Malaise and fatigue following mitral valve repair: case report

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Introduction
Mitral valve regurgitation is the second most common valvular heart disease. In primary, degenerative mitral regurgitation (MR), valve repair is the preferred treatment option.

Case presentation
We present a case of a 73-year-old man who was admitted to our cardiology department with progressively worsening shortness of breath (New York Heart Association-Classification IV) and fatigue 2 months after surgical mitral valve repair for MR. Transthoracic and transoesophageal echocardiography showed a remaining severe MR and mitral valve stenosis II. Laboratory results showed an extra-corpuscular, mechanical, and haemolytic anaemia. After exclusion of other causes of haemolytic anaemia and the lack of clinical and laboratory improvement, the patient underwent valve replacement with a biological valve. Haemolysis parameter normalized and the clinical status improved.

Discussion
Although haemolysis after mechanical prosthetic mitral valve replacement is frequently recognized, haemolytic anaemia after mitral valve reconstruction is still an underestimated complication, and there are only a small number of reported cases. This case demonstrates the clinical diagnostic steps for excluding other causes of haemolytic anaemia after mitral valve repair in patients with a history of heart surgery.

Keywords
Case report • Haemolysis • Mitral valve repair • Mitral regurgitation • Annuloplasty

Learning points
• It is important to include the diagnosis of haemolytic anaemia after mitral valve repair in patients with a history of heart surgery.
• Mechanical haemolysis rarely resolves spontaneously.
• In most valvular cases, patients will have to undergo valve replacement.

Introduction
Haemolysis after prosthetic mitral valve replacement is frequently recognized, but represents a rare complication following mitral valve repair. In this case report, we describe a patient who developed severe mechanical haemolysis 2 month after mitral valve repair.

Case presentation
A 73-year-old man was admitted to our cardiology department with progressively worsening shortness of breath (New York Heart Association-Classification IV) and fatigue 2 months after surgical mitral valve repair for mitral regurgitation (MR). His recovery had been prolonged due to a haemothorax requiring re-thoracotomy in combination with severe enterococcal sepsis treated with Vancomycin for 2 weeks and a right frontal watershed stroke with a residual paresis of the left arm. The patient’s medical history was significant for hypertension, atrial fibrillation, and coronary artery disease, for which he had undergone coronary artery bypass graft surgery in the context of mitral valve repair in November 2016.
Upon arrival, he had pale conjunctivae and a holosystolic 3/6 apical murmur radiating to the axilla. Mild bibasilar crackles were audible, and there were bilateral lower extremity oedema. Blood pressure was 130/80 mmHg, heart rate was 80 beats per minute, temperature and there were bilateral lower extremity oedema. Blood pressure was 36.6/130 mmHg with a palpable holosystolic 3/6 apical murmur radiating to the axilla. Mild bibasilar crackles were audible, and there were bilateral lower extremity oedema. Blood pressure was 130/80 mmHg, heart rate was 80 beats per minute, temperature was 36.6°C, and chest X-ray showed a slight pleural effusion of the left lung without infiltrates.

The patient’s blood test showed haemoglobin and haptoglobin reduced and lactate dehydrogenase increased as hallmarks of an intravascular extracorpuscular anaemia (Table 1). The haemoglobin level was 6.6 g/dL (normal range 13.6–17.2 g/dL), haematocrit was 0.20 (normal range 0.39–0.49), and reticulocyte count was elevated. Haptoglobin was reduced to 0.01 g/L (normal range 0.5–2.4 g/L), total bilirubin was 1.4 mg/dL (normal range 0.3–1.2 mg/dL) with an indirect hyperbilirubin of 0.4 mg/dL (normal range <0.2 mg/dL), and lactate dehydrogenase level was increased to 1382 U/L (normal range 100–247 units per litre). Brain natriuretic peptide levels were increased to 374.2 pg/mL (normal range <100 pg/mL). Creatinine and cardiac enzyme levels were within normal limits. Given that mechanical haemolysis is rarely observed in patients treated with valve repair, it was essential to exclude other potential causes of haemolysis. The most important differential diagnosis was autoimmune haemolytic anaemia with antibodies against erythrocyte structural components. Autoimmune haemolytic anaemia may develop after severe infections or post-surgery. To exclude autoimmune haemolytic anaemia, a coombs test was performed as well as a specific coombs test and cold agglutination, which were negative, thus supporting the diagnosis of surgery related haemolysis.

Transthoracic and transoesophageal echocardiography revealed a high-grade mitral valve regurgitation and mitral valve stenosis II (Figure 1A, B. Supplementary material online, Videos S1 and S2). Mean pressure gradient was 7 mmHg with an eccentric jet. Pulmonary hypertension and second degree tricuspid valve regurgitation were detected.

The patient developed recurrent transfusion-requiring conditions. At four occasions, a total of eight erythrocyte concentrates were transfused. After reaching a satisfactory general state of health, the patient was referred to valve replacement with a 27 mm biological prosthesis (Sorin Pericarbon) and tricuspid repair with annuloplasty ring implantation (30 mm Contour 3D Medtronic). The histopathological examination revealed a myxoid mitral valve degeneration.

Haemolysis parameters normalized and the clinical status improved quickly. Within 3 months, all parameters were within normal ranges. Upon his most recent follow-up visitation, 9 month after valve replacement, the patient reported a significantly improved exercise capacity. Control echocardiography showed a good result after mitral valve replacement without residual regurgitation (Figure 1C).

### Discussion

Mitral regurgitation is the second most common valvular heart disease and can be subdivided in a primary, degenerative form and a secondary mitral valve regurgitation due to ischaemia and dilatation. In primary MR, valve repair is the preferred treatment option. Valve repair provides lower impairment of the left ventricular heart function, lower complication rates, and operative mortality, better long-term results and, no need for anticoagulation. The surgical steps include annuloplasty, shortening of tendinous chords in combination with prolapse and leaflet resection. Patients developing haemolysis after mitral valve repair generally present within the first 2–6 months after surgery. Possible mechanisms of haemolysis after valvular repair are fragmentation of the regurgitant jet by a annuloplasty ring dehiscence, collision of the regurgitant jet into the prosthetic ring, and rapid acceleration of a jet through a small para-ring leakage. The regurgitant jet itself perpetuates haemolysis by delaying endothelialization of the valvuloplasty ring, leading to further fragmentation and collision. In approximately 35%
of all patients, haemolysis is caused by incomplete initial repair or technical error and has been described even in cases of mild MR.\textsuperscript{8}

Timely echocardiographic evaluation is important to diagnose the pathology and may lead to re-repair or replacement of the valve. Guideline recommendations concerning the treatment of haemolysis after mitral valve repair are currently not available, but mechanical haemolysis rarely resolves spontaneously. In most cases, patients will have to undergo valve replacement. This clinical case shows the importance of including the diagnosis of haemolytic anaemia after mitral valve repair in patients with a history of heart surgery.

**Supplementary material**

Supplementary material is available at European Heart Journal - Case Reports online.

**Consent:** The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

**Conflict of interest:** none declared.

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**Figure 1** (A, B) 2D and 3D transoesophageal echocardiography showed incomplete repair after mitral valve surgery with an eccentric jet (arrow). (C) Post-operative transoesophageal echocardiography with a good result after mitral valve replacement without regurgitation.