

Echocardiographic assessment of Lutembacher syndrome

Ocena echokardiograficzna w zespole Lutembachera

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A 59-year-old woman was a candidate for lumbar disc hernia surgery. She was complaining of shortness of breath on mild exertion. Preoperative cardiac assessment was performed. On physical examination, her blood pressure was 110/60 mm Hg and heart rate was 75 bpm. A grade II/VI holosystolic murmur and a diastolic rumble were heard at the cardiac apex. Electrocardiography (ECG) showed normal sinus rhythm with normal axis (Fig. 1A). Chest X-ray showed

prominent hilar vasculature (Fig. 1B — red arrow), right atrial enlargement (Fig. 1B — black arrow), and absence of left atrial enlargement (Fig. 1B). Evaluation by two-dimensional (2D) transthoracic echocardiography (TTE) revealed coexistence of ostium secundum atrial septal defect (ASD) and rheumatic mitral stenosis. Mitral valve leaflets appeared thickened and restricted in motion but calcification and subvalvular involvement could not be defined exactly (Figs. 2A, B). Planimetric mitral valve area (MVA) was 2.1 cm², maximum diastolic gradient was 9 mm Hg, and mean gradient was 3 mm Hg. Also there was a mild-to-moderate mitral regurgitation. The diameter of ASD was estimated to be 2.2 cm² and colour flow mapping revealed left-to-right shunt across the defect (Figs. 2C, D).

Subsequently, 3D TTE study was performed in order to achieve more accurate assessment of mitral valve apparatus. By 3D echocardiography (Figs. 2E, F), planimetric MVA was 1.7 cm² and subvalvular thickening including calcification was seen, indicating that limitations of 2D echocardiography should be recognised and determination of planimetric MVA should be performed accurately by 3D echocardiography before treatment. Soon after, the patient was referred for open-heart surgery. Lutembacher syndrome is a rare combination of congenital ASD and acquired mitral stenosis. Due to the limitations of 2D echocardiography, particularly for distinguishing calcification and subvalvular involvement and for accurate determination of planimetric MVA, mitral valve apparatus should be assessed by 3D echocardiography before treatment.

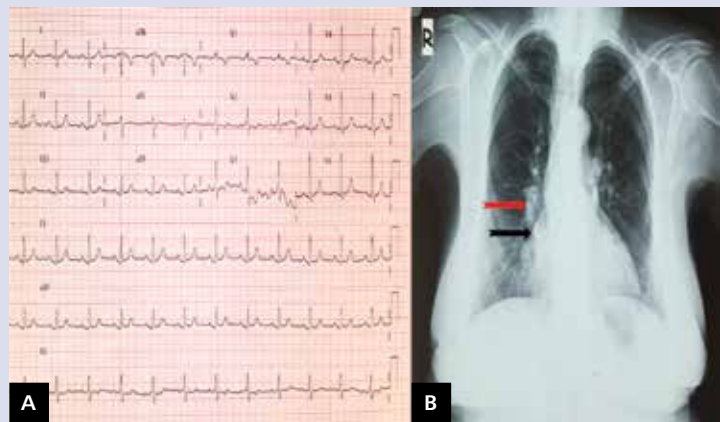


Figure 1. ECG (A) and chest X-ray (B)

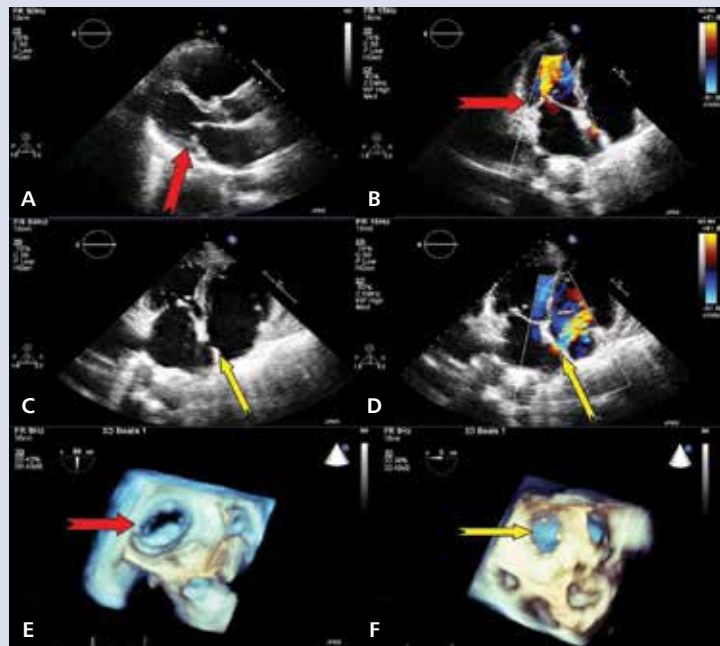


Figure 2. A-F. 2D/3D TTEs

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