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CLINICAL RESEARCH

Accuracy of computed tomography in detection of great vessel stenosis or hypoplasia before superior bidirectional cavopulmonary connection: Comparison with cardiac catheterization and surgical findings

Performance du scanner dans la détection de la sténose ou l'hypoplasie des gros vaisseaux préalable à la connexion cavo-pulmonaire bidirectionnelle supérieure : comparaison avec le cathétérisme cardiaque et les données peropératoires

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Abbreviations: BCPC, Bidirectional Cavopulmonary Connection; CI, Confidence Interval; CT, Computed Tomography; DAP, Dose-Area Product; DLP, Dose-Length Product; IQR, Interquartile Range.

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KEYWORDS
Single ventricle;
Computed tomography;
Bidirectional Glenn

Summary
Background. — Cardiac catheterization is the gold-standard modality for investigation of cardiovascular morphology before bidirectional cavopulmonary connection, but requires general anaesthesia and is associated with procedural risk.
Aims. — To assess the diagnostic accuracy and safety of computed tomography in diagnosing great vessel stenosis/hypoplasia compared with cardiac catheterization and surgical findings.
Methods. — Twenty-seven patients (10 after Norwood stage I) underwent computed tomography before surgery between January 2010 and June 2016; 16 of these patients also underwent cardiac catheterization. Proximal and distal pulmonary artery, aortic isthmus and descending aorta measurements, radiation dose and complications were compared via Bland-Altman analyses and correlation coefficients.
Results. — The accuracy of computed tomography in detecting stenosis/hypoplasia of either pulmonary artery was 96.1% compared with surgical findings. For absolute vessel measurements and Z-scores, there was high correlation between computed tomography and angiography at catheterization ($r = 0.98$ for both) and a low mean bias (0.71 mm and 0.48; respectively). The magnitude of intertechnique differences observed for individual patients was low (95% of the values ranged between −0.9 and 2.3 mm and between −0.7 and 1.7, respectively). Four patients (25%) experienced minor complications from cardiac catheterization, whereas there were no complications from computed tomography. Patients tended to receive a higher radiation dose with cardiac catheterization than with computed tomography, even after exclusion of interventional catheterization procedures (median 2.5 mSv [interquartile range 1.3 to 3.4 mSv] versus median 1.3 mSv [interquartile range 0.9 to 2.6 mSv], respectively; $P = 0.13$). All computed tomography scans were performed without sedation.
Conclusions. — Computed tomography may replace cardiac catheterization in identification of great vessel stenosis/hypoplasia before bidirectional cavopulmonary connection when no intervention before surgery is required. Computed tomography carries lower morbidity, can be performed without sedation and may be associated with less radiation.

MOTS CLÉS
Ventricule unique ;
Scanner cardiaque ;
Glenn bidirectionnel

Résumé
Contexte. — Le cathétérisme est l'examen de référence pour l'évaluation morphologique cardiovasculaire avant la dérivation cavopulmonaire bidirectionnelle supérieure, mais il nécessite une anesthésie générale et comporte des risques liés à la procédure.
Buts. — Notre objectif était de comparer la performance diagnostique et la sécurité de la tomodensitométrie cardiaque avec le cathétérisme et l'examen peropératoire pour le diagnostic des sténoses et/ou hypoplasies des gros vaisseaux en comparaison.
Méthodes. — Vingt-sept patients (10 après une intervention de Norwood) ont eu une tomodensitométrie cardiaque avant la chirurgie entre janvier 2010 et juin 2016. Seize d'entre eux ont également eu un cathétérisme cardiaque. Les mesures proximales et distales des artères pulmonaires, de l'isthme aortique et de l'aorte descendante, ainsi que l'irradiation et les complications per procédurales ont été comparées entre les 2 investigations à l'aide des coefficients de Bland-Altman et de corrélation.
Résultats. — La précision de la tomodensitométrie pour la détection de sténoses/hypoplasies des artères pulmonaires était 96,1 % en comparaison avec l'évaluation peropératoire lors de la chirurgie. Pour les mesures des vaisseaux et les Z-scores, il y avait une excellente corrélation entre la tomodensitométrie et l'angiographie lors du cathétérisme (0,98 pour les 2), peu de biais (0,71 mm vs 0,48, respectivement). La difference de magnitude observée entre les 2 techniques pour chaque individu est faible ainsi que reflètent les intervalles a 95 % des valeurs mesurées allant de −0,9 à 2,3 mm et de −0,7 à 1,7, respectivement. Quatre patients ont souffert de complications mineures lors du cathétérisme alors qu'il n'y a eu aucune complication liée à la tomodensitométrie. Les patients ont eu tendance à recevoir une irradiation plus importante lors du cathétérisme que lors de la tomodensitométrie (médiane 2,5 mSv [IQR 1,3 à 3,4 mSv] vs 1,3 mSv [0,9 à 2,6 mSv], respectivement; $P = 0{,}13$). Toutes les tomodensitométries ont été effectuées sans sédation.
Conclusions. — La tomodensitométrie peut potentiellement remplacer le cathétérisme pour identifier les sténoses et/ou hypoplasies des gros vaisseaux avant la dérivation cavopulmonaire bidirectionnelle, à condition qu'il n'y ait pas besoin d'intervention. La tomodensitométrie a une morbidité moindre, ne nécessite pas de sédation et elle est moins irradiante.

Background

Patients with single ventricle physiology require detailed vascular imaging before superior bidirectional cavopulmonary connection (BCPC). Unrecognized stenoses or hypoplasia of the great vessels can have deleterious consequences for early or medium-term outcome [1,2].

The majority of these single ventricle patients undergo initial neonatal surgery to secure a balanced pulmonary and systemic blood flow, with a systemic-to-pulmonary shunt, pulmonary artery banding or a complex procedure, such as the Norwood stage I operation, incorporating extensive reconstructive surgery to the aortic arch. These procedures may result in hypoplasia or stenosis of the pulmonary arteries and/or aorta [3,4].

Cardiac catheterization is the gold-standard imaging modality in the assessment of vascular anatomy before BCPC; it also provides the opportunity to measure pulmonary artery pressure and resistances, and to perform interventional procedures [5]. Cardiac magnetic resonance imaging is also used extensively [6,7], but requires general anaesthesia, which carries a risk of complications [8], and is time consuming and expensive. Although echocardiography is the first-line imaging technique of choice, it has been shown to be unreliable in detecting great vessel stenosis, even with sedation [9,10].

Recent advances in cardiac computed tomography (CT) have led to such high spatial and temporal resolution that it has become an ideal imaging modality for preoperative planning for patients of all ages with congenital heart disease, without the need for general anaesthesia [11]. In our institution, we often use CT angiography to clarify the anatomy of congenital heart defects in all age groups, from neonates to adults [12]. Patients with single ventricle physiology undergo cardiac catheterization and/or CT angiography according to clinical requirements.

We hypothesized that if haemodynamic assessment or intervention is not required, CT might be superior to cardiac catheterization in patients before BCPC, with equivalent diagnostic accuracy and decreased complications (both procedural and anaesthetic). Therefore, the aim of this study was to assess the accuracy and safety of CT compared with cardiac catheterization and surgical findings in detecting stenosis of the pulmonary arteries and aorta.

Methods

We performed a retrospective study in all patients who underwent BCPC and had a previous CT scan in our institution between January 2010 and June 2016. The study conforms to the ethical guidelines of the 1975 Declaration of Helsinki.

The primary endpoint was to assess the accuracy of CT in detecting pulmonary artery and aortic arch stenosis, and to compare this with surgical and angiographic findings at cardiac catheterization. Secondary outcomes were to assess the safety of CT, in terms of ionizing radiation, the need for anaesthesia or sedation and complications, compared with cardiac catheterization.

Patient demographic data, the need to use anaesthesia or sedation for CT and cardiac catheterization, complications, ionizing radiation exposure, amount of contrast, perioperative findings and follow-up data were collected from patients' medical and imaging records.

CT technique

CT scans were performed on a dual source (2 × 128 slice) CT scanner (Somatom Definition Flash; Siemens Healthcare, Erlangen, Germany), using weight-based paediatric CT angiography protocols. Ungated high-pitch spiral acquisitions (''flash thoracic CT angiogram'') were obtained after administration of intravenous contrast medium (Visipaque™ 320 [iodixanol]; GE Healthcare, Cork, Ireland) up to 2 mL/kg, pump-injected at 0.5—5 mL/s depending on cannula size and location) from the lung apices to immediately below the diaphragm. Tube voltage was set at 80 or 100 kV, depending on patient weight, and tube current was automatically modulated according to the scan projection radiograph. Images were reconstructed at 0.75 mm slice thickness at an interval of 0.5 mm.

General anaesthesia is not used for any elective patient undergoing a CT scan in our institution. When the patient was small enough, gross patient movement was limited using a vacuum immobilization device. It is our local practice to attempt CT, in all instances, without sedation. If the child is too anxious for unsedated CT, light sedation is administered.

Cardiac catheterization technique

Cardiac catheterization was performed under general anaesthesia for all cases. Pulmonary arteries and the aorta were catheterized antegradely through a femoral vein access or through a femoral artery access in patients with Blalock-Taussig shunt. If necessary, additional jugular vein access was used. Pulmonary angiography was performed using orthogonal biplane angiography (Axiom Artis dBC; Siemens Healthcare, Erlangen, Germany) at a frame rate of 30 frames/s after injection of contrast (Visipaque™ 320; 1—1.5 mL/kg), with an injection time of 1—1.5 seconds.

Measurement of the pulmonary arteries and aorta

CT and cardiac catheterization angiographic measurements were performed by two experienced observers blinded to the other modality findings (S. K. has 12 years and L. V.-G. has 5 years of paediatric cardiology experience). Inter- and intraobserver variabilities were compared by reanalysing 10 randomly chosen cases.

On CT, each vessel was viewed on multiplanar reconstructions, and measured in two perpendicular dimensions (OsiriX software, version 6.5; Pixmeo, Bernex, Switzerland), and an average was then calculated. Using cardiac catheterization angiograms, each vessel segment was measured on still two-dimensional images (Medcon database software) in postero-anterior or anterior oblique and lateral angiograms paused at maximal vessel diameter. Examples of CT and cardiac catheterization measurement are shown in Fig. 1A—D.

For measurement of the pulmonary arteries, two different sites were considered: proximal (immediately after the pulmonary trunk bifurcation or proximal to the modified Blalock-Taussig); and distal (before the pulmonary lobar bifurcation). The aorta was measured at two sites: at the

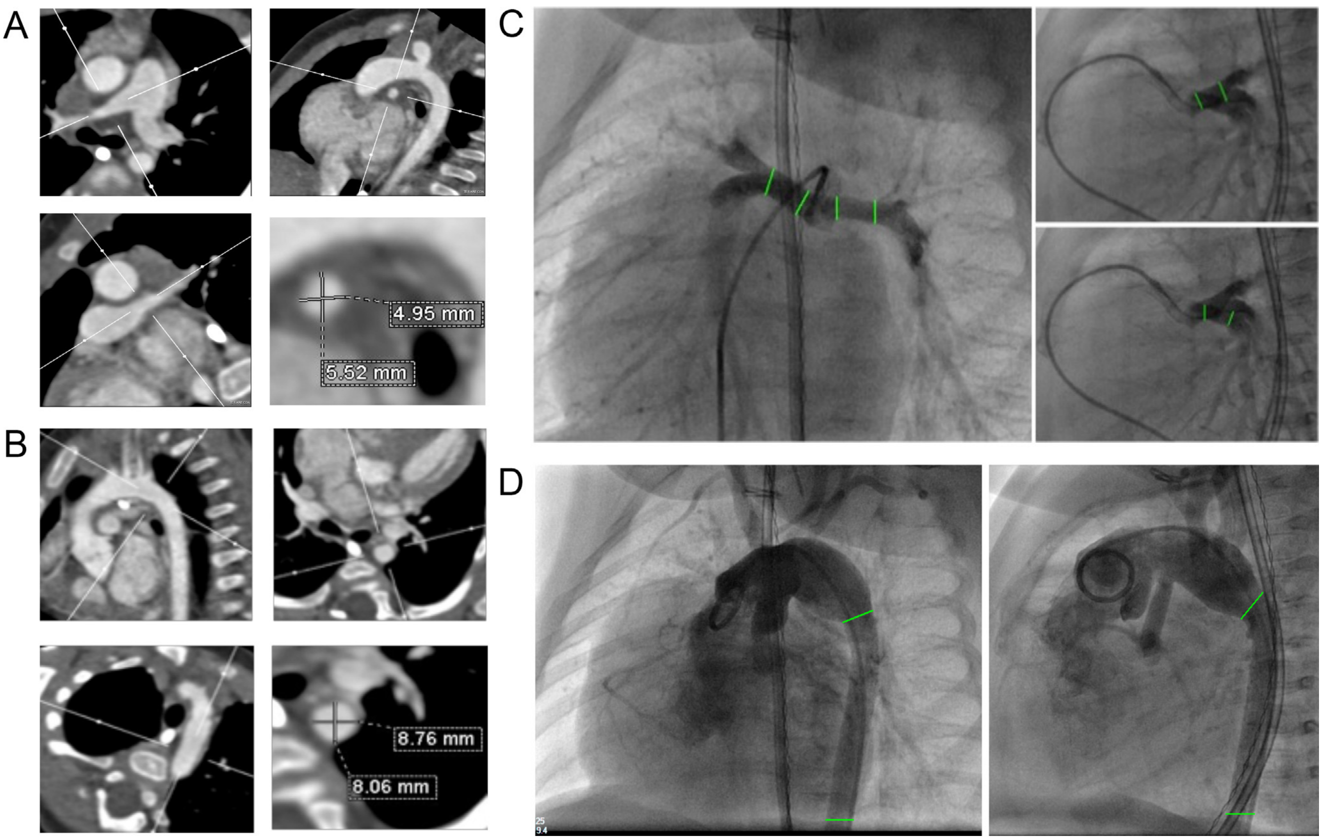

Figure 1. A, B. Measurement of the right pulmonary artery (A) and the aorta (B) on computed tomography multiplanar reformat. C, D. Measurement of the pulmonary arteries (C) and the aorta (D) from angiography at catheterization.

level of the aortic isthmus (the narrowest part of aorta distal to the subclavian artery); and at the level of the diaphragm.

During four cardiac catheterizations, we experienced difficulty in obtaining adequate measurements with lateral angiograms, and considered only the measurement in the posteroanterior or oblique view for the analysis.

Z-scores for CT and catheterization measurements were calculated for each vessel segment using published CT normal values [13,14].

Pulmonary artery branches and the aortic isthmus were considered stenotic when the ratio of proximal and distal measurements was < 0.75 [4,15]. Pulmonary artery branches were classified as hypoplastic if the Z-score of both proximal and distal parts was < -2.

Patients underwent both CT and cardiac catheterization between any initial palliation and BCPC based predominantly on clinical indication. Therefore, in some patients there was a longer time interval between the two investigations. For the purposes of vessel segment comparison, we compared only patients who had CT and cardiac catheterization within 120 days (median 41 days, [interquartile range (IQR) 31 to 63 days]).

Measurement of the radiation dose

CT angiography

In CT, the examination dose-length product (DLP) for a 32 cm diameter dose phantom, expressed in units of mGycm, and patient weight were collected. Effective dose was estimated using the conversion factors from the DLP (16 cm dose phantom) to effective dose [16] available for the chest region for phantoms of a newborn, 1-year-old and 5-year-old [17]. Conversion factors for a 32 cm diameter dose phantom were derived by multiplying published conversion factors by 2.3 (taken from the manufacturer's dosimetry data). The conversion factor for each patient was selected from the best-fit logarithmic function to these conversion factors, plotted against phantom weight. The effective dose for each patient was then estimated by multiplying the DLP by this factor.

Cardiac catheterization

In cardiac catheterization, total dose-area product (DAP), expressed in units of μGym^2, and patient weight were collected. Effective dose [16] was estimated using conversion factors from DAP to effective dose. Conversion factors were derived using the PCXMC computer programme, version 2.0 (Stuk, Helsinki, Finland). The interventional unit was modelled using the typical imaging geometries and X-ray beam qualities used for children in fluoroscopy and acquisition modes. The most frequently used projections were simulated for phantoms of a newborn, 1-year-old and five-year-old. A typical conversion factor for each of the phantoms was estimated by averaging over projections and modes. The conversion factor for each patient was selected from the best-fit logarithmic function to these data plotted against phantom weight. The effective dose for each patient was then estimated by multiplying the total DAP by this factor.

Surgical assessment

CT and/or cardiac catheterization findings were presented and discussed with the surgeons before surgery. In all patients, the right pulmonary artery was dissected as part of the BCPC, and was visualized directly by the surgeon. The left pulmonary artery was explored using a Hegar dilator.

Branch pulmonary artery stenosis was diagnosed if the narrowed segment was < 25% of the diameter of the adjacent segment; hypoplasia was diagnosed if the Z-score diameter of the vessel was < −2.

Statistical analysis

Categorical data are presented as frequencies (percentages). Population data are expressed as median and full range, and all CT and catheterization absolute measurements and Z-scores are expressed as median [IQR]. The differences in matched radiation doses and amount of contrast between CT and cardiac catheterization were tested using the Wilcoxon signed-rank test. Sensitivity, specificity, positive and negative predictive values and accuracy were calculated to assess the ability of CT to detect vessel stenosis/hypoplasia against surgical findings. Accuracy was calculated as the ratio of detected existing defaults divided by all existing defaults, i.e. true positive findings/(true positive + false negative findings). True positive findings were considered when branch pulmonary stenosis/hypoplasia found on CT was confirmed by the surgeon at the time of surgery, and false negative when branch pulmonary stenosis/hypoplasia found at the time of surgery had not been detected at presurgical CT scan. Bland-Altman analysis [18] was performed, and Pearson correlation coefficients were calculated for comparison of all CT and catheterization measurements. Intra- and interobserver reproducibilities were assessed using intraclass correlation. Statistical significance was set at $P < 0.05$, and the analysis was performed using Stata software, version 14.1 (StataCorp LP, College Station, TX, USA).

Results

Eighty-eight children underwent BCPC between 01 January 2010 and 30 June 2016. Before BCPC, 57 patients (65%) underwent catheterization, 27 patients underwent CT (16 of whom also underwent catheterization) and four patients were imaged only with echocardiography.

We studied the group of 27 patients who underwent CT angiograms with or without cardiac catheterization before BCPC; 17 (63%) were male. The underlying diagnoses were as follows: hypoplastic left heart syndrome ($n = 9$); double inlet left ventricle ($n = 4$); tricuspid atresia ($n = 3$); double outlet right ventricle ($n = 2$); left atrial isomerism ($n = 2$), including complete atrioventricular septal defect in one case and hypoplastic left ventricle with double outlet right ventricle in another case; right atrial isomerism ($n = 2$), with complete atrioventricular septal defect and double outlet right ventricle in both cases; double inlet right ventricle ($n = 1$); hypoplastic right ventricle and aortic arch ($n = 1$); hypoplastic tricuspid valve and right ventricle ($n = 1$); pulmonary atresia with intact ventricular septum ($n = 1$); and transposition of the great arteries with pulmonary stenosis ($n = 1$). Three patients had no intervention before BCPC. The Norwood operation with Sano shunt was performed in six patients, Norwood with Blalock-Taussig shunt in four patients, Blalock-Taussig shunt alone in seven patients (right in two, left in one and bilateral in four), pulmonary artery banding in five patients and balloon septostomy in two patients.

BCPC was performed at a median age of 229 days [range, 96 days to 3.2 years], and at a median weight of 8.3 kg [range, 4.0—13.4 kg]. Bilateral BCPC was performed in six patients.

Comparison of CT and cardiac catheterization

All CT scans were performed electively, and none of the patients required sedation.

Complications

Four patients (25%) experienced minor complications during or after cardiac catheterization: transitory complete heart block ($n = 1$); femoral artery pseudoaneurysm treated with thrombin injection ($n = 1$); transient loss of femoral artery pulse requiring heparin infusion ($n = 1$); and femoral vein thrombosis ($n = 1$). There were no complications from the CT scan.

Interventional procedures

At the time of cardiac catheterization, three patients underwent interventional procedures: balloon angioplasty of stenotic pulmonary arteries that was anticipated by echocardiography; balloon angioplasty of recoarctation of the aorta anticipated by CT; and aortopulmonary collaterals occlusion.

Radiation doses and contrast

Sixteen patients underwent both CT and cardiac catheterization (15 had radiation dose data available). The median CT exam DLP was 19.0 mGycm [IQR 12.0 to 30.0 mGycm]. Conversion factors from DLP (32 cm dose phantom) to effective dose ranged between 0.090 and 0.041 mSv/mGycm for a newborn to a 5-year-old phantom, respectively. Median fluoroscopy time and DAP for cardiac catheterization were 13.9 minutes [IQR 11.1 to 29.7 minutes] and 92.9 μGym^2 [IQR 72.0 to 209.0 μGym], respectively. Conversion factors from DAP to effective dose ranged between 0.032 and 0.008 $mSv/\mu Gym^2$ for a newborn to a 5-year-old, respectively. When comparing CT and cardiac catheterization, we found that the radiation dose and amount of contrast were significantly higher for the cardiac catheterizations ($P = 0.035$ and $P = 0.001$, respectively). However, when the three patients who underwent catheter intervention were excluded, the difference in dose was no longer statistically significant ($P = 0.13$) (Table 1).

Table 1 Comparison of amount of contrast (mL) and effective radiation dose (mSv) between computed tomography and cardiac catheterization.

	CT	Catheterization	*P*
Amount of contrast (mL) ($n = 16$)	15.0 (12.0 to 16.8)	41.0 (30.0 to 65.0)	0.001
Effective radiation dose (mSv) ($n = 15$)	1.2 (1.0 to 1.9)	2.6 (1.6 to 3.5)	0.035
Effective radiation dose (mSv) ($n = 12$)[a]	1.3 (0.9 to 2.6)	2.5 (1.3 to 3.4)	0.13

Vessel dimensions are expressed as median (interquartile range); CT: computed tomography.
[a] Patients undergoing intervention excluded.

Table 2 Computed tomography absolute measurements and Z-scores of the vessel segments, acquired from the whole study group ($n = 27$).

Measurement	Vessel segment	CT
Absolute measurement (mm)	RPA distal	7.4 (6.0 to 9.7)
	RPA proximal	6.5 (5.6 to 7.2)
	LPA proximal	5.6 (4.6 to 8.8)
	LPA distal	7.0 (5.6 to 7.9)
	Aorta isthmus	8.9 (7.6 to 10.8)
	Aorta abdominal	7.8 (6.8 to 8.4)
Z-score	RPA distal	−0.3 (−1.5 to 1.0)
	RPA proximal	−1.2 (−2.0 to −0.5)
	LPA proximal	−2.0 (−2.7 to 0.2)
	LPA distal	−0.9 (−2.0 to −0.1)
	Aorta isthmus	1.3 (0.7 to 2.4)
	Aorta abdominal	0.7 (0.2 to 1.6)

Vessel dimensions are expressed as median (interquartile range); CT: computed tomography; LPA: left pulmonary artery; RPA: right pulmonary artery.

Agreement in vessel diameter measurements

Table 2 shows the absolute measurements and Z-score values in all 27 patients who underwent CT.

Scatter and Bland-Altman plots comparing the absolute measurements and Z-scores are shown in Fig. 2A—D, with high correlation ($r = 0.98$ for both) and low mean bias (0.71 mm and 0.48, respectively). The magnitude of intertechnique differences observed for individual patients was low, as reflected in the fact that 95% of values ranged between −0.9 and 2.3 mm and between −0.7 and 1.7, respectively. The bias tended to be higher when the measurement was higher. Table 3 shows the comparison of absolute measurements and Z-scores, broken down into individual segments.

The intraobserver intraclass correlation coefficients for CT and catheterization measurements were 0.99 (95% confidence interval [CI] 0.98—0.99) and 0.91 (95% CI 0.85—0.95), respectively. The interobserver intraclass correlation coefficient for CT measurements was 0.96 (95% CI 0.93—0.97) compared with 0.95 (95% CI 0.92—0.97) for catheterization.

Comparison of CT and surgical findings

Stenosis or hypoplasia of the pulmonary arteries was identified in 10 patients on the right and 12 patients on the left (seven patients had bilateral stenosis/hypoplasia) by CT. CT identified all cases of stenosis/hypoplasia of the pulmonary arteries. There were two cases in which pulmonary artery hypoplasia (one on the right side and one on the left) was diagnosed by CT, but not confirmed intraoperatively at the time of BCPC. One patient with coarctation of aorta underwent successful balloon angioplasty after CT (this patient's aorta was excluded from the comparison of CT and surgical findings). No other patients were found to have aortic coarctation on CT or catheter angiography and, as such, surgical inspection of the aorta was not performed at the time of surgery in any of the 27 patients identified.

The specificity, sensitivity, positive and negative predictive values and accuracy of CT for detecting stenosis/hypoplasia of the pulmonary arteries are shown in Table 4. The accuracy of CT in detecting stenosis/hypoplasia of either pulmonary artery was 96.1% compared with surgical findings.

During the median follow up of 1.7 years [IQR 161 days to 3.4 years], none of the patients required intervention because of pulmonary artery stenosis or coarctation of the aorta.

Comparison of stays in intensive care unit

Comparison of the mean duration of stay in an intensive care unit between patients undergoing cardiac catheterization ($n = 73$) and the rest of the patients (after CT or echocardiography only, $n = 15$) revealed no significant difference

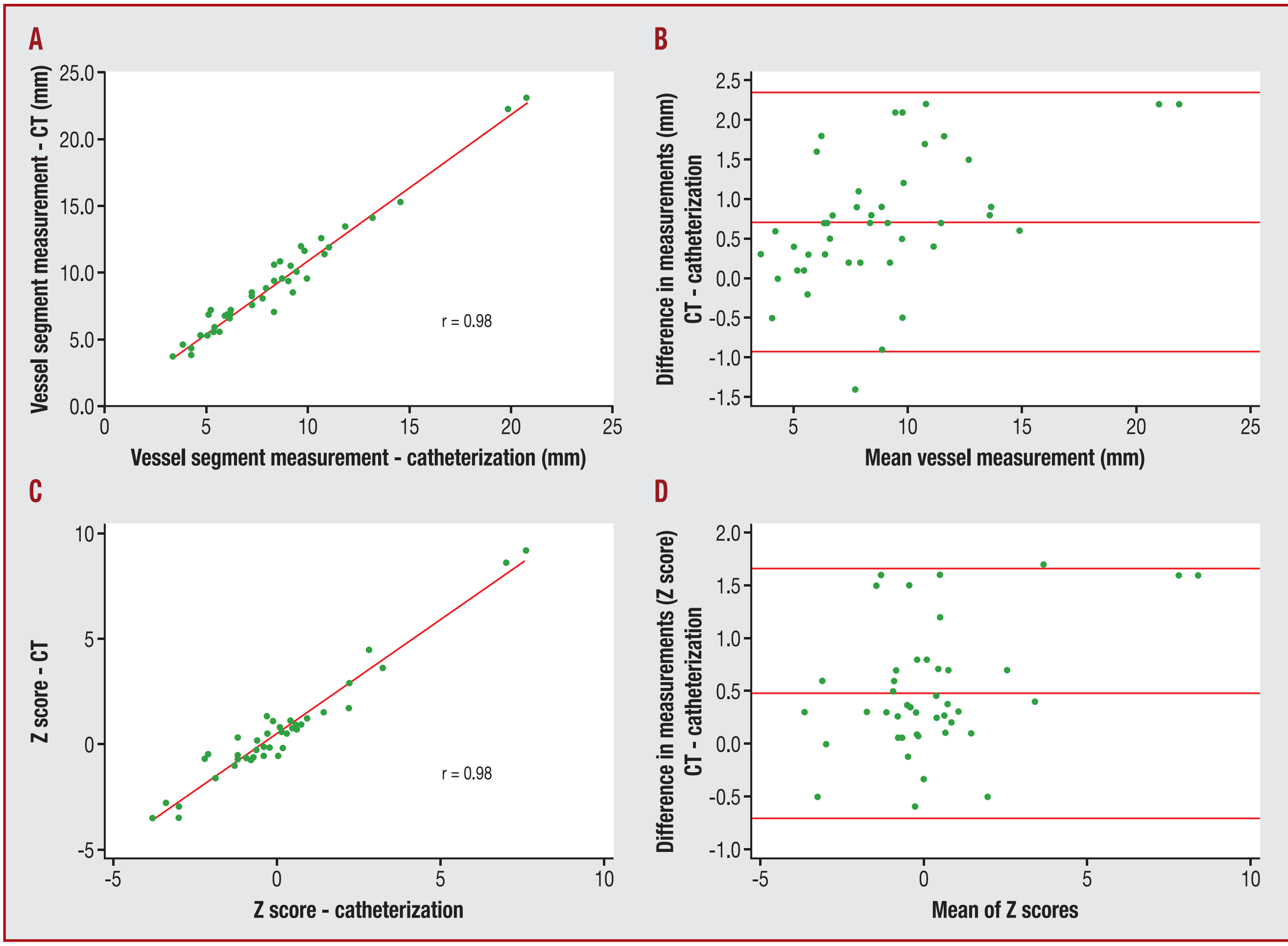

Figure 2. A, B. Scatter plot (A) and Bland-Altman plot (B) comparing absolute vessel segment measurements in patients undergoing both computed tomography (CT) and cardiac catheterization ($n = 10$). C, D. Scatter plot (C) and Bland Altman plot (D) comparing Z-score segment measurements in patients undergoing both CT and cardiac catheterization ($n = 10$). *r*: correlation coefficient.

Table 3 Comparison of each vessel segment measurement by computed tomography and cardiac catheterization.

Vessel	Bias	Lower limit agreement	Upper limit agreement	*r*
RPA (mm)	0.93	−0.7 (−1.4 to 0.0)	2.5 (1.8 to 3.2)	0.98
LPA (mm)	0.87	−0.8 (−1.5 to −0.1)	2.6 (1.9 to 3.3)	0.99
Aorta (mm)	0.44	−1.1 (−1.7 to −0.5)	2.0 (1.4 to 2.6)	0.93
RPA (Z-score)	0.72	−0.5 (−0.9 to −0.2)	1.9 (1.6 to 2.3)	0.96
LPA (Z-score)	0.69	−0.7 (−1.4 to 0.0)	2.1 (1.4 to 2.8)	0.99
Aorta (Z-score)	0.16	−0.4 (−1.0 to 0.2)	0.8 (0.2 to 1.4)	0.89

Limits of agreement from Bland-Altman analyses are displayed with 95% confidence intervals; LPA: left pulmonary artery; *r*: correlation coefficient; RPA: right pulmonary artery.

Table 4 Sensitivity, specificity, negative and positive predictive values and accuracy of computed tomography to detect proximal stenosis and/or hypoplasia for each vessel segment (analysed against surgical findings).

Vessel	Sensitivity	Specificity	NPV	PPV	Accuracy
RPA	90 (56 to 100)	100 (81 to 100)	94 (73 to 100)	100 (66 to 100)	96 (89 to 100)
LPA	92 (62 to 100)	100 (78 to 100)	94 (70 to 100)	100 (72 to 100)	96 (89 to 100)

All data are displayed with 95% confidence intervals; LPA: left pulmonary artery; NPV: negative predictive value; PPV: positive predictive value; RPA: right pulmonary artery.

(6.4 ± 7.1 vs 4.9 ± 4.0 days, respectively; $P = 0.46$). Four patients who underwent cardiac catheterization died (7, 20, 370 and 602 days after BCPC) postoperatively; none of the deaths was related to any undiagnosed coarctation or pulmonary artery stenosis.

Discussion

In the present study, we analysed the accuracy and safety of CT angiography in assessing great artery stenosis or hypoplasia before BCPC. The accuracy of CT in detecting stenosis/hypoplasia was excellent compared with surgery, and no stenosis/hypoplasia was missed using CT. CT and catheterization measurements were similar, the amount of contrast was lower with CT and the radiation dosage was not significantly different between CT and cardiac catheterization. No patients required general anaesthesia or sedation for CT, and procedural complications were only experienced during or after cardiac catheterization.

Detection of great vessel stenosis or hypoplasia using CT versus surgery

We have shown that, in our cohort, CT accurately identified all cases of pulmonary artery stenosis/hypoplasia in patients before BCPC. There were no examples of coarctation of the aorta identified by CT, and thus the aorta was not inspected at the time of surgery. Importantly, during the follow-up period, no intervention was required for pulmonary stenosis or aortic coarctation, and six patients have subsequently undergone successful Fontan completion.

Detection of great vessel stenosis or hypoplasia using CT versus cardiac catheterization

There was high correlation between CT and cardiac catheterization for measurements of the aorta and the pulmonary artery, and a low magnitude of intertechnique differences was observed for individual patients. Furthermore, there was agreement in intra- and interobserver variabilities regarding CT measurements. The three-dimensional dataset allows reconstruction of vessels in any plane, together with isotropic measurements, with a spatial resolution up to 0.3 mm and temporal resolution up to 75 ms [19]. At present, CT is the best non-invasive method for obtaining reproducible data. Cardiac catheterization allows vessel measurements in only two dimensions, and requires general anaesthesia in most centres. Cardiac magnetic resonance imaging allows the acquisition of a three-dimensional dataset, but with lower spatial and temporal resolution than CT (approximately 1.7 mm), and also requires general anaesthesia [20]. Echocardiography is the least reproducible method, as the great arteries may have an oval shape, and measurements are made in only one two-dimensional plane [9].

Radiation dose

Our study group has shown that CT at BCPC carries a low radiation dose. However, the effective dose with cardiac catheterization in our centre was also low, at a median 2.6 mSv (including three patients undergoing interventional catheterization; median 2.5 mSv when these patients were excluded). For comparison, the worldwide average natural background carries an effective radiation dose of 2.4 mSv/year [21], and one simple paediatric chest X-ray carries 0.005 mSv [22]. The estimated lifetime cancer risks [22] for CT scans and cardiac catheterization are approximately 0.01 and 0.03%, respectively. However, the true risk from low-level diagnostic radiation exposure is not clear, and is a topic of much debate. Until more data on the risk of medical radiation exposure are available, exposures should be kept to the minimum possible to obtain sufficiently diagnostic images [23].

The only available comparable study [24] showed a similar effective radiation dose to ours with CT (1.1 mSv), but a significantly higher radiation dose with cardiac catheterization (14.0 mSv). This may be explained by a shorter catheterization time, with a focus on the acquisition of only essential information at our centre. It does illustrate that, in the current era, low radiation exposures are achievable with cardiac catheterization, although at a slightly higher level than with CT.

General anaesthesia and sedation

All patients undergoing cardiac catheterization had general anaesthesia without complications. No anaesthesia or sedation was required for CT. Other centres have reported the use of sedation [24] in patients aged < 1 year, but we have achieved good image quality without any form of sedation, probably as a result of the high-pitch acquisition and the use of vacuum immobilization devices.

Adverse events from the contrast

The complications of low osmolality iodinated contrast are relatively rare, but include hypotension and acute kidney injury, both of which are somewhat dose dependent. Although very rare in children, it should be noted that the risk of hypotension and contrast-induced acute kidney injury during cardiac catheterization increases with higher contrast volumes [25] and with intracardiac administration [26]. Acute contrast-related anaphylactic reactions, although extremely rare, can lead to death from circulatory collapse and severe bronchospasm, even with tiny doses [27]. Nevertheless, no adverse reactions were observed in any of our patients, although the amount of contrast required for cardiac catheterization was significantly greater than for CT.

Imaging work-up before BCPC

We believe that, in most cases, relatively low-cost, low-dose and low-complication CT angiography accurately identifies abnormalities of the great vessels, while overall intracardiac morphology, atrioventricular valve regurgitation, the presence of interatrial communication and ventricular function can be assessed accurately by echocardiography. Our data also showed that there was no significant difference in length of stay in paediatric intensive care units between patients who had catheterization as part of the work-up before BCPC and those who had CT or echocardiography only. Thus, the main indication for cardiac catheterization is likely to be related to interventional procedures, such as balloon angioplasty or abnormal vessel embolization, and indications for such interventional procedures can be diagnosed with a CT scan. The length of time — and therefore radiation, contrast and general anaesthetic doses — required for these catheter measurements and interventions could probably be reduced by preprocedural CT for overall assessment of the cardiovascular anatomy. From our experience, patients with hypoplastic left heart syndrome should still undergo cardiac catheterization before Glenn, before proceeding to the second stage of palliation, because they are very likely to benefit from interventions such as collateral occlusion, balloon dilatation of pulmonary stenosis, stenting of Sano shunt and balloon dilation of aortic arch obstruction. Conversely, other cases of single ventricle (especially tricuspid atresia or double inlet left ventricle) can undergo only cardiac CT.

Study limitations

Besides the retrospective design of our study, the main limitation was the relatively small number of patients. This is a reflection of a gradual increase in the number of patients investigated by CT over a 6-year period. Nevertheless, our results are very encouraging and confirm the value and safety of CT angiography in the preoperative assessment of paediatric patients with complex congenital heart defects.

Finally, conversion of the collated radiation dose metrics to effective dose is inevitably associated with some uncertainties, which could introduce systematic errors of up to 50% in the estimate of the effective dose. Nevertheless, the methodology used for these purposes is widely accepted [16,17].

Conclusions

This study has shown that, in a selected group of patients not requiring intervention before BCPC, CT angiography was accurate in the identification and measurements of great vessel stenosis/hypoplasia. In addition, CT carried a lower radiation exposure and no morbidity. No patient undergoing CT required any sedation. Further multicentre studies are needed to confirm the accuracy of CT for assessment before BCPC.

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Disclosure of interest

The authors declare that they have no competing interest.

References

[1] Feinstein JA, Benson DW, Dubin AM, et al. Hypoplastic left heart syndrome: current considerations and expectations. J Am Coll Cardiol 2012;59:S1—42.
[2] Januszewska K, Kozlik-Feldmann R, Kordon Z, et al. Significance of the residual aortic obstruction in multistage repair of hypoplastic left heart syndrome. Eur J Cardiothorac Surg 2011;40:508—13.
[3] Fraisse A, Colan SD, Jonas RA, Gauvreau K, Geva T. Accuracy of echocardiography for detection of aortic arch obstruction after stage I Norwood procedure. Am Heart J 1998;135:230—6.
[4] Lemler MS, Zellers TM, Harris KA, Ramaciotti C. Coarctation index: identification of recurrent coarctation in infants with hypoplastic left heart syndrome after the Norwood procedure. Am J Cardiol 2000;86 [697—9, A9].
[5] Nakanishi T. Cardiac catheterization is necessary before bidirectional Glenn and Fontan procedures in single ventricle physiology. Pediatr Cardiol 2005;26:159—61.
[6] Brown DW, Gauvreau K, Powell AJ, et al. Cardiac magnetic resonance versus routine cardiac catheterization before bidirectional glenn anastomosis in infants with functional single ventricle: a prospective randomized trial. Circulation 2007;116:2718—25.
[7] Muthurangu V, Taylor AM, Hegde SR, et al. Cardiac magnetic resonance imaging after stage I Norwood operation for hypoplastic left heart syndrome. Circulation 2005;112:3256—63.
[8] Stockton E, Hughes M, Broadhead M, Taylor A, McEwan A. A prospective audit of safety issues associated with general anesthesia for pediatric cardiac magnetic resonance imaging. Paediatr Anaesth 2012;22:1087—93.
[9] Krupickova S, Muthurangu V, Hughes M, et al. Echocardiographic arterial measurements in complex congenital diseases before bidirectional Glenn: comparison with cardiovascular magnetic resonance imaging. Eur Heart J Cardiovasc Imaging 2017;18:332—41.
[10] Krupickova S, Quail MA, Yates R, Gebauer R, Hughes M, Marek J. The comparative role of echocardiography and MRI for identifying critical lesions in patients with single-ventricle physiology, before bidirectional cavopulmonary connection. Cardiol Young 2016;26:1373—82.
[11] Han BK, Rigsby CK, Hlavacek A, et al. 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—92.
[12] Sriharan M, Lazoura O, Pavitt CW, et al. Evaluation of high-pitch ungated pediatric cardiovascular computed tomography for the assessment of cardiac structures in Neonates. J Thorac Imaging 2016;31:177—82.
[13] Akay HO, Ozmen CA, Bayrak AH, et al. Diameters of normal thoracic vascular structures in pediatric patients. Surg Radiol Anat 2009;31:801—7.
[14] Hegde SV, Lensing SY, Greenberg SB. Determining the normal aorta size in children. Radiology 2015;274:859—65.
[15] Stern KW, McElhinney DB, Gauvreau K, Geva T, Brown DW. Echocardiographic evaluation before bidirectional Glenn operation in functional single-ventricle heart disease: comparison to catheter angiography. Circ Cardiovasc Imaging 2011;4:498—505.
[16] 1990 Recommendations of the International Commission on radiological protection. Ann ICRP 1991;21:1—201.
[17] Shrimpton PC, Hillier MC, Lewis MA, Dunn M. National survey of doses from CT in the UK: 2003. Br J Radiol 2006;79:968—80.

[18] Bland JM, Altman DG. Statistical methods for assessing agreement between two methods of clinical measurement. Lancet 1986;1:307—10.
[19] Siemens company website. Available at: https://www.healthcare.siemens.co.uk/.
[20] Steeden JA, Pandya B, Tann O, Muthurangu V. Free breathing contrast-enhanced time-resolved magnetic resonance angiography in pediatric and adult congenital heart disease. J Cardiovasc Magn Reson 2015;17:38.
[21] United Nations Scientific Committee on the effects of atomic radiation. UNSCEAR 2008 Report: sources and effects of ionizing radiation. New York: United Nations 2010 [Available at: http://www.unscear.org/unscear/en/publications.html].
[22] Wall BF, Haylock R, Jansen JTM, Hillier MC, Hart D, Shrimpton PC. Radiation risks from medical X-ray examinations as a function of the age and sex of the patient. Chilton. UK: Health Protection Agency 2011 [Available at: https://pdfs.semanticscholar.org/d90f/22c2c119c618c6822eb094087464e5ca3c4b.pdf].
[23] The 2007 Recommendations of the International Commission on radiological protection. ICRP publication 103. Ann ICRP 2007;37:1—332.
[24] Han BK, Vezmar M, Lesser JR, et al. Selective use of cardiac computed tomography angiography: an alternative diagnostic modality before second-stage single ventricle palliation. J Thorac Cardiovasc Surg 2014;148:1548—54.
[25] Keaney JJ, Hannon CM, Murray PT. Contrast-induced acute kidney injury: how much contrast is safe? Nephrol Dial Transplant 2013;28:1376—83.
[26] ACR Committee on Drugs and Contrast Media. ACR Manual on Contrast Media. Version 10.3 2017 [Available at: https://www.acr.org/-/media/ACR/Files/Clinical-Resources/Contrast_Media.pdf].
[27] Rigby ML. Diagnostic cardiac catheterization and angiography. In: Daubeney PEF, Rigby ML, Niwa K, Gatzoulis MA, editors. Pediatric heart disease. A practical guide. Oxford: Wiley-Blackwell; 2012. p. 46—53.