

Doubly committed subarterial ventricular septal defect: new morphological criteria with echocardiographic and angiocardigraphic correlation

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SUMMARY To gain a better understanding of the anatomy of doubly committed subarterial defects and its relation to findings at cross sectional echocardiography and angiocardigraphy, eight necropsy specimens from patients with this condition were examined, and preoperative echocardiograms and angiocardigrams from 313 patients with surgically or necropsy confirmed outlet defect were reviewed. Of these, 48 had doubly committed subarterial defects. Morphological review showed that doubly committed defects are roofed by the arterial valves in fibrous continuity because of lack of both the outlet septum and the "septal" aspect of the subpulmonary infundibulum. Angiocardigraphy had a lower sensitivity (50%) than echocardiography (95%) for diagnosis of doubly committed defects, but each was highly specific. In five (14%) of 35 available echocardiograms the arterial valves were normally offset, but in the remainder they were at the same level. The ventriculoarterial connection was concordant in 37/48 (77%), discordant in five (10%) of 48, and double outlet right ventricle in six (13%) of 48. Displacement of the fibrous raphe between the arterial valves in relation to the ventricular septum below was associated with outflow obstruction in 14 patients (pulmonary in nine and aortic in five). These features were readily identified by echocardiography.

Thus echocardiography is not only a more accurate method than angiocardigraphy of recognising these defects, but also shows that the arterial pole of the heart is architecturally abnormal.

The doubly committed subarterial defect accounts for 5% of isolated ventricular septal defects in white populations¹ and 30% of such defects in oriental populations.² The course may be complicated by prolapse of the aortic leaflets, with or without regurgitation, pulmonary hypertension, aneurysm of the sinus of Valsalva, acquired aortic to ventricular communications, endocarditis, and the need for

aortic valve replacement.³ Prompt surgical intervention is recommended for aortic leaflet prolapse, with or without regurgitation, or if the defect is large.⁴ This emphasises the need for swift and accurate diagnosis.

We reviewed the morphology of these defects to establish the criteria for diagnosis. We then compared the results of echocardiography and angiography in an effort to establish the strengths and weaknesses of the two techniques.

Patients and methods

We reviewed surgical and pathology records from July 1979-December 1986. Three hundred and thirteen patients had septal defects between the

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ventricular outlets confirmed at operation or necropsy. There were two groups. The first contained those with perimembranous or infundibular muscular defects. The second group comprised 48 patients with doubly committed subarterial defects.

Eight necropsy specimens were available from the 10 patients with doubly committed defects who died. The findings from these hearts were correlated with the results of echocardiography and angiocardiography.

Preoperative echocardiographic and angiocardiographic diagnoses of all 313 patients were noted. We reviewed 35 echocardiograms and 40 angiocardiograms that were available from patients with doubly committed defects. We looked for fibrous continuity between the leaflets of the aortic and pulmonary valves, offsetting of these valves, unequal division of the outflow tracts with overriding of one or other arterial valve, and prolapse of the aortic leaflets with or without aortic insufficiency.

Results

MORPHOLOGICAL REVIEW

The normal outlet (or infundibular) septum is the muscle separating the outflow tracts of the left and right ventricles. This extends cephalad to the tricuspid valve and ends opposite the attachment within the left ventricle of the right coronary leaflet of the aortic valve. It is usually a small structure. The remaining right ventricular outflow tract is a sleeve of muscle supporting the leaflets of the pulmonary valve. Piercing this muscle produces a passage leading outside the heart. The presence of this extensive subpulmonary infundibulum creates normal offsetting of the arterial valves, which is readily visualised by cross sectional echocardiography (fig 1).

In contrast, all eight hearts with doubly committed subarterial defects showed complete lack both of the outlet (infundibular) septum along with the "septal" aspect of the sleeve of subpulmonary infundibulum. In consequence, the interventricular communication was roofed by fibrous continuity between the leaflets of the aortic and pulmonary valves (fig 2). In one case there was considerable offsetting of the leaflets. The structure producing the offsetting was the wall of one aortic sinus rather than the muscular subpulmonary infundibulum, as in the normal hearts (fig 3). In some cases, there were remnants of infundibular muscle near the level of continuity between the valves that did not form a continuous muscle rim to roof the defect. Major associated anomalies in these eight hearts included double outlet right ventricle (one case), complete transposition (two cases), overriding aortic and dysplastic pulmonary valve (one case), and

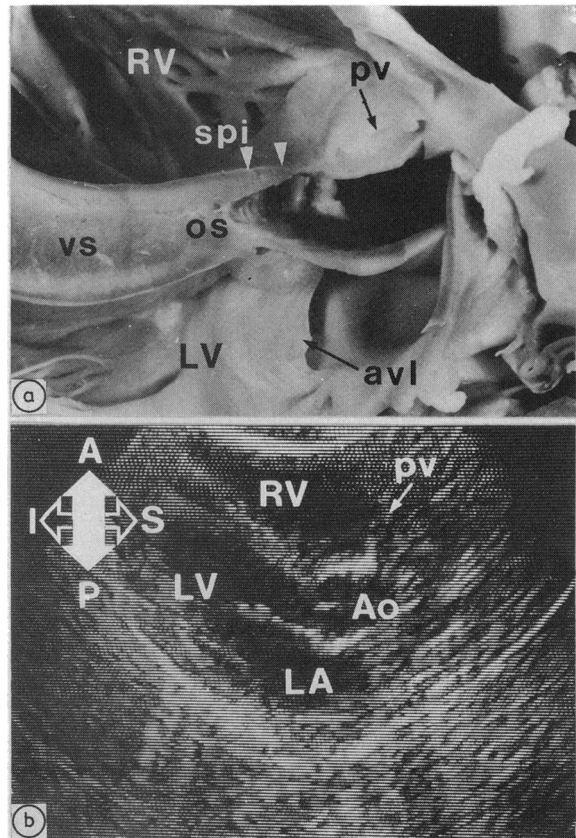


Fig 1 (a) Photograph of a normal heart that represents a long axis echocardiographic view through the arterial valves and shows the relation of the left and right ventricular outflow tracts. The pulmonary valve is more cephalad than the aortic valve because of the extensive subpulmonary infundibulum. Only a relatively small area can be considered as outlet septum (the muscle separating the subpulmonary and subaortic outflow tracts). (b) Parasternal long axis echocardiogram of a normal heart showing the anatomy in (a). A, anterior; Ao, aorta; avl, aortic valve leaflet; I, inferior; LA, left atrium; LV, left ventricle; os, outlet septum; P, posterior; pv, pulmonary valve; S, superior; vs, ventricular septum; spi, subpulmonary infundibulum.

interruption or coarctation of the aortic arch (three cases).

In the unrepaired hearts, obstruction of ventricular outflow was associated with displacement (relative to the outflow tracts) of the fibrous raphe between the aortic and pulmonary valves with overriding of the unobstructed arterial valve, or alternatively, with thickening or dysplasia of the arterial valve leaflets themselves or both. One case showed additional redundant valve tissue adherent to the pulmonary valve producing "subvalvar pulmonary stenosis". In those with a displaced

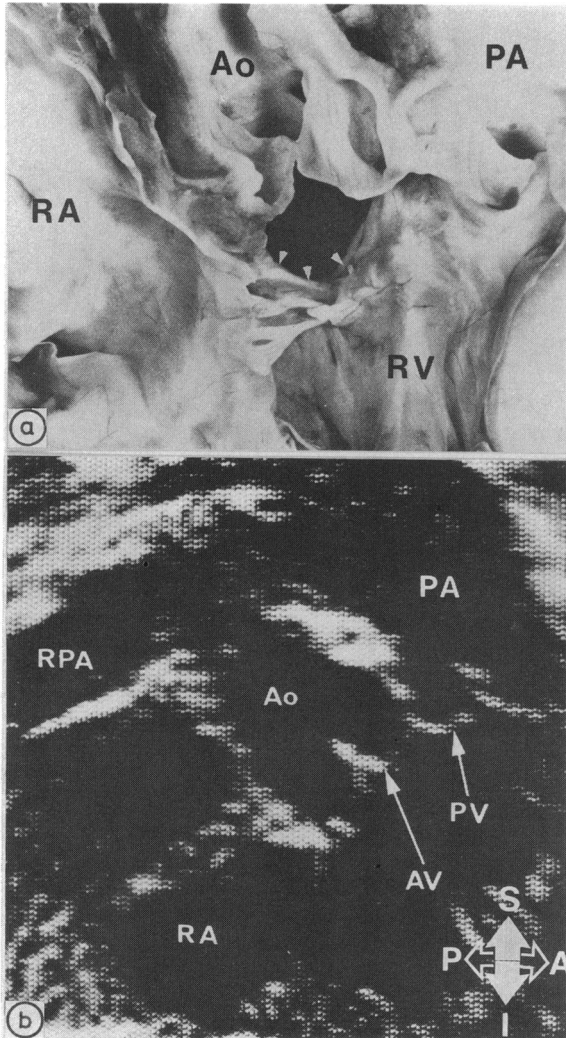


Fig 2 (a) Photograph of the right ventricular aspect of a heart with normal connections showing a doubly committed ventricular septal defect with its superior margin formed by the arterial valves in fibrous continuity, an absent outlet septum, and a deficient subpulmonary infundibulum. (b) Subxiphoid right ventricular long axis echocardiogram in the same patient. There is no outlet septum, a fibrous continuity of the arterial valves, and deficiency of the "septal" aspect of the subpulmonary infundibulum. Arrowheads indicate the inferior rim of the ventricular septal defect. RA, right atrium; AV, aortic valve; PA, pulmonary artery; RPA, right pulmonary artery. See fig 1 for other abbreviations.

aortopulmonary raphe the associated cardiac abnormalities were consistent. Thus when the aortic valve overrode, it was the subpulmonary outflow that was obstructed and the pulmonary arteries were hypoplastic. Overriding by the pulmonary valve was associated with aortic stenosis and obstructive lesions of the aortic flow pathway (fig 3).

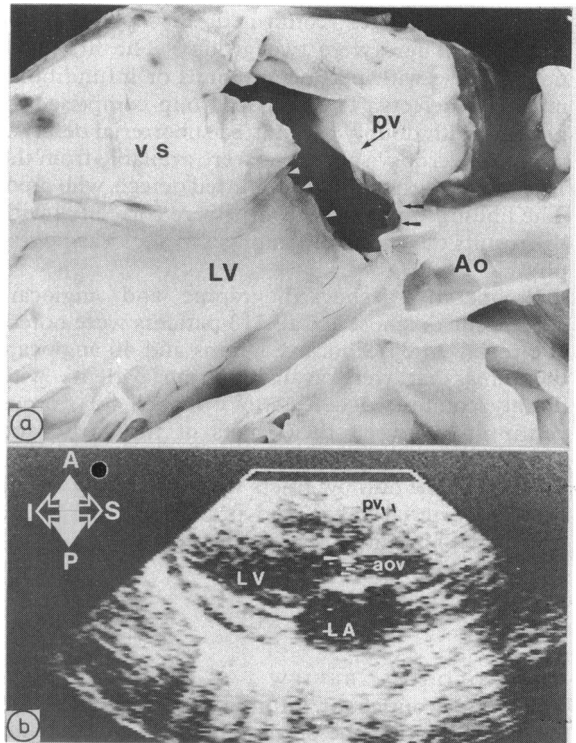


Fig 3 (a) Long axis view of the left ventricle of a heart with interrupted aortic arch and a doubly committed subarterial defect. The lower margin of the defect is indicated by white arrowheads. The superior margin is formed by the arterial valves in continuity. The pulmonary valve overrides the septal defect and the aortic valve is bifoliate. The offsetting of the arterial valve leaflets despite fibrous continuity is shown by black arrows. (b) Long axis echocardiogram of this patient showing absent outlet septum, fibrous continuity of the arterial valves, deficient subpulmonary infundibulum, and overriding pulmonary valve. Note the offsetting of the arterial valves caused by interposition of aortic sinus. The arrowheads indicate the arterial valve leaflets. aov, aortic valve. See figs 1 and 2 for other abbreviations.

ECHOCARDIOGRAPHIC/ANGIOCARDIOGRAPHIC REVIEW

Preoperative angiographic diagnosis was available in 307 of 313 patients (261 of 265 in the first group and 46 of 48 of those with doubly committed defects). Preoperative echocardiographic diagnosis was available in 271 of 313 patients (234 of 265 in the first group and 37 of 48 of those with doubly committed defects). There were 225 of 265 patients in the first and 37 of 48 in the second group with both echocardiographic and concurrent angiographic diagnoses.

Major associated defects in those with doubly committed defects included five (10%) with complete transposition, six (13%) with double outlet

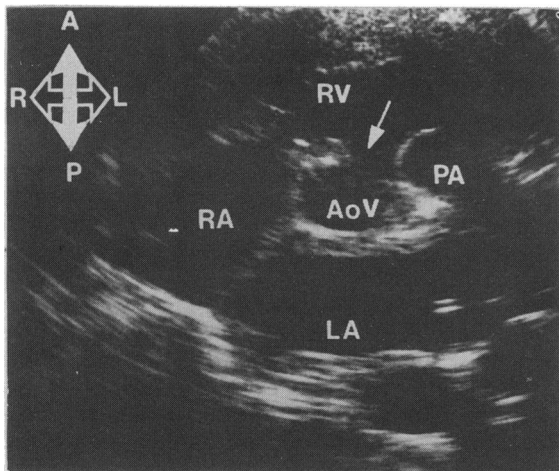


Fig 4 Short axis echocardiogram in a patient with doubly committed subarterial ventricular septal defect (arrow). AoV, aortic valve. See figs 1 and 2 for other abbreviations.

right ventricle, nine (19%) with overriding aortic valve and right ventricular outflow obstruction, and five (10%) with obstruction to the left ventricular outflow. The aortic valve was trifoliate in 45 (94%) and bifoliate in three (6%) patients each of whom had an abnormal aortic arch.

Echocardiographic sensitivity and specificity for prospective differentiation of doubly committed from other outlet defects were 95% and 99% respectively. There were three false positive and two false negative diagnoses. These arose from technically inadequate recordings that resulted in errors in interpretation. Retrospective analysis showed that all echocardiograms from those with doubly committed defects showed absence of the outlet septum and deficiency of the subpulmonary infundibulum. This was shown by the presence of fibrous continuity between the facing leaflets of the aortic and pulmonary valves. None the less, there was offsetting of the facing leaflets in five (14%) of 35 (fig 3), and two (6%) of 35 showed perimembranous extension of the ventricular septal defect. Best visualisation of the fibrous continuity and the outflow anatomy was obtained in 25 (71%) of 35 studies in the subxiphoid long axis view of the right ventricle (fig 2b). In six studies there were technically inadequate subxiphoid views. Parasternal long and short axes (fig 4) were the next most helpful views, particularly when there was rotation from one view to the other.⁵ Prolapse of the right coronary leaflet was diagnosed in eight studies (fig 5).

Angiocardiographic sensitivity and specificity for differentiation of doubly committed from other outlet defects were 50% ($p < 0.001$ versus echo-

cardiography) and 97% respectively. There were 23 false negative and seven false positive diagnoses. Of 40 biplane cineangiograms available for review, left ventriculogram projections were in the long axis¹ in 37 (93%) and in the anteroposterior axis in three (7%). Unequivocal evidence of continuity of the aortic and pulmonary leaflets was established in only 19 (48%) of 40 patients. Frequently, the defect was not well defined and the position of the pulmonary valve was difficult to assess.

Aortography demonstrated prolapse of one or more aortic valve leaflets in 14 (38%) of 37 patients with ventriculoarterial concordance. In 12 there was prolapse of the right coronary leaflet and in two there was prolapse of both the right and non-coronary leaflets. There was aortic regurgitation in 11 (79%) of 14 but this was not present before the age of 2.3 years. Aortic regurgitation did not occur in any patient who did not have aortic leaflet prolapse.

Discussion

MORPHOLOGY

Previous description of doubly committed subarterial defects presumed that the normal outlet septum was the muscle mass extending beyond the tricuspid valve up to the pulmonary valve.¹ The criterion for diagnosis was the absence of the "outlet septum" as thus defined, with the defect being roofed by the leaflets of the aortic and pulmonary valves in fibrous continuity.^{6,7} This study shows that more detailed analysis of the right ventricular outflow tract

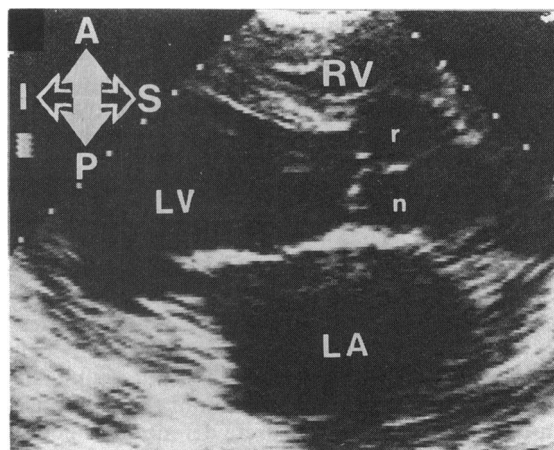


Fig 5 Parasternal long axis echocardiogram in a patient with an isolated doubly committed subarterial defect and prolapse of the right coronary aortic leaflet. The leaflet is prolapsed through the defect into the right ventricular outflow tract. n, non-coronary leaflet; r, right coronary leaflet. See figs 1 to 4 for other abbreviations.

is needed before the morphology of the doubly committed defect can be understood.

In the normal heart, the true outlet septum, that portion of muscle interposing between the outflow component of the left and right ventricles cephalad to the tricuspid valve, is small. More importantly, there is a sleeve of subpulmonary infundibulum which supports the leaflets of the pulmonary valve, yet separates the right ventricular outflow tract from the outside of the heart rather than from the cavity of the left ventricle. This arrangement produces the normal offsetting between the leaflets of the aortic and pulmonary valves. Presence of a doubly committed defect, therefore is based not only on absence of the septum but also on absence of the "septal" aspect of the subpulmonary infundibulum. It is only in these circumstances that the leaflets of the aortic and pulmonary valves are seen in fibrous continuity. Offsetting of the aortic and pulmonary valves can still occur when there is a doubly committed defect in the absence of the subpulmonary infundibulum. This occurs when the wall of an arterial sinus is interposed between the valves. This anatomical feature was recently described² and is endorsed by our findings. The morphological criteria suggested above for the recognition of a doubly committed defect hold good irrespective of the ventriculoarterial connection.

VENTRICULAR OUTFLOW OBSTRUCTION

"Fallot's tetralogy" is often described as occurring with doubly committed subarterial ventricular septal defect.^{8,9} The morphology of tetralogy, however, includes an outlet malalignment ventricular septal defect, overriding of the aortic valve, and subpulmonary infundibular narrowing. If we chose a single morphological criterion for the tetralogy of Fallot it would be anterocephalad deviation of the septal insertion of the muscular outlet septum. In doubly committed subarterial defects, the outlet septum is absent and the subpulmonary infundibulum incomplete; thus there is shortening of the narrowed right ventricular outflow tract and, indeed, obliteration of its "septal" aspect. Therefore, coexistence of Fallot's tetralogy, as defined, and a doubly committed defect is not tenable. This is, of course, dependent upon the definition chosen. None the less, we submit that the description of overriding of the aortic valve in the presence of aortic-pulmonary valvar continuity and obstruction of the pulmonary outflow tract is a more accurate definition of doubly committed subarterial ventricular septal defect in association with obstruction of the pulmonary outflow tract.

Our pathological review has shown that restriction of the right or left ventricular outflow tracts is associated with displacement of the fibrous raphe

between the aortic and pulmonary valves along with overriding of the unobstructed valve. In addition, there may be thickening or dysplasia of the leaflets of the arterial valves and accessory tissue adherent to them or any combination of these features. The major associated abnormalities reflect the haemodynamic consequences of the presence of outflow tract obstruction. Our findings may explain why subvalvar obstruction occurs after patch repair in cases with overriding arterial valves.⁸

CLINICAL IMPLICATIONS

Cross sectional echocardiography is more sensitive than axial cineangiography in the diagnosis of the doubly committed defect. Echocardiography offers a wide variety of views of interest, allowing investigation of the ventricular septum, outflow tracts, and associated anomalies. Since 1984, some patients with isolated doubly committed defects have undergone corrective surgery without preoperative angiography.¹⁰ Angiography none the less remains helpful in more closely defining abnormalities of the aortic valve, pulmonary arteries, and aortic arch.

Conclusion

This study challenges the presently accepted concept of the "outlet septum" in the normal heart and expands the definition of the doubly committed subarterial defect. The diagnosis requires an incomplete subpulmonary infundibulum as well as absence of the outlet septum and fibrous continuity of the aortic and pulmonary valves. These features are more readily demonstrated by echocardiography than by angiography. This information facilitates planning of surgical strategy. Closure of the isolated doubly committed defect can usually be performed through the pulmonary artery with low morbidity, even in small infants, without the need for invasive investigation.¹⁰

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