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## Congenital heart diseases: post-operative appearance on multi-detector CT—a pictorial essay

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**Abstract** Echocardiography is considered as an initial imaging modality of choice in patients with congenital heart disease (CHD), and magnetic resonance (MR) imaging is preferred for detailed functional information. Multi-detector computed tomography (CT) plays an important role in clinical practice in assessing post-operative morphological and functional information of patients with complex CHD when echocardiography and MR imaging are not contributory. Radiologists should understand and become familiar with the complex

morphology and physiology of CHD, as well as with various palliative and corrective surgical procedures performed in these patients, to obtain CT angiograms with diagnostic quality and promptly recognise imaging features of normal post-operative anatomy and complications of these complex surgeries.

**Keywords** Congenital heart disease · Multidetector CT · Cardiac CT

### Introduction

Non-invasive imaging is pivotal in the follow-up of patients with congenital heart disease (CHD) who have undergone palliative or corrective surgical procedures. Trans-thoracic echocardiography is considered as an initial imaging modality of choice for assessment of CHD. However, it is limited in the reliable assessment of these procedures because of operator dependency, a small field of view and acoustic window, and poor assessment of the right heart, intra-cardiac baffle and extra-cardiac complex vascular anatomy [1]. Although magnetic resonance (MR) imaging is considered the imaging technique of choice in patients with CHD because of the high temporal and spatial resolution allowing excellent functional and anatomical information [2], it is time consuming, requires local expertise, repeated multiple breath-holding and prolonged sedation or general anaesthesia in children, and is usually more cumbersome to perform in acutely ill patients.

Because of the higher spatial and temporal resolutions and shorter imaging time of 64-slice multi-detector computed tomography (CT) compared with older scanner generations, combined with improved capabilities for

simultaneous assessment of intra-cardiac anatomy, coronary arteries, extra-cardiac vascular structures, cardiac function and lung parenchyma in a single data acquisition, this modality plays an important role in clinical practice in assessing post-operative morphological and functional information of patients with complex CHD, when echocardiography and MR imaging are inadequate, but at the cost of patients' exposure to ionising radiation [3].

The purpose of this article is to make a pictorial presentation of the typical appearances and potential complications of common surgical procedures performed for palliation or correction of a wide spectrum of CHD on 64-slice CT.

### Scanning technique

All cardiac studies were performed on a 64-slice CT system (Siemens Sensation 64, Erlangen, Germany) and protocols were optimised by the supervising cardiac radiologist on a patient-by-patient basis. Caution was taken in paediatric and juvenile patients to avoid excessive radiation exposure and electrocardiogram (ECG)-gated cardiac studies were

obtained only if clinically indicated. For paediatric patients with intra-cardiac abnormalities, supra-cardiac aortic and pulmonary artery diseases or surgeries, coronary artery disease (Kawasaki, anomalous coronaries) and central shunts, we prefer ECG-gating. Patients underwent ECG-gated helical volumetric data acquisition from above the level of the aortic arch to the diaphragm following intravenous injection of 1–1.5 ml/kg body weight of iodinated contrast medium (Omnipaque 350; Amersham, GE) at a rate of 3 ml/s [5 ml/s for coronary CT angiography (CTA)] with subsequent injection of 30 ml of saline at the same rate. Imaging parameters were 330-ms gantry rotation time, 100- to 120-kV tube voltage (100 kV in paediatric patients), 400 mA (250 mA for weight less than 50 pounds), 0.6-mm collimation and a pitch of 0.2. Although an initial fixed milliamperage was chosen, the CARE DOSE software (Siemens, Erlangen, Germany) allows variable milliamperage to be delivered during helical data acquisition, and therefore, it further reduces the total milliamperage and radiation exposure. In addition, dose modulation (ECG pulsing) was employed for all patients. After data acquisition, image post-processing (sagittal, coronal and coronal-oblique reformats) was performed on-line (CT console, Leonardo, Siemens, Erlangen, Germany) and the best diastolic and systolic phase axial and multiphase images were transferred to a three-dimensional (3D) workstation (Vitrea 3.6; Vital Images, Minneapolis, Minn.) for further post-processing and evaluation of coronary arteries.

Images were reconstructed at 0.75-mm slice thickness with a 0.4-mm overlap interval (at the best diastolic and systolic phase in ECG-gated studies) for the axial source and at 1.5-mm slice thickness with a 0.6-mm overlap interval for multi-planar reformats to assess the conduits and great vessels. To assess cardiac functional parameters, multiphase data reconstructed at 1.0-mm slice thickness

with 0.5-mm overlap and a 10% increment through the cardiac cycle were used.

## Palliative procedures

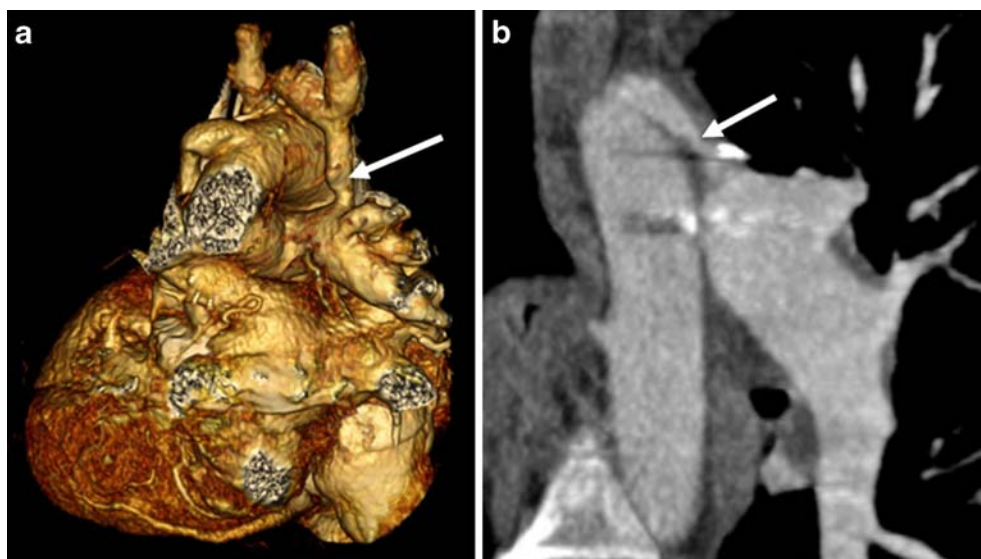
### Blalock–Taussig (BT) shunt

The classic BT shunt (Fig. 1) is a direct end-to-side anastomosis of a subclavian artery with the ipsilateral pulmonary artery. In a modified BT shunt, connection between the subclavian and pulmonary arteries is provided by a graft [4]. This procedure is performed in patients with right-sided cardiac obstruction or hypoplasia, such as right ventricular outflow tract obstruction (RVOTO), pulmonary stenosis or atresia as in Tetralogy of Fallot (ToF), to increase the pulmonary blood flow. Better growth of the pulmonary arterial tree and less distortion of the pulmonary arterial anatomy are advantages of the modified BT shunt [5]. Ipsilateral pulmonary artery enlargement, left ventricular hypertrophy, pseudo-aneurysm, infected shunt, shunt stenosis and seroma around the graft are potential complications of this procedure.

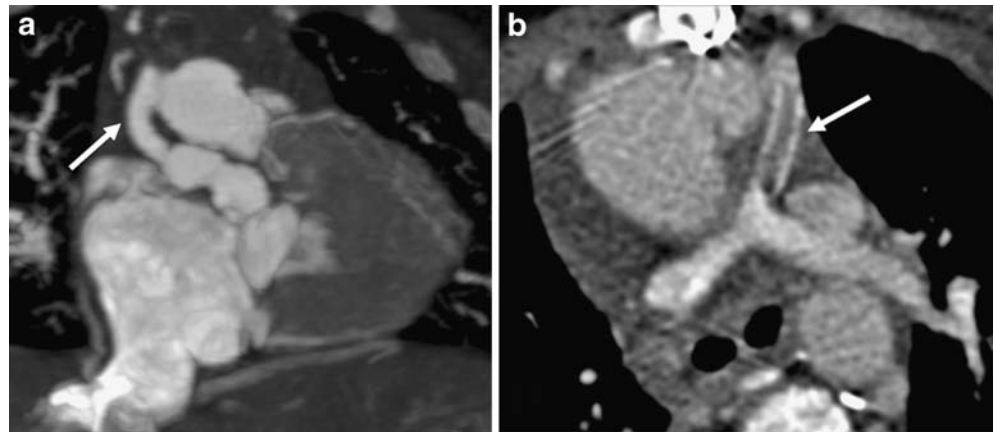
### Davidson or Central shunt

In this procedure a Gore-Tex interposition shunt graft is used to anastomose the ascending aorta to the main pulmonary artery for a more profound increase in the pulmonary circulation (Fig. 2) [5]. The procedure may be performed in patients with transposition of the great arteries (TGA), ventricular septal defect (VSD) with RVOTO, pulmonary atresia, and pulmonary artery stenosis/hypoplasia. Common complications of this procedure may include shunt thrombosis, pulmonary artery stenosis/

**Fig. 1** Blalock–Taussig shunt. **a** Volume-rendered image (posterior view) in a 43-year-old man with a history of ToF demonstrates a patent, classic, right BT shunt with mild distal narrowing (*arrow*). **b** Oblique coronal MIP image in a different patient with ToF shows stenosis of the left classic BT shunt (*arrow*). In a modified BT shunt where the connection between the subclavian and ipsilateral pulmonary artery is created by a graft, blood supply to the ipsilateral upper extremity is preserved via the subclavian artery



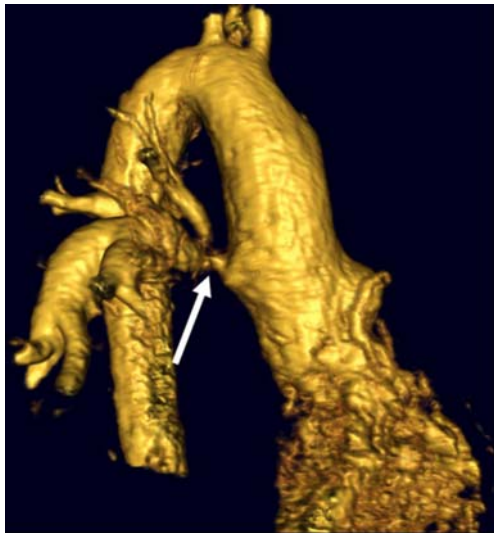
**Fig. 2** Davidson (Central) shunt. **a** Oblique coronal MIP image in a 16-year-old girl with hypoplastic left heart syndrome and status post-Davidson (Central) shunt demonstrates a patent Gore-Tex shunt graft connecting the ascending aorta to the main pulmonary artery (*arrow*). **b** Axial MIP image in a 14-year-old boy with a history of TGA, a single AV canal and subaortic stenosis shows a thrombosed Davidson (Central) shunt (*arrow*)



distortion at the anastomotic site, and congestive heart failure as a result of excessive pulmonary blood flow.

#### Waterston–Cooley shunt

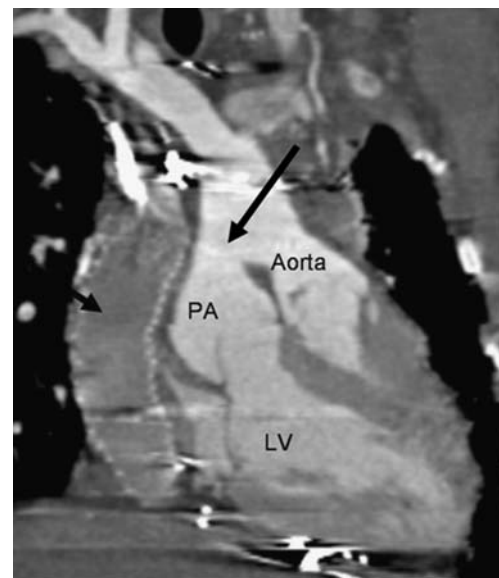
With the Waterston–Cooley shunt (Fig. 3), a communication is created between the posterior wall of the ascending aorta and the anterior wall of the right pulmonary artery to improve pulmonary perfusion in patients with diminished pulmonary flow, such as pulmonary atresia, ToF with RVOTO and tricuspid atresia [6]. Because of the high rate of complications, such as shunt obstruction, kinking, narrowing, thrombosis, preferential distribution of shunt flow to the right lung with pulmonary artery distortion and embolism, Waterston–Cooley shunts have been widely replaced by other procedures.



**Fig. 3** Waterston–Cooley shunt. Volume-rendered image in a 14-year-old boy with a history of ToF and pulmonary atresia demonstrates a Waterston–Cooley shunt extending from the posterior wall of the ascending aorta and the anterior wall of the right pulmonary artery. Note the mild narrowing in the mid portion of the shunt (*arrow*)

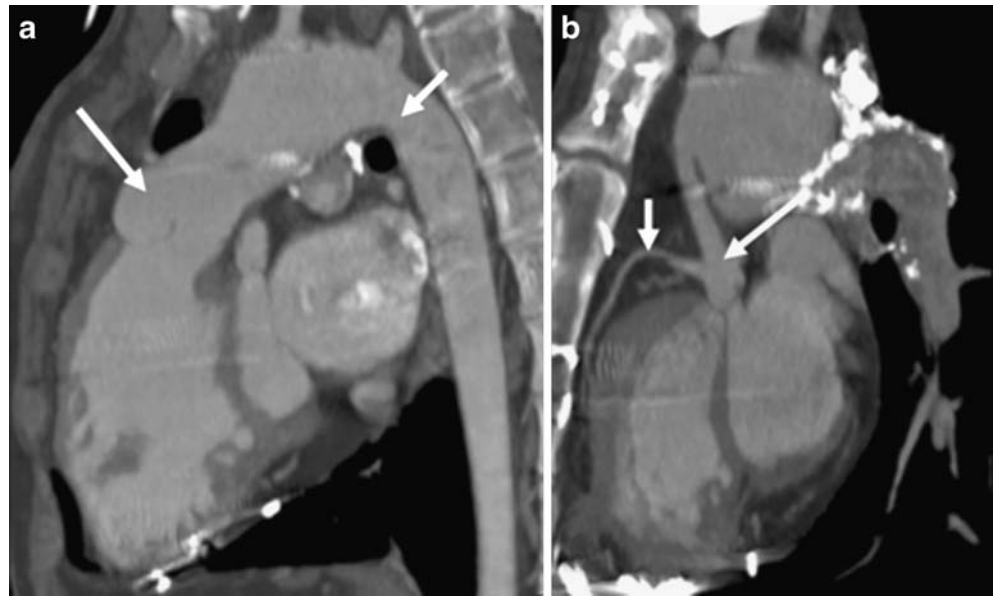
#### Damus–Kaye–Stansel shunt

Here, the main pulmonary artery is transected and connected to the ascending aorta by an end-to-side anastomosis (Fig. 4). This operation is performed to relieve the systemic ventricular outflow tract obstruction in patients with a double outlet right ventricle (RV), a univentricular heart with subaortic stenosis and TGA with VSD [7]. These patients usually have cavopulmonary shunts to return the systemic venous blood to the pulmonary circulation. Complications of the procedure include pulmonary valve regurgitation as a result of distortion of the pulmonary valve and recurrence of systemic ventricular outflow obstruction.



**Fig. 4** Damus–Kaye–Stansel shunt. Oblique coronal MIP image in a 14-year-old boy with a history of TGA shows the transected main pulmonary artery with connection to the ascending aorta by an end-to-side anastomosis (*large arrow*). Note the extra-cardiac Fontan shunt (*small arrow*; not opacified). A later phase image (not shown) demonstrated contrast opacification of the Fontan without thrombosis

**Fig. 5** Norwood–Sano procedure. Oblique sagittal MIP images in a 20-year-old woman with a history of hypoplastic left heart syndrome demonstrate ascending aortic reconstruction using the proximal main pulmonary artery (**a**, *large arrow*). The main pulmonary artery continues as the aortic arch and descending aorta (**a**, *small arrow*). Hypoplastic conventional ascending aorta arises from the left ventricle (**b**, *large arrow*). The conventional origin of the right coronary artery is also noted (**b**, *small arrow*)

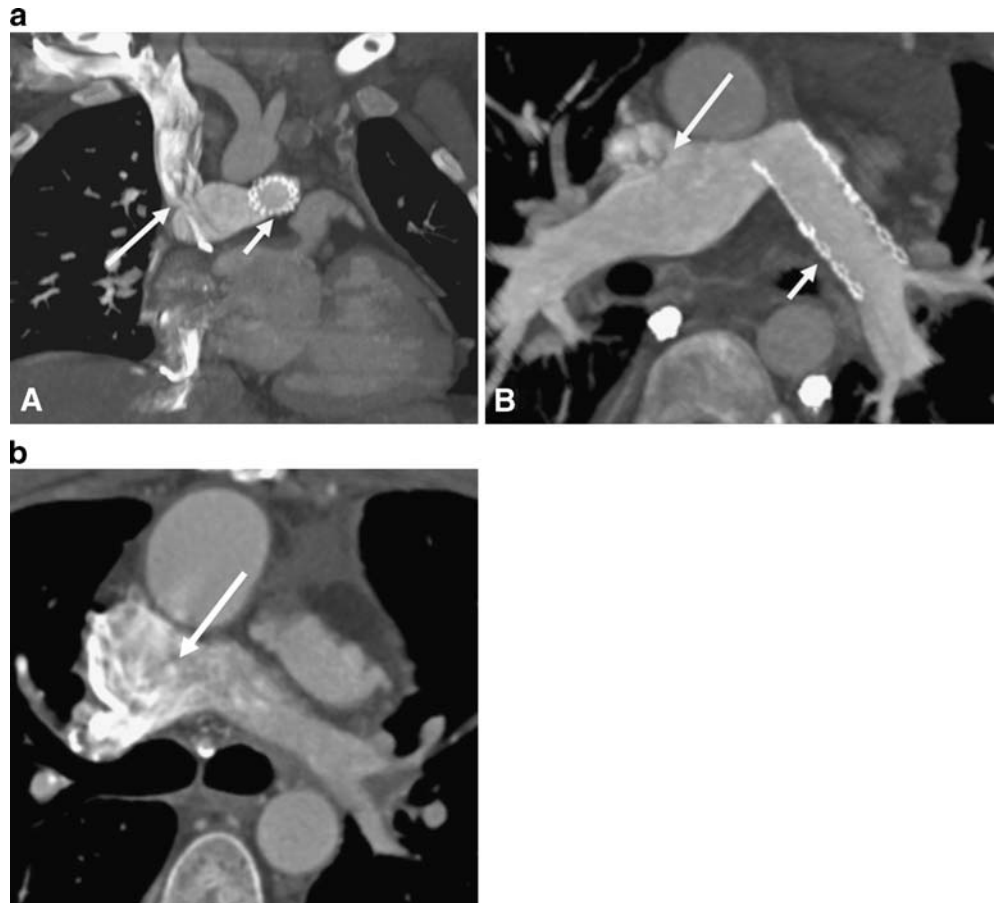


### Norwood–Sano procedure

A stage one Norwood procedure includes aortic reconstruction using the proximal main pulmonary artery and

creation of a systemic–pulmonary artery shunt (i.e. BT shunt) (Fig. 5). In the modified Norwood–Sano procedure, an extra-cardiac allograft valved or non-valved conduit is placed between the right ventricle and distal stump of the

**Fig. 6** **a** Bidirectional Glenn shunt with a left pulmonary artery stent. Oblique coronal and axial MIP images in a 19-year-old man with tricuspid atresia and an atrioventricular canal defect demonstrate anastomosis of the SVC to the right main pulmonary artery (**A** and **B**, *large arrow*). Noted is a left main pulmonary artery stent without in-stent re-stenosis (**A** and **B**, *small arrow*). **b** Bidirectional Glenn shunt. Axial MIP image in a 24-year-old man with a history of TGA demonstrates anastomosis of the SVC to the main pulmonary artery confluence (*arrow*)



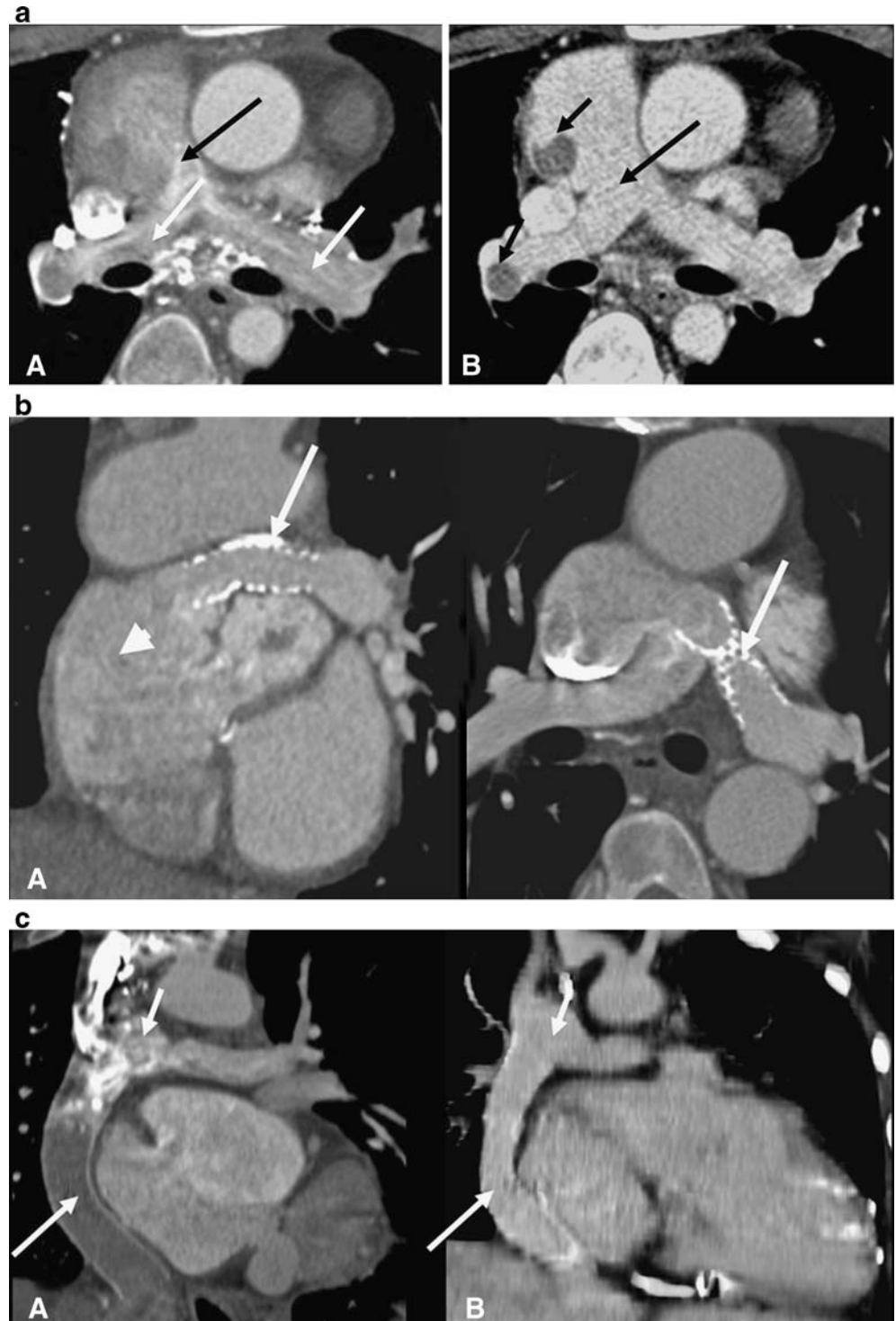
pulmonary artery to secure balanced and stable systemic and pulmonary circulation, and to prevent the reduced diastolic blood flow in the coronaries associated with the BT shunt [8]. Indications include hypoplastic left heart

syndrome (HLHS), double-outlet right ventricle with aortic or subaortic stenosis and aortic atresia. A stage-one Norwood procedure is associated with high mortality as a result of pulmonary over-circulation and haemodynamic instability.

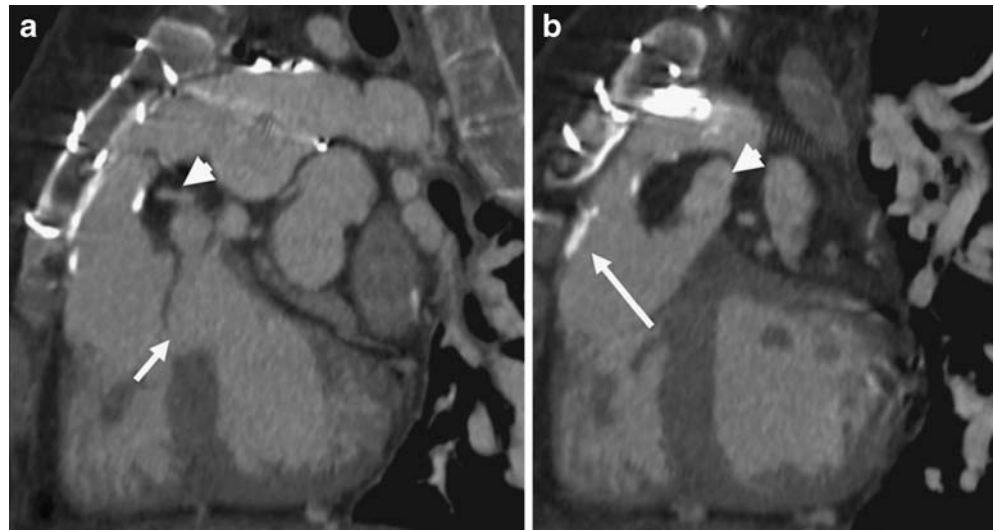
**Fig. 7 a** Direct classic Fontan shunt. Axial MIP images in a 26-year-old man with a history of ToF demonstrate direct anastomosis of the RA to the main PA confluence (*A* and *B*, *black arrow*). In the early phase (*A*), there is a thrombosis-like appearance of bilateral PAs because of mixing artefact (*A*, *white arrows*). In the later phase (*B*), thromboemboli are clearly seen in the RA and the right PA (*B*, *small black arrows*) with good opacification and absent mixing artefact of the main PAs.

**b** Modified Fontan shunt. Oblique coronal MIP image in a 35-year-old man with a history of TGA and tricuspid atresia demonstrates the connection between the right atrium (*A*, *arrowhead*) and the main PA confluence through a Gore Tex graft (*A*, *arrow*) that is peripherally calcified. Noted is a left main PA stent (*B*, *arrow*).

**c** Total cavopulmonary connection with extra-cardiac Fontan shunt. Oblique coronal MIP images in a 25-year-old man with a history of TGA demonstrate the anastomosis of the IVC (*A* and *B*, *large arrow*) to the main PA confluence (*A* and *B*, *small arrow*). In the arterial phase (*A*) the Fontan shunt is not opacified, resembling a thrombosis. In the venous phase (*B*), normal opacification of the shunt is noted, indicating patency. The patient also has a bidirectional superior vena cava (SVC) Glenn shunt. In this procedure, the SVC flow is directed mostly to the right pulmonary artery, and the IVC flow to the left PA



**Fig. 8** Rastelli procedure. Oblique sagittal MIP images in a 30-year-old woman with a history of complex CHD demonstrate the connection of the pulmonary artery confluence to the right ventricle with an extra-cardiac, peripherally calcified conduit without significant stenosis (**a** and **b**, *large arrow*). Note the atretic main pulmonary artery (**b**, *arrowhead*). The VSD is closed by a Gore Tex graft (**a**, *small arrow*) and the LV is connected to the aorta. The right coronary artery is also noted to originate from the aorta (**a**, *arrowhead*)



### Glenn shunt

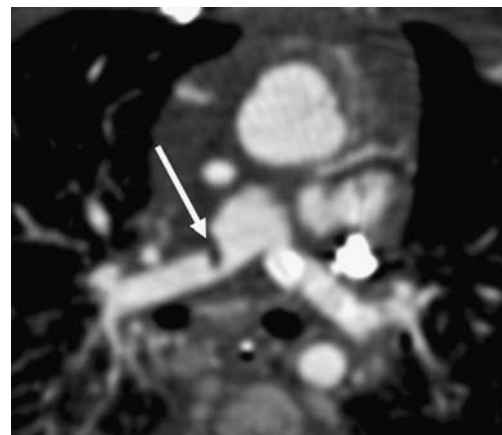
In this procedure, the systemic venous return is re-directed to the pulmonary circulation, bypassing the right heart (Fig. 6). With the classic (unidirectional) Glenn shunt, the distal end of the superior vena cava (SVC) is ligated, and the side of the SVC above the ligation is anastomosed with the distal end of the divided right pulmonary artery, providing unilateral (right) lung perfusion [9]. In a modified (bidirectional) Glenn or hemi-Fontan shunt, anastomosis is created between the transected end of the SVC and the side of the undivided right pulmonary artery, providing balanced bilateral pulmonary circulation [9].

Indications include palliation of a variety of cyanotic CHD that eventually lead to a single anatomical or functional ventricle (i.e. tricuspid atresia), right or left ventricular hypoplasia (i.e. HLHS), and as an initial step before the Fontan form of a total right heart bypass. The Glenn shunt is usually the first stage in early childhood before the Fontan procedure, given the disproportionate blood flow of the SVC in the early part of life. As a child grows, the blood flow from the lower body increases disproportionately to the SVC flow, mandating a follow-up Fontan procedure. Complications of the Glenn shunt include SVC syndrome, shunt and pulmonary artery thrombosis, stenosis of the cavopulmonary anastomosis, sinus node injury with subsequent rhythm disturbance, pulmonary arteriovenous malformation (AVM) and aorto-pulmonary collaterals.

### Fontan shunt

In the Fontan procedure, the systemic venous blood is directed to the pulmonary arteries bypassing the RV (Fig. 7) [9]. With the classic Fontan, the anastomosis is created between the right atrium (RA) or atrial appendage,

and the pulmonary artery (PA) (atriopulmonary connection) using a homograft, patch, or valved conduit (indirect Fontan), or direct anastomosis (direct Fontan). In lateral tunnel (intra-atrial tunnel) Fontan, the right atrial wall is used to create a baffle to direct the blood flow from the inferior vena cava (IVC) to the lower portion of the SVC, which is further drained to the PA. In extra-cardiac Fontan, the systemic venous blood is directed to the main PA confluence through an extra-cardiac conduit. Total cavopulmonary connection includes either the lateral tunnel or extra-cardiac Fontan procedure combined with the modified Glenn shunt, while the pulmonary trunk is disconnected from the heart, and right and left PAs are interconnected. In this procedure, SVC flow is directed mostly to the right pulmonary artery, and IVC flow to the left PA. In these patients, CT pulmonary angiography should be performed with simultaneous foot and arm injections, or two-phase (arterial and delayed venous) or delayed phase



**Fig. 9** Pulmonary artery banding. Axial MIP image in a 15-year-old girl with a history of hypoplastic left heart syndrome shows proximal right pulmonary artery banding (*arrow*)

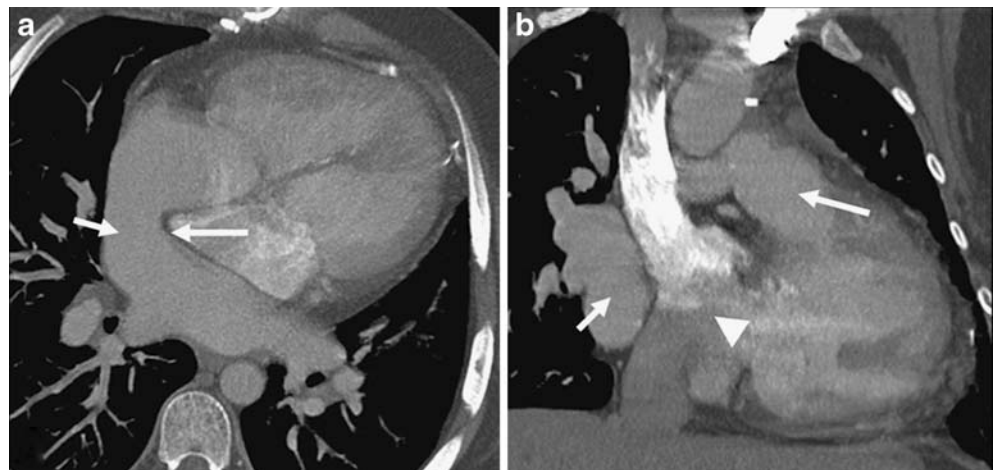


**Fig. 10** Unifocalisation. Oblique coronal MIP image in a 14-year-old girl with a history of ToF with pulmonary atresia and major aortopulmonary collaterals shows right unifocalisation (*large arrow*) with Gore Tex graft extension (*small arrow*) into left unifocalisation (*arrowhead*) and the right ventricular outflow tract (*black arrow*)

only CT acquisitions to assess for conduit patency and pulmonary embolism without mixing artefacts.

In fenestrated Fontan, there is communication between the Fontan conduit and the right atrium to reduce systemic venous pressure in the post-operative period, serving as a “pop-off” valve to increased pulmonary pressure. Indications include cyanotic CHD with an anatomical or functional single ventricle such as tricuspid atresia, pulmonary atresia with intact ventricular septum, double inlet left ventricle, HLHS and an unbalanced atrioventricular septal defect. Radiological complications of the Fontan shunt include shunt and pulmonary artery thrombosis, pulmonary thromboembolism, right atrial enlargement, atrioventricular valve regurgitation, pulmonary artery stenosis, cavopulmonary anastomotic narrowing, pulmonary arteriovenous malformation (AVM), and major aortopulmonary collaterals.

**Fig. 11** Atrial switch repair. A 19-year-old man’s status post-Mustard procedure for double outlet right ventricle and TGA. Oblique axial MIP image demonstrates an inter-atrial baffle (*a, large arrow*) separating the pulmonary venous chamber and the systemic venous chamber. The main pulmonary artery arises from the double-outlet right ventricle (*a, small arrow*). Oblique coronal MIP image demonstrates the pulmonary venous chamber (*b, arrow*) and drainage of the IVC and SVC to the systemic venous chamber (*b, arrowhead*)



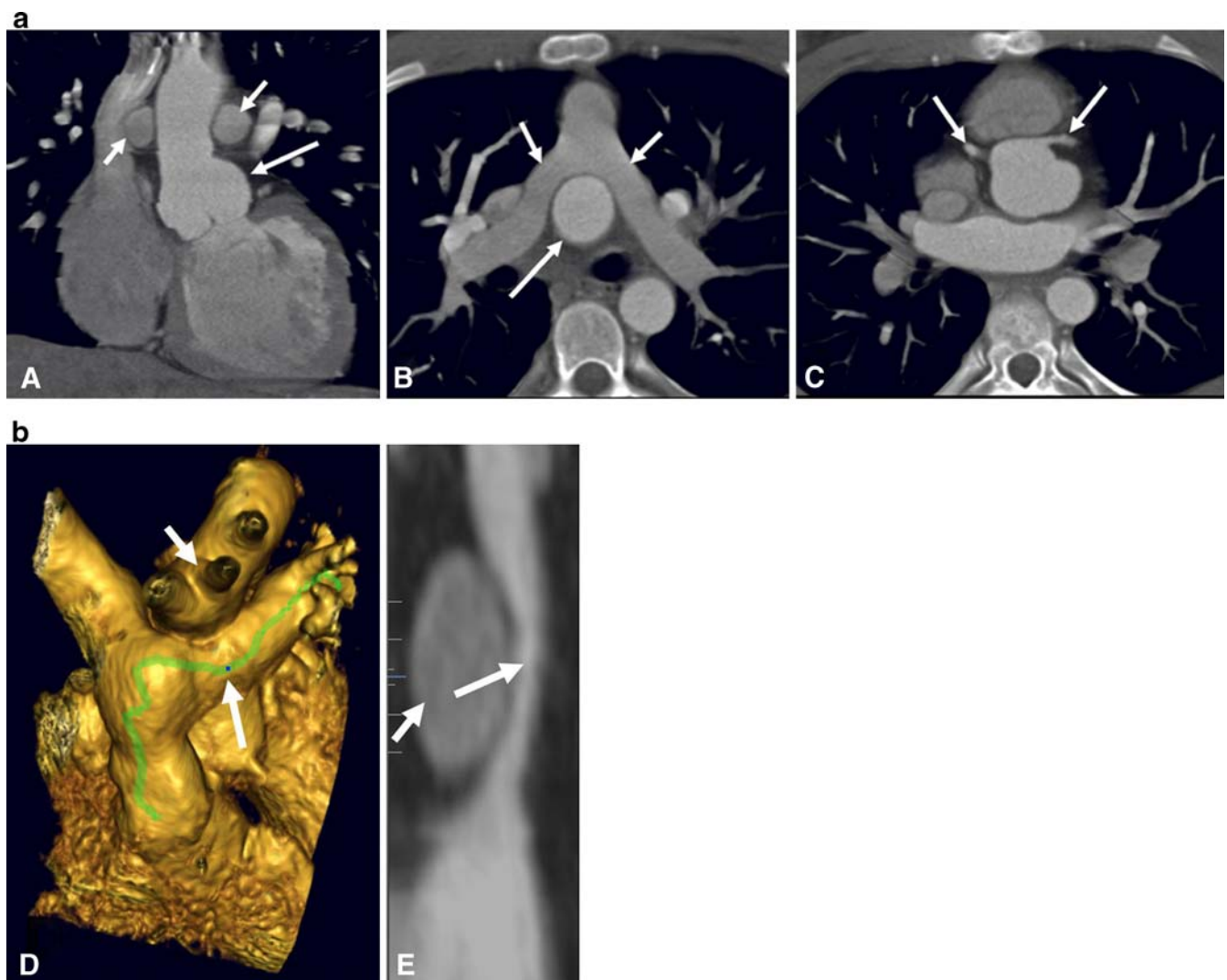
## Rastelli procedure

In this procedure, an external right ventricle to the PA conduit and an intraventricular left ventricle to the aorta via the VSD tunnel are created (Fig. 8). The pulmonary valve may be atretic and is surgically closed at the time of the procedure [10]. Most frequent indications include double outlet right ventricle, TGA associated with VSD and left ventricular outflow tract (LVOT) obstruction, and rarely ToF with pulmonary stenosis or atresia. Complications of the Rastelli procedure include extra-cardiac conduit stenosis (with or without regurgitation), calcification, kinking and aneurysm. The tunnel patch from the left ventricle to the aortic valve may also be complicated by leakage, obstruction, stenosis, or aneurysm. Other complications are branch pulmonary artery stenosis and biventricular dysfunction. In congenitally corrected TGA (atrioventricular discordance and ventriculo-arterial discordance—the aorta arises from the malpositioned RV and the main pulmonary artery from the malpositioned LV) with associated severe pulmonary valve stenosis, a valved or non-valved pulmonary conduit from the LV to the main pulmonary artery is usually created to bypass the blood from the right ventricular outflow obstruction. The valved conduit may undergo severe regurgitation over time, which can be better demonstrated and quantified on MR imaging.

However, CTA can be used to show conduit patency, stenosis, calcification and the distance between the back of the sternum and the mediastinal vessels/heart chambers and the conduit for the pre-surgical planning of a possible re-operation.

## Pulmonary artery banding

This procedure involves palliative restriction of increased pulmonary perfusion in patients with excessive left-to-right shunting who cannot undergo corrective surgery, cyanotic CHD such as complicated VSD, multiple VSDs, VSD with coarctation of aorta, HLHS and D-TGA to prevent pulmonary overcirculation leading to eventual pulmonary arterial



**Fig. 12 a** Jatene procedure (arterial switch repair). An 18-year-old man with a history of D-TGA and status post-Jatene procedure. Oblique coronal and axial MIP images demonstrate switching of the aorta (*A* and *B*, *large arrow*) and pulmonary artery (*A* and *B*, *small arrows*) along with the transposition of the coronary arteries (*C*, *arrows*) from the aorta (or neo-pulmonary artery) to the pulmonary

artery (or neo-aorta). Note that the neopulmonary artery is located anterior to the neo-aorta (Lecompte manoeuvre). **b** Jatene procedure (arterial switch repair). Volume-rendered (*A*) and oblique axial MIP image (*B*) in a 22-year-old man with a history of D-TGA and status post-Jatene procedure. Note the left pulmonary artery stenosis (*A* and *B*, *large arrow*) by the neo-aorta (*A* and *B*, *small arrow*)

hypertension, and subsequent right ventricular failure (Fig. 9) [11]. The band is ideally placed in the mid portion of the pulmonary trunk to avoid injury to the pulmonary valve. Associated complications include branch pulmonary artery stenosis as a result of impingement, migration, pulmonary artery pseudoaneurysm, fibrosis and scarring of the band site, and subaortic narrowing.

#### Unifocalisation

Unifocalisation of major aortopulmonary collateral arteries (MAPCAs) is performed in patients with VSD and pulmonary atresia, ToF with pulmonary atresia, pulmonary atresia with intact inter-ventricular septum, single ventricle, right

isomerism and severely unbalanced common atrioventricular canal (CAVC), to reroute the misdirected blood vessels into a single vessel (or into the pulmonary artery if it is present), which is then attached to the right ventricle through a conduit (Fig. 10) [12]. Complications of unifocalisation include thrombosis, calcification and stenosis of the conduit.

#### Corrective procedures

##### Atrial switch repair

This procedure is performed for physiological correction of TGA in which an atrial baffle (the Mustard procedure uses



pericardial tissue or a Gore-Tex graft, and in the Senning procedure, the atrial septum is reconstructed to form the baffle) redirects systemic venous blood through the mitral valve to the anatomical left ventricle (sub-pulmonary), and oxygenated pulmonary venous return through the tricuspid valve to the anatomical right ventricle (sub-aortic) (Fig. 11) [13]. Complications include baffle leakage and obstruction, right ventricular hypertrophy and failure because of systemic volume and pressure overload, caval and pulmonary venous obstruction, and dilatation of the great arteries [14].

### Jatene arterial switch procedure

The procedure of choice for anatomical correction of D-TGA in neonates is the Jatene operation, where the aorta and main pulmonary artery are transected above their corresponding valves and switched so that the distal aortic segment is anastomosed with the proximal pulmonary root to form a neo-aorta (Fig. 12) [14]. The distal pulmonary artery is brought anterior to the neo-aorta (the Lecompte manoeuvre) to be anastomosed with the proximal aortic root to form the neopulmonary artery. Coronary arteries are re-implanted to the neo-aorta. The most common long-term complication is pulmonary artery stenosis because of a neo-aortic aneurysm

or inadequate growth of the pulmonary arteries. Other complications include aortic root dilation, right ventricular outflow obstruction, supravalvular aortic stenosis, and ischaemic complications because of coronary artery translocation.

### Conclusion

Radiologists and cardiac imagers including cardiologists should understand and become familiar with the complex morphology and physiology of congenital heart diseases, as well as with various palliative and corrective surgeries performed for these patients to obtain diagnostic quality cardiac CTA and to promptly recognise the imaging features of normal and abnormal post-operative appearance seen in this patient population. Failure to understand the principles of post-surgical anatomy and complications of CHD might subject these patients to unnecessary radiation exposure and suboptimal CTA studies, thereby resulting in the imaging physicians not being able to recognise post-operative anatomy and its potential complications.

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