



Another common postoperative problem is the tricuspid valve. After the atrial switch procedure, the tricuspid valve remains the systemic atrioventricular valve and must tolerate systemic pressures. Due to changes in RV morphology and abnormal chordal attachments, the tricuspid valve is prone to become dysfunctional and develop significant regurgitation.

Significant coronary lesions, such as occlusions or stenoses, occur in 6.8% of patients who have had the arterial switch procedure. These lesions are likely related to suture lines or kinking at the time of reimplantation of the coronary arteries into the neo-aorta. Systemic LV function is usually normal. LV dysfunction is associated with coronary anomalies.

Clinical Presentation

In the repaired adult with an atrial switch procedure, the physical examination may reveal a murmur consistent with tricuspid valve insufficiency and a prominent second heart sound due to the anterior position of the aorta. Patients who have had an atrial switch procedure tend to have worsening functional status as the length of follow-up increases. They often have resting sinus bradycardia or a junctional rhythm. Palpitations due to atrial arrhythmias are common, occurring in up to 48% of patients 23 years after the atrial switch procedure.

In those who undergo the arterial switch procedure, the physical examination may reveal a murmur of neo-aortic or neo-pulmonic regurgitation. These patients usually have normal function status, but because of denervation of the heart, myocardial ischemia may manifest as atypical chest discomfort.

Diagnosis

After the atrial switch procedure, the ECG may show a loss of sinus rhythm with evidence of RV hypertrophy. Chest radiographs may show an enlarged cardiac silhouette in those with a dilated systemic right ventricle. An echocardiogram can demonstrate qualitative systemic RV size and function and the degree of tricuspid regurgitation. MRI is often used to accurately quantify systemic RV size and function, tricuspid valve function, and atrial baffle anatomy.

Echocardiography is used to assess pulmonary artery and branch pulmonary artery stenosis, neo-aortic and neo-pulmonic valve regurgitation, and ventricular function. MRI or computed tomography may be used to assess the anatomy of the branch pulmonary arteries. An exercise stress test is often used to evaluate myocardial ischemia.

Treatment


Treatment options are limited for adults with complete TGA repaired by atrial switch who have failing systemic right ventricles or significant tricuspid regurgitation, and evidence of significant benefit is lacking. However, potential treatments include medical therapy, revision of atrial baffles, pulmonary artery banding, resynchronization therapy, ventricular assist devices, and possible transplantation.

After the arterial switch procedure, catheter-based or surgical reintervention for pulmonary artery stenosis may be required in 5% to 25% of patients, Coronary artery revascularization is rarely required (0.46% of patients), as is neo-aortic valve repair or replacement (1.1% of patients).

Prognosis

Long-term follow-up studies after the atrial switch procedure show a small but ongoing attrition rate, with numerous intermediate- and long-term complications. Long-term complications include systemic RV dysfunction and tricuspid valve regurgitation, loss of sinus rhythm with the development of atrial arrhythmias (50% incidence by age 25), endocarditis, baffle leaks, baffle obstruction, and sinus node dysfunction requiring pacemaker placement. Intermediate-term complications include coronary artery compromise, pulmonary outflow tract obstruction (at the supra-avalvular level or takeoff of the peripheral pulmonary arteries), neo-aortic valve regurgitation, endocarditis, and neo-aorta dilation.

As a result of the long-term complications associated with the atrial switch procedure, the arterial switch operation has been the procedure of choice since 1985. Long-term data on the survival after the arterial switch operation do not exist, but intermediate-term results are promising: 88% at 10 and 15 years.

 For a deeper discussion on this topic, please see Chapter 69, "Congenital Heart Disease in Adults," in Goldman-Cecil Medicine, 25th Edition.

SUGGESTED READINGS

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