Iatrogenic Lutembacher’s Syndrome Revisited

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Lacerations or tears of the interatrial septum are a recognized complication of mitral valve replacement surgery; however, the first report of iatrogenic Lutembacher’s syndrome was by Dr. John Ross Jr. and colleagues from the National Institutes of Health. Their series included three patients who had acquired mitral stenosis and developed a persistent atrial septal defect after transatrial septal catheterization. At the time, persistent atrial septal defect was an unusual complication of transatrial septal catheterization because it had not been seen, previously, in more than 800 of these procedures performed at the National Institutes of Health. Two articles in Circulation call our attention to a new iatrogenic cause of Lutembacher’s syndrome, balloon valvuloplasty of the mitral valve by the transatrial septal approach. Originally, Lutembacher’s syndrome was the combination of congenital atrial septal defect and acquired rheumatic mitral stenosis. Subsequently, a few cases have been described with severe acquired mitral stenosis with high left atrial pressures and presumed stretching of a patent foramen ovale leading to left-to-right shunting. In the typical Lutembacher’s syndrome, the atrial-septal defect is large, usually larger than 1.5 cm in diameter. The incidence of the syndrome is approximately 4% of cases of atrial septal defect undergoing cardiac catheterization, but fewer than 1% of mitral stenosis cases. Therefore, the occurrence of mitral stenosis in patients with atrial septal defect is more than would be expected by the chance occurrence of the two conditions in the same individual. Presumably, the cases described in the articles in this issue and other reported cases of atrial septal defect after balloon mitral valvuloplasty are not because of preexistent Lutembacher’s syndrome or stretching of a patent foramen because of left atrial hypertension. It is highly likely that they represent a variant of the iatrogenic production of atrial septal defect described by Ross et al.

Balloon valvuloplasty, especially for isolated mitral stenosis in a young individual with little calcification of the valve, is becoming a popular alternative to surgical commissurotomy. Patients requiring valve replacement because of severe valve calcification or significant chordal fusion are not good candidates for balloon valvuloplasty. Currently, the most popular technique involves transatrial septal catheter advancement, which is accomplished by piercing the intra-atrial septum with a Brockenbrough needle and advancing the balloon catheter through the interatrial septum to the mitral valve. The original technique involved advancing one balloon to the mitral valve but, more recently, better hemodynamic results have been obtained by using double-balloon techniques. The positioning of two balloons can be accomplished through one or two interatrial septostomies. Studies in animals and humans have shown that atrial septostomy produces a 4–5-mm slit in the atrial septum. If two balloons, however, need to be advanced through one septostomy, the hole is usually enlarged to permit the passage of both balloon catheters in a deflated configuration. Thus, it is likely that the two-balloon technique produces a larger atrial septal defect size.

The articles in Circulation address the incidence and course of atrial septal defect after balloon mitral valvuloplasty. Each study used a different balloon valvuloplasty technique and different methods for detecting the presence of atrial septal defect. Although not strictly comparable, the reports do agree on several issues. First, atrial septal defect is common immediately after the procedure; second, the shunts are usually of small magnitude; third, they tend to get smaller or disappear with time; and finally, no major clinical sequelae of these iatrogenic shunts have been documented.

The initial incidence of atrial septal defect seems to be related to the detection method. Yoshida et al used transesophageal color flow echocardiography and detected an 87% incidence of atrial septal defect immediately after balloon mitral valvuloplasty using a single-balloon technique. Cequier et al used right heart oximetry and indicator-dilution curves to detect a 62% incidence within the first day after balloon valvuloplasty using a one-septostomy two-balloon technique. Either oximetry or indicator dilution alone, in Cequier et al’s study, resulted in only a 20–25% incidence. In Yoshida et al’s series, trans-thoracic echocardiography was only able to detect atrial septal defects in 8% of patients. Thus, it seems that transesophageal color flow echocardiography is
the most sensitive procedure for the detection of these small atrial septal defects after balloon valvu-
loplasty. Also, as Yoshida and colleagues2 point out, the color flow jet can be used to estimate the size of 
the atrial septostomy. In their study, the mean diam-
eter of the atrial puncture was 1.0 mm. This is smaller 
than the anatomic findings at operation in humans 
and at necropsy in animals but there might be a 
difference between the functional hole size and the 
anatomic defect.8 Conversely, the color flow tech-
nique might underestimate the hole size.

The reason that Yoshida et al2 demonstrated only 
a 20% incidence of left-to-right shunts across the 
atrial septum at 6 months by transesophageal echo-
cardiography, as compared with 48% in Cequier et 
al's patients3 by oximetry and dye-dilution methods, 
is probably related to the balloon valvuloplasty tech-
nique. Cequier et al3 used a one-septostomy dual 
balloon technique and might have created a larger 
hole that resolved more slowly or persisted more 
frequently after balloon valvuloplasty. Cequier et al,3 
however, also documented recurrent mitral stenosis 
in some of their patients who had persistent atrial 
septal defects, suggesting that persistent defects 
might also be related to increased left atrial pressure. 
It might be that the low incidence of persistent atrial 
septal defects in Yoshida et al's study2 was because of 
a better long-term result from balloon mitral valve-
uloplasty. Thus, there seems to be two causes of a 
persistent defect, that is, a large initial hole size and 
inadequate long-term relief of mitral stenosis.

A large initial septal hole size might be related to 
certain technical misadventures.12 First, the tail of 
the balloon might be across the atrial septum and result in 
further enlargement of the atrial septostomy during 
balloon inflations. There is some support for this pos-
sibility in Cequier et al's series3 because they noted that 
small left atria were more likely to be associated with 
persistent atrial septal defects. Another potential tech-
nical problem might be the failure to fully deflate the 
balloons before withdrawing them back through the 
atrial septum. Also, if both balloons are withdrawn at 
the same time through one hole, this may create a 
greater defect. In Cequier et al's study,3 they specifi-
cally commented that only one balloon was advanced 
across the atrial septum at a time but the withdrawal 
technique was not specified. Finally, sawing of the 
 intra-atrial septum with the guide wires has been impli-
cated. The high incidence of persistent atrial septal 
defects in Cequier et al's series3 could be related to 
technical difficulties because they used the double-
balloon one-septostomy technique, and their report 
encapsulated their initial experience with balloon 
mitral valvuloplasty at the Montreal Heart Institute. 
Thus, it is possible that further refinements in the 
technique might lessen the incidence of persistent 
defects. One solution might be the perfection of the 
retrograde approach, which avoids atrial septostomy.13

The major issue concerning atrial septal defects 
after balloon valvuloplasty is the observed or poten-
tial clinical consequences. The major concern is 
whether the creation of an atrial septal defect will 
lead to the development of pulmonary hypertension 
and the subsequent clinical problems associated with 
elevated pulmonary pressures. Pulmonary hyperten-
sion is universally seen in patients with Lutembach-
er's syndrome; however, they have large atrial septal 
defects and relatively severe mitral stenosis. Pulmo-
nary hypertension has not been observed after bal-
loon mitral valvuloplasty, probably because the atrial 
septal defects are small and the mitral stenosis has 
been relieved. Thus, this does not seem to be a major 
clinical problem. The creation of a small atrial septal 
defect can lead to a continuous murmur as reported 
by Ross and colleagues,1 which can cause confusion 
in the examination of patients after this procedure. 
This potential finding, however, has not been 
reported after balloon valvuloplasty, probably 
because the mitral stenosis is relieved. During long-
term follow-up, however, if the mitral stenosis should 
recur and a small atrial septal defect remains, then 
a continuous murmur could be appreciated. The pre-

cence of an atrial septal defect might be one of the 
reasons the Doppler assessment of mitral stenosis 
has been reported to be unreliable immediately after 
balloon valvuloplasty.14 The opportunity for blood to 
move across the atrial septum might invalidate the 
assumptions underlying the pressure half-time deter-
mination of mitral orifice size. The presence of an 
atrial septal defect of any significance could also 
delay the clinical recognition of mitral restenosis 
because it would tend to decompress the left atrium.7 
Atrial arrhythmias could potentially be caused by 
either irritation of the atrium from the laceration or 
progressive right atrial enlargement. Atrial arrhyth-
mias, however, have not been reported after atrial 
septostomy. Finally, paradoxical embolization is a 
potential complication of a persistent atrial septal 
defect; however, this also has not been reported 
during long-term follow-up. Thus, it seems that there 
are probably no clinical consequences of these small 
atrial septal defects when the mitral stenosis is 
effectively relieved. Because mitral stenosis eventu-
ally recurs after balloon valvuloplasty, there might be 
difficulties encountered in patients with persistent 
atrial septal defects.

Small atrial septal defects with pulmonary-to-
 systemic shunt ratios of less than 1.5 are common 
after balloon mitral valvuloplasty using the trans-
atrial septal approach, especially if highly sensitive 
techniques such as transesophageal color flow echo-
cardiography are used for detection. These defects 
and the associated left-to-right shunting usually 
decrease or are eliminated with time. Some patients, 
however, can have persistent shunting, which might 
be because of the size of the initial defect created or 
the unsuccessful relief of mitral stenosis. Currently, 
there do not seem to be any major clinical sequelae of 
these defects even when they persist; however, longer 
follow-up of more patients is necessary to clarify 
some of the potential clinical consequences.
References

(Circulation 1990;81:1422–1424)
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_Circulation_. 1990;81:1422-1424
doi: 10.1161/01.CIR.81.4.1422

_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1990 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/81/4/1422.citation

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