Mitral stenosis with left-to-right shunt at atrial level

A diagnostic challenge

Rajendra Tandon, Subhash C. Manchanda, and Sujoy B. Roy

From the Department of Cardiology, All India Institute of Medical Sciences
New Delhi-16, India

The difficulties in the recognition of mitral stenosis associated with atrial septal defect have been emphasized. It should be suspected if the patient has a combination of an unusually large heart for an uncomplicated secundum atrial defect, a systolic thrill at the upper left sternal border, and a long and loud delayed diastolic murmur with right and left atrial hypertrophy in the electrocardiogram, even in the absence of left atrial enlargement on the skiagram.

One case of mitral stenosis with partial anomalous pulmonary venous drainage and intact atrial septum has been presented, the published reports reviewed, and the differences from Lutembacher's syndrome highlighted. The diagnosis of additional left-to-right shunt should be thought of in a patient with findings typical of mitral stenosis, if there is an ejection systolic murmur at the second left interspace with or without a thrill and if the chest x-ray shows increased vascularity, suggestive of left-to-right shunt, localized to one side or to an area of one lung. The diagnosis mainly depends on a high index of suspicion combined with cardiac catheterization.

The association of atrial septal defect with mitral stenosis carries the eponym of Lutembacher's syndrome (1916), though according to Soulié, Bouvrain, and Sibille (1954), it had been described a century earlier by Corvisart. In the past 5 years, we have come across 6 proven cases of this combination and 1 of mitral stenosis with partial anomalous pulmonary venous drainage. The differences in the presentation of the two varieties of cases, with open or intact atrial septum, have prompted this report. The purpose of this communication is to highlight the difficulties in the diagnosis of (i) mitral stenosis in the presence of an atrial septal defect, and (ii) left-to-right shunt in the presence of mitral stenosis when the atrial septum is intact.

Clinical profile

Seven patients with mitral stenosis and a left-to-right shunt at the atrial level were seen in the Cardiology Department at the All India Institute of Medical Sciences, New Delhi. Their age, sex, clinical diagnosis, and the findings at operation or necropsy are presented in Table 1.

The radiological and electrocardiographic findings of the first 6 patients were similar, and thus

only the representative electrocardiogram and chest x-rays of Case 1 are being presented, while the data of all the patients are tabulated (Table 2).

Case reports

Case 1 A 12-year-old boy had polyarthritis at the age of 10 years followed by palpitation and exertional dyspnoea. Paroxysmal nocturnal dyspnoea appeared 3 months before coming to the hospital. He had been noted to become blue after running in earlier childhood. Physical examination showed cyanosis, praecordial bulge with gross cardiac enlargement, a hyperkinetic parasternal impulse, and a systolic thrill at the upper left sternal border. The first sound was accentuated and the second widely split and 'fixed' with a loud, palpable pulmonary component. A grade 4/6 ejection systolic murmur was heard along the upper left sternal border, and a long delayed diastolic murmur, without presystolic accentuation, was audible along the left sternal border and transmitted to the apex. The electrocardiogram showed right axis deviation ($+130^\circ$), first-degree atrioventricular block, right and left atrial hypertrophy, and right ventricular hypertrophy with incomplete right bundle-branch block (Fig. 1). The skiagrams of the chest showed cardiomegaly with pulmonary plethora, right ventricular and atrial enlargement, and absence of left atrial enlargement (Fig. 2). Cardiac catheterization find-

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ings (Table 3) showed bidirectional shunting at the atrial level with mild pulmonary arterial hypertension. Twenty-four hours after catheterization, the patient suddenly became hypotensive and died. At necropsy he had a single atrium with tight rheumatic mitral stenosis.

Case 2 A 23-year-old female patient was admitted with progressively increasing exertional dyspnoea, palpitation, fever, clay-coloured stools, and yellow discoloration of the conjunctivae of 10 days' duration. She had polyarthritides 4 years previously and again 4 months earlier. Physical examination showed an orthopnoeic, jaundiced, and emaciated young woman with considerable pallor and peripheral oedema but without cyanosis or clubbing. There was gross cardiac enlargement, with the apex in the sixth left interspace in the anterior axillary line and a systolic thrill at the third and fourth left interspaces. The first sound was accentuated and the second very much so,

and palpable. The splitting and movement of the components of the second sound with respiration were not clear. A grade 4 ejection systolic murmur all over the praecordium and a loud delayed diastolic murmur from the left sternal border to the apex without presystolic accentuation were heard. The lungs were full of coarse moist rales. The neck veins were distended up to the angle of the mandible, with a dominant 'v' wave, and the liver was 10 cm below the right costal margin. It was smooth, tender, and pulsatile. Ascites was present. A portable chest x-ray showed gross cardiac enlargement and was suggestive of pulmonary plethora. The electrocardiogram showed right axis deviation ($+105^\circ$), and right atrial and ventricular hypertrophy with incomplete right bundle-branch block. Because of her poor general condition and jaundice she could not be investigated further. She became hypotensive and died 10 days after admission. At necropsy she had a large atrial septal defect measuring 6.8 cm in

TABLE 1 *Clinical diagnosis and findings at operation or necropsy*

Case No.	Age (yr)	Sex	Clinical diagnosis	Findings at necropsy or operation
1	12	M	Cyanotic congenital heart disease, severe pulmonary hypertension, $L \rightarrow R$ shunt site?	Single atrium with rheumatic mitral stenosis
2	23	F	Tricuspid regurgitation with pulmonary hypertension; ? cause	Atrial septal defect, secundum, with rheumatic mitral stenosis, multiple pulmonary emboli
3	16	F	Atrial septal defect (secundum)	Atrial septal defect, secundum, rheumatic mitral stenosis
4	26	M	Lutembacher's syndrome	Atrial septal defect, secundum, rheumatic mitral stenosis
5	30	F	Atrial septal defect (secundum); ? Lutembacher's syndrome	Atrial septal defect, secundum, rheumatic mitral stenosis
6	10	F	Lutembacher's syndrome	1 cm sinus venosus atrial septal defect; right-sided pulmonary veins to right atrium close to defect, mitral stenosis
7	13	F	Rheumatic mitral stenosis	Rheumatic mitral stenosis, right middle and lower lobe pulmonary veins to right atrium, intact atrial septum

TABLE 2 *Electrocardiographic and radiological findings*

Electrocardiographic findings								Radiological findings						
Case No.	Rhythm	PR interval (sec)	QRS duration (sec)	Axis QRS	QRS in <i>V</i> ₁	'P' pulmonale	'P' mitrale	Cardiac enlargement	Main pulm. art. segment	Right atrium	Left atrium	Right ventricle	Left ventricle	Pulm. vasculature
1	Sinus	0.26	0.09	+130	rsR'	+	+	+++	++	++	o	+++	o	+++
2	Sinus	0.18	0.07	+105	qR	+	o	+++	++	Only a portable PA view available			o	++(?)
3	Sinus	0.16	0.08	+90	rsR'	+	+	+++	+++	++	o	+++	o	+++
4	Sinus	0.18	0.10	+110	rsR'	+	+	+++	+++	++	o	+++	o	+++
5	Sinus	0.16	0.08	+100	rsR'	+	±	++	++	++	o	++	o	+++
6	Sinus	0.17	0.07	+110	qR	+	+	+++	++	++	o	++	o	++
7	Sinus	0.19	0.06	+120	Pure R	o	o	++	++	±	++	++	o	±

diameter, with severe rheumatic mitral stenosis, multiple pulmonary emboli, and active rheumatic carditis.

Case 3 A 16-year-old girl was admitted with the complaint of palpitation. She denied a history suggestive of rheumatic fever and dyspnoea. Physical examination showed a praecordial bulge with diffuse pulsations, a hyperkinetic impulse, and a systolic thrill at the upper left sternal border. The cardiac apex was in the sixth left interspace in the anterior axillary line. The first sound was moderately accentuated and the second widely split and 'fixed' with the pulmonary component much accentuated. A grade 4 ejection systolic murmur was audible at the upper left sternal border and a loud delayed diastolic murmur, without presystolic accentuation, from the left sternal border to the apex. The electrocardiogram showed a mean frontal QRS axis of $+90^\circ$, right and left atrial hypertrophy, and right ventricular hypertrophy with incomplete right bundle-branch block. Skiagrams of the chest showed cardiomegaly with increased pulmonary vasculature, prominent main pulmonary artery segment, right atrial and ventricular enlargement, with absence of left atrial enlargement (Table 2). Haemodynamic findings (Table 3) showed a left-to-right shunt at the atrial level, mild pulmonary arterial hypertension, and equal left and right atrial pressures. An end-diastolic gradient of 4 mmHg in the withdrawal tracing from left ventricle to left atrium was missed before operation and was noted on superimposition of tracings after operation. At operation for a secundum atrial septal defect, a large atrial septal defect was found, measuring about 3 cm, together with rheumatic mitral stenosis of moderate severity admitting only the tip of the index finger.

Case 4 A 26-year-old man was seen with a history of progressively increasing dyspnoea and palpitation of 4 years' duration. There was no history suggestive of rheumatic fever. Physical examination showed gross cardiomegaly with the apex in the sixth left interspace almost in the anterior axillary line. The second sound and a systolic thrill were palpable at the second left interspace. The first sound was loud and the second widely split and 'fixed' with the pulmonary component much accentuated. An opening snap was not audible. A grade-3 ejection systolic murmur was audible at the upper left sternal border and there was a loud and long delayed diastolic murmur without presystolic accentuation along the left sternal border well heard up to the apex. The electrocardiogram showed right axis deviation ($+110^\circ$), with right and left atrial hypertrophy and right ventricular hypertrophy with incomplete right bundle-branch block. The chest skiagrams showed cardiomegaly with prominent main pulmonary artery segment, right atrial and ventricular enlargement, plethoric lungs, and absence of left atrial enlargement (Table 2). The haemodynamic evaluation confirmed the presence of atrial septal defect and the transmitral gradient

FIG. 1 Electrocardiogram of Case 1 showing a QRS axis of $+130^\circ$, first-degree atrioventricular block, right and left atrial and right ventricular hypertrophy.

FIG. 2 Skiagrams of Case 1. (A) and (B) are posteroanterior and right oblique views, respectively. There is cardiac enlargement with prominent right atrium, right ventricle, and pulmonary artery segment. There is no evidence of left atrial enlargement.

(Table 3). At operation there was a secundum atrial septal defect, 1.5×2 cm, with critical mitral stenosis.

Case 5 A 30-year-old mother of one child was seen with the complaint of exertional dyspnoea and palpitation starting after a miscarriage 3 years earlier. She gave a history suggestive of rheumatic fever 8 years and 4 years earlier and of paroxysmal nocturnal dyspnoea for the past few months. Physical examination showed absence of cyanosis and congestive failure. There was a hyperkinetic praecordial impulse with a systolic thrill at the second left interspace and moderate cardiomegaly. The first sound was accentuated and the second widely split and 'fixed' with a loud pulmonary component. An opening snap was not detected. A grade 4 ejection systolic murmur at the upper left sternal border and a long, fairly loud delayed diastolic murmur without presystolic accentuation from the left sternal border to the apex were heard. The electrocardiogram showed a mean frontal QRS axis of $+100$ with right atrial and ventricular hypertrophy with incomplete right bundle-branch block. The skiagrams of the chest showed cardiomegaly with prominent main pulmonary artery segment, pulmonary plethora, right atrial and ventricular en-

largement, and absence of left atrial enlargement (Table 2). The patient had to be catheterized twice as at the first instance a transmitral gradient could not be obtained. The haemodynamic findings confirmed the presence of atrial septal defect and the transmitral gradient (Table 3).

At operation she had a large atrial septal defect of the secundum variety with severe rheumatic mitral stenosis, the opening being about 0.7 cm.

Case 6 A 10-year-old girl was admitted with the complaint of dyspnoea on exertion and palpitation of one year's duration. She gave a history suggestive of rheumatic fever two years earlier. On examination, there was no evidence of congestive failure or cyanosis. There was a praecordial bulge with gross cardiac enlargement, heaving left parasternal impulse, and a systolic thrill at the upper left sternal border. The first sound was accentuated and the second widely split and 'fixed', with the pulmonary component moderately accentuated. A grade 3 ejection systolic murmur was audible at the second left interspace and a long and loud delayed diastolic murmur from the left sternal border to the apex, without presystolic accentuation. During inspiration a venous hum like a continuous murmur was also audible in the second and third left inter-

TABLE 3 Haemodynamic findings

Case No.	Saturations (%)							Pressures (mmHg)						
	SVC	RA	RV	Pulm. art.	Pulm. art. wedge	LA	System. art.	SVC	RA	RV	Pulm. art.	Pulm. art. wedge	LA	LV
1	50	75	77	75	—	77	77	(10)	a = 13 v = 12 (9)	43/11	38/17	—	(10)	—
3	54	80	83	88	—	97	97	(3)	a = 8 v = 7 (3.5)	38/6	32/15	—	a = 10 v = 7 (4)	102/6
4	68	85	84	84	92	93	93	(5)	a = 8 v = 6 (5)	30/8	30/12	(10)	(11)	100/5
5 (a)*	69	89	88	92	98	98	98	(10)	a = 13 v = 11 (10)	35/10	35/20	(13)	(13)	—
5 (b)*	57	86	87	87	97	—	97	—	a = 12 v = 10 (7)	47/10	46/20	(10)	—	138/7
6	76	91	91	91	99	—	99	(6)	a = 7 v = 6 (4)	35/7	35/22	(16)	—	95/2
7	51	71	71	73	—	—	99	(8)	a = 11 v = 9 (8)	68/9	68/35	—	—	—

SVC = Superior vena cava.

RV = Right ventricle.

LA → RA = Withdrawal from left to right atrium.

LVd LA or PAW = Simultaneous left ventricular diastolic and left atrial or pulmonary arterial wedge pressures.

mgr = Mean gradient.

LV = Left ventricular systolic and end diastolic pressures.

RA = Right atrium.

LA = Left atrium.

e.d. gr = End diastolic gradient.

* Patient catheterized on two different occasions as the mitral gradient was not obtained in the first study.

spaces by the side of the sternum (which disappeared during expiration). The electrocardiogram showed a mean frontal QRS axis of $+110^\circ$ with right and left atrial hypertrophy, right ventricular hypertrophy, and incomplete right bundle-branch block. Skiagrams of the chest showed considerable cardiomegaly with prominent main pulmonary artery segment, right atrial and ventricular enlargement, plethoric lungs, and absence of left atrial enlargement. The haemodynamic findings indicated the presence of a left-to-right shunt at the atrial level with a 10 mmHg pressure gradient between the two atria and a left atrioventricular mean diastolic pressure gradient of 13 mmHg. Because of the absence of left atrial enlargement and the presence of continuous murmur, she was felt to have a small atrial septal defect. At operation she had a sinus venosus atrial septal defect slightly less than 1 cm in diameter. The mitral opening was 0.6 to 0.7 cm.

Case 7 A 13-year-old girl was referred to the hospital with a diagnosis of Lutembacher's syndrome. She gave a history of dyspnoea on exertion, cough, and palpitation of one year's duration. There was no history suggestive of rheumatic fever. Physical examination disclosed a praecordial bulge with moderate cardiomegaly, a heaving

left parasternal impulse, and a faint systolic thrill at the second left interspace. The first sound was loud and the second widely split, but moving with respiration with a loud pulmonary component. An opening snap, moderately close to the second sound, and a long delayed diastolic murmur with presystolic accentuation were audible at the apex. There was a grade 2 ejection systolic murmur at the second left interspace. The electrocardiogram showed right axis deviation ($+120^\circ$) and right ventricular hypertrophy. Skiagrams of the chest showed cardiac enlargement with prominent main pulmonary artery segment, left atrial and right ventricular enlargement, and in retrospect pulmonary plethora confined to the right side (Fig. 3). At cardiac catheterization, the catheter could not be wedged in the pulmonary artery. Further attempts to document the mitral stenosis were not made as the clinical data were thought to be sufficient for the diagnosis. The catheter entered the right middle and lower lobe pulmonary veins from the right atrium. She was sent for an operation with a diagnosis of Lutembacher's syndrome. At operation she had a tight rheumatic mitral stenosis with an opening of 0.4 to 0.5 cm. The interatrial septum was intact and the right middle and lower lobe pulmonary veins entered the right atrium directly.

System. art.	LA→RA	LVd LA or PAW	Pulm. flow (l./m/m ²)	System. flow (l./m/m ²)
114/67	(10)→(10)	—	—	—
102/66	(4)→(3)	—	9.6	2.0
100/60	(11)→(5)	mgr=8 e.d. gr=6	11.5	4.1
—	(12)→(10)	—	19.5	4.0
140/92	—	mgr=6 e.d. gr=3	11.7	2.9
95/60	—	mgr=13 e.d. gr=13	10.8	3.7
105/75	—	—	3.7	2.1

FIG. 3 Posteroanterior view of the skiagram of Case 7 showing moderate cardiac enlargement, prominent main pulmonary artery segment, and increased pulmonary vasculature on the right side. The vasculature is suggestive but not diagnostic of left-to-right shunt.

Figures in parentheses indicate mean pressure.

Discussion

The clinical picture of Lutembacher's syndrome depends on the severity of mitral obstruction restricting the forward flow, whereas the size of the atrial defect and the pulmonary vascular resistance determine the magnitude of the left-to-right shunt. The existence of the classical findings of mitral stenosis accompanied by a continuous murmur of the left-to-right shunt because of the small size of the septal defect have been described by Marshall and Warden (1964), Goldfarb and Wang (1966), and Ross *et al.* (1963). In the present study Case 6 belongs to this category. This, however, is not the characteristic finding in the majority of patients with Lutembacher's syndrome, who present with features suggestive of a large left-to-right shunt, with little evidence for the mitral obstruction as seen in the first 5 patients. Thus, the clinical picture of patients with Lutembacher's syndrome is variable.

Mitral stenosis, at the bed-side, is recognized by the combination of a loud first sound, normally split second sound, an opening snap, and a delayed diastolic murmur with presystolic accentuation. An associated atrial septal defect decompresses the left atrium, resulting in the absence of presystolic accentuation of the diastolic murmur. Then flow across the tricuspid valve, which is relatively large because of the mitral obstruction, results in a loud delayed diastolic murmur. The mitral diastolic murmur becomes inseparable from the tricuspid flow murmur and both together are heard as a loud delayed diastolic murmur from the left sternal edge to the apex. The diastolic murmur in all of the first 6 patients was loud, transmitted all along the left sternal border and to the apex. A loud first sound, probably due to a loud tricuspid closure (Leatham and Gray, 1956), is usual in patients with atrial septal defect with a large left-to-right shunt. There is no significant change in the second sound due to additional mitral stenosis, which continues to show wide 'fixed' splitting with a loud pulmonary component. The loud pulmonary component is audible at the apex in the majority of patients with atrial septal defect irrespective of the presence or absence of pulmonary arterial hypertension (Sutton, Harris, and Leatham, 1968). Separation of the pulmonary component of the second sound and the opening snap at the bed-side is, therefore, difficult. Thus, the auscultatory findings may not always be helpful in diagnosing additional mitral stenosis. However, the presence of three unusual findings occurring together should suggest its possibility: (i) a heart size

relatively too large for an uncomplicated secundum atrial defect; (ii) a systolic thrill at the upper left sternal border; and (iii) delayed diastolic murmur which is loud, long, and heard along the left sternal border and well transmitted to the apex.

The electrocardiogram shows right axis deviation and rsR' in the right chest leads in over 90 per cent of patients with secundum atrial septal defects (Wood, 1956; Nadas, 1963; Kaplan, 1968). The morphology of the QRS complex in the right chest leads is not altered significantly to assist in the diagnosis of additional mitral stenosis. The 'P' waves are normal in patients with uncomplicated atrial septal defect of the secundum type (Barber, Magidson, and Wood, 1950; Burch and DePasquale, 1959; Kaplan, 1968). However, 'P' pulmonale may be present with high pulmonary vascular resistance (Martins de Oliveira and Zimmerman, 1958; Wood, 1956). Evidence for right and left atrial hypertrophy in 4 out of the 6 patients in the present series was the most helpful single electrocardiographic criterion for the presence of additional mitral disease.

Two of the chief radiological aids for the recognition of mitral stenosis are evidence of left atrial enlargement and pulmonary venous hypertension. None of the first 6 patients showed left atrial enlargement or presence of pulmonary venous hypertension. As such, radiological evaluation of mitral obstruction in the presence of atrial septal defect is unrewarding unless mitral calcification is present.

Cardiac catheterization shows evidence of a left-to-right shunt with a variable degree of pulmonary arterial hypertension. In the presence of a large atrial septal defect, there is no pressure gradient between the two atria, whereas with a small defect it may be present. While Cases 1, 3, and 5 had minimal gradients, Cases 4 and 6 had significant gradients (Table 3). In the presence of a normal left atrial pressure and the absence of a gradient between the two atria, the mitral obstruction may be overlooked, as happened in Case 3 in the present study or Case 2 in the series of cases reported by Steinbrunn, Cohn, and Selzer (1970). It is well known that smaller gradients during the diastolic period may be missed in a withdrawal tracing especially if there is marked respiratory variation in the ventricular end-diastolic pressure. Hence, unless simultaneous left atrial and ventricular diastolic pressures are recorded it may not be possible to document a small gradient across the mitral valve.

Thus, the clinical and haemodynamic diag-

nosis of mitral stenosis in the presence of an atrial septal defect is not so simple. Nevertheless, if a patient with an atrial septal defect has much cardiomegaly, a systolic thrill at the upper left sternal edge, an unusually long, loud, and widely transmitted delayed diastolic murmur, and electrocardiographic evidence of right and left atrial hypertrophy, the presence of additional mitral stenosis should be suspected even in the absence of radiological evidence for left atrial enlargement. The presence of a left atrioventricular diastolic pressure gradient should be confirmed at cardiac catheterization studies by simultaneous pressure records.

On the other hand, patients with partial anomalous pulmonary venous drainage, intact atrial septum, and mitral stenosis present clinically with findings typical of mitral stenosis with relatively few features to suggest the presence of left-to-right shunt. In Case 7, there was no difficulty in arriving at the clinical diagnosis of mitral stenosis because of the classical auscultatory, electrocardiographic, and radiological findings. The evidences for the additional left-to-right shunt were few. The unusual findings were: (a) the faint systolic thrill and murmur at the second left interspace, (b) the second sound was widely split in expiration and widened further on inspiration, and (c) the skiagram was suggestive of pulmonary plethora on the right side; it was, however, not diagnostic enough and was recognized only after cardiac catheterization. In a review of the 16 cases of this combination reported in English, 3 (Hughes and Rumore, 1944; Dimond and Gonlubol, 1953) were diagnosed at necropsy; 8 (Sepulveda, Lukas, and Steinberg, 1955; Nichols, Woldow, and Goldberg, 1956; Lendrum and Lichtman, 1957; Varnauskas *et al.*, 1963; Adler *et al.*, 1964; Bruschke and Bloch, 1969) were diagnosed after cardiac catheterization; 3 (Zion, 1956; Wassermil and Hoffman, 1962; Goldfarb and Wang, 1966) at the time of operation; and in 1 (Kalke *et al.*, 1967) details are not available. In the last it is probable that a correct clinical diagnosis was made since the patient had a 'scimitar' type of drainage to the inferior vena cava. A correct clinical diagnosis was suspected in one case (Aldridge and Wigle, 1965) because of unilateral hilar pulsations at fluoroscopy and confirmed by cardiac catheterization and operation.

Of these 16 cases, clinical details are available in 12. An ejection systolic murmur at the upper left sternal edge was noted in 11. Splitting of the second sound has been variously described, though the pulmonary com-

ponent was accentuated in all – in 5 normally split, in 3 wide and 'fixed' (in 1 as 'fixed splitting within physiological limits'), and in 4 it was not commented upon. In all, a clinical diagnosis of mitral stenosis was made by physical examination whereas an additional left-to-right shunt was suspected on the basis of a widened supracardiac shadow or increased vasculature in the skiagram in only 6. Six patients were catheterized twice before the diagnosis of anomalous venous drainage with intact atrial septum was reached.

In the present case, the clinical diagnosis was mitral stenosis, though she was referred with a diagnosis of Lutembacher's syndrome. The left atrial or pulmonary arterial wedge pressure was not obtained at the time of catheterization, and as the anomalous veins were entered directly and the saturation data suggested a left-to-right shunt at the atrial level, the presence of atrial septal defect was assumed incorrectly. This indicates that in patients with partial anomalous pulmonary venous drainage in association with mitral stenosis and intact atrial septum, it is the left-to-right shunt which is likely to be missed, specially if small.

There thus appears a justification in separating these two conditions, contrary to the suggestion of Goldfarb and Wang (1966), to broaden the concept of Lutembacher's syndrome to include partial anomalous pulmonary venous drainage with mitral stenosis and intact atrial septum in the same group. The presence of an atrial septal defect decompresses the left atrium and alters the clinical, radiological, electrocardiographic, and haemodynamic picture. It is also not justifiable to include patients with atrial septal defect with mitral regurgitation in this group as their clinical presentation is also quite different. They present as a large left-to-right shunt at the atrial level with peripheral signs of tricuspid regurgitation (Steinbrunn *et al.*, 1970).

Wassermil and Hoffman (1962) state that mitral stenosis with left-to-right shunt at the atrial level in the presence of a significant gradient between the two atria indicates partial anomalous pulmonary venous drainage with intact atrial septum. This is not supported by the findings of Cases 4 and 6 where a significant interatrial gradient was seen in the presence of an atrial septal defect. If, however, different pulmonary artery wedge pressures are obtained at different sites – high pressure confirming left atrial pressure (normally draining pulmonary veins) and low pressure indicating that the anomalous veins are joining the right atrium – partial anomal-

ous pulmonary venous drainage with intact atrial septum and mitral stenosis can be diagnosed with accuracy, as suggested by these authors.

As a rule, patients with uncomplicated atrial septal defects of the secundum variety do not have left atrial enlargement, though in older patients it can occur (Tikoff *et al.*, 1965). The 6 patients of Lutembacher's syndrome in this series were all below 30 years and did not have left atrial enlargement in spite of mitral stenosis. Steinbrunn *et al.* (1970) found left atrial enlargement in patients with Lutembacher's syndrome. Their patients belonged to an older age group. As such, left atrial enlargement in patients with atrial septal defect seems to be related to age irrespective of the presence or absence of mitral stenosis. This suggests that in the younger patients absence of left atrial enlargement does not exclude associated mitral stenosis, whereas in older patients, presence of left atrial enlargement does not necessarily indicate additional mitral narrowing though it should be looked for.

It thus appears that patients with mitral stenosis, partial anomalous venous drainage, and intact atrial septum differ significantly from patients with Lutembacher's syndrome in their clinical presentation. In the former it is the clinical recognition of the left-to-right shunt which is difficult, whereas in the latter it is the mitral obstruction which is likely to be missed. Patients with Lutembacher's syndrome also have a variable clinical picture depending on the size of the atrial septal defect, pulmonary vascular resistance, and the degree of mitral narrowing. The diagnosis of mitral narrowing is important, since, if missed, it may result in high left atrial and pulmonary venous pressures and postoperative difficulties after surgical closure of the atrial septal defect.

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