

Ventricular septal defects

Two dimensional echocardiographic and morphological correlations

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SUMMARY To evaluate the ability of two dimensional echocardiography to identify and classify ventricular septal defects, 280 infants and children with clinically significant ventricular septal defects were studied. Multiple precordial and subcostal echocardiographic planes were scanned in each patient in an attempt to identify the defects. Defects visualised were classified on the basis of the structures which formed their margins. Subsequent correlation of this information with angiographic (280 patients), surgical (130 patients), and pathological (31 patients) data confirmed that defects in the following sites produced a specific two dimensional echocardiographic pattern. (a) Perimembranous inlet, (b) perimembranous outlet, (c) muscular inlet, (d) single trabecular, (e) muscular outlet, and (f) doubly committed subarterial. A defect was identified and correctly classified in 252 patients. Individual defects were identified with varying degrees of accuracy. All subarterial (24 patients) defects were correctly identified and classified, as were muscular defects of the inlet (18 patients) and outlet (six patients) septa. Of the 185 perimembranous defects, 182 were identified. Only 23 of the 43 single trabecular defects were identified. Small multiple ("Swiss cheese") defects (four patients) were not identified.

We conclude that two dimensional echocardiography provides a reliable non-invasive method of identifying and classifying the following ventricular septal defects: (a) perimembranous defects, (b) doubly committed subarterial defects, and (c) muscular defects of the inlet and outlet septa. In our experience it fails consistently to visualise defects in the trabecular septum.

When investigating a ventricular septal defect it is not enough simply to diagnose the presence of a hole in the ventricular septum. The ideal investigative technique should consistently identify the defect and should define its site, its size, and the structures forming its margins, thus providing the information that will allow accurate morphological classification. Two dimensional echocardiography ought to provide this information, as the precision of the technique is such that it can image cardiac morphology in detail and can identify small defects in the ventricular septum.¹⁻³ It should therefore be possible to classify defects by determining whether they are (i) roofed in part by the

central fibrous body and aortic valve (perimembranous defects), (ii) are entirely surrounded by muscle (muscular defects), or (iii) are roofed by conjoint aortic and pulmonary valves (doubly committed subarterial defects). This report describes an echocardiographic investigation of 280 patients known to have ventricular septal defects in one or other of these sites and the morphological studies carried out to validate our echocardiographic classification.

Subjects and methods

Two hundred and eighty patients with clinical evidence of ventricular septal defect, existing as either an isolated lesion or as part of a complex malformation, were studied by two dimensional echocardiography. The echocardiograms were recorded using either a

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2.4 MHz Toshiba phased array sector scanner or a 3.0 or 5.0 MHz Advanced Technology Laboratories mechanical sector scanner. The patients' ages ranged from day 1 of life (at 36 weeks gestation) to 17 years, with the majority of patients between the ages of 1 week and 7 years. All the patients attended either the Regional Cardiothoracic Unit, Freeman Hospital, The Hospital for Sick Children, Great Ormond Street, or the Royal Hospital for Sick Children, Edinburgh. In 31 patients the hearts were subsequently available for pathological sectioning and these were supplemented by the examination of selected hearts from the Cardiopathological Collection of the Cardiothoracic Institute, Brompton Hospital, London.

Of the 280 patients studied, 224 had a ventricular septal defect as the sole cardiac lesion. In the remaining 56 patients the septal defect was part of a more complex malformation such as tetralogy of Fallot (21 patients), double outlet right ventricle (17 patients), complete transposition (seven patients), truncus arteriosus (two patients), and pulmonary atresia (nine patients). These patients were included because the same echocardiographic information could be used to differentiate between perimembranous, muscular, or subarterial types of defect.

All patients had atrioventricular concordance. We excluded patients with any of the following lesions: (i) atrioventricular discordance, (ii) ambiguous atrioventricular connection, (iii) abnormal modes of atrioventricular communication (for example common, straddling, or imperforate atrioventricular valve), and (iv) univentricular heart, as in each of these the ventricular septal defect is only one component of a complex combined atrioventricular junction-septal malformation.

Patients were studied either prospectively or retrospectively. Group 1 comprised 149 patients who were known to have a ventricular septal defect from previous angiography. Group 2 comprised 131 patients who were studied prospectively and in whom the site of the defect was proven at subsequent angiography using the angled views of Soto *et al.*⁴ Thus each patient had angiographic correlation of the presence of a defect. Of the patients, 130 had additional surgical description of their defect.

A standard echocardiographic procedure was used throughout the study. The heart was visualised from both the precordial and subcostal echocardiographic windows and the following standard echocardiographic planes⁵ scanned and recorded, from the precordial approach (a) four chamber (apical and parasternal), (b) four chamber plus aortic root (apical and parasternal), (c) short axis of the great vessels, (d) short axis at ventricular level, (e) long axis and, subcostally (x) four chamber, (y) four chamber plus aortic root, and (z) right ventricular outflow (Fig. 1). An

essential part of each investigation was a scan from one plane to another, building up in the process a three dimensional model of the heart.

The echocardiograms recorded were subsequently reviewed by a second investigator and the findings of both compared before a final decision was made as to the presence of a defect and its classification. (In patients already known to have a defect from previous angiographic investigation, the echocardiographer was unaware of the result of the earlier study.)

The two dimensional echocardiographic feature used to identify a ventricular septal defect was the presence of a discrete area of septal "drop out" persisting throughout systole and diastole in one or more of the echocardiographic planes recorded. In every case where such a defect was visualised an attempt was made to assess the extent of the defect by noting into which of the neighbouring echocardiographic planes it extended and to identify the structures which formed its margins. Having determined the above echocardiographic features, defects could then be classified into one of the following morphological groups: (a) perimembranous defects, (b) muscular defects, or (c) doubly committed subarterial defects (Fig. 2).

This classification of ventricular septal defects is based on that proposed by Moulart *et al.*,⁶ subsequently modified by Soto *et al.*,⁷ and further refined for use in this study.

MORPHOLOGICAL SUBDIVISION OF VENTRICULAR SEPTAL DEFECTS

(a) *Perimembranous defects*

These defects always involve the whole or part of the area which is occupied by the membranous septum in the normal heart. They are roofed in part by the central fibrous body and aortic root. Virtually all extend into one or more of the septal components which border the membranous septal area. Thus the whole group are referred to as perimembranous defects. These defects are subdivided depending on whether they extend primarily into the inlet, trabecular, or outlet septa (Table). Large perimembranous defects may involve all three of the surrounding septal components.

(b) *Muscular defects*

Defects surrounded entirely by muscular rims may be subdivided into those localised to the inlet, trabecular, or outlet components of the septum. Trabecular defects may be of single or multiple (Swiss cheese) types (Table).

(c) *Doubly committed subarterial defects*

The characteristic feature of these defects is that they

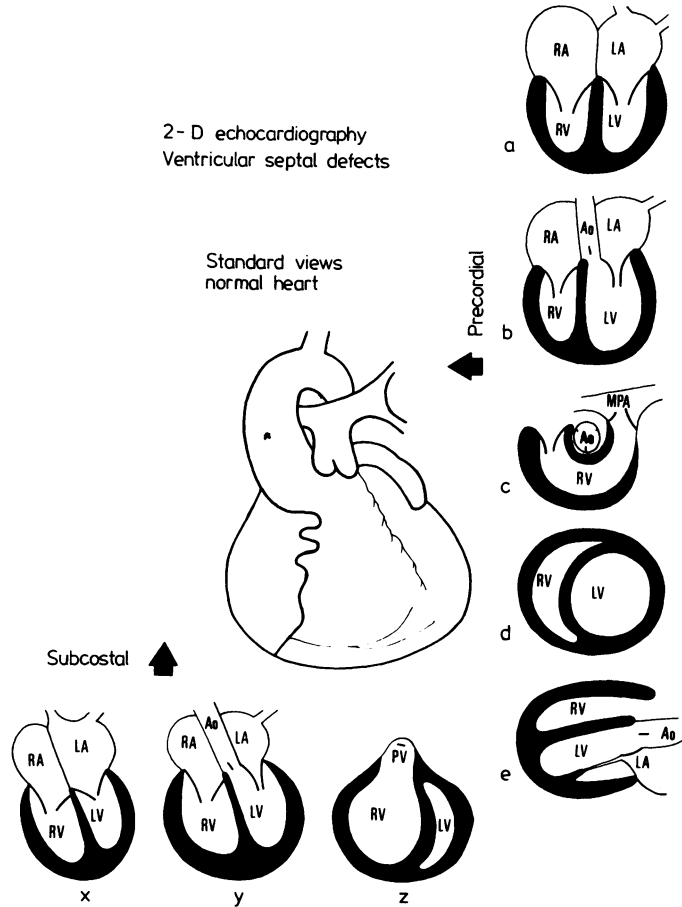


Fig. 1 The two dimensional echocardiographic planes discussed in this study are as follows. Precordial: (a) four chamber; (b) four chamber plus aortic root; (c) short axis great vessels; (d) short axis at ventricular level; (e) left ventricular long axis. Subcostal: (x) four chamber; (y) four chamber plus aortic root; (z) right ventricular outflow tract (an identical sequence is used in Fig. 4b, 5b, 6b, 7b, 8b, and 10b). RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; Ao, aorta; PV, pulmonary valve; MPA, main pulmonary artery.

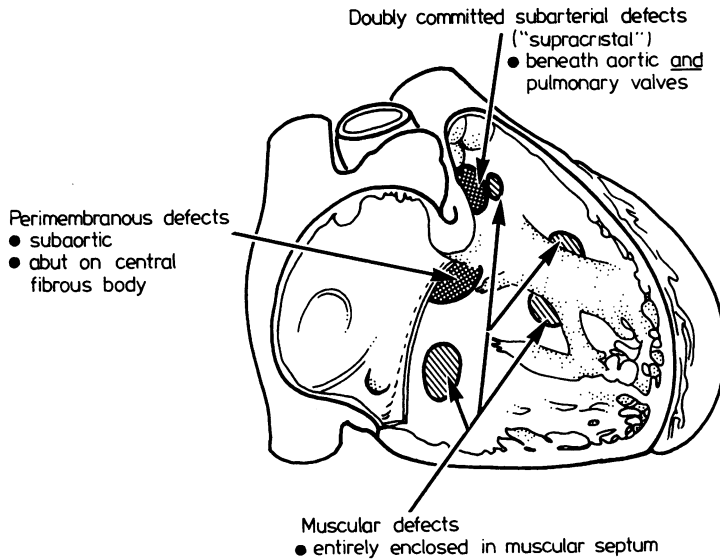


Fig. 2 A diagram of the right ventricular septal surface illustrating the three main groups of defects.

Table Two dimensional echocardiography of ventricular septal defects

Types of defect	No. studied	No. correctly identified	% correctly identified
<i>Perimembranous (extending mainly into)</i>			
(a) Inlet septum	43	43	100
(b) Outlet septum	84	83	98
(c) Trabecular septum	19	16	84
(d) All of the above	39	39	100
<i>Muscular</i>			
(a) Inlet	18	18	100
(b) Outlet	6	6	100
(c) Single trabecular	32	23	72
	11	0	0
(d) Multiple trabecular	4	0	0
<i>Doubly committed subarterial</i>			
(a) Not involving membranous septum	15	15	100
(b) Extending into membranous septal area	9	9	100
Total	280	252	

are roofed by aortic and pulmonary valve rings which lie at the same level and are in fibrous continuity. This abnormal situation is caused by absence of the normal infundibular septum which supports the pulmonary valve ring. Doubly committed subarterial defects may be subdivided into those that extend posteriorly to involve the membranous septum and those that are separated from it by a muscle bar (Table).

Having classified each ventricular septal defect on the basis of the two dimensional echocardiographic features, the results were then correlated with angiographic, surgical, and pathological findings to determine if individual defects could be reliably identified by a specific echocardiographic pattern.

Results

Correlation of the two dimensional echocardiographic information with the angiographic, surgical, and pathological findings disclosed that each type of ventricular septal defect was identified by a specific echocardiographic pattern.

TWO DIMENSIONAL ECHOCARDIOGRAPHIC FEATURES WHICH DIFFERENTIATE SPECIFIC VENTRICULAR SEPTAL DEFECTS

(1) *Perimembranous defects* (185 patients)

In the normal heart the interventricular portion of the membranous septum is bounded superiorly by the atrioventricular portion of the membranous septum, anteriorly by the subaortic outlet septum, inferiorly by the trabecular septum, and posteriorly by the inlet septum. Two dimensional echocardiography will begin to visualise the two portions of the membranous septum at the most anterior extent of a scan through the four chamber plane (either precordial or subcostal) (Fig. 3). In addition, at the point at which this plane merges with the four chamber plus aortic root

plane and the aortic valve is first visualised, the area of septum lying immediately below the aortic root is the interventricular membranous septum. It is consistently identified in the normal heart as a distinct short thin portion of the ventricular septum taking origin superiorly from the aortic root and expanding inferiorly to fuse with the much thicker trabecular system (Fig. 3). These features are better visualised from the subcostal than the precordial position, as the latter view foreshortens the extent of the membranous septum.

In 181 of the 185 perimembranous defects studied a discrete segment of septal echocardiographic drop out was recorded which involved the area of the interventricular membranous septum. In each of the 181 patients, the echocardiographic appearance clearly showed that the roof of this defect was that part of the aortic root which fused posteriorly with the central fibrous body. In the remaining four patients each with surgical confirmation of the type of defect (three with a perimembranous trabecular defect and one with a perimembranous outlet defect) the ventricular septum appeared intact in every echocardiographic plane.

In each of the 181 patients with a defect in the membranous septum, however, the area of septal drop out was found to extend into one or other of the neighbouring septal areas. Perimembranous defects could then be subdivided into four distinct groups on the basis of the differing echocardiographic patterns created by this extension of the defect.

(a) *Perimembranous inlet defects* (43 patients)

In every case the area of septal drop out involving the membranous septal area extended a variable distance posteriorly into the four chamber plane towards the crux of the heart. The roof of this posterior extension was formed by tissue separating the septal aspect of the tricuspid and mitral valve annuli (Fig. 4a). This tissue is comprised anteriorly of the central fibrous

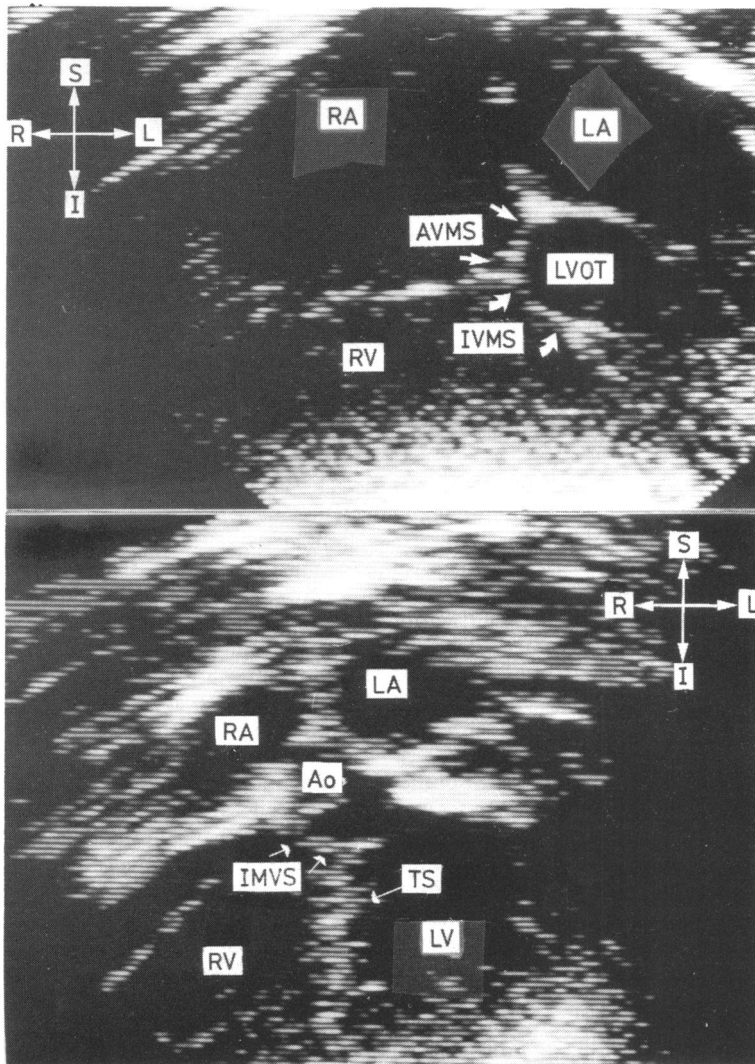


Fig. 3 A normal intact membranous septum. Upper panel: a precordial four chamber plane at its junction with a four chamber plus aortic root plane. Straight arrows, atrioventricular membranous septum; curved arrows, interventricular membranous septum. Lower panel: a four chamber plus aortic root plane at aortic valve level. The interventricular membranous septum (arrowed) is bounded superiorly by aortic root and inferiorly by trabecular septum. Ao, aorta; LV, left ventricle; LVOT, left ventricular outflow tract; RV, right ventricle; RA, right atrium; LA, left atrium; TS, trabecular septum; AVMS, atrioventricular membranous septum; IVMS, interventricular membranous septum.

body and atrioventricular muscular septum and posteriorly by the septal atrioventricular fibrous plane. In every case two dimensional echocardiography clearly showed that there was no remnant of inlet muscular septum projecting downwards from the atrioventricular junction to roof the defect.

Perimembranous inlet defects were consistently visualised (Table) using both the subcostal and precordial four chamber planes. In our experience, where the septum appears intact throughout a four chamber scan this effectively excluded an inlet component to any perimembranous defect.

From the patients studied it was evident that the larger the defect (synonymous with the greater the degree of posterior extension towards the crux) the

more frequently were the atrioventricular valve septal leaflets attached at a common level to the base of the primum atrial septum (that is to the central fibrous body) throughout the extent of the defect (Fig. 4a). Pathological sectioning confirmed that this abnormal echocardiographic appearance results from an extension of the inlet component of the defect into the atrioventricular muscular septum either considerably reducing it in size (in small inlet perimembranous defects) or totally removing it (in large defects). This excavation of the atrioventricular muscular septum, by removing the structure that supports the oblique insertion of the atrioventricular valve rings to the septum, creates the common level of septal valve attachment. This was a constant feature of the perimem-

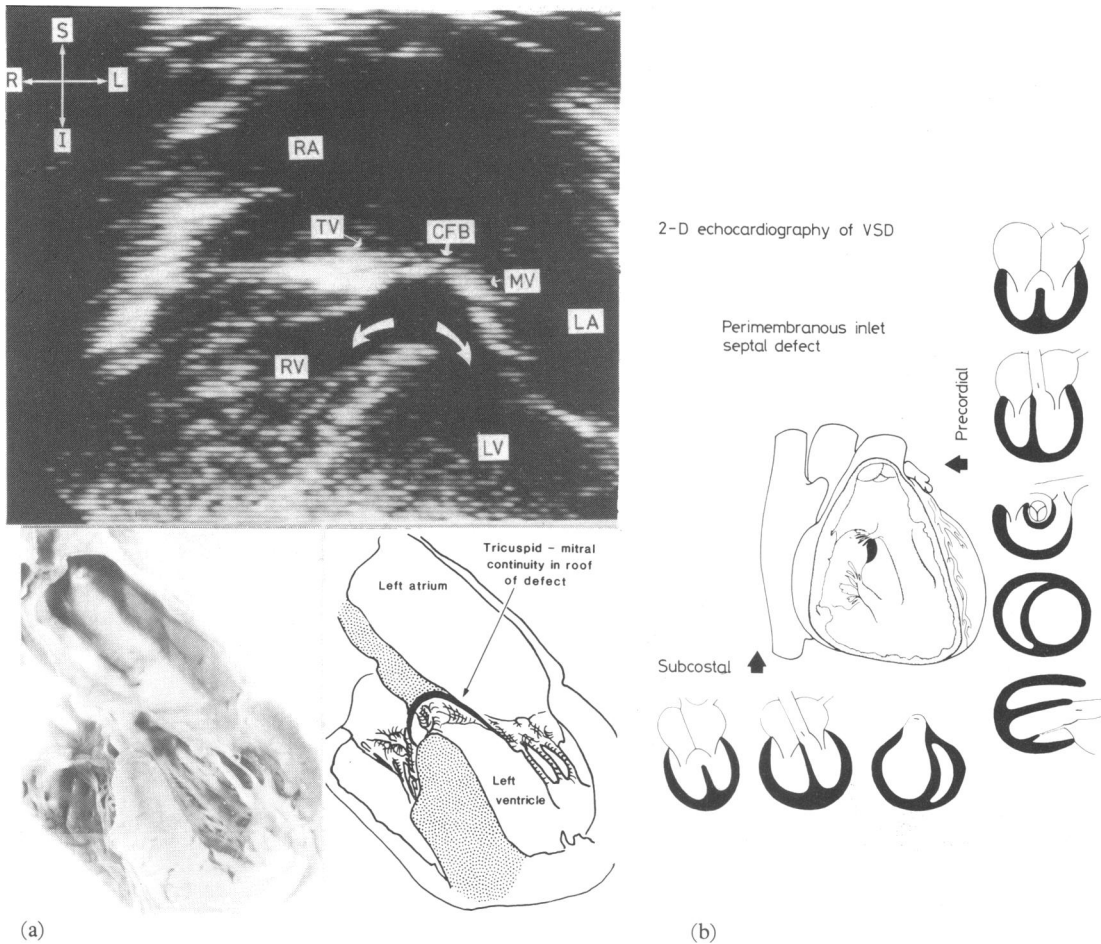


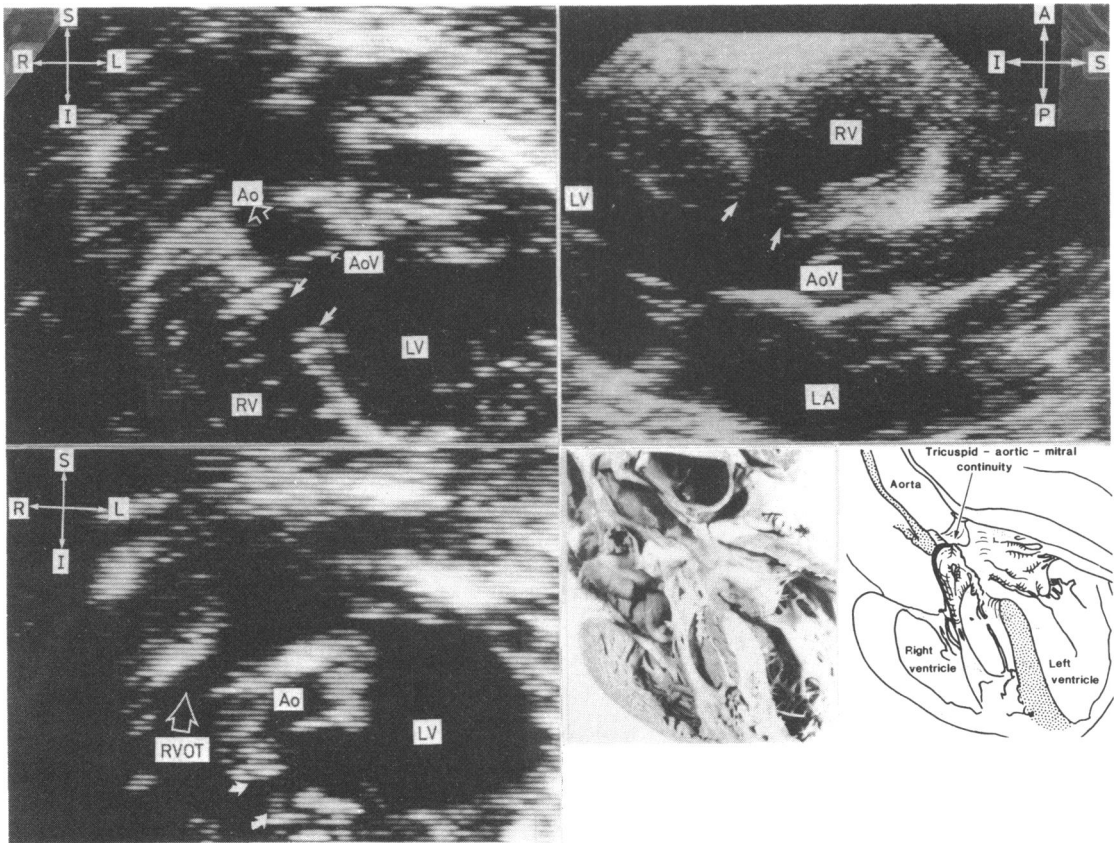
Fig. 4 (a) A perimembranous inlet defect. Upper panel: a subcostal four chamber plane. The defect (arrowed) is bounded superiorly by central fibrous body and inferiorly by trabecular septum. Note the common level of insertion of the atrioventricular valve septal leaflets to central fibrous body. Lower panels: the equivalent specimens and diagram of a perimembranous inlet defect sectioned in a four chamber plane. RA, right atrium; LA, left atrium; RV, right ventricle; LV left ventricle; MV, mitral valve septal leaflet; TV, tricuspid valve septal leaflet; CFB, central fibrous body. (b) The echocardiographic pattern diagnostic of an inlet perimembranous defect.

branous inlet defects studied. The two dimensional echocardiographic pattern and features diagnostic of a perimembranous inlet defect are illustrated in Fig. 4b.

(b) *Perimembranous outlet (subaortic or subpulmonary) defects* (84 patients)
 Eighty-four patients with such a defect were studied. In 83 of them, an area of membranous septal drop out was seen to extend anteriorly as the echo beam was swept anteriorly into the four chamber plus aortic root plane and then anteriorly across the posterior great vessel root from either the subcostal or precordial position. This echocardiographic feature confirmed the extension of the membranous defect into the outlet

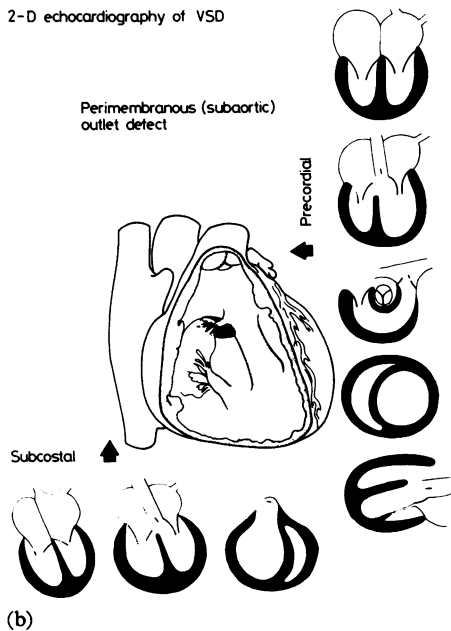
septum. (Where the posterior great vessel is the aorta then such defects are defined to be perimembranous (subaortic) outlet defects; where the posterior great vessel is the pulmonary artery then the defect is a perimembranous (subpulmonary) outlet defect.) The corresponding echocardiographic and morphological features of a perimembranous (subaortic) outlet defect are shown in Fig. 5a.

Large perimembranous outlet defects which excavate anteriorly beneath the whole posterior great vessel root were consistently visualised in the left ventricular long axis plane (Fig. 5a). Smaller defects did not extend into this plane. Perimembranous outlet defects were poorly visualised by short axis scanning. In each of the 83 patients in whom a defect was



(a)

2-D echocardiography of VSD



(b)

Fig. 5 (a) A perimembranous outlet (subaortic defect) which extends into muscular outlet septum. Upper left panel: the defect (arrowed) visualised in a four chamber plus aortic root plane (membranous septum is absent). Upper right panel: a precordial left ventricular long axis plane. The defect (arrowed) has excavated anteriorly beneath the whole aortic root. Lower left panel: a subcostal right ventricular outflow plane with the defect (arrowed) extending into the muscular outlet septum. Lower right panel: the corresponding specimen and diagram of a perimembranous outlet defect sectioned in a four chamber plus aortic root plane. RV, right ventricle; LV, left ventricle; Ao, aorta; AoV, aortic valve; LA, left atrium; RVOT, right ventricular outflow tract. (b) The echocardiographic pattern diagnostic of a perimembranous outlet defect.

identified the roof of the defect was clearly seen to be formed by the valve ring of the posterior great vessel and not by a downward projection of muscular outlet septum. The floor of the defect was seen to be part of the trabecular septum. These features allowed perimembranous outlet defects to be accurately classified with great consistency.

The two dimensional echocardiographic pattern and features diagnostic of a perimembranous (subaortic) outlet defect are illustrated in Fig. 5b.

(c) *Perimembranous trabecular defects* (19 patients)

Small membranous defects which extended inferiorly into the trabecular septum were only visualised at the most posterior aspect of the four chamber plus aortic root plane using either the subcostal or precordial view. They could also be recorded in a short axis scan but this was rare (three patients). Such defects did not extend anteriorly below the aortic valve for any significant distance. They were never visualised in the left ventricular long axis plane.

In the normal heart two dimensional echocardiography cannot determine the precise junction of membranous and trabecular septa. Thus, in theory it should prove difficult to identify when the trabecular septum becomes involved in a membranous defect. This is borne out in practice. Correlating echocardiographic with morphological findings, however, suggested two non-specific features that indicate trabecular septal involvement: (i) a blunt upper margin of the interventricular septum which forms the floor of the defect and (ii) where a membranous defect extends apically for more than half the aortic root diameter.

Nineteen patients with perimembranous trabecular defects were studied. The defect was correctly identified in 16 patients. In the three patients previously described, the ventricular septum appeared intact in every echocardiographic plane.

(d) *Large perimembranous defects* (39 patients)

Morphologically these defects all extend from the membranous septum into the inlet, trabecular, and outlet septa. These defects were correctly identified in each of the 39 patients studied and were the largest defects recorded during the entire series. It proved impossible in each case to determine which of the surrounding areas was predominantly involved and thus subdivision of these defects was not practical. The individual features of each component of such a defect were consistently visualised in the appropriate echocardiographic planes.

MUSCULAR DEFECTS (71 patients)

As indicated, the morphological feature diagnostic of a muscular defect is that it is entirely surrounded by

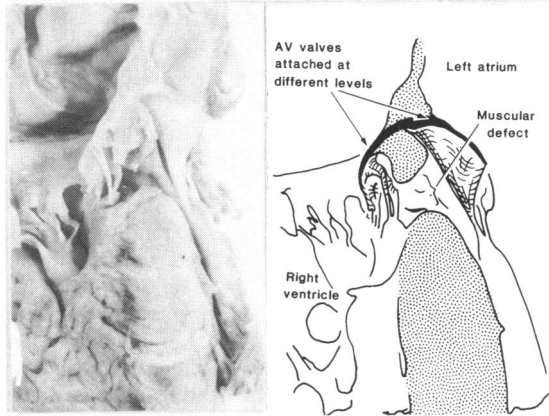
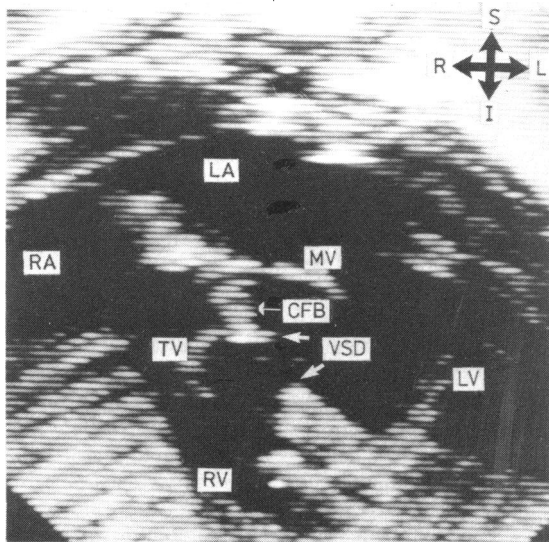
muscle and is not directly bordered by either central fibrous body or by an arterial or atrioventricular valve. The corresponding two dimensional echocardiographic features were used to identify muscular defects. Seventy-one patients with muscular defects were studied. Defects in the inlet and outlet muscular septa were consistently identified but trabecular defects were inconsistently identified. The differing echocardiographic patterns diagnostic of each type of defect are described below.

(a) *Inlet muscular defects* (18 patients)

Inlet muscular defects were consistently visualised when scanning through the four chamber plane (either subcostal or precordial) in each of the 18 patients studied. These defects were not visualised in any other plane. They always appeared larger when visualised from the subcostal approach because this view profiles the inlet septum without foreshortening it. The following echocardiographic features distinguished these defects from perimembranous inlet defects: (i) The downwards projection of a bar of muscle from the atrioventricular junction which formed the upper muscular rim of the defect and which separated the defect from the central fibrous body, (ii) a normal pattern of septal atrioventricular valve insertion (inlet muscular defects do not excavate into the atrioventricular muscular septum in contrast to inlet perimembranous defects), and (iii) when scanning anteriorly from the four chamber plane across the area of the membranous septum the muscular inlet defect is lost and the normal continuity of central fibrous body, aortic root, and membranous septum remains intact. The two dimensional echocardiographic and morphological features of a muscular inlet defect are illustrated in Fig. 6a and b.

(b) *Outlet muscular defects* (six patients)

Six defects with entirely muscular rims were located in the outlet septum by two dimensional echocardiography. The full extent of these defects was only visualised from the subcostal position when scanning anteriorly from the four chamber plus aortic root plane to the right ventricular outflow plane. Morphological sectioning of the normal heart confirmed that this is the only echocardiographic approach which will accurately profile both the subaortic and subpulmonary portions of the outlet septum. In all six patients the defect was seen to be located predominantly in the anterior aspect of the right ventricular outflow plane in a subpulmonary position but separated from the pulmonary valve by a long muscular infundibulum (Fig. 7a). In four of these patients the defect extended a short distance posteriorly into the four chamber plus aortic root plane, indicating involvement of the muscular septum below the aortic



(a)

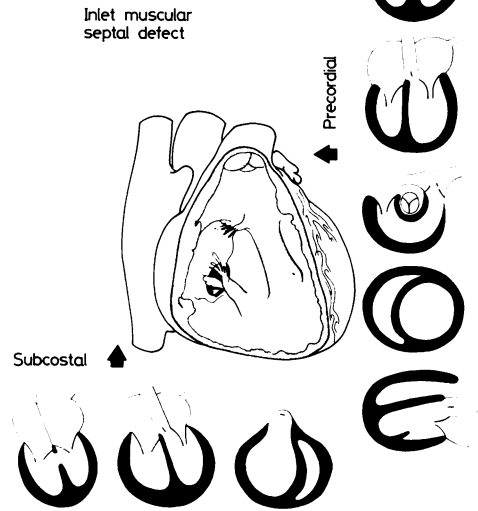
root. In each case the defect was separated from the aortic root by a bar of muscle. No defect extended posteriorly to the level of the membranous septum. Fig. 7c shows the corresponding morphological features of such a defect. The two dimensional echocardiographic features and pattern diagnostic of an outlet muscular defect are illustrated in Fig. 7a and b.

(c) *Trabecular muscular defects* (47 patients)

Defects in the trabecular portion of the muscular septum were inconsistently identified by two dimensional echocardiography.

Single trabecular septal defects (43 patients) could be recognised. These defects were best visualised on short axis scanning (either precordial or subcostal). They could be clearly located in either the anterior or posterior trabecular septum and never involved the

2-D echocardiography of VSD

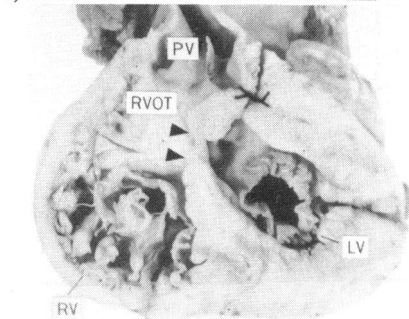
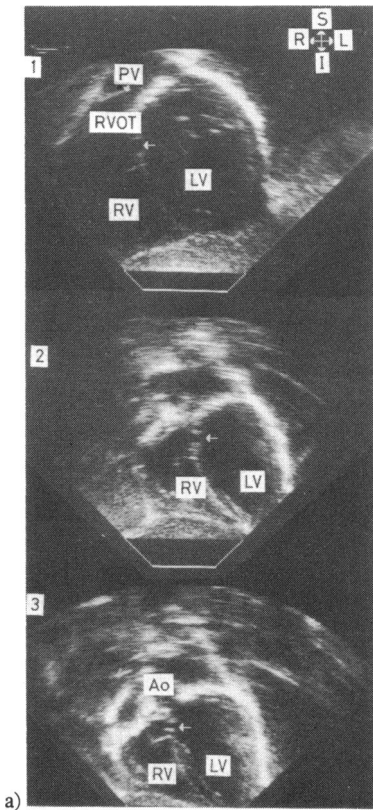


(b)

Fig. 6 (a) An inlet muscular defect shows the following. Upper panel: the defect (between arrows) visualised in a four chamber plane. Note the atrioventricular valve septal leaflets are well separated by the intact atrioventricular septum. A muscle bar projects downwards from central fibrous body to roof the defect. Lower panel: the corresponding pathological specimen and diagram of an inlet muscular defect. RA, right atrium; LA, left atrium; RV, right ventricle; LV, left ventricle; TV, tricuspid valve septal leaflet; MV, mitral valve septal leaflet; CFB, central fibrous body. (b) The echocardiographic pattern diagnostic of an inlet muscular defect.

membranous septum (Fig. 8a and b). Where the defect was recognised on short axis scanning it was usually also identified on four chamber or long axis scanning but this was not always the case. Some surprisingly large single trabecular defects ($Qp:Qs > 1.5:1$) were missed, however, on two dimensional scanning. Small single trabecular defects ($Qp:Qs < 1.5:1$) were never identified in this series.

Multiple small trabecular defects (four patients) (the so-called Swiss cheese defect) were not identified in this study. In each of the four patients studied the septum appeared intact in every plane recorded. The problems encountered in defining multiple small trabecular defects (and single small trabecular defects) were explained by examining their morphology. Fig. 9a shows a heart with multiple small trabecular defects easily visualised when viewed from the right ventricular surface of the septum. Fig. 9b shows the



2-D echocardiography of VSD

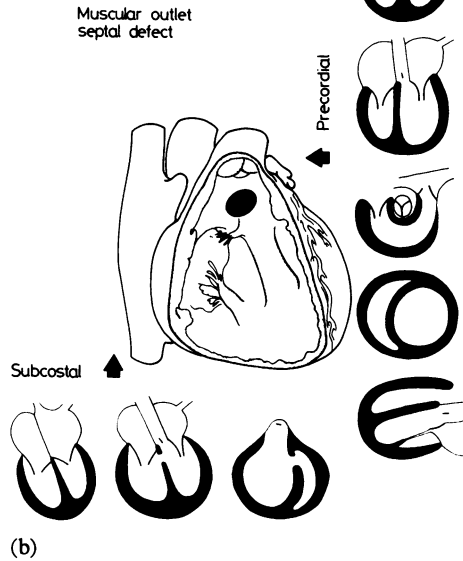


Fig. 7 (a) An outlet muscular defect (extending posteriorly to involve trabecular septum). A composite slide of three still frames from a subcostal right ventricular outflow → four chamber plus aortic root scan, illustrating the advantage of the scan technique. The defect (arrowed) is initially visualised in frame 1 lying within the muscular outlet septum and separated from the pulmonary valve by infundibular septum. In frame 2, an intermediate plane, the defect remains bounded by muscle. In frame 3 (four chamber plus aortic root plane) the most posterior extent of the defect lies within trabecular septum below an intact membranous septum. PV, pulmonary valve; RVOT, right ventricular outflow tract; RV, right ventricle; LV, left ventricle; Ao, aorta. (b) The echocardiographic pattern diagnostic of a muscular outlet defect. (c) A pathological specimen of a muscular outlet defect (defect arrowed) sectioned in a subcostal right ventricular outflow plane. Note how precisely two dimensional echocardiography (Fig. 7a) defines the morphological features. PV pulmonary valve; RVOT, right ventricular outflow tract; RV, right ventricle; LV, left ventricle.

same heart sectioned in the four chamber echocardiographic plane. Note that in this view the septum will appear grossly intact to the echocardiographer. The small trabecular defects are seen to burrow through the septal mass in a spiral or oblique fashion. Such defects do not produce a complete echocardiographic window across the width of the septum and thus the septum will always appear intact to the echo beam. Sectioning hearts with similar defects in a series of echocardiographic planes confirmed the validity of this explanation. From these findings it seems unlikely that two dimensional echocardiography can reliably identify multiple trabecular or small single

trabecular defects.

DOUBLY COMMITTED SUBARTERIAL DEFECTS (24 patients)

The morphological feature which defines a doubly committed subarterial defect is that it is roofed by conjoint aortic and pulmonary valve rings that lie at the same level. These features were well defined by two dimensional echocardiography (Fig. 10a and b). In each of the patients the ventricular septum appeared intact throughout the four chamber plane. When scanning anteriorly to the aortic root, however, both aortic and pulmonary valves were simultane-

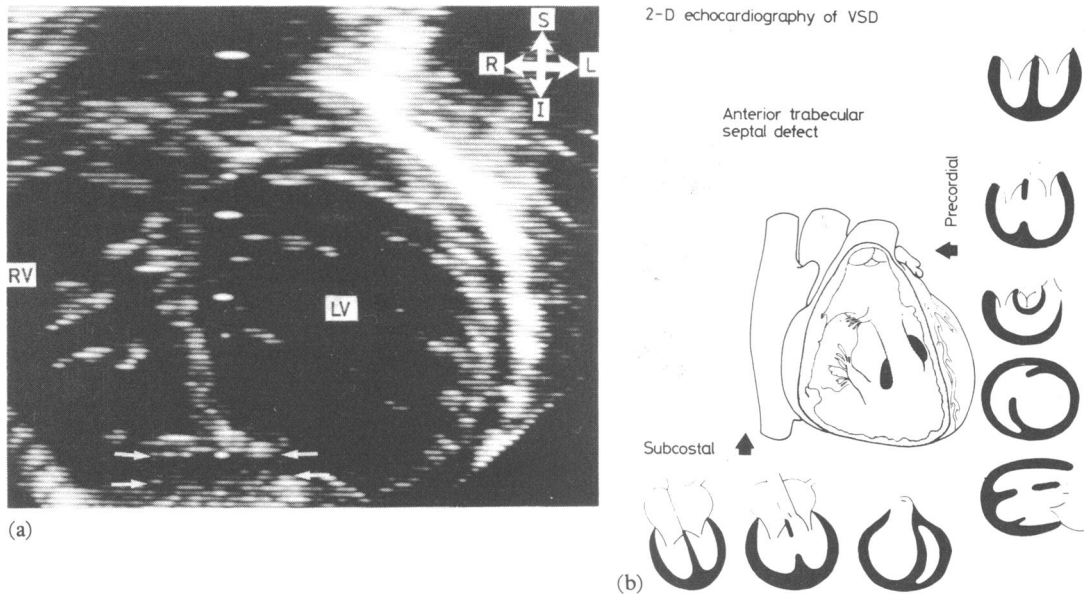


Fig. 8 (a) An anterior trabecular defect (between arrows) visualised in a precordial short axis section. (b) The echocardiographic pattern diagnostic of a large anterior trabecular defect.

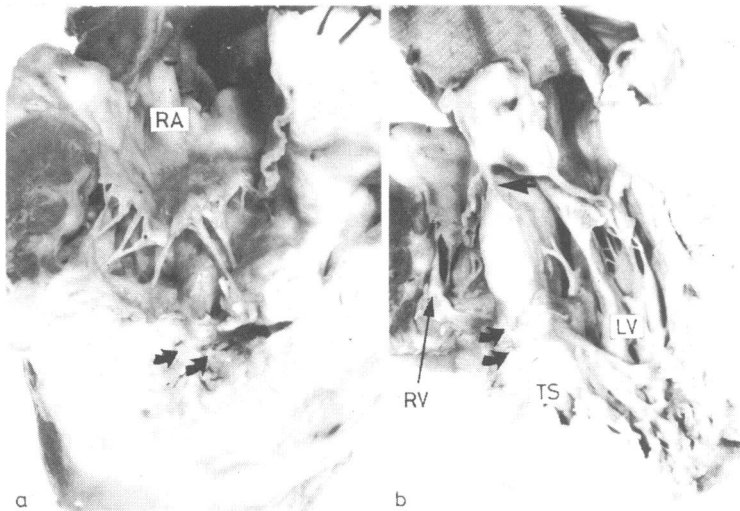
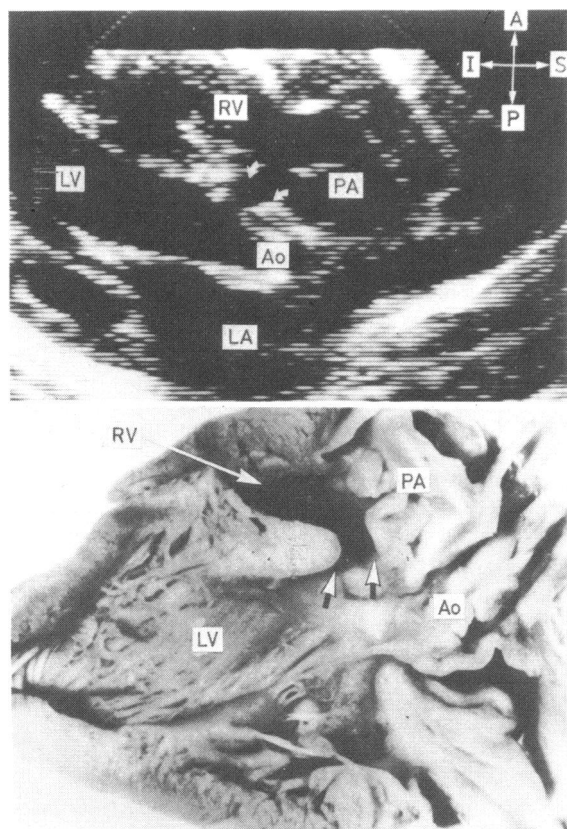


Fig. 9 A pathological specimen showing multiple small trabecular defects. (a) The right ventricular septal surface with the area of multiple defects arrowed. (b) The same heart sectioned in a four chamber plane. Note the defects (small arrows) burrow through the septum in an oblique manner. There is no discrete area where the whole septal width is absent. The septum will appear intact on two dimensional echocardiography. RA, right atrium; RV, right ventricle; LV, left ventricle; TS, trabecular septum (large arrow indicates intact membranous septum).

ously visualised lying at the same level, in the same echocardiographic plane and in a side-by-side relation. These conjoint arterial valves roofed a large ventricular septal defect whose inferior margin was formed by the crest of the trabecular septum. In nine patients this subarterial defect extended posteriorly to involve the area of the membranous septum. In the remaining 15 patients the membranous septum appeared intact and the posterior rim of the defect was

clearly formed by muscle. Thus, two dimensional echocardiography could clearly define doubly committed subarterial defects and subdivide them into two types: (i) those that involve the membranous septum and (ii) those separated from it by a muscle bar.

Doubly committed subarterial defects were consistently recognised using a scan through the four chamber plus aortic root plane (either subcostal or precordial) (Table). The area of septal drop out was



(a)

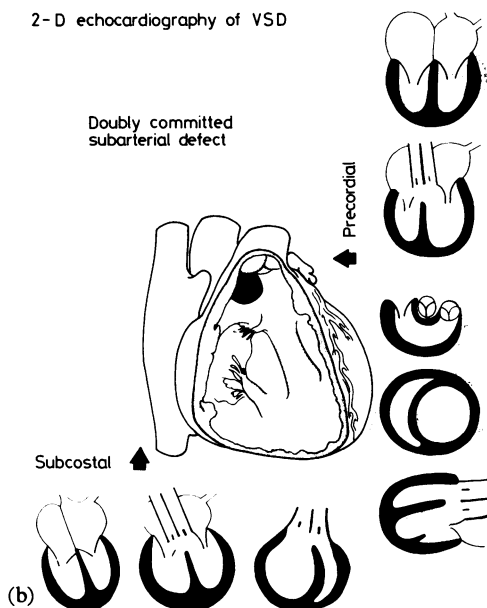
consistently visualised in a subaortic position on precordial long axis scanning, but this view did not always clearly show conjoint aortic and pulmonary valve rings. These defects were not well categorised by short axis scanning.

Discussion

Although the ventricular septum is formed from a number of morphologically distinct subunits it appears as a virtually homogeneous structure on two dimensional echocardiography. The septum may vary in width in different echocardiographic planes but little distinction can be made between individual subunits. The only portion that can be consistently differentiated in the normal heart is the interventricular portion of the membranous septum. Thus, precise localisation of defects within the ventricular septum will only be possible if we appreciate which septal subunits form the composite septal structure visualised in each echocardiographic plane.

Where a defect is identified by an area of persistent septal "drop out" it can be determined through which echocardiographic planes this area extends and where it appears within the septum in each plane. This

2-D echocardiography of VSD



(b)

Fig. 10 (a) A doubly committed subarterial defect. Upper panel: a left ventricular long axis plane. Note the conjoint aortic and pulmonary valve rings which roof the defect (arrowed). Lower panel: the corresponding pathological specimen sectioned in a left ventricular long axis plane. Ao, aorta; PA, pulmonary artery; LV, left ventricle; RV, right ventricle. (b) The echocardiographic pattern diagnostic of a doubly committed subarterial defect.

information will determine the position and extent of a septal defect but will not accurately classify it. How then should we best classify ventricular septal defects using the information which two dimensional echocardiography can provide?

The increased resolution of two dimensional systems now allows specific features of cardiac morphology to be imaged in detail. The aortic root and valve, the central fibrous body (that is the area enclosed by the fibrous annuli of the tricuspid, mitral, and aortic valves and so comprised of the right fibrous trigone and the atrioventricular membranous and muscular septa), the interventricular membranous septum, the infundibular septum, and the pulmonary valve are all readily identified. With this degree of resolution it is now appropriate to base any echocardiographic classification on the identification of all the structures that form the rim of a defect. Thus the morphological classification adopted prospectively was that initially proposed by Moulaert⁶ which subdivided defects on precisely this basis. This classification was subsequently modified to a minor degree during the investigation in view of our findings (Fig. 2).

To determine if this method of classification was both practical and reliable we reviewed 280 patients

with a ventricular septal defect. Using a multiplane scanning technique to construct an approximate three dimensional model of the heart we identified a defect in 252 patients. In each case the structures forming the margins of a defect were readily identified, thus allowing accurate classification into one of three main groups (Fig. 2). The further subdivision of these main groups, based on which septal subunit is predominantly involved, was determined by the echocardiographic planes in which the defect was visualised (for example a defect roofed in part by the central fibrous body but otherwise surrounded by muscle is a perimembranous defect; where such a defect is only visualised in a four chamber plane which profiles the inlet septum it is subclassified as a perimembranous inlet defect). Defects frequently extend across the boundaries of individual septal components, however, and this information should be included in any description of a defect (for example the majority of muscular outlet defects studied were not restricted to the right ventricular outflow plane but were traced posteroinferiorly into the four chamber plus aortic root plane where they burrowed into the trabecular septum. Such a defect should therefore be described as a muscular outlet defect which extends posteroinferiorly into the trabecular septum). In our experience, two dimensional echocardiography accurately subdivided septal defects within each of the three main groups on the basis of which septal subunit was predominantly involved, and in addition accurately identified any extension into an adjoining septal subunit.

Different defects were identified with different degrees of accuracy (Table). Perimembranous inlet and outlet defects, doubly committed subarterial defects, and inlet and outlet defects of the muscular septum were all identified with remarkable consistency, whereas defects of the trabecular septum were identified with varying degrees of success. Multiple small (Swiss cheese) trabecular defects were not identified in this series. Single trabecular defects were inconsistently visualised, with some correlation existing between defect size and defect recognition. When divided into two groups on the basis of intracardiac shunting (small defects with a Qp:Qs < 1.5:1 and large defects with a Qp:Qs > 1.5:1, Table) it was clear that small defects were not visualised at all compared with 72% recognition of large defects. Some surprisingly large trabecular defects, however, were not visualised.

The above findings show broad agreement with the results of previously published series.^{2,3} Both these series, however, have confined their investigation to the identification of septal defects in infants and have restricted their echocardiographic approach to the subcostal window. Though a subcostal approach will yield more information when compared with a pre-

cordial approach (the muscular outlet septum is only profiled from the subcostal window) it can be extremely difficult to record in older children and adolescents. In these cases a precordial approach may be the only means of visualising a defect. Information can be gained from the precordial approach, however, which is not readily obtainable from the subcostal window (for example the anterior extension of a perimembranous subaortic defect beneath the whole aortic root was clearly visualised only in a left ventricular long axis plane; similarly, anterior trabecular defects were best visualised by precordial short axis scanning, being frequently missed from the subcostal approach).

We suggest that the composite approach adopted in this study in which the heart is visualised from both the subcostal and precordial window and where a scanning technique is used to determine the extent of any defect will provide the maximum amount of information. It will accurately identify and classify the majority of ventricular septal defects in infants, children, and adolescents, and thus offers considerable advantages over previously described echocardiographic techniques for studying the ventricular septum.

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