing pressure. Certainly it would be interesting and helpful to know (1) whether the incidence of congenital cardiac deformity is increasing, because this might have a bearing upon possible means of prevention; and (2) the percentage of children suffering with congenital cardiac deformity that might be amenable to surgical treatment. Clearly this proportion is likely to increase, year by year, as surgical and anaesthetic techniques expand and improve.

Congenital Cardiovascular Lesions

Some light can be thrown upon the incidence of congenital cardiac deformity by studying the paediatric reports of maternity hospitals; but even the most detailed of these cannot give a certain answer, because the clinical signs of the handicap may not appear until the child has left hospital; and it must be remembered that a systolic murmur can be heard in 1% to 2% of all newborn babies, and may disappear again within a few weeks. In a series of 6,053 newborn infants (live births and stillbirths) reported by Richards *et al.* (1955) it was found that 7.6 per 1,000 of the live-born had congenital cardiac deformities, and 10 of these babies (26%) died in the neonatal period. At the Liverpool Maternity Hospital the incidence of congenital abnormality of all types (not cardiovascular only) is about 20 per 1,000 live births; and an analysis of the 20,339 live births in the hospital during the eight-year period 1946–53, inclusive, shows that 56 (or 2.7 per 1,000) almost certainly had a congenital cardiovascular lesion, while an additional 43 were “possible” cases. I estimate from these figures that the incidence of congenital heart deformity in the Liverpool Maternity Hospital is probably about 3 to 4 per 1,000 live births.

It is worth noting that of the 56 definite cases 13 (23%) died during the first two months of life, and that this figure compares closely with the 26% noted in the American study as dying in the neonatal period. It would appear, therefore, that about one-quarter of all congenital cardiacs born alive have very serious, and often multiple, anomalies in the heart and large vessels, and sometimes abnormalities elsewhere; and that these cases are seldom suitable for surgical treatment, especially when there are complications such as pneumonia or atelectasis.

The deaths of congenital cardiacs during the first five years of life reduce the incidence of congenital cardiac deformity to a figure that has been thought to be about 2 per 1,000 during school age, which would represent about 13,000 affected children in England and Wales. But it may be that this estimate is too small, and I am indebted to Dr. J. A. Scott, Medical Officer of Health to the London County Council, for the following information. In 1954 some 5,000 children out of a new intake of about 41,000 to the primary schools were examined to determine whether special educational treatment was necessary, and 23 cases of congenital cardiac malformations were found—an incidence of 4.6 per 1,000.

Here is a considerable problem for the medical and educational services, as well as for the affected children and their relatives. Two practical questions at once suggest themselves: (1) Is it possible to take any steps to prevent congenital cardiac deformities?; and (2) How many of the affected children can be helped, or perhaps cured, by surgical treatment? The answer to the first question, unfortunately, is no. It seems probable that many of the cases are due to genetic imperfections, though they are seldom familial. Rubella affecting the mother early in her pregnancy is a rare cause. Probably there are other maternal infections, as well as metabolic disorders and faulty implantation, that disturb the normal nutrition of the early embryo, but we do not know what they are.

Surgical Treatment

In regard to the operative treatment of congenital cardiovascular disorder, an uncomplicated persistent ductus arteriosus presents little difficulty to the experienced surgeon except in the early months of life and when the ductus is unusually wide; pulmonary stenosis (whether valvular or infundibular) is being tackled with increasing success; and operative treatment will often relieve the handicap suffered by a child with Fallot's tetralogy. Techniques are being devised and are now in use to close interatrial septal defects. Coarctation of the aorta lends itself fairly well to operative treatment, and the constricting effect of an aortic ring can often be removed.

So far we have no reliable figures giving the percentage occurrences of these various types of lesion in clinical practice, but interatrial and interventricular defects are the most frequent, followed by persistent ductus arteriosus, pulmonary stenosis, Fallot's tetralogy, and aortic coarctation in that order. Gasul and Fell (1956) have reported their findings in a series of 1,395 congenital cardiacs, 90% of whom were under 16 years of age; the patients were examined with much care, and the diagnoses were confirmed, when necessary, by angiocardiograms (850) and cardiac catheterization (450). There were also 300 necropsies. The frequency of the commoner malformations were: ventricular septal defects, 20.5%; tetralogy of Fallot, 11.1%; persistent ductus arteriosus, 10.7%; atrial septal defects, 9.6%; undiagnosed (mostly either atrial or ventricular septal defects) 8.1%; coarctation of the aorta, 5.5%; complete transposition of the great vessels, 4.3%.

Dr. J. D. Hay, who has charge of the cardiac clinic at the Royal Liverpool Children's Hospital, has kindly furnished me with the following information based upon his detailed examination of 1,324 children referred to him for his advice (in some cases his diagnosis is “provisional,” but I have grouped these cases with the “definite” diagnoses):

“Functional” systolic murmur	..	..	249
Rheumatic heart	..	..	102
Persistent ductus arteriosus	..	..	222
Ventricular septal defect	..	..	207
Pulmonary stenosis	..	..	99
Tetralogy of Fallot	..	..	93
Atrial septal defect and common atria	..	..	88
Aortic and subaortic stenosis	..	..	67
Transposition of great vessels	..	..	36
Aortic coarctation	..	..	32
Miscellaneous	..	..	129

1,324

*Read at a Plenary Session at the Annual Meeting of the British Medical Association, Brighton, 1956.

* These figures would appear to suggest—though only in very general terms—that at the present time about one-third of all children referred to a cardiac clinic may be cured or at least helped by surgical treatment.

Rheumatic Carditis

The incidence of acute rheumatism in children, as recorded in successive reports of the Ministry of Health, has been decreasing fairly steadily, though there are indications that in some areas an increase has occurred during the past year or so. The number of patients admitted to hospital annually for a particular illness cannot be taken as an accurate estimate of the prevalence of that illness in the area, but it may be a pointer. In 1943 185 children were admitted to Alder Hey Children's Hospital, Liverpool, for rheumatic infection (with and without carditis), and subsequently there was a fairly steady decline to 44 in 1950. Since then there has been a gradual increase to 63 in 1955.

Dr. J. A. Scott has informed me that in 1954 146 children were admitted to Queen Mary's Hospital, Carshalton, under the London County Council's rheumatism scheme, representing an incidence of 0.33 per 1,000 of the total school population. But it should be noted that only 30 (20%) of these 146 patients were diagnosed as suffering from rheumatic carditis. Dr. Peter Henderson, Principal Medical Officer to the Ministry of Education, has kindly given me the results of an analysis, made during 1955, of the school medical records of the 14,000 children in Oxford City schools. Only 37 (or 2.6 per 1,000) were found to have organic heart disease (including deformities) or a history of acute rheumatism. Twenty of the 37 had congenital heart lesions, and the remaining 17 had had one or more attacks of acute rheumatism; but only 6 of the 17 had permanently damaged hearts. There is little doubt, however, that in the northern industrial districts the incidence of rheumatic infection is higher than this; and it must be remembered that the report of the Registrar-General for 1954 shows that the total deaths in England and Wales, at all ages, certified as due to acute rheumatism and rheumatic heart disease, numbered 8,895.

Sir John Parkinson (1945) estimated that there were at that time at least 200,000 persons between the ages of 18 and 41 years in Great Britain suffering from acquired heart disease—mostly rheumatic, and usually contracted during childhood. This estimate is supported by Kerley's (1949) observation that in the mass radiography of 3,000,000 adults during the period 1945 to 1948, 4 per 1,000 were found to have undoubted radiological evidence of rheumatic carditis, and he thought that the correct figure would probably be nearer to 6 or 7 per 1,000. At the same time, Kerley noted that 20% of the cases detected were unaware of any disability and gave no history of rheumatic infection. Even if the incidence of acute rheumatism in childhood is only 0.3 per 1,000 children of school age (as indicated by the statistics from areas where acute rheumatism is notifiable and shown in the Ministry of Health Report, Part ii, for 1954), the total number of new cases in each year must be about 2,000. We do not know what proportion of these patients will develop permanent carditis, but we can be sure that even the present incidence of rheumatic infection provides a terrible load of mischief for a nation to carry.

Prevention and Treatment

Is it possible to prevent not only the first attack of rheumatic infection, but also the recurrences which are so liable to occur when the individual's defences have been breached, especially during the early years following the initial infection? On this subject a committee of the American Heart Association (1955) has prepared a statement stressing the paramount importance of controlling group A haemolytic streptococcal infections and claiming that adequate and early penicillin treatment over a period of 10 days will eliminate streptococci from the throat and will usually prevent an attack of rheumatic fever. The dose recommended is one

intramuscular injection (600,000 units) of benzathine penicillin G; or one intramuscular injection of procaine penicillin (300,000 to 600,000 units) in oil with 2% aluminium monostearate, given every second or third day for three doses. Oral penicillin is also recommended in a dose of 250,000 units three times daily for 10 days. For the prevention of rheumatic recurrences in subjects who have had the initial attack (whether rheumatic fever or chorea or heart damage) it is suggested that a daily dose of 0.5 to 1 g. of sulphadiazine be taken each morning, throughout the year, and continued for years. Alternatively penicillin may be used, either by mouth in a dose of 200,000 to 250,000 units once daily, before breakfast; or as a monthly injection of 1,200,000 units of benzathine penicillin G.

Markowitz and Hemphill (1955) found that a daily dose (oral) of benzathine penicillin G, 200,000 units, was more effective than a sulphonamide in preventing rheumatic recurrences and caused fewer toxic reactions. When rheumatic patients are in hospital wards, and therefore in possible contact with streptococcal carriers or patients with active streptococcal infection, particular attention must be paid to the accepted techniques for avoiding cross-infections.

Those who may feel opposed to ordering a prolonged course of either sulphonamide or antibiotic treatment—and I confess that I am somewhere on the fringe of this group—should remember the sinister character of rheumatic recurrences, which are often initiated by quite mild recurrences of streptococcal infection; yet they invade the heart with ever-increasing damage, as a river in mounting flood erodes its banks. In practice one frequently finds that a prolonged course of prophylaxis breaks down because the parents forget to give the drug regularly. A less common difficulty is that the patient shows toxic reactions. Fortunately, there is at present little evidence that beta-haemolytic streptococci become resistant to penicillin.

Another important aspect of the prevention of rheumatic infection must be mentioned under the comprehensive heading of "home circumstances"; for there is little doubt that social status, poverty, housing conditions, overcrowding, and the prevalence of respiratory infections play a part in determining the incidence of rheumatic infection.

Care of Cardiac Cripples

To make adequate provision for the care of cardiac cripples—whether due to congenital cardiovascular deformity or to rheumatic infection—we must consider the needs of each individual patient from a number of angles. This is, of course, true for all types of handicap. A civilized community takes care of its weaker members; it considers their happiness, their medical attention, their education, their training for a suitable occupation, their future economic position, and their spiritual needs. It gives thought to the human and financial strains that may develop in a household that includes a handicapped child, and to the wider social and economic implications of crippling. It makes plans to prevent, in every way possible, the cause or causes of the disability. It should, moreover, keep very much in mind the duty of each individual, whether healthy or handicapped, to make some contribution to the welfare of society. Perhaps the most important, and certainly the most difficult, task of a community in accepting these obligations is to administer its benefits and services in such a way that they do not undermine the sense of responsibility of each family group to face its particular problems bravely and intelligently.

It needs little imagination to realize that a child crippled by cardiac disease or deformity is an individual to whom the handicap is little short of a tragedy, for it interferes with the activity of a full normal life. Depressing psychological reactions are almost inevitable, but they may be modified by understanding and appropriate management. This point was well brought out by Dr. Morris Fishbein (1955) in the following words:

"The poet John Donne wrote that no man is an island. Never was this more apt than when applied to the person who is suffering with a crippling disease. For him isolation is disastrous. If intelligent and left too much alone, he becomes embittered, sullen and perhaps an enemy of society. If unintelligent, he deteriorates slowly and steadily into a parasitic, vegetable-like existence, which may eventually sap the lives of all about him. The modern approach to the problem emphasizes adjustment of the handicapped person to the family, the community, and even the nation. All these forces are brought to bear from the social, economic and psychologic points of view, so that such a person is able to live an existence as nearly normal as possible. He contributes to rather than lives upon the society that encompasses him."

The adjustment to which Dr. Fishbein refers should be, in fact, mutual, at all events in respect of the handicapped child and his family, and the doctor should seek every opportunity of explaining to the child's parents the nature and extent of the patient's crippling, so that they may be the better able to accommodate themselves to the situation. This medical opinion should be based mainly upon the child's functional incapacity; we have all known patients who have been condemned to live a life of invalidism when, in fact, they were well capable of enjoying a more active life. Complete rest is essential during recurrences of rheumatic carditis; but subsequently it should be remembered that in childhood the heart, like most other organs, possesses a remarkable capacity for recovery, and that with graduated exercises and physiotherapy there are many cardiac patients who can be gradually promoted towards a more normal life. In short, the doctor should maintain an optimistic outlook, using as yardsticks of the patient's condition such tests as heart rate, liability to dyspnoea and fatigue, diminution of oedema and venous congestion, and the child's appetite and feeling of well-being.

Institutional or Home Care

The pros and cons of institutional as against home care need careful thought in each case separately. Institutions for cardiac cripples have proved their usefulness, particularly for children whose home conditions are unfavourable. In addition to the medical and nursing care that the children receive in them, education can be provided without difficulty, physiotherapy and occupational therapy can be made available to them, and group interests, like games, concerts, television, etc., can be satisfied more easily than when the patient is in bed at home. A library service is another much-appreciated recreation.

On the other hand, institutional life has its disadvantages, especially for those children who seem predisposed to develop the invalid outlook; and is it not true that for most children "there's no place like home"? The family doctor who is well acquainted with the home situation is in the best possible position for giving advice to the parents, and he is able to call upon the ancillary services provided by the local authority, such as home nursing, home helps, health visitors, and so forth. In particular, he knows the child and the parents as individuals, and he can ensure that the various services available to the child are suitably balanced. The need for overall and continuing supervision of the handicapped child by one doctor—the captain of the team—is properly receiving emphasis in these days when so many different services, often specialized, are available. It is a need which certainly holds good for the cardiac cripple.

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SADISM AND POWER

A PHYSICIAN ON DE SADE

"Unlimited power is apt to corrupt the minds of those who possess it," the first Earl of Chatham told the House of Lords in 1770. The same moral was drawn by Dr. A. H. DOUTHWAITE, senior physician to Guy's Hospital, when he spoke on sadism at a meeting of the London Jewish Hospital Medical Society on January 31. Dr. Douthwaite used to illustrate his theme the lives of the Marquis de Sade, the eponym of our words sadism and sadistic, and of Gilles de Rais, de Sade's evil inspiration. Both men, he said, had possessed great power, largely by virtue of their wealth.

Basis of de Sade's Reputation

Sadism and sadistic were words used freely by both the profession and laity, said Dr. Douthwaite, yet many of those who did so knew little of the man from whom they were derived. The Marquis de Sade (1740–1814), the son of aristocratic parents, had shown himself a brave soldier as a young man, attaining the rank of lieutenant-general at the close of the Seven Years War; and his sexual appetites at first had been normal. But at the age of 23, threatened with imprisonment if he disobeyed, he was forced by his father into a distasteful marriage with the elder daughter of the de Montreuil family. The tragedy was sharpened by the fact that the woman de Sade wanted to marry was his wife's beautiful sister, Louise, whom later he was to seduce, carry off to Italy, and debauch there. It was his wife's large dowry that made possible the excesses of his subsequent life. These landed him often in prison, and although on occasions de Sade indulged in abnormal sexual practices the latter were not so gross or frequent as to justify the universal execration in which his name was now held. It was his books.

Living in the Bastille in great comfort, thanks to the lavish table provided for him by his mother-in-law, de Sade in 1784 wrote his most notorious work, *Cent-vingt Journées de Sodom*, whose three volumes he completed in 37 days. It was kept in the hands of his family until its publication in 1904. The book told of a group of aristocrats who used their extreme power and wealth to abduct men, women, girls, and boys to a castle where was practised every conceivable sexual abnormality amid bouts of extreme cruelty. It was a most boring and repetitious exercise in pornography. Other books followed, of which *Justine* was the best known. It was on his pen, much more than on his deeds, that de Sade's posthumous reputation rested. Of his 74 years of life, the first 28 had been unexceptionable for a rich young man of his times. The last 36 years were spent largely in prison, and of the ten years when he was free to practise the cruelties attributed to him—many of them exaggerated—de Sade was in exile for at least four.

Influence of Gilles de Rais

What influence, then, had led de Sade to imagine atrocities he could never have practised? The answer, said Dr. Douthwaite, lay in his reading. De Sade had enjoyed Suetonius's lives of the Roman emperors and revelled in the cruelties of those days. But above all he had been fascinated by the shocking cruelty and unbridled sexual abnormalities of Gilles de Rais.

Gilles de Rais, Marshal of France, born more than three centuries earlier than de Sade in 1404, was like him in his wealth, his aristocratic origin, and his personal bravery. Like de Sade also he had studied the lives of the Roman emperors. But de Rais was a sadist in the fullest sense of that term.

As a young man Gilles de Rais had earned a good name for his behaviour at court, and when in 1429 Joan of Arc had appeared there he supported her cause. He went further still. He was her squire at Orleans, where, although wounded,