Isolated anterior mitral cleft

Two dimensional echocardiographic assessment and differentiation from "clefts" associated with atrioventricular septal defect

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SUMMARY Five patients with isolated clefts in the anterior leaflet of the mitral valve, unassociated with atrioventricular septal defects, are described. All had significant mitral regurgitation, with the cleft being the only abnormality in three. Two patients had an associated ventricular septal defect, one with a straddling right atrioventricular valve. Angiocardiography in four showed moderate regurgitation, but was not able to delineate the aetiology. Two dimensional echocardiography showed a constant defect in the anterior leaflet, pointing towards the left ventricular outflow tract. This differed from 30 cases with atrioventricular septal defects where the "cleft" pointed towards the interventricular septum and was situated between the anterior and posterior bridging leaflets. All cases with isolated clefts had surgical correction, with minimal residual regurgitation on follow-up examination in two cases. Our current policy in patients with uncomplicated isolated cleft involves non-invasive assessment of these children and surgical correction if the regurgitation is significant.

Most cardiologists and surgeons have adopted a conservative approach to the child with mitral regurgitation. This is justified as the prospect of mitral valve replacement in this age group is not to be considered lightly, in view of the problems associated with prosthetic valves.¹ Reconstructive surgery may prove to be possible, but usually this cannot confidently be decided before operation on the basis of traditional investigational techniques. So the child goes for operation in an atmosphere of uncertainty, with all those involved hoping, but usually not knowing, that reconstructive surgery will be possible.

Part of the basis for uncertainty is the wide variability of anatomical substrates for congenital mitral regurgitation. This may be produced by redundant and deficient chordae tendineae, parachute mitral valve and its variants, anomalous mitral arcade, floppy mitral valve, and, rarely, Ebstein's anomaly of the mitral valve.²⁻³

An isolated cleft in the anterior (aortic) leaflet of the mitral valve is an uncommon cause of regurgitation, the cleft always pointing towards the left ventricular outflow tract. But it is an important lesion because it is potentially correctable.⁴

In the past, M-mode echocardiography has provided functional information about mitral regurgitation, but apart from mitral valve prolapse has yielded little morphological detail as to the exact nature of the lesion.⁵ Angiocardiography has permitted recognition of some specific varieties of congenital mitral valve disease, but has proved singularly unsuccessful in the recognition of isolated mitral cleft.⁶ In contrast, two dimensional echocardiography should enable recognition of an isolated cleft.

We have treated a group of children where the mitral regurgitation was the result of a localised cleft in the anterior leaflet, unassociated with atrioventricular septal defects. We now describe their preoperative assessment, surgical management, and our current approach to the problem.

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Subjects and methods

All patients with isolated anterior mitral clefts attended the congenital heart clinics of The Hospital for Sick Children, Great Ormond Street, or the National Heart Hospital, Westmoreland Street. In all cases a clinical diagnosis of mitral regurgitation had been made and radiological evidence of left ventricular enlargement was present. Of the five patients studied, four had angiocardigraphic evidence of moderate to severe mitral regurgitation. The fifth patient, in view of our previous experience, was submitted to operation without prior invasive studies. Two cases had an associated ventricular septal defect, one with a straddling right atrioventricular valve diagnosed by echocardiography. In the others, the anterior mitral cleft was an isolated defect.

All patients had both M-mode and two dimensional echocardiographic assessments. The patients were studied with either an Advanced Technology Laboratory mechanical sector scanner using a 3.0 MHz transducer, or a Smith Kline mechanical scanner with a 2.25 MHz scan head. The apical and subcostal four chamber views were used to assess the leaflets, as was the short axis view at the level of the aortic root. In the latter cut, a scan from the aortic root to the apex of the left ventricle enabled an accurate assessment of the anterior and posterior mitral leaflets and associated papillary muscles.

In all patients M-mode studies were performed to assess the mitral valve motion and left ventricular cavity size. Two cases had preoperative, and three post-operative pulsed Doppler studies performed by sampling in the left atrium. All were submitted for surgical repair of the cleft anterior mitral leaflet.

To clarify the difference between an isolated anterior mitral cleft and that associated with an atrioventricular septal defect, a further group of 30 patients with the latter abnormality was included in the study. All of these patients had either angiocardigraphic or surgical confirmation of the defect.

PREOPERATIVE ECHOCARDIOGRAPHIC STUDIES

Patients with an isolated mitral cleft

In all cases there was a localised cleft in the anterior (aortic) leaflet of the mitral valve (Fig. 1 and 2). This was best appreciated in a short axis scan from the aortic root to the apex of the left ventricle. Just below the aortic root, a position is obtained wherein the left ventricular outflow tract and the area of aortic to mitral valvar fibrous continuity can be visualised. When the mitral valve is normal, the anterior leaflet is seen as a continuous dense echo (Fig. 3), but in patients with an anterior cleft there is a break in this echo (Fig. 4). In all cases the edges of the cleft

appeared as denser echoes suggesting that they were rolled and thickened, this observation being confirmed at operation (Fig. 4). During real-time study, an abnormal pattern of movement of the anterior leaflet existed throughout the assessment. So, during systole the two portions of the anterior leaflet floated together, whereas during diastole, the edges of the cleft parted widely. Most importantly, the cleft extended into the area of aortic-mitral continuity, and was not directed towards the interventricular septum. All cases had two papillary muscles with no other abnormality of the mitral valve being seen. In particular, the chordae from the anterior mitral leaflet were normally situated.

In the apical four chamber view the effective mitral orifice is frequently not visualised, as the anterior mitral leaflet crosses the plane of the transducer beam before inserting into the posteromedial papillary muscle group. When a localised anterior cleft is present, an echo free area is readily visualised in this cut, representing the break in the leaflet. This appearance can also be visualised in the subcostal four chamber view (Fig. 5).

In all cases the inferior limbus of the interatrial septum was intact, and there was a normal atrioventricular muscular septum (Fig. 5). Thus, the atrioventricular valves were in all cases attached to the septum

Fig. 1 A diagrammatic representation of the surgeon's view of the left atrioventricular valve via a left atrial incision. Note the difference between the cleft in a normal anterior mitral leaflet as compared with the "cleft" seen in an atrioventricular septal defect. In the latter case the "cleft" exists between the anterior and posterior bridging leaflets.

Fig. 2 Necropsy specimen from a patient with an isolated cleft in the anterior mitral leaflet. The picture on the left shows the cleft anterior leaflet. The picture on the right shows the cleft pointing towards the left ventricular outflow tract. AML, anterior mitral leaflet; CL, cleft; LA, left atrium; LV, left ventricle; LVOT, left ventricular outflow tract. This figure is reproduced by kind permission of Professor Anton Becker, Amsterdam.

Fig. 3 The upper panel shows a normal mitral valve at the level of the left ventricular outflow tract. Note the anterior mitral leaflet is in fibrous continuity with the aorta. The specimen on the right is cut in the same plane to indicate the echocardiographic features. The lower panel is a lower short axis cut at the level of the mitral leaflets. Note the fishmouth appearance of the mitral valve. The specimen on the right is cut on the same plane as the echocardiogram. MV, mitral valve; RV, right ventricle; RVOT, right ventricular outflow tract; TV, tricuspid valve. See Fig. 2 for remaining abbreviations.

Fig. 4 The top picture is a short axis cut at the level of the left ventricular outflow tract in a patient with an isolated cleft in the anterior mitral leaflet. Note the cleft, indicated by the two arrows pointing towards the outflow tract, and not the right ventricle. The lower picture is a short axis cut at the level of the mitral valve. Note again the cleft indicated by the arrows, pointing towards the outlet of the heart and not the right ventricle. IVS, interventricular septum. See Fig. 2 and 3 for remaining abbreviations.

at different levels, with the tricuspid valve being attached more inferiorly. The membranous and inlet septa were intact in all patients, including the one with a ventricular septal defect and straddling right (tricuspid) atrioventricular valve. Surprisingly, the defect in the latter patient was found to be in the muscular trabecular septum with chordae from the septal tricuspid leaflet straddling to be attached in the morphologically left ventricle. In this case, previously described,⁷ a posterior interventricular septum going to the crux was present. In the other case the defect was perimembranous. M-mode assessments of the mitral valve showed a normal "M" shaped pattern with some associated fluttering of the anterior leaflet during diastole. The remaining features were those of atrial and ventricular enlargement.

The left ventricular end-diastolic volume was greater than two standard deviations above the mean for age in every case and the left atrial size was also significantly increased.

Patients with atrioventricular septal defects

In all cases the short axis cut showed that the "cleft" was situated between the anterior and posterior bridging leaflet pointing towards the interventricular septum and not the left ventricular outflow tract (Fig. 6).

INTRAOPERATIVE FINDINGS AND MANAGEMENT OF ISOLATED MITRAL CLEFTS

All patients were operated on cardiopulmonary bypass, with moderate hypothermia and cold cardioplegia. The left atrium was opened posterior to the

Fig. 5 The upper panel is a subcostal four chamber cut in a case with an isolated cleft in the anterior mitral leaflet. Note the division in the anterior leaflet indicated by the two arrows. The posterior mitral leaflet is seen in the upper part of the picture. The lower picture is from the same patient showing the presence of an atrioventricular muscular septum indicated by the two arrows. PML, posterior mitral leaflet; VAS, ventriculoatrial septum. See Fig. 2, 3, and 4 for remaining abbreviations.

Waterston groove. The mitral valve was tested by injecting cold saline under pressure into the left ventricular apical vent. The left atrium was moderately enlarged in all patients. A universal finding was a deep cleft in the anterior leaflet, pointing towards the aortic root and extending to the annulus (Fig. 1 and 2). The edges of the cleft were thickened and rolled. The anterior part of the leaflet had chordal attachments to the anterior papillary muscle. The subvalvular apparatus was normal in all patients.

The repair consisted in all cases of closing the cleft with interrupted sutures starting at the annulus and progressing towards the free edge of the valve. The regurgitation at operation was completely abolished in

all cases but one, where a small central jet remained. In one patient in whom the regurgitation was abolished at operation, on follow-up there was a soft pansystolic murmur at the apex with no diastolic murmur. In these two patients pulsed Doppler ultrasound showed a regurgitant jet in the left atrium. Three of the patients had no residual murmur of mitral regurgitation, and this absence of regurgitation was confirmed in one patient by pulsed Doppler ultrasound.

In the patient with the trabecular ventricular septal defect and straddling right (tricuspid) atrioventricular valve, the defect was closed through the tricuspid valve, accepting a small defect around the papillary

Fig. 6 These pictures are from a case with an atrioventricular septal defect. The upper picture is a precordial short axis cut at a slightly higher level than that seen in the lower panel. Note that the "cleft" between the anterior and posterior bridging leaflets points towards the interventricular septum and not the outflow tract of the left ventricle. The bottom panel, at a lower level, again indicates the cleft between the leaflets pointing towards the interventricular septum. The specimen on the right is from a patient with an atrioventricular septal defect and partitioned orifices. The cut is in the same plane as the bottom left echocardiogram. Note the cleft, indicated by the arrow pointing towards the interventricular septum. LAV, left atrioventricular valve; RAV, right atrioventricular valve. For remaining abbreviations see Fig. 3 and 4.

muscle attached to the left side of the septum.

Discussion

These findings throw light on two largely separate questions, one to do with descriptive anatomy and one clinical. Firstly, what is the difference between isolated cleft of the anterior mitral leaflet and the "mitral cleft" found in atrioventricular septal defects?

Secondly, how may patients with isolated cleft of the anterior mitral leaflet be distinguished from patients with mitral regurgitations from other causes?

CLEFTS AND ATRIOVENTRICULAR SEPTAL DEFECTS

The normal mitral valve consists of two leaflets, the anterior or aortic leaflet and the posterior or mural one. These are divided by the anterolateral and posteromedial commissures which are supported by paired left ventricular papillary muscles, the anterolateral and posteromedial group^{8,9} (Fig. 1). "Clefts" are usually described as part of the anatomy of atrioventricular septal defects. In these cases, a normal mitral valve does not exist and the left sided valve is formed from components of three of the five

basic leaflets, which may be found in all atrioventricular septal defects. The "left sided cleft" in such hearts actually lies between the anterior and posterior bridging leaflets as they insert along the crest of the inter-ventricular septum¹⁰ (Fig. 1 and 6).

Isolated clefts of the mitral valve are rare, but have previously been described in the anterior and posterior leaflets unassociated with atrioventricular defects.⁴ The cleft usually involves the anterior leaflet and divides it in its entirety (Fig. 1). Anomalous chordal attachments are frequently a complicating feature though we found none in these patients. It must be stressed, however, that if the cleft involves the anterior leaflet and is to be regarded as isolated, it must point towards the left ventricular outflow tract. In those with an atrioventricular septal defect, because of the very nature of the leaflet pattern the "cleft" can in no sense divide the anterior leaflet¹⁰ as an isolated mitral cleft does. And because it does not divide the anterior leaflet it points to the inter-ventricular septum (compare Fig. 1, 4, and 6) rather than to the left ventricular outflow tract.¹¹

These considerations lead us to suppose that, if the very rare conditions of atrioventricular septal defect with intact atrial and ventricular septa were encountered, the cleft in the "left atrioventricular valve" would still point towards the septum. Furthermore, the other echocardiographic stigmata of atrioventricular septal defects would still be present, notably the presence of two atrioventricular valves at the same level, resulting from absence of the atrioventricular septum.^{12 13}

DIFFERENTIATION FROM OTHER MITRAL ANOMALIES, AND CONSEQUENCES FOR MANAGEMENT

Mitral valve abnormalities in young children are almost always of congenital origin, though mitral regurgitation may also be secondary to primary left ventricular disease, or to anomalous origin or atresia of the left main coronary artery. In older children, rheumatic mitral disease is more common. The echocardiographic features of congenital mitral stenosis have previously been described.^{14 15} In patients in whom the predominant lesion is regurgitation, the majority have no gross malformation of the valve, but possess redundant leaflets and a dilated annulus. Others have grosser abnormalities such as single papillary muscles. Over the past few years, particularly since the advent of M-mode echocardiography, prolapse of the mitral leaflets has been suggested as a likely cause of this regurgitation. Normally the posterior leaflet is involved, but occasionally the anterior one is.^{16 17}

The majority of children with gross structural abnormalities of the mitral valve who present in early

childhood have associated defects which frequently mask the signs of mitral valve disease. Indeed, it is rare for them to have only isolated mitral regurgitation. In the older age group cardiac failure is rare and the lesion is usually detected during routine examination. Even though mitral regurgitation is severe, the combination of good left ventricular function and frequently massive left atrial enlargement ensure that patients maintain a relatively normal left atrial pressure and have little in the way of symptoms, at least in their first decade. Because the condition is well tolerated, and it has not been possible to guarantee that surgery will not involve mitral valve replacement, the tendency has been to postpone surgery until symptoms demand it. Cardiac catheterisation and angiocardiology have in our experience often not provided any more information than was obvious from clinical and non-invasive investigation.

We believe that these results show the possibility of a radical change in management for at least one subgroup of patients with congenital mitral regurgitation. This is because two dimensional echocardiography provides the necessary detailed information as to the precise anatomical abnormalities of the leaflets and subvalvar apparatus. Such simple structural abnormalities as localised clefts are readily visualised. These patients can then be selected for early surgical repair on the basis that a good result can be anticipated, thereby avoiding the problems of left ventricular dysfunction which may supervene in patients with long standing mitral regurgitation.¹⁸ We believe that these patients reflect the fact that the mitral regurgitation is largely, if not entirely, the result of the cleft. Repair of the cleft produces a normal or near normal anterior mitral leaflet (Fig. 7).

This situation is quite different from that pertaining to "left sided clefts" in atrioventricular septal defects. Here the "cleft" is but one cause of left atrioventricular valve regurgitation, and indeed may not cause any regurgitation at all. Repairing the "cleft" in no way restores a normal "anterior mitral leaflet", and may actually result in left ventricular inflow obstruction.^{10 19 20}

From a small series such as this one cannot be dogmatic about the overall functional results of surgery. They do compare favourably, however, with the results of mitral repair for all congenital mitral anomalies reported by Carpentier and colleagues.²¹ These authors found a persistent apical systolic murmur in 22 out of 34 patients, and a significant reduction in heart size in only seven.

Our present policy is therefore to perform two dimensional echocardiographic studies in all patients with clinical evidence of mitral regurgitation. If an isolated cleft is diagnosed and no other complicating suspected features are present, such as coarctation of

Fig. 7 This is a postoperative short axis cut in a patient with a repaired localised cleft in the anterior mitral leaflet. Note the suture line indicated by the dense echo in the centre of the anterior mitral leaflet. See Fig. 2 and 3 for abbreviations.

the aorta or severe pulmonary vascular disease, the degree of regurgitation is assessed clinically, radiographically, and by measuring left atrial and left ventricular sizes. If it is graded as moderate to severe, then the patients are submitted for surgical repair of the valve. Careful follow-up studies of mitral valve function in a large series of patients will obviously be necessary.

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References

- Berry BE, Ritter DG, Wallace RB, McGoon DC, Danielson CK. Cardiac valve replacement in children. *J Thorac Cardiovasc Surg* 1974; 68: 705-10.
- Davachi F, Moller JH, Edwards JE. Disease of the mitral valve in infancy. An anatomic analysis of 55 cases. *Circulation* 1971; 43: 565-79.
- Ruschhaupt DG, Bharati S, Lev M. Mitral valve malformation of Ebstein type in absence of corrected transposition. *Am J Cardiol* 1976; 38: 109-12.
- Goodman DJ, Hancock EW. Secundum atrial septal defect associated with a cleft mitral valve. *Br Heart J* 1973; 35: 1315-20.
- Schwartz DC, Kaplan S, Meyer RA. Mitral valve prolapse in children: clinical, echocardiographic and cineangiographic findings in 81 cases (abstract). *Am J Cardiol* 1975; 35: 169.
- Macartney FJ, Bain HH, Ionescu MI, Deverall PB, Scott O. Angiocardiographic/pathologic correlations in congenital mitral valve anomalies. *Eur J Cardiol* 1976; 4: 191-211.
- Smallhorn JF, Tommasini G, Macartney FJ. Detection and assessment of straddling and overriding atrioventricular valves by two-dimensional echocardiography. *Br Heart J* 1981; 46: 254-62.
- Lam JHC, Ranganathan N, Wigle ED, Silver MD. Morphology of the human mitral valve. I. Chordae tendineae: a new classification. *Circulation* 1970; 41: 449-58.
- Ranganathan N, Lam JHC, Wigle ED, Silver MD. Morphology of the human mitral valve. II. The valve leaflets. *Circulation* 1970; 41: 459-67.
- Carpentier A. Surgical anatomy and management of the mitral component of atrioventricular canal defects. In: Anderson RH, Shinebourne EA, eds. *Paediatric cardiology* 1977. Edinburgh, London: Churchill Livingstone, 1978: 477-90.
- Beppu S, Nimura Y, Sakakibara H, et al. Mitral cleft in ostium primum atrial septal defect assessed by cross-sectional echocardiography. *Circulation* 1980; 62: 1099-107.
- Hagler DJ, Tajik AJ, Seward JB, Mair DD, Ritter DG. Real-time wide-angle sector echocardiography: atrioventricular canal defects. *Circulation* 1979; 59: 140-50.
- Smallhorn JF, Tommasini G, Anderson RH, Macartney FJ. Assessment of atrioventricular septal defects by two-dimensional echocardiography. *Br Heart J* 1982; 47: 109-21.
- Driscoll DJ, Gutgesell HP, McNamara DG. Echocardiographic features of congenital mitral stenosis. *Am J Cardiol* 1978; 42: 259-66.
- Smallhorn JF, Tommasini G, Deanfield J, Douglas J, Gibson D, Macartney FJ. Congenital mitral stenosis. Anatomical and functional assessment by echocardiography. *Br Heart J* 1981; 45: 527-34.
- DeMaria AN, King JF, Bogren HG, Lies JE, Mason DT. The variable spectrum of echocardiographic manifestations of the mitral valve prolapse syndrome. *Circulation* 1974; 50: 33-41.
- Bisset GS, Schwartz DC, Meyer RA, James FW, Kaplan S. Clinical spectrum and long-term follow-up of isolated mitral valve prolapse in 119 children. *Circulation* 1980; 62: 423-9.
- Kirklin JW. Replacement of the mitral valve for mitral incompetence. *Surgery* 1972; 72: 827-36.
- Griffiths SP, Ellis K, Burris JO, Blumenthal S, Bowman FO Jr, Malm JR. Postoperative evaluation of mitral valve function in ostium primum defect with cleft mitral valve (partial form of atrioventricular canal). *Circulation* 1969; 40: 21-9.
- Losay J, Rosenthal A, Castaneda AR, Bernhard WH, Nadas AS. Repair of atrial septal defect primum. Results, course, and prognosis. *J Thorac Cardiovasc Surg* 1978; 75: 248-54.
- Carpentier A, Branchini B, Cour JC, et al. Congenital malformations of the mitral valve in children. Pathology and surgical treatment. *J Thorac Cardiovasc Surg* 1976; 72: 854-66.

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