Case Report
Anaesthetic Management for Minimally Invasive approach for ASD closure, Mitral valve replacement and Tricuspid valve repair in a patient with Lutembacher syndrome

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Abstract
Lutembacher syndrome is a combination of congenital atrial septal defect with acquired mitral stenosis which poses a perioperative challenge to the anaesthesiologist. We herein describe a the case of a 28 year old lady with Lutembacher syndrome for whom an ASD closure, mitral valve replacement and tricuspid valve repair was performed with a minimally invasive approach with fast-track cardiac anaesthesia.

Keywords: Lutembacher syndrome, minimally invasive cardiac surgery, fast-track cardiac anaesthesia.

Introduction
The Lutembacher syndrome is named after physician René Lutembacher who gave the first comprehensive account of this condition in 1916¹. This syndrome entails the presence of a congenital atrial septal defect (ASD) with an acquired mitral stenosis (MS). The ASD may be associated with mitral regurgitation (MR). Previously the preferred method of treatment included open heart surgery involving ASD closure and mitral valve commisurotomy or valve replacement². But in recent years, minimally invasive cardiac surgery has become an increasingly used option for managing patients with valve pathology³.

Here we present a case report describing the perioperative course of a 28 year old female patient with Lutembacher syndrome (LS) for whom an ASD closure, mitral valve replacement and tricuspid valve repair was done using a minimally invasive approach through a right mini thoracotomy.

Case Report
A 28 year old lady, diagnosed as a case of Lutembacher syndrome during her second pregnancy 5 years back, managed conservatively previously, presented with NYHA class III complaints of dyspnea for 6 months. The preoperative transthoracic echo showed a large ostium secondum type ASD (2.5 cm) with a Left to right shunt; a severe MR, velocity 3m/s; moderate MS (Mitral valve peak gradient – 10mmHg, Mitral valve mean gradient–5mmHg); severe tricuspid regurgitation with pulmonary hypertension (pulmonary artery systolic pressure-61 mmHg), a D shaped left ventricle (LV) with a paradoxical septal motion but normal LV systolic
function (Ejection fraction– 60%). The chest x-ray showed cardiomegaly and the ecg showed sinus rhythm.

Patient was assessed under ASA III. She was premedicated with T. Ranitidine 150mg and T. Diazepam 5mg prior to surgery. A large bore peripheral IV access and a right radial arterial line was secured before induction. She was induced using Inj Thiopentone 4mg/kg, Inj Fentanyl 10 mcg/kg and relaxant Inj Vecuronium 0.1mg/kg, then intubated with a left sided double lumen tube (DLT) of 35 Fr. A central venous catheter of 7 Fr and a 15 Fr cannula was secured in the left internal jugular vein and right internal jugular vein (for venous drainage from the superior vena cava) respectively along with a left femoral arterial line with an 18G cannula simultaneously. A transoesophageal echo (TEE) probe was place and used to ensure & secure correct cannula placement, removal of air and adequacy of surgical procedure. The patient was maintained on an oxygen-air-isoflurane gas mixture with Inj Vecuronium 2mg boluses given intermittently. Analgesia cover was provided with Inj Morphine 0.05mg/kg at the start of the procedure along with Inj Fentanyl 50mcg boluses given at skin incision, pericardiectomy and valve placement. Surgeon annulated the right femoral artery and vein with 15 Fr and 17 Fr cannula respectively for cardiopulmonary bypass. After right lung isolation a right mini thoracotomy was performed in the 4th right intercostal space. Pericardial patch was harvested. Cardiopulmonary bypass (CPB) was instituted post systemic heparinisation. The right atrium was opened and through the ASD, the native diseased mitral valve was inspected and excised. A 29mm TTK Chitra valve was implanted with the ASD closed using the autologous pericardial patch. The tricuspid valve annuloplasty was done with 32 mm felt patch. An intercostals block was administered using 0.125% Bupivacaine for post operative pain relief. The patient was weaned off CPB uneventfully and the DLT replaced with an endotracheal tube 7.5 size. Patient was shifted to the cardiothoracic-ICU where she was weaned and extubated within 5 hours. Post op pain was managed with oral T. Paracetamol 20mg/kg and T. Ketorolac 10mg TDS. She was shifted to the ward after drain removal once she was mobilized on post operative day 1. She was then later discharged on post operative day 4 with stable hemodynamics and adequate PT/INR values.

**Discussion**

Lutembacher syndrome is a very rare condition with scanty epidemiological data. According to one study published in the American Heart Journal in 1997 LS incidence is 0.001/100000. It is more likely to be prevalent in areas with higher prevalence of rheumatic heart disease. The hemodynamic effects of this syndrome are because of relative effects of ASD & MS. Features and natural history of the patient depends on :-size of ASD, severity of MS, compliance of right ventricle, degree of pulmonary vascular resistance.

The MS augments the left to right shunt through the ASD decreasing the left atrial pressure gradient causing a decrease in transvalvular mitral gradient resulting in an ameliorating effect on the clinical expression of the mitral stenosis. But there is also an unfavorable influence on the progress of the natural history of the ASD which augments the left to right shunt predisposing to atrial fibrillation and right ventricular failure. Early diagnosis and surgical correction of helps give a better prognosis to the patient. The success of the operation is closely linked to the effective perioperative management of the pulmonary hypertension and the left ventricular function.

As per the Society of Thoracic Surgeons any procedure not performed with full sternotomy and cardiopulmonary bypass is a minimally invasive cardiac surgery (MICS). It includes a wide range of and a variety of techniques and approaches maybe used for the same. These include:-partial sternotomy, thoracotomy, video assisted procedures, robot assisted procedures.
The challenges faced by the anaesthesiologist in MICS which includes:- re-establishing the diagnosis to assess the eligibility for MICS, provision of optimal surgical field, application of appropriate monitoring modalities consistent with cardiac lesions, approach to the patient, cannulations, cardioplegia, single lung ventilation (for thoracotomy approach), TEE6.

There is heavy emphasis on the use of TEE in these surgeries due to reduced cardiac exposure and percutaneous cannulation to confirmation of adequacy of surgical procedures6. TEE is critical for monitoring of cardiac distension and removal of air. In successfully surgeries the patient would enjoy improved cosmesis and decreased bleeding, wound infection, pain and a shorter hospital stay. Even with ample advantages to the patient there are certain drawbacks to MICS which include:- difficulty converting to a sternotomy in case of problems, complications of femoral artery cannulation, arterial dissection, infection, hematoma, increased time of aortic cross clamping.

There is a clear significant learning curve to provide an equivalent surgical outcome with a MICS versus a traditional strategy. Minimal invasiveness requires maximum awareness and vigilance from both the surgeon as well as the anaesthesiologist equally. Continuous coordination and communication among the surgeon, anaesthesiologist and the perfusionist is essential at every step of MICS to deal with perfusion associated emergencies6.

Fast-tracking in cardiac surgeries is a complex intervention for appropriately selected patients which incorporates several components of care during cardiac anaesthesia and in the postoperative period. These components of care include administration of low dose opioid general anaesthesia, use of time directed extubation protocol or both7,8. This is all done with the ultimate aim of- early tracheal extubation after surgery, decreased length of ICU and hospital stay. Safe and effective fast track cardiac care in turn helps reduce hospital costs as well as the burden on hospital resources. However incidence of fast-track failure after cardiac surgery ranges from 11 – 16 %9,10. So an appropriate patient selection is of immense importance.

**Conclusion**

Perioperative management of Lutembacher syndrome in cardiac surgery poses a significant challenge both for the anaesthesiologist and the operating surgeon. Intraoperative coordination between surgeon, anaesthesiologist and the perfusionist helped reduce the aortic cross clamping time which on one hand reduces the associated complications as well as decreased the need for post operative ventilation. Also administration of low dose opioid general anaesthesia and time directed extubation protocol helped us to fast track the patient's recovery from the ICU to the ward and then back home.

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