Multiplanar review analysis of three-dimensional echocardiographic datasets gives new insights into the morphology of subaortic stenosis

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Aims Associated left ventricular structures may play a role in progression and recurrence of discrete subaortic stenosis. The availability of a new 3D echocardiography tool, multiplanar review (MPR), allows comprehensive analysis of datasets in infinite planes, and detailed examination of anatomy. We sought to evaluate the role of MPR in defining the morphology of subaortic stenosis.

Methods Consecutive patients underwent detailed 2 and 3D echocardiographic examination using MPR.

Results Sixteen patients aged 0.7–15.9 years (median 4.57) with diagnosis as follows: isolated subaortic stenosis in nine, additional defects in seven (coarctation of aorta, VSD, mitral, or aortic stenosis). Position and extent of subaortic stenosis was clearly described by multiplanar review in all patients. Additional MPR findings were: abnormalities of mitral valve leaflet or chordal apparatus attachments (14 patients), abnormal ventricular muscle band (11), abnormal increased aorto-mitral separation (two). The aortoseptal angle was significantly decreased in subaortic stenosis, mean 141 ± 12°, vs. normal subjects, mean 153 ± 6°, P = 0.02. Surgical findings correlated well with MPR findings.

Conclusions MPR analysis of 3D datasets is a sensitive and accurate mode for delineation of morphological details of discrete subaortic stenosis, providing additional information to 2D echocardiography.

KEYWORDS
Subaortic stenosis; 3D echocardiography

Introduction
Discrete subvalvar aortic stenosis (SAS) is produced by a fixed lesion, which may be caused by ¹ a fold or ridge of endocardium and fibrous tissue, ² a circumferential fibromuscular ring in the left ventricular outflow tract (LVOT), or³ a long diffuse, tunnel-like fibromuscular narrowing in the LVOT (Figure 1). The first two types account for the majority of SAS, and the third type is rare. As it rarely presents in the newborn period or infancy, SAS is thought to be an acquired, rather than congenital, cardiac abnormality. However, the substrate for development of SAS, and the pathophysiological mechanisms involved in its progression and recurrence, remain poorly understood. Recurrence after surgical excision is a recognized problem. While the potential for recurrence may be inherent in the individual’s tissues, flow disturbance, for example caused by associated left ventricular structures, may also play a role. Diagnosis of SAS by conventional 2D echocardiography is well established. The availability of a new real time 3D echocardiography tool, Multiplanar Review (MPR), allows detailed and comprehensive analysis of datasets in infinite planes. In this study, in an attempt to better understand the functional anatomy of SAS, we sought to evaluate the role of MPR in assessing morphological details of SAS in our patients.

Methods
Consecutive patients with SAS were recruited on the paediatric cardiology ward or outpatient clinic. All patients had a routine 2D echocardiographic examination, including continuous wave and colour flow Doppler. Significant lesions were further studied with 3D MPR. Patients with structurally normal hearts undergoing echocardiographic examination for other reasons, such as innocent murmurs, acted as controls. A commercial real-time 3D imaging system, Philips Sonos 7500 or IE33 (Philips Co, Netherlands) with a 3–5 MHZ matrix phased array transducer was used to perform echocardiography. All patients were awake, unsedated and breathing spontaneously. Stored full volume 3D echocardiographic datasets were assessed off-line by two investigators.
blinded to 2D echocardiographic findings, using Q lab software version 4.1 for the earlier patients, and version 5.2 when this became available. Analysis was carried out in the MPR mode. This mode allows the operator to view the moving 3D dataset in three orthogonal planes simultaneously, and to review the image in infinite planes by moving each of the three planes through the dataset. Each plane is referenced in each of the other two planes being simultaneously viewed, allowing intracardiac structures to be examined in their entirety in three dimensions. This has an advantage over 2D echocardiography, in that the heart is not examined in fixed single planes, with detail missing between the planes viewed, but rather the heart can be examined completely by moving each plane gradually throughout the dataset. This also has an advantage over more commonly used ‘cropping’ of 3D datasets for examination of cardiac anatomical detail, in that structures are not viewed overlying each other, or partially hidden behind superimposed structures.

In addition to the position and extent of subaortic stenosis, abnormalities which may contribute to mechanical stress or flow disturbance in the LVOT were sought. The following parameters were recorded:

- position and extent of subaortic stenosis
- mitral valve leaflet and chordal apparatus attachments
- presence of muscle bundles in left ventricular cavity
- aorto-mitral separation
  - defined as the shortest distance between the non-coronary cusp of the aortic valve and the mitral annulus
- aortoseptal angle
  - defined as the angle formed by the long axis of the ascending aorta and the plane of the ventricular septum
- the correct plane for measurement of this angle in 3D MPR was defined by positioning one plane along the long axis of the aortic root, and one plane through the ventricular septum (with each plane being referenced in each of the other two planes simultaneously viewed), and then measuring the angle between the planes
- associated abnormalities

These findings were compared to 2D echocardiographic findings, and in order to validate the 3D MPR findings, were compared to surgical findings in those patients who routinely underwent corrective surgery during the study period.

Values are expressed as mean values plus or minus one standard deviation.

Approval from the local research ethics committee was obtained prior to carrying out the study.
Results
Sixteen patients were identified, aged 0.7–15.9 years (median 4.57). The diagnosis on 2D echocardiographic examination was isolated SAS in nine patients (56%), and additional defects were coarctation and VSD in three, coarctation in two, ventricular septal defect in one, mitral and aortic valvar stenosis in one patient (Shone complex). Patients with atrioventricular septal defects were excluded from the analysis.

On 2D echocardiography with continuous wave Doppler, the mean gradient across the LVOT was 51 mmHg, range 20–88 mmHg.

Complete analysis by MPR was possible in all patients, and the findings are summarized below (Table 1).

Position and extent of subaortic stenosis
All patients had discrete SAS; no patients with diffuse tunnel-like SAS were identified. In nine of the 16 patients (56%), the SAS was in the form of a fibrous ridge, in the other seven, a circumferential fibromuscular ring in the LVOT was identified. In the nine patients with a fibrous ridge, five of these had a discrete ridge remote from the aortic valve at the septal aspect of the LVOT, two had a discrete ridge closely related to the non-coronary cusp of the aortic valve at the septal aspect of the LVOT, two had a ridge closely related to the aortic valve, with extension to the anterior leaflet of the mitral valve (this had not been noted on 2D echocardiography in one patient). In the seven patients with a circumferential ring, the ring was remote from the aortic valve in all patients except one, in whom the ring was intimately related to the valve and restricted the opening of all three cusps (Figure 2).

In one patient, 2D echocardiography had been unable to determine the level of LVOT obstruction, and this was clearly defined by MPR.

At least one additional subaortic abnormality was found in all except one patient by 3D MPR analysis, but only detected in one patient by 2D echocardiography. Details are given below.

Mitral valve leaflet and chordal apparatus attachments
In four patients (25%), the anterior leaflet of the mitral valve had an abnormal attachment to the septum (Figure 3). In 10 patients (63%), there was an abnormal accessory chordal attachment of the mitral valve to the septum (Figure 4). These patient groups did not overlap; in patients in whom the anterior mitral valve leaflet was attached directly to the septum, there was not an accessory chordal attachment to the septum. In the patient who had previously had a mitral valve replacement, it was not possible to comment on the mitral valve attachments and chordal arrangement.

Presence of muscle bundles in left ventricular cavity
In 11 patients (69%), there was an abnormal muscle band situated apically.

Aorto-mitral separation
There was abnormal increased aorto-mitral separation in two patients (13%), both of whom had SAS occurring in isolation (one with a discrete ridge and the other with a circumferential ring). In both these patients, the aorta was displaced cranially by 5 mm, and there was no significant dextroposition of the aorta. There was no aorto-mitral separation in any controls.

Aortoseptal angle
The aortoseptal angle was significantly decreased in patients with SAS, mean $141 \pm 12^\circ$, compared to normal subjects, mean $153 \pm 6^\circ$, $P = 0.02$. There was no significant

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<th>Position of SAS relative to AV</th>
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<th>Abnormal MV chordal attachment</th>
<th>Muscle bundles in LV cavity</th>
<th>Aorto-mitral separation (mm)</th>
<th>Aorto-septal angle</th>
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AV, aortic valve; LV, left ventricle; MV, mitral valve; SAS, subaortic stenosis.

*Patient who had previously had mitral valve replacement surgery
difference in the aortoseptal angle between patients who had SAS in the form of a discrete ridge, or as a circumferential ring.

Correlation with surgical findings

Eight patients underwent surgery during the study period, of which one, in addition to resection of SAS, had a mitral valve replacement. Five of these were patients with a circumferential ring, and three had a fibrous ridge. In all cases, surgical findings correlated well with MPR findings, with no unexpected findings, and no discrepancies.

Discussion

Echocardiography is usually the only necessary diagnostic investigation required in the diagnosis, follow-up and surgical planning of SAS. Three-dimensional echocardiography has now evolved to become a rapid and accurate adjunct to 2D echocardiography, and aids in surgical planning. Real-time 3D echocardiography gives new insight into the dynamic changes of intra-cardiac structures during the cardiac cycle, by providing the clinician with immediate 3D images, which can be manipulated and cut in different planes to pinpoint the area of interest. Three-dimensional datasets take only seconds to acquire, and can be processed away from the bedside, which is of enormous benefit in the examination of children. The MPR modality of studying 3D datasets also allows the operator to return to the dataset and study it at leisure, and in infinite planes. Datasets acquired by one operator, or a trainee or technician, may be studied in full detail by the clinician, even if he or she is not present at the time of data acquisition.

Two-dimensional echocardiography remains a valuable and necessary tool, and in particular Doppler assessments of LVOT gradients by 2D echocardiography are a fundamental part of the complete echocardiographic examination. However, 3D echocardiography is a valuable adjunctive investigative tool, as it provides additional information that may not be readily appreciated on 2D echocardiography alone.

We have shown here that visualization of the LVOT by MPR in 3D echocardiography is accurate and feasible. Comprehensive demonstration by MPR pre-operatively allows the nature of the lesion in relation to the proximity and attachment to mitral or aortic valve, and its extent, to be accurately outlined prior to surgical exploration, and this may reduce operative time. Analysis by MPR allows investigation of the whole 3D dataset, and so is an excellent modality for detection of details that may not be observed in standard 2D imaging planes. We report here a higher incidence of additional subaortic abnormalities detected on echocardiography than has been previously reported by Marasini and colleagues, and it may be that MPR allows an increased rate of detection of these abnormalities. All of these subaortic abnormalities may be visualized using 2D imaging alone, but by allowing detailed examination of the entire dataset in infinite planes, MPR increases the ease of their detection, even in patients with suboptimal imaging quality. In their series of 73 consecutive patients, Cohen and colleagues reported 48% with one or more mitral valvar abnormalities. They
surmised the incidence of these abnormalities may have been underestimated previously and recommended that they be searched for systematically in all cases of subaortic stenosis.

It is of interest that patients who had an abnormal attachment of part of the anterior mitral valve leaflet directly to the ventricular septum, did not have an accessory chordal attachment to the septum; either one or the other of these abnormalities was found in certain patients. This may suggest the failure of delamination of the anterior leaflet from the septum in the pathogenesis of SAS. Either of these abnormalities was found in the presence of an abnormal apical muscular band. These findings raise the possibility that discrete subaortic stenosis is a primary mitral valve abnormality, and further morphological study should address this question, including examination of the implantation of the mitral valve papillary muscles, which we have not addressed in this study.

MPR has the additional benefit that it allows the investigator to check each plane against the other two planes being simultaneously viewed. This ensures that the investigator never gets ‘lost’ in the dataset, as each structure can be referenced in three different planes simultaneously. This also allows accurate measurement of intra-cardiac structures and angles, as each structure can be accurately delineated. Our findings of steeper aortoseptal angle in patients with SAS agree with previous reports. Kleinert reports a mean aortoseptal angle of 131 ± 6° in patients with SAS, versus 144 ± 5° degrees in normal controls.\textsuperscript{15} Aortoseptal angles may be measured on 2D echocardiography, but we believe that measuring the angle on MPR ensures accuracy by allowing the operator to check the correct plane for measurement against the other two planes being concurrently viewed.

We have found a lower incidence of aortic-mitral separation than previous investigators,\textsuperscript{14,15} although when it is present in our patients, the extent of separation that we have measured agrees with previous reports (Rosenquist and Kleinert report mean separations of 4.9 and 5.1 mm in patients with SAS). We had the advantage of being able to use MPR to define very accurately the presence or absence of aortic-mitral continuity, and also that 3D echocardiography allows complete examination prior to surgery (Rosenquist’s original description of aortic-mitral separation was from post mortem specimens mainly derived from deaths following surgery for SAS).

We have also found that MPR analysis is possible in most subjects in whom imaging quality is not adequate for 3D volume rendering or ‘cropping’. Although the image quality may be too poor to allow clear visualization of anatomical structures in a cropped dataset, the multiplanar modality allows examination of the structures of interest without interference by surrounding artefact. It is also far less sensitive to artefact created by patient movement or respiration during acquisition. With current software, the resolution of 3D datasets is not yet adequate to allow examination by cropping alone. If we wish to reconstruct a 3D image, we do this only after aligning the MPR planes in an attitudinally appropriate manner, and then cropping in these predetermined planes.
Conclusion

MPR analysis of 3D echocardiographic datasets is a sensitive and accurate mode for diagnosis of discrete subaortic stenosis, providing additional information to 2D echocardiography.

References


