Percutaneous Transseptal Mitral Valvuloplasty in the Presence of Undegenerated Septum Primum

Chang-Min Chung, MD; Cheng-Wen Chiang, MD; Nye-Jan Cheng, MD; Po-Hsien Chu, MD

A 37-year-old woman had progressive shortness of breath and mitral stenosis was diagnosed. Despite the unusual finding of undegenerated septum primum on echocardiography and angiography, percutaneous transseptal mitral commissurotomy was successfully performed in this patient with rheumatic mitral stenosis under the guidance of online transesophageal echocardiography. (Circ J 2002; 66: 302–304)

Key Words: Percutaneous transseptal mitral commissurotomy; Rheumatic mitral stenosis; Septum primum

Normally the dorsocranial portion of the septum primum degenerates or disappears during fetal development! We report a case of rheumatic mitral stenosis treated by percutaneous transseptal mitral commissurotomy (PTMC) under transesophageal echocardiography (TEE) guidance in the presence of undegenerated septum primum.

Case Report

A 37-year-old woman was hospitalized for increasing dyspnea. She did not have a history of clinical systemic embolism. A diagnosis of mitral stenosis had been made 1 month earlier when she had a sudden onset of palpitation; she remained in New York Heart Association functional class II prior to receiving treatment.

On physical examination, her blood pressure was 100/52 mmHg and her heart rate was regular at 75 beats/min. No jugular venous distension was noted and both lung fields were clear. Cardiac auscultation revealed a loud first heart sound, an opening snap, and a grade 2 diastolic rumbling murmur. No systolic murmurs were heard. Her electrocardiogram revealed normal sinus rhythm, and her chest X-ray showed left atrial enlargement. Two-dimensional echocardiography demonstrated mitral stenosis with a mitral valve area of 1.2 cm² by planimetry² and of 1.5 cm² by the pressure half-time Doppler method³. Using the Wilkins score system⁴ mitral valve calcification was graded as 1, leaflet mobility as 2, leaflet thickening as 2, and subvalvular involvement as 2. The end systolic anteroposterior diameter of the left atrium was 40 mm. Color Doppler detected mild mitral regurgitation, and mild tricuspid regurgitation was also recorded with a peak velocity of 2.2 m/s. An extraneous linear echo continuous with the septum primum in the fossa ovale and coursing to the dorsocranial wall of the left atrium was noted (Fig 1). It created small extraneous chambers in the left atrium. Color Doppler signal was not used during the TEE examination and did not show any color filling in the extraneous chambers.

After discussing the therapeutic options with the patient, percutaneous transseptal balloon mitral valvuloplasty was performed. A transseptal puncture site in the fossa ovalis was selected under online TEE guidance. The puncture needle was carefully steered so as to keep away from the undegenerated portion of the septum primum. After septal puncture, a spring-tip guide wire was coiled into the left atrium, a 26-mm Inoue balloon catheter was advanced into the left atrium along the spring-tip guide wire and balloon inflation was initiated at 22 mm. Successful, uncomplicated stepwise dilatation of the mitral valve in 0.5 cm³ increments was performed until a final 28 mm balloon size was reached. The mitral valve area, as determined by the Gorlin formula, increased from 1.1 cm² to 2.3 cm², without any increase in the severity of mitral regurgitation or left-to-right shunting, as demonstrated by oximetry after the procedure. There was no gradient between the pulmonary veins and the left atrium before or after the procedure.

After PTMC, right upper pulmonary vein (RUPV) cineangiography demonstrated a filling defect and a bizarre-shaped dorsocranial portion of the left atrium. No extraneous small chambers could be visualized, suggesting that there was not communication between the main left atrial cavity and the extraneous chambers (Fig 2). However, additional findings derived from dynamic 3-dimensional echocardiography with the use of a contrast agent or color Doppler signal would have outlined it more clearly.

Discussion

During fetal development, partitioning of the atrioventricular canal and the primitive atrium and ventricle begins around the middle of the fourth week of gestation! The septum primum, a thin, crescent-shaped membrane, grows toward the fusing endocardial cushions from the dorsocranial wall, or roof, of the primitive atrium. Before the foramen primum is obliterated, perforations that appear in the dorsal part of the septum primum coalesce to form another opening, the foramen secundum. Concurrently, the free edge of the septum primum fuses with the left side of the fused endocardial cushions, obliterating the foramen
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Circulation Journal Vol.66, March 2002

The cranial part of the septum primum, initially attached to the roof of the left atrium, gradually disappears or forms the abnormality of degenerating septum primum (Fig 3).1,5,6

To the best of our knowledge, finding an undegenerated septum primum on echocardiography has not been reported and even pathology textbooks have rarely described it in detail. We found that it was continuous with the septum primum in the fossa ovale and coursing to the dorsocranial wall of the left atrium. (B) Simplification of (A) showing the relationship of the fossa ovale, extraneous chamber, and undegenerated septum primum. (C) Three-dimensional echocardiogram from the left atrium toward the right atrium: no extraneous chambers can be seen, suggesting that there is no communication between the extraneous chambers and the main atrial cavity. LA, left atrium; RA, right atrium.

Performing PTMC in the presence of an undegenerated septum primum has the potential of being problematic. If it is penetrated during septal puncture it may interfere with the steering of the balloon catheter thus making the procedure more difficult. Furthermore, thrombi may develop within the extraneous chamber, predisposing to embolism. In this context, online TEE guidance can monitor the whole procedure and prevent inadvertent puncture of the undegenerated portion of the septum primum.

References


