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IMAGES IN CARDIOLOGY

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Mid aortic syndrome: a rare cause of reversible cardiomyopathy

A 7 month old girl (body weight 7 kg, length 73 cm) was presented with tachypnoea, hepatomegaly, vomiting, decreased peripheral perfusion, systolic heart murmur, and sinus tachycardia of up to 205 beats/min. She had a short history of decreasing activity for about four weeks. Radiography showed an enlarged heart, and echocardiography showed the left ventricular function was depressed with severe mitral regurgitation and mild tricuspid regurgitation. M mode showed left ventricular end diastolic diameter (LVEDD) 28 mm, fractional shortening (FS) 19%, and ejection fraction (EF) 49%. The aortic valve was without pathological findings and an aortic coarctation in the isthmus was excluded. Coronary arteries were normal; anomalous left coronary artery from pulmonary artery (ALCAPA) syndrome was excluded. Femoral pulse deficit and a difference of 50 mm Hg in systolic blood pressure between the arms and the legs pointed to the diagnosis, and magnetic resonance imaging verified stenosis of the distal thoracic aorta up to the diaphragm. There were no collaterals. Percutaneous

transluminal angioplasty with inflation pressure up to 8 atm was not successful (panel A). Subsequently surgery was performed without cardiopulmonary bypass through left lateral thoracotomy. A 10 mm diameter Gore-Tex prosthesis was implanted with end-to-side anastomosis. Panel B shows that the length of the bypass is oversized to allow for patient growth. Cardiac function improved within three days and mitral regurgitation vanished completely.

Mid aortic syndrome is a rare variety of aortic coarctation, located in the distal thoracic or the abdominal aorta, often involving the renal and intestinal vessels and presenting predominantly as untreatable arterial hypertension. In the later course it ends in congestive heart failure, mimicking dilatative cardiomyopathy.

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Angiography: the stenosis is 2 cm long with a minimum diameter of 3 mm; the diameter of the aorta proximal and distal to the stenosis is 8 mm.

A surgical view in the left hemithorax; a 10 mm diameter Gore-Tex prosthesis oversized in length was implanted to allow for anatomical growth.