Iatrogenic Lutembacher’s Syndrome Revisited

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acereations 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.1 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 transaistral septal approach.2,3 Originally, Lutembacher’s syndrome was the combination of congenital atrial septal defect and acquired rheumatic mitral stenosis.4,5 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.6 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.7 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.1

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.8,9 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.10,11 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.12 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 al2 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 al3 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,3 resulted in only a 20–25% incidence. In Yoshida et al’s series,2 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 valvuloplasty. Also, as Yoshida and colleagues\(^2\) point out, the color flow jet can be used to estimate the size of the atrial septostomy. In their study, the mean diameter 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 technique might underestimate the hole size.

The reason that Yoshida et al\(^2\) demonstrated only a 20% incidence of left-to-right shunts across the atrial septum at 6 months by transesophageal echocardiography, as compared with 48% in Cequier et al’s patients\(^3\) by oximetry and dye-dilution methods, is probably related to the balloon valvuloplasty technique. Cequier et al\(^3\) 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 study\(^2\) was because of a better long-term result from balloon mitral valvuloplasty. 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 possibility in Cequier et al’s series\(^3\) because they noted that small left atria were more likely to be associated with persistent atrial septal defects. Another potential technical 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 specifically 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 implicated. The high incidence of persistent atrial septal defects in Cequier et al’s series\(^3\) could be related to technical difficulties because they used the double-balloon one-septostomy technique, and their report encompassed 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 potential 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 hypertension is universally seen in patients with Lutembacher’s syndrome; however, they have large atrial septal defects and relatively severe mitral stenosis. Pulmonary hypertension has not been observed after balloon 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 presence 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 determination 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 arrhythmias, 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 eventually 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 echocardiography 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


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