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# Cardiac computed tomography angiography in the paediatric population: Expert consensus from the Filiale de cardiologie pédiatrique et congénitale (FCPC) and the Société française d'imagerie cardiaque et vasculaire diagnostique et interventionnelle (SFICV)

## *Tomodensitométrie (TDM) cardiaque en pédiatrie : consensus d'experts de la Filiale de cardiologie pédiatrique et congénitale (FCPC) et de la Société française d'imagerie cardiaque et vasculaire diagnostique et interventionnelle (SFICV)*

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*Abbreviations:* CCTA, Cardiac Computed Tomography Angiography; CHD, Congenital Heart Disease; CT, Computed Tomography; MAPCAs, Major aortopulmonary Collateral Arteries; MRI, Magnetic Resonance Imaging; TTE, Transthoracic Echocardiography.

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**KEYWORDS**
Cardiac computed tomography scan;
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**Summary** This paper aims to provide a paediatric cardiac computed tomography angiography expert panel consensus based on the opinions of experts from the Société française d'imagerie cardiaque et vasculaire diagnostique et interventionnelle (SFICV) and the Filiale de cardiologie pédiatrique congénitale (FCPC). This expert panel consensus includes recommendations for indications, patient preparation, computed tomography angiography radiation dose reduction techniques and postprocessing techniques. We think that to realize its full potential and to avoid pitfalls, cardiac computed tomography angiography in children with congenital heart disease requires training and experience. Moreover, paediatric cardiac computed tomography angiography protocols should be standardized to acquire optimal images in this population with the lowest radiation dose possible, to prevent unnecessary radiation exposure. We also provide a suggested structured report and a list of acquisition protocols and technical parameters in relation to specific vendors.

**MOTS CLÉS**
TDM cardiaque ;
Cardiopathies congénitales ;
Dose d'irradiation ;
Protocoles d'acquisition

**Résumé** Notre papier vise à offrir un consensus d'experts sur les modalités de réalisation d'un scanner cardiaque chez l'enfant sur la base de l'expérience de la Société française d'imagerie cardiaque et vasculaire diagnostique et interventionnelle (SFICV) et de la Filiale de cardiologie pédiatrique congénitale (FCPC). L'article inclus des recommandations sur les indications, sur la préparation du patient, sur les techniques pour réduire la dose de radiations livrée au patient et sur les techniques de post-traitement des images. On est profondément convaincus que pour exploiter le potentiel de la technique de TDM cardiaque tout en protégeant les enfants d'une exposition dangereuse aux rayons X une expertise médicale dédiée et un entrainement spécifique sont nécessaires. D'autant plus, les protocoles d'acquisition nécessitent une standardisation visée à éviter toute exposition inutile chez la population pédiatrique. À la fin de l'article, un exemple de compte rendu est suggéré, avec aussi des propositions des protocoles d'acquisition selon les différentes industries.

## Background

In the paediatric population with congenital heart disease (CHD), cardiac computed tomography angiography (CCTA) has enhanced the applicability of cross-sectional anatomical imaging, and is now often used as a complementary diagnostic tool to echocardiography, cardiac magnetic resonance imaging (MRI) and cardiac catheterization. However, when the radiologist is faced with a CHD case, adapted protocols can be formulated for the specific indication, by applying general principles of computed tomography (CT) acquisition and contrast administration. Individual modalities have specific attributes for the assessment of CHD, but no single imaging method is comprehensive in any given patient. Therefore, referrers and cardiac imagers must recognize the limitations of each imaging technique, and endeavour to use a multimodality approach. CCTA can be used to obtain isotropic volume data, and high-quality two- and three-dimensional multiplanar reformatted images can be created to accurately and systematically delineate the normal and pathological morphological features of the cardiovascular system. CCTA can be technically challenging and demanding in uncooperative young children. Recent technical developments in CCTA, notably decreasing radiation dose, have improved patient medical care. Therefore, paediatric CCTA protocols should be standardized to acquire optimal images in this population with the lowest radiation dose possible, to prevent unnecessary radiation exposure [1].

We propose a paediatric CCTA expert panel consensus based on the opinions of experts from the Société française d'imagerie cardiaque et vasculaire diagnostique et interventionnelle (SFICV) and the Filiale de cardiologie pédiatrique congénitale (FCPC). This expert panel consensus includes recommendations for indications, patient preparation, CTA radiation dose reduction techniques and postprocessing techniques.

## Patient preparation

Careful patient preparation is a prerequisite for successful performance of paediatric CCTA. The patient should be quiet and relaxed. Babies should be comfortably installed in a specially designed bed with blankets and bands to avoid motion and keep warm. Electrocardiogram electrodes are stuck on the chest outside the examination zone to avoid artefacts. The examination and breath hold should be explained to small children. Parents, with protection against radiation exposure, may stay with their child during imaging. The use of a power injector is preferred to manual injection of contrast media. Patients need to have a good injection site (antecubital vein, foot or head). For children aged < 6 years, it is recommended to perfuse them a few hours before the CCTA, to keep them calm. It is recommended to apply an Emla® patch (AstraZeneca AB, Södertälje, Sweden) at the puncture site 1 h before perfusion. To obtain optimal vascular enhancement in subjects with superior cavopulmonary connection, leg vein injection is recommended. For imaging of total cavopulmonary connection or Fontan pathway, simultaneous injection of contrast agent through arm and leg veins is preferred. General anaesthesia is not necessary if newborns and infants can drink their baby bottle before the examination, to fall asleep. Teenagers and children aged > 6 years are able to keep calm and follow instructions provided by staff. However, for younger children, light sedation is occasionally required, according to the paediatric cardiologist's or paediatric anaesthetist's practice. In very rare cases, general anaesthesia is necessary; scheduling of a pre-anaesthesia classical evaluation is required in this situation.

## CCTA techniques

A variety of scan techniques can be used to image children with CHD. Non-electrocardiogram-gated helical scans and retrospectively electrocardiogram-gated scans have been abandoned, to be replaced by prospectively electrocardiogram-triggered sequential scans or one-shot acquisition.

The scan length is defined according to clinical indication: limited to the heart for coronary indications or extended to the whole thorax for the vascular anatomy studies.

Precise acquisition parameters should be adapted locally, depending on the CT scanner model that is available. The main difficulty of CCTA is that the cardiac phase with the lowest coronary artery motion is strongly dependent on the heart rate, and is specific to each vessel [2]. A biphasic injection of iodinated contrast (270—300 mgI/L < 40 kg; 320—350 mgI/L > 40 kg) followed by a saline flush (1 cc/kg) is injected using a power injector. The amount of iodinated contrast is based upon the weight of the patient and varies from 1.5 to 2 cc/kg. Acquisition timing should not be automatic, but should be decided by the operator, according to the indication and specific pathology of the patient.

### Electrocardiogram-gated prospective acquisition

Electrocardiogram-gated prospective acquisition is recommended for acquiring cardiac images in children [3,4]; it is applicable to any heart rate condition and can even be used during free breathing. Beta-blockers may be used, not to decrease the heart rate, but to stabilize it, especially in infants aged > 4 years.

First, anteroposterior and lateral scout views of the chest are performed; these are used for both acquisition planning and dose modulation. It is recommended to use a field of view adapted to the paediatric population.

A manual exposure window and target phase are selected before the scan, according to heart rate and heart rate variability. Low heart rates (< 70 beats/min) and high heart rates (> 120 beats/min) tend to be less variable, and a target phase of 75% and 40% of the cardiac cycle, respectively, is recommended [5]. The exposure window is often not necessary in these cases. For heart rates between 75 and 120 beats/min and in all cases of high heart rate variability it is often difficult to preview the right target phase; an exposure window of 200—400 ms is recommended to provide a reconstruction window of 30—80% of the R-R cycle. During the acquisition, a 0.5 mm × 80 rows volume is acquired every first to third beat: one beat for the acquisition, the next beats for table motion. Multiple volumes are acquired to cover the entire heart, and automatic adaptive blending is used to stitch the scanned volumes into one reconstructed volume dataset. The selected Kv should be as low as possible for the machine and the patient, and mA are adapted according to patient weight. Based on prospectively electrocardiogram-triggered examinations, automated algorithms have been developed to determine the optimal cardiac phase in a short prospective multiphasic acquisition [6—8]. The clinical efficiency of electrocardiogram-gated prospective acquisition has been demonstrated in patients with fast heart rates as well as in paediatric patients, while keeping the radiation dose low [6,8].

### One-shot acquisition

One-shot acquisition allows acquisition of the whole heart within one rotation, and therefore with no step artefacts; it is an axial acquisition. The target phase should be chosen as described previously for general prospective acquisition protocols. Kv and mA should be set as low as possible.

## Dose reduction

Specific care to decrease radiation exposure as low as possible is crucial in children with CHD, who are susceptible to potential life-long exposure to irradiating procedures. The choice of scanning protocol should be selected for and tailored to the patient and their clinical characteristics (heart rate, heart rate variability, weight and thorax morphology). The kV and mA should be adapted to the patient's morphology. Dose modulation during acquisition must be used, if available. Moreover, all vendors have developed several reconstruction algorithms that allow the radiation dose to be lowered. All these techniques should be combined to lower the radiation dose (see section entitled ''Postprocessing''). Several examples of electrocardiogram-gated sequential acquisitions are given in Appendix B.

## Postprocessing

Over the past decade, postprocessing imaging has evolved rapidly to perform accurate image reconstruction while keeping the scan dose to a minimum.

### Iterative reconstruction and motion correction software

Iterative reconstruction algorithms and specific post-treatment software to reduce coronary motion artefacts should be used when available [5,9—20].

### Reconstruction kernel

The reconstruction kernel is a main parameter that affects image quality because of the trade-off between spatial resolution and noise for each kernel. A smooth kernel generates images with low noise, but with reduced spatial resolution. A sharp kernel generates images with higher spatial resolution, while increasing the noise [21,22].

As a result of the wide variety of clinical situations requiring paediatric CCTA, specific attention must be paid to the selection of the reconstruction kernel. Smooth kernels will be used for native structures to reduce image noise and enhance low contrast detectability, whereas sharper kernels will rather be used in case of stents, metallic devices or prosthetic valves [23,24].

The modulation of slice thickness and the settings of standard reconstruction kernels differ considerably between the different vendors [25].

### Windowing

Low tube voltage, commonly used for paediatric CCTA, leads to a different image aspect if the window setting is not adapted, mainly because of increased attenuation of high-density structures, such as iodinated vessels. Wider and higher window settings may minimize this effect and correct the image noise appearance [26,27]. Images are commonly processed with standard soft tissue (e.g. width, 400—450 HU; level, 40—50 HU) and lung (e.g. width,—1600 to—1800 HU; level,—450 to—550 HU) window settings [28].

### Multiplanar reconstructions

The interpretation of a CCTA scan traditionally starts with axial plane review. Because of the wide variety of clinical situations and anatomies, paediatric CCTA very often requires multiplanar reconstructions or specific volume and kinetic rendering techniques.

Two-dimensional multiplanar reconstructions can be generated in any plane with resolution comparable with that of axial images. Coronal and sagittal images provide information about cardiovascular structures, particularly structures that traverse the z-axis, which may not be apparent on axial images. This method is therefore directly dependent on slice thickness and on the overlap of the slices. In order to improve the quality, axial CCTA images should be reconstructed using thinner reconstructed sections overlapped by at least 50% [29—37].

Curved multiplanar reconstructions enable the two-dimensional display of the complete course of a vessel in a single image, by fitting a surface along the vessel centreline to assess maximum diameters, length or stenosis, or along the central bronchi to assess vascular airway compression [33]. A drawback of the curved multiplanar reconstruction technique is that only a single branch can be displayed at a time [29].

### Maximum intensity projection and volume rendering

Comprehensive review may require a more integrated approach. Two postprocessing techniques are detailed below: maximum intensity projection and volume rendering.

### Maximum intensity projection

Maximum intensity projection consists of projecting the voxel with the highest attenuation value on every view throughout the volume onto a two-dimensional image [35]. A single volumetric parameter from the original data is used to reconstruct customizable images. This method tends to display bone and contrast material-filled structures preferentially, making it a particularly suitable tool for the study of heart structures and large vessels. Thus, maximum intensity projection is widely used in paediatric CCTA because of its simplicity of usage. Maximum intensity projection sections of variable thickness are excellent for assessing the size and location of vessels, including the coronary arteries. However, the technique has a major drawback, in that the depth and occlusion information cannot be perceived in the output images, leading to confounding spatial relationships [37].

### Volume rendering

Volume rendering defines the optical properties of voxel densities encountered along the ray. A specific colour is assigned to the density of each voxel. The results vary according to the segmented area, the chosen colour, opacity and lighting settings, and are therefore not standardized [29]. This technique is sensitive to image noise, so increased slice thickness will improve image quality without raising the radiation dose [32]; it provides an overview of the whole heart anatomy, with particularly detailed visualizations of high-density objects, including contrast-enhanced vessels and highly vascularized structures [36]. A number of articles have addressed the accuracy of volume rendering in various applications, such as vascular stenosis, producing realistic images suitable for referring physicians or surgeons [38—40], and useful for three-dimensional printing, computational modelling and virtual computed procedures [41—44]. Novel techniques, such as cinematic rendering, are emerging to produce photorealistic images, but without paediatric application as yet [7].

## Cine CT

Cine CT is not recommended in children; the use of echocardiography or cardiac MRI is preferred. Indeed, radiation exposure increases substantially without current tube modulation during the acquisition [28,39]. Even if functional cardiac assessment in CT is highly concordant with cardiac MRI, and improves diagnostic accuracy for complex anatomy [42,45], non-irradiant techniques should be always chosen as the first imaging option when possible.

## Indications

In the paediatric population with CHD, the main indications for CCTA are:

- when echocardiography is incomplete or suboptimal, and more anatomical details are necessary to define the diagnosis and;
- when extracardiac anatomical details are needed for clinical management of the patient and patient cardiac care.

We would like to highlight that cardiac MRI should always be considered as the first option in children with CHD, and eventually excluded if contraindicated or not adequate to answer the clinical question.

More specifically, several indications are recognized in the paediatric population, and are detailed below.

### Coronary artery diseases

CCTA in patients with a suspicion of an anomalous origin of a coronary artery has good accuracy [46—48]; it provides good visualization of coronary ostia, coronary dominance, angulation from the aortic root, ostial narrowing, length of intramural course and presence of coronary fistulas. In newborns and infants, the use of CCTA is limited, because transthoracic echocardiography (TTE) can accurately diagnose anomalous origin of a coronary artery in the majority of cases. CCTA could be complementary to TTE in cases of anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) and anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA). Symptoms of anomalous origin of the coronary artery from the opposite sinus of Valsalva (ACAOS) happen later in life, usually after infancy. The main indications for performing a CCTA to visualize coronary arteries in newborns are large coronary fistulas, absence of one coronary branch, postsurgical coronary complications (as in postswitch intervention) or unusual forms of anomalous origin of coronary artery. Furthermore, CCTA is useful in the follow-up of patients with Kawasaki disease or coronary aneurisms, or after surgery, to check distal coronary branches or surgical sutures, avoiding repeated invasive angiography.

### Aortic coarctation

In complex cases of aortic coarctation, CCTA provides important anatomical details not shown by echocardiography, especially in distal lesions. CCTA can be performed at the time of the 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 specified. After endovascular treatment, CCTA is suited to evaluating stent patency in children [49]. 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 CHDs [50]. In the majority of complex arch anomalies, CCTA is mandatory to diagnose the type of anomaly and the relationship between the trachea and oesophagus, which are surrounded by vascular structures. Eventual anomaly or compression of respiratory tree can also be identified.

### 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. CCTA allows visualization of the entire aorta, and is a reliable modality to demonstrate the extent of the supravalvular aortic stenosis. CCTA is able to determine the permeability of the coronary ostia, especially in Williams syndrome, avoiding the risk of invasive coronarography. With the electrocardiogram-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 $< 0.1\%$ of CHDs. Non-invasive evaluation with TTE may not demonstrate the communication in up to 37% of cases [51]. CCTA demonstrates the communication between the aorta and the pulmonary artery, as well as signs of pulmonary hypertension. It can show the precise size and exact location of the defect, and the relationship with the origin of the coronary arteries. CCTA can play an important role and be a significant help in planning the surgical strategy.

## Pulmonary vessels and aortopulmonary collaterals (major aortopulmonary collateral arteries [MAPCAs])

In all forms of pulmonary obstruction with suspicion of distal anomalies of pulmonary arteries associated or not with the presence of MAPCAs, CCTA may define the distal anatomy of pulmonary branches and also show the precise anatomy of the MAPCAs. Invasive angiography is still performed in association with CCTA, especially to define the relationship between MAPCAs and native pulmonary branches and their eventual communication. CCTA corroborates invasive data and adds important anatomical details. Three-dimensional reconstructions of vessels and trachea permit an understanding of their reciprocal relationship, allowing the surgeon to plan the surgical strategy. In case of other complex anomalies, such as retrotrachea pulmonary artery (pulmonary artery sling, absent left pulmonary artery), CT can show the precise distal anatomy and associated airway anomalies [52].

## Pulmonary venous anomalies

In complex anomalies of pulmonary veins, such as subdiaphragmatic or mixed total anomalous pulmonary venous return or scimitar syndrome, CCTA visualizes and shows the precise anatomy of all pulmonary vein connections. When TTE is insufficient to diagnose a total anomalous pulmonary drainage in the coronary sinus or in the superior vena cava/innominate veins, CCTA should be performed. When the anomalous drainage is subdiaphragmatic or mixed type, and the clinical status of the patient is not critical, CCTA is important to define the complete anatomy of pulmonary veins.

## Transposition of the great arteries

Patients with transposition of the great arteries who have undergone an arterial switch operation at birth are at risk of coronary artery complications in late follow-up [53—56]. After an arterial switch operation at the age of 5—6 years, coronary CCTA permits analysis of the coronary ostia and the proximal part of the coronary arteries. Coronary artery complications after an arterial switch operation usually concern the ostium and the proximal part of the vessel. The position of the reimplantation of coronary ostia is also important to define the risk of late complication [57,58].

## Intracardiac anatomy in complex CHD

In some cases of complex forms of CHD, when TTE is lacking concerning intracardiac details, CCTA is performed, especially to define the relationship between the great vessels and interventricular septal defect for surgical strategy. Three-dimensional modelling from CCTA data enables the cardiovascular anatomy to be reconstructed and eventually printed to help with planning the surgical strategy [59].

## Conclusions

Cardiac CT imaging is largely used in the paediatric population with CHD. Technological advances have resulted in marked improvements in the spatial and temporal resolution of CCTA, with a concomitant increase in speed of data acquisition and a reduction in radiation dose. However, in newborns and infants, CCTA should be performed as a second-line strategy after echocardiography, only if strictly required, and the benefit/risk balance should be kept in mind. In adolescents operated on for CHD, MRI remains the first-choice imaging modality (except for comprehensive study of the coronary arteries), especially to avoid repeated radiation exposure during follow-up. In case of specific contraindications, electrocardiogram-CCTA can replace cardiac MRI, and should be reserved for situations in which it is expected to provide important diagnostic information and pose less risk than other modalities.

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## Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at https://doi.org/10.1016/j.acvd.2020.03.016.

## Disclosure of interest

The authors declare that they have no competing interest.

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