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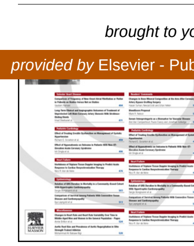

REVIEW

Computed tomography imaging in children with congenital heart disease: Indications and radiation dose optimization

CT dans les cardiopathies congénitales des enfants : indications et optimisation des doses

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Summary Computed tomography (CT) technology is acquiring a key role in the diagnostic process of complex cardiac congenital anomalies. Recent advances and improvements in spatial and temporal resolution and radiation dose are encouraging the use of CT scanning in children. Paediatric cardiologists should have a good knowledge of the potential of CT techniques and their limitations to plan and properly perform CT examinations without forgetting radiation concerns. In this paper, we will discuss the principal indications for CT scans in newborns and children in our clinical practice. We will also outline the most-used strategies for dose reduction.

Abbreviations: 3D, Three-dimensional; ACAOS, Anomalous origin of coronary artery from opposite sinus of Valsalva; ALCAPA, Anomalous origin of left coronary artery from pulmonary artery; ARCAPA, Anomalous origin of right coronary artery from pulmonary artery; CHD, Congenital heart disease; CT, Computed tomography; ECG, Electrocardiogram; kV, Kilovoltage; MAPCAS, Major aortopulmonary collateral arteries; MRI, Magnetic resonance imaging; TTE, Transthoracic echocardiography.

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Basic knowledge about the various CT techniques is crucial, not only to perform, but also to interpret CT results, thus helping the medical and surgical management of patients.

MOTS CLÉS
Tomodensitométrie ;
Cardiopathies congénitales complexes ;
Dose de rayonnement ;
Pédiatrie

Résumé Le scanner est devenu un examen diagnostique clé dans les cardiopathies congénitales complexes. L'amélioration de la résolution spatiale et temporelle et les techniques permettant une diminution des doses autorisent son utilisation dans la population pédiatrique. Les cardiopédiatres doivent connaître les bases techniques et les limites du scanner, sans oublier l'optimisation des doses, afin de le prescrire et le réaliser à bon escient. Dans cette revue, nous exposerons dans une première partie les principales indications du scanner cardiaque chez les nourrissons et les enfants en pratique clinique et dans une seconde partie, l'optimisation des doses. Ceci apparaît important afin de réaliser mais aussi d'interpréter les images pour la prise en charge médicale et chirurgicale des patients.

Introduction

The introduction of helical computed tomography (CT) about 30 years ago has changed the diagnostic medical approach around the world, with a considerable increase of the number of CT examinations in both paediatric and adult patients.

Transthoracic echocardiography (TTE) and cardiac catheterization serve as the mainstay modalities in the diagnostic process of complex congenital heart diseases (CHD). TTE with colour Doppler provides excellent definition of intracardiac anatomy, including haemodynamic evaluation. However, its performance may be lacking in terms of visualization of extracardiac structures such as the distal aorta, aortic arch branches, distal pulmonary arteries, pulmonary veins, or other associated vascular structures and airways abnormalities. Invasive catheter-directed cardiac angiography has been, for many years, the gold standard for the anatomical assessment of CHD in children, having the potential to couple anatomical and haemodynamic evaluation. However, its disadvantages include the risk of complications of an invasive procedure (vessel dissection, stroke, pseudoaneurysm formation, costs of hospital stay, patient discomfort) [1,2]. Moreover, cardiac catheterization is not informative regarding associated airway pathology.

Consequently, cardiac electrocardiogram (ECG)-CT scanning, with all the recent advances and improvements in spatial and temporal resolution, is acquiring a key role in the diagnostic process of complex cardiac congenital anomalies. However, the increase in radiation exposure from CT in children has become a great concern. Although the link between medical radiation exposure and future cancer risk is still controversial, children have higher radiation sensitivity and a longer life expectancy. These factors justify a maximal effort to have scanning protocols specifically designed for the paediatric population and to restrict CT indication to specific cases when CT scan results are considered essential for patient management.

In the first part of this review, we will discuss the principal indications in clinical practice for paediatric CT scans. In the second part, we will discuss some optimization methods for reducing radiation dose exposure.

Principal indications for CT scanning in children

CT scanning is an accurate, non-invasive imaging modality to visualize cardiac, vascular, extracardiac and extravascular structures. After contrast injection, the heart and vessels can be evaluated in detail. With isotropic reformatted images created in multiple different planes, CT scans provide excellent anatomical detail of the cardiovascular system and any abnormalities. CT scans may provide accurate information to assess complex spatial relationships of vascular airway compression frequently associated with CHD [3]. In the paediatric population, TTE contributes — in the majority of cases — to the definition of anatomical details and the relationships between the great vessels. In some complex cases, however, CT scanning is necessary to complete the diagnosis with precise anatomical elements, which are also required by surgeons to define the surgical strategy before intervention.

Previous data [4—6] have suggested that CT scanning is a reliable modality for complex CHD in infants [7], but it is not yet recommended for routine evaluation in newborns and young infants. Use of CT varies according to the age of the patient for several technical reasons. Here, we will discuss the principal indications in clinical practice.

Systemic vessels

Aortic coarctation

In complex cases of aortic coarctation (Video 1), CT scans may provide important anatomical details not shown by

Figure 1. Sagittal multiplanar reformatted image of the descending aorta with a stent in the isthmic region. It shows its perfect apposition and patency.

Figure 2. Maximum intensity projection axial reformatted image of a double aortic arch. It shows the relationship between the trachea and oesophagus, which are surrounded by vascular structures.

echocardiography, especially in distal lesions. CT scans can be performed at the time of diagnosis, as a complement to TTE, and during follow-up after surgical correction or catheter intervention when complications such as restenosis, residual stenosis, aneurysm or pseudoaneurysm should be assessed.

CT scanning allows measurement of the diameter and length of the stenosis and the distance between supra-aortic trunks. CT scans allow calculation of the diameter — in truly orthogonal planes — of the vessel axis to make the segments comparable. After endovascular treatment, CT scanning is suited to evaluate stent patency in children [8] (Fig. 1). For patients suspected to have aortic arch hypoplasia, CT provides information about the exact location, shape and length of the hypoplastic segment, as well as the course of the collateral vessels.

Complex arch anomalies

Aortic arch anomalies account for 0.5—3% of the population [9]. Corone and Vernant [10] proposed a system that used a hypothetical embryonic double aortic arch to explain the various congenital anomalies of the aortic arch determined by the site of interruption or atresia of the embryonic arch system. In the majority of complex arch anomalies, CT is mandatory to diagnose the type of anomaly and the relationship between the trachea and oesophagus, which are surrounded by vascular structures (Fig. 2, Video 2).

Supravalvular aortic stenosis

Supravalvular aortic stenosis is a focal or diffuse narrowing of the aorta starting at the sinotubular junction and often involving the entire ascending aorta. CT allows visualization of the entire aorta and is a reliable modality to demonstrate the extent of the supravalvular aortic stenosis. CT scans are able to determine the permeability of the coronary ostia, especially in Williams syndrome, avoiding the risk of invasive coronary angiography. With an ECG-gated technique, myocardial hypertrophy and bicuspid valve can be depicted.

Aortopulmonary window

Aortopulmonary window is a communication between the ascending aorta and the pulmonary trunk or right pulmonary artery. It is a rare entity, representing less than 0.1% of CHD cases. Non-invasive evaluation with TTE may not demonstrate the communication in up to 37% of cases [11]. CT scans can demonstrate the communication between the aorta and the pulmonary artery, as well as signs of pulmonary hypertension. CT can accurately identify the size and the exact location of the defect, and its relationship with the origin of the coronary arteries. CT scans can play an important role and be of significant help for the surgeon to plan the surgical strategy.

Pulmonary vessels and aortopulmonary collaterals

In all forms of pulmonary obstruction with suspicion of distal anomalies of pulmonary arteries — associated or not to the presence of major aortopulmonary collateral arteries (MAPCAS) — CT scans may define the distal anatomy of pulmonary branches and also precisely identify the anatomy of MAPCAS.

Invasive angiography is still performed in association with CT scans, especially to define the relationship between MAPCAS and native pulmonary branches and their eventual communication. CT scans corroborate invasive data and complement them with important anatomical details.

Three-dimensional (3D) reconstructions of vessels and the trachea permit understanding of their reciprocal relationship (Video 3), allowing the surgeon to plan the surgical strategy. This kind of analysis should be performed before planning surgical interventions because anatomical details of pulmonary anatomy and collateral vessels are crucial for long-term prognosis. In case of other complex anomalies, such as retrotrachea pulmonary artery (pulmonary artery sling, absent left pulmonary artery), CT may provide details of distal anatomy and associated airway anomalies [12] (Fig. 3).

Pulmonary venous anomalies

In complex anomalies of pulmonary veins, such as subdiaphragmatic or mixed total anomalous pulmonary venous return or scimitar syndrome, CT can visualize and precisely identify the anatomy of all pulmonary vein connections.

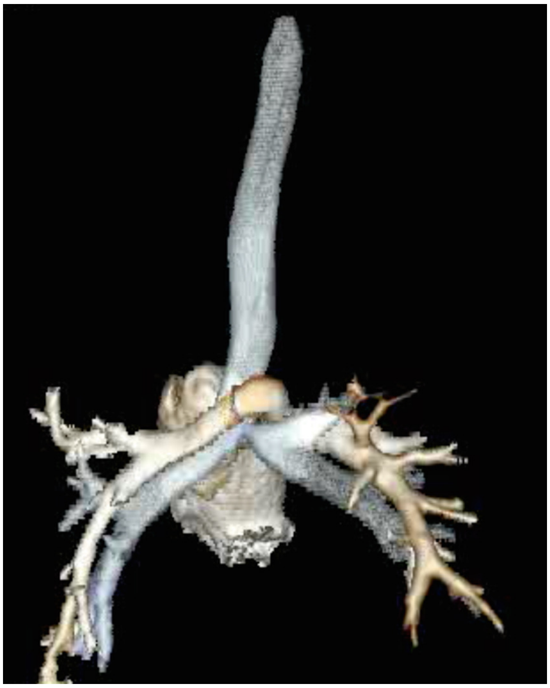

Figure 3. Posterior view of a volume-rendering reconstruction of the great vessel and trachea in a patient with left pulmonary artery sling.

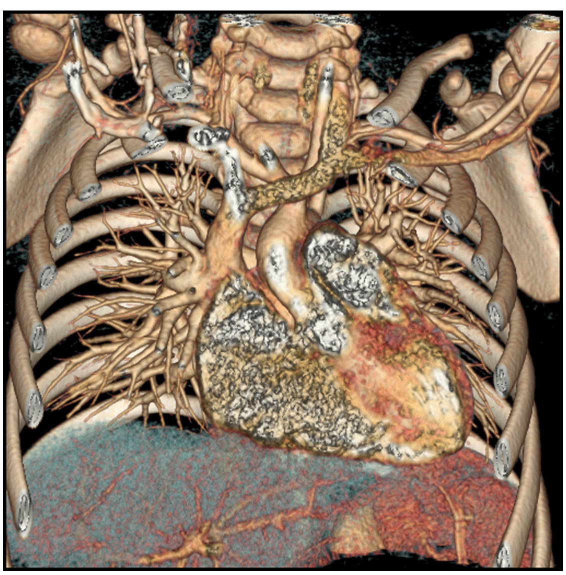

Figure 4. Posterior view of a volume-rendering reconstruction of a sub-diaphragmatic anomalous venous pulmonary return in the inferior vena cava. The left pulmonary venous collector is red, the right cavity is blue and the left cavity, limited to the left ventricle, is yellow.

When the anomalous pulmonary drainage is totally in coronary sinus or in superior vena cava/innominate vein, TTE is often sufficient to make the right diagnosis. When the anomalous drainage is subdiaphragmatic or of mixed type and the clinical status of the patient is not critical, CT is important to define the complete anatomy of the pulmonary veins (Fig. 4).

Coronary arteries

CT scanning in patients with suspected anomalies of coronary arteries is well described and has good accuracy [13—15]. It provides good visualization of coronary ostia, coronary dominance, angulation from the aortic root, ostial narrowing, length of intramural course and fistulae.

Newborns and infants

In newborns and infants, the use of coronary CT is limited because TTE can accurately diagnose coronary anomalies in the majority of cases. In the more common cases of anomalous origin of left coronary artery from pulmonary artery (ALCAPA) and anomalous origin of right coronary artery from pulmonary artery (ARCAPA), TTE is generally sufficient to provide good images for diagnosis in newborns and infants. Suspicion of anomalous origin of coronary artery from opposite sinus of Valsalva (ACAOS) happens later in life, usually after infancy.

The rare indications to perform a CT scan to visualize coronary arteries in newborns are large fistulae, absence of one coronary branch, post-surgical coronary complications (as in post-switch intervention) or unusual forms of anomalous origin of coronary artery (Figs. 5 and 6).

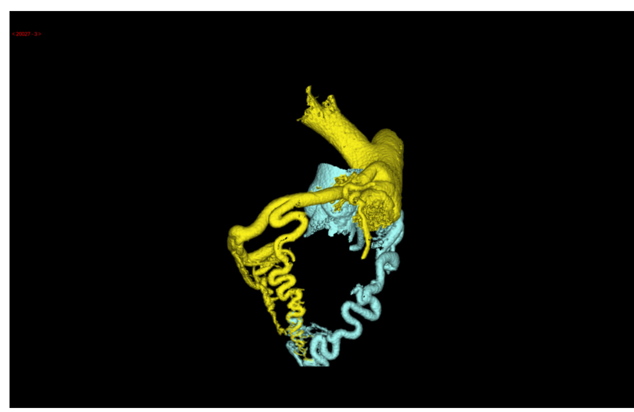

Figure 5. Volume-rendering reconstruction of anomalous origin of right coronary artery from pulmonary trunk (ARCAPA). The pulmonary trunk and right coronary artery are blue, the aorta and left coronary artery are yellow.

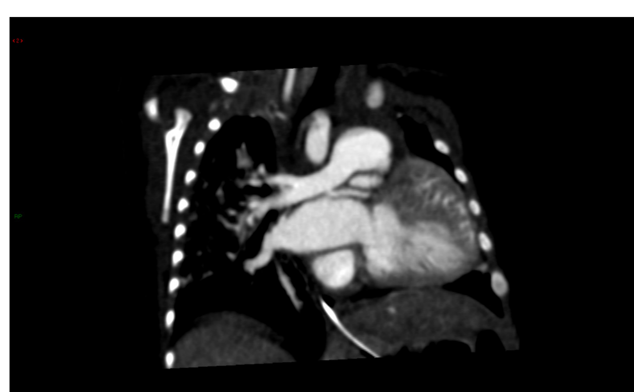

Figure 6. Maximum intensity projection coronal reformatted image of anomalous origin of left coronary artery from right pulmonary artery.

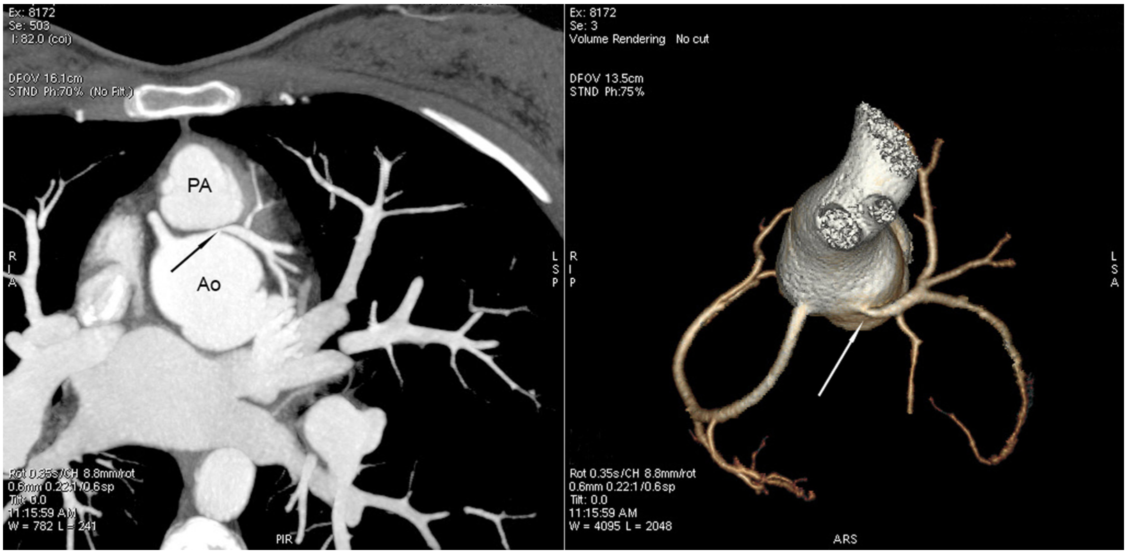

Figure 7. Maximum intensity projection axial reformatted image and volume-rendering reconstruction show the stenosis of left coronary ostium after arterial switch operation (courtesy of Diala Kraiche).

Children > 1 year old

In children with a suspected anomaly of the origin of coronary arteries (e.g. ACAOS), CT provides good visualization of coronary ostia (Fig. 7) and intramural course between the vessels. Coronary CT is also used for follow-up after surgery. It is also useful for follow-up of patients with Kawasaki disease and coronary aneurisms to check distal coronary branches, avoiding repeated invasive angiography.

Transposition of the great arteries

Patients with transposition of the great arteries who have been operated with arterial switch at birth are at risk for coronary artery complications in later life [16—19]. In our institution, we screen all patients after arterial switch operations at about the age of 5—6 years using coronary CT scanning. The objective of this examination is to analyse the coronary ostia and the proximal part of the coronary arteries (Video 4). Coronary artery complications after arterial switch operation usually concern the ostium and proximal part of the vessel. The position of the reimplantation of coronary ostia is also important to define the risk of late complications [20,21] (Fig. 8).

Complex congenital heart disease

In some cases of complex forms of CHD, when TTE lacks intracardiac detail, we perform cardiac CT especially to define the relationship between the great vessels and interventricular septal defect for surgical strategy.

Adolescents

In adolescents who have undergone surgical repair of CHD, CT is often replaced by cardiac magnetic resonance imaging (MRI) to avoid repeated radiation exposure during follow-up. MRI provides reproducible and reliable functional and morphological information. In case of specific contraindications, ECG-CT scans can replace cardiac MRI. CT should be reserved for situations in which it is expected to provide important diagnostic information and less risk than other modalities.

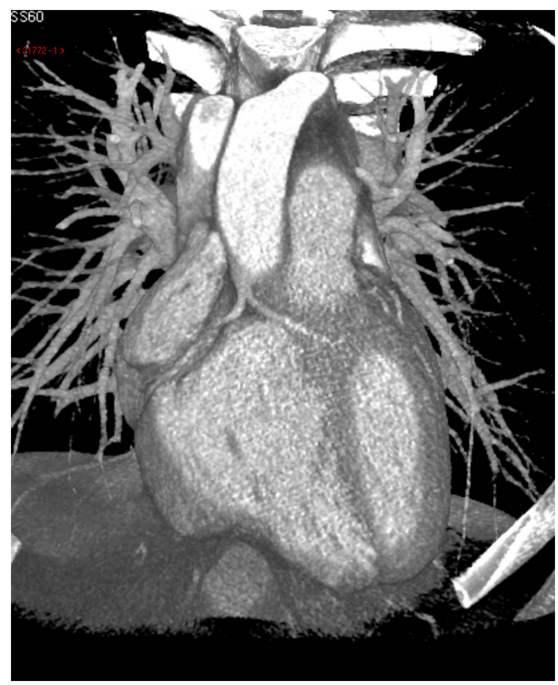

Figure 8. Thick maximum intensity projection coronal reformatted image of anomalous origin of left coronary artery from right coronary ostium with intramyocardial course. Ao: aorta; PA: pulmonary artery.

Optimization of radiation dose

The effect of dose reduction is of utmost importance in younger and more radiosensitive patients [22—33]. Diagnostic reference levels may identify high-dose practices where dose reduction techniques would have the greatest effect.

Unfortunately, the only diagnostic reference level data published in France are about thoracic CT [34], not specifically for cardiac CT.

Radiation is directly proportional to kilovoltage (kV), tube current, scan time, slice thickness and field of view. Protocols vary depending on the type of scan, so it is not possible to provide parameters for all types of CT scan.

In recent years, a number of dose-reduction techniques have been introduced. These include the availability of low tube potential (70—100 kV) settings, ECG-based tube current modulation and anatomic-based tube modulation. The iterative reconstruction algorithms allow for a reduction in tube current and radiation dose while maintaining acceptable dose properties. Improvements in detector technologies have also allowed radiation exposure to be lowered.

Non-ECG-gated spiral scanning

Non-ECG-gated spiral scanning is a simple technique to perform CT in patients with CHD to visualize the cardiac anatomy, with the exception of the coronary arteries. Cardiac CT with this scan technique has been widely used since the introduction of multi-slice CT. This scan technique may be acquired during either breath holding or free-breathing. The scan time is short and sedation is often not needed. However, contrast injection is always required. The site of contrast injection should be properly discussed in advance with the cardiologist because it varies according to different pathologies (e.g. the leg vein should not be used in patients who have undergone bidirectional cavopulmonary connection; simultaneous injection of diluted contrast agent through the leg and the arm may be used for the evaluation of Fontan patients).

In our experience, in a paediatric population, the manual scan acquisition is preferable to an automatic one. Contrast density in the vascular system is difficult to be automatically evaluated in the paediatric population because of different weights and heights. The radiologist should define the time of the acquisition according to the pathology. They should supervise each scan acquisition according to the smart preparation, where the contrast in a target vessel should be visualized. Then, they manually specify the scan acquisition time after contrast injection. This kind of acquisition allows them to tailor the best contrast density for a specific patient, but it requires a good level of expertise. When appropriate CT parameters are used, the delivered dose of this scan technique is usually in the range of 0.5—2 mSv in children (unpublished data).

Conventional helical scan protocol

Conventional helical scan protocol, also named retrospective-ECG gating mode, uses ECG-gated tube current modulation to irradiate the heart in all cardiac phases. Despite this, a large portion of the information acquired during the scan is not used in the interpretation process. Rather, only one or two of the best phases are used. Thanks to recent improvements and the development of prospective-ECG gating technology, we prefer to reserve this protocol for the evaluation of ventricular function and for detailed coronary artery assessment when arrhythmia is present and when no other cardiac imaging modality is possible.

Table 1 The delivered dose of coronary CT angiography in our series [49].

Age (years)	Retrospective gating ED (mSv)	Prospective gating ED (mSv)
0—4	3.8	0.7
5—7	2.9	0.6
8—18	2.7	0.7

CT: computed tomography; ED: effective dose.

Prospective-ECG gating technology

Prospective-ECG gating technology shows really promising results in radiation dose reduction, for heart rates up to 120 bpm, offering similar imaging quality and diagnostic value [35—38] compared to conventional helical techniques. It uses prospectively triggered axial step-and-shoot scans in which X-rays are turned on only during the required heart phase and turned off completely at all other times. Tailored premedication with betablockers, especially in children > 4 years of age, allows the use of this protocol with really low-dose radiation (about 0.2 mSv if the heart rate is < 65 bpm). Even in case of higher heart rate (65—120 bpm), prospective gating allows a good image quality with low-dose radiation.

Applying iterative reconstructions, the dose can be considerably reduced while image quality is maintained compared to conventional reconstruction methods [39—48]. The iterative-reconstruction method refines the current estimate of the solution by modelling photon statistics and the scanned object, and thus extracts noise in the final image.

The accuracy of iterative reconstruction relies on the way it models the signal-generation process in the estimation of the synthesized projection and in the adjustment of the current estimate. Iterative reconstruction provides significant benefits in clinical practice, especially for low signal/noise examinations. Image quality can be significantly improved; alternatively, the dose can be lowered while image quality is maintained. The delivered dose of coronary CT angiography in our series [49] is presented in Table 1.

Future developments

For the past decade, it has been possible to create 3D models from medical images, especially from MRI and CT scans [50—53]. CT allows rapid and non-invasive examination and provides quality images that can be post-treated to create 3D models. 3D models influence treatment by providing an opportunity to perform detailed and precise preoperative planning. As the technology matures and the cost decreases, 3D models will be increasingly used in surgery.

Conclusions

CT techniques are continuously evolving. Cooperation between radiologists and paediatric cardiologists is crucial to optimize CT performance and radiation dose. The risks and benefits of cardiac imaging should be discussed with the referring cardiologist before cardiac CT. The radiologist should tailor radiation delivery to obtain the minimum image quality sufficient to provide the required diagnostic information. If appropriate protocols and strategies for dose reduction are applied, CT could become a low-irradiation technique. Thanks to its simplicity and high performance, cardiac CT scanning will improve its role in congenital cardiac imaging, even in uncooperative children (no need for general anaesthesia or hospitalization).

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at http://dx.doi.org/10.1016/j.acvd.2015.11.003.

Disclosure of interest

The authors declare that they have no competing interest.

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