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Biatrial enlargement: 6,000 ml intracardiac blood volume

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A 42-year-old man (height 202 cm, body weight 120 kg) was admitted to our intensive care unit in cardiogenic shock and acute respiratory failure. Nine years before, the patient had undergone mitral valve replacement with a

mechanical prosthesis because of severe degenerative mitral valve regurgitation in another hospital. Recent pulmonary function indicated a restrictive ventilation disorder and ECG showed permanent atrial fibrillation. Chest radiography and CT scan revealed an impressive cardiomegaly (Fig. 1). Transthoracic echocardiography showed a massive biatrial enlargement (Fig. 2e). Transesophageal echocardiography confirmed the extreme dimensions of both atria with spontaneous echo contrast (Fig. 2a–d).

The CT scan had to be performed in the prone position, as the patient no longer tolerated a supine position because of compression of the airways. This scan revealed a grotesque biatrial enlargement. The calculated intracardiac blood volume corresponded to 6,060 ml (Fig. 1d). Surgical reduction of both atria was indicated for cardiac and pulmonary reasons (Fig. 3a, b) and resulted in improved respiratory function and hemodynamics (Fig. 4).

Etiology of the disease remained uncertain since previous genetic analyses showed no abnormalities for fibrillin 1 (FBN1) nor transforming growth factor β -I/II (TGF- β -I/II).

The patient was informed and gave his written consent to the publication of this case.

Conflicts of interest The authors declare that they have no competing interests.

Fig. 1 **a** CT scout image showing massive cardiomegaly. **b** Left and right atrium dominating the thoracic cavity with diameters of 20 cm and 25 cm, respectively. Mitral valve prosthesis shows the entrance to the normally dimensioned left ventricle (arrow). **c** Thoracic computed tomography demonstrates both left and right atrial enlargement, dilated coronary arteries, and compressed main bronchus. **d** Software-based segmentation of the four heart chambers using multiplanar reformation, MPR (*F, A, L*) and volume rendering technique (VRT) for comprehensive 3D visualization (*A, L*) and volumetry (left atrium, 3,598 ml; right atrium, 2,109 ml; left ventricle, 98 ml; right ventricle, 255 ml)

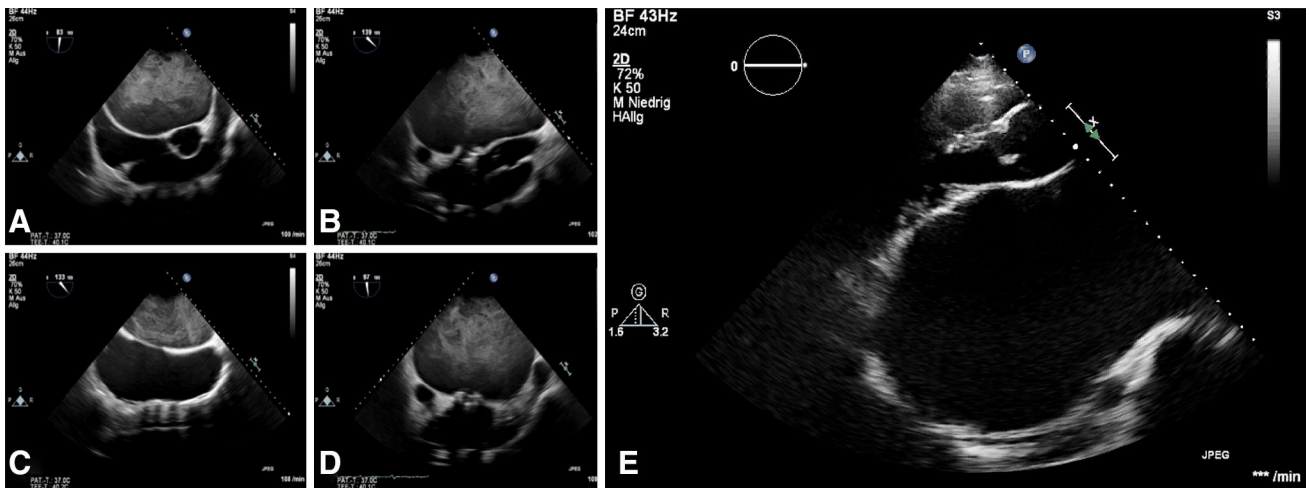
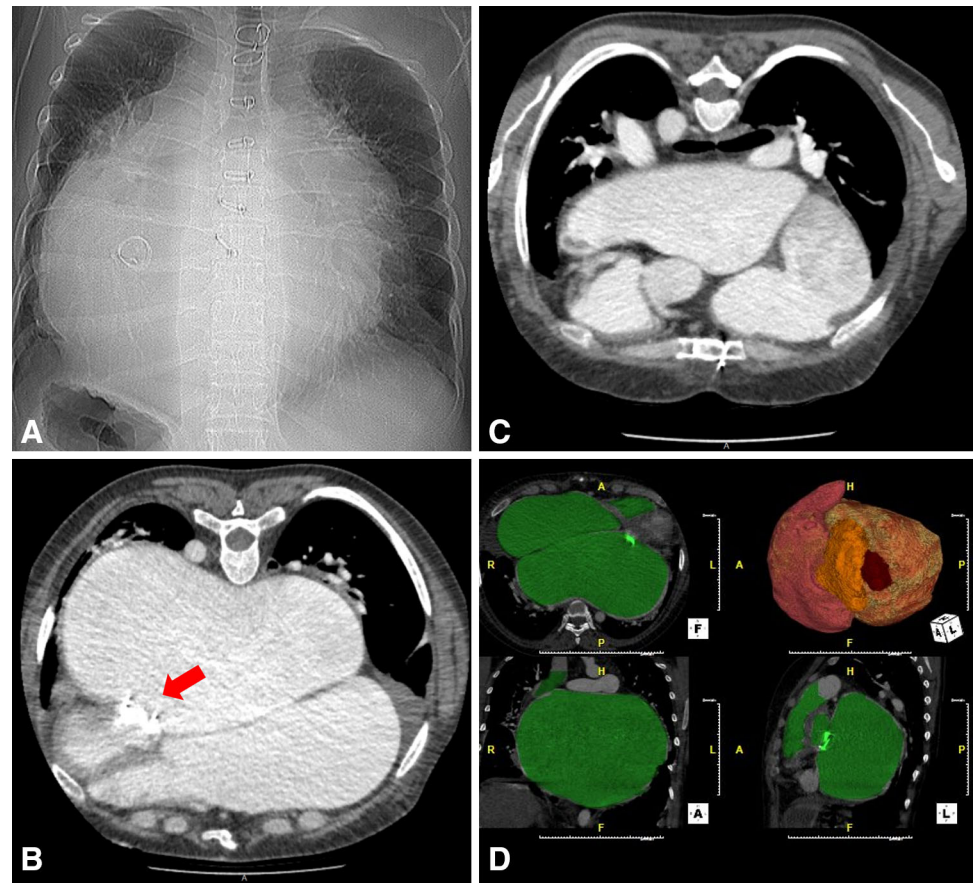


Fig. 2 Transesophageal echocardiography modified midesophageal long axis view (**a**) at 83° dilated left and right atrium, (**b**) at 139° dilated left atrium, mechanical mitral valve and aortic root, (**c**) at 133° parts of the left atrium and dilated right atrium, (**d**) at 97° left

atrium with implanted mechanical mitral valve. Transthoracic echocardiography (**e**) parasternal long axis view with massive dilated left atrium with a maximum diameter of 13 cm

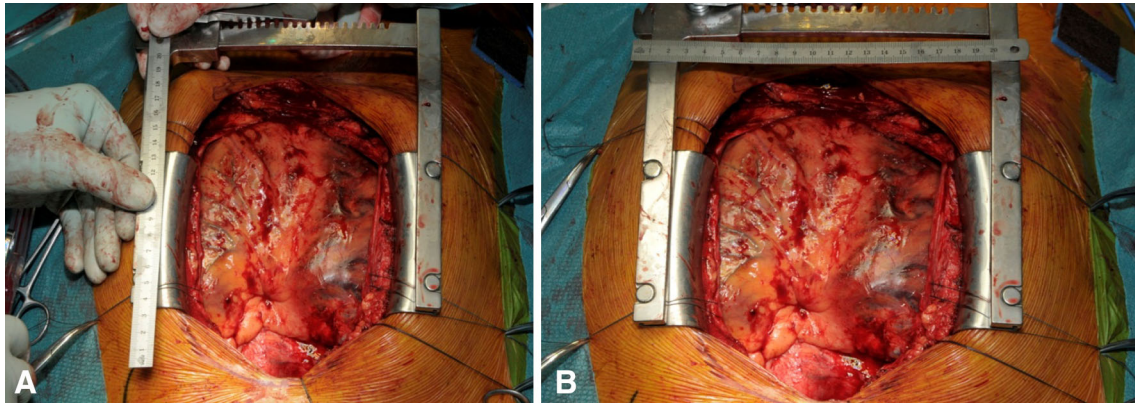


Fig. 3 a, b Intraoperative view after opening of the pericardium. The retrosternal space is dominated by the gigantic right atrium with sternal retractor maximally open



Fig. 4 a Preoperative conventional chest radiography showed cardiothoracic ratio of 0.86. **b** Postoperative imaging shows regular dimensions in the computed tomography with mechanical mitral prosthesis in place and **c** improved cardiothoracic ratio of 0.57 in the conventional chest radiography