HANDBOOK OF PEDIATRIC CARDIOLOGY

Second Edition

By

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University Park Press Baltimore

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preface to the second edition

When we decided to revise this *Handbook* we anticipated a quick and easy task. After all, we reasoned, there had been relatively few changes in the field of pediatric cardiology in the last nine years. We would need to revise the sections on surgical results in most of the chapters, since there had been significant improvements in surgery for small infants. We anticipated that it would require at the most a few months of spare time.

As we began to examine the details of what we were going to change we found that there were six entities that we had not discussed in the first edition that warranted separate chapters. Thus we have added chapters on single ventricle, hypertension, atherosclerosis, mitral valve prolapse, hypoplastic left heart syndrome, and hypoplastic right heart syndrome. There were three others that we had discussed briefly that now merited separate chapters of their own: arrhythmias, pericarditis, and problems associated with cyanotic heart disease. It also seemed desirable to include a chapter on a new and exciting diagnostic tool, echocardiography. In order to maintain the handy size of the previous edition, we thought it necessary to reduce our coverage in other areas. For example, the chapter in the first edition entitled "Natural History of Ventricular Septal Defect" was combined with the chapter on Ventricular Septal Defect. We have also eliminated or shortened sections on other subjects that we thought to be of lesser importance.

Although we had anticipated a few of these changes, we were astonished at how many other changes were needed, especially in areas where we thought our notions were relatively stable. In a surprising number of instances we found ourselves saying, in effect, "Did we really say that?" We found that in a number of ways our opinions and handling of even some common anomalies had changed over the years. By the time we finished, there were changes in all except two of the chapters and over a year had elapsed since we started. The number of changes in this revision are testimony to the growth of pediatric cardiology since the first edition of the *Handbook*.

acknowledgments

"For this relief, much thanks."
—Hamlet

The authors would like to acknowledge the aid of several colleagues who reviewed chapters relating to their areas of special expertise and knowledge. In particular we would like to thank Drs. Carlos R. Ayers (Chapter 35, Hypertension), Karen S. Kuehl (Chapter 36, Atherosclerosis), J. Owen Hendley (Chapter 37, Infective Endocarditis), and Thaddeus Kelly (Chapter 41, Connective Tissue Disorders). Our thanks also to Dr. Marius Hubbell for his aid in preparing many of the new illustrations and to Ralph Davis for his excellent typing and his patience with the changes wrought by having three authors.

contents

PART I
Elements of Cardiology

This section of the Handbook contains materials relating to the basics of heart disease, with the emphasis on congenital cardiac anomalies. Information regarding genetic and teratogenic aspects of congenital heart disease, as well as discussions on cardiac anatomy and the hemodynamics of heart disease, is included.

The latter part of the section concerns the tools available to the physician. We have limited our discussion to those parts of the physical and laboratory examinations that are pertinent to cardiology. The concluding chapter deals with aspects of congestive heart failure.

This section may be read in its entirety, or it may be skipped over at this time and attention directed to the sections on congenital and acquired heart disease. The reader may then refer back to these chapters to gain a further understanding of the parts of the cardiovascular examination as they relate to the descriptions of the cardiac lesions in the latter parts of this book.

chapter 1

ANATOMY OF THE HEART

POSITION AND EXTERIOR APPEARANCE

An understanding of cardiac anatomy includes an awareness of the heart's position within the thorax. Correct interpretation of plain chest roentgenograms as well as angiocardiograms demands this precise knowledge.

As seen in Figure 1, the apex of the heart is directed anteriorly, inferiorly, and to the left. The right border of the cardiac silhouette is made up of the superior vena cava, the right atrium, and the inferior vena cava. The right ventricle does not ordinarily form any part of this border. The left border is formed predominantly by the left ventricle. The left atrium lies in the midline

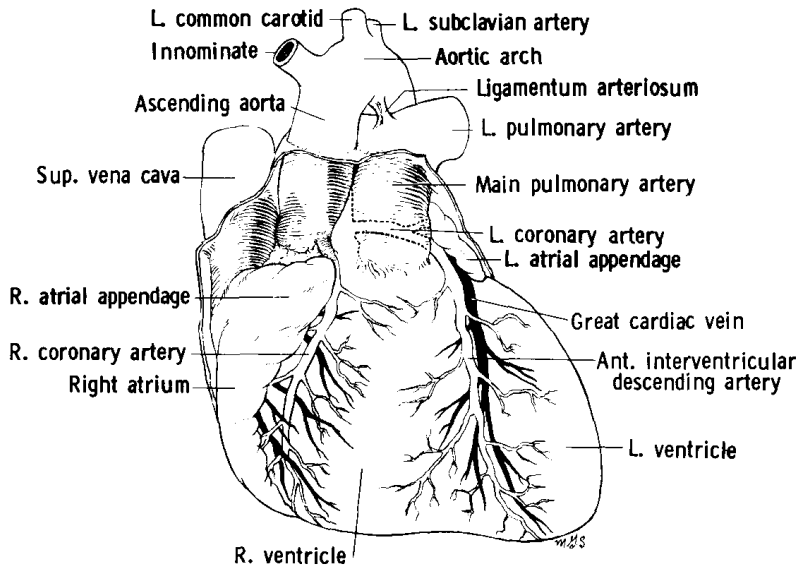

Figure 1. Anterior view of in situ heart and great vessels. The pericardium has been removed and its cut edge is shown reflected from the great vessels. Coronary veins are shown in solid black and coronary arteries in outline.

posteriorly and, because of the short, fixed pulmonary veins, anchors the heart.

The right ventricle is separated from the right atrium by the right atrioventricular groove and from the left ventricle by the anterior interventricular groove. The right coronary artery and the anterior descending branch of the left coronary artery, respectively, lie in these grooves. The coronary sinus lies posteriorly in the left atrioventricular groove.

Figure 1 also illustrates the origin of the pulmonary trunk—it rises anteriorly from the upper left border of the heart. After bifurcating, the right pulmonary artery runs rightward behind the ascending aorta and superior vena cava, and the left pulmonary artery runs directly left and somewhat posteriorly. The aorta originates centrally within the heart, with the aortic valve being inferior, posterior, and slightly to the right of the pulmonic* valve. After the origins of the innominate, the left common carotid, and the left subclavian arteries, the arch of the aorta passes behind the bifurcation of the main pulmonary artery.

Ordinarily, four pulmonary veins—two on either side—enter the left atrium. The right veins pass behind the right atrium to enter the right side of the left atrium and the left veins enter the left side of the left atrium.

RIGHT HEART (Figures 2 and 3)

The superior vena cava enters the right superior portion of the right atrium slightly forward, so that it aims toward the tricuspid orifice. The inferior vena cava enters the lower right atrium in a more medial position, slightly forward and leftward, so that it aims toward the fossa ovalis. Anterolateral to the orifice of the inferior vena cava lies the Eustachian valve, and anteromedial to this is the coronary sinus ostium. The anterior portion of the right atrium is trabeculated and thin walled, as is the right atrial appendage, which extends anteriorly to lie over the root of the aorta.

The atrial septum lies posteriorly and medially. A thin, fibrous depression, the fossa ovalis, is found in the center of the septum, and the muscular portion of the septum forms the limbus fossa ovalis.

The tricuspid valve leaflets are placed anteriorly, posteriorly, and medially. The medial, or septal, cusp lies in close relation to the membranous portion of the ventricular septum.

The inflow portion of the right ventricle is heavily trabeculated. Several papillary muscles attach to the tricuspid valve. The anterior papillary muscle originates from the moderator band and its chordae tendineae attach to the anterior and posterior cusps. The posterior papillary muscle attaches to the

*Pulmonic in this handbook refers to the valve, and pulmonary refers to the lungs. For example, pulmonic insufficiency means regurgitation through the valve, whereas pulmonary insufficiency refers to ventilatory insufficiency of the lungs.

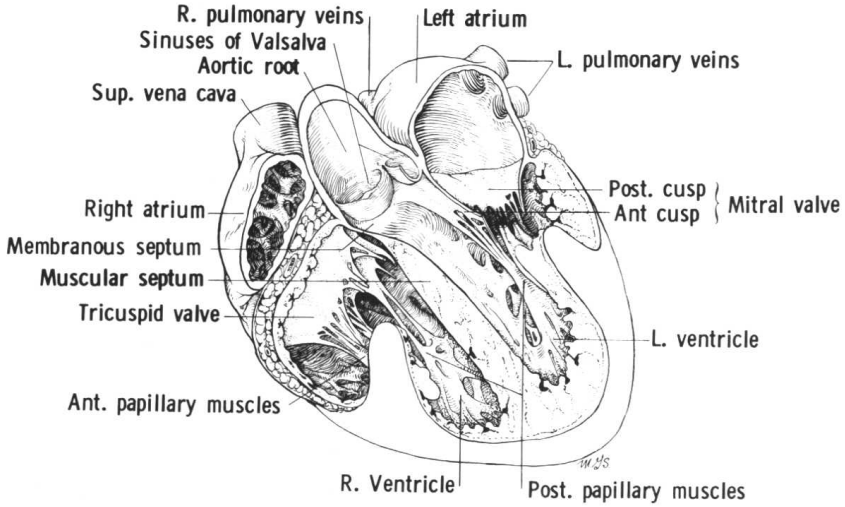

Figure 2. Semifrontal section of the heart. Note the intimate relation of the mitral and aortic valves. Note also the rightward angulation of the membranous portion of the ventricular septum.

posterior and septal cusps. A small, medial papillary muscle arises from the crista supraventricularis at its junction with the septal band and attaches to the anterior and septal cusps.

The outflow tract, or infundibulum, of the right ventricle is a smooth-walled, tubular structure delineated from the inflow portion by four prominent muscular bands: the crista supraventricularis and the parietal, moderator, and septal bands.

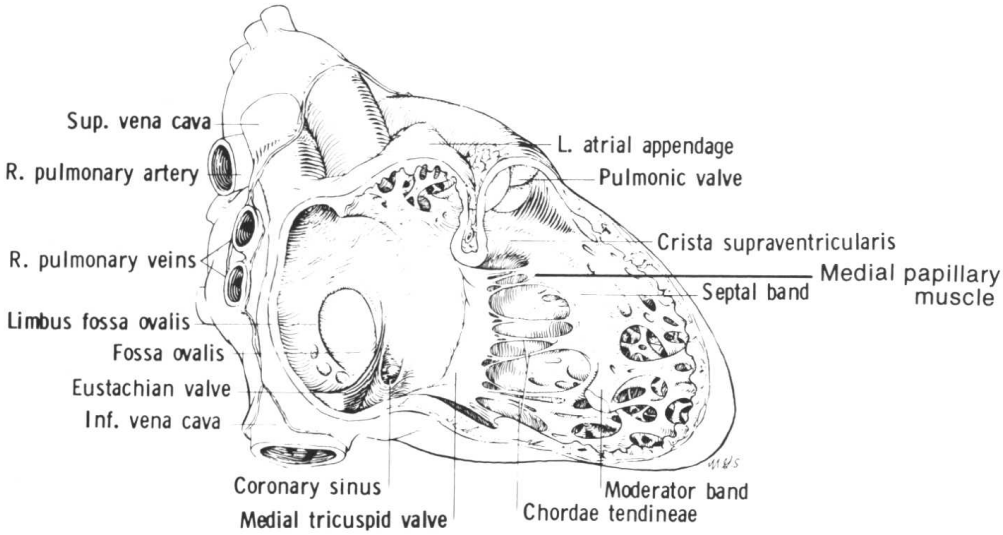

Figure 3. Semischematic view of the opened right side of the heart, illustrating both the right atrium and the right ventricle. Note the heavy trabeculation of the right ventricle in contrast to the smoother left ventricle shown in Figure 4.

The pulmonic valve, which lies at the top of the right ventricular outflow tract, has three cusps: right, left, and anterior.

LEFT HEART (Figures 2 and 4)

The left atrial wall is thicker than the right, and its internal surface is smoother. The left atrial appendage arises superiorly and anteriorly, and extends to the left of the pulmonary trunk to form a small portion of the left upper border of the cardiac silhouette, as viewed *en face*.

The mitral valve consists of two major cusps: a large anterior, or aortic, cusp and a posterior cusp. Usually, two small commissural cusps can also be identified.

The left ventricle has the shape of a megaphone, with the small end forming the apex. The septal surface of the left ventricular cavity is smooth and the free (parietal) wall is trabeculated, although the trabeculae are finer than those in the right ventricle.

Two papillary muscles—the anterior and the posterior—are present. Each attaches via its chordae to both the anterior and the posterior mitral valve cusps.

The ventricular septum is predominantly muscular. It presents a concave surface to the left ventricle and a convex surface to the right ventricle. The small membranous portion is located below the right and posterior aortic valve cusps, as viewed from left ventricle. The medial cusp of the tricuspid valve inserts across the membranous ventricular septum, dividing it into two portions. One portion lies between the left ventricle and the right atrium, the other between the left ventricle and right ventricle. The rightward angulation of the upper part of the ventricular septum forms the floor of the left ventricular outflow tract.

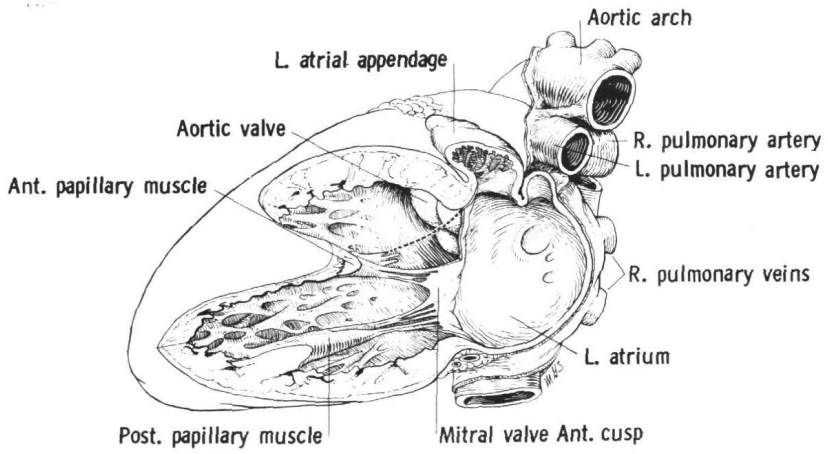

Figure 4. Semischematic view of the opened left side of the heart. A portion of the anterior leaflet of the mitral valve has been cut away (----) in order to view the aortic valve.

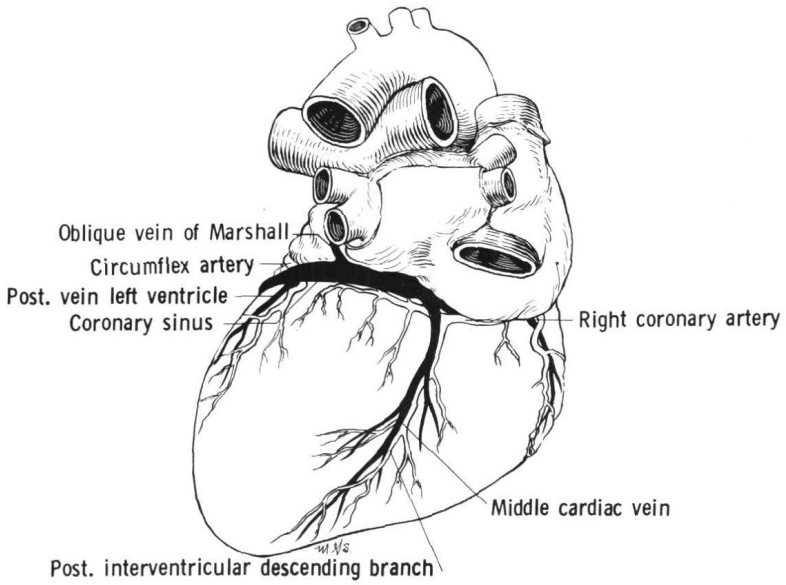

Figure 5. View of diaphragmatic (posterior, inferior) aspect of the heart and great vessels. Coronary vessels are indicated as in Figure 1.

The aortic valve consists of three semilunar cusps: right, left, and posterior. The coronary ostia lie within the sinuses of Valsalva of the right and left cusps. The left posterior cusps are in fibrous continuity with the anterior cusp of the mitral valve.

CORONARY VESSELS (Figures 1 and 5)

The right coronary artery arises from the right aortic sinus of Valsalva. It gives off several branches to the right atrium and right ventricle, including the posterior interventricular descending branch, which runs in the posterior interventricular groove. The left coronary artery originates from the left aortic sinus of Valsalva. It bifurcates within 1 or 2 cm into an anterior interventricular descending artery, which runs in the anterior interventricular groove, and a circumflex artery, which lies in the left atrioventricular groove. The branching pattern of the coronary arteries is subject to a great deal of variation, including an occasional third ostia in the right aortic sinus of Valsalva.

The major coronary veins are the great cardiac vein, the middle cardiac vein, and the posterior vein of the left ventricle. These all enter the coronary sinus from the left atrium, as does the oblique vein of Marshall.

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