

## Brief Report

# Mitral perforation: a rare cause of congenital mitral regurgitation

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**Abstract** In a 4-year-old boy with severe mitral regurgitation, cross sectional echocardiography combined with Doppler interrogation confirmed the presence of isolated perforation of the aortic leaflet of the mitral valve. The perforation was closed with a patch of fresh autologous pericardium. Serial echocardiograms taken postoperatively showed no regurgitation across the mitral valve.

**Keywords:** Congenital cardiac disease; echocardiography; mitral incompetence

**C**ONGENITAL MITRAL REGURGITATION IS VERY rare in children.<sup>1,2</sup> We report here a child who had mitral regurgitation caused by a hole in the aortic leaflet of the mitral valve that was diagnosed using cross sectional and color Doppler echocardiography and repaired surgically.

### Case report

A 4-year-old boy was referred to our outpatient clinic for investigation of a cardiac murmur. The history failed to reveal any evidence of fever, fatigue, loss of appetite, arthritis, or any significant recent surgical procedure. On admission, he weighed 15 kg, on the 25th centile for his age. His temperature was normal, there were no stigmas of Marfan or Ehler Danlos syndrome or metabolic disease, and he was neither tachypneic nor tachycardic. The cardiac pulsation was prominent, and a systolic regurgitant murmur of grade 3 out of 6, was audible at the apex, with transmission to the left axilla. Electrocardiographic tracings were normal, but the chest X-ray showed cardiomegaly, with a cardio thoracic ratio of 0.55, involving the left ventricle. Echocardiographic examination revealed

dilation of the left atrium and left ventricle. An isolated perforation, measuring 0.5 by 0.7 mm, was found in the aortic leaflet of the mitral valve, with Doppler interrogation revealing significant regurgitation through the hole (Fig. 1). The cordal anatomy, and the arrangement of the papillary muscles, were normal. Other laboratory tests for acute phase reactants, and the titre of antistreptococcal antibodies, were also normal.

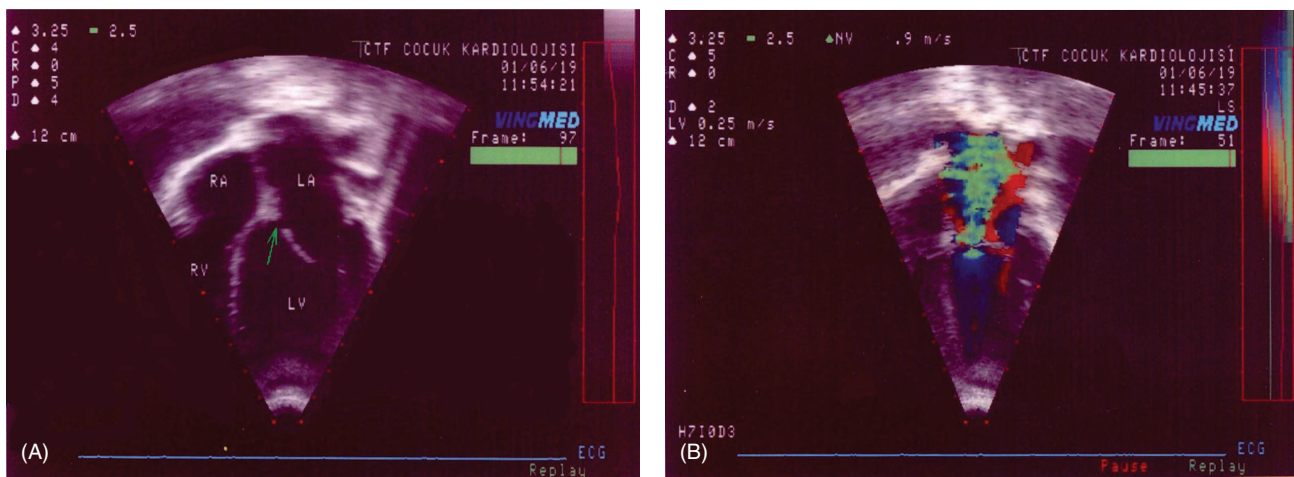
Surgery was performed via a median sternotomy, using cardiopulmonary bypass, and the defect was closed with a patch of fresh autologous pericardium (Fig. 2). After weaning from bypass, the valve was found to be competent, and there was no need either for annuloplasty or to use an annular ring. His postoperative course was uneventful. Serial echocardiograms in the first postoperative days, and eight months later, showed no regurgitation across the mitral valve.

### Discussion

Isolate congenital mitral insufficiency is an extremely uncommon cardiac condition. Mitral insufficiency is often found in association with other congenital cardiac defects, connective tissue disorders, metabolic or storage diseases. Secondary or acquired mitral regurgitation may be seen following trauma, myocarditis, rheumatic fever, endocarditis, Kawasaki disease, or collagen vascular disorders.<sup>1,2</sup>

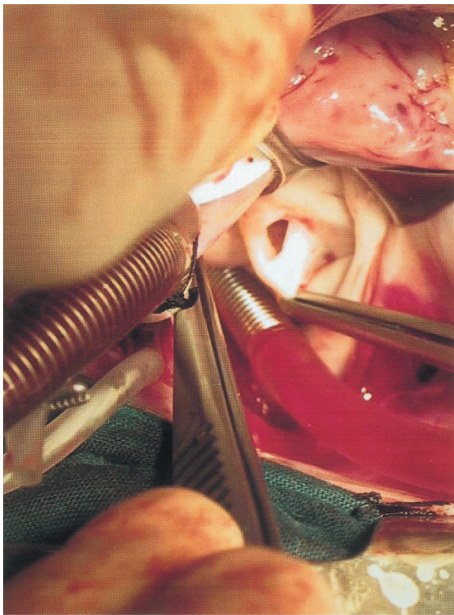
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**Figure 1.**

The apical four chamber echocardiographic section (A) shows a perforation in the aortic leaflet of the mitral valve away from the line of coaptation (arrow). Color Doppler interrogation (B) shows severe regurgitation through the perforation. RA: right atrium; LA: left atrium; RV: right ventricle; LV: left ventricle.



**Figure 2.**

Intraoperative view of the perforation in the aortic leaflet of the mitral valve.

The congenital causes of mitral valvar insufficiency are reported as prolapse, double orifice, isolated clefting, severely hypoplastic leaflets, the hammock valve, multiple leaflets, a long and billowing aortic leaflet with thin and elongated tendinous cords, and the Ebstein like malformation.<sup>1–8</sup> In our patient, we discovered none of these lesions, nor did we find any history of trauma, endocarditis, or rheumatic fever. Normal laboratory test ruled out the presence of an inflammatory process. The echocardiographic

investigations revealed a defect with regular margins in the aortic leaflet of the mitral valve in the setting of normal cordal anatomy. The aetiology of the perforation remains unknown, but we believe it is probably congenital.

Review of the literature revealed only one case previously reported with a congenital perforation of the mitral valve, the diagnosis being made at surgery.<sup>9</sup> In our case, of course, the diagnosis was made preoperatively, encouraging us to proceed to a corrective operation. The operative results were quite pleasing. Thus, to the best of our knowledge, our patient is the only one with a congenital perforation of the mitral valve diagnosed preoperatively.

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