

SURGICAL TECHNIQUE

Surgical Correction of Lutembacher's Syndrome in a Patient with Severe Pulmonary Artery Hypertension

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ABSTRACT **Lutembacher's syndrome is an uncommon combination of a congenital ostium secundum atrial septal defect (ASD) with acquired mitral stenosis (MS). The incidence of this condition is very rare. The symptoms are dependent upon the size of the ASD, severity of the MS, compliance of the right ventricle and pulmonary artery hypertension. We describe a patient with Lutembacher's syndrome with severe pulmonary hypertension who underwent successful surgical repair.** doi: 10.1111/jocs.12340 *(J Card Surg 2014;29:569–571)*

Lutembacher's syndrome is a rare cardiac anomaly comprising a congenital atrial septal defect (ASD) and acquired mitral stenosis (MS).[1] The treatment in such patients can be either surgery or percutaneous intervention. We describe a case of Lutembacher's syndrome with severe pulmonary hypertension (PAH), which was treated successfully by surgery.

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(Institutional Review Board permission was obtained to present this case.)

A 51-year-old male presented with chest pain and dyspnea on exertion and New York Heart Association Class III symptoms. On clinical examination, there was a 3/6 mid-diastolic murmur at the mitral area and a parasternal heave. Electrocardiogram demonstrated normal sinus rhythm with complete right bundle branch block. Chest radiography revealed cardiomegaly as well as increased pulmonary vascularity (Fig. 1). Transesophageal echocardiography (TEE) showed severe MS with a mitral valve area of 0.8 cm^2 by planimetry, a large ostium secundum ASD of 39 mm and a dilated right atrium, right ventricle and main pulmonary artery (PA) (Fig. 2). Color flow Doppler by TEE showed a left to right shunt at the atrial septum. Additionally, there was moderate tricuspid regurgitation with a systolic PA pressure of 80 mmHg.

Right heart catheterization demonstrated that the pulmonary arterial pressure was 76/31 mmHg with a mean of 46 mmHg at a systemic pressure of 118/70 mmHg. The baseline pulmonary to systemic flow ratio (Qp/Qs) was 2.7 and after oxygen Qp/Qs was 4.1. The baseline pulmonary vascular resistance was 5.8 and after oxygen it was 3.7. The pulmonary arterial pressure came down to 69/27 mmHg with a mean of 44 mmHg on oxygen. Coronary angiography showed normal epicardial coronaries. Since the ASD was too large with a deficient inferior septal rim and associated with severe PA pressures, it was decided to perform a surgical repair instead of percutaneous balloon mitral valvotomy and device closure of the ASD.

Following a midline sternotomy, cardiopulmonary bypass (CPB) was instituted using standard aortic and bicaval cannulation with moderate hypothermia (28 °C). After aortic cross-clamping, antegrade cold blood cardioplegia was instituted supplemented with topical myocardial cooling. The right atrium was opened obliquely and extended over the top of the right atrium to the common wall of the right and left atrium and the roof of the left atrium. A large ASD was seen. The mitral valve was found to be severely stenosed and the mitral valve was replaced using a 29 mm St. Jude mechanical valve (St. Jude Medical, Inc., St. Paul, MN, USA) with posterior mitral leaflet preservation. As the

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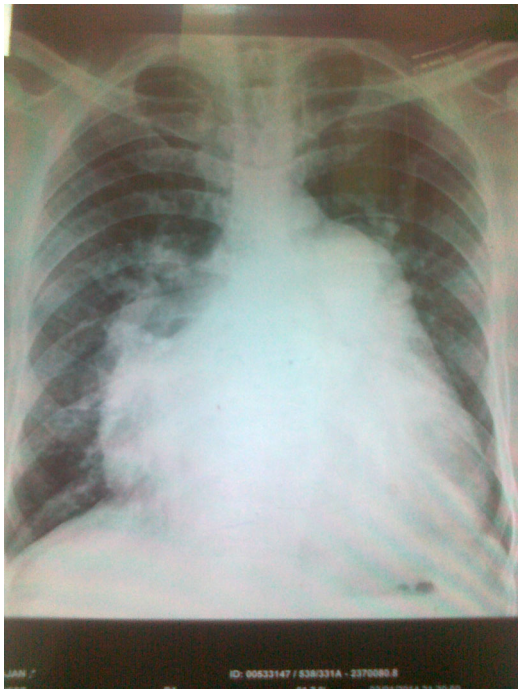

Figure 1. *Preoperative chest X-ray showing cardiomegaly and increased pulmonary vascularity.*

PA pressures were high, to improve the outcome after repairing the septal defect it was decided to leave a small interatrial communication to provide a pop-off during periods of elevated right-sided pressures. Hence, the ASD was closed using a Dacron patch with a 4-mm fenestration in the central area. The patient was weaned off from CPB uneventfully.

In view of the severe pulmonary pressures, the patient was electively ventilated for 2 days. Postoperatively his PA pressures were 34/21 mmHg and he was discharged on the 8th postoperative day on anticoagulative medications. Follow-up echo was done after 3 months and showed a small residual ASD with a left to right shunt with a systolic PA pressure of 30 mmHg.

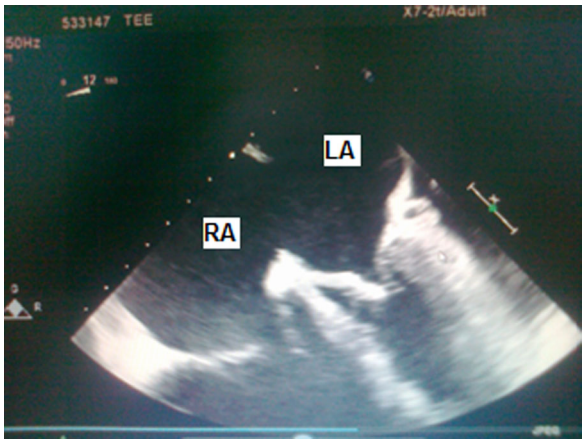

Figure 2. *TEE showing severe MS with ASD.*

DISCUSSION

Lutembacher's syndrome is an uncommon combination of a congenital ostium secundum ASD with acquired MS, which is usually rheumatic and was first described by Rene Lutembacher.[2] The incidence of this condition is very rare, 0.001/1,000,000.[1] The hemodynamics of the disease is dependent upon the size of the ASD, severity of MS, compliance of the right ventricle, and PA hypertension. In cases of MS and an ASD, left atrial (LA) pressure is decreased. The incidence of MS in patients with ASD is estimated at 4%, and conversely, an ASD in patients with MS is 6% to 7%.[3,4]

In Lutembacher's syndrome, the presence of MS augments the left to right shunt through the ASD instead of going backward into the pulmonary veins, which decompress the left atrium, reducing the mitral gradient, thus avoiding pulmonary venous congestion. Thus, patients with this syndrome may have a relatively benign clinical course until the development of PAH. Development of Eisenmenger's syndrome or irreversible pulmonary vascular disease is uncommon in the presence of a large ASD and elevated LA pressure because of MS.[1]

The treatment in Lutembacher's syndrome is predominately open-heart surgery. But with the introduction of percutaneous balloon mitral valvuloplasty and transcatheter closure of ASDs, Lutembacher's syndrome can be safely and effectively treated percutaneously in selected patients with suitable septal anatomy and mitral valve morphology.[4,5] But in our case as the ASD was large with a deficient rim it was decided to perform a surgical repair.

In the presence of severe PA pressures, closing the septal defects may cause right ventricular dysfunction, which can be fatal. Hence, a fenestration of the patch while closing the ASD can potentially serve as a pop-off during periods of elevated right-sided pressures where the resultant right to left shunt through the interatrial communication prevents acute right ventricular failure. Afterwards it can be intervened percutaneously to close this communication if the PA pressures subside and a left to right shunt persists.[6]

If a patient is diagnosed at a late stage with severe PAH, the prognosis is poor.[5] In Lutembacher's syndrome with severe PA pressures with ''border-line operability,'' mitral valve replacement with ASD closure leaving a small interatrial communication has a good prognosis and prolongs survival.[6] This approach may help to salvage these patients in the short- to mid-term but the long-term effects on the natural history of patients with long-standing severe PAH are unknown.

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