Clinical Cardiology

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3rd edition

3rd Edition

Clinical Cardiology

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Clinical Cardiology

This book is dedicated to
Margaret McIlroy and the memory of Ethel Sokolow

Preface

The large amount of published research in cardiology in the past few years and the favorable response to our book from colleagues, readers, and reviewers have encouraged us to make a fairly thorough revision for this third edition. Major revisions are in the chapters on cardiac physiology, noninvasive investigations, coronary disease, and congenital heart disease. Although our emphasis has always been on clinically relevant new material, we have included, when pertinent, the physiologic underpinnings necessary to enhance understanding.

We have attempted to integrate new data from the literature with our experience as clinicians and academicians. In an attempt to make access to the literature easier for the student, we have arranged the references in the larger chapters under subheadings. We want to affirm our belief that cardiology is a scholarly field of medicine; that cardiology cannot be practiced today as it was practiced ten years ago or even five years ago—or in some instances one year ago; and that the practitioner who hopes to achieve and maintain competence in cardiology must continue to hound the library for new data as they become available for publication.

We again wish to thank our friends, readers, and reviewers for their comments and criticisms. We take their help seriously and make every effort to balance their opinions with our own. We hope that they will continue to let us know their views.

We wish to thank our colleagues Dr Elias H. Botvinick, Dr Erik Carlsson, Dr Gordon Gamsu, Dr Mervin J. Goldman, Dr Robert Grover, Dr Arthur Hollman, Dr John Hutchinson, Dr Oscar Rambo, Dr Nelson Schiller, and Dr Norman Silverman for providing echocardiograms, electrocardiograms, pathologic data, radioisotope studies, and x-rays from their files.

We wish to thank our publisher, Dr Jack Lange, and chief editor, Dr James Ransom, for their valuable advice and contributions to the success of this book. We also wish to thank Ms Diane Pederson and Ms Lorraine Matthews for valuable organizational and secretarial assistance.

A translation of *Clinical Cardiology* has been completed in Spanish; and French, German, Greek, Italian, Polish, Portuguese, and Turkish translations are in preparation.

Maurice Sokolow
Malcolm B. McIlroy

San Francisco
September, 1981

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Anatomy & Physiology of the Circulatory System | 1

ANATOMY OF THE HEART

The normal heart lies within its pericardial sac in the middle of the thorax slightly to the left of the midline. The low-pressure right atrium and right ventricle occupy the anterior portion of the heart and the higher pressure left ventricle and atrium lie posteriorly. The long axis of the heart, from the apex of the left ventricle to the root of the aorta, runs upward and backward at an angle of about 30 degrees from the horizontal plane and 45 degrees from the sagittal plane of the body (Fig 1–1). The apex of the heart rests on the upper surface of the diaphragm, which lies close to the posterior and inferior surfaces of the heart. The lie of the heart varies with the build of the patient and with respiration. It assumes a more vertical position during inspiration and in tall, thin persons, and a more horizontal position during expiration and in persons of heavier build.

EXTERNAL APPEARANCE

Anterior Aspect

As viewed from the front (Fig 1–1), the largest area of the surface of the heart is formed by the triangle-shaped right ventricle, with the pulmonary trunk arising from the apex of the triangle. Above and to the right of the right ventricle, one can see the right

Figure 1–1. Anterior view of the heart.

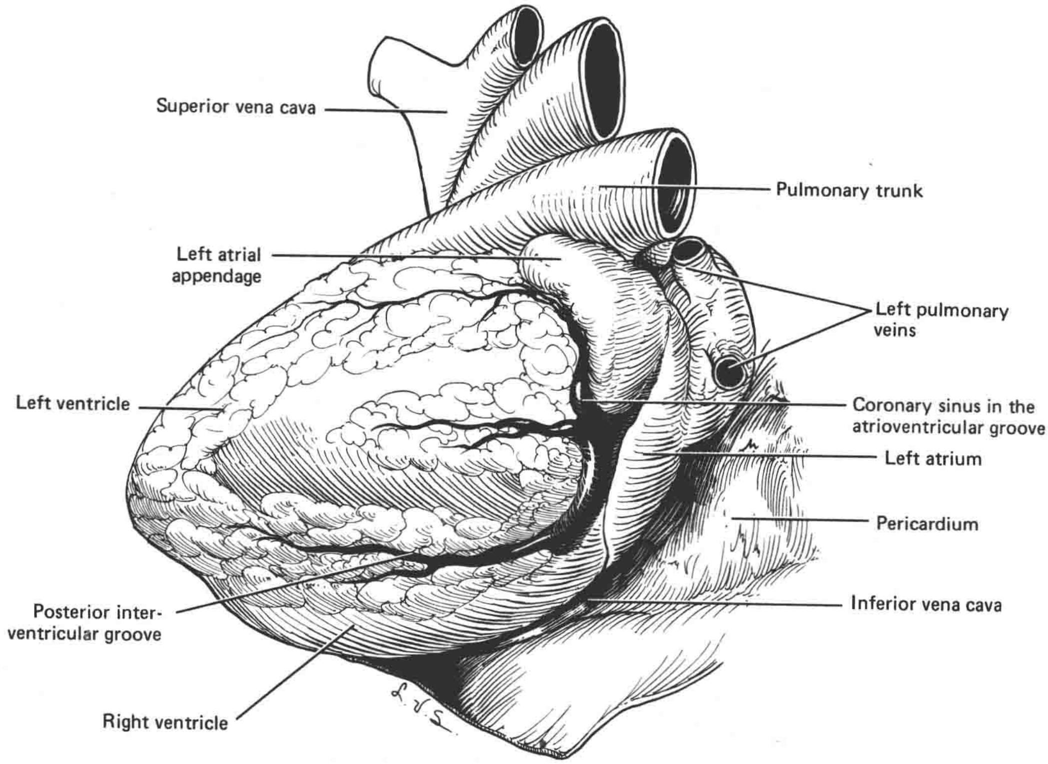

Figure 1–2. The heart viewed from the left side with the apex raised.

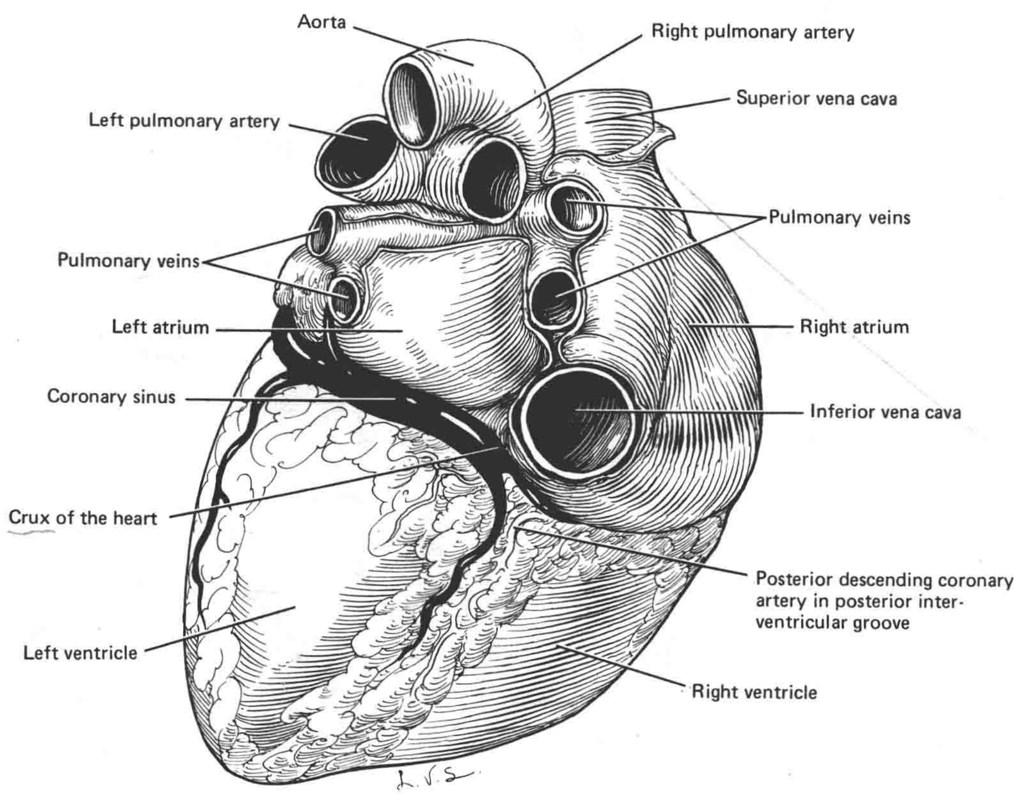

Figure 1–3. The heart viewed from below and behind.

atrium—or, more specifically, the right atrial appendage—as an ear-shaped structure overlying the root of the aorta. The groove between the right atrium and ventricle (coronary sulcus) is often filled with fat and is occupied by the right coronary artery. Above the right atrium, the superior vena cava is seen entering the right atrium through the pericardium. The inferior vena cava lies on the diaphragmatic surface of the heart and enters the right atrium from the back. The anterior aspect of the heart reveals only a small part of the left ventricle, lying to the left of the right ventricle and forming the apex of the heart. The anterior interventricular sulcus often contains fat and is occupied by the anterior descending branch of the left coronary artery. The only portion of the left atrium visible from the front is the left atrial appendage, which lies above the ventricle and curves around the left side of the origin of the pulmonary trunk. The lungs normally cover most of the anterior surface of the heart, especially during inspiration, leaving only a small area apposed to the back of the sternum and left ribs.

Left-Sided Aspect

As viewed from the left side (Fig 1–2), the left ventricle and left atrium occupy most of the surface of the heart. The posterior interventricular groove separates the left ventricle above from the right ventricle below. The posterior descending branch of the right coronary artery lies in this groove. The atrioventricular groove runs almost vertically in this view, separating the left ventricle from the left atrium. The coronary sinus and the circumflex branch of the left coronary artery lie in this groove and complete the ring of blood vessels forming the base of the corona (crown) after which the blood vessels supplying the heart are named.

Posterior Aspect

The back of the heart mainly rests on the diaphragm and is largely occupied by the left atrium and ventricle plus portions of the right atrium and ventricle, as shown in Fig 1–3. The point at which all 4 chambers meet posteriorly is called the crux of the heart because of the cross-shaped pattern of blood vessels lying at the junction of the posterior interventricular groove and the atrioventricular groove. The vessels forming the cross are the coronary sinus and the posterior descending coronary artery. This latter vessel may be a branch of either the right or the circumflex branch of the left coronary artery depending on whether the right or left coronary artery is the larger (dominant) vessel. The pulmonary veins enter the back of the left atrium. The pattern may vary, but 2 right and 2 left pulmonary veins are normally present.

Right-Sided Aspect

When viewed from the right side, the right atrium and ventricle occupy most of the surface, as shown in Fig 1–4. The superior and inferior venae cavae enter the atrium at the back, and the aorta runs upward from the middle of the heart. The outflow tract of the right ventricle and the pulmonary trunk form the upper border of the heart in this view.

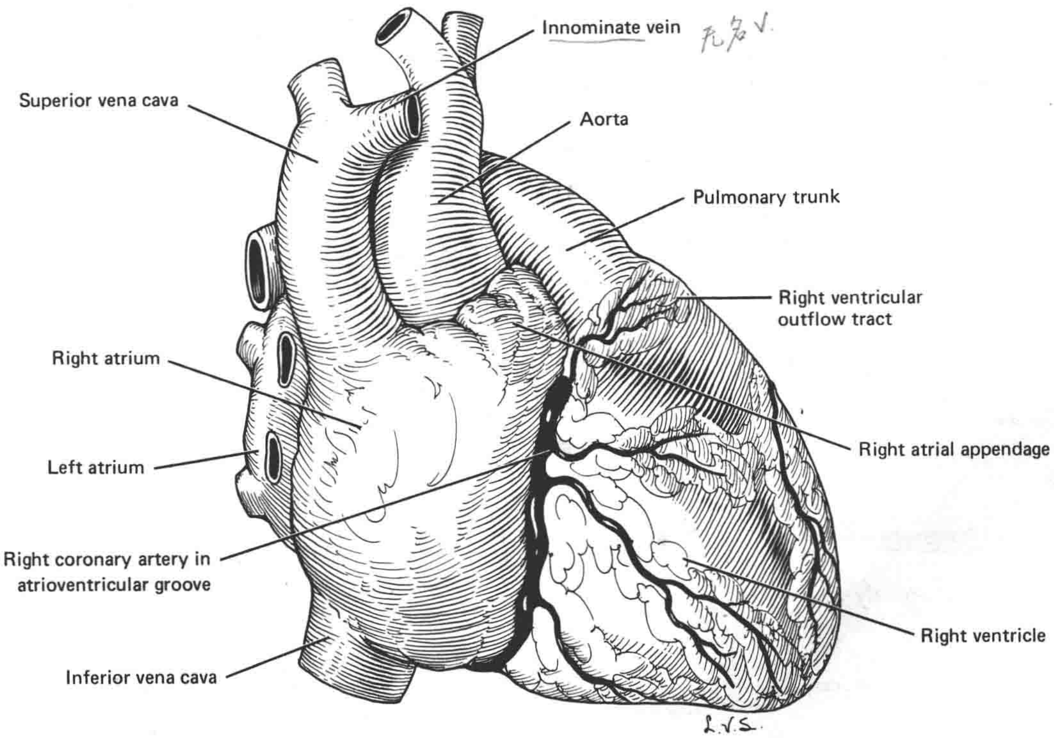

Figure 1–4. The heart viewed from the right side.

The Great Vessels

The main pulmonary artery (pulmonary trunk) runs upward and to the left in front of the aorta and leaves the pericardial sac before dividing into its right and left branches. The left pulmonary artery continues to arch backward in the same line as the main trunk, while the right branch turns laterally behind the ascending aorta and the superior vena cava to reach the hilum of the right lung. The bifurcation of the pulmonary artery lies on the roof of the left atrium and above the left main bronchus.

The aorta arises deep within the heart, and its proximal portion is covered by the right atrial appendage. It runs upward beside the superior vena cava before giving off its first and largest (innominate) branch, which shortly divides into the right common carotid and right subclavian branches. The aortic arch passes backward and to the left, giving off its left common carotid and left subclavian branches before crossing the left pulmonary artery. The close relationship between the left pulmonary artery and the aorta is due to the lie of the ductus arteriosus in the fetal circulation. This vessel, which runs from the left pulmonary artery to the descending aorta in an almost direct line in the fetus, persists as a remnant—the ligamentum arteriosum—in adults. The point at which it joins the aorta is termed the isthmus of the aorta because there is sometimes a narrowing at this level. The aorta is weakest at this point, and traumatic aortic tears usually occur at this level.

THE CHAMBERS OF THE HEART

The Right Atrium

The right atrium consists of 2 embryologically distinct portions, as shown in Fig 1–5. The more posterior thin-walled portion into which the venae cavae and coronary sinus empty is formed from the sinus venosus and is composed of tissue similar to that of the great veins. The more anterior muscular portion includes the right atrial appendage and the tricuspid valve ring. The fossa ovalis lies in the middle of the thin-walled portion and is the site of the foramen ovale. This interatrial communication, which is present during fetal life, permits the flow of oxygenated placental blood from the inferior vena cava into the left heart. The foramen ovale remains open or potentially open in about 15% of normal subjects, but since it is a flap valve that only allows flow from right to left, it is normally functionally closed.

The Right Ventricle

The right ventricle is triangular in shape and forms a crescentic, shallow structure wrapped over the ventricular septum. It can be divided, as shown in Fig 1–6, into a lower inflow portion, containing the tricuspid valve and its chordae, and an upper outflow tract, from which the pulmonary trunk arises. The line of demarcation between the 2 portions consists of bands of muscle formed by the crista supraventricularis, the parietal band, the septal band, and the moderator band. The outflow tract of the right ventricle is derived from the embryologically distinct bulbus cordis—in contrast to the inflow portion, which arises from ventricular tissue.

The Left Atrium

The left atrium, like the right, is composed of a veinlike portion, into which the pulmonary veins drain, and a more muscular anterior portion, which includes the left atrial appendage. Its wall is slightly

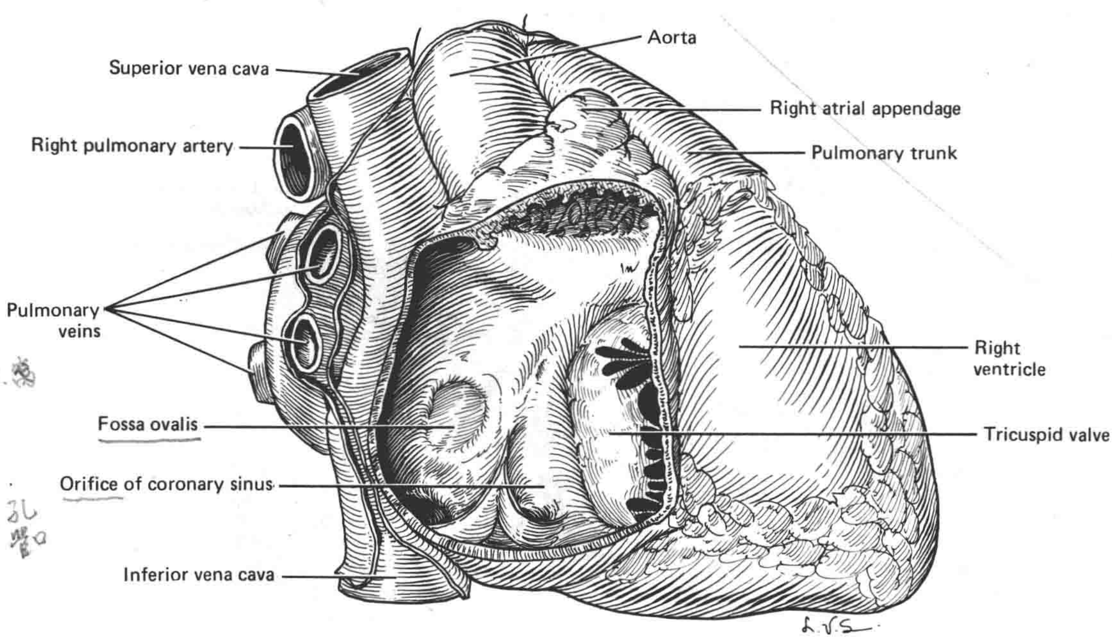

Figure 1–5. View of the right heart with the right wall reflected to show the right atrium.

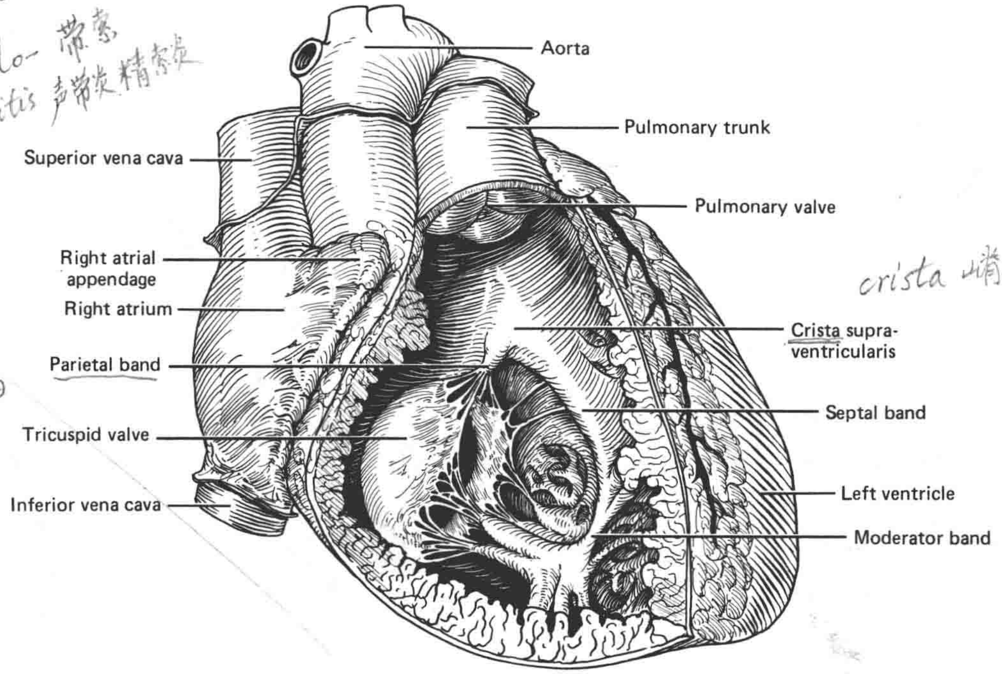

Figure 1–6. Anterior view of the heart with the anterior wall removed to show the right ventricular cavity.

thicker than that of the right atrium, and the thinner area, corresponding to the fossa ovalis, can be seen on its right upper surface (Fig 1–7).

The Left Ventricle

The left ventricular cavity is shaped like an egg. The base of the egg is formed by the mitral valve ring. The wall of the left ventricle is 3–4 times as thick as that of the right ventricle and accounts for about 75% of the mass of the heart. The aortic and mitral valve rings lie close to one another, with the larger anterior mobile cusp of the mitral valve adjacent to the left and posterior cusps of the aortic valve. The posterior immobile cusp of the mitral valve is shorter and, together

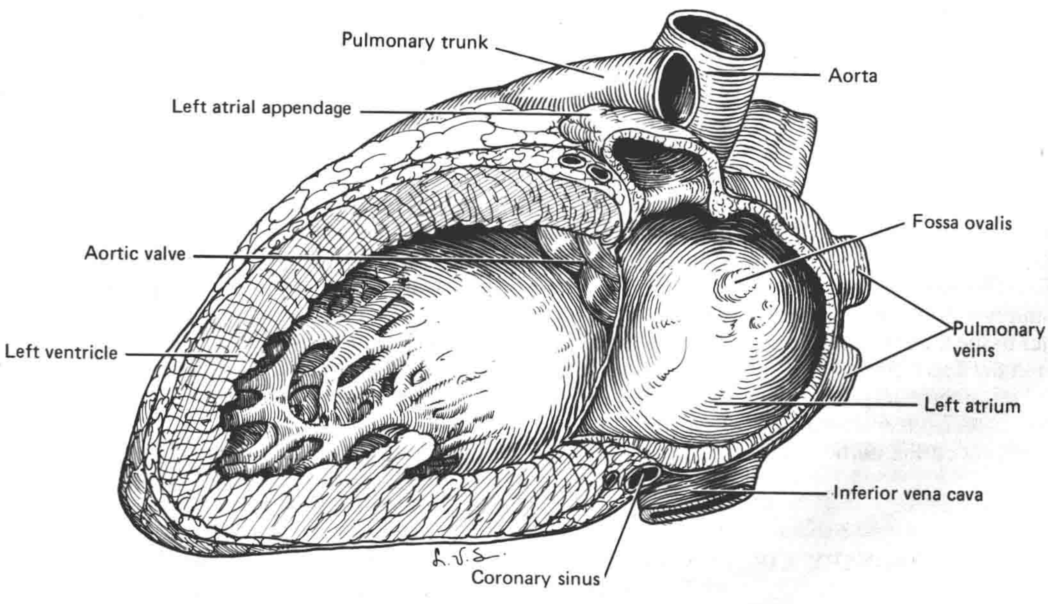

Figure 1–7. View of the left heart from the left side with the left ventricular free wall and mitral valve cut away.

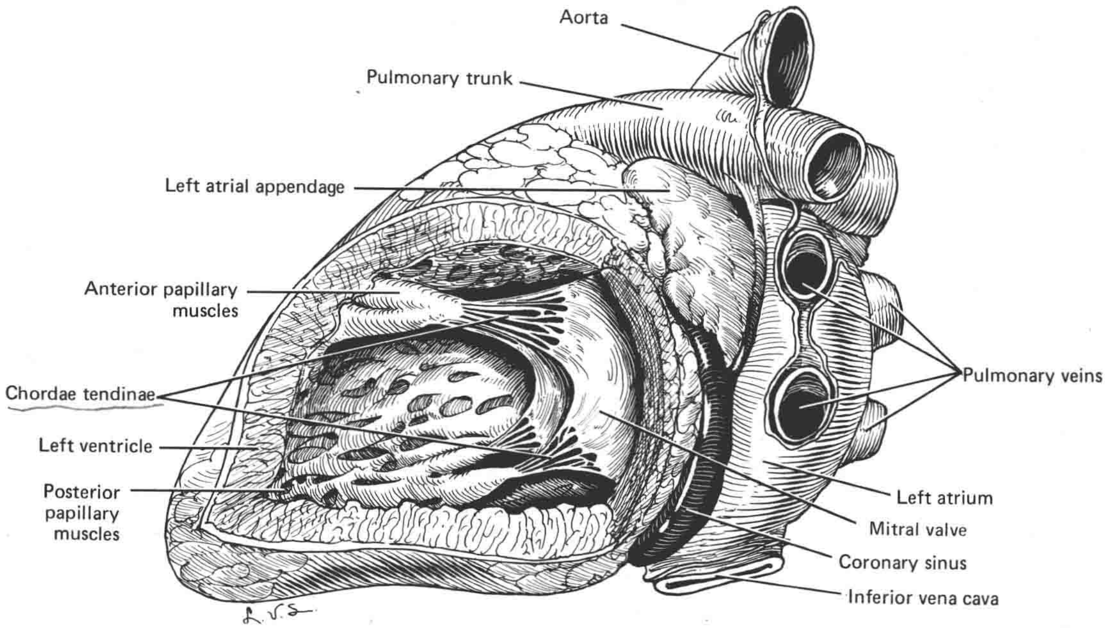

Figure 1–8. View of the left heart with the left ventricular wall turned back to show the mitral valve.

with the anterior cusp, is tethered to the anterior and posterior papillary muscles in a parachutelike fashion by chordae tendineae, some of which are shared by the 2 cusps as seen in Fig 1–8. The interventricular septum, which forms the anterior aspect of the left ventricle, bulges into the right ventricle, making the cross section of the mid portion of the left ventricle circular in shape.

CARDIAC VALVES

The **tricuspid valve** is a thin, filmy tripartite structure with anterior, posterior, and medial cusps. The membranous portion of the interventricular septum lies beside its medial cusp. The **mitral valve** is thicker than the tricuspid valve and is shaped like a bishop's hat (miter) in which the anterior surface (anterior cusp) is longer and wider than the posterior surface. The **pulmonary valve** is composed of pocketlike anterior right and left cusps. It is constructed of thinner tissue than the **aortic valve,** which lies farther down in the heart. It too has 3 cusps—the right (coronary), left (coronary), and posterior (noncoronary) cusps—associated with corresponding dilatations of the aorta called the aortic sinuses or sinuses of Valsalva.

THE CORONARY CIRCULATION

The coronary arteries are more variable in pattern than any other part of the cardiac anatomy. The 2 main coronary arteries—left and right—arise from the right and left aortic sinuses within the pockets of the aortic valve cusps. Either vessel may predominate and supply the posteroinferior portion of the heart. In 30% of persons the **left coronary artery** is the smaller of the 2. The left coronary artery is likely to be dominant in patients with congenital aortic stenosis or bicuspid aortic valve. The left coronary artery runs behind the main pulmonary artery as a short main stem about 1 or 2 cm long before dividing into an **anterior** and a **circumflex branch.** The anterior branch usually has a **descending branch** that follows the interventricular groove. The circumflex branch follows the atrioventricular groove, curving around to the posterior surface of the heart. The area between these 2 vessels, each of which is defined by a course within a groove, is supplied by branches from one artery or the other. Thus, the left coronary artery usually consists of 3 branches, with the mid branch arising from one of the more readily definable arteries. The circumflex branch is larger in persons with a dominant left coronary pattern. In this case, the vessel may run as far as the crux of the heart and even give off the posterior descending branch, which runs in the posterior interventricular groove.

The right coronary artery runs in the atrioventricular groove, downward and to the right, before curving around to the back of the heart to reach the crux, giving off a posterior descending interventricular branch. An anterior right atrial branch usually arises near the origin of the right coronary artery. It usually supplies a branch to the sinoatrial node. The atrioventricular node is also commonly supplied by a branch of the right coronary artery that arises from the posterior descending branch.

Most of the coronary venous drainage is into the **coronary sinus.** The few veins that drain directly into the cardiac chambers are called thebesian veins. The main venous drainage of the left ventricle is via the great cardiac vein, which runs with the anterior descending branch of the left coronary artery before joining with the posterior cardiac vein to form the coronary sinus.

The anatomy of the coronary vessels is of great importance in the interpretation of coronary arteriograms and in coronary artery surgery. The subject is discussed in more detail in Chapter 8.

CONDUCTION SYSTEM

The **sinoatrial node,** which initiates the normal cardiac impulse, lies at the junction of the superior vena cava and the right atrium. The **atrioventricular node** is located in the right posterior portion of the interatrial septum near the base of the tricuspid valve. Poorly defined anterior, middle, and posterior atrial tracts connect the 2 nodes and conduct the cardiac impulse through the atrial tissue. The atrioventricular node is continuous with the **bundle of His,** which divides into a left and a right bundle branch at the top of the interventricular septum. The left branch divides again into anterior and posterior branches, and all 3 branches run subendocardially close to the septum before ramifying into the **Purkinje fibers,** which spread to all parts of the ventricular myocardium. The details of abnormal conduction pathways are given in Chapter 14.

LYMPHATICS

The lymphatics of the heart are arranged in 3 plexuses: subendocardial, myocardial, and subepicardial. The drainage is outward to the subepicardial plexus, where the vessels unite to form drainage trunks that follow the coronary arteries. They eventually form a single vessel that leaves the heart on the anterior surface of the pulmonary artery to reach a lymph node between the superior vena cava and the innominate artery. Few valves can be found in the cardiac lymphatics, and it appears that cardiac contraction provides the force that drives the lymphatic flow from the heart. No certain role for cardiac lymphatics has been established in disease. It has been suggested that endomyocardial fibrosis might be related to lymphatic obstruction. Cardiac transplantation, which inevitably severs the cardiac lymphatics, does not seem to produce any deleterious effect, but regeneration of lymphatic vessels is known to occur within 2 or 3 weeks after they have been divided.

CARDIAC NERVES

The heart is innervated both by cholinergic fibers from the vagus nerve and by adrenergic fibers arising from the thoracolumbar sympathetic system and passing through the superior, middle, and inferior cervical ganglions. The efferent cholinergic supply is confined to the atria. Fibers from the right vagus nerve supply the sinoatrial node and serve to control the heart rate and the force of atrial contraction. Fibers from the left vagus nerve supply mainly the atrioventricular node, but there is usually some cross-innervation. The atria also receive sympathetic fibers, but most of the adrenergic nerves pass to the ventricles, where they serve to increase the force of cardiac contraction. The heart also has an autonomic sensory innervation via small, mainly nonmedullated sympathetic fibers. These are thought to respond to nociceptive stimuli and to constitute the pathway through which cardiac pain is mediated.

Vagally innervated receptors are also widely distributed in the atria and ventricles. The atrial receptors discharge into myelinated fibers and send impulses up the vagus nerve that reduce sympathetic output to the kidneys. Their effect is to cause an increase in urinary volume and sodium excretion. The ventricular receptors are served by nonmedullated fibers. The endings are thought to be mechanoreceptors that respond to changes in ventricular pressure and reinforce the effects of the carotid and aortic baroreceptors.

MICROSCOPIC ANATOMY OF THE HEART

The basic heart muscle cell forms part of a syncytium in which the individual cells are joined together in an irregular fashion in bands and spirals without the well-defined tendons and bony attachments characteristic of skeletal muscle. The heart muscle cell differs from the skeletal muscle cell also in that it possesses inherent rhythmicity. This property varies with different types of cardiac muscle; it is most marked in nodal tissue and least notable in peripheral muscle cells. The subcellular arrangement of cardiac muscle cells (Fig 1–9A) is similar to that of skeletal muscle. The cells are about 30×10 μm in size and contain about 20–50 fibrils. Each fibril is about 1 μm in diameter and is composed of a series of sarcomeres, the basic muscle units. The cell contains a nucleus and numerous mitochondria. The limiting membrane is the sarcolemma, from which a sarcoplasmic reticulum invaginates the cell to form a complex tubular (T) system surrounding each fibril. The electrical activity triggering the contraction of each sarcomere passes through this complex membranelike structure.

The Sarcomere

The structural unit of the sarcomere is shown in Fig 1–9B. Its banded appearance results from overlap-

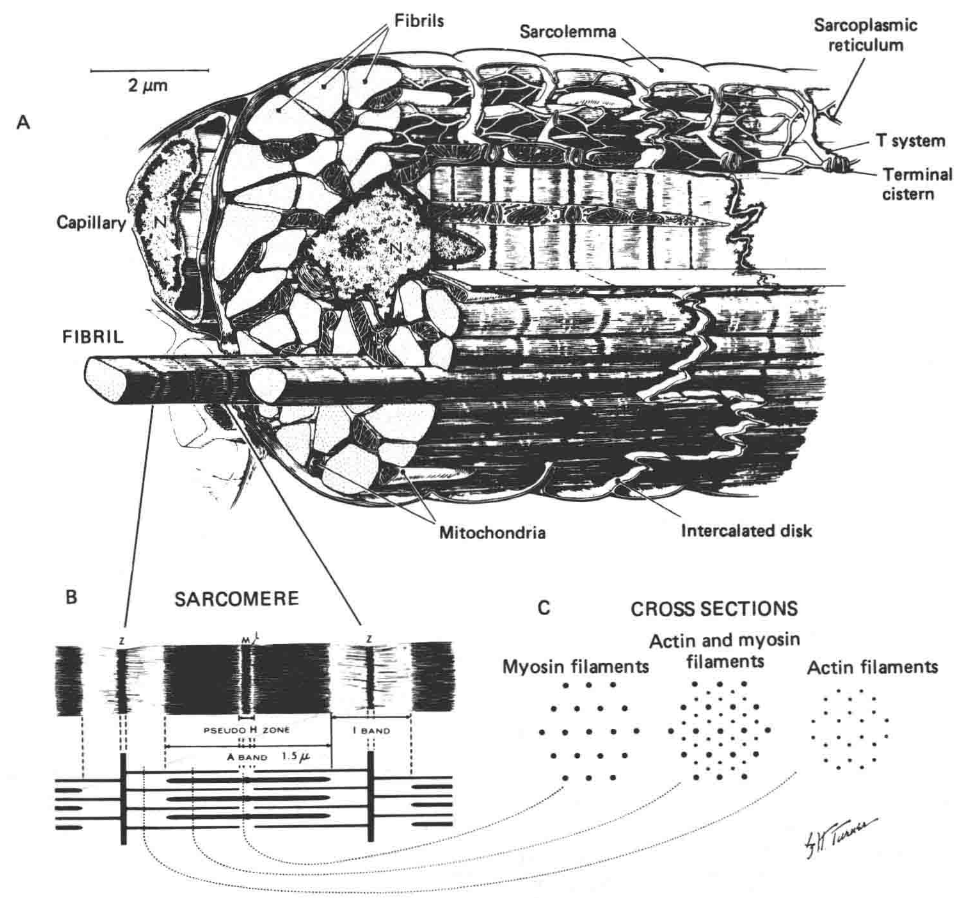

Figure 1–9. Diagram of cardiac muscle as seen under the electron microscope. *A:* A myocardial cell showing the arrangement of the multiple parallel fibrils. *B:* An individual sarcomere from a myofibril. A representation of the arrangement of myofilaments that make up the sarcomere is shown below. *C:* Cross sections of the sarcomere, showing the specific lattice arrangement of the myofilaments. N, nucleus. (Reproduced, with permission, from Braunwald E, Ross J Jr, Sonnenblick EH: Mechanisms of contraction of the normal and failing heart. *N Engl J Med* 1967;**277**:794.)

ping of the 2 major muscle proteins—actin and myosin—which accounts for the striated appearance. The wide dark A bands are formed by overlapping of the thicker myosin elements with the thinner, lighter actin filaments. The thinner dark Z lines indicate the end of one sarcomere and the beginning of the next. The lighter I bands are seen in areas in which only the actin filaments are present. The pattern of the sarcomere seen by electron microscopy varies with contraction and relaxation of the sarcomere. With contraction, as shown in Fig 1–10, the I band becomes shorter and the A band more dense. The Z lines come to lie closer together as the muscle contracts. When the muscle fibril is cut in cross section, a specific lattice pattern is seen (Fig 1–9C). In the zone in which the actin and myosin overlap (S zone), each thick myosin fiber is surrounded by 6 actin fibers. This hexagonal pattern is also seen in the lighter I band region. In the center of the sarcomere, where only myosin is present (M zone), the individual myosin filaments are arranged in a lattice pattern. A similar pattern is seen at the Z lines.

EMBRYOLOGY OF THE HEART

The embryology of the heart is as complex as that of any organ in the body. The process of development of the heart takes place mainly during the period between the second and sixth weeks of gestation; thus, the factors responsible for the development of congenital heart lesions probably operate in most cases before the diagnosis of pregnancy is clinically certain.

Primitive Heart Tube

The heart is formed by the folding of the primitive vascular tube, which appears in the splanchnic mesodermal tissue near the pericardial cavity at about the start of the third week of gestation. At first the primitive heart tube is straight, but differential growth soon forms a cardiac loop, as shown in Fig 1–11. Three more or less distinct portions of the tube can be distinguished, and it is convenient to describe them separately even though their development proceeds in parallel. The 3 portions are (1) the sinus venosus,

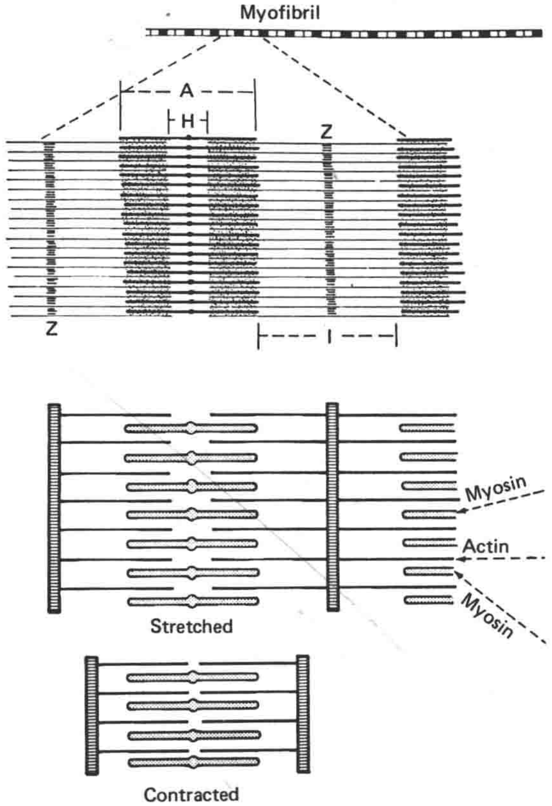

Figure 1–10. Muscular contraction. The myofibrils are composed of overlapping thick myosin filaments and thin actin filaments. The amount of overlap is diminished during stretching and increased during contraction. (Reproduced, with permission, from Rushmer RF: *Cardiovascular Dynamics,* 2nd ed. Saunders, 1970.)

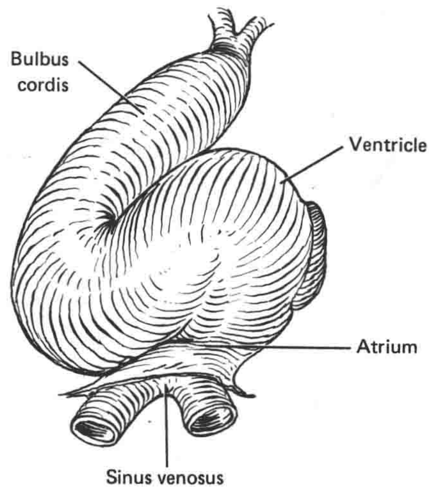

Figure 1–11. Formation of the cardiac loop.

(2) the cardiac loop, and (3) the aortic and branchial arches.

The Sinus Venosus

The most caudad portion of the primitive heart tube gives rise to the sinus venosus. As shown in Fig 1–12, this is an independent chamber during the early stage of development of the heart. It originally consists of 2 horns, each receiving a duct of Cuvier. The umbilical veins are formed from this structure, which ultimately gives rise to the superior and inferior venae cavae, the pulmonary veins, the coronary sinus, and the posterior portions of the right and left atria.

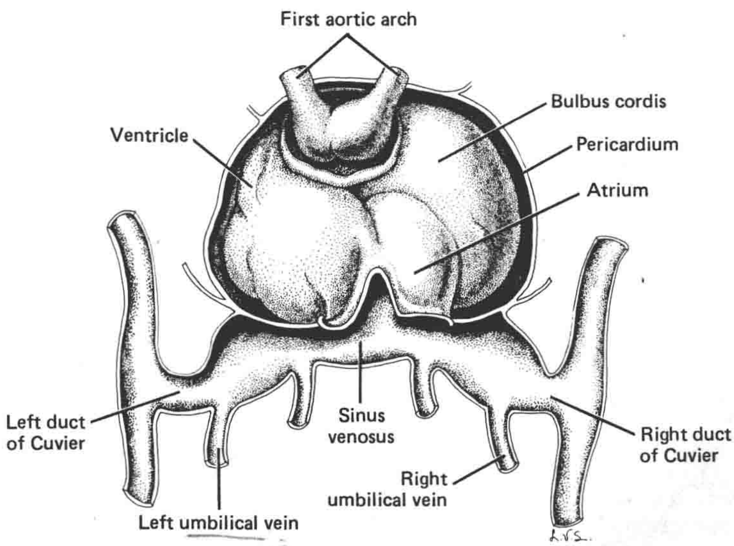

Figure 1–12. The sinus venosus, atrium, and ventricle as seen from the dorsal surface of an embryo at about the fourth week of gestation. (Modified and reproduced, with permission, from Davies J: *Human Developmental Anatomy.* Ronald Press, 1963.)

The Cardiac Loop

The intermediate portion of the primitive heart tube bends to form the cardiac loop, which twists on itself to form 3 distinct portions: the primitive atrium, the ventricle, and, more distally, the bulbus cordis. In the process of twisting, the primitive heart comes to lie in close apposition to its surrounding pericardial sac, as shown in Fig 1–12. The cardiac chambers are at first single; septation to form separate right- and left-sided atria and ventricles occurs at a later stage.

Aortic Arches

The most distal portion of the primitive heart tube forms the aortic sac; distal to this sac, 6 paired aortic arches appear sequentially. Some disappear but others persist to give rise to the great vessels. The basic original pattern of arches is shown in Fig 1–13, with the persisting vessels outlined. The third arch persists as the internal carotid artery; the left fourth arch forms the arch of the aorta; and the sixth arch gives rise to the pulmonary arteries and the ductus arteriosus.

Septation

The most complex stage of cardiac embryology is septation of the various parts of the heart. Septation in the atrium is depicted in Fig 1–14. A septum extends downward and forward toward the center of the heart where the endocardial cushions are located and from which the atrioventricular valves subsequently develop. This is the septum primum. A second atrial septum—the septum secundum—grows on the right side of the septum primum. A hole develops in the septum primum in the middle of the atrium, and atrial septation is never complete. The septum secundum does not extend all the way forward and downward to the endocardial cushions, and a persistent interatrial communication between the 2 atrial septa persists as the foramen ovale until birth.

Separation of the primitive ventricle into right and left chambers is accomplished by the development of an interventricular septum, which grows from the anterior wall of the common ventricle. Its free margin is aligned slightly to the right of the midline, toward the region of the endocardial cushions.

The most distal part of the primitive ventricle, the bulbus cordis, dilates to form the truncus arteriosus, which develops between the ventricles and the aortic arches. A spiral septum forms within the bulbus cordis

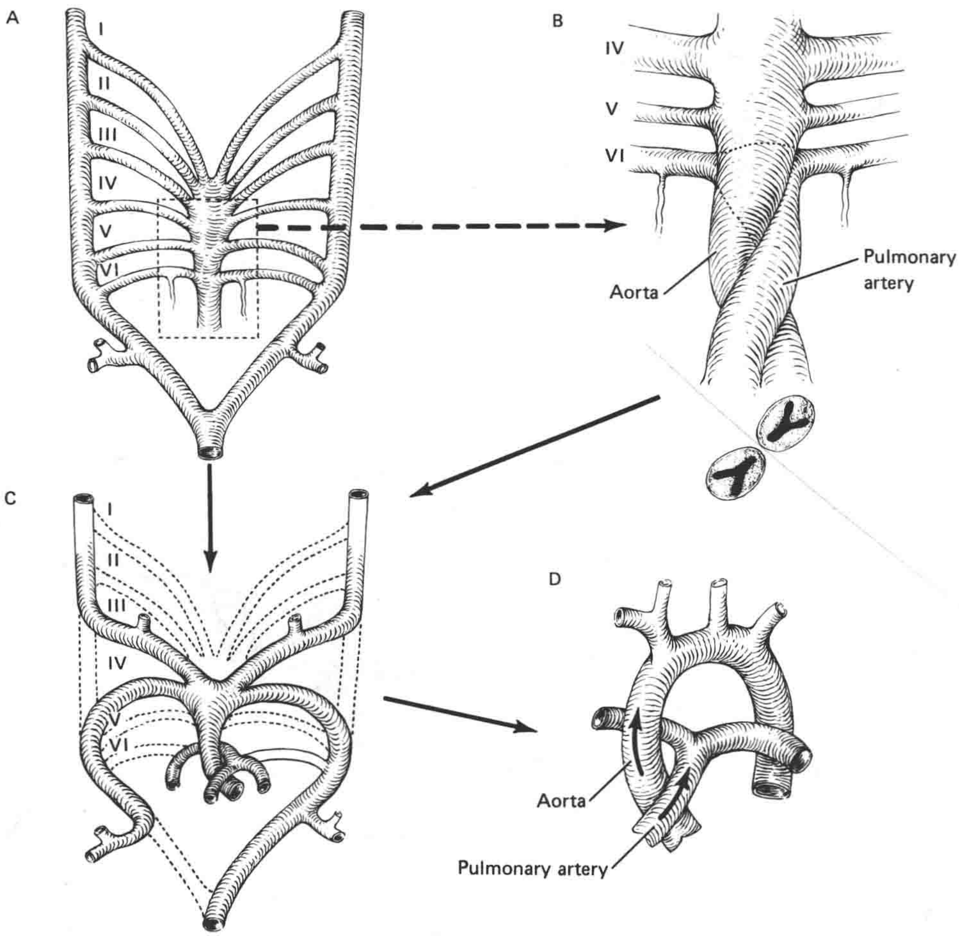

Figure 1–13. In *A,* the primitive arches are shown as paired structures. In *B,* the rotation and septation of the great vessels are seen. *C* shows the persistence of the third, fourth, and sixth arches to give the adult pattern shown in *D.*