The Anatomy of Congenital Pulmonary Stenosis

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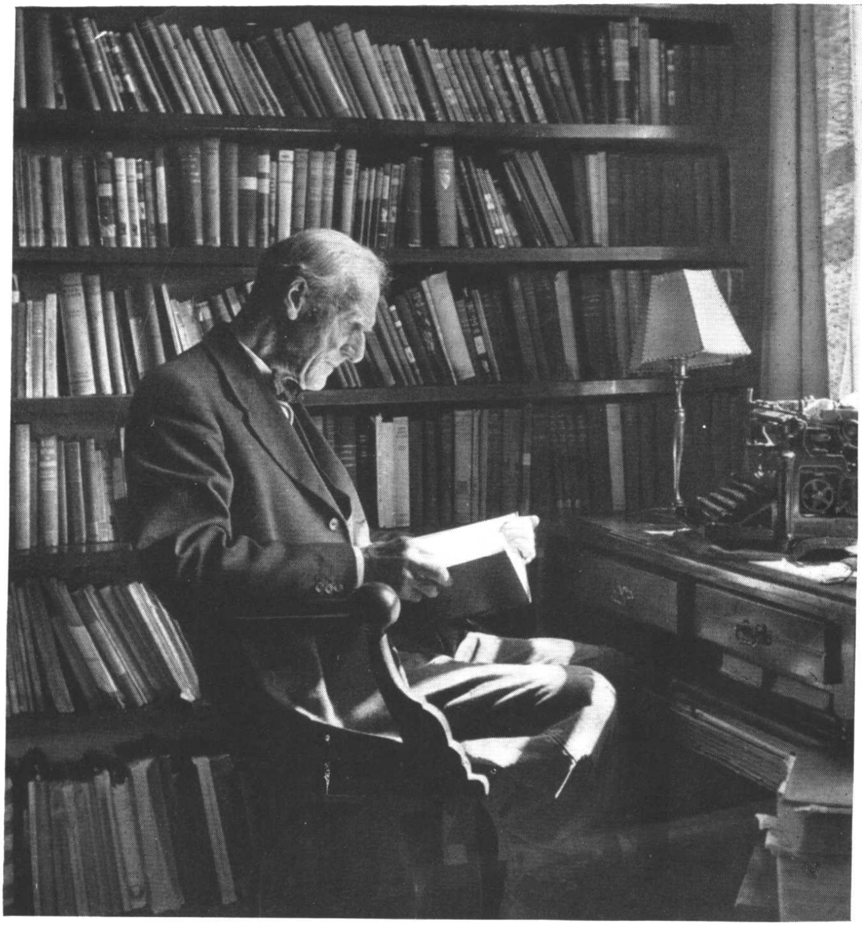

Sir Arthur Keith

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INTRODUCTION

THIS account of the anatomy of congenital pulmonary stenosis was written in its greater part over five years ago at the time when great interest had recently been aroused in the condition and many cardiologists, surgeons, radiologists and others were engaged in the investigation and treatment of cases of pulmonary stenosis. Its publication was, however, deliberately delayed because I realised that with a steadily increasing experience in the management and surgical treatment of these cases I was acquiring much additional information. Although, in the meantime, a number of accounts of the anatomy of pulmonary stenosis have appeared, I have not regretted this delay for the fuller experience has been of greater value.

The term 'pulmonary stenosis' is itself, of course, not precise; in this book it is used as a comprehensive term covering all varieties of obstruction to the blood-flow to the lungs from congenital malformation. The individual varieties of pulmonary stenosis will be indicated by their appropriate names.

My interest in the development and surgical anatomy of the condition arose from two reasons. First, it is more satisfactory to know why a thing occurs and it seemed obvious that there must be an explanation for the common occurrence of such a peculiarly constant form of maldevelopment as pulmonary stenosis, even though its varieties are several. Second, my interest in the relief of pulmonary stenosis by direct operation made it essential to understand the anatomical formation both of the stenosis itself and of the muscular structure of the ventricular walls in its neighbourhood. As a corollary to this it is necessary to emphasise the need for careful and complete study of the normal morbid anatomy of a disease process when it is being investigated and treated. This remark may be superfluous but the fact remains that many people are handling cases of pulmonary stenosis, including performing operations upon them, without any more than a second-hand and often rather hazy picture of the anatomical condition present. This is quite apart from the practice of making an empiric diagnosis of 'pulmonary stenosis' and advising and performing a stereotyped operation for its indirect relief without attempting, at operation, to make the diagnosis of the particular variety as exact as possible. The information generally available about the anatomy of the condition is, moreover, commonly accepted without any personal verification or study. This is obvious from the popular conception of the nature of the obstruction, especially the infundibular form.

In any case the introduction of such an important step as operation for the relief of pulmonary stenosis demands a reinvestigation of the whole subject and verification of past observations and descriptions. What might have been acceptable as a general description for a condition which was of little interest other than as an untreatable clinical state, or as a range of congenital malformations, can be inadequate if exact surgical attack is to be based on it. It is imperative in such new circumstances to re-open the whole problem and to confirm, amplify or modify the older descriptions and conceptions.

It is also important to realise that there may be a great difference between the anatomical state of affairs as observed after death in a specimen in a condition of muscular contraction aggravated by fixation and hardening, and the state that exists during life in an active, functioning muscular structure such as the heart. The surgeon who observes at operation the condition both outside and inside the living heart will see definite and significant differences in the same specimen after death. We must remember this difference between the anatomy of the living heart and the dead heart and that, in so far as this feature is concerned, post-mortem examination may be deficient and misleading instead of being the acme of truthful observation it is usually thought to be.

The ordinary conception of the right ventricle is often that of a rather simple muscular structure which expands to receive blood from the atrium and then contracts to expel it. In fact the arrangement is much more complex than this, as will appear from the succeeding description. The formation, structure and function of the outflow portion of the right ventricle in particular is quite complex and is of great practical importance in relation to the stenoses that affect it.

The general paucity of knowledge about the anatomy of pulmonary stenosis at the time of the introduction of surgical treatment was surprising because reference to the literature reveals a whole wealth of first-class information that is interesting, informative and even enthralling. Much of this has remained neglected or forgotten and to the surgeon seeking practical information it is rather like digging and finding gold. In this connection one must mention especially the writings of Sir Arthur Keith which, as always, are charmingly and simply written and full of scholarship, clear observation and deduction. No one interested in the subject should fail to read the series of lectures published in the *Lancet* between 1904 and 1924. As long ago as 1904 Keith wrote in the introductory paragraph to his Hunterian Lecture at the Royal College of Surgeons on *The evolution and action of certain muscular structures of the heart*: 'It is with a feeling of keen disappointment that men who seek to extend our knowledge of the origin and development of the human heart lay down any of the standard treatises on the function and diseases of that organ. Their labour as far as one may judge from the perusal of such works lies outside the field of practical medicine and is useless to physician and physiologist alike.'

Keith had to wait nearly half a century to see his work on the nature and development of pulmonary stenosis bear fruit, but it is now possible to say that his observations and explanations have been of immense help in planning and developing a logical surgical approach to the direct operative relief of pulmonary stenosis.

I have to thank Professor T. B. Johnston for reading through my manuscript, and making numerous valuable suggestions and correcting several errors.

My secretary, Miss Chris Jones, has spent many tiring hours on the preparation and scrutiny of manuscript and proofs and I am very grateful to her.

Sir Arthur Keith gave permission to use certain of his diagrams and also selected the frontispiece from among his several photographs; it was taken by Mr. John Miller of Barnet. Messrs. Edward Arnold and Company have given permission to reproduce Figures 1 and 6 from *Human Embryology and Morphology*.

Messrs. Baillière, Tindall, and Cox have given permission to reproduce Figure 9.

I am very grateful to Dr. Prinzmetal for his kindness in sending me a short strip from one of his splendid films which demonstrates the peristaltic-like movements of the infundibulum.

I am also grateful to the various colleagues who have given indispensable aid by recording electromanometric pressures both at cardiac catheterisation and at operation. Among these are Dr. D. C. Deuchar, Dr. R. J. Shephard, Dr. R. Gibson, Dr. P. Fleming and Dr. L. Brotmacher. Dr. Maurice Campbell has kindly allowed me to use Figure 34 from one of his articles. Figure 51 was first published in the *British Heart Journal*.

I acknowledge with gratitude the help so readily and skilfully given with the preparation of many of the photographs, drawings and pressure records by the Members of the Department of Medical Illustration of Guy's Hospital and of the Photographic Department of the Brompton Hospital.

CONTENTS

ILLUSTRATIONS

CHAPTER I

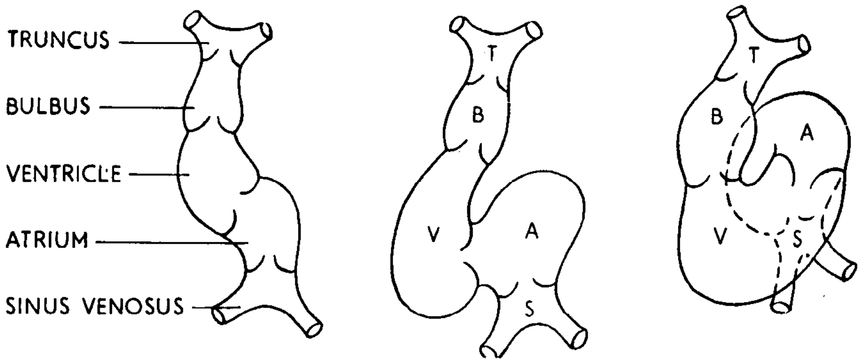
The comparative anatomy and embryology of pulmonary stenosis

It is easier to understand the embryology of pulmonary stenosis from a simple knowledge of the comparative anatomy. The complicated developmental features of the human heart are difficult to follow until it is appreciated that in its development the heart passes through most of the stages of formation of the heart of the lower orders of animals. In fact most of the congenital malformations represent the heart, fixed as it were, at a more primitive stage of development: one that can be recognised as belonging to that commonly seen in certain of the lower orders, with the addition of adaptations resulting from the arrested development.

Maude Abbott (1936) in her classical *Atlas of Congenital Cardiac Disease* remarks: '. . . The most bizarre combinations of defects can usually be interpreted quite simply, as due to early arrest of development, marked it may be by ingenious structural adaptation of growth'; and 'Much light is cast upon the development of the mammalian heart, and incidentally, upon the period at which arrest of growth has taken place in cardiac anomalies of the graver sort by a comparative study of the adult fish, amphibian and reptilian organ. The truly extraordinary way in which these various orders in the ascending vertebrate scale mirror the successive stages through which the human heart passes in very early intrauterine life is one of nature's most spectacular and impressive feats, presenting as it does a complete review of this organ's evolution down to the closure of the cardiac septa in the eighth week of foetal life.'

The following account does not pretend to be original; the description is entirely second-hand although illustrated by a number of comparative anatomical specimens personally collected and studied. It is not possible for a practising surgeon to survey and verify the whole of this vast comparative and developmental problem; it is essential to rely upon the work of others. The examination, dissection and description of the human material is, however, the result of personal work.

The primitive tubular heart consists essentially of five parts; the sinus venosus, the atria, the ventricles, the bulbus cordis and the truncus arteriosus (Fig. 1). It



1. Diagrams to show the primitive tubular heart of five chambers and how, as the heart lengthens and becomes folded on itself, the bulbus comes to lie beside the atrium and ventricle. (*After Keith, 'Human Embryology and Morphology', Arnold.*)

can be said at once that what concerns us most in the present problem is the formation and fate of the bulbus cordis. Thus Keith (1909) states 'one of the greatest discoveries since Peacock's time is now only dawning, but every year increases our assurances of its truth—viz. that there is a fourth part or chamber in the mammalian heart which hitherto we have taken no cognisance of . . . The fourth part is the bulbus cordis . . . We have good reason for believing that, in the same manner as the sinus venosus has become incorporated in the right auricle, the bulbus has become included in the right ventricle, forming the part loosely termed its infundibulum.' Although Keith mentions only four parts of the primitive heart, later (1933) he includes the truncus arteriosus as a fifth element and it is really more correct to say that the bulbus is absorbed partly into the ventricles and partly into the truncus, of which it forms the pericardial part. He goes on to say, 'The credit of this discovery belongs to Alfred Greil, prosector in the University of Innsbruck. He traced the fate of the bulbus by a prolonged study of the hearts of developing vertebrates (Greil 1903). Independently of him I had reached the same conclusion from an investigation of malformed human hearts and of the hearts of vertebrate animals. A large number of the very commonest malformations of the human heart are due to an arrest of the process which ends in the incorporation of the bulbus cordis in the right ventricle. The great majority of cases of congenital stenosis of the pulmonary artery are of this nature.'

The bulbus cordis is best shown in fishes, i.e. gill breathers; Figures 2–4 show examples in the skate, shark and angler fish and demonstrate that it varies much in size and shape in the different classes. Quain (1929) makes some interesting comments on its nomenclature, about which there is sometimes confusion. The names that have been used for it are *conus arteriosus*, *bulbus arteriosus*, *truncus arteriosus* and *bulbus cordis*.

Walmesley in *Quain's Anatomy* pointed out that, '*conus arteriosus* is now used in human anatomy for a definite part of the right ventricle; *truncus arteriosus* is the name now usually applied in human embryology to that part of the common ventral vessel from which the pulmonary aorta and the proximal part of the systemic aorta are derived and the structure of whose wall, from the beginning, is that of a vessel. In the Teleostei this section of the ventral vessel is enlarged and its wall is thickened by a great development of smooth muscle and elastic tissue fibres, but it is entirely free from cardiac muscle; the name *bulbus arteriosus* is then used for it.

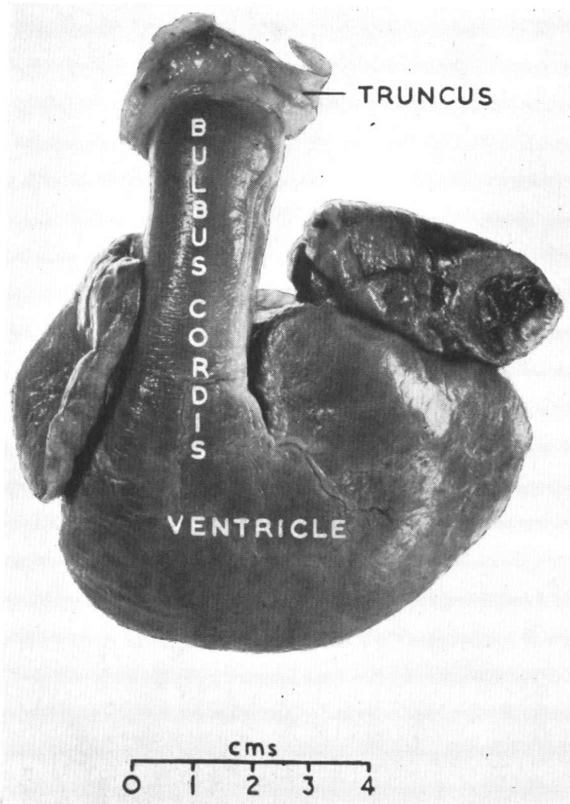
'The name *bulbus cordis* is thus the most suitable to be applied to that region which is the most distal part of the heart and which yet forms the most proximal part of the exit tube from the heart and it is to be considered a cardiac chamber for in its interior it carries the bulbar valves and in its wall there is cardiac muscle. It is present in all vertebrate embryos but in most forms it undergoes an ontological retrogression through the absorption of its proximal part into the ventricle and of its distal part into the truncus arteriosus; it remains persistent as a clearly separable chamber of the heart only in the Elasmobranchii, Ganoids, Dipnoi and amphibia.'

According to Waterston (1918) Langer first suggested the name bulbus cordis and it was then generally recognised and adopted.

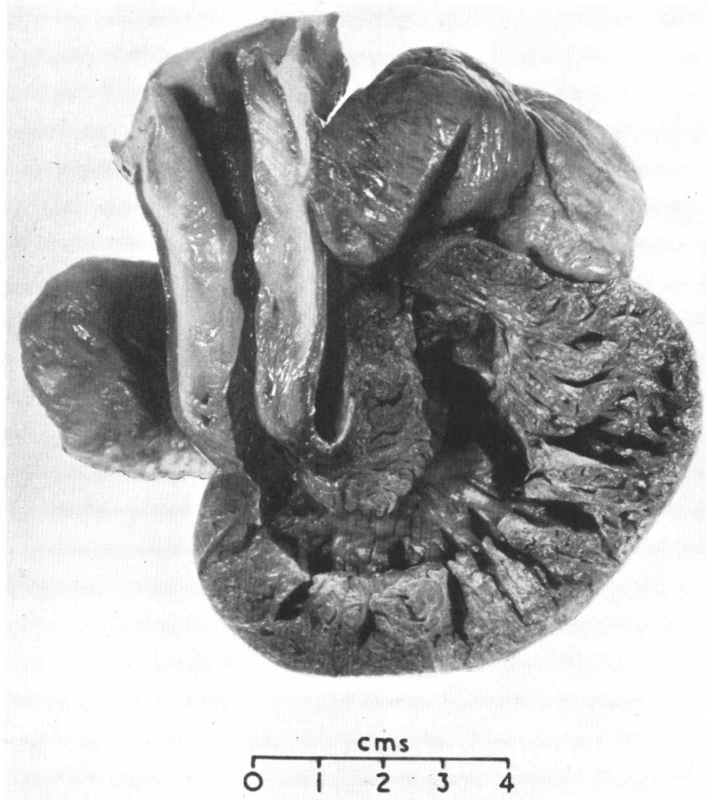
The specimens depicted in Figures 2–4 clearly show the muscle in the wall of the bulbus cordis, especially Figure 3 (the heart of a basking shark) in which the muscle is darker. The triple row of bulbar valves is also seen. The apparent constriction where the bulbus joins the ventricle should be noted (Fig. 2*b*) in view of what will be said later about constriction at the bulboventricular junction in cases of arrested fusion of the bulbus in man. The bulboventricular spur should also be noted.

In contrast to the large size of the bulbus cordis in the fish heart there is no separate bulbus cordis in reptiles, as is shown in the turtle's heart in Figure 5.

It is necessary to consider the reptilian heart because of other striking features which it presents. The turtle's heart shown in Figure 5 has two atria, the right being much larger than the left, and one common ventricle. The most striking feature is that three large vessels leave the common ventricle; a pulmonary artery and two systemic aortae, a right which supplies the abdominal viscera and the lower extremities and a left which supplies the head and the upper extremities.

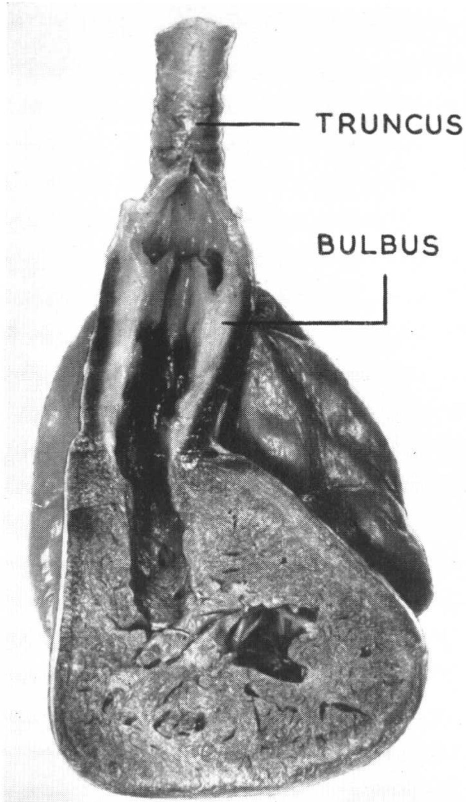


a

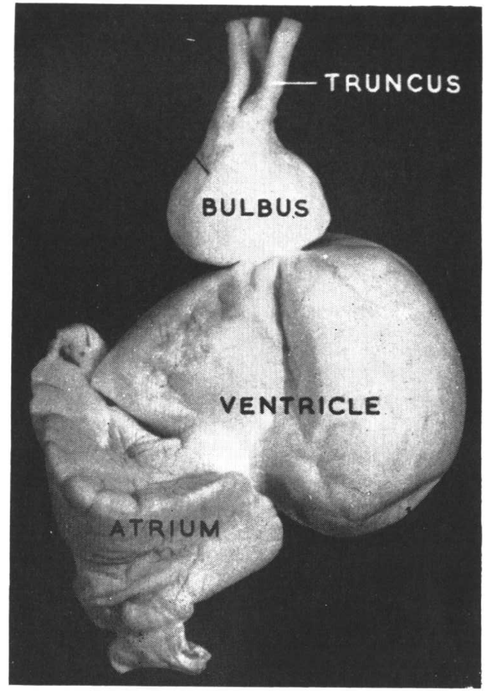


b

2. The heart of a skate to show the common ventricle and bulbus cordis. Note the bulboventricular spur.



3. The heart of a basking shark to show the common ventricle, the bulbus cordis and the truncus arteriosus. The continuity of ventricular and bulbar muscle is well shown.



4. The heart of an angler fish (Museum of Royal College of Surgeons). The bulbus forms a solid almost globular mass. The atrium and ventricle are well seen.