Color Doppler Echocardiography of Isolated Cleft Mitral Valve*

Roles of the Cleft and the Accessory Chordae

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To study the mechanism of altered mitral function in the presence of an isolated cleft mitral valve (ICMV) with regard to the relative roles of the cleft and of the accessory chordae, seven patients with ICMV were studied with color Doppler echocardiography. Mitral insufficiency ranging from mild to severe was demonstrated in six cases. The regurgitant jet originated in each case from the site of the cleft; in five patients the regurgitant jet had a narrow base originating exactly from the cleft; in the sixth patient, the regurgitant flow presented as a broad-based jet suggesting that accessory chordae restricted the motion of the anterior mitral leaflet. Turbulent flow in the left ventricular outflow tract, starting at the level of the accessory chordae, was found in one patient in whom a pressure drop of 44 mm Hg was detected with continuous-wave Doppler imaging. The altered function of the mitral valve cleft stems from two elements, the cleft itself and the accessory chordae. Color Doppler flow imaging showed that the cleft was the main factor causing mitral insufficiency. The accessory chordae played an additional pathogenetic role in two patients by causing restricted mitral motion or left ventricular outflow tract obstruction. (Chest 1992; 101:12-15)

ICMV = isolated cleft mitral valve

Cleft mitral valve not associated with a septal defect of the endocardial cushion defect type (atrioventricular septal defect), namely isolated cleft mitral valve (ICMV), is a rare cause of congenital mitral insufficiency. Previous reports of ICMV have shown the usefulness of two-dimensional echocardiography in demonstrating the cleft as well as additional anatomic features, such as accessory chordae and thickening of the edges of the cleft with increasing age. The introduction of color Doppler echocardiography provides a sensitive tool for studying the qualitative and quantitative aspects of mitral insufficiency. We used color Doppler echocardiography to study a group of patients with ICMV. The aim of this article is to report our findings with particular reference to the relative roles of the cleft and of the accessory chordae in the pathogenesis of mitral insufficiency and of left ventricular outflow tract obstruction.

MATERIALS AND METHODS

Five male and two female patients aged 1 to 45 years, were studied. In each case ICMV was diagnosed by visualizing with two-dimensional echocardiography the cleft dividing in diastole the anterior mitral leaflet in two portions and by excluding associated septal defect of the endocardial cushion defect type (ostium primum atrial septal defect and inflow ventricular septal defect). Four patients did not present any additional cardiac anomalies, two had a supraventricular or perimembranous ventricular septal defect, and one had d-transposition of the great arteries. Four patients belonged to families in which several members were affected by various forms of endocardial cushion defect including ICMV. Two of these four patients were the index cases, and two were diagnosed following echocardiographic screening of other family members. The pedigrees of three of these families have been reported previously.

Echocardiograms were performed with an Aloka (Tokyo) SSD 870 phased-array echocardiographic system or a Hewlett-Packard (Andover, Mass) Ultrasound Imaging System (model 77020AC) using 5-, 3.5-, and 2.5-MHz transducers. A complete two-dimensional, M-mode, pulsed and continuous Doppler examination was performed from the parasternal and apical windows in each patient; subcostal views were also recorded in five children but not in the adult patients. Parasternal long- and short-axis and apical views were obtained with color Doppler flow imaging in each patient; subcostal views were also obtained in the children. In these views the regurgitant jet of mitral insufficiency appeared bluish with a central core showing a mosaic of blue and yellow.

In order to obtain an optimal image of the entire regurgitant flow, the gain of the color Doppler signal was adjusted at the maximal level just before the appearance of the noise artifacts. In each view the presence, the point of origin, the width of the base, the extension, and the direction of a regurgitant jet were recorded. The degree of mitral regurgitation was estimated by using the jet area left atrial area ratio according to Helmcke et al. With this method, a ratio of <0.2 denotes mild regurgitation, a ratio of 0.2 to 0.4 moderate regurgitation, and a ratio of >0.40 severe regurgitation. The left ventricular outflow tract was accurately examined to detect the presence and the exact starting point of any turbulent flow, which appeared as a mosaic of yellow and blue with a red background in the parasternal views and as a bluish background in the apical and subcostal views.

RESULTS

Clinical Aspects

Three patients with other anomalies (not of the endocardial cushion defect type) presented with the clinical and auscultatory features of the basic anomaly (complete transposition of the great arteries and ventricular septal defect). Two patients (the index cases of those families in which several members were
affected) had an apical 3/6 pansystolic murmur. The two remaining patients, who were identified during the family study, did not have evidence of mitral insufficiency by auscultation.

Two-dimensional Echocardiographic Aspects

In three patients the cleft was located in the middle of the anterior mitral leaflet. In three patients the lateral portion of the anterior leaflet was wider than the medial portion, while in one patient, who had transposition of the great arteries, the medial portion was wider. Mild systolic billowing of both halves of the anterior mitral leaflet was present in six patients (Fig 1), but not in a patient with severe mitral regurgitation and a broad-based regurgitant jet in which the motion of the medial portion of the anterior mitral leaflet was restricted (Fig 2). Accessory chordae were detected in each case. In one patient redundant accessory chordae appeared to obstruct the left ventricular outflow tract (Fig 3). Short-axis views in this

![Figure 1](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21638/)

**Figure 1.** Systolic frame of color Doppler flow imaging (left) and corresponding two-dimensional echocardiogram (right) in the short-axis parasternal view at the level of the left ventricular outflow tract (LVOT) in a patient with ICMV. The anterior mitral leaflet is shown at its full extension between the commissures. From the site of the cleft (arrow), accessory chordae (arrowhead) extend to the interventricular septum. The regurgitant jet of mitral insufficiency has a narrow base originating from the cleft. LA = left atrium.

![Figure 2](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21638/)

**Figure 2.** Systolic frame of color Doppler flow imaging (left) and corresponding two-dimensional echocardiogram (right) in the short-axis parasternal view in a patient with ICMV (open arrow). The broad-based regurgitant jet stems from the entire area between the medial commissure and the cleft. The medial portion of the anterior mitral leaflet is straight, while the lateral portion shows normal systolic billowing. In real-time imaging the body of the medial portion of the anterior leaflet appeared connected to accessory chordae, and its systolic motion was clearly restricted. The lateral portion moved normally and did not go beyond the plane of the mitral annulus, thus excluding prolapse. LA = left atrium; LVOT = left ventricular outflow tract.
A patient clearly showed that the obstructive tissue was made of several isolated chordae, not a membrane.

**Color Doppler Flow Imaging**

Mitral insufficiency was demonstrated with color Doppler flow imaging in six of the seven cases (Fig 1 and 2). Mitral insufficiency was mild in two patients, moderate in two, and severe in two. The regurgitant jet originated in each case from the area of the cleft; this was particularly well demonstrated in the parasternal short-axis view. The regurgitant jet was directed inferiorty and toward the center of the left atrium in five cases and inferolaterally in the sixth case.

The regurgitant jet had a narrow base, originating exactly from the cleft in five cases (Fig 1); in one case the regurgitant flow was depicted as a broad-based jet stemming from the medial half of the anterior mitral leaflet between the cleft and the commissure (Fig 2).

Unrestricted diastolic flow through the mitral valve was found in each case. This flow was characterized by laminar, red colored flow that entered the left ventricle through the entire mitral valve opening. In the parasternal short-axis view the diastolic flow was seen entering the left ventricular outflow tract through the open halves of the anterior mitral leaflet.

Turbulent flow starting in the area of the accessory chordae, giving a mosaic appearance to the left ventricular outflow tract on color Doppler flow images, was found in one patient (Fig 3). With continuous-wave Doppler, a pressure drop of 44 mm Hg was measured. This was the only patient in whom evidence of left ventricular outflow obstruction was found.

**Discussion**

Color Doppler echocardiography proved to be an especially useful technique in correlating altered function to anatomic abnormalities. A few publications have reported color Doppler flow images of cleft mitral valve associated with an endocardial cushion defect, but no color Doppler echocardiographic study of ICMV has been reported. This condition is clinically important, although rare, since it may cause significant functional impairment. Two-dimensional echocardiography is superior to cardiac angiography in diagnosing ICMV and it may be expected that the addition of color Doppler flow imaging may further improve the diagnostic accuracy of the noninvasive evaluation.

The altered function of the cleft mitral valve arises from two elements, the cleft itself and the accessory chordae. The cleft is the primary factor causing mitral insufficiency, and the accessory chordae may occasionally cause left ventricular outflow tract obstruction. The mechanism of mitral regurgitation in mitral cleft is not always as simple as might be intuitively thought. It is usually true that incomplete coaptation of the two halves of the cleft anterior leaflet causes mitral insufficiency, especially when a wide cleft is demonstrated. Pathologic studies in the past suggest, however, that restriction of the motion of the anterior mitral leaflet caused by accessory chordae may be an additional mechanism. The importance of this latter mechanism in limiting the success of mitral valve repair once the cleft has been sutured has long been recognized, but it may also play a role in the pathogenesis of mitral insufficiency of the native valve as well. In the
preoperative evaluation of these patients, it is potentially important to establish whether leaking through the cleft is the only mechanism involved or whether restricted leaflet motion due to accessory chordae is also present. All these points are clarifiable by combining color Doppler flow imaging with conventional two-dimensional echocardiography, thus showing the site of origin and the extension of the regurgitant jet as well as the presence and the exact location of the left ventricular outflow tract turbulence.

The short-axis parasternal view was the most useful for investigating the mechanism of mitral insufficiency. In this view the total surface area of the mitral valve is shown, with the anterior and posterior leaflets fully displayed between the valve commissures. The origin of the regurgitant jet could therefore be accurately located in all six cases in this view and could be identified with the cleft itself, confirming that the absence of perfect leaflet coaptation in this area was the cause of the regurgitation. The lack of any regurgitant flow through the remaining "normal" commissures does not support the possible role of accessory chordae in producing mitral insufficiency in the majority of our patients. If the accessory chordae had restricted the motion of the mitral leaflets, they would have produced incomplete coaptation along the main commissures as well as along the cleft. In the single case in whom the regurgitant flow spurted through the main commissures, the systolic motion of a portion of the mitral valve connected to accessory chordae was restricted, suggesting that these chordae may indeed have added to the valvular regurgitation (Fig 2). In another patient, color Doppler flow imaging clearly showed left ventricular outflow tract obstruction starting at the site of the accessory chordae. Interestingly, in this patient the redundant accessory chordae did not restrict mitral valve motion.

Our observations with color Doppler echocardiography in a series of patients with ICMV confirm the concept that mitral regurgitation is primarily due to leakage through the cleft. Accessory chordae may be an additional pathogenetic factor causing left ventricular outflow tract stenosis and contributing to the mechanism of mitral regurgitation. Although these conclusions may be limited by the fact that the number of patients studied was small, due to the rarity of this anomaly, and by the unavailability of anatomic confirmation, we strongly feel that the clearness of the echocardiographic images (mainly in real-time recording) nicely demonstrated our interpretation. Color Doppler flow imaging may thus add important information to that achieved with conventional two-dimensional echocardiography, contributing to a better understanding of the altered function in the presence of an ICMV. The findings of color Doppler flow imaging may be taken into consideration in planning the surgical repair of mitral cleft (eg, whether the repair should be limited to suturing the cleft or should also include resection of accessory chordae that may be obstructing the flow or preventing adequate mitral valve closure).

REFERENCES
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