Chapter 21

D-Transposition of the Great Arteries (Arterial Switch)

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Case Scenario

A 10-year-old male presents for elective upper endoscopy and colonoscopy. He has a 2-week history of persistent vomiting and weight loss with new-onset diarrhea. He has a history of dextro-transposition of the great arteries and underwent an arterial switch operation shortly after birth. He has been followed since by his cardiologist for residual pulmonic stenosis. He and his family report he is able to participate in sports but has begun to tire more quickly than his peers; he has felt considerably more fatigued since the start of the vomiting and diarrhea. His current vital signs include heart rate 100 beats/minute, respiratory rate 18 breaths/minute, and SpO_2 99% on room air. His abdomen is soft, mildly distended, and nontender to palpation. He has mild eczema on his extremities.

Transthoracic echocardiography 1 week earlier showed the following:

- *Peak velocity of 3.5 m/s and peak gradient of 50 mm Hg across the pulmonary valve*
- *Right ventricular hypertrophy*
- *Qualitatively normal biventricular function*

Key Objectives

- Understand the anatomy of dextro-transposition of the great arteries.
- Describe the arterial switch operation and identify potential sequelae.
- Describe the preoperative assessment and anesthetic management of a patient with repaired dextro-transposition.
- Understand considerations regarding performing procedures in remote or offsite locations in patients with repaired congenital heart disease.

Pathophysiology

What is transposition of the great arteries?

Transposition of the great arteries (TGA) is most accurately an umbrella term for all congenital cardiac lesions whereby the aorta arises from the right ventricle (RV) and the pulmonary artery (PA) arises from the left ventricle (LV). This is termed ventriculoarterial discordance. Most typically the term TGA is used to denote dextro (d)-TGA, whereas other forms of TGA are called by their specific names, as discussed in the text that follows.

What is d-TGA?

D-transposition of the great arteries is one of the most common cyanotic congenital heart lesions, characterized by ***ventriculoarterial discordance***, the origination of the great vessels from the incorrect ventricles, in a heart with otherwise normal connections. Deoxygenated systemic blood flows from the vena cavae to the right atrium (RA), on to the RV, and then to the aorta. Oxygenated pulmonary venous blood enters the left atrium (LA), flows to the LV, and exits the PA. This anatomy results in a parallel circulation. (See Figure 21.1.) Fetal shunts between the parallel circulations, including a patent foramen ovale (PFO) and patent ductus arteriosus (PDA), are required to provide mixing between the separate circulations and thus some degree of oxygen delivery to the tissues. Without intervention, d-TGA is uniformly fatal in infancy, with a 30% mortality in the first week of life and 50% in the first month. Transposition represents 5% of all congenital heart lesions (32 per 100,000 live births) with a 2:1 predilection for males.

What are other types of TGA and how are they distinct from d-TGA?

D-transposition is a morphologically distinct entity wherein the "d" denotes dextroposition of the bulboventricular loop embryologically, resulting in atrioventricular concordance, ventriculoarterial discordance, and a typically anterior and rightward position of the aortic valve.

Congenitally corrected, or levo-TGA (l-TGA), is also characterized by ventriculoarterial discordance; however, the addition of atrioventricular discordance results in a series circulation, with the left-sided morphologic RV