REFRESHER COURSE FOR GENERAL PRACTITIONERS

CONGENITAL HEART DISEASE—II

BY

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Cyanotic Heart Disease

Diagnosis has recently become of great practical importance in the cyanotic group because of the enormous help that can be given to many of these children by a subclavian–pulmonary anastomosis or other surgical treatment. Without this the outlook is poor for most of these patients; this is strongly supported by the age incidence of the first 340 cases seen in 1947–8, as it shows a steeply diminishing curve.

Age	No.	Age	No.	Age	No.
0– 2 years	60	15–17 years	14	27–29 years	6
3– 5 ,,	68	18–20 ,,	15	30–32 ,,	4
6– 8 ,,	68	21–23 ,,	10	33–35 ,,	4
9–11 ,,	48	24–26 ,,	8	Over 35 years	7
12–14 ,,	29				

Only one in two reach the age of 7; one in five reach the age of 14; and less than one in ten reach the age of 21. The risk of death during the first four years is greater than would appear from these figures, as fewer infants have been seen. A heavy mortality rate continues up to the age of 18 or 21, and the small number surviving beyond this are relatively mild cases with a fair expectation of life. Even slight cyanosis recognizable in infancy generally means severe cyanosis later, with a relatively bad prognosis.

Apart from the general frailty of these children making them apt to die from relatively slight upsets, they have a special risk of cerebral infection because organisms that have entered the blood stream are able to pass to the brain instead of being dealt with by the lungs. Any indication that such a child is losing ground and becoming able to walk shorter distances than before is a sign of serious significance, and if anything is to be done it must be done quickly, as such deterioration is generally progressive until the child dies.

This, unfortunately, applies only to children over the age of 3. However bad the outlook in young children, and this is often because of frequent attacks of increased cyanosis, sometimes with unconsciousness, the risk of operation is greater, and the chance of improvement does not justify surgical intervention. The result is likely to be better if operation can be deferred till after the child is 5 years of age. Exact diagnosis is often impossible in infancy without special investigations which are not justifiable till later.

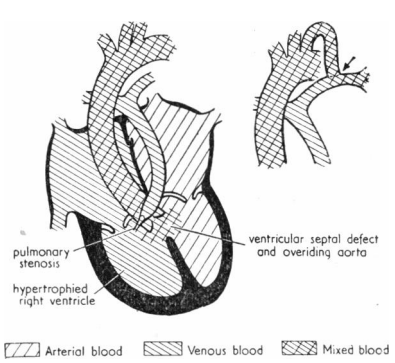

FIG. 4.—Fallot's tetralogy and diagram of Blalock–Taussig operation (end-to-side subclavian–pulmonary anastomosis).

Fallot's Tetralogy (*Fig. 4*).—Fortunately, the combination of pulmonary stenosis (generally infundibular) with an overriding aorta, a high ventricular septal defect, and a right ventricular hypertrophy—known as Fallot's tetralogy, though it should be called after Peacock, who described it clearly in 1858—is the most common group of congenital malformations and is present in two-thirds of these patients. This most probable diagnosis is supported if cyanosis has been present from birth or from the first 18 months of life; if there is a systolic murmur, often with a thrill, in the pulmonary area and a diminished or normal pulmonary second sound; if there is no diastolic murmur; if the heart is not enlarged; if there is no prominence of the pulmonary arc or pulmonary arteries (though less than half of them show a real sabot-shaped heart, the remainder having a relatively straight left border or even slight prominence of the pulmonary arc due to dilatation of the infundibulum beyond the stenosis); and, most important of all, if the density of the lungs on radioscopy is lighter than usual. These criteria should enable the diagnosis of Fallot's tetralogy to be made in most cases, and it is here that the operation of subclavian–pulmonary anastomosis is most likely to help. It is curious that the habit of squatting should be so common in Fallot's tetralogy; it occurs in at least four-fifths of these cases, but is seen in other forms of cyanotic heart disease, even in Eisenmenger's complex in which there is no pulmonary stenosis.

Pulmonary Atresia.—Cases are generally included as Fallot's tetralogy whether there is pulmonary stenosis or pulmonary atresia. The last group must now be differentiated, because the small and thin-walled pulmonary artery that is present beyond the atresia makes subclavian–pulmonary anastomosis more difficult, but not always impossible. This diagnosis must be considered when in a case otherwise resembling Fallot's tetralogy there is no systolic murmur and a rather loud second sound in the pulmonary area.

Tricuspid Atresia (*Fig. 5*).—When a case resembles Fallot's tetralogy in general, but the electrocardiogram shows left ventricular preponderance and also a rather larger heart with hypertrophy of the left ventricle, the diagnosis is generally tricuspid atresia with a small or non-functioning right ventricle and a defect in the auricular septum. These cases can be helped by subclavian–pulmonary anastomosis because the anomalies cause a reduction in the pulmonary blood flow, but the degree of improvement to be expected is less than in ordinary Fallot's tetralogy.

Pulmonary Valvular Stenosis.—In the oligaemic group Fallot's tetralogy is the most common condition, but pure

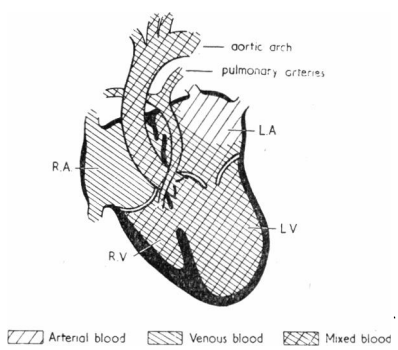

FIG. 5.—Tricuspid atresia with small undeveloped right ventricle.

pulmonary valvular stenosis with a right-to-left shunt through a patent foramen ovale is not uncommon, and this diagnosis may be suspected if there is great enlargement of the right ventricle, and if cyanosis develops later, sometimes not till early adult life. When this happens the downward course is generally rapid and the possibility of operation should be considered at once, direct surgical relief of the stenosis being indicated. In these cases the electrocardiogram will show right ventricular preponderance, often with deep inversion of T in Leads II and III. The physical signs are the same as in the acyanotic cases, but in the more severe cases the murmur may be less and the thrill may be trivial.

Pleonaemic Cases.—The division of those in whom the blood flow to the lungs is diminished (oligaemic) and those in whom it is increased (pleonaemic) is best made by radioscopy, but physical examination may often give a clue, as enlargement of the heart or the presence of a diastolic murmur, generally in the pulmonary area sometimes towards the apex, is much more common in this group. Even more important is the pulmonary second sound: if this is much increased and booming, or clearly duplicated, or if it is palpable, it is almost certain that large pulmonary arteries will be seen on radioscopy, and that the patient already has an adequate blood supply to the lungs, so that he cannot be helped by subclavian–pulmonary anastomosis. In the pleonaemic group accurate diagnosis is at present of less practical importance. Transposition of the aorta and pulmonary artery with septal defects, and Eisenmenger's complex, are probably the commonest lesions, the latter being the more likely diagnosis in adult patients.

Surgical Treatment in Cyanotic Heart Disease

The outlook for these children has been completely changed by the introduction of subclavian–pulmonary anastomosis (Blalock–Taussig operation) and by Brock's more direct relief of the stenosis. The increased blood flow to the lungs allows more blood to be oxygenated and so the disability and cyanosis are both greatly relieved. When the disability is enough to justify it, operation should be advised between the ages of 5 and 10 years.

In infants the small size of the vessels produces a surgical problem of the greatest difficulty and the operation is much more dangerous and sometimes less lasting in its good effect, because the anastomosis is too small as the child grows larger. For this reason the operation should never be performed under 3, except as a life-saving emergency, and generally not till 5 years of age. From 5 to 10 is perhaps the ideal age, but there seem to be no special difficulties in older children, though the difficulties and risks are greater in those over 20.

The operation is a serious one, with a mortality of about 15%, but two-thirds of all the children are enormously improved, so that they may walk a mile or more instead of 100 yards or so, and get about all day almost normally. The polycythaemia disappears quickly and the clubbing of the fingers more slowly. The arm to which the subclavian has been divided gives no trouble.

It is important for the parents to realize that the heart condition has not been cured, and that though the outlook is much improved it is still uncertain for how long this will last. Naturally, with the increased work given to the heart, there is some increase in size; but this does not seem to be progressive, and follow-up studies so far are very encouraging and suggest that the improvement may be lasting.

In the first 100 cases operated on by Mr. R. C. Brock at Guy's Hospital 15 died, 7 could not have an anastomosis done (2 at least because of a mistake in diagnosis), and 14 were only slightly or moderately improved; the remaining 64 were enormously improved almost at once, and their condition has continued satisfactory in all except two—one with tricuspid atresia and one with a single ventricle. Many hundreds of children have now been operated on at various centres and the procedure may be regarded as an established one. The relative position to be taken by Brock's operations for valvulotomy and resection of the infundibulum still remains to be established. It may well prove to be the operation of choice in those cases in which stenosis is the main feature, which is suggested when the disability is much worse than the cyanosis. At present the relative risk seems greater, but there is no doubt that the improvement may be just as striking and dramatic, and possibly in the successful cases the good result may last for even longer.

Illustrated by Miss Sylvia Treadgold.

GENERAL PRACTICE: A NEW PERSPECTIVE

THE REPORT OF A COMMITTEE OF THE ASSOCIATION

Two years ago the British Medical Association published a report entitled *The Training of a Doctor*, with proposals for the improvement of the undergraduate curriculum. It was received with such appreciation that the Council forthwith appointed another Committee under the same chairman, Sir Henry Cohen, to carry the subject further by considering the postgraduate education of the general practitioner. The new Committee looked at this subject from a wider viewpoint than the term "postgraduate education" ordinarily suggests, and reached a proposal that there should be a post-registration period of training for general practice—a new conception in this country and one scarcely formulated in any other. It considered also the continuing education of the established general practitioner, not only by means of refresher courses and lectures, but by clinical conferences, hospital work, and consultations with specialists, as well as self-education.

It was necessary to consider the status of the general practitioner, the scope and organization of his practice, and the personal qualities his work demands. Thus the report appears under an extended title, *General Practice and the Training of the General Practitioner.*[1] It presents general practice in a new perspective, freed from certain ideas which in recent times have obscured its role and diminished its importance.

The Committee, which has put in 18 months' intensive work, consisted of 33 members, eight of whom were chosen because they were general practitioners in representative fields of practice—rural, small town, and large town. A similar number of members were associated with university teaching, and others represented the consulting branches of the profession, hospital work, and the administrative side of medicine. The six principal officers of the Association, two of whom are general practitioners, were also on the Committee.

Apothecaries and Family Doctors

At the outset of its work the Committee was faced with the objection that conditions of practice now are such that many practitioners cannot take advantage of whatever postgraduate facilities are offered. Their immersion in routine details of practice allows them no time for study, or, if they have time, the energy and mental detachment are lacking.

The answer which the Committee makes is that its recommendations will really ease the lot of the general practitioner

[1] *General Practice and the Training of the General Practitioner. The Report of a Committee of the Association.* London: British Medical Association, B.M.A. House, Tavistock Square, W.C.1. 1950. Price 7s. 6d.