Two Dimensional Echocardiographic Profile of Left Ventricular Outflow Tract in Transposition of the Great Arteries

by

K.U. Aziz*, Milton H. Paul** and A.J. Muster***

Angiographic (1,2,3) and M-mode echocardiographic (4, 5) profiles of the left ventricular outflow tract in patients with transposition of the great arteries have recently been presented. These studies indicate that angiographic and echocardiographic imaging can distinguish between the anatomically fixed and dynamically functional types of left ventricular outflow tract stenosis so that appropriate selection of surgical procedures can be undertaken.

The dynamic form of left ventricular outflow tract stenosis is caused by posterior systolic septal bulging and concomitant systolic anterior motion of the mitral valve (5). The pressure gradient across the left ventricular outflow tract is generally mild to moderate, nevertheless it has been implicated in the development of symptoms such as cyanotic spells, due to inadequate interatrial mixing, following an anatomically adequate baloon atrial septostomy in the newborn period (6,7). The dynamic form of left ventricular outflow tract stenosis is apparantly not amenable to surgical resection since no true septal hypertrophy is present and the obstruction is phasic, i.e. systolic only.

The anatomically fixed types of stenosis are variously deseribed as consisting of either a fibrous ridge, at times also associated with features of the dynamic types of left ventricular outflow tract stenosis(6), a more extensive fibromuscular collar, or rarely a long muscular tunnel(8). A type of subpulmonary stenosis due to malalignment of the infundubular septum has been described in patients with an associated ventricular septal defect (8). Left venticular outflow tract obstruction due to atrioventricular valve leaflet tissue is less often appreciated (8, 9, 10), however it is important to recognize these more complex types of left ventricular outflow tract stenosis so that appropriate surgical procedure can be planned.

The purpose of the present study is to present correlative angiographic and two dimensional echocardiographic profiles of the types of sub pulmonary left ventricular outflow tract obstruction observed in patients with transposition of the great arteries.

MATERIALS AND METHODS

Fifty Five patients with transposition of the great arteries ranging in age from 3 months to 14 years were studied (Table-1)

All had cardiac catheterization, cineangiography and two dimensional echocardiographic studies. Hemodynamically significant sub

* Pediatric Cardiologist, NICVD, Karachi.

** Children's, Memorial Hospital, Chicago, Ill, U.S.A.

Table I: Clinical profile of Fifty patients with Transposition of the Great Arteries studied by 2-D Echocardiography.

	Pre M.O.	*Post M.O.*	*Sequential*	*Total*
	N	*N*	*N*	*N*
Group I. (IVS)	0	12	6	18
Group II (VSD)	3	7	4	14
Group III (LVOTS)	8	9	6	23
	11	28	16	55

Abbreviations.

- IVS = Intact Ventricular septum.
- LVOTS = Let Ventricular outflow tract Stenosis
- MO = Mustard operation
- N = Number of PatMents.
- VSD = Ventricular Septal defect.

pulmonary stenosis was diagnosed in patients having > 20 mm Hg pressure difference across the left ventricular out flow tract. Eighteen patients had intact ventricular septum (Group-I), 14 had ventricular septal defect (Group-II), and 23 had left ventricular outflow tract stenosis with or without a ventricular septal defect (Group III). Eleven patients were studied before, 28 patients after, and 16 sequentially before and after the Mustard operation (Table 1). Two dimensional sector scans were obtained without seuation with patient in supine position. Various transducer positions were employed using an Advanced Technology Laboratories sector scanner with a 3.5 mHZ transducer. (11,12,13). The images were videotaped and stop-frame 35 mm photographs were obtained for analysis.

RESULTS

Two dimensional echocardiographic and angiographic features of dynamic type of left ventricular outflow tract stenosis were noted in 8/18 patients with intact ventricular septum (Group-1). The M-mode echocardiogram of these patients showed all the features of dynamic stenosis which was characterized by systolic anterior motion of the mitral valve and posteriorly bulging ventricular septum in early systole. The pressure difference across the left ventricular outflow tract ranged between 4 to 16 mm Hg (mean 8.4 ± 5.5) (Table II).

Table II: Hemodynamic Data of Patients with Left Ventricular Outflow Tract Obstruction

Type	*N*	*ΛP (LV—MPA) Range mm Hg*
Dynamic (ΛP < 20 mm Hg)	7	4—16
Dynamic (ΛP < 20 mm Hg)	6	25—16
Anatomic Lvots.		
Fibromuscular Ridge	4	21, 32, 40 120
Muscular Band/Collar	7	59—92
Anatomic AV Valve Tissue		
Tricuspid	4	30, 52, 94, 98
Mitral	1	85, 98

Abbreviations.

- LV = Let Ventricle
- LVOTS = Left Ventricular outflow tract stenosis
- MPA = Main pulmonary artery
- ΛP = Pressure difference across the left Ventricular outflow tract
- N = Number of Patients.

Eleven of the 14 patients with ventricular septal defect (Group II) had a Mustard operation with closure of the ventricular septal defect. Two of these patients postoperatively showed angiographic and echocardiographic features of dynamic stenosis with peak systolic left ventricular outflow tract gradient of 28 and 32 mm Hg and two-dimensional echo showed wide open left ventricular outflow tract.

In all, twenty three of the total 55 patients studied, had significant hemdynamic (> 20 mm Hg) left ventricular outflow stenosis (Group III). The dynamic type of left ventricular outflow stenosis (Fig. 1) was noted in 6 patients with pressure differences across the left ventricular outflow tract ranging from 25 to 50 mmHg (31 ± 12).

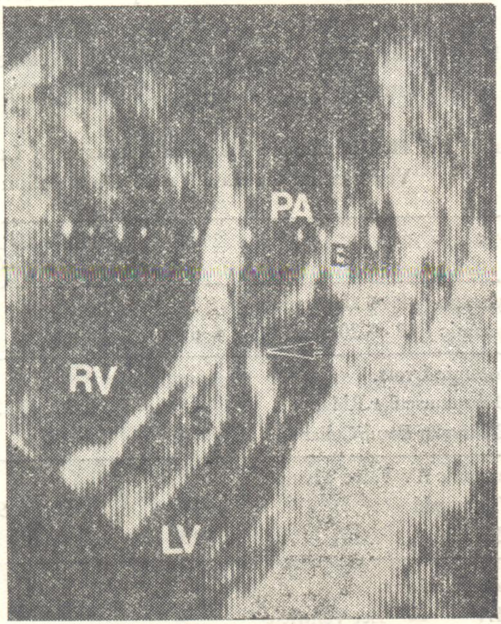

Fig. 1. LV long-axis view sector scan obtained from the left parasternal location in a patient with transposition of the great vessels and intact ventricular septum following Mustard operation. Right ventricle (RV) is enlarged and moderate posteriorly bulging interventricular septum is seen in systole. Mitral valve showed marked systolic anterior motion (arrow) toward the ventricular septum producing dynamic narrowing of the outflow tract. A 20 mm Hg pressure differance was noted between left ventricular and pulmonary artery.

Abbreviations: B=intraatrial baffle; LV=left ventricle; PA=pulmonary artery

Seventeen patients had angiographic and echocardiographic fectures of an anatomically fixed type of left ventricular outflow stenosis In 4/17 patients an obstructive fibrous or fibromuscular ridge was demonstrated in the left ventricular outflow tract with the pressure gradient ranging from 21—120mm Hg. (Table II). Two dimensional echocardiogram, on long axis view, showed thick linear echo band at the septal and/or mitral valve region extending variably into the left ventricular outflow tract (Fig. 2, 3). In 7/17 patients the thickened echoes beneath the base of the pulmonary valve annulus corresponded well with the angiographic appearances of a fibromuscular ridge or collar (Table II). In one patient the features of both dynamic left ventricular outflow obstruction and anatomic fibrous ridge abstruction could be well seen on the long axis sector view and the M-mode echocardiogram confirmed the features of the dynamic form of left ventricular outflow stenosis. On angiography Muscular Subpulmonary left ventricular outflow tract stenosis was characterised by a cushion of negative contrast beneath the pulmonary valve. Two-dimensional echocardiograms showed thick cushion of echoes guarding the exit of the ventricular outflow tract. (Fig. 4).

The left ventricular outflow tract stenosis due to portions of atrioventricular valves was noted in 6/17 patients, tricuspid valve in 4 and mitral valve in 2. On 2-dimensional sector scan imaging the septal leaflet of the tricuspid valve was seen, on the long axis and subxiphoid views, to prolapse as a large pouch into the left ventricular outflow tract during systole and retract back into the right ventricle during diastole (Fig. 5,6). The systolic pressure

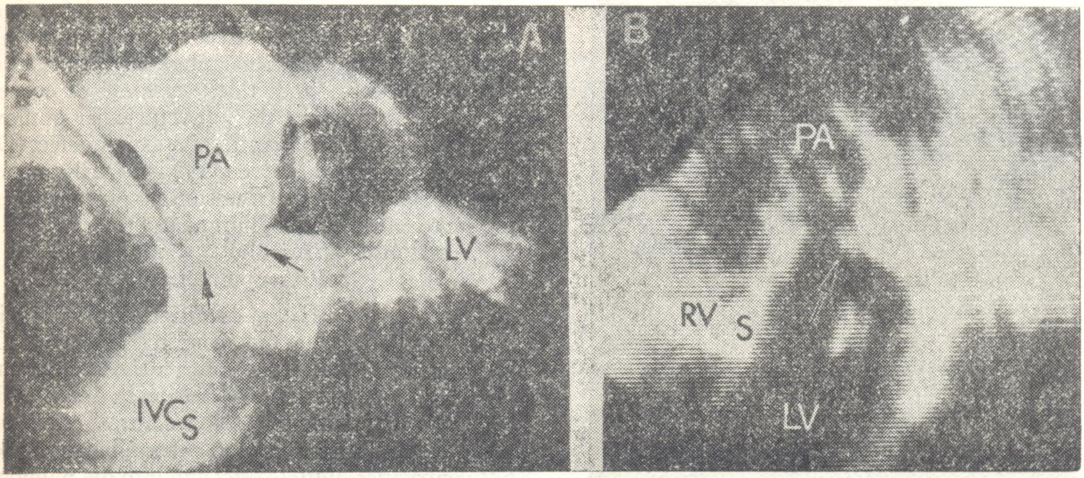

Fig. 2. A, Anteroposterior systemic venous angiogram and *B*, left ventricular long axis view sector scan echocardiogram obtained from parasternal location in a patient with transposition and intact ventricular septum. Moderate ridge formation is seen in the outflow tract (arrow, A and B) below the pulmonary artery (PA) producing a 44 mm Hg pressure difference between left ventricle and pulmonary artery.

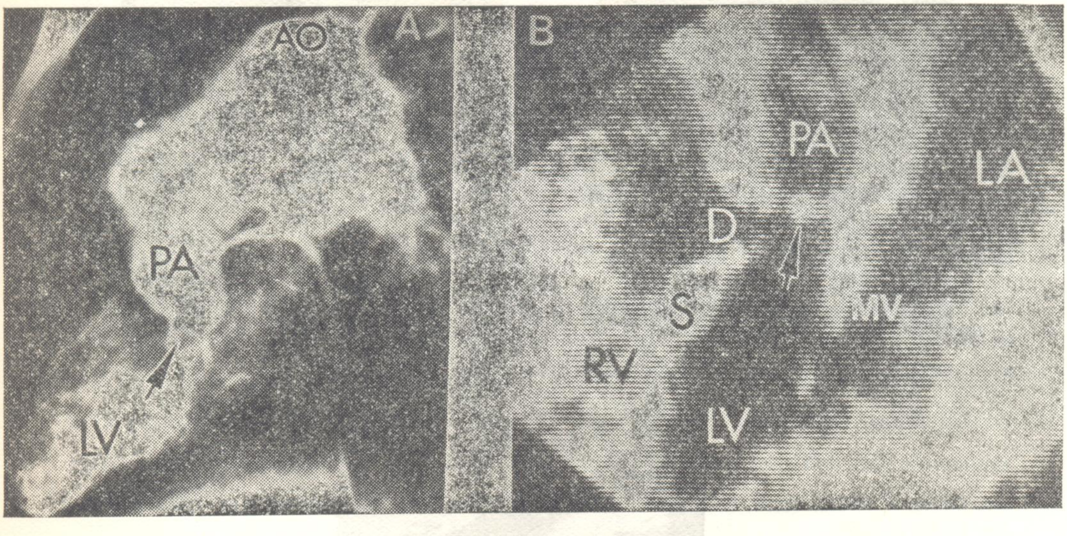

Fig. 3. A, Lateral view left ventricular angiogram in a patient with transposition of great arteries and ventricular septal defect. *B*, LV long axis view sector scan echocardiogram obtained form parasternal location. Note thickened echoes from a fibrous ridge displayed transversely across the left ventricular outflow tract (arrow) above the ventricular septal defect(D). Thickened echoes show muscular component of the stenosis on both septal and mitral aspects. A 47 mmHg pressure difference was noted across the left ventricular outflow tract.

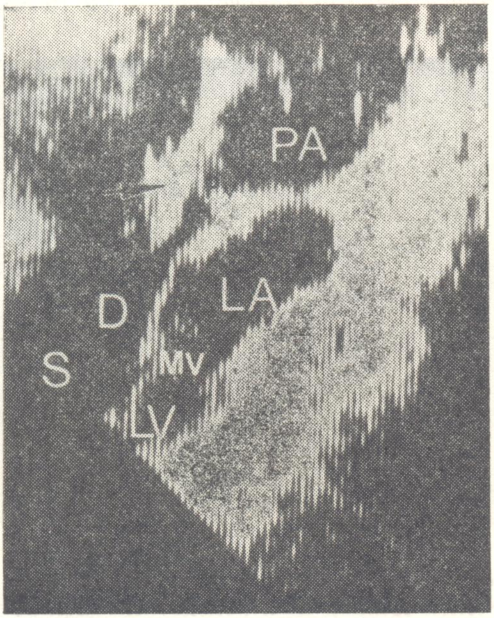

Fig. 4. LV long axis view obtained from parasternal location in a patient with transposition of the great arteries, ventricular septal defect and left ventricular outflow tract stenosis following Waterston shunt. Note markedly thickened pulmonary valve and thick cushion of echoes →markedly obstructing the left ventricular outflow tract (arrow). Large ventricular septal defect (D) is present and mitral valve (MV) is partially open. The pressure difference was 65 mm Hg between the left ventricle and Pulmonary artery.
Abbreviations: LA = left atrium;
PA = pulmonary artery.

Fig. 5. A Left ventricular cineangiogram in a patient with transposition, (anteroposterior view). *B.C*, LV long axis view sector scan echocardiograms obtained from subxiphoid location in diastole (B) and systole (C). Note negative contrast of tricuspid valve pouch (TV arrow) protruding into the left ventricular outflow tract and into the pulmonary artery (A). A tongue like projection of tricuspid valve (open arrows) prolapsing tissue is imaged on the sector scan showing motion toward the pulmonary artery during systole (C).
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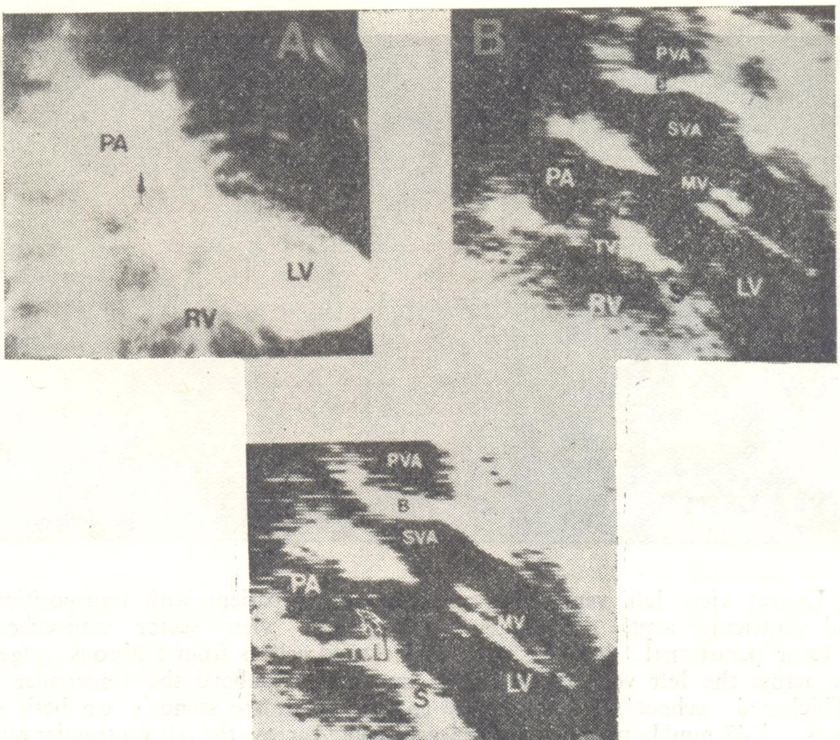

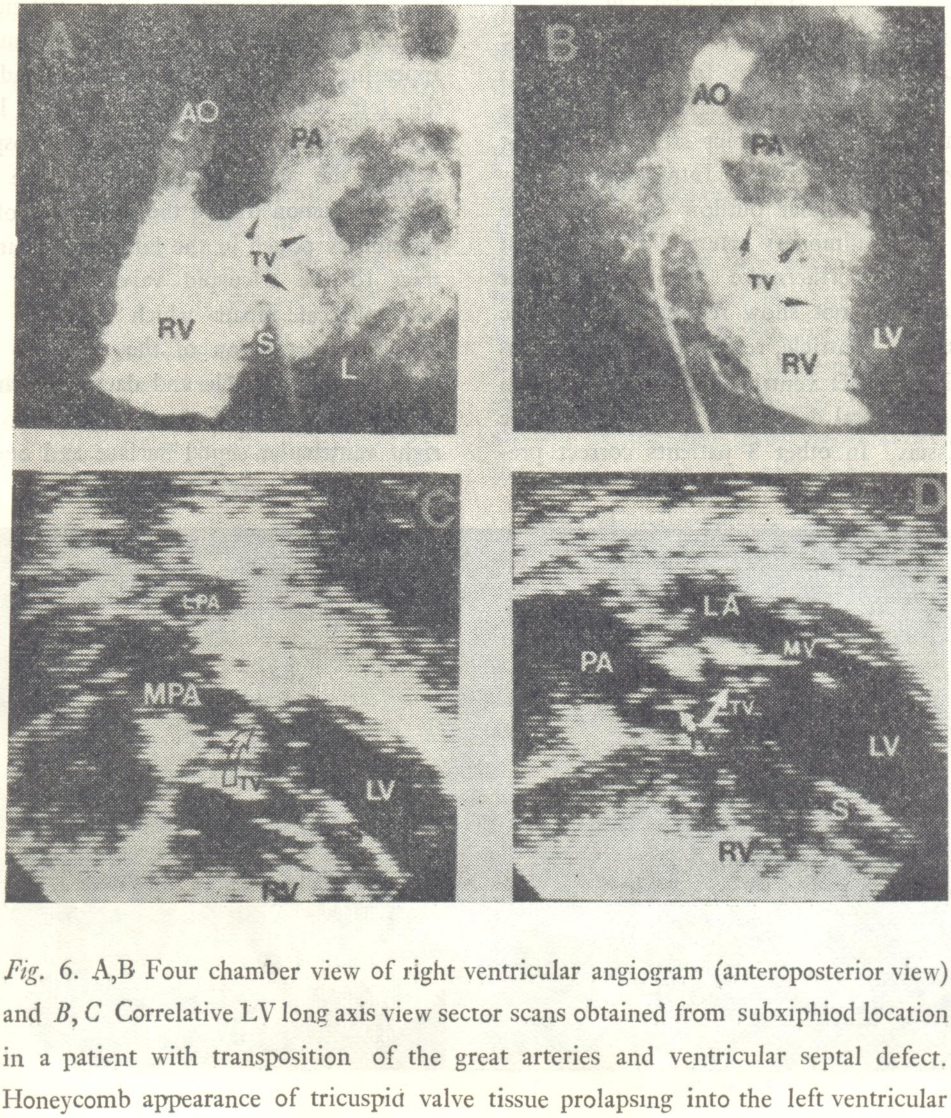

Fig. 6. A,B Four chamber view of right ventricular angiogram (anteroposterior view) and *B, C* Correlative LV long axis view sector scans obtained from subxiphiod location in a patient with transposition of the great arteries and ventricular septal defect. Honeycomb appearance of tricuspid valve tissue prolapsing into the left ventricular outflow tract can be seen on the angiogram (arrows). Sector scan shows a thickened echo of tricuspid valve imaged in the left ventricular outflow extending from the right ventricle across the ventricular septal defect (C.D). During systole branching pattern of the tricuspid valve echoes are seen obstructing the left ventricular outflow tract(D).

difference across the left ventricular outflow tract ranged from 30 to 98 mm Hg and the left ventricular pressure was above systemic pressure in two of these four patients. In one of these four patients the nature of the left ventricular outflow tract stenosis was not recognized pre-operatively. Intraoperative view of the left ventricular outflow tract from the opened main pulmonary artery did not suggest obstruction. Furthermore right ventricular angiograms did not show regurgitation of dye into the left ventricle, resulting in an erroneos diagnosis of intact ventricular septum, when in fact a large septal defect was obstructed by the valve tissue. In other 3 patients correct pre-operative diagnosis was established and right ventricular angiograms in these patients were particularly helpful in defining the honeycomb-appearing tricuspid valve pouch extending into the left ventricular outflow tract. In 3 of these 4 patients echocardiography prospectively defined the antomic diagnosis of the obstruction by demonstrating the continuity of the sub pulmonary pouch in the left ventricular outflow tract to the tricuspid valve. Three patients had surgical repair which included Mustard operation, retraction of the tricuspid valve in to the right ventricle and dacron patch closure of the ventricular septal defect, two on the right ventricular septal surface and one on the

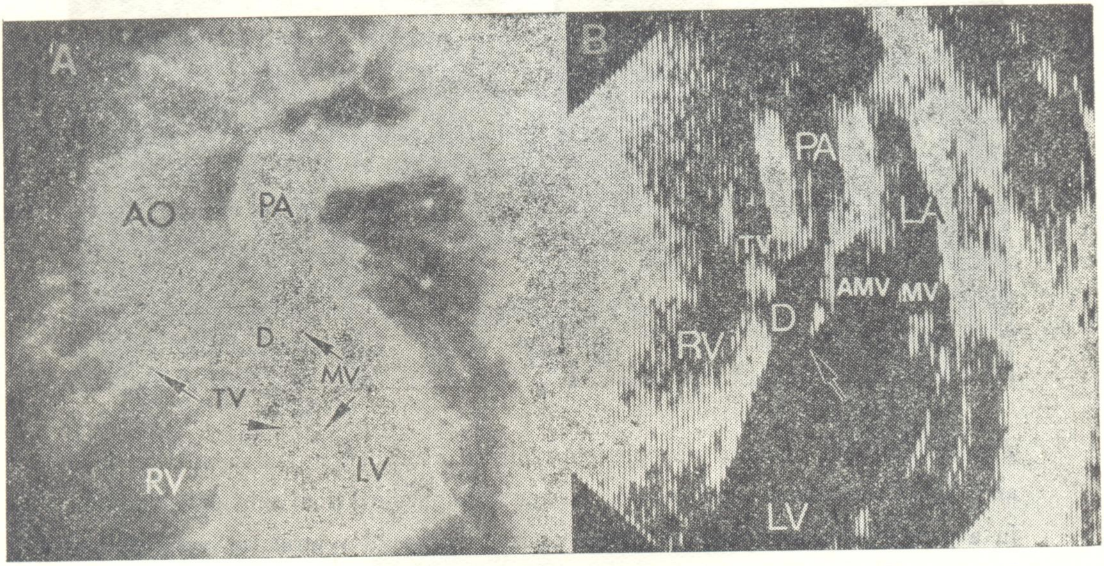

Fig. 7. A, left ventricular angiogram, left anterior oblique view, and *B*, Correlative long axis view sector scan obtained from the left parasternal location. A large ventricular septal defect (D) extends from the base of the pulmonary artery (PA) to the crest of the ventricular septum. Overriding of the pulmonary artery is seen above the VSD. Tricuspid valve partially covers the VSD from the right ventricular side. Mitral valve is seen is closed position (MV). Accessory mitral valve tissue (AMV) has malattachments in the left ventricular outflow tract below the pulmonary artery and to the left ventricular surface of the septum across the Ventricular septal defect (arrows).

left ventricular septal surface. In this later patient, post operatively, the patch continued to produce left ventricular outflow stenosis and a hemolytic anemia appeared which was severe and uncontrolled. Subsequently a left ventricle to pulmonary artery conduit was successfully placed to bypass the left ventricular outflow stenosis. One surgical death in these four patients was attributed to a hypoplastic left ventricle and a form of cardiomyopathy.

In 2 patients portions of the mitral valve were malattached into the left ventricular outflow tract and across the ventricular septal defect onto the left side of the ventricular septum. The pressure difference across the left ventricular outflow tract were 85 and 98 mm Hg. The long-axis sector scan view clearly demonstrated malattachments of the accessory or redundant mitral valve tissue to the left ventricular outflow tract and ventricular septum (Fig. 7). The normally located mitral valve was seen at the atrioventricular annulus. One of these patients underwent successful surgical repairs with Mustard operation and placement of the left ventricle to pulmonary artery conduit to bypass the left ventricular outflow stenosis (10), the second patient remains unoperated.

DISCUSSION

Our experience indicates that two dimensional sector scan echocardiography is a powerful tool for visualising the left ventricular outflow tract from various planer projections. The nature of left ventricular outflow stenosis can be elucidated and its effect assessed throughout the cardiac cycle. We have previously reported the usefulness of quantitative M-mode echocardiography for distinguishing between the dynamic and anatomic forms of left ventricular outflow tract stenosis and for the features of the dynamic type i.e. posterior systolic septal bulging, anterior systolic motion of the mitral valve, and systolic motion abnormalities of the pulmonary valve which are more easily appreciated and quantitatively assessed using M-mode echocardiography (5). Sector scan echocardiography can be used to evaluate septal bulging, but careful stop frame analysis is required to appreciate the motion abnormality of the mitral valve. The pulmonary valve can be well seen in the sector scan images, however, early systolic closure and coarse systolic flutter of the valve cannot be well appreciated.

The major diagnostic advantage of sector scan imaging is the elucidation of the nature of the anatomically fixed types of stenosis and the obstruction due atrioventricular valve leaflet tissue. Fibrous ridges can be clearly seen traversing the left ventricular outflow tract and the extent of these ridges can be assessed by observing whether the attachments are restricted only to the septum (partial circumferential), or additionally involving the anterior mitral valve (complete circumferential). The fibromuscular collar type of obstruction below the pulmonary valve appears as broader cushions of thick Echoes. The degree of stenosis can be evaluated by observing the orifice size or passage between these cushions. The position of the ventricular septal defect and the its relationship to obstructing structures can also be clearly visualized. Since the pulmonary valve motion can be seen during systole and diastole so that infrequent occurence of significant pulmonic valve stenosis in patients with transposition of the great vessels can also be diagnosed. We have previously reported patients having fixed fibrous ridge pathology with additional feature of dynamic obstruction by M-mode echocardiography and we have observed this combined type again with two-dimenional echocardiography imaging(5). It seems likely that there may be a cause-effect relationship. The early systolic posterior bulging of the ventricular septum results in abnormally increased blood flow velocity through tne left ventricular outflow tract and may result in jet trauma to the ventricular septum. This "jet" trauma coupled with 'Kissing' lesion caused by the diastole opening of the mitral valve, may initiate the grafting of fibrous ridge at the Septum.

Left ventricular outflow tract obstruction due to aneurysm of the membranous septum has been reported by Vidne et al. The report does not clarify whether the aneurysm was exclusively formed by the membranous septum or by the tricuspid valve tissue (14). In patients with transposition of the great arteries the left ventricular outflow tract stenosis caused by atrioventricular valve tissue is uncommon, (8, 9, 10). We have observed 6 cases with this pathology in a series of 100 patients with complete transposition of the great arteries studied with both angiography and echocardiography in our laboratories.

On right ventricular angiogram the tricuspid valve form of obstruction reveals a "pouch" with a honeycomb appearance, which extends through the ventricular septal defect into the left ventricular outflow tract. Sector scan echo images in each of our patients clearly showed the continuity of these protrusions to the septal leaflet of the tricuspid valve. It is important to preoperatively recognize these cases on echocardiographic examination since the tricuspid valve in some cases may effectively close the ventricular septal defect during systole resulting in the ventricular septal defect not being visualized on angiography.

Surgical experience remains limited in this malformation, however, it has been feasible to close the ventricular septal defect after retraction of the obstructing tricuspid valve tissue into the right ventricle. If the nature of obstruction is not recognized, surgical resection of this tissue can be disastrous. Mitral valve mallatachments into the left ventricular outflow tract and to the interventricular septum can result in severe obstruction of the left ventricular outflow tract. The overall cardiac pathology here has been described as representing variant form of double outlet right ventricle (Taussig-Bing anomaly) with straddling mitral valve(10). Resection of the obstructing valve tissue is not usually feasible without introducing gross mitral dysfunction, therefore a conduit bypass operation such as left ventricle to pulmonary artery conduiting has been successfully employed(10). Sector Scan echocardiography in our experience has provided reliable diagnosis in these cases.

It is concluded that 2-dimensional sector scan echocardiography is a useful tool in elucidating various forms of left ventricular outflow tract stenosis in patients with transposition of the great arteries.

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