Introduction

Lutembacher syndrome is a combination of congenital atrial septal defect (ASD) and acquired mitral stenosis (MS), which was first described by Lutembacher, a French physician, in 1916.1 The interatrial shunt decompresses the left atrium but increases the pulmonary blood flow.2,3 Surgical correction has previously been the treatment of choice. Nowadays, both diseases are amenable to transcatheter treatment without the need for surgery. Here, we describe a 28-year-old female with Lutembacher syndrome who benefited from combined transcatheter therapy of balloon valvuloplasty for MS and device closure for ASD with an Amplatzer septal occluder. [J Chin Med Assoc 2007;70(6):253–256]

Case Report

A 28-year-old female came to Taiwan from Indonesia 6 years ago. Her first baby was born smoothly via vagina 5 years ago. She developed breathlessness on exertion, palpitation, and chest tightness 3 years previously, but paid it no attention. She was found to have a heart murmur last year, 3 months into her second pregnancy. Transthoracic echocardiography revealed moderate MS, moderate-sized ASD, and dilatation of the right ventricle and both atria. At 36 weeks of pregnancy, she developed hemoptysis and an induction was carried out in our cardiac intensive care unit, where she delivered a 1,990 g female baby. Two months after delivery, the patient was referred to the department of pediatric cardiology for transcatheter therapy. She had had several episodes of tonsillitis-associated fever in childhood but showed no signs of arthritis, chorea, subcutaneous nodules or erythema marginatum. Physical examination revealed a grade 3/6 systolic murmur over the left upper sternal border and a grade 2/6 diastolic murmur over the apex. The lungs were clear and the liver was impalpable. Exercise capacity was New York Heart Association class III. Chest radiogram showed significant cardiomegaly with increased pulmonary vascularity and a cardiothoracic ratio of 0.65 (Figure 1A). Transesophageal echocardiography showed a Wilkins score of 8 including mitral valve thickening (2), reduced leaflet motility (2), subvalvar thickening (3) and trivial calcification (1).11 The mitral valve area was 1.49 cm² using the planimetry method and 2.49 cm² using the Doppler pressure half-time method. There was trace mitral regurgitation.

Key Words: atrial septal defect, heart catheterization, Lutembacher syndrome, mitral stenosis
After informed consent was obtained, routine right and left heart catheterizations were performed. The pulmonary-to-systemic flow ratio (Qp/Qs) was 5.14 as measured by Fick’s principle. The pulmonary pressure was 56/23 mmHg with a mean of 37 mmHg. The mean pressure of the left atrium was 23 mmHg. The mean diastolic pressure gradient across the mitral valve was 16 mmHg. After a pig-wire was put in the left atrium, the partially inflated Inoue-balloon (Toray Marketing & Sales Inc., Houston, TX, USA) could not be advanced into the left ventricle due to it easily floating back to the right atrium. Therefore, we advanced a 6 Fr Judkins right catheter into the left ventricle (Figure 2A) and then placed the pig-wire in the left ventricle (Figure 2B). At that time, mild hypotension was noted, but blood pressure returned to normal soon after the Inoue-balloon catheter was advanced into the left ventricle and the pig-wire was retracted. The mitral valve was dilated sequentially with the balloon diameters set at 22, 23 and 24 mm, respectively (Figure 2C). After valvuloplasty, the mean pressure of the left atrium decreased to 11 mmHg and the mean diastolic mitral pressure gradient decreased to 4 mmHg. Rechecked Qp/Qs decreased to 2.40. The stretched diameter of the ASD as measured with a sizing plate was 18 mm. Under intracardiac echocardiographic guidance, the ASD was closed with a 19-mm Amplatzer septal occluder as detailed in our previous report (Figure 2D). Intracardiac echocardiography revealed no residual atrial shunt. Transthoracic echocardiography showed that the mitral valve area was dilated to 2.35 cm² using the planimetry method and showed mild mitral regurgitation. The patient was discharged uneventfully the following day. Chest radiogram 1 month later showed significant improvement of cardiomegaly, with a cardiothoracic ratio of 0.55 (Figure 1B). There were no complications during the 4-month follow-up, and the patient’s exercise capacity improved to New York Heart Association class II.

Discussion

MS is usually caused by rheumatic fever, which is rare in developed countries today. In America, a review paper indicated that MS is most commonly found in patients who have emigrated from areas where rheumatic fever is still endemic. It is generally believed to be an autoimmune attack of the heart in response to streptococcal infection. Medical management for patients with MS includes antibiotic prophylaxis for infective endocarditis and diuretics for mild symptoms. When MS is complicated by atrial fibrillation, rate control and anticoagulation should be started. If more than mild symptoms exist or asymptomatic pulmonary hypertension occurs, mechanical relief of MS is indicated, which includes balloon valvuloplasty, surgical valvuloplasty or valve replacement. Although the presented case had no definite past history of rheumatic fever, the MS still seemed to be rheumatic because of characteristic features shown on echocardiography and migratory history from an endemic area.
The original case of Lutembacher syndrome was a 61-year-old woman who had been pregnant 7 times. Female predominance has been noted in both ASD and MS, and thus Lutembacher syndrome has a predilection for females. The incidence of MS in patients with ASD is 4%, and conversely, ASD in patients with MS is 0.6–0.7%. “Iatrogenic” Lutembacher syndrome refers to patients with MS who have undergone balloon mitral valvuloplasty which created an ASD during the procedure. The incidence of left-to-right atrial shunts following mitral valvuloplasty is estimated at 11–12%. Most of these ASDs are small and insignificant hemodynamically.

In Lutembacher syndrome, MS augments the left-to-right shunt through the ASD while the nonrestrictive ASD decompresses the LA, reducing the diastolic mitral pressure gradient. That means the Doppler pressure half-time method would underestimate the degree of MS; this occurred in our patient. The planimetry method yields accurate estimates of mitral valve area in Lutembacher syndrome.

Concerning treatment, transcatheter closure of ASD was first reported by King et al in 1976. Since then, several devices have been developed to close ASDs. The Amplatzer septal occluder is the first and only device ever to receive full approval from the United States Food and Drug Administration for clinical use in patients with secundum ASD. Transcatheter balloon mitral valvuloplasty with transseptal technique has been in development for years. The Inoue-balloon catheter was introduced in 1984 and was declared to be the procedure of choice rather than open commissurotomy for the treatment of MS in 1998. The balloon was specially designed to be inflated in 2 stages. The distal portion of the balloon inflates first, and this allows seating of the balloon on the valve. Further inflation of the proximal portion makes an hourglass shape. This unique shape centers the balloon.

Figure 2. (A) A 6-Fr Judkins right catheter was advanced into the left ventricle. (B) A pig-wire was placed in the left ventricle. (C) The mitral valve was dilated with a 24-mm Inoue balloon. (D) Under intracardiac echocardiographic guidance (arrowhead), the atrial septal defect was closed with a 19-mm Amplatzer septal occluder (arrow).
on the valve and prevents migration when dilating the mitral valve. These features make it popular for mitral valvuloplasty.

The combined transcatheter therapy as a rescue procedure for Lutembacher syndrome was first described in 1992 by Ruiz et al, who utilized Lock’s clamshell occluder, mitral and aortic balloon valvotomies. They reported a patient with Lutembacher syndrome and pulmonary hypertension who received palliative transcatheter therapy and died before surgery. Successful combined transcatheter therapy was first described by Joseph et al in 1999. He used the Amplatzer septal occluder for ASD and Joseph mitral balloon catheter for MS. Since then, several successful cases have been reported.

To our knowledge, this is the first case of a patient with Lutembacher syndrome who received combined percutaneous therapy in Taiwan. We used the Inoue balloon antegrade through an existing ASD to dilate the stenotic mitral valve and closed the ASD with an Amplatzer septal occluder. Based on our limited experience, combined transcatheter therapy of Lutembacher syndrome is safe and effective, with a short hospital stay and without the complications of open heart surgery.

References