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Editor

Cardiac Pathology

A Guide to Current Practice

Second Edition

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To the family of Lucy Atherton, and all similar families, who have allowed the use of autopsy images for the education of all those practicing cardiac pathology.

Preface to the Second Edition

It is some 8 years since the first edition was published. Since then, I have been grateful to receive helpful comments and suggestions as to how to improve the publication. It is maintained that good cardiac pathology knowledge is always to be balanced alongside awareness of embryological and developmental matters, current therapeutics, modern imaging techniques, and a reasonable understanding of cardiac electrophysiology. These subjects have kept their place in the publication, along with the other standard cardiac pathology chapters. The rising use of postmortem radiology prompts its appearance within the autopsy chapter.

As always, thanks go to the chapter authors for their unfaltering dedication to the project. They have built upon the previous edition and the work of other practitioners. Likewise, I am indebted to my colleagues who have shared their interesting cases, thereby adding to the photomicrographs and macroscopic images in the publication. As previously, the support and understanding from the publishers have been valued, and my grateful thanks are also given to Seonaid Ashby for the secretarial support. Lastly, my ongoing thanks go to Grace, Miranda, and Elara who have given me the time to work on this project.

Sheffield, UK
October 2019

S. Kim Suvarna

Contents

The Normal Adult Heart and Methods of Investigation

S. Kim Suvarna

1 Introduction

The heart is a complex, folded and hollow muscular structure situated just to the left side of the mid-low sternum when viewed from the front of the body, being enclosed by the pericardial sac and joined to the great vessels [1–3]. It develops its shape from embryological folding of the cardiac tissues (see chapter "Embryology of the Heart") [4]. The surface/external landmarks of the cardiac tissues, viewed from the front of the body, are the right and left parasternal second intercostal spaces down to the right sixth costal cartilage with the apex of the heart being in the fifth left intercostal space mid-clavicular line.

To appreciate any cardiac disorder one must first appreciate normality. This simple statement is deceptive since the heart is an organ with a complex 3-dimensional architecture, along with mechanical functionality. It is made mostly of muscle, although the muscular tissue varies in format across the chambers, with a dynamic microscopic layout and an elegant electrophysiological function. There are non-muscular tissues are also present within the heart, namely blood vessels, nerves, connective tissues and fat. These are normal, but excessive amounts of fat and fibrous tissues may point to a pathological process, as discussed later in this book.

In this chapter the normal heart is considered purely from the macroscopic and histological perspective, along with attention to specialised cardiac tissues and structures. It should be appreciated that the images have been derived following standard autopsy and referred examinations [5, 6]. This often involves separating the mid-ventricular tissues in transverse section through to the apex. Other views in this chapter, and elsewhere in the book, show cardiac tissues with the ventricles intact.

There is no single, or perfect, method to consider and assess cardiac tissue. Indeed, all examinations should be guided by the anatomical or clinical query to be considered by the pathologist and/or other medical practitioner. However, any cardiac sample examination and assessment requires a clear understanding of relevant clinical data—which may often include results of a variety of tests (see chapters on "Cardiac Imaging" and "Electrophysiology") along with "Medication History".

2 Pericardium

The pericardium is a serous cavity/sac-like structure (Figs. 1 and 2), with slight distensibility, encasing the heart and its large vascular connections. It measures about 1 mm in thickness although peripheral mediastinal fat may make it appear more prominent. At the superior boundary it may abut the thymus. It has a well-defined dense fibrous wall with collagenised tissue. As stated, a variable amount of fat lies immediately adjacent. There is normally only a scanty amount of clear (serous) fluid within the pericardial sac, generally less than 0.5 ml. This fluid allows for heart movement freely during contraction (systole) and relaxation (diastole), as well as movement of the chest.

The pericardial sac (containing the heart) is in direct continuity with the adjacent mediastinal soft tissue structures of the oesophagus, thymus and the lungs on either side [2]. The inferior aspect of the pericardial sac is bounded by the diaphragm. The superior aspect, comprising the great vessel tributaries and soft tissues, runs up to the thoracic inlet [2].

Histologically the pericardium, and the outer surface of the heart, is lined by a monolayer of bland cuboidal mesothelial cells (Fig. 3). The wall of the pericardial tissues comprises dense fibrous connective tissue with some adjacent fatty connective tissue. There is a limited blood and lymphatic vascular network present with scanty nerve fibres. There is normally no inflammatory cell component.

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Fig. 1 The pericardium has been opened and shows the heart within the smooth-surfaced pericardial sac

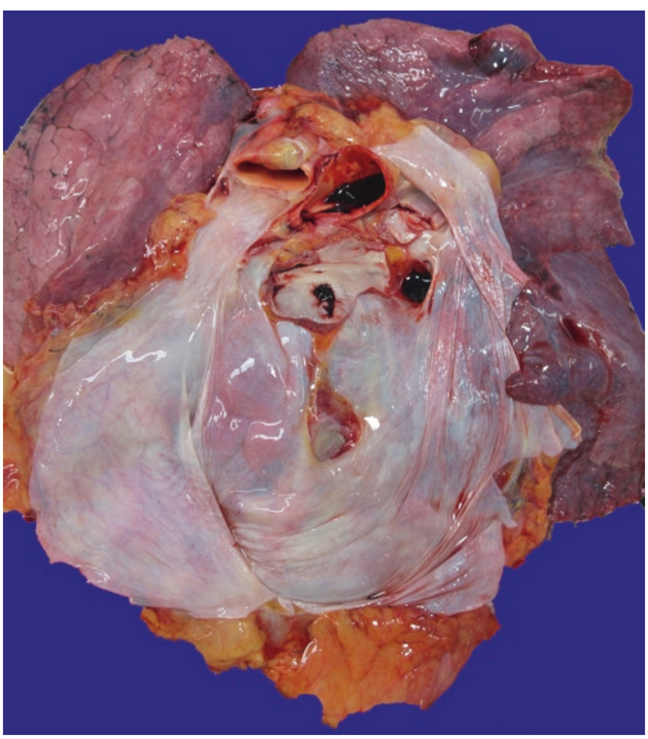

Fig. 2 The back of the pericardium is seen, after heart tissue removal. The openings of the venae cavae, aorta, pulmonary artery and pulmonary veins are visible

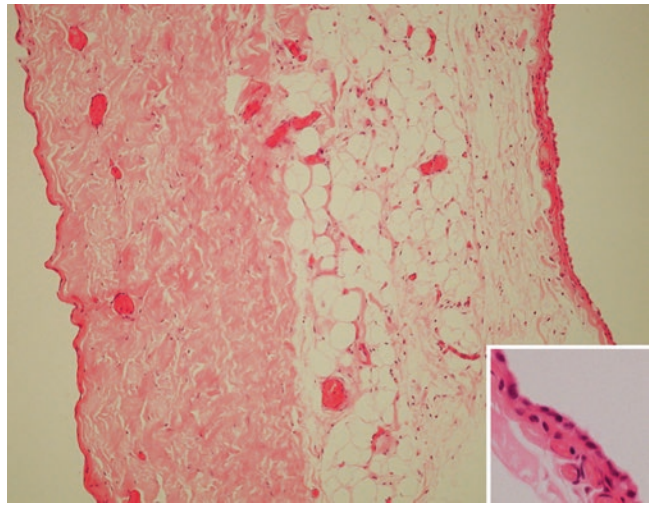

Fig. 3 Histology of the pericardium demonstrates mesothelium either side, reflecting the pericardial and pleural aspects. There are scattered vascular channels and some fibrous and fatty connective tissues. The high magnification (low right inset) shows detail from the regular monolayer of mesothelial cells (Haematoxylin & eosin)

3 External Cardiac Morphology

The external (epicardial) surface has a smooth aspect with some fat usually being evident (Fig. 4). Rarely, for those with significant systemic disease/malnutrition, these may be no appreciable fat. There may be minor fibrous thickening on the anterior epicardial surface of the right ventricle where the heart 'rubs' against the pericardium/chest wall (Fig. 5). A moderate and variable amount of fat is usually seen running parallel/alongside the coronary blood vessels and in the grooves between chambers. There is a small lymphatic vascular network present along with a few lymphoid cells often to be found in the fat.

The heart is generally regarded about the size of the individual's fist in health, growing from childhood through to adult status. However, it may be considerably enlarged in varying states of disease. However, the concept of 'fist size' is unreliable various and measurements may give a better method for assessment. Indeed, the weight of a normal heart may be assessed only if empty of blood and detached from adjacent tissue. There exist tables to compare cardiac weight against body mass, which may assist analysis—with predicted weights and a population range [7, 8]. Alternatively, one may make a basic calculation against the body mass, with expected values of 0.45% and 0.4% for males and females respectively [9]. However, the standardised charts and calculation derivations might be argued to be dated and thereby not reflective of current diets and exercise habits. Some caution always needs to be exercised when considering the cardiac weight. Indeed, there is some evidence to

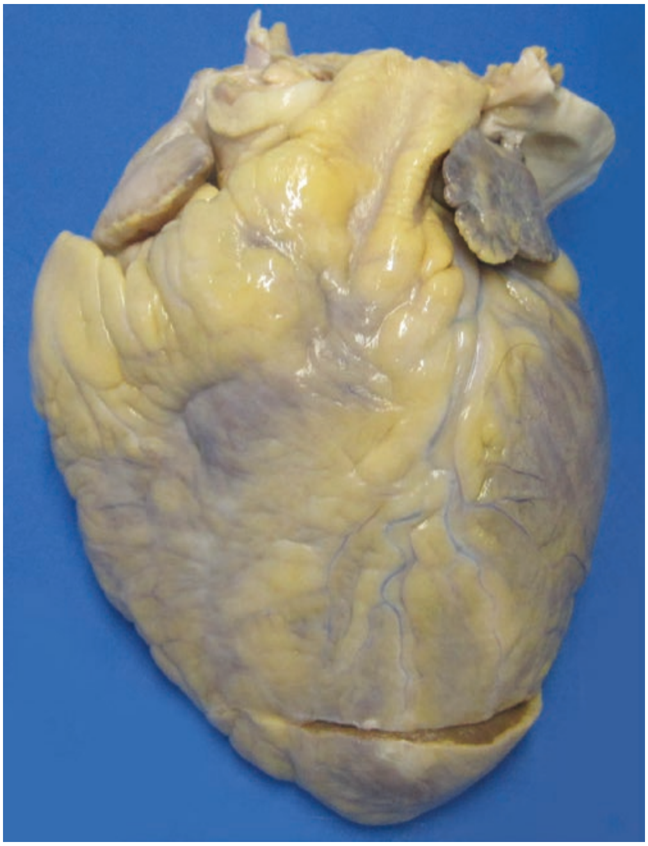

Fig. 4 The heart is seen from the anterior aspect with two auricles at the top. The ventricular chambers are seen with coronary vessels coursing across the surface

Fig. 5 The pericardium often has a small area of semi-opaque fibrous tissue. This reflects rubbing of the heart against the pericardium during contractions in life

suggest that a higher figure (possibly 0.51% of body mass) might be more appropriate [10, 11].

In simple terms, there are four chambers with great vein and artery connections evident when viewed from the outside. The two atrial and two ventricle compartments are separated at the level of the coronary sulcus by the central fibrous body. There is internal septation into right and left halves by the inter-atrial and inter-ventricular tissues. Whilst often displayed in planar two-dimensional view, it should be appreciated that, when the individual is standing, the thoracic anatomy points the heart partially downwards and towards the left side of the body [2, 12]. Thus, when the individual is standing upright, the base of the heart is mostly the right atrium and right ventricle.

The chambers are best considered in sequential format, and convention has it that all analysis follows "the flow of the blood" [1, 5, 13]. One 'starts' in the right atrium, then considers the right ventricle, left atrium and left ventricle in turn.

4 Right Atrium and Tricuspid Valve

Externally, this roughly round chamber is noted to have an external slightly triangular appendage, or auricle (Fig. 6).

Fig. 6 The heart specimen is seen from the upper right oblique aspect, allowing inspection of the right atrial appendage in detail. The appendage has a roughly triangular structure

The right atrium derives venous blood flow from the superior and inferior venae cavae, the coronary sinus and other minor cardiac veins. The venae cavae enter the posterior and basal aspect of the right atrial chamber. There is some returning cardiac venous blood inflow from the coronary sinus, usually covered in part by a flap of tissue (Eustachian valve) (Fig. 7). The sinus may have a vestigial 'web' present, being a normal anatomy variant (Fig. 8). This sinus receives blood from the great cardiac vein, but there are small anterior (or minor) cardiac veins draining the anterior wall of the right atrium (Fig. 7).

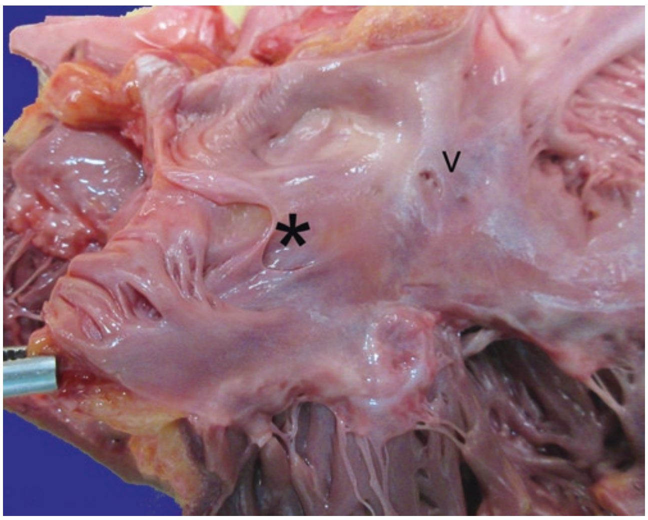

Fig. 7 The right atrium has been opened and laid almost flat revealing the fossa ovalis at the top with the coronary sinus below (*). The trabeculated atrial appendage is seen, opened onto both sides of the dissection. One can also see several small coronary veins (v) entering from the septal tissue

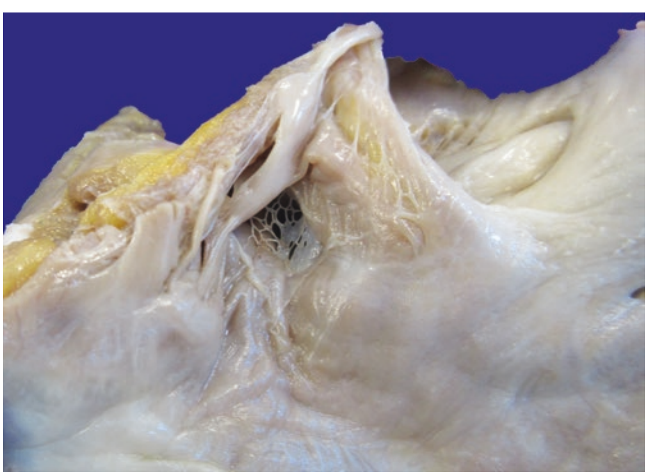

Fig. 8 The coronary sinus may have a congenital web of fibrous tissue within the opening. This has no functional deficit with regard to the heart and is not normally a focus for infective endocarditis

The chamber internally consists of three parts, termed venous, trabeculated and vestibular, as indicated (Fig. 9). There is a smooth walled posterior part (venous inlet), joining the superior and inferior venae cavae, which also derives blood from the coronary sinus. On the anterior aspect and fully involving the appendage are parallel, trabeculated muscle bands. The auricular appendage itself consists of the triangular projection running anteriorly across the upper anterior cardiac tissues (Fig. 6). There is an ill-defined groove externally (sulcus terminalis) that highlights the division of the venous (posterior) and trabeculated (anterior) zones internally with a ridge (crista terminalis). The other part of the chamber is the smooth surfaced vestibule (at the base of the chamber) being the support tissue for the tricuspid valve and the outflow for this chamber (Fig. 9).

The inter-atrial septum shows the fossa ovalis, being an oval depression/residuum from closed-off gestational/antenatal blood flow connection (foreman ovale) (Figs. 7 and 9). In about 15–20% of the population there is probe patency of part of the septal tissues (Fig. 10). This is a normal anatomical variation and usually without functional deficit—unless the hole remains patent into childhood/adulthood, allowing 'shunting of blood' across the septum (see chapter "Congenital Heart Disease"). The coronary sinus/Eustachian valve tissues are noted posterior-laterally in the right atrium (Figs. 7 and 9).

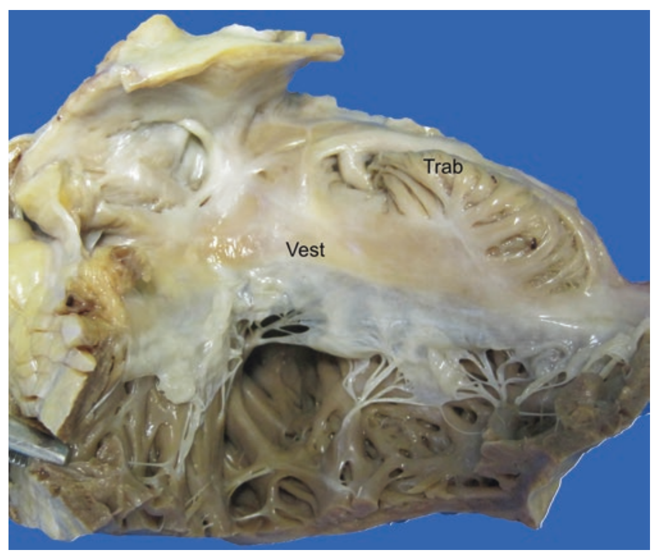

Fig. 9 The right atrium has been opened, from the posterior aspect with a cut between the superior and inferior venae cavae and with a further cut running alongside the atrial and ventricular septum. Focally there is the trabeculated appendage/auricular architecture. There are smooth endothelial surfaces elsewhere. The trabeculated (Trab) and vestibular areas (Vest) are marked. The venous part sits next to the entrance points of the superior and inferior vena cava vessels. On the left side one can see the atrial septum and fossa ovalis. The tricuspid valve is present at the base. The valve chordae are noted to run from edge of the valve to the papillary muscle tissues of the ventricle

Fig. 10 This autopsy specimen shows forceps demonstrating the probe patency of the atrial septum, passing through the foramen ovale

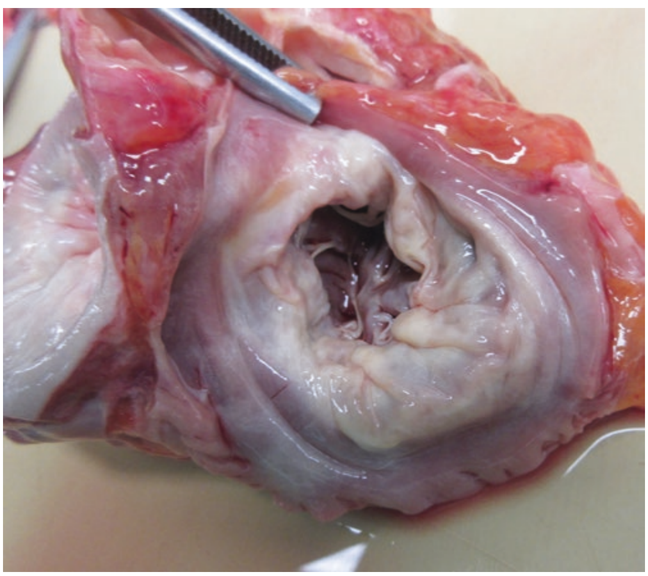

Fig. 11 The unopened tricuspid valve, seen from above, shows the flaccid valve leaflets with chordae radiating downwards into the ventricle tissues. The mitral valve is seen towards the left side of the image

Blood flows out from the right atrium across the atrioventricular orifice/tricuspid valve (Figs. 7 and 11). This usually has a circumference of 10–12 cm. It allows blood to enter the posterior aspect of the right ventricle. The valve is generally defined as three parts (anterior, posterior and septal). The tricuspid valve (Fig. 11) has a thin pliable tri-leaflet structure (less than 1 mm thickness), anchored onto the myocardial wall tissues and by chordae tendinae onto the right vestibular wall (Fig. 9). The right side of the atrio-ventricular orifice, from above, is part of the fibrous skeleton of the heart.

5 Right Ventricle and Pulmonary Valve

The second cardiac chamber is the right ventricle (Fig. 12). This makes up the anterior and inferior part of the heart (when standing erect) and has a vaguely crescentic architecture on transverse section (Fig. 13) with some features suggesting an inverted cone shape, in the longitudinal axis. There is a variable fat (adipocytic) component, often best appreciated by histology (Fig. 14). Internally, there are three parts evident, described as inflow, trabecular and outflow components (based on the path the blood takes passing through the chamber).

There are coarse trabeculations across the chamber evenly, apart from the outflow tract (conus), that is relatively smooth walled (Fig. 15). The right ventricle is thinner than the left (Fig. 13). Mature fat (adipose tissue) in variable amounts is normally present in the free wall of the chamber (Fig. 14). This should not be confused with the fat/fibrous tissue replacement of arrhythmogenic (right ventricular) cardiomyopathy (see chapters "Cardiomyopathies" and "Sudden Cardiac Death"). Given that the ventricular wall varies markedly in thickness around the circumference and has variable fat content, it is often found that different pathologists, assessing the same right ventricle tissues, will derive different wall thicknesses. Consequently, it may be pragmatic to assess right ventricle wall thickness in the outflow tract,

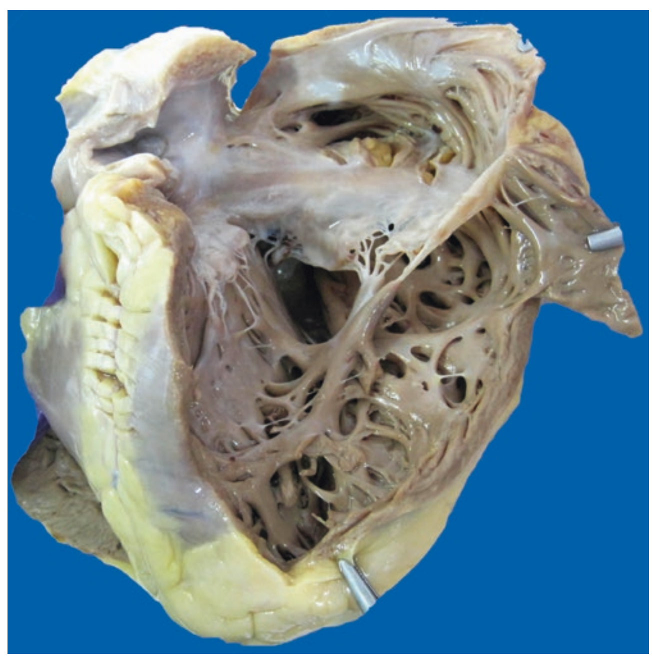

Fig. 12 The right ventricle is seen from the posterior aspect with the atrium above. The coarse trabeculations are noted and the chordae/papillary muscles are also evident. The inlet is seen immediately below the tricuspid valve. The ventricular trabeculated part is clearly demarcated and the blood passes towards the outlet/conus

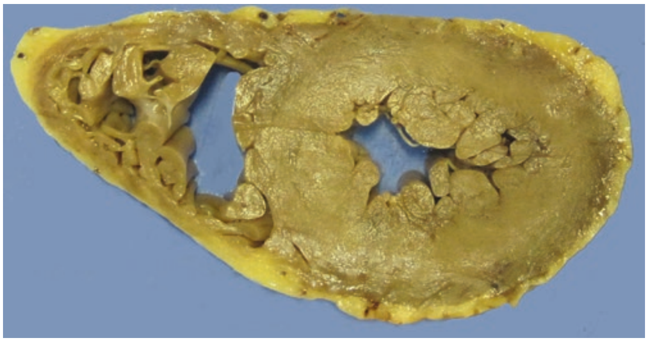

Fig. 13 A transversely sliced view of right and left ventricle tissues. The variably trabecular architecture can be fully appreciated in this view along with comparative detail of the two ventricular wall thicknesses. This view of the cardiac tissues (standard in autopsy examinations) allows the cross section of the chamber diameters can be defined. The coronary arteries are seen running through the epicardial fat peripherally

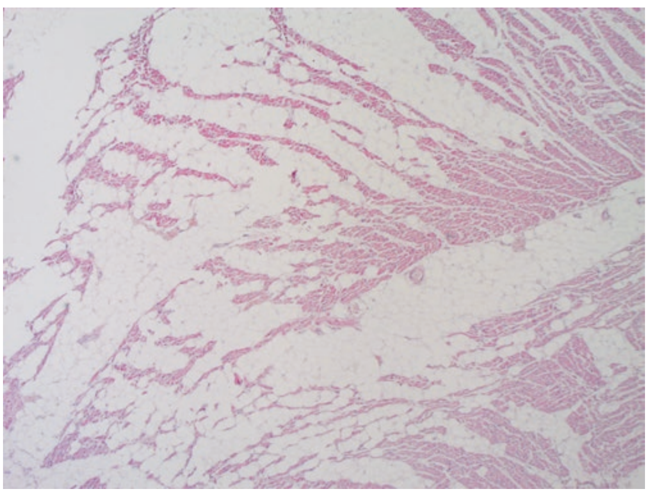

Fig. 14 The right ventricle can be very fatty and with relatively few myocytes. This should not be regarded as indicative of arrhythmogenic cardiomyopathy (see Chap. "Cardiomyopathies")

approximately 10 mm below the pulmonary valve—being a smooth area, which is easy to define and measure (Fig. 15). This area also has less peripheral fat, along with a smooth inner surface allowing for ease of assessment. The normal right ventricle outflow tract measures 2–4 mm.

There are three papillary muscles, running from the walls of the right ventricle that anchor into the free margins and ventricular undersurfaces of the tricuspid valve by fibrous cords (chordae tendinae) (Fig. 12).

The septomarginal trabeculum (Fig. 16) is a notable/well-defined band of muscle crossing the cavity of the right ventricle from the septum to the anterior papillary muscle, importantly carrying part of the ventricular bundle of conducting tissue. This ensures that the papillary muscles have already contracted (tensioning the chordae tendinae) when right ventricular contraction commences. This permits efficient closure of the tricuspid valve during systole.

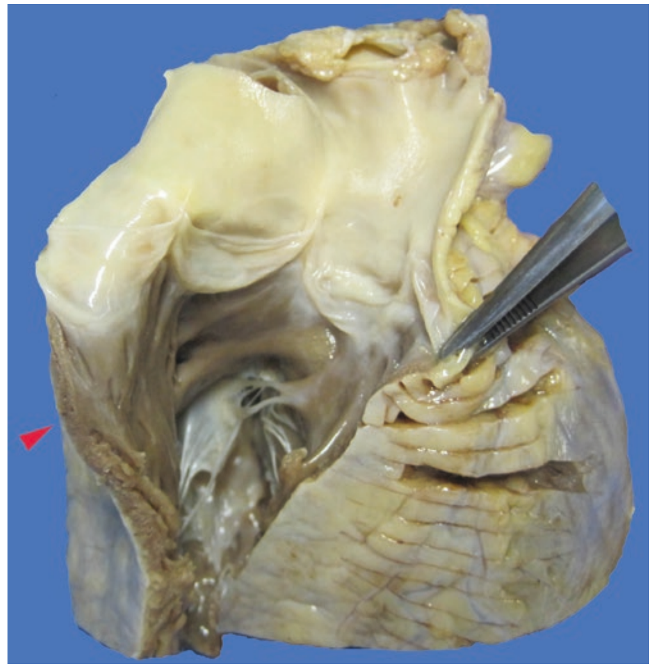

Fig. 15 The right ventricle outflow tract has been opened anteriorly allowing inspection of the pulmonary valve, smooth aspect conus tissue and proximal pulmonary artery. Measurement of the right ventricle outflow tract at this point is recommended (arrow) as this thickness is easy to define and repeat—particularly as no trabeculations are present

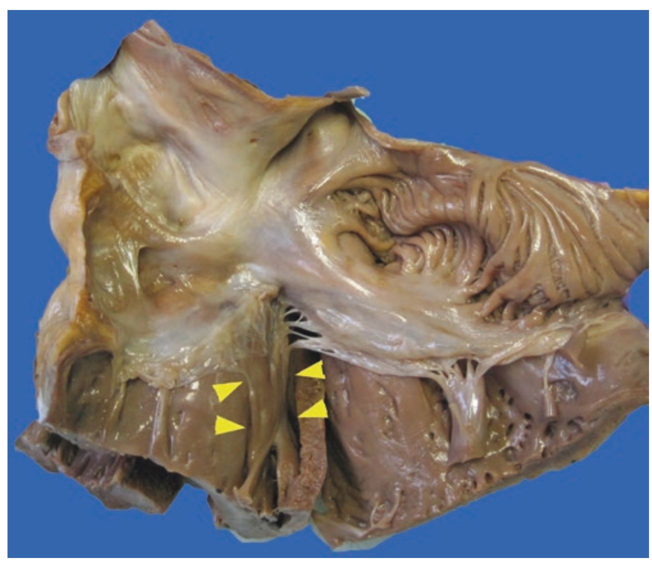

Fig. 16 The right ventricle and right atrial tissues has been opened to allow definition of the septomarginal band of myocardial parenchyma (arrowed) as it runs from the high septal tissue towards the right ventricle trabeculations and papillary muscles. This carries right bundle branch fibres to assist with effective right ventricular contraction

At the apex of the outflow tract is the pulmonary valve (Figs. 15, 17, and 18). This valve has three, roughly equal

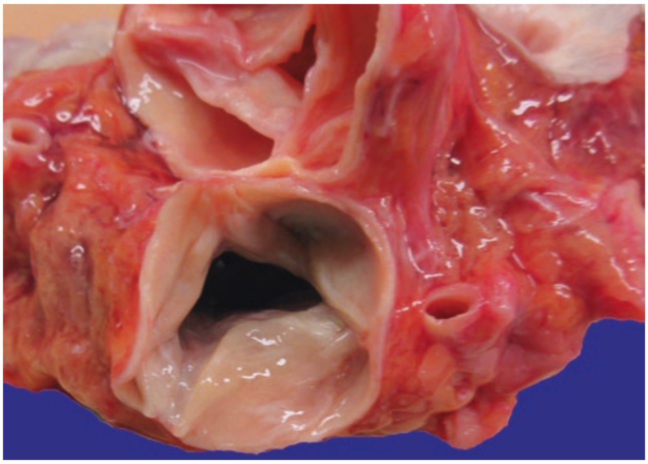

Fig. 17 This is the pulmonary valve seen from above in a freshly dissected heart showing the thin and translucent tri-leaflet architecture without coronary ostia. The aortic valve is seen at the top of the image

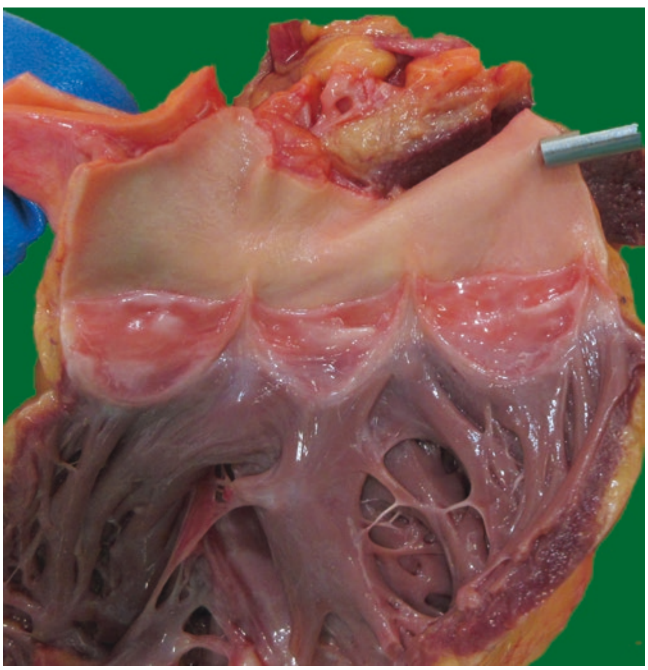

Fig. 18 The pulmonary artery root shows three normal valve cusps similar in quality with a bland intimal aspect for the pulmonary artery/trunk. One notes the absence of coronary ostia. The trabeculations of the right ventricle are coarse and thick, as compared with the left side

semi-lunar leaflets joined minimally, by commissures, to each other at the wall. The ring circumference is generally 5.5–7.5 cm. The valve function permits unidirectional deoxygenated blood to flow into the pulmonary artery and thence towards the alveolated tissues. The pulmonary artery is discussed later in this chapter.

6 Left Atrium and Mitral Valve

Oxygenated blood returns from the lungs to the left atrium through the four pulmonary veins, two from each lung. These enter the venous inlet (akin to the right atrium anatomy) high/posteriorly in the chamber. For the left atrial chamber itself, there are no specific landmarks externally—apart from the oblong (rather 'dog-ear') appendage (Fig. 19). Otherwise, the left atrium has a broadly rounded architecture.

Internally, the left atrium (Fig. 20) has a mainly smooth endocardial surface, although the left side appendage is trabeculated. The only other landmark aside from the veins and appendages, is the closed fossa ovalis. This is seen on the inter-atrial septal aspect, being the closed foramen ovale. The left arterial chamber (Fig. 20) can also be divided into the three parts, analogous to the right atrium. These regions are termed venous inlet or posterior, appendicular and vestibular, the latter supporting the adjacent valve. After atrial contraction, blood passes across the left atrioventricular orifice/mitral valve into the left ventricle. The mitral valve (Figs. 20 and 21) has a normal adult ring circumference of 8–10 cm. Contrasting with the right atrium, there are only two mitral (bicuspid) valve leaflets present. The anterior cusp/leaflet is longer in circumference and larger than the posterior, with the valve opening having a slightly curved architecture (Fig. 21) when viewed from above [6]. These

Fig. 19 The left atrium is seen from the cardiac specimen from the upper left oblique aspect. The appendage has a somewhat square architecture

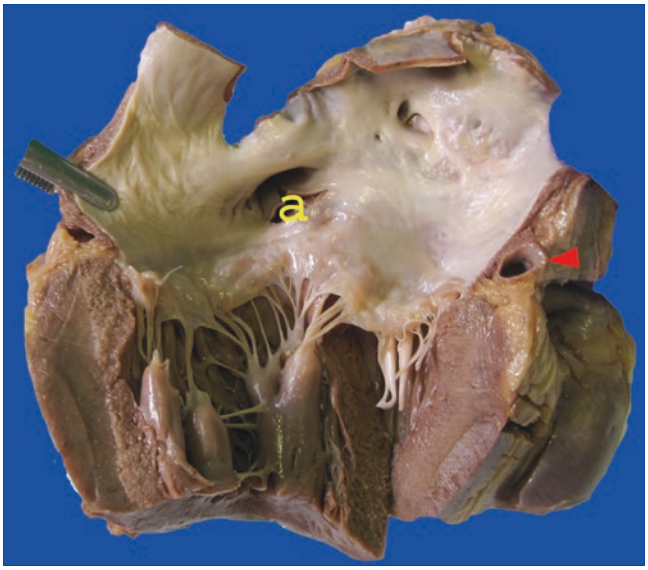

Fig. 20 The left atrium has been opened by cuts interlinking the pulmonary veins and with a further cut running alongside the atrial and ventricular septum. The two leaflets of the mitral valve are seen with adjacent chordae and papillary muscles. The auricle (a) can be inspected directly. The left atrium is largely smooth surfaced

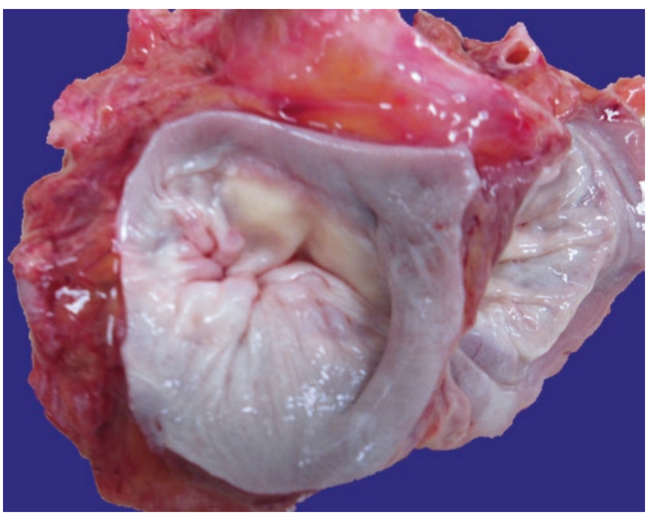

Fig. 21 The dissected heart is seen with the mitral valve, from above. The tricuspid valve is seen adjacent, towards the right side of the image

valve leaflets also have a similar structure and attachments as that of the tricuspid valve, but with two papillary muscle anchors.

7 Left Ventricle and Aortic Valve

The last chamber to be considered is the left ventricle, on the left side of the heart (Fig. 4). There is normally only scanty fat present peripherally, generally distributed on the epicardial aspect of the heart mostly alongside vascular channels (Figs. 1, 4 and 22). Readily appreciable fat within the left ventricle wall and septum often points to a pathological process, such as cardiomyopathy or prior infarction.

The left ventricular chamber is longer than the right, and has a rounded transverse cross section, with an inverted cone architecture. The chamber has trabeculations throughout, apart from the very apical part of the outflow tract (making this area useful for estimating wall thickness and possible hypertrophy). However, some prefer to estimate left ventricle wall thickness by measurements taken in the posterior, lateral, anterior free wall and septum. The left ventricle trabeculations are somewhat finer than those of the right ventricle, but are most marked towards the apex. There are two large papillary muscles, attached via chordae tendinae to the mitral valve (Fig. 22). The chordae insert into the free margins and ventricular surfaces of the mitral valve (Fig. 23).

Within the chamber (Fig. 24) there is the comparable inlet, trabecular and outflow ventricular compartment subdivisions. The aortic outflow tract/vestibule is thin muscle and, in part, fibrous. The left ventricle outflow tract passes behind the right ventricle outflow tract, approximately in perpendicular fashion, to finish at the aortic valve. The aortic valve ring has an average circumference of 5.5–8.5 cm. It is centrally positioned within the fibrous body/skeleton of the heart. Indeed, it is sometime described as wedged (centrally within a triangle) between the pulmonary valve

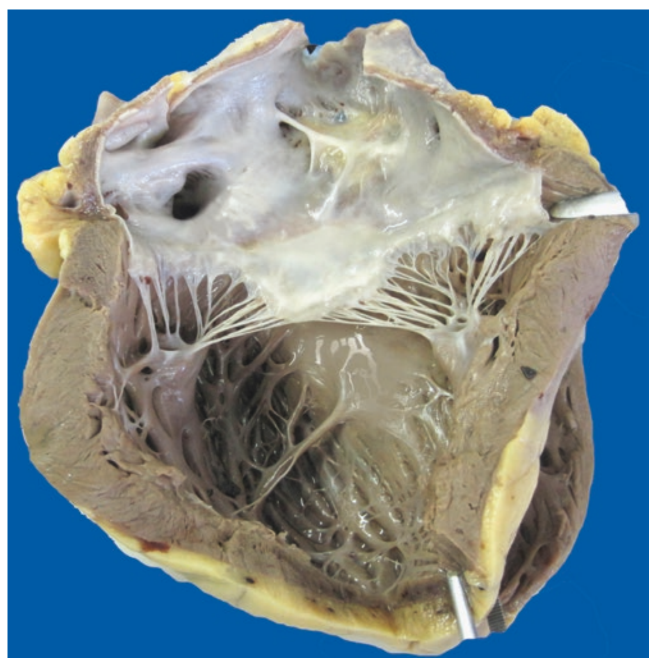

Fig. 22 The left ventricle has been opened from the back to demonstrate the atrial and ventricular chambers. As with the right side, there is the inlet, ventricular trabeculated and outlet components evident. The trabeculations are slightly finer than those on the right side. Papillary muscles are seen at the ventricular base/side walls