Atrial Baffle Stenosis: A Late Complication after Mustard Repair for d-TGA
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Clinical History
A 37 year old male with a history of d-transposition of the great arteries and Mustard atrial switch operation as an infant presented with abdominal distension and facial swelling. Cardiac MRI and CT were performed for evaluation of the atrial baffle and biventricular function.

Findings
The Cardiac MRI demonstrated significant stenosis at the superior and inferior limbs of the baffle (figure 1). The atrial baffle path for pulmonary vein flow was widely patent. The systemic right ventricle was hypertrophied with low normal systolic function (RVEF=45%). Cardiac CT demonstrated an anomalous left circumflex artery arising from a common trunk with the right coronary artery from the posterior sinus of Valsalva and anteriorly located ascending aorta relative to the main pulmonary artery (figure 2). Ascites and liver cirrhosis due to chronic right atrial hypertension were also visualized.

The patient underwent cardiac catheterization followed by successful angioplasty and stenting of the stenotic segments in the atrial baffle. Post procedure, his neck fullness resolved and ascites markedly improved.

Discussion
D-transposition of the great arteries (d-TGA) refers to the dextroposition of the bulboventricular loop and ventriculoarterial discordance (1). Due to failure of spiral septation of the truncus arteriosus, the great arteries course parallel to each other rather than crossing. The aorta arises from the morphologic right ventricle and the main pulmonary artery from the morphologic left ventricle. Communication between the pulmonary and systemic circuits is necessary for infant survival either via an atrial septal defect, patent ductus arteriosus or ventricular septal defect. Ventricular septal defects are the most common associated anomaly, observed in almost half of the cases. Pulmonary outflow tract obstruction and coartation of the aorta may also be seen (2).

Atrial septostomy was the original surgical procedure first performed in the 1960s. The atrial switch procedure (Mustard or Senning types) then became the most common surgical treatment. Right ventricular dysfunction, tricuspid regurgitation, atrial arrhythmias and heart block are frequently encountered in the follow-up of these patients. The arterial switch procedure has become the preferred technique to repair d-TGA. It involves the transection and switching of the great arteries with reimplantation of the coronary arteries, thus restoring normal ventriculo-arterial concordance. Baffle leaks and obstruction are common late complications of atrial switch surgery.

Baffle leaks should be considered in patients with stroke or paradoxical embolism and baffle obstruction should be considered in patients with signs and symptoms of systemic venous hypertension. Percutaneous angioplasty or surgery is the main treatment strategy in baffle obstruction.

REFERENCES