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Case Report

Single Operator Percutaneous Balloon Mitral Valvotomy with Subsequent Device Closure of Atrial Septal Defect in a Case of Lutembacher Syndrome under Trans-Esophageal Echocardiography Guidance

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Abstract

Lutembacher Syndrome is an uncommon disease entity. Case reports of non- surgical treatment using team approach have been described. We describe our experience in a patient where a single operator carried out the entire procedure using Trans-esophageal guidance, periodically switching between the catheterization laboratory table and the echocardiography machine to guide the procedure. Logistic difficulties in developing world lead to inevitable modifications to overcome difficult circumstance. Twin procedure in the case described was done safely and effectively as a modification of the standard procedure.

Keywords: Congenital Heart Disease; Rheumatic Mitral Stenosis; Non-surgical treatment

Introduction

Lutembacher Syndrome(LS) represents a combination of congenital heart disease; Atrial Septal Defect (ASD) with acquired rheumatic heart disease, mainly Mitral Stenosis (MS). Surgical treatment has been the cornerstone of this condition with patch closure of ASD along with mitral valve repair and/or replacement for MS. There have been several case reports of non- surgical percutaneous treatment of LS. In some cases, per-cutaneous treatment has been shown to be less morbid [1]. A multi-disciplinary team approach including interventional cardiologists, cardiac anesthetist and trained personnel in trans-esophageal echocardiography are usually required for performing the procedure. Developing world scenarios in catheterization laboratory may pose logistic challenges for which locally devised innovations may be necessary.

Case Report

The patient was an 18 year old girl who presented with complaints of shortness of breath and palpitation of several years duration. She weighed 38 kilograms and was 148 cm tall. She was married early and had a girl child at the age of 16 years. During pregnancy and peri-partum course, she had noticed worsening of her symptoms. She was treated with diuretics with symptomatic benefit.

For the past few months, she again noticed worsening of symptoms for which an echocardiographic evaluation was done in a remote place. She was diagnosed with ASD and moderately severe MS of rheumatic etiology. ASD was measured in a trans-thoracic echocardiography at 24 mm with large left to right shunt. Her mitral valve area (MVA) was calculated by

planimetry was 1.1 cm^2. There was no mitral re-gurgitation. Due to large pre-tricuspid shunt, MVA by pressure half time over-estimated the MVA and was considered unreliable [2, 3].

Surgical option of mitral valve repair or replacement with patch closure of ASD was offered to the patient. She wanted to have one more child to complete the family and given the unwillingness and practical limitations to monitor prothrombin time and non- availability of institutional care for future child birth, it was decided to treat her non surgically by percutaneous method.

The patient underwent a trans-esophageal echocardiographic (TEE) exam to assess suitability of the defect for device closure. The ASD has good margins all around with maximum size of 28 mm in the short axis view at the upper esophageal level. The MVA was estimated to be 1.2 cm^2 in the trans-gastric short axis view by planimetry. The left atrium (LA) and left ventricle (LV) were found to be small. The right atrium, right ventricle and pulmonary arteries were dilated. The shunt was left to right with calculated Qp/Qs ratio of 2.9. There was minimal tricuspid regurgitation with estimated pulmonary artery systolic pressure of 38 mm Hg.

Procedure

Patient was shifted to the catheterization laboratory after informed and written consent. General anesthesia was administered and a TEE probe was introduced and positioned to profile the ASD in short axis at upper esophageal level. The echocardiography machine was placed near the operator on the same side of the catheterization laboratory since the regular echocardiographer failed to turn up. The femoral vein access was taken on the right side using modified Seldinger technique and after adequate dilatation of the right groin area; an 11 French (F) cannula was placed in the right femoral vein. The left femoral artery was accessed using similar method and a 5F arterial monitoring sheath was introduced and transduced for continuous arterial pressure monitoring peri-procedure. Unfractionated heparin @ 80 units/ kg was administered. The inter-atrial septum was crossed using Judkin's right (JR) 6F catheter over regular 0.035" J tip wire. The 6F JR was then left in the LA and attempts were made to park the 0.021" coiled guide wire in the LA. Due to small size of the LA, the coiled guide wire kept buckling back into the right atrium. Keeping the coiled guide wire in the RA, a stretched Acura 23-26 mm balloon (Vascular Concepts, UK) was then advanced into the RA. Subsequently a 0.035" J tip wire was used to place the balloon in the LA. This type of modified passage of balloon over the wire into the LA has been described earlier [4]. Initial attempts to introduce the Acura balloon into the LV were unsuccessful due to small LA size. The stylet was then taken out and given a very narrow 180 degree curve. The balloon was then entered into the LV, this was confirmed on TEE, and an inflation of 25 mm was given (Figure 1). Both commissures were then seen to split adequately. Subsequent attempts with 26 mm inflation were given but the balloon would come out in inflated state out of the MV. This suggested adequate MV opening, MVA using trans-gastric short axis view was estimated to be 2 cm^2. There was trace mitral regurgitation.

Figure 1.Acura Balloon in inflated position across the mitral valve.

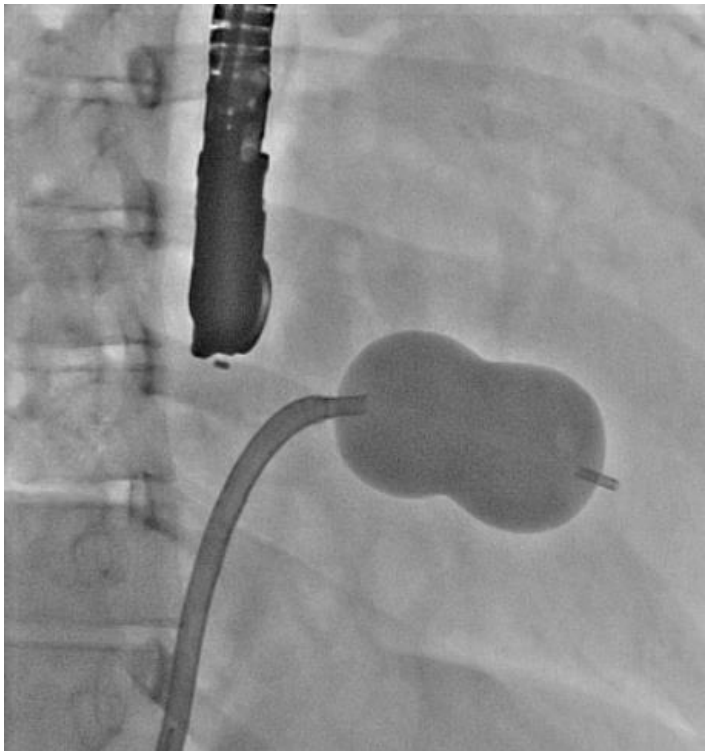
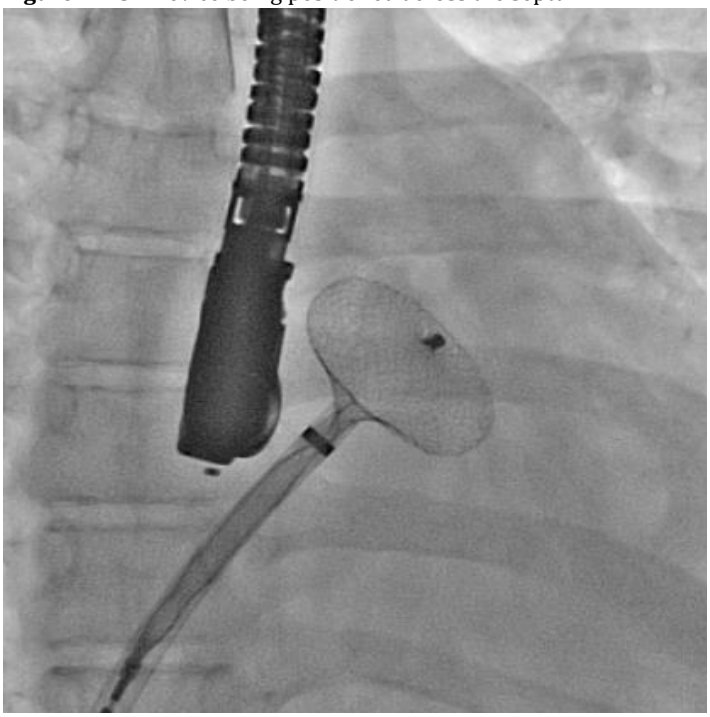
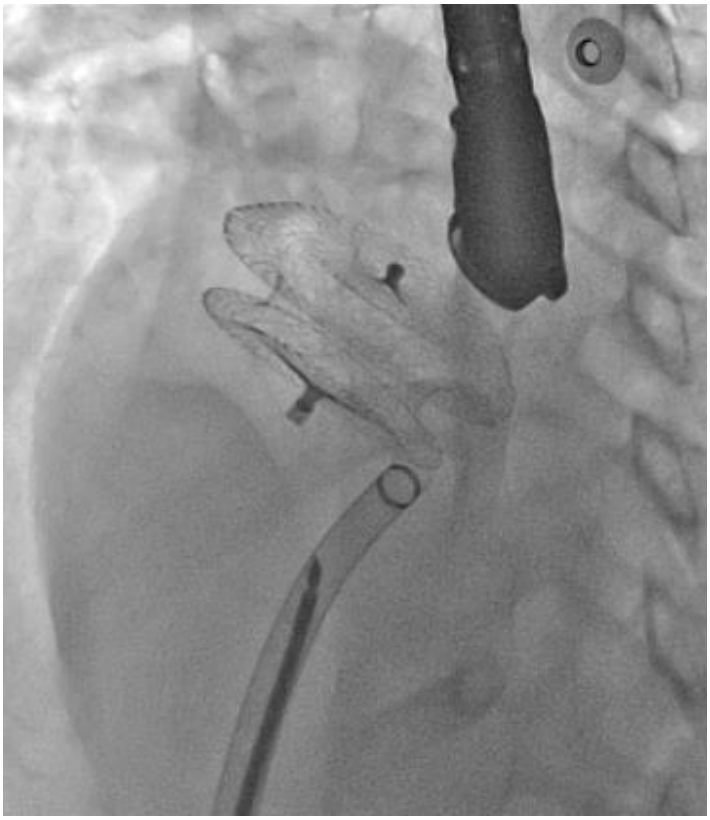


Figure 2. ASD Device being positioned across the septum.



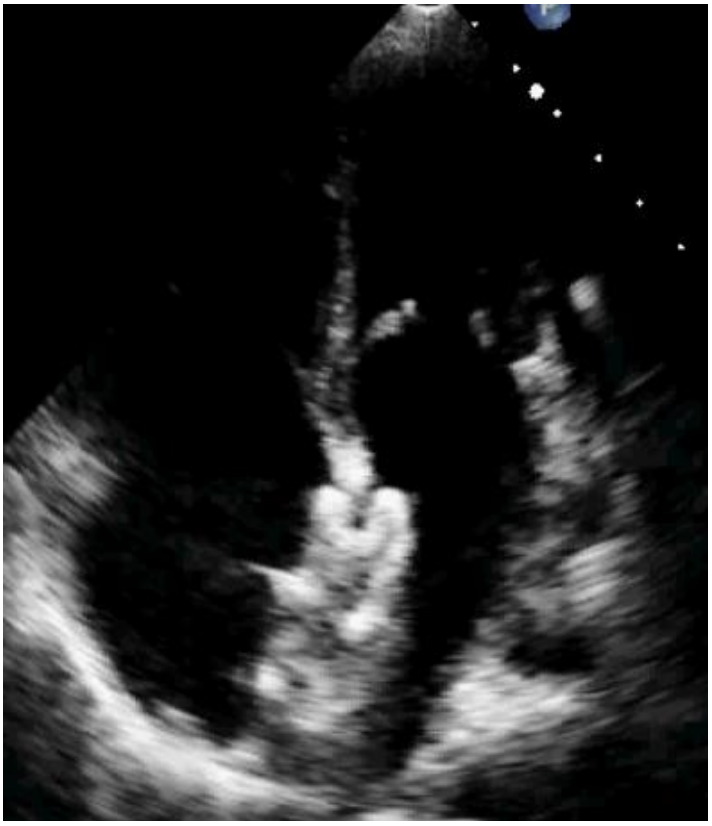
Next the balloon was removed into the RA over coiled guide wire, stretched and removed keeping the coiled guide wire in the RA. A JR catheter was then advanced into the RA and coiled guide wire removed. The JR was then advanced into the LA over 0.035" J tip wire, which was then exchanged for 12 F Amplatzer TM TorqVueTM delivery system (St. Jude Medical, MN, USA) over 260 cm super stiff wire. A 30 mm Cocoon ASD device (Vascular Innovations, Thailand) was then loaded and attempts were made to deploy the device (Figure 2). The device position was ascertained with the help of TEE images, the operator switching intermittently between the TEE machine and catheterization laboratory table for accurate placement of device. The TEE machine was strategically positioned on the same side as the operator, next to the catheterization laboratory table, to facilitate these movements. After repeated attempts to place the device went unsuccessful using the left superior pulmonary vein approach, the Amplatzer delivery system was given half a clockwise turn to face the right superior pulmonary vein. Deployment from this position was satisfactory and no residual shunt could be seen from across the margins (Figure 3).

Figure 3. Device in fully deployed position.



The patient was then shifted to the intensive care unit after anesthesia reversal. Post procedure hospital course was uneventful, electrocardiogram was normal and she was discharged after 48 hours after a post procedure TTE (Figure 4) revealed no residual shunt across the defect with MVA of 2 cm^2.

Figure 4. Post procedure TTE image (Apical 4 chamber view) showing device in position and mitral valve adequately opened up.



Discussion

LS is an uncommon disorder where congenital heart disease physiology of ASD gets modified by acquired rheumatic heart disease, mainly MS. As a result, the left to right shunt due to ASD gets exaggerated; over-estimating the ASD related shunt and under-estimating the severity of MS. Surgical treatment has been considered the standard of care, till several operators reported feasibility of non- surgical closure of ASD after treating MS with balloon valvotomy [5-12]. A percutaneous approach was considered in the patient described due to high costs of surgery (Balloon valvotomy with ASD device implant typically costs about 80% of open heart surgery in our hospital), higher risks involved in subsequent pregnancies with artificial valve necessitating institutional care which was not locally available and patient's unwillingness for open heart surgery. A per-cutaneous approach would eliminate these risks. Moreover, in the event of valve re-stenosis post balloon valvotomy, surgical option could be exercised at a later date allowing sufficient time to complete her family.

Unusual technical feature of balloon mitral valvotomy relates to small size of the LA due to ASD shunting away most of the blood into right heart chambers, not allowing LA to dilate. This presents a technical challenge which pertains to directing the Acura balloon into the LV, which was overcome in the case by

modification of the shaper device to a smaller and more acute bend. Subsequent ASD closure represents another technical challenge due to altered lie of the atrial septum necessitating maneuvering the Amplatzer sheath clockwise so that if faces the right sided pulmonary veins for optimal device delivery. Other technical issues due to combined disorder have been recently described in an article by Bhambhani et al [13].

A unique challenge in this particular case was the absence of trained personnel to assist on the TEE for guidance during the procedure. The echocardiographer called sick the very last moment. This was overcome by strategic position of the TEE system on the same side of the catheterization laboratory as the primary operator, thus facilitating operator switch from the TEE system to catheterization laboratory table and vice versa. This added extra minutes to the procedure and had sterility issues which were handled with care using extra pair of gloves and gowns every time a switch was made. Intra-cardiac echocardiography is a reasonable alternative in such circumstance, unfortunately it was not available. A cardio-thoracic surgical colleague was kept standby during the entire procedure.

Periodic non availability of well-trained support staff represents a practical challenge in the developing world; this was overcome by modification of the available resources and manpower without compromising the safety or the efficacy of the procedure.

Conclusion

LS is an uncommon clinical condition. For selected individuals with favorable anatomy, a percutaneous treatment with balloon mitral valvotomy followed by ASD device closure is an option. The procedure is feasible using single operator with strategic placement of equipment in the catheterization laboratory.

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