

## SHORT CASES IN CARDIOLOGY

## Effect of percutaneous fenestration of the atrial septum on protein-losing enteropathy after the Fontan operation

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Protein-losing enteropathy after the Fontan operation is an uncommon and serious complication that is difficult to manage.<sup>1-3</sup> The abnormal loss of protein in the gut is believed to be related to a chronic increase in pressure in the superior vena cava, resulting in impaired lymphatic drainage from the gastrointestinal tract. Overt protein-losing enteropathy is characterised clinically by oedema, ascites, immunodeficiency, and hypocalcaemia.

Surgical fenestration of the atrial septum improved clinical outcome among high risk patients who had a modified Fontan operation.<sup>4</sup> Fenestration reduced complications, such as unremitting pleural effusions, that were related to congestion. The right to left shunt through the fenestration decreased the right atrial pressure and increased the cardiac output because it increased the preload of the single ventricle.<sup>5</sup> This, however, occurs at the expense of a decrease in arterial saturation and a risk of paradoxical (septic) emboli. None the less, patients are less limited by cyanosis than by congestive heart failure. Because protein-losing enteropathy is caused by chronic venous congestion, a reduction in systemic venous pressure should be beneficial.

We performed a percutaneous fenestration of the atrial septum in a 29 year old woman 4 years after a right atrial-pulmonary artery Fontan connection for tricuspid atresia had been constructed. One year after surgery our patient presented with symptoms of protein-losing enteropathy that did not respond to conventional medical treatment with diuretics, digoxin, a high protein diet with low sodium and low fat (with supplements of medium chain triglycerides), corticosteroid treatment, and intermittent intravenous replacement of albumin. The symptoms of peripheral oedema with abdominal congestion and crushing chest pain (without ST segment changes on electrocardiogram) became intolerable. Her exercise capacity was severely limited (New York Heart Association class III) and normal daily activity became impossible. Reactive depression with suicidal thoughts developed.

Haemodynamic evaluation showed low systemic venous pressures (mean 9 mm Hg) with no obstruction of the pulmonary arter-

ies. Cardiac output was low (1.8 l/min/m<sup>2</sup>) with a very slow washout of contrast: 5 minutes after injection, contrast was still whirling in the lower parts of the right atrium.

The interatrial septum was fenestrated by puncturing it with a Brockenbrough needle. The puncture hole in the septum was dilated with an 8F Mullins sheath and then with progressively larger balloons. Each inflation of the balloon lasted for more than two minutes. We started with a 6 mm balloon and increased the size of the balloon until oxygen saturation measured by percutaneous pulse oximetry reached about 85%. We used a 10 mm balloon for the final dilatation. The right atrial pressure remained unchanged, but cardiac output increased by 24%. There were no complications related to the procedure.

After the procedure there was a remarkable immediate clinical improvement with the disappearance of all abdominal and thoracic complaints. Protein loss, as measured by a chromium-51 albumin test, decreased from 29% to 18% (normal < 1% in 5 days) the blood concentration of albumin increased from a mean of 2.5 g/100 ml under optimum medical treatment to 4.1 g/100 ml with only diuretic treatment. Intravenous albumin and chronic steroid treatment were no longer needed and 5 months after the procedure the patient's condition was stable. Her quality of life was much improved, exercise capacity had improved (NYHA II) and she was no longer depressed.

Fenestration of the interatrial septum should be considered in patients with a poor clinical result after Fontan operation, before proceeding to Fontan take-down or heart transplantation. We believe that a late fenestration is indicated in Fontan patients with a low cardiac output, venous congestion, and complications that are related to congestion, such as protein-losing enteropathy. The procedure is not feasible after Fontan operations in which a Gore-tex intra-atrial conduit or a complex baffle have been used. Stenting of the atrial connection can be used to prevent spontaneous closure of the fenestration. The feasibility of this technique is being tested on mongrel dogs and the preliminary results seem very promising.

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## Diagnosis, treatment, and long-term follow up of a patient with a hydatid cyst of the left ventricle

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An 18 year old woman was admitted with fever and atypical chest pain. A hydatid cyst had been removed from her liver 13 years before. A 2/6 systolic heart murmur was audible in the mitral valve area. The electrocardiogram showed deep negative T waves in leads I, II, III, aVF, and V3-V6. A chest x ray showed a calcified cyst adjacent to the left ventricle. The echocardiogram showed a spherical calcified cyst (4 cm) containing multiple intracystic trabeculations (fig 1). It was attached to the free wall of the left ventricle near the apex and occupied three quarters of the chamber. Computed tomography and magnetic resonance imaging confirmed the diagnosis of hydatid cyst and showed a second cyst in the liver (fig 2). The intact intracardiac cyst was surgically removed and the patient made an uneventful recovery. The postoperative echocardiogram was normal and the electrocardiogram returned gradually to normal over the next two years.

Interesting aspects of this case are the unusual site,<sup>1</sup> young age of the patient, and the successful removal of the intact cyst. The most dangerous complication of cardiac hydatid cysts is early rupture, which can induce embolism or lifethreatening anaphy-

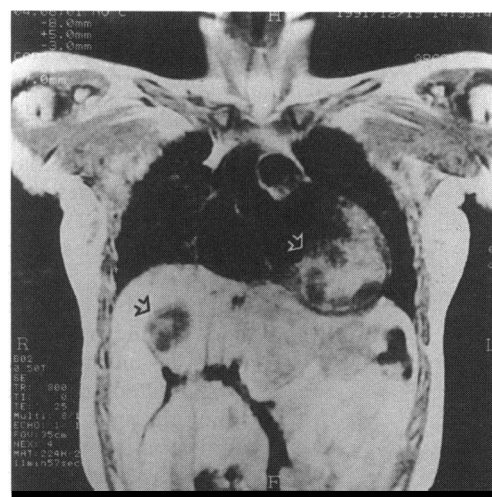


Figure 2 Magnetic resonance imaging of thorax and abdomen showing two hydatid cysts, one in the left ventricle and another in the liver (arrows).

lactic shock.<sup>2</sup> Echocardiography is the most reliable diagnostic method,<sup>3</sup> but computed tomography and magnetic resonance imaging can also show cysts elsewhere in the body.<sup>4</sup> We would like to emphasise the importance of the negative T waves in the electrocardiogram that led to further investigation and diagnosis.

We believe that all patients with echinococcosis should have echocardiography to exclude cardiac involvement and to prevent subsequent lifethreatening complications.

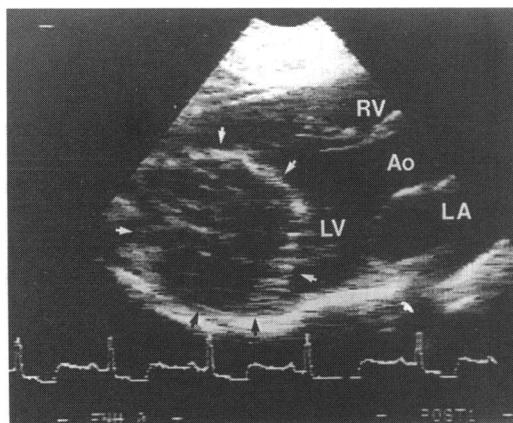


Figure 1 Cross sectional echocardiogram showing a hydatid cyst in the left ventricle. It contained daughter cysts, arose from the free wall of the ventricle, and occupied three quarters of its chamber.

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