Chapter 1

The Anatomy of the Normal Heart

1.1 Introduction

Self-evidently, before tackling an abnormal heart, knowledge of normal cardiac anatomy and histology is useful. This chapter examines the structure of the normal heart at the gross, microscopic and ultrastructural levels. Chapter 2 gives details of dissection of the heart and includes a detailed description of sequential segmental analysis. Chapter 3 describes the formation of the normal heart.

A Brief Note on Terminology

As is now common practice in the United Kingdom, I have anglicised many anatomical terms. Thus, for example, I have used the terms "superior caval vein" rather than "superior vena cava" and "arterial duct" rather than "ductus arteriosus". I have, however, baulked at the use of such neologisms as "atriums" and "septums", preferring the original, shorter and infinitely more elegant Latin plurals "atria" and "septa". I fully accept that there is, thus, inconsistency, but it is, at least, consistent inconsistency. The point of language is to communicate information and I do not believe that the terms I have employed in any way impair that communication.

1.2 Anatomy

1.2.1 Situation

The heart sits in the mediastinum, more on the left side than the right. Inferiorly it rests on the central part of the diaphragm and superiorly the aortic arch rises almost to the neck (Figures 1.1 and 1.2). Posteriorly, there is the descending thoracic aorta, oesophagus and vertebral column, and anteriorly, the thymus and sternum. The lungs lie on either side, and their anterior extensions interpose between the heart and the anterior chest wall.

1.2.2 Pericardium

The heart is anchored by its attached structures (caval and pulmonary veins and the aorta and pulmonary trunk) and is surrounded by a dense fibrous covering – the pericardium. The pericardium encloses the heart and great vessels but is reflected off the anchoring structures (Figure 1.3). Externally, the parietal layer of pericardium is adherent to the diaphragm, sternum and costal cartilages. The bilateral symmetry of the pulmonary veins combined with asymmetrical arrangement of the caval veins (lying on the right side, and not the left) means that there is a blind invagination of pericardium behind the left atrium between the right pulmonary veins and the inferior caval vein – the oblique sinus of the pericardium (Figure 1.4). A transverse sinus runs from side to side posterior to the aorta and pulmonary trunk and anterior to the bodies of both atria (Figure 1.5).

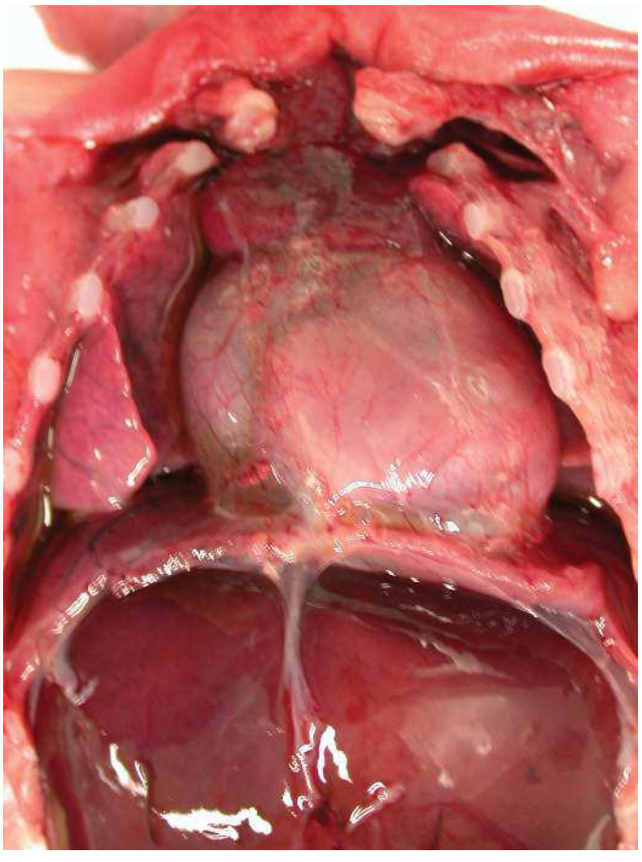

Figure 1.1 Normal heart. A post-mortem in a neonate. The chest has been opened and the sternum removed by cutting through the costal cartilages. The heart is enclosed in the pericardium. Above is the thymus, below the diaphragm separates it from the liver. The pleural cavities are open, and the lungs are visible.

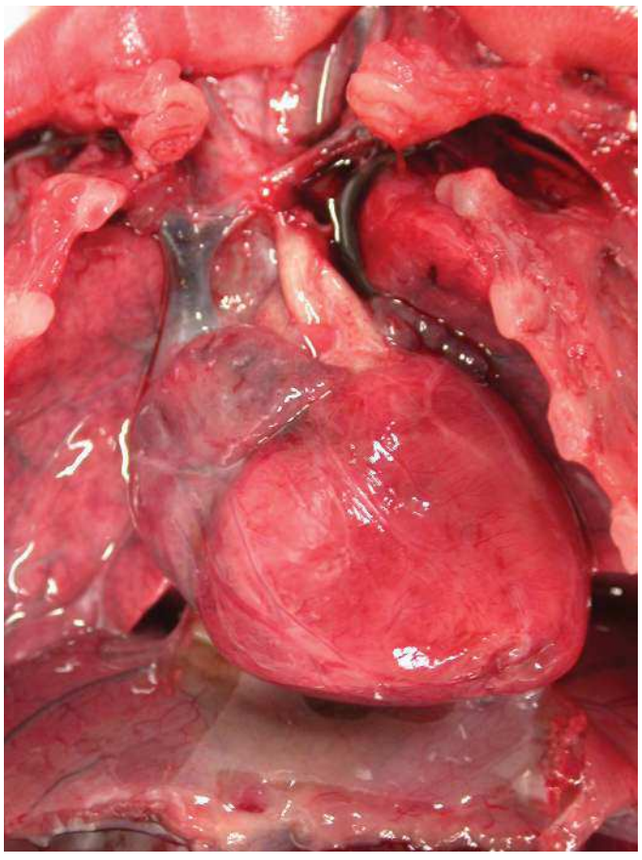

Figure 1.2 Normal heart after removal of the thymus and pericardium. The greater part of the ventricular mass visible in this view is the right ventricle. The right atrial appendage lies above, and above this again the superior caval vein. The pulmonary artery arises from the right ventricle, and the aorta is just visible behind it. The left atrial appendage is just visible to the left of the arterial pedicle.

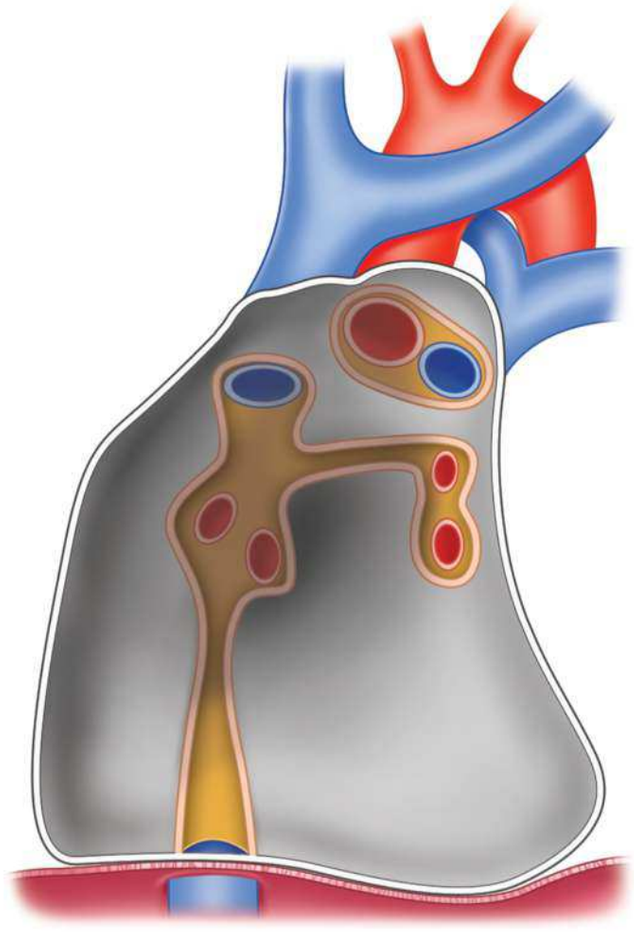

Figure 1.3 Diagram of attachments of pericardium. The sites of attachment are coloured red. The pericardial cavity encloses the most proximal parts of the superior and inferior caval veins and the pulmonary veins, and also the most proximal parts of the aorta and pulmonary trunk and proximal part of the arterial duct.

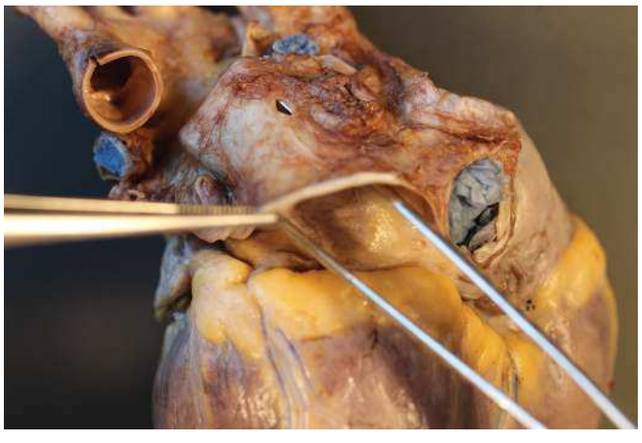

Figure 1.4 Oblique sinus of the pericardium. The heart viewed from behind. One pair of forceps grabs the cut edge of the parietal pericardium. Another pair is inserted into the oblique sinus. The right margin is formed by the pericardial attachment between the inferior caval vein and right pulmonary veins, and the superior blind end is closed by the attachment of pericardium between the upper pulmonary veins.

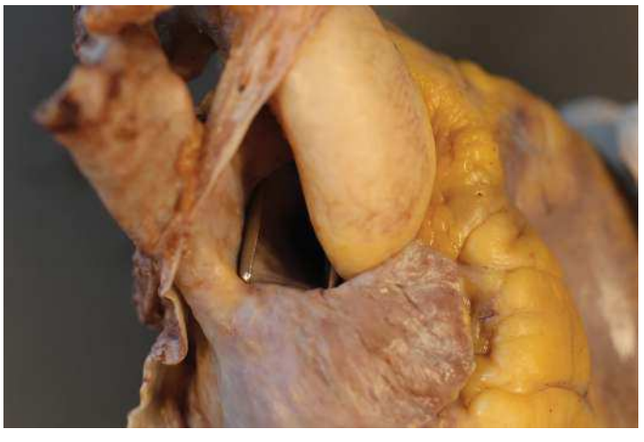

Figure 1.5 Transverse sinus of pericardium. The heart is viewed from the right side. A pair of forceps has been inserted from the left side between the arterial pedicle and the atria. The tip can be seen emerging on the right side between the junction of the superior caval vein and right atrium posteriorly and the aorta anteriorly. The cut edge of the pericardial reflection is seen above.

A small triangular fold of the pericardium is reflected from the left pulmonary artery to the left upper pulmonary vein – the fold of the left caval vein (vestigial fold of Marshall) (Figure 1.6). It contains a fibrous strand, called the ligament of the left caval vein, that is a remnant of the left common cardinal vein (left duct of Cuvier) and that extends downwards in front of the root of the left lung to the back of the left atrium where it is continuous with the oblique vein of the left atrium. The fold frequently forms the anterior wall of a small blind recess, the mouth of which is directed to the left. In the undissected state connective tissue joins the aorta and pulmonary trunk, and there is no cavity between them. The pulmonary end of the arterial duct lies within the pericardial cavity; its distal part is outwith the pericardium. The pericardium is not essential to life, nor the efficient working of the heart, which operates adequately even when the pericardium is removed. The phrenic nerves descend on the outer lateral aspects of the pericardial sac, one on each side.

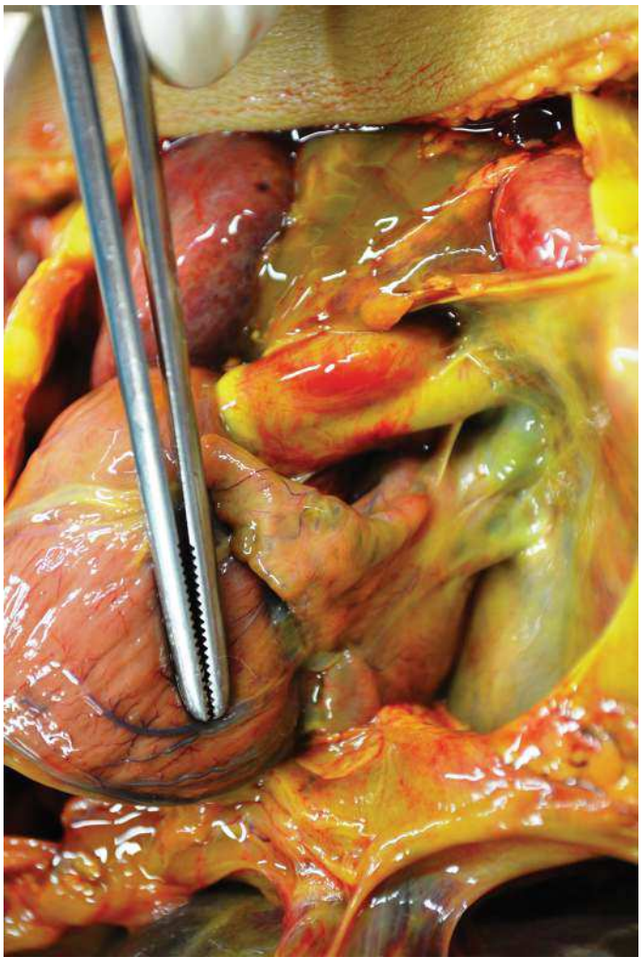

Figure 1.6 Fold of Marshall. The heart viewed from the left side and displaced by forceps to the right to display the left pulmonary artery and the left pulmonary veins. A fold of pericardium runs from the inferior surface of the left pulmonary artery to the upper border of the left upper pulmonary vein. This is the vestigial fold of Marshall, and it is continuous with the oblique vein of the left atrium (not visible in this picture). The transverse sinus of the pericardium lies anterior to the fold. Posterior to it is a blind-ending recess of the pericardial cavity.

1.2.3 The Right and Left Atrium

The right atrium comprises three components: a smooth-walled venous component; an atrial appendage; a vestibule supporting the tricuspid valve.

1. The venous component lies between the orifices of the superior and inferior caval veins, encompasses the orifice of the coronary sinus and is smooth walled (Figure 1.7). Embryologically it derives from the sinus venosus and is separated from the atrial appendage externally by the terminal groove and internally by the terminal crest (crista terminalis) (Figure 1.8). The terminal crest originates on the right atrial aspect of the interatrial septum and passes anterior to the mouth of the superior caval vein onto the lateral wall of the atrium and extends downwards to pass anterior to the orifice of the inferior caval vein where it is continuous with the eustachian valve.
2. The right atrial appendage is triangular in shape and has a broad junction with the atrium (Figure 1.9). It contains multiple parallel trabeculations – pectinate muscles – that extend around the greater part of the orifice of the tricuspid valve and are limited by the terminal crest (Figure 1.8). Externally, the junction of the crest of the appendage with the superior caval vein marks the site of the sinoatrial node (Figure 1.10).
3. The vestibule supports the tricuspid valve, and it is smooth. Situated in the vestibule, between the orifice of the coronary sinus, the attachment of the tricuspid valve and the membranous septum (see Section 1.2.7), lies the

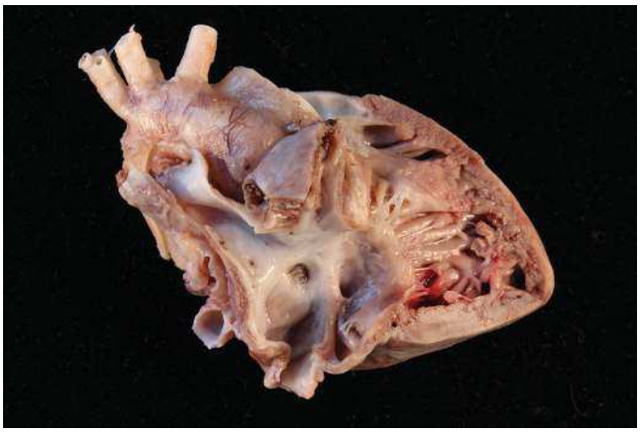

Figure 1.7 The right heart dissected to show the septal structures. The parts of the right atrium shown lie between the orifices of the superior and inferior caval veins. Visible are the interatrial septum, including oval fossa, vestibule of the tricuspid valve, and coronary sinus orifice. In this view only the origin (just above the oval fossa) and insertion (from the junction with the inferior caval vein, eustachian valve, and above the coronary sinus) of the terminal crest are seen, and the muscular trabeculations and appendage are not included. This part of the atrium derives from the sinus venosus and is smooth walled.

atrioventricular (AV) node. The bundle of His exits the node anteriorly to penetrate the membranous septum and divide astride the crest of the muscular interventricular septum giving rise to the right and left bundle branches.

The interatrial septum is smaller than it appears. The true septum comprises only the oval fossa with its rim (Figure 1.11). The remainder of the party wall with the left atrium is formed by infolding of both atrial walls with a sandwich of extracardiac adipose tissue (Figure 1.12). The oval fossa is closed by a flap valve. About 20% of the population have a valve that is

Figure 1.8 Terminal crest. The right atrium in a simulated four-chamber view of the heart. The interatrial septum runs diagonally across the field. A little beneath it, and separated from it by the orifice of the superior caval vein (not seen in this view), is the terminal crest – a solid rounded bar of atrial muscle that runs from superior to inferior delimiting the atrial appendage, and from which muscular trabeculations arise at right angles and extend to the vestibule of the tricuspid valve.

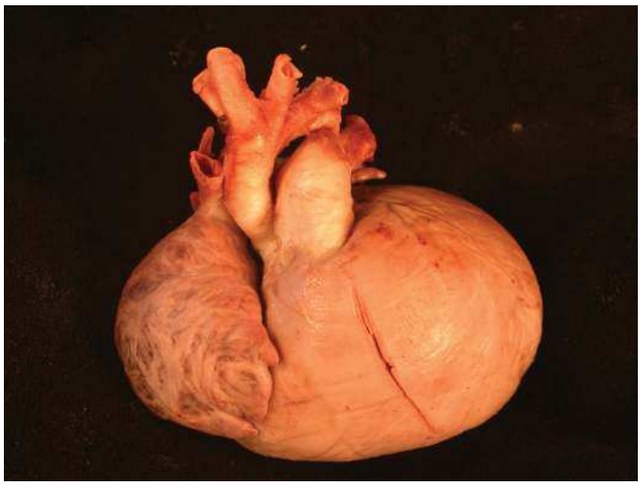

Figure 1.10 The sinoatrial node lies at the junction of the superior caval vein and the crest of the right atrial appendage.

Figure 1.9 Right atrial appendage. Viewed from the outside. The right atrial appendage is roughly pyramidal in shape, has a broad junction with the right atrium and contains parallel muscular trabeculations that extends into the body of the atrium around the vestibule of the tricuspid valve.

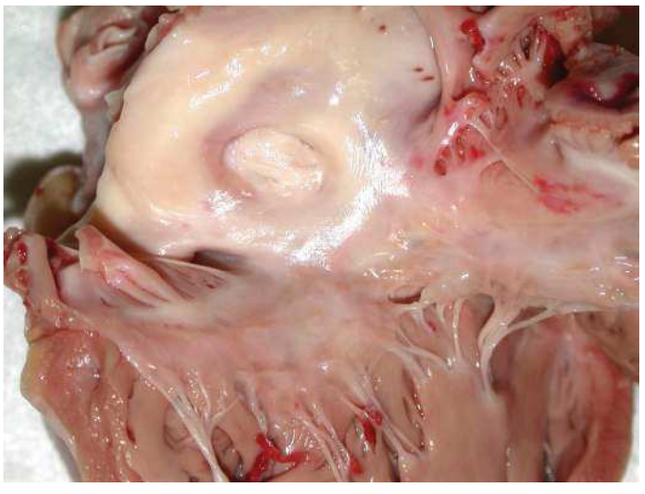

Figure 1.11 Oval fossa. The right atrium and right ventricle have been opened, and the heart is viewed from the right side. The opened orifice of the inferior caval vein is to the left midfield and the unopened orifice of the superior caval vein to the upper midfield. Lying between them is the oval depression of the oval fossa. Its rim is smooth and the flap valve completely closed. Beneath it, the orifice of the coronary sinus can be seen.

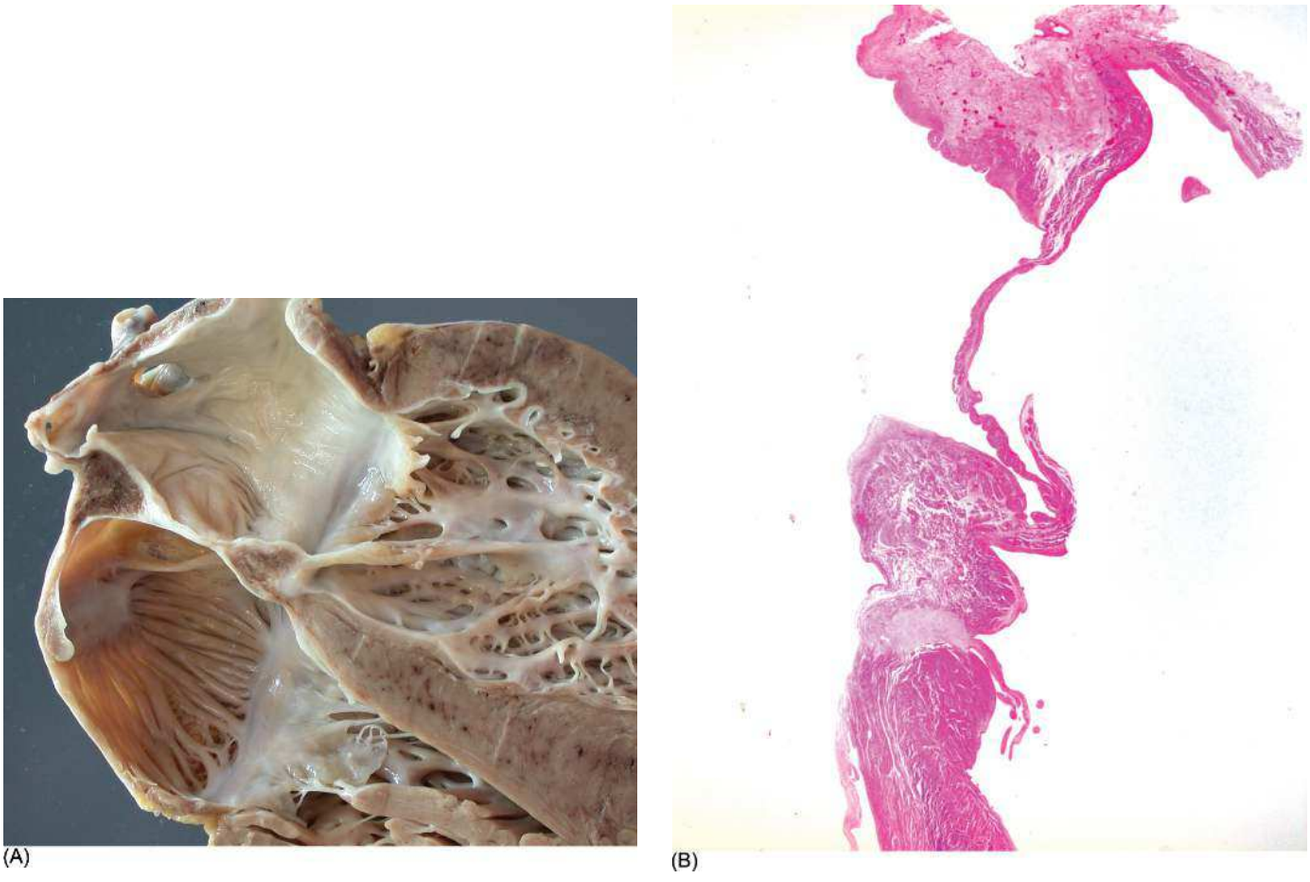

Figure 1.12 Infolding of atrial septum. **(A)** A post-mortem heart from a case of idiopathic dilated cardiomyopathy showing the interatrial septum cut vertically through the oval fossa. The rim of the fossa is muscular, but the remainder of the apparent septum is formed of infolding of extracardiac fibrous and adipose tissue. Note also the extension of muscular trabeculations around the right atrioventricular junction while the left is smooth. **(B)** The histological section shows more clearly the infolding of fibrous tissue between the two atrial walls, especially superiorly. The right atrium is on the left of the field and the left atrium to the right.

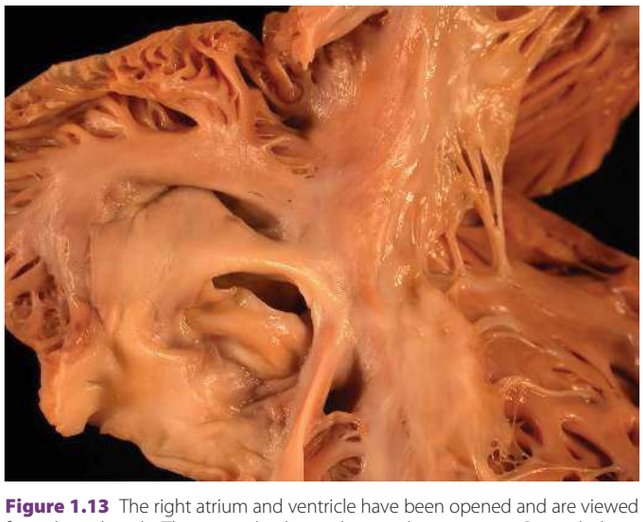

Figure 1.13 The right atrium and ventricle have been opened and are viewed from the right side. The opened right atrial appendage is on top. Beneath this is the slightly distorted orifice of the superior caval vein. Beneath this again is the oval fossa. There is persistence of the oval foramen with the flap valve not closing the defect anterosuperiorly.

probe patent at its anterosuperior margin (sometimes termed persistent foramen ovale (PFO)) (Fig 1.13).

The right atrium contains a eustachian valve of variable prominence (Figure 1.7) – a relic of the structure that in fetal life directed the venous duct (ductus venosus) blood from the inferior caval vein through the oval foramen. In some instances, the valve is a thick muscular ridge. The coronary sinus may be guarded by a thin membrane: the thebesian valve. The valve is usually attached at the postero-inferior margin and is variably fenestrated (Figure 1.14). The area between the orifices of the inferior caval vein and the coronary sinus is termed the sinus septum and is traversed by the tendon of Todaro. Between the eustachian valve and the attachment of the septal leaflet of the tricuspid valve is an area known by electrophysiologists as the isthmus. It contains a pouch-like area beneath the orifice of the coronary sinus termed the sub-thebesian recess [1].

A Chiari network may be present (Figure 1.15). This is a netlike structure in the right atrium connected to the terminal crest or atrial septum and to the eustachian or thebesian valve.

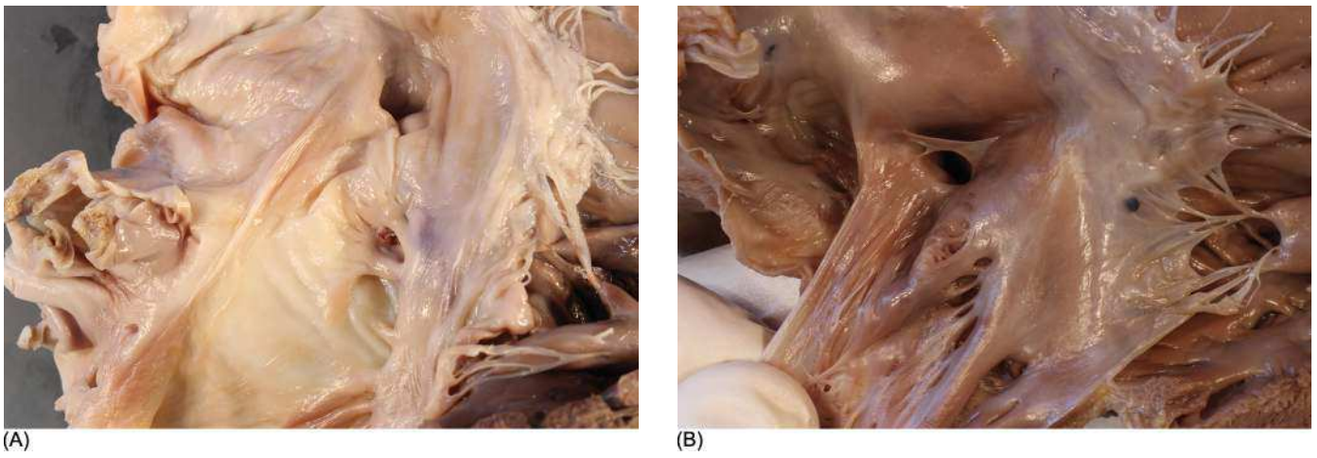

Figure 1.14 Eustachian and thebesian valves. **(A)** The right atrium and ventricle have been opened and are viewed from the right side. The oval fossa is at the top of the picture. The eustachian valve runs obliquely from bottom left to upper centre. Towards its upper extent is the oval fossa with a slight ridge of tissue postero-interiorly forming the thebesian valve. The valve is very variable in morphology, if it is present at all. **(B)** In a different heart viewed from the same vantage point, the eustachian valve has been grasped and pulled taut. It can be appreciated how it acts as a baffle to direct blood to the oval fossa. The coronary sinus shows a fenestrated thebesian valve.

Figure 1.15 Chiari network. Termination of pregnancy at 18 weeks' gestation for hypoplastic right heart. The right atrium has been opened looking towards the interatrial septum. A filigreed diaphanous structure partly covers the oval fossa extending from the terminal crest down towards the eustachian and thebesian valves. The right heart structures were small but otherwise unremarkable. It is unknown whether this Chiari network was the cause of right heart hypoplasia by obstructing the tricuspid orifice.

Usually it is highly fenestrated, but may be more solid and resemble a spinnaker sail, causing obstruction to forwards flow of venous blood across the tricuspid valve [2]. It represents the remains of the right venous valve of the sinus venosus [3].

Remnants of the left valve of the sinus venosus may be seen as lacelike structures or cords resembling tendinous cords (chordae tendineae) attached to the right side of the atrial septum in the region of the oval fossa (Figure 1.16) [4].

The left atrium is usually smaller than the right and receives the pulmonary veins. The junction between the two is marked externally by a shallow groove running vertically between the superior caval vein and the right pulmonary veins – Waterston's groove (Figure 1.17). Usually there are four: two on the right – one superior and one inferior – and two on the left – one superior and one inferior – but the number can vary. The left side of the oval fossa is generally corrugated and rougher than on the right (Figure 1.18). The endocardium of the left atrium is thicker than that of the right atrium, the thickening being caused by fibroelastic tissue. This should not be mistaken for a pathological change. The left atrial appendage is quite distinct from the right. It is long and tubular and has a narrow junction with the atrium and characteristically has a hooked extremity. Pectinate muscles are confined to the appendage and do not extend onto the atrial wall, nor around the orifice of the mitral valve (Figure 1.19). The coronary sinus runs in the posterior wall of the left atrium at the level of the atrioventricular junction. If there is a persistent left superior caval vein, the sinus is correspondingly larger and may bulge into the left atrium.

1.2.4 The Ventricles

The left ventricle is the thicker walled of the two ventricles and is ellipsoid in shape. The right ventricle is wrapped around its rightward aspect, thus giving it a more complex

Figure 1.16 Remnants of left valve of sinus venosus. The oval fossa viewed from the right atrium. The eustachian valve is grasped by forceps. The lower border of the oval fossa is buttressed by a trabecular network of fibrous cords that represent the incompletely fused remnants of the left valve of the embryonic sinus venosus.

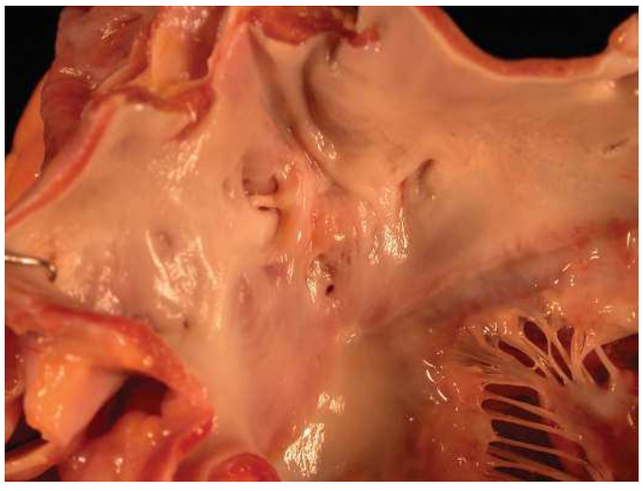

Figure 1.18 Left side oval fossa. The left atrium has been opened to display the left side of the interatrial septum. The mitral valve leaflets are apparent to the lower right. Note the opaque thick pale endocardium characteristic of the left atrium. This is especially evident on the cut edge of the wall at the upper right of the field where the endocardium occupies nearly one-third of the thickness of the atrial wall. No distinct oval structure can be recognised on the left side of the septum. Instead the attachments of the flap valve of the oval fossa are evident as a rugose area in the centre of the field.

Figure 1.17 Waterston's groove. The heart viewed from behind. Running superiorly from the inferior caval vein to the superior caval vein is a shallow groove marking the junction between the right and left atrium – Waterston's groove.

shape (Figure 1.20). From the point of view of descriptive anatomy, the ventricles have three components: an inlet, comprising the atrioventricular valve and its supporting structures; an outlet supporting the arterial valve; and an apical trabecular component linking the two (Figure 1.21). It is important to keep in mind that the components on the right and left side are not perfectly aligned. This apical trabecular component is the most constant and most characteristic feature of the ventricles. On the right side, the septal aspect of the apex shows thick muscle bundles termed trabeculations (trabeculae) that have a roughly parallel orientation along the long axis of the septum. The most prominent of these, the septomarginal trabeculation (trabecula septomarginalis), extends nearly the full length of the septum. Shaped like the letter Y, its stem extends from the apex upwards, its anterior limb extends in the outflow tract to the pulmonary valve and its posterior limb extends backwards, supporting the medial papillary muscle of the tricuspid valve (Figure 1.22). There is considerable normal variation in the posterior limb – in some cases it extends posterior to the membranous septum and in others anterior [5]. The septomarginal trabeculation is usually incorporated into the muscle of the septum analogous to an engaged pillar or pilaster, but may occasionally, at least in the fetus, be a largely free-standing structure (Figure 1.23). On the left side of the septum the trabeculations are fine and typically have an interwoven appearance, and the outflow tract shows a smooth septal surface (Figure 1.24).

Figure 1.19 Left atrial appendage. Explanted heart from a 14-year-old boy with idiopathic dilated cardiomyopathy, cut in a four-chamber view and viewed from behind. The orifice of the right atrial appendage is on the right side and shows extension of the muscular trabeculations from the appendage around the atrioventricular junction. By contrast, on the left side, the junction of the appendage and atrium is narrow and the trabeculations are confined to the appendage, the remainder of the atrial wall being smooth.

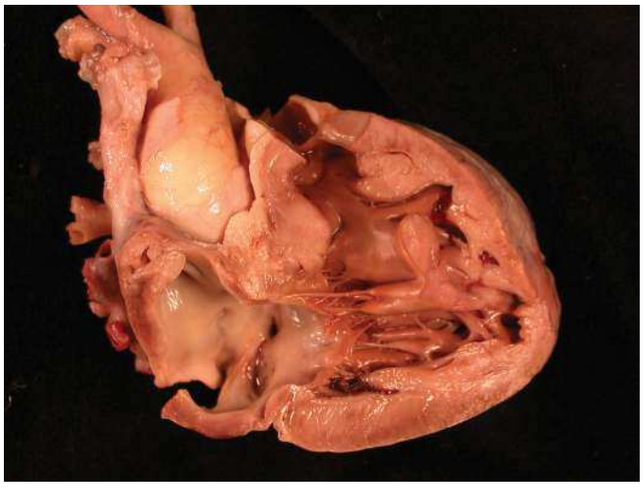

Figure 1.21 Components of septal aspect of right ventricle. The heart has been dissected to demonstrate the right-sided aspect of the interventricular septum. The tricuspid valve together with its tension apparatus (tendinous cords and papillary muscles) occupies the inlet component. Distal to it is the apical trabecular component where the muscular trabeculations are chunky and roughly parallel to one another. The outlet component lies superior to the papillary muscles of the tricuspid valve and is largely smooth. The septomarginal trabeculation occupies much of this component.

1.2.5 Atrioventricular Valves

The right-sided atrioventricular valve, the tricuspid valve, as its name indicates, has three leaflets: septal, anterosuperior and inferior. All three leaflets are anchored by tendinous cords (chordae tendineae) to papillary muscle groups situated at the leaflet commissures. The septal leaflet is also attached by cords directly to the septum (Figure 1.25). Its medial papillary muscle (muscle of Lancisi) is small and arises from the

Figure 1.20 Right ventricle wrapping around left. A short-axis dissection of the heart viewed from the apical aspect. The left ventricle is roughly elliptical in cross section and is at the lower aspect of the picture. The anterior leaflet of the mitral valve occupies much of its cavity. The right ventricle is wrapped around the left and extends from the left of the picture, where the right atrium and tricuspid valve are seen to the right, where it disappears up towards the pulmonary valve. Occupying the "hinge" region is the supraventricular crest. The bulge upwards in the interventricular septum at the site of insertion of the supraventricular crest into the septum represents the stem of the septomarginal trabeculation. The anterior limb of the septomarginal trabeculation extends upwards into the right ventricular outflow tract. The posterior limb is obscured by anterior and septal leaflets of the tricuspid valve. The commissure of the valve is supported by the medial papillary muscle attached to the posterior limb.

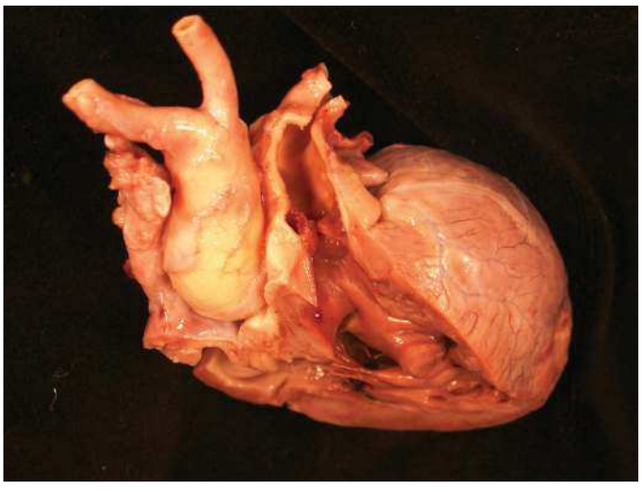

Figure 1.22 Septomarginal trabeculation. The same dissection as in Fig 1.21 rotated forwards to demonstrate the right ventricular outflow tract. The Y-shape of the septomarginal trabeculation can be readily appreciated with the anterior limb of the Y extending up to the pulmonary valve. Inserted between the limbs of the Y is the supraventricular crest, which separates the pulmonary valve from the tricuspid valve and which forms the posterior wall of the subpulmonary infundibulum. Externally, the right coronary artery travels along the upper border of the supraventricular crest. Note the spiral configuration of the aorta and pulmonary artery relative to each other.

posterior limb of the septomarginal trabeculation (Figure 1.20). Frequently there are associated small accessory papillary muscles variably located around the muscle of Lancisi [5]. The anterosuperior leaflet is the largest of the three leaflets of

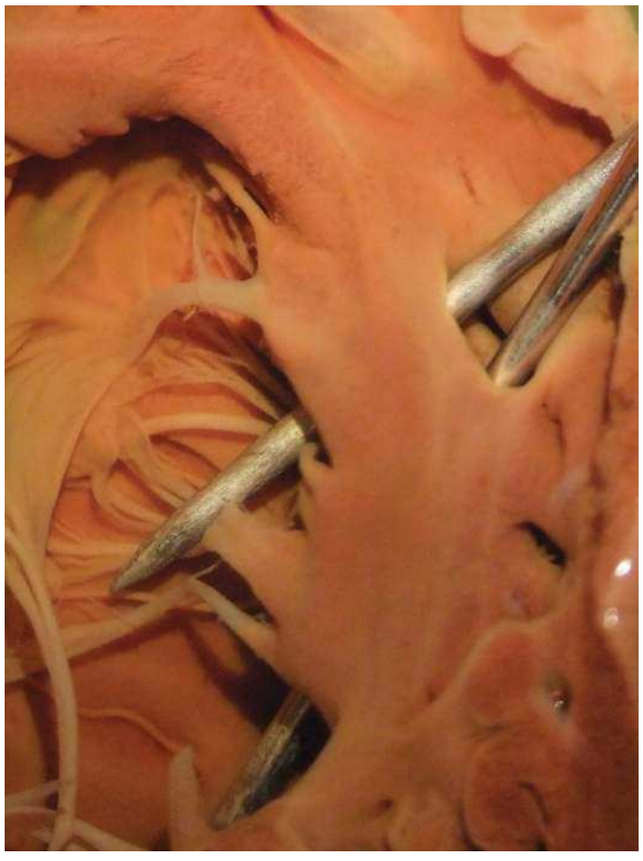

Figure 1.23 Free-standing septomarginal trabeculation. A fetus of 24 weeks' gestation with hypoplastic left heart. The dissection of the right side of the interventricular septum shows a septomarginal trabeculation that is largely free standing. Pins have been inserted between the stem of the trabeculation and the septum to demonstrate the lack of attachment.

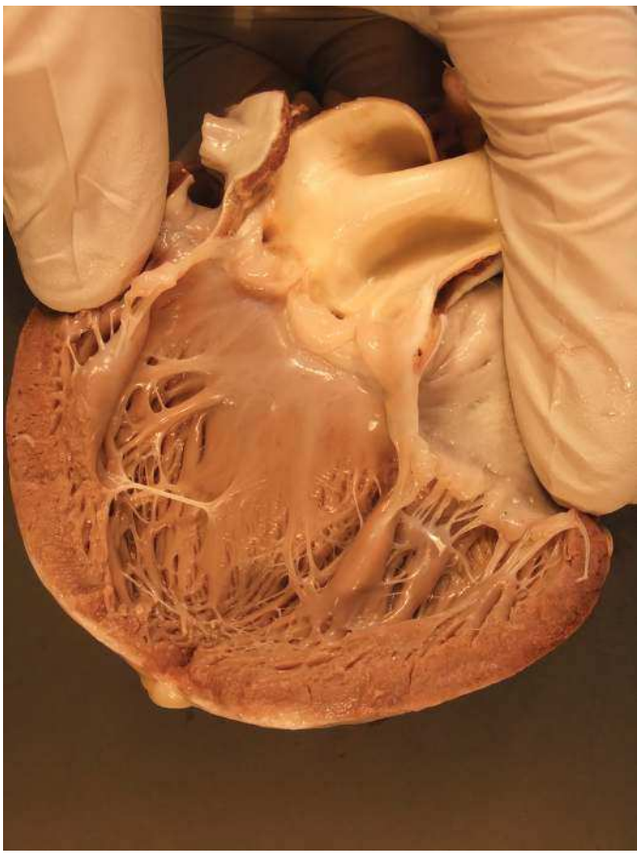

Figure 1.24 Septal aspect of left ventricle. The left ventricle has been opened along its lateral margin and splayed to demonstrate the structures on the left aspect of the interventricular septum. The inlet component is occupied by the mitral valve leaflets, their attached tendinous cords and papillary muscles. The apical component shows fine trabeculations with a criss-cross configuration. The outlet component is smooth.

the tricuspid valve and its anterior papillary muscle is prominent. The inferior leaflet is less conspicuous, as are its papillary muscles.

The left atrioventricular valve – the mitral valve – comprises two leaflets: a large rectangular, anterior (or aortic) leaflet, and a mural leaflet, which is attached to about two-thirds of the atrioventricular junction (Figure 1.26). Two large papillary muscle groups, termed anterolateral and posteromedial, support the commissures of the leaflets. The anterior leaflet is attached to the interventricular septum only on its postero-inferior aspect; the left ventricular outflow tract is interposed between the ventricular aspect of the leaflet and the septum. Thus, there is fibrous continuity via a subaortic fibrous curtain between the anterior mitral leaflet and the non-coronary cusp and part of the left coronary cusp of the aortic valves (Figure 1.27). The two lateral margins of this area of fibrous continuity show fibrous thickening, the so-called right and left fibrous trigones, the right fibrous trigone being in continuity with the membranous septum and the left fibrous trigone anchoring the fibrous curtain to the muscular septum.

In a small percentage of normal hearts a small band of muscle separates mitral and aortic valves [6].

The attachment of the mitral valve to the left side of the interventricular septum is higher than the attachment of the tricuspid valve to the right side of the septum, a feature termed offsetting, and easily detected on echocardiography and useful for identifying the ventricles (Figure 1.28). This means that there is an area between the two attachment sites where there is a potential communication between the left ventricle and the right atrium – the so-called atrioventricular septum.

Especially in the neonate, small blood-filled cysts may be present on the leaflets of the atrioventricular valves (see Section 1.3.4 for more detailed discussion). Yellow thickenings may be seen on the anterior leaflet of the mitral valve even at a young age (Figure 1.27) [7].

1.2.6 Interventricular Septum

The septum between the two ventricles is predominantly muscular and usually of a similar thickness to the rest of the left

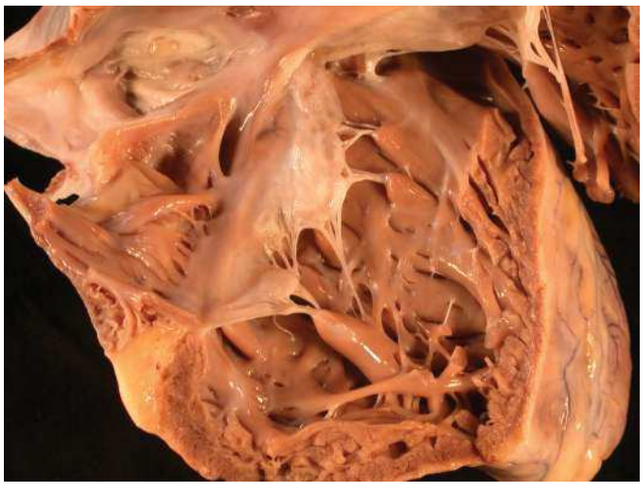

Figure 1.25 Tricuspid valve. A heart opened to display the tricuspid valve. The septal leaflet, as its name implies, is attached to the spetum and shows short cord-like attachments to it. The medial papillary muscle is a small structure at the top centre of the field. The inferior leaflet has been cut through to open the heart. Nonetheless, its papillary muscle is visible towards the bottom centre. The anterosuperior leaflet is the largest and occupies the upper right field. Its papillary muscle is just visible.

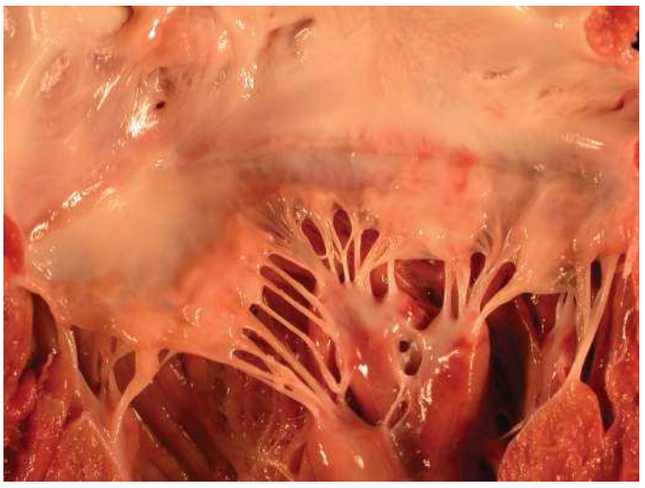

Figure 1.26 Mitral valve. The mitral valve has been opened between the lateral junction of the anterior leaflet (to the left of the field) and the mural leaflet. The posteromedial papillary muscle group occupies the centre of the field and the anterolateral group has been divided with components on the extreme right and left of the lower part of the field. Note that the anterosuperior leaflet is attached to about only one-third of the valve circumference but has a greater depth. Thus, when closed, the anterosuperior leaflet occupies the greater part of the cross sectional area of the orifice and is encompassed on three sides by the mural leaflet.

Figure 1.27 Subaortic fibrous curtain. The left ventricular outflow tract has been opened and the anterosuperior leaflet of the mitral valve retracted to the right of the field. The proximal aorta, aortic valve and septal aspect of the left ventricular outflow are visible. The cut passes through the left coronary cusp of the aortic valve. The two intact leaflets are the right coronary leaflet to the left of the field and the non-coronary leaflet in the centre. In the fibrous triangle between the right and non-coronary cusps lies the membranous septum. The mitral valve is in fibrous continuity with the non-coronary cusp and part of the left coronary cusp – the so-called subaortic fibrous curtain. That part of the fibrous curtain adjacent to the membranous septum is thickened as the right fibrous trigone. The left fibrous trigone attaches to the muscular interventricular septum. Note that even though this child was only five years old at the time of death, there are fatty streaks in the fibrous curtain.

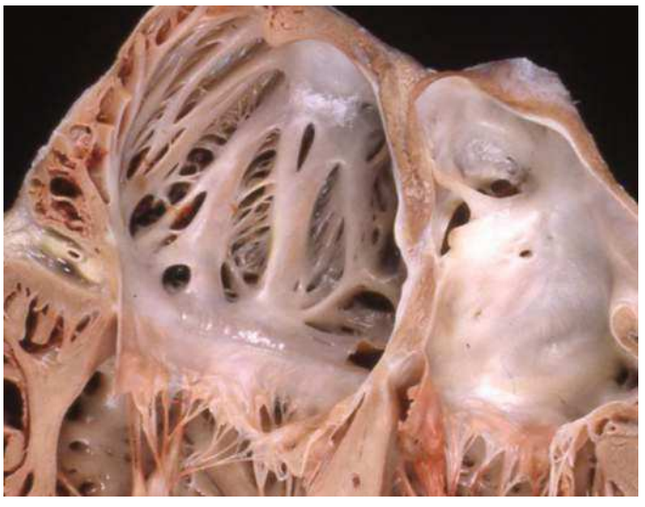

Figure 1.28 Tricuspid–mitral offsetting. Heart cut in a simulated four-chamber view. The anterosuperior leaflet of the mitral valve is attached to the septum at a higher level than the septal leaflet of the tricuspid valve. The area of the septum lying between the two attachments is the atrioventricular septum.

ventricular wall. The right ventricular aspect has already been discussed in detail above. The left ventricular aspect has a finely trabeculated apical aspect. The upper part of the left side of the interventricular septum is usually smooth and forms the left ventricular outflow tract (Figure 1.24). In this area and immediately beneath the aortic valve there is a small fibrous area situated between the right and non-coronary cusps of the aortic valve and extending beneath the non-coronary cusp, where the septum is very thin and completely fibrous – the membranous septum (Figure 1.29). This can be dramatically demonstrated by transillumination (Figure 1.30). On the right