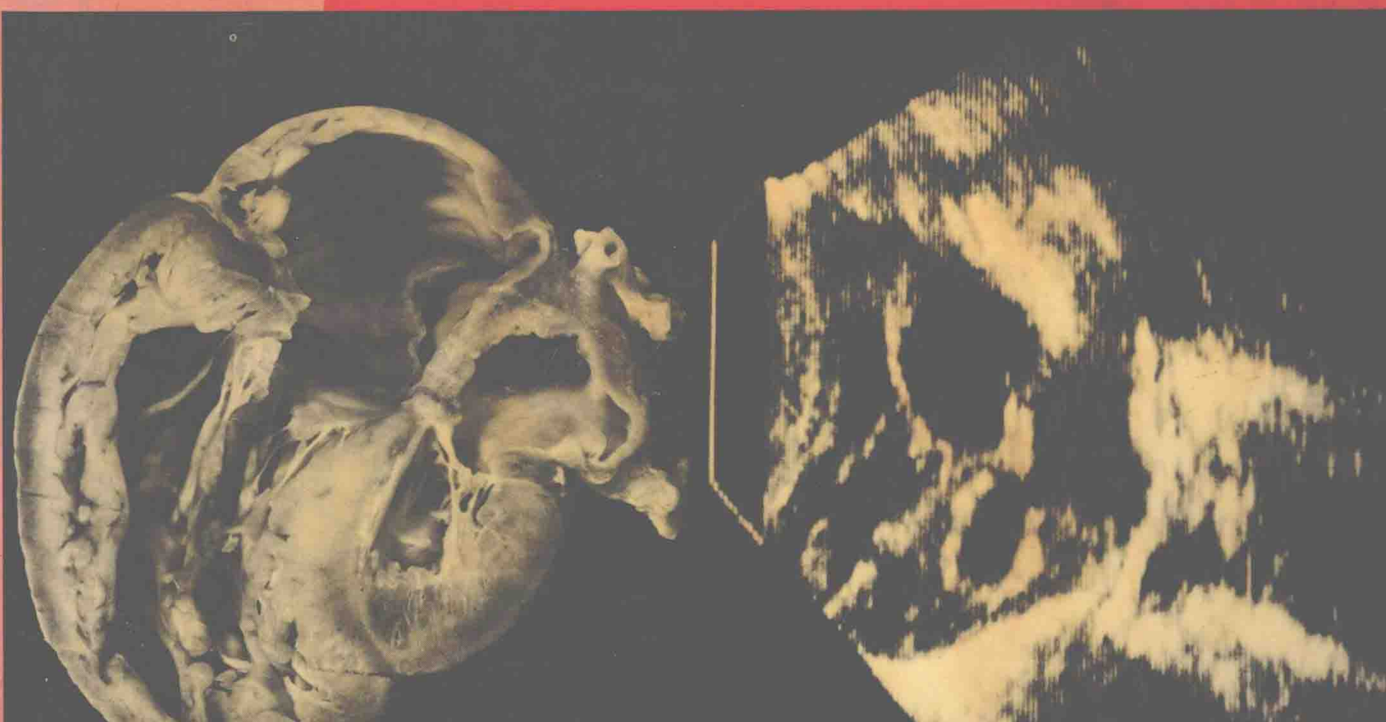
MODERN PAEDIATRIC CARDIOLOGY

# Congenital Heart Disease

## Morphologic Echocardiographic Correlations

Elma J. Gussenhoven / Anton E. Becker

Foreword by Dirk Durrer



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# CONGENITAL HEART DISEASE

*Morphologic Echocardiographic Correlations*

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# CONGENITAL HEART DISEASE

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# FOREWORD

'A bad foreword will considerably lengthen a bad book'. This remark from a French author makes one reluctant to accept an invitation to write some introductory notes. I took courage from the fact that this is an excellent book, as could be expected by those who are familiar with the scientific activities of both authors. Dr Gussenhoven and Professor Becker, experts in clinical echocardiography and cardiac pathology respectively, have co-operated closely for several years. However, this book is not the sum of their individual expertise: it has become a synthesis. It is evident that each author has learned much from the other. Several factors have influenced the ultimate result of their work, some of which will become apparent while reading this study, but other factors, which are less evident, deserve some comments. The authors were fortunate in having a great deal of material at their disposal, which was largely obtained in a short period of time, thanks to invaluable help from many sources. In particular six academic cardiology departments and paediatric cardiology departments referred echocardiographic recordings and interesting anatomic specimens to them. This help came about as a result of the invaluable assistance of the Interuniversity Cardiology Institute which gladly supported this study and I feel some remarks about the structure of this Institute are appropriate here.

In 1965 a law of interuniversity institutes made it possible to integrate the research activities of similar departments of cardiology into one institute. The Interuniversity Cardiology Institute is without walls—there is no central physical building. It only integrates the cardiology research activities. One of the major projects in echocardiography was undertaken in Rotterdam in 1974 because of the great expertise in this field there. As a result of the project several clinical studies were published and recently a study of normal echocardiographic values of healthy children and young adolescents from 4 to 17 years appeared*.

This book is a logical consequence of that initiative. Although the authors were certainly very tempted to describe all their data, they wisely restricted

* P J Voogd, H Rijsterborgh, G van Zwieten, J Lubsen Percentiles of echocardiographic dimensions in healthy children and young adolescents. In: Lancée C T (ed) Echocardiology. Martinus Nijhoff, The Hague, p 299–307.

this book to those congential vitia commonly encountered. The authors fully used the resources of the Institute so wholeheartedly given. However, for the outstanding merits of this present book they alone are responsible. The scientific board of the Interuniversity Cardiology Institute is proud of their achievement. Sometimes it is worthwhile to live in a small country, where daily contact is relatively easy.

1983 D.D.

# PREFACE

In the past decade much has been achieved regarding the understanding of congenital heart disease. The sequential analysis of the congenitally malformed heart has been widely accepted as a most useful method, particularly in cases of complex anomalies. The crux of this approach is the analysis of the cardiac junctions. It is here that echocardiography comes in to its own, since two-dimensional techniques demonstrate the actual connexions and valve morphologies at both atrioventricular and ventriculo-arterial junctions.

While utilizing the segmental approach in echocardiographic practice and while teaching it to our colleagues it became clear that no book existed which set out in clear and unambiguous fashion the steps employed. This is our attempt to fulfill this need.

We have on purpose restricted ourselves to the correlation of anatomy with echocardiography and, hence, have refrained from the inclusion of functional features. For the same reason we have not included contrast echographic studies. This, of course, is not to deny their significance in making a final and more complete diagnosis. We felt that the inclusion would have detracted from our prime message. Moreover, there are numerous textbooks on echocardiography, some excellent, that concentrate on these aspects.

Furthermore, we have abstained from the temptation to cover all congenital heart anomalies and have limited our descriptions to the more common malformations that occur within the heart. Anomalies, such as abnormal pulmonary venous drainage, have not been included. Likewise, the scope of the book does not permit a full description of the pathomorphology of congenital heart defects.

A volume like the present work could not have been accomplished without the help of many colleagues and friends. One of us (EJG) has had the opportunity to work for a considerable length of time with Professor V. H. de Villeneuve, Department of Paediatric Cardiology, Sophia Kinderziekenhuis, Rotterdam and with Professor J. Rohmer, Department of Paediatric Cardiology, University of Leiden. The opportunity to study their patients has been of great help in compiling the data to complete this book. Echographic tracings which were lacking from our collection have most generously been put at our disposal by F. J. ten Cate, Department of Cardiology, Erasmus University, Rotterdam; O. Daniëls, Department of Paediatric Cardiology, University of Nijmegen; W. van Lommel, Hervormd Diaconessenhuis, Arnhem; M. S. J. Naeff, Department of Paediatric Cardiology, Academic Medical Centre, Amsterdam; J. Roelandt and W. B. Vletter, Department of Cardiology, Erasmus University, Rotterdam and P. J. Voogd, Department of Cardiology, University of Leiden.

The majority of specimens used for the cross-sectional correlations have been collected from the Cardiovascular Registry of the Department of Pathology, Wilhelmina Gasthuis, University of Amsterdam. Supplementary specimens have most generously been provided by our friend and colleague Dr C. E. Essed, Department of Pathology, Erasmus University, Rotterdam and by Professor A. Oppenheimer-Dekker, Department of Anatomy and Embryology, University of Leiden.

We thank the numerous colleagues, pathologists, cardiologists and cardiac surgeons, who over the years have contributed to our experience by sending one of us (AEB) so many specimens of congenital heart disease for evaluation. They have served a most useful purpose. Over the years the help of Mr M. J. Klaver in preparing all these specimens has been invaluable.

The assistance of the Audiovisual Centre of the Erasmus University, Rotterdam, has been much appreciated, and in particular we are indebted to Cees de Vries and Bea Grashoff who prepared all the diagrams.

Professor R. H. Anderson has been kind enough to read the manuscript, correcting the English when necessary. We thank Dr F. J. ten Cate, for his most valuable contribution to the section on hypertrophic cardiomyopathy.

We are much indebted to the Interuniversity Institute of Cardiology for putting so many facilities at our disposal and we are particularly grateful to the Professors N. Bom, D. Durrer and P. G. Hugenholtz for their continuous support.

Finally, we would like to emphasize that completion of a book such as this is only possible when secretarial help is freely given and is nigh-on perfect. Such assistance was provided by Marsha Schenker, Corrie de Bruijn and Ria Willemstein.

1983

*Elma J. Gussenhoven*
*Anton E. Becker*

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# CONTENTS



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### Contrast technique

Injection of acoustic contrast agents into the bloodstream creates large quantities of small bubbles. These bubbles act as strong reflectors thus causing the blood to become highly 'visible' with echo techniques.

Contrast techniques can be performed in a routine laboratory with ambulatory patients. The method is particularly suited to detect or confirm abnormal bloodstream dynamics such as occur, for instance, in ventricular septal defect. In this situation the passage of contrast agent from the right ventricle through the defect into the left ventricle is conclusive evidence for the diagnosis of a right-to-left shunt.

## PRINCIPLES

### M-mode principles

For better understanding of the possible pitfalls and limitations of M-mode echocardiography it may be helpful to describe first the basic recording and display methods. In ultrasonic echo techniques, short sound pulses are transmitted at high repetition rate by a piëzo-electric transducer. Structures in the sound beam may cause reflections. Echoes are received by the same transducer and will be displayed as function of the travel time of the sound pulse. The M-mode principle is schematically illustrated in Figure 1.1. The echoes may be displayed in A-mode (Amplitude modulation) or B-mode (Brightness modulation). When B-mode information is projected on moving light-sensitive paper, the M-mode (Motion-mode) is obtained. The

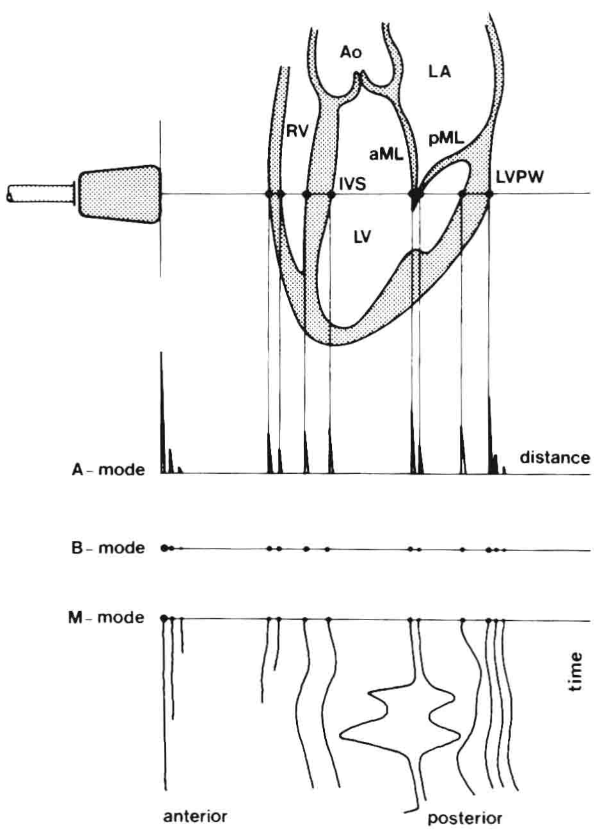


**Fig. 1.1** Echoes originate at the interface between tissues with different acoustic impedances. Echoes may be displayed in amplitude or brightness mode on the oscilloscope. Documentation of echo dots as function of time results in M-mode registration.

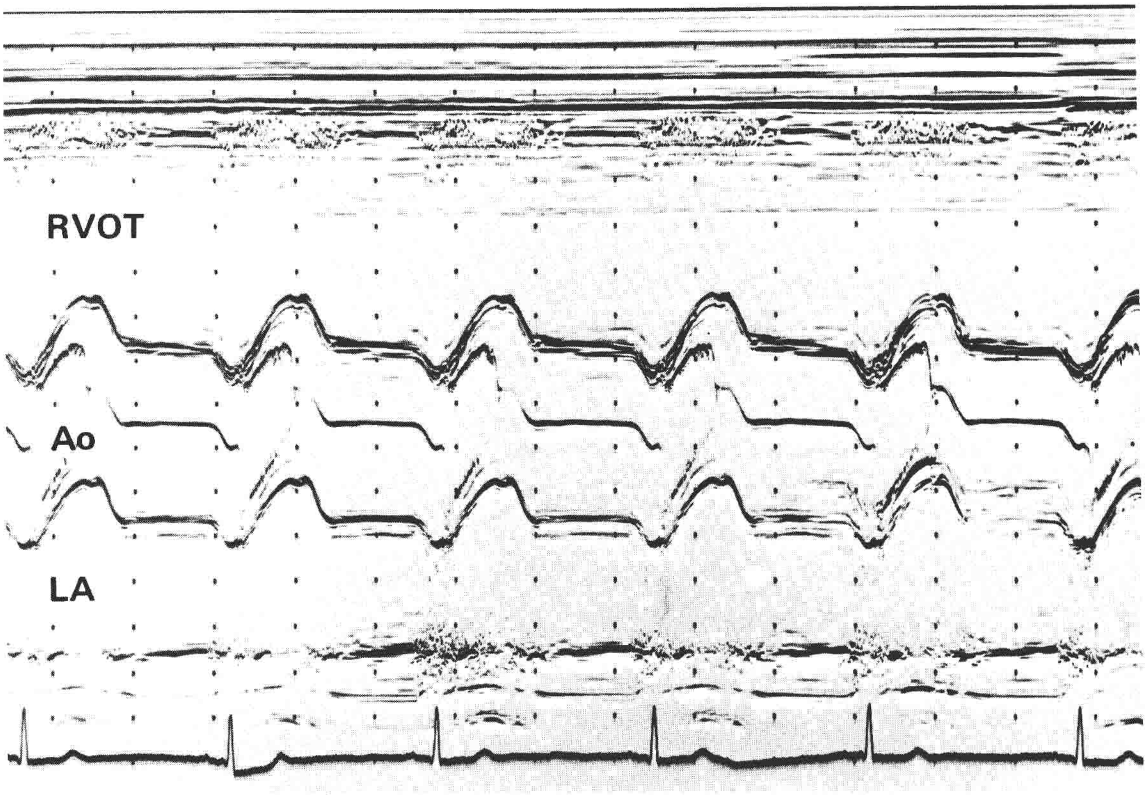


**Fig. 1.2** Example of an M-mode echocardiogram obtained with the sound beam aimed through the aortic region. RVOT = right ventricle outflow tract; Ao = aorta; LA = left atrium.

transducer is kept stationary and the recording only displays the motion of structures along the beam axis. In Figure 1.1 the transducer is positioned to study the mitral valve in the longitudinal cardiac cross-section. The M-mode yields the graphic representation of changes in range with respect to the transducer as fixed reference point.

The M-mode strip is usually displayed horizontally. When the transducer sound beam is aimed through the aortic region the sound beam passes through the right ventricle, the aorta, the aortic valve cusps and the left atrium. The corresponding recording is shown in Figure 1.2. If the beam direction of the transducer is not tilted these are the only cardiac structures documented. In order to obtain more information about cardiac structures and their interrelationships an M-scan is made. The M-scan is achieved by sweeping the sound beam slowly from the aorta towards the apex during the recording.

## Two-dimensional real-time principles

Presently a number of two-dimensional real-time systems is available. These may be subdivided into mechanical versus electronic beam steering or sectorial image versus rectangular image format systems. The principle of all these systems is that an acoustic beam scans the cross-sectional plane at high rate so as to yield instantaneous information about two-dimensional geometrical structures. The rapid scanning rate allows study of moving structures in their two-dimensional relationship (Kisslo et al, 1978). The

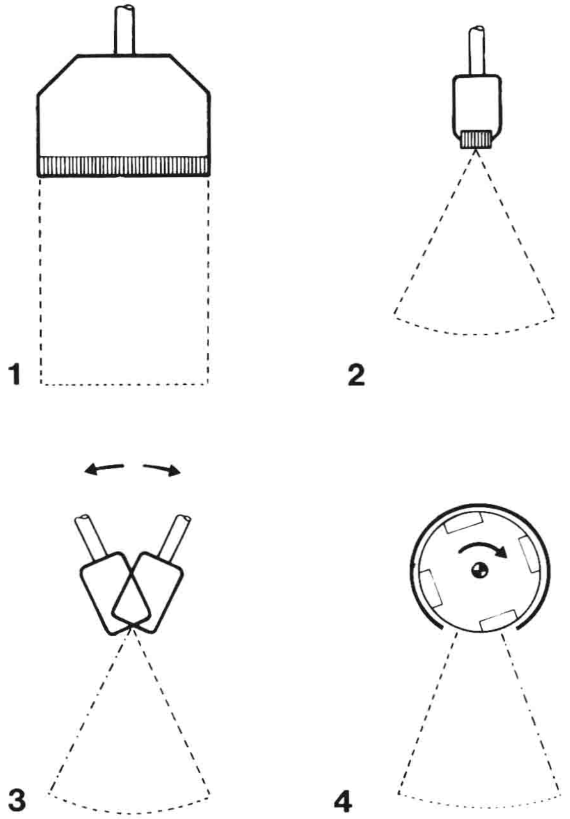


Fig. 1.3 Schematic diagram of commonly used two-dimensional real-time imaging systems: (1) linear array scanner, (2) phased array scanner, (3) mechanical sector scanner, (4) spinning wheel scanner.

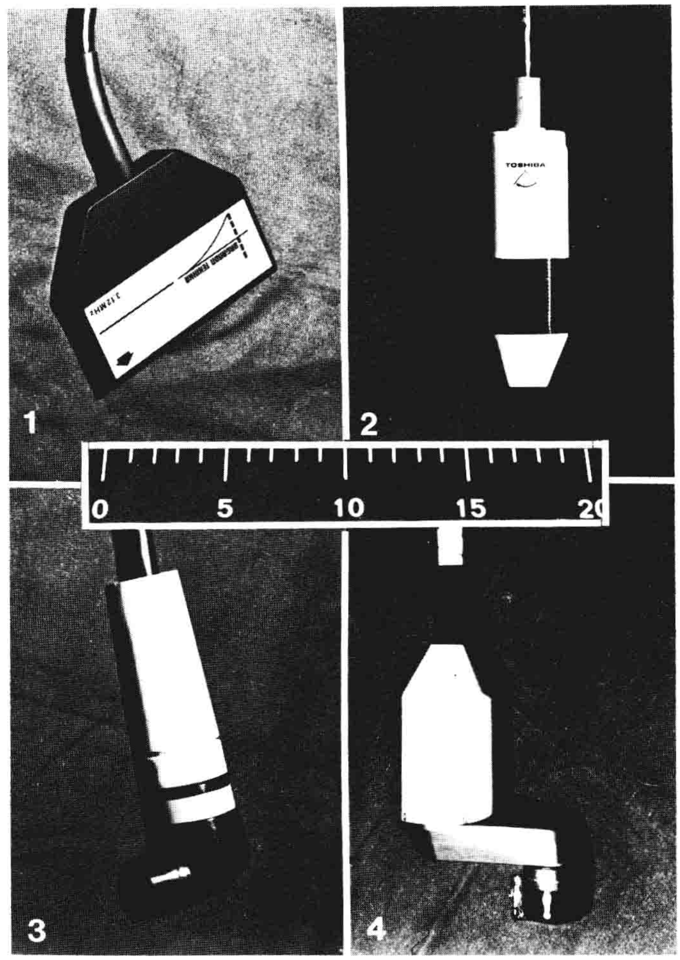


Fig. 1.4 Photograph of transducer assemblies from commercially available systems corresponding with the diagrams shown in Figure 1.3. Courtesy: (1) Organon Teknika, (2) Toshiba, (3) Smith-Kline and (4) Advanced Technology Laboratories.

four main principles of ultrasonic imaging systems are shown schematically in Figure 1.3.

Corresponding transducer assemblies of representative examples of each method as available in 1981 are illustrated in Figure 1.4. Photographs have been printed to the same scale. The various techniques include:

1. The linear array method where a number of elements have been positioned in a row. A subgroup of elements is used to form a sound beam. Adjacent and overlapping subgroups produce parallel sound beams. Rapid scanning results in high image rate. The characteristics of this system are a rectangular image format produced with no mechanical motion of the transducer head, but requiring a rather large acoustic window.

2. Phased array sector scanners also have a stationary transducer with multiple small elements in a row. All elements are activated at once to produce a single sound beam. Rapid sectorial beam steering is then introduced by electronic time delay processing. The transducer is relatively small. The characteristics are a sectorial image format again with no mechanical motion of the transducer head and now with excellent probe manoeuvrability.

3. The mechanical sector scanner where pivoting motion is applied to a single transducer. Commonly a small motor and the transducer are combined in one housing. As an alternative to mechanical pivoting systems, either vibration, magnetic deflection or rotation mechanism are used to cause sectorial deflection of the transducer. The characteristics are a sectorial image format, sometimes with slight mechanical vibration of the transducer assembly and the possibility of point entry through a small window.

4. The spinning wheel method. With three or four transducers on a rotating wheel, it is possible to create sectorial beam deflection at a high repetition rate. Each transducer is active when it passes an acoustic window in the housing. The characteristics are a sectorial image format with mechanically driven rotating transducers and possibility of point entry. The total transducer assembly is usually larger compared to other systems.

## Contrast principles

First applications of contrast injections were directed towards cardiac cavity identification. Microbubbles are formed when an acoustic contrast agent is push-injected into another fluid such as blood. Low pressure in the jet stream may be one of the microbubble forming mechanisms. In blood the bubbles persist for 15 to 30 seconds. This time is sufficient to allow the acoustic contrast to travel from the site of injection such as a peripheral vein in the

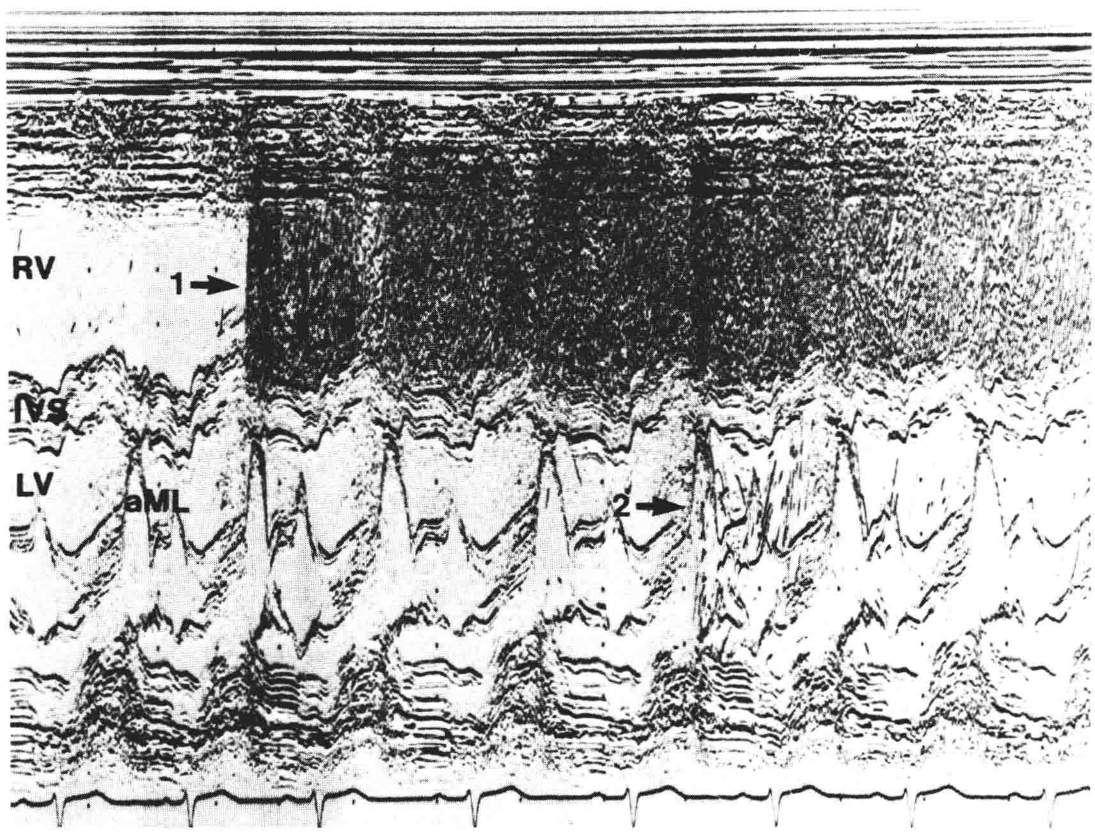


**Fig. 1.5** Contrast injection in a patient with secundum type atrial septal defect. Contrast appearance and timing are the important parameters in M-mode techniques. Contrast appears in the right ventricle (RV; see arrow 1) and four cardiac cycles later in the mitral valve orifice (see arrow 2). IVS = interventricular septum; LV = left ventricle; aML = anterior mitral valve leaflet.

right arm to the right ventricular cavity. However, most of the early studies with ultrasound contrast injections were carried out in the catheterization laboratory where the contrast fluid was directly injected into the cardiac cavity. Presently the procedure is carried out in the echo laboratory by injection of 2 to 10 ml contrast fluid into a suitable vein, for example the right basilar vein. When the blood is acoustically 'labelled' it can then be followed for a short period, allowing the detection of shunts or the recognition of complex cardiac malformations (Hunter, 1980).

With M-mode echocardiography, information on absence or appearance of contrast bubbles can be studied in the single sound beam area only. With M-mode contrast studies therefore it is necessary to aim the beam precisely through the areas in which contrast may be anticipated to appear. Contrast appearance, absence and timing are the available parameters. It is not possible to record or obtain direct information on the actual geometrical path of the contrast fluid since only velocity vectors along the sound beam are recorded.

In Figure 1.5 the appearance of contrast in the right ventricle is shown by the arrow. In this patient with a secundum type atrial septal defect, contrast appears in the left ventricle through the mitral orifice four cardiac beats after the appearance of contrast in the right ventricle (see arrow). It may be inferred that the contrast has reached the left ventricle via an atrial septal defect, but the septal defect has not unequivocally been demonstrated.

Use of two-dimensional echography circumvents this disadvantage. Here the entire cross-sectional cardiac plane is observed. From this information it is possible to follow the path of contrast through the chamber recorded in the echographic plane. With correct selection of the two-dimensional cardiac plane all cavities may be observed at once. This allows careful study of blood passage from one chamber to another, this being difficult if not impossible with M-mode techniques.

## LIMITATIONS

Increasing use of any ultrasound system will inevitably show its limitation. In this section we will describe a variety of limitations such as resolution problems, gain setting errors and aiming problems. These may occur in both M-mode and in two-dimensional techniques.

### Resolution limitations

The resolution of an ultrasound system can be defined as the system's ability to distinguish closely related independent structures. In general, resolution is divided into *axial* resolution and *lateral* resolution. The axial resolution is the power of the system to separate closely lying structures along the beam axis. The lateral resolution is the power of the system to separate closely lying structures in a plane perpendicular to the beam axis.

Axial resolution is determined by the length of the ultrasound pulse generated by the transducer. The length of the pulse should be short in order to obtain a good axial resolution. This can be achieved by choosing a high ultrasound frequency. However, effective penetration depth decreases with

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