



## Guidelines

## Computed Tomography Imaging in Patients with Congenital Heart Disease Part I: Rationale and Utility. An Expert Consensus Document of the Society of Cardiovascular Computed Tomography (SCCT) **Endorsed by the Society of Pediatric Radiology (SPR) and the North American Society of Cardiac Imaging (NASCI)**



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## ABSTRACT

This is an expert consensus document created to provide information about the current use of cardiovascular computed tomography (CT) in patients of all ages with proven or suspected congenital heart disease (CHD). The discussion and recommendations are based on available literature and the judgment of a diverse group of subspecialists with extensive experience in the use of CT imaging in CHD. The field of CHD CT imaging is evolving rapidly with the availability of new scanner technology. In addition, the prevalence of palliated CHD has increased with marked improvements in patient survival. We believe it is important to review the clinical indications, strengths, limitations, and risks of cardiovascular CT in this patient population. This is the first of two complementary documents. It will concentrate on the disease entities and circumstances in which CT may be used. The second document will focus on recommendations for the technical performance of cardiovascular CT in patients with CHD.

Successful cardiovascular CT imaging of CHD requires an in depth understanding of the core teaching elements of both cardiology and radiology. The ability to perform and interpret high quality congenital cardiovascular CT in a clinical context requires focused time and effort regardless of the previous background of the cardiac imager. This is reflected by a writing committee that consists of pediatric and adult radiologists and cardiologists, all whom have extensive experience in performing CT in this patient population. Cardiovascular CT is complementary to other imaging modalities and its optimal use will be in centers where all diagnostic modalities are available. The choice of modality for an individual patient should be determined by age, diagnosis, clinical condition, clinical question and patient preference.<sup>1–4</sup> Use of CT in CHD should be reserved for situations in which it is expected to provide unique diagnostic information for the individual patient or clinical indication, and/or less risk than other modalities. This multi-disciplinary document is intended to guide the optimal selection of CHD patients for cardiovascular CT.

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The goals of this document apply to both pediatric and adult CHD patients and are to:

- 1) Review the current use of cardiovascular CT.
- 2) Assess the most up to date information on risks, benefits, as well as limitations of cardiovascular CT.
- 3) Provide disease-specific indications for cardiovascular CT imaging.
- 4) Outline a consensus opinion on the essential skills and knowledge needed to optimally perform and interpret cardiovascular CT.

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## 1. Introduction: why CT is increasingly used in CHD

### 1.1. Changing CT technology

The first clinical CT scanners had limited use for cardiac applications due to poor spatial and temporal resolution, and long scan times. There has been a marked improvement in CT scanner technology in the past decade. Current generation multidetector CT (MDCT) scanners allow rapid coverage of large anatomic volumes, submillimeter isotropic spatial resolution and temporal resolution as low as 66 ms. These advances provide diagnostic images of small cardiovascular structures, even at the high heart rates encountered in a pediatric population.<sup>1–13</sup> Data acquisition now requires only a portion of the cardiac cycle or at most several cardiac cycles. The highest pitch scan modes and volumetric scanners provide full anatomic coverage of a pediatric thorax in less than a second or a single heartbeat, freezing respiratory motion. This rapid image acquisition eliminates or reduces the need for sedation and anesthesia in those unable to cooperate with a short breath hold.<sup>10,14</sup> The prospective delivery of radiation to a limited portion of the cardiac cycle and post processing approaches such as iterative reconstruction allow for significantly reduced radiation doses while maintaining or improving image quality.<sup>15–17</sup>

The use of cardiovascular CT has been described in patients of all ages with cardiac malformations of all levels of complexity. Detailed coronary artery imaging is possible in nearly every patient using a current generation CT scanner. Retrospectively ECG gated scans may be performed for quantification of ventricular function with an accuracy that is equal to cardiovascular magnetic resonance (CMR), the modality most often used as the reference standard.<sup>18</sup> Non-cardiovascular anatomy, including airway, lung parenchyma, and skeletal anatomy is clearly seen. Cardiovascular CT also provides excellent visualization of stents, conduits, and metallic objects and can be performed in patients with implanted pacemakers and defibrillators.<sup>19</sup>

Limitations of cardiovascular CT include poor myocardial tissue characterization, inability to quantify valve regurgitation in patients with more than one regurgitant lesion or shunt, and exposure to ionizing radiation. Additionally, intravenous administration of iodinated contrast with its attendant risks is required in almost all patients for vascular opacification. Although breath holding is no longer required for many indications on high pitch and volumetric scanners, it is still needed for images acquired over several heart beats, including functional imaging and detailed coronary artery imaging at high heart rates. For this reason anesthesia is still required for specific indications in young or critically ill patients who are unable to cooperate with breath holding.

Advanced diagnostics in the current era are primarily non-invasive, and cardiovascular CT is increasingly used as an adjunct

to echocardiography when CMR is considered high risk, contraindicated, or unlikely to provide images suitable-quality to answer the clinical question.<sup>20,21</sup>

### 1.2. Changing patient population and diagnostic paradigm

Congenital heart disease is the most common congenital anomaly. Survival after surgical intervention for all forms of CHD is now expected for most patients.<sup>22–25</sup> The Society of Thoracic Surgeons database reports a national overall mortality for all CHD operations of 3.5% for 2010–2013.<sup>26</sup> The average age of patients with CHD is continually increasing and has reached adulthood. In fact, 2/3 of patients with CHD are now adults, and the number of those reaching  $\geq 60$  years of age is increasing rapidly.<sup>24,27–29</sup> Much of the cardiac morbidity occurs in older patients; and mortality in CHD has shifted from infants towards adults.<sup>22,29–31</sup> These complex patients often have residual hemodynamic lesions requiring repeat intervention throughout life, and therefore, require close surveillance and have high utilization of health care resources.<sup>32,33</sup>

It is widely recognized that catheterization is no longer required for the diagnosis and management of most forms of CHD. It is now reserved for patients needing invasive hemodynamic evaluation or catheter based intervention.<sup>34</sup> Non-invasive imaging can establish the details of anatomy with the degree of certainty required for surgical intervention for most CHD indications.<sup>35–38</sup> The availability of cardiovascular CT and CMR that is supervised, performed, and interpreted by physicians with expertise and training in congenital heart disease has become an essential component of regional pediatric and adult CHD centers.<sup>23</sup>

Echocardiography remains the standard initial imaging modality in CHD, and has excellent diagnostic accuracy when performed by skilled practitioners.<sup>20,35</sup> Echocardiography routinely visualizes intracardiac anatomy, and is both portable and widely available. A significant limitation of echocardiography is its poor reproducibility in quantifying single or right ventricular size and systolic function and valve regurgitation.<sup>39,40</sup> Echocardiography is often unable to adequately assess distal pulmonary arteries, complex systemic or pulmonary venous anatomy, and cavo-pulmonary anastomoses in patients with single ventricle physiology.<sup>39,40</sup> Given the reliance on transmission and receipt of ultrasound energy through the chest, echocardiographic images may be inadequate due to limitations of the “acoustic window” in patients with large and small body habitus, or those with scoliosis, metallic implants, or other alterations in the chest wall.

CMR is the modality most often used as an adjunct to echocardiography for CHD when further information is needed.<sup>20,21,41</sup> It is excellent for 3D visualization of complex anatomy, reproducible quantification of single and right ventricular function, quantification of valve regurgitation, myocardial tissue characterization and stress imaging.<sup>42–46</sup> CMR is considered the standard for

quantification of ventricular size, systolic function and valve regurgitation to which other modalities are compared. CMR uses no ionizing radiation, and gadolinium-based contrast is needed for angiography only where the non-contrast 3D sequences are inadequate. In institutions with access to both cardiovascular CT and CMR, CMR is used more commonly due to the favorable risk profile and excellent diagnostic quality for most indications and patients.<sup>20,21</sup>

Although advances such as real-time cine imaging,<sup>47</sup> single heartbeat delayed enhancement imaging<sup>48</sup> and highly accelerated parallel imaging<sup>49</sup> allow completion of even the most complex CHD patients within 1 h, CMR still usually requires relatively long imaging times.<sup>50,51</sup> Children younger than eight years of age, and developmentally delayed patients of all ages require sedation or anesthesia for the MR study. Intravenous (IV) gadolinium contrast is needed only for certain indications. Due to the risk of nephrogenic systemic fibrosis, its use is contraindicated in patients with acute or chronic severe renal (kidney) disease defined as glomerular filtration rate <30 mL/min/m<sup>2</sup>; or renal dysfunction due to the hepato-renal syndrome.<sup>52–54</sup>

Many patients with CHD require placement of metallic devices such as coils and stents that may degrade CMR image quality due to susceptibility artifact. The combination of an endovascular coil and a stent has been shown to decrease the diagnostic utility of CMR in Fontan patients to <10%.<sup>55</sup> Pacemaker and defibrillator use is common in patients with CHD.<sup>19,56–58</sup> While MRI safe pacemakers are now available and some non-pacemaker dependent patients with older devices may undergo MRI,<sup>59,60</sup> imaging artifact from the device may continue to obscure anatomy, such as epicardial devices with leads directly adjacent to the heart.<sup>61</sup>

## 2. Systematic review of the use of CT in CHD

CT has been used to assess complex CHD for over two decades. There are many excellent comprehensive review articles on the use of cardiovascular CT for the pre- and post-operative evaluation of patients of all ages with congenital cardiovascular disease.<sup>1,3,5,7,62–73</sup> Cardiovascular CT has been used for detection and follow-up of extracardiac vascular lesions, intracardiac lesions, and pericardial diseases.<sup>2–4,7,16,69,74–89</sup> The next section provides a lesion-specific review of the use of CT for the most commonly referred CHD diagnoses.

### 2.1. Coronary imaging

Coronary CTA is well established in adult patients for coronary artery imaging.<sup>90–93</sup> The Society of Cardiovascular Computed Tomography has established guidelines for both the acquisition and reporting in coronary artery CT for atherosclerotic heart disease in adults.<sup>94,95</sup> Increasing evidence supports coronary CT for evaluation of the coronary arteries in patients with pediatric and congenital cardiovascular disease, including congenital coronary anomalies, coronary fistula, Kawasaki disease, and after CHD surgical repair requiring coronary artery manipulation.<sup>96–109</sup>

### 2.2. Use of CT for congenital coronary artery anomalies

Congenital coronary anomalies are the second most common cause of sudden death in young athletes<sup>110,111</sup> and are present in 0.2–2% of the population.<sup>112,113</sup> Echocardiography can accurately diagnose coronary anomalies in children when performed by a skilled echocardiographer, but it is limited in its ability to fully characterize coronary anatomy in many adult patients and is prone to false negatives.<sup>114–117</sup> Coronary artery dominance, angulation from the aortic root, ostial narrowing and presence and length of

intramural course cannot be reliably determined by echocardiography. These anatomic features are considered by some to be linked to prognosis. Precise anatomic definition with advanced imaging aids in surgical planning when indicated.<sup>118–120</sup> Coronary CTA in patients of all ages with coronary anomalies is well described and has superior accuracy for this indication because of the ability to simultaneously visualize the coronary arteries as well as the great vessels.<sup>117,119,121–127</sup> CMR has been shown to be useful for congenital coronary anomalies in older children and adolescents, but is less useful in the youngest patients because image quality is inversely related to both patient age and heart rate.<sup>128,129</sup> Also, CMR is often unreliable in the evaluation of the distal coronary artery anatomy to determine coronary dominance in young patients.<sup>128,129</sup> Coronary anomalies are common in patients with CHD, and precise anatomic definition prior to surgical intervention is often indicated since it may alter the surgical course.<sup>130</sup> Current guidelines recommend that all patients that have previously undergone surgical coronary artery manipulation have complete angiographic assessment at least once in adulthood, and that intervention on the RVOT should be preceded by unambiguous definition of coronary artery anatomy.<sup>23</sup> The modality chosen for coronary assessment will depend on the age of the patient and institutional capabilities. See lesion-specific sections below for additional detail (TOF, transposition complexes).

### 2.3. Use of coronary CTA for acquired coronary artery disease in pediatric and CHD patients

As long term survival in CHD continues to improve, patients may acquire coronary artery disease that can affect outcome for congenital surgeries performed in adulthood, and concomitant CABG may be required.<sup>27,131</sup> Cardiac CTA is useful for simultaneous evaluation of congenital anatomy and of coronary arterial pathology in adult patients with symptoms that may be attributable to coronary artery disease. High risk CHD patients undergoing cardiac intervention should have preoperative evaluation of the coronary arteries.<sup>23</sup>

Kawasaki disease (KD) is the most common acquired cardiac disease in children in the United States. Despite adequate treatment, 3–5% percent of patients develop coronary artery aneurysms or ectasia.<sup>132</sup> While aneurysms in these patients are frequently identified in the proximal coronary arteries on transthoracic echocardiography by a skilled pediatric echocardiographer, commonly-occurring mid and distal coronary artery aneurysms and coronary stenoses are poorly visualized.<sup>98–104</sup> In long-term follow-up, patients with a history of KD and aneurysms may develop coronary stenoses, occlusions, calcifications, thrombi and embolic infarction.<sup>132</sup> Coronary CTA offers comparable accuracy to conventional coronary angiography<sup>105,106</sup> which has historically been considered the reference standard for evaluation of coronary aneurysms and stenoses in patients with KD. Recent reports of both calcification and plaque identification by coronary CTA in high risk patients with a history of KD may identify a subset of patients at higher risk of future adverse event.<sup>107</sup> CMR is described for definition of coronary aneurysm, but is less sensitive than coronary CTA for identification of stenoses, calcifications, ectasia, and distal coronary artery disease.<sup>105,108,109</sup>

### 2.4. Thoracic vasculature abnormalities (pulmonary/systemic venous abnormalities, aortic/pulmonary arterial anomalies and vascular rings/slings)

Cardiovascular CT evaluation of pulmonary venous anomalies is well described and has been shown to be highly specific when compared to operative findings.<sup>133–136</sup> Cardiovascular CT is

accurate for determining the site of anatomic obstruction in anomalous pulmonary venous return and for the diagnosis of pulmonary vein stenosis.<sup>137–142</sup> Systemic venous anomalies are also well visualized by cardiovascular CT.<sup>143–145</sup>

Cardiovascular CT has also been shown to accurately visualize congenital aortic anomalies including interrupted aortic arch and aortic coarctation in both pediatric and adult patients.<sup>146–153</sup> It is particularly useful for evaluation of the aortic arch after endovascular intervention (stent or stent graft) where aneurysm, aortic wall injury or recurrent arch obstruction are relatively common.<sup>154–159</sup> It is recommended that every patient with repaired or unrepaired aortic coarctation have comprehensive evaluation of the thoracic aorta, and that those who have undergone intervention should undergo serial evaluation by cardiovascular CT or CMR throughout adulthood.<sup>23</sup>

Cardiovascular CT may be considered the optimal diagnostic modality for evaluation of suspected vascular rings and slings, and for assessment of vascular anatomy and associated tracheobronchial narrowing. The ability of cardiovascular CT to simultaneously image vascular structures and airway structures makes it an ideal imaging modality prior to surgical intervention. In addition to detailing the precise anatomy and measurements, cardiovascular CT allows the imager to describe and quantify involvement of the trachea and bronchi including assessment for complete cartilaginous rings. Multiple groups have shown that cardiovascular CT accurately characterizes tracheomalacia and vascular anatomy resulting in airway compression pre- and postoperatively in patients with symptomatic thoracic vascular anomalies.<sup>147,148,160–168</sup> In symptomatic patients seeking surgical relief, CT facilitates planning of the surgical approach and helps determine whether tracheal reconstruction or aortopexy will be necessary as well.<sup>169</sup>

Cardiovascular CT is well established in the evaluation of pulmonary artery anomalies. Ductal continuation of the pulmonary artery with subsequent ductal closure and pulmonary artery isolation is optimally imaged with CT since the lung parenchyma and associated anomalies can be evaluated.<sup>170</sup> This lesion can be asymptomatic into adulthood.<sup>171–174</sup> Abnormal arterial supply to the lung segments such as seen in scimitar syndrome with sequestration is also well seen by cardiovascular CT.<sup>175–178</sup> Cardiovascular CT is the imaging modality of choice to determine anatomic substrate and interventional planning for complex lung lesions such as intralobar or extralobar pulmonary sequestration.<sup>179,180</sup> In a study evaluating the utility of cardiovascular CT for surgical planning in these patients, treatments were correctly planned using cardiovascular CT with 100% accuracy, sensitivity and specificity.<sup>181</sup>

### 2.5. Septal defects, including ASD/VSD/AVSD

Advanced imaging is rarely needed to evaluate atrial or ventricular septal defects unless associated with systemic or pulmonary venous anomalies. Cardiac CT may be considered prior to device placement in patients with large atrial septal defects (ASD) who have poorly visualized inferior–posterior rims on echocardiography.<sup>182</sup> The retroaortic course of an anomalous circumflex coronary artery from the right facing sinus can be identified prior to device placement additionally. CT evaluation of atrial and venous anatomy in symptomatic patients after Amplatzer device occlusion of ASD has been reported.<sup>183</sup> The region of patch closure of a septal defect may become calcified with aging. Since these lesions are the most common congenital cardiac abnormalities, previously unrecognized septal defects may be first identified by cardiac CT in patients undergoing scanning for other indications.<sup>184</sup> Relief of left ventricular outflow tract obstruction and AV valve revision or replacement are the most common re-interventions after repair of atrioventricular septal defects (AVSD).<sup>185–188</sup> Cardiac CT may be

useful in these patients when echocardiography is not fully diagnostic, although literature regarding cardiovascular CT specific to this diagnosis is not reported.

### 2.6. Tetralogy of Fallot (TOF with pulmonary stenosis or pulmonary atresia)

The anatomic targets assessed with cardiovascular CT in patients with tetralogy of Fallot (TOF) include main pulmonary arteries and pulmonary conduits, branch pulmonary arteries, aorto-pulmonary collaterals, postoperative shunts, coronary arteries, and the aortic root.<sup>189–196</sup> Multimodality imaging is often needed for complete assessment and serial evaluation in these patients. MRI is considered by many experts to be the modality of choice for investigation after TOF repair unless detailed coronary imaging is needed.<sup>197</sup>

The ability to reliably visualize pulmonary arterial supply makes cardiovascular CT an excellent imaging modality in patients with TOF who are not adequately imaged by echocardiography prior to repair, particularly in those with pulmonary atresia. Pulmonary blood flow in patients with TOF with pulmonary atresia (TOF-PA) may be supplied via a patent ductus arteriosus, aorto-pulmonary collaterals or both. In comparison to conventional angiography, cardiovascular CT has excellent accuracy in defining aorto-pulmonary collaterals in these patients prior to surgical unifocalization.<sup>195,198,199</sup> On the other end of the spectrum, TOF with absent pulmonary valve, which is the least common form of tetralogy, often has severe pulmonary artery dilation. This dilation may cause bronchial compression, which is well visualized by cardiovascular CT as is any associated air trapping.<sup>164</sup>

After complete repair of TOF, patients with residual pulmonary regurgitation often develop right ventricular dilation and dysfunction. Furthermore, during adulthood 20% of patients with repaired TOF develop left ventricular systolic dysfunction which may improve after pulmonary valve replacement.<sup>200–202</sup> Right and left ventricular size, systolic function and pulmonary regurgitant fraction are routinely evaluated in order to determine the optimal timing and type (surgical or catheter based) of pulmonary valve or conduit replacement.<sup>203–205</sup> Adult patients with repaired TOF are at increased risk of sudden death after the second decade of life and many eventually meet criteria for placement of a defibrillator, making CMR a relative contraindication.<sup>206–208</sup> For these select patients who are contraindicated for CMR, cardiovascular CT assessment of ejection fraction and ventricular volumes provides comparable information.<sup>209–212</sup> In the absence of shunts or other significant valve disease, pulmonary regurgitation can be estimated from differences between right and left ventricular stroke volumes. Stroke volume differences to estimate pulmonary regurgitation has been reported with adequate correlation to 3 T MRI in this patient subset.<sup>213</sup> Careful correlation to echocardiography should be used when interpreting stroke volume differences to determine valvular regurgitation since flow sequences cannot be used to verify findings as in MRI. If both tricuspid regurgitation and pulmonary insufficiency are present, the total stroke volume can be reported but not the severity of each lesion.

Methods for trans-catheter pulmonary valve placement have been developed for those who meet criteria for intervention. These valves have been primarily used in previously placed conduits with a diameter of 16 mm or larger. Recently they have been placed in the native right ventricular outflow tract (RVOT) and in smaller patients.<sup>214–216</sup> Precise anatomic definition is required for optimal valve sizing and successful valve placement.

Congenital coronary anomalies are relatively common in patients with TOF, and it is important to define the coronary anatomy prior to surgery; particularly in patients with an anomalous



coronary that crosses the RVOT.<sup>217</sup> A preoperative study in 100 patients under one year of age with TOF demonstrated that cardiovascular CT provided 100% sensitivity and specificity for coronary artery anatomy compared to surgical findings, with a radiation dose less than 1 mSv.<sup>189</sup> Coronary artery anatomy must be clearly defined prior to repeat intervention on the RVOT.<sup>23</sup> This is essential in patients undergoing repeat sternotomy with a substernal coronary artery, and also for those being considered for transcatheter pulmonary valve placement due to potential for coronary artery compression with device placement.<sup>218–220</sup> It is important to recognize that the aortic root is commonly dilated in patients with TOF, measuring  $\geq 40$  mm in 28.9% of patients in a recent multi-institutional study.<sup>221</sup> Aortic root dimensions should be measured and reported in all TOF patients whenever a cardiovascular CT is performed.

## 2.7. Transposition complexes

### 2.7.1. Atrial switch (Mustard or Senning Procedure)

The atrial switch was the procedure of choice for *d*-transposition of the great arteries (*d*-TGA) prior to development of the successful coronary artery reimplantation techniques that facilitate the arterial switch operation. Most patients who have undergone the atrial switch are young- to middle-aged adults. In these patients, systemic right ventricular (RV) failure and tricuspid regurgitation are common and are the primary predictors of mortality. For this reason, measurement of systemic RV function is critical, and recent guidelines are based on quantitative evaluation of ejection fraction.<sup>222,223</sup> The pacemaker insertion rate is relatively high in this patient population, with a minority of patients in sinus rhythm 20 years after intervention.<sup>224,225</sup> Complete, pre-procedural assessment of systemic venous anatomy to facilitate potential placement of stents or devices is recommended. In a single-center catheterization study, over 50% of patients had baffle complications; and pre procedural echocardiograms had a positive predictive value of only 37% compared to catheterization findings, underscoring the need for improved pre-procedural diagnostics.<sup>226</sup> In a large cohort of adult patients who had undergone atrial baffle creation, systemic venous baffle obstruction rate was significantly higher for patients who had undergone a Mustard vs Senning operation (risk ratio 3.5).<sup>227,228</sup> Cardiovascular CT is able to visualize systemic and pulmonary venous baffles and identify baffle obstruction, evaluate RV size and function, and estimate tricuspid regurgitation using stroke volume differences from functional analysis.<sup>209,229–232</sup> If stroke volume differences are used for estimation of regurgitant fraction, findings must be correlated with echocardiographic Doppler evaluation. Baffle leaks are difficult to reliably visualize using cardiovascular CT unless there is differential opacification showing a negative or positive contrast jet between the atria. This is problematic when a biventricular contrast injection protocol is used and there is similar contrast density in both atria. Cardiovascular CT has been described for follow-up evaluation of both baffle stents and EP device placement.<sup>231</sup> Another potential indication for cardiovascular CT in this population includes pacemaker dependent patients who are referred for cardiac resynchronization therapy. Pre-procedural evaluation of coronary sinus and coronary venous anatomy by cardiovascular CT can help determine the procedural approach for EP device lead placement.<sup>233,234</sup>

### 2.7.2. Arterial switch

The arterial switch operation (ASO) for *d*-TGA was first described by Jatene in 1975, and has been widely applied since the 1980s.<sup>235</sup> The first patients to undergo arterial switch are now young adults.

With *d*-TGA, abnormalities of coronary origin and course are common and impact immediate and long-term surgical outcomes.<sup>236</sup> Overall, survival after an ASO is excellent. Nevertheless, late deaths resulting from coronary ischemia and arrhythmias have been documented, and the rate of reintervention is relatively high.<sup>237–239</sup> Major complications that may occur after the arterial switch operation include coronary ostial stenosis, neo-pulmonary artery and branch pulmonary artery stenosis, and neo-aortic root stenosis, dilatation or insufficiency. The most common indication for intervention is for relief of supravalvar pulmonary stenosis.<sup>240</sup> As noted above, cardiovascular CT performs well for visualization of the all aspects of the right ventricular outflow tract, and branch pulmonary arteries.

Since myocardial perfusion via reimplanted coronary arteries is the primary determinant of mortality and long-term outcomes in these patients, imaging of the coronary anastomoses is recommended in symptomatic patients, as well as at least once during adolescence or early adulthood in asymptomatic patients.<sup>23,241,242</sup> A recent cardiovascular CT evaluation of 190 patients 5–16 years of age found 8.9% of patients with coronary lesions (defined as  $>30\%$  narrowing to occlusion) confirmed by invasive angiography.<sup>243</sup> Other studies have shown similar rates of coronary compromise, primarily in asymptomatic children.<sup>241,244,245</sup> Cardiovascular CT has been shown to be highly accurate in evaluating coronary arteries before and after an ASO.<sup>130,245,246</sup> No accepted standard for routine interval coronary evaluation exists. Cardiovascular CT evaluation of coronary artery stenosis after the ASO has been shown to correlate well with invasive angiography, and is preferred since catheter placement may alter the coronary ostium. CT also provides information on the underlying mechanism of coronary luminal narrowing.<sup>245</sup> Evaluation after bypass grafting for coronary stenosis resulting from an ASO in children using cardiovascular CT has been reported as well.<sup>247,248</sup>

### 2.7.3. Complex transposition repair (Rastelli and Nikaidoh)

In some patients with complex transposition, such as those with *d*-TGA with a VSD and left ventricular outflow tract obstruction or certain patients with double outlet right ventricle, an ASO is not feasible. The Rastelli procedure is the most common surgical intervention in this situation. This includes closure of the VSD using a patch from the crest of the ventricular septum to the distant aorta and placement of a right ventricle to pulmonary artery conduit. As with all other forms of surgery requiring placement of a conduit, there is potential for stenosis or insufficiency of the conduit, and the relationship of the coronary arteries should be evaluated prior to repeat intervention. A multi-institutional study performed in 2010 revealed a survival of 58%, and an event free survival of 26%, at 20 years after a Rastelli procedure.<sup>249</sup> There is a relatively high reintervention rate for both right and left ventricular outflow tract obstruction, and a majority of patients will require pulmonary conduit replacement.<sup>249–251</sup> The pathway between the left ventricle and aorta may become obstructed, and determining the anatomic substrate is crucial to determine the method of intervention, when indicated. The Nikaidoh procedure, which has gained popularity in recent years, is a procedure in which the aorta is translocated closer to the VSD. Since the aorta must be moved leftwards towards the VSD, right coronary artery lesions are a potential complication.<sup>252–254</sup> All of the anatomic lesions seen after the Rastelli and Nikaidoh procedures can be readily assessed with cardiovascular CT, although literature for this indication is limited to descriptive case reports.<sup>232</sup>

### 2.7.4. Congenitally corrected transposition

Patients with congenitally corrected transposition (also known

as *l*-TGA have a high rate of complete heart block requiring pacemaker placement. The most simple form (with no associated intracardiac defect) may be first uncovered as an incidental finding in CT performed for another indication.<sup>255,256</sup> Cardiovascular CT can be used to assess atrial, ventricular and arterial relationships and to evaluate systemic right ventricular function.<sup>257–259</sup>

Some patients with *l*-TGA can be managed without surgery, and those that undergo surgery may either have a “physiologic” or “anatomic” repair. The “physiologic repair” keeps the right ventricle as the systemic ventricle, occasionally with a left ventricle to pulmonary artery conduit, while an “anatomic repair” consists of an atrial switch plus an arterial switch or Rastelli procedure. Both surgical options have similar medium term outcomes, except for those with significant tricuspid regurgitation, which is better tolerated in an anatomic repair with a systemic left ventricle.<sup>260</sup> Complications, follow-up, and cardiovascular CT imaging are similar to what is described above based on type of repair (atrial switch, arterial switch, Rastelli).

## 2.8. Single ventricle heart disease

Patients with a functionally single ventricle, including those with tricuspid atresia, pulmonary atresia, hypoplastic left heart syndrome, double inlet left ventricle, and unbalanced atrioventricular septal defects, generally follow a palliative surgical pathway with 2–3 stages. The first stage, if necessary and dependent on the physiology, is performed as a neonate and usually involves a Norwood procedure or systemic to pulmonary arterial shunt. Some centers advocate a “hybrid” approach utilizing a catheter-placed ductal stent and pulmonary artery bands. The most common systemic to pulmonary arterial shunt is the Blalock-Taussig shunt, but right ventricle to pulmonary artery shunts (Sano shunt) and central shunts (from ascending aorta) are other common types. At 4–6 months of age the shunt is taken down and the superior vena cava is anastomosed to the pulmonary artery (Glenn or Hemi-Fontan procedure). With the third stage (Fontan completion) the inferior vena caval flow is directed into the pulmonary arteries. In the current era, the third stage palliation is typically performed between 18 months and three years of age. Clinical outcome is dependent upon the morphology of the single ventricle and patients with a systemic left ventricle do better during second stage palliation, have improved ejection fraction, lower rates of valvular regurgitation and fewer long term complications.<sup>261–263</sup> Although patients with a systemic right ventricle do not do as well, it is now expected that 70% will survive to adulthood and most patients do well clinically.<sup>264,265</sup> A single institution reported a median radiation exposure of 25.7 mSv through the Fontan operation using primarily catheter based diagnostics.<sup>266</sup>

### 2.8.1. Prior to stage 1 surgery

While many patients with single ventricle anatomy can be imaged adequately using echocardiography prior to stage 1 surgery, cardiovascular CT is occasionally necessary to define complex systemic or pulmonary venous, aortic, or pulmonary artery anatomy, particularly in patients with atrial isomerism. Given that pulmonary venous anomalies are a significant risk factor for survival in these patients, it is critical that the pulmonary venous anatomy is defined accurately prior to intervention.<sup>267–269</sup> Cardiovascular CT is excellent for this application, and can be performed with minimal or no sedation in most cases. (See section above on thoracic vascular abnormalities).

### 2.8.2. After stage 1 surgery (Norwood, systemic to pulmonary arterial shunt, hybrid)

Between stage 1 and 2 surgery, systemic and pulmonary artery stenoses are relatively common, and are often insufficiently visualized with echocardiography.<sup>39</sup> Patients with systemic to pulmonary artery shunts occasionally experience shunt thrombosis, resulting in acute, profound cyanosis. Shunt thrombosis can be challenging to identify with echocardiography, but cardiovascular CT, given its easy accessibility and short imaging time, is an excellent imaging modality to identify this problem and identify when intervention is necessary.<sup>270,271</sup>

In most centers, cardiac catheterization is performed in preparation for a stage 2 procedure. A recent comparison of cardiovascular CT and catheterization prior to stage 2 palliation revealed excellent correlation to surgical findings for both modalities and no difference in surgical outcome to hospital discharge.<sup>272</sup> The estimated cardiovascular CT radiation dose (both age and size adjusted) was 1 mSv compared to a catheterization dose estimate of 14 mSv. Additionally, the catheterization group had higher contrast dose, required central vascular access and general anesthesia in all cases, and had a relatively high rate of adverse events. A prior study randomized pre-stage 2 patients to CMR or catheterization, and found no difference in surgical outcomes or medium term outcomes for patients followed a median of 8 years.<sup>273</sup> Some centers now propose a completely non-invasive diagnostic pathway for patients with single ventricle heart disease through third stage palliation.<sup>274,275</sup> The single ventricle patient population is high risk for adverse event with anesthesia.<sup>276–278</sup> For single ventricle patients that may require anesthesia for CMR, cardiovascular CT may be a reasonable alternate imaging modality if performed with no or minimal sedation.

### 2.8.3. After stage 2 surgery (Glenn or Hemi-Fontan procedure) or stage 3 surgery (Fontan)

Cardiovascular CT has been shown to adequately visualize all aspects of the Glenn or Fontan circuit after single ventricle palliation.<sup>279–286</sup> Thrombus formation after the Fontan procedure is relatively common, and thrombi have been visualized by cardiovascular CT in the Fontan conduit, residual ventricle or in the residual PA stump after pulmonary artery ligation.<sup>287–290</sup> Pulmonary embolism has also been identified by cardiovascular CT.<sup>291</sup> Care must be taken, however, to optimize the contrast injection technique both to avoid a false positive diagnosis of pulmonary embolism and to optimally opacify the Fontan circuit.<sup>232,285,292</sup> Unopacified venous blood from the hepatics mixing with a lower extremity injection, or inferior vena cava mixing with contrast from an upper extremity injection can be mistaken as clot or embolism. As with TOF or TGA, quantification of ventricular function by cardiovascular CT may be warranted in patients with metallic implants and contraindications to CMR.

## 2.9. After Ross procedure

The Ross procedure is performed for children and adults as an alternative to prosthetic aortic valve placement. In this procedure, the pulmonary valve and root are harvested and placed in the aortic position, with re-implantation of the coronary arteries and placement of a right ventricle to pulmonary artery conduit. Evaluation after the Ross procedure requires visualization of the neo-aortic root, reimplanted coronary arteries and pulmonary conduit. As stated in prior sections above and in additional publications, cardiovascular CT performs well for these indications.<sup>293–296</sup>

## 2.10. Other complex CHD (stent, VAD, ECMO)

Cardiovascular CT may be considered for determination of stent integrity, diagnosis of aneurysm and other complications of stent placement and assessment of either airway or coronary compression from mass effect after intravascular intervention for aortic or pulmonary abnormalities.<sup>157,297–301</sup> Evaluation of coronary and vascular stents using cardiovascular CT is highly accurate when compared to traditional angiography.<sup>157,298,302–304</sup> Beam hardening artifact and the partial volume effect may degrade in-stent evaluation for vessels less than 3 mm, although iterative reconstruction and appropriate kernel selection can improve image quality.<sup>305–307</sup> Pediatric vascular stents were evaluated in an *in vitro* model found excellent correlation to conventional angiography in pediatric patients, despite at low tube potential settings and small stent sizes.<sup>298</sup>

Cardiovascular CT also provides diagnostic information (cannula positions, presence of thrombus, driveline infection) in patients on ECMO or with ventricular assist device (VAD) support, a population in whom conventional imaging may be challenging.<sup>308,309</sup>

## 2.11. Use of CT for functional imaging

Echo and CMR are first line non-invasive modalities to assess ventricular function in patients with CHD, with cardiac CT offering an accurate alternative for this application in patients with CHD when contraindications or limitations to these modalities exist.<sup>211,212,310</sup> Heart Failure is increasingly common in adults with CHD and serial evaluation is sometimes needed to help with medical management and advanced therapy decisions.<sup>311</sup>

Cardiac CT may be used for ventricular function analysis when data acquisition (“ECG trigger or ECG gating”) and image reconstruction are synchronized to the ECG and then reconstructed in a multiphase dataset. With a retrospectively ECG-gated helical protocol, ECG-based tube current modulation allows full radiation during a specified short portion of the cardiac cycle while tube current is reduced during the remainder of the cardiac cycle. This dose modulation protocol allows reconstruction of an end systolic dataset with sufficient image quality to detect the endocardial contours in addition to a high quality diastolic dataset. Prospectively ECG-triggered datasets can also be used for functional analysis, as long as the data acquisition window captures both end-systole and end-diastole. Several publications have demonstrated that both right and left ventricular systolic function can be measured by cardiac CT with accuracy comparable to CMR.<sup>18,257,312–316</sup> An assessment of MRI vs DSCT found that the function results were considered interchangeable.<sup>210,317,318</sup> When estimates of ventricular volumes and calculation of ejection fraction were compared to known volumes using a moving heart phantom and standard clinical imaging protocols, DSCT performed better than both MRI or 64 slice CT. The accuracy of 64 slice CT was dependent on heart rate, however.<sup>319</sup> The accuracy of CT for functional analysis will depend on the temporal resolution available on the scanner platform and scan sequence used. For functional imaging, beta blockade is not typically required unless high resolution distal coronary imaging is also needed from the fully radiated phase of the dataset.

Since 7–10 mm slice thickness is sufficient for both RV and LV quantification, lower-dose scans yield evaluable datasets for this purpose.<sup>320,321</sup> Heart failure associated with ventricular pacing is the largest indication for biventricular pacing in pediatric patients and CHD.<sup>322</sup> Cardiac CT has been shown to evaluate regional wall motion associated with ventricular pacing in a small cohort of patients.<sup>323</sup>

### 2.11.1. Valvular stenosis, regurgitation, prosthetic valve and perivalvular leak

Many patients with CHD need repeat valve intervention, commonly on more than one valve.<sup>131</sup> In young patients undergoing mitral valve replacement, 50% will require re-replacement within 10 years, and 15% require pacemaker placement within one month of valve placement.<sup>324,325</sup> There are several studies showing the utility of cardiac CT for evaluation of native and mechanical valve stenosis and insufficiency, perivalvular leak, thrombosis, abscess and endocarditis.<sup>326–333</sup> Stroke volume differences between ventricles calculated from a functional dataset may be used to quantify the severity of valvular regurgitation if correlated closely with echocardiographic findings.<sup>213</sup>

Quantification of regurgitation is not possible in single ventricle patients or in patients with more than one regurgitant lesion or intracardiac shunt. The total difference in stroke volume is evaluable, with reliance on other modalities for assessment of the contribution of each lesion (see section on function imaging). Assessment of coronary artery anatomy in relationship to the mitral valve (occasionally supraannular) is needed for surgical planning at the time of replacement. In patients with pacemakers or previously replaced mechanical valve, complete pre-operative assessment of anatomy, including coronary artery anatomy, is often required and can be performed with cardiovascular CT.

### 2.12. Sternal re-entry in high risk patients

It has long been appreciated that reoperation in patients with CHD carry increased risk of serious vascular injury upon sternal reentry, sometimes requiring emergent peripheral cannulation for cardiopulmonary bypass.<sup>334,335</sup> While a recent study suggests that the risk has decreased with improved surgical techniques, presence of right ventricle to pulmonary artery conduits and increasing number of sternotomies remain risk factors for injury.<sup>336</sup> It is critical to define the proximity of the coronary arteries and cardiac structures to the posterior sternum prior to repeat sternotomy for consideration of peripheral bypass at the time of sternal entry.<sup>337</sup> Some authors advocate cardiovascular CT prior to repeat sternotomy in select patients.<sup>338</sup> While CMR can identify the relationship between vascular structures and the sternum, artifact resulting from sternal wires limits its ability to define this relationship with the clarity afforded by cardiovascular CT.

## 3. Risk of CT in the current ERA

### 3.1. Sedation/anesthesia

The time required to image the thorax ranges from 0.25 to 10 s for 64–320 slice CT scanners depending on factors such as the need for ECG gating and scan length. The newest generation scanners acquire the dataset in a fraction of a second or a single heartbeat, reducing or eliminating the need for sedation and suspended respiration for a majority of indications.<sup>14</sup> Studies have shown that images acquired without sedation in neonates and with conscious sedation in toddlers and young children yield adequate to excellent image quality.<sup>10,14</sup> When image acquisition requires breath holding, only a single and short duration suspension of respiration is needed.<sup>10,14,189,339</sup> Most patients age  $\geq 7$  years can reliably cooperate with scanning instructions. General anesthesia (GA) may be required for the youngest patients who cannot sustain a breath hold when required for scan sequences that acquire data over several heart beats. This includes detailed coronary artery imaging at high heart rates and ventricular function measurement. When GA is needed, no change in ventilator management or specialized equipment is needed for cardiovascular CT acquisition. For older

generation scanners with image acquisition times of 6–10 s, sedation or anesthesia may still be required in children and developmentally delayed patients of any age unable to cooperate with breath hold instructions to eliminate motion artifact even for non-ECG triggered scans.

In pediatric patients, general anesthesia (GA) confers risks of both procedural complication as well as the potential for long-term adverse neurodevelopmental outcome.<sup>340</sup> In a multi institutional study, those with congenital heart disease in anesthesia class 3 or above were at highest risk for a procedural adverse event with GA.<sup>277,341</sup> The risk of cardiac arrest with GA is highest in the youngest patients and in those with unrepaired single ventricle heart disease, pulmonary hypertension, left ventricular outflow tract obstruction and cardiomyopathy.<sup>276,341</sup> In patients with congenital heart disease, the risk of GA has been shown to be higher when performed outside of the operating room.<sup>341,342</sup> In a study examining the complications associated with CMR, the use of GA significantly increased the risk of adverse events, with an odds ratio of 3.9.<sup>342–344</sup>

Anesthesia exposure in young patients may adversely affect long term cognitive and behavioral outcomes, particularly those exposed to prolonged or multiple anesthetics before age 2 years.<sup>345–351</sup> This concern is most relevant for CHD patients who will undergo multiple diagnostic evaluations and palliative interventions in the first year of life that will require anesthesia, such as patients who have TOF with pulmonary artery atresia or single ventricle heart disease. For patients who do require anesthesia the length of anesthesia for a CT study will be much shorter than for either CMR or cardiac catheterization.<sup>340,349</sup>

Infants with CHD referred for advanced cardiac imaging often require vasoactive infusions and/or mechanical ventilation in the intensive care unit. CMR in such patients requires conversion to MRI-compatible equipment, followed by 1–2 h of anesthesia with multiple breath holding sequences.<sup>342</sup> Poor thermoregulation is also a challenge in small patients with prolonged anesthesia. CT can be performed quickly, patients can be returned to the intensive care unit within 15–30 min of leaving, and there is no need for conversion to and from specialized equipment. During the scan, patients are more accessible during computed tomography.<sup>340</sup> CT can even be performed on patients on ECMO.<sup>308</sup>

### 3.2. Vascular access

IV access is required for contrast administration, usually delivered with a power injector. There is a low complication rate (0.2–0.4%) using power injectors with contrast injected via many different venous access devices in pediatric patients.<sup>352</sup> Power injection through central lines using low pressure limits and longer injection times is considered safe, but provides inadequate contrast opacification in patients weighing over 30 kg.<sup>353</sup> A more recent study of peripheral IV power injection for CT examinations in children using 22 gauge angiocatheters in 443 of 557 children (range 18–24 gauge) at a median flow rate of 1.5 ml/s reported two episodes of contrast extravasations treated conservatively (0.3%).<sup>354</sup> Safe use of power injectors in neonates at low flow rates has been reported.<sup>10</sup>

### 3.3. Contrast exposure

Almost all cardiovascular examinations are performed with iodinated contrast. Typical exams require 1–2 ml/kg of contrast volume. The rate of adverse reaction from iodinated contrast administration is very low in adult patients, ranging from 0.1% to

1% in several studies.<sup>355–358</sup> In several large reviews of pediatric age range patients the incidence of contrast reaction is also low (0.18–0.46%).<sup>359,360</sup> The incidence of contrast induced nephropathy is highest in patients with severely reduced renal function and use of pre-scan hydration may decrease the incidence of adverse renal effects.<sup>361</sup> If CT is required in the setting of renal failure, every effort should be made to withdraw nephrotoxic drugs, select low or iso-osmolar contrast media, use as little contrast as possible, and consider pre-scan hydration.<sup>362</sup> Gadolinium-enhanced CT studies have been reported in iodine allergic patients and the fairly low *k* edge of gadolinium is particularly suited to reduced kV imaging.<sup>363</sup>

### 3.4. Medications to lower heart rate

For coronary artery imaging, a heart rate below 60 beats per minute maximizes the potential for obtaining high quality images within a single diastolic interval and requires the lowest radiation exposure.<sup>364,365</sup> Even with beta blockade the heart rate will be elevated in small patients. Significant heart rate variability during the monitoring or acquisition phase of certain scan sequences will automatically widen the acquisition window. Pre-procedural medication can be used to decrease the overall heart rate or the variability with respiration. Different protocols for beta blockade have been described and are effective for decreasing the heart rate and thus the required radiated interval of the cardiac cycle.<sup>366–368</sup> Protocols specific to children have been described with effective heart rate control and excellent safety.<sup>10,369</sup> The safety of heart rate lowering medications must be assessed for each patient prior to administration. Hemodynamically unstable children and those with pulmonary hypertension may not tolerate the effects of beta blockade and are often imaged at their intrinsic heart rate. For heart rates above 60 beats per minute, images with minimum cardiac motion artifact may be obtained during either the end systolic or end diastolic phase of the cardiac cycle. Some scans obtained at higher heart rates will require a widened acquisition window and higher radiation doses.<sup>370–372</sup>

### 3.5. Radiation exposure

Ionizing radiation is fundamental to image creation with CT, and radiation exposure is thought to increase the risk of future development of cancer.<sup>373–376</sup> A linear no threshold model has been adopted for medical radiation exposure. The risk of radiation exposure is particularly relevant for young patients due to both their longer expected lifespan and greater radiation sensitivity compared to adults. The radiation exposure required for cardiac CT has decreased significantly in the last number of years with the introduction of a number of dose reduction techniques.<sup>377,378</sup> These include the availability of low tube potential (e.g. 70, 80, 90 or 100 kV) settings, ECG-based tube current modulation and anatomic-based tube current modulation.<sup>10,11,13,379–381</sup> The introduction of iterative reconstruction algorithms allow for a reduction in tube current and radiation dose while maintaining acceptable noise properties.<sup>15,78,339,382–384</sup> Improvements in detector technologies have also allowed radiation exposures to be lowered. To achieve a lower estimated dose in smaller patients, tube potential and tube current should be adjusted to patient size with 70 or 80 kV as the default tube potential. Retrospective ECG-gated helical scans should be reserved for evaluation of ventricular function and for detailed coronary artery assessment when arrhythmia is present.<sup>13,365,385,386</sup>

The most advanced CT scanner platforms can routinely achieve



**Table 1**

Optimal imaging environment for cardiovascular CT in CHD.

- Alternate cardiac imaging modalities are available so that the test with the least risk can be performed for a specific clinical indication
- Close collaboration & communication is present among surgeons, clinical cardiologists and imagers
- All patient clinical information is accessible to allow understanding of the clinical indication and potential management options for the patient
- Scan protocols can be designed and adjusted to extract maximum clinical information at minimum procedural risk
- Technologists are experienced in cardiac CT and comfortable with varied cardiac scan modes
- Easy access to pacemaker programming to allow rate and mode adjustment
- Nursing support to facilitate administration of medication for heart rate control when necessary in patients with and without permanent pacemakers, and to provide appropriate monitoring for any side effects
- Access to all forms of prior imaging (echocardiography, angiography, nuclear, CMR) so that a targeted evaluation may be performed for an individual patient
- Post-processing workstations capable of handling large multiphase data sets for advanced reconstructions
- High-speed network to transfer large volume data sets from scanner to workstation
- Immediate availability of advanced resuscitation equipment and resuscitation team appropriate for the size and age of the patient

an effective radiation dose estimate of less than 1 millisievert (mSv) for many congenital cardiac applications, even when using the size and age adjusted CT dose volume index (CTDIvol) and chest conversion factors.<sup>10–13,16,17,387</sup> The radiation dose estimate for an infant will be increased by a factor of seven over the standard adult estimate if the smaller phantom (16 cm vs 32 cm) is used to determine CTDIvol, and an age adjusted chest conversion factor is

used to convert this value into mSv.<sup>10–13,16,198,387,388</sup> Size specific dose estimate (SSDE) is another radiation measurement used by some centers to estimate an individual organ dose from scanner output and patient specific chest measurements. The estimated organ dose cannot be used to determine a patient dose.<sup>389</sup> When pediatric radiation dose estimates are reported, it should be stated which phantom size is used to estimate CTDIvol (16 cm or 32 cm)

**Table 2**

Relevant knowledge for the performance of cardiovascular CT in CHD.

**Cardiology/CHD knowledge required**

- Anatomy & physiology of CHD – natural and repaired
- Surgical procedures used to palliate or repair CHD
- Catheter interventions used to palliate or repair CHD
- Material composition of the surgical materials or catheter devices used and the artifact produced in different imaging modalities (MRI and CT)
- Common residual hemodynamic lesions following initial CHD repair
- Indications for re-intervention (AHA/ACC/ESC/CCSHRS guidelines)
- Normal coronary anatomy
- Congenital coronary anomalies and the indications for and methods of repair
- Basic ECG knowledge and arrhythmia recognition (and impact on imaging strategy)
- Pediatric and adult doses for heart rate lowering medications and sublingual nitroglycerin, and contraindications to these medications

**CT technique specific knowledge required**

- Training and experience in congenital cardiac CT (there are no current educational standards for CHD CT)
- Scanning principles and scan modes, including the different capabilities of individual scanner platforms
- Contrast injection protocols adjusted for both patient size and cardiac pathology
- Prophylaxis against and treatment of minor and major contrast reactions
- Radiation physics and basics of radiation dose measurement
- Radiation dose reduction strategies and individualized scan planning
- Familiarity and competence with post-processing methods and software
- Familiarity with standards for quantification and reporting in CHD

**Table 3**

Situations in which cardiovascular CT may be considered in CHD.

- Presence of CMR incompatible implant or foreign body (retained pacing leads, non-MR compatible pacemaker/defibrillator, neurostimulator)
- Poor CMR image quality (known or expected) due to metallic artifact
- Unable to fit in MRI scanner due to obesity, or severe claustrophobia
- Neonate or young patient requiring evaluation of complex anatomy, particularly if considered higher risk for adverse event with sedation or anesthesia required for CMR, and the CT scan can be performed with no or limited sedation
- Critically ill patient of any age that may not tolerate breath holding or length of CMR scan
- Evaluation of ventricular assist device or ECMO cannula positioning
- Patient requiring CT for evaluation of extra-cardiac anatomy in addition to CHD (e.g. lung parenchyma, airway, skeletal abnormality)
- Pre-operative patients with prior sternotomy considered high risk for vascular injury with sternal reentry due to an anterior coronary artery, conduit, or sternal adhesions
- Evaluation of prosthetic valve function or structural integrity (calcification, stenosis, coaptation defect, leaflet immobility, paravalvular leak, endocarditis or clot)
- Evaluation of calcification within vessels and surgical conduits prior to catheter-based intervention (e.g. balloon angioplasty, transcatheter valve replacement, stent placement).
- Coronary artery imaging in CHD:
  - a) Patient needing detailed pre-operative coronary artery evaluation in addition to assessment of complex cardiac anatomy
  - b) Patient with symptoms and signs suggestive of atherosclerotic coronary artery disease and a history of CHD, prior coronary intervention, or high risk Kawasaki disease
  - c) Young symptomatic patients with known or suspected coronary anomaly, particularly if CMR is unlikely to provide complete assessment or more likely to require anesthesia
  - d) Delineation of coronary anatomy prior to surgical or percutaneous pulmonary valve implantation
  - e) Evaluation of coronary artery after any surgery requiring coronary artery manipulation or reimplantation

and which age and chest conversion factors are used. This will allow equivalent comparisons of radiation dose estimates despite the variability in reporting of scanner output and calculation of dose in pediatric patients.

ECG-gated cardiac CT studies have historically yielded higher radiation doses using older retrospective ECG triggering techniques.<sup>390</sup> A study published in 2011, however, shows consistently lower doses for 64-slice CT than for conventional angiography.<sup>391</sup> A recent study documented a median effective dose of 1 mSv for ECG-gated helical and ECG-triggered axial coronary CTA in a wide range of pediatric patients.<sup>10</sup> CT now has the potential to deliver 10–15 fold less radiation than cardiac catheterization when used by experienced users at centers with modern 64-slice or greater CT scanners and with careful attention to scan parameters.<sup>391,392</sup> However, if scan parameters are not carefully adjusted for clinical indication and patient size, radiation doses may be significantly higher. A recent publication estimates cumulative radiation exposure from cardiac diagnostics for patients less than six years of age,

required for a detailed coronary artery scan is not needed for the majority of CHD scan indications. Recent recommendations suggest that the risks and benefits of cardiac imaging should be discussed as part of informed consent, and that radiation parameters should be included in procedural reporting.<sup>394</sup>

#### 4. Recommendations for CT imaging in CHD

Cardiac CT will have an important role in the future of congenital cardiac imaging. The Tables 1–3 below summarize consensus recommendations for the imaging environment and required knowledge to perform high quality CT in patients with CHD, and clinical scenarios where CT may be used.

#### Conflict of interest

#### Authors' Reported Industry/Other Relationships

Last name	First name	MI Role	Reported Industry/Other Relationships
Abbara	Suhny	Writing Group	Grant/Research Support: Siemens (Institutional support), Philips (Institutional support), NIH. Textbook royalties: Elsevier/Amirysis
Bardo	Dianna	Writing Group	Consultant: Koninklijke Philips NV; Speakers Bureau: Koninklijke Philips NV
Crean	Andrew	Writing Group	Nothing to Disclose
Ghoshhajra	Brian	Writing Group	Consultant: Siemens Healthcare, USA
Han	B. Kelly	Writing Group	Grant/Research: Siemens Healthcare (Institutional support)
Hlavacek	Anthony	Writing Group	Grant/Research: Siemens Healthcare (Institutional support)
Leipsic	Jonathon	Writing Group	Grant/Research: Heartflow, Edwards Lifesciences – Core lab services; Consultant: Circle CVI, Heartflow, Edwards, GE Healthcare
Lesser	John	Writing Group	Nothing to Disclose
Nicol	Edward	D Writing Group	Nothing to Disclose
Raman	Subha	Writing Group	Nothing to Disclose
Rigsby	Cynthia	Writing Group	Nothing to Disclose
Siegel	Marilyn	Writing Group	Consultant: Spouse GE Healthcare Consultant Advisory Board

#### Discussion of off label use.

Last name	First name	MI	Role	Off label use
Leipsic	Jonathon		Writing Group	Sapien 3 THV 2. Heartflow FFRCT

and shows relatively high radiation exposure for patients undergoing repeat diagnostic examinations such as transplant recipients and those with single ventricle heart disease.<sup>393</sup> In this study, ECG gated CT scans were estimated to deliver twice the radiation dose of cardiac catheterization. Such findings underscore the importance of meticulous attention to dose reduction techniques with every cardiovascular CT examination to minimize both procedural and cumulative radiation exposure. Scanner output recommendations are often for high resolution coronary artery imaging. For evaluation of larger cardiac structures this level of detail (and radiation dose) are not needed for clinical decision making. Congenital cardiac CT imagers should communicate with referring cardiologists to determine the minimum image quality required that will deliver the diagnostically important information. The image quality

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