THE PATHOLOGY OF THE HEART

SECOND EDITION

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Preface to the First Edition

The aim of this book has been to bridge the gap between voluminous texts on cardiac pathology and the insufficiently detailed chapters included in many standard textbooks of pathology. This relatively short volume has been designed to provide practising pathologists and clinicians engaged in the study of the heart with a quick reference, thereby enabling understanding of the morphological background of clinical features and accurate interpretation of the findings at necropsy. I hope that this book will be of value to those involved in teaching, and to postgraduate students preparing themselves for a higher degree examination. I also hope that undergraduates may find it helpful.

The book is based on my seminars given to members of the various disciplines engaged in cardiac work at The Mount Sinai School of Medicine (New York City) and the Royal Postgraduate Medical School, and the National Heart Hospital and Cardiothoracic Institute (London). The pathological features and their effect on the rest of the heart are described in detail. As these alone would be meaningless, I have therefore endeavoured to relate them to clinical manifestations, radiological appearances and findings at surgical operation.

Each subject is described in a uniform manner, starting with a definition, a range of incidence wherever possible, and ending with the common causes of death. In each instance I have selected one classification – the most comprehensive in my opinion – and have summarised clinical manifestations and pathological appearances at macroscopical, histological, and where appropriate at histochemical and ultrastructural levels. Embryological considerations, associated conditions and complications, with an indication of frequency of occurrence, are listed. Aetiologies and theories of aetiologies are briefly discussed; treatment, including indications for surgical intervention where appropriate, are summarised.

Where possible only key references have been included (which necessitated, however, an occasional text in German or French), and I have also included 'Letters to the Editor' when relevant. My aim has been to cite predominantly those pertinent contributions which have appeared in major journals.

The first chapter deals with the normal heart, in order to provide a basis for comparison. In the chapter on cardiomyopathies I have introduced a new, short, clear classification. Pulmonary hypertension, a common complication of heart disease, has been discussed in a separate chapter, in which is included a short section on the primary or idiopathic form.

I am most grateful for the co-operation of authors, co-authors, editors and publishers of the various journals, who have permitted me to reproduce previously published illustrations and tables.

I am particularly grateful to my secretary, Mrs D. Wilman, who typed the manuscript and has been an immense help through the various stages of the writing of this book.

I am grateful to Mr Duncan Mackintosh of

Intercontinental Medical Book Corporation for his many helpful suggestions during the production of this book.

My thanks are also due to the Department of Photomicrography of the Royal Postgraduate Medical School, and the Photographic Departments of the National Heart Hospital and the Cardiothoracic Institute, London.

Last, but by no means least, I am grateful to all those who have attended my seminars, who through open discussion and questioning have acted as a constant stimulus.

E. G. J. Olsen, MD

Preface to the Second Edition

The aims I had set out in the preface to the first edition of this book and had hoped to achieve, have been largely fulfilled, but since its publication in 1973, great advances in the study of the heart have continued to be made. It was felt, therefore, that a second edition at this stage was timely. The general acclaim of the reviews and the continued contact with all those involved in the various disciplines concerned with the study of the heart, throughout the world, have encouraged me to proceed with this new edition.

The format of the book and the arrangement of the various chapters proved to be popular and have therefore been retained. Although this volume is almost double the size of the previous book, the numerous sections have been kept brief and concise. Much of the original text has been retained, as this formed the framework for the basic understanding of the pathology of the heart, to which succinct accounts of the advances that have been made in recent years have been added. Every section within each chapter has been critically analysed and brought up to date. Over one thousand new references and a number of new photographs have been added. Other illustrations have been exchanged to improve technical standards. Newer classifications have been listed and cogent reasons for their acceptance have been stated; alternatively, reasons for retention of an older classification have been justified. Some additional clinical aspects have been highlighted, without which pathological description would often be meaningless.

Several of the additions to this volume are worthy of special mention, such as the assessment of the size and volume of cardiac chambers, in the first chapter as well as the anatomy of muscle bundles, the modern concept and nomenclature of the ultrastructure of the normal heart, greater details of the arterial supply and lymphatic drainage of the heart.

The second chapter, dealing with hypertrophy of the heart, has been entirely rewritten and the controversy whether hypertrophy and hyperplasia, or hypertrophy alone occurs under pathological condition, has been fully discussed. Experimental approaches and the pathogenetic mechanisms of cardiac dilatation and heart failure have also been included.

Newer concepts of carcinoid heart disease, amyloid and necrosis have been detailed.

An entirely new chapter has now been devoted to the mitral valve prolapse syndrome and Marfan's disease.

I have fully analysed the monoclonal theory of atherogenesis, the natural history of arteriosclerosis and the problem of the primary or secondary role of thrombus in the pathogenesis of myocardial infarction.

New concepts of rheumatic heart disease and the more recent theory of the pathogenesis of Aschoff nodules have been added, so have rarer forms of infective endocarditis and the problems besetting haemodialysis.

For clarification, a brief section has been added introducing the seven chapters devoted to congenital heart disease and deals with the early development of the heart and the more recent proposals on nomenclature in this

rapidly changing subject. Newer concepts of embryology and classification of congenital heart disease have been included in each relevant section of each chapter. 'New' conditions, such as double outlet left ventricle have been added.

The great accumulation of knowledge in the field of cardiomyopathy has been summarised. The latest definition and classification has been detailed and each entity now included under 'specific heart muscle disease' (previously known as secondary cardiomyopathy) has briefly been described. Advances of possible aetiology, such as the role of the eosinophil in endomyocardial disease have also been included.

The increasing use of the bioptome to obtain fresh endomyocardial tissue and the various analyses that are being carried out on this tissue are succinctly reviewed.

Recent proposals for pathological description of pulmonary vascular hypertensive changes and pulmonary veno-occlusive disease have been emphasised.

The advances and achievements in cardiac transplantation have been brought up to date.

I hope that this new edition will continue to be useful to those who have found the first edition of value. I also hope that those workers engaged in the study of the heart, whether practising pathologists, physicians or surgeons, teachers or postgraduate or undergraduate students who are not familiar with the previous edition, will find the book valuable.

I would like to express my gratitude to the members of my staff for their forbearance during the preparation of the work.

The co-operation of authors, co-authors, editors and publishers of the various journals, permitting the inclusion of published material is also gratefully acknowledged.

I would like to thank Mr C. Fry and Mr P. Milford of The Macmillan Press Ltd for their helpful advice and co-operation.

I would like to thank Mrs D. Wilman for typing the manuscript and Mrs R. Christie for typing the index. I would also like to thank Mr Barry Richards of the Cardiothoracic Institute, University of London, for additional photographs.

As in the preface to the first edition my last but not least grateful acknowledgement must go to all participants of my seminars, who by their continued support and searching discussions have been a constant stimulus.

E. G. J. Olsen, MD, FRCPath

Contents

Part 1

Normal Heart and Structural-functional Changes; Degenerations and Pigmentations

1

The Normal Heart

The Removal of the Heart

In routine post-mortem examination, a midline incision from the neck to the symphysis pubis is usually made. For display of the heart, the thoracic cage is exposed by undercutting the overlying skin and other structures, so that the ribs and intercostal muscles are all that remain. A small cut - if a pneumothorax is not suspected – should then be made in the 6th intercostal space and the parietal pleura in the region of the costochondral junction on both sides; this allows the lungs to fall away. By inserting the index finger, any possible adhesions can then be divided and the adjacent spaces can be cleared before the knife is again inserted. This procedure should be continued until all intercostal spaces have been incised. The costochondral junctions must then be cleared from muscle and then divided by a sharp curved knife in an upward direction towards the operator; bone-cutting scissors may also be used. This procedure is carried out until the sternum is free, care being taken to avoid damage to the lungs, should injection of radio-opaque material be required (see below). The thoracic viscera can then be removed *en bloc*. The heart can be removed separately before the removal of the lungs.

Either method may be adopted to avoid cutting through any unsuspected abnormalities. The following steps of examination should be rigidly adhered to:

(1) Examine the outer surface of the pericardium.

(2) Follow and display the systemic venous drainage, by blunt dissection if necessary.

(3) Examine the relationship of the great vessels.

(4) Identify the pulmonary veins.

If an infected pericardial effusion is suspected, a suitable area should be selected and a hot searing iron applied to sterilise the pericardium in order to aspirate fluid for bacterial examination. The pericardium should then be incised and any fluid carefully collected and measured. The pericardium should then be opened sufficiently to allow the index finger to enter, exploring the cavity gently, dividing any recent adhesions. Dense adhesions or obliteration of the pericardial cavity may be found, especially after cardiac operations. Sharp dissection may then be necessary, but once the plane between the visceral and parietal pericardium is reached, separation is usually possible.

When the parietal pericardium is free, the pericardium can be opened. Further steps should now be undertaken:

(5) Note the position of the atrial appendages.

(6) Gently palpate the coronary arteries.

(7) Identify the ductus arteriosus.

(8) Make a small cut into the anterior surface of the pulmonary trunk about 4 cm above the position of the pulmonary valve to ascertain whether or not an embolus is present.

(9) Make an incision into the superior vena cava and extend the cut into the innominate vein and the right azygos vein.

If no abnormal features are present the heart

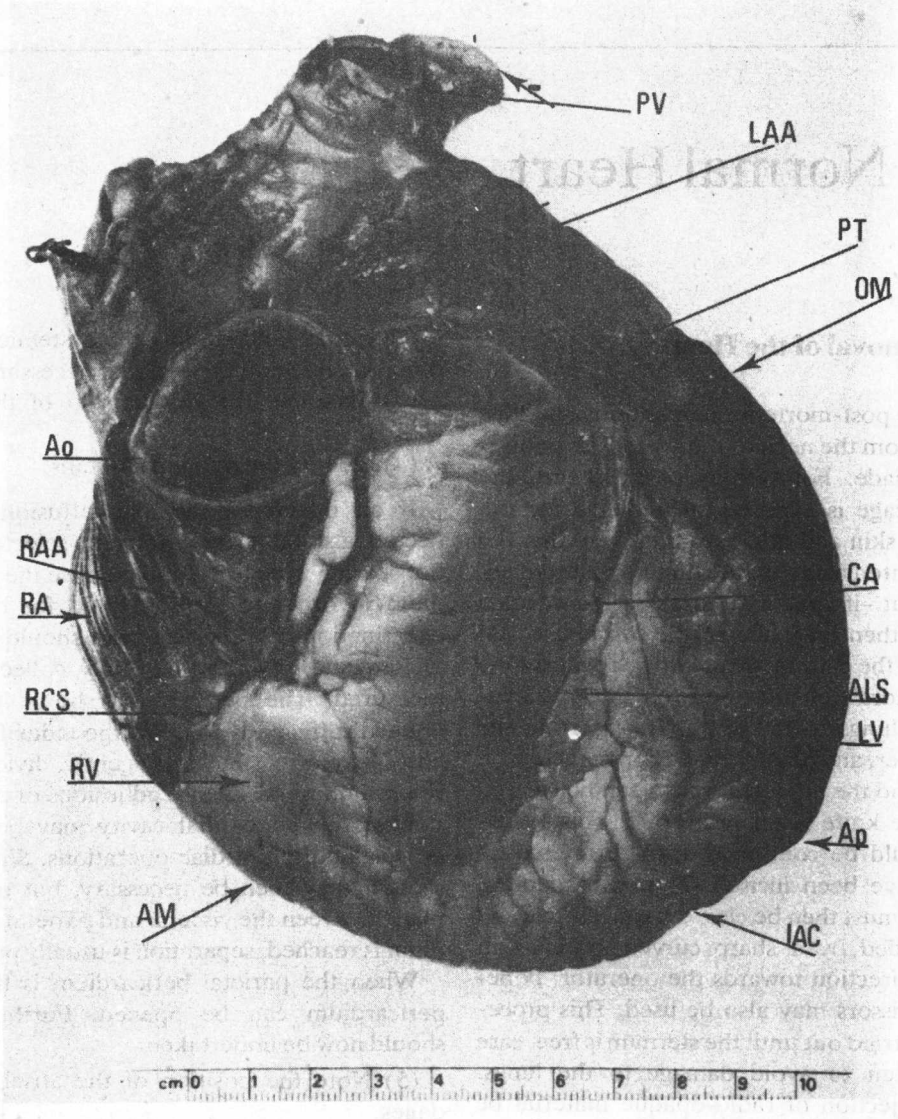

Figure 1.1 External landmarks (ventral view).
Ao, aorta; RAA, right atrial appendage; RA, right atrium; RCS, right coronary sulcus; RV, right ventricle; AM, acute margin; PV, pulmonary veins; LAA, left atrial appendage; PT, pulmonary trunk; OM, obtuse margin; CA, conus arteriosus; ALS, anterior longitudinal sulcus; LV, left ventricle, Ap, apex; IAC, incisura apicis cordis.

can now be removed. Trim the pericardium around the inferior vena cava. If the examiner is right handed, slip the left index and the middle fingers behind the pulmonary trunk and the aorta and cut with scissors through the space between the two fingers of the left hand. This procedure allows inclusion of uniform portions of the great vessels which are fairly constant when the heart is weighed. Grip the left ventricle and lift the heart so that the pulmonary veins are under gentle strain. Blunt dissect, if necessary, the left veins and then divide them about 1 cm from the left atrium. Next, cutting the pericardial reflections over the left atrium, work

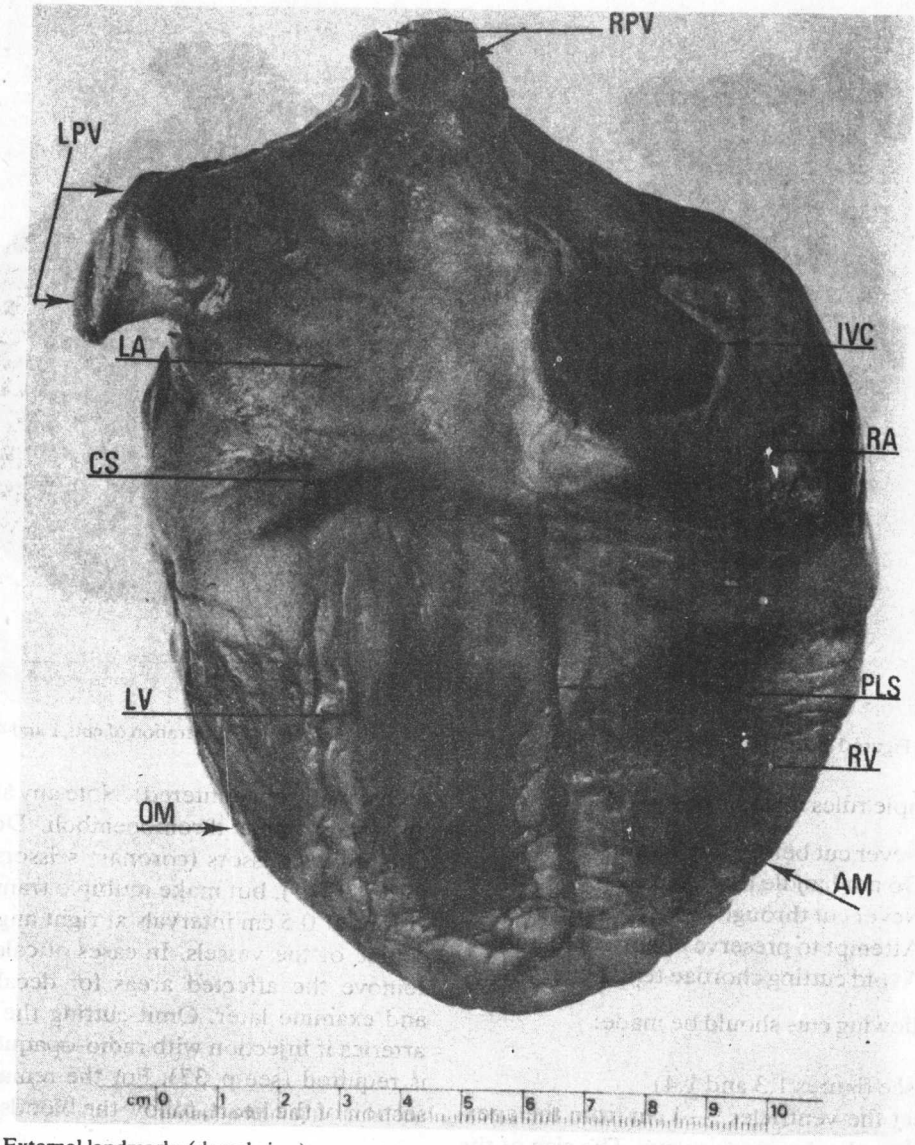

Figure 1.2 External landmarks (dorsal view).
LPV, left pulmonary veins; LA, left atrium; CS, coronary sulcus; LV, left ventricle; OM, obtuse margin; RPV, right pulmonary veins; IVC, inferior vena cava; RA, right atrium; PLS, posterior longitudinal sulcus; RV, right ventricle; AM, acute margin.

towards the right side – avoiding damage to the right pulmonary artery – cut the superior vena cava, then the right pulmonary veins, and finally cut the inferior vena cava if the thoracic contents have been left *in situ*. The heart is now free. The external landmarks are seen in figures 1.1 and 1.2.

Examination of the Heart

There is no standard method in opening the heart and slight variations are found in the various texts. The method to be described is slightly modified from that of Hudson (1965*a*) and has been found most useful by the author. A

Figure 1.3 Illustration of cuts 1, 2 and 3.

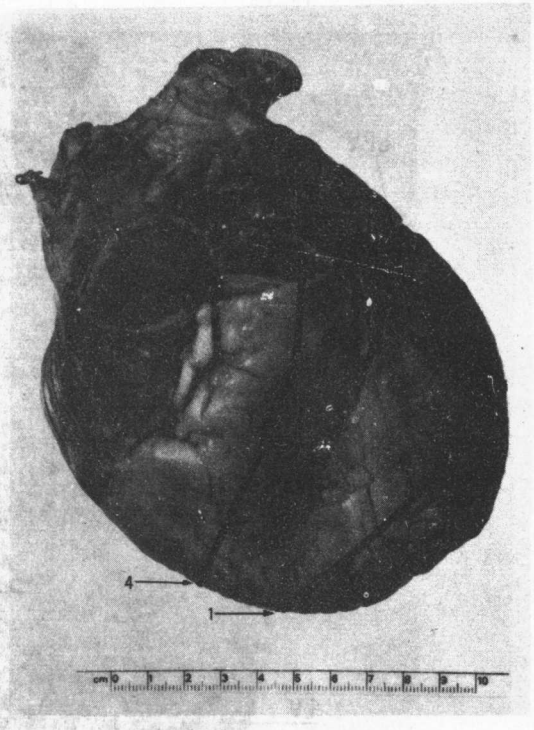

Figure 1.4 Illustration of cuts 1 and 4

few simple rules must be observed.

(1) Never cut before inspection.
(2) Do not handle the valve cusps.
(3) Never cut through valve commissures.
(4) Attempt to preserve abnormal features.
(5) Avoid cutting chordae tendineae.

The following cuts should be made:

Cut 1 (see figures 1.3 and 1.4)
Transect the ventricles 3–4 cm from the apex, parallel to the coronary sulcus. The size of the ventricular cavities can then be assessed and laminar infarction, if present, seen.

Examination of Coronary Arteries

Begin by inspecting the ostia, suitably incising the stump of the aorta. Inspect the coronary ostia, note their position, size and number (two orifices for left or right coronary arteries are occasionally encountered). Note any abnormality, for example thromboemboli. Do *not* use fine-angled scissors (coronary scissors) (Crawford, 1977*a*), but make multiple transverse incisions at 0.5 cm intervals at right angles to the course of the vessels. In cases of calcification, remove the affected areas for decalcification and examine later. Omit cutting the coronary arteries if injection with radio-opaque material is required (see p. 37). For the remaining dissection of the heart, follow the bloodstream.

Cut 2 – opening of the right atrium (see figure 1.3)
This is best done by scissors, starting about 0.5 cm away from the orifice of the inferior vena cava, cutting 0.5 cm above and parallel to the atrioventricular junction. Extend the cut to the tip of the right atrial appendage (inferior margin). This cut can be further extended at right angles towards the superior vena cava; avoid cutting through the orifice. The right atrial

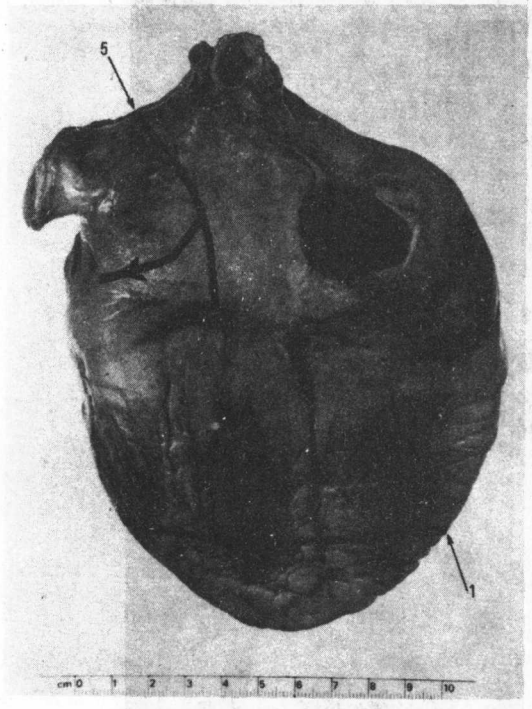

Figure 1.5 Illustration of cuts 1 and 5.

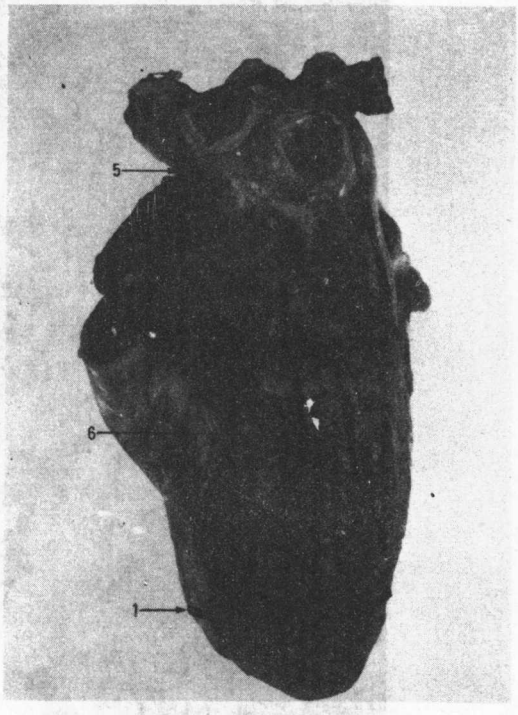

Figure 1.6 Illustration of cuts 1, 5 and 6

cavity is now displayed. Identify the following features (see figure 1.8):

(1) Limbus of the fossa ovalis.
(2) Fossa ovalis.
(3) Crista terminalis.
(4) The valve guarding the inferior vena cava (Eustachian valve).
(5) The valve guarding the coronary sinus (Thebesian valve).

Additional features are the opening of the Thebesian veins and Chiari's net. Inspect the atrial aspect of the tricuspid valve.

Cut 3

Using scissors, bistoury or a knife, cut through the tricuspid valve and the right margin of the right ventricle towards the apex, until the first cut (transverse cut of the ventricles) is met (figure 1.3).

Cut 4

Identify the anterior papillary muscle of the right ventricle and cut slightly to the right of this structure through the conus and pulmonary valve (avoiding the commissure) to the pulmonary trunk (figure 1.4). The right ventricle is now open (figure 1.8). Identify the following features:

(1) Tricuspid valve.
(2) Anterior and posterior papillary muscles.
(3) Papillary muscle of the conus.
(4) Crista supraventricularis.
(5) Pulmonary valve.

} (figure 1.9)

The opening of the left atrium should next be undertaken.

Cut 5

Using scissors, cut through the left atrial appendage near its base, extending the cut posteriorly across the roof of the left atrium, aiming to cut midway between the right and left pulmonary veins. At about the midpoint of the posterior surface of the left atrium, direct the scissors so

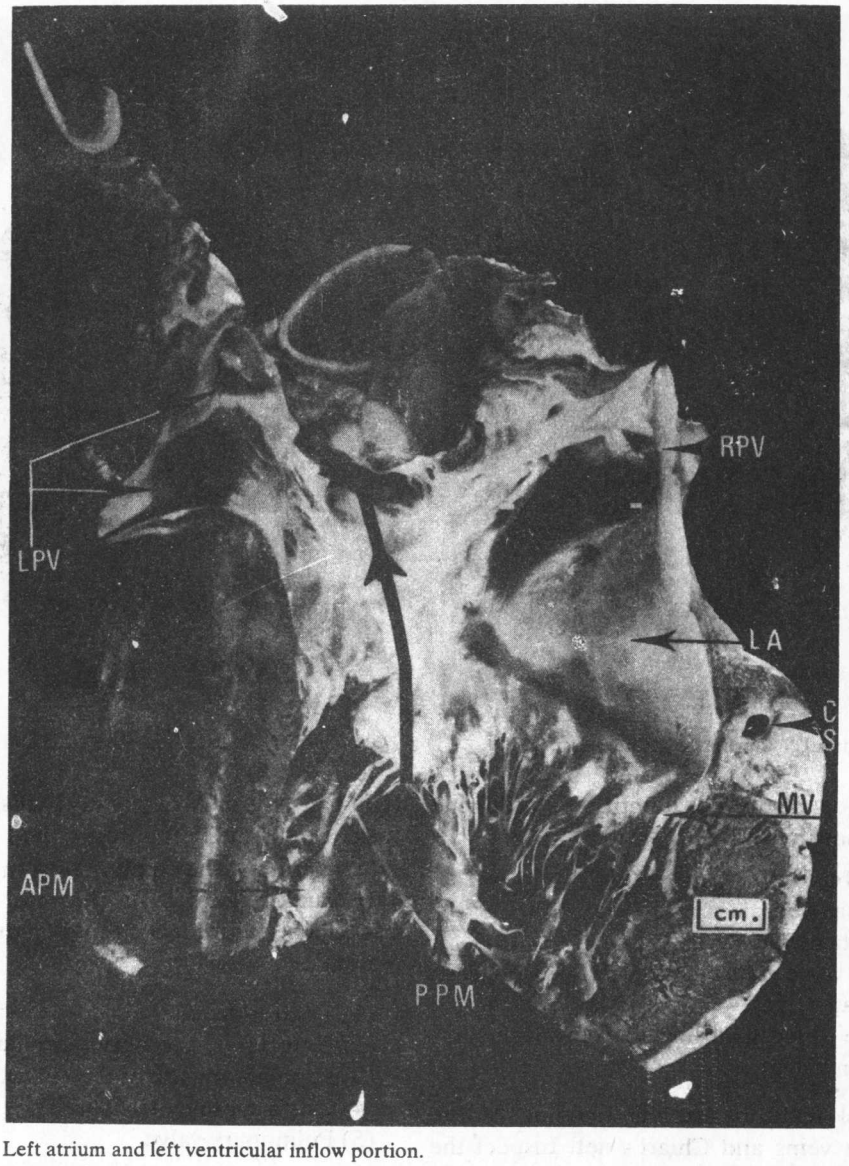

Figure 1.7(a) Left atrium and left ventricular inflow portion.
LPV, left pulmonary veins; APM, anterior papillary muscle; PPM, posterior papillary muscle. RPV, right pulmonary veins; LA, left atrium; CS, coronary sinus; MV, mitral valve (posterior leaflet).

that the cut is made slightly anterior or posterior to the left commissure of the mitral valve, as the atrioventricular junction is reached (figures 1.5 and 1.6). The following features should be identified:

(1) Orifice of the pulmonary veins.
(2) The region of the fossa ovalis.
(3) The mitral valve.

Cut 6

Cut along the obtuse margin of the left ventricle, extending Cut 5 to the region of Cut 1

Figure 1.7(b) The left ventricular outflow portion (leaving the mitral valve intact).
RCAO, right coronary artery ostium; PPM, posterior papillary muscle; APM, anterior papillary muscle; Ao, aorta; Av, aortic valve; MV, mitral valve (anterior leaflet).

(figure 1.6). This is best done with a knife or scissors. (Great care should be exercised not to damage the chordae tendineae.) Identify the following (figure 1.7a):

(1) Anterior and posterior mitral valve cusps.
(2) Anterior and posterior papillary muscles.

Cut 7

If no abnormalities of the anterior mitral valve cusp are seen, a cut through this structure can then be made (after measuring the valve ring) to display the aortic valve (figure 1.7a). Alternatively, if it is desirable to leave the anterior mitral valve cusp intact, cut slightly to the left