
STAPLES

CONGENITAL HEART DISEASE

by

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STAPLES PRESS

STAPLES PRESS LIMITED
Mandeville Place, London

STAPLES PRESS INCORPORATED
70 East 45th Street, New York

FIRST PUBLISHED 1939
SECOND EDITION 1950

This Book is set in 10 on 11 point 'Monotype' Times New Roman

Made and printed in England by
STAPLES PRESS LIMITED
at their St. Albans, Herts, establishment

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ACKNOWLEDGMENTS

Fig. 1 Pichon, E., Nouveau Traité de Medicine, 1933, Vol. X, p. 616 (Paris. Masson.)

2 *and* 3 Lev and Saphir, *J. Tech. Meth.*, 1937, 17, figs. 18 and 19.

4 Auricular Septum: 'Quain's Anatomy', 1929, Vol. IV, part iii, p. 40. *Adapted from.*

5 Formation of Aortic Vestibule: 'Gray's Anatomy', 1932, 25th Ed., figs. 188 and 189. (Longmans, Green.)

6 Fish Hearts: 'Quain's Anatomy', 1929, Vol. IV, part iii, p. 5, fig. 3.

8 Reptile Heart: 'Quain's Anatomy', 1929, Vol. IV, part iii, p. 9, fig. 7.

9 Evolutionary Outlines: Kingsley, J. S. 'Comparative Anatomy of Vertebrates', 1912, fig. 287. (John Murray.)

10 Modified from Still.

11 *and* 12 Deficient Pericardium. Courtesy of Mr. N. R. Barrett.

19 Types of Coarctation: figs. from Evans, W., *Quart. J. Med.*, New Series, **2**, 1, *et seq.*

21 Injected Arteries in Coarctation. Courtesy of Mr. Chesterman.

22 Coarctation: *Lancet*, 1934, **2**, 924, figs 2 and 3.

23 From Campbell, M., and Suzman, S., 1947. *Brit. Heart J.*, **9**, 185.

26 Types of Coarctation: figs. from Evans, W., *Quart. J. Med.*, New Series, **2**, 1, *et seq.*

28 Courtesy of Mr. L. Fatti. Transactions Med. Soc. London (1943-45), 64, 261.

29 High Aortic Arch. Courtesy Mr. N. R. Barrett.

31 Ex Bedford and Parkinson, 1936, *Brit. J. Radiol.*, **9**, 777, fig. 1.

32 Abnormalities of the Aortic Arch, after Edwards, J. E., 1948, *Med. Clin. N. Amer.*, 32, 925.

33 Abnormalities of the Aortic Arch, after Edwards, J. E., 1948, *Med. Clin. N. Amer.*, 32, 925.

35 Ex Bedford and Parkinson, 1936, *Brit. J. Radiol.*, **9**, 781.

44 Courtesy of Dr. M. Matthews.

46 Ex Bedford, Papp and Parkinson, 1941, *Brit. Heart J.*, **3**, 7.

49 Courtesy of Dr. M. Matthews.

50 *and* 51 'British Encyclopedia of Medical Practice', Vol. VI, p. 225, Plate 2.

55 Ex Muir and Brown, 1934, *Arch. Dis. Childh.*, 9, 35, figs. 5 and 6.

61 Courtesy of Dr. M. Matthews.

62 Infra Valvular Stenosis: Laubry, Ch., Nouveau Traité de Pathologie Interne, 1930, 715. (Paris, Doin.)

65 Ex 'British Encyclopedia of Medical Practice', Vol. VI, p. 228, fig. 32.

67 Ex 'British Encyclopedia of Medical Practice', Vol. VI, p. 229, fig. 34.

70 Ex Brown, J. W., *Postgraduate Med. J.*, **25**, 514, 1949.

74 Ex Brown, J. W., *Postgraduate Med. J.*, **25**, 514, 1949.

75 Courtesy of Dr. D. V. Hubble.

79, 82 *and* 85 Ex Brown, J. W., *Arch. Dis. Childh.*, 1936, **11**, 275, figs. 1, 2 and 3.

80 Ex Brown, J. W., *Postgraduate Med. J.*, **25**, 518, 1949.

Fig. 83 Ex Brown, J. W., *Postgraduate Med. J.*, **25**, 518, 1949.
88 Ex Brown, J. W., *Postgraduate Med. J.*, **25**, 518, 1949.
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93 *and* 94 Ex Humphreys, *Arch. Path.*, 1932, 14, 671, fig. 1.
96 Kerwin, J., *Tech. Meth.*, 1936, **15**, 45.
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102 From Marshall, R., 1943, *Brit. Heart J.*, **5**, 193.
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106 From Dadds and Hoyle, 1949, *Brit. Heart J.*, **11**, 390.
107 From Micks, R. H., 1940, *Brit. Heart J.*, **2**, 63.
110 Herson, R., and Simons, M., 1946, *Brit. Heart J.*, **8**, 125.
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118 Ex 'British Encyclopedia of Medical Practice', Vol. VI, p. 215, fig. 26.
121 Radiological Anatomy of the Heart: ex Parkinson, J., *B.M.J.*, 1933 (ii), 591, figs. 1, 2 and 3.

PREFACE TO SECOND EDITION

The ten years that have elapsed since the first appearance of this work have witnessed many important advances in the field of congenital heart disease. The impetus furnished by the development of the surgical treatment in certain cardiovascular anomalies has awakened intense interest in the clinical study of these patients. New work is constantly appearing and fresh methods are being applied to the elucidation of heart abnormalities. To embrace these new developments the book has been for the most part rewritten. Even then, the author feels that in the present state of constantly changing knowledge the last word has not been said. He therefore offers his apologies if at the time of appearance of this work certain parts are not as complete as the reader would like them to be. In particular, such topics as angiocardiography are but briefly mentioned because the author has had little practical experience in this ever advancing field. The writing of a book such as this involves many references to the current literature, and as far as possible information has been credited to its respective author. Apologies are offered for any inadvertent omission.

The author owes much to his colleagues who have worked with him in various clinics and who have referred cases and specimens to him. Particular gratitude is expressed to Dr. D. C. Muir, of Hull, where much of the clinical study has been carried out and where observations on the natural history of congenital heart disease are in progress; to Dr. J. C. H. MacKenzie, Mr. T. Holmes Sellors, and Mr. Gordon Cruikshank, of the Leicester Chest Unit, where the surgical aspects have been especially considered; to Drs. Evan Jones, R. Daley, and Mr. N. R. Barrett, at St. Thomas's Hospital, for much help in connection with cyanotic cases; and to Dr. T. Lodge, of Sheffield, for much radiological advice. It would be impossible to name all those others who have helped me with advice, criticism, and have furnished me with specimens. I am grateful to them all.

My deepest thanks are due to Mrs. Neata Brown, who has conscientiously and patiently helped me in the preparation of this work, and with the proofs. My thanks are again due to the publishers for their interest and co-operation in all matters connected with this book.

J. W. B.

Grimsby, 1950.

ERRATA

Page 66, line 2.
For posterior intercostal spaces, *read* costo-clavicular spaces.

Page 329, line 57.
For Suzman. W., *read* Suzman. S.

Page 45, Caption to Fig. 15.
For as fig. 11, *read* as fig. 14.

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CHAPTER I

INTRODUCTION

Congenital heart disease has become of recent years an important branch of cardiovascular pathology. Largely owing to the impetus of the work of Abbott (1927) an increasing interest has developed not only in the embryological aspects, but also in the clinical features and anatomical diagnosis of certain lesions. With the development and more universal application of radiological and cardiographic methods, many hitherto unrecognized features have been brought forward and a reasonably accurate diagnosis has become possible in many abnormalities. The recent advances in the surgical treatment of certain anomalies has made correct diagnosis of paramount importance. Despite these advances the published cases are still comparatively rare where clinical, X-ray and electrocardiographic data have been correlated with post-mortem findings. In the minds of many there still lingers the conception of a puny cyanotic child, often of indifferent mentality, as the typical picture of congenital heart disease. This is an erroneous idea, for in point of numbers acyanotic congenital heart disease greatly exceeds in incidence the cases of cyanotic disease. The acyanotic case is frequently overlooked because it may be entirely symptomless, and its recognition purely fortuitous. Further, there is a tendency to designate any unexplained cardiac murmur as 'functional' or as the result of a rheumatic carditis without arthritis, the possibility of a congenital etiology being overlooked.

Certain features of congenital heart disease at once command attention. In the first place there is a fixed anatomical lesion with structural alterations in the heart or great vessels that cannot generally be removed. Consequently the course of the patient is largely moulded by the factor of adaptation, and the interplay of various mechanical forces in order to assure an adequate circulation despite the handicap of a gross anatomical abnormality. Exceptionally, some lesion such as an isolated interventricular septal defect, which may be of considerable importance when the heart is small, may become, as the heart enlarges with normal growth, unimportant relative to the size of the heart as a whole, and its physical signs may disappear. Equally, a lesion such as coarctation may become more important as age advances because growth of the defect may not be commensurate with growth of the aorta. This furnishes an impetus to the development of a collateral circulation. In most cases a congenital lesion imposes a strain on either

the left or the right side of the heart, and, with certain rare exceptions, these sites of strain are exposed to the lethal effects of a superimposed bacterial endocarditis.

The various factors of adaptation are well known in coarctation of the aorta where a collateral circulation may be the means of prolonging life to its normal span, despite such a gross lesion as occlusion of the aorta in the distal part of its arch. Again the polycythaemia of the morbus caeruleus is a classical example of adaptation by the individual to a gross anatomical handicap.

The maintenance of a pulmonary circulation in the severe anomalies must always be a matter of paramount importance in view of the necessity of a supply of aerated blood if life is to be maintained. In some severe anomalies, and in transposition of the vessels, the persistence of one or all of the foetal orifices, the ductus arteriosus, the foramen ovale, or a defect of the interventricular septum, at least assures that a certain proportion of the blood-stream is deflected through these channels to gain access to the lungs. Where transposition is associated with a common ventricle, the phenomena of the turtle heart are plainly visible. Not only is admixture of blood slight in the common ventricle, but the very fact of transposition tends to bring the vessels adjacent to their appropriate auriculoventricular orifices. In Spitzer's theory the morphological similarity of the deformed heart to that of the reptile is clearly exemplified. In pulmonary atresia the role of a patent ductus as the sole source of blood to the pulmonary circulation is only too clearly emphasized. It is remarkable that late closure of the ductus may take place in pulmonary atresia with lethal effects to the individual. Other anatomical adjustments, again made necessary by the need for a pulmonary circulation adequate to the patient, are the hypertrophied bronchial arteries evident in some cases of pulmonary atresia, some cases of persistent truncus arteriosus, and in a few cases of transposition of the vessels.

In spite of these adaptations there occurs from time to time a case where the anatomical conditions present are such that the greatest difficulty is experienced in determining how an adequate supply of aerated blood reaches certain organs.

The blood supply of the heart itself may be imperilled in certain abnormalities. The possible sources of a blood supply to the heart have been summarized by Mahon (1936) in relation to a case of aortic atresia, where the aorta was a cul-de-sac of 2 mm. diameter. The coronary arteries arose in their usual situation, and blood reached the ascending aorta through a widely patent ductus arteriosus. Wearn (1933) and his co-workers showed connections between the coronary arteries and the cavities of the heart by means of celloidin injections and wax plate reconstruction. They noted two types of vessels, arterioluminal and arteriosinusoidal. The latter, in virtue of their course

amongst muscle bundles and fibres, appear to be connected with the nourishment of muscle. In Bellet and Gouley's (1932) case of aortic atresia with closed septa, such small channels were demonstrated, and no doubt if material was more frequent and examination more minute, it would be possible to add other examples of such a mechanism. An additional blood supply to the heart may be furnished through the extracardiac anastomoses of Gross (1921). Beck (1935) has shown experimentally that the response to this mechanism is greatest when there is most need for it. It is possible that some blood supply to the heart may be acquired through the coronary sinus and its ramifications. Lastly the coronary arteries may have abnormal origins as shown elsewhere (page 90). Other aspects of adjustment of the circulation are discussed under the respective lesions.

Certain congenital cardiac defects have been considered by Holman (1937) in regard to their role as intracardiac fistulae. Where there are septal defects and some associated obstructive lesion, as for example, pulmonary stenosis, the general result is that the volume flow of blood is unequally distributed amongst the chambers, thus producing notable effects. Not only may cyanosis be caused by abnormal venous arterial shunts, but the increased flow of blood through the chamber leads to its hypertrophy and dilatation. The dilatation is simply due to the increased volume of blood that the chamber has to accommodate. Hypertrophy is the result of the work performed by the heart in ejecting this increased volume of blood, and in overcoming an abnormal peripheral resistence imposed by the defect. Thus a dextroposed aorta lying astride a high ventricular septal defect leads to the right ventricle sharing in the work usually undertaken by the left. Similarly, an abnormal communication such as a patent ductus arteriosus, where up to 70 per cent. of the blood ejected into the aorta may pass into the pulmonary artery, there is a gradual hypertrophy of the left ventricle which may be reflected both in the electrocardiogram and in the radiological picture. It is singular that the patent ductus does not produce more marked symptoms and signs of cardiac embarrassment. A peripheral arteriovenous fistula leads to marked cardiac enlargement because blood escapes into a capacious venous system at low pressure. In the patent ductus, constriction of the smaller pulmonary branches probably creates an area of peripheral resistance, tending to limit flow into the pulmonary artery by maintaining a degree of pressure in the latter vessel.

With a view to testing that an increased volume flow was a productive stimulus to ventricular hypertrophy, Holman experimentally created ventricular septal defects in dogs. The effects produced were dependent upon the size of the defect created. Where the defect was large there appeared dilatation of those chambers through which the shunt passed, the changes being most apparent in the left ventricle, right ventricle,

pulmonary artery, and the left auricle. Dilatation was confirmed by serial X-ray pictures, and the presence of hypertrophy established at autopsy. The converse may be true, and a diminished volume flow leads to hypoplastic changes from functional inactivity. Such changes are apparent in the left ventricle in mitral atresia, or in the small aorta of aortic atresia, and of some cases of ventricular septal defect.

THE COR PULMONALE. Congenital heart disease involving the right side of the heart, pulmonary stenosis and dilatation, tricuspid incompetence, and to a certain extent the long-standing patent ductus arteriosus, may be the cause of the cor pulmonale. Fineberg and Wiggers (1936), using a method of compression of the pulmonary artery, showed that when the lumen of the artery was reduced up to 58 per cent., there was a fall in venous pressure, and the right ventricle became distended. At the same time, owing to less blood reaching the pulmonary circulation, and hence the left ventricle, there was a diminished output from the left ventricle and a fall in aortic pressure. Brill and Robertson (1937) indicate that in these circumstances there is less blood available for the coronary circulation, and a vicious circle is established which accelerates failure of the right ventricle. Death may result from congestive failure, or from failure of the pacemakers before there is time for congestive failure to appear. Thompson and White (1936) estimate that hypertrophy of the right ventricle appears within two months of obstruction in the pulmonary circuit. The same authors state that the most frequent cause of right ventricular hypertrophy is left ventricular strain and failure, for when the left ventricle fails the lungs become congested and thereby place a load on the right ventricle.

The cor pulmonale develops quite insidiously, and may be present for years before actual failure appears. The signs of failure are those of congestion, distended veins in the neck, an enlarged liver, and oedema. The physical signs are those of enlargement of the right ventricle, together with the signs of the causal valvular or other disease. The pulmonary second sound is accentuated and the blood-pressure tends to be low. The assessment of the enlargement of the right ventricle may be difficult owing to its anatomically anterior position. It may, however, be radiologically recognized by alterations in the contour of the heart, or by right-axis deviation in the electrocardiogram. Although dilatation of the pulmonary artery may be apparent on the screen, it is not always present at autopsy. This dynamic dilatation of the pulmonary artery has been noted by Brill and Robertson (1937), Schwedel and Epstein (1936), and others. Cough and dyspnoea are often present as well as symptoms produced by congestion of individual viscera. In congenital heart cases, death often occurs without the intervention of marked congestive failure. In the cyanotic types, where there is some

lesion of the pulmonary tract, death is often the result of some respiratory infection which adds embarrassment to an already embarrassed right side. Death with congestive failure and auricular fibrillation is practically only seen in the auricular septal defect, and perhaps in this case may be the result of the commonly associated valvular lesion of mitral stenosis.

PARADOXICAL EMBOLISM. When thrombotic or other material from the right side of the heart reaches the systematic circulation through a defect of the auricular septum, paradoxical embolism is said to occur. Rostan (1874), who observed seven cases at autopsy where he thought that an embolus derived from the right side of the heart had passed through the foramen ovale, referred to the condition as 'crossed embolism'. The term 'paradoxical embolism' was subsequently used by von Recklinghausen. The classical conception is of its occurrence only in cases of patent foramen ovale or other auricular septal defect. Actually it may take place in cases where there is dextroposition of the aorta and an interventricular septal defect, or in the patent ductus arteriosus. Abbott has reported twelve cases in which paradoxical embolism occurred in the above anatomical conditions. The typical case shows a coiled thrombus passing through the auricular septal defect into the left auricle. The embolus is usually derived from a thrombus in the femoral or other veins. Zahn (1881) was among the first to report such a typical case, and others are those of Barnard (1930) and Thompson and Evans (1930). The latter authors, and Beattie (1925), discuss the changes that may favour paradoxical embolism in the auricular septal defect. It appears that attacks of pulmonary embolism precede the paradoxical embolus. A sudden occlusion of the pulmonary artery, or of one of its important branches, leads to a rise in the pulmonary pressure and a steep fall in the systemic blood pressure. Coincidentally there is a rise in pressure in the right auricle which favours the possible transmission of the embolus through a defect of the auricular septum. Normally the pressure in the left auricle is sufficient to keep closed the valve-like flap of the foramen ovale in cases that are potentially patent. In parenthesis, it may be remarked that a similar sharp fall in the blood pressure may occur in coronary thrombosis and may give rise to difficulty in the differentiation of this condition from pulmonary embolism, particularly in post-operative cases. There is the experimental evidence of Haggart and Walker (1923) to show that a sudden occlusion of the left branch of the pulmonary artery causes a rise of 29 per cent. in the pulmonary pressure, and that in total occlusion of the trunk a rise in pressure up to 250 per cent. may be anticipated. These rises in pulmonary pressure were shown to be accompanied by a sharp fall in systemic blood pressure. Consequently, if there is a defect of the auricular septum, clot and thrombotic material may pass from the right auricle to the left. According to Thompson