

Lutembacher Syndrome: Dilemma of Doing a Tricuspid Annuloplasty

Abstract

We discuss the case of a 24-year-old woman with Lutembacher syndrome and severe tricuspid regurgitation (TR) who underwent surgical closure of atrial septal defect and mitral valve replacement without tricuspid annuloplasty despite a severe TR and a large tricuspid annulus on preoperative echo. The pathophysiology of Lutembacher syndrome is discussed below. The utility of perioperative echocardiography in assessing the annular diameter, tenting area and coaptation depth and thus providing insights into the functioning of the tricuspid valve will also be emphasized.

Keywords: Coaptation, Lutembacher syndrome, tenting height, tricuspid annuloplasty

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Introduction

Lutembacher syndrome is a rare clinical condition characterized by the presence of a congenital atrial septal defect (ASD) and acquired mitral stenosis (MS).^[1] It was first described in 1916 by Lutembacher in a 61-year-old woman.^[2] The hemodynamic effects depend on the severity of MS, size of ASD, pulmonary vascular resistance, and right ventricular compliance. MS augments the left to right shunt caused by the ASD, while the ASD reduces the left atrial pressures due to MS and hence the transmitral gradient.^[3] This leads to right ventricular volume overload, consequent dilatation of tricuspid annulus and a functional tricuspid regurgitation (TR). Although traditionally corrected surgically, both lesions are amenable to transcatheter correction.

Case Report

A 24-year-old woman presented with complaints of New York Heart Association class III dyspnea and exertional palpitations since 2 years and worsening of symptoms in the past 4 months. She had orthopnea and anasarca at presentation and also gave a history of recurrent lower respiratory tract infection since childhood.

Transthoracic echocardiography showed a large 29 mm ostium secundum ASD with left to right shunt, severe MS, and calcified mitral valve (mitral valve area of 0.9 cm² and transmitral gradient of

8 mmHg), severely dilated right atrium and right ventricle (RV), and a severe TR with a tricuspid annulus of 53 mm. Chest X-ray showed cardiomegaly (RV type apex) and electrocardiogram showed right bundle branch block with the left atrial enlargement.

The patient was optimized with medications before surgery. She was on verapamil 40 mg thrice daily, frusemide 40 mg twice daily, and digoxin 0.125 mg once daily which were continued.

On the day of surgery, the patient was premedicated with lorazepam 1 mg and omeprazole 20 mg orally 1 h before surgery. A peripheral venous line and radial arterial line was started before induction. The patient was induced with 5 mcg/kg of fentanyl and 0.2 mg/kg etomidate intravenously, and tracheal intubation was facilitated by 0.1 mg/kg vecuronium using a 7 mm ID endotracheal tube. An 8.5 French quadrilumen central venous catheter was inserted into the internal jugular vein under ultrasound guidance. A transesophageal echo (TEE) probe was inserted, and intraoperative examination confirmed thickened calcified mitral leaflets with severe MS, a large ASD and the tricuspid annulus measuring 47 mm with severe TR.

Intraoperatively, mitral valve replacement using a size 31 St. Jude bileaflet mechanical valve was done. The ASD was closed with a large Dacron patch. The aortic cross clamp was released, and the heart picked up spontaneously in a sinus rhythm.

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Once adequate deairing was done under TEE guidance, the patient was weaned off bypass with adrenaline and milrinone. The TEE showed a tricuspid annulus of 4.7 cm, leaflets coapting well with a coaptation depth of 7.2 mm [Figure 1]. Since the leaflets were coapting surface to surface, appeared healthy and the right ventricular contractility was good, it was decided against a tricuspid annuloplasty despite the moderate TR jet [Figure 2]. A TTE done on postoperative day 5 showed that the TR was only mild.

Discussion

Lutembacher syndrome refers to a rare combination of congenital ASD with acquired MS. It is more common in females. The long-term natural course of ASD is unfavorably influenced by MS, which augments the left to right shunt leading to right ventricular dilatation and functional TR.

The natural history of unrepaired functional TR post adequate restoration of left-sided valve lesions is often difficult to predict. Usually, there is enduring dysfunction after repair. Reoperation for isolated TR carries significant surgical risk. Current guidelines (ESC/EACTS and ACC/AHA) recommendations based on expert experience favor surgical repair of tricuspid valve during the left heart surgery if tricuspid annulus is $> 40 \text{ mm}/21 \text{ mm/m}^2$.^[4,5] These recommendations are for left side valve lesions which cause pulmonary hypertension and right ventricular pressure overload. Lutembacher syndrome essentially leads to RV volume overload by the augmented shunt across the ASD. There are no clear recommendations regarding management of functional TR for such lesions.

Tricuspid valve is an asymmetric structure, and its annulus is stiffer due to surrounding muscular tissue. Normally, the tricuspid valves coapt surface to surface (5–10 mm surface contact between the leaflets during systole) at or just below the tricuspid annular plane.^[6] RV dilatation causes its elliptical structure to become more planar with tenting of the valve leaflets into the RV side.^[6] Furthermore, the coaptation becomes edge to edge. With significant tethering, there may be no coaptation of the leaflets during systole. Tenting

area (the area enclosed by the annular plane and septal and anterior leaflet) and coaptation depth (the distance between the coaptation point of the septal and anterior leaflets and the tricuspid annular plane) must also be considered in addition to annular diameter before deciding on tricuspid annuloplasty, considering the three-dimensional (3D) structure. A tenting area $>1.6 \text{ cm}^2$ and a coaptation depth $>8 \text{ mm}$ indicate severe TR.^[6,7] The use of 3D echocardiography to assess tricuspid valve tethering in terms of tenting volume is considered a more accurate parameter for decision-making. A tenting volume of $>2.3 \text{ ml}$ has a sensitivity of 100% and specificity of 84% in predicting residual TR after tricuspid annuloplasty.^[8]

In our case, we found that the coapting depth was less than the recommended cut-off. Furthermore, the valve was healthy and coapting well. RV was contracting well. Hence, it was decided against a tricuspid annuloplasty despite the annular size and moderate TR. A transthoracic echo done during follow-up showed that the TR had become mild.

Conclusion

Lutembacher syndrome has a unique pathophysiology and mechanism of functional TR. The presence of an ASD with a left to right shunt causes a volume overload in the right side of the heart, increase in pulmonary blood flow and pulmonary artery hypertension. These dynamics are even further exaggerated with the presence of an MS. All these can contribute to an increase in the severity of TR. With the closure of the ASD, the hemodynamics start normalizing, with a decrease in the amount of TR in an otherwise normal TV. Before deciding for tricuspid annuloplasty, a comprehensive TEE examination of the tricuspid valve including annular diameter, tenting area, and coapting depth may provide insight to the functioning of tricuspid valve. The appropriate assessment can potentially obviate the need for a tricuspid annuloplasty in select cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The

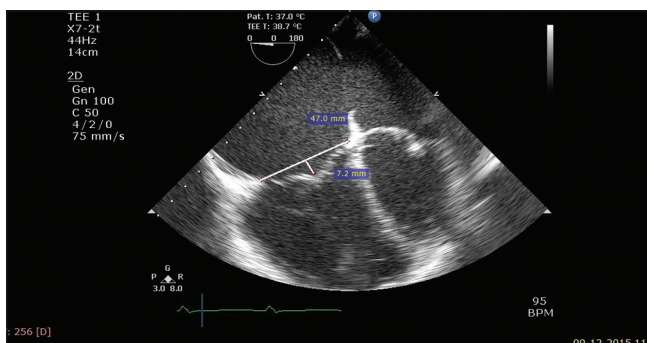


Figure 1: Tricuspid annulus of 47 mm and depth of coaptation 7.2 mm. Tricuspid leaflets are seen coapting surface to surface

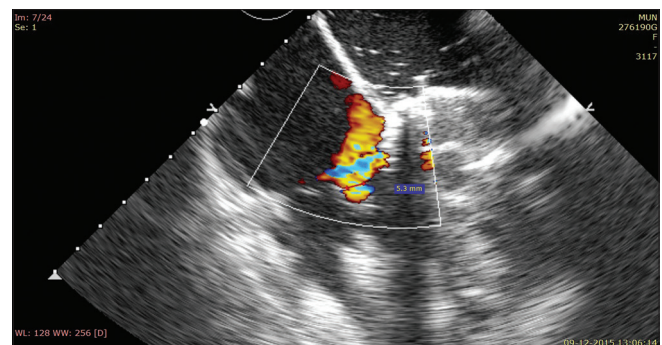


Figure 2: Moderate tricuspid regurgitation with vena contracta of 5.3 mm

patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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