3

Cardiac anatomy and examination of specimens

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Cardiac anatomy

Owing to the quality of prenatal ultrasound and the expanded experience of prenatal diagnosticians, it is possible to observe congenital heart malformations in increasingly greater detail and at an ever earlier stage of gestation.[1] Since it is on the basis of ultrasound findings that decisions to terminate pregnancies are made, there is a critical need for monitoring and confirmation of the prenatal diagnosis. This need can only be adequately met only by autopsy.

The diagnosis of congenital heart malformations is considered to be difficult. In fact, with the proper systematic approach, most cases of congenital heart malformation are relatively easy to diagnose, although there are cases that are extremely difficult.[2,3] For this reason, it is necessary that the system for describing and diagnosing such conditions be both simple and comprehensive, on the one hand, and applicable to all cases, on the other. The goal of a descriptive system of the heart is to allow the heart atria and ventricles, the aorta and the pulmonary trunk to be identified and distinguished from each other.

In this chapter, the characteristic features of the various components of the normal fetal heart are described.

The morphologically right atrium

In the normal heart, the morphologically right atrium forms the right, front part of the cardiac mass (Figure 3.1). The morphologically right atrium consists of the following components: the venous component, the vestibule, the septum and the appendage.

The external wall of the atrium consists of two parts: the venous component (sinus venarum cavarum, or posterior part), into which the superior and inferior caval veins, as well as the coronary sinus, open; and an anterior part, the appendage, which extends forward and surrounds the right wall of the aorta.

The morphologically right appendage has a broad, triangular shape. Internally, it is connected with the smooth-walled venous component of the atrium. This

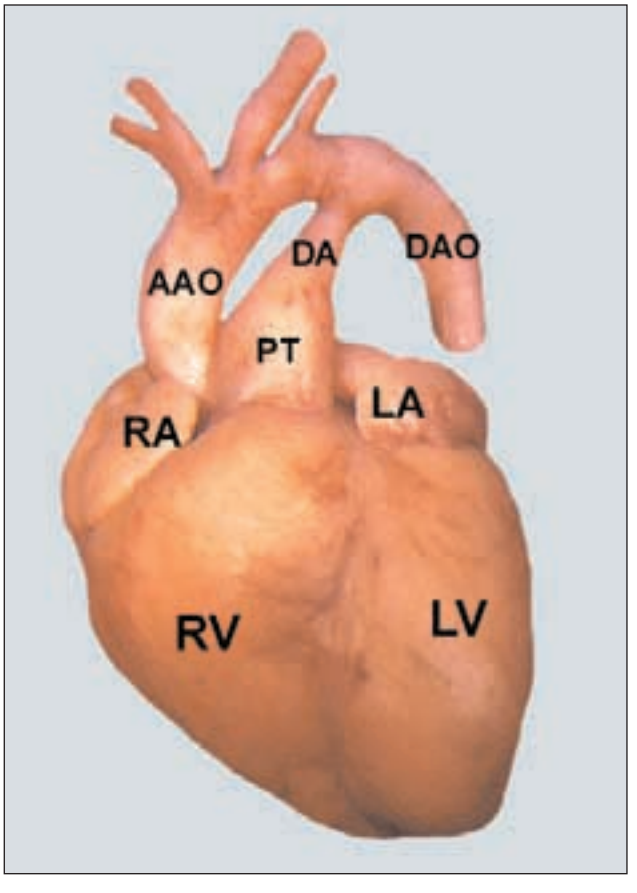

Figure 3.1
Frontal view of the heart, showing the various components of the normal fetal heart. AAO, ascending aorta; DA, ductus arteriosus; DAO, descending aorta; PT, pulmonary trunk; RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle.

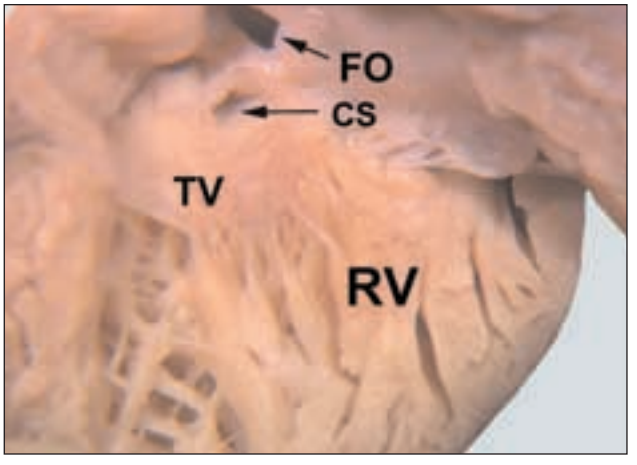

Figure 3.2
Dissection showing the typical atrioventricular junction of the right heart. FO, fossa ovalis; CS, coronary sinus; TV, tricuspid valve; RV, right ventricle.

connection is marked in the morphologically right appendage by a prominent crest with pectinate muscles.

The ostium of the tricuspid valve points diagonally to the right and can be considered as the base of the vestibule. The ostium is smooth-walled and the leaflets of the tricuspid valve are attached to its edge (Figure 3.2).

The terminal crest runs laterally, forming the connection between the venous component and the right appendage. It emerges from the anterior part of the septal surface and curves in front of the orifice of the superior caval vein. The superior caval vein is located in the roof of the atrium and enters the right atrium between the terminal crest and the superior rim of the oval fossa. The border of the crest is reinforced by fibrous structures. These structures separate the orifices of the inferior caval vein and the coronary sinus from the atrial appendage, becoming the venous valves of the inferior caval vein (Eustachian valve) and the coronary sinus (Thebesian valve). The Eustachian valve can be prominent, but it can also be completely lacking; in individual cases it can form a network with pronounced fenestration, which is called the Chiari network. The Eustachian valve is usually small; it protects the mouth of the inferior caval vein and may extend to the inlet of the superior caval vein. The tendon of Todaro is the continuation of the venous valves. The triangle between the tendon and the tricuspid valve marks the location of the atrioventricular conduction tissue.[4] The septal surface consists of the floor of the oval fossa and the atrioventricular septum.

The coronary sinus opens above the posterior interventricular groove into the right atrium. The so-called "septum secundum" does not belong to the septum proper, but simply forms the surrounding wall of the atrial chambers.

The morphologically left atrium

The morphologically left atrium is the most posterior chamber of the heart. It consists of the following components: the venous component, a septal surface, the vestibule and an appendage.

At the top, the venous component admits the four pulmonary veins, two on each side. It is considerably larger than the appendage.

The septal surface of the left atrium is at an angle; it is in part uneven and consists of the left atrial surface of the oval fossa. The valve overlaps the surrounding atrial wall (the septum secundum) from above.

The surface of the vestibule of the left atrium is smooth; the leaflets of the mitral valve are attached to it. The pectinate muscles are less pronounced here than in the right atrium, and do not project into the body of the atrium. In contrast with the right atrium, the pectinate muscles do not extend into the area of the atrioventricular junction.

The morphologically left appendage has a tubular, hooked shape. It has characteristic grooves at the edge and partly surrounds the pulmonary artery. The connection between the smooth-walled venous component and the left appendage is narrower than in the right atrium and it is not limited by a crest. As with the right appendage, internally the left appendage is trabeculated, but the trabeculations are finer.

The morphologically right ventricle

The morphologically right ventricle occupies the greatest part of the ventral mass of the heart. It consists of the following: the inlet, the apical trabecular and the outlet components, which go from the lower right to the upper left in the ventricular mass. The inlet component surrounds and provides support for the leaflets and the tension apparatus of the tricuspid valve, which extends dorsally to the crux cordis. The leaflets of the tricuspid valve are in the septal, anterior-superior and inferior (or mural) position. All the leaflets arise at the atrioventricular junction. The most characteristic feature of the tricuspid valve is the presence of tendinous cords which fix the septal leaflet to the ventricular septum.

The border between the inlet and the trabecular component is formed by the attachment of the papillary muscles. The trabecular zone reaches to the apex of the heart. Characteristic of the apical trabecular component of the right ventricle are coarse trabeculations (Figure

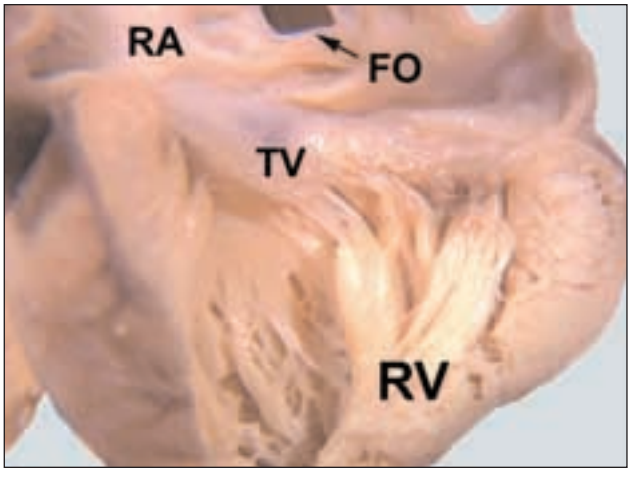

Figure 3.3
Section showing the typical coarse trabeculations of the right ventricle (RV). RA, right atrium; FO, fossa ovalis; TV, tricuspid valve.

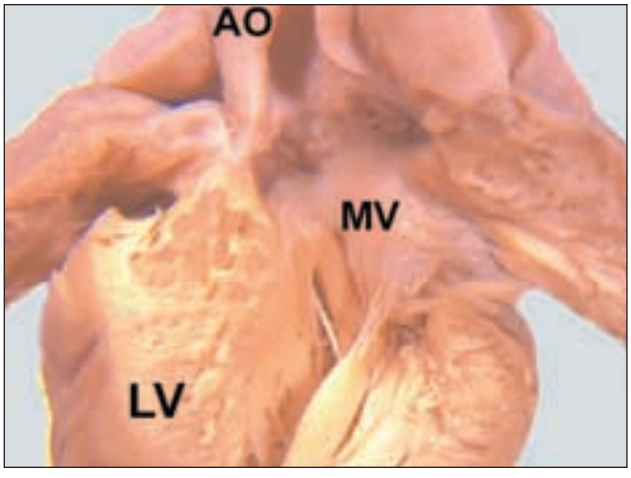

Figure 3.4
Specimen in which the left ventricle (LV) is displayed to show the typical fine criss-crossed trabeculations. AO, aorta; MV, mitral valve.

3.3), which provide the best criterion for identification of the morphologically right ventricle in cases in which there is no inlet component. The inlet and outlet components are separated from each other in the roof of the ventricle by a prominent muscular crest, the crista supraventricularis. The outlet component (infundibulum) of the right ventricle is a muscular tube, which generally has smoother walls than the trabecular component. The three leaflets of the pulmonary valve are attached to a completely muscular infundibulum. The posterior wall of the infundibulum is separated from the aorta by an extracardiac space.

The septum of the normal heart has a muscular and a membranous component, with the membranous component being very narrow. The trabecula septomarginalis is a powerful ridge that abuts the right-ventricular surface of the septum.

The morphologically left ventricle

As with the right ventricle, the left ventricle consists of an inlet, an apical trabecular component and an outlet component.

The inlet component contains and surrounds the mitral valve, which has an aortic (anterior or septal) leaflet and a posterior (or mural) leaflet. The leaflets are separated from each other by the anterolateral and posteromedial commissures. The two mitral valve leaflets are connected to two groups of papillary muscles, which occupy a posteromedial and anterolateral position below the commissural areas. The most characteristic feature of the mitral valve is that it has no cordal attachments to the ventricular septum.

The trabecular component of the left ventricle extends from the origin of the papillary muscles to the apex of the heart. The apical trabecular part of the left ventricle has fine criss-crossed trabeculations (Figure 3.4).

The inlet and outlet components of the left ventricle form an acute angle, with the inlet and outlet components being separated by the anterior of the mitral valve.

The outlet component of the left ventricle contains the aortic valve. The component consists partially of muscular, partially of fibrous tissue. Dorsally it is incomplete, so that the mitral and aortic valves are connected to each other by fibrous tissue. The aortic valve has three semilunar leaflets called the right coronary, left coronary and noncoronary leaflets, which are attached to the root of the aorta.

The interventricular septum

The interventricular septum consists mainly of muscular tissue, called the muscular septum, and only to a small extent of fibrous tissue, the membranous septum (Figure 3.5).

The muscular septum is divided into the inlet, apical trabecular component and outlet component. On the right-ventricular side, the inlet area septum is bordered by the cordal attachments of the septal tricuspid leaflet. On the left-ventricular side, this line is connected with the dorsal end of the smooth-walled septum. On the right-

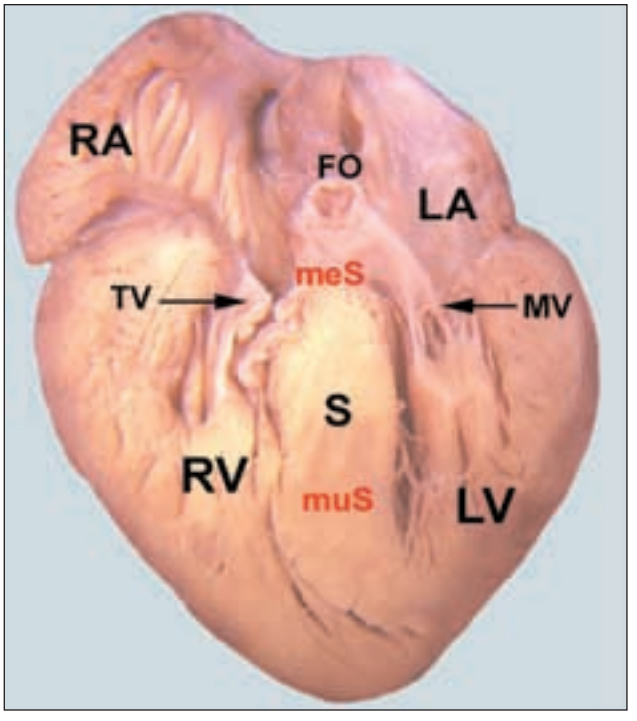

Figure 3.5
Specimen sectioned along the cardiac long axis, showing the four cardiac chambers, the two atrioventricular valves and the interventricular septum. RA, right atrium; FO, fossa ovalis; LA, left atrium; TV, tricuspid valve; meS, membranous septum; MV, mitral valve; S, septum; RV, right ventricle; muS, muscular septum; LV, left ventricle.

ventricular side, the trabeculations of the apical trabecular component of the septum are rough, whereas they are fine on the left-ventricular side. The outlet component of the septum lies below the distal part of the infundibulum, separating the outlet paths of the right and left ventricles.

The membranous septum lies at the point where the inlet, apical trabecular component and outlet component of the muscular septum come together.

The aorta

The aorta leaves the heart in the cranial direction (this section is called the ascending aorta), curves dorsally to the left (this section is called the aortic arch) and then runs caudally from in front of the thoracic spine (this section is called the descending aorta).

The ascending aorta emerges from behind the pulmonary trunk out of the conus arteriosus of the left ventricle and then runs intrapericardially somewhat to the right. The ascending aorta has only two branches: the

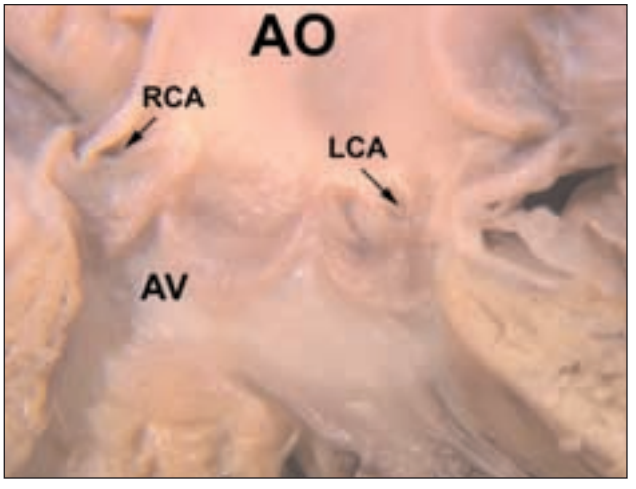

Figure 3.6
Longitudinal section through the aortic valve (AV) showing the ostium of the left coronary artery (LCA) below the left coronary leaflet and the ostium of the right coronary artery (RCA) below the right coronary leaflet. AO, aorta.

right and left coronary arteries. These emerge immediately behind the aortic valve in the area of the bulbus aortae, the left coronary artery in the sinus aortae below the left coronary leaflet and the right coronary artery in the sinus aortae below the right coronary leaflet (Figure 3.6).

The ascending aorta passes over into the aortic arch. The great branches of the aorta emerge from the convex side of the aortic arch: the brachiocephalic artery (which divides into the right subclavian artery and the right common carotid artery), the left common carotid artery and the left subclavian artery.

The descending aorta designates that part of the vessel which extends from the ostium of the arterial duct to the aortic bifurcation.

The pulmonary trunk

The pulmonary trunk emerges from the conus arteriosus of the right ventricle, terminating there at the pulmonary valve. Below the aortic arch it divides into right and left pulmonary arteries. In the fetal circulation the arterial duct emerges from the bifurcation of the pulmonary trunk, which connects the pulmonary trunk with the descending aorta. The arterial duct demarcates the isthmus of the aorta between the site of emergence of the left subclavian artery and the aortic insertion of the duct. The arterial duct is structurally different from the aorta and the pulmonary artery. The wall of the arterial duct is thicker than that of both the aorta and the pulmonary

trunk. The luminal surface of the duct in the newborn is less smooth than that of the great arteries and has irregular ridges running lengthwise along it. After birth, the duct closes and becomes the arterial ligament.

The longer right pulmonary artery passes under the aortic arch and behind the inferior vena cava to the right bronchus. The shorter left pulmonary artery reaches the left bronchus over a direct path in front of the ascending aorta.

Examination of specimens

By applying the method of sequential segmental analysis of the heart a relatively simple and reliable diagnosis of complex congenital heart malformations can be achieved.[5,6] This requires a description of the atrial arrangement, the atrioventricular junction and the ventriculoarterial junction, in this order.

The normal heart consists of three segments: the atria, the ventricular mass and the great arteries. Each of these three sections of the normal heart has a right and a left side. Congenital heart anomalies can affect one or more of these segments or the great veins.

To determine which of the cardiac chambers are normal, the structure of their constant components has to be evaluated morphologically.[7] This is followed by a description of the position of the heart in the thorax, the orientation of the apex of the heart and any associated malformations.

Atrial arrangement

The most constant component of the atrium is the atrial appendage. The right and left atrial appendages are different.[8] The shape of the morphologically right appendage is that of a broad triangle, whereas the left appendage is tubular and hooked. The most relaible feature differentiating between the right and left appendages, however, is the nature of the junction between the appendage and the smooth-walled venous component of the atrium. In the morphologically right appendage this junction is wide, and marked by an extensive crest with pectinate muscles extending all round the atrioventricular junction; in the morphologically left appendage this junction is narrow, and has no crest, and no pectinate muscles.

The first step in the sequential analysis is the determination of the arrangement of the atrial chambers. There are four different possibilities here (Figure 3.7). In the usual arrangement, the so-called situs solitus, the morphologically right atrium is located to the right of the morphologically left atrium. The mirrored position, the situs inversus, is rare. In some cases the lateralization of the atria is lacking, so that there are two morphologically right appendages (right-isomerism) or two morphologically left appendages (left-isomerism). Generally, left- or right-atrial isomerism occurs in cases of visceral heterotaxy. Since the judgement as to whether isomerism exists

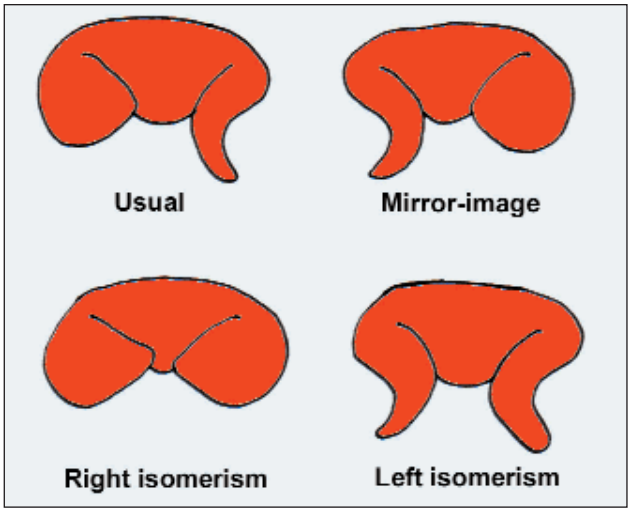

Figure 3.7
Diagram showing the four possible arrangements of the atria. Reproduced from *The Heart*. Gower Medical: London, 1992, with permission.

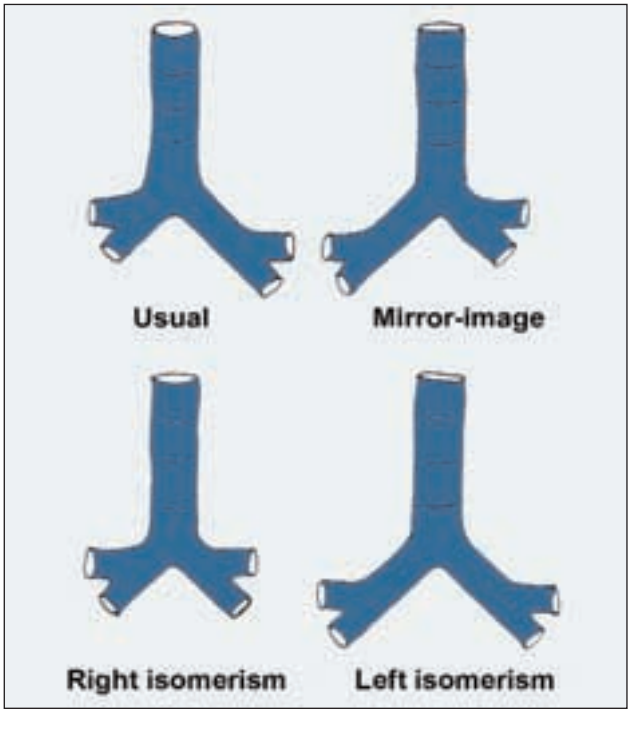

Figure 3.8
Diagram showing the four variants of bronchial morphology usually correlating with atrial arrangement. Reproduced from *The Heart*. Gower Medical: London, 1992, with permission.

is normally made on the basis of the atrial appendages, the expressions "isomerism of the right atrial appendage" and "isomerism of the left atrial appendage" are also used.

The atrial arrangement almost always corresponds to the bronchial morphology. There are four different possibilities here (Figure 3.8). This is especially important in cases in which the arrangement of the atrial chambers is not clear. The identification of the bronchi is based on the fact that the morphologically left bronchus is almost twice as long as the morphologically right bronchus. In addition, the morphologically left bronchus is crossed by the left pulmonary artery before the bronchus divides, which is not the case for the morphologically right bronchus. Therefore, a long hyparterial bronchus on the left means situs solitus, whereas a long hyparterial bronchus on the right is a sign of situs inversus. Bronchi of equal length are an indication of atrial isomerism. The relationship of the bronchi to the pulmonary arteries is the basis for distinguishing between right and left isomerism.

Variation of the atrioventricular junction

This section describes the junction (also called "connection") of the atria with the ventricles, the morphology of the atrioventricular valves and of the ventricles, as well as the interrelationship of the ventricles. There are two groups of atrioventricular junction: biventricular and univentricular.

In biventricular connections each atrium is connected to one ventricle. If the morphologically right atrium is connected to the morphologically right ventricle and the morphologically left atrium is connected to the morphologically left ventricle, both atrioventricular connections are called concordant. An atrioventricular connection is called discordant when the morphologically right atrium is connected to the morphologically left ventricle or the morphologically left atrium is connected to the morphologically right ventricle (Figure 3.9). Atrioventricular concordance and discordance can each occur together with either situs solitus or situs inversus of the heart. If the atrial appendages are isomeric, the biventricular connections cannot be classified in this way and are called ambiguous. In cases of an ambiguous atrioventricular connection the ventricular topology should be described. Here one has to distinguish between a right-hand and a left-hand topology (Figure 3.10).

In describing the spatial relationship of the ventricles to each other the following terms are used for the position of the right ventricle with regard to the left ventricle: anterior, posterior, superior, inferior, right and left.

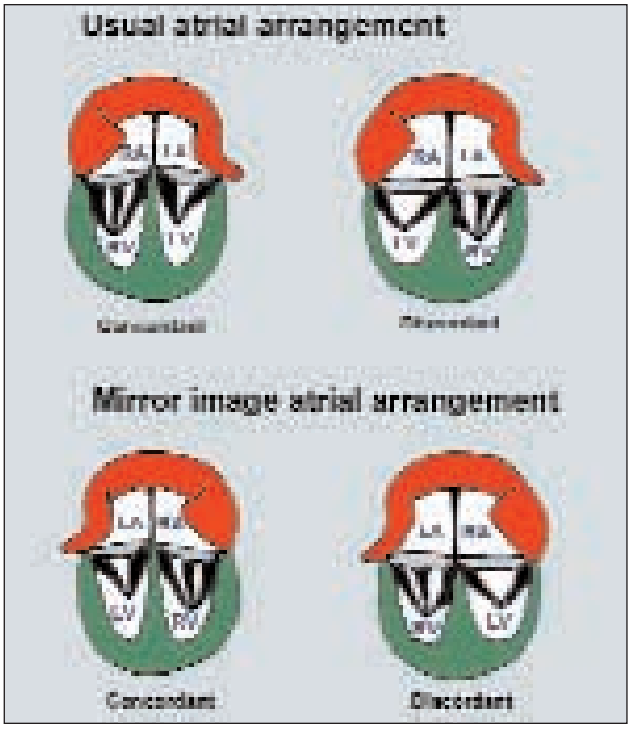

Figure 3.9
Diagram showing concordant and discordant atrioventricular connections in the usual and mirror image arrangements. Reproduced from *The Heart*. Gower Medical: London, 1992, with permission.

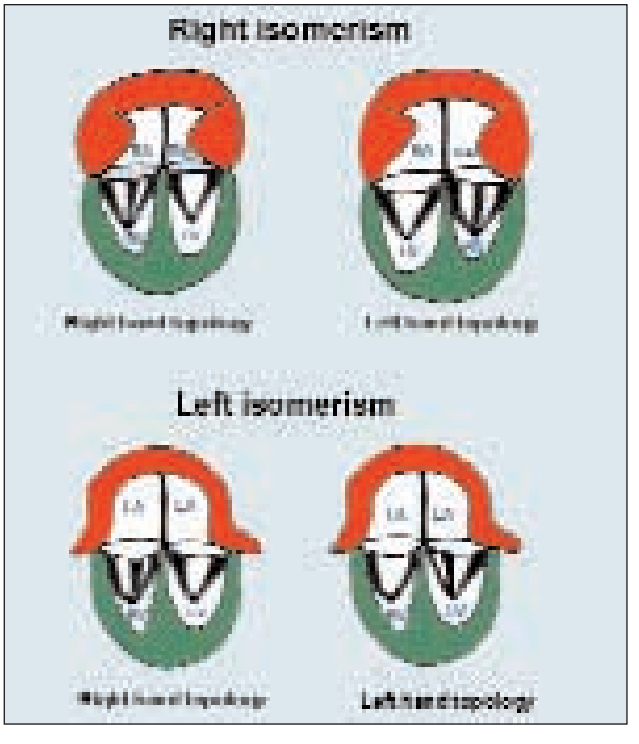

Figure 3.10
Diagram showing the isomeric arrangement of the atrial appendages. The atrioventricular junctions are concordant, but they are ambiguous. Reproduced from *The Heart*. Gower Medical: London, 1992, with permission.

The second group of atrioventricular junctions is called "univentricular." This connection is characterized by a double ventricle inlet (i.e. both atria open into a single ventricle) or the absence of an atrioventricular junction on the right or the left side. The atria can be connected to a dominant left ventricle (the right ventricle is rudimentary or incomplete); to a dominant right ventricle (the left ventricle is rudimentary or incomplete); or to a solitary ventricle of indeterminate morphology. Rudimentary right ventricles are normally located anterosuperior; rudimentary left ventricles are normally in the posteroinferior position within the ventricular mass. In such cases there are either two atrioventricular valves or a single common valve. If there are two atrioventricular valves, one of them can be stenotic, regurgitant, imperforate or straddling, or sit astride the septum. If there is one atrioventricular valve sitting astride the septum, the heart is intermediate between having a completely biventricular and a completely univentricular connection. There are no defined intermediate categories.

If less than 50% of one valve overrides the septum the connection is described as biventricular. The connection is called univentricular only when more than 50% of both valves open into one ventricle.[9] Frequently the valves can be stenotic, incompetent or imperforate. If one atrioventricular connection is missing, there is only a solitary valve that can be incompetent, straddling or overriding. The relationship between the ventricles does not depend on the atrioventricular connection.

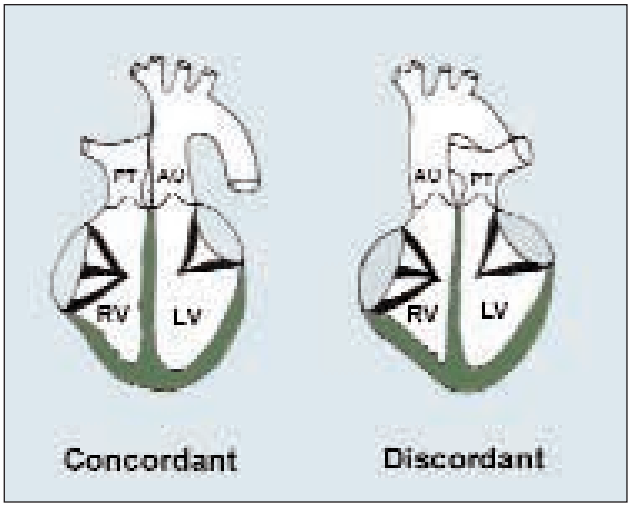

Figure 3.11
Diagram showing concordant and discordant ventriculoarterial junctions. When the aorta and pulmonary trunk arise from the appropriate ventricles, these are concordant. Origins from inappropriate ventricles produce discordant junctions. Reproduced from *The Heart*. Gower Medical: London, 1992, with permission.

Variation of the ventriculoarterial junction

The connection of the arterial trunks to the ventricular mass is here described. The possible relationships of the arterial valves to one another and the relationships of the arterial trunks, as well as the morphology of the infundibular structures, is described.

The arterial trunks are the third segment of the heart. They are identified by their branching pattern. In normal hearts the aorta gives rise to the systemic and the coronary branches and the pulmonary trunk divides into the right and left pulmonary arteries.

Terms used for describing the ventriculoarterial junctions of the heart are: concordant, discordant, double outlet and single outlet (Figure 3.11). In a concordant or discordant ventriculoarterial connection, each great arterial trunk originates in a separate ventricle. If one of the great arterial valves overrides the ventricular septum, it is assigned to the ventricle associated with its greater part.

The ventriculoarterial connection is concordant when the morphologically right ventricle is connected to the pulmonary trunk and the morphologically left ventricle is connected to the aorta. The term "ventriculoarterial discordance" is used to describe the reverse connection. When more than half of both arterial valves are connected to the same ventricle it is called a double outlet ventriculoarterial connection (or, simply, "double outlet").[9] The ventricle may be of right, left, or indeterminate morphology. If the ventricular mass is connected to only one arterial trunk, the arterioventricular connection is called a single outlet ventriculoarterial connection (or, simply, "single outlet"), i.e. it is connected to a single common arterial trunk or to an aorta with an atretic pulmonary trunk lacking a connection to the ventricle, or to a pulmonary trunk with an atretic aorta lacking a connection to the ventricle (Figure 3.12).

With regard to the arrangement of the ventriculoarterial junction, the arterial valves can be perforate, or one of the arterial valves can be imperforate. One or both of the arterial valves may override the ventricular septum. If there is a common arterial valve, either it overrides the ventricular septum or it is committed to one ventricle.

The structure separating the two arterial valves and the two ventricle outlet paths is the infundibular septum. The most frequent arrangement with regard to the infundibular morphology is that the right ventricle has a complete, muscular infundibulum which is lower than its arterial valve. A fibrous continuity exists between the arterial valve issuing from the left ventricle and the left atrioventricular valve. Another possibility is that on both sides a complete,

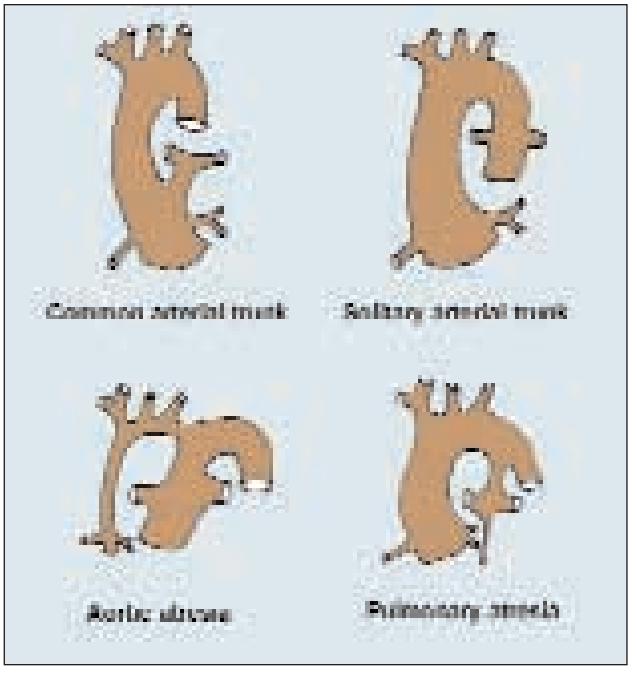

Figure 3.12
Diagram showing the four variants of a single arterial trunk from the heart. Reproduced from *The Heart.* Gower Medical: London, 1992, with permission.

muscular infundibulum lies below the arterial valves or there is a fibrous continuity on both sides between the arterial valves and the atrioventricular valve.

Position of the heart

On describing the position of the heart in the chest it is necessary to give the basic position of the heart and the orientation of the cardiac apex. The heart can lie primarily within the left chest (levocardia), primarily within the right chest (dextrocardia), or in the midline. The cardiac apex can point to the left, to the right or to the middle. The orientation of the cardiac apex is independent of the position of the heart.

An unusual cardiac position does not represent an abnormality of the cardiac morphology. A morphologically normal heart can be abnormally located. The basic position of the heart should be described separately from the cardiac morphology.

In very rare cases the heart can have an extrathoracic position (ectopia cordis).

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