Multimodality Imaging Guidelines of Patients with Transposition of the Great Arteries: A Report from the American Society of Echocardiography Developed in Collaboration with the Society for Cardiovascular Magnetic Resonance and the Society of Cardiovascular Computed Tomography

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Abbreviations

ASE = American Society of Echocardiography

ASO = Arterial switch operation

AtrSO = Atrial switch operation

AV = Atrioventricular

BAS = Balloon atrial septostomy

CMR = Cardiovascular magnetic resonance

CT = Computed tomography

EF = Ejection fraction

IV = Intravenous

IVC = Inferior vena cava

LGE = Late gadolinium enhancement

LV = Left ventricular

PDA = Patent ductus arteriosus

PET = Positron emission tomography

PH = Pulmonary hypertension

Qp/Qs = Pulmonary-to-systemic flow ratio

RV = Right ventricular

SPECT = Single-photon emission computed tomography

SVC = Superior vena cava

TEE = Transesophageal echocardiography

TGA = Transposition of the great arteries

3D = Three-dimensional

TTE = Transthoracic echocardiography

2D = Two-dimensional

VSD = Ventricular septal defect

EXECUTIVE SUMMARY

Transposition of the great arteries (TGA) is a congenital heart defect with ventriculoarterial discordance in which the aorta is aligned with the right ventricle and the pulmonary artery is aligned with the left ventricle. When atrioventricular (AV) concordance is present, this anatomy results in cyanosis because the systemic and pulmonary circulations are in parallel. The clinical diagnosis and management of patients with TGA has improved dramatically over the past three decades because of the evolution and availability of multiple imaging modalities and strides made in the surgical management of these patients. Despite these technical advances, patients with TGA require long-term surveillance because of ongoing anatomic and hemodynamic abnormalities. The purpose of this report is to present guidelines for multimodality imaging in this cohort of patients.

Goals of Imaging

The goals of imaging in patients with TGA are to provide accurate and reproducible anatomic and hemodynamic information that facilitate medical and surgical planning and to provide surveillance imaging to evaluate potential issues related to the type of surgical operation that has been chosen.

Imaging Modalities

We review each imaging modality used in the diagnosis and follow-up of patients with TGA including the strengths, limitations, and clinical utility. We also define an optimal imaging protocol for each modality. In addition, an integrated multimodality imaging algorithm is defined and discussed.

Echocardiography

Echocardiography remains the main diagnostic imaging modality for TGA because of its widespread availability and portability. Transthoracic echocardiography (TTE) with two-dimensional (2D) and Doppler echocardiography provides comprehensive anatomic and hemodynamic evaluation in the majority of patients with TGA and is usually the only modality required for preoperative evaluation. For postoperative imaging, echocardiography is often used to assess for residual, recurrent or new pathology. Transesophageal echocardiography (TEE) is indicated in patients with poor windows, during intraoperative imaging, and in patients (usually adolescents or adults) who require cardioversion for arrhythmia.

Cardiovascular Magnetic Resonance

Cardiovascular magnetic resonance (CMR) plays a major role in the evaluation of patients with TGA. It is used primarily to image patients after surgical intervention. It provides important information regarding myocardial performance and viability as well as quantitative assessment of valvar function and accurate evaluation of baffles, conduits, and extracardiac structures such as the branch pulmonary arteries and the aortic arch.

Cardiovascular Computed Tomography

Multidetector computed tomography (CT) is typically used in patients with TGA who cannot undergo CMR. Adults with TGA who have had the atrial switch operation (AtrSO) frequently have pacemakers; thus, CT is an alternative imaging modality to provide incremental information to echocardiography.

Nuclear Scintigraphy

The primary use of nuclear imaging in patients with TGA is to assess myocardial viability or to assess blood flow to the branch pulmonary arteries after the arterial switch operation (ASO). Imaging can be performed at rest and during stress (exercise or pharmacologic) to determine if there are myocardial perfusion defects.

Exercise and Stress Imaging

Exercise testing and stress imaging are predominantly used to assess for myocardial perfusion problems in patients with TGA, particularly after the ASO. In patients with concern for coronary ischemia, stress imaging can unmask issues that are not present at rest in this population.

Cardiac Catheterization and Angiography

Diagnostic cardiac catheterization is rarely used in the preoperative evaluation of TGA but is required when balloon atrial septostomy (BAS) is performed to improve mixing and alleviate cyanosis. Some institutions use angiography to diagnose or confirm coronary artery (CA) anatomy before an ASO. Postoperative angiography is performed to assess for CA stenosis after CA reimplantation or during interventions such as branch pulmonary artery balloon dilation and stent placement. In the AtrSO, angiography is used to assess for baffle leaks or narrowing of the systemic or pulmonary venous pathways.