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*Brief Report*

# Mitral valvar regurgitation in a child with Sweet's syndrome

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**Abstract** We report the very unusual perforation of the mitral valve in the setting of Sweet's syndrome, or acute febrile neutrophilic dermatosis, in a boy aged 5 years. Surgical repair was uneventful, and follow-up showed no residual anomalies. Acute or delayed valvitis, with damage to either the mitral or aortic valves, should be screened for in this rare disease.

Keywords: Neutrophilic dermatosis; valvitis

In developing countries, acquired mitral regurgitation in childhood is mostly related to rheumatic diseases. Isolated congenital mitral regurgitation is rare, and is usually due to malformations of the valvar apparatus, typically a cleft leaflet. Infective endocarditis may cause also cause damage to the valve. Here, we report an unusual instance of perforation of the aortic leaflet of the mitral valve in a child with Sweet's syndrome, also known as acute febrile neutrophilic dermatosis.

## Case report

A 5-year-old boy was referred to our institution because of significant mitral regurgitation. He was diagnosed to have Sweet's syndrome, which is acute febrile neutrophilic dermatosis, when he was two years old. Since the age of 4, he was asymptomatic and took no medication. Physical examination was normal except for an intense holosystolic murmur at the apex.

Echocardiography showed a dilated left atrium and left ventricle, with left ventricular end-diastolic diameter of 44 millimetres. Important regurgitation was found across the valve, and was shown to be due to a perforation of 3 millimetres in the aortic leaflet of the valve, located 5 millimetres below the hinge of the aortic valvar leaflets. The leaflets of the mitral valve themselves were thin, with no prolapse, nor was there any clefting of the leaflets. The tendinous cords and papillary muscles were normal. No vegetation was seen. The aortic, tricuspid and pulmonary valves were all normal, and no additional anomalies were found (Fig. 1).

We thought that the mitral regurgitation would worsen with time and lead to thickening of the borders of the perforation. We decided, therefore, to recommend surgical repair. The perforation was closed with a pericardial patch under cardiopulmonary bypass. The borders of the perforation were already thickened, and the surgeon also repaired a ruptured tendinous cord from the mural leaflet. No lesion was seen that would have favoured previous but burnt-out endocarditis. The postoperative period was eventless, and patient could be discharged at home on the eight day. Echocardiography after one year showed no residual mitral regurgitation and normal left ventricular function.

## Discussion

Mitral perforation is a well known complication of mitral endocarditis.[1,2] The occurrence in children, in contrast, of congenital perforation or non infectious acquired perforation is very rare.[3] Acute febrile neutrophilic dermatosis, or Sweet's syndrome, is a rare disease in infancy Associated conditions include malignancies, infections, drug reactions, and autoimmune diseases. In our patient, investigations prior to the diagnosis of mitral valvar perforation, and during

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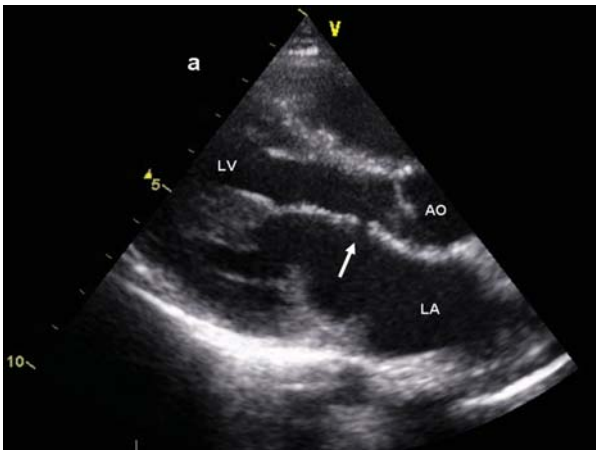
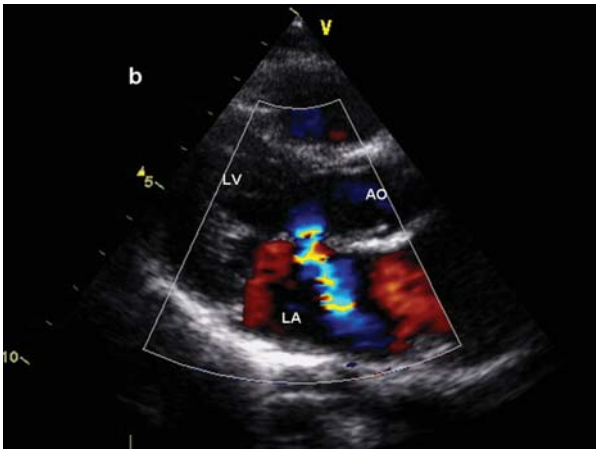



(**a**)

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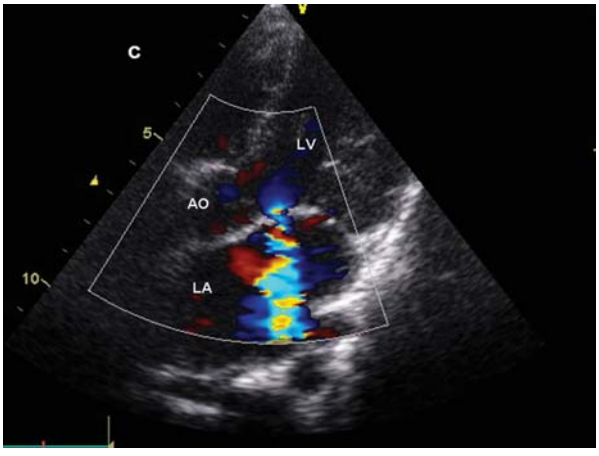


(**c**)

**Figure 1.**
*The long axis view of the heart (**a**) shows a perforation of 3 millimetres in the aortic leaflet of the mitral valve (white arrow). Colour Doppler shows the regurgitation through the perforation (**b**), and also in the five chamber view (**c**). Abbreviations: LA: left atrium; LV: left ventricle; AO: aorta.*

follow-up, failed to reveal evidence of underlying malignancy or a chronic systemic illness. Cardiac involvement in this syndrome has been previously described in both adults and children, with myocardial infiltrates and diffuse vascular disease described as involving the aorta, pulmonary, and coronary arteries.[4–6] Valvitis of both the mitral and aortic valves has also been reported in adults.[7,8] We did not perform microscopic examination in our patient, as the surgeon repaired the mitral valve without any resection. The clinical history, and the absence of other causes for this unusual mechanism for mitral valvar regurgitation, is in favour of mitral valvitis associated with Sweet's syndrome.

## References

1. Burgess BC, Armstrong WF. Echocardiographic evaluation of mitral valve endocarditis and perforation. Echocardiography 2005; 22: 363–364.
2. De Castro S, d'Amati G, Cartoni D, et al. Valvular perforation in left-sided infective endocarditis: a prospective echocardiographic evaluation and clinical outcome. Am Heart J 1997; 134: 656–664.
3. Oztunc F, Saltik IL, Turkoglu H. Mitral perforation: a rare cause of congenital mitral regurgitation. Cardiol Young 2003; 13: 472–474.
4. Shimizu K. Neutrophilic infiltration of myocardium in a patient with myelodysplastic syndrome. Am J Hematol 1998; 58: 337–338.
5. Guia JM, Frias J, Castro FJ, Gracian M. Cardiovascular involvement in a boy with Sweet's syndrome. Pediatr Cardiol 1999; 20: 295–297.
6. Muster AJ, Bharati S, Herman JJ, Esterly NB, Gonzales-Crussi F, Holbrook KA. Fatal cardiovascular disease and cutis laxa following acute febrile neutrophilic dermatosis. J Pediatr 1983; 102: 243–248.
7. Dorenkamp M, Weikert U, Meyer R, Schwimmbeck PL, Morguet AJ. Heart failure in acute febrile neutrophilic dermatosis. Lancet 2003; 362: 1374.
8. Hayashi I, Hosoda Y, Kawasaki S, Yamamoto T, Dohi S, Kawai S. Aortic and mitral valve replacement in a patient with acute neutrophilic dermatosis (Sweet's syndrome): report of a case. Surg Today 2001; 31: 810–813.

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