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Conotruncal Cardiac Defects: A Clinical Imaging Perspective

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Abstract Conotruncal cardiac defects make up a significant portion of congenital heart disease. For proper diagnosis and subsequent care of patients with these defects, different and sometimes multiple imaging modalities are needed at various stages of care. This article reviews the characteristics of some of the most common conotruncal defects and the imaging options available along with the advantages and disadvantages of each. Intricate knowledge of the capabilities of each modality will aid the practitioner in making optimal clinical decisions.

Keywords Cardiac imaging · Congenital heart disease · Conotruncal heart defects

Introduction

Defects of the cardiac outflow tract make up the conotruncal cardiac defects. Due to errors during embryogenesis, a spectrum of various abnormalities can result. Of all non-syndromic congenital heart disease (CHD), up to 25–30% are conotruncal cardiac defects [11, 17]. The most common conotruncal cardiac defects include tetralogy of Fallot (TOF), truncus arteriosus, transposition of the great vessels (TGV), and double-outlet right ventricle (DORV). Each lesion has multiple variations and subtle nuances that must be clearly defined because the clinical decisions for therapeutic intervention will be dependent on these details. Thus, proper imaging of these defects is imperative.

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Numerous cardiac imaging modalities are available for the initial diagnosis and subsequent follow-up of patients with cardiac defects. Some modalities provide a complete picture of the lesions, whereas others give indirect clues to the diagnosis or clinical status of the patient, therefore requiring different or complementary imaging techniques. Although one method may be acceptable for initial diagnosis, the optimal method may change throughout the patient's course while monitored throughout childhood into adolescence and adulthood (Table 1).

This article reviews the salient features and useful imaging techniques for the most common conotruncal anomalies seen on a regular basis in congenital cardiac centers.

Imaging Modalities

Chest X-Ray

Chest radiography (CXR) is the simplest technique for imaging the heart. An abnormal cardiac silhouette can guide the practitioner toward the need for further diagnostic cardiac imaging. CXR is used frequently to aid in monitoring the general cardiorespiratory status of the patient and complications that may arise during clinical care or intervention. However, the CXR alone rarely provides a final detailed diagnosis needed for making decisions in the cases of complex CHD, such as that found with conotruncal cardiac defects.

Echocardiography

Fetal Echocardiography

With technical advances in echocardiography, fetal echocardiography has become a growing field that may afford a

Table 1 Advantages and disadvantages of cardiac diagnostic imaging modalities

Modality	Advantages	Disadvantages
CXR	Quick, easily accessible, noninvasive, minimal radiation	Nondiagnostic
TTE	Relatively quick, easily accessible, noninvasive, nonradiating, excellent image quality in younger patients	Limited FOV, image quality less in older patients, 2D
TEE	Excellent intracardiac images, intraoperative assessment of surgical repair	Invasive, sedation/anesthesia, difficult to image extracardiac structures, 2D
Catheterization	Assess physiology, interventions possible	Invasive, radiation, sedation/anesthesia, 2D
CMR	Noninvasive, nonradiating, unlimited FOV, assess flow and volumes/function, tissue characterization, 3D	Sedation/anesthesia in younger patients, less accessible, implanted device contraindications
CCT	Relatively quick, noninvasive, 3D	Radiation, contrast, less accessible

FOV field of view

basic and often detailed depiction of a conotruncal defect. However, unless the pregnant mother or fetus has been judged to be at high risk, many will not undergo detailed fetal echocardiography. Often, the fetus with a conotruncal defect that undergoes routine prenatal ultrasound will have four relatively normal chambers, and the investigation stops there. Only with more detailed imaging of the outflow tracts and the great arteries, and the use of Doppler ultrasound in experienced hands performing a thorough fetal echo, will some of these defects be detected [5]. Here, the focus will be on postnatal imaging of conotruncal defects.

Transthoracic Echocardiography

Transthoracic echocardiography (TTE) is often the first modality applied to an infant with suspected CHD. It is an ideal tool in this population because of its accessibility and portability as well as its safety (nonradiating) and noninvasive nature. Because acoustic imaging windows are usually excellent, a thorough diagnosis needed for providing critically important care can usually be provided in infants with conotruncal cardiac defects. In patients for whom more information may be needed, TTE can lead the cardiologist to decide on the next best imaging modality.

As the patient ages, TTE imaging windows can become poor due to scar tissue, body habitus, or other causes, and the field of view may be limited. This may especially limit visualization of extracardiac structures. TTE may be adequate for a general picture of cardiac function, but it may not provide all desired details. Many older children will require additional imaging modalities (Table 1).

Transesophageal Echocardiography

Transesophageal echocardiography (TEE) is minimally invasive and can provide great detail of the intracardiac anatomy and cardiac function, but it is not often performed in the awake child or used for diagnosis or frequent monitoring of CHD. TEE is most often used in the operative setting, at the time of repair of many conotruncal defects, to assess the adequacy of repair and to determine if there are any residual defects significant enough to require further surgery before completing the procedure (Table 1). Although many centers are investigating the application of three-dimensional (3D) echocardiographic techniques, echocardiography largely remains a tool that easily offers two-dimensional (2D) anatomical and functional information.

Cardiac Catheterization and Angiography

In the infant who requires further imaging and urgent intervention, cardiac catheterization and angiography can be quite valuable. With advances in noninvasive imaging techniques, catheterization has become less frequently used solely for diagnostic purposes. With progress and enhancement of interventional devices that may be used or placed during cardiac catheterization, the catheterization laboratory is now used more for these necessary interventional procedures. Cardiac catheterization remains a reliable technique for accurately assessing pulmonary vascular resistance, which cannot be accurately discerned noninvasively and can become of considerable concern if a patient is first diagnosed with a significant conotruncal defect beyond infancy. It may also help better define the coronary artery anatomy and small anomalous extracardiac vascular anomalies that would otherwise be difficult to identify. Catheterization is also a valuable to tool to gather additional physiologic information when a patient's clinical course is not consistent with their diagnosis or is not progressing as it should. Like echocardiography, angiography provides 2D information. However, it is invasive, requires sedation or general anesthesia, and exposes the patient to ionizing radiation (Table 1)

Cardiac Magnetic Resonance

Cardiac magnetic resonance (CMR) is an optimal 3D-imaging modality that is noninvasive, nonradiating, and can provide information on structural and functional details, including anatomical relationships, ventricular volumes and function, flow in selected vessels and chambers, myocardial perfusion and viability, and tissue characteristics [2] (Table 1).The field of view is virtually unlimited. The contrast agent, gadolinium, is very safe in children. CMR is now considered the "gold standard" for ventricular volume analysis in adult patients [3]. In some patients with complex conotruncal heart defects, CMR offers the 3D information needed preoperatively to determine the appropriate surgical course for a patient with a complex defect, such as the decision to attempt a two-ventricle repair or resort to a palliative single-ventricle series of operations.

Adult CHD is a class I indication for CMR, meaning that it provides relevant information; it may be used as an initial imaging technique; and its use has substantial support in the literature [10]. In adolescents and adults with repaired conotruncal lesions and residual defects or complications, CMR reveals the functional data required to help determine optimal timing of repeat surgery, such as the placement of a pulmonary valve in a patient with TOF. Special CMR techniques allow for tissue characterization and evaluation of myocardial perfusion, viability, and the presence of fibrosis and/or fat.

In younger patients, sedation or general anesthesia may be required due to long scanning times during which the patient must lie in the same position throughout the scan. CMR is limited in some patients with vascular stents or coils, which can cause artifacts from metallic-induced inhomogeneities in the magnetic field. A variety of pulse sequences can be used to minimize this artifact. CMR is also considered a relative contraindication in a patient with a pacemaker or defibrillator. The spatial resolution of CMR approaches that of cardiac computed tomography, but it can be more limited in the setting of a very fast heart rate and excess cardiac motion.

Cardiac Computed Tomography

CCT has the ability, like CMR, to provide 3D imaging and is noninvasive (Table 1). Submillimeter spatial resolution is possible due to increased speed and resolution available with multidetector computed tomography during the last 10 years. CCT provides detailed images of metal stents within vascular structures, and patients with pacemakers usually can be safely imaged [14] (Fig. 1).

Scanning time is quick, but intravenous contrast is required for adequate results, and imaging must be timed carefully to ensure that the contrast bolus is at its peak in the heart and related vessels. This requires the patient to breath-hold and be perfectly still for the scanning time, thus in many young patients sedation or anesthesia is still required. The other limitation of CCT is exposure to ionizing radiation, to which infants and children are particularly sensitive. The exposure can be similar to or much greater than cardiac catheterization. Functional imaging, with less temporal resolution than CMR, can be performed, but it requires electrocardiogram (ECG) gating and a concomitant significant increase in radiation. The preferred heart rate for optimal imaging is 60 beats/min, and beta-blockers are routinely administered to achieve this heart rate in older patients. In patients with very fast heart rates, ECG gating may not be possible. Newer scanners that overcome these limitations and deliver decreased radiation are on the horizon.

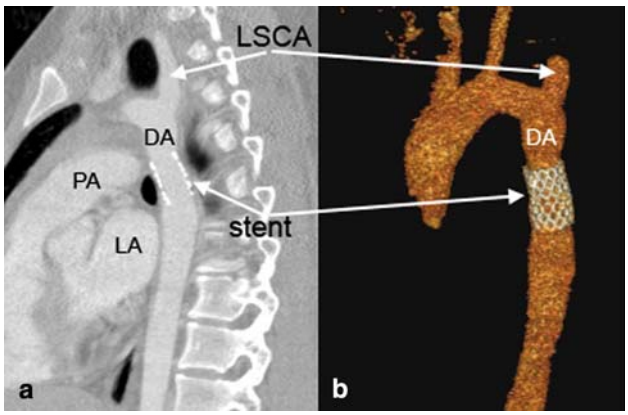

Fig. 1 CCT of stent. Stent is well visualized in DA. **a** Sagittal view of stent in aorta. **b** 3D reconstruction of aorta and stent. *DA* descending aorta, *LSCA* left subclavian artery, *PA* pulmonary artery, *LA* left atrium

Although knowledge and experience with CMR and CCT are becoming more widespread, not all centers have access to these technologies or to individuals with expertise in these areas.

Conotruncal Defects

Tetralogy of Fallot

TOF is the most common form of cyanotic CHD and comprises 3.5% to 9% of patients with CHD [11, 15]. The four components of TOF include the following: (1) malalignment ventricular septal defect (VSD), (2) overriding of the aorta, (3) right ventricular (RV) outflow tract obstruction/multilevel pulmonary stenosis, and (4) concomitant RV hypertrophy. Right aortic arch may occur in approximately 25% of patients. Absent pulmonary valve occurs in

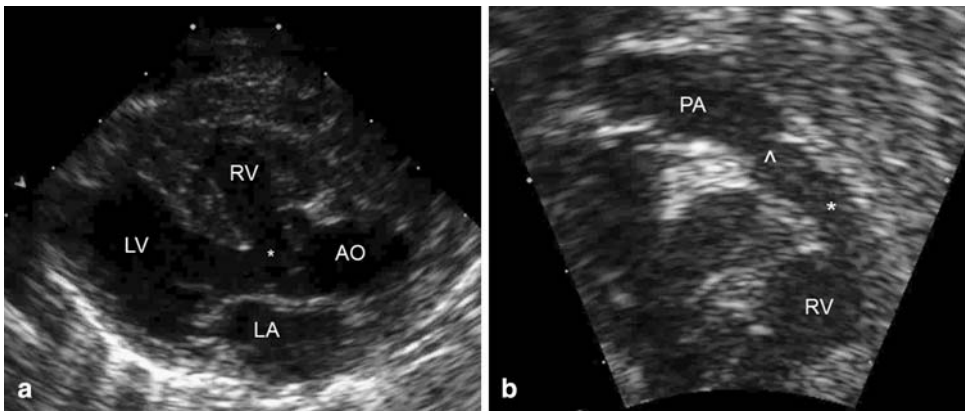

Fig. 2 TTE of TOF. **a** Parasternal long-axis view demonstrates VSD (*), AO, and RV hypertrophy. *AO* overriding aorta, *LA* left aorta. **b** Subcostal angle to outflow tract shows pulmonary stenosis. * Infundibular stenosis; ^ valvar stenosis; *PA* pulmonary artery

2.5% of TOF patients [24]. In this case, the pulmonary valve is rudimentary or incompletely formed and is usually both stenotic and regurgitant, and the pulmonary arteries are significantly dilated.

CXR will not provide details of the TOF diagnosis, but it may demonstrate an upturned cardiac apex caused by RV hypertrophy, concavity of the upper left heart border caused by underdevelopment of the pulmonary area, decreased lung field markings with diminished pulmonary blood flow, and possibly a right aortic arch.

At initial presentation during infancy, ECG is typically employed to define the components of TOF (Fig. 2). In a patient who requires a modified Blalock-Taussig shunt from the subclavian artery to the ipsilateral pulmonary artery to augment pulmonary blood flow, Doppler imaging be used to assess the shunt, and the modified Bernouli equation is used to determine the pressure gradient. TEE is used to assess the TOF repair intraoperatively.

Routine cardiac catheterization and angiography before repair is not necessary, but it may be used if there is concern for a significant coronary abnormality or very hypoplastic pulmonary arteries that are not well visualized by echo.

Patients with TOF face a lifetime of possible residual complications, including pulmonary stenosis, pulmonary regurgitation, RV dilation and dysfunction, life-threatening arrhythmias, and sudden cardiac death. Pulmonary valve replacement is often required and can decrease the incidence of these long-term complications. As the patient ages, acoustic windows by echocardiography can become poor. The abnormal shape, size, and function of the RV can be difficult to fully visualize and define accurately with echocardiography. Pulmonary regurgitation must also be reliably assessed. CMR has become the workhorse for reliably evaluating the adolescent and young adult with TOF to assess all of the above-mentioned problems. It is considered the clinical "gold standard" for right and left ventricular volumes [3]. An accurate pulmonary regurgitant fraction can be determined (Fig. 3). Differential pulmonary flow can be measured, and 3D reconstructions of the pulmonary arteries can reveal the need for further intervention [2, 15, 21]. Based on clinical findings and CMR parameters, pulmonary valve replacement can be recommended [1, 18]. The myocardium can also be assessed for fibrosis and viability.

Because surgical options lead to relatively good outcomes, many patients with TOF are surviving into adulthood. Patients require close monitoring throughout their lifetime and will likely encounter the need for a multitude of imaging approaches depending on the stage of their heart disease.

Truncus Arteriosus

Truncus arteriosus is a very uncommon CHD and occurs in 1.1% to 2.5% of cases, without significant sex predominance [11]. Rather than a separate aorta and pulmonary artery arising from the heart, in truncus arteriosus, there is a single arterial trunk, which is often dilated. The pulmonary arteries arise together or separately from the truncal root (Fig. 4a). One may arise more distally. The truncal valve is usually abnormal, with two to five cusps (or unicommissural) and a varying degree of stenosis and/or regurgitation (Fig. 4b). An interrupted aortic arch, typically between the left carotid and subclavian arteries (type B), may also occur [20]. This defect is accompanied by a large VSD due to deficient conal septum, and abnormalities of the atrioventricular valves may coexist. A right aortic arch may be found in $\leq$ 35% of patients.

Operative repair of truncus arteriosus was first developed approximately 30 years ago, and now most patients undergo correction during infancy. The initial surgery requires removal of the pulmonary arteries from the truncal root and placement of a conduit, with confluency, from the RV to the pulmonary arteries. The truncal valve may also require repair. Lifelong surveillance is required, and multiple interventions may be needed. Appropriate imaging is necessary to follow-up these patients.

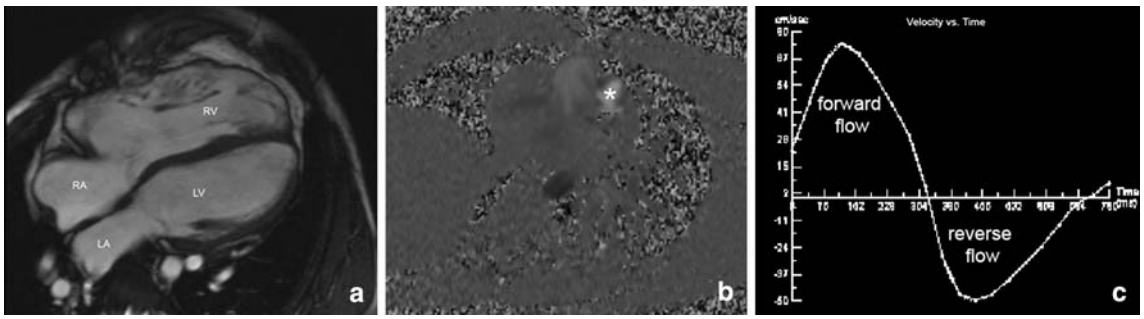

Fig. 3 Repaired TOF with 60% regurgitant fraction. **a** CMR four-chamber view with dilated RV. **b** CMR pulmonary artery flow with velocity-encoded phase image. * Pulmonary artery. **c** CMR graph of forward- and reverse-flow velocity in main pulmonary artery. *RA* right atrium, *LA* left atrium

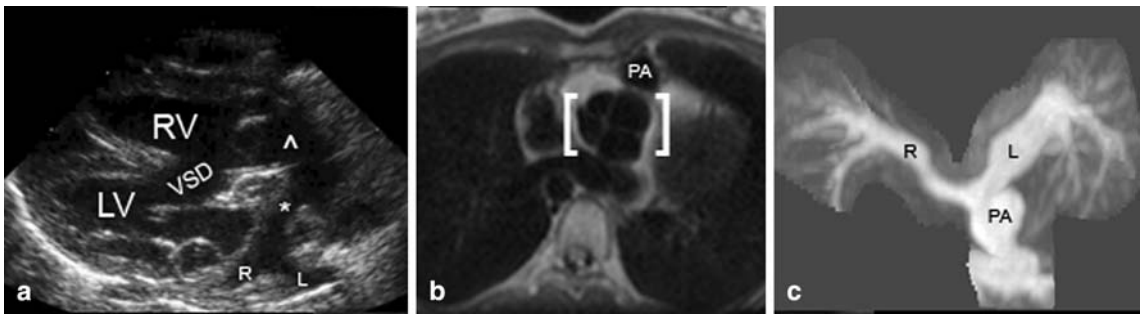

Fig. 4 **a** TTE of truncus arteriosus with pulmonary arteries (*) arising from truncal root (^). **b** CMR dark blood images of a quadricuspid aortic/truncal valve [] and anterior pulmonary conduit (PA). **c** CMR contrast 3D reconstruction of pulmonary arteries in repaired truncus. *R* right pulmonary artery, *L* left pulmonary artery

The initial diagnosis in the newborn is made by TTE. If there is any difficulty or uncertainty in locating the origin of the pulmonary arteries or identifying the aortic arch and its branches by echocardiography, further imaging with cardiac catheterization is typically used. At centers with experience in CMR, this alternative may be used to avoid an invasive procedure and exposure to radiation. More commonly, CMR is used in the older patient who has undergone surgical repair many years earlier. TEE is used in the operative suite. Postoperatively, TTE is used in the young patient to monitor the conduit and time its replacement as well as to monitor for residual truncal stenosis or insufficiency that will require intervention. If imaging windows are not adequate to make appropriate clinical decisions, CMR is then employed. This is most often the case in the older child or young adult. Detailed anatomical images, ventricular volumes and function, flow in the major arteries, and 3D reconstructions will assist the clinician in deciding on the need for further procedures (Fig. 4c), As the patient ages and echocardiographic acoustic imaging windows fail to provide thorough information, CMR is appropriate as a complementary or primary imaging method for follow-up. CCT or cardiac catheterization may be used if there are contraindications to CMR or if metal implants create artifacts that cannot be otherwise overcome. Because of the invasive nature, the need for anesthesia, and the exposure to radiation, most would choose to avoid catheterization unless an interventional procedure is contemplated.

Transposition of the Great Vessels

Transposition of the great vessels accounts for 5% to 7% of all CHD. There is a strong male predominance (60% to 70%), and extracardiac anomalies are less frequent than in other conotruncal defects. Up to 80% of cases are considered simple transposition, which, in addition to ventriculoarterial discordance, may include a VSD or intact septum, coronary abnormalities (Fig. 4), Left ventricular outflow obstruction, and patent ductus arteriosus [22]. TTE is the method of choice for the initial diagnosis of TGV and can accurately diagnose most of the important associated anomalies. Echocardiographic guidance and/or cardiac catheterization may be used in the infant requiring balloon atrial septostomy. Patients must be followed-up throughout life for residual defects.

Until approximately 20 years ago, the technique of repair was atrial baffling, i.e., the Mustard or Senning procedure. This redirects the returning systemic blood flow to the left ventricle (LV), and the pulmonary venous return drains to the RV, which remains the systemic pumping chamber. Many of these patients have now reached early

adulthood and require continued monitoring for systemic RV heart failure, superior vena cava baffle obstruction, baffle leak, and other complications [4, 7, 13]. Imaging with echocardiography is usually attempted, but imaging windows are often limited at this stage. CMR will allow a thorough anatomical and functional assessment, which is considered more accurate for assessing the RV [3, 6] (Fig. 5). In the case of a pacemaker or defibrillator or other contraindication for CMR, CCT or catheterization can be used to assess overall ventricular size, ventricular function, and details of baffle pathways (Fig. 6).

The arterial switch procedure has replaced the atrial baffle for simple TGV. Of particular concern after this operation is pulmonary artery stenosis, or branch pulmonary artery stenosis, due to the LeCompte maneuver performed during the arterial switch procedure. This maneuver places the right pulmonary artery and proximal main pulmonary artery anteriorly, i.e., draped over the aorta. The tension in this region may lead to poor growth and flattening of the main pulmonary artery or stenoses of the branches. With better fields of view, CMR or CCT may be better able than echocardiography to provide the details to determine the necessity for cardiac catheterization or reoperation to intervene in the presence of significant findings for a patient undergoing either type of repair.

Double-Outlet Right Ventricle

DORV accounts for $\leq$ 1.5% of CHD, and there is no significant sex difference. Both great arteries arise from the RV. There is loss of fibrous continuity between the AV valves and either semilunar valve. There is usually a VSD, which provides the only outflow for the LV. DORV is categorized based on the type of VSD present: subaortic, subpulmonic, doubly committed, or remote. The great arteries may be normally related, side-by-side, or malposed. Associated anomalies are suspected and further detailed. With the most common type of DORV—a subaortic VSD and side-by-side great arteries—pulmonary stenosis can be a concern. The next most common is Taussig-Bing anomaly: DORV with subpulmonary VSD, side-by-side great arteries, and often subaortic stenosis with an aortic coarctation or an interrupted aortic arch. A doubly committed VSD is less common [16]. A remote VSD is rare but may occur with a complete AV canal defect and complex heterotaxy syndromes. DORV may also occur in the setting of AV valve straddling or a significantly hypoplastic ventricle.

TTE most often allows important details of DORV to be delineated in infancy. In rare complex cases, cardiac catheterization with angiography or CMR may be used to provide complementary information during the preoperative planning stage. Depending on the nature of initial lesions and repair performed, various residual defects and possible complications will require lifelong care and imaging to assist in appropriate clinical management. As the patient ages, CMR or CCT may provide better image quality than echocardiography. CMR will provide more accurate functional information and does not expose the patient to radiation. Catheterization may be chosen for imaging especially if intervention is likely.

Dilated Aortic Root in Conotruncal Defects

Structural abnormalities have been detected by light and electron microscopic analyses in the great arterial walls of

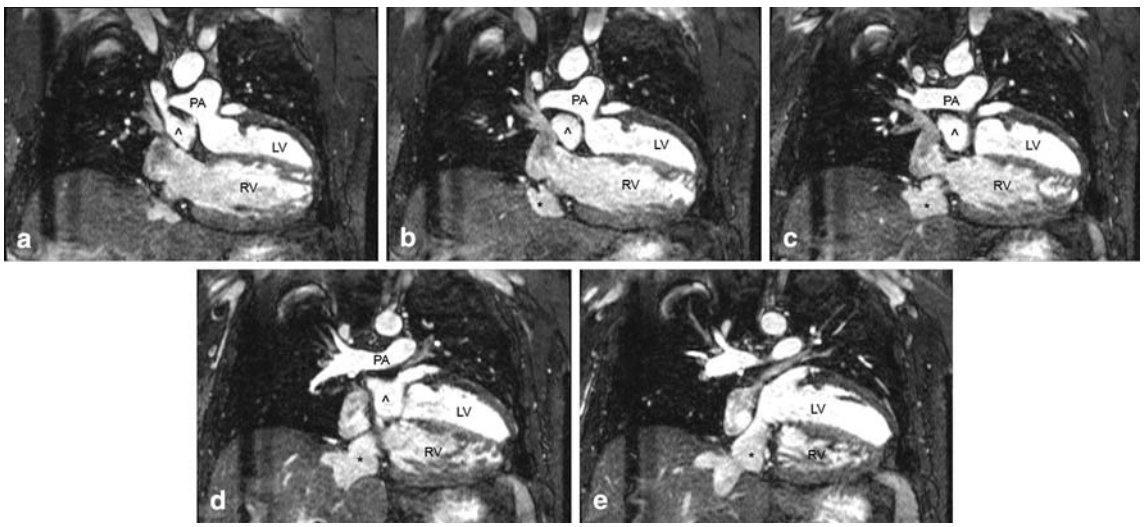

Fig. 5 **a–e** CMR of SVC (^) and IVC (*) baffle to the LV in a patient with Mustard repair for TGV. Pathways are widely patent. *PA* pulmonary artery

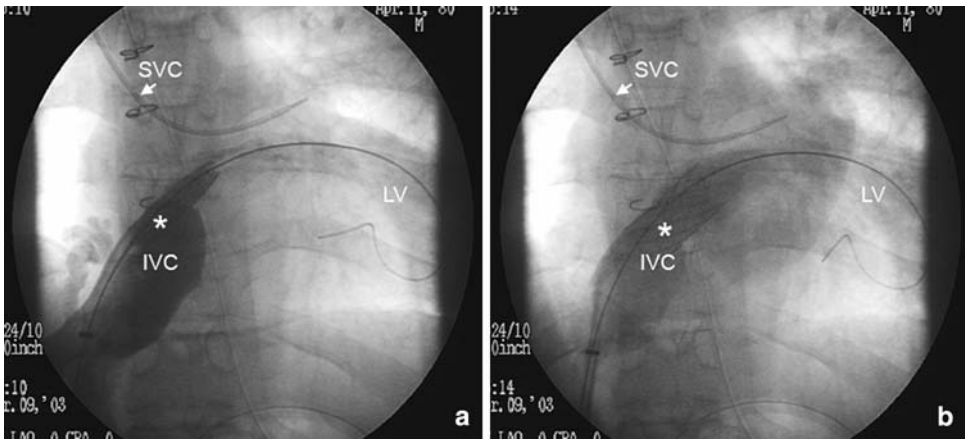

Fig. 6 Catheterization with angiography. **a** Narrow IVC (*) Mustard baffle with nonexpanded stent in place. **b** Stent expanded across IVC (*) Mustard baffle. *IVC* inferior vena cava, *SVC* superior vena cava

patients with conotruncal cardiac defects and may predispose these patients to dilatation, aneurysm, and rupture [8, 9]. Progressive aortic root dilatation has been reported in nearly half of patients with TOF [19]. Dissection of the aorta has been reported in a patient with repaired TOF [12]. Aortic root dilatation has also been reported in patients after repair of truncus arteriosus and TGV [23]. TTE and TEE images of the aorta in older patients may be suboptimal, whereas CMR provides an ideal 3D noninvasive ability to measure the true aortic diameter and allow for close monitoring in this patient population. CCT will provide excellent images as well, but it does introduce the risks associated with ionizing radiation.

Summary

Imaging of conotruncal defects begins early, with echocardiogram as the cornerstone for initial newborn diagnosis. Fetal echocardiogram, TTE, and TEE each have their place in management and follow-up of these often complex defects. To aid in assessing the intricacy of associated cardiac lesions, complementary modalities may include cardiac catheterization with angiography, CMR, or CCT. As the patient matures, the optimal imaging methods will change, depending on the patient characteristics and clinical findings. At each occasion, the advantages and disadvantages of each technique should be weighed (Table 1) so that optimal information can be provided to assist the clinicians caring for patients with conotruncal heart defects throughout their lifetime.

Future Perspectives

The ideal imaging technique would be one that is easily accessible, quick, noninvasive, nonradiating, requires no sedation or anesthesia, has no contraindications, and provides all needed diagnostic details. Currently, no single modality meets all of these criteria. It is likely that cardiac diagnostic imaging techniques will continue to be complementary and essential rather than one becoming all-encompassing. Technological advances will allow improvements in all modalities. Echocardiography is making advances in image capability, including 3D imaging. CMR continues to develop faster imaging sequences and is now being proven safe in patients with implanted devices. CCT is progressing in its ability to produce high-quality images with less radiation. Interventional catheterization procedures are allowing more definitive treatment without surgery. All of these advances will lead to higher-quality and safer diagnostic imaging capabilities that will continue to be needed in the field of CHD.

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