Transposition of the Great Arteries

Congenital Heart Diseases in Adults: Imaging and Diagnosis

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Contents

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1 Introduction

Transposition of the great arteries (TGA) is the second most common cyanotic congenital heart disease with a prevalence of 3.5/10,000 live births in Europe and a greater than twofold male predominance (Pexieder et al. 1995). It is characterized by a ventriculo-arterial discordance with the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle. In case of an atrioventricular concordance (with the right atrium connected to the right ventricle and the left atrium connected to the left ventricle) it is called dextro-TGA (d-TGA) due to the right-sided position of the ascending aorta. In case of an atrioventricular discordance (with the right atrium connected to the left ventricle and vice versa) it is called congenitally corrected-TGA (cc-TGA) or simplified levo-TGA (l-TGA) due to the mostly left-sided position of the ascending aorta. However, the use of the term "l-TGA" for this congenital heart disease (CHD) is unprecise as in cc-TGA with situs inversus the aorta is in "d"-position and there are also other CHD with "l"-position of the aorta like the double-outlet right ventricle (DORV).

Both forms of TGA have their own diagnostic and therapeutic challenges. In this chapter we focus on typical adult cardiac magnetic resonance (CMR) and computed tomography (CT) findings that result from the different types of surgical repair and which have a significant value for the early detection of typical long-term complications.

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2 Etiology

The etiology of TGA is still not fully understood and there are two major theories trying to explain the altered morphologies (Ferencz et al. 1995).

One theory suggests that the underlying mechanism is an alteration of the normal clockwise rotation of the aorta towards the left ventricle caused by an abnormal persistence of the subaortic conus (Goor and Edwards 1973). Following this theory TGA can be interpreted as one end in the spectrum of dextroposition of the aorta (Unolt et al. 2013).

The second theory sees the underlying cause in a rather linear than normal spiral development of the aortopulmonary septum putting the future aorta in contact with the anterior conus, which is connected to the right ventricle (de la Cruz et al. 1981).

In epidemiologic studies, TGA showed a higher prevalence with maternal infection, maternal intake of pesticides and ibuprofen, and ionizing radiation and in infants with diabetic mothers, whereas the periconceptional intake of folic acid may have a protective effect (Loffredo et al. 2001).

D-TGA can be accompanied by additional cardiac anomalies of which a ventricular septal defect (VSD) is most common with almost 50%. Furthermore, right ventricular outflow tract (RVOT) obstruction, valvular stenosis, and, less commonly, coarctation of the aorta (5%) can be found. Extracardiac anomalies are rare and mostly include the kidneys and the brain.

In cc-TGA additional cardiac anomalies are very common (>95%) and determine the clinical course of the disease. Besides a VSD, which is present in about 25% of patients, stenoses of the left ventricular outflow tract (LVOT) and the pulmonary arteries, hypoplasia of the ventricles, and tricuspid insufficiencies can be found. Moreover, due to the atrioventricular discordance, also the cardiac conduction system is altered and up to 30% of the patients are diagnosed with an atrioventricular (AV) block (Warnes 2006).

3 Clinical Presentation

Unlike other congenital heart defects d-TGA does not present an alternative model of blood circulation. With the systemic and pulmonary circulation running in parallel there is no blood oxygenation after closure of the ductus arteriosus unless there is an additional shunt. If not treated, d-TGA is the leading cause of cardiac death in neonates and infants (Samanek 2000). The children develop progressive cyanosis and have an average life expectancy of 2 months.

In contrast to d-TGA there is a physiologic situation with the systemic and pulmonary circulation running in row in cc-TGA. However, in difference to normal hearts, the systemic ventricle is not the left, but the right ventricle.

The clinical presentation of cc-TGA shows a broad spectrum depending on the type and degree of additional anomalies. In the rare absence of additional cardiac anomalies (except of the cardiac conduction system) cc-TGA can remain undetected for a long time until it becomes clinically apparent in adulthood with the symptoms of systemic heart failure. Survival to the seventh and eighth decades has been reported. Although the right ventricle can compensate the systemic pressure for many years by hypertrophy, it is designed as a subpulmonary volume pump with a complex triangular shape and not as a concentric pressure pump like the left ventricle. Moreover the coronary artery anatomy is concordant and the hypertrophic RV is perfused by a single right coronary artery, which might cause an imbalance between oxygen demand and oxygen supply. Increasing tricuspid regurgitation is common in cc-TGA patients.

Pulmonary stenosis can be found in about 40% of patients and is mostly subvalvular, but also valvular stenosis occurs. In combination with a VSD varying degrees of cyanosis can be found.

4 Treatment Options and Prognosis

4.1 D-TGA

In the industrialized countries d-TGA can be determined during ultrasound screening in pregnancy in many cases and a precise prenatal diagnosis decreases both the infantile and the maternal risk and improves outcome. Still, d-TGA is one of the most difficult CHD to diagnose prenatally

because of the relatively normal appearance in the four-chamber view (Jouannic et al. 2004).

For patients without a prenatal diagnosis, CHD should be considered if there is cyanosis not responding to oxygen in the immediate newborn period. In fetal d-TGA diagnosis, birth should be planned in a center where a balloon atrial septostomy (BAS) can be performed.

Therapy of d-TGA consists of pharmacological therapy, initial palliative procedures to improve oxygenation by connecting the systemic and pulmonary circulations, and an early corrective surgery.

4.1.1 Prostaglandin and Balloon Atrial Septostomy

In hypoxic neonates prostaglandin is administered to maintain patency of the arterial duct. Additionally this large-volume shunt can cause left atrial hypertension and enlargement which is stretching the foramen ovale, resulting in improved mixing.

If prostaglandin therapy is insufficient a BAS has to be considered. Usually a BAS results in an acute and substantial improvement in oxygen saturation and can be performed under echocardiographic as well as fluoroscopic control. The benefits of a BAS have to be weighed against the rare but threatening risks like vascular trauma, arrhythmias, and atrial perforation.

4.1.2 Surgical Therapy

Corrective surgery of d-TGA is performed a few days after birth after transition from fetal to neonatal circulation and initiation of enteral nutrition. It is common to plan surgery within the first 2 weeks after diagnosis. This approach might avoid some adverse long-term effects of the unstable d-TGA circulation.

Before the anatomical correction became procedure of choice for surgical treatment of patients with d-TGA, the atrial switch operation was performed to achieve a physiological correction of blood flow since the 1960s. This type of correction involves the creation of a two-way systemic venous tunnel (baffle) at the atrial level to direct the deoxygenated systemic venous return towards the mitral valve, the LV, and eventually the pulmonary artery. Accordingly pulmonary venous blood is directed through the pulmonary venous baffle towards the tricuspid valve, the RV, and eventually the aorta. Depending on the used material for the baffle this procedure is also called Senning (use of autologous tissue) or Mustard (use of synthetic material) operation after the surgeons who described the procedures first. The long-term benefits of this procedure are moderate, and there are specific long-term complications that are associated with increased morbidity and mortality (Williams et al. 2003). In addition to the development of arrhythmias, systemic ventricular dysfunction is the most important sequela, being mainly caused by the nonphysiological systemic position of the right ventricle (RV). Moreover, hemodynamically relevant stenosis or leakage of the systemic venous baffle, particularly of the superior limb, can occur.

Since the late 1980s the arterial switch operation (ASO) has replaced the atrial switch procedure. The first successful ASO was performed in 1975 by Jatene and is now the procedure of choice in all centers (Jatene et al. 1976).

Although the concept of ASO is simply the restoration of the normal anatomic arrangement where the arteries are transferred to the proper ventricle, it is still one of the most complex and technically challenging operations in CHD. During this procedure the aorta and the pulmonary artery are transected above the sinuses and switched into their new position. The pulmonary trunk is hereby placed anterior to the ascending aorta (Lecompte maneuver) and the pulmonary arteries embrace the aorta (Fig. 1). The ASO includes the detachment of the coronary arteries along with a "button" from the aortic sinus and their transfer to the sinus of the neo-aorta.

The surgical technique of the ASO determines its potential short- and long-term complications. Particularly the meticulous reinsertion of the coronary artery origins is of highest importance for a successful procedure. Stenosis of the coronaries can result in immediate ischemia and early mortality is mostly due to difficulties with this transfer. However obstruction of the coronary arteries by growth can also be a long-term complication and visualization of the course of the proximal coronaries is an important component of lifelong follow-up care. Further potential long-term

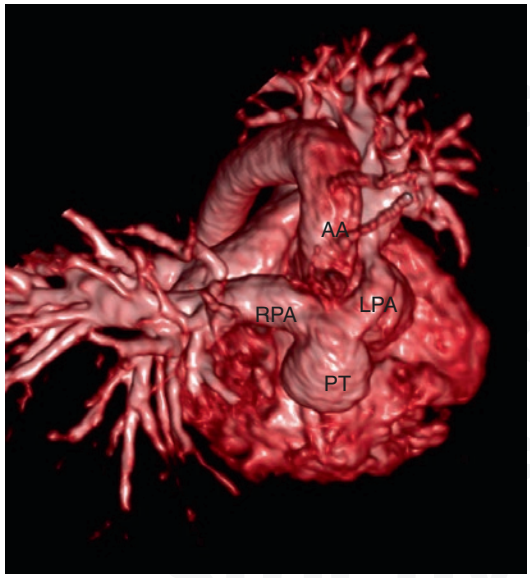

Fig. 1 3D volume rendering after arterial switch operation. The pulmonary artery branches are encompassing the ascending aorta. The pulmonary trunk is in anterior position after Lecompte maneuver. *AA* ascending aorta, *PT* pulmonary trunk, *RPA* right pulmonary artery, *LPA* left pulmonary artery

consequences are neo-pulmonary stenosis, neo-aortic regurgitation, and neo-aortic root dilatation.

Survival into adulthood is common and adults after ASO represent a large group in adult congenital heart disease clinics (Fricke et al. 2012).

When a d-TGA coexists with a pulmonary stenosis and a large subaortic VSD a Rastelli procedure can be performed. In this operation a patch is placed to conduct blood from the LV through the VSD to the aorta (Fig. 2). Additionally a valved conduit conducts blood from the RV to the pulmonary artery. The Rastelli procedure has the advantage that the LV is in systemic position. However, there are also typical long-term sequelae of which conduit degeneration and stenosis are of foremost importance.

4.2 CC-TGA

Therapy of cc-TGA depends on the type and severity of associated cardiac anomalies. If isolated, pharmacological therapy focuses on

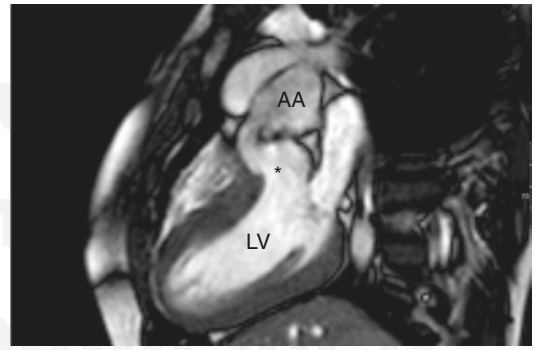

Fig. 2 Steady-state free-precession LVOT view after Rastelli procedure. A patch is placed to conduct blood through the VSD to the aorta (*asterisk*). *AA* ascending aorta, *LV* left ventricle

treatment and the prevention of RV failure. The basic principles do not differ from treatment of LV failure and focus on reduction of the pre- and afterload and improvement of ventricular contractility.

As the RV is in systemic position, systemic pressure causes hypertrophy which is followed by a dilatation of the RV as well as the tricuspid annulus. This mechanism causes the steady increase in tricuspid regurgitation over time. Long-term prognosis of adults with even mild systemic RV dysfunction or tricuspid regurgitation is poor and patients can develop severe tachyarrhythmia unpredictably (Filippov et al. 2016). In high-degree AV block the implantation of a pacemaker has to be considered.

As in many CHD surgical management of cc-TGA has evolved over the last decades. In the early 1960s the physiological repair was introduced. It consisted of repair of associated cardiac anomalies and surgical palliations to improve the prognosis. However in this type of repair the RV remains the systemic ventricle and long-term follow-up showed limited outcome in these patients.

During the 1990s the physiological repair was replaced by the double-switch operation. In this approach a Senning/Mustard repair is performed on the atrial level. In the rare cases in which the LV is in systemic or close to systemic pressures as a result of a VSD and

pulmonary stenosis, the atrial switch procedure can be combined with a Rastelli operation and an RV to PA conduit. More often, however, the LV has to be prepared for the systemic position before an additional arterial switch procedure can be performed. This preparation is achieved by a pulmonary artery banding procedure to induce LV hypertrophy. Perioperative mortality is reported to vary between 0 and 10% (Duncan et al. 2003). Late results after double-switch procedure are satisfactory and 10–20-year survival after surgery was reported as up to 85% with the majority of survivors in a good functional outcome (New York Heart Association functional class I or II) (Murtuza et al. 2011).

5 Imaging Goals

All patients with TGA are in need of regular follow-up examinations. Here, the knowledge of the type of surgical correction is crucial for postoperative imaging as each type of repair has its specific long-term complications and imaging protocols need to be adapted accordingly.

5.1 D-TGA After Atrial Switch

After atrial switch (Senning or Mustard) one imaging goal is to evaluate the status of the systemic right ventricle. This includes determination of RV size and function as well as the assessment of RV hypertrophy. It is known that a certain degree of RV hypertrophy is beneficial for maintaining RV systolic function (Grothoff et al. 2012). An excessive degree of hypertrophy, however, can deteriorate RV function as the single coronary artery is unable to provide a sufficient oxygen supply.

A second aim is to visualize the complex baffle anatomy. Shrinking and stenoses particularly of the superior limb of the systemic venous baffle can occur due to scarring and can cause upper venous congestion. Stenoses of the inferior limb are less common. Small baffle leaks are even more common than stenoses. Although hemodynamically insignificant in most cases they bear the risk of paradoxical embolus and stroke.

5.2 D-TGA After Arterial Switch

After arterial switch operation imaging has to focus on the reinserted coronary arteries as well as dilatation of the neo-aortic root with aortic regurgitation. Particularly the visualization of the proximal course of the coronaries using noninvasive cross-sectional imaging can avoid repeated diagnostic catheterization in this delicate patient group. Further attention has to be put on distortion of the RV outflow tract and stenoses of the pulmonary arteries that are encompassing the ascending aorta.

5.3 D-TGA After Rastelli

Besides the assessment of biventricular function imaging after Rastelli procedure has to exclude residual ventricular septal defects as well as subaortic stenosis. Furthermore the function of the ventriculo-pulmonary conduit has to be evaluated.

5.4 CC-TGA

If isolated, the diagnosis is often overlooked. Typical imaging findings of cc-TGA are the characteristics of the RV in the left position. These include the moderator band, the pronounced trabeculation of the myocardium, and the funnel-shaped muscular outflow tract (conus or infundibulum). Furthermore the parallel arrangement of the pulmonary trunk and the ascending aorta (side-by-side position) is common (Fig. 3). If surgically corrected the findings in cc-TGA depend on the type of repair (double switch or atrial switch with Rastelli procedure).

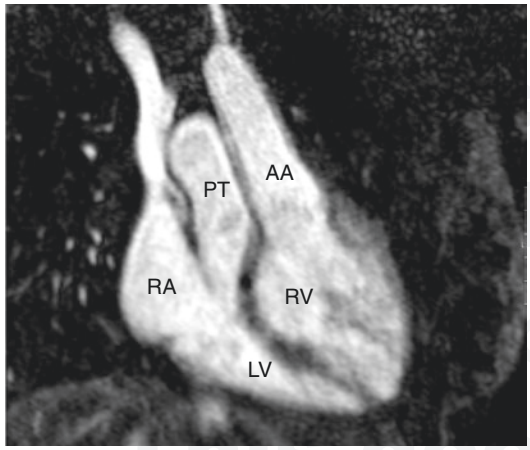

Fig. 3 MR angiography demonstrates the side-by-side position of the ascending aorta and the pulmonary trunk—here in cc-TGA with additional atrioventricular discordance. *RA* right atrium, *LV* left ventricle, *PT* pulmonary trunk, *AA* ascending aorta, *RV* right ventricle

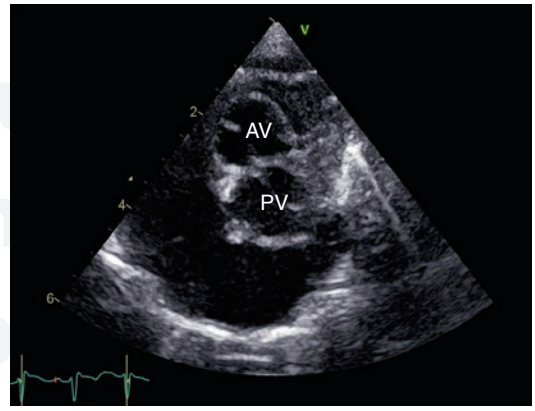

Fig. 4 Parasternal short-axis echocardiography showing the aortic valve anterior and rightward from the pulmonary valve. *AV* aortic valve, *PV* pulmonary valve

6 Noninvasive Imaging Techniques

6.1 Echocardiography

Echocardiography is the first-line imaging tool in CHD. In the hands of an experienced examiner it can provide a broad spectrum of morphological and functional information. Also in TGA it is used for the initial diagnosis and for surgical planning. In postoperative follow-up it should be complemented by cross-sectional imaging, predominantly by MRI.

In the preoperative assessment it is key to define the ventriculo-arterial connection. Here the subcostal imaging planes are particularly valuable. Sagittal imaging planes typically show the aorta positioned anterior to the pulmonary artery. In a short-axis orientation the aortic and the pulmonary valve can be visualized simultaneously, which confirms the ventriculo-arterial discordance (Fig. 4). Associated ventricular septal defects are best visualized by subcostal and parasternal short-axis planes. If the pressure in both ventricles is equal additional ventricular septal defects can be undetectable if only Doppler echocardiography is used. Therefore careful B-mode imaging is always required. For visualizing the aortic arch to exclude aortic coarctation (which is present in 5–10% of patients) a suprasternal approach is suitable. Preoperative visualization of the coronary arteries is challenging as there are numerous anatomic variations. Multiple imaging planes are required, of which the parasternal short-axis, the apical four-chamber, and the left oblique subcostal views are the most useful ones.

In postoperative imaging ultrasound has to address the complications that are inherent to the type of correction as mentioned above. These can occur immediately after correction or years later. After arterial switch, again, visualization of the coronary arteries is of great importance and should be performed with both two-dimensional B-mode and Doppler echocardiography. However, diagnostic imaging of the coronaries is not always possible due to acoustic window constraints. Assessment of myocardial contractility might be helpful but cannot differentiate whether a preserved contractility is caused by a normal coronary artery supply or by a collateral circulation (Taylor et al. 2005). Clearly, other imaging modalities are necessary to overcome these limitations in late follow-up examinations. As diagnostic cardiac catheterization is invasive and therefore not the ideal modality for regular follow-up examinations in adolescents and young adults, noninvasive coronary MR- or CT-angiography should be considered.

Besides stenoses of the coronaries also stenoses of the branch pulmonary arteries following the Lecompte maneuver have to be excluded in follow-up. In ultrasound these can be best visualized with a high parasternal short-axis view. In Doppler echocardiography gradients of >25 mmHg indicate a relevant stenosis and need further intervention.

After atrial switch (Senning or Mustard) systemic and pulmonary venous baffles have to be examined for obstruction or leaks. Here transesophageal echocardiography (TEE) is superior to transthoracic echocardiography (TTE). Midesophageal (ME) views using sweeps and particularly ME four-chamber views with flexion and rotation of the probe allow visualization of both baffles. Doppler echocardiography may determine the severity of stenoses, which mostly occur at the superior limb of the systemic venous baffle. Also injection of ultrasound contrast agent into an upper body peripheral vein can assess the function of the upper systemic venous limb. In the absence of an obstruction the contrast rapidly moves from the superior vena cava towards the tricuspid valve and no contrast can be seen in the inferior limb of the baffle. In the presence of a relevant baffle obstruction the contrast is also directed through collaterals to the lower limb of the baffle.

Assessment of systemic RV function, size, and hypertrophy can be difficult in echocardiography due to acoustic window constraints when the RV is in the right position. In TTE the most useful planes are the parasternal long-axis (PLAX) and parasternal short-axis (PSAX), subcostal long-axis, and parasternal RV inflow views. Accurate assessment of the systolic function of the RV is achieved by measuring one or many echocardiographic indices like the tricuspid annular plane systolic excursion (TAPSE), which is defined as the excursion of the tricuspid annulus from diastole to systole, and is measured typically at the lateral annulus using M-mode. For a better reproducibility of the RV status and imaging without limitations caused by the retrosternal position of the RV and patient constitution, MRI should be used in regular follow-up examinations to determine size and function.

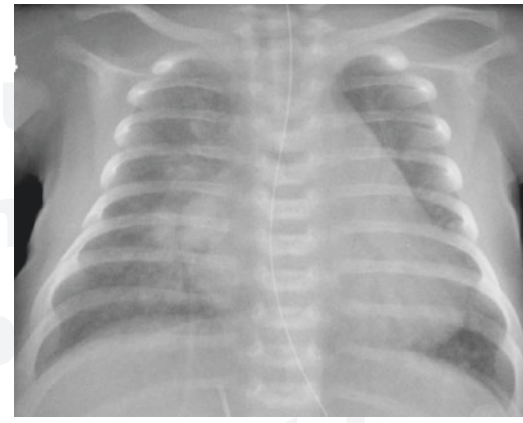

Fig. 5 Radiograph in uncorrected d-TGA. Note the absence of the ascending aorta on the right and the absence of the descending aorta on the left side

6.2 Radiography

With the advent of echocardiography and the cross-sectional imaging modalities the role of chest radiography in TGA has changed. It is no longer used for diagnosis purposes but to provide an overview over the general cardiopulmonary status. Typical findings in uncorrected TGA are an abnormally straight vascular pedicle caused by the loss of the normal arterial relationships. The ascending aorta is not visible on the right side and the aortic knob as well as the descending aorta is not visible on the left side (Fig. 5). In cc-TGA frequently dextrocardia can be found.

In the early postoperative period chest radiography is used for diagnosing early postoperative complications and is the method of choice for the close-meshed follow-up of the cardiopulmonary status. In late follow-up radiography is furthermore helpful in the evaluation of stent integrity after stent implantation in the superior limb of the systemic venous baffle.

6.3 Computed Tomography

In the era of low-dose computed tomography (CT) the spectrum of indications for this modality has extended, even in CHD. With prospective ECG-triggering techniques detailed morphologic information at reasonable radiation doses can be

obtained. Today the radiation dose in coronary computed tomography angiography (CCTA) is lower than that in standard percutaneous coronary angiography and the complication rate is lower (Meinel et al. 2015). A recent study showed that the high temporal and spatial resolution of CCTA can be used to reliably visualize the coronaries after arterial switch procedure in adolescents and young adults (Szymczyk et al. 2018). In knowledge of the great variance of the reimplanted coronaries and their importance for the individual outcome the visualization of the proximal course is part of most routine follow-up protocols.

Also after atrial switch CT can be helpful in long-term follow-up, particularly after stent implantation, e.g., in the upper limb of the systemic venous baffle. Here, MRI as the alternative cross-sectional modality has problems in visualizing the in-stent lumen due to susceptibility artefacts.

Independent of the type of surgery CT is the first-line modality in perioperative complications like hemothorax, pneumothorax, and venous thrombosis. These CT scans can be performed with low radiation exposures of approximately 0.5 mSv effective dose.

In complex vessel anatomy CT is also used for preoperative planning. The relationships of the arteries and the surrounding structures can be depicted in a submillimeter spatial resolution. A 3-dimensional volume rendering gives the surgeon an impression of the operative situs. Contrast agent is administered adapted to weight in a single bolus. The acquisition of a single phase with contrast in all major vessels is sufficient in most cases.

6.4 Magnetic Resonance Imaging

Cardiac MRI is the imaging modality that can acquire the widest range of information in children and adults with CHD. It is also the ideal method for regular follow-up examinations as it is free of ionizing radiation and not limited by patient constitution or a sufficient acoustic window. CMR is only rarely used for preoperative imaging. Without sedation it requires a certain degree of patient compliance over a time of at least 30–40 min. In children this is usually possible from the age of 8–9 years.

6.4.1 Morphology

In CHD, CMR imaging should start with an overview over the postoperative situs in a transverse orientation using black-blood or steady-state free-precession (SSFP) images. This gives a first impression of the dimensions of the chambers and great vessels as well as the degree of ventricular hypertrophy and reveals the type of repair and modifications. In a next step the direct acquisition of freely angulated planes in the three-dimensional space is possible. In d-TGA after atrial switch procedure, this is particularly helpful in visualizing the upper and lower limb of the systemic venous baffle. This is best achieved by a paracoronal view with an angulation from the superior to the inferior vena cava (Fig. 6a, b). If angulated properly, both limbs of the baffle can be visualized at once and obstructions can be detected. Implanted stents in the upper limb can cause susceptibility artifacts and make the assessment of stent patency impossible (Fig. 6c).

After arterial switch procedure the transverse imaging planes nicely demonstrate the anterior position of the pulmonary trunk after Lecompte maneuver (Fig. 7) and potential stenoses of the branch pulmonary arteries.

A substantial improvement in spatial resolution was the introduction of the three-dimensional whole-heart sequence. This ECG-gated sequence was developed to enable CMR coronary imaging and provides isotropic submillimeter voxels in free breathing. The use of acceleration techniques, of which parallel imaging is the most common one, results in acquisition times of about 10 min for a complete 3D dataset of the cardiovascular structures of the mediastinum during systole and/or diastole. In d-TGA after arterial switch 3D whole-heart imaging can reliably visualize the course of the proximal reinserted coronary arteries up to a heart rate of 100 beats/min and can replace the invasive and radiation-intense cardiac catheterization (Greil et al. 2017).

Furthermore, the 3D whole-heart sequence is usually performed without the administration of a contrast agent and may therefore replace the contrast-enhanced MR angiography (CE-MRA) as far as the central vessels, e.g., the pulmonary branches after Lecompte maneuver, are concerned. This is particularly helpful in an era of uncertainty regarding the possible deleterious effects of gadolinium deposition in the central nervous system (see also below).

6.4.2 Function

Functional assessment is one of the most important elements in follow-up examinations in patients with TGA. Impaired function in combination with other parameters may lead to an indication for reoperation or reintervention. Therefore the meticulous determination of ventricular size and ejection fraction and the early detection of tendencies are part of any CMR follow-up examination. Assessment of RV dimensions and function is

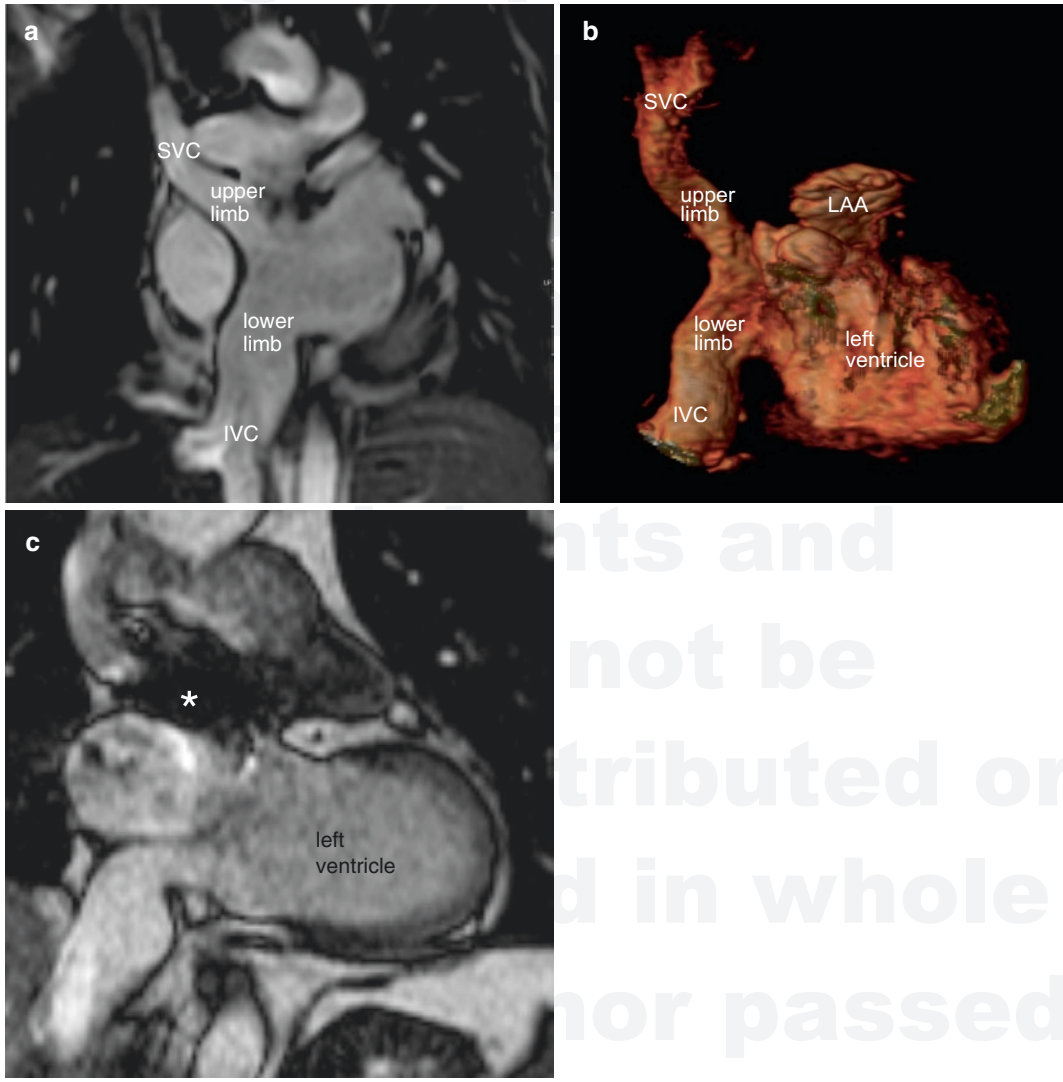

Fig. 6 (**a**) Steady-state free-precession paracoronal view after atrial switch operation shows the upper and lower limb of the systemic venous baffle conducting blood from the superior and inferior vena cava to the left ventricle. (**b**) 3D volume rendering of the same patient. (**c**) Stent in the upper limb of the systemic venous baffle after treatment of stenosis causing artefacts

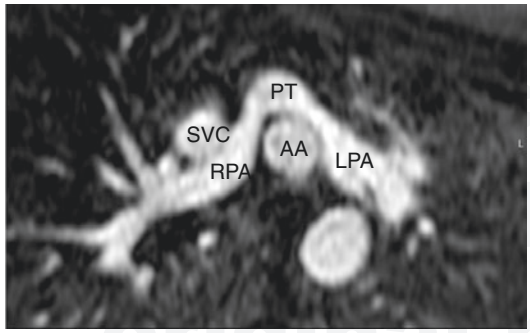

Fig. 7 MR angiography after arterial switch procedure. The proximal RPA and LPA are predilection sites for postoperative stenoses. *AA* ascending aorta, *PT* pulmonary trunk, *RPA* right pulmonary artery, *LPA* left pulmonary artery, *SVC* superior vena cava

difficult by echocardiography due to its retrosternal position and its complex chamber shape, which does not allow for application of an accurate geometric model. This is even truer for right ventricles after atrial switch procedure or in cc-TGA in which the systemic RV and its tricuspid valve are exposed to systemic arterial afterload. Therefore, the most commonly used two-dimensional echocardiography provides rather qualitative than quantitative information on RV dimensions and volume indices.

CMR has become the gold standard in the assessment of biventricular volume and mass. Mostly steady-state free precession (SSFP) sequences with a high contrast between myocardium and blood pool are used. Classical gradient echo sequences provide a lower contrast-to-noise ratio but can be helpful in reducing artifact, particularly banding artifacts in 3T imaging.

In the symmetrical LV CMR shows excellent reproducibility, and in the asymmetrical RV delineation of the contours is more challenging but reproducibility is still good (Kilner et al. 2010). For RV volume measurement various stack orientations can be used, each of them offering particular advantages and disadvantages. The short-axis orientation offers a good delineation between the diaphragm and the thin inferior wall of the RV but has problems with the delineation of the complexly shaped tricuspid valve. The four-chamber-view orientation can clearly depict the tricuspid valve, but lacks differentiation between inferior myocardium and diaphragm. A good compromise is the use of transversal slices, typically with a slice thickness of 6 mm without gap.

However, no matter what orientation is used, consistency in measurements is most important especially when RV parameters are assessed over many years in regular follow-up examinations and measurements are performed by various investigators. Consensus criteria and a training of postprocessing physicians and technicians are helpful in reaching consistency and a good interobserver variability at least within one institution. Commercially available automated or semiautomated contour detection programs for the geometrically complex RV still do not show satisfying results and time-consuming manual correction is mandatory.

When delineating the borders of the RV, the endocardial contour should be drawn along the compacted layer of the RV myocardium and exclude the trabeculations. Certainly this approach results in a systematic error namely an overestimation of the blood pool and an underestimation of the myocardial mass, particularly in hypertrophied systemic RVs in cc-TGA and d-TGA after atrial switch. However, it is impracticable and time consuming to delineate the numerous RV trabeculations and an attempt will result in lower intra- and interobserver reproducibility (Kilner et al. 2010). The myocardial mass is best assessed in end diastole as the hypertrophied trabeculations appear to merge in end systole and cannot be demarcated from the compacted layer.

In d-TGA after atrial switch procedure functional SSFP imaging can also be used for the detection of baffle leaks. With an adequate in-plane visualization the shunt jets can directly be visualized. Small leaks, however, may not be detectable and CMR is inferior to contrast-enhanced echocardiography in the detection of leaks (Lu et al. 2012).

6.4.3 Flow Measurement

Phase-contrast (PC) flow measurement is part of any CMR protocol in CHD. It is the only modality which can acquire quantitative hemodynamic

information noninvasively. In clinical routine mostly two-dimensional PC CMR is used to assess flow velocity and regurgitation. In TGA there are numerous applications for this method depending on the type of surgery: After arterial switch operation PC CMR is used to quantify stenoses of the branch pulmonary artery after Lecompte maneuver in a paraxial in-plane orientation. Furthermore PC flow can quantify the regurgitation fraction in neo-aortic insufficiencies. After atrial switch procedure flow measurement is helpful in quantifying stenoses of the upper limb of the systemic venous baffle and in quantifying shunts caused by baffle leaks by comparing the aortic and pulmonary forward flow. After Rastelli procedure PC CMR focuses on the assessment of stenoses or insufficiencies of the pulmonary conduit and obstructions of the left ventricular outflow tract.

A further development in PC flow measurement is time-resolved three-directional velocity encoding (4D PC CMR). Data acquisition can be performed in free breathing within 10–12 min. 4D flow provides full volumetric coverage of the great arteries and can improve hemodynamic evaluation in complex postsurgical anatomy. Postprocessing can be performed retrospectively and data quality is no longer dependent on correct pre-acquisition angulations as it is in 2D flow. Furthermore new flow parameters like helicity and vorticity of the flow can be visualized. Although robust and available for some years, 4D flow is still mostly used in scientific settings and reference values from larger cohorts are not available yet. Nevertheless, it will be the future of flow measurement in CHD.

6.4.4 Tissue Characterization

Systemic dysfunction is a common sequel in the long-term follow up of TGA patients with systemic RVs and there has been much debate for the reason of RV impairment. Tricuspid insufficiency, a complete atrioventricular block, and arrhythmias have been identified as risk factors. Also myocardial scarring assessed by late gadolinium enhancement (LGE) imaging can have a deleterious effect on RV function. Over the past years LGE techniques have evolved and been established as reliable and robust tools to visualize biventricular myocardial scars with high reproducibility. Phase-sensitive inversion-recovery (PSIR) DE sequences provide a better and more consistent image quality compared to DE sequences with magnitude detection. In d-TGA patients with atrial switch procedure performed in the late 1970s and the 1980s (before the arterial switch procedure became the method of choice) myocardial scars were found rarely in both the RV and the LV (Preim et al. 2013). Also in uncorrected cc-TGA scars are uncommon. In older patient cohorts after atrial switch scars were found more frequently, probably due to different surgical techniques, and also correlated with RV ejection fraction (Babu-Narayan et al. 2005). In this context it is important to notice that hyperenhancement adjacent to prosthetic patches, e.g., after closure of ventricular septal defects or Rastelli procedure, must not be considered as myocardial scar but rather as adhesion of contrast agent to the patch (Fig. 8).

LGE can only detect adjacent fibrosis with a volume of >0.5 mL but is unable to detect diffuse fibrosis. T1 mapping techniques might overcome this constraint as they are able to detect fibrosis by measuring tissue-specific relaxation times. After atrial switch diffuse fibrosis is more pronounced in the myocardium of the

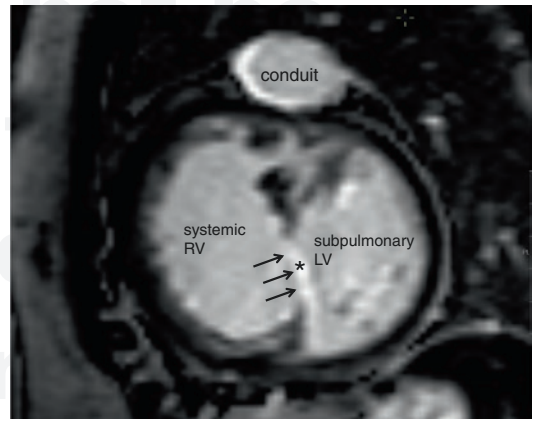

Fig. 8 Phase-sensitive inversion recovery (PSIR) late gadolinium enhancement image in d-TGA after atrial switch procedure with a conduit to the pulmonary trunk demonstrating adhesion of contrast (*arrows*) at a large VSD patch (*asterisk*). *LV* left ventricle, *RV* right ventricle

systemic RV of adults as compared with controls in equilibrium contrast mapping (Plymen et al. 2013). However, to quantify the extracellular volume, which is enlarged in diffuse fibrosis, the application of gadolinium is necessary. In our institution we try to avoid the repeated administration of gadolinium in regular follow-up examinations in children, adolescents, and young adults as it is unclear whether deposition in the central nervous system might have adverse long-term effects.

An overview over the key points in imaging is given in Box 1.

Box 1 Key Points in Imaging TGA

- Radiography:
  - Overview cardiopulmonary status and course
- Echocardiography:
  - First-line imaging modality, prenatal diagnosis, preoperative planning, follow-up examinations

Preoperatively:
- Detection of ventriculo-arterial discordance
- Parallel position of great arteries
- Detection of associated cardiac anomalies (e.g., VSD, pulmonary artery stenosis, coarctation of the aorta)

Postoperatively:
- Assessment of biventricular function
- *After arterial switch procedure*: detection of pulmonary branch artery stenoses
- *After atrial switch procedure (Senning/Mustard)*: evaluation of systemic RV (function and hypertrophy), detection of baffle stenoses and leaks, assessment of tricuspid insufficiency
- *After Rastelli procedure:* exclusion of residual VSD and subaortic stenosis, assessment of pulmonary conduit function

- Computed tomography
  - Imaging of perioperative complications, preoperative planning
- Cardiac magnetic resonance
  - Routine in follow-up examinations
  - Gold standard in evaluation of biventricular function and mass
  - Imaging of scar and fibrosis
  - Quantification of stenoses, insufficiencies, and shunts
  - *After arterial switch*: detection of coronary artery and pulmonary branch artery stenoses
  - *After atrial switch*: evaluation of systemic RV, detection of baffle stenoses and leaks, evaluation of tricuspid insufficiency
  - *After Rastelli:* exclusion of residual VSD and subaortic stenosis, assessment of pulmonary conduit function

Conclusion

Echocardiography is the first-line imaging modality in TGA and can already detect the ventriculo-arterial discordance during screening examinations in pregnancy. Also preoperative planning is mostly done by echo. After surgical correction imaging is an essential part of follow-up examinations for the detection of morphologic and hemodynamic changes. The postoperative imaging protocol strongly depends on the type of repair of which each has its specific long-term complications. Limitations in echocardiography in visualizing the RV and the coronary arteries can be overcome by CMR. This modality provides the most detailed information in morphology, function, and tissue characterization and is routinely performed in all tertiary care centers. Pathologies associated with a systemic RV can be imaged in freely angulated slices or in 3D without limitations by patient constitution. Modern imaging sequences like T1 mapping and 4D PC CMR may provide additional information but have to be evaluated in regard

to their clinical significance. Computed tomography can be valuable in detailed imaging of the morphology and is the first-line modality in perioperative complications.

References

Babu-Narayan SV, Goktekin O, Moon JC et al (2005) Late gadolinium enhancement cardiovascular magnetic resonance of the systemic right ventricle in adults with previous atrial redirection surgery for transposition of the great arteries. Circulation 111:2091–2098

Duncan BW, Mee RB, Mesia CI et al (2003) Results of the double switch operation for congenitally corrected transposition of the great arteries. Eur J Cardiothorac Surg 24:11–19

Ferencz C, Brenner JI, Loffredo C, Kappetein AP, Wilson PD (1995) Transposition of the great arteries: etiologic distinctions of outflow tract defects in a case control study of risk factors. In: Clark EB, Markwald RR, Takao A (eds) Developmental mechanism of heart disease. Futura, Armonk, pp 639–653

Filippov AA, del Nido PJ, Vasilyev NV (2016) Management of systemic right ventricular failure in patients with congenitally corrected transposition of the great arteries. Circulation 134:1293–1302

Fricke TA, d'Udekem Y, Richardson M et al (2012) Outcomes of the arterial switch operation for transposition of the great arteries: 25 years of experience. Ann Thorac Surg 94(1):139–145

Goor DA, Edwards JP (1973) The spectrum of transposition of the great arteries: with special reference to developmental anatomy of the conus. Circulation 48:406–415

Greil G, Tandon A, Vieira MS et al (2017) 3D whole heart imaging for congenital heart disease. Front Pediatr 5:36

Grothoff M, Hoffmann J, Abdul-Khaliq H et al (2012) Right ventricular hypertrophy after atrial switch operation: normal adaptation process or risk factor? A cardiac magnetic resonance study. Clin Res Cardiol 101:963–971

Jatene A, Fontes V, Paulista P et al (1976) Anatomic correction of transposition of the great vessels. J Thorac Cardiovasc Surg 72:364–370

Jouannic JM, Gavard L, Fermont L et al (2004) Sensitivity and specificity of prenatal features of physiological shunts to predict neonatal clinical status in transposition of the great arteries. Circulation 110:1743–1746

Kilner PJ, Geva T, Kaemmerer H et al (2010) Recommendations for cardiovascular magnetic resonance in adults with congenital heart disease from the respective working groups of the European Society of Cardiology. Eur Heart J 31:794–805

de la Cruz MV, Arteaga M, Espino-Vela J, Quero-Jimenez M, Anderson RH, Diaz GF (1981) Complete transposition of the great arteries: types and morphogenesis of ventriculoarterial discordance. Am Heart J 102:271–281

Loffredo CA, Silbergeld EK, Ferrencz C, Zhang J (2001) Association of transposition of the great arteries in infants with maternal exposures to herbicides and rodenticides. Am J Epidemiol 153:529–536

Lu JC, Dorfman AL, Attili AK et al (2012) Evaluation with cardiovascular MR imaging of baffles and conduits used in palliation or repair of congenital heart disease. Radiographics 32:E107–E127

Meinel FG, Henzler T, Schoepf UJ, Park PW, Huda W, Spearman JV et al (2015) ECG-synchronized CT angiography in 324 consecutive pediatric patients: spectrum of indications and trends in radiation dose. Pediatr Cardiol 36:569–578

Murtuza B, Barron DJ, Stumper O, Stickley J, Eaton D, Jones TJ, Brawn WJ (2011) Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience. J Thorac Cardiovasc Surg 142:1348–1357

Pexieder T, Blanc O, Pelouch V, Ostadalova I, Milerova M, Ostadal B (1995) Late fetal development of retinoic acid-induced transposition of great arteries: morphology, physiology and biochemistry. In: Clark EB, Markwald RR, Takao A (eds) Developmental mechanism of heart disease. Futura, Armonk, pp 297–307

Plymen CM, Sado DM, Taylor AM et al (2013) Diffuse myocardial fibrosis in the systemic right ventricle of patients late after Mustard or Senning surgery: an equilibrium contrast cardiovascular magnetic resonance study. Eur Heart J Cardiovasc Imaging 14:963–968

Preim U, Hoffmann J, Lehmkuhl L et al (2013) Systemic right ventricles rarely show myocardial scars in cardiac magnetic resonance delayed-enhancement imaging. Clin Res Cardiol 102(5):337–344

Samanek M (2000) Congenital heart malformations: prevalence, severity, survival and quality of life. Cardiol Young 10:179–185

Szymczyk K, Moll M, Sobczak-Budlewska K et al (2018) Usefulness of routine coronary CT angiography in patients with transposition of the great arteries after an arterial switch operation. Pediatr Cardiol 39:335–346

Taylor AM, Dymarkowski S, Hamaekers P et al (2005) MR coronary angiography and late-enhancement myocardial MR in children who underwent arterial switch surgery for transposition of great arteries. Radiology 234:542–547

Unolt M, Putotto C, Silvestri LM et al (2013) Transposition of the great arteries: new insights into the pathogenesis. Front Paediatr 1:11

Warnes CA (2006) Transposition of the great arteries. Circulation 114:2699–2709

Williams WG, McCrindle BW, Ashburn DA, Jonas RA, Mavroudis C, Blackstone EH (2003) Outcomes of 829 neonates with complete transposition of the great arteries 12-17 years after repair. Eur J Cardiothorac Surg 24:1–9

Further Reading

Gutberlet M, Boeckel T, Hosten N, Vogel M, Kühne T, Oellinger H, Ehrenstein T, Venz S, Hetzer R, Bein G, Felix R (2000) Arterial switch procedure for D-transposition of the great arteries: quantitative mid-term evaluation of hemodynamic changes with cine MR imaging and phase-shift velocity mapping-initial experience. Radiology 214(2):467–475

Grothoff M, Fleischer A, Abdul-Khaliq H, Hoffmann J, Lehmkuhl L, Luecke C, Gutberlet M (2013) The systemic right ventricle in congenitally corrected transposition of the great arteries is different from the right ventricle in dextro-transposition after atrial switch: a cardiac magnetic resonance study. Cardiol Young 23(2):239–247. https://doi.org/10.1017/S1047951112000790 Epub 2012 Jun 14