Dual Source Computed Tomography in the evaluation of Pediatric Congenital Heart Diseases: A Pictoral essay

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Learning objectives

1. To establish optimal protocols for pediatric imaging on DSCT for the evaluation of CHD.
2. To identify the ideal image processing and reformatting techniques to analyse the acquired data.
3. To illustrate a systematic process for image analysis to delineate the spectrum of pathologic morphological features in the evaluation and management planning of pediatric CHD using DSCT.
4. To be aware of the advantage and disadvantage of DSCT in the evaluation of CHD.
Background

In India, CHD affects about 8-10 per 1000 live births and is a leading cause of infant mortality. The burden of CHD in India is enormous due to the very high birth rate thus emphasizing its importance. It is known that 180,000 children are born with CHD each year in India [1]. With current advances in cardiac surgery, a large majority of these infants are expected to reach adulthood. A vital component in the multidisciplinary management of congenital heart disease patients is adequate imaging of the heart and circulation. [2]

Traditionally, invasive angiography was used to evaluate patients with congenital heart disease. However catheter cardioangiography is limited by the overlapping of adjacent vascular structures, difficulty in demonstrating systemic and pulmonary vascular systems simultaneously, catheter-related complications (especially in young children), and relatively high doses of ionizing radiation and iodinated contrast material. [3, 4]

Today, physicians increasingly turn to noninvasive methods, such as echocardiography, magnetic resonance imaging (MRI), and, more recently, multislice computed tomography. Although echocardiography is the initial diagnostic modality for patients with suspected congenital heart disease, it is limited by a small, limited field of view, an acoustic window, operator dependence, and poor depiction of extracardiac vascular structures. [2,3,4] Thus in the assessment of an entire congenital repair, which may include extra cardiac anatomic bypasses or systemic shunts, it is often suboptimal. [2]

MR Imaging despite its excellent anatomic and functional assessment is often time-consuming requiring long sedation and hence is limited in seriously ill or uncooperative patients. In addition, contraindications include patients with a pacemaker. [4, 5]

CT has the advantages of widespread availability and short acquisition times. Recent developments with dual source CT have increased scanning speed with higher spatial resolution, coupled with electrocardiographic (ECG) data and high-quality two- and three-dimensional multiplanar reformatted images allowing accurate delineation of rapidly moving cardiac and paracardiac structures thus increasing its clinical application in the evaluation of patients with congenital heart diseases [6]. In addition, CT can obtain functional data about ventricular wall and cardiac valve motion [7,8,9]. Drawbacks of CT that must be borne in mind include radiation exposure and the inherent risks of iodinated contrast material, particularly in pediatric patients.

This pictoral essay, attempts to illustrate the typical pathologic conditions (extracardiac, cardiac and connection abnormalities) seen on CT in patients with CHD, and the advantages and disadvantages of CT in this setting.
DSCT was performed on 120 pediatric congenital heart disease patients, aged between 0 to 16 years, following clinical evaluation and echocardiography. All patients underwent a detailed examination on a 64 slice DS CT scanner (SOMATOM definition; Siemens medical solutions, Germany). Sedation with 50-75 mg/kg of oral chloral hydrate was given barring few cases where intravenous sedation was required.

The CT protocols were modified depending on the case based clinician's question and echocardiography findings.

The thorax was scanned in a craniocaudal direction with ECG synchronization during quiet breathing, where breath holding was not possible. Pediatric low dose protocols were used at 120 kV, 30-80mA, pitch of 1.4mm and slice thickness 0.6mm, reducing tube voltage to 80 kV if possible (3). Vascular and cardiac opacification, was obtained using non-ionic contrast agent, optiray (Ioversol 300mg of iodine per milliliter, diluted 2:1). 3ml/kg body weight of diluted contrast was injected intravenously at a rate of 1.5-2.5mL/sec followed by a saline bolus chasing technique. Scanning was started manually when contrast was seen in the main pulmonary artery.

Depending on the clinical question and structures targeted postprocessing techniques, included multiplanar reformatting, maximum and minimum intensity projections and volume rendering on a dedicated workstation (Syngo In Space 4D; Siemens).

The Fortis Escorts Heart Institute and Research Centre is a super speciality cardiac research centre specializing in congenital heart disease surgeries. The high quality 3D DSCT images guide further intervention, clinical and surgical management.

**Imaging Findings:**

A segmental and systematic step by step analysis of cardiac and extracardiac structures was made to determine extracardiac, intra cardiac and connection abnormalities in these patients.

**Extracardiac Abnormalities**

**Aortic Anomalies**

Beyond the aortic valve, echocardiography is limited in its capability to evaluate the ascending aorta, aberrant vessel anatomy, and aortic coarctation. DSCT permits excellent delineation of complex congenital aortic arch anomalies particularly due to the availability of CT bolus-tracking techniques and volume-rendered image reconstructions. [4, 5]
Interrupted Aortic Arch is defined as a separation between the ascending and descending aorta.[10,11]. Based on the site of interruption it is classified as: Type A, distal to the left subclavian artery; Type B, between the left carotid and the ipsilateral subclavian arteries and Type C, between two carotid arteries. Each of these is further subdivided into Subtype 1 with a normal subclavian artery; Subtype 2 with an aberrant subclavian artery; and Subtype 3 with an isolated subclavian artery arising from the ductus arteriosus. In addition evaluation of the distance between the proximal and distal segments, the size of a patent ductus arteriosus (PDA), the narrowest dimension of the left ventricular outflow tract, and other cardiac structural abnormalities are important for surgical planning. [3 ].

Coarctation of the Aorta maybe a localized discrete narrowing of the aortic lumen, usually located just distal to the left subclavian artery with associated arch and isthmus hypoplasia [12]. Rarely, coarctation may be proximal to the left subclavian artery compromising the same. Associated aberrant right subclavian artery may arise at or below the coarctation. There is often dilatation of the aorta just distal to the coarctation. Tubular hypoplasia involving a long segment of the aortic arch is more frequent in neonates. A localized coarctation and tubular hypoplasia may coexist or may occur independently.[3]

Hypoplastic left heart Syndrome is characterized by underdevelopment of the left side of the heart with hypoplasia/atresia of the ascending aorta, aortic valve, left ventricle and mitral valve.[12].

Pulmonary Artery Anomalies

In older children, echocardiographic evaluation of the pulmonary artery anomalies is limited. Further a number of congenital heart defects, require complex surgical pulmonary artery reconstructions that have traditionally been evaluated with catheter angiography [13].

Today DSCT with its volume rendering capability is the preferred noninvasive imaging alternative to delineate complex anomalies of the pulmonary arteries in CHD. Pulmonary sling is a rare anomaly in which the left pulmonary artery arises from the right main pulmonary artery. [5]

Truncus Arteriosus and Aortopulmonary Window. In truncus arteriosus, a single arterial trunk arises from the ventricle via a single arterial valve to supply the systemic, pulmonary, and coronary arterial circulations. Aortopulmonary window is a communication between the ascending aorta and the pulmonary trunk in the presence of separate aortic and pulmonary valves.[3]

Venous Anomalies

Where echocardiography is limited in the evaluation of systemic and pulmonary venous return DSCT accurately displays complex venous anatomy and is increasingly being used
in assessing these complex congenital venous anomalies eg in heterotaxia syndromes with complicated systemic venous return due to interruption of the inferior vena cava [14].

**Coronary Artery Anomalies** have traditionally been evaluated by catheter angiography, DSCT permits excellent and accurate depiction of the coronary artery .[5]

**Patent Ductus Arteriosus (PDA)** is persistent post natal patency of the normal prenatal connection from the pulmonary artery to the proximal descending aorta. PDA maybe part of a complex CHD as hypoplastic left heart syndrome, D Transposition and right ventricular outflow obstruction.[12]

**Total Anomalous Pulmonary Venous Connection** is characterized by connection of the pulmonary veins from both lungs to form a confluence behind the left atrium and connection of a venous channel from this confluence to a systemic vein, the right atrium, or both being described as supracardiac, cardiac, infracardiac, or mixed depending on the site or sites of connection. Supracardiac and cardiac types are rarely obstructive, but bilateral infracardiac types are almost always obstructive because the blood passes through the hepatic sinusoids. [3]

**Partial Anomalous Pulmonary Venous Connection** occurs when the pulmonary veins from some portions of both lungs show an anomalous connection. The sites of connection include the SVC, the right atrium or the IVC. The right pulmonary vein to the IVC connection is called the "scimitar vein" due to its curved configuration. [15, 16]

**Left SVC** is an abnormality draining into the right atrium via a dilated coronary sinus, recognition of its presence is important for surgical planning. [3]

**Intracardiac Abnormalities**

DSCT accurately demonstrates typical appearances of various CHDs including stenosis or obstruction, defect, or connection problem.

**Tetralogy of Fallot** includes subpulmonary infundibular stenosis, ventricular septal defect (VSD), overriding of the aorta, and right ventricular hypertrophy. [17]

**Pulmonary Atresia with VSD** demonstrates morphologic characteristics of extreme Tetralogy of Fallot. [18,19,20,21,22]. There is no pulmonary blood flow hence detailed morphologic evaluation of the presence or absence of the central pulmonary artery, confluence or non confluence of the branch pulmonary arteries and various pulmonary arterial feeding vessels including PDA and major aortopulmonary collateral vessels is important for surgical planning. [3]

**Ebstein Anomaly** is characterized by downward displacement of the septal and posterior leaflets of the tricuspid valve with resultant atrialization of the inlet of the right ventricle causing hemodynamically a severe tricuspid valve regurgitation. [20, 23]
Tricuspid atresia results in absence of direct communication between the right atrium and the right ventricle [20]. There are two types of tricuspid atresia. In the more common type, the right atrioventricular connection is absent and areolar sulcus tissue occupies the gap ventricle [13]. In the rarer type, an atretic tricuspid valve is present.

Ventricular Septal Defect encompasses a hole or holes of variable size in the interventricular septum classified as either perimembranous, muscular, inlet or outlet supracristal committed juxtaarterial defects.[3]

Atrioventricular Septal Defect is a defect in the atrioventricular septum with variable involvement of the adjacent atrial and ventricular septa resulting in abnormal arrangement of the atrioventricular valve leaflets, and associated anterosuperior displacement of the aortic valve from its normal wedged position. The left ventricle has a short inlet and an elongated outlet. [3, 20]

Connection Problems

Complete Transposition of the Great Arteries or D Transposition is a combination of atrioventricular concordance and ventriculoarterial discordance. The systemic and pulmonary circulations are parallel, independent closed circuits necessitating mixing of blood through the VSD, PDA, ASD to prevent severe cyanosis and metabolic acidosis. [3, 12, 20, 21]

Congenitally Corrected Transposition of the Great Arteries or L Transposition involves both the atrioventricular and ventriculoarterial discordance resulting in a hemodynamically normal condition in the absence of any other cardiac anomaly. [3, 12, 20, 21]

Double Outlet Right Ventricle involves the aorta and the pulmonary artery arising from the morphologic right ventricle secondary to maldevelopment of the conotruncus. [3, 20, 21]

Viscero-atrial Situs and Heterotaxia Syndromes include disturbances in the normal Right-Left asymmetry in the position of the thoracic and abdominal organs. Viscero-atrial situs designated as S (Solitus or normal) and I (Inversus or mirror image of normal). Always associated on the same side are major lobe of the liver, IVC, anatomic right atrium, trilobed lung and eparterial bronchus versus spleen, stomach, descending aorta, anatomic left atrium, bilobed lung and hyparterial bronchus. Any arrangement other than the situs solitus and inversus is termed as heterotaxy. [12]

The two major subtypes are double right sidedness or asplenia syndrome characterized by absence of the spleen, IVC and aortic juxtaposition, bilateral SVC, absent coronary sinus, right isomerism of the atrial appendage, common atrium, bilateral trilobed lung, bilateral eparterial bronchi, associated cyanosis and anomalies of pulmonary venous connection.[12]
Polysplenia syndrome includes double left sidedness with multiple spleens, bilateral SVCs connected to the coronary sinus, left isomerism of the atrial appendages, common atrium or large ostium primum ASD, bilateral bilobed lungs, bilateral hyparterial bronchi, associated abnormalities of the systemic venous connections and less severe CHD. [12]
**Fig. 0:** Fig. 5. CTA MIP Image: Pulmonary Artery hypoplasia with RVOT stenosis

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Fig. 0: Fig.6. 3D VR Image: Subvalvular pulmonary stenosis

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**Fig. 0:** 3D VR Image: Sizeable aortopulmonary collateral in a patient with stenosis of the RVOT

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**Fig. 0:** Fig. 7. 3D VR Image: Major aorto pulmonary collaterals in a patient with Tetralogy of Fallot and right sided aortic arch

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**Fig. 0:** Fig.8. 3D VR Image: Transposition of the great Arteries (D Transposition) with the aortic root and the pulmonary trunk parallel to each other in the sagittal plane.

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Conclusion

High temporal and spatial resolution with multiplanar imaging are essential for satisfactory evaluation of CHD. Faster scanning, thinner slices, and improvement in intravenous contrast enhancement of DSCT now offer the unique opportunity to acquire, reformat, volume render and analyse complex data in pediatric thoracic CT angiography. This pictoral essay depicts our experience with dual source CT in 120 cases of congenital heart disease and highlights a systematic approach towards step by step analysis of extracardiac, cardiac and connection abnormalities in pediatric congenital heart diseases.

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