Dextro-Transposition of the Great Arteries
Long-term Sequelae of Atrial and Arterial Switch

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KEYWORDS

• Transposition of great arteries • Arterial switch • Atrial switch • Congenital heart disease

KEY POINTS

- Patients who have undergone the Mustard/Senning operation are at risk of heart failure, atrial arrhythmias, and sudden cardiac death that requires long-term follow-up.
- Compared with the Mustard/Senning operation, the long-term survival and event-free survival rates in the arterial switch operation (ASO) are superior, though these patients still require routine follow-up.
- Echocardiography is recommended in the long-term follow-up of both the atrial and arterial switch. For Mustard/Senning patients, evaluation of the systemic right ventricle and integrity of the baffle should be assessed. Coronary stenosis, neo-aortic regurgitation and branch pulmonary artery stenosis can be seen after ASO.
- Routine CT angiography or MRI is recommended in patients with D-TGA after atrial switch for patency of the baffles, baffle leak, and systemic right ventricular function.
- Chest pain or wall motion abnormalities should be evaluated promptly in the arterial switch patient, given the increased risk of coronary complications.

OVERVIEW

This article focuses on complete transposition of the great arteries, otherwise commonly referred to as dextro-transposition of the great arteries (D-TGA) and the 2 primary ways used to repair the lesion: the atrial switch procedure and the arterial switch operation (ASO).

TERMS

TGA is a congenital heart defect in which the aorta arises from the morphologic right ventricle (RV) and the pulmonary artery (PA) arises from the morphologic left ventricle.

In the term *D-TGA*, the *D* refers to either the ventricular loop or the spatial relationship of the aortic and pulmonary arteries. For this article, D-TGA represents situs solitus, atrioventricular (AV) concordance, and ventriculo-arterial discordance.

Atrial switch is the creation of an atrial baffle to direct venous flow to the contralateral AV valve and ventricle.

Senning operation is an atrial switch operation that creates a baffle out of autologous tissue.

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Mustard operation is an atrial switch operation that creates a baffle out of synthetic material.

ASO is surgery that restores normal physiologic relationships in D-TGA. The PA and aorta are transected above the sinuses and reimplanted to positions to restore normal blood flow. The coronary arteries are also detached and sewn onto the neo-aorta.

Complete Transposition of the Great Arteries

The most common form of TGA is a cyanotic heart lesion. The prevalence of TGA is between 20 and 30 per 100,000 live births.[1] It composes approximately between 5% and 7% of all congenital heart disease.[1]

In the most common form of transposition, also known as complete transposition or D-TGA, there is ventriculo-arterial discordance. Thus, the aorta arises from the morphologic RV and the PA arises from the morphologic left ventricle (LV), which creates 2 parallel circuits for blood flow. In one circuit, the deoxygenated blood returns via systemic venous return to the right atrium, travels to the RV, and then flows out to the body via the aorta. In the other circuit, the oxygenated blood returns from the lungs to the left atrium, flows into the LV, and then flows back to the lungs via the PA (Fig. 1A).

Because the blood runs in 2 parallel circuits, there must be a communication between the circuits to sustain life in the form of an atrial septal defect, ventricular septal defect (VSD), or patent ductus arteriosus. These communications allow mixing of oxygenated and deoxygenated blood.

If D-TGA is present with no other cardiac abnormalities, it is called *simple TGA*. If there are other anomalies present, that is, a VSD or LV outflow tract obstruction (LVOTO), this is often referred to as *complex TGA*. VSDs and LVOTO occur in approximately 50% and 25% of patients with D-TGA, respectively.[1]

Congenitally Corrected Transposition of the Great Arteries

There is another form of TGA: levo-TGA (L-TGA). Most patients with L-TGA also have ventricular inversion; thus, the commonly applied term for L-TGA is congenitally corrected TGA. In this defect, in addition to the transposed great arteries, there is ventricular inversion, with the LV receiving inflow from the right atrium and the RV receiving inflow from the left atrium. In this circuit, deoxygenated blood returns via the right atrium, goes into the morphologic LV, and then goes to the lungs through the PA. Once the blood is oxygenated in the lungs, it flows back into the left atrium through the pulmonary veins, into the morphologic RV, and then out to the aorta. Because there is also ventricular inversion, along

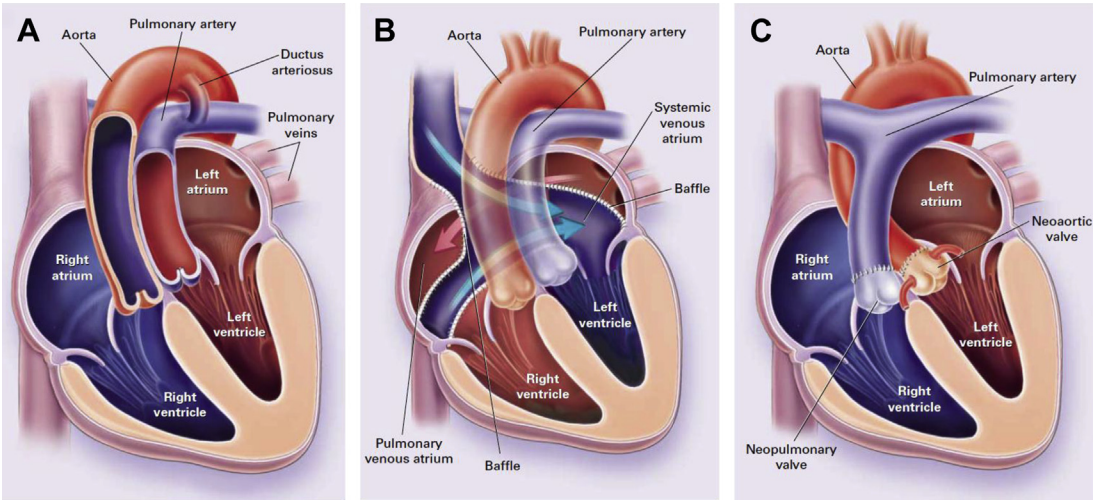

Fig. 1. (*A*) Diagram of typical TGA. The atria and ventricles are in their usual position, but the aorta arises anteriorly from the RV and the PA arises posteriorly from the LV. (*B*) Diagram of atrial-level repair for TGA. Both the Mustard and Senning repairs create a baffle within the atria that redirects the caval blood to the mitral valve and the pulmonary venous blood to the tricuspid valve. (*C*) Diagram of arterial repair for TGA. The PA and the aorta are excised from their respective positions and resewn into their correct anatomic positions. The coronary arteries are also excised and sewn onto the neo-aorta. (*From* Brickner ME, Hillis LD, Lange RA. Congenital heart disease in adults. Second of two parts. N Engl J Med 2000;342(13):338; with permission.)