Lutembacher Syndrome in a 35-Year-Old Filipino Woman: A Case Report

Arch Anthony Reynoso, MD | Josephus Sibal, MD; Jose Donato Magno, MD | Angelito Medina, MD | Myrna Reblando, MD | Melissa Perez, MD
Angeles University Foundation Medical Center, Angeles, Pampanga, Philippines

Corresponding author:
Arch Anthony Reynoso, MD
Angeles University Foundation Medical Center, Angeles, Pampanga, Philippines
Email: anthonyreynosomd@yahoo.com.ph

Declaration of conflicts of interest: None.

INTRODUCTION

Lutembacher syndrome (LS) is an uncommon cardiac abnormality characterized by any association of a congenital or iatrogenic atrial septal defect (ASD) and a congenital or acquired mitral stenosis (MS).[1] The hemodynamic effects of this syndrome are a result of the interplay between the relative effects of the ASD and MS.[2]

CASE REPORT

A 35-year-old woman presented to the cardiology clinic with a 1-month history of easy fatigability, exertional dyspnea, and productive cough with whitish phlegm. She has no edema, orthopnea, paroxysmal nocturnal dyspnea, or fever. Two weeks before admission, she developed palpitation and still with persistence of symptoms. She sought consultation and was prescribed with clopidogrel 75 mg one tablet once a day, sildenafil 100 mg one tablet once a day, and verapamil 40 mg one tablet once a day, which provided relief. She has no history of hypertension, diabetes, cerebrovascular disease, coronary artery disease, or thyroid or renal disease. She is a nonsmoker and an occasional alcoholic drinker.

On general physical examination, blood pressure was 120/80 mm Hg; cardiac rate, 72 beats per minute; and respiratory rate, 20 cycles per minute. She was afebrile and had distended jugular veins with rales at the left lung base. Abdominal examination revealed neither organomegaly nor shifting dullness. There was neither cyanosis nor pedal edema.

On cardiac examination, dynamic precordium and no visible skin lesions or mass were noted. Apical impulse was displaced at the sixth intercostal space left midclavicular line. The left parasternal lift was noted. Rhythm was regular. The following were also noted: loud S1, widely split and fixed S2, grade 4/6 crescendo-decrescendo systolic ejection murmur heard at the left upper sternal border grade 4/6 short, and rumbling mid-diastolic murmur at the third intercostal space LPSB. Carvallo sign was noted.

Electrocardiogram revealed normal sinus rhythm, normal axis, left atrial abnormality, left ventricular hypertrophy by voltage criteria, incomplete right bundle-branch block, and occasional premature atrial contraction (Figure 1). Chest x-ray anteroposterior view showed dilated central pulmonary arteries, cardiomegaly predominantly showing right ventricular enlargement, and marked left atrial enlargement (Figure 2). Transthoracic echocardiogram revealed interatrial septal defect, large secundum type with significant left-to-right shunt with Qp/Qs of 5.9:1, and possible mild mitral valve stenosis (Figure 3). Transesophageal echocardiogram revealed possible LS: congenital heart disease: interatrial septal defect, secundum type with deficient retroaortic rim and significant left-to-right shunt, and rheumatic heart disease, possible mild mitral valve stenosis (Figure 4). Hemodynamic studies revealed ASD with bidirectional shunting, severe pulmonary hypertension (pulmonary vascular resistance: 3733 dyn/s per cm^5 and 2993 dyn/s per cm^5 before and after O_2 supplementation, respectively), and moderately severe pulmonary vascular disease with Rp:Rs of 0.71 and 0.60 before and after O_2 supplementation, respectively. She received nondihydropyridine calcium-channel blocker, phosphodiesterase inhibitor 2, and P2Y12 inhibitor. She is currently stable and on follow up at the cardiology clinic.

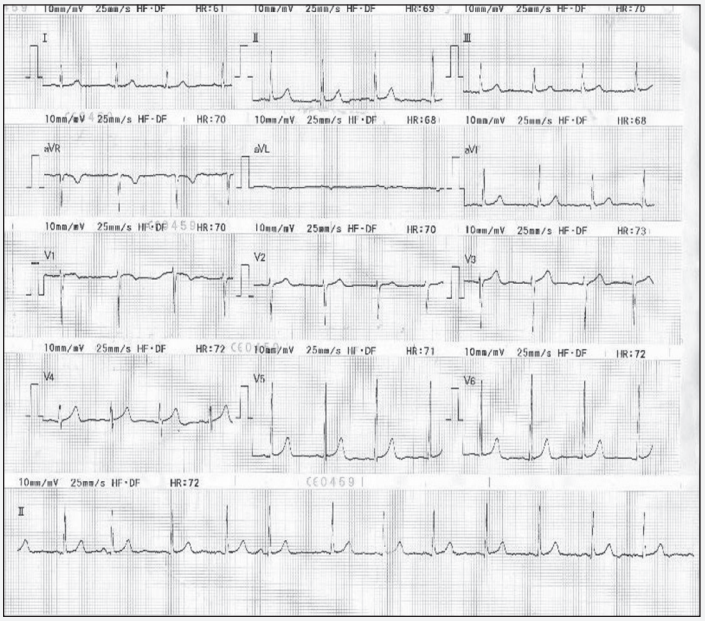

Figure 1. Electrocardiogram: normal sinus rhythm, normal axis, left atrial abnormality, left ventricular hypertrophy by voltage criteria, incomplete right bundle-branch block, and occasional premature atrial contraction.

DISCUSSION

The objective is to present the case of a 35-year-old woman with large secundum-type ASD with mitral valve stenosis or LS. It is important to report such cases to determine the therapeutic approach in this case. Lutembacher syndrome primarily has high left atrial pressure due to MS assumed to stretch open the patent foramen ovale, initiating left-to-right shunt and producing another outlet for the left atrium.[3] There is insufficient epidemiology, and its occurrence is generally undetermined, but existing estimations propose that it could be more common in areas of elevated endemicity for rheumatic fever.[4] Incidence is more common in female than in male patients. Hemodynamic features and natural history of patients with LS may vary and depend on the severity of MS, size of the ASD, pulmonary vascular resistance, and the compliance of the right ventricle.[5] Prompt diagnosis and surgical treatment on present studies showed a good prognostic value. A bad prognosis is anticipated if diagnosis is done at a late stage and when pulmonary hypertension and right-sided heart failure develop.[6]

Lutembacher syndrome is uncommon, and appropriate management and investigation are important to improve

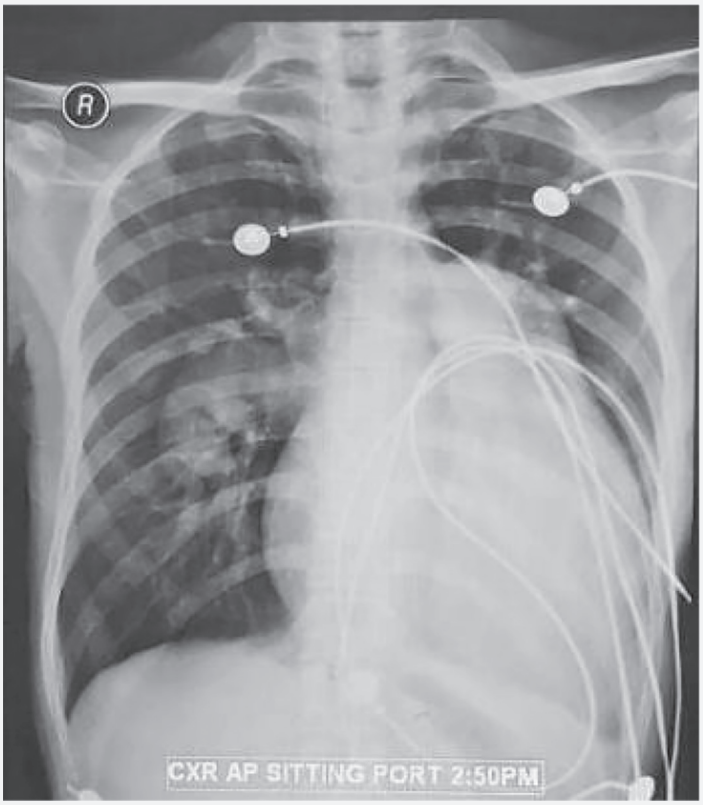

Figure 2. Chest x-ray anteroposterior view: Dilated central pulmonary arteries, cardiomegaly predominantly showing right ventricular enlargement and marked left atrial enlargement.

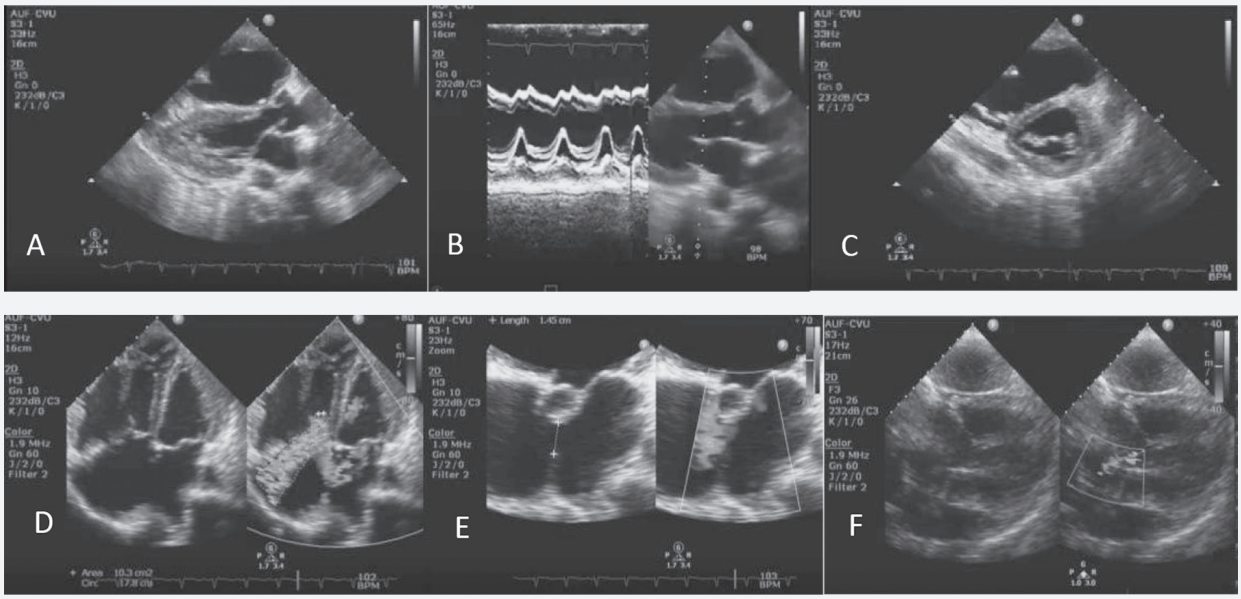

Figure 3. Transthoracic echocardiogram. A, PLAX view: the anterior mitral valve leaflet is thickened at the tip, with subtle diastolic doming motion; the commissures, however, are not yet fused. B, M-mode: showing sclerosis of the anterior mitral leaflet. C, PSAX mitral valve level: possible mitral stenosis, with thickened anterior mitral valve leaflet. D, Apical 4-chamber view: ASD, large secundum type: abnormal color flow noted across the interatrial septum, secundum type. Moderate to severe tricuspid regurgitation: mosaic color flow noted across tricuspid valve (jet area = 10.3 cm^2, vena contracta = 0.60 cm) during systole. E, Apical-5 chamber view: ASD, large secundum type: abnormal color flow noted across the interatrial septum, secundum type with defect measuring 1.4 cm in its widest diameter with a predominantly left-to-right shunt with Qp/Qs of 5.9:1 using flow across the RVOT and LVOT (RVOT VTI = 12 cm, LVOT VTI = 9.6 cm). F, Subcostal view: ASD, large secundum type: abnormal color flow noted across the interatrial septum, secundum type.

LVOT=left ventricular outflow tract; PLAX=parasternal long-axis view; PSAX=parasternal short-axis view; RVOT=right ventricular outflow tract; VTI,=velocity time integral.

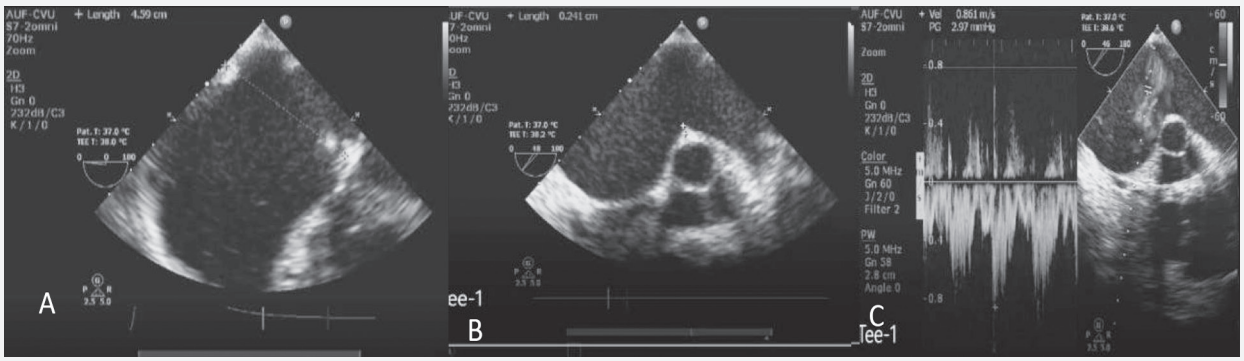

Figure 4. Transesophageal echocardiogram. A, Atrial septal defect, large secundum type: echo drop out in the midportion of the interatrial septum: interatrial septal height: 4.6 cm. B, Atrial septal defect, large secundum type: retroaortic rim: 2.41 mm. C, Atrial septal defect, large secundum type: abnormal color flow across the septum (peak pressure gradient = 3 mm Hg).

outcomes. There are neither large case series nor studies that report its natural course, and it is essential to report such cases to establish the timing of management. Transthoracic echocardiography remains the standard of imaging with the aid of transesophageal echocardiography to exclude other concomitant cardiac pathologies. Hemodynamic evaluation, including indices of right ventricular function, is of utmost importance in decision-making. Diagnosing at an early stage is beneficial and appropriate in deciding interventional management..

REFERENCES

1. Aminde LN, Dzudie A, Takah NF, Ngu KB, Sliwa K, Kengne AP. Current diagnostic and treatment strategies for Lutembacher syndrome: the pivotal role of echocardiography. Cardiovasc *Diagn Ther* 2015;5(2):122–132.
2. Uguru SU, Raphael SC, Idoko PO, Onuh JA, Amusa GA. Lutembacher's syndrome: a rare combination of congenital and acquired heart disease—a case report and review of literature. *Jos J Med* 2016;10(1).
3. Kulkarni SS, Sakaria AK, Mahajan SK, Shah KB. Lutembacher's syndrome. *J Cardiovasc Dis Res* 2012;3:179–181.
4. Budhwani N, Anis A, Nichols K, et al. Echocardiographic assessment of left and right heart hemodynamics in a patient with Lutembacher's syndrome. *Heart Lung* 2004;33:50–54.
5. Yadav DK, Choudhary S, Gupta P, Kumar P. Lutembacher syndrome: masquerading as rheumatic mitral stenosis. *Congenit Cardiol Today* 2012;10(3):1–6.
6. Levin AR, Spach MS, Boimeau JP, Canent RV Jr, Capp MP, Jewett PH. Atrial Pressure flow dynamics and atrial septal defects (secundum type). *Circulation* 1968;37:476–488.