

# MDCT of Congenital Heart Disease in Children

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Nowadays, with advanced technology of computed tomography (CT), high quality 2D and 3D images can be obtained which precisely show morphologic abnormalities of congenital heart disease. The abnormalities of the aorta, the pulmonary arteries, the pulmonary veins, the cardiac chambers, the ventriculo-arterial connection, the relationship between upper lobe bronchi and the pulmonary arteries, the coronary arteries, the systemic veins and the visceral situs can be evaluated without technical difficulty. Cardiac CT is a non-invasive study and provides unique information of the anatomy of the heart and great vessels that cannot be currently obtained by traditional methods. The disadvantage of cardiac CT is ionizing radiation and risks of adverse reaction to contrast media. The other problem is we have to perform cardiac CT under general anesthesia to control respiration in patients who cannot hold their breath, especially in cases of CT angiography of coronary artery. Even though MDCT has a limited role in the assessment of cardiac function, the evaluation of congenital heart disease with MDCT as a complementary imaging tool after echocardiogram should be the future direction of congenital heart disease imaging if physiological data is not necessary in that patient. This article highlights the role of MDCT in the evaluation of congenital heart disease in certain problematic aspects.

## INTRODUCTION

Cardiovascular imaging is essential to diagnose and treat all forms of pediatric congenital heart disease. Diagnostic imaging of congenital heart disease has undergone a remarkable change over the past two decades. Advances in echocardiography in the 1990s, magnetic resonance imaging (MRI) in the late 1990s and multi-slice computerized tomography in early 2000s allow a comprehensive noninvasive assessment of the cardiovascular structure and function in many children with cardiovascular anomalies. These technical advances contribute to the continued decline in the use of invasive catheterization procedures performed solely for diagnostic purposes. Echocardiography is the initial diagnostic modality of choice for a patient with suspected congenital heart disease. Its strengths and limitations have been

recognized. The well known limitations are small field of view, poor acoustic window, operator dependence and its poor depiction of extra-cardiac vascular structures such as great arteries and intrapulmonary vessels. The traditional method of using conventional angiography as a complementary imaging modality of echocardiography for assessment of complex congenital heart disease have to be changed since catheter angiogram is not only invasive, but also has limitations such as overlapping of adjacent cardiovascular structures, difficulties in simultaneously depicting the systemic and pulmonary vascular systems and catheter related complications. Cross-sectional imaging with MRI and computed tomography may help overcome the limitations of echocardiography and conventional angiogram. Despite the great capabilities of MRI for anatomic and functional assessment of the heart, an examination of this modality is time consuming and requires lengthy period of general anesthesia in patients who cannot hold their breath.

MDCT has the advantages of short acquisition time, and a higher spatial resolution as compared to MRI. Its capabilities for simultaneous evaluation of cardiovascular structures and lung parenchyma, has increased the clinical application of CT for the evaluation of patients with congenital heart diseases especially for morphologic assessment purposes. With ECG gating, MDCT can accurately delineate rapidly moving cardiac structures and allow assessment of coronary anomalies and coronary artery disease. In addition ventricular function can be obtained using MDCT coupled with ECG gating. Its drawbacks are risks of ionizing radiation, and risks of iodinated contrast material. These disadvantages must be kept in mind when considering the use of MDCT in children.<sup>1</sup>

## Scanning protocol

With our 64-slice scanner, we use a detector collimation of 0.6mm. Slice thickness is 1 mm, and pitch is 1.2. We image in either the cranial-caudal direction or the caudal-cranial direction. Contrast administration consists of 2 cc/kg body weight of nonionic contrast media. Injection rate is adjusted to get an injection duration of not more than 8-10 seconds if the heart rate is more than 100/min, and not more than 13-15 seconds

if the heart rate is less than 100/min. We do not use bolus tracking except for the patient who has adult size. Scan delay is injection duration plus 8 seconds to reduce inflow streak artifacts in SVC or right atrium. If contrast media used is less than 10 cc. or contrast injection is through the central line, the contrast media will be injected by hand and scan delay is about 12-15 seconds. Contrast access site could be arm or leg. For the patients who cannot hold their breath, we use general anesthesia to perform apnea in the patient whose respiratory rate is more than 25/min and use sedation in patients whose respiratory rate is less than this. We use ECG gating only in coronary CTA.

When viewing the images, we derive information from axial planes, 2D multi-planar reconstruction (MPR), maximum intensity projections (MIP) and 3D volume rendering (VR). The usefulness of MDCT is CT volume data which can be reformatted in unlimited planes for interpretation<sup>2,3</sup>. In addition, minimum intensity projection which is very helpful for evaluation of airways, curved reformatted images for evaluation of bending vessels such as pulmonary artery can be selectively retro-reconstructed<sup>2,4-6</sup> for additional information in some cases. The time used for post image processing is about 1 hour.<sup>2,7</sup>

### Clinical application

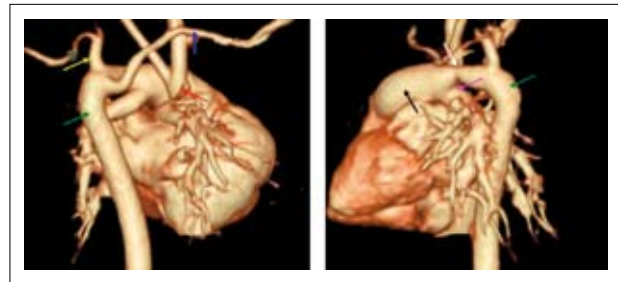
Most of the MDCT studies were performed to answer specific anatomical questions raised by inconclusive echocardiographic or angiographic evaluation. The common indications are evaluation of pulmonary artery in severe obstruction on the right side of heart, assessment of systemic collateral blood supply to the lungs, demonstration of abnormal pulmonary venous connection, postoperative evaluation of shunts and complications, evaluation aortic arch anomalies and coronary artery abnormalities.

Today MDCT is accepted as a diagnostic modality of choice for evaluation of coronary artery anomalies. The simultaneous visualization of coronary arteries and the heart greatly help a cardiac surgeon for preoperative planning, such as planning how to approach and avoid injury of coronary arteries when seeing it crosses the right ventricular outflow tract on 3D MDCT images in Tetralogy of Fallot for total correction. Coronary anomalies can be correctly characterized because the origin, course and destination of coronary arteries are better defined than by catheter angiogram.<sup>8,9</sup>

### Application Examples of MDCT

#### Aortic Arch Anomalies

Although echocardiography can demonstrate aortic arch anomalies, such as co-actation and interruption of the aortic arch, it may be limited for evaluation of the ascending aorta, aberrant vessel anatomy and collateral vessels. MDCT is an effective and rapid imaging modality for the morphological assessment of the aortic branching pattern and an interrupted gap distance in an interrupted aortic arch (Fig 1). In addition CT is a valuable tool when considering different therapeutic treatments (balloon catheterization, stent placement or surgery) in patients with severe aortic arch co-actation by evaluation of the degree of stenosis, its relationship to the aortic branch and collateral vessels. (Fig 2)<sup>6,10</sup> The more complex anomalies such as interrupted aortic arch associated with truncus arteriosus in which



**Fig 1.** A 3-day-old boy with left side heart failure. Interrupted aortic arch was seen at echocardiography. MDCT was performed to evaluate the interrupted gap and aortic arch branching pattern for surgical planning.

**A.** Right posterior oblique 3D volume rendered image shows interruption of aortic arch between left subclavian artery (yellow arrow) and common trunk of right and left common carotid artery (red arrow). Right subclavian artery originates aberrantly as the 3<sup>rd</sup> branch of aortic arch (blue arrow). The interrupted gap can be accurately measured (not shown)

**B.** Left lateral 3D volume rendered image shows ductus arteriosus with mild focal stenosis (white arrow) supplying descending aorta (green arrow). Black arrow= MPA, pink arrow= LPA

echocardiography cannot depict all anomalies accurately can be precisely assessed by MDCT. (Fig 3)

### Pulmonary Artery Anomalies

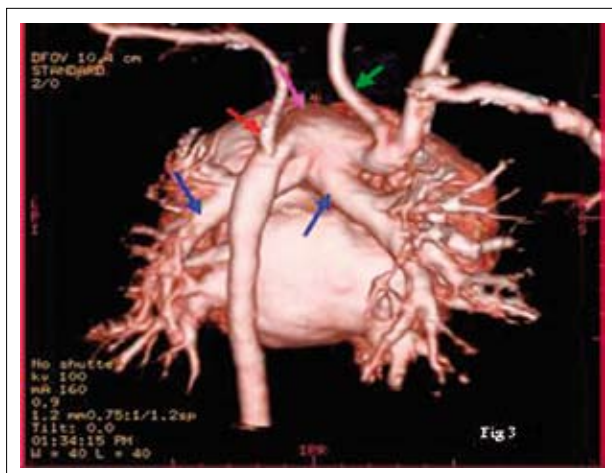
In older children, echocardiographic assessment of the right heart and the pulmonary artery and its branches is technically more difficult because it is located deeper in the thorax. These limitations are particularly



**Fig 2.** A 4-year-old boy with stronger radial than femoral pulses. Echocardiogram suggested the presence of coarctation. MDCT was requested to evaluate coarctation and collateral anatomy before considering treatment.

**A.** Left posterior oblique 3D volume rendering image shows severe narrowing of aortic arch (green arrow) just distal to left subclavian artery with mild poststenotic dilatation (red arrow) of descending aorta. The descending aorta is supplied by collateral vessels from internal mammary arteries through intercostal arteries (blue arrow), costocervical trunk arteries (pink arrow) and transverse scapular artery (yellow arrow). Noted the stenotic area is too close to left subclavian artery ostium and difficult to avoid its occlusion if performing stent placement.

**B.** Posterior coronal MIP image shows enlarged intercostal arteries (violet arrow) carry blood from internal mammary arteries to descending aorta. There are also collateral vessels (orange arrow) from transverse scapular artery carries blood through connecting branches to intercostal arteries to descending aorta.



**Fig 3.** A 1-month-old girl presented with dyspnea. Posterosuperior 3 D volume-rendered image shows single arterial trunk exits from the heart (pink arrow). The right and left pulmonary arteries (blue arrows) are close to each other but arise from the trunk separately from the posterior aspect of the trunk (truncus arteriosus type II). There is associated interosseous aortic arch between the left common carotid (green arrow) and left subclavian arteries (red arrow).

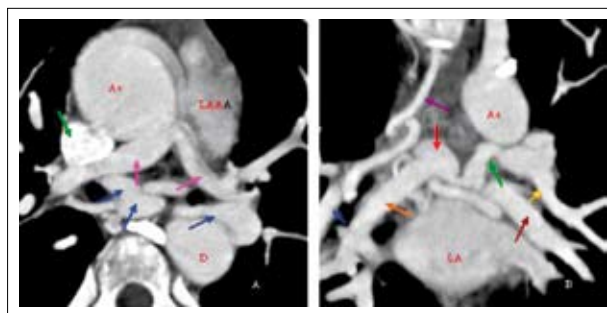
important in the evaluation of pulmonary artery anatomy in severe obstruction of the right side of the heart such as TOF and pulmonary atresia with ventricular septal defect (PA-VSD), especially post palliative surgery when considering total corrective surgery. These patients experienced many cardiovascular imaging investigations especially invasive methods such as conventional angiogram in their early life, the later suitable imaging modalities should be non-invasive methods which might be either MDCT or MRI. At our hospital, most MDCT examinations for congenital heart disease are requested for evaluation of the pulmonary artery anatomy, and the systemic to pulmonary arterial collateral vessels in severe obstruction of the right side of the heart such as pulmonary atresia, and TOF after palliative surgery (Fig 4).<sup>3,11</sup> MDCT and its capability of volume rendering for viewing in any plane and angle can overcome superimposition of vascular structures which usually is the problem of biplane conventional angiograms. (Fig 5)

### Venous Anomalies

The cardiologist commonly encounters difficulty in evaluation of systemic and pulmonary veins by echocardiography. The poor visualization is usually due to the complexity of various types of abnormal pulmonary venous connection and their courses which are usually within lungs. In heterotaxy syndrome, systemic venous return is often complicated including bilateral superior vena cava, inferior vena cava interruption, direct draining of the hepatic veins into the right atrium and an echocardiographer often cannot visualize all drainage into the right atrium. MDCT is increasingly used in assessing these complex anomalies as the whole problem can be disclosed by the single examination. (Fig 6)<sup>12</sup>

### Coronary Artery Anomalies

Coronary artery anomaly has traditionally been reserved for catheter angiography. The advanced technology of MDCT nowadays has allowed successful evaluation of a variety of coronary artery anomalies in



**Fig 4.** A 12-year-old girl, known case of PAVSD having right BT shunt in early life. MDCT was performed for evaluation of pulmonary artery and systemic collateral vessels to lungs before corrective surgery.

**A.** Axial MIP image shows small RPA and LPA with central confluence (pink arrow). Large mapca (blue arrow) originates from left anterior wall of aorta (D) supplies both lungs. (LAA= left atrial appendage, Ao= ascending aorta, green arrow= SVC)

**B.** Coronal MIP image shows large mapca seen on end (red arrow) gives large branch (orange arrow) to RLL, another branch (green arrow) connects to LPA (yellow arrow) with focal stenosis at origin and connecting site with LPA. Another mapca (brown arrow) from descending aorta supplies LLL. Smaller collateral artery (violet arrow) from RSA supply RML. It is very difficult for conventional angiogram to demonstrate numerous major aortopulmonary collateral arteries (mapcas) because many injections have to be performed instead of single peripheral injection as in MDCT. (PAVSD= pulmonary atresia with ventricular septal defect, MIP= maximum intensity projection, LPA= left pulmonary artery. LLL=left lower lobe, RSA=right subclavian artery, RML=right middle lobe)

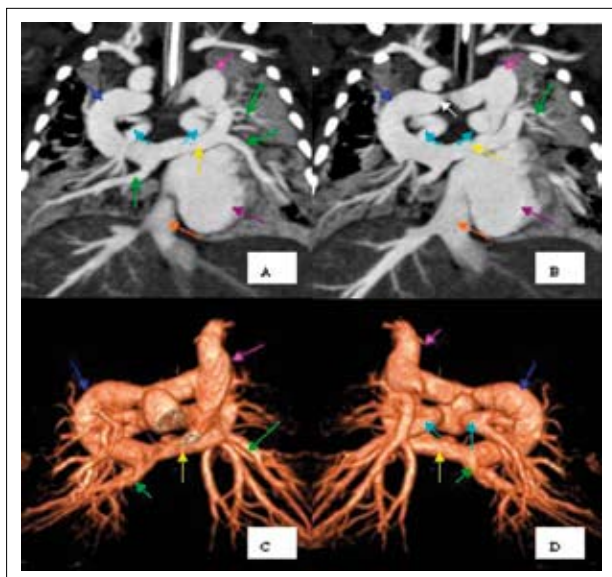
patients with congenital heart disease. It is now accepted that MDCT is the modality of choice for coronary angiography because it is non-invasive and the quality of image is comparable to conventional angiograms. In addition, intracardiac lesions, extracardiac vessels and ventricular function can be evaluated from the same



**Fig 5.** TOF with large MAPCA

**A.** Superoanterior 3D volume rendering image shows more anterior aorta (Ao) with overriding over ventricular septum (pink arrow). The RV infundibulum (green arrow) and MPA (blue arrow) as well as its branches (yellow arrow) are hypoplastic.

**B.** Right posterior oblique 3D volume rendering image shows large collateral artery (red arrows) originated from descending aorta supplying left lung by connecting to descending branch (brown arrow) of LPA (yellow arrow) at left hilum. There is focal stenosis (violet arrow) before connecting to LPA.



**Fig 6.** 17-day-old girl with heterotaxy syndrome and asplenia. TAPVR was suspected by echocardiography. MDCT was requested to evaluate venous drainage.

**A.** Coronal MIP image shows all pulmonary veins (green arrows) from both lungs join to common pulmonary vein (yellow arrow) and drain through the vertical vein (blue arrow) running upwardly on the right side before emptying into left SVC (pink arrow).

**B.** Coronal MIP image more anterior than A shows severe focal stenosis (white arrow) of the vertical vein before emptying into left SVC. (Light blue arrows= pulmonary artery, orange arrow= IVC, violet arrow=right atrium)

**C.** Total anomalous pulmonary venous return seen from front of 3D volume rendering image with heart removed.

**D.** Total anomalous pulmonary venous return seen from back of 3D volume rendering image. Volume rendering images can be rotated in any direction and angle and very helpful for surgeon to see the distance between atrium and common pulmonary vein for surgical planning when considering corrective surgery.

study. It is also very helpful for a surgeon to consider the technique of surgery preoperatively to avoid collateral injury because of the coronary artery and its course in relation to cardiac chambers are seen simultaneously. (Fig 7)<sup>8,9,13,14</sup>

## CONCLUSION

MDCT application for congenital heart disease in children is increasing dramatically because it shows the heart, the extra-cardiac vessels, the lungs, airways and the thoracic cage simultaneously in high quality images. It is a non-invasive, very rapid image acquisition and is very suitable for unstable patients. If the clinical question is anatomy, CT is very appropriate. MDCT still has a limited role for functional assessment.

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**Fig 7.** A-7-year-old boy, known case of TOF. Coronary anomaly was suspected at echocardiography. MDCT was requested to determine coronary artery anomaly and its course in relationship to RVOT before corrective surgery.

**A.** Left anterosuperior view of the 3D volume rendering image (pulmonary artery removed) shows single coronary artery arising from left aortic sinus (violet arrow) and giving RCA and LCA. The right coronary artery (blue arrow) runs anteriorly between ascending aorta and pulmonary artery to the right atrioventricular groove. Conal branch (yellow arrow) arises from RCA is rather large and passes over RVOT (white arrow) which may interfere with ventriculotomy. LCA gives LCX (red arrow) and LAD (green arrow) which have normal course.

**B.** Superoanterior 3D volume rendered image (pulmonary artery included) shows normal size pulmonic valve (light blue arrow), hypoplastic MPA (orange arrow) and infundibular stenosis (white arrow). The course of conal branch over RVOT is well seen in this view. (RV=right ventricle, LV=left ventricle, RA=right atrium, AO=aorta, PA=main pulmonary artery, LA=left atrium, RA=right atrium)

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