Review Article

Current diagnostic and treatment strategies for Lutembacher syndrome: the pivotal role of echocardiography

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Abstract: Lutembacher syndrome (LS) is a rare cardiac abnormality characterized by any combination of a congenital or iatrogenic atrial septal defect (ASD) and a congenital or acquired mitral stenosis (MS). Clinical features and hemodynamic effects of LS depend on the balance of effects of the MS and the ASD. Prognosis is influenced by several factors [pulmonary vascular resistance, right ventricle (RV) compliance, size of ASD and MS severity] but the occurrence of secondary pulmonary hypertension and congestive heart failure is commonly associated with poor outcome. Echocardiography remains the gold standard for diagnosis and evaluation of LS. Timely diagnosis is critical for modifying the natural course, by allowing patients to benefit from currently available percutaneous trans-catheter therapies with favorable effects on the outcomes. This article is a review of published literature on the current diagnostic and therapeutic modalities for LS, focusing on the pivotal role of echocardiography as the key diagnostic tool. Clinical suspicion of LS should prompt extensive investigation with non-invasive and where possible, invasive technics. Multicenter registers have a potential to assist the evaluation of long term outcomes of percutaneous trans-catheter therapies in patients with LS.

Keywords: Lutembacher syndrome (LS); mitral stenosis (MS); atrial septal defect (ASD) diagnosis; echocardiography; trans-catheter therapy

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Introduction

Cardiovascular disease (CVD) is a leading cause of death and a major public health challenge worldwide (1). The proportion of CVD due to congenital heart diseases is increasing as exemplified by the increasing demand for cardiac interventions both invasive and non-invasive to treat both acquired and congenital CVD (2). This has major public health and financial implications. The situation is likely worse in low- and middle-income countries (LMIC) and those in sub-Saharan Africa in particular owing to the high endemicity of rheumatic heart disease (3). In some rare instances, congenital and acquired heart diseases co-existing.

Lutembacher syndrome (LS) is a rare clinical condition characterized by the presence of a congenital, secundum-type atrial septal defect (ASD) and an acquired mitral stenosis (MS) (commonly of rheumatic origin) (4). LS has been reported to be generally associated with long-term unfavorable natural course (5). The ASD in LS can also be

iatrogenic, secondary to trans-septal puncture during mitral valvuloplasty for acquired MS. Generally, ASD in LS should have a diameter greater than 1.5 cm which later causes left to right shunt, progressive pulmonary hypertension (PH) and development of Eisenmenger syndrome (4). Another form described in the literature is the 'reverse LS'; with instead a predominant pulmonary-to-systemic or right-to-left shunting of blood in the context of an ASD and severe tricuspid stenosis (6). Some authors have argued that LS could involve any mitral valve lesion (stenosis, insufficiency or mixed). The definition of LS has undergone several changes and adaptations with time but, currently any combination of ASD (congenital or iatrogenic) and MS (congenital or acquired) is referred to as LS (7).

There is scanty epidemiological data on LS and its prevalence is generally unknown, but current opinions suggest that it could be more common in areas of high endemicity for rheumatic fever (8). In a case series by Bashi *et al*. in 1987, a history of acute rheumatic fever was found in up to 40% of patients with LS in developing countries (5). Congenital MS is a rare entity, accounting for only 0.6% of cases of congenital heart diseases in autopsy studies. In patients with ASD, up to 4% have MS while the incidence of ASD in patients with MS is 0.6-0.7% (9). Nevertheless, some data suggests that the incidence of LS is 0.001 per million populations, while the proportion of iatrogenic LS stands at 11-12% (10). In a necropsy study carried out in the United States, five cases out of 25,000 autopsies conducted had a combination of MS and ASD (11). LS is common among young adults (4) and a female preponderance has also been noted (9) with the first case reported in the literature being in a 61-year-old woman (12). In this review, we focus on the role of echocardiography in the contemporary diagnostic and therapeutic modalities for LS.

Diagnosis of LS

Clinical presentation

Generally, patients with LS are known to remain asymptomatic for several years. In up to 40% of cases from developing countries, there is a history of rheumatic fever (5,13). Clinical features are usually due to ASD and variations in symptoms and signs are dependent on the size of ASD. Thus, the hemodynamic changes seen in LS result from the balance effects of the ASD and the MS, as well as compliance of the right and left ventricles which are the main determinants of the direction of blood flow (14). Commonly, patients present with fatigue, exercise intolerance and palpitations. Fatigue and exercise intolerance are due to the reduced systemic blood flow which is a result of the MS and the systemic to pulmonary shunting of blood across the ASD in diastole, thereby reducing blood flow into left ventricle (9,10). The MS and increased left atrial pressure from augmented left-to-right shunt of blood lead to atrial dilatation. This predisposes patients to atrial arrhythmias commonly atrial fibrillation (AF) and most patients present with palpitations (9).

In patients with non-restrictive ASD, the features of pulmonary congestion present late in the course of the disease. With moderate to severe MS, patients would present mainly with features of right ventricle (RV) overload and right heart failure (RHF). Patients with restrictive ASDs and moderate to severe MS, present much earlier and usually with features of pulmonary congestion from MS (5). Thus, factors which influence the natural history as well as haemodynamic features of LS include: pulmonary vascular resistance, RV compliance, severity of MS and the size of the ASD (5,14). Due to the non-specific features at the early stage of the disease, patients almost always present to hospital in advanced states (15).

Physical examination

Physical examination of patients with LS reveals signs in relation with the co-contributing lesions (i.e., ASD and MS):

(I) Arterial pulses are generally of small volume with regular rhythm due to the decreased left ventricular stroke volume (SV). In those patient who already have AF, the pulse is irregular (15);

(II) Jugular venous pulse (JVP): in non-restrictive ASD, both atria function as a single chamber, hence, height and contour of left atrial pressure is transmitted to right atrium and internal jugular vein. Thus, patients with LS have elevated JVP (even in the absence of RHF) as well as elevated jugular venous a-wave in absence of PH (9);

(III) The precordium: a left parasternal lift is observed due to the transmitted RV and pulmonary impulses commonly in patients with a non-restrictive ASD. The left ventricle impulse is usually not appreciated due to reduced filling from MS (16). The diastolic thrill from MS is unusual due to the comparatively

low mitral valve flow velocity (17);

(IV) Heart auscultation: a loud first heart sound is heard as well as early-to-mid diastolic murmur from the MS and also presence of pulmonary hypertension [leading to increased right atrial (RA) pressure and consequently left atrial pressure] causing increased trans-mitral pressure gradient (16). Unfortunately, these are sometimes attenuated by the ASD causing decompression of left atrium as well as further dilation of the RV in pulmonary hypertension which decreases trans-mitral pressure gradient (9,16,17). Splitting of second heart sound (S_2) also occurs for the following reasons; late closure of the pulmonic component of S_2 (from increased right heart flow of ASD) and early closure of the aortic component of S_2 (from decreased LV and aortic flow as a result of MS and ASD) (16,17). However, when a non-restrictive ASD occurs with a mild MS, the auscultatory features resemble those of an ASD. Right ventricular third and fourth heart sounds may be audible at the left sternal border and marked on inspiration (9);

(V) Murmurs and additional heart sounds: systolic murmurs (SM) may also be observed in LS (13,15) and occur due to the following reasons: (i) increased flow at pulmonic valve would lead to a systolic murmur at the upper left parasternal area from ASD; (ii) holosystolic murmur at left parasternal area from tricuspid regurgitation (TR) which may be misinterpreted as due to mitral regurgitation (MR). To differentiate, the murmur usually increases with inspiration (Carvallo sign); and (iii) Systolic murmur of TR at lower left sternal border due to RV dilation and consequent displacement of tricuspid valve (9,16). Mid diastolic murmurs may arise due to: (i) increased flow across tricuspid valve from ASD or associated tricuspid stenosis (at left lower sternal border); (ii) MS (best appreciated with the bell of stethoscope, patient coughing briskly and in left lateral position) (13,15-17). Continuous murmurs may also be appreciated in LS in which there is a restrictive ASD and severe MS usually heard at the lower right sternal area. This occurs due to the continuous shunting of blood across the small ASD (13,14);

(VI) Other features of overt RHF (pedal edema, hepatomegaly and ascites) are frequently noticed on presentation (15).

Investigations

Chest radiography

LS patients with a small or restrictive ASD have X-ray features consistent with MS. This is usually marked by significant left atrial enlargement and pulmonary venous congestion (9,10). Patients with a non-restrictive ASD and MS have prominent enlarged pulmonary artery and blood flow (pulmonary plethora) (9,16). Due to decompression of the left atrium via the large ASD, there is only mild enlargement of the left atrium. Other features are RA enlargement and RV enlargement are seen when there is already significant RV dilation from pulmonary hypertension (10,13). In more advanced disease, patients with LS may also develop mitral valve calcification (9).

Electrocardiography (ECG)

ECG features in LS vary depending on whether it's a restrictive or non-restrictive ASD. With a restrictive ASD, the ECG features are consistent with those of MS whereas in case of non-restrictive, features of ASD prevail. Overall, ECG abnormalities in patients with LS range from rhythm to P-wave and QRS morphology changes and finally axial abnormalities:

(I) With respect to rhythm, the common features identified are AF (15) and in some cases a sinus rhythm may be found (13);

(II) For P-wave morphology, a tall, broad and or bifid wave form may be seen in lead II and a deep negative force in lead V_1 suggesting bi-auricular abnormalities (13,17). Isolated left atrial abnormalities may also be observed in restrictive ASD with severe MS (13);

(III) Regarding QRS morphology changes, the following have been reported: complete or incomplete right bundle branch block (11,13,16), right ventricular hypertrophy (8,9). A right axis deviation is also observed in some cases (13,16).

Doppler echocardiography

Doppler echocardiography is the gold standard technique to establish the diagnosis of LS. It is non-invasive, available in several clinical settings, and accurate for diagnosing both ASD and MS. Also, it is important in the assessment of degree of valve regurgitation and flow, volume/pressure changes (13). At varying stages of LS, the 2-dimensional (2D) transthoracic echocardiography (TTE) determines

the following; Left atrial enlargement, enlargement of right side cavities, ASD, pulmonary artery enlargement and mitral valve stenosis. Color flow and Doppler imaging are also important in confirming and assessing the severity of ASD, mitral valve stenosis and regurgitation as well as TR and pulmonary pressure changes (18).

Assessment of MS

Echocardiography serves to confirm the diagnoses of MS, assess the severity, and characterize the valve anatomy. The following methods have been proposed; diastolic pressure gradient (DPG) method, pressure half-time method, continuity equation method, planimetry method and the proximal isovelocity surface area method (18).

The diastolic pressure gradient is estimated from the transmitral velocity flow curve using the simplified Bernoulli equation ($\Delta P=4v^2$) and has been shown to have a good correlation with measurements from trans septal catheterization, thus a reliable estimation (19). DPG is preferably estimated using continuous wave Doppler (CWD) owing to maximal recording of velocities. However, with pulse-wave Doppler, the sample volume should be placed at or just after the leaflet tips (17). In cases of severe valvular and subvalvular apparatus deformity, color Doppler is very useful to identify eccentric diastolic mitral jets. Whilst Doppler reliably assesses mitral gradient, this method has shortcomings [dependence on cardiac output, associated MR, heart rate which influence transmitral flow and also the mitral valve area (MVA)] which limit its efficacy (20).

The pressure half-time ($T_{1/2}$) method measures the interval in milliseconds between the maximum mitral gradient in early diastole and the time point where the gradient is half the maximum initial value. A reduction in diastolic transmitral blood flow velocity is inversely proportional to valve area (cm^2), and hence MVA is determined via the formula: MVA $=220/T_{1/2}$ (21). $T_{1/2}$ is then obtained by tracing the deceleration slope of the E-wave on Doppler spectral display of transmitral flow and the valve area is then automatically calculated by in-built software in most recent echo machines. In the presence of bimodal deceleration slope in early diastole, the slope is preferably traced at mid-diastole (22). This method is widely used for simplicity but is limited by the fact that it overestimates MVA (thus underestimating severity of MS), and also the presence of factors like left ventricular diastolic function, severe aortic regurgitation (AR), and degenerative calcific MS, which influence the relationship between $T_{1/2}$ and MVA (8,23-26).

The continuity equation method states that the filling volume of diastolic mitral flow is equal to aortic SV as exemplified in the following formula for MVA; MVA = $\pi(D^2/4)(VTI_{Aortic}/VTI_{Mitral})$, where D is the diameter of the left ventricular outflow tract (LVOT) and velocity time integral (VTI) is in cm (27). Some reports have suggested a good correlation with 2D TTE direct valve area measurement (28), though its accuracy and reproducibility is weak due to cummulative errors from the different measurements required to obtain MVA (18).

The proximal isovelocity surface area method is based on the hemispherical shape of the convergence of diastolic mitral flow on the atrial side of the mitral valve seen on color Doppler. Here, the MVA is estimated by dividing the obtained mitral volume flow by the maximum velocity of diastolic mitral flow on CWD (18). Here, MVA = $\pi(r^2)(V_{aliasing})\cdot$Peak $V_{mitral}^{-1}\cdot\alpha/180°$, where r is the radius of convergence hemisphere (cm), $V_{aliasing}$ is the aliasing velocity (cm/s), Peak V_{mitral} is the peak CWD velocity of the mitral inflow (cm/s) and α is the opening angle of mitral leaflets relative to flow direction. This method is however technically demanding due to the multiple measurements required (29).

Planimetry method involves a direct measurement of MVA using 2D echocardiography. It is obtained by tracing the mitral orifice and opened commissures (where applicable) on parasternal short-axis view. The measurement plane is also expected to be perpendicular to the mitral orifice with an elliptical shape (28). The recommended time of the cardiac cycle to perform planimetry is mid-diastole and is best done using the cineloop mode on a frozen image. In patients with AF and those with incomplete commissural fusion (moderate MS or after commissurotomy), several measurements are required since valve area may be influenced by changes related to flow conditions (30,31). The only limitation here even for experienced sonographers reported in about 5% of situations, is in the event of a poor acoustic window or severe valve structural malformation (especially from calcification) wherein, its estimation of valve area is relatively less accurate (32). Whilst this exists, recent advances with 3D echo and 3D-guided biplane imaging helps in optimizing the position of measurement plane (improving reproducibility) and enhances accuracy of planimetry conducted even by less experienced sonographers (33,34).

Reports have suggested the superiority of 3D TTE over 2D echography in accurate planimetric assessment of MVA and in distinguishing calcification from subvalvular

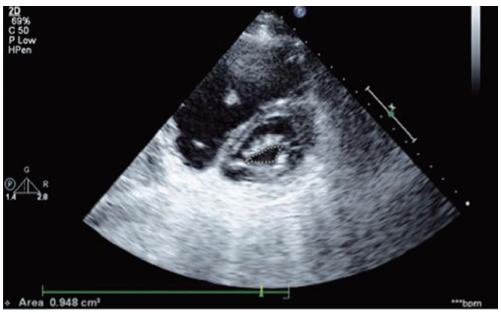

Figure 1 2D echocardiographic short axis view showing severe MS. Adapted with permission from Arora *et al*. (16). 2D, two-dimension; MS, mitral stenosis.

Table 1 Classification of MS severity (18)

Items	Mild	Moderate	Severe
Specific findings			
Valve area (cm^2)	>1.5	1.0-1.5	<1.0
Supportive findings			
Mean gradient (mmHg)	<5	5-10	>10
Pulmonary artery pressure (mmHg)	<30	30-50	>50

MS, mitral stenosis.

apparatus involvement (35). In a study comparing four echo methods in determining MVA, planimetry in practice was demonstrated to correlate best with true anatomical valve area (28). Planimetry is thus considered the best method for determination of MVA in the assessment of MS (36,37).

The normal mitral valve has an area of 4-6 cm^2 and a funnel shaped orifice favouring RV filling with no DPG. With the onset of MS, there is reduction of MVA and progressive increase in DPG (8). *Figure 1* demonstrates the severity of MS using MVA (16) and *Table 1* shows the classification of MS severity (18). Other parameters factored in the evaluation of MS include mitral valve resistance, systolic pulmonary artery pressure which reflects more the consequences of MS and not severity, though is pivotal in clinical decision making (18).

Transeosophageal echocardiography (TEE) is also helpful especially when TTE is of poor quality, for detection of associated lesions (left atrial thrombosis before balloon mitral commissurotomy or following a thromboembolic event) (38) and some authors suggest considerable accuracy of TEE for assessing valve morphology and degree of commissural fusion though this needs to be supported by more evidence (28,36). Stress or Exercise echocardiography monitors gradient and pulmonary pressures during increasing workload. It is however important in patients with symptoms discordant with severity of MS (36,37).

Assessment of ASD

The 2D TTE (*Figure 2*) is the preferred imaging technique for ASD (38). The more flexible and compliant nature of the RV over the left one favors shunting from left to right leading to RA and subsequent right ventricular dilatation. These are commonly the frequent features identified in patients with a previously unidentified ASD (as is the case in LS). TTE also characterizes the hemodynamic importance of the defect (8,13,39). In the event of severe RV volume overload, a paradoxical anterior septal motion may be noted. Color flow Doppler however demonstrates the shunt across the defect (18) while the gradient across the ASD may provide additional Doppler estimation of left atrial pressure (major determinant of clinical features) in a patient with LS (8). Secundum-type ASD require the following precise evaluation prior to percutaneous intervention with TEE: rim size and quality, sizing, exploration of septum morphology and confirmation of normal pulmonary circulation (18).

Doppler echocardiographic assessment in LS

In the evaluation of MS as seen above, plannimetry by 2D-echo is the best method for MVA estimation as exemplified by studies. This is important as MS severity influences the hemodynamics, clinical presentation and natural history of patients with LS. Color flow mapping demonstrates the regurgitation at the MS and also the shunt across the ASD. The size of ASD is crucial prior to therapeutic interventions as studies have shown that ASDs with diameters >38 mm are usually ineligible for percutaneous therapy but rather open heart surgery (40). Cognizant of the fact that most ASD may either be round, oval or crescentric, a single dimension of the ASD is not usually appropriate. In case of an oval ASD, the longest diameter may be well greater than the 38 mm cut-off, but the shorter diameter less, and such a patient still eligible for transcatheter therapy. Thus, where available, 3D-echo is important for LS patients in distinguishing calcification and subvalvular involvement in MS (*Figure 3*). It is also envisaged that with 3D echo measurements like circular index of ASD (ratio of maximal diameter to minimal diameter), indications for transcatheter therapy in LS would be reviewed (41). The role of TEE cannot be overemphasized especially in excluding other pathologies like left atrial

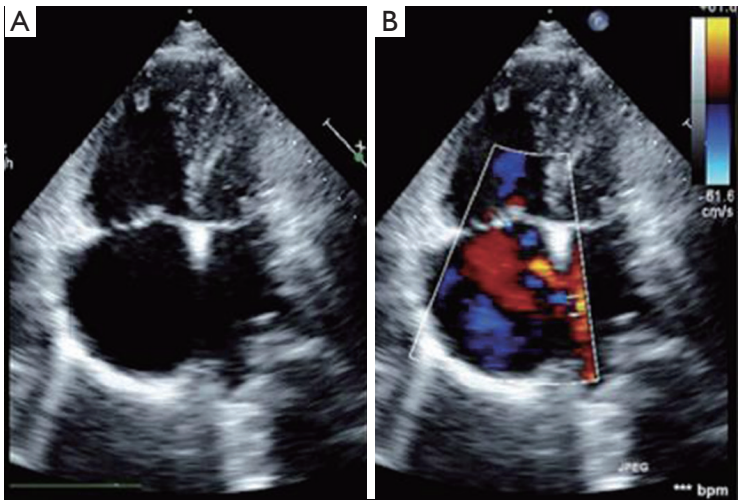

Figure 2 (A) 2D echocardiographic apical view showing a large ASD; and (B) a colour flow mapping demonstrating shunt across. Adapted with permission from Arora *et al*. (16). 2D, two-dimension; ASD, atrial septal defect.

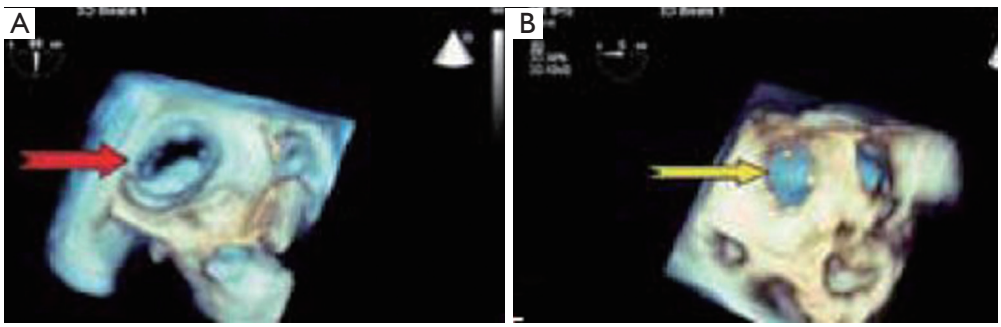

Figure 3 3D echocardiogram showing mitral valve and sub-valvular apparatus. Adapted with permission from Tezcan *et al*. (35). 3D, three dimension.

thrombi (contraindication to percutaneous therapy).

While obvious features of MS in association with enlargement of RV with an abnormal septal motion are suggestive of pulmonary hypertension and a functional TR [group 2 pulmonary hypertension: i.e., pulmonary hypertension due to left heart disease (42)] on echo, it should be considered however that, in the absence of clinical and echocardiographic features of pulmonary hypertension, a combination of MS with right ventricular volume overload, should alert the clinician to the possibility of LS (43). Basically, 2D and Doppler echo which are relatively widely available are essential and the mainstay in establishing diagnosis of LS. Prior to percutaneous interventions however; further assessment with 3D echo and TEE will be paramount.

Cardiac catheterization

Performing right heart catheterization (RHC) in patients with LS is not routinely recommended due to its invasive nature. However, it may be crucial in evaluating the severity of ASD, measuring MVA using the Gorlin formula (though it can be questionable in case of low output or just after balloon mitral commissurotomy), when echocardiography is not conclusive or is discordant with clinical findings (36,37). RHC is useful for detecting pulmonary vascular resistance, presence of reversible pulmonary hypertension and finally evaluating the presence of coronary artery disease in high risk patients (36,37).

Other investigations

Computerized tomography (CT) scan (contrast enhanced,

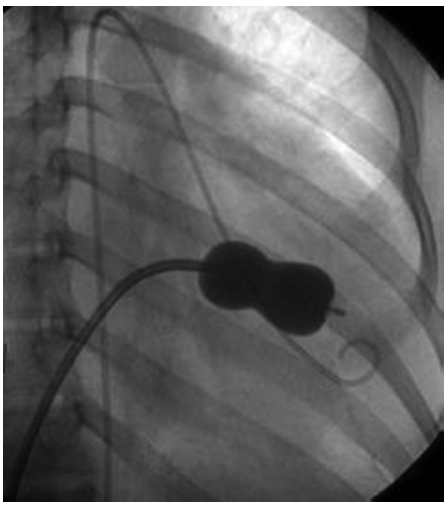

Figure 4 Inoue balloon for mitral valve dilation. Adapted with permission from Arora *et al.* (16).

3D ultrafast CT), cardiac magnetic resonance imaging (MRI) have also been used in investigating patients however, most of the necessary details they provide for LS diagnosis are obtainable by TTE (36,39). Nuclear imaging (equilibrium and first-pass radionuclide angiocardiography) has also been employed in patients with ASD and MS. However, these investigations are more expensive, require technical expertise and are also not widely available (39).

Current management of LS

Medical therapy

This involves symptomatic relief and prophylaxis for sub-acute bacterial endocarditis (SBE).

Symptomatic relief of RHF: diuretics such as furosemide are generally used to ameliorate symptoms of RHF (15).

Atrial arrhythmias: cardiac glycosides, beta blockers and calcium channel blockers may be used for rate control while drugs like amiodarone, besides rate control, also help in achieving and maintaining a normal sinus rhythm (2).

SBE prophylaxis: LS patients who have undergone repair with prosthetic device usually need antibiotic prophylaxis for the first six months following the procedure (44).

Percutaneous trans-catheter therapy

Open heart surgery had been the preferred method of treatment of patients with LS involving ASD closure and mitral commisurotomy or valve replacement (16). Recently, progress in interventional cardiology has significantly changed the treatment of LS with trans-catheter therapies (in eligible patients) with impressive success rates (16,45). The first reports of successful trans-catheter therapy was about two decades ago by Ruiz *et al.* (46) and later Joseph *et al.* (47). Since then, several successful reports have been published (10,45,48-50). Whilst several techniques have been proposed for trans-catheter therapy in LS, the most widely used today is the Inoue balloon for percutaneous balloon mitral valvuloplasty (PBMV) for mitral valvuloplasty and the Amplatzer septal occluder for percutaneous closure of ASD. The first successful combination of these techniques was reported by Chau *et al.* (49). In India, the Cocoon septal occluder is still very much in use with high success rates recorded (17,51).

Indications and contraindications for percutaneous therapy (16,17)

A number of situations in which percutaneous therapy can be conducted include: ASD with Qp/Qs ratio >1.5 with adequate rims, symptomatic moderate to severe MS with valve morphology favorable for PBMV, and any degree of pulmonary hypertension, with the exclusion of patients with Eisenmenger syndrome (irreversible pulmonary hypertension). However, the following clinical situations currently are contraindications to percutaneous therapy in LS: presence of left atrial thrombus, absence of adequate rims around the septal defect and presence of anomalous pulmonary drainage, grade 3 MR or higher, bicommissural calcification, and finally lack of expertise.

The technique

Several reports on the successful treatment of LS using the Inoue balloon (*Figure 4*) for mitral valvuloplasty and Amplatzer septal occluder (*Figure 5*) for closure of ASD, have led to their consideration as treatment of choice for LS (10). Through the femoral vein, access is made through to the right atrium, and through ASD to the left atrium, subsequently to the MS. Prior to this, anti-platelet aggregants (clopidogrel, low dose aspirin are administered) and following vascular access, 2,500-5,000 units of heparin and prophylaxis for infective endocarditis also administered (44). With the help of Mullin sheath dilator, the pig tail Inoue guide wire is inserted. Due to instability from large ASD, J-tip Amplatz extra stiff wire (ESW) is inserted through to the left ventricle. Next, Mullin sheath is then removed and Inoue balloon inserted and stabilized

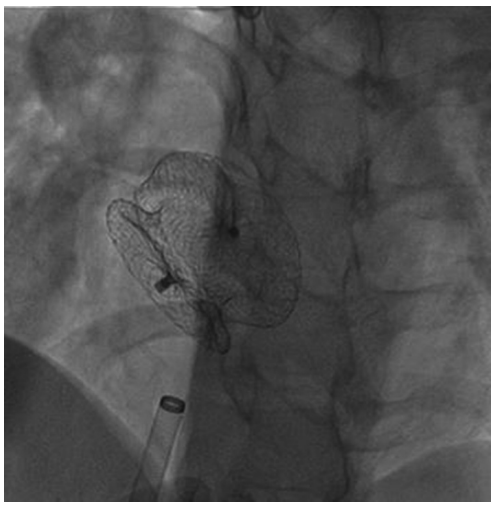

Figure 5 Amplatzer septal occluder for ASD. Adapted with permission from Arora *et al.* (16). ASD, atrial septal defect.

in the left ventricle. This is followed by inflation of balloon which dilates the stenosed valve. Upon withdrawal of the balloon, plannimetry with TTE is performed to determine the new MVA and Doppler assessment is also done for significant MR (10,51-53). Following this, the amplatzer delivery catheter is then positioned across the ASD, the left atrial disc with the centered connecting stalk is delivered. The device is then withdrawn provided that the connecting stalk is within the ASD and the left disc is adherent to atrial septum. Next, the RA disc is delivered, and the delivery wire is finally disconnected from the device, leaving the device closing the ASD. These procedures are done with fluoroscopic guidance or trans-oesophageal echo (under general anesthesia) (10,44,51). Hence in a single catheterization procedure, LS can be treated.

Challenges/complications of percutaneous therapy

Some practical difficulties which may be encountered are: (I) floating of Inoue catheter due to large ASD, hence the inability of delivery wire to go through stenosis to the left ventricle; (II) tendency to mitral restenosis: because of this, some authors advise to allow a lapse of a 24-48 hours after PBMV prior to ASD closure to give room for mitral valve assessment (51); (III) embolization or slippage of septal occluder probably due to large size ASD (52); (IV) persistent ASD after mitral valvuloplasty either due to presence of tail of balloon at atrial septum during inflation (increasing atrial septostomy) or inadequate deflation of balloon prior to withdrawing it (48).

Despite the above mentioned challenges associated with percutaneous therapy, current success rates of complete ASD closure with septal occluder during combined procedure stand at 93-97% (10) while success rates for PBMV with Inoue technique of 98.5% have been reported (54). Combined percutaneous therapy is documented to reduce morbidity and mortality following open heart surgery, decrease psychological trauma from thoracotomy scar and finally significantly reduces length of hospital stay following surgery (10).

Conclusions

LS remains a rare entity and echocardiography assessment is the current diagnostic modality of choice with 3D echo and TEE further helpful in excluding co-existent cardiac pathologies. Planimetry by Doppler echo remains the best method for assessing MVA. Whilst open heart surgery is frequently the treatment modality of choice in case of co-existent cardiac malformations, LS is currently an example of an exception, with recent percutaneous trans-catheter therapies, combining PBMV with Inoue balloon technique for MS and the Amplatzer Septal occluder for ASD closure. With most of these conclusions drawn essentially from case reports, we propose prospective multicenter registries to evaluate trans-catheter therapies and its long term outcome in patients with LS.

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