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Late anastomotic stenosis after correction of totally anomalous pulmonary venous connection

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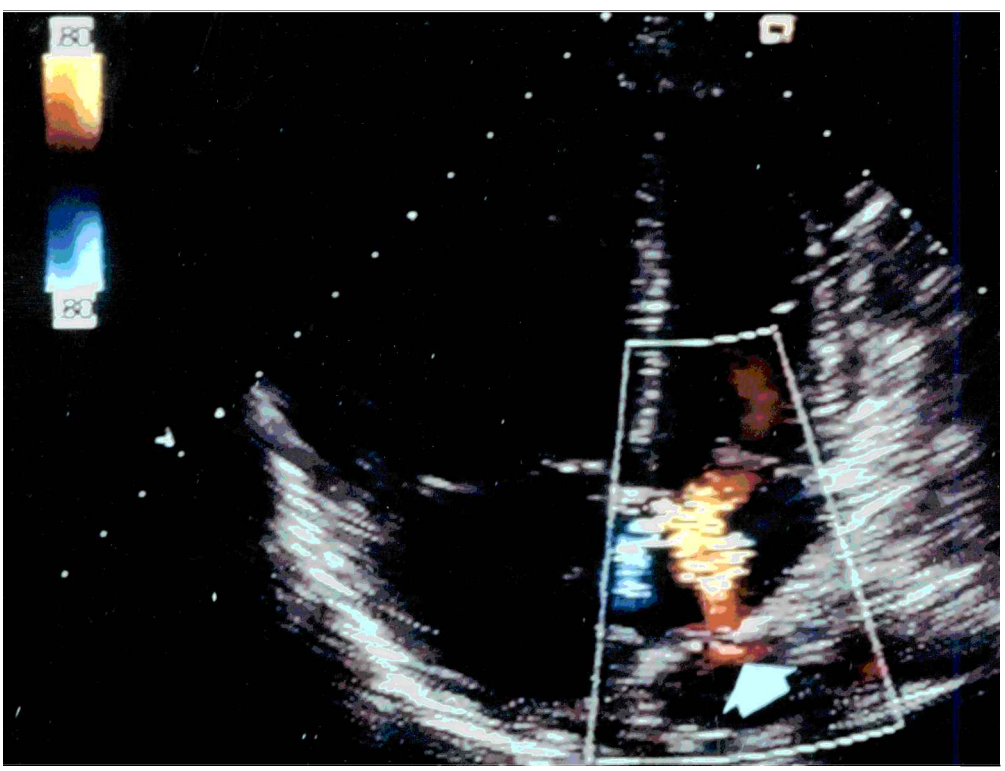

A 14 YEAR OLD BOY WHO HAD SUCCESSFUL repair of infracardiac totally anomalous pulmonary venous connection as a neonate was symptom free until he presented with a 1 month history of dyspnoea. He had clinical signs of pulmonary hypertension, but no murmurs. This apical 4 chamber view of the echocardiogram shows a dilated, hypertrophied right ventricle with severe stenosis (arrow) at the original anastomosis between the pulmonary venous channel and the left atrium. Doppler echocardiography revealed a flow gradient of 40 mmHg, giving an appearance very similar to divided left atrium ("cor triatriatum"). The anastomosis was enlarged using a pericardial patch, and the patient made an uncomplicated recovery. There was no residual

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obstruction at follow up 6 weeks later, when his symptoms had resolved completely, although the right ventricular dilation had not fully regressed. The long-term outlook for patients after correction of totally anomalous pulmonary venous connection is usually excellent. Pulmonary venous stenosis related to the anastomotic site is well recognised, but almost invariably occurs early postoperatively. When obstruction does occur late, it is most frequently related to progressive pulmonary veno-occlusive disease, and this responds very poorly to surgical treatment. It is difficult to explain why our patient had been symptom free, and still playing soccer, until only a few weeks before his presentation with severe pulmonary venous and arterial hypertension. The case illustrates that an excellent early result from surgery does not necessarily guarantee late freedom from reoperation.

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