Mitral Cleft in Ostium Primum Atrial Septal Defect Assessed by Cross-sectional Echocardiography

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SUMMARY We attempted to detect mitral deformities in ostium primum atrial septal defect using real-time cross-sectional echocardiography. Transverse sections of the anterior mitral leaflet echo were examined in 11 patients with this malformation who subsequently received surgical treatment. The section for observing the transverse view of the anterior leaflet was along the sagittal plane of the body, because of the deformity of the mitral annulus. Each echocardiographic finding was compared with the surgical and angiographic findings. On the echocardiogram, the superior and inferior parts of the anterior mitral leaflet separated into two parts during diastole in all patients with mitral cleft. Thin linear echoes connected the ridges of the cleft and the ventricular septum in seven patients in whom the accessory chordae at that area were revealed at surgery. The systolic configuration of the anterior leaflet echo varied among the patients. The severity of the mitral regurgitation seemed to relate not only to the size of the cleft but also to the systolic configuration of the anterior mitral leaflet. After surgery, diastolic separation of the anterior leaflet echo was no longer observed. However, the abnormal systolic configuration of the anterior leaflet was unchanged.

IN OSTIUM PRIMUM atrial septal defect (ASD), absence of the membranous septum and a cleft in the anterior mitral leaflet are the characteristic features resulting from maldevelopment of the endocardial cushions. The success of the surgery depends on the repair of the mitral regurgitation due to mitral cleft, because mitral regurgitation sometimes remains after surgery.1-5 Mitral repair in this malformation is difficult due to the complex deformities of the mitral valve and the deformity of the mitral annulus. At present, however, there is no adequate method to demonstrate the details of the mitral deformities before surgery or the results of the repair of the cleft after surgery. Angiocardiography reveals the presence of the mitral cleft and regurgitation but reveals few details about the leaflet and chordae tendineae.6-8 Although the mitral anomalies can be seen at surgery, the width of the cleft and the configuration of the mitral apparatus are modified in the arrested heart, and the exact orientation of the mitral apparatus is obscure. Therefore, a method of evaluating the condition of the mitral apparatus in the beating heart is needed.

As echocardiography offers unique information about the arrangement and movement of the intracardiac structures, it has come to be an indispensable examination in diagnosing congenital and acquired heart diseases. The echocardiographic findings in endocardial cushion defect, including ostium primum ASD, have been reported.9-17 In mitral cleft, the systolic multiple echoes of the mitral valve were reported as specific findings using M-mode and multcrystal echocardiography.11,12,14 Mitral cleft has been observed with real-time cross-sectional echocardiography.16,17 In the present study our goal was to detect the mitral cleft, to show the abnormal configuration of the anterior mitral leaflet using real-time cross-sectional echocardiography and to compare the echocardiographic findings with the surgical and angiographic findings in patients with ostium primum ASD.

Subjects and Methods

We studied only subjects who later underwent surgery to permit the comparison of echocardiographic and anatomic findings. Ten patients with ostium primum ASD and one patient with a transitional form of endocardial cushion defect18 were included. The latter patient was included because her ventricular septal defect was small and was considered clinically to be the same as ostium primum ASD. There were five males and six females, ages 4-45 years. At surgery, 10 patients had mitral cleft and the other one did not. For the sake of convenience, the degree of the mitral cleft was defined as complete or incomplete, according to its extension to the mitral annulus. Seven of the 10 had complete cleft and the other three had incomplete cleft. In nine patients with mitral cleft, the cleft was sutured directly; one patient underwent mitral valve replacement. The accessory chordae were revealed at surgery in five patients and were resected partially in two. The mitral regurgitation before and after surgery was diagnosed by cineangiography using the criteria of Sellers et al.19 The angiographic and surgical findings for individual patients are listed in table 1.

All patients were examined using real-time cross-sectional echocardiography before surgery and all

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Received February 21, 1979; revision accepted May 6, 1980.

TABLE 1. Angiocardiographic and Surgical Findings

<table>
<thead>
<tr>
<th>Pt</th>
<th>Age (years)</th>
<th>Before surgery MR (cine)</th>
<th>During surgery</th>
<th>Accessory chordae</th>
<th>Method</th>
<th>After surgery MR (cine)</th>
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<td>24</td>
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<td>Cleft: Complete</td>
<td>+</td>
<td>Suture</td>
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<td>+</td>
<td>Sutureculated</td>
<td>2+</td>
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<tr>
<td>11</td>
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<td>1+</td>
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<td>+</td>
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</tbody>
</table>

*Transitional form of endocardial cushion defect.
†No mitral regurgitant murmur.
‡The accessory chordae were partially resected.

Abbreviations: MR = mitral regurgitation; MVR = mitral valve replacement; + = present; - = absent.

except the patient who underwent mitral valve replacement were also examined after surgery. The echocardiographic findings of the mitral configuration and motion were compared with those in healthy subjects and patients without abnormalities in the mitral valve.

The echocardiograph used was a Toshiba SSH-11A or a Varian V-3000, which are electronic phased-array sector systems. In the Toshiba SSH-11A, the transducer has 32 elements with a frequency of 2.4 MHz. The scanning angle is 78° and the line density is 224 lines/frame. In the Varian V-3000, the transducer has 32 elements with a frequency of 2.25 MHz. The scanning angle is 84° and the line density is 85, 128 or 255 lines/frame at the depth level of 21, 15 or 7 cm, respectively. Real-time cross-sectional echocardiograms were recorded on 8-mm cine film or a videotape recorder. The cross-sectional echocardiograms at a given cardiac phase were recorded on Polaroid film or obtained later from 8-mm cine film.

Echocardiographic examinations were performed with the patient in the supine position. The cross-sectional plane was set to observe the transverse view of the anterior mitral leaflet. In healthy subjects and patients with cardiac disease other than ostium primum ASD, the section was set along the short axis, as the mitral annulus is parallel to the short axis. However, in ostium primum ASD, the right side of the mitral annulus is displaced toward the apex and the anterior part of the annulus is parallel not to the short axis but to the sagittal plane of the body, as shown in the left ventriculogram (fig. 1A). Therefore, the section for observing the transverse view of the anterior mitral leaflet was set to be the sagittal plane of the body so that the section was parallel to the anterior part of the mitral annulus. First, the transducer was placed in the third, fourth or fifth interspace, and the echocardiographic section was set along the long axis of the left ventricle (fig. 1B-1). Then, the transducer was rotated clockwise to the sagittal plane of the body while the echo of the anterior mitral leaflet was set to be the center of the cross-sectional image (fig. 1B-2). When the echo of the posterior mitral leaflet or the left ventricular posterior wall was observed, the transducer was tilted medially to ensure that the chordae tendineae were not being examined (fig. 1B-3). Though this section was oblique to the left ventricle, it was perpendicular to the anterior mitral leaflet in ostium primum ASD. To scan the entire area of the anterior leaflet, the transducer was tilted carefully from the direction of the mitral annulus to that of the tip of the anterior leaflet.

In three patients, contrast studies with cross-sectional echocardiography at cardiac catheterization were available. Isotonic saline was injected into the left ventricular cavity and the location of the regurgitation was examined.

Results

Normal Anterior Mitral Leaflet

In healthy subjects and patients with cardiac diseases other than ostium primum ASD, the echo of the anterior mitral leaflet was a curved line parallel to the ventricular septum echo during diastole and straight or slightly concave toward the left atrium and lying between the left ventricular outflow tract and the left atrium during systole (fig. 2). There was neither interruption of the leaflet echo nor asynchronous motion in any part of the leaflet throughout the cardiac phase.

Ostium Primum ASD

The superior and inferior portions of the anterior mitral leaflet separated into two parts during diastole, and the superior portion narrowed the left ventricular outflow tract (figs. 3 and 4). Each part of the leaflet
ECHO OF MITRAL CLEFT IN OSTIUM PRIMUM ASD/Beppu et al.

**Figure 1.** (A) Frontal view of the angiocardiogram in ostium primum atrial septal defect showing that the right side of the contour of the left ventricle (LV) consists of the mitral annulus. The correct section at which to observe the transverse view of the anterior mitral leaflet by cross-sectional echocardiography is indicated by the triangle as a spatial plane. The white arrows indicate mitral regurgitation. The black arrow points to the nonopaque notch that indicates mitral cleft. (B) Schematic drawings indicating the procedure to set the section. AO = aorta; Ao.v = aortic valve.

**Figure 2.** Transverse views of the anterior mitral leaflet (AML) in a healthy subject showing an uninterrupted curve during diastole (left) and a straight line during systole (right). RV = right ventricle; IVS = ventricular septum; LV = left ventricle; LA = left atrium.
had a different fluttering motion in diastole. These findings during diastole were found in all patients with mitral cleft but not in the patient without cleft (patient 6). The extent of the cross section in which the diastolic separation of the anterior mitral leaflet echo was observed varied among the patients. In patients with a complete cleft, the diastolic separation was easily detected in the wide area. In patients with an incomplete cleft, the diastolic separation was not detected near the annulus.

During systole, the echo of the two parts of the leaflet met at the center and formed a knot echo, somewhat resembling two hands pressed together. The size of the knot echo varied among the subjects from well-defined (fig. 4) to scarcely defined (fig. 3). In the patients with a well-defined knot echo, the angiocardiograms showed a wide nonopaque notch indicating mitral cleft. In patient 8 there was an opening between the ridges of the two parts of the leaflet during systole.

In five patients, the echo line of the anterior mitral leaflet was not straight, but bent toward the ventricular septum with the angle at the contact site (fig. 3). The echo of the anterior leaflet was almost straight in four patients (fig. 5), one of whom did not have the cleft. In one of the other two patients, the edge of one part of the leaflet slipped down the edge of the other part so that they did not meet symmetrically, and the echo line of the leaflet was not straight (patient 3) (fig. 4). In the other, patient 4, the prolapse of the anterior mitral leaflet was shown by echocardiography and the configuration of the anterior leaflet during systole was not assessed.

In seven patients, thin linear echoes were detected between the ventricular septum and the ridges of the two parts of the divided leaflet echoes (fig. 3). At surgery, accessory chordae were found in the area concerned. In some patients, these echoes were not very strong and could not be recognized clearly on the real-time image, but were seen on the 8-mm frame-by-frame display. All patients with systolic bending of the anterior leaflet echo had accessory chordae, which looked as if they were preventing the anterior leaflet...
from being straight during systole (fig. 3). Conversely, the patients with the accessory chordae did not always show the systolic bending of the anterior leaflet echo. The cross-sectional echocardiographic findings before surgery are listed in table 2.

Eight patients had complete cleft, but the severity of mitral regurgitation varied. In patient 11, whose echocardiogram showed the thin linear echoes and systolic bending of the anterior mitral leaflet echo, angiocardiography showed minimal mitral regurgitation. In patient 7, who did not have the accessory chordae and whose anterior leaflet echo was straight, angiocardiography showed severe mitral regurgitation. In this patient, a contrast study showed regurgitation from the site between the two parts of the leaflet (fig. 5). The patient who had an opening between the ridges of the divided leaflet echo had severe mitral regurgitation and the same finding in the contrast study as in patient 7.

In all patients, after surgical repair of the cleft, the anterior mitral leaflet echo did not show the diastolic separation (fig. 6). It seemed that the motion of the
Table 2. Echocardiographic Findings

<table>
<thead>
<tr>
<th>Pt</th>
<th>Diastolic separation*</th>
<th>Thin linear echoes</th>
<th>Systolic bending</th>
<th>Diastolic separation</th>
<th>Thin linear echoes</th>
<th>Systolic bending</th>
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<td>-</td>
<td>+</td>
<td>*</td>
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</tbody>
</table>

*++ and + mean that this finding was observed from near the annulus to the tip of the leaflet and only near the tip of the leaflet, respectively.
†The systolic configuration was not assessed because of the prolapse of the anterior mitral leaflet.
Abbreviation: MVR = mitral valve replacement.

The anterior mitral leaflet was not restrained by the suture site. The knot echo was observed throughout the cardiac phase, indicating the suture site. In three patients who showed systolic bending of the anterior leaflet echo before surgery and whose accessory chordae were not resected, the thin linear echoes were detected at the same area as in the preoperative state, and the degree of systolic bending of the anterior leaflet was unchanged (fig. 6). In these patients, the severity of the mitral regurgitation by angiocardiography was unchanged in two and diminished in one. In one of two patients who showed systolic bending of the leaflet before surgery and whose accessory chordae were partially resected at surgery, the echo of the anterior leaflet was almost straight and mitral regurgitation had decreased (patient 10). In two patients who had neither the accessory chordae nor systolic bending of the anterior leaflet mitral regurgitation was not revealed. The severity of mitral regurgitation was not reduced in the patient with mitral valve prolapse.

FIGURE 5. Contrast study in ostium primum atrial septal defect (patient 7). The echo of the anterior leaflet is almost straight. An arrow indicates contact site of the separated anterior leaflet (left). Immediately after saline injection into the left ventricle (LV), contrast echoes filled almost the entire ventricular cavity and some of them are observed in the left atrium (LA) from the contact site of the two parts, as indicated by the arrow (middle). In the following phase, the contrast echoes in the left atrium have increased (right). RV = right ventricle; IVS = ventricular septum.
mitral cleft is diagnosed by the nonopaque notch in the mitral area. This notch would correspond to the knot echo formed by the ridges of the divided leaflet in the cross-sectional echocardiogram.

The Echo of the Accessory Chordae

The thin linear echoes between the ridges of both parts of the anterior leaflet and the ventricular septum are believed to be the echo of the accessory chordae because there was neither a false-positive nor a false-negative case. To detect these echoes in some patients required careful observation. However, the presence of accessory chordae could not be detected by any means except echocardiography.

In some patients, the accessory chordae seemed to restrain the anterior mitral leaflet from being straight during systole, causing it to bend toward the ventricular septum. This condition was shown pathologically. The accessory chordae may prevent the two parts of the anterior leaflet from prolapsing into the left atrium at the site of the cleft.

Apex four-chamber echocardiography has been recommended as being possibly superior for showing the accessory chordae, but it does not show the mitral cleft at the same time. Because it is necessary to show the presence of the accessory chordae and their relationship to the mitral cleft, we consider the sagittal plane the superior view.

Mitral Regurgitation in Ostium Primum ASD

Mitral regurgitation in ostium primum ASD is thought to be produced at the site of the cleft. In this study, the degree of the mitral regurgitation was not directly proportionate to the size of the cleft. In patient 11, the mitral regurgitation was minimal even though the cleft was complete; in patient 7, however,
the mitral regurgitation was severe even though the cleft was incomplete. Therefore, several possible factors causing regurgitation must be considered (table 3). In ostium primum ASD, there are two coaptation parts in the mitral valve. First, regurgitation depends upon the coaptation of the two parts of the anterior mitral leaflet divided by the cleft. The width of the gap between the two parts during systole is the prime factor. In patient 8, who had a complete cleft and a relatively wide space between the two parts of the anterior leaflet during systole, mitral regurgitation was severe. The width of the cleft in diastole did not correlate with the degree of mitral regurgitation. Another factor is the absence of accessory chordae. If they are absent, the edges of the cleft may slip out of the proper position for the coaptation of the two parts of the anterior leaflet. In patients 3, 7 and 8, who did not have the accessory chordae, the mitral regurgitation was severe. In patient 7, the echo of the anterior mitral leaflet was almost straight, as in the normal leaflet. However, this finding may indicate that there is no coaptation between two parts of the anterior leaflet. This hypothesis was supported by the contrast study, which showed that contrast echoes flowed into the left atrium from the site of the cleft. Conversely, even in the patients with a complete cleft, the accessory chordae seemed to aid the coaptation between the two parts of the anterior leaflet and reduce mitral regurgitation (patients 1, 2, 5, 9 and 11). Other factors, such as the site of the papillary muscle or the deformity of the annulus, may cause imperfect coaptation of the two parts of the anterior mitral leaflet. In this study, no suggestive finding was obtained concerning these factors.

Second, in ostium primum ASD, as well as in other cardiac diseases, the coaptation between the anterior and posterior leaflets seems to be related to the regurgitation. While perhaps preventing the edges of the cleft from prolapsing, as mentioned above, the accessory chordae restrain the anterior leaflet from being straight during systole. This condition may interfere with the proper coaptation between the anterior and posterior leaflets. If the mitral regurgitation occurred only from the site of the cleft, it would be corrected merely by suture of the cleft. However, mitral regurgitation remains in some patients even after surgery. In this study, the patients with systolic bending of the anterior leaflet after surgery were shown to have mitral regurgitation, and all but one patient without systolic bending had no mitral regurgitation, even if the accessory chordae were not resected. In addition, as in other cardiac diseases, mitral valve prolapse may be the cause of the mitral regurgitation, as in patient 4.

### Conclusion

This study showed that real-time cross-sectional echocardiography demonstrates not only the presence of the mitral cleft, but also the configuration of the anterior mitral leaflet and the presence of the accessory chordae. This information has not been shown in the beating heart. Not only the cleft but all these conditions may relate to the mitral regurgitation in this malformation. Although we did not study a large number of patients, one explanation of the cause of mitral regurgitation was proposed from the echocardiographic viewpoint. Real-time cross-sectional echocardiography should become indispensable in assessing mitral deformities before surgery.

### References

Dicrotic Pulse After Open Heart Surgery

ROBERT CHARLES ORCHARD, M.D., AND ERNEST CRAIGE, M.D.

SUMMARY Pre- and postoperative echophonocardiograms (EPCGs) and preoperative hemodynamic data of 108 patients who underwent valve replacement were reviewed to establish the frequency and significance of a dicrotic pulse (DP) postoperatively. DP occurred almost exclusively in patients who underwent valve replacement for regurgitant lesions (20 of 28 with aortic regurgitation, nine of 25 with mitral regurgitation, and four of six with both aortic and mitral regurgitation). These patients were divided into dicrotic and nondicrotic groups. Preoperatively, the dicrotic group had significantly larger end-diastolic volumes ($p < 0.01$) and end-systolic volumes ($p < 0.01$) and significantly lower ejection fractions ($p < 0.01$). Echocardiographically, the dicrotic group had larger left ventricular dimensions, both systolic ($p < 0.01$) and diastolic ($p < 0.05$), reduced percentage fractional shortening of the left ventricular cavity ($p < 0.01$) and poor thickening properties of the left ventricular posterior wall ($%\Delta Th-LVPW$) ($p < 0.01$). Postoperatively the dicrotic group had a slightly larger end-diastolic dimension ($p = NS$) and markedly depressed $%\Delta Th-LVPW$ ($p < 0.001$) compared with the nondicrotic group.

On follow-up EPCG the persistence of a DP correlated with continued left ventricular dysfunction by echocardiographic and hemodynamic studies and an extremely poor clinical course. DP after valve replacement is therefore an important prognostic sign.

A DICROTIC PULSE is characterized by two palpable pulsations (fig. 1). As opposed to anacrotic and bisferiens pulses, in which the two pulsations are systolic in timing, the second pulsation of the dicrotic pulse is diastolic, immediately after the dicrotic notch. The dicrotic pulse has long been recognized, particularly as a feature of febrile states such as typhoid fever. More recently the dicrotic pulse has been associated with a variety of cardiac diseases that have in common a low cardiac output. These include cardiomyopathies, pericardial tamponade, constrictive pericarditis and pulmonary embolism. There is only one report, however, of dicrotism after open heart surgery, and the significance of this finding is unknown. At our institution, pre- and postoperative echophonocardiograms are routinely performed on most adult patients who undergo cardiac surgery, so we had the opportunity to observe the frequent, although not uniform, presence of a dicrotic pulse after open heart surgery. The present study was undertaken to establish the frequency and significance of this clinical sign in patients who undergo cardiac surgery for valvular disease.

Methods

Subjects

The patients were identified by a computerized data retrieval system as having undergone pre- and postoperative echophonocardiograms for valvular surgery between January 1973 and December 1978. The patients studied were all age 45 years or younger, and any patient who developed prosthetic malfunction was excluded. After technically inadequate carotid pulse tracings were excluded, 108 patients remained. Postoperative tracings were performed 7-10 days after surgery.

Echophonocardiography

Phonocardiograms were obtained from the four standard precordial areas using either an Irex 101 or a Cambridge MC IV multichannel recorder and Leatham suction microphones. Indirect carotid artery pulse recordings were made with an air-filled funnel.
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Circulation. 1980;62:1099-1107
doi: 10.1161/01.CIR.62.5.1099

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