# Demonstration of Coronary Arteries and Major Cardiac Vascular Structures in Congenital Heart Disease by Cardiac Multidetector Computed Tomography Angiography

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## ABSTRACT

**Background.** Detection of coronary artery is important when considering surgical treatment of a congenital heart disease (CHD) such as tetralogy of Fallot (TF). Cardiac catheterization plays an important supplementary role in the evaluation of patients with CHD. In a few reports, it has been proposed that multidetector computed tomography (MDCT) can be helpful for the visualization of coronary arteries. We sought to demonstrate the feasibility and usefulness of MDCT angiography for anatomical evaluation of coronary arteries in CHD patients with suspected coronary artery anomalies.

Materials and Methods. A total of 10 patients, 9 pediatric and 1 adult, underwent MDCT angiography for the investigation of coronary artery anomalies and mediastinal vascular structures. Seven patients had TF; 5 of these patients were suspected of having coronary artery anomalies and 2 were suspected of having pulmonary artery atresia or a nonconfluent pulmonary artery. The other 3 patients had truncus arteriosus and severe left pulmonary artery stenosis (n = 1), double outlet right ventricle (n = 1), and Kawasaki disease (n = 1) with suspected coronary artery aneurysms. The entire heart, major vascular structures, and coronary artery anomalies were preoperatively scanned in patients with cyanotic heart disease. Examinations were performed by 16-MDCT with 1-mm slice thickness. A breath-holding test was performed in 5 patients. Nonionic iodinated contrast material (2 cc/kg) was administered by a power injector.

**Results.** Major vascular structures and the proximal part of the right and left coronary arteries were visualized successfully in all patients. Mid and/or distal segments of the coronary arteries were visualized in 5 patients with TF. Pulmonary vascular bed findings were also confirmed during surgery in patients with TF and in one patient with truncus arteriosus type I and severe left pulmonary artery stenosis. Kawasaki disease was diagnosed by the presence of aneurysms in one patient. Pulmonary artery atresia was confirmed in one patient and diameter of the pulmonary arteries (4 mm and 4.5 mm) was determined in the other 2 patients by MDCT.

**Conclusion.** The advantage of MDCT for cardiac imaging is the shortened scanning time for imaging the entire heart without long breath-holding times. Selective conventional coronary angiography is invasive and technically difficult in pediatric patients. We suggest that MDCT angiography can be performed as a noninvasive method in patients with CHD for the evaluation of coronary artery anatomy and anomalies and mediastinal vascular structures.

## INTRODUCTION

Surgical management of congenital heart disease (CHD), especially in patients with tetralogy of Fallot (TF), may be complicated by anomalies in the course and distribution of coronary arteries and in the pulmonary vascular structures and pulmonary arterial bed. Total repair of cyanotic heart disease, especially in TF, depends on the absence of certain unfavorable anatomic features. Surgical and postmortem studies have shown that important coronary course and distribution anomalies occur in 2% to 10% of patients with TF [Meng 1965; Howe 1970; Berry 1973]. The most common anomaly is the left anterior descending coronary artery arising from the right coronary artery and crossing the right ventricular outflow tract. This type of anomaly greatly affects the surgical outcome in young infants undergoing total correction. Invasive methods such as selective coronary angiography [Dabizzi 1980] and flush aortography [Fellows 1975, 1981] have been used for the detection of coronary artery anatomic variations and distribution. Cross-sectional echocardiography may also be used as a noninvasive method [Berry 1988; Jureidini 1989]; however, it requires an experienced cardiologist and distal coronary visualization can be difficult in most cases.

Multidetector computed tomography (MDCT) is developing rapidly as a noninvasive diagnostic technology for assessing coronary artery anatomies and for calculating various parameters of cardiac function. Recently, the

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Patient No.	Age	Angiocardiographic Diagnosis	Surgery	Heart Rate	Beta Blocker Administration
1	14 mo	TF	Left mB-T shunt	118	Yes
2	47 mo	TF	Total repair	90	Yes
3	15 mo	TF and nonconfluent left PA	Left mB-T shunt	88	No
4	8 y	TA Type I	Left pulmonary artery banding	84	No
5	16 y	Kawasaki disease (Cardiac angiography was not performed. Diagnosis was made with echocardiography and physical examination.)	Clinical follow-up	78	No
6	20 y	DORV and bilateral mB-T shunt	Surgery was planned	74	No
7	6 y	TF and pulmonary atresia	Intrapericardial mB-T shunt	86	No
8	18 mo	TF	Right mB-T shunt	116	Yes
9	16 mo	TF	Total correction	104	Yes
10	36 mo	TF	Left mB-T shunt	96	Yes

#### Patient Characteristics\*

\*TF indicates tetralogy of Fallot; mB-T, modified Blalock-Taussig; PA, pulmonary artery; TA, truncus arteriosus; DORV, double outlet right ventricle.

clinical usefulness of this method has been described for the detection of coronary artery occlusive disease in adult patients.

We present the findings of MDCT angiography in 10 patients. The coronary artery course and distribution and presence of aneurysms were evaluated. The aim of this study was to investigate the feasibility and usefulness of MDCT in patients with CHD.

## PATIENTS AND METHODS

A total of 10 patients, 3 female and 7 male, were evaluated for coronary artery course and distribution anomalies in TF, coronary artery aneurysm in Kawasaki disease, the main and branch pulmonary arteries in cases with TF and truncus arteriosus, and the patency of a modified Blalock-Taussig (mB-T) shunt in one adult case. Patient ages ranged from 14 months to 20 years. Mean age of the pediatric patients was  $6.8 \pm 1.2$  years. There were 7 patients with TF. The other 3 patients had double outlet right ventricle (DORV) and bilateral Blalock-Taussig shunt operation, truncus arteriosus type I and severe left pulmonary artery stenosis, and Kawasaki disease with suspected coronary artery aneurysms (patient characteristics are summarized in the Table).

## Prior Diagnostic Evaluation and Surgical Approach

Electrocardiography (ECG), chest x-ray, and routine examinations were performed for all patients. Transthoracic echocardiography and left and right heart catheterization were performed in the division of pediatric cardiology in all patients except one, who had Kawasaki disease. Modified left, right, or central (intrapericardial) shunts were placed with the use of a polytetrafluoroethylene graft because of an unsuitable pulmonary artery bed and a significant McGoon index rating in 5 patients with TF. Total correction was performed in the remaining patient with TF. Pulmonary artery banding was performed in the patient with truncus arteriosus type I. All patients had an uneventful recovery after surgery. We planned

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surgery in one patient with DORV due to severe stenosis of bilateral mB-T shunts. We decided on clinical follow-up for the patient with Kawasaki disease and multiple coronary artery aneurysms.

# **MDCT Angiography Protocol**

Informed consent was obtained for all patients. Examinations were performed by 16-MDCT using ECG-gating (Somatom Sensation 16; Siemens Medical Systems, Munich, Germany). Technical parameters were a slice thickness of 1 mm, detector configuration of  $12 \times 0.75$  mm, table speed of 3.4 mm/rotation, and a gantry rotation time of 420 milliseconds. After administration of an intravenous contrast medium, the entire heart was scanned during a single breath hold. Imaging time was less than 15 seconds in pediatric patients. Two radiologists blinded to the study analyzed the images. A beta-blocker agent (propranolol) was given to decrease the heart rate in patients with tachycardia before MDCT scanning, and an intramuscular sedative drug was given to small children to avoid artifacts. Scanning was initiated by the modified bolus tracking method as described previously. MDCT data were reviewed on a distant workstation (Leonardo; Siemens) using interactive scrolling, multiplanar reconstructions, and volume-rendering and maximumintensity projection algorithms.

## RESULTS

The proximal right and left coronary artery systems and the branch pulmonary arteries were successfully visualized in all patients. The mid and distal part of coronary arteries were visualized successfully in most cases (Figure 1). However, in 2 patients we could not successfully visualize the pulmonary branches by cardiac catheterization. There were coronary artery anomalies in 2 patients with TF (Figure 2). In 2 cases, a coronary artery branch originating from the left anterior descending artery was noted to cross the right ventricular outflow tract. Appropriate measurements of the pulmonary

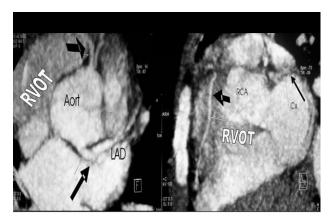


Figure 1. Axial multidetector computed tomography images display the findings of an abnormal coronary artery course and distribution in a patient with tetralogy of Fallot. RCA indicates right coronary artery; LAD, left anterior descending coronary artery; Cx, circumflex coronary artery; RVOT, right ventricular outflow tract.

artery and its branches were clearly provided for the mB-T shunt operation in 4 patients with TF (during the operation we confirmed that there were no coronary artery course or distribution anomalies in these patients). Pulmonary artery branches were seen with MDCT to a better extent than with conventional cardiac catheterization, and the MDCT also provided objective measurements.

There were multiple coronary artery aneurysms in the patient with Kawasaki disease. Aneurysms of the left anterior descending artery and right coronary artery were clearly and successfully visualized in this patient (Figure 3). We observed severe stenosis of the left and the right mB-T shunt and patency of the left and right coronary system, including distal parts in a 20-year-old patient with DORV (Figure 4).

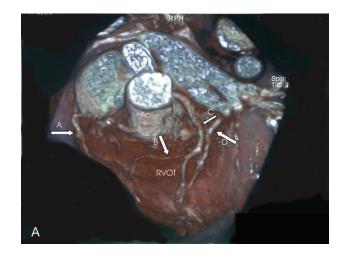
In patients with a suitable branch pulmonary artery, mB-T shunt operations were done and the MDCT findings were confirmed during the operation. The size of the main pulmonary artery and its branches measured about 10 mm, 4 mm, and 4.5 mm in these patients. There was no mortality or morbidity related to surgery, and the patients were discharged home in good condition after the surgery.

## DISCUSSION

Findings of this study showed that 16-MDCT coronary angiography can be performed preoperatively performed as a noninvasive method for detection and evaluation of coronary artery course and/or distribution anomalies in patients with CHD. These results also clearly displayed that valuable data about the pulmonary arterial trunk and its bed may be obtained by the use of this technology in the preoperative period.

Selective coronary angiography has been the gold standard of coronary imaging since its introduction in 1959 [Sones]. However, this technical option is difficult to perform, especially in small infants. Flush aortography has also been described as an invasive procedure for the evaluation of the coronary arterial tree [Fellows 1975, 1981]. However, although it is possible to identify abnormal coronary vessels across the right ventricular outflow tract, 2-dimensional projection images of invasive angiography may preclude visualization of the coronary anatomy in a significant proportion of infants. Berry et al reported that it was not possible to see the coronary arteries well enough in over 20% of their patients with TF [Berry 1988].

Noninvasive techniques and modalities such as transthoracic and transesophageal echocardiography and magnetic resonance imaging (MRI) have been described in previous studies for the visualization of coronary anomalies. Even so, echocardiographic investigation requires an experienced cardiologist, especially for small infants. Echocardiography



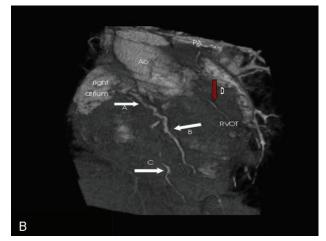


Figure 2. The anomalous coronary artery crossing the right ventricular outflow tract in 2 cases. A branch coronary artery was separated from the left anterior descending artery and crossed the right ventricular outflow tract. For part A, Ao indicates aorta; A, right coronary artery (RCA); B, abnormal branch that crossed the right ventricular outflow tract; C, left anterior descending artery; D, anterior interventricular vein. For part B, A and B indicate RCA; C, branch RCA; D, abnormal coronary artery; PA, pulmonary trunk; Ao, aorta.

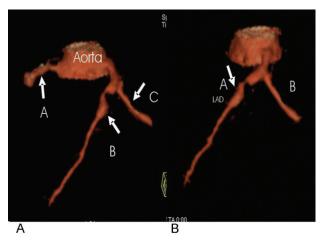


Figure 3. This patient was an 8-year-old boy with Kawasaki disease. A, The right coronary artery (arrow A) and a left anterior descending coronary aneurysm (arrow B) formation is clearly shown. B, The aneurysm of the left anterior descending artery (arrows A and B).

can analyze only the very proximal part of anomalous coronary vessels [Gianoccoro 1993; Fernandes 1993; Zeppilli 1998]. Several studies involving 5 to 19 patients and a number of case reports have demonstrated reliable visualization of coronary anomalies by magnetic resonance angiography [Vliegen 1997; Taylor 2000]. But when we compare MRI and computed tomography (CT) technology, CT has some advantages over MRI, including the simplicity of the examination and short examination times. Moreover, 16-MDCT can be performed with a small amount of contrast medium, and high-resolution images can be obtained in small infants.

Anomalous vessels are not always detectable during the operation, especially when the coronary arteries are obscured by the overlying myocardium and epicardial fat [Friedman 1960; White 1972] or during reoperation due to epicardialpericardial adhesions in patients who previously underwent palliative surgery. We suggest that MDCT angiography can be easily used in these patients as a noninvasive diagnostic technique when the coronary artery anatomy can not be displayed during cardiac catheterization.

The first multislice spiral CT scanner was introduced into the clinical routine in 1998. All currently available MDCT scanners allow for the simultaneous acquisition of at least 4 parallel slices in the z-direction with an image quality of at least equal to that of a single-slice spiral CT. However, the MDCT has higher spatial and temporal resolution compared with single-slice CT. With retrospective ECG synchronization, a 3-dimensional volume data set of the entire heart can be obtained for the different phases of the cardiac cycle, which corresponds to 4-dimensional data acquisition with time as the fourth dimension [Nieman 2001]. The coronary artery and its segments can be visualized optimally during the different phases of the cardiac cycle [Lu 2001]. Using current reconstruction algorithms, the projections necessary for image calculation are collected from several gantry rotations around the patients. We used a nonionic monomeric x-ray agent

administered by an automatic injector as the contrast material.

Four-slice MDCT requires a longer period of breath holding, which depends on the extent to which the patient can cooperate as well as on the oxygenation and ventilation condition. These limitations are removed with the use of the 16-slice MDCT. We obtained cardiac and coronary artery images in a short time (within approximately 30 seconds for all patients). Good correlation of the CT results with catheter angiography has been demonstrated for the proximal segments [Lu 2001].

Most authors report the use of MDCT for detection of coronary artery disease in adult patients [Georg 2001; Herzog 2001; Lu 2001; Nieman 2001; Fernandez-Valls 2004]. But a limited number of studies have been reported in the English literature of MDCT use for cardiac evaluation in CHD or coronary anomalies [Westra 1996; Fernandez-Valls 2004]. To our knowledge, no prior report matches this description of detection of coronary artery anomalies or the evaluation of the pulmonary artery branches or its peripheral bed using 16-MDCT.

MDCT findings of the coronary artery and the pulmonary vascular trunk have been confirmed surgically in our study. We suggest that the 16-MDCT is a reliable and noninvasive method and can be preoperatively used in small infants with cyanotic heart disease. Also, this technology might be combined with routine cardiac catheterization in some patients with CHD who are suspected of having pulmonary artery confluency, like our described patient.

A congenital coronary artery anomaly has been detected and successfully shown in an adult patient by the use of 16-MDCT by Hong [2004]. This patient had no coronary arteriosclerosis but had an exercise-related angina pectoris, and a benign left main coronary artery crossing anomaly was detected in this patient.

Coronary artery aneurysms are rare disorders defined by a vessel diameter >1.5 times larger than the present artery. An aneurysm can be characterized by abnormal dilatation of a localized or diffuse arterial segment. Potential complications

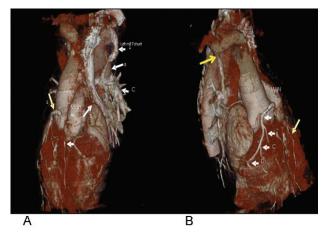


Figure 4. Severe stenosis of the distal part of the left modified Blalock-Taussig shunt and proximal part of the right modified Blalock-Taussig shunt are shown (B arrow in part A and unlabelled arrows in part B). In part A, arrow E indicates the origin of the right coronary artery and arrows A, B, C, and D indicate the right coronary artery and its branches. Arrow B shows the left pulmonary artery in part A, and RPA indicates the right pulmonary artery in part B.

associated with these abnormalities include thrombosis, distal embolization, and rarely rupture and sudden death. In our study, we also evaluated coronary artery aneurysms in a patient with Kawasaki disease. There was aneurysmatic dilatation of the left main and the left circumflex artery, but the patient had no ischemic symptoms and there was no evidence of distal embolic events. We propose that 16-MDCT can be used as a diagnostic modality for the clinical follow-up and detection of coronary artery aneurysms in patients with Kawasaki disease.

We successfully visualized the appropriate left and right pulmonary arteries in our infant patients to evaluate the feasibility of a surgical option. There was a suspicion of a nonconfluent pulmonary artery in one patient with TF. A severe left pulmonary artery hypoplasia was diagnosed by the use of cardiac catheterization in a patient with truncus arteriosus type I. Extrapericardial and intrapericardial pulmonary artery diameter were visualized successfully with a high-quality image, and normal coronary artery anatomy was confirmed in these patients at the same time. In addition, we visualized the entire heart and detected severe stenosis of the bilateral mB-T shunt in a 20-year-old patient. This patient had been admitted to our clinic with perioral cyanosis and exercise intolerance. Coronary MDCT angiography rather than cardiac catheterization was the first diagnostic choice for this patient on referral to our hospital.

In conclusion, we propose that MDCT may be useful for the visualization of the entire heart as a noninvasive method in infants with CHD because it can be performed with lowdose contrast material and in a short examination time.

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