Three-dimensional CT scanning: a new diagnostic modality in congenital heart disease

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New generation multislice CT technology has changed the approach to non-invasive assessment of congenital heart disease, in both paediatric and adult patients. This is mainly because of rapid advances in spatial and temporal resolution and in post-processing capability. At Hôpital Necker-Enfants Malades, CT with multiplanar and three-dimensional reconstruction has become a routine examination in the evaluation of congenital heart disease planning surgery, complex interventional catheterisations and for follow-up. It has proved to be an invaluable diagnostic and decision-aiding methodology in these situations, as a complement to echocardiography and, increasingly, as a substitute for diagnostic angiography (which is usually associated with higher-dose radiation and longer sedation times, as well as occasional morbidity). This review illustrates the current status of 64-slice CT in congenital heart diseases, including assessment of the aorta, the coronary arteries, the pulmonary arteries, the systemic and pulmonary veins, and other intra- and extracardiac malformations.

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multislice CT scanning in the assessment of congenital heart disease.^{7–9} Currently, cardiac ultrasound is the leading diagnostic modality for the interrogation of intracardiac structures in children. However, ultrasound is usually insufficient for the study of certain structures such as the coronary arteries, the aorta and branches of the pulmonary artery, or for systemic and pulmonary venous structures. For visualising these structures, CT allows better diagnostic capability, as a result of the high-precision, three-dimensional imaging it provides.⁹ Three-dimensional imaging is particularly useful for diagnostic investigation of complex forms of congenital heart disease, preparation of complex interventional procedures involving cardiac catheterisation, and the postoperative assessment of surgical reconstructions.

We provide here some examples of the main forms of congenital heart disease where valuable diagnostic information can be obtained from 64-slice CT scanning with three-dimensional volume rendering, using commercially available equipment.

TECHNICAL ASPECTS

The examinations illustrated were carried out with a multislice CT-scan device (GE, LightSpeed VCT, Milwaukee, USA). The acquisition protocols take account of the patient's age, the malformation to be studied, and the ratio of image quality to radiation dose.

Children under 5 years

This group includes neonates, infants and young children unable to achieve a 5-second breath hold and whose heart rates are high, generally above 100 beats/min. In this group of children, good image quality is difficult to obtain owing to respiratory and cardiac movement artefacts, which are all the more marked when the child is anxious or agitated. The rule therefore is to acquire images as quickly as possible while the child is calm or lightly sedated. This implies exploiting the speed of the CT scan along with optimal injection of contrast medium. ECG gating is avoided as it lengthens acquisition time. Contrast medium injection and image acquisition are carefully calculated to achieve optimal image quality. The acquisition delay is automatically determined by placing a region of interest in the aorta or pulmonary artery (depending on whether systemic or pulmonary arterial structures are to be examined), with an automatic triggering threshold at 100 HU. The contrast medium bolus is then delivered by an automatic injector to ensure that all the contrast medium passes at a sufficiently high and uniform rate during the short image

Multislice CT has seen rapid technological advances over the past 3–5 years, particularly with the recent emergence of 64-slice devices. Indeed since 2000 there has been a doubling of detector rows approximately every 2 years, with 4, 16, 32 and then 64 rows in 2004. This latest technology allows much faster acquisition times, improved spatial resolution (with thinner slices) and with fewer motion artefacts, owing to improved ECG gating. CT scanning is thus becoming a leading modality for non-invasive imaging of cardiovascular diseases in adults, particularly with CT coronary angiography.^{1,2} Indeed, in cardiovascular imaging in adults, multislice CT provides very high image quality equal or even better than that of conventional invasive angiography, for the proximal epicardial vessels. CT scanning is also a valuable diagnostic tool for the diagnosis of pulmonary embolism, complex arteriovenous malformations, aneurysms, aortic dissections and even coronary arteries in certain circumstances. In such clinical situations, CT scanning often obviates the need for conventional angiography.^{3–6}

Although many reports of cardiac CT imaging in adults have been published, there is little such information about imaging in paediatric cardiology. Only a few papers have examined the place of

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Figure 1 Anomalous left coronary artery rising from the pulmonary artery.

acquisition period. Injection concludes with saline rinsing of the tubes, to avoid enhancement artefacts generated by the persistence of contrast medium in the superior vena cava. Finally, radiation dose must be minimised by continuous attention to the voltage and current being applied, keeping these aspects reduced as far as possible without degrading image quality.

For this group of children we use the following protocol:

- *Patient preparation:* sedation (oral or intrarectal pentobarbitol or chloral hydrate, or both, 30 minutes before the examination); preparation of the neonate or infant, wrapped comfortably in Velpau strapping if necessary; insertion of a 22–24G peripheral venous line in the elbow, back of the hand or foot.
- *Scan parameters:* rotation time 400 ms; high pitch 0.984, slice thickness 0.625 mm; low voltage 80 kV; current varied during acquisition and according to the weight of the child.
- *Injection parameters:* volume of contrast medium (Iopamerol 300 mg/ml; Schering SA, Berlin, Germany) 1 ml/kg, followed by 5 ml saline solution; flow rate 0.5–1 ml/s for neonates; 1–2 ml/s for older children.

Children over 5 years

For this older group of children, breath holding with appropriate and sympathetic coaching is generally possible. Sedation

should be avoided if good cooperation from the child is required to ensure breath holding during the acquisition period. Choice of acquisition protocol depends above all on the malformation to be explored. Study of the great vessels (aorta, pulmonary arteries, systemic or pulmonary venous return) is relatively straightforward: it is carried out by fast acquisition times without ECG gating. Study of the coronary arteries, on the other hand, is more difficult. The best image quality is obtained with ECG-gated acquisition in a child whose heart rate is maintained stable and slow by administration of a β blocker (<70 beats/min). In all cases, the same precautions should be taken for contrast medium injection parameters and radiation dose as already described.

For this group of older children we use the following protocol:

- *Patient preparation:* no sedation; β blocker in the case of CT coronary angiography (propranolol, 1–2 mg/kg orally 2 hours before the examination); 20–22 G peripheral venous line in the elbow, back of the hand or foot.
- *Scan parameters:*
 - Examination of the great arteries: rotation time 400 ms; high pitch 0.984; slice thickness 0.625 mm; voltage 80–100 kV depending on body weight; current varied during acquisition and adapted to the weight of the child.
 - Examination of the coronary arteries: rotation time 350 ms; short pitch 0.2; slice thickness 0.625 mm; voltage 80–100 kV depending on body weight; current varied during acquisition and adapted to the weight of the child.
- *Injection parameters:* volume of contrast medium (Iopamerol 300 mg/ml; Schering SA, Berlin, Germany) 1.5 ml/kg maximum 70 ml, followed by 5–10 ml saline solution; high flow rate 2–3.5 ml/s.

IMAGE PROCESSING

The acquisition volume is analysed on dedicated workstations. Routine analyses are carried out on standard slices in various planes, multiplanar and three-dimensional reconstructions. This post-processing stage is partly facilitated by vascular analysis algorithms with an interface that is interactive and simplified for the user. For CT coronary angiography, analysis is more refined and requires reconstructions at different phases of the cardiac cycle: a first set of images is obtained systematically at 75% of the R–R interval (corresponding to late diastole); further image reconstructions are performed at every 10% of the R–R interval when artefacts are present.

Figure 2 Coarctation associated with hypoplasia of the aortic arch in a neonate. The coarctation can be located (arrow) and the degree and extent of hypoplasia of the aortic arch defined. DA, ductus arteriosus.

Figure 3 Vascular ring. Left panel: double aortic arch. Right panel: severe compression of the trachea (arrow).

CLINICAL APPLICATIONS

Coronary arteries

Indications for CT coronary angiography in the diagnosis of congenital abnormalities of the coronary arteries are now well established in adults.^{10–11} In congenital heart disease, the coronary arteries are important if anomalous, or reimplanted after surgery in certain circumstances. The main advantage of CT scanning is to provide a precise description of the three-dimensional anatomy of the coronary arteries, in particular their origin and course (fig 1). CT coronary angiography is especially useful in screening for complications after coronary re-implantation in cases of anomalous left coronary artery arising from the pulmonary artery or after the arterial switch operation for transposition of the great arteries. With even four-slice CT scanning we demonstrated excellent negative predictive value of this technique, allowing CT coronary angiography to be used for screening for coronary lesions after the arterial switch procedure and reserving selective coronary angiography for confirmation in cases of suspected abnormality detected by CT.¹² The ability of 64-slice CT scan to provide accurate imaging of the proximal coronary vessels is better than

four-slice scanning, and may even replace selective coronary angiography. This still awaits prospective validation; our preliminary results, however, are extremely promising.¹³

Other non-irradiating techniques—for example, MRI—are theoretically preferable for repeated screening examinations, particularly in children. Nevertheless, MRI is currently limited in congenital heart imaging by relatively poor temporal resolution (particularly for assessment of the coronary arteries) and often-complex acquisition protocols requiring a long examination so that general anaesthesia is often necessary in young children.¹⁴

Aortic arch abnormalities

Imaging methods using slices—CT and MRI—are the preferred techniques for morphological study of the aorta—for example, in the investigation of coarctation and congenital degenerative diseases of the aorta. Being complementary to echocardiography, CT and MRI allow precise pre- and postoperative assessment of the thoracic aorta.^{15–16} In our experience, CT scan is often extremely useful for neonates with isthmus coarctation associated with hypoplasia of the aortic arch (fig 2). It precisely

Figure 4 Mapping of pulmonary vascularisation in a child with pulmonary atresia with ventricular septal defect. Visualisation of major aortopulmonary collateral arteries, which show a seagull shape (in blue).

Figure 5 Pulmonary arteries supplied by systemic pulmonary anastomoses. Three-dimensional CT makes it possible to check the patency of palliative anastomoses (arrows). Note the deformation of the origin of the right pulmonary artery, at the level of the implantation of the right Blalock anastomosis (posterior view).

Figure 6 Partial anomalous pulmonary venous return. Note the anomalous pulmonary venous return from the right superior pulmonary vein, which drains into the superior vena cava. LA, left atrium; LIPV, left inferior pulmonary vein; LSPV, left superior pulmonary vein; RIPV, right inferior pulmonary vein; RSPV, right superior pulmonary vein; SVC, superior vena cava.

identifies the site of the coarctation, determines the degree of narrowing and, above all, defines precisely the extent of hypoplasia of the aortic arch, thereby assisting the choice of surgical technique (simple left posterolateral thoracotomy or midline sternotomy). CT is preferable to MRI for this group of children owing to the simplicity of the examination and the rapidity of image acquisition, generally less than 2 seconds for neonates.

For vascular rings, multiplanar and three-dimensional reconstruction clearly demonstrates the origin and course of the great vessels and the relationship of the vessels to the adjacent airway (fig 3).¹⁷

Some authors have suggested that MRI is the “gold standard” for the evaluation of vascular rings,¹⁸ however, MRI quality is commonly affected by respiratory artefacts in neonates and infants. In our experience, MRI examination in children under 7 years requires profound sedation or general anaesthesia. Thus, for practical reasons, MRI is only used to study older children, who can hold repeated breath during an examination which often lasts for 30 minutes.¹⁹

Pulmonary arteries

Precise anatomical assessment of the right outflow tract and the pulmonary arteries is essential in the investigation of certain types of cyanotic congenital heart diseases such as Fallot's tetralogy or pulmonary atresia with ventricular septal defect (PA-VSD). In such cases, echocardiography is often inadequate for the study of the pulmonary arteries beyond their proximal segments. CT scanning, by contrast, is well suited for precise three-dimensional mapping of the pulmonary arterial tree and any aortopulmonary collateral arteries (fig 4). We currently perform routine CT scanning in the preoperative investigation of PA-VSD.^{20, 21}

In our experience, CT is also useful for monitoring the growth of the pulmonary arteries after successive palliative interventions in children (fig 5). Measurement of the size of the pulmonary arteries before dilatation or stenting procedures

Figure 7 Supracardiac total anomalous pulmonary venous drainage. Volume rendered CT showing the supracardiac collecting vein draining all four pulmonary veins.

allows accurate sizing of the devices to be used and often obviates the need for selective invasive angiography in such patients.

Venous return

Pulmonary CT venography has been recently described in adults.²² In our practice, CT is used as a complement to echocardiography whenever there is a suspicion of anomalous systemic or pulmonary venous return in the context of complex heart disease. The most common indication concerns the preoperative investigation of a sinus venosus defect, which is commonly associated with a partially anomalous pulmonary venous return, with the right superior pulmonary vein draining

Figure 8 Infradiaphragmatic partial anomalous pulmonary venous drainage. Volume rendered CT showing the right superior pulmonary vein draining into the inferior vena cava.

Figure 9 Pulmonary arteriovenous fistula. Volume rendered CT showing the fistula (big arrow) and identifying the feeding artery and receiving vein. Please note other fistula that had been closed with coils (small arrows).

abnormally into the superior vena cava (figs 6–8). Diagnosis of pulmonary arteriovenous fistulae is also facilitated by CT scanning (fig 9). CT locates the malformation and identifies the feeding artery and receiving vein, thereby guiding treatment. Precise definition of the anomalous systemic venous return is often crucial in determining the appropriate approach for cardiac catheterisation, particularly in cases of isomerism, and for the planning of cardiopulmonary bypass strategies.

Limitations

Multislice CT scanning undoubtedly represents a major advance in the imaging of congenital heart disease. Its application to small children, in particular, is revolutionary. However, it is equally true that for a large number of simple and common malformations, such as atrial septal defect or ventricular septal defect, echocardiography is sufficient for complete definition of the disease, and cardiac catheterisation provides essential haemodynamic data. Experience with this new technology should be acquired prospectively and its benefits must be precisely evaluated through comparative trials against the current standard methods.

Efforts should be made to minimise radiation dose, particularly in children for whom successive examinations are necessary. In our institution, we strictly follow the ALARA (as low as reasonably achievable) principles²³ with regard to radiation exposure. Indeed, CT examinations in children are performed at low kilovoltage (80–100 kV, instead of 120 kV, the standard kilovoltage setting). In adults, such precautions associated with strict examinations according to protocol allowed reduction of the radiation dose in comparison with conventional invasive angiography.²⁴ Further studies are required to compare radiation exposure during paediatric cardiac CT scan with cardiac catheterisation; the latter is known to be associated with high radiation doses because of prolonged fluoroscopy time, multiple cine planes and the complexity of the procedures.²⁵

It is too early yet to regard multislice CT as a substitute for invasive angiography, although this may be demonstrable in future. Its current use is as a complementary diagnostic modality. In young adults with congenital heart disease, in whom ultrasound windows are often suboptimal, CT scanning may be particularly valuable.

CONCLUSION

Three-dimensional imaging obtained with the latest generation of CT-scan devices represents a real advance in the morphological study of cardiovascular malformations in children. In our institution, CT now has a vital diagnostic and decision-aiding function in the assessment of congenital heart disease, as a complement to echocardiography and, more and more often, as a substitute for invasive angiography.

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IMAGES IN CARDIOLOGY

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Increasing severity of functional mitral regurgitation by left bundle branch block

A 70-year-old man with a history of hypertension and diabetes was admitted for new onset congestive heart failure. Three days later, at the time of the diagnostic echocardiography, he exhibited intermittent left bundle branch block (LBBB) and mild mitral regurgitation (MR) (panels A and B).

Narrow- and LBBB-QRS complex corresponding shells of identical velocity proximal to the regurgitant orifice obtained 34 seconds apart are shown in panels C and D, respectively. The continuous wave Doppler derived jet velocity corresponding to the narrow-QRS isovelocity shell (panel E) was higher than those corresponding to the LBBB-QRS isovelocity shell (panel F) because the narrow-QRS complex was associated with a steeper increase of transmitral pressure. Thus, the effective regurgitant area of MR increased from 0.09 cm² (narrow-QRS) to 0.17 cm² (LBBB-QRS).

LBBB related features which might have contributed to this finding include a decrease in left ventricular (LV) ejection fraction and maximal rate of systolic pressure rise from 48% and 1236 mm Hg/s to 43% and 641 mm Hg/s, respectively, and an increase in electromechanical delay between the myocardial segments adjacent to the papillary muscles (as assessed by tissue Doppler velocity analysis) from +4.6 ms to +70.3 ms (not shown).

Resynchronisation therapy in patients with LBBB and severely impaired LV systolic function acutely reduces the severity of MR, thus indirectly supporting the concept of LBBB-related MR aggravation. Our images directly illustrate this concept in a patient with preserved LV ejection fraction and mild MR.

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