# Echocardiographic assessment of left and right heart hemodynamics in a patient with Lutembacher's syndrome

Navin Budhwani, MD,[a] Ather Anis, MD,[a] Kelly Nichols,[a] and Muhamed Saric, MD, PhD,[a] Newark, New Jersey

We present a case of a 53-year-old woman with intractable shortness of breath that was originally ascribed to bronchiolitis obliterans organizing pneumonia. Subsequently evaluation by echocardiography and cardiac catheterization revealed that she had Lutembacher's syndrome, an uncommon combination of congenital atrial septal defect (ASD) and acquired mitral stenosis that is difficult to diagnose clinically. Our case illustrates the pitfalls and advantages of echocardiographic assessment of the mitral valve area (MVA) and the left atrial pressure (LAP). The pressure half-time method used most commonly for estimating MVA echocardiographically is inaccurate and may lead to underestimation of the severity of mitral stenosis in patients with Lutembacher's syndrome. On the other hand, the presence of ASD provides an additional method of calculating LAP, the most important determinant of symptoms in patients with mitral stenosis. (Heart Lung® 2004;33:50-4.)

## CASE REPORT

A 50-year-old woman, a former smoker, came to the emergency room with complaint of shortness of breath on minimal exertion. She also reported 5-pillow orthopnea, paroxysmal nocturnal dyspnea, and cough productive of white sputum over the preceding month. She denied any chest pain, fever, or chills.

Her past medical history included rheumatic fever as a child, systemic hypertension and type II diabetes mellitus. She had been previously hospitalized on several occasions for severe shortness of breath thought to be caused by either bronchial asthma or presumed bronchiolitis obliterans organizing pneumonia. Her recent cardiac catheterization revealed normal left ventricular ejection fraction and no significant coronary artery disease.

On examination she was afebrile. Her blood pressure was 160/88 mm Hg and pulse was regular at a rate of 126 beats per minute. She was tachypneic with a respiratory rate of 40 breaths per minute and oxygen saturation of 86 to 88% on room air.

Jugular venous pressure was elevated at 12 cm of water. Lung auscultation revealed rales at both lung bases. On cardiac exam, S1 and S2 were normal in intensity. In addition, a grade II/VI holosystolic murmur and a diastolic rumble were heard at the cardiac apex.

Liver edge was palpable 2 cm below the costal margin. Exam of lower extremities revealed palpable pulses and ankle edema bilaterally.

Electrocardiogram was notable for sinus tachycardia. Chest x-ray showed diffuse bilateral air space disease suggestive of either pneumonia or pulmonary edema.

Transthoracic echocardiography revealed normal left ventricular systolic function with an ejection fraction of 65%. Mitral valve leaflets and subvalvular apparatus appeared thickened and partly calcified. Both mitral leaflets were restricted in motion and the anterior mitral leaflet had the characteristic hockey-stick appearance. As a consequence, there was moderate mitral regurgitation and significant mitral stenosis.

Invasive hemodynamic evaluation and all echocardiographic estimates of the mitral valve area except for the pressure-half time method were consis-

From the Department of Medicine, New Jersey Medical School, Newark, New Jersey.

Reprint requests: Muhamed Saric, MD, Director, Echocardiography Lab, Division of Cardiovascular Diseases, New Jersey Medical School, 185 South Orange Avenue I-538, University Heights, Newark, NJ 07103.

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**Table I**
Estimates of mitral valve area

| Method | Mitral Valve Area ($cm^2$) |
|---|---|
| I. Echocardiography | |
| Pressure Half-time | 1.3-1.6 |
| Continuity Equation | 1.1 |
| PISA Method | 0.8 |
| II. Cardiac Catheterization | |
| Gorlin Formula | 0.9 |

Normal mitral valve has an area of 4-6 $cm^2$.

tent with severe mitral valve stenosis (Table I). Furthermore, both noninvasive and invasive techniques revealed a severely elevated mean diastolic pressure gradient across the mitral valve that was directly proportional to the heart rate (Table II).

Echocardiographic estimates of elevated resting pulmonary systolic and diastolic artery pressures (47/23 mm Hg) were confirmed by subsequent cardiac catheterization (Table III).

On transesophageal echocardiogram, a secundum atrial septal defect (ASD) was noted having a maximum diameter of at least 5 mm and a predominant left-to-right shunt. Mean pressure gradient across the ASD was unusually high and was estimated echocardiographically at 14 mm Hg. The ratio of pulmonic to systemic blood flow was 1.2:1 and 1.3:1 by cardiac catheterization and echocardiography, respectively (Table IV).

On the basis of the combined finding of rheumatic mitral valve stenosis and a secundum ASD, we established the diagnosis of Lutembacher's syndrome. After medical management brought her heart failure under control, she was referred for open-heart surgery. Intraoperatively, the native mitral valve leaflets were noted to be deformed and heavily calcified. The chordae were markedly foreshortened whereas the papillary muscles were scarred. The severely stenotic mitral valve was replaced with a 23-mm St. Jude prosthesis and primary closure of ASD was performed.

## DISCUSSION

In 1916, Lutembacher first described in detail a combination of congenital ASD with acquired mitral stenosis. The hemodynamic features and natural history of patients with Lutembacher's syndrome are variable and depend on the size of ASD, severity of mitral stenosis, pulmonary vascular resistance and the compliance of right ventricle.

The hemodynamic consequences of ASD in patients with Lutembacher's syndrome are exacerbated by high LAP caused by mitral stenosis. This leads to an increase in the shunt across the ASD and results in pulmonary flow higher than what otherwise would be in the absence of mitral stenosis.

When mitral stenosis is severe and atrial septal defect is small, it usually presents clinically as pure mitral stenosis. On the contrary, when the atrial septal defect is large, the signs and symptoms of ASD dominate the clinical picture, despite significant mitral stenosis.

In either case, the severity of mitral stenosis may be underestimated. Failure to recognize mitral stenosis has been related to mortality from intractable pulmonary congestion following closure of the atrial septal defect.

The normal mitral valve has a funnel-shaped orifice with an area of 4.0 to 6.0 $cm^2$ and permits filling of the left ventricle from the left atrium without significant diastolic pressure gradient. As mitral stenosis develops, mitral valve area (MVA) decreases and the diastolic pressure gradient (DPG) increases.

Severe mitral stenosis is characterized by a MVA $< 1.0$ $cm^2$ and/or mean DPG $> 10$ mm Hg. Cardiac catheterization has been traditionally used for hemodynamic evaluation of mitral stenosis. However, 2-dimensional and Doppler echocardiography are currently the method of choice and thus cardiac catheterization in no longer mandatory in the evaluation of a patient with mitral stenosis. [5-8]

There are at least 4 different echocardiographic techniques for determination of MVA: pressure-half time, planimetry, continuity equation, and proximal isovelocity area method. Because of its relative simplicity, the pressure half-time method using Doppler echocardiography is the most commonly used technique and allows for the simultaneous determination of mean DPG.

In patients with isolated mitral stenosis, all 4 methods provide similar estimates of MVA. However, in patients with Lutembacher's syndrome the pressure half-time (P ½ is an inaccurate method to determine the mitral valve area. In these patients, the pressure half-time across the stenotic mitral valve is abnormally shortened because of the simultaneous blood flow across the atrial septal defect. Therefore P ½ method should be used with caution in patients with Lutembacher's syndrome since it overestimates MVA (Table I).

**Table II**
Mean diastolic pressure gradients across mitral valve

| Method | Heart Rate (beats per minute) | Mean Pressure Gradient (mm Hg) |
|---|---|---|
| Echo | 68 | 10 |
| | 97 | 12-15 |
| Cardiac catheterization | 80 | 12 |
| | 102 | 16 |

Normal mitral valve area exhibits virtually no diastolic pressure gradient.

**Table III**
Left and right heart pressures by cardiac catheterization and echocardiography

| | Cardiac Catheterization | Echocardiography |
|---|---|---|
| Right atrium | | |
| Mean | 5 | |
| Right ventricle | | |
| Systolic | 50 | |
| End-diastolic | 6 | |
| Pulmonary artery (PA) | | |
| Systolic | 48 | 47 |
| Diastolic | 24 | 23 |
| PA wedge pressure | | |
| Mean | 21 | |
| Left ventricle | | |
| End-diastolic | 6 | |

Pressures are expressed in millimeters of mercury.

**Table IV**
Estimates of atrial septal defect shunt fraction

| Method | Pulmonic Flow (Qp) (L/min) | Systemic Flow (Qs) (L/min) | Qp:Qs Ratio |
|---|---|---|---|
| Echocardiography | 7.11 | 5.39 | 1.3:1 |
| Cardiac catheterization | 6.02 | 5.02 | 1.2:1 |

Although the MVA estimate is an important indicator of the severity of mitral stenosis, the degree of left atrial pressure (LAP) elevation ultimately determines the patient's symptoms.

In all patients with mitral stenosis, LAP can be estimated both invasively and noninvasively from either the mean DPG across the stenosed mitral valve or from the pulmonary artery diastolic pres-

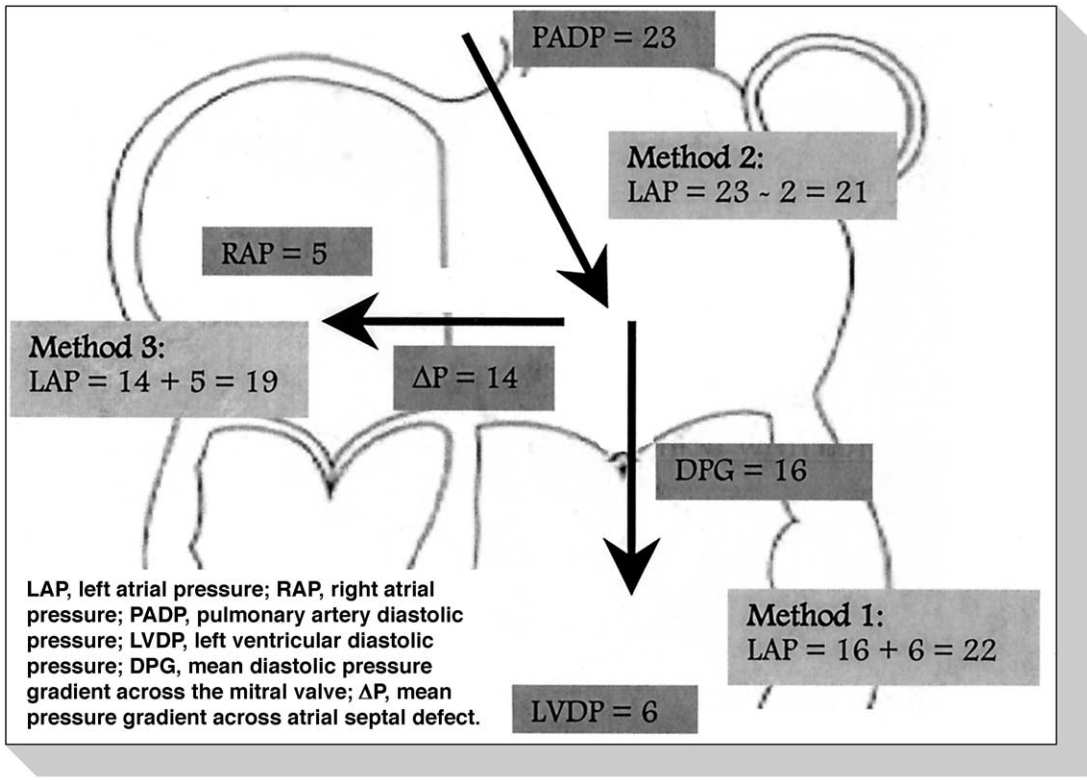


**Fig 1.** Estimates of Left Atrial Pressure in a patient with Lutembacher's syndrome. LAP, left atrial pressure; RAP, right atrial pressure; PADP, pulmonary artery diastolic pressure; LVDP, left ventricular diastolic pressure; DPG, mean diastolic pressure gradient across the mitral valve; ΔP, mean pressure gradient across atrial septal defect.

sure (Methods 1 and 2, respectively in Fig. 1). In our patient, LAP was estimated to be greater than 20 mm Hg by both of these methods.

In Method 1, LAP was estimated by adding left ventricular diastolic pressure determined on cardiac catheterization (6 mm Hg) to the mean DPG (16 mm Hg by cardiac cath, or 15 mm Hg by echocardiography). In Method 2, LAP was obtained by subtracting the gradient between the pulmonary artery and the left atrium in diastole (about 2 mm Hg) from the pulmonary artery diastolic pressure estimated by echocardiography (23 mm Hg) or cardiac catheterization (24 mm Hg).

The presence of ASD in a patient with Lutembacher's syndrome provides an additional echocardiographic method of measuring left atrial pressure. In such a patient, LAP can be determined from the Doppler measurement of the gradient across the atrial septal defect (Method 3 as shown in Fig. 1).

For example, the pressure gradient across the ASD (14 mm Hg), added to the right atrial pressure (5 mm Hg) at cardiac catheterization or Doppler echocardiography estimated LAP of 19 mm Hg, which is consistent with the estimated LAP using Methods 1 and 2 as shown in Fig. 1.

Lutembacher's syndrome is an uncommon condition that is difficult to diagnose clinically. Clinical suspicion of Lutembacher's syndrome should be raised by history of rheumatic heart disease, heart failure and atrial septal defect. The role of 2D and Doppler echocardiography in identifying Lutembacher's syndrome is well documented and it is suggested that this may be the only diagnostic technique needed before surgical correction of the lesions in younger patients.[14,15]

Limitations and unique advantages of echocardiography in the setting of Lutembacher's syndrome should be recognized. Although the Doppler-derived pressure half-time method is commonly used to estimate MVA, this method may lead to underestimation of the severity of mitral stenosis in patients with ASD. On the contrary, the presence of ASD provides an additional method of calculating

LAP, the most important determinant of symptoms in patients with mitral stenosis.

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