TWO-DIMENSIONAL ECHOCARDIOGRAPHY IN CONGENITAL HEART DISEASE

A THESIS

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by

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C O N T E N T S

A C K N O W L E D G E M E N T S

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has been frequently delayed from his work as I examined his patients, and the continuing encouragement of Dr. R.A. Shanks is much appreciated. I am under greatest obligation to Dr. E.N. Coleman, physician in administrative charge of the Department of Cardiology, Royal Hospital for Sick Children, Glasgow, who first suggested that I should become interested in echocardiography as a clinical and research commitment. He encouraged me to persevere with the initial two-dimensional studies before the results suggested that they would be of real clinical value, and devoted a great deal of time to the discussion of the results and further useful clinical studies.

S U M M A R Y

This thesis will describe the author's early experience in the study of congenital heart disease using two-dimensional echocardiography, which was, at the start of the work, a new diagnostic technique.

A brief description will be given of the equipment, a real-time two-dimensional ultrasonic sector scanner developed by a team of electronic and mechanical engineers in the Department of Clinical Physics and Bio-engineering, Glasgow. The limitations of the original design will be outlined and the subsequent modifications which improved the image quality and were necessary to provide satisfactory studies from small infants will be described.

Application of the technique first required that the normal two-dimensional echocardiographic anatomy be established; techniques which were developed for this, scanning planes which were considered to give a full echocardiographic display of the anatomy of the heart, and the appearances in a group of normal infants and children will be described.

A technique of examination having been established, infants and children with the following types of complex congenital heart disease were studied: transposition of the great arteries, corrected transposition of the great arteries, double outlet right ventricle, persistent truncus arteriosus, tetralogy of Fallot, pulmonary atresia with and without a ventricular septal defect, aortic atresia, and total anomalous pulmonary venous drainage. For each condition the echocardiographic appearances will be presented, their significance discussed, and, where possible, diagnostic criteria will be described.

The findings in patients with atrioventricular canal

malformations will be considered separately; echocardiographic features considered to identify the partial atrioventricular canal and the type A and the type C complete atrioventricular canal with standard and contrast echocardiography will be reported.

The findings in the initial studies were applied in the study of a consecutive series of 64 cyanosed infants examined before cardiac catheterisation and angiocardiography; the results will be assessed and the diagnostic accuracy of two-dimensional echocardiography evaluated.

Finally, the results described in this thesis and by other workers will be applied to suggest an approach to the diagnosis of congenital heart disease with two-dimensional echocardiography based on the principles of sequential chamber localisation, and to outline the present place of two-dimensional echocardiography in the investigation of patients with congenital heart disease.

ABBREVIATIONS

ASD	atrial septal defect
AV	atrioventricular
CTGA	complete transposition of the great arteries
MPA	main pulmonary artery
PDA	patent ductus arteriosus
RVO	right ventricular outflow
TAPVD	total anomalous pulmonary venous drainage
VSD	ventricular septal defect

CHAPTER I

I N T R O D U C T I O N

Congenital malformations have come to assume an increasingly important role in paediatrics as improvements in social conditions and advances in medical care have resulted in a progressive fall in infant and child mortality. The incidence of individual malformations differs in different races, but in north west Europeans it is the central nervous and cardiovascular systems which are most often malformed to a degree which constitutes a danger to life (McKeown & Record, 1960).

The feasibility of surgical correction of congenital abnormalities of the great arteries was demonstrated by the successful ligation of a ductus arteriosus in 1938 (Gross & Hubbard, 1939) and by the resection of coarctation of the aorta in 1944 (Crafoord & Nylin, 1945) and 1945 (Gross & Hufnagel, 1945). In 1944 when Blalock and Taussig showed the favourable effect of palliation of the tetralogy of Fallot by the anastomosis of the subclavian artery to the pulmonary artery (Blalock & Taussig, 1945), the possibility of surgical treatment of other congenital cardiac anomalies became apparent. However until 27 years ago there was no corrective surgical treatment for the child with complex congenital heart disease and the exact diagnosis was therefore largely a matter of academic interest. In 1953 Gibbon pioneered the use of the heart lung machine with the successful closure of an ostium secundum atrial septal defect (Gibbon, 1954). This introduced a new era in cardiac surgery and indicated the practicability of the repair of complicated intracardiac anomalies. Since then rapid advances in cardiac surgery and extra-corporeal oxygenation and circulation have transformed the outlook for the patient with congenital heart disease. Open heart

surgery is now feasible even in the infant (Pierce et al., 1971) and there are few children with serious cardiac abnormalities who cannot benefit from some form of surgical therapy (Mustard et al., 1964; Gomes et al., 1971; Shams et al., 1971).

This surgery demands an accurate diagnosis with precise knowledge of the anatomical abnormality and assessment of the severity of the associated haemodynamic disturbances which can be obtained only by cardiac catheterisation and angiocardiography. Cardiac surgery, whether corrective or palliative, must therefore be preceded by cardiac catheterisation and angiocardiography, but to reduce the need for repeating this examination it should, if possible, be delayed until the time when surgical treatment is necessary. Furthermore, for optimal management of the child with congenital heart disease, the physician must have as much knowledge as possible of the nature of the defect to allow planning of the timing of the cardiac catheterisation and angiocardiography. Over the age of 12 months it is usually possible to make a reasonably accurate diagnosis of acyanotic lesions from clinical examination, radiology, and the electrocardiogram. In such patients cardiac catheterisation and angiocardiography need only be performed when an operation is thought necessary. However, the most serious forms of congenital heart disease present in infancy and particularly in the neonatal period. Richards et al. (1955) reported that of babies born alive with congenital heart disease 15% would die from cardiac causes within the first month and a further 12% between one and twelve months, making a total of 27% within the first year. McMahon et al. (1953) found a higher incidence of deaths and estimated that approximately 34%

were dead by the end of the first month and a total of 60% by the end of the first year. These figures were obtained before the development of modern cardiac surgery and indicate that early recognition, diagnosis and treatment should be an important factor in reducing the death rate from congenital heart disease.

In infants, particularly the newborn, it is seldom possible to reach an exact diagnosis with the simple non-invasive techniques, and in the ill baby the possible need for an emergency procedure for the support of the circulation therefore demands that urgent investigation be undertaken. Cardiac catheterisation in the newborn infant is a much more hazardous procedure than in older patients and carries a mortality of 6% (Braunwald, 1968) and a morbidity of approximately 8% (Rudolph, 1968). The purpose of the procedure is to provide an accurate anatomical diagnosis and it is almost always essential to combine cardiac catheterisation with angiocardiography and several different angiocardiographic injections are usually required. The risk to life from the two procedures is greater than from either alone, but angiocardiography causes the major hazard. Furthermore, the majority of infants subjected to this investigation are either in severe cardiac failure or cyanotic and acidotic and they therefore tolerate catheter manipulation and contrast injection poorly. The morbidity and mortality can be reduced by early detection and referral, and the rapid performance of the investigation with the absolute minimum of contrast injections to avoid the problems of improper catheter siting, excess osmolality, or sodium overload. To perform the procedure swiftly with a minimum of angiocardiograms it is therefore essential that the cardiologist has as much

knowledge as possible about the anomaly in advance of cardiac catheterisation and angiocardiography. Assessment of congenital heart disease prior to cardiac catheterisation is based on clinical examination combined with non-invasive investigations, the most useful of which would generally be recognised as being electrocardiography and radiology.

Clinical examination indicates the presence of cardiac failure and allows the distinction to be made between the two broad groups of congenital heart disease, cyanotic and acyanotic. Cardiomegaly can be detected by palpation, but this occurs in a wide variety of conditions and clinical examination does not always indicate which cardiac chamber is enlarged. Murmurs may be diagnostic in acyanotic heart disease, but in the cyanotic infant are frequently due to the presence of a ventricular septal defect, great artery stenosis, or patent ductus arteriosus which, although important, may be secondary to a more severe underlying condition. Splitting of the second heart sound is definite evidence of two outlet valves and therefore two patent great artery roots; however, in the ill child with tachypnoea it can be difficult to be certain about the presence or absence of splitting. Thus in the cyanotic infant it is often impossible to reach an accurate assessment of the underlying cardiac defect with clinical examination alone.

Radiological examination allows determination of the visceral situs and demonstration of the contour of the heart and great arteries and the effect of the anatomical abnormality on the pulmonary vasculature. It is the method of choice for determining the visceral and thus atrial situs; should there be any difficulty,

penetrated views showing the anatomy of the larger bronchi will determine the morphology of the lungs and thus by implication that of the atria (Van Mierop et al., 1970). The postero-anterior chest x-ray provides a quantitative assessment of the heart size. Radiological enlargement may be the result of pericardial effusion, myocardial disease or volume overload due to shunts, valvar regurgitation or high output states; the x-ray does not allow differentiation of the specific causes of enlargement. However serial measurements of heart size provide a useful index of the improvement, deterioration or stabilisation of cardiac function in these conditions. Postero-anterior, lateral and oblique views can indicate the size of the venae cavae, all four cardiac chambers and the great arteries. Contour evaluation is most accurate when the usual relationships between the chambers and arteries apply, but it is less informative in cases of complex congenital heart disease where there may be an absent chamber or a malpositioned or atretic artery or valve. In infants the shape in the postero-anterior view can be altered considerably by the presence of a large thymus; lateral views frequently show that its shadow lies in the upper anterior mediastinum but it may merge with and be indistinguishable from the shadow of the heart itself. The straight postero-anterior chest x-ray is essential for assessment of pulmonary vascularity. Where there is a significant left to right shunt this shows increased pulmonary blood flow and in the later stages the lack of peripheral vascular markings may indicate irreversible pulmonary hypertension. Reduced pulmonary vascular markings are found with decreased pulmonary blood flow but this does not give a precise indication of the level of the obstruction

or the size of the pulmonary artery. Pulmonary venous congestion may be seen in lesions associated with obstruction to the outflow of blood from the lungs or with lesions producing left ventricular failure. Assessment of pulmonary vascularity is essential in the diagnosis of congenital heart disease and the correlation of the appearance with the presence of or absence of hypoxaemia allows considerable reduction in the number of possible differential diagnoses.

The electrocardiographic pattern may be suggestive of the underlying condition in some anomalies such as uncomplicated mirror image dextrocardia, corrected transposition of the great arteries, tricuspid atresia, or endocardial cushion defects. However with each condition the pattern will vary considerably in the presence of associated anomalies such as a ventricular septal defect or valvar stenosis. The standard electrocardiogram can indicate the presence of chamber hypertrophy and the vector-cardiogram has been shown to provide a quantitative estimate of the magnitude of the myocardial loading process (Ellison & Restieaux, 1972). Each defect is usually associated with fairly characteristic secondary structural and physiological alterations within the heart which in turn are often reflected in the electrocardiogram; however in most cases a specific congenital cardiac defect does not produce a specific electrocardiographic abnormality.

Thus, in the infant with congenital heart disease, the history and clinical examination suggest a number of probable differential diagnoses. The simple non-invasive technique, electrocardiography and chest radiology may provide information which will help the

cardiologist reach a final likely diagnosis and assess the severity of the changes associated with this. All these techniques however, provide only presumptive information as to the internal cardiac anatomy; it is therefore difficult to reach a diagnosis without cardiac catheterisation and angiocardiography in patients with complex congenital heart disease, and in cyanotic disease virtually impossible. Although cardiac catheterisation and angiocardiography are the accepted techniques for determining the internal cardiac anatomy they may not always provide all the information required by the surgeon. Difficulty may be experienced in the case of complex congenital heart disease in demonstrating the ventricular or atrial septa or the atrioventricular valves. For example mistakes are frequently made with the differentiation of the different forms of the atrioventricular canal malformation (Rastelli et al., 1967). Many of these problems can be overcome by the use of axial views with cineangiocardiography as described by Bargeron et al. (1977). There is, however, a place for another technique for determining the internal cardiac anatomy which could be complimentary to angiocardiography in the investigation of some types of congenital heart disease.

In recent years, ultrasonic methods have been applied to the study of heart disease. These have the unique advantage of providing information about the intracardiac structure by a safe non-invasive technique. Various terms were initially used to describe the application of pulsed reflected ultrasound to cardiology and the term echocardiography was finally formulated as one which could be universally accepted. The development of

satisfactory techniques for ultrasonic imaging of the heart has been more difficult than for other organs because of problems in providing permanent records of the dynamic information obtained from the beating heart. The first technique to be developed, the M-mode system, allows a recording to be made over a period of time, but this is obtained from a single ultrasound beam and it can be difficult to ascertain the exact path of the sound beam and therefore to determine the relationship of structures. Systems were subsequently designed to obtain a stationary cross-sectional image over a number of cardiac cycles; this demands an absolutely stationary patient, is slow, and does not show the alteration in shape and relationship of structures throughout the cardiac cycle. More recently real-time two-dimensional echocardiography has been developed to provide a moving cross-sectional picture of the heart in motion.

This thesis will describe the application of one of these systems, a mechanical sector scanner, to the study of certain aspects of congenital heart disease. The two-dimensional scanning equipment will be briefly described and the limitations of the original design and the subsequent modifications which improved its diagnostic efficiency outlined. Application of the technique first required that the normal two-dimensional echocardiographic anatomy be established. The techniques which were developed for this, the scanning planes which were considered to give a full echocardiographic profile of the heart, and the appearances in a group of normal infants and children will be described. The findings in patients with different types of complex or cyanotic congenital heart disease will be presented

and the diagnostic features of each condition discussed. In patients with atrioventricular canal malformations the echocardiographic features considered to allow the elucidation of the different anatomical abnormalities will be reported, including the appearances using contrast echocardiography to determine the level of left to right shunting. The findings have been applied in the study of a consecutive series of cyanosed infants examined before cardiac catheterisation and angiocardiography; the results obtained will be described and, on their basis, the diagnostic accuracy of the technique will be assessed. Finally a segmental approach to the diagnosis of complex congenital heart disease with two-dimensional echocardiography will be suggested and the author's opinion on the place of the technique in paediatric cardiology outlined.

CHAPTER II

PRINCIPLES OF ECHOCARDIOGRAPHY

DISPLAY TECHNIQUES

By definition ultrasound is sound above the audible range, that is with a frequency greater than 18 kilohertz. In practice frequencies in the range of 1.5 to 10 megahertz (MHz) are used for medical diagnostic purposes. The frequency used depends upon the organ being studied. As a rule the deeper the surface to be explored the lower the frequency used since, for almost all biological material, absorption of ultrasound increases linearly with its frequency; thus the higher the frequency of the ultrasound the less its penetrating ability. However, the higher the frequency the better the resolution of the ultrasound beam, and the highest possible frequency able to penetrate the whole organ is therefore chosen. In cardiac ultrasound in adults a frequency of approximately 2.5 MHz is used, but in children it is possible to use higher frequencies, often up to 7 MHz for some small newborn infants. In contrast to audible sound, ultrasound is readily directed in a beam, more faithfully obeys the laws of reflection and refraction and is reflected by objects of very small size (Kossoff, 1966). Ultrasound travels through a homogenous medium in a straight line. When the beam meets an interface between two different media it undergoes reflection and refraction; the proportions reflected and refracted depend on the relative acoustic impedances of the media, the acoustic impedance of a substance being a measure of its resistance to movement imparted by the sound wave. The amount of sound reflected at an interface is proportional to the amount of the acoustic impedance difference between the structures and the angle of incidence of the beam as it strikes the interface;

the greater the impedance difference and the closer the angle of incidence to the perpendicular, the greater is the amount of sound reflected. Ultrasonic examination utilises piezoelectric transducers; these are made from a substance (quartz, barium titanate or lead zirconate) which changes shape under the influence of an electric field. As a crystal of the substance expands and contracts it produces compressions and refractions, or sound waves, in the air. The reverse is also true for when the crystal is struck by a sound wave it produces an electrical impulse.

The equipment used to produce ultrasonic images consists of a transducer, transmitter, receiver, signal processor and cathode-ray tube. The transducer, which is usually in contact with the patient, essentially consists of a piezoelectric crystal which both sends out the ultrasound and receives the returning echoes. The transmitter intermittently feeds electrical energy into the crystal which thus sends out short bursts of ultrasound waves. Following this the transducer becomes a receiver and records any reflected ultrasound waves. After a relatively longer period of time another burst of ultrasound is emitted and the cycle repeated. This is repeated at a rate of 1,000 to 2,000 pulses per second in most commercial equipment designed for echocardiography. The transducer converts the returning echoes to electrical impulses which are fed into the receiver and signal processer and displayed on the cathode ray tube. If the velocity of sound through the medium and the time from transmission to return of an echo is known, the distance to the reflecting surface can be calculated. The ultrasound equipment is calibrated for the

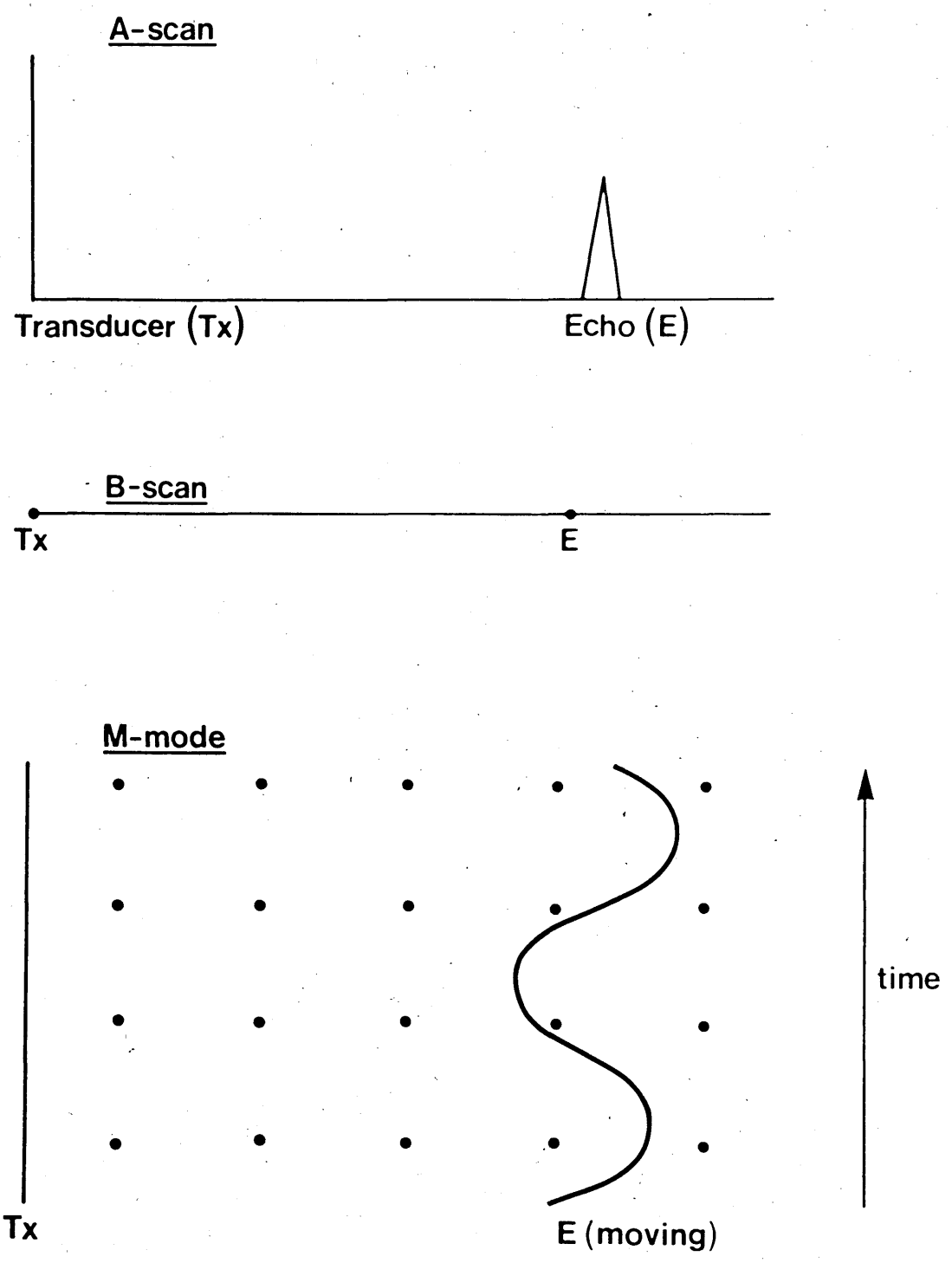


Figure 2, 1 Diagramatic representation of the basic methods of displaying the ultrasound echoes. The returning echo is displayed as a vertical displacement from the horizontal axis in the A-scan, as a spot on this axis in the B-scan, and as a moving line on the M-mode scan.

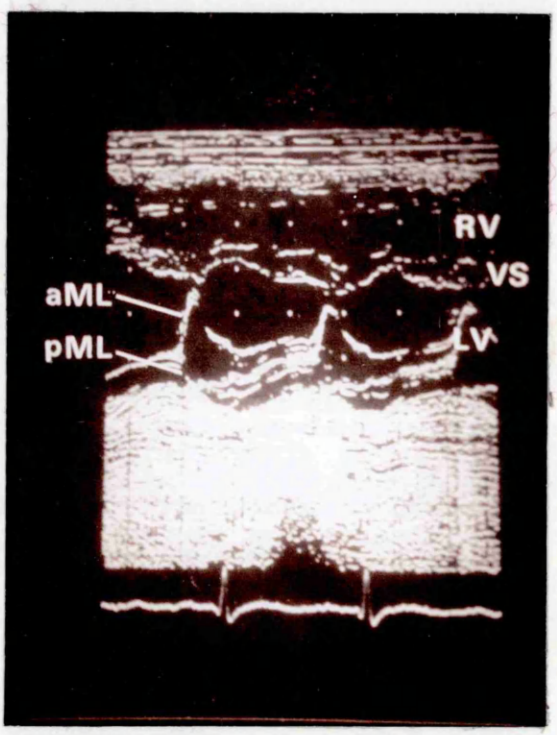


Figure 2, 2 M-mode recording at the level of the mitral valve from a 2 year old child. RV, right ventricle; VS, ventricular septum; LV, left ventricle; aML, anterior mitral valve leaflet; pML, posterior mitral valve leaflet.

velocity of sound in the medium being examined (a mean value of 1540 metres per second for soft tissue) and converts the time for the echo to return to distance from the transducer. The returning signal as displayed on the cathode ray tube indicates the distance of the reflecting interface from the transducer. Four methods of displaying the returning echoes are used for echocardiography; the A-scan, the M-mode scan, the compound B-scan and the real time two-dimensional display.

II, 1:1 A-scan (A = amplitude modulated)

The A-scan displays returning echoes as vertical displacements from a horizontal axis and the height of the returning echo represents its strength (Figure 2, 1); if the structure is moving this echo can be seen to move along the horizontal axis. With this system it is difficult to obtain permanent records and the exact nature of many of the echo producing structures is difficult to ascertain.

II, 1:2 M-mode (M = motion)

The M-mode system is a development of the A-scan and records the motion of the echoes from a moving surface over a period of time. The returning echoes are first displayed as spots along the horizontal axis, the intensity of the spots being related to the intensity of the returning echo; this is known as the B-scan (B = brightness modulated). In the M-mode record the horizontal axis becomes a time base and the movement of the echoes with time are displayed by slowly sweeping the time base across the oscilloscope screen (Figure 2, 1). Figure 2, 2 is an M-mode recording through the left ventricle and mitral valve of a two year old child; this allows the distance between different interfaces and

the movement of intracardiac structures to be measured. M-mode echocardiography thus allows the exact measurement of the rate of movement of echo producing structures and the distances between different ones lying along the axis of the ultrasound beam. The determination of the relationships of structures not lying along a single beam is dependent on the operator's assessment of small changes he makes in transducer angulation; the technique is difficult, particularly when it is applied to the rapidly beating heart of a restless infant. Structures are recognised by their characteristic pattern of motion and those with little intrinsic motion are almost impossible to identify with certainty. Furthermore, tissue which lies in a plane approximately parallel to the ultrasound beam cannot be recognised and the atrial septum and main pulmonary artery are therefore difficult to study.

II, 1:3 ECG triggered B-scan (B = brightness modulated)

To improve the information available from diagnostic ultrasound, systems producing stationary two-dimensional or cross-sectional images have been developed for studying the abdominal organs. In this technique, known as compound B-scanning, the images consist of numerous B-scan lines; the probe is moved by hand and as each B-scan line of information is obtained a sensor in the equipment calculates its position in two dimensions and writes it in the appropriate position on a storage oscilloscope. The images are built up over a period of seconds and the technique is thus unsuitable for examining the rapidly beating heart. If conventional B-scanning equipment is fitted with an electrocardiographic module each B-scan line can

be triggered to a chosen time in the cardiac cycle and a stationary image can thus be produced over a number of heart cycles (King, 1972). This technique is time consuming, requires the patient to lie completely still while the image is built up over a considerable period of time, and does not produce clear images in the presence of an arrhythmia. Even sinus arrhythmia will result in distorted diastolic images.

II, 1:4 Real Time Two-dimensional Display

Real time two-dimensional scanning systems are modifications of the compound B-scanning technique. They provide cross-sectional images of the heart in motion, that is, almost in real time, by rapidly sweeping a single or multiple ultrasound beams through a plane, either by electronic or mechanical means. The results obtained in clinical cardiology have been reported with three of these systems; the multiple element system, the phased array system and the mechanical sector scanner.

The first to be described was the multiple element system developed by Bom et al. (1973). The prototype transducer consisted of 20 small piezoelectric crystals in a linear array 80 mm long. Each of these elements was excited in sequence and an appropriate display of the returning echoes produced, in a rectangular format, a continuous two-dimensional image of the tissue underlying the transducers. The system as initially described generated 160 frames per second, each frame consisting of 20 ultrasound lines. The value of such equipment in the study of children was limited by the need to utilize small transducers which lack the sensitivity and resolution provided by the larger ones used for B-scanning or M-mode echocardiography. The use of

20 small transducers produced a line density of slightly less than three per centimetre. The length of the scanning head posed problems due to difficulty in maintaining ultrasonic contact with an irregular chest wall or interference from underlying calcified rib or sternum or aereated lung. Furthermore although the scanning plane could be tilted in a direction perpendicular to the original plane, the plane itself could not be tilted along its axis to visualise areas lying under bone or lung. More recent developments with the use of increased numbers of smaller elements and line interpolation have improved the line density and reduced the size of the transducer, and the use of dynamic focusing has improved the resolution in the plane of the transducer in similar systems which are now being developed commercially. In the early device the out of plane resolution was poor and the information was obtained from a relatively thick "slice" of tissue; this has been improved by the provision of a degree of fixed focusing in the plane perpendicular to the scanning plane by curvature of the elements or the use of acoustic lenses. An M-mode record can be obtained from any individual transducer element, either from the moving display or by stopping it and selecting the element from which the record has to be taken.

The phased array system, a multiple crystal system with improved line density and resolution within the scanning plane, was described by Von Ramm and Thurstone (1976). The equipment as described consisted of a linear array of 16 ultrasound transducer elements. The delays between pulsing successive transducer elements are small, being in the order of one microsecond

compared to 250 microsecond intervals between pulses in the other multiple crystal system. The energy from the individual elements thus combines to form a single ultrasound beam the direction of which can be controlled by varying the delay between the pulsing of the individual elements. It is therefore possible to produce lines in any direction and in the system described these were displayed in the format of a 60^{o} sector. To produce this image the pattern of time delays between successive elements requires complex electronics involving the use of a computer and makes the system very expensive. For a maximal range of 15 cm the system Von Ramm & Thurstone described produced images of 128 individual B-scan lines at a frame rate of 30 per second although faster or slower scanning rates, with fewer or more lines respectively, could be obtained. With this system the beam can be focused to produce high resolution images at any chosen depth in the scanning plane. In the prototype the out of plane resolution was poor but this has been improved with further electronic modifications in more recent commercial systems based on this principle. An important limitation in paediatric cardiology of the prototype of Von Ramm & Thurstone was that artefact produced by the array obliterated the first three or four centimeters of the final image but this has also been improved in commercial equipment and is not likely to pose a significant problem. With the steered beam system an M-mode recording can be obtained from any individual line simultaneously with the two-dimensional display.

Another approach to the problem, allowing the use of a conventional high sensitivity narrow beam transducer, was to rapidly oscillate such a transducer through an arc of a circle.

Several scanners based on this principle have been produced, all using an electric motor to produce oscillatory movement of the transducer. Different systems for indicating the transducer angle are electronically coupled to standard or modified ultrasound equipment and each B-scan line of information is displayed in the appropriate position on an oscilloscope, thus producing the image in the format of a sector of a circle. The display in any one system is made up of a sector of a given angle filled with lines which may be variable in number and spaced by varying amounts. These are interdependent and are related to the rate at which the transducer oscillates and the transmitter is pulsed, and the maximum depth of tissue to be examined. The higher the transmission frequency the greater the number of B-scan lines obtained per second, but the deeper the structures being studied the longer the time interval from transmitting to receiving the echo and thus the fewer the number of lines which can be produced. For a given transmitter frequency and selected depth of scan a maximum number of lines can be produced. However, at the centre of the sector the oscillating transducer travels more rapidly than at the edges, and if the display is to consist of evenly spaced lines the final number of lines per second will by necessity be below this theoretical maximum. The faster the transducer oscillates and the larger the angle of scan the less the line density of the display. Thus in any sector scanner display the final image is characterised by the angle of sector scanned, the rate of scanning, the number of lines displayed, and the spacing between the lines. The results obtained in cardiac imaging with several different types of mechanical sector scanners have been

reported. In the system described by Griffith and Henry (1975) the transmitter was pulsed at 2 KHz and scanned a 30° angle to produce 30 frames of 66 lines per second. The scanner is hand held on the chest wall allowing it to be easily rotated or tilted but the oscillating transducer is in contact with the skin and may cause discomfort. The small scanning angle (30°) produces an image of high line density but a limited view of the heart, which may cause difficulty in determining the relationship of different structures. By using a transmitter frequency of 3.6 KHz Nishimura et al. (1976) were able to generate 30 to 36 frames per second, the effective image being a 65° sector composed of 100 to 120 lines. Acoustic coupling cannot be maintained throughout this large scanning angle and in clinical use the transducer has to be immersed in an oil bath placed on the surface of the chest. This technique makes an examination more complicated and time consuming and causes difficulty in the manipulation of the transducer.

Another form of mechanical two-dimensional scanner uses more than one (usually four) different transducers arranged in a circular fashion like the spokes of a wheel. The whole transducer assembly is made to rotate and each individual transducer in turn emits and receives ultrasound pulses, but only while passing through a chosen arc of the circle This rotating mechanical sector scanner can produce a faster scanning rate and larger scanning angle than an oscillating one. Some form of a permanent cover containing acoustic coupling material has to be incorporated within the scanning head to permit it to be placed on the skin without the rotating transducers causing discomfort. Commercial sector

scanners based on this principle are now available. As with the oscillating mechanical scanner a high quality M-mode echocardiagram cannot be obtained simultaneously with the two-dimensional record.

THE GLASGOW SECTOR SCANNER

The mechanical sector scanner used for the work described in this thesis was produced by a development team from the Department of Clinical Physics and Bio-engineering, Glasgow. The basic principle was first reported by McDicken et al. (1974) and the initial design described by Shaw et al. (1976) with subsequent modifications detailed by Houston et al. (1977b). Full technical details of the system are described in the publications by Shaw et al. (1976) and Houston et al. (1977b) enclosed in this thesis. The scanning system consists of two basic parts, the scanning head and electronic interface unit, and was specially designed to connect directly to a commercially available ultrasound machine, the Nuclear Enterprises Diasonograph.* The scanning head is essentially an electric motor which oscillates a single ultrasound probe through a 60^{o} arc at a variable rate up to 12.5 cycles per second. To maintain continuous acoustic coupling between the probe and the skin through a 60^{o} arc a constant volume oil filled cell with a front diameter of 30 mm is interposed in the 3 mm gap between the face of the probe and the front cover of the instrument. The oil filled cell thus separates the patient from the movement of the transducer and avoids discomfort. The principal function of the electronic interface unit, which couples the scanner to the display, is to control the sector scan display, making it follow the scanning action of the probe. In the initial design the scanner was fitted with a 2.5 MHz transducer, the display consisted of 32 lines of

* Diasonograph NE 4102 or 4200, Nuclear Enterprises Limited.

information for each cycle and there was no indication of the time in the cardiac cycle. It became apparent that this design did not provide satisfactory echocardiograms from small children and four modifications were therefore undertaken. Initially the scanning head was modified in order that different transducers could be used and four different ones produced for work with adults, children and infants; the specifications of these are 2.5 MHz 15 mm diameter, 2.5 MHz 12.5 mm diameter, 3.5 MHz 15 mm diameter and 5 MHz 6 mm diameter focused for optimum performance in the range 8 to 12 cm, 5 to 10 cm, 3 to 7 cm and 2 to 4 cm respectively. Secondly the display was improved to consist of 128 equally spaced lines of information in each complete oscillation of the transducer; at maximum speed of 12.5 cycles per second 25 frames of 64 lines are generated per second. Thirdly the display was modified to allow the scale of the images on the oscilloscope to be altered; five different scales are now available from one third to five thirds life size. Fourthly electrocardiographic information was included in the telerecording by means of a light flashing in time with the R wave of the electrocardiogram and by a digital indication of time elapsed after the electrocardiographic pulse.

A diagramatic representation of the equipment is shown in Figure 2, 3 and a photograph of it in use in Figure 2, 4. The moving echocardiogram is normally viewed from the Diasonograph oscilloscope through a television camera and monitor system and recorded on a video recorder controlled with a foot operated switch. Information is frequently more easily appreciated from the moving echocardiogram or by observing a series of individual

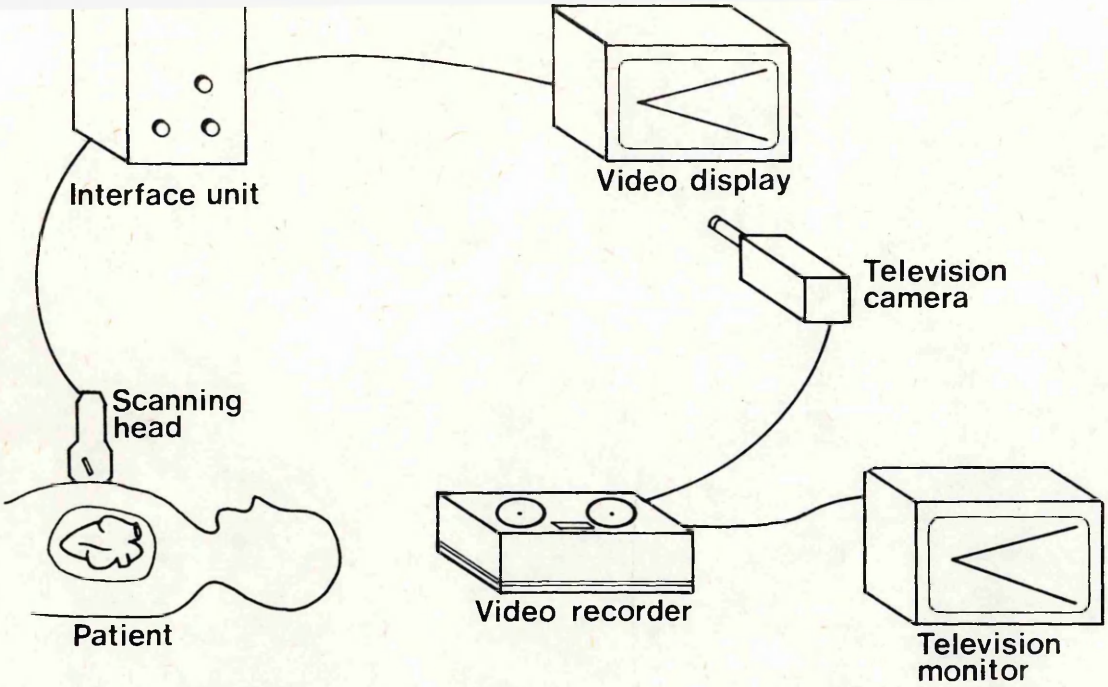


Figure 2, 3 Diagramatic representation of the scanning equipment. The ultrasound signals returning from the ultrasonic transducer are processed in the interface unit and displayed on the oscilloscope of the ultrasound machine. The moving image can be viewed either directly on the oscilloscope or, by using a television camera, on a TV monitor and recorded on videotape.

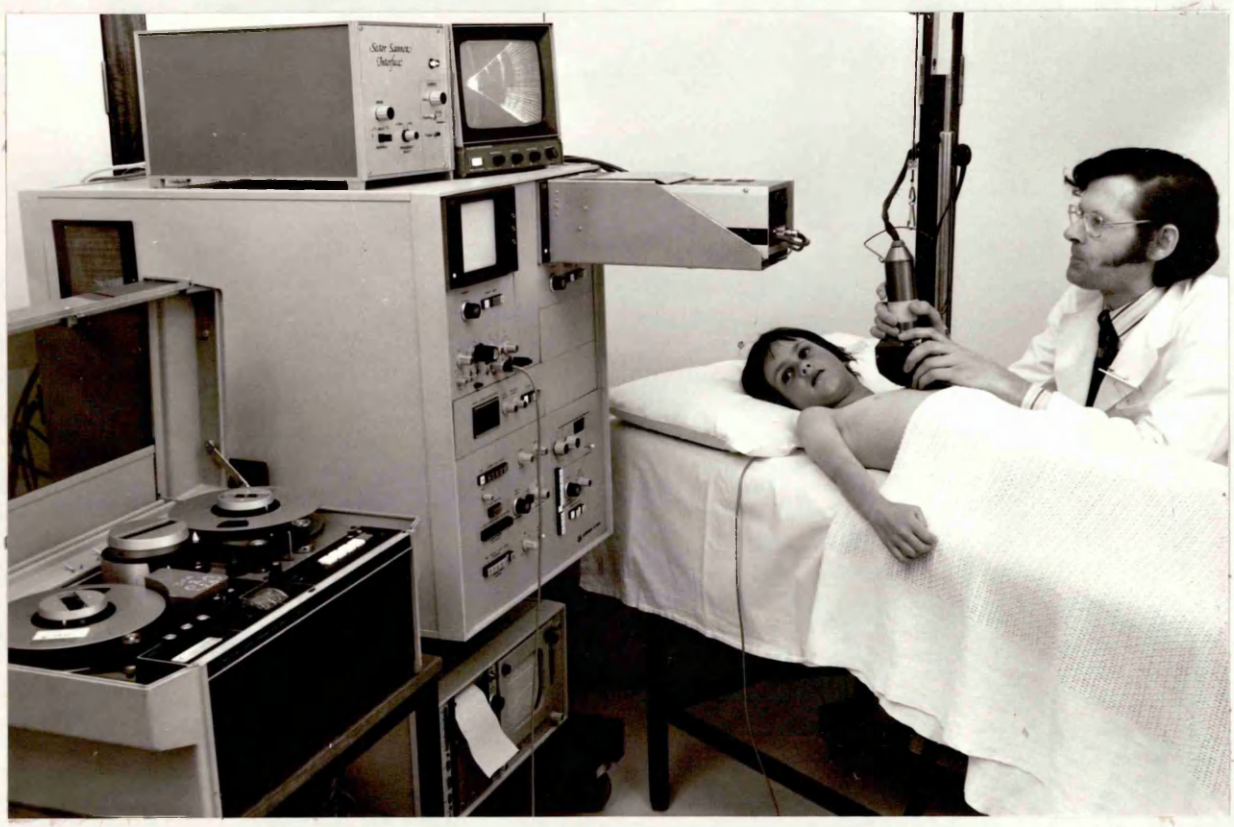


Figure 2, 4 The scanning equipment in use. The scanning head is hand held; the Diasonograph 4200 is in the centre of the photograph with the TV camera protruding from the front face, and the interface unit and TV monitor on top; the video tape-recorder is on the left.

frames from the record, but still photographs are required as a permanent record of the appearances and have been used to illustrate the findings in this thesis. Still frame pictures can be obtained from the videotape record by stopping the recorder on the chosen frame and photographing the picture on the television monitor with Polaroid film. An alternative viewing system, which provides a static view of the heart and permits immediate photography, is provided by electrocardiograph controlled gating of the storage monitor. The display may be re-written once in each heart cycle, at any point in the cycle, and when an appropriate view has been selected it can be stored for inspection or photography. In normal use the patient lies supine and the weight of the scanner is supported by a spring tension cord device which hangs from a simple track above him. This system permits free tilting and rotation of the scanner to any normally required position. This and the use of a foot operated switch on the tape recorder allows the examination to be carried out by a single operator.

The different real-time two-dimensional scanning systems are still in the developmental stages and their full potential is as yet unknown. No comparative study of the value of the different types of real-time systems has yet been reported. While each will have a place in ultrasonic examination, those which have a relatively small contact area and are readily rotated and tilted may be more appropriate to the study of the heart. The author's experience has principally been the use of the mechanical 60° sector scanner in studying the heart of infants and children and a detailed discussion of the relative merits of the different systems is therefore outwith the scope of this thesis.

CHAPTER III

THE STANDARD TWO-DIMENSIONAL ECHOCARDIOGRAM

III, 1 METHODS

III, 1:1 The Standard Views

The feasibility of any medical ultrasonic examination is determined by the characteristics of the tissue between the skin on which the transducer is placed and the organ being studied. The absorption of ultrasound by bone and air is considerably higher than that of soft tissue or blood, and the presence of bone or aerated lung between the transducer and the heart effectively screens it from the ultrasound beam. This limits echocardiographic study in adults to the second to fifth left interspace within three or four centimetres of the left sternal edge. In young children the costal cartilage, ribs, and even the sternum may not be calcified, and the main obstacle to ultrasonic examination in these cases is the underlying lung. Satisfactory echocardiograms from infants and young children can often be obtained from any left parasternal position from approximately the second left interspace downwards, laterally towards the mid-clavicular line and medially often over the sternum. This area may be considerably reduced by the intervention of lung in the tachypnoeic infant with cardiac or respiratory disease. Within these limitations to transducer placement an almost unlimited number of cross-sectional echocardiograms can be obtained. To allow comparison between different individuals it is therefore essential that standard echocardiographic views are used. Because of variability in the chest wall shape and in the axis and position of the heart these views cannot be based on standard thoracic cage sites, but rather on approximately standard planes through the heart which are obtained by altering the position and angulation of the scanner

in such a way as to make allowances for differences in cardiac position and chest configuration. If a satisfactory view is not obtained in the original position the scanning head is moved to an adjacent one and in each position it is rotated and tilted to obtain an optimal view.

Scanning planes are chosen such that a minimum number show all the major cardiac structures, and that when structures are in apposition a single plane will demonstrate them simultaneously to permit accurate assessment of their relationships. Initial experience indicated that these requirements could be met by four standard planes of scan; each provides a landmark from which the scanner can be tilted and if necessary rotated to show the entire heart.

These scanning planes have been entitled the following:

Longitudinal view

Low transverse view

High transverse view

Oblique or four chamber view

In this chapter the procedure for obtaining the standard views and the typical findings will be described.

III, 1:2 Techniques of Contrast Echocardiography

For each of the standard views the identity of the cardiac chambers (or great artery) was confirmed by the injection of ultrasonic contrast medium at the time of cardiac catheterisation. This technique, known as contrast echocardiography, was first described by Gramiak et al. (1969) who noted that following the rapid injection of indocyanine green multiple echoes appeared and

could be visualised by using suitable ultrasonic gain settings. He also found that the use of blood, saline or dextrose produced a similar appearance allowing multiple injections to be performed without hazard to the patient. The rapid hand injection of 2 ml of blood or saline through the cardiac catheter was therefore used for these studies. In each case the position of the catheter was first determined by the usual techniques of fluoroscopy, pressure recording and oximetry. In all studies the information is for the most part more readily visualised on the moving display than from still frames. This is particularly apparent following contrast injections where there are fewer frames from which to choose the best illustration. Furthermore, although the technique is described as a real-time study, the ultrasound lines of information are not, in fact, recorded simultaneously. All the illustrations of the contrast studies have been obtained following a single upward or downward sweep of the scanner in order to minimise the differences in time between upper and lower portions of the scan (not more than 40 milliseconds).

III, 1:3 Study of Normal Subjects

After the standard scanning planes had been developed and experience gained in obtaining the required views, a study of 40 normal subjects was undertaken. Each subject was considered to have a normal heart on the basis of history and clinical examination. The 40 infants and children were divided as follows, on the basis of age, into four groups of 10 designated A, B, C and D.

Group A Newborn infants.

Age range 20 hours to 23 days (mean 7 days).

Weight range 2.4 kg to 3.7 kg (mean 3.2 kg).

Group B Infants aged 1 to 12 months.

Age range 5 weeks to 11 months (mean 5 months).

Weight range 3.1 kg to 9.8 kg (mean 6.9 kg).

Group C Children aged 1 to 6 years.

Age range 19 months to 5 years 11 months (mean 3 years 6 months).

Weight range 11.0 kg to 23.8 kg (mean 15.0 kg).

Group D Children over 6 years of age.

Age range 6 years 1 month to 13 years (mean 9 years).

Weight range 19.8 kg to 36.7 kg (mean 27.1 kg).

The examinations were performed with the intention of determining how easily structures could be visualised in a routine investigation and the examination was therefore performed as rapidly as possible, usually in less than 5 minutes. The 5.0 MHz transducer was used for newborn infants and the 3.5 MHz transducer for older infants and children. If a structure was not definitely recognised on review of the videotape recording the examination was not repeated. It is possible that the use of a different transducer (particularly the 5.0 MHz one for some infants between 1 and 12 months) or repeat examination might have increased the recognition rate for certain structures but it was considered that, for the purposes of the study, a single examination would be performed and not repeated.

The limitations of an ultrasonic examination are such that failure to recognise a structure does not mean it is absent and diagnostic criteria with echocardiography must therefore be based on the positive identification of structures. No attempt was made to extend this investigation of healthy subjects in such a way as to measure, in statistical terms, the reliability of the method in its capacity to identify the various anatomical features. The

study should therefore be considered only as an attempt to show whether a structure is likely to be recognised or may not always be seen.

Values for the size of both ventricles, the aorta and the left atrium in healthy children (Feigenbaum, 1972) and infants (Hagan et al., 1973; Solinger et al., 1973) based on M-mode echocardiography have been reported. This technique allows exact measurement of the diameter of the great arteries but provides only one dimension for estimating the volume of the cardiac chambers in which the size varies in three dimensions. Particular attention has been given to the estimation of left ventricular size and function with M-mode echocardiography in adults (Gibson and Brown, 1973) and children (Kaye et al., 1975). Although there is good correlation between the left ventricular volume calculated from the angiocardiogram and the cube of the left ventricular internal diameter measured with M-mode echocardiography at the level of the mitral valve cusps, there is a large standard error and caution must be exercised in interpreting the echocardiographic data. Furthermore this correlation does not hold when the ventricle is dilated (Fortuin et al., 1971; Ludbrook et al., 1973). Gehrke et al. (1975) used ECG triggered two-dimensional echocardiographic images of the longitudinal view of the left ventricle to estimate its volume and concluded that the technique should be more reliable than M-mode volume calculations where the ventricle is large or irregular. Real-time two-dimensional echocardiography provides images which should allow a calculation of the volume of the cardiac chambers to be made more rapidly and accurately than with the ECG triggered or M-mode techniques. This

aspect of real-time two-dimensional echocardiography will merit further study but has not been undertaken for the purposes of this thesis. Where the relative sizes of cardiac chambers are quoted they have been estimated in a similar way to that in which they are estimated on viewing an angiocardiogram. In studying patients with narrowing of the right ventricular outflow (usually tetralogy of Fallot) or a hypoplastic pulmonary artery, an accurate estimate of the severity of the obstruction cannot always be made with M-mode echocardiography. Thus in the study of normal subjects the diameter of the right ventricular outflow just anterior to the aortic root has been measured where possible, and the ratio of this diameter to that of the aortic root calculated to allow a comparison with patients.

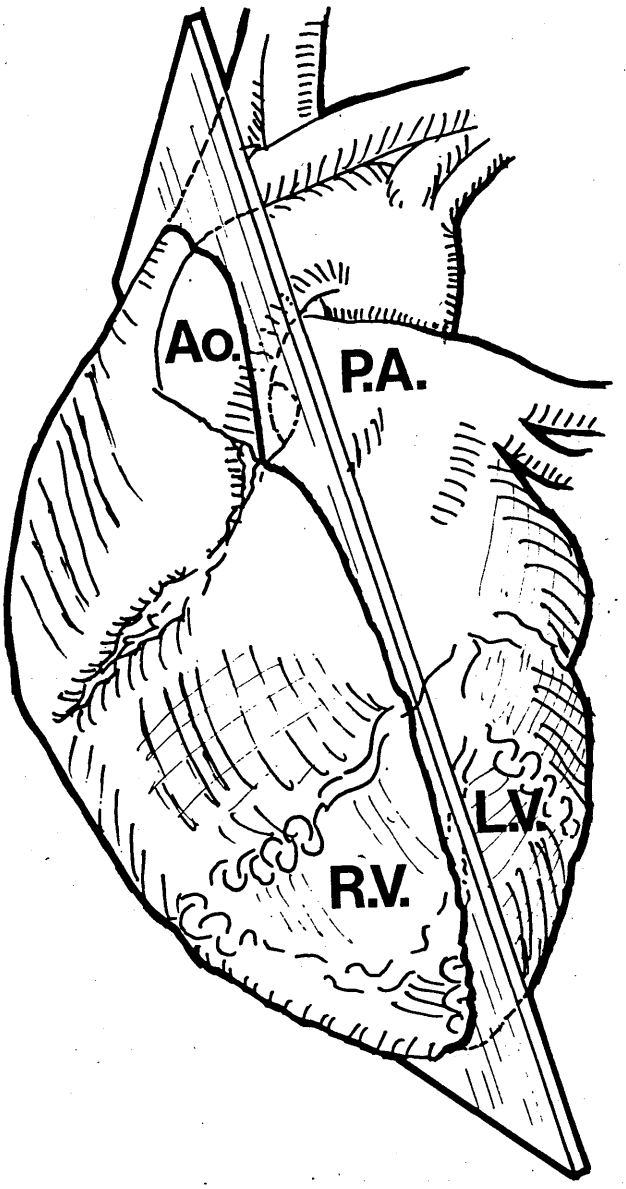
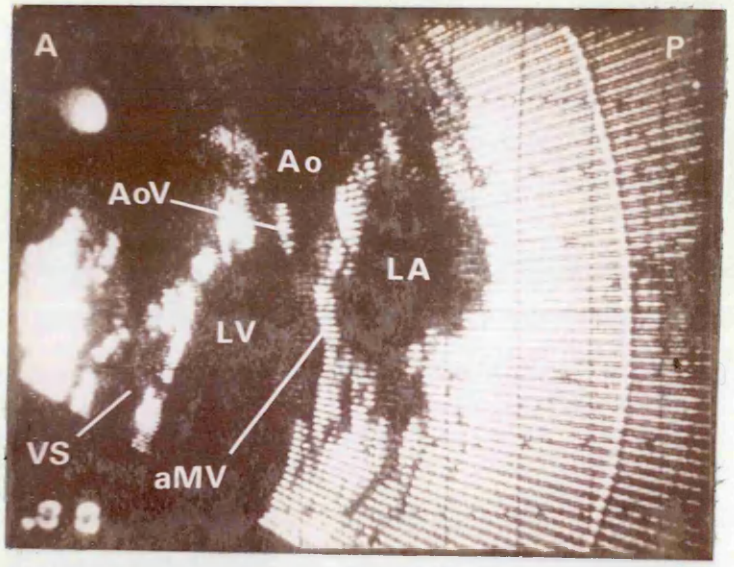
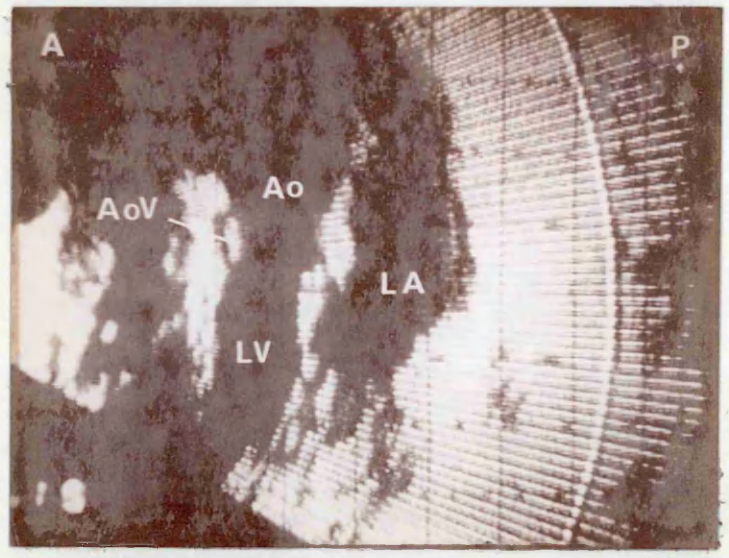


Figure 3, 1 Diagramatic representation of the heart and the longitudinal scanning plane. Ao, aorta; PA, pulmonary artery. Other abbreviations as before.

(a)



(b)



(c)

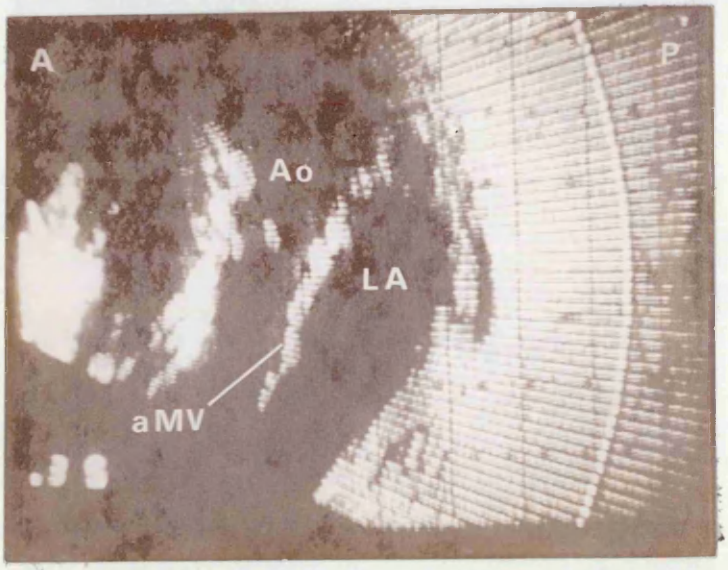


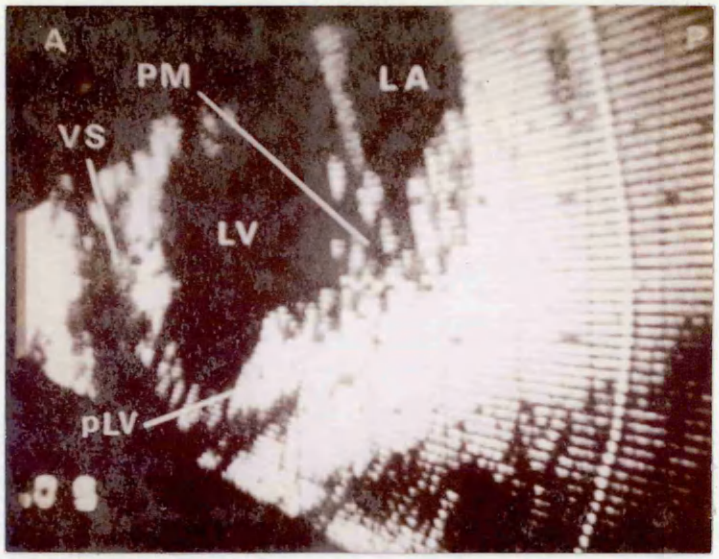
Figure 3, 2 Longitudinal views through the aortic root and body of the left ventricle from an infant (2.9 Kgs) during (a) pre-ejection period (at onset of R wave), (b) systole (0.15 seconds after R wave), and (c) diastole (0.35 seconds after R wave). The motion of the aortic and mitral valves can be appreciated. The marker arcs are 5 cm apart; the anterior one is obscured in (a) and (b). A = anterior; P = posterior; AoV = aortic valve cusps; aMV = anterior mitral valve leaflet; LA = left atrium. Other abbreviations as before.

LONGITUDINAL VIEWS

Standard Longitudinal View

The standard longitudinal view provides a saggital section through the long axis of the left ventricle from the cardiac apex to the aorta. It is most frequently obtained with the scanning head on the fourth left interspace at the left sternal edge and the scanning plane perpendicular to the chest wall and directed towards the right shoulder (Figure 3, 1). The rapidly moving anterior mitral valve leaflet is used as an initial landmark and the plane adjusted to lie along the long axis of the heart showing the aortic root, left atrium, left ventricle, ventricular septum and mitral and aortic valves; all these structures cannot always be seen in a single plane and minor alterations in transducer angulation may be required to display these structures optimally. Figures 3, 2 and 3, 3 are such views obtained from a 2.9 kg baby with a patent ductus arteriosus. The illustrations of the longitudinal view will be displayed in such a way that they are most easily interpreted if the reader imagines he is at the patient's left side seeing the heart cut along the plane indicated in Figure 3, 1; the orientation is similar to that of a lateral chest x-ray. In all such views the anterior chest wall will be towards the left. In the illustrations white represents echo-producing structures and black represents blood filled spaces. In the upper part of Figures 3, 2ab&c the aorta is demonstrated with its anterior and posterior walls continuous with the ventricular septum and anterior mitral leaflet respectively. The ventricular septum forms the anterior wall of the left ventricle and if the scanner is tilted inferiorly

(a)



(b)

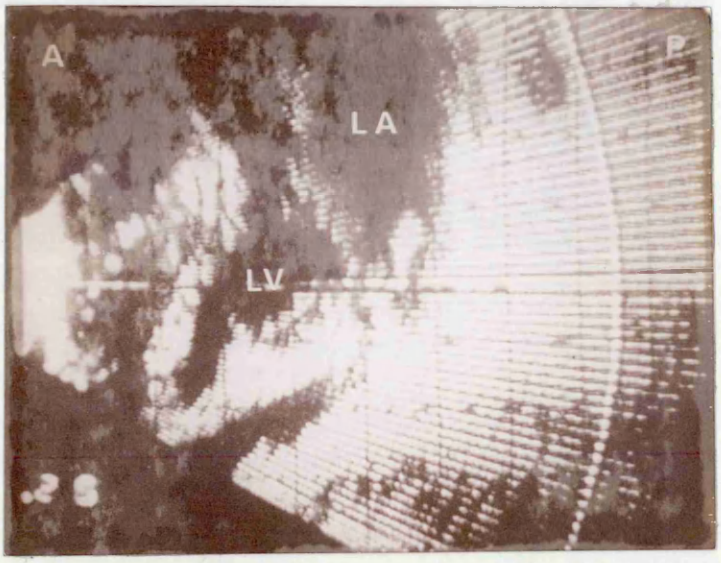


Figure 3, 3 Longitudinal views through the apex and body of the left ventricle from the same infant as Figure 3, 2 obtained in (a) early systole (0.05 seconds after the R wave) and (b) late systole (0.25 seconds after the R wave). The contraction of the ventricle is more clearly demonstrated than in Figure 3, 2. PM, papillary muscle; pLV, posterior wall of left ventricle. Other abbreviations as before.

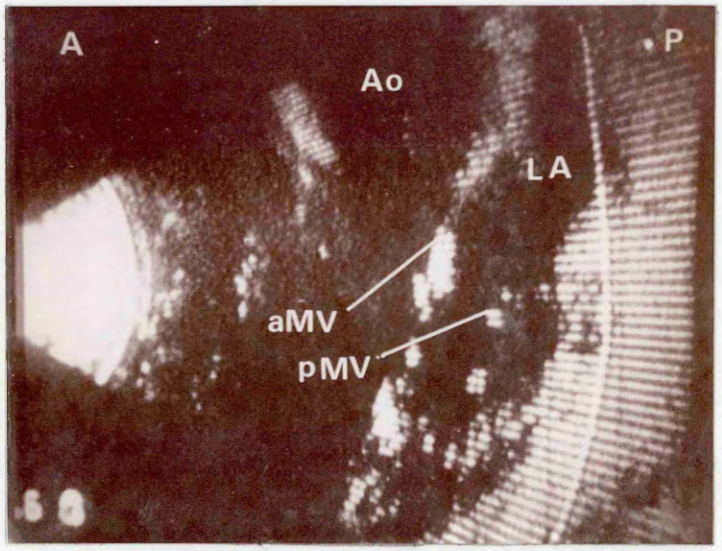


Figure 3, 4 Longitudinal views from a 6 year old child demonstrating both the anterior and posterior mitral valve leaflets. pMV, posterior mitral valve leaflet. Other abbreviations as before.

it can be seen to pass downwards and join the posterior wall at the cardiac apex (Figures 3, 3a&b). The left atrium lies posterior to the aortic root, the posterior wall of which forms the anterior atrial wall. The left atrium opens through the mitral valve into the left ventricle. The smaller posterior leaflet of the mitral valve, which delineates the transition from the left atrium to the left ventricle, is not seen in Figure 3, 2c but is shown clearly in Figure 3, 4 obtained from an older child. Attached to and passing down from the tips of the mitral leaflets echoes from the chordae tendineae can be shown to attach to papillary muscle (Figure 3, 3a). The contraction of the left ventricle and motion of the mitral and aortic valves, best appreciated in the moving (real-time) pictures, can be demonstrated in still frames obtained at several different time intervals through the cardiac cycle. The white dot in the top left corner of Figure 3, 2a is the flashing light which indicates that the picture was obtained at the time of the R wave on the ECG and the digital display in the bottom left hand corner indicates time elapsed after the R wave. Thus Figure 3, 2a is taken at the R wave and Figure 3, 2b 0.15 seconds and Figure 3, 2c 0.35 seconds after the R wave. Figure 3, 2a obtained in the pre-ejection period shows the closed aortic valve as a linear echo in the centre of the aortic root; the mitral valve is closed and the anterior and posterior cusps are in apposition. During systole (Figure 3, 2b) the aortic valve cusps are drawn back against the aortic walls and only the anterior one is seen as a separate echo; the mitral valve remains closed. In diastole (Figure 3, 2c) the central linear echo of the aortic valve

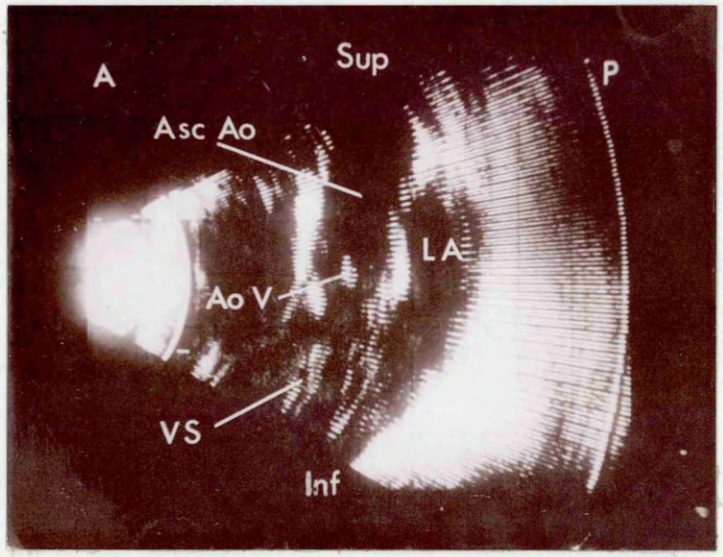
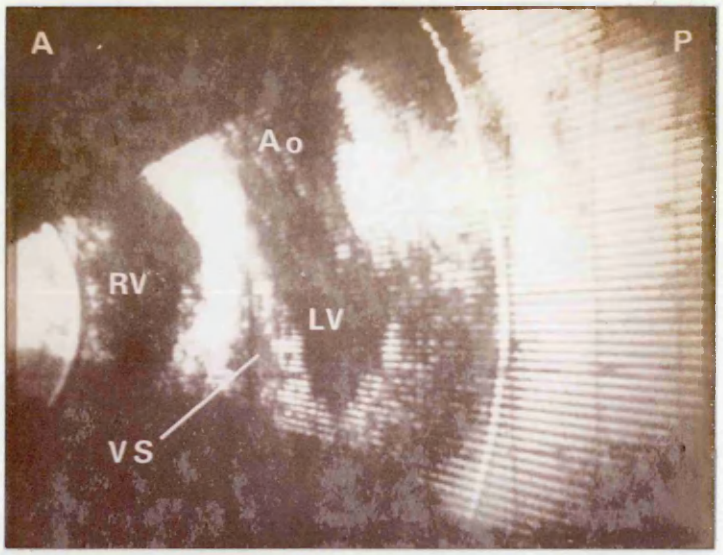


Figure 3, 5 High longitudinal view from a newborn infant showing the ascending aorta. Sup, superior; Inf, inferior; Asc Ao, ascending aorta. Other abbreviations as before.

(a)



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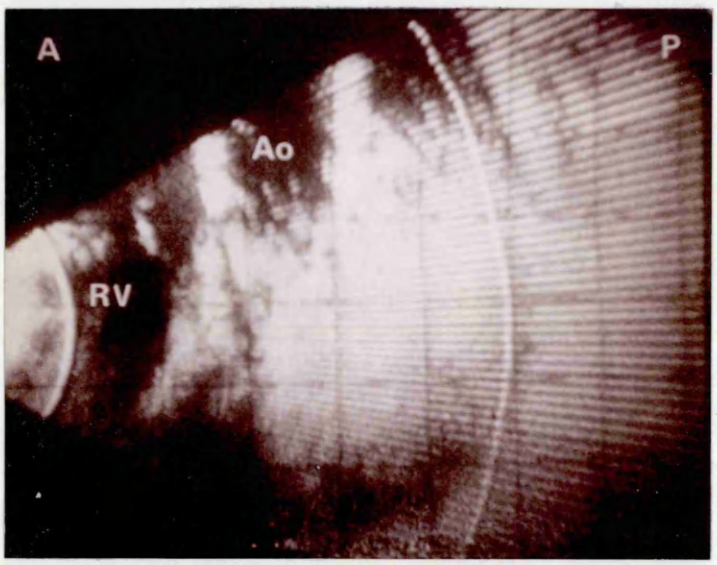


(c)

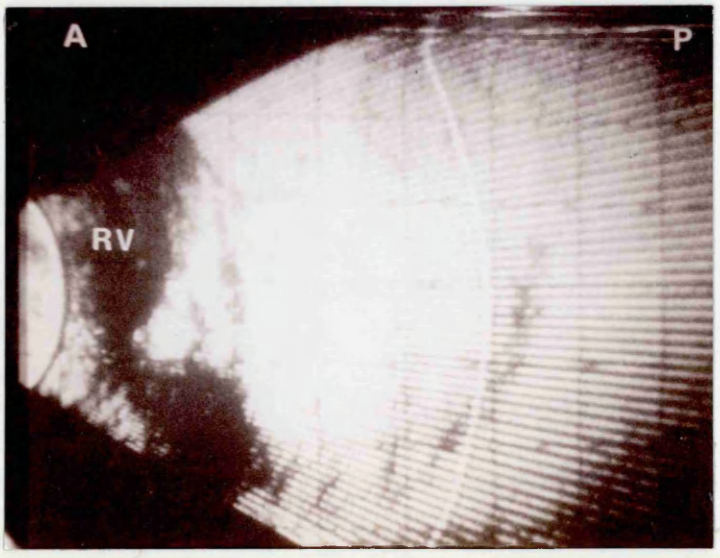


Figure 3, 6 Longitudinal views from a 6 year old child following the injection of blood into left atrium. Ultrasonic contrast is seen first in (a) the left atrium only, then (b) in the left ventricle with subsequent diastole, and finally (c) in the aortic root with subsequent systole. Abbreviations as before.

returns and as the mitral valve opens its anterior cusp moves anteriorly towards the ventricular septum. Left ventricular contraction can be appreciated by comparing the shapes in early (Figure 3, 3a, 0.05 seconds after the R wave) and late (Figure 3, 3b, 0.25 seconds after the R wave) ventricular systole. Similarly the left atrium contracts from its maximal size at end ventricular systole to its smallest at end diastole (i.e. after atrial systole).

High Longitudinal View

The high longitudinal view of the ascending aorta is a modification of the longitudinal view of the left ventricle. The scanning head is tilted or edged superiorly and adjusted to show the approximately parallel anterior and posterior walls of the aorta passing upwards from the aortic valve cusp level (Figure 3, 5). When the ascending aorta is shown clearly the left ventricle and mitral valve are frequently sectioned obliquely.

Contrast Studies

The appearances in the longitudinal views are illustrated in Figures 3, 6ab&c which were obtained following the injection of blood into the left atrium in a 6-year-old patient with partial anomalous pulmonary venous drainage. Echoes were first visualised proximal to the mitral valve (i.e. in the left atrium) (Figure 3, 6a); with ventricular diastole the echoes appeared distal to the mitral valve (i.e. in the left ventricle) (Figure 3, 6b) but the aortic root remained free of contrast until ventricular systole when it also opacified (Figure 3, 6c). The space anterior to the ventricular septum remained clear following the injections into the left atrium and left ventricle and was identified as the

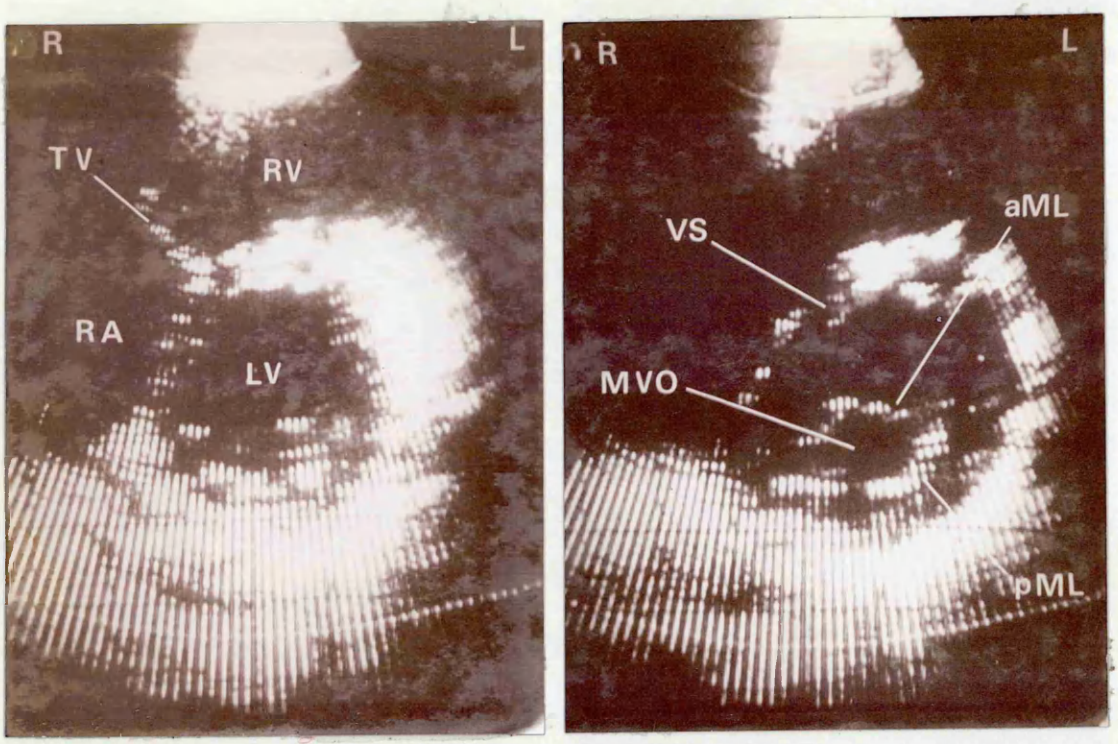
right ventricle by injection of contrast directly into its cavity. These studies thus confirmed the identification of the left atrium, left ventricle, aortic root and right ventricle and, by implication, that of the mitral and aortic valves and ventricular septum.

Normal Subjects

In the longitudinal view the aortic root, aortic valve cusps, ventricular septum and the anterior mitral valve leaflet were recorded in all 40 subjects. It was not possible to demonstrate the anterior mitral chordae and papillary muscle in three infants (2, group A; 1, group B) or the posterior mitral valve leaflet in 22 subjects (7, group A; 8, group B; 5, group C; 2, group D). The cardiac apex could not be convincingly shown in 14 subjects (2, group A; 3, group B; 5, group C; 4, group D). Careful adjustment of gain settings were performed in an attempt to show the anterior heart wall but this structure could not be imaged in 22 patients (2, group A; 6, group B; 8, group C; 6, group D). There appeared to be an echo-free space posterior to the left atrium in 9 patients (4, group A; 2, group B; 2, group C; 1, group D). A similar appearance is shown in Figure 3, 2c. It has not yet been possible to provide a satisfactory explanation for this appearance.

The high longitudinal view in all 40 subjects showed the parallel echoes from the anterior and posterior walls of the ascending aorta passing upwards from the aortic root. In most cases it was not possible to determine the point at which the ascending aorta arched posteriorly to become the aortic arch either because the aorta turned outwith the scanning plane or

because of difficulty imaging the upper parts of the ascending aorta. In 39 subjects the posterior aortic wall could be followed upwards from the lower edge of the aortic cusp echo for a distance at least as great as the diameter of the aortic root. In 1 infant (group B) the anterior and posterior walls of the ascending aorta could be followed upwards for only a short distance.



(a) (b)

Figure 3, 7 Transverse views through the left ventricle at the level of the mitral valve tips obtained (a) in systole with the mitral valve closed, and (b) in diastole with the valve open and its orifice clearly seen. MVO, mitral valve orifice; RA, right atrium; TV, anterior leaflet of the tricuspid valve. Other abbreviations as before.

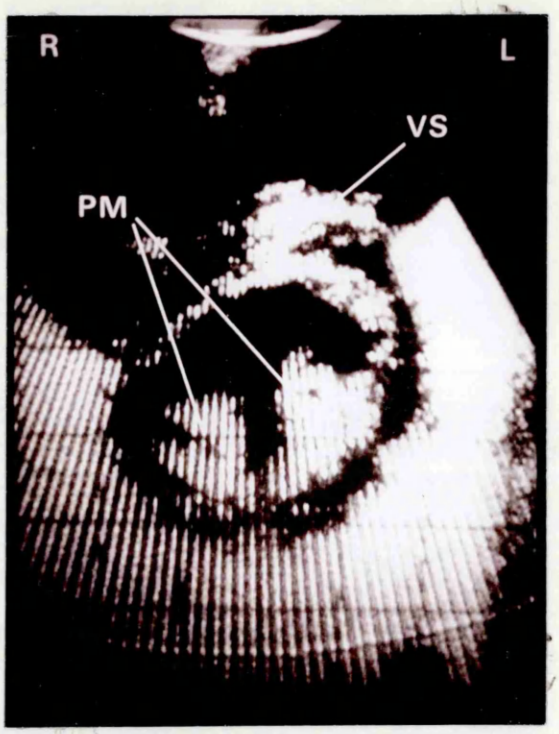


Figure 3, 8 Low transverse view through the left ventricle at the level of the papillary muscles. Abbreviations as before.

TRANSVERSE VIEWS

Low Transverse View

The low transverse view provides a transverse section through the body of the left ventricle. It is obtained by positioning the scanning head on the same chest wall site from which a satisfactory longitudinal scan was obtained and rotating the scan through approximately 90^{o}. This plane lies through the short axis of the left ventricle and shows it as an almost circular shape with the anterior and right walls formed by the ventricular septum. Figures 3, 7a&b are low transverse views through the left ventricle at the level of the mitral valve cusps shown as if the heart is being viewed from the patient's feet. Thus the apex of the sector is anterior and the left of the illustration represents the patient's right side. Tilting the plane towards the apex and then up to the aortic root provides the view of all parts of the left ventricular wall. The tips of the mitral valve cusps are seen in the mid-position of this sweep. In systole the valve is closed and the leaflets, in apposition, appear as confluent echoes close to the posterior wall of the left ventricle (Figure 3, 7a); in diastole the valve opens and the valve orifice is seen as an oval space between the anterior and posterior cusps (Figure 3, 7b). Below the level of the mitral valve the chordae tendineae are visible. In the lower left ventricle the papillary muscles are seen as two protrusions from the ventricular wall (Figure 3, 8). The right ventricle lies anterior and to the right of the left ventricle; the scanner may have to be tilted to the right to show it clearly. The right wall of the right ventricle is difficult to visualise

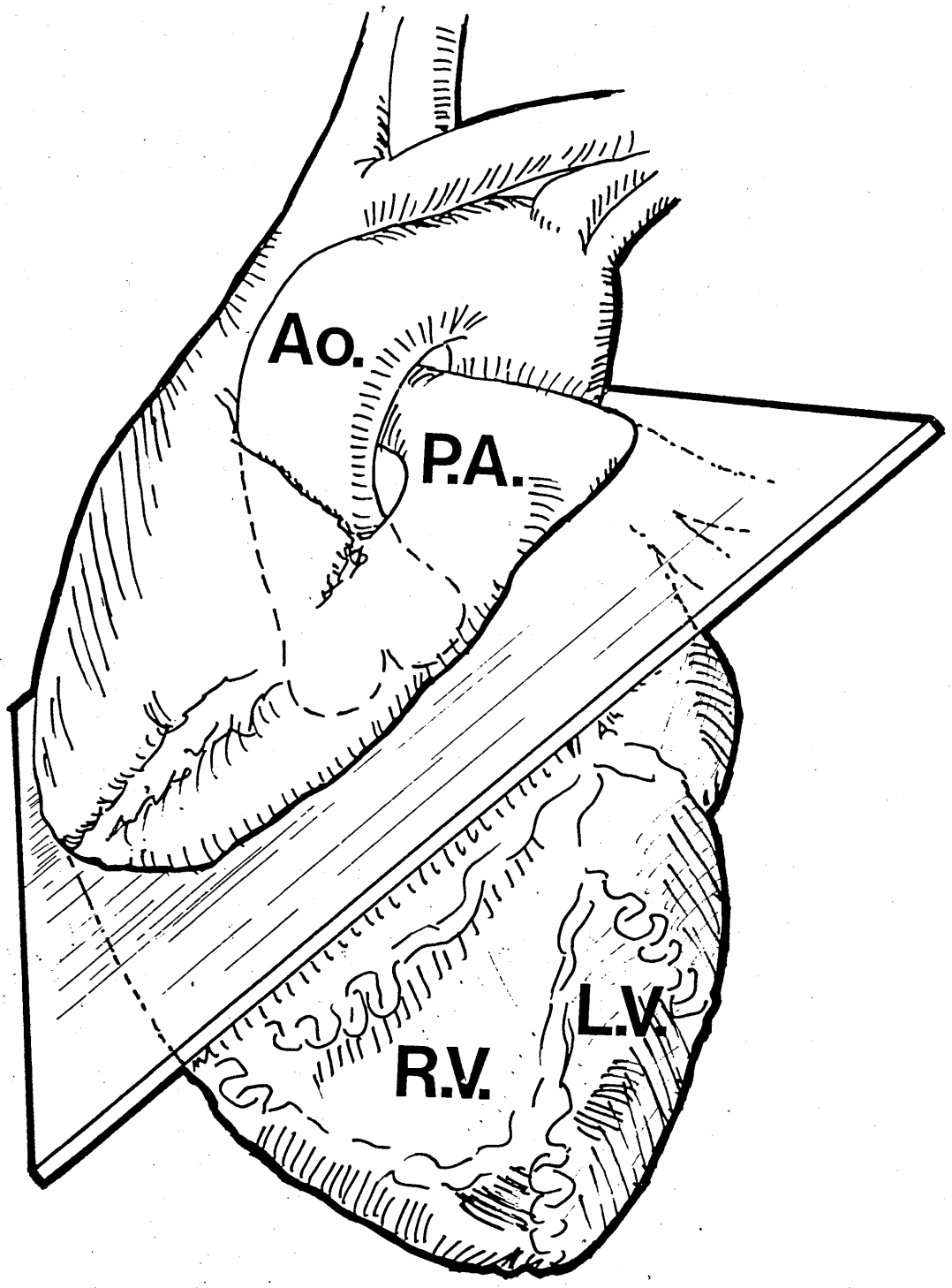


Figure 3, 9 Diagramatic representation of the heart and the high transverse scanning plane. Abbreviations as before.

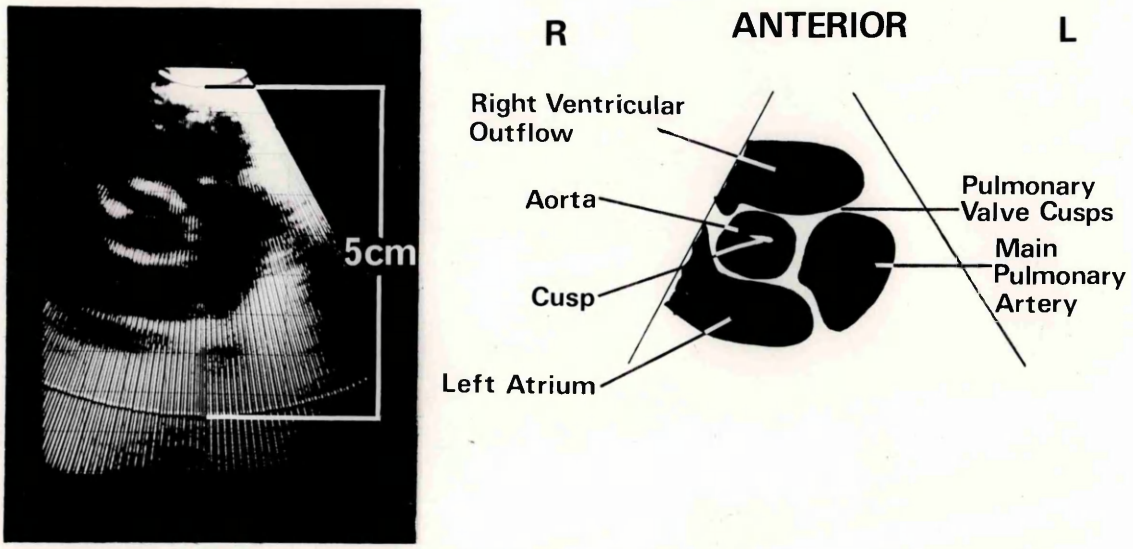


Figure 3, 10 High transverse view, in diastole, from a newborn infant. The scanning plane, which lies along the right ventricular outflow and main pulmonary artery and through the pulmonary valve, passes through the aortic root transversely.

because of the oblique angle of the ultrasound beam and interference due to calcified sternum. The anterior tricuspid leaflet which lies to the right of the ventricular septum exhibits motion similar to that of the anterior mitral leaflet and separates the right ventricle (anterior) from the right atrium (posterior) (Figure 3, 7a).

High Transverse View

The high transverse view shows the relationship of the great arteries at their origins and also the atria and, in some cases, the atrial septum. It is most often obtained with the scanner one interspace higher than the position from which the low transverse scan is obtained. In the normal heart the right ventricular outflow crosses the aortic root obliquely before passing posteriorly and to the left as the main pulmonary artery. For assessment of relationships and sizes of the great arteries the high transverse plane is adjusted to lie through the aortic root and along the right ventricular outflow and main pulmonary artery by angling the scanner towards the lateral third of the left clavicle and tilting it superiorly (Figure 3, 9). Figure 3, 10 is such a view in diastole from a 2.9 kg baby. The illustrations are displayed in such a way that they are most easily interpreted if the reader imagines he is at the patient's feet looking headwards and seeing the heart cut along the plane indicated in Figure 3, 9. In all such views the anterior chest wall will be upwards and the patient's left side to the right of the illustration. The scanning plane is first adjusted to position the aorta at the level of the valve, as identified by its cusps, in the centre of the scanning arc.

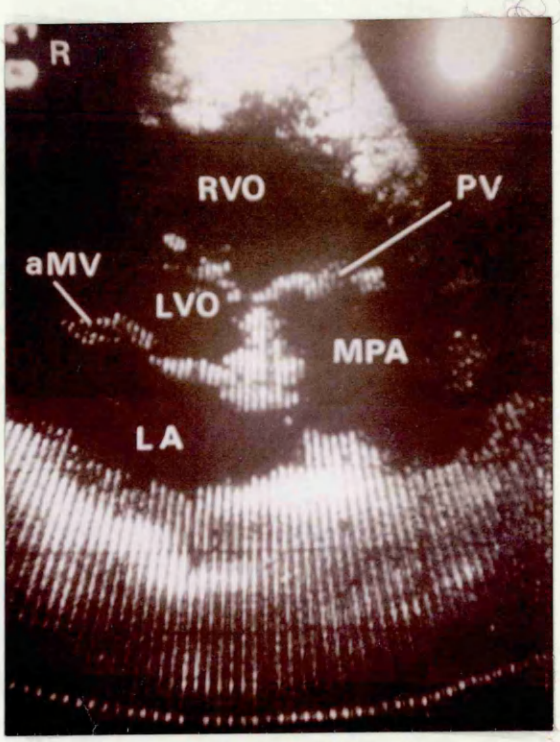


Figure 3, 11 High transverse view with the scanning plane lying along the right ventricular outflow and main pulmonary artery and through the pulmonary valve. The plane passes through the left ventricular outflow and not the aortic root. LVO, left ventricular outflow; MPA, main pulmonary artery; RVO, right ventricular outflow. Other abbreviations as before.

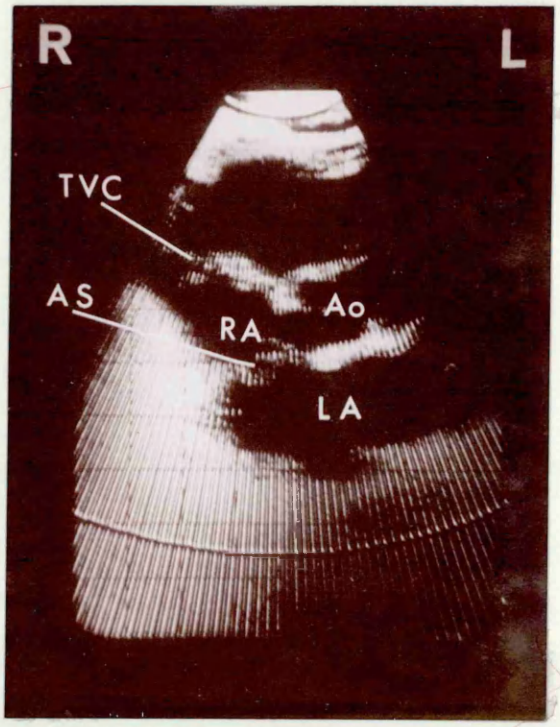
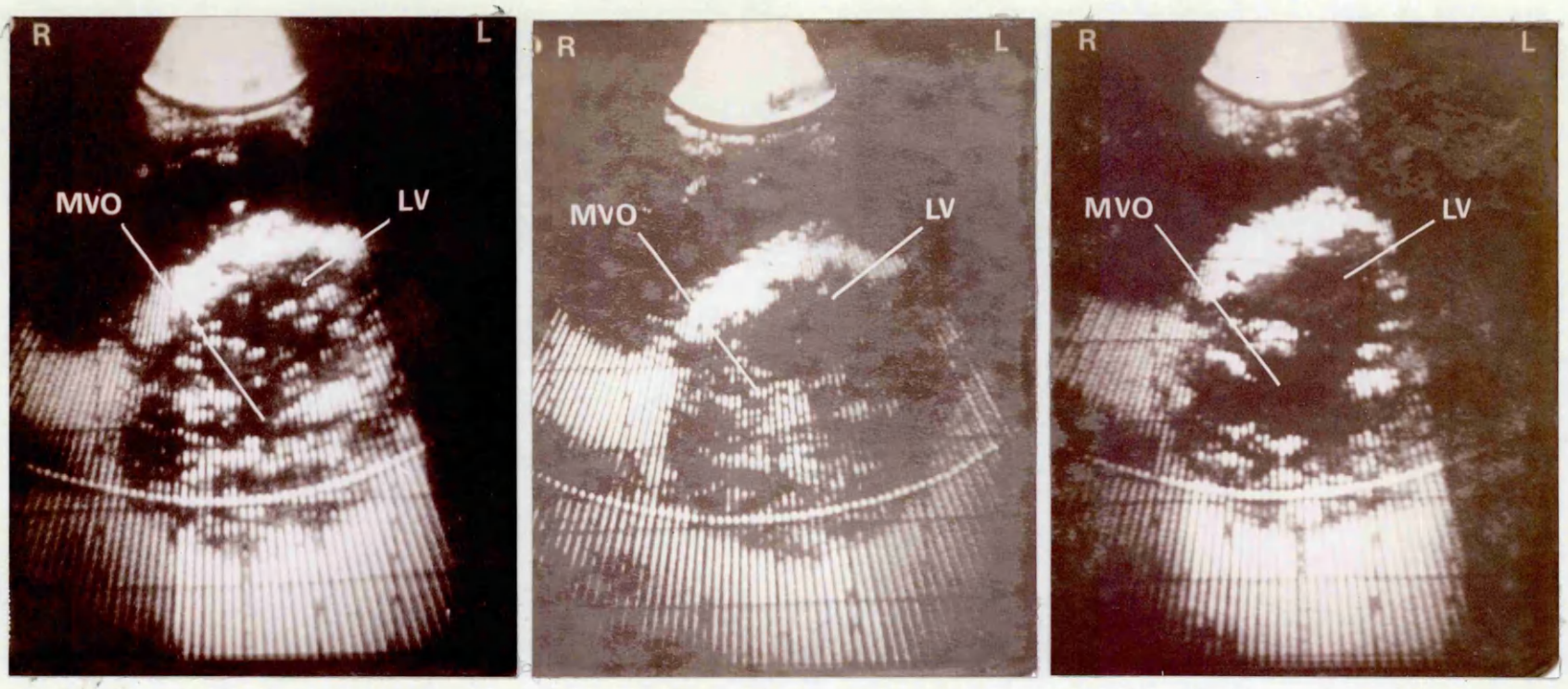


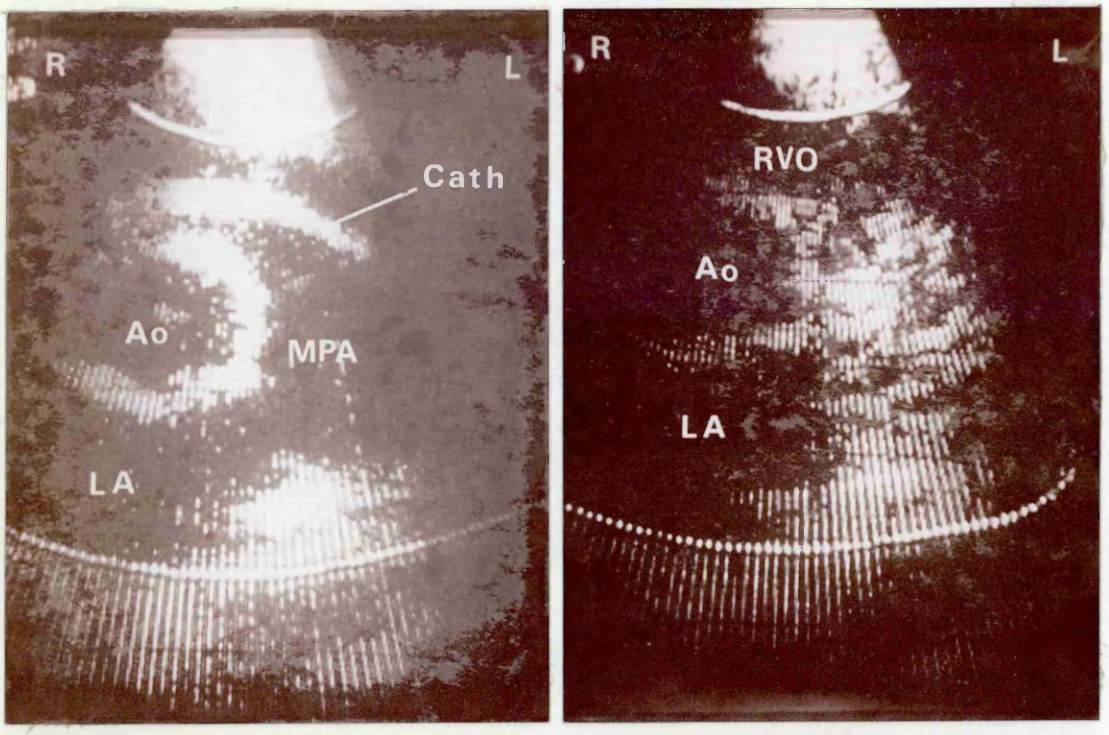
Figure 3, 12 High transverse view from the infant in Figure 3, 10 adjusted to show the atrial septum. AS, atrial septum; TVC, anterior tricuspid valve leaflet. Other abbreviations as before.

The aorta cut in transverse section is seen as an approximately circular space with the valve cusps as a central linear echo in diastole which disappears in systole. The right ventricular outflow appears as an echo-free space passing anterior to and then to the left of the aorta as the main pulmonary artery, with the echoes from the cusps of the pulmonary valve indicating the transition between them. The pulmonary valve lies anterior and to the left of the aorta. In some cases the right ventricular outflow and main pulmonary artery may cross the aortic root more obliquely and the course is better imaged in a more longitudinal plane which crosses the left ventricular outflow rather than the aortic root. In Figure 3, 11 the right ventricular outflow, pulmonary valve and main pulmonary artery are clearly seen in such a plane; this is more directly longitudinal than indicated in Figure 3, 9 and lies to the left of the aortic root thus showing the right ventricular outflow in relation to the ventricular septum and anterior mitral valve leaflet. Thus a scan through the right ventricular outflow and main pulmonary artery may section the aorta somewhat obliquely rather than transversely, or may not show it at all, and minor alterations in the plane of scan are then required to show the aorta in direct transverse section. The left atrium lies directly posterior to the aortic root. Its anterior wall is formed by the posterior aorta and the medial one by the atrial septum. The atrial septum is not always seen in this view which shows great artery origins optimally, but it may be demonstrated by adjustment of the scanner position (Figure 3, 12). The right atrium lies to the right of the atrial septum and posterior to the tricuspid valve. This view



(a) (b) (c)

Figure 3, 13 Low transverse views at the level of the mitral valve orifice at approximately the same time (ventricular diastole) during different cardiac cycles. (a) Before contrast injection; (b) immediately following contrast injection into the left atrium, showing opacification of the mitral orifice; (c) subsequent cardiac cycle showing contrast in the body of the left ventricle as well as the mitral valve orifice. Abbreviations as before.



(a) (b)

Figure 3, 14 High transverse views obtained at the time of cardiac catheterisation. In (a) artefact from the cardiac catheter is seen in the RVO and MPA, while in (b) following contrast injection the MPA opacifies but the catheter, which moves with each heart beat, has moved outwith the scanning plane. Cath, cardiac catheter. Other abbreviations as before.

frequently shows the anterior tricuspid leaflet just to the right of the aortic root (Figure 3, 12).

Contrast Studies

The appearances in the low transverse view are illustrated in Figures 3, 13ab&c which were obtained following the injection of blood into the left atrium in a 7 year old patient. All three illustrations were obtained at approximately the same time in the cardiac cycle (ventricular diastole) when the mitral valve was in the open position. Figure 3, 13a, before injection, shows the mitral valve orifice and the anterior part of the left ventricular cavity (the left ventricular outflow). Contrast first appeared in the mitral valve orifice (Figure 3, 13b) and the left ventricular outflow remained echo free until later in the cardiac cycle when it also opacified (Figure 3, 13c). These studies thus confirmed the identification of the mitral valve orifice and left ventricular outflow.

The appearances in the high transverse view are illustrated in Figures 3, 14a&b which were obtained from a 6 year old patient in whom the cardiac catheter was passed from the right ventricle into the main pulmonary artery. Figure 3, 14a shows the echo artefact from the cardiac catheter passing along the right ventricular outflow anterior to the circular aorta and then curving to the left into the main pulmonary artery. The opacification of the main pulmonary artery following injection just distal to the pulmonary valve is shown in Figure 3, 14b; the right ventricular outflow, aorta and left atrium remained clear following this injection. Similar studies confirmed the identity of the right ventricular outflow, left atrium and aorta.

Normal Subjects

In the low transverse view the entire circumference of the left ventricle at the level of the mitral valve cusps was imaged in all but 3 patients (1, group A; 2, group B) in whom the lower part of the right wall (part of the ventricular septum) could not be clearly defined. The anterior mitral and tricuspid leaflets were demonstrated in all subjects but it was not possible to identify with certainty the posterior mitral valve leaflet in 11 (5, group A; 3, group B; 1, group C; 2, group D) or the papillary muscles in 17 (6, group A; 5, group B; 4, group C; 2, group D).

In the high transverse view the aortic root and central aortic cusp echoes were apparent in all 40 subjects. The right ventricular outflow and main pulmonary artery were clearly seen simultaneously with a transverse section of the aortic root in 27 and in a more longitudinal plane above the left ventricular outflow in 12 (2, group A; 2, group B; 1, group C; 7, group D). In one infant (group B) a satisfactory image of the right ventricular outflow and main pulmonary artery was not obtained because of intervening echoes (probably from lung). The pulmonary valve could not be recorded in 3 infants (group B) but its motion was clearly discernible in all other subjects. A clearly demarcated edge representing the upper wall of the right ventricular outflow (RVO) and pulmonary artery was not recognised in 8 subjects (3, group B; 4, group C; 1, group D). The ratio of the RVO to the aortic root in the other 32 subjects was:

Group A, range 0.9 to 1.4 (mean 1.1)

Group B, range 0.8 to 1.1 (mean 1.0)

Group C, range 0.8 to 1.0 (mean 0.9)

Group D, range 0.9 to 1.0 (mean 1.0)

The atrial septum was seen in 17 subjects (5, group A; 5, group B; 4, group C; 3, group D).

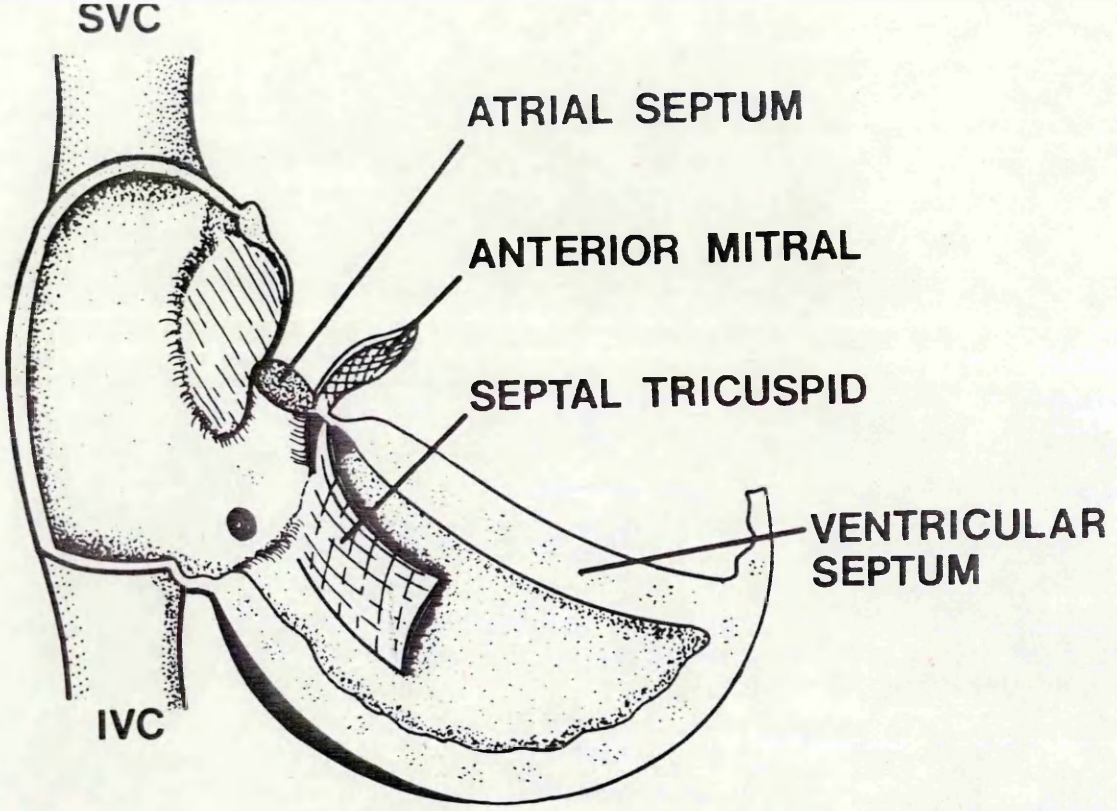


Figure 3, 15a Diagramatic representation of the heart as seen from above and sectioned along the plane of the four chamber view. The ventricular and atrial septa are continuous, and the tricuspid valve lies antero-inferior to the mitral, being separated from it by part of the membranous septum.

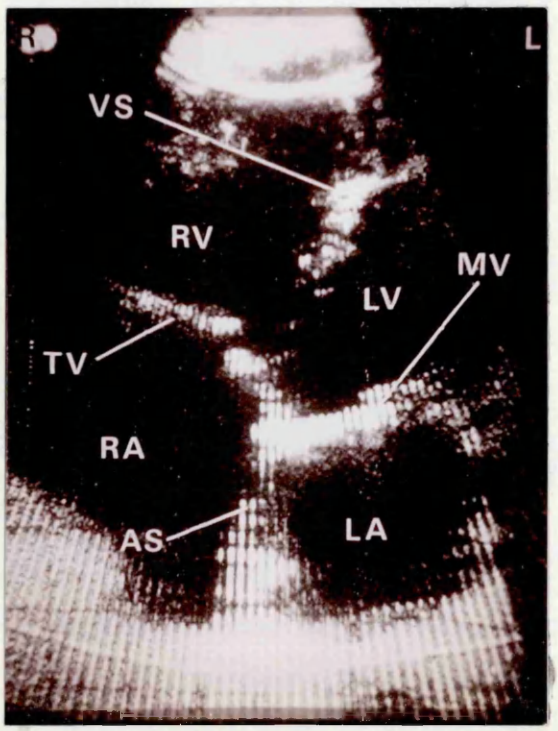
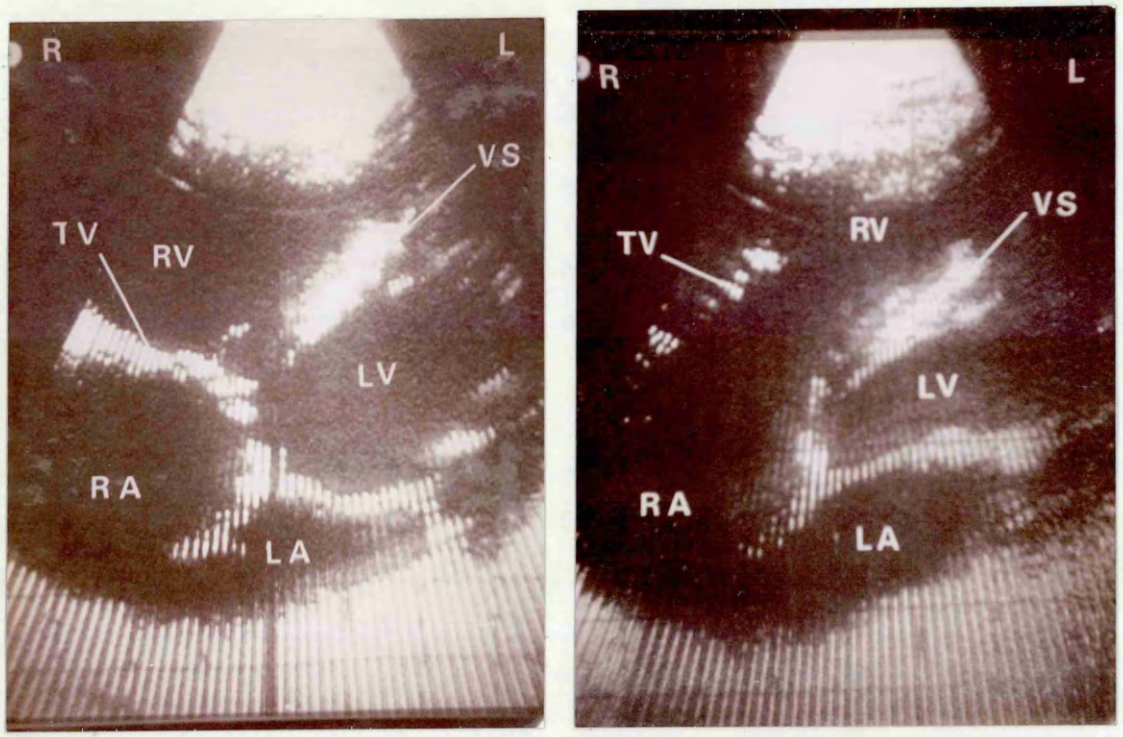


Figure 3, 15b Four chamber view from a 6 year old child with a normal heart. The ventricular and atrial septa are continuous and the tricuspid valve impinges on the ventricular septum anterior to the mitral. Although the whole ventricular septum was clearly seen in the moving display there is some echo drop-out from its posterior margin in this still frame. MV, anterior leaflet of the mitral valve. Other abbreviations as before.

OBLIQUE OR FOUR CHAMBER VIEW

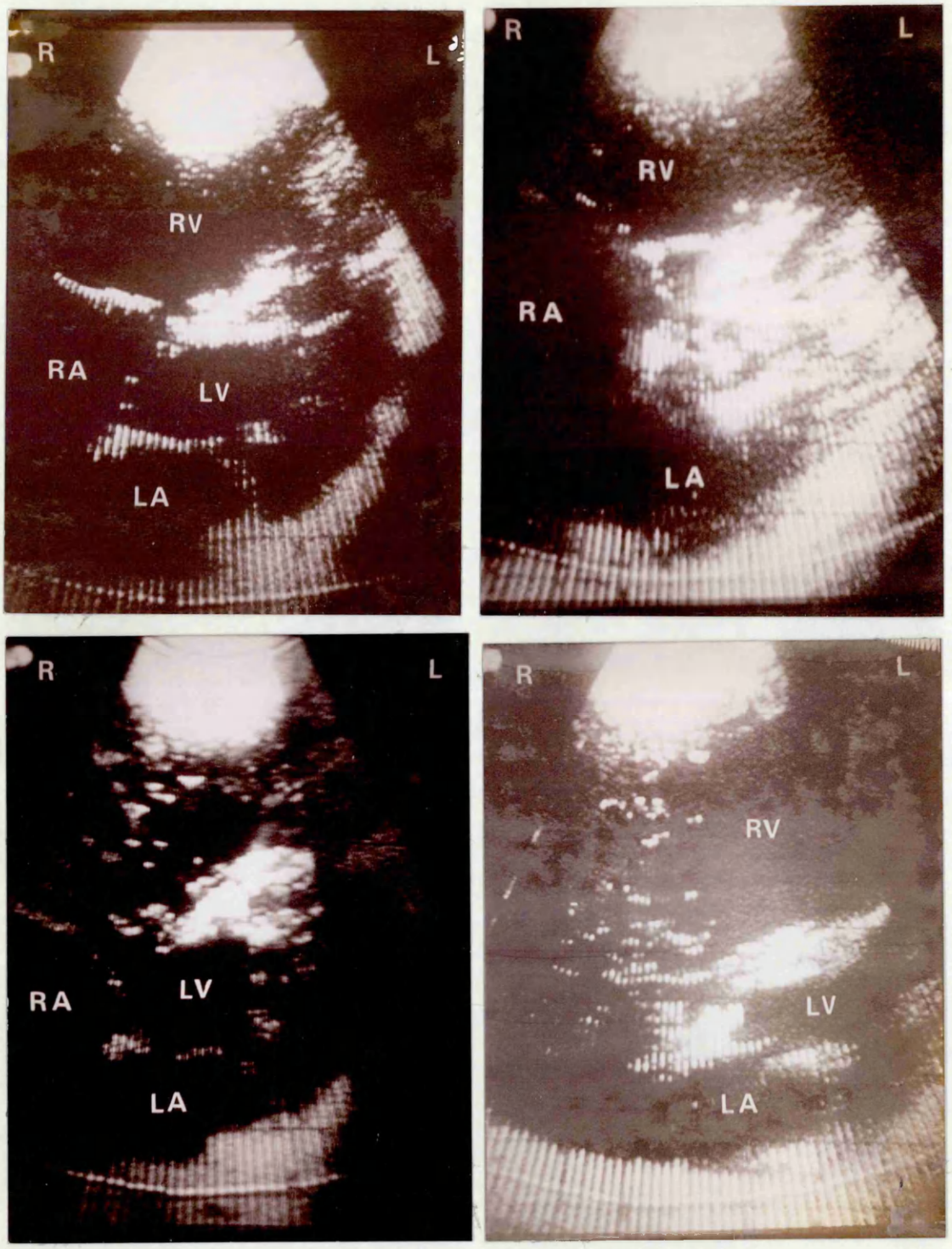
This view is designed to show the junction of the ventricular and atrial septa and anterior mitral and tricuspid leaflets. The scanner is placed and orientated as for the low transverse scan, showing the anterior and posterior mitral leaflets and the posterior left ventricular wall, and then tilted to the right to bring the anterior tricuspid leaflet into view; satisfactory views are frequently obtained with the scanning plane almost directly transverse. The plane is then angled upwards until the posterior left ventricular wall and posterior mitral valve leaflets are no longer seen and then adjusted to show the atrial septum. The scanning head often has to be moved to a lower interspace to obtain an optimal four chamber view. Figure 3, 15a is a diagramatic representation of the structures seen with this view. The ventricular septum lies in the antero-inferior position, curves posteriorly and to the right, and is continuous with the atrial septum (inferior limb of the fossa ovalis). The anterior mitral and septal tricuspid leaflets are attached to the left and right sides respectively of the ventricular septum at different levels, the tricuspid leaflet being the lower. Figure 3, 15b is an echocardiogram from a 6 year old child with a normal heart showing this four chamber view. It is orientated in a similar manner to the transverse views, that is it is viewed as if looking up from below. The apex of the sector thus represents an antero-inferior position and the base a postero-superior one. The ventricular septum is seen as a relatively thick structure which curves posteriorly from the left side and joins the atrial septum. The atrioventricular



(a) (b)

Figure 3, 16 Four chamber view from a 4 year old child with an ASD. (a) In systole the atrioventricular valves are closed, while (b) in diastole both anterior leaflets move anteriorly, the tricuspid also moving away from the ventricular septum. Note that in (a) there is a demarcated posterior edge to the atrial septum, possibly indicating the edge of the septal defect. Abbreviations as before.

(a) (b)



(c) (d)

Figure 3, 17 Contrast echocardiography and the four chamber view. (a) Before contrast injection, and following injection into (b) left ventricle, (c) right ventricle, and (d) the right atrium. Abbreviations as before.

valve leaflets are represented by transverse linear echoes with the tricuspid leaflet adjacent to the right side of the ventricular septum and the anterior mitral valve leaflet to the left. The septal tricuspid leaflet is small and in most cases the tricuspid valve echo seen in this view represents the anterior tricuspid leaflet. This is illustrated in Figures 3, 16a&b. In systole (Figure 3, 16a) the transverse echo from the tricuspid valve is apparent and in diastole it moves anteriorly (Figure 3, 16b). The echo is relatively long with no apparent attachment to the ventricular septum suggesting that it does not represent the septal tricuspid leaflet, and since the echo moves anteriorly in diastole it must represent the anterior rather than the posterior tricuspid valve cusp. The tricuspid leaflet impinges on the ventricular septum in a more anterior and inferior position than the anterior mitral leaflet and this separation is clearly demonstrated. This view therefore visualises all four cardiac chambers simultaneously. It also shows the exact relationship of the atrial and ventricular septa and the anterior leaflets of both atrioventricular valves at their junction point.

Contrast Studies

The appearances in the four chamber view are illustrated in Figures 3, 17abc&d which were obtained from a 6 year old child with an atrial septal defect. Figure 3, 17a, prior to injection, shows all four chambers, the anterior atrioventricular valves, ventricular septum and the anterior part of the atrial septum. Opacification of the chambers following contrast injection into the left ventricle, right ventricle and right atrium is demonstrated in Figures 3, 17bc&d respectively. Contrast injection into the

left atrium showed it to opacify immediately and echoes were subsequently seen in the left ventricle and right atrium. These studies thus confirmed the identity of the four cardiac chambers and thus, by implication, that of the septa and anterior atrioventricular valves.

Normal Subjects

In all subjects the anterior mitral and tricuspid valve cusps were demonstrated with the tricuspid leaflet anterior to the mitral leaflet. Echoes from the whole length of the ventricular and atrial septa were apparent in 31 subjects but it was not possible to define clearly the posterior margin of the ventricular septum in 4 (1, group A; 1, group B; 2, group D) or the central and posterior parts of the atrial septum in 3 (1, group C; 2, group D). In 2 subjects (group C) the posterior margins of both the ventricular and atrial septa were not clearly demonstrated.

DISCUSSION

The author's initial experience in the longitudinal view and low and high transverse views has previously been published (Houston et al., 1977b) and his description of the four chamber view reported (Houston et al., 1978b).

The appearances in the longitudinal and low transverse views were first described using a multiple element two-dimensional system by Kloster et al. (1973) and an appraisal of their clinical usefulness reported by Roelandt et al. (1974). The findings were subsequently confirmed using the steered beam system (Kisslo et al., 1976) and a mechanical sector scanner (Nishimura et al., 1976). Kisslo et al. (1976) were the first group to report the demonstration of the papillary muscles in the longitudinal and transverse views. The findings reported by these workers correspond to those described in this thesis and previously reported by the author and his colleagues (Houston et al., 1977b). The two groups of workers who initially studied the high transverse view in patients with normally related great arteries found differences in the appearances. Henry et al. (1975) and Maron et al. (1975) showed an appearance similar to that initially reported with this system; the aorta was seen in transverse section with the right ventricular outflow crossing anterior to it before turning posteriorly to its left as the main pulmonary artery. Sahn et al. (1974a) were able to show both the aorta and main pulmonary artery as circular structures in a directly transverse section and the right ventricular outflow and main pulmonary artery in a more longitudinal view crossing the aorta or left ventricular outflow tract obliquely. Experience subsequent to the initial report using the present scanning

equipment has shown that, although in the majority of cases the right ventricular outflow and main pulmonary artery are shown when the aorta is cut in a directly transverse section, in others they are better seen in a more longitudinal view which crosses the aorta or left ventricular outflow obliquely. It has never been possible to show the great arteries at cusp level simultaneously as circular spaces in any subjects with normally related great arteries; the difference between these findings and the appearance noted with the multiple element system may be explained by the better lateral resolution of single element scanners.

A four chamber view with two-dimensional echocardiography was first reported by Beppu et al. (1976) using an ECG-triggered B-scan image to study normal subjects and patients with atrio-ventricular canal abnormalities or ostium secundum atrial septal defects. Similar four chamber views can be obtained from the parasternal or apical positions with real-time echocardiography; the author and his colleagues, with a mechanical sector scanner, reported the appearances from the parasternal position (Houston et al., 1978b) and, with the phased array system, Silverman and Schiller (1978) described the findings from an apical position with the patient lying on his left side, and Tajik et al. (1978) from both the parasternal and apical positions. In accord with the findings described here the insertion of the mitral and tricuspid leaflets into the ventricular septum at different levels was also noted by Beppu et al. (1976) and Tajik et al. (1978) but Silverman and Schiller (1978) considered the leaflets joined the ventricular septum at the same point.

The appearances which have been found in the standard views thus concur with those reported by most other workers. The most comprehensive description of the appearances in the normal two-dimensional echocardiogram has been that of Tajik et al. (1978) who described 20 different views. Their report included all but one of the basic views and their modifications described in this chapter; they did not describe a high longitudinal view of the ascending aorta which, as will be described in Chapter IV, the author considers to be essential in identifying the great arteries in cases of complex congenital heart disease. In addition they used subxiphoid and suprasternal transducer placements which have not been used in the basic studies in this thesis but which should add to the information obtained by two-dimensional echocardiography by providing a better view of the atrial septum and showing the inferior vena cava, the aortic arch, and right pulmonary artery. The author has been using the subxiphoid view for studies conducted after completion of the work described in this thesis but has found that with the large prototype scanner satisfactory suprasternal images cannot be regularly obtained. Nevertheless the views described in this chapter permit visualisation of all four cardiac chambers, the atrioventricular and semilunar valves and the proximal parts of the great arteries, and allow exact determination of the relationships of different structures. These views are therefore considered to provide a basic anatomical profile of the heart which allows comparisons to be made with anatomical abnormalities found in many types of congenital heart disease. The studies in the 40 normal subjects have shown that it is not always possible

to recognise all the cardiac structures; this emphasises the fact that, with echocardiography, any diagnostic criterion must be based on the positive identification of anatomical structures, whereas any criterion based on a negative finding would have to be regarded with extreme caution.

CHAPTER IV

COMPLEX CONGENITAL HEART DISEASE

This chapter describes the findings with two-dimensional echocardiography in groups of patients with different forms of complex congenital heart disease. One of the most relevant clinical applications of these studies is the preliminary assessment of children with congenital heart disease. The information obtained can be used to decide the timing of cardiac catheterisation, and then to assist with the planning of the procedure and the interpretation of the findings. Chapter VI will evaluate the results obtained in the initial assessment of a consecutive series of 64 infants suspected of having cyanotic congenital heart disease. However it was first considered necessary to establish the echocardiographic appearances in the more common types of complex congenital heart disease; this chapter details these findings in patients examined before or after angiocardiography. A preliminary study was carried out to establish the echocardiographic appearances of the great arteries in a series of 19 patients with known great artery abnormalities and 11 cyanotic patients with suspected congenital heart disease; these results have previously been described (Houston et al., 1977a). The results from these patients will be included in this chapter. In addition a detailed report of the findings in 17 patients with complete transposition of the great arteries has been published separately (Houston et al., 1978a).

The scanner was initially fitted with a 2.5 MHz transducer and the display consisted of only 32 ultrasound lines. It became apparent that the initial design of the system rendered it unable to produce images of diagnostic quality in most infants. However further developments, with the introduction of the 3.5 MHz and

5 MHz focussed transducers, the improved 128 line/cycle display and the potential for greater magnification, greatly increased its diagnostic power in small children. Thus the findings in the neonates will include only those obtained with the 5 MHz transducer and the early unsuccessful studies with the 2.5 MHz one will be omitted. When the author started to perform two-dimensional echocardiographic studies in 1975 the examination was based on previous reports of the longitudinal view of the left ventricle (Kloster et al., 1973; Sahn et al., 1974a) and on two which disagreed on the appearances in a high transverse view (Sahn et al., 1974a; Henry et al., 1975). As experience was gained in the clinical use of the equipment, modifications in the application of these basic views and the introduction of others further increased its diagnostic potential. It thus became apparent that an optimal view of the great arteries or endocardial cushions requires positioning and angulation of the transducer to provide scanning planes which had not been developed when the early studies were performed. Subsequent experience demonstrated that the supravalvar course of the aorta could be seen in a high longitudinal view (Houston et al., 1978a), and that the junction of the atrial and ventricular septa and anterior atrioventricular valves was clearly demonstrated in the four chamber view (Houston et al., 1978b; Silverman and Schiller, 1978). Since these views were not used in the early studies and were therefore not always recorded, significant observations relating to them will not apply to all patients. The observations were made on patients in whom a definitive diagnosis was obtained by cardiac catheterisation and angiocardiography or autopsy. Where

possible the echocardiographic examination was performed and an assessment reached before angiocardiography. However, many of the older patients had undergone previous cardiac catheterisation. The author's clinical commitment included the management of these patients when they attended the cardiac out-patient clinic or were admitted to the wards and it was therefore impractical to perform the echocardiographic examination before reading their case notes. In these children the echocardiographic examination was carried out with prior knowledge of the angiocardiographic findings with the intention of establishing the echocardiographic appearances for the structural abnormality and confirming (or questioning) the previous diagnosis. Furthermore in some other patients it was not possible to perform echocardiography before angiocardiography for technical (e.g. scanner repairs or modifications) or other (e.g. absence of the operator) reasons. When possible confirmation of the exact anatomical abnormality was sought from information gained at surgery or autopsy. If the echocardiographic findings had been incorrectly interpreted the videotape record was reviewed and, where possible, the patient re-examined in an attempt to explain and correct the misinterpretation.

In the initial part of the study nine patients with complete transposition of the great arteries, four with double outlet right ventricle, four with tetralogy of Fallot, one with persistent truncus arteriosus, and one with primitive ventricle attended the hospital at the author's request specifically for the echocardiographic study. With these exceptions the examination was performed on patients attending the cardiology out-patient

clinic or admitted to the Royal Hospital for Sick Children for cardiac investigation or treatment. It cannot, therefore, be a comprehensive work on the findings in complex congenital heart disease but includes most of the common abnormalities.

IV, 1 COMPLETE TRANSPOSITION OF THE GREAT ARTERIES

Thirty seven infants and children with complete transposition of the great arteries (CTGA) were studies in two sub-groups. Initially 10 patients aged four weeks to 16 years in whom the diagnosis was known were investigated with the 2.5 MHz transducer. This study was undertaken to use the high transverse view to confirm published reports of recognition of the parallel great arteries characteristic of the transposition complexes (Henry et al., 1975) and to try to identify the main pulmonary artery (MPA) by the visualisation of its branches. The findings in these 10 patients and 4 newborn infants in whom the diagnosis was unknown at the time of echocardiographic examination have previously been described (Houston et al., 1977a). When the aorta and MPA could be identified directly in a high longitudinal view a further group consisting of two of these children and another 23 infants and children less than 2 years old was studied with the 3.5 MHz or 5 MHz transducer in an attempt to make this identification; the preliminary results of the study have also been reported (Houston et al., 1978a). The examination was carried out prior to cardiac catheterisation and angiocardiography in 15 newborn infants less than 13 days old and one infant aged 12 weeks, and following angiocardiography in 9 children between the ages of 2 weeks and 20 months; their weights ranged from 2.2 kg to 8.7 kg.

Results

In all cases the appearance in the standard longitudinal view was similar to the normal. The ventricular septum separated the right and left ventricles and was continuous with the anterior

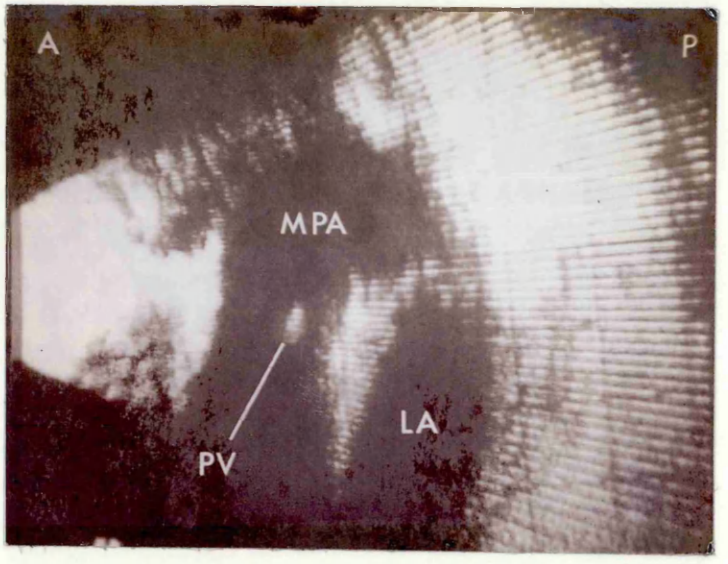


Figure 4, 1 Longitudinal view from a 13 day old infant with complete transposition of the great arteries. The main pulmonary artery rises from the left ventricle and turns posteriorly immediately above the valve cusps. PV, pulmonary valve cusps. Other abbreviations as before.

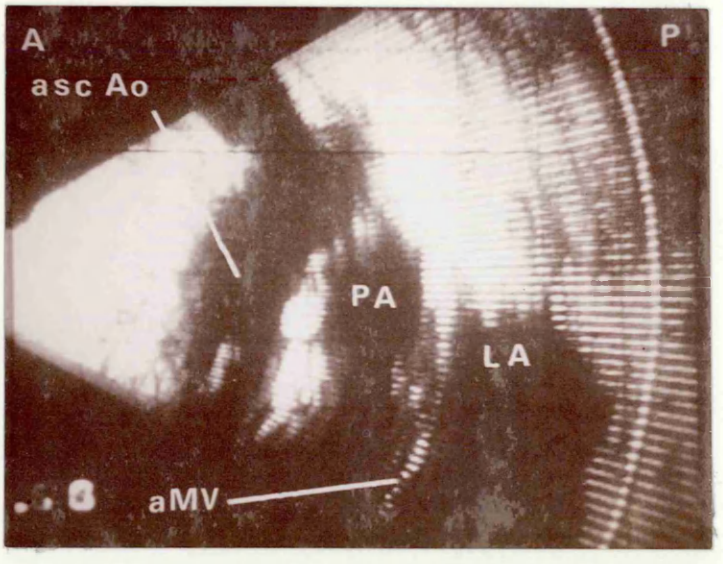
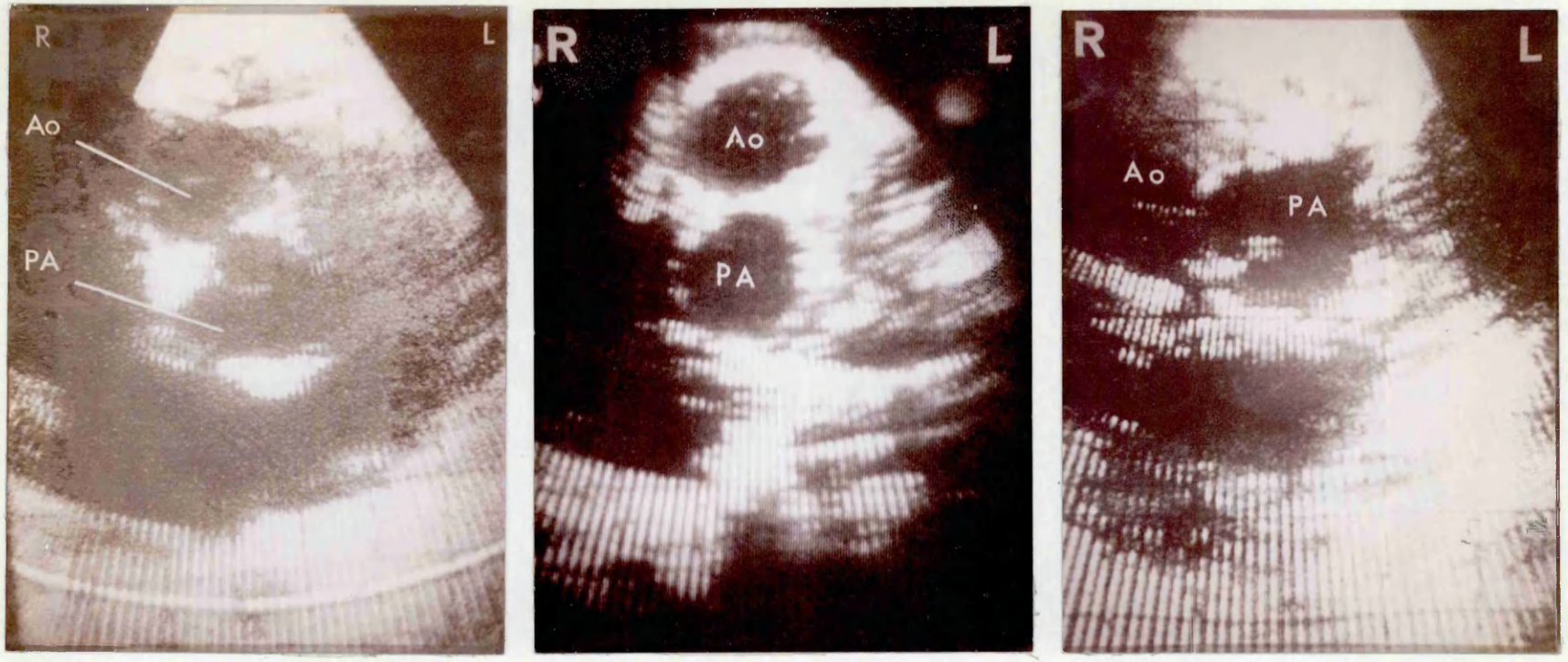


Figure 4, 2 Longitudinal view from a 10 day old infant with complete transposition of the great arteries. The anterior aorta turns posteriorly above the main pulmonary artery which is sectioned obliquely. The relations of the great artery root are shown in Figure 4, 3a. Abbreviations as before.

wall of the great artery arising from the left ventricle. The mitral valve leaflets were normally situated, the anterior leaflet being in continuity with the posterior wall of this great artery root. At the level of the semilunar valve the appearance of the origin of this great artery (the MPA) was indistinguishable from that of a normal aortic root. In the initial study the supravalvar part of the great artery was not sought. In all patients in the second group tilting or moving the scanner upwards to obtain the high longitudinal view demonstrated that the supravalvar part of the great artery arising from the left ventricle turned posteriorly almost immediately beyond the valve (Figure 4,1). The position of the other great artery origin was determined from the high transverse view (see below) and a longitudinal view of it obtained; this showed it to have a variable upward course but in all cases to turn posteriorly at a higher level than the MPA (Figure 4,2). Both great arteries could be clearly shown in a single longitudinal view only in patients where the spatial relationship between the great arteries was directly antero-posterior.

The low transverse view demonstrated the ventricular septum and both ventricles and atrioventricular valves, an appearance indistinguishable from patients with normally related arteries. In the high transverse view the origin of the great artery arising from the left ventricle was shown as a circular space with a central linear cusp echo in diastole, but the anterior crescentic space representing the crossing of a normal right ventricular outflow (RVO) and MPA could not be demonstrated in any case. In all patients modifying the plane of scan (usually to a more



(a) (b) (c)

Figure 4, 3 High transverse views in complete transposition of the great arteries from infants aged 10 days, 1 month, and 13 days respectively. (a) The aorta lies anterior and to the right of the pulmonary artery; (b) the aorta lies directly anterior to the pulmonary artery; (c) the great arteries lie side-by-side. Abbreviations as before.

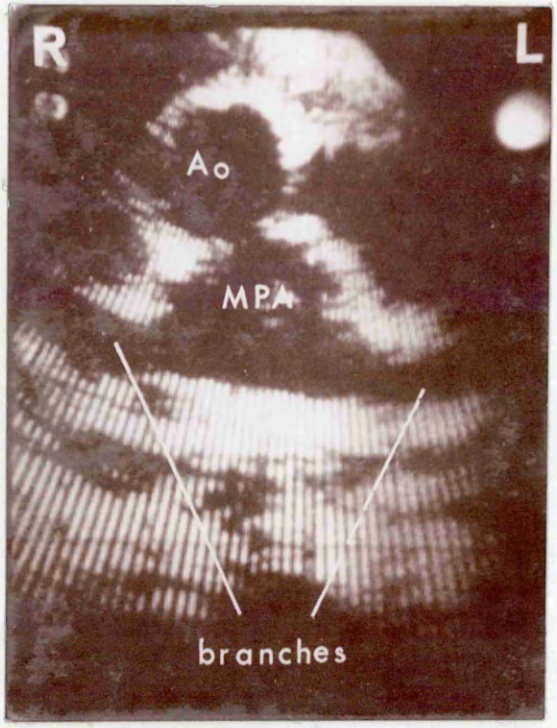


Figure 4, 4 High transverse view from the infant in Figure 4, 3b with the scanning plane tilted upwards to show branching of the pulmonary artery. Abbreviations as before.

directly transverse position) demonstrated another circular space (Figure 4, 3) which was confirmed as representing a great artery origin by the presence of a central linear cusp echo present in diastole but disappearing in systole. The demonstration of the great artery roots as two circular spaces in a transverse scan indicated that the arteries did not cross but lay parallel at their origin. The relationship of the arteries determined from this view corresponded with that found at angiocardiography; in 30 the aorta was anterior to the right of the MPA (Figure 4, 3a) in 6 directly anterior (Figure 4, 3b) and in one side by side to the right (Figure 4, 3c). At this point the scanner was tilted towards the head and in 24 patients the artery taking origin from the left ventricle was demonstrated to give rise to an elongated transverse echo-free space which, on the right side, passed posterior to the aorta (Figure 4, 4); this was considered to represent the bifurcation of the pulmonary artery and to confirm that the artery arising from the left ventricle was the main pulmonary artery.

The four chamber view was introduced after the initial studies had been performed and was used in only the 14 most recent examinations. In all 14 infants the appearance was similar to normal with continuity of the atrial and ventricular septa and separate anterior atrioventricular valves, the right one being antero-inferior to the left.

Discussion

In complete transposition of the great arteries there is atrial situs solitus, atrioventricular concordance with normally positioned ventricles and transposition of the great arteries

(Shinebourne et al., 1976); the aorta arises from the right sided morphological right ventricle and the main pulmonary artery from the left sided morphological left ventricle. With rare exceptions the great arteries lie parallel at their origins and do not cross. The most common relationship of the great artery roots in CTGA is that of an anterior right aorta and posterior left MPA (Lev et al., 1961). Criteria for echocardiographic diagnosis of CTGA have been based largely on the demonstration of this abnormal relationship of the semilunar valves with M-mode echocardiography (Gramiak et al., 1973; Dillon et al., 1973) or of the origins of the great arteries with ECG gated B-scanning (King et al., 1973) or real-time two-dimensional systems (Sahn et al., 1974a; Henry et al., 1975; Maron et al., 1975). These techniques do not identify the great arteries individually but suggest the identity of the aorta (or its valve) by its anterior right position relative to the pulmonary artery. But in 21 percent of cases of CTGA this relationship does not hold (Nadas and Fyler, 1972). Attempts have been made to distinguish the great arteries with M-mode echocardiography by comparing systolic time intervals (Solinger et al., 1974; Fouron et al., 1976) or the outside diameters of the great artery roots (Solinger et al., 1974) or most recently by using contrast echocardiography (Mortera et al., 1977). Measurement of electromechanical systole, ventricular ejection times and pre-ejection periods allows the great arteries to be differentiated, but only when the pulmonary vascular resistance is low (Solinger et al., 1974; Fouron et al., 1976). Although the echocardiographic measurement of the outside diameter of the pulmonary artery root has been reported to be

larger than that of the aortic root in normal infants (Solinger et al., 1973; Hagan et al., 1973) this observation is likely to be of little practical value in differentiating the great arteries in CTGA, where in those with no ventricular septal defect or pulmonary stenosis Shaher (1973) found them to be equal in 54 percent. Mortera et al. (1977) used contrast echocardiography with injection into a systemic vein and suprasternal positioning of the transducer to show both great arteries simultaneously with M-mode echocardiography. With this approach the artery nearer the probe is the aorta and in patients with an intact ventricular septum and CTGA this was shown to opacity more intensely, suggesting that it arose from the right ventricle. This technique was considered to be ineffective when there was a large ventricular septal defect.

Two-dimensional echocardiography, with a transverse scan, can demonstrate the parallel great arteries characteristic of the transposition complexes. Furthermore, Sahn et al. (1974a) suggested that in a longitudinal scan the long retrosternal course of the aorta might allow it to be recognised, but the pulmonary artery was not positively identified. A more universally applicable technique was therefore required to allow reliable diagnosis of CTGA with echocardiography. Furthermore, to assess the great artery anatomy in more complex forms of congenital heart disease it is essential that the great arteries be identified individually. The initial finding that in CTGA the high transverse view could be used to demonstrate the bifurcation of a great artery and thus identify it as the main pulmonary artery provided such diagnostic information but in approximately one third of cases (13 out of 37)

this could be convincingly shown for the purpose of precise diagnosis. The high longitudinal view has allowed differentiation of the aorta from the MPA in all 25 patients in whom this was sought. In accord with the suggestion of Sahn et al. (1974a) the long retrosternal course of the aorta has been demonstrated, but this course is subject to variation. To prove that this artery is the aorta it should be shown to turn posteriorly at a level above the MPA, which can be independently identified by its posterior course immediately beyond the pulmonary valve. Continuity between the anterior leaflet of the left atrioventricular valve and the great artery implies that the valve is the mitral valve and that the left sided ventricle is the morphological left ventricle (Shinebourne et al., 1976). Recognition that the MPA arises from the left ventricle and aorta from the right ventricle allows definitive echocardiographic diagnosis of CTGA. In all 16 infants with CTGA examined before cardiac catheterisation and angiocardiography the diagnosis was reached and the anatomy of the proximal great arteries correctly determined by echocardiography.

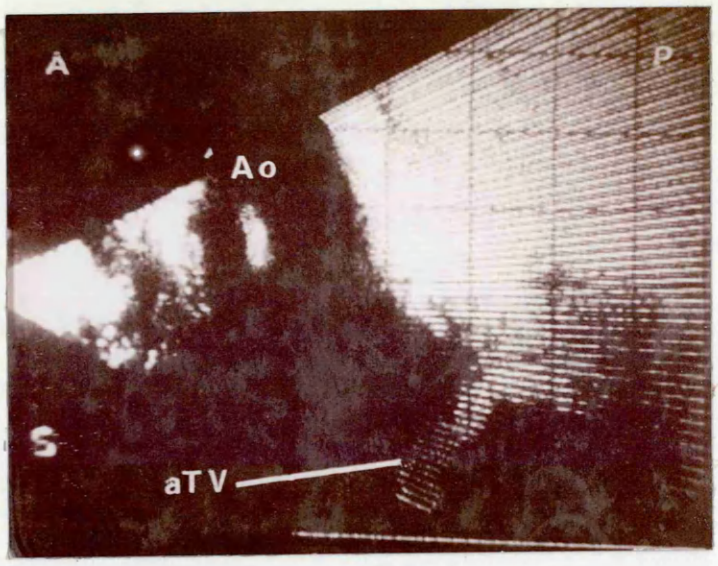


Figure 4, 5 Longitudinal view from a child with corrected transposition. The posterior wall of the aorta is not continuous with the anterior leaflet of the tricuspid valve. aTV, anterior tricuspid valve leaflet. Other abbreviations as before.

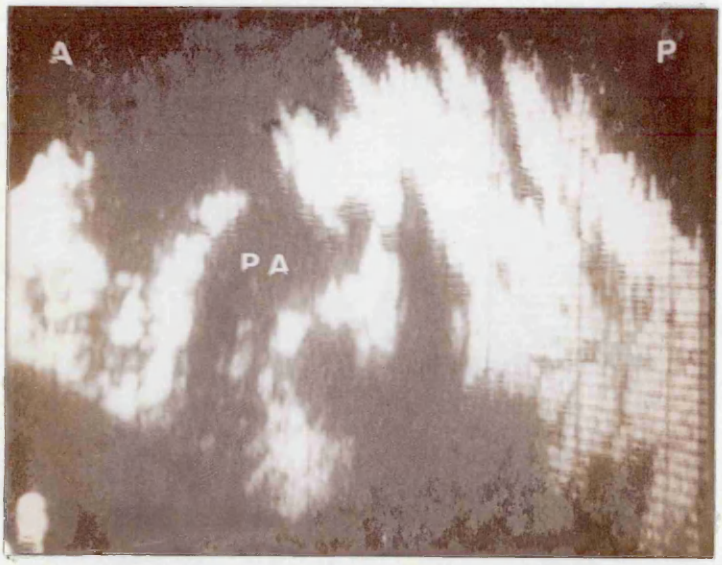


Figure 4, 6 Longitudinal view from a child with corrected transposition. The posterior wall of the pulmonary artery is continuous from the anterior leaflet of the mitral valve. Abbreviations as before.

IV, 2 CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Four children (aged 5 to 11 years) with corrected transposition of the great arteries were examined following angiocardiographic diagnosis. All were initially studied before the four chamber view had been developed and one was re-examined after its introduction.

Results

The aortic root lay in an anterior position in all patients, and to demonstrate it the standard longitudinal plane had to be modified to lie to the left of, and almost parallel to, the left sternal edge. In two patients considerable difficulty was experienced in obtaining a clear image of the aortic root which was best seen when the patient held his breath in expiration. The posterior aortic wall was continuous with echoes from tissue showing no intrinsic movement throughout the cardiac cycle but which, in turn, was continuous with a structure showing the movement characteristic of the anterior leaflet of an atrioventricular valve (Figure 4,5). In two the ventricular septum was not clearly seen in this scanning plane. A longitudinal view of the other great artery was obtained by determining its position from the high transverse view (to the right and posterior as described below) and rotating the scanner until the scanning plane lay along the great artery root. This demonstrated that the posterior wall was continuous with the anterior leaflet of a right sided atrioventricular valve and identified the vessel as the main pulmonary artery (MPA) by its posterior turn at a lower level than the left sided great artery (Figure 4,6). Thus the right sided MPA was continuous with the right sided atrioventricular valve and the left sided aorta was

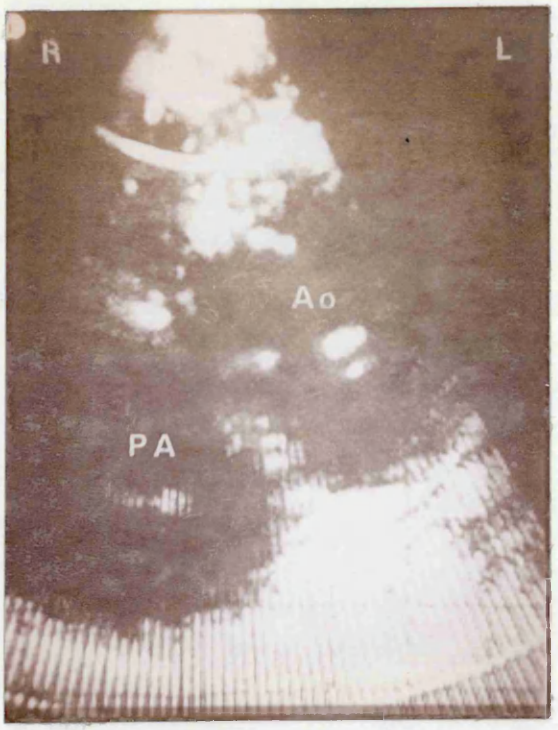


Figure 4, 7 High transverse view through the great artery roots from a child with corrected transposition. The aorta lies anterior and to the left of the pulmonary artery (l-transposition). Abbreviations as before.

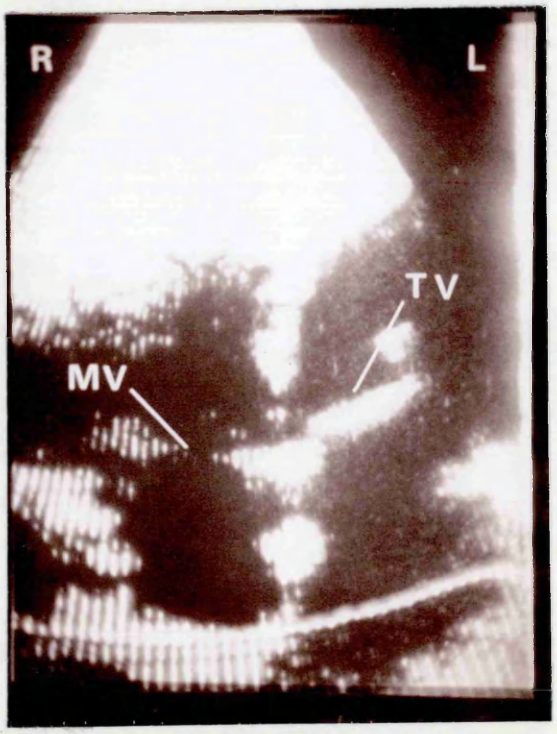


Figure 4, 8 Four chamber view from a child with corrected transposition. The tricuspid valve lies on the left side of the ventricular septum and impinges on it in an antero-inferior position with respect to the mitral valve. Abbreviations as before.

discontinuous with the left sided atrioventricular valve.

The low transverse view showed the cusps of both atrioventricular valves. In two patients, the left atrioventricular valve appeared to have three separate cusps but in the other two cases only two cusps could be clearly demonstrated. In three patients the orientation of the ventricular septum was more directly antero-posterior than in the normal heart and in two it was only shown with difficulty by moving the transducer to the left and tilting the scanning plane to the right. In the fourth patient the position of the ventricular septum was indistinguishable from normal. In all patients the high transverse view demonstrated two circular spaces with central cusp echoes (confirming that they were great artery roots) with the left one (the aorta from a longitudinal view) just anterior to the other (Figure 4,7).

The four chamber view had not been developed when the four patients were initially examined. However, after it had been introduced the author had the opportunity to re-study one of them. The ventricular and atrial septa were in continuity and the anterior leaflet of the left atrioventricular valve (the tricuspid) was attached to the ventricular septum anterior to that of the right atrioventricular valve (the mitral) (Figure 4,8).

Discussion

In corrected transposition of the great arteries there is atrial situs solitus, atrioventricular discordance and transposition of the great arteries. The MPA rises from a right sided morphological left ventricle and aorta from a left sided morphological right ventricle. In addition the great arteries do not cross but lie parallel at their origin, the aorta being positioned to the left

of the MPA (L-transposition). The echocardiographic finding of continuity of the anterior leaflet of the right sided atrioventricular valve with a great artery (the MPA) indicates that the ventricle has no infundibulum, and suggests that the valve is the mitral valve and thus that the right sided ventricle is a morphological left ventricle (Shinebourne et al., 1976). Similarly discontinuity between the left sided atrioventricular valve and the aorta points to the presence of an infundibulum, with the valve the tricuspid and the left sided ventricle a morphological right ventricle (Shinebourne et al., 1976). Similar findings to these have been reported based upon M-mode echocardiography (Rey and Lablanche, 1976; Beardshaw et al., 1976). However there is a spectrum of mitral aortic separation from 0 to 7 mm in the normal heart (Rosenquist et al., 1976) and this can be even greater (up to 11 mm) in patients with discrete subaortic stenosis (Rosenquist et al., 1979). Furthermore in some malformed hearts the morphological left ventricle may possess an infundibulum (Van Praagh and Van Praagh, 1967), and it is therefore important to confirm the identity of atrioventricular valves by other features. The low transverse view provides direct visualisation of the orifices of the atrioventricular valves but in only two cases could three separate cusps of the left sided tricuspid valve be visualised with certainty. Furthermore, in corrected transposition of the great arteries the left atrioventricular valve (tricuspid) can show considerable morphological variation and may not be clearly divided into three separate cusps (Schiebler et al., 1961). In the patient studied with the four chamber view the left atrioventricular valve (tricuspid) was shown to lie

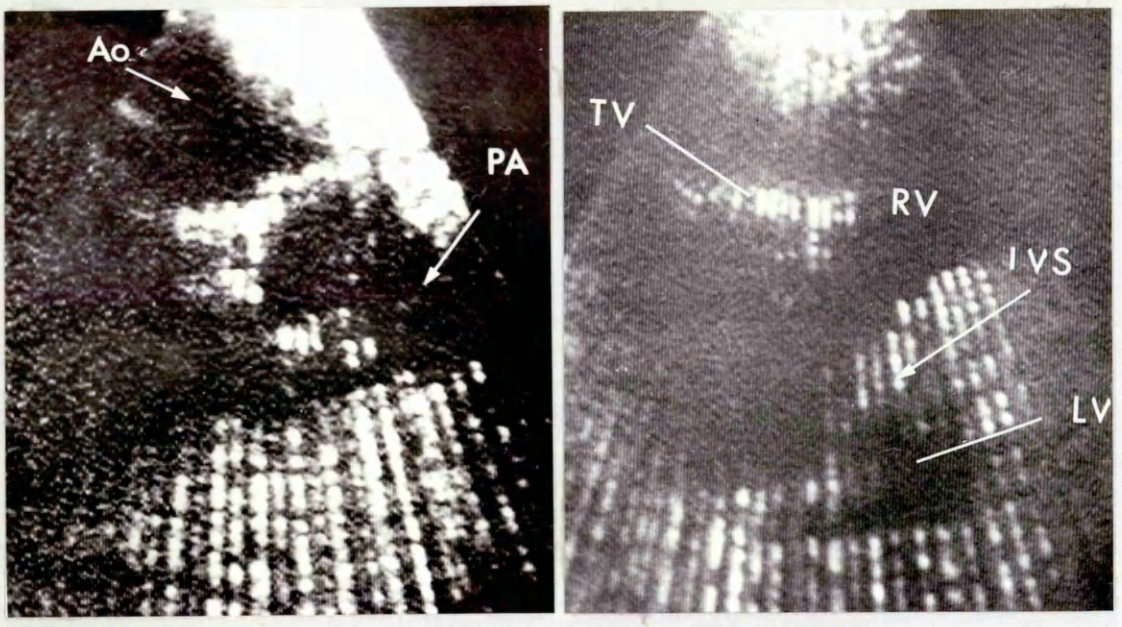
antero-inferior to the right (mitral). Thus the antero-inferior position of the tricuspid valve clearly demonstrated in the normal heart may hold where there is ventricular inversion and allow the left atrioventricular valve to be identified as the tricuspid and thus the left sided ventricle as the morphological right ventricle. Tajik et al. (1978) suggested that this might be true but showed no echocardiographic or pathological data to confirm this and further studies are therefore required to ascertain its validity.

DOUBLE OUTLET RIGHT VENTRICLE

Five patients with an angiographic diagnosis of double outlet right ventricle (DORV) were examined. Their ages ranged from 1 to 13 years. In 4 examined before the high longitudinal view had been developed the aorta lay anterior and to the right of the pulmonary artery, while in the other patient the pulmonary artery lay anterior and just to the left of the aorta.

Results

The standard longitudinal view showed a great artery root, the ventricular septum and the left atrioventricular (mitral) valve; the left ventricle was relatively small. In three patients the anterior mitral leaflet was continuous with the great artery and in the other two this continuity was interrupted by intervening tissue. When this great artery root and the ventricular septum were viewed simultaneously their relationship was seen to vary throughout the cardiac cycle, the septum moving posteriorly with respect to the great artery root during systole. The diastolic position was considered to give the best indication of the relationship of the great artery to the ventricles; the upper ventricular septum was posterior to the mid point of the great artery root in 3 patients and anterior in the other 2. In the only patient examined with the high longitudinal view the long upward course of this great artery identified it as the aorta. The other great artery (which lay anterior and to the left in the high transverse view) turned posteriorly at the lower level, confirming it was the main pulmonary artery.



(a) (b)

Figure 4, 9 Transverse views from a child with double outlet right ventricle obtained as the scanning plane is tilted downwards from the level of (a) the great artery roots to (b) the ventricular septum. As this manoeuvre is performed it can be appreciated that the great artery roots lie anterior to the ventricular septum and therefore take origin from the right ventricle. IVS, interventricular septum. Other abbreviations as before.

The appearance in the low transverse view was normal except for a relatively small left ventricle. In all five patients high transverse views showed two circular spaces with central cusp echoes in diastole indicating the valves of parallel great arteries. In all patients the artery seen in the standard longitudinal view was posterior, the other lying anterior and to its right in four but anterior and just to its left in one. Tilting the scanning plane downwards from the high transverse view towards the low transverse view brought the ventricular septum into view. In all patients the anterior great artery appeared to be anterior to the septum (indicating that it took origin from the right ventricle) but in only three was more than half the other (posterior) great artery anterior to the ventricular septum in diastole (Figure 4,9).

Discussion

For the diagnosis of double outlet right ventricle as defined by Lev et al. (1972) more than one-and-a-half great arteries should arise from the right ventricle. Echocardiographic examination showed that the relationship of the great arteries to the ventricular septum varied throughout the cardiac cycle. It was considered that the position in diastole was likely to show the closest agreement with findings at post mortem or surgery and was used in the assessment of their relationship. In two of the patients more than half of one great artery appeared to be posterior to the septum, the inferior margin of the ventricular septal defect being used in this assessment. Subsequent review of the material has however led to the suggestion that the superior and lateral margins of the septal defect may have been

posterior to both great arteries (Houston et al., 1977a). Such margins should, however, have been visualised in the sweep downward from the great artery roots to the low transverse view, as exemplified in Figure 4,9. Using a similar technique Henry et al. (1977) were able to demonstrate that the centre of both great arteries was anterior to the ventricular septum in all 5 patients with DORV where satisfactory echocardiograms were obtained. In the two present patients in whom this was not demonstrated the angiocardiographic studies had been carried out some years before the echocardiography and subsequent review of the angiocardiograms showed that it was difficult to be certain that the diagnosis was DORV, and further angiocardiographic studies may confirm the echocardiographic findings.

The demonstration of discontinuity between the mitral valve and posterior semilunar valve with M-mode echocardiography has been suggested as a diagnostic criterion for double outlet right ventricle (Chesler et al., 1971), but a post mortem study has, in a minority of cases, found continuity (Cameron et al., 1976). However, as has already been discussed under corrected transposition of the great arteries, there is a spectrum of mitral aortic discontinuity even in the normal heart and discontinuity between the left atrioventricular valve and the great artery is found with corrected transposition of the great arteries and other more complex abnormalities (Van Praagh and Van Praagh, 1967). Whilst in corrected transposition of the great arteries the anterior position of the great artery should prevent confusion between the two abnormalities, such a distinction may not always be possible with echocardiography in other anomalies.

Thus the recognition that, in diastole, at least one-and-a-half great arteries are anterior to the ventricular septum should be diagnostic of double outlet right ventricle, while the demonstration of discontinuity between the mitral valve and the posterior great artery root would merely be supportive of the diagnosis.

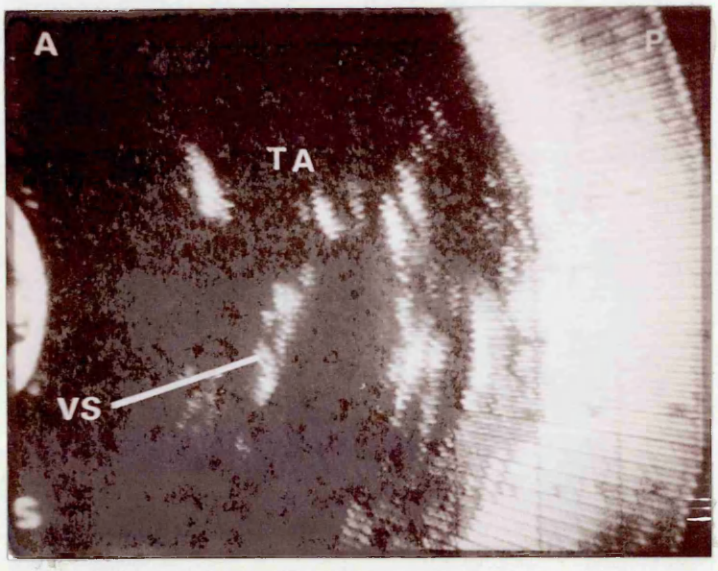


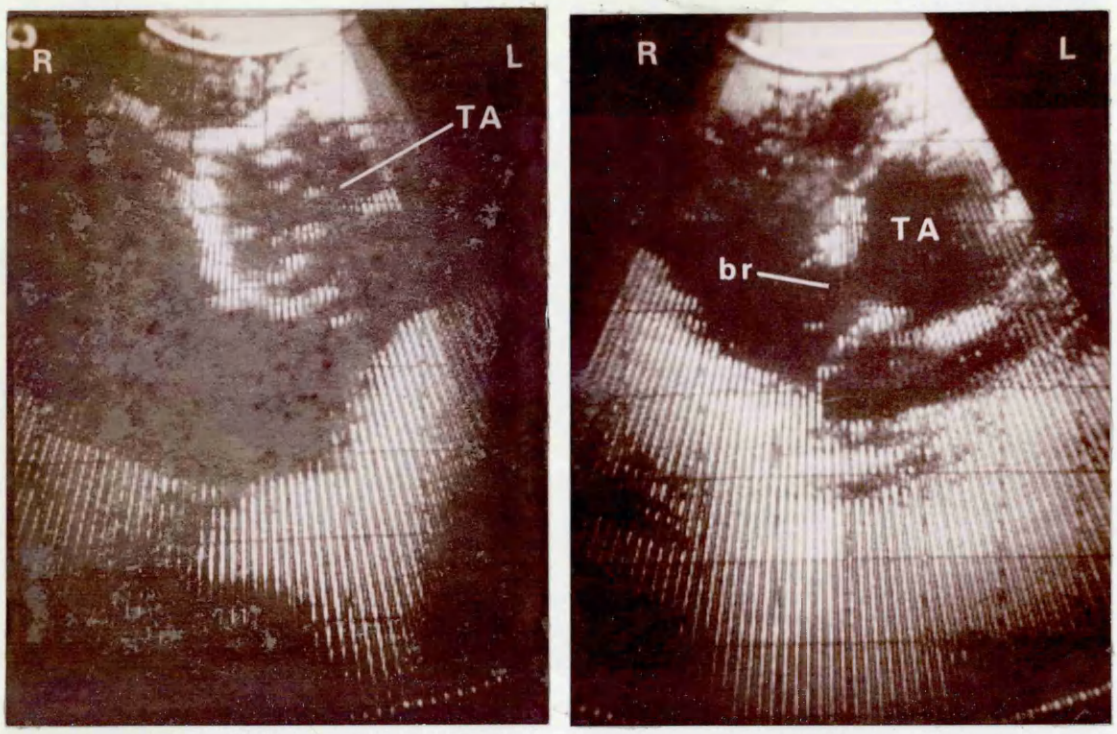
Figure 4, 10 Longitudinal view from a one day old infant with a persistent truncus arteriosus. The great artery root over-rides the ventricular septum and multiple echoes are recorded from its valve cusps. TA, truncus arteriosus. Other abbreviations as before.

PERSISTENT TRUNCUS ARTERIOSUS

Three patients (ages 1 day, 8 weeks and 15 years) with an angiocardiographic diagnosis of persistent truncus arteriosus were examined. Both infants were examined prior to angiocardography. The angiocardiographic diagnosis in the 15 year old was Type II truncus (pulmonary artery branches arising separately from the posterior wall) (Collett and Edwards, 1949) with pulmonary branch stenosis, and in the 8 week old was Type I (main pulmonary trunk arising from the posterior wall); in the day old infant it was uncertain whether the abnormality was Type I or II. Autopsy confirmed the diagnosis in both infants and showed the day old to have Type II truncus. Both had 4 truncal valve cusps; all were of approximately equal size in the 8 week old infant and 3 were of approximately equal size with a small posterior right cusp in the other.

Results

In all patients the standard longitudinal view showed the ventricular septum, both ventricles and mitral valve leaflets. The anterior mitral leaflet was continuous with the posterior wall of the great artery. The upper ventricular septum and anterior wall of the great artery were discontinuous and moved asynchronously; in diastole the position of the septum approximated to the mid point of the great artery (Figure 4,10). In the newborn infant unusual echoes were apparent behind the posterior wall of the great artery just above valve level; these could not be identified with certainty as representing a great artery or arterial branches. The high longitudinal view had not been developed when the 15 year old patient was studied. In both



(a) (b)

Figure 4,11 High transverse views from the infant in Figure 4, 10 with a type II persistent truncus arteriosus, obtained as the scanning plane is tilted upwards from the level of (a) the truncal valve, which can be seen to have four cusps, to (b) the origin of the pulmonary artery branches. br, right pulmonary artery branch. Other abbreviations as before.

infants it showed the great artery to follow a long upward course typical of the aorta.

The low transverse view showed the ventricular septum, both ventricles and both atrioventricular valves. In all three patients the high transverse view showed a single large circular great artery and no evidence of a crossing or parallel artery. In the older patient a single central cusp echo was not apparent, but double central echoes and several eccentric ones suggested the presence of multiple truncal valve cusps. This examination was performed with a 2.5 MHz transducer on the 32 line display and it was not possible to identify the valve cusps in any greater detail. In both infants in diastole there were atypical cusp echoes which suggested there were more than three, probably four, cusps (Figure 4,11a). In the day old infant tilting the scanning plane headwards showed two branches arising from the posterior wall of the great artery (Figure 4,11b) which could be followed further upwards; these were considered to be the right and left pulmonary arteries. In the other two patients branching of the great artery could not be shown with certainty.

Discussion

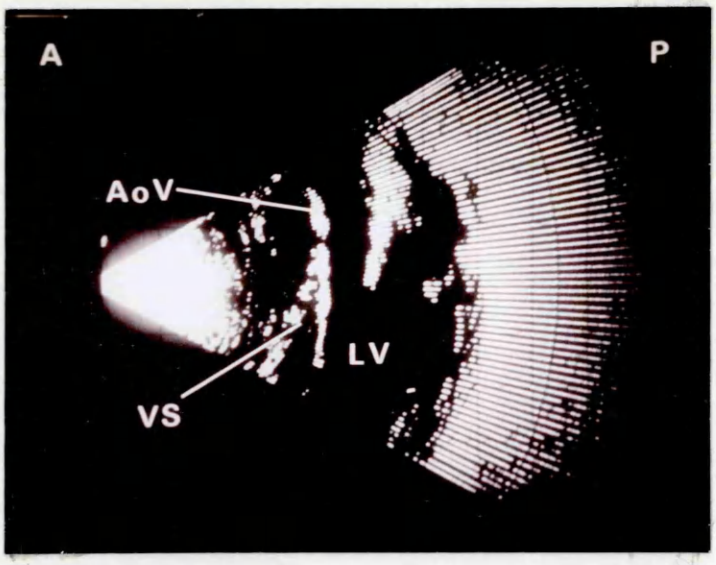
Persistent truncus arteriosus implies persistence of the fetal common arterial trunk with a single great artery arising from the heart, receiving the outflow from both ventricles and supplying the coronary, systemic, and pulmonary circulations. In Type I truncus a single pulmonary artery takes origin from the base of the trunk and then bifurcates to form the pulmonary arteries and in Type II the pulmonary arteries arise independently from the common arterial trunk (Collett and Edwards, 1949). The number of

semilunar valve cusps varies from 2 to 6; these are often thickened and of an abnormal appearance.

With M-mode echocardiography it has been suggested that demonstration of an overriding aorta and failure to show the normal pulmonary valve echo may allow the diagnosis of persistent truncus arteriosus to be made (Chung et al., 1973a). However, in severe tetralogy of Fallot or pulmonary atresia with a ventricular septal defect the pulmonary valve echo cannot be recorded and it has subsequently been reported that this criterion is inadequate for the positive diagnosis of truncus arteriosus with M-mode echocardiography alone, although its association with a large left atrial diameter would be strongly suggestive of the diagnosis (Assad-Morell et al., 1976). Even if only one great artery root has been recorded it is conceivable that another is present but has merely escaped detection; in these circumstances it is essential to confirm or rule out the possibility of persistent truncus arteriosus. Sahn et al. (1974a), using a multiple element two-dimensional system, have suggested that a transverse view of the great artery root showing multiple and unusual cusp echoes may be characteristic of persistent truncus arteriosus and this appearance was confirmed in the 15 year old patient. In the two infants more than 3 valve cusps (probably 4) were identified and subsequently confirmed at post mortem examination. A transverse view of a normal aortic root with two-dimensional echocardiography usually shows the valve as a single linear echo in diastole although three cusps may be recognised. In the infants the visualisation of the four separate cusps may have been made possible by their thickening and abnormal orientation. The recognition of more

than three semilunar cusps would be diagnostic of persistent truncus arteriosus but this is not possible in approximately 75% of cases where there is a bicuspid or tricuspid truncal valve (Bharati et al., 1974). In both infants the long upward course of the great artery suggested it was an aorta but in one the transverse view showing two branches just above cusp level indicated that this was not an aorta but a Type II persistent truncus arteriosus. This feature may allow the diagnosis of persistent truncus arteriosus to be reached in cases where abnormal semilunar cusp echoes cannot be identified. In the infant in whom the pulmonary artery branches were not clearly demonstrated the pulmonary artery arose from the truncus immediately above the valve. Because the infant was ill and tachypnoeic the examination was performed as rapidly as possible and the baby died before the echocardiogram could be repeated. It may be that a more assiduous examination technique with particular regard to the transverse view of the great artery just above valve level could have demonstrated the pulmonary artery in this infant and may improve the recognition rate of the pulmonary artery branches in future.

(a)



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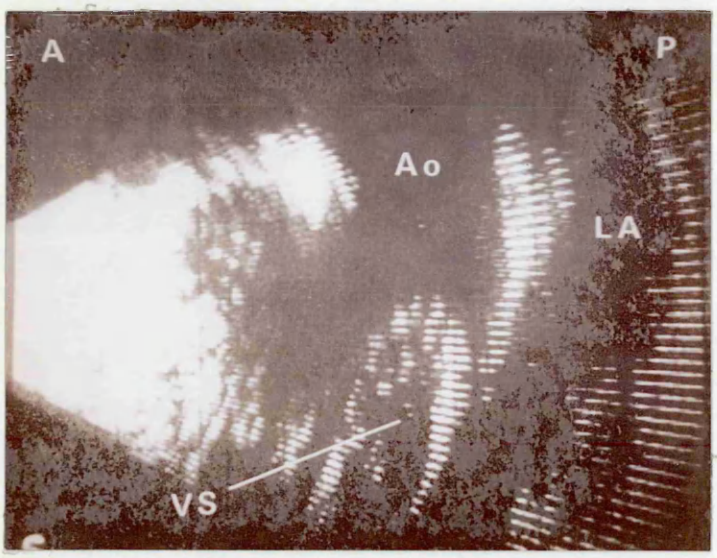


Figure 4, 12 Longitudinal views from children with tetralogy of Fallot showing the aorta over-riding the ventricular septum. In (a) the ventricular septum appears to be anterior to the mid point of the great artery root, while in (b) their exact relationship is difficult to ascertain. Abbreviations as before.

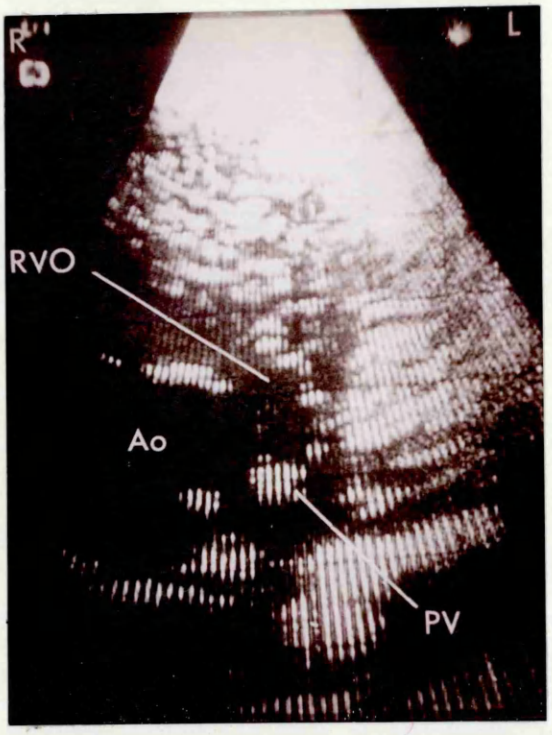


Figure 4, 13 High transverse view from an infant with tetralogy of Fallot and severe infundibular stenosis. The very narrow right ventricular outflow crosses the aortic root in the normal manner. The lateral walls of the aortic root were recognisable in the moving display but are not clearly seen in this still frame. Abbreviations as before.

TETRALOGY OF FALLOT

Twenty-one children (aged 6 weeks to 17 years) with a cardiac catheterisation and angiographic diagnosis of tetralogy of Fallot were studied. Examination was carried out before angiocardiography in 10 and following it but before total surgical correction in 11.

Results

The standard longitudinal view, in all cases, demonstrated the ventricular septum, both ventricles and the mitral valve leaflets; the anterior mitral leaflet showed normal continuity with the posterior wall of the aorta. In 20 the upper ventricular septum and anterior wall of the aorta were discontinuous, the anterior aortic wall being anterior to the septum throughout the cardiac cycle (Figure 4, 12); the separation was greatest during ventricular systole, the septum showing little movement while the aorta moved anteriorly. In all patients the mid-point of the upper ventricular septum in diastole appeared to be anterior to the central position of the aortic root. In one patient the ventricular septum and anterior wall of the aorta showed apparent continuity in diastole but separated in systole, the aorta moving forward.

The low transverse view showed the ventricular septum, both atrioventricular valves and both ventricles, the left being relatively small. The high transverse view demonstrated the normal appearance of the aortic root as a circular echo-free space with a central cusp echo in diastole. The right ventricular outflow (RVO) and main pulmonary artery (MPA) were normally related to the aorta (Figure 4, 13); in 4 patients they were more clearly demonstrated in the view directed above the left

ventricular outflow. Pulmonary valve echoes were recognised in 18 but in the remaining 3 could not be identified with certainty. In 12 of the 18 patients where the pulmonary valve echoes could be clearly seen these echoes remained in the same position throughout the cardiac cycle suggesting little movement of the cusps. In the other 6 patients the pulmonary valve echoes disappeared in systole, indicating relatively mobile cusps. In 20 patients there appeared to be a generalised narrowing of the RVO, pulmonary valve ring and the proximal MPA, all being of approximately the same size (Figure 4, 13); the ratio of the diameters of the RVO to the aorta at valve level ranged from 0.18 to 0.57 (mean 0.40). In the remaining patient, in whom there was apparent aortic-septal continuity in diastole, the MPA was the same size as the aorta and the pulmonary valve cusps showed movement indistinguishable from normal.

Discussion

In the tetralogy of Fallot there is a dextroposed aorta over-riding the ventricular septum with a large ventricular septal defect and a narrowed infundibulum of normal or increased length (Becker and Anderson, 1977). The longitudinal view with two-dimensional echocardiography can demonstrate the over-riding aorta; in 20 of the patients the aortic-septal over-ride was seen throughout the cardiac cycle and in the other this became apparent in ventricular systole. Limitations of ultrasonic imaging are such that failure to display a structure does not mean that it is absent and thus an echocardiographic appearance of discontinuity between the aorta and ventricular septum cannot be interpreted with

certainty as a break in their continuity. However marked discontinuity between the anterior aorta and the ventricular septum and asynchronous movement between them are likely to indicate true discontinuity and thus the presence of a high sub-aortic septal defect. M-mode studies in the tetralogy of Fallot (Chung et al., 1973b), using a headward sweep, have shown the position of the echo from the anterior aorta to be anterior to that of the upper ventricular septum, suggesting discontinuity between them with an over-riding aorta. However it has been pointed out that if the transducer is placed in a high medial position the upper ventricular septum will be further from it than the anterior aorta, and rapid downwards and medial angulation may give an incorrect impression that the anterior wall of the aorta is anterior to the ventricular septum and thus over-riding it (Kotler et al., 1977). Furthermore the ventricular septum and anterior aorta cannot be imaged simultaneously with this technique and minor degrees of over-riding and asynchronous movement may not be shown. Two-dimensional echocardiography, by giving precise information on the relative positions of these structures throughout the heart cycle, should allow the presence of over-riding to be determined with greater certainty. In differentiating between tetralogy of Fallot and double outlet right ventricle it is necessary to determine whether more or less than half of the aorta takes origin from the left ventricle. This is clearly apparent in the longitudinal view in the majority of patients, but in some the ventricular septum may appear to be positioned below the mid-point of the aortic root and two-dimensional echocardiography may not then provide definitive assessment of

the ventriculo-arterial connections. This is exemplified by considering Figure 4, 12b. If a demarcating line is drawn up the centre of the ventricular septum and continued to meet the aortic root the aorta would appear to arise from the right ventricle. It might seem more appropriate to draw an approximately horizontal line to demarcate the aortic root then use another perpendicular to it at its mid point to ascertain the connections of the aorta. However it is frequently not possible to determine the position of the aortic root with complete certainty with two-dimensional echocardiography and thus any such division would be an approximation. It would therefore be inappropriate to rely upon it as a definite diagnostic criterion and in this thesis decisions as to the ventriculo-arterial connections in these cases have been made subjectively. Thus, where a diagnostic label is based on such an assessment the echocardiographic appearance (i.e. ventricular septum under approximately the mid point of the great artery root) should be described; this has the additional advantage of defining the anatomical relationship of the great artery root and ventricular septum with greater precision.

In the tetralogy of Fallot with severe infundibular and valve stenosis the pulmonary valve shows little intrinsic movement and is difficult to record with M-mode echocardiography. If this is not possible the pulmonary artery cannot be identified with this technique and a certain distinction between the tetralogy of Fallot and persistent truncus arteriosus is not then possible with echocardiography alone (Assad-Morell et al., 1976). Using a 30^{o} mechanical sector scanner Henry et al. (1975) showed that

in the tetralogy of Fallot two-dimensional echocardiography can demonstrate the narrow right ventricular outflow and main pulmonary artery and confirm that the great arteries are normally related. This is in accord with the present findings. The characteristic narrowing was apparent and the severity of this and the size of the main pulmonary artery could be estimated. In this study the degree of RVO narrowing was assessed by comparing the ratio of its diameter with that of the aortic root; in all except one of the patients this ratio was well below the normal range. However in the tetralogy of Fallot the aorta is enlarged (Morris et al., 1975) and it might be more useful in future to measure the absolute diameter of the RVO. The ratios of the diameters were measured just after performing the echocardiographic examinations on the normal subjects and those with tetralogy of Fallot and a record of every two-dimensional echocardiogram was not stored. Furthermore in others the still frame records do not clearly show both marker arcs and it is therefore not possible to obtain an absolute measurement of the size of the RVO in retrospect.

In the one patient where, according to echocardiography, RVO narrowing could not be recognised and discontinuity between the ventricular septum and the anterior aortic wall was apparent only in systole, the angiocardiographic diagnosis was made 5 years before echocardiography and a pulmonary valvotomy was performed shortly after the angiocardiographic study. The author subsequently reviewed the angiocardiogram with the radiologist. There appeared to be little RVO narrowing and, although the left ventricular injection had been recorded in only the antero-posterior and lateral planes and the exact relationship of the ventricular septum to the anterior aorta was not clearly shown, it was considered that the degree of aortic over-ride of the ventricular septum was minimal.

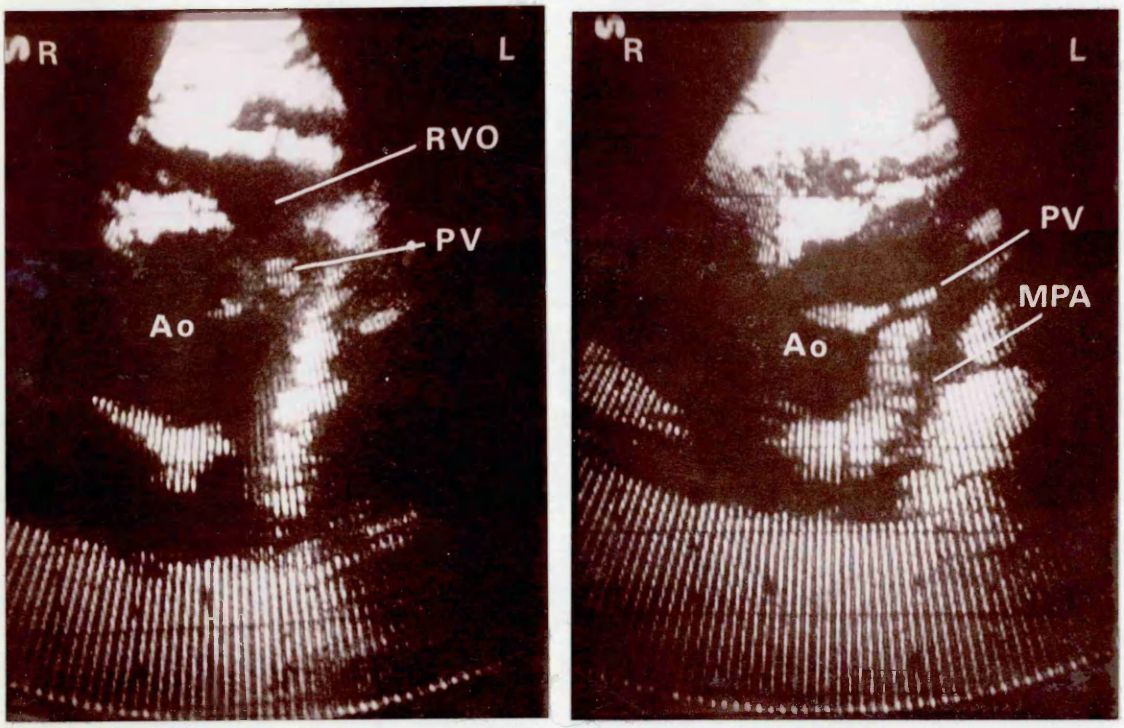
IV, 6 PULMONARY ATRESIA WITH VENTRICULAR SEPTAL DEFECT

Two patients (aged 5 days and 11 years) with Fallot type pulmonary atresia were studied. The infant was examined before cardiac catheterisation. In both, angiocardiography showed a ventricular septal defect with over-riding aorta and narrow right ventricular outflow (RVO); the pulmonary arteries filled through a patent ductus arteriosus (PDA) in the infant and via bronchial artery anastomoses in the older child. Autopsy confirmed the diagnosis in the infant; the main pulmonary artery (MPA) was shown to be hypoplastic (diameter approximately 2 mm) and the pulmonary valve to be atretic.

Results

The appearances in the longitudinal views were similar to patients with Fallot's tetralogy. A great artery, whose posterior wall was continuous with the anterior mitral valve, over-rode the ventricular septum. The long upward course of this artery was shown in the infant but not in the older child. The left ventricle and atrium were relatively small in both patients.

The low transverse view showed a small left ventricle and normally positioned mitral valve. The high transverse view showed the typical appearance of the aorta as a circular space with a central linear cusp echo in diastole. This was crossed anteriorly from right to left by a very narrow right ventricular outflow and main pulmonary artery. A persistent linear echo which moved slightly distally with ventricular systole but did not exhibit doming was considered to represent the atretic pulmonary valve. In the infant the right ventricular outflow and main pulmonary artery could not be clearly seen in a single plane but minor alterations in transducer angulation showed each individually



(a) (b)

Figure 4, 14 High transverse view from a 5 day old infant with pulmonary atresia and a ventricular septal defect. (a) The aortic root, right ventricular outflow, and pulmonary valve are clearly seen simultaneously, but (b) the main pulmonary artery is only demonstrated by adjustment of the scanning plane. Abbreviations as before.

and indicated they were continuous but separated by pulmonary valve tissue (Figure 4, 14a&b). The ratio of the RVO to aorta was 0.15 in the infant and 0.1 in the child.

Discussion

The anatomical abnormality in pulmonary atresia with VSD and hypoplastic MPA is very similar to that of severe tetralogy of Fallot. The echocardiographic findings in the two patients were almost identical to those in severe tetralogy of Fallot where pulmonary valve movement could not be seen, although the ratio of the diameter of the RVO to the aorta was smaller than in any of the patients with the tetralogy. If the echocardiographic examination was to be combined with injection of ultrasonic contrast into a peripheral vein it is theoretically possible that in severe tetralogy of Fallot the MPA could be shown to fill from the RVO. However, in the infant with pulmonary atresia the RVO and MPA were very narrow and did not lie in a single plane so they could not be demonstrated simultaneously. Thus if following contrast injection the MPA opacified it might be difficult to determine whether the contrast filled the MPA through the pulmonary valve or in a retrograde fashion from a PDA. It is thus unlikely that two-dimensional echocardiography, even with contrast echocardiography, will allow a certain distinction to be made between severe Fallot's tetralogy and Fallot type pulmonary atresia where there is a small patent distal MPA. Since the two conditions cannot be differentiated with certainty the points made in the discussion of tetralogy of Fallot apply equally to this type of pulmonary atresia. Where there is pulmonary atresia with complete atresia of the RVO and

absence of the proximal MPA the longitudinal view with echocardiography may resemble that in truncus arteriosus. The demonstration of more than three cusps or of a branch arising just above the great artery root would be diagnostic of truncus arteriosus. If these findings could not be demonstrated the diagnosis of pulmonary atresia could then only be inferred by the recognition of a relatively small left atrium as described with M-mode echocardiography by Assad-Morell et al. (1976).

IV, 7 PULMONARY VALVE ATRESIA WITH INTACT VENTRICULAR SEPTUM

Three infants (all aged less than 24 hours) with pulmonary valve atresia and an intact ventricular septum were studied; two were examined prior to cardiac catheterisation and angiocardiography and the third before the angiocardiographic information obtained at another hospital had been made known to the author. In two angiocardiography disclosed a right ventricle of approximately normal size with tricuspid regurgitation and a large right atrium, and retrograde filling of the main pulmonary artery (MPA) through a patent ductus arteriosus. The third had a right ventricle smaller than normal with a competent tricuspid valve, the ventricle emptying through myocardial sinusoids; the pulmonary artery branches filled through a small patent ductus but the MPA was not identified on angiocardiography. At operation all three infants had an imperforate pulmonary valve with a main pulmonary artery of approximately normal size.

Results

The longitudinal view in each patient showed the ventricular septum and anterior mitral leaflet which were continuous with the anterior and posterior walls respectively of the posterior great artery, which followed the long upward course characteristic of the aorta. The right ventricular size was slightly smaller than normal in the patient with a competent tricuspid valve and within normal limits in the two patients with tricuspid regurgitation.

The low transverse view was basically normal in all three patients but the right atrium appeared larger in the two patients with tricuspid regurgitation. The high transverse view showed normally related great arteries with the circular aorta crossed

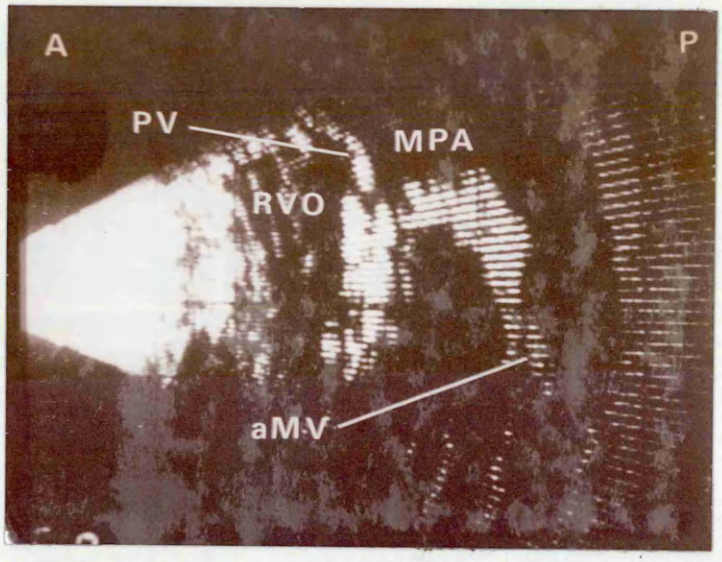
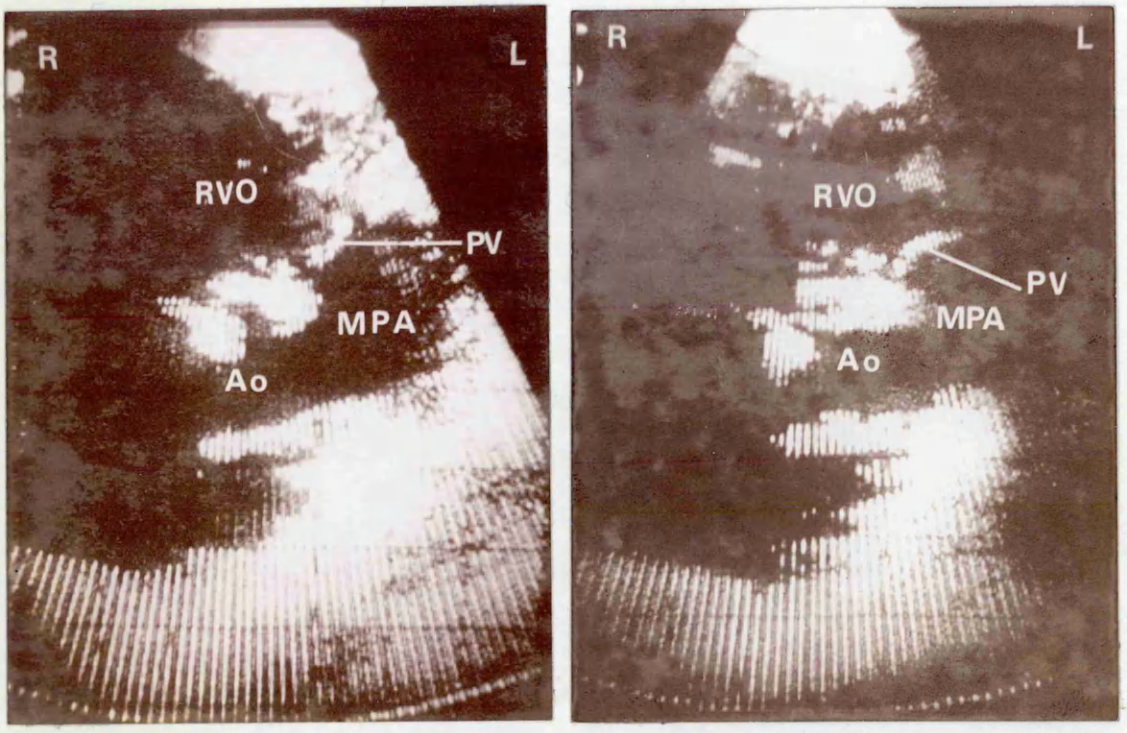


Figure 4, 15 View in systole from an infant with pulmonary atresia and an intact ventricular septum. The scanning plane lies along the right ventricular outflow and main pulmonary artery and through the left ventricular outflow. The pulmonary valve echo persists and appears to dome in systole. Abbreviations as before.



(a) (b)

Figure 4, 16 High transverse views from the infant with pulmonary atresia with an intact ventricular septum and marked tricuspid regurgitation. The pulmonary valve echo exhibits (a) doming with atrial systole and (b) reverse doming with ventricular systole. Abbreviations as before.

by the crescentic right ventricular outflow (RVO) and main pulmonary artery of approximately normal size. Cusp motion characteristic of a pulmonary valve was not however apparent. A persistent transverse linear echo situated in the normal pulmonary valve position was considered to represent the immobile fused pulmonary valve cusps. In two patients this linear echo moved distally within the main pulmonary artery and showed apparent doming in ventricular systole (Figure 4, 15). In the other infant (severe tricuspid regurgitation) the pulmonary valve echo moved distally and domed with atrial systole, but with ventricular systole moved proximally and showed "reversed doming" (Figure 4, 16a&b).

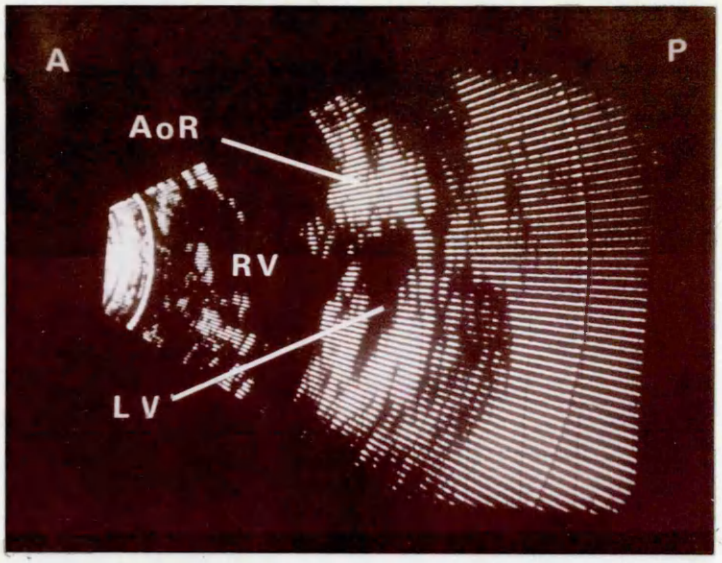
Discussion

In pulmonary valve atresia with an intact ventricular septum, the RVO and MPA are usually of from one third to normal size and are separated by a diaphragm formed by the fused pulmonary valve cusps. With echocardiography a great artery is routinely recognised by the characteristic pattern of active movement of its valve cusps. An atretic pulmonary valve, having little intrinsic cusp movement, does not present this pattern and a pulmonary artery with an atretic valve is therefore difficult to identify with M-mode echocardiography. Where the pulmonary valve is imperforate and there is no VSD Godman et al. (1974) reported that the diagnosis could be suspected from a small right ventricular diameter on M-mode echocardiography. However, in pulmonary valve atresia with tricuspid regurgitation the cavity of the right ventricle is normal or dilated (Davignon et al., 1961). Two of the patients studied had a right ventricle of normal size in the

presence of tricuspid regurgitation. Where there is an imperforate pulmonary valve of approximately normal diameter two-dimensional echocardiography has shown it as a transverse echo which persists throughout the cardiac cycle. The different patterns of movement of the pulmonary valve echo (doming and reverse doming) seen in these patients are likely to be related to tricuspid valve function and to reflect the relative pressures in the right ventricle and main pulmonary artery. Where tricuspid regurgitation is absent or only moderate the right ventricular systolic pressure rises above that in the main pulmonary artery with resultant distal movement (doming) of the valve diaphragm as seen in two patients. In the other who had severe tricuspid regurgitation doming was only seen just before ventricular systole; this coincided with atrial systole and the mechanism is therefore likely to be similar to that of the "a" wave on the M-mode echocardiogram, which is considered to be due to the rise in the right ventricular pressure above that in the MPA with atrial contraction (Weyman et al., 1974). Subsequent reversed doming in diastole would have resulted from the fall in the right ventricular pressure below that in the main pulmonary artery as a result of severe tricuspid regurgitation. The pulmonary artery was not entered at cardiac catheterisation and the haemodynamic proof of this is therefore not available. Weyman et al. (1977) with two-dimensional echocardiography have reported the recognition of valve doming in pulmonary valve stenosis and it is possible that the appearance of the cusp echoes that has been found in pulmonary atresia with fused cusps and moderate or no tricuspid regurgitation may be closely similar to those of severe stenosis. However,

Weyman et al. (1977) were able to recognise the valve orifice at the apex of the dome whereas no such orifice was apparent in the present subjects. In addition to showing the fused valve cusps the high transverse view with two-dimensional echocardiography can clearly demonstrate the size of the main pulmonary artery, while in infants with pulmonary valve atresia and a small patent ductus arteriosus this diagnostic information may not always be obtained from cardiac catheterisation and angiocardiography.

(a)



(b)

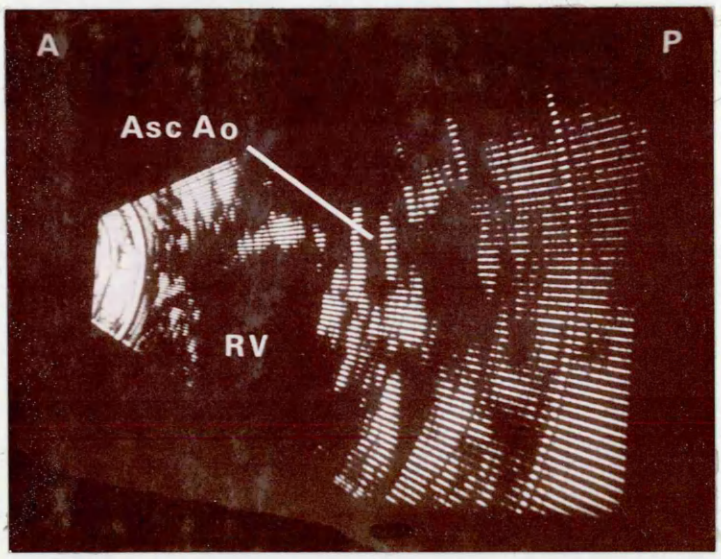


Figure 4, 17 Longitudinal views from an infant with aortic atresia and a hypoplastic left ventricle. (a) The low view shows the small left ventricle, and (b) the high one the tiny aorta. The aortic valve cusps are not clearly demonstrated. AoR, aortic root. Other abbreviations as before.

AORTIC ATRESIA

Five infants (aged 2 days to 5 weeks) with aortic atresia and a hypoplastic ascending aorta were studied. On angiocardiography two were considered to have a hypoplastic left ventricle and ascending aorta with aortic and mitral atresia although the left ventricle was not demonstrated in either case. Neither underwent postmortem examination. In one infant angiocardiography and autopsy showed aortic atresia with a very small ascending aorta and a small but not hypoplastic left ventricle with a ventricular septal defect (VSD) and normal mitral valve. Two others were examined by two-dimensional echocardiography on referral to the Royal Hospital for Sick Children and on the basis of the echocardiographic diagnosis of hypoplasia of the left ventricle and ascending aorta it was decided not to proceed with cardiac catheterisation and angiocardiography. Autopsy showed both to have hypoplasia of the left ventricle and ascending aorta with aortic atresia; the mitral valve orifice was very small but probe patent.

Results

The echocardiographic findings in the standard longitudinal view were similar in the four infants with hypoplasia of the left ventricle. The ventricular septum appeared as a thick structure separating a large anterior right ventricle from a very small posterior left ventricle (Figure 4, 17a). Within the right ventricle the extensive movement of the echo from the anterior tricuspid leaflet was seen. A linear echo which showed little movement separated the left atrium from the left ventricle but there was no clearly discernible mitral valve movement. In the

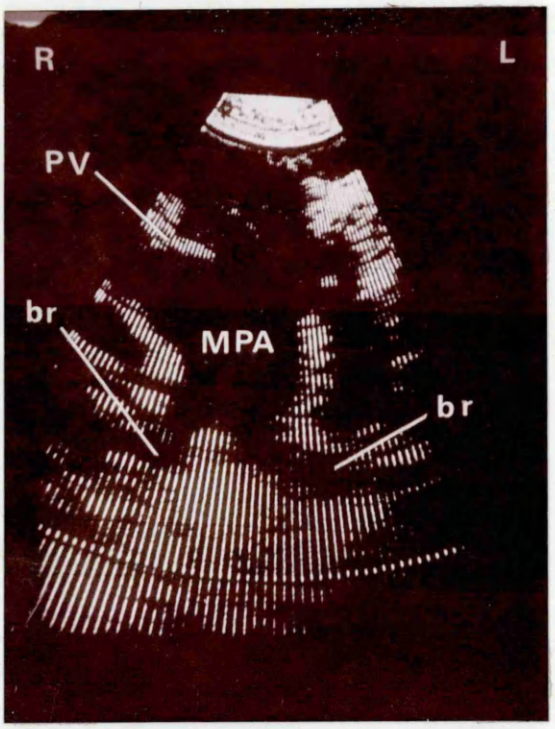


Figure 4, 18 High transverse view from the infant in Figure 17 showing the large pulmonary artery and its branches. br, pulmonary artery branches. Other abbreviations as before.

infant with a VSD echocardiography showed the left ventricular size and the mitral valve movement to be normal. In two patients the left ventricle was separated from the ascending aorta by a short segment of dense echoes suggesting absence of a lumen (Figure 4, 17a) and in the other three patients the aortic valve was represented by a central linear echo which persisted in systole. In the high longitudinal view two parallel linear echoes were seen to pass upwards from the structure considered to represent the atretic aortic origin (Figure 4, 17b). These parallel echoes could be followed upwards to a level above the main pulmonary artery indicating that they represented the ascending aorta. The internal diameters of the ascending aortas measured from echocardiographic still frames ranged from 2 to 5 mm.

The low transverse view showed a large anterior right ventricle containing the tricuspid valve echo in all 5 patients. In the 4 infants with severe left ventricular hypoplasia it was possible to demonstrate the tiny left ventricle but mitral valve motion could not be discerned. In the patient with a VSD the left ventricular size and mitral valve movements appeared normal. In all 5 patients it was possible to demonstrate the very large right ventricular outflow and main pulmonary artery crossing the tiny aorta in the normal manner and branching (Figure 4, 18).

Discussion

The majority of patients with aortic valve atresia and a hypoplastic aorta have a hypoplastic left ventricle with an intact ventricular septum, while a few have a left ventricle of normal or intermediate size related to a VSD (Roberts et al., 1976). Those

with a hypoplastic left ventricle have an abnormal mitral valve which is either atretic or hypoplastic while most of those with a larger left ventricle and a VSD have a normal mitral valve (Roberts et al., 1976).

In each infant studied here echocardiography identified the very small ascending aorta and permitted measurement of its internal diameter. These dimensions (2 to 5 mm) correspond with those (3 to 6 mm) reported with M-mode echocardiography and left heart hypoplasia (Farooki et al., 1976). The combination of the hypoplastic aortic root and absence of movement of the aortic valve cusps in all cases suggested aortic atresia but, as with pulmonary atresia, it was not possible to be absolutely certain from two-dimensional echocardiography whether there was atresia or severe stenosis. The apparently normal left ventricular size and mitral valve motion in the infant with a VSD allowed a clear distinction to be made from those with a hypoplastic left ventricle. The appearance suggested the possibility of a VSD although none was identified. The absence of mitral valve motion in those with a hypoplastic left ventricle indicated an abnormal valve but it was not possible to say whether it was atretic or hypoplastic. A hypoplastic left ventricle and ascending aorta can be recognised with M-mode echocardiography (Godman et al., 1974; Farooki et al., 1976) and it may be that two-dimensional echocardiography does not add to the diagnostic information which may be obtained with the single beam technique in this condition; indeed M-mode echocardiography may allow more accurate assessment of the movement of the mitral valve and permit a cleaner distinction between atresia and severe stenosis (Lundstrom, N-R., 1972) than

was obtained from these two-dimensional studies. However two-dimensional echocardiography allows the great arteries to be identified independently; this could theoretically be important should there be a patient with transposition of the great arteries and pulmonary atresia, although there should be no difficulty in making the distinction on clinical and radiological grounds.

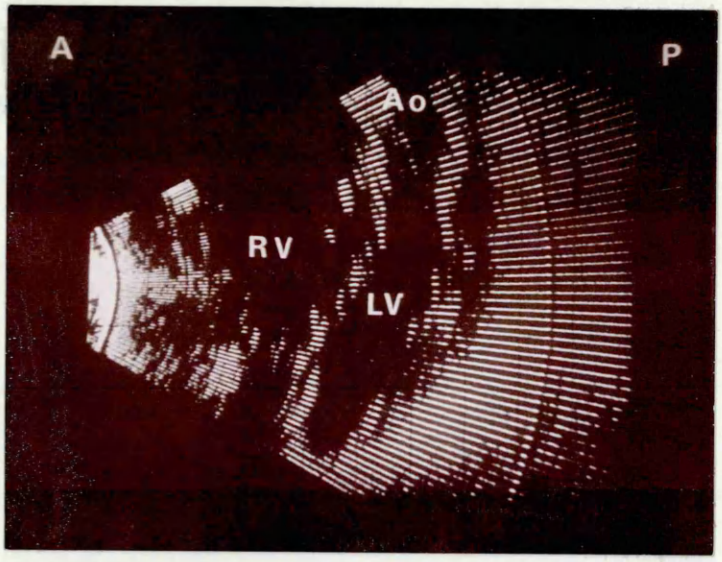


Figure 4, 19 Longitudinal view from an infant with non-obstructed TAPVD (left sided veins draining to the innominate vein and right sided ones to the coronary sinus). The left atrium, left ventricle and aorta are small and the right ventricle relatively large. A posterior echo-free space cannot be seen in this illustration but was demonstrated by adjustment of the scanning plane and ultrasonic gain settings. Abbreviations as before.

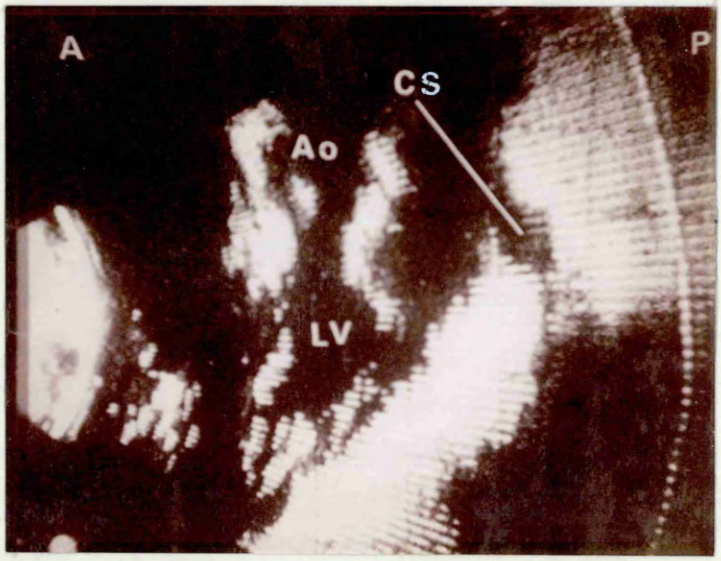


Figure 4, 20 Longitudinal views from an infant with non-obstructed TAPVD to the coronary sinus showing a circular posterior echo-free space considered to represent the coronary sinus. CS, coronary sinus. Other abbreviations as before.

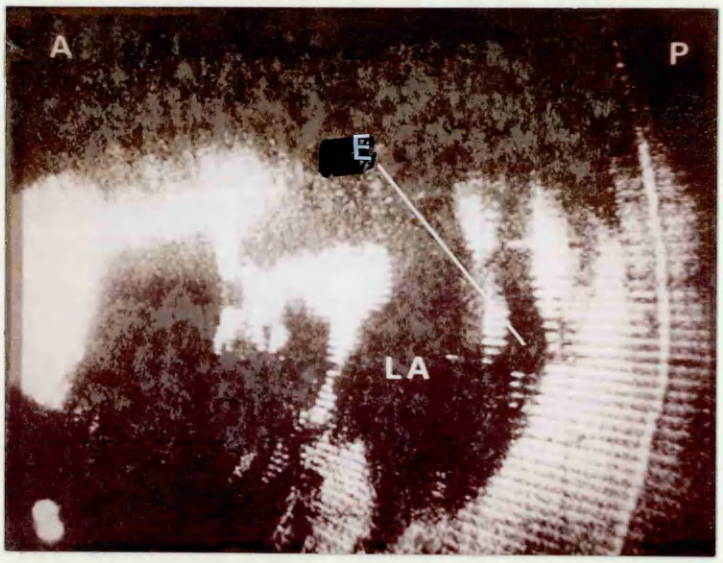
TOTAL ANOMALOUS PULMONARY VENOUS DRAINAGE

Seven infants (aged 1 day to 5 months) with total anomalous pulmonary venous drainage (TAPVD) were studied. Six were examined before angiocardiography and the exact diagnosis was ascertained at surgery in all seven. Pulmonary venous drainage was obstructed in three with the pulmonary veins entering a common pulmonary venous trunk which led to a hepatic vein (2) or to the superior vena cava (SVC) just above the heart (1). There was no obstruction in four patients. In two all the pulmonary veins entered a single common pulmonary venous trunk which in turn entered the coronary sinus. In the other two there was a mixed type of abnormality with blood draining to the coronary sinus or innominate vein; in one the left upper lobe vein, and in the other both the left sided veins, entered the innominate vein while in both all the other veins entered the coronary sinus.

Results

In all cases the structures seen in the normal longitudinal scan were identified in their usual relationships. The left atrium, left ventricle and aorta were relatively small and the right ventricle large (Figure 4, 19). An echo-free space could be clearly demonstrated posterior to the left atrium in five patients although it was not noted on initial examination in two of them. This space lay behind the lower part of the left atrium and was approximately circular in shape in four patients in whom the anomalous pulmonary drainage was at least partially to the coronary sinus (Figure 4, 20); in two of these (all four veins to the coronary sinus, and left upper pulmonary vein to the innominate

(a)



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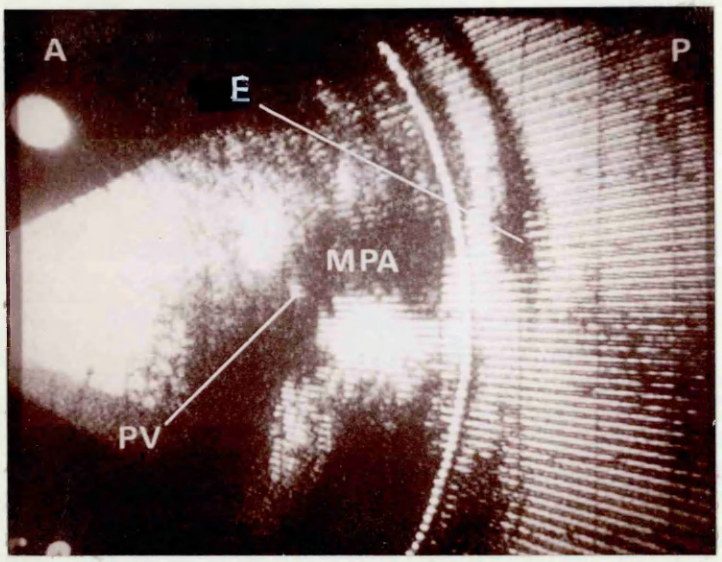


Figure 4, 21 Views with the scanning plane lying along the right ventricular outflow and main pulmonary artery and adjusted to show a posterior echo-free space apparently passing upwards. The space is considered to represent the left upper pulmonary vein draining downwards in (a) (all veins to coronary sinus) and upwards in (b) (left upper pulmonary vein to innominate vein and others to the coronary sinus). E, echo-free space. Other abbreviations as before.

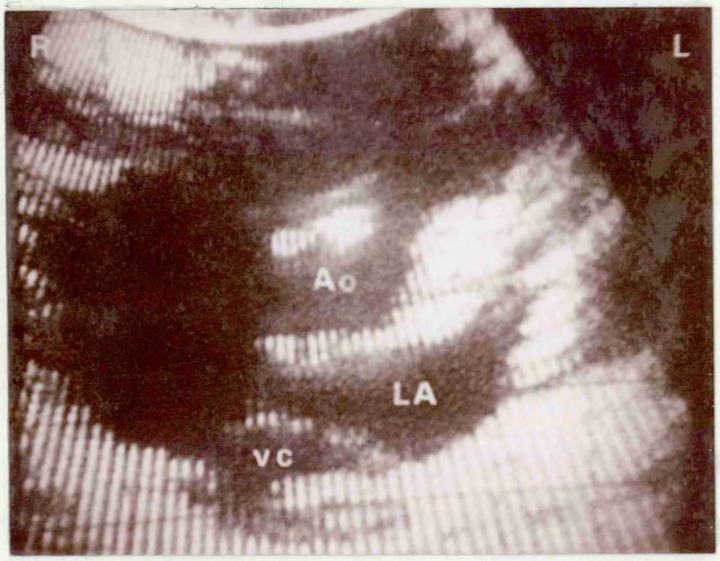


Figure 4, 22 High transverse view from an infant with non-obstructed TAPVD to the coronary sinus showing a posterior echo-free space considered to represent the common venous chamber or coronary sinus. VC, common venous chamber. Other abbreviations as before.

vein and others to the coronary sinus) adjustment of the scanning plane to lie along the right ventricular ourflow and main pulmonary artery showed the space to be elongated and apparently pass upwards and slightly to the left (Figure 4, 21). In the other (obstructed drainage to the SVC just above the heart) the echo-free space lay behind the upper part of the left atrium and was elongated in a superior-inferior direction. A clear echo-free space was not demonstrated in the two infants with subdiaphragmatic obstructed TAPVD; in one a space with indistinct boundaries could be discerned behind the left atrium and ventricle, but in the other no space was demonstrated by echocardiography although a wide pulmonary venous trunk was found at surgery.

The low transverse view was similar to normal with a small left ventricular cavity. In the high transverse view the great arteries were shown to be normally related; the aorta was small with a large right ventricular outflow and main pulmonary artery. An echo-free space was demonstrated posterior to the left atrium in the patients in whom a similar space was apparent in the longitudinal view (Figure 4, 22).

The four chamber view had not been developed when the first four patients were examined and it was used only in three infants (all veins to the coronary sinus (2) or left upper vein to innominate vein and others to the coronary sinus). In all three patients the ventricular septum and anterior atrioventricular valve cusps were recorded. However, in two the atrial septum could not be clearly demonstrated and separate insertions of the mitral and tricuspid valves into the ventricular septum at different levels could not be shown; in the first of these

patients to be examined the appearance was initially misinterpreted as representing a partial atrioventricular canal with an absent atrial septum (see Chapter V).

Discussion

There are numerous anatomical variations of total anomalous pulmonary venous drainage (Goor and Lillehei, 1975) but in all the four pulmonary veins drain into the right atrium, either directly or via its systemic venous tributaries. All four veins may enter a common venous trunk and thence the systemic venous circulation or individual veins or combination of veins may enter at different sites. Where there is no venous obstruction there is resultant volume overload of the right ventricle, recognised with M-mode echocardiography as a large ventricle and paradoxical septal motion (Tajik et al., 1972). However with severe pulmonary venous obstruction the pulmonary arterial and right ventricular pressures become markedly elevated with the result that significant right ventricular volume overload is not present and paradoxical septal motion is not observed (Hagler, 1976). With M-mode echocardiography a common venous trunk may be identified as a well defined echo-free space behind the left atrium (Paquet and Gutgesell, 1975) and where there is total anomalous pulmonary venous drainage to the coronary sinus a similar space behind the atrioventricular junction can represent the dilated coronary sinus (Orsmond et al., 1978; Aziz et al., 1978). However a large pulmonary venous trunk is not always present posterior to the left atrium and will not then be demonstrated on echocardiography. It might be expected that where there is obstruction to pulmonary venous drainage a pulmonary venous trunk would become dilated and

would be more easily recognised with echocardiography. However, of the three infants with obstructed drainage, a posterior echo-free space was seen clearly (although not noted on initial examination) in only one, shown with indistinct margins in another, and not demonstrated in the third although a dilated common venous trunk was found at surgery. In the other four infants where an echo-free space was clearly seen two or more of the pulmonary veins entered the coronary sinus and this space probably represented a dilated coronary sinus and not a pulmonary venous trunk. In two cases a long echo-free space was recognised in a view lying along the right ventricular outflow. In one (all veins to the coronary sinus) it was initially thought that this space might represent a venous trunk draining upwards to the innominate vein; reinterpretation of the echocardiogram after the diagnosis was made by angiocardiography suggested that this probably represented the left upper pulmonary vein draining downwards. However in the other (left upper lobe vein to the innominate vein and others to the coronary sinus) it is not clear what this space represents, and it may also be the left upper pulmonary vein.

It is difficult to give a satisfactory explanation as to why an echo-free space could not be shown in the patients in whom a dilated common venous trunk was subsequently demonstrated at surgery. At present the only explanation for this is that the examinations were performed rapidly and sufficient care may not have been taken. Three of the four infants in whom difficulty was experienced in showing a common venous trunk (or dilated coronary sinus) had obstructed TAPVD and were consequently very ill and the examination was therefore performed as fast as possible. The other infant, in whom the coronary sinus was not shown initially, was examined

during a busy out-patient clinic and the author spent a minimum of time in the study and, in fact, made the incorrect diagnosis of atrioventricular canal. It was possible to re-examine two of these four infants after angiocardiography and spaces presumed to represent a common venous trunk and the coronary sinus respectively were clearly demonstrated. However the significance of the interpretation of an echo-free space as representing the common venous trunk or a dilated coronary sinus must be questioned in view of the fact that similar spaces can sometimes be shown posterior to the left atrium in subjects with normal pulmonary venous drainage (Bozio and Davignon, 1976). A similar though less well circumscribed space was seen in 9 of the 40 normal subjects reported in Chapter III. It is likely that further studies, particularly contrast injections at the time of cardiac catheterisation, will be required to elucidate the nature of this echo-free space and confirm the identity of those seen in patients with TAPVD. The appearance of the atrioventricular valves in the four chamber view which was similar to the partial atrioventricular canal with a large atrial septal defect may be explained by the fact that in TAPVD the left ventricle is small and the mitral valve ring is therefore positioned very close to the tricuspid. Furthermore when the atrial septum cannot be recognised the distinction cannot be made with certainty between the membranous septum (which separates the insertions of the atrioventricular valves) and the anterior mitral leaflet; the structure considered to represent the anterior mitral leaflet may therefore have consisted of both the anterior mitral leaflet and the membranous septum. It has been reported that subxiphoid transducer placement

provides a better image of the atrial septum (Bierman and Williams, 1979b) and of a common venous trunk in TAPVD (Bierman and Williams, 1979a). The application of this view should ensure that the atrial septum is clearly demonstrated and improve the diagnostic ability of two-dimensional echocardiography in patients with TAPVD.

CONCLUSION

As a conclusion to this chapter the distinguishing features for each condition studied will be summarised. Definite diagnostic features will be given where possible and where none have been found the appearances suggestive of the diagnosis will be described; when the findings quoted concern only some of the standard views it should be assumed that the appearance in the others must be shown to be normal. The reliability of any diagnostic criterion will be evaluated and an attempt made to assess the frequency with which it should be demonstrated with two-dimensional echocardiography. However on occasions it can be difficult to obtain an echocardiogram of diagnostic quality in every view, and on the basis of rather small groups of patients in some of the conditions studied a definitive conclusion as to the frequency with which any echocardiographic feature is likely to be demonstrated cannot always be reached. Thus an estimate of the probability of demonstrating a specific diagnostic feature must to some extent depend on the author's subjective assessment of the difficulty in showing it.

Complete transposition of the great arteries

The great artery arising from the left ventricle must be identified as the main pulmonary artery and that from the right ventricle as the aorta. The MPA can be distinguished from the aorta in a longitudinal view by its posterior turn at a lower level or in a high transverse view by the demonstration of its bifurcation. Either of these features will allow the great arteries to be identified and thus a definite diagnosis to be made, but while it should be possible to demonstrate the posterior

turn of the MPA in almost all patients, its branching may be shown with certainty in no more than two-thirds of them.

In the high transverse view the demonstration of the great arteries as two circular spaces with central cusp echoes in diastole would indicate that the great arteries lie parallel at their origins and would support the diagnosis of CTGA.

In addition the ventricular septum and both atrioventricular valves must be demonstrated in their normal positions to exclude other lesions associated with malposition of the great arteries.

Corrected transposition of the great arteries

Where there is situs solitus and levocardia the left sided ventricle must be identified as the morphological right ventricle and be shown to give rise to the aorta, and the right sided ventricle as the morphological left ventricle giving rise to the MPA. The morphological right ventricle may be distinguished from the morphological left ventricle in the longitudinal view by the presence of an infundibulum (semilunar-atrioventricular valve discontinuity) or in the four chamber view by the antero-inferior position of the left atrioventricular valve relative to the right one. The presence of an infundibulum in one ventricle and not in the other should provide reliable identification of the ventricles in corrected TGA but will not always do so in other rare and complex conditions. Ventricular identification by the situation of the atrioventricular valves requires further study before an estimate of its applicability can be given. The great arteries should be identified by the criteria described under complete TGA.

In the high transverse view the demonstration of the great arteries as circles with the anterior one (the aorta) to the left

of the other would indicate parallel great arteries and support the diagnosis.

In addition the ventricular septum must be demonstrated to exclude univentricular heart with malposition of the great arteries.

Double outlet right ventricle

More than one and a half great arteries must be demonstrated to arise from the right ventricle. This can best be achieved by using the longitudinal view to show that the central part of the ventricular septum is, in diastole, posterior to the mid point of the posterior great artery, and a transverse sweep to show that the other great artery is anterior to the ventricular septum. This will allow the diagnosis to be ascertained with certainty.

The demonstration that both atrioventricular valves are discontinuous with the semilunar valves would be suggestive but not diagnostic of double outlet right ventricle.

Persistent truncus arteriosus

A single great artery should be shown to arise from the heart and this must be positively identified as a persistent truncus arteriosus by the demonstration of more than three valve cusps or a long upward course with a branch (or branches) arising from it just above cusp level. More than three valve cusps will be present in only 25% of cases of persistent truncus arteriosus and it is possible that even when four or more are present they may not always be clearly recognised. Although branches arising from the main arterial trunk should be demonstrated more frequently than a valve with more than three cusps, they may not always be

seen. A definite assessment of this cannot be made on the basis of the present small study.

Tetralogy of Fallot

Over-riding of the aorta and infundibular pulmonary stenosis must be demonstrated. The longitudinal view will show the anterior wall of the aorta to be discontinuous with the ventricular septum and to over-ride it in such a way that in diastole less than half the aortic root is anterior to the mid point of the upper septum. In the high transverse view the crescentic right ventricular outflow and main pulmonary artery crossing the aortic root will indicate normally related great arteries, and a narrow RVO the indundibular pulmonary stenosis. The demonstration of an over-riding aorta and narrow RVO will allow a definite diagnosis of tetralogy of Fallot to be reached but whilst the over-riding aorta should always be shown a very narrow RVO may not always be clearly defined. Echocardiography alone does not permit distinction between tetralogy of Fallot with severe infundibular and valve stenosis and atresia of the pulmonary valve.

Pulmonary atresia with an intact septum

The pulmonary valve echo must be shown as a continuous transverse echo which separates the right ventricular outflow and main pulmonary artery and does not disappear in systole. The pulmonary valve echo will either move distally (doming) during ventricular systole (moderate to no tricuspid regurgitation) or move distally with atrial systole and proximally with ventricular systole (severe tricuspid regurgitation).

In addition the ventricular septum must be shown to be

continuous with the anterior wall of the aorta. The immobile pulmonary valve echo should be demonstrated in all infants but echocardiography alone may not permit a definite distinction between pulmonary valve atresia and severe stenosis.

Aortic atresia

The walls of a hypoplastic aorta will appear, in longitudinal view, as parallel linear echoes very close together, and where there is aortic atresia the aortic root will contain an immobile linear echo or appear to have an absent lumen. Other findings will depend on the different abnormalities in each individual case. The size of the left ventricle can be assessed; a hypoplastic ventricle should be readily apparent and will suggest an intact ventricular septum, while one of normal size will be found where there is a VSD. Mitral or aortic atresia cannot with certainty be distinguished from severe stenosis with echocardiography but will be suggested by an immobile echo in the valve position.

Total anomalous pulmonary venous drainage

No completely reliable diagnostic criterion has been found on two-dimensional echocardiography for the diagnosis of TAPVD. The demonstration of relatively small left heart chambers and an echo-free space behind the left atrium (representing a common venous trunk or coronary sinus) would strongly suggest the diagnosis. However, it may not always be possible to demonstrate this echo-free space and the finding of small left heart chambers would then support a clinical diagnosis but would not be diagnostic of TAPVD. Where the pulmonary artery pressure is not elevated M-mode echocardiography can provide further support for the diagnosis if right ventricular volume overload (paradoxical septal motion) can be shown.

CHAPTER V

THE ELUCIDATION OF ATRIOVENTRICULAR CANAL MALFORMATIONS

The hepato-clavicular view with angiocardiography has recently been developed to provide improved visualisation of four cardiac chambers simultaneously, and thus allow more accurate assessment of atrioventricular (AV) canal abnormalities. The four chamber view with two-dimensional echocardiography provides a similar demonstration of the endocardial cushion region of the heart; it not only shows the atrial and ventricular septa but has the advantage over angiocardiography of directly imaging the anterior leaflets of the AV valves and of showing their chordal insertions. Thus two-dimensional echocardiography should be uniquely suited to the diagnosis of AV canal malformations. This chapter will describe the findings on two-dimensional echocardiography in 13 infants and children with AV canal malformations and compare them with a control group consisting of normal subjects described in Chapter III and patients with a ventricular septal defect (VSD), atrial septal defect (ASD), left ventricle to right atrium septal defect or pulmonary stenosis.

The terminology used will be that of McMullan et al (1972) based on the anatomical report of Rastelli et al. (1966). Common to all types is an endocardial cushion defect characterised centrally by deficiency of septal tissue involving both the atrial septum (septum primum) and base of the ventricular septum. They are classified on the basis of the anterior AV valve abnormalities into partial AV canal (separate anterior AV valve leaflets adherent to the edge of the ventricular septum) and complete AV canal (anterior common leaflet not directly adherent to the ventricular septum). All forms of complete AV canal have an anterior common leaflet and sub-types are differentiated by

the anatomy of the leaflet and its chordal insertions. In types A and B the anterior common leaflet is divided by a midline cleft into mitral and tricuspid portions each of which has medial and lateral chordae. The lateral mitral and tricuspid chordae are inserted into papillary muscles in the left and right ventricles respectively. The medial chordae are attached to the rim of the ventricular septal defect in type A, and to an abnormal papillary muscle in the right side of the ventricular septum in type B. In type C the anterior common leaflet is undivided and free floating with no chordal attachment to the septum.

Subjects

13 patients (1 week to 14 years) with an angiographic diagnosis of AV canal malformation were examined. The exact anatomy ascertained at autopsy (2 infants) or surgery (8 children) was partial AV canal (7) or complete AV canal type A (1) or type C (2). In one child, surgery confirmed the recurrence of a low ASD following earlier repair of a partial AV canal. In the remaining 2 patients, the angiocardiographic diagnosis was partial AV canal. The echocardiogram was repeated after surgical repair of the defect in 5 patients. A control group of 25 patients in whom the abnormality was ascertained by angiocardiography was also studied; 11 had a membranous or muscular VSD (1 month to 10 years), 8 had an ostium secundum ASD (3 to 8 years), one a left ventricle to right atrium septal defect (7 years) and 5 pulmonary stenosis with no intracardiac shunting. In addition the 40 normal subjects described in CHAPTER III will be considered as control subjects. A standard two-dimensional echocardiographic examination was performed on all subjects and in addition echocardiography with left ventricular injection of

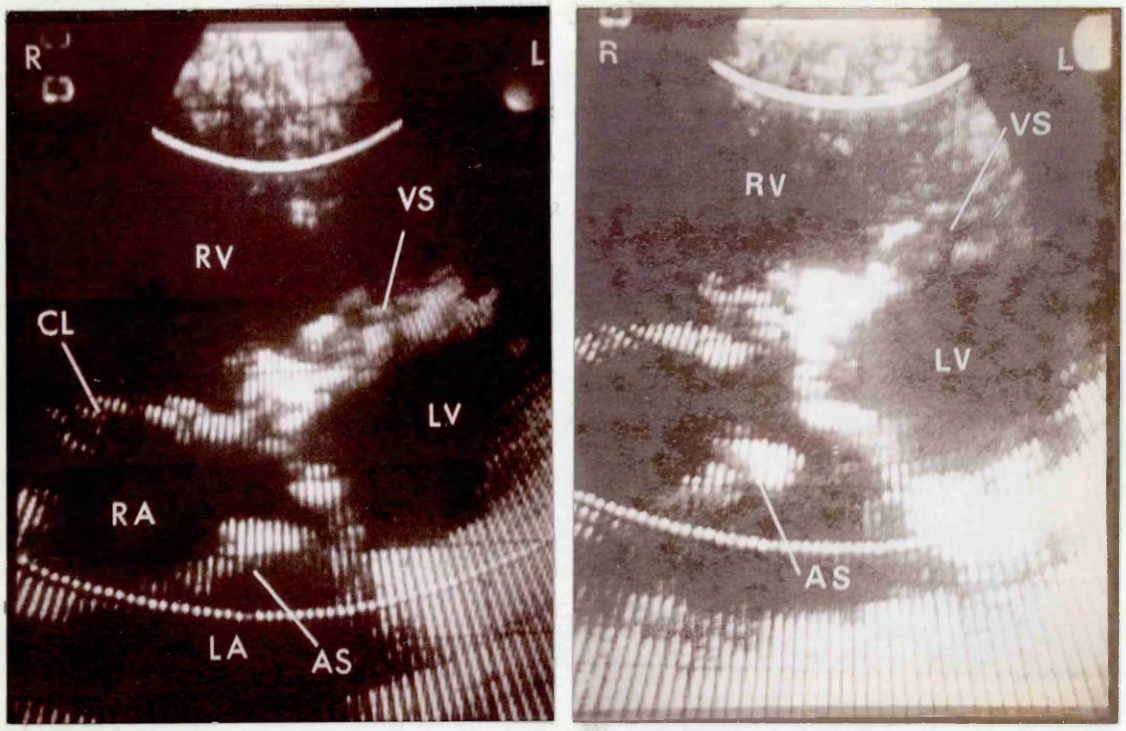
ultrasonic contrast (saline or blood) was performed at cardiac catheterisation in three patients with pulmonary stenosis, three with a VSD, two with an ASD, two with a partial AV canal and one with a complete AV canal type C.

Results

The important diagnostic features were seen in the four chamber view.

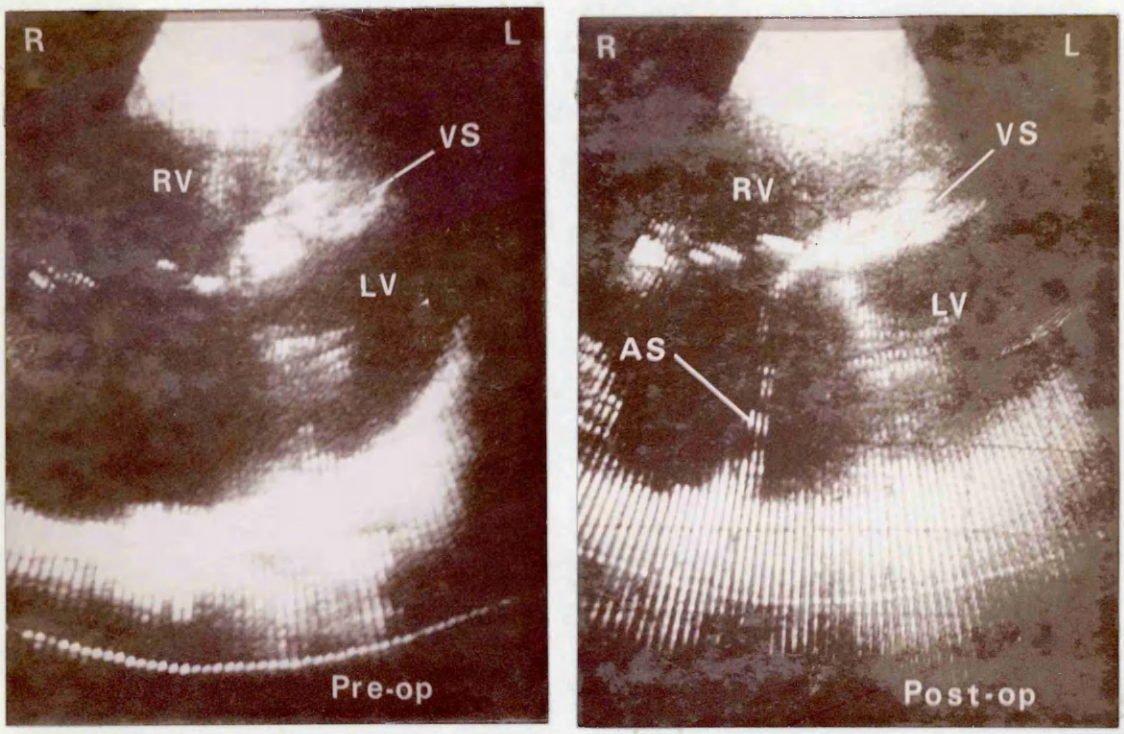
Control group

The appearance of the four chamber view was similar in all cases and was described in detail in CHAPTER III. The ventricular septum appeared as a thick structure which curved posteriorly from left to right and joined the thinner atrial septum which continued backwards to the posterior heart margin. In some patients, echoes could not be recorded in the central or posterior parts of the atrial septum; this echo drop-out may have represented the region of the fossa ovalis or the ASD in some cases (Silverman and Schiller, 1978) but this could not always be clearly shown even in the patients with ostium secundum ASDs. The anterior mitral and tricuspid leaflets were represented by transverse linear echoes impinging on the left and right sides respectively of the ventricular septum, the anterior tricuspid leaflet being situated in an antero-inferior position relative to the mitral leaflet. Contrast echocardiography was performed with injection of ultrasonic contrast (blood or saline) into the left ventricle while the four chamber view was recorded in 8 of the control patients; in 3 with no intracardiac shunting and 2 with an ostium secundum ASD contrast was seen only in the left ventricle while in the 3 with a VSD opacification of the right ventricle followed



(a) (b)

Figure 5, 1 Four chamber views in systole from a child with a partial atrioventricular canal showing discontinuity between the atrial and ventricular septa. In (a) it appears that there may be a common anterior leaflet but in (b) there are clearly separate mitral and tricuspid components attached to the edge of the ventricular septum. CL, anterior common leaflet. Other abbreviations as before.



(a) (b)

Figure 5, 2 Four chamber views from a child with a partial AV canal, (a) before and (b) after surgical repair. The atrial septum initially appears to be absent, but is clearly seen in the post operative study. Abbreviations as before.

that of the left. Contrast was not visualised in the atria in any of these patients.

AV canal malformation

In the four chamber view the ventricular septum was recorded in all 13 patients and the atrial septum in 10. In those where both the ventricular and atrial septa were seen there was apparent discontinuity between their contiguous margins (Figure 5, 1a). These structures moved asynchronously through the cardiac cycle; in ventricular diastole the edges were almost antero-posterior whilst in ventricular systole the atrial septum moved to the left of the posterior margin of the ventricular septum. The lack of continuity in the echoes from the septa and the asynchronous movement between their edges were considered to represent true discontinuity between the base of the ventricular septum and the low atrial septum and thus the confluent septal defect. The 3 patients in whom echoes from the atrial septum could not be visualised (Figure 5, 2a) were found at subsequent surgery to have very large atrial septal defects, one having a common atrium and two only a small rim of atrial septal tissue; in the latter 2 patients echoes from part of the atrial septum could be recorded posterior to the aortic root in a standard high transverse view. The echocardiogram in the 5 patients studied after reconstructive surgery showed continuity of the atrial and ventricular septa throughout the cardiac cycle. In the 3 patients in whom the atrial septum was not seen in the pre-operative echocardiogram the atrial septum constructed at operation was clearly demonstrated as attached to the edge of the ventricular septum (Figure 5, 2b).

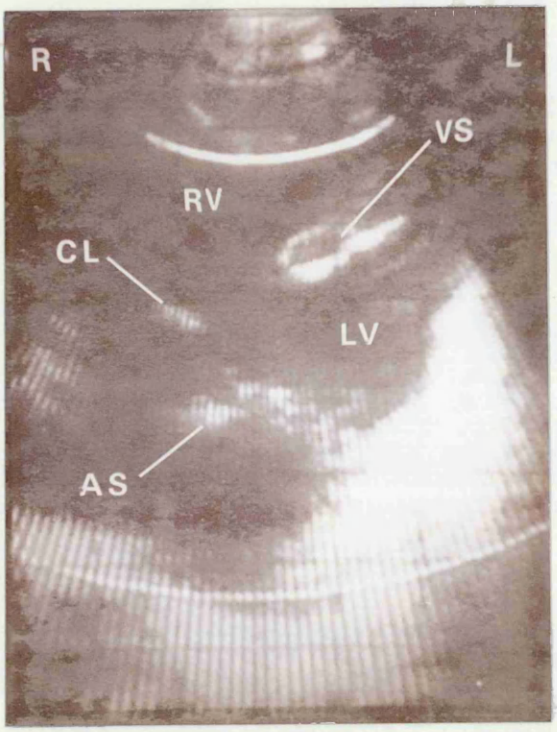
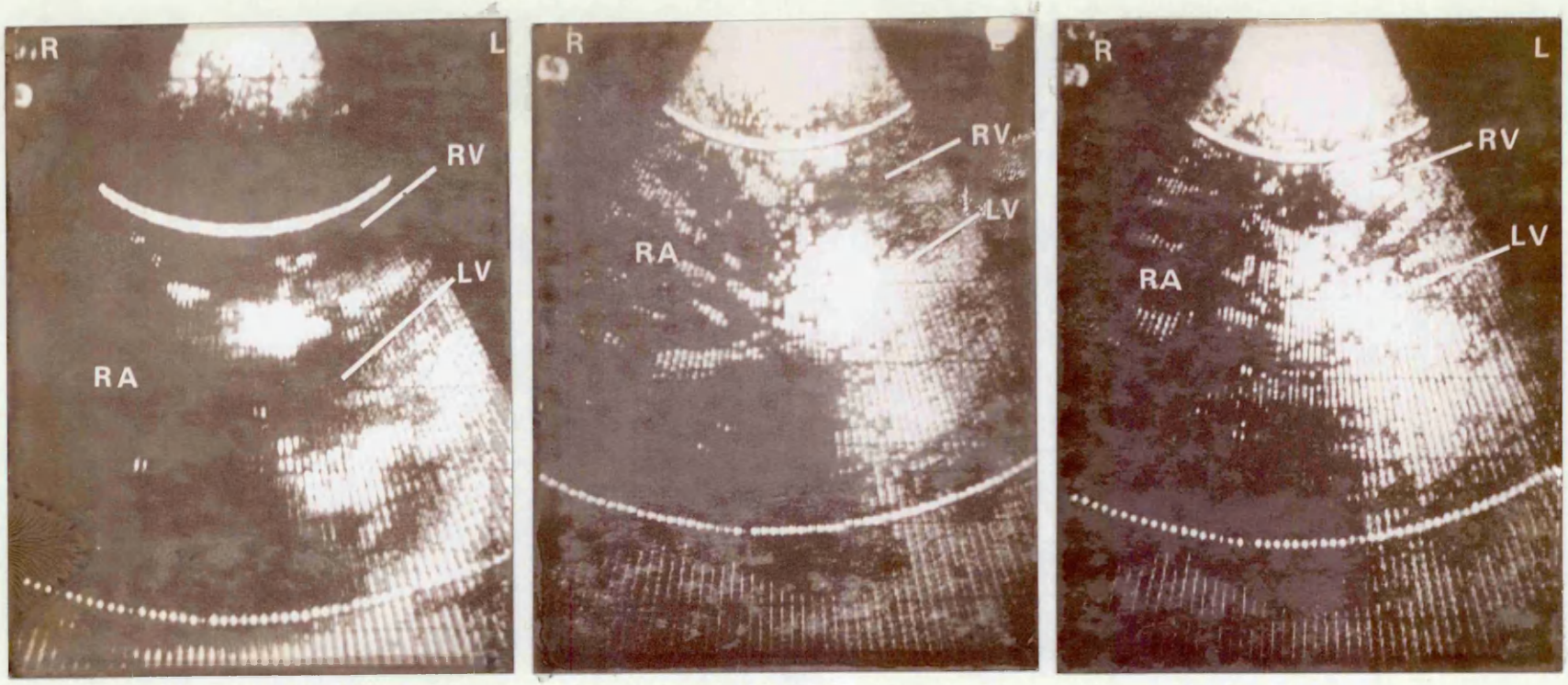


Figure 5, 3 Four chamber view in systole from a child with a complete atrioventricular canal type C. The anterior common leaflet is posterior to the ventricular septum with no apparent attachment to it. Abbreviations as before.



(a) (b) (c)

Figure 5, 4 Four chamber views at approximately the same time in the cardiac cycle from a child with partial AV canal and mitral regurgitation; (a) before and (b) and (c) after injection of blood into the left ventricle. Following injection (b) the left ventricle and right atrium opacify with (c) contrast appearing in the right ventricle only with the following cycle, suggesting the right ventricle opacifies as a result of mitral valve regurgitation and not through a ventricular septal defect.

The anterior atrioventricular valve leaflets (or common leaflet) were seen as a continuous echo passing from the right ventricle to the left ventricle and separate insertions of the mitral and tricuspid components into the ventricular septum were not apparent. Two distinct appearances were noted during ventricular systole. In the 10 patients with partial AV canal (8 proven at operation or autopsy and 2 diagnosed by angiocardiography) and one infant with a type A complete AV canal (confirmed at surgery) the lateral margins of the anterior leaflets moved to a posterior position with respect to the central parts; the leaflets showed a tented appearance with the apex apparently attached to the edge of the ventricular septum (Figure 5, 1b). In the 2 patients with type C complete AV canal the entire anterior leaflet moved posteriorly and there was no apparent attachment to the ventricular septum (Figure 5, 3). The post-operative echocardiogram in patients with both partial and complete AV canal showed a similar appearance with the anterior mitral and tricuspid leaflets attached to the ventricular septum at the same level, its junction with the atrial septum.

Left ventricular injection of ultrasonic contrast was performed in 2 patients with a partial AV canal and cleft mitral leaflet and one with a type C complete AV canal. In those with a partial AV canal the left ventricle first opacified and contrast was subsequently seen in the atria and then in the right ventricle (Figure 5, 4ab&c); this suggested that there was AV valve regurgitation with the contrast subsequently reaching the right ventricle through the tricuspid valve. In the patient with type C complete AV canal contrast appeared in the left ventricle and then

the right ventricle but the atria did not opacify; this suggested that there was a ventricular septal defect but no AV valve regurgitation.

Discussion

The elucidation of the exact abnormality in patients with endocardial cushion defects can be difficult with conventional cineoangiocardiography. Rastelli et al. (1967) found that with antero-posterior and lateral angiocardiography an incorrect assessment was made in more than 20% of cases. The subsequent development of the hepato-clavicular view (Bargeron et al., 1977) has improved angiocardiographic visualisation of the atrioventricular junction and should increase diagnostic accuracy in patients with endocardial cushion defects, but this is still unlikely to provide exact determination of AV valve attachments.

It has been demonstrated that M-mode echocardiography can show abnormalities associated with AV canal malformations and criteria for the diagnosis of partial (ostium primum ASD) and complete AV canal have been suggested (Williams and Rudd, 1974; Pieroni et al., 1975; Eshaghpour et al., 1975). Bass et al. (1978) studied the M-mode echocardiogram and suggested that it should be possible to distinguish partial from complete AV canal in 90% of patients by considering different findings, but that there was no certain distinguishing feature. Using a prototype two-dimensional scanner Sahn et al. (1974) were able to demonstrate abnormal anterior atrioventricular valve appearances in AV canal malformations and in some cases a large common anterior leaflet was shown. However, the exact relationship of the anterior atrioventricular valve to the ventricular septum could not be

determined and the anomalies could not be divided on this basis into the subgroups of endocardial cushion defects. With B-scan echocardiography Beppu et al. (1976) described a view showing all four cardiac chambers simultaneously and demonstrated the different appearances between normal subjects and those with AV canal abnormalities, the latter having a characteristic deficiency of the low atrial septum. A preliminary report of this present study using real time two-dimensional echocardiography has previously been presented (Houston et al., 1978b); in accord with the findings of Beppu et al. (1976) this study has shown continuity between the atrial and ventricular septa and separate insertions into the ventricular septum of the anterior mitral and tricuspid leaflets (the tricuspid being anterior) not only in the normal subjects, but also in patients with an ostium secundum atrial septal defect, ventricular septal defect, and left ventricle to right atrium septal defect. In those patients with an AV canal and a septal defect of moderate size discontinuity between the echoes from the atrial and ventricular septa was apparent. However the limitations of ultrasonic examination are such that failure to demonstrate a structure does not necessarily indicate its absence, and the presence of a septal defect is not certain on this criterion alone. The edges of the ventricular and atrial septa moved asynchronously and the author considers that the combination of this and the apparent discontinuity between them represents true discontinuity, and thus the presence of a confluent septal defect characteristic of all forms of the AV canal anomaly. This appearance has also been confirmed by Silverman and Schiller (1978) and Hagler et al. (1979). In patients with a common atrium or very large atrial component of a combined septal defect the failure to demonstrate the atrial

septum in continuity with the ventricular septum does not, with certainty, exclude its presence. However in the three patients in whom an atrial septum could not be shown in the four chamber view an AV canal abnormality was suggested by the demonstration of an anterior common leaflet or anterior mitral and tricuspid leaflets inserting into the ventricular septum at the same level. Furthermore, in two patients part of the atrial septum was recorded posterior to the aortic root in a higher view. The abnormal appearance of the anterior AV leaflet or leaflets is evidence of a complete or partial AV canal anomaly respectively, and the failure to show the atrial septum in the four chamber view is therefore strongly suggestive of an AV canal anomaly with a large septal defect.

The individual types of AV canal anomaly are identified by the abnormalities of the anterior AV valves (or common valve) and their relationship to the ventricular septum. This is most clearly visualised in systole when ventricular contraction causes the valve leaflets to be pushed posteriorly, away from the ventricular septum. In a partial AV canal the anterior mitral and tricuspid leaflets are inserted directly into the edge of the ventricular septum and this attachment was clearly demonstrated with echocardiography. Silverman and Schiller (1978) and Hagler at al. (1979) reported that in patients with types A and B complete AV canal a division of the anterior common leaflet into mitral and tricuspid components could usually be demonstrated. However, in one patient with type A complete AV canal Hagler et al. (1979) found an appearance indistinguishable from that in partial AV canal; this was explained by the fact that the VSD was tiny.

An appearance similar to that of partial AV canal was noted in the only patient in this series (4 weeks old, 3.9 Kg) with type A complete AV canal and in whom the VSD was of moderate size at surgery. The differentiation of partial from type A complete AV canal without contrast echocardiography is dependent on identifying the tissue attached to the rim of the ventricular septum as the atrioventricular valve leaflets (partial defect) or chordae (type A defect). It seems that this distinction may not always be apparent, particularly where there is a very small VSD or with small infants. In type B complete AV canal defects where the chordae pass to the right of the edge of the ventricular septum such diagnostic difficulties may not arise. In type C complete AV canal the absence of any attachment of the valves or their chordae to the ventricular septum is manifest on echocardiography as a free floating anterior leaflet completely separated from the ventricular septum in systole.

The injection of ultrasonic contrast into the left ventricle allowed the levels of intracardiac shunting to be correctly identified. Opacification of the atria was taken to indicate regurgitation of the mitral component and therefore to suggest a cleft mitral valve leaflet in the partial AV canal. This method should further allow the distinction to be made between type A complete AV canal (where there is shunting at ventricular level) and partial AV canal in cases where this distinction is unclear with the standard two-dimensional echocardiogram.

It has thus been shown that in AV canal abnormalities the four chamber view with two-dimensional echocardiography can demonstrate and estimate the size of the confluent septal defect

and elucidate abnormalities of the anterior atrioventricular valve leaflets. The absence of attachment of the common leaflet to the ventricular septum allows complete AV canal type C to be differentiated from the other anomalies. The recognition of a cleft in the common leaflet as reported by Silverman and Schiller (1978) and Hagler et al. (1979) should allow differentiation of complete AV canals type A and B from partial defects but such distinction may be difficult where there is a very small ventricular septal defect or in small infants. The apparently normal appearance in patients with a defect between the left ventricle and right atrium will permit the separation of this abnormality from that of an AV canal. Combination of the echocardiographic examination with left ventricular injection of ultrasonic contrast demonstrates atrioventricular valve regurgitation and interventricular shunting and may allow the anatomical abnormalities to be more accurately differentiated. The author therefore considers that in patients with AV canal anomalies two-dimensional echocardiography is an important investigation which may provide diagnostic information not readily obtained from conventional angiocardiography.

CHAPTER VI

THE ASSESSMENT OF THE CYANOSED INFANT

Before two-dimensional echocardiography can be used as a definitive investigation on which to base therapeutic decisions it is essential that the cardiologist has a clear understanding of its abilities and limitations. Such knowledge can best be obtained by an assessment of its diagnostic accuracy in patients in whom the operator has no prior knowledge of the anatomical diagnosis. This chapter will make such an evaluation by comparing the preliminary echocardiographic diagnosis with the subsequent angiocardiographic or pathological one in a consecutive series of 64 cyanosed infants referred to the Royal Hospital for Sick Children, Glasgow (RHSC) with suspected congenital heart disease.

Chapter IV detailed the echocardiographic appearances on two-dimensional echocardiography in a number of the more common forms of complex congenital heart disease. In many of these conditions patients will not survive infancy without corrective or palliative surgery, and in some no surgical help is possible and the baby will die in the first few months of life. Thus the more severe abnormalities causing cyanotic congenital heart disease will only be apparent in infancy and the findings of many of the infants in this study have therefore been included in Chapter IV. However, in some patients the echocardiogram was initially misinterpreted and the diagnostic features became apparent only on subsequently reviewing or repeating the echocardiographic examination. The author therefore feels it is important that an evaluation of the diagnostic accuracy of two-dimensional echocardiography should be made independently from the findings in Chapter IV. In the cases where the

echocardiographic appearances have been previously discussed the findings will be mentioned only briefly, while in the others they will be described in more detail, particularly where the echocardiogram was misinterpreted.

This description of the author's experience with two-dimensional echocardiography in the initial assessment of the cyanosed infant should indicate conditions in which a confident diagnosis can easily be made with echocardiography, and others in which the diagnosis must at present be made with reservations. The results of this study have previously been reported (Houston and Coleman, 1980).

Subjects and methods

64 cyanosed infants (including 44 newborns) referred to the RHSC between May 1st, 1977 and June 10th, 1978 with suspected congenital heart disease were studied. At the time of the echocardiographic examination their ages ranged from 12 hours to 6 months and their weights from 2.1 to 5.8 Kg. 53 were subsequently shown to have congenital heart disease and 11 considered to have no structural heart defect, the cyanosis being due to pulmonary disease or persistence of the fetal circulation. In all except one patient a diagnostic statement based on the echocardiographic findings was made before the exact diagnosis was reached by cardiac catheterisation and angiocardiography or autopsy, or before the subsequent clinical course indicated the cyanosis was of non-cardiac origin. The remaining infant (pulmonary atresia with intact ventricular septum) underwent echocardiographic examination before the angiocardiographic data obtained at another hospital had been made available to the author.

Final diagnosis	Total number	Echocardiographic diagnosis		
		Basically correct	Incorrect	Incomplete
Transposition of the great arteries	16	16	-	-
Persistent truncus arteriosus	2	2	-	-
Tetralogy of Fallot	5	5	-	-
Pulmonary atresia with intact septum	3	3	-	-
Severe pulmonary stenosis	2	1	-	1
Left heart hypoplasia	4	4	-	-
Atrioventricular canal	3	3	-	-
Univentricular heart	7	5	2	-
Total anomalous pulmonary venous drainage	5	4	1	-
No organic heart disease	11	10	1	-
Miscellaneous	4	1	1	2
	62	54	5	3

Table VI Echocardiographic assessment compared with the final diagnosis in the 62 infants from whom satisfactory echocardiograms were obtained.

Because the author was not always available he could not examine every cyanosed infant referred to the hospital during the study period and, therefore, the group does not consist of 64 consecutive referrals to the RHSC. Nevertheless, every infant referred for cardiological assessment when the author was available to perform an echocardiogram is included, and the group thus represents an unselected series of 64 infants.

The scanning planes have been fully described in Chapter III. The four chamber view was developed several months after the start of the study period and was not therefore used in the examination of the infants who presented in the early months.

Results

Satisfactory echocardiograms were recorded from 62 of the 64 infants. In the other two it was impossible to obtain echocardiograms of diagnostic quality because of poor co-operation or technical difficulties. Table VI summarises the findings.

Complete transposition of the great arteries (TGA)

The correct diagnosis was reached with echocardiography in all 16 infants with TGA. In the longitudinal view the main pulmonary artery (MPA) was identified as arising from the left ventricle and the aorta from the right ventricle. In the high transverse view the great arteries were shown to lie parallel at their origins in all cases, and the bifurcation of the main pulmonary artery was recognised in seven. Other complex lesions with great artery malposition were excluded by demonstrating that the ventricular septum was continuous with the anterior wall of the pulmonary artery and that there were two atrioventricular valves with the left one continuous with the posterior wall of the pulmonary artery.

Persistent truncus arteriosus

The correct diagnosis of persistent truncus arteriosus was made by echocardiography in the two infants with this anomaly. A single great artery with a long upward course over-rode the ventricular septum and was identified as a persistent truncus arteriosus by the demonstration of four valve cusps in both cases and of pulmonary artery branches arising from it in one of them.

Tetralogy of Fallot

This group included 4 infants with tetralogy of Fallot and one with tetralogy of Fallot and pulmonary atresia. The basic anatomical abnormalities were correctly interpreted in all patients. The longitudinal view demonstrated a great artery over-riding the ventricular septum. The high transverse view showed normally related great arteries with a narrow right ventricular outflow (RVO) and MPA. In all infants the pulmonary valve echo was immobile and with echocardiography it was not possible to establish whether it was patent or atretic.

Pulmonary valve atresia with intact ventricular septum

The correct diagnosis was reached with echocardiography in the three infants with pulmonary valve atresia and intact ventricular septum. The presence of a diaphragm formed by the fused pulmonary valve cusps was recognised by the persistence and doming of the pulmonary valve echo as described in Chapter IV, 7. The size of the pulmonary valve ring and the main pulmonary artery was correctly estimated in all patients, including one in whom the main pulmonary artery was not demonstrated on angiocardiography.

Pulmonary stenosis

The echocardiogram was interpreted as showing pulmonary valve

stenosis or atresia in the two infants with pulmonary stenosis. In one who had pulmonary stenosis and regurgitation with a normal sized infundibulum and a ventricular septal defect with the aorta over-riding the ventricular septum, echocardiography showed a thick and relatively immobile pulmonary valve (but not doming), aortic over-ride, and a normal sized RVO; pulmonary regurgitation was not suspected on echocardiography. The other patient had infundibular and valve stenosis but no ventricular septal defect; echocardiography demonstrated septal aortic continuity, a narrow RVO and MPA and a persistent immobile pulmonary valve echo which could not be seen to dome with systole. In both cases the immobile pulmonary valve echo suggested an abnormal pulmonary valve but on the basis of the echocardiogram it was not possible to be certain whether it was stenotic or atretic.

Left heart hypoplasia

The correct diagnosis was established with echocardiography in all four patients with a hypoplastic left ventricle and aorta and aortic valve atresia. The diagnostic features were the tiny left ventricle, aortic root, and ascending aorta in the longitudinal and transverse views. The diagnosis was confirmed by angiocardiography in two and by autopsy in the other two in whom the echocardiogram was considered to be diagnostic and angiocardiography was not performed.

Atrioventricular canal

Two patients with a partial atrioventricular (AV) canal and one with a type A complete AV canal and moderate sized VSD were studied; the findings are included in Chapter V. In all three the basic diagnosis of an AV canal was made with echocardiography but

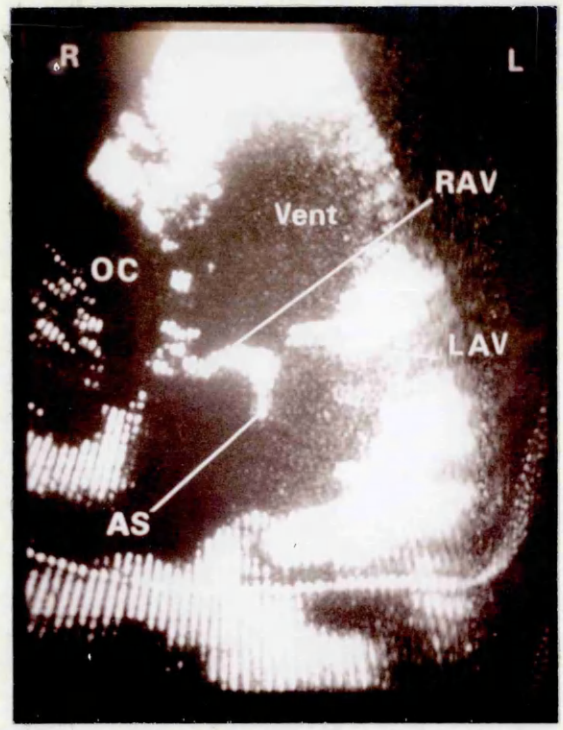
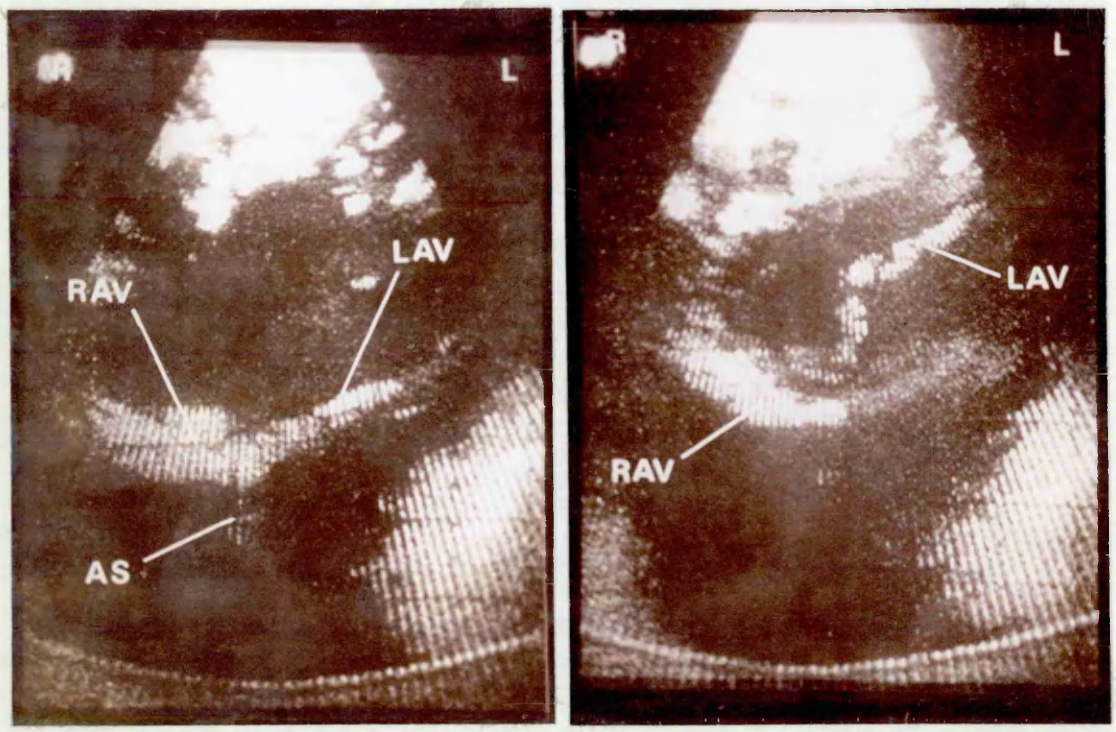


Figure 6, 1 Four chamber view from an infant with double inlet univentricular heart. The "ventricular" septum is not continuous with the atrial septum, but lies to the right of both atrioventricular valves and separates the ventricle from the right-sided outlet chamber. RAV, right atrioventricular valve; LAV, left atrioventricular valve; Vent, ventricle. Other abbreviations as before.



(a) (b)

Figure 6, 2 Four chamber viewsfrom an infant with double inlet univentricular heart and right atrioventricular valve atresia. The anterior leaflet of the right atrioventricular valve appears a little thicker than the left, and it does not exhibit typical anterior motion from (a) systole to (b) diastole shown by the left atrioventricular valve. Abbreviations as before.

it was not possible to distinguish the different types. The four chamber view showed discontinuity and asynchronous movement between the edges of the ventricular and atrial septa and the apparent insertion of the mitral and tricuspid leaflets into the edge of the ventricular septum at the same level. This suggested the diagnosis of partial AV canal in all three patients; the failure to identify the infant with a type A complete AV canal may have been due to the relatively small size of the VSD.

Univentricular heart

Seven infants with a univentricular heart were studied. Four had malpositioned great arteries; five an outlet chamber (4 anterior of right ventricular type and 1 posterior of left ventricular type); four right AV valve atresia, one left AV valve atresia and two had two patent AV valves. In 5 the interpretation of the echocardiogram corresponded with the angiocardiographic findings. A univentricular heart was diagnosed when the "ventricular" septum could not be shown in any view, or separated the main ventricular chamber from another with which less than half an AV valve appeared to connect. An outlet chamber was recognised on longitudinal or four chamber view (Figure 6, 1) as an echo free space separated from the ventricle by a septal structure and not containing an AV valve but giving rise to a great artery. Patent AV valves were suggested by their characteristic anterior motion in diastole on transverse or four chamber views (Figure 6, 2a&b). Absence of an AV connection was presumed in one where no evident or potential communication could be demonstrated between an atrium and the ventricle, and AV valve atresia in three where an immobile structure was recognised between an atrium and the ventricle in the position

of an AV valve (Figure 6, 2a&b). The great arteries were identified and their positions determined by the criteria described under transposition of the great arteries (Chapter IV, 1).

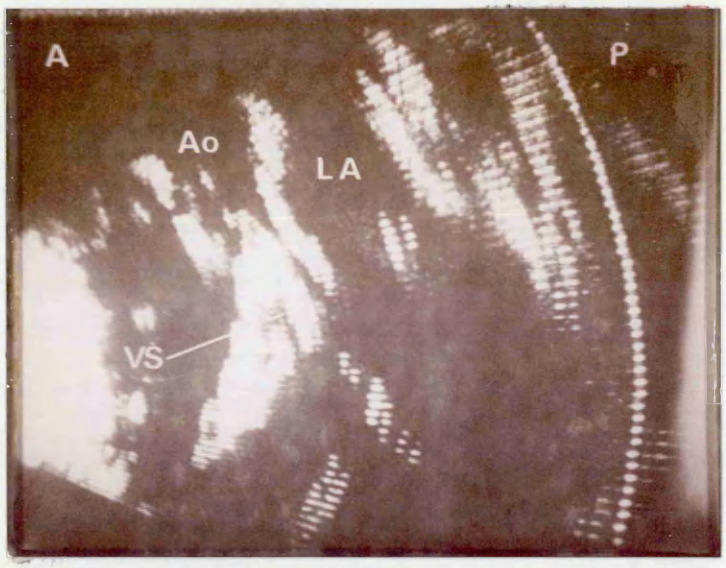
The echocardiographic findings were misinterpreted in two infants. In one the appearance was considered to be essentially normal with a relatively small right ventricle and RVO. Angiocardiography showed the diagnosis to be situs solitus, univentricular heart of left ventricular type with two atrioventricular valves and a large subpulmonary outflow chamber with normally related great arteries. Subsequent review of the echocardiogram showed that the "tricuspid" valve echo had not been demonstrated in the "right ventricle" in the transverse view and that in the four chamber view where both AV valves and the atrial septum were seen a ventricular septum was not demonstrated between the AV valves; the only explanation for this mistake is observer error which was apparent on reviewing the echocardiogram. On angiocardiography the other infant was considered by some observers to have situs solitus, univentricular heart of indeterminate type without an outlet chamber, right AV valve atresia and a persistent truncus arteriosus. On echocardiography a structure resembling the ventricular septum passed downwards from the anterior aortic wall and no AV valve could be demonstrated anterior to it; the space anterior to the "septum" was considered to represent a pouch. Review of the echocardiogram following cardiac catheterisation revealed that a heart wall had not been recorded anterior to this structure, which therefore represented the anterior heart wall, and that there was no pouch or outflow chamber. In addition the echocardiogram showed a great artery with the long upward course

characteristic of an aorta and, posterior to it, parallel echoes which suggested a hypoplastic MPA. On later review of the angiocardiogram with the author the radiologist considered that there was a large anterior aorta with pulmonary atresia and a hypoplastic posterior MPA which filled through a patent ductus arteriosus.

Total anomalous pulmonary venous drainage

Five infants with total anomalous pulmonary venous drainage (TAPVD) were included in the study and the findings have been discussed in detail in Chapter IV, 9. In three the pulmonary venous drainage was obstructed with the pulmonary veins entering a common venous trunk which lead to the hepatic veins (2) or to the superior vena cava just above the heart (1). In two there was no obstruction; in one all four veins entered a common venous trunk and thence the coronary sinus, while in the other the left upper lobe vein entered the innominate vein and the others led to a common venous trunk and thence the coronary sinus. In 4 of the 5 patients, the diagnosis of TAPVD was considered likely on the basis of normal relations and connections of the heart chambers and great arteries with a small left atrium and ventricle and aorta and a large right ventricle, the left heart chambers being larger than would be found with left heart hypoplasia. On the initial examination a definite echo-free space could be demonstrated behind the left atrium in only two infants (non obstructed drainage to the coronary sinus and obstructed drainage to the hepatic veins). Repeat examination after angiocardiography in one of the infants in whom an echo-free space was not seen initially showed the presence of a space considered to represent the common venous

(a)



(b)

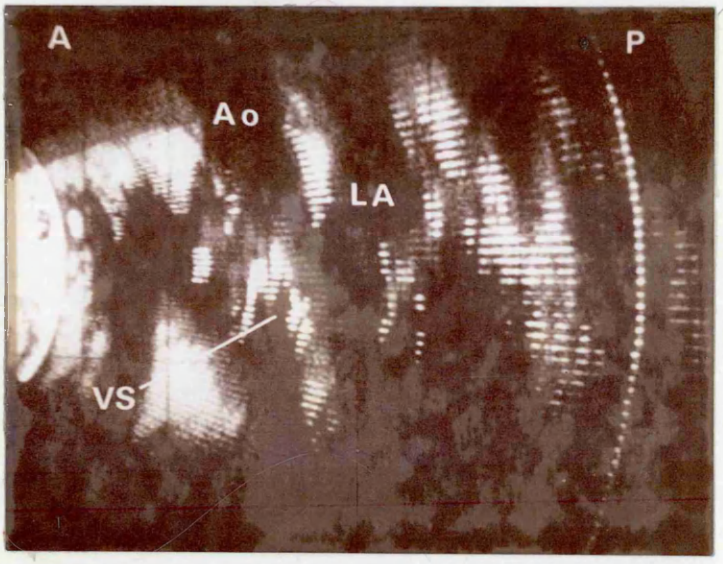


Figure 6, 3 Longitudinal views with slightly different transducer position and angulation from the infant with severe pulmonary hypertension due to upper airway obstruction. In both views the aorta appears to over-ride the ventricular septum. The unusual echoes between the anterior aortic wall and what appears to be the upper ventricular septum are more clearly seen in (a) than in (b). Abbreviations as before.

trunk (drainage to the SVC just above the heart). In the fifth infant the four chamber view was misinterpreted as demonstrating a partial atrioventricular canal. The atrial septum was not clearly shown and the anterior mitral and tricuspid leaflets appeared to be attached to the edge of the ventricular septum at the same level. Re-examination demonstrated normal insertions of the AV valves into the ventricular septum and a posterior echo free space which was thought to represent the coronary sinus (drainage of three veins into the coronary sinus).

No organic heart disease

Eleven of the infants referred for assessment of probable cyanotic congenital heart disease were subsequently shown to have no organic heart disease. In 10, the echocardiogram was considered to show no anatomical abnormality. Five of these underwent cardiac catheterisation; 4 were found to have a high pulmonary vascular resistance with right to left shunting at atrial or ductal level, and one sequestration of the lung. The subsequent clinical course in the remaining 5 infants suggested the cyanosis had not been due to cardiac disease. The echocardiogram was misinterpreted in one infant (aged 4 months) with Pierre-Robin syndrome who at cardiac catheterisation and angiocardiography was shown to have severe pulmonary hypertension secondary to upper airway obstruction. The echocardiogram showed normally sited and connected chambers with normally related great arteries and a large main pulmonary artery. The right heart chambers were large and the left relatively small. In the longitudinal view there appeared to be marked over-riding of the ventricular septum by the anterior aortic wall (Figure 6, 3) which was interpreted as indicating a definite anatomical abnormality,

a ventricular septal defect with double outlet right ventricle. There was no narrowing of the right ventricular outflow, pulmonary artery, or pulmonary valve to suggest pulmonary stenosis and account for the small left sided chambers, and it was postulated that there might be anomalous pulmonary venous drainage, although no definite echo-free space was identified posterior to the left atrium. Review of the echocardiogram showed echoes between the anterior aortic wall and the structure considered to represent the free upper margin of the ventricular septum. As this appearance has never been recorded from patients with true aortic-septal discontinuity and over-ride, it is likely that these echoes represented the upper ventricular septum. The demonstration of this appearance may indicate that there is not discontinuity between the anterior aortic wall and the ventricular septum, and prevent similar misinterpretation of the echocardiogram in the future.

Miscellaneous

The abnormalities in four infants do not readily fit into any of the groups which have been discussed and are best considered individually.

Echocardiography showed one infant to have two AV valves and ventricles with the septum lying in an antero-posterior position. The aorta took origin from the right sided ventricle and was anterior to the MPA, which arose from the left sided ventricle; the left sided AV valve was discontinuous with the posterior wall of the MPA (suggesting a subpulmonary conus) while the relationship of the right AV valve to the aortic root was not clearly shown. The angiocardiogram was interpreted as showing atrial situs solitus,

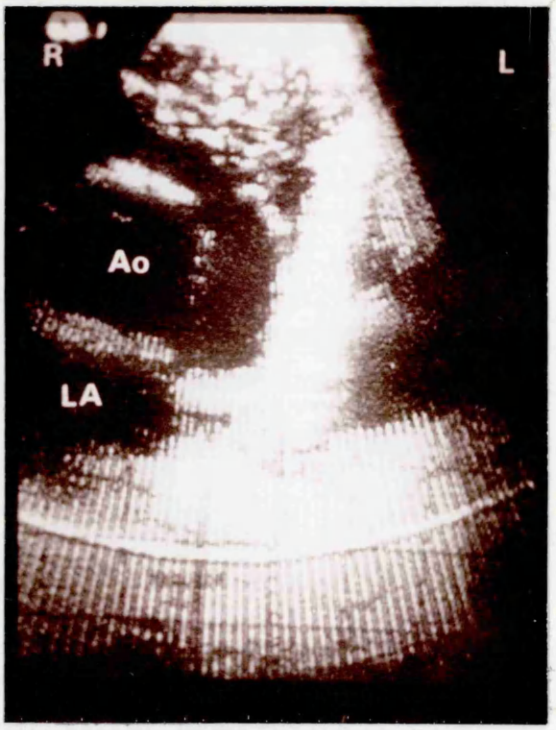


Figure 6, 4 High transverse view from the infant with absence of the right ventricular outflow tract and proximal main pulmonary artery. An echo-free space, probably the left atrial appendage, lies anterior and to the left of the aortic root in the position of a normal right ventricular outflow and pulmonary artery, but a pulmonary valve echo is not seen at any stage in the cardiac cycle. Abbreviations as before.

univentricular heart with left AV valve atresia, transposition of the great arteries with a subaortic outflow chamber, and a hypoplastic aortic arch. Autopsy showed the abnormality to be situs solitus with left juxtaposition of the atrial appendages, atrioventricular concordance with a straddling right AV valve and small right ventricle, a bilateral conus and transposition of the great arteries with a hypoplastic aortic arch. Echocardiography therefore demonstrated the left atrioventricular valve and the subpulmonary conus which were not apparent on angiocardiography; the straddling AV valve was not recognised, probably because this was one of the early studies when the author was relatively inexperienced and the four chamber view was not in use.

In another infant the echocardiogram was incorrectly interpreted as showing a univentricular heart with atresia of the right AV valve, no outlet chamber, and normally related great arteries with a narrow RVO and MPA. Angiocardiography, surgery and autopsy showed a very hypoplastic right ventricle with a hypoplastic but patent tricuspid valve and pulmonary atresia with absence of the right ventricular outflow tract and proximal MPA. Review of the echocardiogram showed that the ventricular septum had been indistinctly shown. In addition, although an echo-free space had been recorded anterior to the aortic root in a position similar to that of a normal RVO and MPA (Figure 6, 4), a pulmonary valve echo had not been demonstrated and the "RVO" had not been shown to be continuous with a ventricle. Re-examination of the autopsy specimen showed that both atrial appendages were large and almost met anterior to the aortic root and it is therefore likely that the echo free space demonstrated with echocardiography

represented the left atrial appendage.

In the third patient there was dextrocardia and echocardiography suggested a type C complete AV canal with a small diameter pulmonary artery continuous with the right sided portion of the AV valve and arising from the right sided ventricle. The aorta arose from the left sided ventricle and was discontinuous with the AV valve. Chest X-ray showed dextrocardia and the bronchial anatomy suggested situs inversus. The echocardiogram was therefore interpreted as showing dextrocardia, situs inversus (from chest X-ray), a type C complete AV canal and transposition of the great arteries with a hypoplastic MPA. The angiocardiogram was interpreted by some observers as showing dextrocardia, situs inversus, univentricular heart of left ventricular type with left AV valve atresia, l-transposition of the great arteries with a subaortic outflow chamber, and pulmonary stenosis. In addition there was total anomalous pulmonary venous drainage with the left pulmonary veins entering a left sided superior vena cava while the right ones were obstructed and their site of entry to the systemic venous circulation was uncertain. Review of the echocardiogram showed no posterior echo-free space and thus TAPVD could not be suspected from echocardiography. Repeat catheterisation at the age of 20 months showed that there was situs inversus, AV concordance with a complete AV canal and transposition of the great arteries. The intracardiac anatomy thus corresponded with the initial echocardiographic assessment.

In the fourth infant the echocardiogram showed a large aorta over-riding the ventricular septum and what appeared to be a very narrow RVO and MPA. This was interpreted as showing pulmonary atresia with a ventricular septal defect or a very severe form

of tetralogy of Fallot. Angiocardiography confirmed the presence of a high VSD and over-riding aorta. However, the right lung was shown to fill from the ascending aorta and the left one through a patent ductus arteriosus; an RVO tract or proximal MPA was not shown on angiocardiography. The infant died shortly after the catheterisation and postmortem permission was refused. It is therefore uncertain whether there was a hypoplastic RVO and proximal MPA as suggested by echocardiography.

Discussion

In the management of the infant with cyanotic heart disease echocardiography is largely used before cardiac catheterisation and angiocardiography to allow better planning of the procedure. If echocardiography is going to influence therapeutic decisions, such as the degree of urgency with which cardiac catheterisation needs to be performed, the cardiologist must be aware of its abilities and shortcomings. However, much of the published work has simply described the echocardiographic features in patients known to have specific types of congenital heart disease. The author is aware of only one study of the accuracy of echocardiography in cyanotic congenital heart disease in which the authors specifically state that the echocardiogram was performed and a diagnostic opinion reached before the diagnosis was established by other means (Bass et al., 1978); these authors concluded that in transposition of the great arteries a certain diagnosis could not always be reached with M-mode echocardiography. The present study provides a prospective evaluation of the capabilities of two-dimensional echocardiography in the diagnosis of the cyanosed infant. It is not possible, however, to assess only the technique,

but rather the technique, the equipment, and the operator together. Although the author had some experience with two-dimensional echocardiography at the beginning of the study, he had not encountered all the conditions which presented during it and had little experience in small infants since the early studies with two-dimensional echocardiography were carried out with 2.5 MHz transducer, which was unsatisfactory for precise diagnosis in the newborn. Thus, the continued learning process throughout the study period allowed the diagnostic opinion to be reached more rapidly and more accurately.

In making an evaluation of two-dimensional echocardiography it is also important to consider its advantages and disadvantages over the M-mode technique. The equipment required for real-time two-dimensional studies is considerably more expensive than that for M-mode echocardiography and justification of its use therefore requires that it has definite advantages. This study was not designed to compare M-mode and two-dimensional echocardiography and the assessment was made independently of findings on M-mode echocardiography. However, it appears that in the study of the cyanosed infant two-dimensional echocardiography is of particular value in the identification of the great arteries, assessment of their relationships, and elucidation of anomalies with a single patent great artery. When two great arteries have been recognised they can be identified by their characteristic supravalvar course and an exact picture of their anatomy can be reached; where satisfactory echocardiograms were obtained no mistakes were made in the identification of the great arteries or the determination of their relations. The advantage of two-dimensional echocardiography

in this respect is illustrated by the findings in the infants with complete transposition; this study suggests that the basic anatomical diagnosis can be made with certainty with two-dimensional echocardiography, whereas Bass et al. (1978) reported that the M-mode echocardiogram was not always diagnostic. Furthermore in some patients where only a single great artery can be recognised with the M-mode technique, two-dimensional echocardiography may demonstrate another one or, if only one can be shown, identify it as a persistent truncus arteriosus or an aorta. However, the use of two-dimensional echocardiography alone limits the information which can be obtained from echocardiography. The M-mode technique provides a more accurate assessment of the movement of structures, such as the ventricular septum and atrioventricular valves, and permits measurement of right and left ventricular systolic time intervals from which an assessment of the pulmonary vascular resistance (Spooner et al., 1978) or pulmonary artery diastolic pressure (Riggs et al., 1978) can be obtained. Thus by combining both techniques a more detailed echocardiographic picture of cardiac structure and function should be obtained.

In considering the diagnostic value of two-dimensional echocardiography, it must be remembered that the technique employed in this study does not give a full anatomical profile of the heart and great arteries. It was not possible to determine the presence or absence of a patent ductus arteriosus, coarctation of the aorta, ostium secundum atrial septal defect or, with the exception of truncus arteriosus and tetralogy of Fallot, a ventricular septal defect. With the relatively large prototype scanner it was found that, in small infants, satisfactory images

could not be obtained from the suprasternal position as has been described for the diagnosis of coarctation of the aorta (Weyman et al., 1978; Sahn et al., 1977). In addition it has not been possible to reproduce the results of Sahn and Allen (1978) who reported the demonstration of the patent ductus arteriosus with the scanner in a parasternal position. Modification of the equipment or the use of other two-dimensional scanners may allow these anomalies to be demonstrated. Furthermore, valve regurgitation could not be recognised and a certain distinction could not be made between valve stenosis or atresia. Nevertheless within these limitations the correct basic diagnosis was reached with echocardiography in 54 infants (84%). Although mistakes were made in the interpretation of some of the echocardiograms this preliminary study suggests that with two-dimensional echocardiography it should be possible to reach a confident and accurate diagnosis in infants with transposition of the great arteries, tetralogy of Fallot, pulmonary valve atresia or severe stenosis (although not distinguish them with certainty), left heart hypoplasia, atrioventricular canal, univentricular heart and most of those with persistent truncus arteriosus.

Two-dimensional echocardiography would make an important contribution to patient management if it were possible to rule out structural heart disease. With the exception of the child with severe pulmonary artery hypertension, the echocardiographic appearances in the infants with respiratory disease or persistence of the fetal circulation were considered to be normal. Similarly, in 4 of the 5 infants with total anomalous pulmonary venous drainage, this was thought to be the likely diagnosis from

echocardiography. However the significance of the echo-free space behind the left atrium seen in 4 of the infants with TAPVD is uncertain. A similar space can also be seen on occasions in normal subjects and the differentiation of infants with TAPVD from those with normal anatomy was based on the demonstration of relatively small left heart chambers and aorta, the echo-free space only being considered as corroborative of the diagnosis. This is unlikely to be sufficient in all cases for the certain distinction of TAPVD from respiratory disease on the basis of echocardiography alone. Thus in the third child in the "Miscellaneous" group there was pulmonary stenosis and therefore reduced pulmonary blood flow and the presence of TAPVD could not be suspected on echocardiography even on reviewing the study. Perhaps further studies to determine whether the common pulmonary venous trunk (when present) can always be demonstrated in the subxiphoid view (Bierman and Williams 1979a) will allow these conditions to be distinguished with greater certainty.

It is disturbing that an incorrect assessment should be made in 5 patients. Review of the findings show that in 3 (univentricular heart (2) and TAPVD) the observer misinterpreted the appearances due to insufficient care and experience in examining the echo-cardiogram; further experience and meticulous examination technique should ensure that such errors are not repeated. In the infant with pulmonary atresia and absence of an RVO and MPA the echo free structure which was considered to represent them was probably the left atrial appendage, and the failure to demonstrate the pulmonary valve echo should have suggested that the interpretation was incorrect. Furthermore the RVO was not shown to be in communication

with the right ventricle and this indicates that a view demonstrating this continuity should always be obtained. The incorrect diagnosis of structural congenital heart disease in one infant with severe pulmonary hypertension may have been related to alterations in the haemodynamics, the systolic pressure being higher in the right ventricle than in the left. It would seem that care may have to be exercised in interpreting the significance of apparent aortic over-ride where severe pulmonary hypertension is suspected. Consideration of the echocardiogram in the light of clinical and other non-invasive findings should permit the correct interpretation of the echocardiogram in such cases.

The importance of two-dimensional echocardiography as a diagnostic technique is exemplified in several cases by the provision of information not obtained by cardiac catheterisation and angiocardiography. In one of the patients with pulmonary valve atresia the main pulmonary artery was not demonstrated by angiocardiography, but echocardiography showed it to be of almost normal size, allowing the correct surgical approach to be undertaken. The type of truncus arteriosus was not certain on angiocardiography in the infant with the type II abnormality but echocardiography established the correct diagnosis. Furthermore, in some cases interpretation of the angiocardiogram has been questioned on the basis of the echocardiographic findings. In the infant in whom some observers considered the diagnosis to be univentricular heart with persistent truncus arteriosus, echocardiography showed a posterior hypoplastic (and presumably atretic) MPA. The patient died suddenly at home the week before he would have been admitted

for repeat angiocardiographic studies and a postmortem examination was not performed. Although the full diagnosis was not reached with echocardiography in the patient with juxtaposition of the atrial appendages, the echocardiographic assessment of the defect showed better correlation with the autopsy findings than the angiocardiographic one. Furthermore, in the infant with dextrocardia, echocardiography in the neonatal period correctly determined the intracardiac anatomy, while an incorrect diagnosis was made on angiocardiography. These results indicate that two-dimensional echocardiography is not only useful in reaching the diagnosis by non-invasive means and thus in planning cardiac catheterisation and angiocardiography, but in some conditions it may be the only method by which the correct diagnosis can be reached.

CHAPTER VII

C O N C L U S I O N

This thesis has described a large part of my first three years' experience with two-dimensional echocardiography which, at the start of this period, was a new diagnostic technique. The original design and subsequent modification of the scanning equipment have been outlined, but the main aim has been to establish and describe standard scanning planes and to apply them in the study of congenital heart disease, and to make an assessment of the diagnostic accuracy of the technique. The experience gained from these studies allows certain conclusions to be drawn about the approach to the diagnosis of congenital heart disease with two-dimensional echocardiography and about its diagnostic value; this chapter will consider these two subjects.

VII, 1 THE DIAGNOSTIC APPROACH WITH TWO-DIMENSIONAL ECHOCARDIOGRAPHY

In my early studies of patients with congenital heart disease an attempt was made to demonstrate abnormal findings in the standard longitudinal and transverse views, and then correlate them with specific cardiac defects. It subsequently became apparent that by the use of additional views (high longitudinal and four chamber) a more complete analysis of the cardiac anatomy could be obtained, and this progressive modification of the examination technique improved its diagnostic value as the work progressed. In reviewing my own findings and considering those of other workers, it is apparent that the description of the normal views and their variations in different abnormalities does not provide a clear idea of what I now consider should be the standard examination procedure in congenital heart disease. It is important that anyone learning the technique, providing a clinical service, or undertaking further research with two-dimensional echocardiography should not record simply the conventional views, but should adopt a more detailed diagnostic approach. Such an approach can now be suggested based principally on the work described in this thesis. However, in some aspects of the examination the findings of other workers may provide additional information, and these will therefore be discussed even if the author has not undertaken detailed confirmatory studies.

The approach to the diagnosis of the patient with congenital heart disease with two-dimensional echocardiography should, as far as possible, be based on the same well established principles of sequential chamber localisation which apply to other techniques

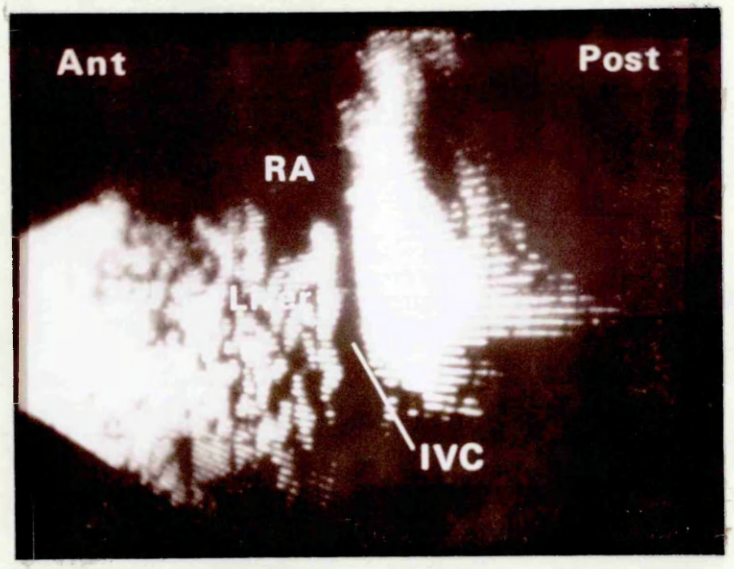


Figure 7, 1 Ultrasonic scan from an infant with the scanning plane adjusted to lie along the inferior vena cava and demonstrate its junction with the right atrium. Ant, anterior; Post, posterior; IVC, inferior vena cava. Other abbreviations as before.

(Shinebourne et al., 1976; Tynan et al., 1979). The aim must be to identify the various cardiac segments, determine their connections and define any associated anomalies. To this end the individual segments which must be identified are the atrial chambers, ventricular chambers, and the great arteries. In the echocardiographic examination the same views may be used both to identify the chambers and to determine connections; the description of this diagnostic approach will follow the lines in which the echocardiographic assessment should be routinely performed.

Identification of the atrial situs

Two-dimensional echocardiography may provide some assistance with the determination of atrial situs by the demonstration of the atrial connection of the suprahepatic inferior vena cava (IVC); as a general rule, except in cases of situs ambiguus, this will be into the morphological right atrium (Van Praagh, 1972). If the scanning plane is positioned over the liver just below the xiphisternum and adjusted to lie in a longitudinal plane, a long axis view of the IVC can be obtained (Figure 7, 1). By rotating the scanning plane through a right angle the IVC is shown in cross section as an approximately circular echo-free space. Its atrial connection is then determined by tilting the scanning plane upwards to obtain a four chamber view of the heart; as this manoeuvre is performed the IVC is followed upwards and the atrium into which it enters can be determined. Where there is situs solitus or inversus this will be, with few exceptions (Black et al., 1964; Gardner and Cole, 1955), the morphological right atrium and, if the atrial septum is visualised in this or another four chamber view, the morphological left atrium can also be identified. The

major limitation to the application of this rule is its unreliability in cases of situs ambiguus (Van Praagh, 1972), in which difficulties in the determination of the atrial situs most commonly occur. Thus two-dimensional echocardiography cannot at present provide a reliable method of determining atrial situs, and a sequential diagnosis with this technique must be based upon the determination of the atrial situs from a chest x-ray (Van Mierop, 1968).

Atrioventricular connections

The atrioventricular (AV) connections are most clearly demonstrated in a four chamber view showing (where present) the ventricular and atrial septa and the anterior leaflets of the AV valves. The relationship of the ventricular septum (where present) to the AV valve rings in diastole will allow the atrioventricular connections to be determined. A normally patent AV valve is recognised by its characteristic pattern of anterior motion and, as described in Chapter VI, an atretic one as an immobile echo in the position of an AV valve.

Ventricular identification

Certain features on two-dimensional echocardiography may suggest the ventricular morphology but none has, as yet, been shown to be conclusive; the significance of two of these features, the presence of a conus and the relative positions of the atrioventricular valves, has been discussed in detail in Chapter IV, 2. The presence of a conus recognised as discontinuity between an AV and semilunar valve in the longitudinal view suggests a morphological right ventricle; similarly AV-semilunar valve continuity suggests the absence of a conus and a morphological

left ventricle. However, as the morphological left ventricle may possess a conus in some instances, this criterion will not always permit determination of ventricular morphology. In the apical four chamber view it has been suggested that one ventricle appears more trabeculated than the other, and can thus be identified as the morphological right ventricle (Silverman & Schiller, 1978); this has not been studied in detail for the purposes of this thesis, but with this scanning equipment such a distinction has not been apparent. With a normal heart the four chamber view shows the tricuspid valve impinging on the ventricular septum in an antero-inferior position with respect to the mitral valve; if this relationship holds in complex abnormalities it may allow more reliable ventricular identification, but only where the ventricular septum and anterior AV valve leaflets are clearly shown.

A univentricular heart is likely to be present when a ventricular septum cannot be seen in any view, and can be diagnosed with certainty where a septum separates the main ventricular chamber from one with which less than half an AV valve connects. At present there is no certain means by using two-dimensional echocardiography to determine the ventricular morphology in univentricular heart. However, if a chamber is recognised in situs solitus, an antero-superior position will suggest a univentricular heart of left ventricular type with a rudimentary chamber of right ventricular type, while a postero-inferior chamber would be in favour of a univentricular heart of right ventricular type with a rudimentary chamber of left ventricular type. If no chamber can be identified the ventricle

is likely to be of indeterminate type although the presence of a small anterior chamber cannot be excluded with certainty.

Ventriculo-arterial connections

When two ventricles, two great arteries, and a ventricular septum are shown their connections are usually easily established. As described in Chapter IV, 5, where a great artery over-rides the ventricular septum it is suggested that their relative positions in diastole are used to determine connections. The longitudinal view most commonly shows the relationship of the ventricular septum to only one of the great arteries (usually the posterior); its relationship to the other can then be determined in a short axis view by sweeping the scanning plane from a low position, showing the ventricular septum, upwards to the great artery roots, and estimating their positions relative to the septum. A great artery is considered to be connected to the ventricle from which more than 50% of the circumference of its root appears to take origin.

Great artery identification

A great artery in longitudinal view is recognised as two parallel lines and in transverse view as a circle. When both great arteries are seen they are differentiated in the longitudinal view by the posterior turn of the main pulmonary artery at a lower level than the aorta. If only one artery is shown it may be possible to identify it as the pulmonary artery by the demonstration, in the transverse view, of the bifurcation. A persistent truncus arteriosus can in a minority of cases be identified by the demonstration of more than three cusps, but it is likely that in the majority of cases the recognition of branches arising from a

vessel with a long upward course will permit the correct diagnosis to be established. To be certain that the parallel lines or circle represent a great artery, the semilunar valve must be recognised. In a longitudinal view a semilunar valve will appear, in diastole, as a central linear echo between the parallel walls of the great artery root. In transverse view the three separate cusps of a normal tricuspid semilunar valve may be apparent although more often only a single linear echo is clearly seen.

Identification of associated anomalies

It may be possible, with two-dimensional echocardiography, to demonstrate some anomalies within the different cardiac segments. Although the IVC and pulmonary veins may be demonstrated on occasions, my observations have not yet ascertained how reliably anomalies of venous return can be identified; the ultrasonic gain settings should however be adjusted to exclude (or identify) a posterior echo-free space (representing a common venous chamber) or a large coronary sinus suggestive of total anomalous pulmonary venous drainage. The atrial septum can be demonstrated in four chamber views; the defect characteristic of an AV canal abnormality is easily identified and the subxiphoid view has been reported reliably to demonstrate an ostium secundum atrial septal defect in infants (Bierman and Williams, 1979b). Longitudinal, transverse, and four chamber views must all be studied for abnormalities in the motion and position of the atrioventricular valves; for instance, a four chamber view showing the tricuspid valve ring in the normal position with anterior displacement of the tricuspid valve leaflets would suggest Ebstein's anomaly (Ports et al., 1978), and an immobile valve echo would suggest atresia. The ventricular

septal defect associated with tetralogy of Fallot or persistent truncus arteriosus can be easily recognised but it has seldom been possible to demonstrate with certainty other types of VSD. Longitudinal views of great artery roots allow their diameter to be assessed, and doming of the semilunar valves may be demonstrated where there is severe stenosis (Weyman et al., 1975; Weyman et al., 1977) or atresia. It has been reported that it is possible to demonstrate a patent ductus arteriosus (Sahn and Allen, 1978), and, by using suprasternal transducer placement, to show aortic narrowing in coarctation of the aorta (Weyman et al., 1978; Sahn et al., 1977).

Thus a number of associated anomalies may be demonstrated with two-dimensional echocardiography but, in this respect, care in interpreting the study is necessary since failure to demonstrate a structure, or a defect in a structure, does not in itself exclude its presence with certainty.

VII, 2 THE PLACE OF TWO-DIMENSIONAL ECHOCARDIOGRAPHY IN PAEDIATRIC CARDIOLOGY

On the basis of the studies described in this thesis it is possible to make an interim assessment of the value of two-dimensional echocardiography in the management of patients with congenital heart disease. As detailed studies of all types of congenital heart disease have not been performed this cannot be fully comprehensive, but rather represents the author's opinion as to the place of two-dimensional echocardiography in the investigation of conditions described in the preceding chapters.

Two-dimensional echocardiography can most easily recognise gross abnormalities of the intracardiac or great artery anatomy and the studies on congenital heart disease described in this thesis have, for the most part, concentrated on this group of patients. There is no doubt that in those with complex congenital heart disease the technique adds significantly to the diagnostic information obtained from other non-invasive investigations such as clinical examination, electrocardiography, radiology, and even M-mode echocardiography. Complex congenital heart disease frequently presents with cyanosis, and it is therefore in the cyanosed infant that the influence of two-dimensional echocardiography on patient management has first become apparent in the Royal Hospital for Sick Children, Glasgow. In the majority of cases a basic diagnosis can be made with two-dimensional echocardiography and when considered with the infant's clinical status this allows an accurate assessment of the urgency with which cardiac catheterisation should be undertaken. With an experienced observer, the diagnosis of left heart hypoplasia with aortic atresia is considered as

definitive and, as surgery is not yet feasible, to preclude the need for further investigation, while that of transposition of the great arteries to warrent urgent cardiac catheterisation with the intention of performing an atrial septostomy. Furthermore, in some other infants (e.g. those with tetralogy of Fallot or univentricular heart), a definite echocardiographic diagnosis has allowed the cardiologist to defer cardiac catheterisation and angiocardiography for some weeks or months until the baby has grown bigger and surgery is considered necessary. When cardiac catheterisation has to be performed the information on the intracardiac and great artery anatomy obtained from echocardiography minimises the number of angiocardiographic contrast injections required and usually allows the examination to be performed more rapidly than it could otherwise be undertaken. It is disappointing that congenital heart disease cannot, as yet, be ruled out with certainty with echocardiography; although the diagnosis of total anomalous pulmonary venous drainage can usually be inferred, the distinction between it and persistence of the fetal circulation or pulmonary disease may not always be possible. Thus in some infants echocardiography may not give the diagnosis with certainty, but the echocardiographic findings can be used to rule out other cardiac anomalies which could cause the cyanosis.

There is, as yet, a need for caution in basing management decisions on echocardiography alone as exemplified by the misdiagnosis in 5 of the infants described in Chapter VI. However it must be remembered that it is not the technique of echocardiography which is "wrong"; the equipment merely displays the returning echoes and its abilities are limited by its design

and the physical characteristics of ultrasound. Any mistakes are therefore due to deficiency in the interpretation of the echocardiogram and are likely to be due to the physician's lack of experience of echocardiography. These incorrect diagnostic interpretations were all attributable to errors which could be explained on review of the echocardiogram, and the same mistakes should not be repeated. Mistaken diagnostic interpretations should not be permitted to bring the technique into disrepute but should indicate the need for further experience with it. Thus, at present, care must be exercised in interpreting the echocardiogram, and if the clinical and x-ray findings are not consistent with the echocardiographic diagnosis, therapeutic decisions should not be based on echocardiography alone.

It is less easy to be certain of the value of two-dimensional echocardiography in the management of the older child, in whom the need for performing cardiac catheterisation and angiocardiography should be apparent on clinical grounds, and speed of catheterisation is of less importance than in the infant. The main aim of echocardiography in this group must be to provide information not readily obtained from angiocardiography or other studies. My initial experience with two-dimensional echocardiography suggests that the outstanding example of its value in addition to angiocardiography lies in the elucidation of atrioventricular valve abnormalities. In atrioventricular canal anomalies two-dimensional echocardiography can provide more detailed definition of the anatomy of the atrioventricular valve (or valves) and their chordal attachment than can be obtained with angiocardiography. Thus the echocardiographic appearances in atrioventricular canal

malformations are now taken into consideration by the surgeons in deciding upon their approach to the correction of the defect. The atrioventricular valve abnormalities associated with the univentricular heart to a large extent determine the feasibility of surgical repair and the approach which must be undertaken. Difficulty may be encountered in defining these abnormalities by angiocardiography and, even with the use of the newer projections, details such as the atrioventricular valve morphology and chordal attachments of the papillary muscles cannot easily be shown. M-mode echocardiography has already been shown to be of value in the study of the univentricular heart (Mortera et al., 1978), but two-dimensional echocardiography, with its ability to demonstrate (where present) the ventricular and atrial septa and both atrioventricular valves simultaneously, should permit more rapid and exact definition of the intracardiac anatomy. The results in newborn infants with univentricular heart described in Chapter VI indicate the importance of further studies in this group of abnormalities.

Two-dimensional echocardiography is not likely to be of such great value in the investigation of the patient with the less complex forms of congenital heart disease. With the technique it has been reported that it is possible to demonstrate valve doming in aortic and pulmonary stenosis (Weyman et al., 1975; Weyman et al., 1977), narrowing of the aorta in coarctation (Sahn et al., 1977; Weyman et al., 1978) and the presence of a patent ductus arteriosus (Sahn & Allen, 1978) and in some cases an ostium secundium atrial septal defect (Bierman & Williams, 1979b). Furthermore by the use of contrast injection through a peripheral

vein intracardiac right to left shunting has been demonstrated in some patients with an ASD or VSD and raised pulmonary artery pressure (Serruys et al., 1979). However in these conditions there is usually less difficulty in making the diagnosis with conventional methods and simple confirmation of this would be of only limited importance as a new investigation. It would however be important if two-dimensional echocardiography could provide an assessment of the severity or of the haemodynamic consequences of these lesions. It has been reported that the orifice area of the stenosed valve and the diameter of a patent ductus arteriosus can be measured accurately with echocardiography, but knowledge of the valve area or defect size is of much less importance than the measurement of pressure gradient over a stenosed valve or the blood flow through a septal defect. Nevertheless by providing a pictorial demonstration of the presence of some of these lesions two-dimensional echocardiography may obviate the need for a diagnostic catheterisation in some cases.

As the work embodied in this thesis progressed two-dimensional echocardiography became an established investigation in the Cardiology Department of the Royal Hospital for Sick Children. It is now becoming accepted as an essential investigative technique for all major paediatric cardiology centres. It is difficult to speculate on the ultimate place of two-dimensional echocardiography in paediatric cardiology. In the next few years it will be important not only to explore other uses of the technique, but also to consolidate the work which has already been done and assess its validity. It is unlikely that two-dimensional echocardiography will supplant cardiac catheterisation and angiocardiography as the

definitive diagnostic technique in paediatric cardiology. However, by combining the information from two-dimensional and M-mode echocardiography with that derived from other relatively new techniques, such as Doppler ultrasound and radionuclide angiography it may in the future be possible to obtain information on such features as the intracardiac anatomy, pulmonary artery pressure, and intracardiac shunt size, which will be sufficiently accurate to make cardiac catheterisation and angiocardiography unnecessary in some conditions.

Over the years other new techniques for the investigation of heart disease such as phonocardiography, indicator dilution studies, and ballistocardiography have been advocated with considerable enthusiasm, but have subsequently proved to be disappointing and to be of only limited value in paediatric cardiology. These techniques, however, have been able to provide only indirect information on cardiac defects, whereas ultrasound gives a precise demonstration of the intracardiac anatomy. It is therefore probable that echocardiography will take its place alongside electrocardiography and chest radiology as a standard and routine non-invasive investigation.

Two-dimensional echocardiography has the unique advantage over other non-invasive investigations in its ability rapidly to provide a precise diagnosis in many forms of complex congenital heart disease. However, increasing confidence in its capabilities must not result in a failure to give full consideration to the results of longer established non-invasive techniques. I or anyone tempted to underestimate their value has only to remind himself that echocardiography cannot yet differentiate between pulmonary atresia

and severe pulmonary stenosis, but the presence or absence of a pulmonary ejection murmur rapidly allows this distinction to be made simply by listening to the heart.

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