Section of Medicine

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DISCUSSION ON THE COURSE AND MANAGEMENT OF CONGENITAL HEART DISEASE

Professor C. Bruce Perry: In this discussion on the course and management of congenital heart disease, consideration should be limited to those types of congenital cardiac defect which are compatible with life and are recognizable clinically. Therefore, I propose to exclude both gross malformations, such as ectopia cordis, and minor defects, in some cases none the less important, such as bicuspid aortic valves.

In considering the prognosis and treatment of cases of congenital anomalies of the heart and the great vessels, which produce clinically recognizable symptoms and signs, we immediately meet two great difficulties. The first is that in many cases it is impossible from the physical signs and symptoms to diagnose accurately the anatomical defects present, even when we are assisted by radiology and electrocardiography. The other difficulty is that the influence of the cardiac abnormality on the patient's life varies widely according to the defect present. We therefore need a more general classification based on symptoms, one which does not, in the first instance, require an accurate diagnosis of the anatomical defect, but which is based on the physiological disturbances produced by the abnormal anatomy. For this clinical classification we are indebted to Maude Abbott (1924 and 1929) whose studies have given us a valuable working conception of congenital heart disease. She divides the cases into two large groups, those with cyanosis, and those without. This classification must be the basis of any attempt to study the course and management of congenital heart disease. It is helpful further to subdivide the acyanotic cases into those in which there is no abnormal communication between the venous and arterial sides of the heart—such as coarctation of the aorta and subaortic stenosis—and those in which there is an arteriovenous shunt. The latter cases normally present no cyanosis, since the only result of the abnormal communication is that some arterial blood is sent to the lungs via the pulmonary artery, as occurs in a case of a patent interventricular septum or patent ductus arteriosus. Such cases, however, may develop cyanosis—*cyanose tardive*—if any pathological process so alters the blood-pressure on the two sides of the heart that the pressure in the right ventricle or the pulmonary artery exceeds that in the left ventricle or the aorta.

Taking this classification as our basis, we find that in the truly acyanotic cases, the malformation produces little or no effect on the activities of the patient, who suffers no disability and may lead a normal life. Some of these patients, particularly those with coarctation of the aorta, appear to be rather liable to sudden death, although the condition is obviously compatible with fairly long life, as shown by Lewis's (1933) series and by Reynaud's (1828) patient who died at the age of 92, having suffered no apparent inconvenience. The great danger for the patient with acyanotic congenital heart disease is the ever-present possibility of developing bacterial endocarditis. Of 60 cases of coarctation of the aorta collected by Maude Abbott, (1927) 12, i.e. 20%, developed a bacterial infection at the site of the constriction. This serious accident is not confined to acyanotic cases; Abbott found 90 cases of bacterial endo-

carditis amongst 395 acyanotic cases (i.e. 22%), while the incidence in cyanotic cases was 39 in 291 cases, i.e. 14%. The probable cause of this difference is merely that the acyanotic patients live longer, and therefore run more risk of superimposed infection. This danger indicates the one great point to be considered in the management of cases of acyanotic congenital heart disease. Patients with this type of congenital heart disease need no restriction of their activities, but every effort should be made to eliminate any possible source of infection. Sepsis in the mouth and upper respiratory tract as Okell and Elliott's (1935) researches have shown, are potent sources of bacteræmia such as might easily lead to an engrafted endocarditis. Apart from this, it is very important that these patients should lead a perfectly normal life. If the position is not simply and clearly explained to the parents, the child may be so pampered and coddled that he or she develops a serious cardiac neurosis.

Those cases with an arteriovenous shunt, which may develop *cyanose tardive,* such as patent interventricular septum and patent ductus arteriosus, need the same management and have the same outlook as the truly acyanotic cases, with the exception that pulmonary disease or infection may be rendered more serious than it might otherwise be by a reversal of the shunt and the development of severe cyanosis. Children with this type of congenital defect, like those with no abnormal communication, should be encouraged to attend school and do games and drill normally, in fact to lead a full and unrestricted life.

When we turn to the cyanotic type of case, of which the commonest group of clinical significance is probably that with a patent interventricular septum, dextroposition of the aorta and pulmonary stenosis—the "Tetralogy of Fallot", we find that the one guide in the treatment and to the prognosis is the depth of the cyanosis and the severity of the symptoms. The earlier in life that cyanosis develops, the more serious is the outlook. Broadly speaking, we may divide the cyanotic cases according to the relationship of the onset of cyanosis to the three critical periods of growth. Those patients who show permanent cyanosis at, or soon after, birth rarely survive five years. If cyanosis is delayed, but appears at the beginning of school life, at five years, the patient will probably not reach puberty. While those who develop permanent cyanosis only at or after puberty, as may happen in the case of a patent interventricular septum with dextroposition of the aorta but no pulmonary stenosis (the Eisenmenger complex), may live a comparatively long, even if restricted, life.

For treatment we can do nothing radical to relieve the condition, and the patient's activities must be determined entirely by his capabilities. While cyanotic patients obviously cannot play games at school or undertake laborious occupations, they may yet lead useful lives, as illustrated by White and Sprague's (1929) musician who lived until his sixtieth year. It is important to remember that the preventive care which must be given to the acyanotic cases, in an effort to prevent the development of bacterial endocarditis, should also be afforded to those with cyanosis.

One other point that I should like to raise in the hope that this discussion may increase our information about it, is the relationship of pulmonary tuberculosis to congenital heart disease. Rokitansky (1855), one of the first students of congenital heart disease, suggested that cyanosis was incompatible with pulmonary tuberculosis, whereas Peacock (1866) shortly afterwards described nine cyanotic cases with pulmonary tuberculosis. This difference of opinion has persisted to the present time. Personally, I wonder how many of the cases labelled pulmonary tuberculosis were really cases of superimposed bacterial endocarditis with pulmonary infarcts; possibly this is a biased view, due to the fact that I have seen one such case sent to a sanatorium as tuberculosis, but there was no tuberculosis, the pulmonary symptoms and signs being due entirely to multiple emboli. Gloyne (1936) has analysed 21 cases of congenital heart disease found among 2,735 post-mortem examinations at the

Victoria Park Chest Hospital. He found that seven of the 21 had pulmonary tuberculosis. In four of these there was no mention of cyanosis; in the other three the notes were lost, but the anatomical defect was in every case one that was unlikely to be associated with cyanosis.

Most of my experience of congenital heart disease has been derived from a study of the condition as seen in school children, and it bears out the main points that have already been mentioned. The majority of these cases are of the acyanotic type. The commonest defect is a patent interventricular septum or *maladie de Roger*, with a loud systolic murmur over the centre of the præcordium, often associated with a palpable thrill. The 87 children with this defect that I have seen, all lead perfectly normal school lives, with no restriction and with no symptoms referable to the heart. The oldest patient with signs of a patent interventricular septum that I have seen was aged 74. Very occasionally a patent interventricular septum is combined with a complete heart-block. In the only case that I have seen with this combination of congenital defects the heart-block appeared to have produced no symptoms. One child has died of bacterial endocarditis occurring on the lateral wall of the right ventricle at the site of impact of the stream of blood passing through the arteriovenous shunt. The hole in the septum in this case was surrounded by a ring of fibrous tissue and looked almost as though the septal defect was being slowly obliterated. I have notes of two cases in which at the ages of 4 and 6 respectively, signs suggesting a patent interventricular septum were found. Both children have been examined every twelve months for five years, and the murmur, at first persistent, became gradually less marked, and at the last two examinations the heart appeared normal. In one other case the patient when first seen, at the age of 4, had a well-marked central thrill, but seven years later the physical signs had so decreased that there is now only a soft indefinite murmur audible over the centre of the præcordium. Muir and Brown (1934) have suggested that this disappearance of the physical signs may be due to the fact that the opening in the septum does not increase in size as the child and the heart grow, and that an opening, which in the small heart of an infant might be considerable, would be negligible compared with the heart in later childhood or adult life. At first it did not seem possible for a congenital defect to correct itself, but the appearance of the opening in the case in which the patient died of endocarditis was so suggestive that I began to wonder if possibly this might happen. French (1918), Parkes Weber (1918), and Stamm (1918) have also reported cases in which the typical central systolic thrill and murmur of a patent interventricular septum have disappeared. Two of the children with a patent interventricular septum developed acute rheumatism, and one of them died from carditis. It is difficult to see how a congenital cardiac defect can in any way predispose to acute rheumatism, but it is interesting, in view of the frequent occurrence of rheumatic heart-disease amongst the cases of patent foramen ovale collected by Roesler (1934). Possibly this discussion may provide further information on this point.

The other common congenital cardiac defect found in school children is that without symptoms or cyanosis but with a systolic thrill and murmur at and above the pulmonary area. It has been suggested that if in such cases the pulmonary second sound is audible and radiology reveals a prominent pulmonary artery, the defect is really a patent ductus arteriosus. This is not so, however, as has been pointed out by Hochsinger, and Miller and Orton (1913). In one such case of mine the patient died of bacterial endocarditis, had a very prominent pulmonary arc radiologically, and post-mortem examination showed that this was due to considerable dilatation of the pulmonary artery beyond the constriction. Forty children with these physical signs lead perfectly normal lives with no disability.

Children with the less common subaortic stenosis, with a systolic thrill and murmur at the aortic area conducted up into the neck, of 10 of whom I have notes, also suffer no disability.

In 4 out of 17 cases of patent ductus arteriosus, however, the patients were unable to take part in school games because of easily induced fatigue and dyspnœa. These children were rather slenderly built and I suspect that their symptoms were due to a certain degree of aortic hypoplasia, such as is sometimes associated with this anomaly and is not actually due to the patent ductus arteriosus itself.

To sum up: The prognosis in acyanotic types of congenital heart disease is good. The patients should live normal lives, but care should be taken to avoid superimposed bacterial infection. Patients with cyanotic heart disease should lead restricted lives. The degree of restriction desirable and the prognosis depend almost entirely on the severity of the cyanosis and other symptoms.

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Dr. Terence East: Professor Perry has suggested the best lines on which to approach this question. Unfortunately the patients with congenital lesions of the heart are uncommon, and we rarely have the opportunity of following them. We can only try to gain some composite picture by blending the manifestations observed at the various ages at which one sees each type.

The diagnosis of the nature of the lesion can usually be made with a fair degree of accuracy nowadays, as far as concerns those defects which permit survival after the first few months. Before that age it is, as a rule, impossible to make the diagnosis.

From a general point of view the division of the cases into cyanotic and acyanotic is of considerable practical value. The presence of cyanosis depends upon two fundamental causes: (1) The hindrance of access of the blood to the lungs, and (2) the other is the presence of a right-to-left shunt allowing the unoxygenated blood to return into circulation. The grade of cyanosis is a fair indication of the importance of the part that either or both of the factors are playing. Atresia of the pulmonary artery is the extreme example of the former factor. In some of these cases the blood reaches the lungs through a patent ductus. In one case in which I carried out the autopsy the ductus was closed and the blood reached the lungs by very large bronchial arteries. There was a large hole in the septum of the ventricles. The patient had worked as a dental mechanic, and died at the age of 33. In this case no murmur was heard. Another man was well known in the hospitals a few years ago, and died at about the same age, with a similar atresia of the pulmonary artery. These two cases are instances of the possibility of survival with gross disability. Patients with the tetralogy of Fallot sometimes live long. One such patient is now 49, but lives a restricted life. Another survived an attack of lobar pneumonia a few years ago, and is now getting married, at the age of 33. Two

patients have died of pulmonary tuberculosis as young adults. There is no doubt that in any cyanotic case the patient must lead a very restricted life, and this life is not likely to be long. One cannot say which cases may do better than others, but the long-lived exceptions offer grounds for hope. The period of active growth at puberty is a critical time ; naturally, a lesion that may be tolerated in the child is likely to be impossible for the adult.

Of the acyanotic lesions, patency of the interventricular septum seems to be the commonest. In one case the murmur had disappeared at about the age of 4 years, and one may presume that the hole had closed or that the flow of blood through it had ceased. The smallest holes appear to cause the most noise ; probably a very large hole causes no murmur. The murmur is sometimes described as being present in diastole as well as systole. Laubry, Routier, and Soulie have described an associated defect of the adjacent aortic valve which causes aortic incompetence. Such a combination would be more serious than a simple patency of the septum. It would be less well tolerated and more liable to infection. Perhaps the cases with this combined defect are those with murmurs in systole and diastole. Although patent septum is apparently only rarely associated with that uncommon abnormality, congenital complete heart-block, yet patients with this defect often have an open septum. The reason for this association is obscure. It seems that the ventricular rate is often high in these patients, and they probably suffer little disability as far as the block is concerned.

In a series of 15 cases the patients, all female, have a systolic murmur in the pulmonary area, usually but not always, accompanied by a thrill. The pulmonary second sound is loud, and the closure of the valves palpable. X-rays show that the pulmonary artery is enlarged. There are no other abnormalities. The ages range from 7 to 52, and seven are over 35. Their lives are not affected, so the lesion cannot be important. There is no need to restrict them. There has been no post-mortem examination on such a case. It seems hardly likely that a patent ductus is present. It is true that in children the murmur is said to be confined to systole, but it is doubtful whether it is so in adults. Perhaps they are similar to those cases described by the opener, with a slight degree of stenosis below the valves, and an enlarged artery above.

Bicuspid aortic valves are of no importance in themselves. Their liability to infection makes them significant later on. In the later years of life there are cases of aortic stenosis with fused calcareous aortic valves. One wonders whether some of these conditions begin as congenital abnormalities. When the stenosis develops there is, of course, a strain on the left ventricle and the patient must be protected from over-exertion.

Coarctation seems to be as well tolerated as any congenital defect. As the patient will probably die as a result of it, some limitation of activity seems to be indicated, in adults at any rate, especially as years go on, and other defects are likely to appear.

As all patients with congenital lesions of the heart are liable to cardiac infections, it seems wise to keep them clear of septic foci. Unfortunately the infection creeps in very insidiously. It may begin as an apparently trivial bout of influenza. Special care is needed for these patients in any mild febrile attack, as it might indicate the escape into the blood-stream of dangerous organisms.

The question of pregnancy may sometimes arise. There does not seem to be any contra-indication in patent ductus. A patient with a patent septum would probably be safe, provided there was no enlargement of the heart ; otherwise, there might be some risk of a right-to-left shunt in the later stages. One would imagine that a cyanotic patient should not run the risk of pregnancy.

Persons with congenital heart lesions occasionally propose themselves for life assurance. An acyanotic case, a case of patent septum, or one of patent ductus might be accepted if the lesion were perfectly tolerated. One would have to impose

a heavy premium to cover the risk of infection, and only recommend acceptance for a limited term of years, not exceeding the age of 40 or thereabouts.

The acyanotic lesions are evidently well borne in youth, but so, very often, are acquired lesions. In the adult body the defect is apt to cause more trouble. In any large group of children with heart lesions the congenital variety turn up fairly often. One gets the impression that adults are seen far less frequently, whether the patient comes as a heart case, or is suffering from something else. By the age of 40 they are rare. Special care is needed in any case of congenital heart disease to protect the patient from infection. As regards activity, acyanotic patients can usually do a good deal, but they should be guarded from extreme forms of exertion. It would be wise to forbid all forms of competitive sport in youth, and laborious occupations later on.

Dr. D. C. Muir: My original group of cases consisted very largely of school children between the ages of 5 and 15. Additions have since been made at both ends of this age-period by the inclusion of babies from municipal clinics and institutions, of children passing out from school into the labour market, and of adults discovered in various ways.

In the very young it has been extremely difficult, or impossible, to make anatomical diagnoses or even to say whether a non-cyanotic baby may not later be found to have a lesion of the cyanotic type. The impression that the *maladie de Roger* may be regarded as a benign lesion during the school period has been fully confirmed. There has never been any reason to regret allowing these children a normal unrestricted school life. Repeated examination over years has not shown any strain on the heart—as revealed by the preservation of a normal contour, a physiological electrocardiogram, and absence of symptoms. Several children have gone through acute illnesses, including pneumonia, without ill-effect or production of cyanosis.

The disappearance of physical signs noted by Weber and others has been observed twice ; in one case a marked thrill and bruit gradually disappeared over a period of seven years. The only undoubted dramatic manifestation seen in the whole group was one case of very severe chorea, requiring four months' treatment in hospital. There were no cardiac sequelæ. The only precaution taken in cases of *maladie de Roger* is to pay particular attention to any possible foci of infection.

The same latitude is not allowed in cases of patent ductus arteriosus. Here there is a more definite stress on the heart, as shown by a dilated pulmonary artery. The aorta is generally hypoplasic and further pressure alterations occur more readily where the shunt takes place above the valves of the heart. Symptoms are more frequent, though not common during the school period. Later in life they may be severe and the activities of one male patient are greatly restricted by a pain which, in its severity and distribution, is typically anginal. Infective endocarditis is more common than in the *maladie de Roger*, and one of the original patients is at present in a sanatorium with active pulmonary tuberculosis.

Absence of symptoms with marked physical signs is characteristic of subaortic stenosis, cases of which have required no special management during the school age. There would seem to be a still greater risk of infective endocarditis. Of four deaths, one occurred suddenly while the patient was cycling, one was due to pulmonary tuberculosis, and two to bacterial endocarditis.

Dr. J. W. Brown : In a group of 291 cases of congenital heart disease attending at five clinics there are 57 (19 per cent.) cases with cyanosis. All the patients in this group are not seen because a certain number are exempted permanently from school or are inmates of various institutions.

The cyanotic patient is generally at least as intelligent as his normal fellow and, given the same educational opportunities, can hold his own at school. It is a grave

mistake to place these children in special schools for the mentally defective and backward, for by so doing an inferiority complex is developed. Each case is considered on its merits and as far as possible the patient is encouraged to go to school, if only for part time. If taught to read and write, these children can at least amuse themselves and later, in some cases, undertake sedentary work. Once a child has been excluded from school it is difficult to get him back and exclusion only leads to invalidism.

Clinical experience suggests a revision of ideas as to prognosis in this group. These patients may survive very much longer than is generally supposed, and observation of individual cases over years has shown that the degree of cyanosis presented remains remarkably constant. The blood-count, which unfortunately has not been ascertained as a routine, may be as high as twelve millions, and this figure does not indicate a rapidly approaching end, as is sometimes taught.

The infectious diseases of childhood—measles and whooping-cough—figure largely in the histories of these children, and are surprisingly well tolerated. Scarlet fever appears to be a dangerous complication and two patients have died from infective endocarditis as the result of this disease. Tuberculosis occurred in four of the elder cases.

In each case the patient had been weighed and measured at the clinic and only a small percentage are below the average in height and weight. Nearly all the cases have been examined with X-rays and a routine electrocardiogram has been taken. These methods of examination have proved helpful. In this the most obvious group of congenital cardiac defects and nearly always with gross interventricular septal defects, no case of congenital heart block has been found.

An interesting result of electrocardiographic examination has been the discovery of 12 cyanotic patients with normal or left axis deviation in the absence of dextrocardia or gross conduction defect. Autopsies in three of these cases have shown tricuspid atresia in one, and a large interauricular septal defect in the other two. The X-ray picture in this latter small group differs from that of the tetralogy of Fallot, which is distinctive.

A case of peculiar interest was that of a woman aged 29, admitted to hospital on account of infective endocarditis. A congenital history was clear. There was no cyanosis or clubbing other than the parrot-beaking of the nails encountered in infective endocarditis. On the physical signs a diagnosis of a patent ductus, with infective endocarditis, was confidently made. The electrocardiogram showed right-axis deviation but the patient was too ill for X-ray examination. Autopsy revealed a typical Fallot's tetralogy, with the pulmonary artery dilated and filled with vegetations distal to a constricted bicuspid pulmonary valve.

Congestive failure is rare in the cyanotic group, as in other groups. There was one case of interauricular septal defect with auricular fibrillation.

Treatment is rarely necessary in this group. Instruction must be given to the patient and his parents as to the amount of effort that can be undertaken with comfort. Digitalis and similar preparations are of no value. Oxygen is of little use, owing to the large shunt and dextraposition of the aorta present in the bulk of these cases. Bleeding may occasionally be of value as a temporary measure.

Supervision, of an effective and friendly nature, seems to be the most desirable factor in the management of these patients and can do much, not only to dispel the air of fatality that surrounds them, but also to increase their happiness, particularly if some small measure of participation in school activities is allowed.

Dr. A. G. Gibson said all would agree that in congenital heart disease the maximum activity should be allowed. He doubted, however, whether competitive athletics in adolescence or adult life were always compatible with safety. In one of his cases, a schoolboy aged 15 who had a good record in his school sports, was found

in an intercurrent illness to have a slightly enlarged heart and a systolic murmur suggestive of *maladie de Roger*. In view of the hypertrophy it was thought proper to curtail athletics. In another case the patient, an undergraduate aged 22, with coarctation of the aorta and aortic regurgitation, desired to be trained as an air pilot. In a third case, a man aged 52, who had been seen on account of symptoms of cirrhosis of the liver, had a to-and-fro murmur in the position suggestive of patent ductus arteriosus; there was no enlargement of the heart and no collapsing pulse. The man had been a soldier and had been through the Great War without any disability.

It was no doubt safe to allow patients with a mild degree of congenital heart disease to undertake activities, with supervision, but if the heart became enlarged, or suffered in any way under the stress, exertion would have to be diminished.

Dr. Evan Bedford said he agreed with previous speakers that most children with congenital heart disease should attend an ordinary school, though, in his experience, in London, the school medical officers usually excluded them and sent them to special schools, where the stigma of invalidism was acquired early in life.

In his opinion a pulmonary systolic thrill and murmur, with a dilated pulmonary arc seen by X-rays, indicated some form of pulmonary stenosis, either valvular or sub-valvular; in a case with these signs necropsy had shown a stenosis of the infundibulum and a dilated pulmonary artery. In his experience congestive failure was exceptional in congenital heart disease, and the cause of death was usually intercurrent disease or some complication. For example, the subjects of coarctation of the aorta often died from rupture of the aorta or from cerebral hæmorrhage, which would suggest that some restriction of physical activity was advisable, as advocated by Dr. Gibson.

A defective interauricular septum was an important and not infrequent congenital lesion encountered in adults. The condition might produce no symptoms in infancy and childhood, and caused neither clubbing nor cyanosis. Patients were able to lead normal lives, play games and have children, but after they reached the age of 30, symptoms began to appear and often, eventually, heart failure occurred. The lesion differed from other congenital defects in several ways. Infective endocarditis was rare, but associated rheumatic infection causing mitral stenosis and auricular fibrillation was common. In most congenital defects the degree of cardiac enlargement was relatively slight, but in cases of patent interauricular septum, there was gross enlargement. The X-ray picture was characteristic and consisted of great enlargement of the right heart-chambers with aneurysmal dilatation of the pulmonary artery and its branches. On the screen the right pulmonary artery pulsated visibly, like an aneurysm. The aorta was unusually small. He might mention two cases in which the diagnosis had been made during life and confirmed by necropsy. Both the patients were women with children, and cardiac symptoms had been delayed until adult life.