Arrhythmias Following the Mustard and Senning Operations for Dextro-Transposition of the Great Arteries

Clinical Aspects and Catheter Ablation

Sherrie Joy Baysa, MD, Melissa Olen, ARNP, Ronald J. Kanter, MD*

KEYWORDS

• Mustard operation • Senning operation • Dextro-transposition of the great arteries
• Sinoatrial node dysfunction • Intraatrial reentry tachycardia

KEY POINTS

- The atrial switch procedures, Mustard and Senning Operations, for dextro-transposition of the great arteries, have been supplanted by the arterial switch operation, meaning that a finite number of adult patients are currently alive with this anatomical arrangement.
- Sinoatrial node dysfunction is common long-term after this operation and, when clinically severe, is successfully treated with pacemaker implantation.
- Atrial tachycardias, especially intraatrial reentry tachycardia, is also prevalent long-term after this operation, and may be successfully ablated in the great majority of cases.
- Although there is known to be an increased risk of systemic ventricular dysfunction and sudden death in this patient group, risk assessment and treatment are only beginning to be understood.

INTRODUCTION

Transposition of the great arteries (TGA) accounts for 5% of all congenital heart defects and is the most common cardiac cause of cyanosis in the newborn period (**Fig. 1**).[1]

Although the introduction of balloon atrial septostomy by Rashkind and Miller[2] in 1966 led to a dramatic change in the early survival of patients, TGA is ultimately a surgical disease. Early attempts at anatomic correction with arterial switch procedures showed dismal results, particularly because of the need for coronary artery transfer.[3] Thus, reversing the venous inflows, or so-called atrial switch procedures, became the mainstay of surgical treatment in dextro-TGA (d-TGA) for many years and remained so for more than a decade after Jatene and colleagues[4] reported the first successful arterial switch operation in 1976.[3]

Because the arterial switch and coronary artery translocation procedure supplanted the Mustard and Senning operations in most Western countries starting in the late 1980s, most surviving patients of the atrial switch are now at least 25 to 30 years old. However, there are rare anatomic conditions for which these operations are still used. These conditions primarily include congenitally corrected transposition in which a double switch (atrial and

Division of Cardiology, Nicklaus Children's Hospital, Miami, FL 33155, USA
* Corresponding author. 3100 Southwest 62 Avenue, 2nd Floor, Ambulatory Care Building, Miami, FL 33155.
E-mail address: Ronald.kanter@mch.com

Card Electrophysiol Clin ■ (2017) ■–■
http://dx.doi.org/10.1016/j.ccep.2017.02.008

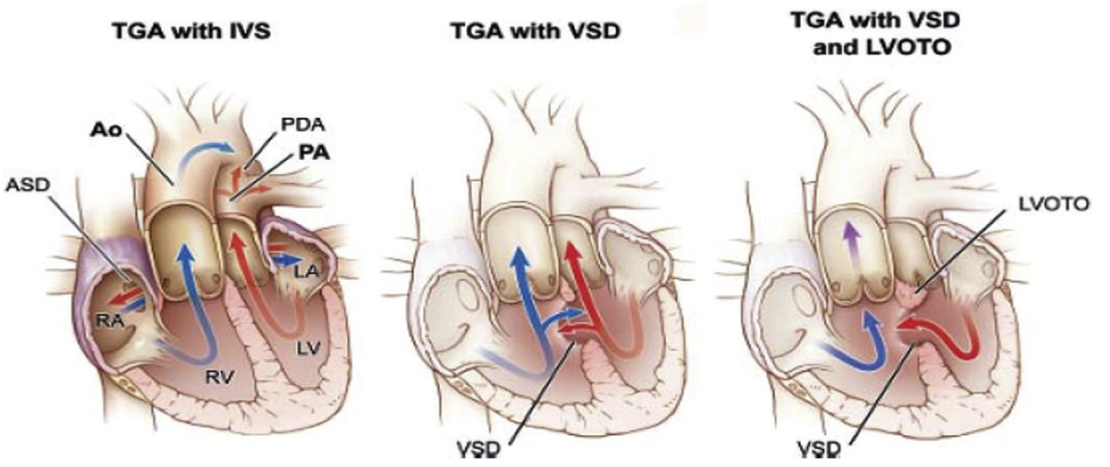

Fig. 1. Examples of anatomic variants of dextro-TGA. Arrows indicate direction of blood flow. Locations where arrows cross one another represents opportunities for mixing of systemic and pulmonary venous returns. It is only at those locations that effective blood flow may occur. Effective blood flow is defined as that portion of systemic venous return that is fully saturated by the lungs. Ao, aorta; ASD, atrial septal defect; IVS, intact ventricular septum; LA, left atrium; LV, left ventricle; LVOTO, left ventricular outflow tract obstruction; PA, pulmonary artery; PDA, patent ductus arteriosus; RA, right atrium; RV, right ventricle; TGA, transposition of the great arteries; VSD, ventricular septal defect. (*From* Qureshi Athar M, Justino H, Heinle Jeffrey S. Transposition of the great arteries. [Chapter 47]. In: Allen HD, Shaddy RE, Penny DJ, editors. Moss and Adams' heart disease in infants, children, and adolescents, including the fetus and young adult. 9th edition. Philadelphia: Lippincottt Williams and Wilkins; 2016. p. 1163; with permission.)

arterial switches) or atrial switch plus right ventricular (RV) outflow tract-to-pulmonary artery conduit are indicated. Even rarer are patients with isolated atrioventricular (AV) discordance, in whom there is AV discordance but ventriculoarterial concordance. In all of these conditions, the atrial switch will establish the anatomic left ventricle as the subaortic pumping chamber and the anatomic RV as the subpulmonic pumping chamber.

SURGICAL ANATOMY

Senning Operation

Senning[5] performed the first atrial switch procedure in 1958. In this procedure, an atrial baffle is created with autologous tissue to direct systemic venous return to the mitral valve and subpulmonic left ventricle. First, a vertical right atriotomy is made anterior to the crista terminalis (Fig. 2A, upper dotted line). Then, the anterior, inferior, and superior margins of the atrial septum are incised, creating an atrial septal flap (Fig. 2B, dotted line). The atrial septal flap is placed down into the left atrium and sutured around the pulmonary veins, separating the pulmonary venous return from the left atrium and forming the floor of the eventual systemic venous pathway (Fig. 2C). The posterior/lateral right atrial wall is then sutured around the superior and inferior vena cavae orifices and onto the anterior edge of the atrial septal remnant between the AV valves (approximation of the 2 dotted lines in Fig. 2C). In this way, an intercaval tube representing the systemic venous pathway to the mitral valve is completed (Fig. 2D). A vertical left atriotomy is then made between the right pulmonary veins and the atrial septum (see Fig. 2A, lower dotted line). Finally, the anterior right atrial wall is used to complete the pulmonary venous pathway from the left atriotomy to the tricuspid valve and systemic RV (see Fig. 2D, dotted line of upper free wall flap is brought down to lower dotted line). Pericardium or artificial material may be required to bridge the gap between those incisions.

Mustard Operation

In 1963, Mustard[6] performed an atrial switch operation, using pericardium to create the intra-atrial baffle; this soon emerged as an alternative to the Senning procedure. In this operation, the atrial septum and most of the limbus are excised to create a large atrial septal defect, extending from the inferior to superior vena cava. An intra-atrial baffle, generally made of pericardium, is then placed, directing the systemic venous flow to the left-sided mitral valve and ultimately to the pulmonary artery (Fig. 3). Surgical details are described later in the section on the electrophysiologic anatomy.

The Mustard and Senning atrial switch procedures provided excellent early and midterm results;