

Pediatric Cardiothoracic CT Guideline Provided by the Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group: Part 2. Contemporary Clinical Applications.

Title: Pediatric Cardiothoracic CT Guideline Provided by the Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group: Part 2. Contemporary Clinical Applications.

Publication year: 2021

Journal: Korean journal of radiology

Authors: Department of Radiology and Research Institute of Radiology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea. ghw68@hanmail.net.

Society: European Respiratory Society

Origin country: International (Europe)

Target countries: Europe

DOI: 10.3348/kjr.2020.1332

PMID: 33987995

Source: <https://pubmed.ncbi.nlm.nih.gov/33987995/>

Korean J Radiol Korean J Radiol 659 kjr 100956096 KJR Korean Journal of Radiology 1229-6929 2005-8330 Korean Society of Radiology PMC8316776 PMC8316776.1 8316776 8316776 33987995 10.3348/kjr.2020.1332 1 Pediatric Imaging Review Article Pediatric Cardiothoracic CT Guideline Provided by the Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group: Part 2. Contemporary Clinical Applications <https://orcid.org/0000-0001-6861-5958> Goo Hyun Woo 1 <https://orcid.org/0000-0002-4140-6280> Siripornpitak Suvipaporn 2 <https://orcid.org/0000-0001-6181-011X> Chen Shyh-Jye 3 <https://orcid.org/0000-0003-3802-4801> Lilyasari Oktavia 4 <https://orcid.org/0000-0002-0164-8752> Zhong Yu-Min 5 <https://orcid.org/0000-0001-5806-9113> Latiff Haifa Abdul 6 <https://orcid.org/0000-0003-2123-9187> Maeda Eriko 7 <https://orcid.org/0000-0002-6235-6550> Kim Young Jin 8 <https://orcid.org/0000-0002-4356-918X> Tsai I-Chen 9 <https://orcid.org/0000-0002-5831-7954> Seo Dong Man 10 1 Department of Radiology and Research Institute of Radiology, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Korea . 2 Department of Diagnostic and Therapeutic Radiology, Faculty of Medicine, Ramathibodi Hospital, Mahidol University, Bangkok, Thailand . 3 Department of Medical Imaging, National Taiwan University, Medical College and Hospital, Taipei, Taiwan . 4 Department of Cardiology and Vascular Medicine, Universitas Indonesia, National Cardiovascular Center Harapan Kita, Jakarta, Indonesia . 5 Diagnostic Imaging Center, Shanghai Children's Medical Center, Shanghai, China . 6 Pediatric and Congenital Heart Centre, Institut Jantung Negara, Kuala Lumpur, Malaysia . 7 Department of Radiology, The University of Tokyo, Tokyo, Japan . 8 Department of Radiology, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea . 9 Congenital Heart Disease Study Group Member of the Asian Society of Cardiovascular Imaging, Taichung, Taiwan . 10 Department of Cardiothoracic Surgery, Ewha Womans University Seoul Hospital, Seoul, Korea . Corresponding author: Hyun Woo Goo, MD, PhD, Department of Radiology and Research Institute of Radiology, Asan Medical Center, University of Ulsan College of Medicine, 88 Olympic-ro 43-gil, Songpa-gu, Seoul 05505, Korea. ghw68@hanmail.net 8 2021 04 5 2021 22 8 386639 1397 1415 09 11 2020 27 1 2021 31 1 2021 01 08 2021 03 08 2021 03 08 2021 Copyright © 2021 The Korean Society of Radiology 2021 The Korean Society of Radiology <https://creativecommons.org/licenses/by-nc/4.0/> This is an Open Access article distributed under the terms of the Creative Commons Attribution Non-Commercial License (<http://creativecommons.org/licenses/by-nc/4.0/>) which permits unrestricted non-commercial use, distribution, and reproduction in any medium, provided the original work is properly cited. The use of pediatric cardiothoracic CT for congenital heart disease (CHD) was traditionally limited to the morphologic evaluation of the extracardiac thoracic vessels, lungs, and airways. Currently, the applications of CT have increased, owing to technological advancements in

hardware and software as well as several dose-reduction measures. In the previously published part 1 of the guideline by the Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group, we reviewed the prerequisite technical knowledge for clinical applications in a user-friendly and vendor-specific manner. Herein, we present the second part of our guideline on contemporary clinical applications of pediatric cardiothoracic CT for CHD based on the consensus of experts from the Asian Society of Cardiovascular Imaging CHD Study Group. This guideline describes up-to-date clinical applications effectively in a systematic fashion.

With its increased utility, cardiothoracic CT has replaced diagnostic catheter angiography in patients with congenital heart disease (CHD) [1 2]. CT protocols need to be optimized to prevent outliers with high radiation exposure [3] and, thus, to increase its benefit-risk ratio [4 5 6]. In patients with CHD, the clinical applications of CT have been traditionally confined to the morphologic evaluation of the extracardiac thoracic vessels, lungs, and airways [7 8]. The recent introduction of electrocardiography (ECG)-synchronized CT scan enables us to perform quantitative and functional evaluations of the cardiovascular structures [9 10]. In CHD, combined multiple cardiac defects are commoner than isolated ones and often associated with anomalies of other systems or organs. In such cases, a systematic organ-based approach seems to more appropriately demonstrate clinical applications compared to overlapped descriptions among specific cardiac defects as in the so-called systematic segmental approach commonly used to evaluate patients with CHD.

An expert consensus describing the rationale and utility of CT in patients with CHD was previously provided by the Society of Cardiovascular Computed Tomography [11]. However, ample accounts on the clinical experiences with pediatric cardiothoracic CT in Asian countries [12] may provide more useful information about the practical applications. Furthermore, guidelines need to be updated to include new information. Therefore, we comprehensively discuss the contemporary organ-based clinical applications of pediatric cardiothoracic CT, as the second part of the guidelines provided by the Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group.

Most pulmonary artery anomalies are associated with CHD, and they may have underlying syndromes [13 14]. Cardiothoracic CT is useful for evaluating these pulmonary artery anomalies associated with CHD [11].

In tetralogy of Fallot, the most common congenital cyanotic heart disease, a broad spectrum of pulmonary artery hypoplasia in addition to pulmonary annular and infundibular narrowing is observed on CT [15]. The sizes of these structures are usually close to normal in pink tetralogy of Fallot. In contrast, pulmonary artery hypoplasia tends to be severer in patients with severe cyanosis, and severe infundibular narrowing is observed during hypoxic spells. This information is crucial for planning right surgical ventricular outflow tract reconstruction. In patients with severe pulmonary artery hypoplasia, a palliative shunt may be necessary to increase pulmonary artery flow.

In CHD, pulmonary artery stenosis may involve central or peripheral branches. Central stenosis occurs commonly in a juxtaductal region of the pulmonary artery, most commonly involving the left side. In contrast, peripheral stenosis is often associated with genetic syndromes, such as Williams and Alagille syndromes, and its morphologic types on CT varies from focal narrowing to long-segment hypoplasia. Recently, CT was used to provide an accurate quantitative measure of differential pulmonary vascularity in patients with pulmonary artery stenosis [16].

In pulmonary atresia and ventricular septal defect, multiple pulmonary arterial flow sources originate from the systemic arterial system, including a patent ductus arteriosus (PDA) or major aortopulmonary collateral arteries. CT is useful to accurately map the morphological characteristics, including the size, number, location, and stenosis, of these collateral arteries and small central pulmonary arteries at various stages of unifocalization (Fig. 1A) [17]. The presence and confluence of the central pulmonary artery can be evaluated accurately with CT although the size is small because of its high spatial and contrast resolutions [18]. The presence/absence of

communication between the central pulmonary artery and the systemic arterial collaterals is also important for optimal surgical planning, which should be evaluated with CT (Fig. 1B) and subsequently confirmed on catheter angiography. Stenosis may be observed in the origin or proximal portion of the major aortic collaterals (Fig. 1C), and its presence decreases hemodynamic contribution of the affected collateral artery to total pulmonary arterial flow. The key role of CT is to reduce procedure time and potential diagnostic errors of preoperative diagnostic cardiac catheterization and help optimally plan unifocalization surgery.

In truncus arteriosus, a single great artery with a single semilunar valve supplies the pulmonary, systemic, and coronary circulations. Four types based on the branching pattern of the pulmonary artery can be accurately evaluated with CT, which is helpful for surgical planning (Fig. 2) [19].

Aortopulmonary window is a rare anomaly, characterized by abnormal communication between the ascending aorta and pulmonary artery and classified into the proximal, distal, and total defects depending on the location and extent of the defect (Fig. 3) [20].

In absent pulmonary valve syndrome, CT is useful for assessing vascular bronchial compressions due to markedly dilated pulmonary arteries before and after surgical treatment (Fig. 4) [21].

In left pulmonary artery sling, stenosis of the affected pulmonary artery and associated tracheobronchial abnormalities can be accurately evaluated with CT (Fig. 5) [22]. CT is also useful after reimplantation of the left pulmonary artery with or without tracheoplasty.

In anomalous pulmonary venous returns, where part or all of the pulmonary veins drain anomalously into the right atrium directly or indirectly, CT shows high diagnostic accuracy in delineating pulmonary vein morphology, obstruction of the pulmonary venous drainage, and associated anomalies [23 24].

Partial anomalous pulmonary venous return (PAPVR) involving at least one, but not all, pulmonary veins (Fig. 6) is usually asymptomatic or mildly symptomatic, and it is often detected incidentally. The anomaly most commonly involves the right upper thorax, in which the right upper pulmonary vein anomalously drains into the right atrium or the superior vena cava.

In total anomalous pulmonary venous return (TAPVR), which is commonly associated with right atrial isomerism [23], the anomaly is classified into the supracardiac, cardiac (Fig. 7), infracardiac, and mixed types based on the location of the anomalous drainage [23 24]. When the infracardiac or mixed type is suspected on echocardiography, the upper abdominal region should be included in the scan range for cardiothoracic CT.

Anomalous pulmonary venous returns are frequently associated with atrial septal defect or patent foramen ovale [23 24]. Notably, CT is commonly used to identify PAPVR associated with sinus venosus defect (Fig. 8). PAPVR, or rarely TAPVR, is one of anomalies constituting scimitar syndrome, which commonly involves the right lung [23].

CT is frequently used for the accurate and prompt diagnosis of congenital pulmonary vein obstruction [23 25 26]. Unilateral pulmonary vein atresia is usually asymptomatic and incidentally detected in most cases. In contrast, bilateral pulmonary vein atresia or common pulmonary vein atresia, although extremely rare, cannot be distinguished from obstructive TAPVR clinically, but it can be accurately diagnosed with CT [26].

CT is also useful for evaluating postoperative residual or recurrent pulmonary vein stenosis or occlusion (Fig. 9).

Congenital anomalies of the aorta may occur in isolation or more commonly with other cardiac defects (especially involving the conotruncal region) and chromosomal abnormalities [27].

Supravalvular aortic stenosis may occur in association with or without Williams syndrome, and it may show a discrete narrowing (mimicking hourglass deformity), multiple stenoses, or diffuse hypoplasia on CT (Fig. 10) [28]. Coronary artery abnormalities, including ostial stenosis, diffuse stenosis, dilatation, and inflow obstruction by the aortic valve, the sinotubular ridge, or a combination of both, may be identified on CT [29]. CT is also useful for evaluating post-aortoplasty outcomes [30].

Coarctation of the aorta may present as a discrete juxtaductal stenosis, tubular arch hypoplasia, or a combination of both (Fig. 11) [31]. Interrupted aortic arch, defined as the loss of luminal continuity between the ascending and descending aorta, is divided into type A (distal to the left subclavian artery; most common in Asia), type B (between the left subclavian artery and left common carotid artery; most common in West), and type C (between the left common carotid artery and innominate artery; rare) according to the location of the interruption in the left arch [32]. In these two anomalies, CT can provide detailed aortic arch morphology, left ventricular outlet obstruction associated with posterior malalignment ventricular septal defect, combined cardiovascular defects, and the absence of the thymus in cases of DiGeorge syndrome [31 32]. Additionally, CT is useful for identifying post-treatment complications such as residual or recurrent aortic stenosis, dissection, and pseudoaneurysm (Fig. 12) [31 32].

A vascular ring encircling the trachea and esophagus may cause variable degrees of compression leading to respiratory symptoms and/or dysphagia [33]. The double aortic arch and the right aortic arch with an aberrant left subclavian artery are the two most common vascular rings. CT is the imaging modality of choice for evaluating vascular airway compression and associated tracheomalacia [34 35 36]. In a double aortic arch, the right arch is commonly larger and higher than the left arch (Fig. 13). Less frequently, a distal portion of the smaller arch may be stenotic or atretic [37]. In an aberrant left subclavian artery, a PDA or ligamentum arteriosum completes the vascular ring. At its proximal portion, the Kommerell diverticulum may compress the adjacent airway and esophagus directly.

A persistent fifth aortic arch rarely originates from the distal ascending aorta, which is proximal and opposite to the ostium of the innominate artery and connects to the descending aorta. Typically, a persistent fifth aortic arch traverses under the normally developed fourth aortic arch, and these two arches show a superior-inferior relationship. Both arches may be patent (double-barreled or double-lumen aorta) or the superior arch may show atresia or interruption (Fig. 14) [38 39].

Rarely, pseudocoarctation demonstrates elongation and kinking of the aorta without a significant pressure gradient across the lesion or increased collateral circulation. Patients are usually asymptomatic, and they are followed up for a mild aortic coarctation. CT evaluation may be requested when significant flow acceleration is observed across this anomaly on Doppler echocardiography (Fig. 15) [40].

CT may be used for the morphologic evaluation of a PDA before ductal stent placement in patients with ductal-dependent CHDs, such as tetralogy of Fallot and functional single ventricle with severe pulmonary stenosis or atresia and hypoplastic left heart syndrome (Fig. 16) [41]. CT may be useful for a tortuous or complex PDA that is not clearly delineated on echocardiography [42]. Moreover, CT may be used to detect stent-related complications, such as branch pulmonary artery narrowing, aortic narrowing, stent migration, and in-stent thrombosis. Similarly, CT may be used to assess complications after ductal device closure, such as device embolization or protrusion into the adjacent vasculature. Virtual angiography may help visualize an occlusion device from the inside from the aortic and pulmonary sides [43]. Of note, beam-hardening artifacts caused by the ductal implants may make accurate CT assessment difficult.

The use of pediatric coronary CT angiography is increasing due to improvements in coronary artery visibility [44 45 46 47 48]. Because young children have high heart rates and a limited ability to

follow CT instructions, optimized imaging techniques are important to evaluate coronary arteries [6 49].

Compared with that in the general population, the prevalence of coronary artery anomalies is higher (approximately 5–10%) in patients with CHD, and most of them are closely related to surgical techniques used for specific CHD, most commonly tetralogy of Fallot and transposition of the great arteries [10 50]. The CT-based incidence of coronary artery anomalies associated with tetralogy of Fallot has been reported to be 8.5–12.7%; the proportion of significant anomalies crossing the anterior aspect of the right ventricular outflow tract was 55.6–62.3% (Fig. 17) [51 52]. CT showed a high diagnostic accuracy of 91.7% for identifying various patterns of coronary artery anatomy before arterial switch operation in transposition of the great arteries [53]. Ventriculo-coronary arterial communications are often observed in pulmonary atresia with an intact ventricular septum may lead to a right ventricle-dependent coronary circulation [54]. This fistulous connection can be identified on CT [55]. An anomalous origin of the left coronary artery from the pulmonary artery is a significant cause of acute heart failure in infants [56]. CT can be used to confirm the anomaly when echocardiographic findings are inconclusive [49]. The anomalous origin and course of the coronary arteries may lead to myocardial ischemia and sudden cardiac death, and a detailed morphologic spectrum of the anomaly can be accurately evaluated with CT for surgical planning [57]. Although an anomaly, such as high take-off coronary artery, may initially be hemodynamically minor, it may be considered as hemodynamically significant during cardiothoracic surgical or interventional procedures [10 58]. In addition, the coronary arteries may be compressed by adjacent anatomic structures or therapeutic implants.

The prevalence of a persistent left superior vena cava is higher in patients with CHD (up to 10%) than in the normal population (up to 0.5%) and much higher in patients with heterotaxy syndrome (up to 60–70%) [59]. A persistent left superior vena cava drains into the right atrium through a dilated coronary sinus. It may complicate catheter or pacemaker lead placement. Coronary sinus ostial atresia may be associated with a persistent left superior vena cava, and preoperative CT recognition of this rare anomaly is critically important in patients with a single functional ventricle [60]. The presence and size of a bridging vein between bilateral superior vena cavae should be noted on CT when planning a bidirectional cavopulmonary shunt.

A levoatriocardinal vein is a form of pulmonary systemic connection most commonly observed in patients with left heart obstructive lesions [61]. It is important to differentiate a levoatriocardinal vein from more commonly encountered mimics, such as a vertical vein in an anomalous pulmonary venous return, a persistent left superior vena cava, and a dilated left superior intercostal vein [61]. Although the variant has no functional importance, a subaortic left innominate or brachiocephalic vein needs to be distinguished from the pulmonary artery and persistent left superior vena cava on preoperative CT in patients with CHD [62].

Interrupted inferior vena cava with azygos continuation is frequently (83%) observed in patients with left atrial isomerism [63]. In these patients, pulmonary arteriovenous malformations may be detected on CT in a lung with a deficient hepatic venous flow [64]. Separate hepatic venous drainage is often associated with heterotaxy syndrome (more frequently in right atrial isomerism), and it typically makes Fontan completion technically more difficult [65].

Cardiothoracic CT may be used to identify and monitor catheter-related thrombosis or stenosis in central thoracic veins in patients with CHD (Fig. 18), as demonstrated using CT venography in adults [66].

The morphologic left or right ventricle can be determined based on the presence of the moderator band, the degree of the trabeculations, and the septal attachment of the atrioventricular valve on CT [67]. In addition, abnormal ventriculoarterial and atrioventricular connections, such as a double-outlet right ventricle, discordant connections, a double-inlet left ventricle, and twisted connections, can be assessed with CT [8 67]. Clinically meaningful obstruction of the left or right ventricular outflow tract should be evaluated on end-systolic phase CT showing a minimal

dimension [5 7]. In a double-chambered right ventricle, CT can reveal anomalous muscle bundles leading to the abnormal division of the right ventricle into a high-pressure inlet and a low-pressure outlet [68]. Pseudoaneurysm of the right ventricular outflow tract at the site of patch widening, as a rare postoperative complication, may be detected on CT after a total correction of tetralogy of Fallot [69]. A congenital cardiac ventricular diverticulum is rarely (3.4% for the left ventricle and 0.6% for the right ventricle) diagnosed using CT [70]. An apical diverticulum of the left ventricle and ectopia cordis may present in association with pentalogy of Cantrell [71]. Cardiothoracic CT may be used to characterize the type, size, number, and location of a ventricular septal defect [72]. In a double-outlet right ventricle, CT can show the intracardiac three-dimensional (3D) spatial relationships between a ventricular septal defect and the valves essential for surgical planning (Fig. 19) [73 74]. The transparent-lumen volume-rendering technique is useful for illustrating 3D morphologic features, especially the muscular type, of ventricular septal defects [75].

The degree of ventricular enlargement and hypertrophy resulting from volume and pressure loadings can be evaluated qualitatively on CT. Myocardial infarction may be demonstrated using myocardial delayed-enhancement CT imaging [76]. Furthermore, quantitative measures, including ventricular volume (Fig. 20A), myocardial mass (Fig. 20B), and myocardial strain, can be evaluated using CT in patients with CHD [77 78 79 80 81 82]. Based on a combined evaluation of CT-measured ventricular volume and mass, ventricular remodeling patterns, such as adaptation and maladaptation, may be assessed quantitatively [83].

Unlike in other cardiovascular structures, it is challenging to obtain homogeneous contrast enhancement in the right atrium [84 85]. Slow injection of a contrast agent with a generous delay may help obtain homogeneous right atrial enhancement.

An atrial septal defect is classified into four types, including the secundum, primum, sinus venosus, and coronary sinus types. The secundum is the most common type, whereas the coronary sinus type is the rarest. The sinus venosus type is further divided into superior and inferior subtypes depending on whether the defect is close to the superior vena cava or inferior vena cava, respectively (Fig. 8). The coronary sinus type or unroofed coronary sinus is commonly associated with a persistent left superior vena cava [86 87]. Echocardiography is the mainstay diagnostic modality for evaluating atrial septal defect, particularly in the secundum and primum defects, because the interatrial septum is thin, and it may not always be discernible on CT. CT is mainly useful for the sinus venosus type, and it facilitates the determination of the subtype and the associated PAPVR, which is useful for surgical planning (closing an atrial septal defect and rerouting anomalous pulmonary venous flow into the left atrium [LA]). PAPVR commonly associated with the sinus venosus type of atrial septal defects may be missed on echocardiography. CT is also useful for delineating the location and extent of the coronary sinus type [88].

Cor triatriatum sinister is characterized by the division of the LA into proximal and distal chambers by a fibromuscular membrane located superior to the left atrial appendage [89]. The atrial septal defect, PAPVR, and mitral valvular disease are commonly associated with this anomaly. CT can depict the size and numbers of fenestrations in the fibromuscular membrane and its associated anomalies (Fig. 21) [90]. Because of the similar morphological and clinical characteristics, this anomaly should be distinguished from the supramitral valve ring, in which the obstructive ring is located inferior to the left atrial appendage.

Cor triatriatum dexter is a rare anomaly in which the right atrium is divided into upstream and downstream chambers by a fibromuscular band. Imaging reveals a membrane coursing from the inferior vena cava to the superior vena cava, separating the right atrium into a posterior chamber (upstream), receiving vena caval flow and coronary sinus drainage, and an anterior chamber (downstream), containing the right atrial appendage and connecting the tricuspid valve [91].

CT is the imaging modality of choice for evaluating airway anomalies and abnormalities commonly associated with CHD [92]. Vascular airway compression is frequently observed before and after cardiac surgery. The left main bronchus is frequently compressed between the descending thoracic

aorta and a portion of the pulmonary artery and commonly associated with aortic arch repair [93]. The trachea is typically compressed by vascular rings and slings, including a double aortic arch (Fig. 14), right aortic arch with an aberrant left subclavian artery with or without Kommerell diverticulum, and pulmonary artery sling (Fig. 5) [94]. Tracheobronchomalacia, typically secondary to long-standing extrinsic vascular compression, can be detected with cine or four-dimensional CT [35 36 95]. Airway anomalies, such as tracheal bronchus [96], congenital tracheal stenosis (Fig. 5) [97], and bridging bronchus [22], are more frequently observed in patients with CHD, and they can be depicted accurately on CT. In heterotaxy syndrome, the anatomical relationship between the upper lobe bronchus and the pulmonary artery, which is the eparterial or hyparterial bronchus, on CT is useful for determining atrial situs [67 72].

CT can provide unprecedented lung imaging. In scimitar syndrome, various degrees of lung hypoplasia are associated with cardiac defects, most commonly ASD [98], and rarely the horseshoe lung, characterized by the fusion of the right and left lower lobes across the midline of the body [99]. Upper and lower airway obstructions frequently result in air trapping in young children with CHD. The extent of air trapping can be accurately identified on expiratory-phase CT imaging using mechanical or controlled ventilation with passive breath-holding at end-expiration [95 100] or the less invasive cine imaging technique during free breathing [101].

Heterotaxy syndrome is a complex anomaly presenting with various congenital defects, including CHD and abnormal organ positions. This syndrome has two major categories consisting of bilateral right-sidedness and bilateral left-sidedness; the latter is rarer than the former. Using segmental analysis on CT, various abnormalities observed as part of this syndrome can be characterized [102 103].

In addition to the thorax, we should include the abdomen in the scan range of initial cardiothoracic CT to assess abdominal situs when abdominal sonography is not performed and planned for this assessment. The abdominal situs is likely ambiguous if a midline gallbladder is identified, and a horizontal liver and no spleen constitute the rule, which is consistent with asplenia syndrome closely associated with right atrial isomerism. In contrast, patients with polysplenia syndrome or left atrial isomerism usually have multiple small spleens and interrupted inferior vena cava with azygos continuation.

The morphologic characteristics of the atrial appendages are keys to determining the atrial situs [67]. Bronchopulmonary morphology on CT can also be used to suggest atrial situs: bilateral short and trifurcated bronchi with three lung lobes in right atrial isomerism and bilateral long and bifurcated bronchi with two lung lobes in left atrial isomerism.

When a bridging vein between the bilateral superior vena cavae is small or absent, cavopulmonary connections should be performed on both sides. TAPVR is commonly associated with right atrial isomerism. In heterotaxy syndrome, a morphological right ventricle is usually dominant, whereas a morphological left ventricle is generally underdeveloped.

Most patients with heterotaxy syndrome end up with a staged single ventricular repair [104]. Before and after surgical or interventional treatment, the adequacy of pulmonary flow can be monitored and quantified using CT [105]. To avoid heterogeneous enhancement in a Fontan pathway mimicking vascular stenosis or thrombosis after Fontan operation, contrast enhancement protocols should be carefully optimized [84 106]. Moreover, CT can demonstrate systemic venous collaterals between the upper and lower body parts as well as the systemic arterial collaterals to the lungs, which indicates the elevation of the pulmonary arterial pressure often leading to Fontan failure [107]. Systemic to pulmonary venous collaterals leading to arterial desaturation may also be identified on CT (Fig. 22) [108]. However, cardiac MRI is more frequently used for the comprehensive evaluations of the Fontan pathway.

CT is not used as a primary diagnostic imaging method for evaluating cardiac valves mainly due to its insufficient temporal and spatial resolutions and high radiation exposure required for data acquisition throughout the entire cardiac cycle. Nevertheless, CT may be used for morphologic and functional assessments of cardiac valves in patients with CHD [109]. The number of the semilunar valve leaflets may be evaluated on CT. Valve thickening and doming with restricted opening on CT suggest the presence of valvular stenosis (Fig. 23). In valvular insufficiency, valve cusps fail to coapt during diastole. In muscular pulmonary atresia, the sizable infundibulum and the distance between the main pulmonary artery and the infundibulum can be accurately assessed with CT. CT may be used for the postsurgical comprehensive assessment of prosthetic heart valves [110]. In children, longitudinal coverage of multi-phase CT scanning should be minimized to reduce CT radiation dose (Fig. 24). CT can be used for evaluating commissural malalignment of the semilunar valves in patients with transposition of the great arteries frequently requiring modified coronary transfer techniques during an arterial switch operation [111].

The anatomic and functional assessment of the atrioventricular valves in CHD is primarily performed with echocardiography. A muscular type of tricuspid or mitral atresia can be observed on CT [7], but a membrane type of atrioventricular valve atresia may not be distinguished from the non-atretic atrioventricular valve on CT. In addition, detailed morphologic features of atrioventricular valve abnormalities in the atrioventricular septal defect cannot be evaluated with CT. In a hypoplastic ventricle, CT-based morphometric measures (i.e., diameter and area) of the small atrioventricular valve annulus may be used to plan an optimal surgical strategy [112]. The displaced and tethered tricuspid valve in Ebstein anomaly may be identified on CT [7 8]. Volumetric severity assessment using CT was recently reported to be feasible and useful for characterizing various phenotypes of this anomaly [113].

Cyanotic CHDs are commonly associated with small pulmonary artery size and flow. Therefore, a palliative procedure is performed to promote pulmonary arterial growth before corrective surgery. In this respect, a vascular shunt is commonly created for that purpose by connecting the aorta (a central shunt) or its branch (usually the subclavian artery; a modified Blalock–Taussig shunt) to the pulmonary artery. CT is useful for monitoring pulmonary arterial growth and evaluating potential complications, including shunt thrombosis or stenosis, calcified wall graft, seroma around the shunt, and retraction and stenosis of the pulmonary artery around the shunt insertion [114].

A conduit and a baffle are used for redirecting blood flow in complex CHD. A conduit (frequently containing a valve) is used for extracardiac rerouting of blood flow in outflow tract or semilunar valve obstruction, whereas a baffle is used for intracardiac flow rerouting [115]. A conduit connecting the right ventricle and the pulmonary artery, the so-called Rastelli procedure, is commonly used for tetralogy of Fallot with pulmonary atresia, transposition of the great artery, and severe left ventricular outflow tract obstruction. In addition to the evaluation of conduit patency and complications, CT may be used for evaluating retrosternal proximity of the conduit and close proximity of the conduit to the coronary artery to prevent serious vascular injury during redo surgery or interventional treatment (Fig. 25).

For evaluating an implanted vascular stent, CT is the imaging method of choice [116] because MRI is considerably limited in assessing vascular stents due to ferromagnetic metallic artifacts.

To achieve appropriate managements for patients with CHD, anatomical and hemodynamic information is necessary. Currently, CT can provide accurate anatomical information and some quantitative information quickly, which is useful for expeditious management decisions. Advanced 3D visualization techniques, such as augmented reality, virtual reality, and 3D printing, using 3D cardiothoracic CT data have been widely used in CHD for education, preprocedural planning and simulation, procedural guidance, and development of new insights in embryology and pathology of complex cardiac defects [75 117]. This comprehensive review includes a wide spectrum of contemporary organ-based clinical applications of pediatric cardiothoracic CT for different types of CHD (Table 1). The organ-based approach demonstrated in this review is more effective and more practical in evaluating CHD cases in which the primary diagnosis is typically made with

echocardiography. Pediatric cardiothoracic CT is typically requested to specify which organs need to be assessed. In this regard, CT provides complimentary but important information about cardiac and extracardiac structures, which cannot be clearly answered using echocardiography or catheter angiography.

Conflicts of Interest: The authors have no potential conflicts of interest to disclose.

Author Contributions: Conceptualization: Hyun Woo Goo, Suvipaporn Siripornpitak, Shyh-Jye Chen, Oktavia Lilyasari, Yu-Min Zhong, Haifa Abdul Latiff, Eriko Maeda.

Visualization: Hyun Woo Goo, Suvipaporn Siripornpitak, Shyh-Jye Chen, Oktavia Lilyasari, Yu-Min Zhong, Haifa Abdul Latiff.

Writing—original draft: Hyun Woo Goo, Suvipaporn Siripornpitak, Shyh-Jye Chen, Oktavia Lilyasari, Yu-Min Zhong, Haifa Abdul Latiff.

Writing—review & editing: Hyun Woo Goo, Eriko Maeda, Young Jin Kim, I-Chen Tsai, Dong Man Seo.

Yang JC Lin MT Jaw FS Chen SJ Wang JK Shih TT Trends in the utilization of computed tomography and cardiac catheterization among children with congenital heart disease J Formos Med Assoc 2015 114 1061 1068 25241602 10.1016/j.jfma.2014.08.004 PMC7126232 2 Lee T Tsai IC Fu YC Jan SL Wang CC Chang Y Using multidetector-row CT in neonates with complex congenital heart disease to replace diagnostic cardiac catheterization for anatomical investigation: initial experiences in technical and clinical feasibility Pediatr Radiol 2006 36 1273 1282 17036235 10.1007/s00247-006-0315-y 3 Hui PKT Goo HW Du J Ip JJK Kanzaki S Kim YJ Asian consortium on radiation dose of pediatric cardiac CT (ASCI-REDCARD) Pediatr Radiol 2017 47 899 910 28435986 10.1007/s00247-017-3847-4 4 Goo HW State-of-the-art CT imaging techniques for congenital heart disease Korean J Radiol 2010 11 4 18 20046490 10.3348/kjr.2010.11.1.4 PMC2799649 5 Goo HW Cardiac MDCT in children: CT technology overview and interpretation Radiol Clin North Am 2011 49 997 1010 21889018 10.1016/j.rcl.2011.06.001 6 Hong SH Goo HW Maeda E Choo KS Tsai IC Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group User-friendly vendor-specific guideline for pediatric cardiothoracic computed tomography provided by the Asian Society of Cardiovascular Imaging Congenital Heart Disease Study Group: part 1. Imaging techniques Korean J Radiol 2019 20 190 204 30672159 10.3348/kjr.2018.0571 PMC6342752 7 Goo HW Park IS Ko JK Kim YH Seo DM Yun TJ CT of congenital heart disease: normal anatomy and typical pathologic conditions Radiographics 2003 23 Spec No S147 S165 14557509 10.1148/rg.23si035501 8 Goo HW Park IS Ko JK Kim YH Seo DM Park JJ Computed tomography for the diagnosis of congenital heart disease in pediatric and adult patients Int J Cardiovasc Imaging 2005 21 347 365 16015453 10.1007/s10554-004-4015-0 9 Goo HW Current trends in cardiac CT in children Acta Radiol 2013 54 1055 1062 23104372 10.1258/ar.2012.120452 10 Goo HW Seo DM Yun TJ Park JJ Park IS Ko JK Coronary artery anomalies and clinically important anatomy in patients with congenital heart disease: multislice CT findings Pediatr Radiol 2009 39 265 273 19159923 10.1007/s00247-008-1111-7 11 Han BK Rigsby CK Hlavacek A Leipsic J Nicol ED Siegel MJ 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) J Cardiovasc Comput Tomogr 2015 9 475 492 26272851 10.1016/j.jcct.2015.07.004 12 Tsai IC Goo HW Cardiac CT and MRI for congenital heart disease in Asian countries: recent trends in publication based on a scientific database Int J Cardiovasc Imaging 2013 29 Suppl 1 1 5 23344910 10.1007/s10554-013-0184-z 13 Zucker EJ Cross-sectional imaging of congenital pulmonary artery anomalies Int J Cardiovasc Imaging 2019 35 1535 1548 31175525 10.1007/s10554-019-01643-4 14 Escalon JG Browne LP Bang TJ Restrepo CS Ocazonez D Vargas D Congenital anomalies of the pulmonary arteries: an imaging overview Br J Radiol 2019 92 20180185 30102560 10.1259/bjr.20180185 PMC6435093 15 Lapiere C Dubois J Rypens F Raboisson MJ Déry J Tetralogy of Fallot: preoperative assessment

with MR and CT imaging *Diagn Interv Imaging* 2016 97 531 541 26969119
10.1016/j.diii.2016.01.009 16 Goo HW Computed tomography pulmonary vascular volume ratio in children and young adults with congenital heart disease: the effect of cardiac phase *Pediatr Radiol* 2018 48 915 922 29572746 10.1007/s00247-018-4120-1 17 Meinel FG Huda W Schoepf UJ Rao AG Cho YJ Baker GH Diagnostic accuracy of CT angiography in infants with tetralogy of Fallot with pulmonary atresia and major aortopulmonary collateral arteries *J Cardiovasc Comput Tomogr* 2013 7 367 375 24331932 10.1016/j.jcct.2013.11.001 18 Lin MT Wang JK Chen YS Lee WJ Chiu HH Chen CA Detection of pulmonary arterial morphology in tetralogy of Fallot with pulmonary atresia by computed tomography: 12 years of experience *Eur J Pediatr* 2012 71 579 586
10.1007/s00431-011-1621-4 22083156 19 Koplay M Cimen D Sivri M Güvenc O Arslan D Nayman A Truncus arteriosus: diagnosis with dual-source computed tomography angiography and low radiation dose *World J Radiol* 2014 6 886 889 25431644 10.4329/wjr.v6.i11.886 PMC4241496 20 Yakut K Tokel NK Özkan M Varan B Erdoğlan A Şılamacı S Diagnosis and surgical treatment of aortopulmonary window: our single-center experience *Turk Gogus Kalp Damar Cerrahisi Derg* 2018 26 30 37 32082708 10.5606/tgkdc.dergisi.2018.14772 PMC7018111 21 Zhong YM Jaffe RB Liu JF Sun AM Gao W Wang Q Multi-slice computed tomography assessment of bronchial compression with absent pulmonary valve *Pediatr Radiol* 2014 44 803 809 24706163
10.1007/s00247-014-2898-z PMC4061480 22 Zhong YM Jaffe RB Zhu M Gao W Sun AM Wang Q CT assessment of tracheobronchial anomaly in left pulmonary artery sling *Pediatr Radiol* 2010 40 1755 1762 20490486 10.1007/s00247-010-1682-y 23 Dillman JR Yarram SG Hernandez RJ Imaging of pulmonary venous developmental anomalies *AJR Am J Roentgenol* 2009 192 1272 1285 19380552 10.2214/AJR.08.1526 24 Pandey NN Sharma A Jagia P Imaging of anomalous pulmonary venous connections by multidetector CT angiography using third-generation dual source CT scanner *Br J Radiol* 2018 91 20180298 30004806 10.1259/bjr.20180298 PMC6319858 25 Ou P Marini D Celermajer DS Agnoletti G Vouhé P Sidi D Non-invasive assessment of congenital pulmonary vein stenosis in children using cardiac-non-gated CT with 64-slice technology *Eur J Radiol* 2009 70 595 599 18353590 10.1016/j.ejrad.2008.02.006 26 Goo HW Park SH Koo HJ Cho YH Lee E Atresia of the bilateral pulmonary veins: a rare and dismal anomaly identified on cardiac CT *Pediatr Radiol* 2014 44 942 947 24535116 10.1007/s00247-014-2900-9 27 Hanneman K Newman B Chan F Congenital variants and anomalies of the aortic arch *Radiographics* 2017 37 32 51 27860551 10.1148/rg.2017160033 28 Liu PS St John Sutton MG Litt HI Diffuse supravalvular aortic stenosis: comprehensive imaging with ECG-gated CT angiography *Int J Cardiovasc Imaging* 2007 23 269 272 16821118 10.1007/s10554-006-9126-3 29 Das KM Momenah TS Larsson SG Jadoon S Aldosary AS Lee EY Williams-Beuren syndrome: computed tomography imaging review *Pediatr Cardiol* 2014 35 1309 1320 25139247 10.1007/s00246-014-0998-z 30 Shin HJ Jhang WK Park JJ Goo HW Seo DM Modified simple sliding aortoplasty for preserving the sinotubular junction without using foreign material for congenital supravalvular aortic stenosis *Eur J Cardiothorac Surg* 2011 40 598 602 21345696 10.1016/j.ejcts.2010.12.044 31 Karaosmanoglu AD Khawaja RD Onur MR Kalra MK CT and MRI of aortic coarctation: pre- and postsurgical findings *AJR Am J Roentgenol* 2015 204 W224 W233 25714305 10.2214/AJR.14.12529 32 Yang DH Goo HW Seo DM Yun TJ Park JJ Park IS Multislice CT angiography of interrupted aortic arch *Pediatr Radiol* 2008 38 89 100 17965856 10.1007/s00247-007-0662-3 33 Tola H Ozturk E Yildiz O Ereke E Haydin S Turkvatan A Assessment of children with vascular ring *Pediatr Int* 2017 59 134 140 27454661 10.1111/ped.13101 34 Lee EY Zurakowski D Waltz DA Mason KP Riaz F Ralph A MDCT evaluation of the prevalence of tracheomalacia in children with mediastinal aortic vascular anomalies *J Thorac Imaging* 2008 23 258 265 19204470 10.1097/RTI.0b013e31817fbbdf 35 Goo HW Free-breathing cine CT for the diagnosis of tracheomalacia in young children *Pediatr Radiol* 2013 43 922 928 23417231 10.1007/s00247-013-2637-x 36 Goo HW Four-dimensional thoracic CT in free-breathing children *Korean J Radiol* 2019 20 50 57 30627021 10.3348/kjr.2018.0325 PMC6315071 37 Schlesinger AE Krishnamurthy R Sena LM Guillerman RP Chung T DiBardino DJ Incomplete double aortic arch with atresia of the distal left arch: distinctive imaging appearance *AJR Am J Roentgenol* 2005 184 1634 1639 15855130 10.2214/ajr.184.5.01841634 38 Bernasconi A Goo HW Yoo SJ Double-barrelled aorta with tetralogy of Fallot and pulmonary atresia *Cardiol Young* 2007 17 98 101 17184561 10.1017/S104795110600148X 39 Naimo PS Vazquez-Alvarez Mdel C d'Udekem Y Jones B Konstantinov IE Double-lumen aortic arch: persistence of the fifth aortic arch *Ann Thorac Surg* 2016 101 e155 e156 27106464 10.1016/j.athoracsur.2015.10.014 40 Thodi Ramamurthy M Balakrishnan VK David SAN Korrapati HS Congenital kinking of aorta *BMJ*

Case Rep 2017 2017 bcr2017220896 10.1136/bcr-2017-220896 PMC5535190 28705800 41

Jadhav SP Aggarwal V Masand PM Diaz E Zhang W Qureshi AM Correlation of ductus arteriosus length and morphology between computed tomographic angiography and catheter angiography and their relation to ductal stent length *Pediatr Radiol* 2020 50 800 809 32170350

10.1007/s00247-020-04624-1 42 Rehman R Marhisham MC Alwi M Stenting the complex patent ductus arteriosus in tetralogy of Fallot with pulmonary atresia: challenges and outcomes *Future Cardiol* 2018 14 55 73 29199861 10.2217/fca-2017-0053 43 Hayabuchi Y Mori K Kagami S Virtual endoscopy using multidetector-row CT for coil occlusion of patent ductus arteriosus *Catheter Cardiovasc Interv* 2007 70 434 439 17380493 10.1002/ccd.21108 44 Goo HW Yang DH Coronary artery visibility in free-breathing young children with congenital heart disease on cardiac 64-slice CT: dual-source ECG-triggered sequential scan vs. single-source non-ECG-synchronized spiral scan *Pediatr Radiol* 2010 40 1670 1680 20464385 10.1007/s00247-010-1693-8 45 Paul JF Rohnean A Elfassy E Sigal-Cinqualbre A Radiation dose for thoracic and coronary step-and-shoot CT using a 128-slice dual-source machine in infants and small children with congenital heart disease *Pediatr Radiol* 2011 41 244 249 20821005 10.1007/s00247-010-1804-6 46 Kanie Y Sato S Tada A Kanazawa S Image quality of coronary arteries on non-electrocardiography-gated high-pitch dual-source computed tomography in children with congenital heart disease *Pediatr Cardiol* 2017 38 1393 1399 28689328 10.1007/s00246-017-1675-9 47 Barrera CA Otero HJ White AM Saul D Biko DM Depiction of the native coronary arteries during ECG-triggered high-pitch dual-source coronary computed tomography angiography in children: determinants of image quality *Clin Imaging* 2018 52 240 245 30142610 10.1016/j.clinimag.2018.08.013 48 Goo HW Image quality and radiation dose of high-pitch dual-source spiral cardiothoracic computed tomography in young children with congenital heart disease: comparison of non-electrocardiography synchronization and prospective electrocardiography triggering *Korean J Radiol* 2018 19 1031 1041 30386135 10.3348/kjr.2018.19.6.1031 PMC6201980 49 Goo HW Coronary artery imaging in children *Korean J Radiol* 2015 16 239 250 25741188 10.3348/kjr.2015.16.2.239 PMC4347262 50 Lowry AW Olabiyyi OO Adachi I Moodie DS Knudson JD Coronary artery anatomy in congenital heart disease *Congenit Heart Dis* 2013 8 187 202 23601059 10.1111/chd.12067 51 Kervancioglu M Tokel K Varan B Yildirim SV Frequency, origins and courses of anomalous coronary arteries in 607 Turkish children with tetralogy of Fallot *Cardiol J* 2011 18 546 551 21947991 10.5603/cj.2011.0011 52 Goo HW Coronary artery anomalies on preoperative cardiac CT in children with tetralogy of Fallot or Fallot type of double outlet right ventricle: comparison with surgical findings *Int J Cardiovasc Imaging* 2018 34 1997 2009 30051301 10.1007/s10554-018-1422-1 53 Goo HW Identification of coronary artery anatomy on dual-source cardiac computed tomography before arterial switch operation in newborns and young infants: comparison with transthoracic echocardiography *Pediatr Radiol* 2018 48 176 185 29032431 10.1007/s00247-017-4004-9 54 Giglia TM Mandell VS Connor AR Mayer JE Jr Lock JE Diagnosis and management of right ventricle-dependent coronary circulation in pulmonary atresia with intact ventricular septum *Circulation* 1992 86 1516 1528 1423965 10.1161/01.cir.86.5.1516 55 Séguéla PE Houyel L Loget P Piot JD Paul JF Critical stenosis of a right ventricle to coronary artery fistula seen at dual-source CT in a newborn with pulmonary atresia and intact ventricular septum *Pediatr Radiol* 2011 41 1069 1072 21487673 10.1007/s00247-011-2068-5 56 Peña E Nguyen ET Merchant N Dennie C ALCAPA syndrome: not just a pediatric disease *Radiographics* 2009 29 553 565 19325065 10.1148/rg.292085059 57 Pandey NN Sinha M Sharma A Rajagopal R Bhambri K Kumar S Anomalies of coronary artery origin: evaluation on multidetector CT angiography *Clin Imaging* 2019 57 87 98 31170598 10.1016/j.clinimag.2019.05.010 58 Tsai WL Wei HJ Tsai IC High-take-off coronary artery: a haemodynamically minor, but surgically important coronary anomaly *Pediatr Radiol* 2010 40 232 233 19924411 10.1007/s00247-009-1413-4 59 Batouty NM Sobh DM Gadelhak B Sobh HM Mahmoud W Tawfik AM Left superior vena cava: cross-sectional imaging overview *Radiol Med* 2020 125 237 246 31823296 10.1007/s11547-019-01114-9 60 Kim C Goo HW Yu JJ Yun TJ Coronary sinus ostial atresia with persistent left superior vena cava demonstrated on cardiac CT in an infant with a functional single ventricle *Pediatr Radiol* 2012 42 761 763 21993993 10.1007/s00247-011-2258-1 61 Agarwal PP Mahani MG Lu JC Dorfman AL Levoatriocardinal vein and mimics: spectrum of imaging findings *AJR Am J Roentgenol* 2015 205 W162 W171 26204303 10.2214/AJR.15.14365 62 Takada Y Narimatsu A Kohno A Kawai C Hara H Harasawa A Anomalous left brachiocephalic vein: CT findings *J Comput Assist Tomogr* 1992 16 893 896 1430437 10.1097/00004728-199211000-00012 63 Ugas Charcape CF Alpaca Rodriguez LR Matos

Rojas IA Lazarte Rantes CI Valdez Quintana M Katekaru Tokeshi DA Characterisation of computed tomography angiography findings in paediatric patients with heterotaxy *Pediatr Radiol* 2019 49 1142 1151 31165901 10.1007/s00247-019-04434-0 64 Montesa C Karamlou T Ratnayaka K Pophal SG Ryan J Nigro JJ Hepatic vein incorporation into the azygos system in heterotaxy and interrupted inferior vena cava *World J Pediatr Congenit Heart Surg* 2019 10 330 337 31084312 10.1177/2150135119842869 65 Nakata T Fujimoto Y Hirose K Osaki M Tosaka Y Ide Y Fontan completion in patients with atrial isomerism and separate hepatic venous drainage *Eur J Cardiothorac Surg* 2010 37 1264 1270 20138532 10.1016/j.ejcts.2009.12.026 66 Kim H Chung JW Park JH Yin YH Park SH Yoon CJ CT venography in the diagnosis and treatment of benign thoracic central venous obstruction *Korean J Radiol* 2003 4 146 152 14530642 10.3348/kjr.2003.4.3.146 PMC2698080 67 Goo HW Chapter 3. CT in pediatric heart disease Saremi F Achenbach S Arbustini E Narula J Revisiting cardiac anatomy: a computed-tomography-based atlas and reference Oxford Wiley-Blackwell 2011 76 84 68 Truong QA Yared K Maurovich-Horvat P Siegel E Cubeddu RJ King ME Images in cardiovascular medicine. Double-chambered right ventricle and situs inversus with dextrocardia *Circulation* 2010 121 e229 e232 20212287 10.1161/CIR.0b013e3181d56ebd PMC2990775 69 Inoue Y Igawa O Iitsuka K Aneurysm of the right ventricular outflow tract after surgical repair of Tetralogy of Fallot: three-dimensional computed tomography findings *Europace* 2009 11 130 18829702 10.1093/europace/eun260 70 Nakazono T Jeudy J White CS Left and right ventricular diverticula: incidence and imaging findings on 256-slice multidetector computed tomography *J Thorac Imaging* 2012 27 179 183 21516044 10.1097/RTI.0b013e31821255ae 71 Ojha V Chandrashekhara SH Ganga KP Saxena A Gulati G Congenital left ventricular diverticulum in pentalogy of cantrell: puzzle solved with dual-source CT *Ann Thorac Surg* 2019 108 e205 30922822 10.1016/j.athoracsur.2019.02.050 72 Nakagawa M Hara M Sakurai K Ohashi K Asano M Shibamoto Y Usefulness of electrocardiography-gated dual-source computed tomography for evaluating morphological features of the ventricles in children with complex congenital heart defects *Jpn J Radiol* 2011 29 540 546 21927995 10.1007/s11604-011-0594-6 73 Dydynski PB Kiper C Kozik D Keller BB Austin E Holland B Three-dimensional reconstruction of intracardiac anatomy using CTA and surgical planning for double outlet right ventricle: early experience at a tertiary care congenital heart center *World J Pediatr Congenit Heart Surg* 2016 7 467 474 27358302 10.1177/2150135116651399 74 Yim D Dragulescu A Ide H Seed M Grosse-Wortmann L van Arsdell G Essential modifiers of double outlet right ventricle: revisit with endocardial surface images and 3-dimensional print models *Circ Cardiovasc Imaging* 2018 11 e006891 29855425 10.1161/CIRCIMAGING.117.006891 75 Goo HW Park SJ Yoo SJ Advanced medical use of three-dimensional imaging in congenital heart disease: augmented reality, mixed reality, virtual reality, and three-dimensional printing *Korean J Radiol* 2020 21 133 145 31997589 10.3348/kjr.2019.0625 PMC6992436 76 Goo HW Myocardial delayed-enhancement CT: initial experience in children and young adults *Pediatr Radiol* 2017 47 1452 1462 28534155 10.1007/s00247-017-3889-7 77 Han BK Hlavacek AM Kay WA Pham TDN Grant K Garberich RF Multi-institutional evaluation of the indications and radiation dose of functional cardiovascular computed tomography (CCT) imaging in congenital heart disease *Int J Cardiovasc Imaging* 2016 32 339 346 26474570 10.1007/s10554-015-0775-y 78 Goo HW Serial changes in anatomy and ventricular function on dual-source cardiac computed tomography after the Norwood procedure for hypoplastic left heart syndrome *Pediatr Radiol* 2017 47 1776 1786 28879411 10.1007/s00247-017-3972-0 79 Goo HW Changes in right ventricular volume, volume load, and function measured with cardiac computed tomography over the entire time course of tetralogy of Fallot *Korean J Radiol* 2019 20 956 966 31132821 10.3348/kjr.2018.0891 PMC6536786 80 Goo HW Quantification of initial right ventricular dimensions by computed tomography in infants with congenital heart disease and a hypoplastic right ventricle *Korean J Radiol* 2020 21 203 209 31997595 10.3348/kjr.2019.0662 PMC6992442 81 Goo HW Technical feasibility of semiautomatic three-dimensional threshold-based cardiac computed tomography quantification of left ventricular mass *Pediatr Radiol* 2019 49 318 326 30470863 10.1007/s00247-018-4303-9 82 Buss SJ Schulz F Mereles D Hosch W Galuscky C Schummers G Quantitative analysis of left ventricular strain using cardiac computed tomography *Eur J Radiol* 2014 83 e123 e130 24368011 10.1016/j.ejrad.2013.11.026 83 Goo HW Park SH Pattern analysis of left ventricular remodeling using cardiac computed tomography in children with congenital heart disease: preliminary results *Korean J Radiol* 2020 21 717 725 32410410 10.3348/kjr.2019.0689 PMC7231616 84 Malik SB Kwan D Shah AB Hsu JY The right atrium: gateway to the

heart--anatomic and pathologic imaging findings Radiographics 2015 35 14 31 25590385 10.1148/rg.351130010 85 Goo HW Haemodynamic findings on cardiac CT in children with congenital heart disease Pediatr Radiol 2011 41 250 261 21127855 10.1007/s00247-010-1886-1 86 Saremi F Cardiac shunts: ASD, VSD, PDA Saremi F Cardiac CT and MR for adult congenital heart disease 1st ed New York Springer 2014 305 345 87 Yasunaga D Hamon M MDCT of interatrial septum Diagn Interv Imaging 2015 96 891 899 25981979 10.1016/j.diii.2015.02.011 88 Sun L Juan YH Chen J Zhuang J Xie J Li H Evaluation of unroofed coronary sinus syndrome using cardiovascular CT angiography: an observational study AJR Am J Roentgenol 2018 211 314 320 29949414 10.2214/AJR.17.19128 89 Malik A Fram D Mohani A Fischerkeller M Yekta A Mohyuddin Y Cor triatriatum: a multimodality imaging approach Can J Cardiol 2008 24 e19 e20 18340384 10.1016/s0828-282x(08)70593-2 PMC2649642 90 Thakrar A Shapiro MD Jassal DS Neilan TG King ME Abbbara S Cor triatriatum: the utility of cardiovascular imaging Can J Cardiol 2007 23 143 145 17311121 10.1016/s0828-282x(07)70735-3 PMC2650651 91 Sánchez-Brotons JA López-Pardo FJ Rodríguez-Puras MJ López-Haldón JE Cor triatriatum dexter in adults Rev Esp Cardiol 2010 63 998 999 20738947 10.1016/s1885-5857(10)70196-1 92 Goo HW Evaluation of the airways in patients with congenital heart disease using multislice CT J Korean Pediatr Cardiol Soc 2004 8 37 43 93 Jhang WK Park JJ Seo DM Goo HW Gwak M Perioperative evaluation of airways in patients with arch obstruction and intracardiac defects Ann Thorac Surg 2008 85 1753 1758 18442579 10.1016/j.athoracsur.2008.01.059 94 Etesami M Ashwath R Kanne J Gilkeson RC Rajiah P Computed tomography in the evaluation of vascular rings and slings Insights Imaging 2014 5 507 521 25008430 10.1007/s13244-014-0343-3 PMC4141344 95 Lee EY Zucker EJ Restrepo R Daltro P Boisselle PM Advanced large airway CT imaging in children: evolution from axial to 4-D assessment Pediatr Radiol 2013 43 285 297 23417254 10.1007/s00247-012-2470-7 96 Ming Z Lin Z Evaluation of tracheal bronchus in Chinese children using multidetector CT Pediatr Radiol 2007 37 1230 1234 17909778 10.1007/s00247-007-0636-5 97 Yamoto M Fukumoto K Sekioka A Iwazaki T Sano K Takahashi T Non-operative management of congenital tracheal stenosis: criteria by computed tomography Pediatr Surg Int 2019 35 1123 1130 31410561 10.1007/s00383-019-04532-y 98 Masrani A McWilliams S Bhalla S Woodard PK Anatomical associations and radiological characteristics of Scimitar syndrome on CT and MR J Cardiovasc Comput Tomogr 2018 12 286 289 29550261 10.1016/j.jcct.2018.02.001 99 Goo HW Kim YH Ko JK Park IS Yoon CH Horseshoe lung: useful angiographic and bronchographic images using multidetector-row spiral CT in two infants Pediatr Radiol 2002 32 529 532 12107588 10.1007/s00247-002-0705-8 100 Long FR Castile RG Technique and clinical applications of full-inflation and end-exhalation controlled-ventilation chest CT in infants and young children Pediatr Radiol 2001 31 413 422 11436888 10.1007/s002470100462 101 Goo HW Kim HJ Detection of air trapping on inspiratory and expiratory phase images obtained by 0.3-second cine CT in the lungs of free-breathing young children AJR Am J Roentgenol 2006 187 1019 1023 16985151 10.2214/AJR.05.0895 102 Chen SJ Li YW Wang JK Wu MH Chiu IS Chang CI Usefulness of electron beam computed tomography in children with heterotaxy syndrome Am J Cardiol 1998 81 188 194 9591903 10.1016/s0002-9149(97)00879-5 103 Wolla CD Hlavacek AM Schoepf UJ Bucher AM Chowdhury S Cardiovascular manifestations of heterotaxy and related situs abnormalities assessed with CT angiography J Cardiovasc Comput Tomogr 2013 7 408 416 24331937 10.1016/j.jcct.2013.11.008 PMC3947807 104 Makhija Z Marwah A Mishra S Kumar J Goel A Sharma R Biventricular repair in heterotaxy patients World J Pediatr Congenit Heart Surg 2015 6 195 202 25870336 10.1177/2150135114563772 105 Chen BB Chen SJ Wu MH Li YW Lue HC EBCT-McGoon ratio a reliable and useful method to predict pulmonary blood flow non-invasively Chinese J Radiol 2007 32 1 8 106 Park EA Lee W Chung SY Yin YH Chung JW Park JH Optimal scan timing and intravenous route for contrast-enhanced computed tomography in patients after Fontan operation J Comput Assist Tomogr 2010 34 75 81 20118726 10.1097/RCT.0b013e3181ae292c 107 Egbe AC Reddy YNV Khan AR Al-Otaibi M Akintoye E Obokata M Venous congestion and pulmonary vascular function in Fontan circulation: implications for prognosis and treatment Int J Cardiol 2018 271 312 316 30223363 10.1016/j.ijcard.2018.05.039 PMC6146405 108 Lluri G Levi DS Aboulhosen J Systemic to pulmonary venous collaterals in adults with single ventricle physiology after cavopulmonary palliation Int J Cardiol 2015 189 159 163 25897896 10.1016/j.ijcard.2015.04.065 109 Litmanovich DE Kirsch J Computed tomography of cardiac valves: review Radiol Clin North Am 2019 57 141 164 30454810 10.1016/j.rcl.2018.08.011 110 Faure ME Swart LE Dijkshoorn ML Bekkers JA van Straten M Nieman K Advanced CT

acquisition protocol with a third-generation dual-source CT scanner and iterative reconstruction technique for comprehensive prosthetic heart valve assessment *Eur Radiol* 2018 28 2159 2168 29234912 10.1007/s00330-017-5163-7 PMC5882630 111 Bang JH Park JJ Goo HW Evaluation of commissural malalignment of aortic-pulmonary sinus using cardiac CT for arterial switch operation: comparison with transthoracic echocardiography *Pediatr Radiol* 2017 47 556 564 28243677 10.1007/s00247-017-3786-0 112 Goo HW Park SH Computed tomography-based ventricular volumes and morphometric parameters for deciding the treatment strategy in children with a hypoplastic left ventricle: preliminary results *Korean J Radiol* 2018 19 1042 1052 30386136 10.3348/kjr.2018.19.6.1042 PMC6201981 113 Goo HW Volumetric severity assessment of Ebstein anomaly using three-dimensional cardiac CT: a feasibility study *Cardiovasc Imaging Asia* 2019 3 61 67 114 Tomasian A Malik S Shamsa K Krishnam MS Congenital heart diseases: post-operative appearance on multi-detector CT-a pictorial essay *Eur Radiol* 2009 19 2941 2949 19513718 10.1007/s00330-009-1474-7 PMC2778768 115 Beerbaum P Valverde I Greil GF Babu-Narayan SV Chapter 17. Baffles and conduits Fogel MA Principles and practice of cardiac magnetic resonance in congenital heart disease 1st ed New Jersey Wiley-Blackwell 2010 316 344 116 Eichhorn JG Jourdan C Hill SL Raman SV Cheatham JP Long FR CT of pediatric vascular stents used to treat congenital heart disease *AJR Am J Roentgenol* 2008 190 1241 1246 18430838 10.2214/AJR.07.3194 117 Yoo SJ Saito M Hussein N Golding F Goo HW Lee W Systematic approach to malalignment type ventricular septal defects *J Am Heart Assoc* 2020 9 e018275 33170057 10.1161/JAHA.120.018275 PMC7763733 Fig. 1 Pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries. A. Merged frontal volume-rendered CT image obtained in a 9-month-old girl illustrates color-coded pulmonary artery mapping including two major collateral arteries (red and green) originating from the DA and the central pulmonary arteries (blue) supplied by a 4-mm central shunt (arrow). B. Posterior volume-rendered CT image obtained in a 6-month-old boy shows a connection (asterisk) between a larger aortopulmonary collateral artery (arrows) and the right central pulmonary artery (R). The pulmonary arteries appear dilated and tortuous, suggesting increased pulmonary arterial pressure. C. Oblique coronal reformatted CT image obtained in a 6-day-old boy shows two focal stenoses (arrows) at the proximal portion of one major aortopulmonary collateral artery. AA = ascending aorta, DA = descending aorta

A. In an 11-year-old girl, color-coded volume-rendered CT image with oblique posterior view shows truncus arteriosus type 1, in which the pulmonary trunk (blue) arises from the left lateral aspect (arrows) of the common arterial trunk (red) and divides into the right and left pulmonary arteries (blue). B. In a 1-year-old girl, color-coded volume-rendered CT image with inferior view demonstrates truncus arteriosus type 2, in which the right and left pulmonary arteries (blue) originate separately but close together directly from the posterior aspect (arrows) of the common arterial trunk (red).

Color-coded volume-rendered CT image with superior view demonstrates a small communication (white) between the ascending aorta (red) and the pulmonary trunk (blue) at their distal portion.

A. Axial CT image with maximum intensity projection demonstrates the dilated right (R) and left (L) pulmonary arteries. B. Coronal CT image with minimum intensity projection shows central airway narrowings, most severely involving the left main bronchus (arrow) compressed by the dilated proximal branch pulmonary arteries. Hyperlucent lung areas due to air trapping are also noted.

Color-coded volume-rendered CT image with posterior view demonstrates left pulmonary artery sling (asterisks) and congenital tracheal stenosis (white).

Volume-rendered CT image with posterior view demonstrates that the LUPV anomalously drains into the innominate vein, whereas the other pulmonary veins are normally connected to the left atrium. Notably, the LLPV is diffusely small. LLPV = left lower pulmonary vein, LUPV = left upper pulmonary vein, RLPV = right lower pulmonary vein, RUPV = right upper pulmonary vein

Volume-rendered CT image with posterior view reveals that all pulmonary veins anomalously drain into the dilated coronary sinus (asterisk).

Transverse volume-rendered CT image shows that the RUPV anomalously drains into the RA, whereas the LLPV normally drains into the LA. A sinus venosus atrial septal defect (asterisk) is also seen. LA = left atrium, LLPV = left lower pulmonary vein, LV = left ventricle, RA = right atrium, RUPV = right lower pulmonary vein, RV = right ventricle

A, B. Oblique coronal (A) and oblique axial (B) CT images reveal focal stenosis (arrows) of the left pulmonary veins at the atrio-pulmonary venous junctions. LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle

Volume-rendered CT image demonstrates discrete supra-ventricular aortic stenosis (arrows) and the dilated and tortuous left coronary artery. In contrast, the right coronary artery is barely seen (arrowheads).

Volume-rendered CT image with lateral view shows a discrete juxtaductal narrowing (arrow) and the mildly hypoplastic isthmus. Post-stenotic dilatation of the proximal descending aorta and dilated collateral arteries around the aortic lesion are noted.

Volume-rendered CT image with oblique sagittal view performed 1 year after angioplasty demonstrates a pseudoaneurysm (arrow).

Superior volume-rendered CT image shows double aortic arch with a dominant right arch. A bronchoscopy demonstrates tracheomalacia with abnormal vascular pulsation from the posterior wall.

Volume-rendered CT image reveals coarctation (arrow) of a persistent fifth aortic arch and a type A interrupted fourth aortic arch.

A series of sagittal volume-rendered CT images of the aorta show the tortuous, elongated distal aortic arch with mild narrowing, which is consistent with pseudocoarctation. Cardiac catheterization confirmed no significant pressure gradients across the narrowing.

A. Oblique coronal volume-rendered CT image obtained from a 28-day-old boy with tricuspid atresia and pulmonary atresia, shows a patent ductus arteriosus (arrow) originating from the distal aortic arch immediately distal to the left subclavian artery origin. B. Oblique coronal volume-rendered CT image obtained from a 7-day-old boy with pulmonary atresia and atrioventricular septal defect demonstrates a patent ductus arteriosus (arrow) arising from the proximal aortic arch opposite to the left common carotid artery origin. Various origins and courses of patent ductus arteriosus reflect the effect of ductal flow during fetal life on the development of ductal tissue. Accurate evaluation of morphologic features depicted on CT imaging helps plan ductal stent placement.

Volume-rendered CT image shows a left anterior descending artery (arrows) crossing the right ventricular outflow tract.

Coronal CT image reveals a focal stenosis (long arrow) at the junction between the superior vena cava and the RA. A contrast jet through the stenosis is seen in the RA. Undiluted contrast agent is opacified in the left pericardiophrenic vein (short arrows), as a collateral vein, from the left brachiocephalic vein. A = ascending aorta, LV = left ventricle, RA = right atrium

Oblique coronal transparent-lumen volume-rendered CT image with frontal cut demonstrates that a ventricular septal defect (asterisk), committed to the aortic valve and the outlet septum (arrow), is fused to the left margin of the ventricular septal defect. A short muscular subaortic stenosis is noted. A = ascending aorta, P = pulmonary trunk, RA = right atrium, RV = right ventricle, VIF = ventriculoinfundibular fold

A, B. Short-axis CT images obtained at the end-diastolic phase and the mid-ventricular level show the mildly enlarged RV (98.6 mL/m² of body surface area) with mild right ventricular hypertrophy (25.7 g/m² of body surface area). The right ventricular cavity (pink in A) and right ventricular myocardial mass (purple in B) were segmented using a three-dimensional threshold-based approach. LV = left ventricle, RV = right ventricle

Axial CT image shows a membrane (arrows) dividing the left atrium into a PC and a DC. The pulmonary veins are connected to the PC and the left atrial appendage is connected to the DC. DC = distal chamber, PC = proximal chamber

Volume-rendered CT image with oblique posterior view shows dilated systemic venous collaterals (blue) draining into the LA directly or via the pulmonary veins (arrows). After the coil embolization of these systemic to pulmonary venous collaterals, oxygen saturation was increased. The patient has dextrocardia and a right aortic arch. LA = left atrium

A, B. Oblique axial (A) and oblique sagittal (B) CT images show thickening and doming of the pulmonary valve leaflets (arrows) and restricted valve opening (asterisks) during systole.

A, B. Oblique coronal diastolic (A) and systolic (B) unenhanced CT images obtained with a minimal longitudinal scan range demonstrate a bi-leaflet prosthetic pulmonary valve (arrows) with the valve in closed (A) and open (B) position. Two leaflets show severely limited opening during systole (B) , which is more severe on the right side. Based on the CT finding, urokinase was administered for 72 hours to treat the prosthetic valve malfunction.

A. Sagittal CT image shows extensive calcific degeneration of an RV-PA conduit (asterisks). Pacing wire is noted (arrows). B. Short-axis CT image shows proximity of the left coronary artery (arrow) and the conduit. PA = pulmonary artery, RV = right ventricle

Typically, the essential evaluations are almost always included in every cardiothoracic CT examination and can be promptly completed, while the optional evaluations are performed as needed and take more time and effort.