# A giant left atrium in a patient with Barlow syndrome, abnormal chordae tendineae, and perforation of the anterior mitral leaflet

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An 80-year-old man was admitted to our hospital due to weakness and dyspnea at rest. His medical history included dilated cardiomyopathy, mitral regurgitation (MR), and persistent atrial fibrillation diagnosed 10 years earlier. Physical examination revealed signs of pulmonary congestion, massive bilateral peripheral edema, irregular pulse wave, and systolic grade 3/6 murmur at the apex, radiating to the parasternal region. Marked cardiomegaly and right-sided pleural effusion were seen on chest X-ray (FIGURE 1A). Transthoracic echocardiography showed a giant left atrium (LA) of 13 cm in diameter in the parasternal long-axis view and severe MR. Both leaflets of the mitral valve were thickened and prolapsed into the LA (FIGURE 1B). The medial papillary muscle could not be visualized. Due to suspicion of an abnormal mitral valve apparatus, we performed cardiac magnetic resonance imaging (MRI; Symphony 1.5T, Siemens, Erlangen, Germany) and 3-dimensional transesophageal echocardiography (TEE; Lisendo 880, Hitachi, Tokyo, Japan).

Cardiac MRI revealed a huge LA (195 × 105 × 155 mm), with a volume of 3100 ml (FIGURE 1C). The left ventricle was also enlarged (ejection fraction, 55%), and MR was severe. On 3-dimensional TEE, the mitral annulus was dilated to 55 mm. The lateral papillary muscle exhibited numerous chordae tendineae to both leaflets, and the medial papillary muscle had 2 heads with 1 chorda tendinea to the ventricular septum, 1 to the lateral papillary muscle, and 3 chordae tendineae to the anterior leaflet. Both leaflets were thickened and prolapsing. The anterior leaflet was perforated in the A1 segment (FIGURE 1D).

The case was discussed by a multidisciplinary team. Although mitral valve repair or replacement is considered the gold standard for patients with severe MR,1,2 the patient refused surgery. Thus, we applied standard pharmacotherapy for heart failure that included ramipril, spironolactone, furosemide, carvedilol, and warfarin. Moreover, we performed thoracentesis and daily weight measurements. After 4-month follow-up, the patient was diagnosed with New York Heart Association class II heart failure.

Barlow syndrome is characterized by bilaterally prolapsed or billowing leaflets, elongated chordae tendineae, and annular dilation. In some cases, the lack of leaflet coaptation leads to MR and subsequent LA enlargement. However, its exact etiology is still unknown.3 An LA larger than 8 cm in the long-axis view is considered giant.

We present this case with a giant LA, as its volume on cardiac MRI was estimated at 3100 ml. In the literature, we found only 1 case report of a similar LA volume, which was published at the beginning of the 20th century.4 Abnormal chordae tendineae are also rarely described.5 In our case, 2 chordae tendineae of the medial papillary muscle were abnormally attached, but paradoxically it could improve mitral valve function.

In most cases, transthoracic echocardiography remains the first-choice investigation for Barlow syndrome. However, its usefulness is limited in some patients, and 3-dimensional TEE plays a key role in their evaluation. Alternatively, cardiac MRI can be used to study the morphology

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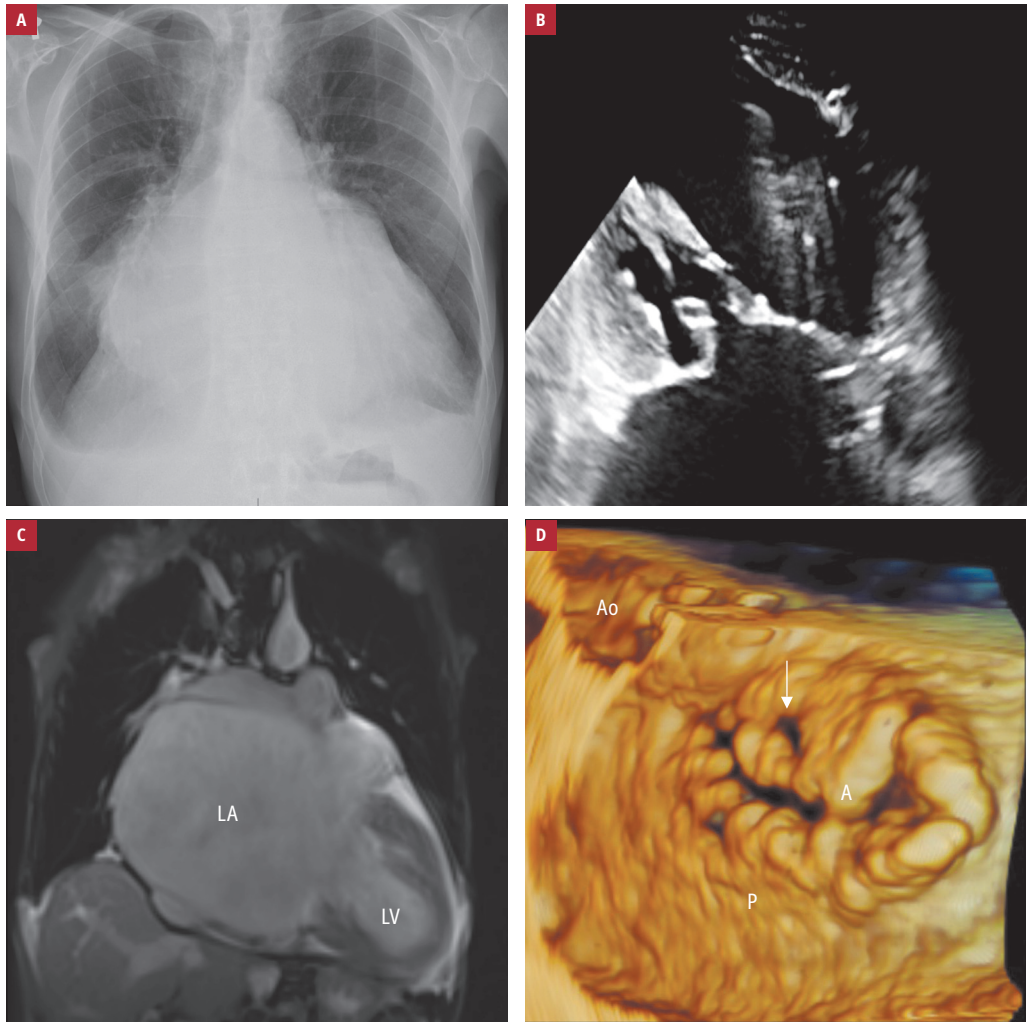


**FIGURE 1** Imaging of a giant left atrium: **A** – chest X-ray; **B** – transthoracic echocardiography; **C** – cardiac magnetic resonance imaging; **D** – 3-dimensional transesophageal echocardiography showing the perforation (arrow)
Abbreviations: A, anterior leaflet; Ao, aorta; LA, left atrium; LV, left ventricle; P, posterior leaflet

and function of the mitral apparatus. In the presented case, clinically relevant findings were evaluated using transthoracic echocardiography, 3-dimensional TEE, and cardiac MRI. Multimodal imaging allowed us to precisely diagnose our patient with a giant LA.

## ARTICLE INFORMATION

**CONFLICT OF INTEREST** None declared.



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