

# Congenital mitral stenosis

## *Anatomical and functional assessment by echocardiography*

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**SUMMARY** Digitised left ventricular echocardiograms were studied in nine children with congenital mitral stenosis to assess the severity of inflow obstruction. In six children the two prime indices of mitral stenosis were abnormal, with a prolonged time from minimum dimension to 20 per cent dimension change and a reduced peak dimension change during diastole. In three, however, these values did not suggest inflow obstruction, despite significant gradients at cardiac catheterisation.

Two-dimensional echocardiography was performed in 10 children with congenital mitral stenosis to determine the mitral annular size and the morphology of the valve and subvalvular apparatus. The annular size and number of papillary muscles could be assessed along with the detection of combined mitral abnormalities.

Two-dimensional studies can reliably delineate the type of mitral abnormality, and should be performed in all cases with congenital heart disease having a high incidence of associated left ventricular inflow obstruction. Digitised M-mode left ventricular echocardiography is in general unreliable in assessing congenital obstruction, though it may be of some value in individual cases.

The diagnosis of congenital mitral stenosis can be difficult, especially in the sick neonate with associated defects which may mask the signs of inflow obstruction.<sup>1</sup>

Standard M-mode echocardiography is unreliable in assessing either the severity or site of obstruction.<sup>2</sup> In acquired mitral stenosis digitised left ventricular echocardiography has proved a more reliable means of assessing severity than measurements of the movement of cusps themselves.<sup>3,4</sup>

Two-dimensional echocardiography permits a more detailed anatomical assessment of orifice size and valvular apparatus in acquired mitral stenosis.<sup>5,6</sup> But the anatomy of congenital mitral stenosis is much more complex,<sup>7</sup> and little information is available as to its demonstration by two-dimensional echocardiography.

The aims of this study were to determine whether congenital mitral stenosis could be assessed by M-mode echocardiography in the same way as the acquired form, and if, by the use of two-dimensional echocardiography, the annular size and morphology of the valve and subvalvular apparatus could be determined.

### Methods

M-mode echocardiograms were performed with an Ekoline 20 ultrasonoscope. Recordings were made on ultraviolet paper from a Cambridge Multichannel recorder at a paper speed of 100 mm/s with a simultaneous electrocardiogram and phonocardiogram.

The patients were studied in a supine position with some slight rotation to the left side. Echocardiograms of the left side of the septum and posterior left ventricular wall were recorded at the level of the mitral valve. Only those beats where both the septum and posterior wall were clear continuous lines were used in the study.

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The echocardiograms were digitised as described by Gibson and Brown<sup>3</sup> in 1973, on a Summographic digitising table and were processed by a Prime 300 computing system. Left ventricular filling pattern was assessed from the peak rate of increase of left ventricular dimension and the time from minimum dimension to 20 per cent of the peak dimension change at the end of the rapid filling period. The periods of isovolumic relaxation (aortic closure to mitral opening) and minimum dimension to mitral opening were also studied.

Two-dimensional echocardiography was performed using an Advanced Technology Laboratory mechanical sector scanner with a 3.5 MHz transducer. The studies were recorded in the supine position using standard views as previously described.<sup>8</sup> A sweep from the apex of the left ventricle to the outflow tract was performed in the short axis view, to assess valvular and subvalvular regions, including the number of papillary muscles.

A subxiphoid four chambered view was used to visualise the papillary muscles, valve, and subvalvular apparatus. In the parasternal long axis projection an assessment of the annular size and mitral excursion was made.

The parasternal long axis cut was standardised by ensuring that the aortic valve cusps were centrally situated in the aortic root. Recordings of the echocardiogram were then made on a Sanyo videorecorder, which was subsequently replayed frame by frame until the point of maximal diastolic excursion of the valve leaflets was recognised. A polaroid photograph was taken of this frame, and the annular dimension measured from the photo-

graph as the distance from a point anteriorly where the anterior mitral leaflet was in continuity with the posterior wall of the aortic root, to the point posteriorly where three structures joined. These were the left atrial and left ventricular free walls, and the posterior mitral leaflet.

### Subjects

Eleven patients whose ages ranged from 3 months to 15 years and who had congenital mitral stenosis without regurgitation were studied.

All patients had the diagnosis confirmed by cardiac catheterisation. In eight cases the precise anatomy was observed, either during open heart surgery (six cases) or at necropsy (two cases).

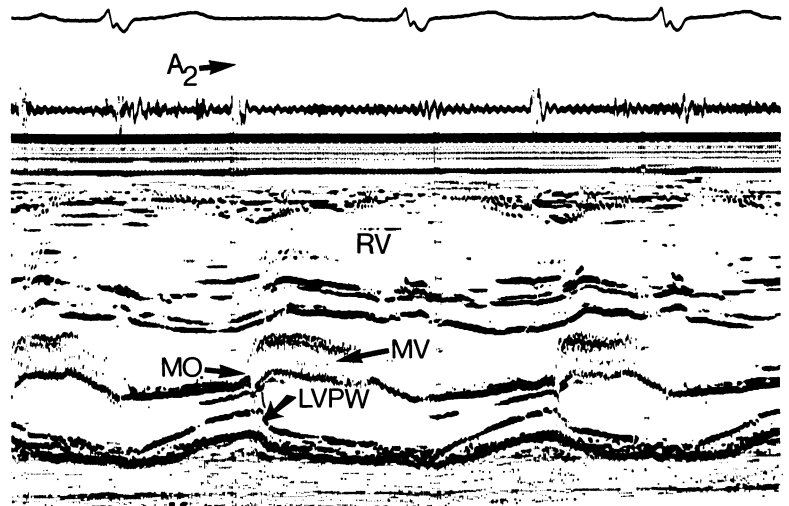
Five patients had previously had associated coarctation of the aorta which had been resected. None had systemic hypertension at the time of the examination.

Two patients had associated ventricular septal defects both with a Qp/Qs of 1.4. All patients had normal-sized left ventricles, since cases of the hypoplastic left heart syndrome were excluded from the study, as were patients with cor triatriatum.

The 11 patients yielded 10 adequate M-mode and two-dimensional examinations. One child had only an M-mode study and another a two-dimensional, but not an M-mode investigation.

Forty-one normal children whose ages ranged from 1 year to 15 years had M-mode echocardiograms to assess their left ventricular filling patterns. Thirty-two normal children whose ages were between 1 month and 15 years had two-dimensional

Fig. 1 M-mode echocardiogram from a patient with congenital mitral stenosis. Coarse vibrations of both leaflets are seen and anterior movement of the posterior leaflet during diastole. The left ventricle fills rapidly during early diastole. A<sub>2</sub>, aortic closure; MO, mitral opening; RV, right ventricle; MV, mitral valve; LVPW, left ventricular posterior wall



echocardiograms to assess their mitral annular size. Informed parental consent was given for all these examinations.

The data were punched onto cards and analysed on a Control Data Corporation 6600 computer (under NOSBE) at the University of London Computer Centre using the *Statistical package for*

*the social sciences* Version 7.0.<sup>9</sup> Logarithmic transformations were used to produce linearity in regression and additivity for analysis of covariance. Multiple stepwise regression was used to predict normal annular size from age, body weight, and body height.

**Results**

**M-MODE**

Of the 10 patients who had adequate M-mode examinations one, aged 3 months, had a posterior left ventricular free wall which moved anteriorly during diastole, the septum moving in the normal way. Because of this paradoxical movement it was not possible to digitise the echocardiogram and obtain reliable information.

All, apart from the above patient, had M-mode mitral valve echocardiograms suggesting the presence of an abnormal valve. Of the nine patients in whom the left ventricular echocardiograms were digitised, six had both a reduced peak rate of dimension change during diastole and a prolonged time to 20 per cent peak dimension change (Table). Three, however, despite having significant obstruction, with gradients between 15 and 17 mmHg, had valves within the normal range for age (Fig. 1 and 2). There was no specific pattern of abnormality in the time intervals minimum dimension to mitral opening and aortic closure to mitral opening (Table).

**TWO-DIMENSIONAL**

In two cases of parachute mitral valve there was an associated supravulvar membrane; in neither was this displayed by two-dimensional echocardiography.

In nine patients adequate parasternal long axis views enabled an assessment of the annular size to be made. The simplest linear prediction of normal annular size was obtained by plotting the  $\log_{10}$  of annular size against  $\log_{10}$  body weight. This explained 85 per cent of the variance in annular size. Though age could be added to the prediction at a statistically significant level, the combination of  $\log_{10}$  weight and age only explained a further 3 per cent of the variance in annular size, so prediction was based on  $\log_{10}$  weight alone. The further contribution of  $\log_{10}$  height was not statistically significant. The values of patients with mitral stenosis were within the normal range for age in all cases (Fig. 3), confirmation being obtained at necropsy in two and at operation in five. The fact that annular size in these patients was in the normal range was also apparent when  $\log_{10}$  annular size was plotted against height.

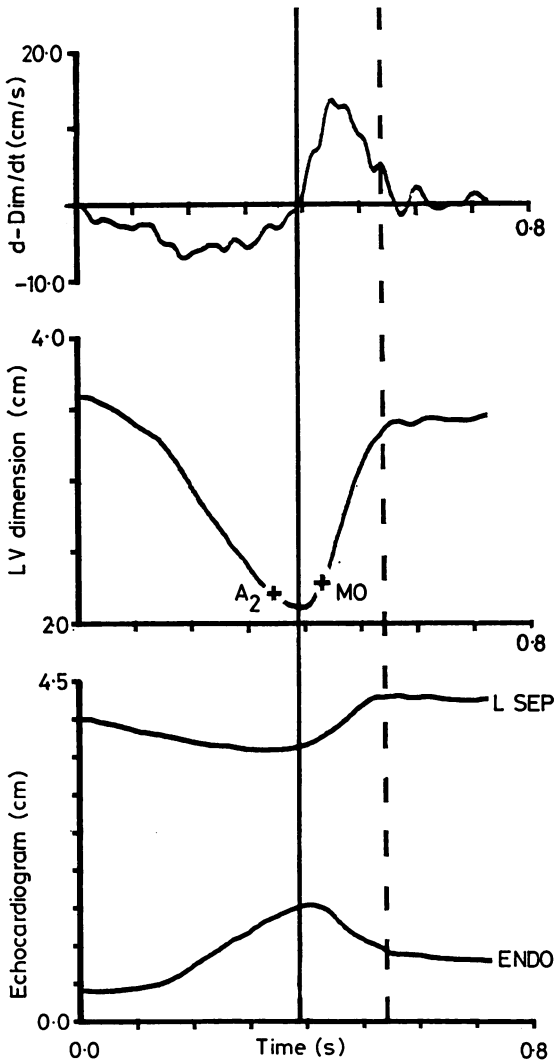


Fig. 2 Digitised left ventricular echocardiogram of Fig. 1. A gradient of 15 mmHg across the mitral valve was present and a supramitral membrane plus associated parachute mitral valve seen at cardiac surgery. The solid vertical line represents minimum dimension and the broken vertical line the time to 20 per cent peak dimension change.  $A_2$ , aortic closure; MO, mitral opening;  $A_2$  to MO represents isovolumic relaxation.

Table Digitised values from left ventricular echocardiograms

	$A_2$ -MD (ms)	$A_2$ -MO (ms)	MD-MO (ms)	Peak d-Dim/dt diastole (cm/s)	Time to 20% PFR (ms)
Control (n=41)	1.5 ±2.7	40.2 ±13.3	42.3 ±22.3	11.6 ±2.9	176 ±28.5
Mitral stenosis (n=9)	12.7 ±2.1	32.2 ±12.0	45.0 ±20.3	9.6 ±4.2	281 ±126.7
	p < 0.5	p < 0.8	p < 0.8	p < 0.1	p < 0.0001

Mean values ± 1 standard deviation.

$A_2$ , aortic closure; d-Dim/dt, rate of change of dimension; MD, minimum dimension; MO, mitral opening; PFR, peak filling rate.

The mitral valve excursion was reduced in all 10 cases, with dense echocardiograms from the leaflets being seen in nine. In all examinations abnormally dense echocardiograms from the subvalvular region were seen in both the parasternal long axis and four chamber view (Fig. 4 and 5). These echocardiograms represented abnormal papillary muscles and thickened chordae, the latter being shortened with partial obliteration of the interchordal spaces by fibrous tissue. As a result of the above abnormalities it was difficult, if not impossible, to differentiate between the various components of the subvalvular region by two-dimensional echocardiography.

In the short axis projection, echocardiograms of a mitral orifice could be visualised at a much lower level than is normally seen in sequential cuts from the level of the upper part of the mitral valve leaflets to the region just above the papillary muscles. In some cases this gave the appearance of a funnel type effect with the orifice decreasing in size towards the apex. Six children were assessed as having two papillary muscles and four as having one (Fig. 6, 7, and 8), confirmation being obtained by surgery or necropsy in eight cases.

## Discussion

Congenital mitral stenosis is a rare condition occurring in 0.6 per cent of necropsied patients with congenital heart disease.<sup>1</sup> It is usually associated with other cardiac defects, particularly left sided. The frequency in coarctation of the aorta may be as high as 19 to 21 per cent.<sup>10</sup> Under 1 month of age in patients with congenital aortic stenosis the incidence of associated mitral stenosis is as high as 30 per cent.<sup>11</sup> The physical signs of the obstruction are often subtle and may go unrecognised because of these other lesions.

The anatomy of congenital mitral stenosis is variable, and can be the result of any or all of combinations of a supravalvular ring,<sup>12</sup> annular hypoplasia,<sup>7, 13</sup> and abnormalities of the leaflets, chordae tendineae, and papillary muscles.<sup>7, 12-14</sup>

By angiocardiography the detection of congenital mitral stenosis may be difficult; even with adequate pictures a failure of diagnosis in 10 out of 21 cases has been reported.<sup>1</sup> In particular, the chordae tendineae cannot be directly visualised, though it is possible to deduce by indirect means some of their anatomical abnormalities.<sup>13</sup>

M-mode echocardiography has enabled earlier diagnosis in some cases, but normal mitral valve appearances can be seen especially in the sick neonate with tachycardia. An assessment of either the site or severity of the obstruction by this technique has been unreliable.<sup>2</sup>

With the advent of digitised left ventricular echocardiograms it has been possible to study the effect of left sided inflow obstruction by measuring the rate of diastolic filling and the associated timing intervals.<sup>3</sup> This technique has provided a reliable method for assessing the extent of the physiological disturbance, and in postoperative evaluation in adults.<sup>15</sup> Classically in acquired mitral stenosis there is a reduced rate of change of dimension in diastole and a prolonged time from minimum dimension to 20 per cent peak dimension change. In addition, it has been shown that the period aortic closure to

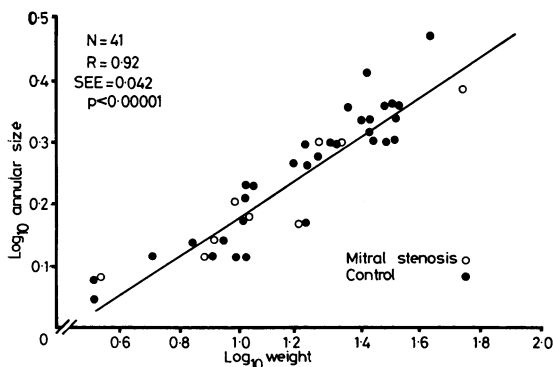


Fig. 3 Plot of the log of mitral annular size versus the log of the patient's weight. A line of identity is shown.

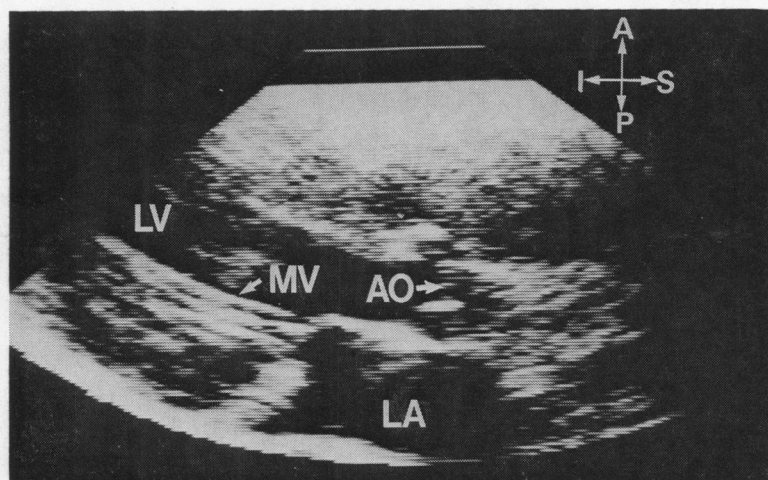


Fig. 4 Parasternal long axis view from a patient with congenital mitral stenosis. The mitral valve and subvalvular echocardiograms are dense, with reduced movement observed during diastole. LV, left ventricle; MV, mitral valve; LA, left atrium; AO, aorta.

mitral opening is shortened and the time from minimum dimension to mitral opening prolonged in acquired mitral stenosis.<sup>16</sup>

In the group studied only six out of the nine patients had features suggesting inflow obstruction, with the rate of change of dimension in diastole and minimum dimension to 20 per cent peak dimension change being the most reliable indices. Three patients, despite having significant obstruction, had normal filling patterns which suggested the stenosis was minimal. In all of these no other significant defects were present, which indicates that as an assessment of the severity of stenosis this technique is unreliable.

An explanation for this is that the M-mode cut is taken at the level of the leaflets where the inter-ventricular septum and posterior wall are seen, but is in a position proximal to the major inflow obstruction which in many cases is caused by

thickened fused chordae. If, during diastole, the mitral leaflets proximal to the obstruction moved outwards, taking the left ventricular wall with them, this could explain the normal filling pattern.

Two-dimensional echocardiography has played an important role in the assessment of acquired mitral stenosis.<sup>5-6</sup> In congenital mitral stenosis there is little information regarding the two-dimensional echocardiographic features.<sup>17</sup> The use of several standard views is essential for the accurate diagnosis and assessment of combined lesions. The presence of a supramitral ring has been shown by two-dimensional echocardiography,<sup>17</sup> but close proximity of the membrane to the mitral valve leaflets and annulus in two of our cases prevented its recognition. The parasternal long axis view was chosen for measuring the mitral annulus as it is easily reproducible in most cases and constant landmarks are identifiable. Though the view only

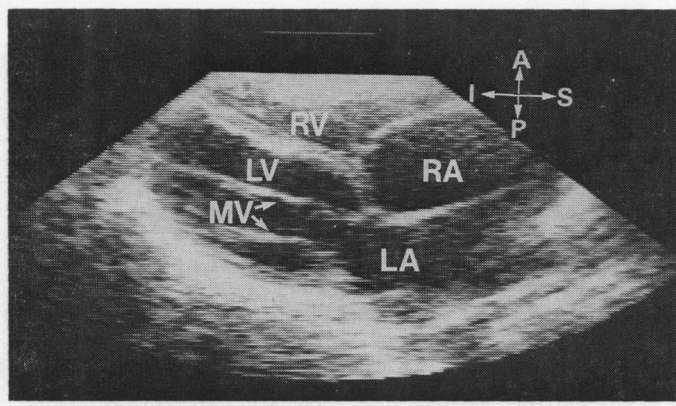
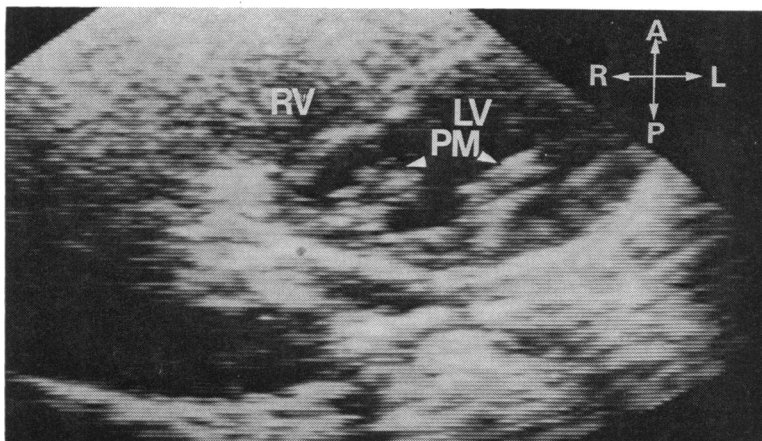


Fig. 5 Subxiphoid four chambered view in a child with congenital mitral stenosis having a dominant posteromedial papillary muscle and rudimentary anterolateral one. This was also seen in the short axis projection. The posterior leaflet is firmly attached to the anterolateral muscle with no normal movement seen during the cardiac cycle in real time. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle; MV, mitral valve.

Fig. 6 Short view of the left ventricle with two papillary muscles in a child with congenital mitral stenosis. The view is slightly more oblique than usual. RV, right ventricle; LV, left ventricle; PM, papillary muscles.



represents a segment of the true annulus, post-mortem and intraoperative observations suggest that if the annulus is small then its dimension should be reduced in all diameters. Furthermore, in two of these cases where the annular dimension was judged to be normal by two-dimensional echocardiography in life, confirmation of this normality was obtained at necropsy. In each of these cases the major obstruction was at the valve and subvalvular level. In five other cases surgical confirmation of the normal annular size was made.

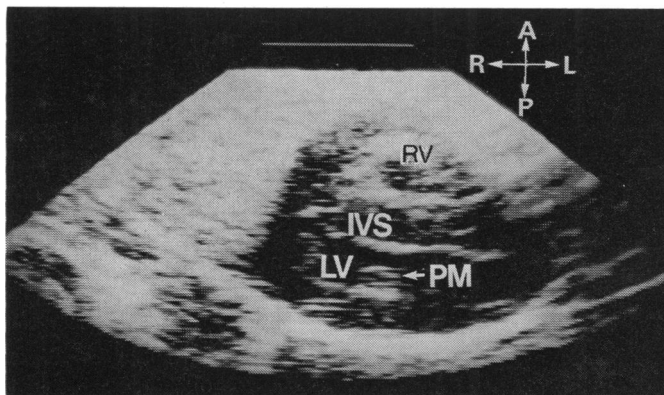
A thin rim of supramitral tissue, not seen by the two-dimensional echocardiogram, obscured intraoperative assessment of the true annulus in one patient and gave the appearance of a small ring caused by the associated mitral valve abnormality. Preoperative assessment by two-dimensional echocardiography, however, showed that the annular size was normal and that the abnormality was the

result of a parachute mitral valve.

In acquired mitral stenosis the short axis view has been reliable in detecting the effective orifice size and correlates well with that seen at operation or necropsy.<sup>6</sup> But in many children with congenital mitral stenosis the effective orifice is not that visualised in the short axis, since it may be asymmetrically situated.<sup>7</sup> This is a result of the thickened valve leaflets and chordae forming a tunnel of mitral valve tissue, which then inserts into the papillary muscles. The effective orifice lies between the papillary muscles and chordae and cannot be visualised by two-dimensional echocardiography.

The dense echocardiograms in the region of the valve and subvalvular apparatus seen in the four chamber and parasternal long axis view represent the above findings and also provide the explanation for the reduced mitral valve excursion observed in our cases.

Fig. 7 Short axis view of the left ventricle in a child with a single papillary muscle. RV, right ventricle; PM, papillary muscle; LV, left ventricle; IVS, interventricular septum.



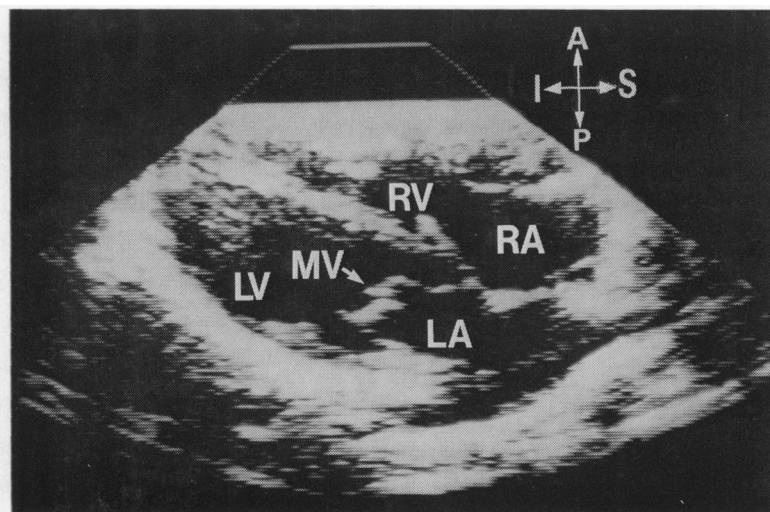


Fig. 8 Subxiphoid four chambered view in a child with a parachute mitral valve having an absent posteromedial papillary muscle. LA, left atrium; LV, left ventricle; MV, mitral valve; RA, right atrium; RV, right ventricle.

In the normal heart the anterolateral papillary muscle is slightly larger than the posteromedial, with a great variation between different hearts. The base to apex length of the papillary muscles and the number of major muscle groups also vary considerably.<sup>18</sup> The short axis and subxiphoid four chamber view are the most reliable for determining the number of muscles, as both groups can be displayed.<sup>8</sup> The subxiphoid is preferred to an apical four chamber view as in the latter only the posteromedial muscles can be seen.

In congenital mitral stenosis a wide spectrum of papillary muscle abnormalities can exist. They may be underdeveloped (Fig. 5) with a reduced inter-papillary distance, in some cases the muscles being very close or fused.<sup>7 14</sup> A single papillary muscle is often seen, the anterolateral one being absent in many cases, but a variable array is possible. In some cases where two papillary muscles are present, the chordae from the mitral leaflets insert totally into only one. Many of the above cases are associated with thickened and short chordae which play a major role in the pathogenesis of the stenosis. To the surgeon the presence of one or two papillary muscles is of great importance in determining the type of surgical procedure possible and the eventual prognosis.<sup>14</sup> In the past it has been difficult to detect reliably the presence of one or two papillary muscles by angiocardiography, with a detection rate of less than 50 per cent even in those without congenital mitral valve abnormalities or left ventricular outflow tract obstruction.<sup>19</sup> In congenital mitral stenosis the figure has been reported to be as low as 20 per cent of cases.<sup>1</sup> With two-dimensional echocardiography accurate detection is possible in all

cases where an adequate picture can be obtained.

Since two-dimensional echocardiography appears to be the most reliable detector of congenital mitral stenosis, we recommend its use not only when congenital mitral stenosis is suspected on clinical grounds, but also as a screening test in patients at risk of having masked mitral stenosis, particularly infants with coarctation of the aorta and aortic stenosis.

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