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Role of dual source multidetector row cardiac computed tomography angiography in diagnosis and management of congenital heart disease patients

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Abstract

Introduction: Primary evaluation of patients with congenital heart disease (CHD) traditionally relies on echocardiography and conventional cardiac angiography (CCA), both of which have potential limitations.
Aim: To test the hypothesis that cardiac computed tomography angiography (CCTA) is useful in the diagnosis and management of these patients.
Material and methods: The 3-year observational, analytical, retrospective, cohort study included a total of 111 tomographic studies of patients with congenital heart disease. Computed tomography scans were read twice and medical records were reviewed. The Aristotle complexity was assessed as well as and the contribution of new data in relation to clinical suspicion and diagnostic change was evaluated by two expert readers who were blinded for clinical outcome in consensus reading. The confidence interval was set at 95% and a *p*-value of < 0.05 was used as the cutoff for statistical significance.
Results: In total, 111 patients were included (56 men and 55 women) with a mean age of 7.2 years (1 day–71 years). The therapeutic procedure was performed without additional tests in 85.8% of patients. New findings were observed in 60.4% of patients and a subsequent change in management in 46.9%. New unexpected findings in CCTA prompted changes in management in 86.8% of patients. There were no significant differences in age between patients with new findings vs. patients without such findings in CCTA suggesting that CCTA-supported diagnosis of CHD is independent of age.
Conclusions: Use of dual-source cardiac computed tomography yields good diagnostic performance in congenital heart disease, prompts changes in management in more than one-third of patients, and reveals new findings in relation to the presumed diagnosis in most patients.
Key words: congenital heart disease, cardiac computed tomography angiography, multidetector row computed tomography.

Streszczenie

Wstęp: Pierwotna ocena pacjentów z wrodzonymi wadami serca tradycyjnie opiera się na echokardiografii i konwencjonalnej angiografii, przy czym obie te metody wiążą się z pewnymi ograniczeniami.
Cel: Sprawdzenie użyteczności angiografii tomografii komputerowej w diagnostyce i postępowaniu u tych pacjentów.
Materiał i metody: Trzyletnie obserwacyjne, analityczne, retrospektywne badanie kohortowe objęło łącznie 111 badań tomograficznych pacjentów z wrodzonymi wadami serca. Obrazy tomografii komputerowej odczytano dwukrotnie i dokonano przeglądu dokumentacji medycznej. Stopień złożoności planowego zabiegu według systemu Aristotle oraz wpływ nowych danych w odniesieniu do podejrzeń klinicznych i zmiany rozpoznania oceniało dwóch specjalistów nieznających wyniku klinicznego. Przedział ufności ustalono na 95%, a jako graniczną wartość istotności statystycznej przyjęto *p* < 0,5.
Wyniki: W badaniu wzięło udział łącznie 111 pacjentów (56 płci męskiej i 55 płci żeńskiej) w wieku średnio 7,2 roku (od 1 dnia do 71 lat). Zabieg leczniczy wykonano bez dodatkowych badań u 85,8% pacjentów. Nowe nieoczekiwane wyniki angiografii tomografii komputerowej uzyskano u 60,4% pacjentów i spowodowały one zmianę strategii leczenia u 46,9% pacjentów. Spośród chorych z nowymi danymi u 86,8% nastąpiła zmiana postępowania. Ponadto nie stwierdzono istotnych różnic wieku między grupą chorych z nowymi danymi a grupą bez nieoczekiwanych znalezisk w tomografii komputerowej, co sugeruje, że rozpoznanie wrodzonych wad serca z zastosowaniem tomografii nie zależy od wieku.
Wnioski: Zastosowanie dwuźródłowej tomografii komputerowej serca jest skuteczną metodą diagnostyczną w przypadku wrodzonych wad serca. Powoduje zmianę postępowania u ponad 1/3 pacjentów i ujawnia nowe fakty w stosunku do zakładanego rozpoznania u większości pacjentów.
Słowa kluczowe: wrodzone wady serca, angiografia tomografii komputerowej serca, wielorzędowa tomografia komputerowa.

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Introduction

Congenital heart disease (CHD) is defined as any structural heart defect with potential clinical significance. Its incidence ranges between 4/1000 live births and 12/1000 live births and the most common CHDs are major congenital defects. The majority of CHDs are due to abnormalities that develop between 3 and 10 weeks of gestation and are dynamic entities that have a different course in each patient. There is a progressive decrease in mortality because of improvement of diagnosis and treatment and thus an increase in survival (approximately 85–90% of CHD patients reach adulthood) [1]. Although it is difficult to determine the influence of imaging techniques on the increase in survival, there has been a demonstrated increase in the detection of CHD following the introduction of multidetector computed tomography (MDCT). The diagnostic information obtained using low-risk techniques is essential for optimal medical and surgical management [2–4] and perioperative imaging is useful in assessing procedural success [5, 6]. Continuing advances in medical imaging have made it possible to diagnose cardiovascular disease in any patient by the use of different methods involving a wide variety of technical requirements, benefits, limitations, and costs. The correct application of each method requires a well-integrated group of experts who collaborate with the clinical diagnostic services [7, 8]. There is a trend toward the decreased use of diagnostic catheterization despite it being the reference diagnostic technique, and its use is now mainly reserved for therapeutic decision-making [9]. The incorporation of cardiac computed tomography angiography (CCTA) led us to reflect on the capacity of this new technique to provide answers to the clinical problems of CHD patients and evaluate its implications for their subsequent management. This article assesses the impact of the introduction of CCTA on the management of CHD patients.

Aim

As far as we know, this is the first study to analyze the clinical application of CCTA in Thailand of mainly pediatric CHD patients, in relation to the reports submitted to clinicians, and to determine the role, accuracy, and safety of this technique for this group of patients.

Material and methods

Patient population

This retrospective study included 111 patients with diagnosis of congenital heart disease by clinical data and initial echocardiography who underwent CCTA between February 2013 and December 2016. The indication for CCTA was pre- or postoperative evaluation of congenital heart disease patients, which is considered an appropriate indication for CCTA, based on the criteria of the American College of Cardiology (ACC) [10]. Exclusion criteria for CCTA included the presence of renal failure and a history of allergic reaction to iodine-containing contrast agents. The present study was approved by the Ethics Committee of the Faculty of Medicine and informed consent was obtained from all patients.

Dual-source MDCT scanning protocol

Short-term IV sedation is often required, particularly in children under 5 years of age, and the imaging can be performed during quiet breathing. Imaging was performed by using a dual-source MDCT scanner (Somatom Definition; Siemens Healthcare, Forchheim, Germany). The radiation dose is kept to a minimum by reducing the kilovoltage and tube current appropriately. For children weighing less than 10 kg, 10–19 kg and 20–30 kg, we use 80 kV and 80 mAs, 80 kV and 100 mAs, and 100 kV and 120 mAs, respectively. Other cardiac CT angiographic parameters were as follows: number of X-ray tubes, two; collimation, 128 detector rows of 0.5 mm each, with double sampling by using rapid alteration of the focal spot in the longitudinal direction (*z* flying focal spot) [11], rotation time ms, 330; tube voltage, 100 kV; and full tube current. Prior to scanning, the pitch was set automatically by the scanner software. Depending on heart rate, pitch was set between 0.2 and 0.43. Automated dose regulation methods such as CARE dose 4D (Siemens Healthcare) may be used to reduce the radiation. Image acquisition was performed during inspiratory breath-hold for older children or adult. To familiarize the patient with the protocol, breath-holding was practiced before the examination. A bolus of iodinated contrast material (350 mg/ml, Omnipaque; GE Healthcare) at a dose of 1.5 ml/kg with a dual-head power injector at a rate of 1.5–2.0 ml/s for a 22-gauge cannula, 3.0 ml/s for a 20-gauge cannula and 4.0–5.0 ml/s for a 18-gauge cannula followed by 10–20 ml of saline flush at the same rate as that of the contrast injection was applied. For timing purposes, automated bolus-tracking software was used, starting the scan automatically 6 s after contrast agent density in the descending aorta reached a predefined threshold of 130 HU [12]. The entire volume of the heart and pulmonary arteries was covered during one breath-hold in approximately 5 s with simultaneous recording of the ECG trace. Patients were scanned in the supine position. The CCTA is performed from the thoracic inlet level to the diaphragm. When there is suspicion of anomalous pulmonary venous drainage, the scan can be extended down to the lower border of the liver.

Cardiac computed tomography angiography image analysis

Cardiac computed tomography angiography image analysis was performed by two cardiovascular and thoracic radiologists in consensus (with respectively 11 and 10 years of experience in examining cardiovascular and thoracic CT scans) and blinded to the clinical data and the results of CCA data. First, all axial image data are evaluated using a 3D post processing workstation with Syngo software (Siemens Healthcare). Various image reformatting techniques including curved planar reconstruction, maximum intensity projection (MIP), minimum intensity projection, and volume-rendering technique (VRT) are used to obtain

Tab. I. Most common specific diagnoses in cardiac computed tomography angiography

Variable	Number of patients
Pulmonary atresia with ventricular septal defect	32
Tetralogy of Fallot	5
Transposition of great arteries	12
Anomalous pulmonary venous return	10
Coarctation of aorta	4
Interrupted aortic arch	2
Anomalous coronary artery	16
Double outlet right ventricle	5
Truncus arteriosus	3
Post bidirectional Glenn shunt	8
Tricuspid atresia	2
Post-arterial switch operation	2
Post-Fontan operation	10

Tab. II. Change in therapeutic approach

Result	Patients, *n* (%)	Total (%)
No change in management:		
Known findings	22 (19.8)	53.1
New findings	37 (33.3)	
Change in management:		
New findings	30 (27.1)	46.9
New diagnosis	22 (19.8)	

all the clinically relevant information. Curved multi-planar reformatting and MIP are primarily used to evaluate curved structures such as the pulmonary arteries and major aorto-pulmonary collateral arteries (MAPCAs). Minimum intensity projection is used to evaluate the airway and lung parenchyma. For 3D reformatting of the complex anatomy, VRT is used. Thin-section multiplanar reformatting is used for quantitative analysis of the structure in question.

Data collected

Population data: age, sex, date of test. Clinical information contained in the test order: purpose and justification of the test and pre- or post-treatment study.

Clinical history: additional pre- and post-CCTA. The complexity of heart disease [13, 14] and of each patient was calculated (Aristotle score) [15, 16]. Anatomical findings were coded by group: aortic disease, pulmonary tree, heart disease, anomalous pulmonary venous drainage, complex CHD, post-procedural complications, pulmonary hypertension, airways, and unexpected findings. We assessed whether the findings prompted a change in clinical management, treatment, or patient lifestyle (scored as 1, known findings; 2, new findings without change in management; 3, new findings with a change in management; 4, change in diagnosis). We also assessed concordance between the presumptive diagnosis and the diagnostic test: existence of expected or unexpected findings. New findings were defined as any anatomical abnormality not found in previous tests.

A change in diagnosis: changes in CHD complexity, chamber sequence, or in previously diagnosed intracardiac or great vessel defects.

Changes in treatment: from surgery to catheterization, from catheterization to surgery, or changes in the catheterization or surgical technique used.

– No changes in treatment.
– No changes in diagnosis.

Unexpected findings were defined as findings that were not predicted by the known natural history of the main anatomical abnormality or changes in its natural history.

Statistical analysis

Continuous data were expressed as mean ± SD. Statistical analyses were performed using SPSS software version 16 (SPSS, Inc., Chicago, IL, USA). A level of $p < 0.05$ was considered statistically significant and all reported *p*-values were two-sided. Means were compared using the unpaired *t*-test, and the Mann-Whitney rank sum test was used when data were not normally distributed. Qualitative variables were compared using the Pearson χ^2 test or Fisher exact test.

Results

In total, 111 patients were included (56 men and 55 women) with a mean age of 7.2 years (1 day–71 years). In total, 89.1% of patients did not undergo further studies. The specific diagnoses in cardiac computed tomography angiography are lists as Table I. The therapeutic procedure was performed without additional tests in 85.8% of patients. Additional catheterization was performed in 10.8% of patients. Of the patients studied, 90% had moderate to severe CHD and 80.2% had a level of complexity = 9.1 (level III) according to the Aristotle score. New findings were observed in 60.4% of patients. Mutually exclusive dichotomous categories were as follows: no change in management (53.1%), subsequent change in management (46.9%), of which 19.8% were due to changes in diagnosis (Tab. II; Figs. 1 and 2). Regarding disease, 44.8% of new findings were complex CHD, with changes in treatment in 17.9% ($p < 0.001$), and changes in diagnosis in complex CHD (14.9%), aortic disease (7.5%) and coronary artery disease (5.9%) (Tab. III). Regarding CHD complexity, there were new findings in 81.8% of patients with severe CHD, with changes in management in 31.2%. There were changes in diagnosis in patients with moderate CHD (18.9%; $p < 0.05$) and changes in diagnosis in patients with greater complexity (mean score: 3.25; $p < 0.05$). All unexpected reports (100%) described new findings prompting changes in management in 86.8% of patients and a new diagnosis in 47.2% ($p < 0.001$). Patient progression after cardiac computed tomography angiography in shown in Table IV.

Discussion

The present study demonstrated that cardiac computed tomography angiography (CCTA) is an excellent diagnostic

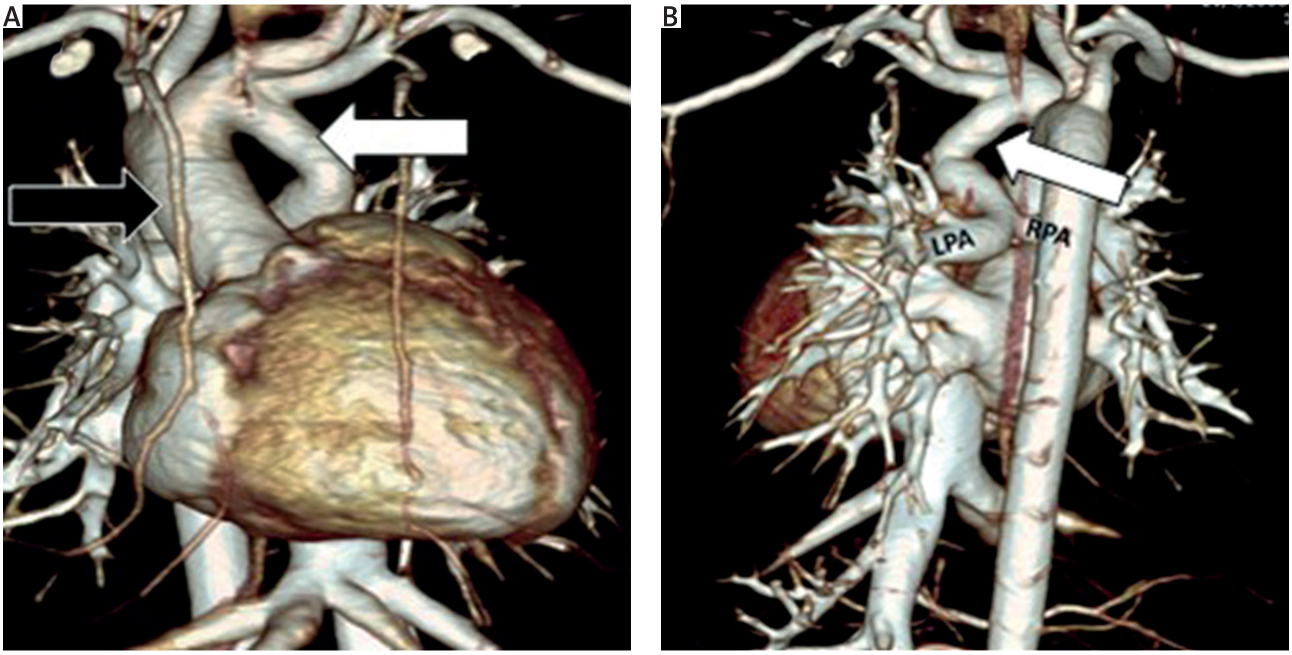

Fig. 1. Cardiac computed tomography angiography (CCTA) of a 7-year-old boy with presumed diagnosis of tetralogy of Fallot. The CCTA showing new unexpected findings as pulmonary atresia with ventricular septal defect (PA-VSD), right-sided aortic arch (**A**; black arrow) and left-sided patent ductus arteriosus (**A** and **B**; white arrow) supplying pulmonary arteries causing a change in diagnosis which changed the therapeutic management from total corrected TOF to staged repair of pulmonary atresia with VSD

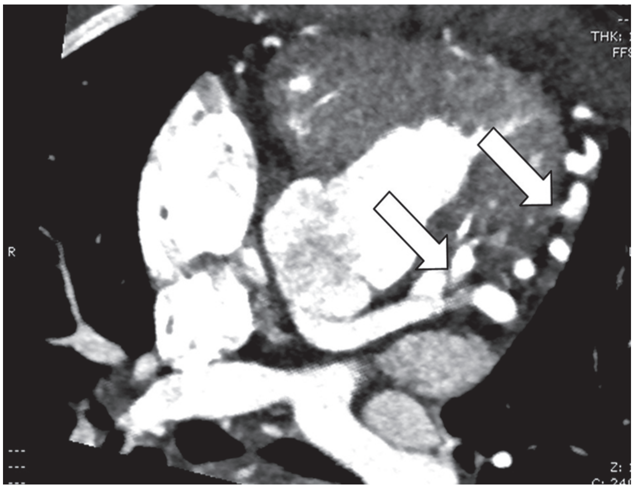

Fig. 2. Cardiac computed tomography angiography (CCTA) of a 5-year-old girl with presumed diagnosis of pulmonary atresia with VSD. The CCTA showing new unexpected findings as single coronary artery and coronary artery fistula from left anterior descending artery to right ventricle (arrows), which changed the therapeutic management

tool for the comprehensive assessment of patients with congenital heart disease using dual-source MDCT. These results are in line with previous published data on the performance of the older generation CT scanner [17–19]. Regarding all unexpected findings, there was a change in management in 46.9% of patients. In an MRI study, Secchi *et al.* [20] found that unexpected reports were associated with more clinically important categories and 85% of unexpected reports

Tab. III. New findings by disease

Disease	New findings without change in management	New findings with change in management	Change in diagnosis
CCHD	18	12	10
APVC	2	8	2
Pulmonary artery disease	8	5	1
Aortic disease	1	3	5
Coronary disease	8	2	4
Total	37	30	22

CCHD – complex congenital heart disease, APVC – anomalous pulmonary venous connection. Data are expressed as number of patients.

Tab. IV. Patient progress after cardiac computed tomography angiography

Variable	Number of patients (%)
Discharge	4 (3.6)
Medical therapy	46 (41.4)
Diagnostic catheterization	12 (10.8)
Therapeutic catheterization	18 (16.2)
Surgery	31 (28)

prompted a change in treatment, lifestyle, or diagnosis. No significant differences were found between study periods, indicating that clinicians used consistent criteria when interpreting the test reports. Tsai-Goodman *et al.* [3] found that 70.3% of patients who underwent MRI did not require additional catheterization. In the present study, 89.2% of patients did not require additional diagnostic studies. Therefore, the values obtained are better and provide sufficient information for decision making. The technique used is non-invasive and avoids additional risks. Eichhorn *et al.* [21] also found that multidetector CT has high diagnostic accuracy, avoiding the need for additional techniques to plan the surgical approach. According to these authors, the diagnostic accuracy of CT is comparable to catheterization and is more accurate in detecting other complications that could put the patient at risk. Lee *et al.* [22] found that additional diagnostic cardiac catheterization was not needed in a group of neonates following 64-MDCT. Vastel-Amzallag *et al.* [23] also suggested in their study on tetralogy of Fallot that 64-MDCT avoids additional angiographic analysis before corrective surgery. These changes in diagnostic approach have also been addressed in recent publications and guidelines on interventional cardiology [24]. The present study assessed the correlation between the presumptive and definitive diagnosis according to the percentage of unexpected results and diagnostic changes. Unexpected reports described new findings in 100% of patients and prompted changes in management in 86.8% of patients. Of these, 47.2% were new diagnoses. There was a change in management in only 19.5% of the patients with an expected report. There were no significant differences by age group or expected/unexpected report, suggesting that a diagnosis of CHD is independent of age. Greater complexity was found in patients with unexpected reports and in younger patients in each age group. The clinical value of CCTA can be appreciated by considering what would have happened to patients with unexpected findings. In patients with CHD, CCTA has several advantages compared to MRI, such as the simultaneous assessment of cardiovascular abnormalities via airways, lung parenchyma, and chest wall, short anesthesia times, greater spatial resolution, wider availability and less sensitivity to metal artifacts, and can be performed in patients with pacemakers or claustrophobia [25, 26]. Although the damage caused by ionizing radiation can lead to cancer and is directly associated with the patient's age, no consistent cause-effect relationship has been found between the incidence of tumors and exposure to radiation for diagnostic purposes [27–29]. There is no absolute contraindication to CCTA except iodine contrast allergy or contrast-induced nephrotoxicity. Once the CCTA study has been approved, the aim is to minimize the radiation dose needed to acquire an image of sufficient quality to obtain a correct diagnosis [30]. Despite promising initial results, our study has potential limitations. First, the retrospective design of the study was ideal for post-hoc statistical analysis. However, we based the definition of predicted data on speculations derived from the clinical information contained in the test order because otherwise it would have been more difficult to make predictions. Secondly, cases of CHD in patients attending a medical school hospital are complex and may have been a source of selection bias, possibly leading to overestimation in the results. Lastly, inter-observer variability was not assessed.

Conclusions

The present study shows that cardiac computed tomography angiography (CCTA) has good diagnostic performance in CHD patients. The use of this technique has an impact on patient progress, prompts a change in patient management in more than a third of patients, and reveals new findings in relation to initial suspicion in 60.4% of patients. All unexpected reports (100%) described new findings that prompted changes in management in 86.8% of patients. Although it would be ideal to conduct more studies of this kind, we consider that the current rapid developments in technology make it difficult to conduct future comparative studies or studies involving other centers.

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Disclosure

Author reports no conflict of interest.

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