Anatomy of the Heart
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Key Points
- External looping and internal septation to form four-chambered heart.
- Relationship of embryologic defects to congenital heart disease.
- Structure-function relationships of the pericardium.
- Distribution of the coronary arteries and anatomic variations.
- Definition of anatomic right and left atria and right and left ventricles.
- Structure of the four cardiac valves and concept of functional valve apparatus.
- Anatomy of the cardiac conduction system and cardiac innervation.

This chapter presents basic features of the anatomy of the heart and great vessels, including the embryologic development of these structures and their configuration in the mature state. Basic knowledge of cardiovascular anatomy is essential for effective diagnosis and treatment of cardiovascular diseases.

Embryologic Development

Basic Embryology

Development of the cardiovascular system occurs in the early first trimester fetus. Beginning at about 3 weeks’ gestation, elements of splanchnic mesoderm differentiate into a primitive cardiac tube and pericardial cavity, and vascular channels form and fuse to form blood vessels. The primitive cardiac tube results from the moving together and fusion of two lateral endothelial heart tubes. Subsequently, the epimyocardial mantle and cardiac jelly develop. These components differentiate into the endocardium, producing the internal endothelial lining of the heart, the myocardium forming the muscular wall, and the epicardium or visceral pericardium producing the outside covering of the heart. The recently formed cardiac tube is a single chambered structure and is composed of the following components, extending from inferior (caudal) to superior (cephalad): the sinus venosus, which connects to the major veins, the atrium, the ventricle, the bulbus cordis or conus, and the truncus arteriosus, which connects through six pairs of aortic arches to two dorsal aortae.

Initially, the single chambered heart is a straight tube residing in the pericardial cavity. The bulboventricular portion grows much more rapidly than the pericardial cavity. As a result, further extension in a longitudinal direction cannot occur, and the heart tube is forced to bend. The cephalic ventricular portion of the tube bends in a ventral and caudal direction and to the right, whereas the caudal atrial portion progresses in a dorsal and cranial direction and to the left. This process, known as dextro-bulboventricular looping, results in the atrial region establishing a position superior to the ventricular region and the cardiac apex being pointed to the left (Fig. 1.1).

The external shape changes are accompanied by a complex process of internal septation that leads to the formation of a four-chambered heart (Fig. 1.2). In the atrium, a septum primum forms and then develops two openings: ostium primum and ostium secundum. A septum secundum then develops on the right of the septum primum. The foramen ovale is formed in the midportion of the developing interatrial septum as a result of the growth and positioning of the septum secundum adjacent to the septum primum. The sinus venosus is incorporated into the superior portion of the developing atria. Ventricular septation is partially accomplished by the upward growth of muscular tissue to form the muscular interventricular septum. Endocardial cushion tissue develops and provides the essential tissue for formation of the atrioventricular valves, the closure of the ostium primum in the atrial septum, and the formation of the membranous interventricular septum.

The primitive bulbus cordis contributes several key components of the forming heart. The proximal third of the
Relationship to Congenital Heart Disease

Knowledge of normal and abnormal embryologic development of the heart and blood vessels provides an essential basis for and understanding of the morphogenesis of congenital heart disease.9,9 The position of the atria is determined by general body habitus. The anatomic right atrium is defined as the atrium receiving the systemic venous drainage. The anatomic left atrium is defined as the atrium receiving the pulmonary venous drainage. In situs solitus, the right atrium is on the right and the left atrium on the left side of the body. In situs inversus, the anatomic right atrium is on the left side and the anatomic left atrium is on the right side of the body. The right and left ventricles are normally connected to the corresponding atria, but the ventricles may be inverted. Inversion of the ventricles is a major feature of a condition known as congenitally corrected transposition of the great vessels. This condition arises when the primitive heart undergoes levo[1]-bulboventricular looping rather than the usual dextro[dl]-bulboventricular looping, hence, the alternative designation of corrected transposition as l-transposition. Other positional anomalies of the heart include dextrocardia and right-sided aortic arch.

Abnormalities in atrial septation give rise to three types of atrial septal defects; in order of frequency they are the ostium secundum defect, ostium primum defect, and sinus venosus defect. Ostium secundum defects are located in the midportion of the interatrial septum and result from inadequate formation of septum secundum and/or septum primum to cover the foramen ovale. Ostium primum defects are located in the inferior portion of the interatrial septum and result from a defect in formation of endocardial cushion tissue. Severe endocardial cushion deficiency can lead to a common atrioventricular canal anomaly with ostium primum atrial septal defect, membranous ventricular septal defect, and abnormal atrioventricular valve. The sinus venosus atrial septal defect is located in the superior portion of the interatrial septum and results from defective incorporation of the primitive sinus venosus into the forming heart; this defect is often associated with partial anomalous pulmonary venous drainage into the right atrium. Abnormalities in ventricular septation give rise to ventricular septal defects, usually in the region of the membranous septum. Formation of the membranous interventricular septum involves contributions from the conal ridges of the bulbus cordis and the endocardial cushions. Deficiencies in contributions from these embryonic structures leads to the formation of membranous ventricular septal defects. Abnormalities in septation of the great vessels give rise to congenital complete transposition of the great vessels, or d-transposition, since the abnormal septation of the great vessels occurs with normal dextro-bulboventricular looping and the atria and ventricles are in normal position. Congenital complete transposition is often accompanied by...
atrial or ventricular septal defects or other congenital lesions. Severe maldevelopment of the heart can give rise to a hypoplastic left heart or hypoplastic right heart and associated valvular atresia. Other anomalies include vascular rings, persistent patent ductus arteriosus, and coarctation of the aorta.

Cardiovascular malformations occur in about 0.8% of live births. However, the incidence of congenital heart disease is significantly higher because cardiac malformations occur 10 times more often in stillborn than in liveborn infants. Children with congenital heart disease are predominantly male. However, specific defects have a definite sex preponderance: females have a higher incidence of patent ductus arteriosus and atrial septal defect; males have a higher incidence of valvular aortic stenosis, congenital aneurysm of the sinus of Valsalva, coarctation of the aorta, tetralogy of Fallot, and congenital complete transposition of the great arteries.
FIGURE 1.3. Frontal plane dissections of developing heart show important relations in establishing aortic and pulmonary outlets. The truncus arteriosus has been drawn with its cut end turned upward, in order to show the absence of truncus ridges in the early stages and their relationships in later stages.

FIGURE 1.4. Schematic drawing to show interrelations of septum primum and septum secundum during the latter part of fetal life. Note especially that the lower part of septum primum is positioned so as to act as a one-way valve at the oval foramen in septum secundum. The split arrow indicates that a considerable part of the blood from the inferior vena cava passes through the foramen ovale to the left atrium while the remainder eddies back into the right atrium to mix with the blood being returned by way of the superior vena cava.
External Anatomy of the Heart and Great Vessels

The anatomy of the heart and great vessels has been well documented in previous publications. The normal location of the heart is in the mediastinum to the left of the midline with the cardiac apex pointed to the left (Fig. 1.5). The heart is rotated and tilted in the chest, and as a result about two thirds of the anterior surface of the heart is composed of the right ventricle, and the left third of the anterior surface is composed of the left ventricle. The right inferior border (obtuse border) of the heart is formed by the right ventricle and the left lateral border is formed by the left ventricle. Located superior to the right and left ventricles are the right and left auricles of the right and left atria, respectively. The anterior superior surface of the heart is constituted by the outflow portion (conus) of the right ventricle and the pulmonary trunk, which extends from right to left as it exits the pericardium. The pulmonary trunk then gives rise to the left and right main pulmonary arteries. The aorta is located posterior to the pulmonary trunk. The aorta takes origin from the left ventricle and is oriented from left to right as it exits the pericardium. The aorta then curves to the left and inferiorly, creating a left-sided aortic arch. The aortic arch gives origin to the right innominate, left common carotid, and left subclavian arteries. The aorta then continues inferiorly as the descending thoracic aorta.

Pericardium

The pericardial cavity is a fluid-filled space that surrounds the heart and proximal great vessels. The pericardial space is enclosed by a thin layer of connective tissue that is lined by a single layer of mesothelial cells. There are two components: the visceral and parietal pericardium. The visceral pericardium covers the epicardium of the heart, and the parietal pericardium forms the outer layer. The superior extent of the pericardial cavity representing the transition of visceral to parietal pericardium occurs approximately 2 to 3 cm superior to the heart at the level of the great vessels, thereby enclosing the proximal aorta and pulmonary trunk in the pericardial cavity (Fig. 1.5). The pericardial cavity normally contains about 20 cc of serous fluid, which serves to lubricate the heart and facilitate cardiac motion.

The pericardium can be affected by a variety of inflammatory and neoplastic conditions. Hemorrhage into the pericardium may occur as a result of cardiac rupture, usually secondary to acute myocardial infarction, or rupture of the proximal aorta following aortic dissection. The severity of cardiac dysfunction secondary to pericardial disease is influenced acutely by the amount and rapidity of fluid accumulation in the pericardial cavity and chronically by the severity of inflammation and fibrosis. Rapid accumulation of 100 to 200 mL of fluid or blood in the pericardial cavity can induce cardiac tamponade, whereas the slow accumulation of several hundred milliliters can be accommodated in the

FIGURE 1.5. Ventral view of heart in situ with the pericardial sac opened.
cavity with stretching of the pericardial lining before impaired cardiac function develops.

Heart

Basic Structure

The anatomy of the heart has been documented in detail. The heart is composed of three layers: the epicardium, the myocardium, and the endocardium. The epicardium consists of fatty connective tissue and is lined by the visceral pericardium. The major coronary arteries and veins traverse the epicardium. The myocardium constitutes most of the mass of the heart and is composed of cardiac myocytes, vessels, and connective tissue. The cardiac myocytes represent approximately 80% of the mass but only 20% of the number of cells in the myocardium. The endocardium is divided into the nonvalvular (visceral) and valvular endocardium. The endocardium consists of thin fibrocellular connective tissue, which is lined by a single layer of endothelial cells.

Cardiac Dimensions

Several sources have provided information regarding dimensions and measurements of the heart. The weight of the heart varies in relationship to body dimensions, including length and weight. Hudson has published a useful guide regarding fresh heart weight in males and females. The adult male heart weight has the following parameters: 0.45% of body weight, average 300 g, range 250 to 350 g. The adult female heart weight has the following parameters: 0.40% of body weight, average 250 g, range 200 to 300 g. Selective measurements of right ventricular and left ventricular weights can be made, and ranges have been established for determination of selective enlargement of right and left ventricles as well as biventricular enlargement. The thickness of the walls of the cardiac chambers is as follows: right and left atria, 0.1 to 0.2 cm; right ventricle, 0.4 to 0.5 cm; left ventricle, 1.2 to 1.5 cm (free wall, excluding papillary muscles and large trabeculae). The average circumferences of the cardiac valves are as follows: aortic, 7.5 cm; pulmonic, 8.5 cm; mitral, 10.0 cm; and tricuspid, 12.0 cm.

Coronary Vasculature

The anatomy and physiology of the coronary circulation have been described in detail. In the normal heart, oxygenated blood is supplied by two coronary arteries that are the first branches of the aorta. The origin of the left and right coronary arteries from the aorta is through their ostia positioned in the left and right aortic sinuses of Valsalva, which are located just distal to the left and right cusps, respectively, of the aortic valve (Figs. 1.5, 1.6, and 1.7). The left main coronary artery is a short vessel with a length of 0.5 to 1.5 cm. The left main coronary artery divides into left anterior descending and left circumflex branches and, occasionally, a left marginal branch. The left anterior descending coronary artery and its left diagonal and septal branches supply the anterior portion of the left ventricles and interventricular septum. The left circumflex coronary artery and its circumflex marginal branches supply the lateral left ventricle. The right coronary artery supplies the right ventricle and, in about 90% of hearts, it extends posteriorly to give rise to the posterior descending coronary artery.

There is considerable variation in the anatomic distribution of the coronary arterial branches. However, in most hearts, branches from both the left circumflex and right coronary arteries contribute to the blood supply of the posterior left ventricle, resulting in a so-called balanced circulation. In about 10% of hearts, the right coronary artery is small, and the left circumflex coronary artery gives origin...
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Cardiac collaterals, and intramural branches, which communicate with the cardiac cavities [arterioluminal vessels]. In the normal adult heart, the collateral vessels are thin walled, small channels, usually less than 50 µm in diameter, and they contribute little to total coronary blood flow.

The myocardial collateral vessels can increase in diameter into the range of 200 to 600 µm or to the posterior descending coronary artery and provides the sole blood supply for the posterior left ventricle, creating a left dominant circulation. Rarely, the converse right dominant circulation exists when the left circumflex is small and the posterior left ventricle is supplied primarily by left ventricular branches of the right coronary artery.

The major blood supply to the sinoatrial node via the sinus node artery is derived from the proximal right coronary artery in about 60% of hearts and from the left circumflex coronary artery in about 40% of hearts. The major atrioventricular nodal artery is derived from the coronary artery that gives rise to the posterior descending branch, which is the right coronary artery in about 90% and the left circumflex coronary artery in about 10% of hearts.

The epicardial coronary arteries deliver oxygenated blood to the intramyocardial arteries, arterioles, and capillaries leading to oxygen and substrate extraction in the myocardium [Fig. 1.8]. A small amount of unoxygenated blood flows directly into the ventricular cavities via the thebesian veins. However, most desaturated blood traverses the myocardial venules and veins into the epicardial veins, which drain into the coronary sinus located in the inferoposterior region of the right atrium.

Collateral blood vessels form during embryologic development of the heart, and they connect different components of the coronary arterial circulation. The coronary collateral system is composed of four types of vessels: intramural branches of the same coronary artery [homocoronary collaterals], intramural branches of two or more coronary arteries [intercoronary collaterals], atrial branches, which connect with the vasa vasorum of the aorta and other vessels [extra-

**FIGURE 1.7.** Dorsocaudal view of the heart with the epicardium removed to expose the injected coronary vessels.

**FIGURE 1.8.** Diagram of the ventricular wall, showing the relationship between the various intramural vascular channels.
greater, develop muscular media, and transport significant amounts of blood flow [Fig. 1.9]. In addition to the types of collateral vessels described above, collateral channels also can develop proximal and distal to a stenosis in a given coronary artery.

Right and Left Atria and Ventricles

The right and left atria and right and left ventricles have distinctive anatomic features (Figs. 1.10 to 1.17). The anatomic right ventricle is characterized as follows: distinct muscular infundibulum [conus] arteriosus, which separates the right semilunar [pulmonary] valve and the right atrioventricular [tricuspid valve]; single large anterior papillary muscle; and coarse trabecular muscles [trabeculae carneae cordis] at the apical and inflow portion of the chamber. Key landmarks of the right ventricular infundibular [conus] region from inferior to superior are the membranous interventricular septum; the crista supraventricularis, an inverted V-shaped structure with parietal and septal limbs; and the pulmonic valve. The anatomic left ventricle has the following features: fibrous continuity of annulus of left semilunar [aortic] valve and anterior leaflet of left atrioventricular [mitral] valve, two well-developed papillary muscles [anterolateral and posteromedial], and fine trabecular muscles at the apical and inflow portion of the chamber. These features allow determination of the anatomic right ventricle and anatomic left ventricle in complex congenital anomalies involving displacement of the various components of the heart.

Cardiac Valves

The four-chambered heart has four valves: the right semilunar or pulmonic valve; the right atrioventricular or tricuspid valve; the left semilunar or aortic valve; and the left
FIGURE 1.11. Right side of the heart opened in a plane approximately parallel to the septa, to show the interior of the right atrium and the right ventricle. A segment of the septal leaflet of the tricuspid valve has been removed to expose more fully the region of the membranous portion of the interventricular septum.

FIGURE 1.12. Ventral view of the heart with the walls of the right atrium and ventricle opened to show their internal configuration. This heart has an unusually well-developed moderator band, and the position of the foramen ovale is somewhat more cephalic than usual.
FIGURE 1.13. Photograph of opened right heart demonstrating actual structures depicted in the illustrations. Note the muscular infundibulum separating the tricuspid and pulmonic valves. CA, conus arteriosus, i.e., infundibulum; CS, coronary sinus; PV, pulmonic valve; RA, right atrium; RV, right ventricle.

FIGURE 1.14. Left side of the heart opened in a plane approximately parallel to the septa, to show the interior of the left atrium and left ventricle. A portion of the anterior leaflet of the mitral valve has been removed to expose more fully the region of the membranous portion of the interventricular septum and the aortic orifice.
FIGURE 1.15. Frontal section through a heart fixed in diastole, showing a ventral view of the dorsal portion. The plane of section passes through the septum membranaceum and both atrioventricular ostia.

FIGURE 1.16. Photograph showing the relationship of the membranous interventricular septum (lighted) to the tricuspid annulus and septal leaflet of the tricuspid valve. While the membranous interventricular septum is entirely contained in the left ventricle, the superior portion of the interventricular septum extends superior to the tricuspid annulus. This anatomic relationship allows for left ventricular to right atrial shunts in certain pathologic states, such as infective endocarditis of the aortic valve. MIS, membranous interventricular septum; SLTV, septal leaflet of the tricuspid valve.

FIGURE 1.17. Photograph of opened left ventricle showing continuity of the anterior mitral leaflet and the annulus of the aortic valve. AML, anterior mitral leaflet; LC, left cusp of aortic valve; NCC, noncoronary cusp of aortic valve; PPM, posterior papillary muscle; RC, right coronary cusp of aortic valve.
atrioventricular or mitral valve (Figs. 1.18 to 1.21). The pulmonic and aortic semilunar valves each have three cusps separated by three commissures, and the cusps insert into a fibrous connective tissue annulus. The aortic valve cusps are designated as the left and right cusps in relationship to the coronary ostia and a third, noncoronary cusp. With valve closure during diastole, the cusps of the aortic and pulmonic valves make contact along a line about 1 to 2 mm below the free margin. The central points of maximal contact are especially prominent and are known as the noduli of Arantius. The most common congenital valve lesion is a malformed aortic valve, either a unicuspid or bicuspid valve.

The tricuspid or right atrioventricular valve has three leaflets (cusps) that are separated by three commissures and insert into a fibrous annulus. The leaflets are designated as the lateral, medial, and anterior leaflets. The leaflets are attached to the ventricular muscle by multiple chordae tendineae, which extend from the ventricular surfaces of the interventricular septum membranaceum, and the mitral, the tricuspid and the aortic valves.
leaflets to the mural ventricular wall as well as a well-developed anterior papillary muscle and a diminutive structure, the papillary muscle of the conus.

The mitral or left atrioventricular valve has a highly developed and coordinated group of structures that are referred to as the mitral valve apparatus (Figs. 1.18 to 1.21). The mitral valve apparatus consists of fibrous annulus, anterior and posterior leaflets, anterior and posterior commissures, multiple chordae tendineae, and anterolateral and posteromedial papillary muscles. Chordae tendineae extend from the ventricular surfaces of both leaflets and attach to both papillary muscles. Chordae tendineae exhibit a branching pattern with primary chordae arising from the papillary muscles and branching into secondary and then tertiary chordae, which insert into the valvular leaflets. Mitral valve dysfunction can result from a wide variety of pathologic processes affecting any component of the mitral apparatus, including the myocardium.

The anterior mitral leaflet is usually a single structure, whereas the posterior leaflet may be divided into two or three scallops (Fig. 1.21). The anterior mitral leaflet normally has a significantly larger area and longer length from annulus to free margin that the posterior mitral leaflet. As a result, the anterior leaflet contributes about two thirds of the area of leaflet tissue involved in closure of the mitral orifice during systole. Expansion of the posterior leaflet to equal the length of the anterior leaflet from annulus to free margin is an important anatomic feature of myxomatous degeneration of the mitral valve. Other features include general redundancy, thickening, and a glistening white appearance of leaflet tissue and chordae tendineae.

Cardiac Conduction System and Cardiac Innervation

The anatomy of the cardiac conduction system has been defined by meticulous study (Fig. 1.22).26,27 The sinoatrial node is located in the superficial subepicardium of the superior right atrium at the junction of the superior vena cava and the right auricular appendage. The sinoatrial node contains myocytes that are specialized for the generation of electrical impulses and constitute the cardiac pacemaker. The electrical impulse propagates selectively along certain paths in the atria; however, the existence of anatomically distinct conduction pathways in the atria is difficult to demonstrate. Atrioventricular conduction is accomplished by specialized structures, the more proximal atrioventricular node (node of Tawara) and the more distal atrioventricular bundle [bundle of His]. The atrioventricular node is positioned in the inferior interatrial septum just anterior and medial to the ostium of the coronary sinus. The specialized conduction tissue then extends through the fibrous skeleton separating the atria and ventricles to connect to the bundle of His, which is located at the apex of the muscular interventricular septum. The His bundle gives origin to the right and left bundle branches, which extend through the subendocardium of the interventricular septum into the right and left ventricular free walls.

The heart has a dual innervation from the sympathetic [thoracolumbar] and parasympathetic [craniosacral] divisions of the autonomic nervous system (Fig. 1.23). These nerves interact with the conduction tissue to provide neural modulation of cardiac function.
FIGURE 1.22. Schematic diagram of heart opened frontally, demonstrating the location and relations of the several parts of the sino-atrial and atrioventricular conduction system.

FIGURE 1.23. Ventral view of thorax to show the nerve supply to the heart.
Summary

This chapter presented a description and illustration of the embryologic and anatomic features of the heart and great vessels. The embryologic development of the cardiac structures was presented, and the relationship of cardiac development to congenital heart disease was discussed. Key components of the mature heart were described and illustrated, including the pericardium, the coronary arteries, the four cardiac valves, the right and left atria and ventricles, and the cardiac conduction system and cardiac innervation. The relationship of structure to function was considered. Understanding cardiovascular anatomy is important for the diagnosis and treatment of cardiovascular diseases.

References
