Substantive treatment of Lutembacher’s syndrome by the trans-septal puncture technique: a case report and review

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ABSTRACT
There are few reports on the primary percutaneous treatment of Lutembacher’s syndrome for which surgery has been the traditionally accepted modality of treatment. Concerns for percutaneous treatment in this entity are: (1) it is technically demanding and the dual lesion renders negotiation of catheters and balloons difficult; (2) redo valvulotomy in the presence of an atrial septal device would be difficult; (3) it may not be suitable for all patients with the condition. We describe percutaneous treatment by a hitherto undescribed technique of separate low septal puncture and review the existing literature.

INTRODUCTION
Lutembacher’s syndrome, a concomitant congenital atrial septal defect (ASD) and rheumatic mitral stenosis (MS), was first described by Lutembacher in 1916.1 Although traditionally treated by open heart surgery,2 it is amenable to percutaneous treatment. However, the reports of definitive percutaneous treatment are still anecdotal.3–11 Mitral valve (MV) interventions performed surgically have comparable rates of restenosis to percutaneous valvulotomy. Apart from the cosmetic reasons, redo surgery poses a higher risk to the patient while primary percutaneous treatment leaves an option for first surgery at a lower risk or the use of other techniques such as retrograde percutaneous valvulotomy. Here we present a case of Lutembacher’s syndrome treated percutaneously by a technique of separate low atrial septal puncture and review the cases described in the literature.

CASE REPORT
A young patient diagnosed with ostium secundum ASD (22 mm) and rheumatic MS (0.9 cm²) was taken for definitive percutaneous treatment. The patient was planned to undergo percutaneous valvulotomy followed by device closure of the ASD. The MV was pliable and suitable for percutaneous valvulotomy. The ASD was 22 mm in diameter with adequate margins. The patient was given standard premedication. The transmitral gradient was not recorded because of the presence of the large ASD. It was decided to make a low septal puncture (figure 1) (separate from the ASD) which would provide support to the shaft and necessary orientation for crossing the mitral valve.

Figure 1 Low trans-septal puncture performed separately from the atrial defect to provide the stability and necessary orientation for crossing the mitral valve.

Figure 2 J-tip guidewire being used to track the balloon into the left ventricle and subsequent inflation.
through to enter the LV. An Accura balloon (23–26 mm) was
tracked over and MV dilation was successfully achieved. The MV
area increased to 1.8 cm² with no mitral regurgitation. In the
second sitting the ostium secundum ASD was successfully closed
with a 26 mm device (Lifetech delivery system 12 F Sercare;
figure 3). The procedure was uneventful, as was the patient’s sub-
sequent hospital stay.

**DISCUSSION**

Multiple structural heart diseases in one patient influence the
natural history and the procedural success of catheter-based treat-
ment unfavourably. The percutaneous treatment of Lutembacher’s
syndrome was first described by Ruiz and colleagues in 1992 as a
palliative rescue procedure.¹² In case studies to date, the success
rate of combined percutaneous treatment is very high with no
short-term or long-term complications (table 1). It is also reported
to reduce the mortality and morbidity associated with cardiac
surgery, physiological trauma and length of hospital stay. However,
the percutaneous approach of treatment in Lutembacher’s syn-
drome has many technical difficulties, such as pushing the Inoue
balloon across the stenosed MV necessitating the use of alternative
and innovative methods. Some of the described methods (although
not specifically in relation to Lutembacher’s syndrome) include the
use of flotation balloon catheters, the reverse loop method,
catheter sliding method, over-the-wire method, modified
over-the-wire technique and reshaping the stylet. Another
important consideration is the size of the septal defect as the
defects can be large and the margins can be thin. The preferred
techniques are an optimum transesophageal echocardiographic
image or measuring the stretched balloon diameter. In patients
with Lutembacher’s syndrome in whom definitive percutaneous
repair is contemplated, a separate low septal puncture can be a
useful strategy to provide the balloon shaft with the desired stabil-
ity for easy manipulation and necessary curve so that entry to the
LV is facilitated. LV entry is facilitated by use of a J-tip exchange
length guidewire which can be used to enter the LV and subse-
quently track the balloon over it as negotiation of the coil wire
into the LV can be difficult. Apart from cosmetic reasons and
avoiding the complications of surgery, primary percutaneous
treatment keeps open the option of surgery in cases of restenosis.
In contrast, surgery offered as the primary treatment in
Lutembacher’s syndrome leaves patients with a comparable risk
of restenosis to balloon mitral valvulotomy and, in addition, the
possibility of high-risk redo surgery. On the other hand, balloon
mitral valvulotomy will be difficult in the presence of a patch in
the septum.

**Table 1 Percutaneous repair of Lutembacher’s syndrome**

<table>
<thead>
<tr>
<th>Year</th>
<th>Authors</th>
<th>Number of cases</th>
<th>Hardwire used</th>
<th>Special challenge/technique</th>
</tr>
</thead>
<tbody>
<tr>
<td>1999</td>
<td>Joseph et al¹³</td>
<td>1</td>
<td>The mitral valve was dilated using a 25 mm diameter cylindrical single balloon (Joseph mitral balloon catheter, Numed, Ontario, Canada). The ASD was closed using a 16 mm Amplatzer septal occluder (AGA Medical, Golden Valley, Minnesota, USA)</td>
<td>Modified over the wire technique, atrial septal aneurysm</td>
</tr>
<tr>
<td>2000</td>
<td>Chau et al⁴</td>
<td>1</td>
<td>The mitral valve was crossed and dilated twice with an Inoue balloon catheter set at 25 mm and 26 mm, respectively; 20 mm Amplatzer atrial septal defect occluder (AGA Medical)</td>
<td>Nil</td>
</tr>
<tr>
<td>2003</td>
<td>Ahmed et al⁵</td>
<td>1</td>
<td>Size 28 Inoue balloon catheter (Toray, Houston, Texas, USA), Amplatzer septal occluder (AGA Medical)</td>
<td>Nil</td>
</tr>
<tr>
<td>2005</td>
<td>Xiang-qian et al⁶</td>
<td>1</td>
<td>26 mm diameter cylindrical single balloon (Jin Chang Medical), 24 mm ASD occluder (Lifetech Scientific)</td>
<td>Balloon flotation catheter used to cross the mitral valve</td>
</tr>
<tr>
<td>2007</td>
<td>Ho et al⁷</td>
<td>1</td>
<td>Inoue balloon (Toray), 19 mm Amplatzer septal occlude</td>
<td>JR 6 F used for LV entry</td>
</tr>
<tr>
<td>2010</td>
<td>Özdemir et al⁸</td>
<td>1</td>
<td>Inoue balloon catheter set (Toray), 16 mm Amplatzer septal occluder device (AGA Medical)</td>
<td>Nil</td>
</tr>
<tr>
<td>2011</td>
<td>Behjatiardakani et al⁹</td>
<td>2</td>
<td>Inoue balloon, 32 mm Amplatzer septal occlude</td>
<td>Nil</td>
</tr>
<tr>
<td>2012</td>
<td>Sena et al¹⁰</td>
<td>1</td>
<td>Inoue balloon catheter 26 mm</td>
<td>Palliative BMV as a bridge to surgery</td>
</tr>
</tbody>
</table>

ASD, atrial septal defect; BMV, balloon mitral valvulotomy; LV, left ventricle.

Figure 3 Device closure of the atrial septal defect. Note the orientation of the device in relation to the septum.
CONCLUSION
Percutaneous interventional treatment of Lutembacher’s syndrome is feasible in suitable patients but can be complex, requiring alternative or modified methods for crossing the MV and accurate sizing of the septal defect. In most cases the procedure can be safely and effectively carried out with presently available techniques and should be considered as an alternative to surgical therapy. A pre-planned strategy with prior anticipation of difficulties in catheter manipulation will improve the chances of success.

Contributors All the authors were actively involved in the care and management of the patient and in the drafting, revision and final approval of the manuscript.

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