Illustrative Three Dimensional CT Spectrum of Congenital Heart Diseases

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Learning objectives

Three dimensional Volume Rendered and Maximum Intensity Projections on MDCT provide enhanced delineation of complex cardiovascular and extra cardiac morphology for instituting the appropriate interventional or surgical management.
Background

Echocardiography is the initial screening modality in a case of congenital heart disease. But the use of this modality is encumbered by the limited ability to delineate great arteries and extra cardiac anomalies, pulmonary veins and coronary arteries. Diagnostic cardiac catheterisation has small but well known risk and it is usually performed if echocardiography fails to provide a confident evaluation.\(^1\) MR imaging is often limited in evaluation of seriously ill or uncooperative patients, is contraindicated for patients with pacemaker, is time consuming and may require sedation.

The role of CT is well established in evaluation of congenital heart disease but little has been reported on the role of three dimensional reconstructions which make it an unparalleled one-stop-modality for extensive and complete delineation of the anomalies.\(^2\) The different anatomic structures (i.e., heart, great vessels, lungs and abdomen) can be evaluated simultaneously in one acquisition.

The technologic advances have produced diagnostic quality images with increased speed, markedly decreased the sedation time, and ease of peripheral venous access without angiography complications. The reconstructed images such as multi planar reformations (MPR), maximum intensity projections (MIP) and volume rendering technique (VRT) with ability to review images by radiologists and the cardiologists repeatedly and in any desired plane are invaluable for planning treatment.

We have a spectrum of Atrial and Ventricular Septal Defects, Tetralogy of Fallot, Tricuspid Atresia, Single Ventricle, Double Aortic Arch, Right sided Aortic Arch, Coarctation of Aorta, Patent Ductus Arteriosus, Total and Partial Anomalous Pulmonary Venous Drainage, Abernethy Syndrome, Situs Ambiguous, Situs Inversus with Dextrocardia, Pulmonary Veno Occlusive Disease, Williams Syndrome and a combination of these defects.
Imaging findings OR Procedure details

All the studies were performed on the reference of the paediatric cardiologists to answer specific questions raised by either an inconclusive echocardiographic or angiographic evaluation, and for evaluation of the pulmonary vasculature and extracardiac anomalies.

Pulmonary Angiography was performed on 128 slice CT. A collimation of 64 X 0.625mm, slice thickness 0.9mm, pitch 0.8, rotation time 0.5second,120 kV,100 mAs was used with a 512 matrix. The dose (1-2mg/kg) of contrast material (Ioversol 300mg) and the rate (2.5ml/second at a PSI of 150-200) of intravenous administration varied according to patient weight using pressure injector. The paediatric patients were sedated with orally administered chloral hydrate (50-100 mg/kg) and occasionally light intravenous sedation with midazolam (0.1-0.2mg/kg) and/or propofol (1-2mg/kg) for 4-5 minutes by an anaesthesiologist.

Segmental analysis of congenital heart disease

It was introduced 25 years ago by Van Praagh\(^3\). This approach is flexible, easy to understand and applicable to any imaging modality. A clear understanding of the anatomy, the visceroatrial situs, ventricular loop, correct identification of the great vessels, atroventricular and ventriculoarterial connections, the pulmonary and systemic venous return patterns, and major aortopulmonary collaterals is needed for planning effective management.

The anomalies are assessed at the cardiac level for presence, size, and location of atrial and ventricular septal defects, degree of any ventricular outflow tract stenosis, followed by evaluation of aorta and pulmonary vasculature for hypoplasia or stenosis.

Cardiac Abnormalities

Atrial Septal Defect (ASD) (Fig. 1 on page 25)

ASD represents 10% of all congenital heart disease and 20-40% of adult congenital heart disease. Imaging features include enlargement of the right atrium and ventricle, dilatation of the pulmonary artery and its branches and an atrial septal defect depending upon the location. There are 4 types of ASD: ostium secundum(75%), ostium primum(15-20 %),
sinus venosus in the upper aspect of the atrial septum (5-10%) and coronary sinus type (<1%).

Fig. 1: 4 year old child with Down’s syndrome with Atrial septal defect. Axial and coronal reformatted images show abnormal communication between the atria. The right atrium is enlarged and left atrium is small in size

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Ventricular Septal Defect (VSD) (Fig. 2 on page 25)

This entity was first described by Roger in 1879. It represents the most common congenital heart disease in the paediatric population and the second most common defect in the adult population. The incidence is 1 in 1000 live births. 50% are associated with other congenital malformations.
Fig. 2: Perimembranous Ventricular septal defect. Axial and sagittal multiplanar reformatted image reveals a small communication between the right and left ventricles at the subaortic septum level.

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

VSD is described as a hole or holes of variable size in the interventricular septum. It is classified in terms of their appearance from the lumen of the right ventricle as perimembranous, muscular and doubly committed juxta arterial defects.

Tetralogy of Fallot (Fig. 3 on page 26, Fig. 4 on page 26)

It is the most common cause of cyanotic congenital heart disease. It occurs in 10% of cases of congenital heart disease. The incidence is 3–6 infants for every 10,000 births. Imaging features includes infundibular stenosis, ventricular septal defect, overriding of the aorta, right ventricular hypertrophy. Antero-superior deviation of the infundibular septum is considered the developmental cause for subpulmonary infundibular stenosis and VSD in tetralogy of Fallot. Associated anomalies include a right-sided aortic arch with mirror imaging branching (25% of cases), atrial septal defect (10% of cases, referred to as Pentalogy of Fallot), coronary artery anomalies (10% of cases) and stenosis of the peripheral pulmonary arteries.
Fig. 3: Axial and oblique sagittal and coronal images showing (A) absence of pulmonary trunk (B) Ventricular septal defect (C) overriding of aorta (D) MAPCA arising from the descending aorta

References:
Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN
Fig. 4: 3D Volume Rendered image showing large MAPCA from descending aorta giving rise to left pulmonary artery

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Major Aortopulmonary Collateral Arteries (MAPCAs) (Fig. 5 on page 27 & Fig. 6 on page 28)

MAPCAs are frequently the main sources of pulmonary flow in patients with Tetralogy of Fallot with pulmonary atresia. These may be seen in cases of pulmonary hypoplasia or stenosis. The vessels may originate from the ascending, arch or descending aorta and may course to the right or left to reinforce the narrowed pulmonary artery.
Fig. 5: Axial and Coronal Maximum Intensity Projections show multiple large collaterals arising from the aortic arch and descending thoracic aorta on either side giving rise to the pulmonary vasculature. The main pulmonary artery, right and left pulmonary arteries are not visualised

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Fig. 6: Volume Rendered Coronal and Oblique Sagittal images show multiple large collaterals arising from the aortic arch and descending thoracic aorta on either side giving rise to the pulmonary vasculature. The main pulmonary artery, right and left pulmonary arteries are not visualised

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Tricuspid Atresia (Fig. 7 on page 28 & Fig. 8 on page 29)
Tricuspid Atresia has a prevalence of 0.3-3.7% of all congenital cardiac diseases. It represents the third most common form of cyanotic congenital heart disease. It results from failure of formation of tricuspid valve with direct communication between the right atrium and right ventricle. Imaging features include fat deposition in the right atrioventricular groove, small right ventricle, large right atrium and supracristal VSD.\textsuperscript{6}

![Fig. 7: Axial and Coronal multiplanar reformatted images show absence of pulmonary trunk, and Blalock Taussig shunt between the right pulmonary artery and right carotid artery. The tricuspid valves are not visualised. The right ventricle is hypoplastic](image)

**References:** Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN
**Fig. 8**: Volume Rendered image showing Blalock Taussig shunt between the Right pulmonary artery and Right carotid artery

**References**: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

**Ebstein anomaly** *(Fig. 9 on page 30)*

It has a prevalence of less than 1% of congenital heart diseases and occurs in 1 in 210,000 live births. Ebstein anomaly is defined as displacement of the attachment of the tricuspid valve leaflets from the atrioventricular junction to the right ventricular cavity with atrialization of the inlet of the right ventricle. Displacement of the attachment of the tricuspid valve usually involves the septal and posterior leaflets and is maximal at the commissure between the two leaflets.
Fig. 9: Oblique Sagittal and Axial reformatted images reveal displacement of septal and posterior leaflets of tricuspid valve from the atrioventricular junction. Volume Rendered image shows dilated right atrium and other chambers

**References:** Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

**Double Outlet Ventricle with Situs Inversus** (Fig. 10 on page 31)

A double outlet ventricle requires that more than 50% of both great arteries to arise from either the morphologic right or left ventricle. Double outlet right ventricle is more common than double outlet left ventricle.
Fig. 10: Volume Rendered, Axial and Coronal reformatted images show single right ventricle, Dextrocardia, right sided aortic arch and descending thoracic aorta, hypoplasia of pulmonary arteries and situs inversus

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Situs inversus is a rare anomaly with a prevalence of 0.01%. There is mirror image of the normal arrangement of the organs in the chest and abdomen. It is usually associated with dextrocardia which has an incidence of 3 - 5% of congenital heart disease. It is most commonly associated with transposition of the great vessels. The incidence of congenital heart disease is 95% in situs inversus with levocardia. Up to 20% of patients with situs inversus can have Kartagener syndrome which comprises a subgroup of primary ciliary dyskinesia.

Extra Cardiac Abnormalities

Double Aortic Arch (Fig. 11 on page 32 & Fig. 12 on page 33)
It is caused by persistence of Right and Left IV branchial arches. Anterior and posterior arches encircle the trachea and esophagus in a tight ring, joining distally to form a common descending aorta. It is the most common of the complete vascular rings, causing tracheo-esophageal compression. It is rarely associated with Congenital Heart Disease. Patients with this anomaly usually have severe respiratory symptoms and some swallowing difficulty.

Fig. 11: Axial and Coronal Maximum Intensity Projections show right and left aortic arches forming a complete ring around the trachea

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN
Fig. 12: Volume Rendered images show right and left aortic arches forming a complete ring around the trachea.

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Coarctation of the Aorta (Fig. 13 on page 34)

It accounts for 5-10% of all congenital cardiac lesions and approximately 7% of critically ill infants with heart disease. Aortic coarctation refers to a constriction of the aortic arch in the region of the embryologic ductus arteriosus. On CT, it is visible as an indentation of the aorta, with prestenotic and poststenotic dilatation of the aorta.5 50-85% of patients may have a bicuspid aortic valve. There is a strong association with PDA, VSD, and subaortic obstruction.
Fig. 13: Sagittal Maximum Intensity Projections and Volume Rendered image show narrowing in supraductal aorta

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Williams Syndrome (Fig. 14 on page 35)

Williams syndrome was first identified in 1961 by J. C. P. Williams. It has an incidence of 1 per 7,500-20,000 births. Associated features include oral abnormalities, short stature (50% of cases), mild to moderate mental retardation, supra valvular aortic stenosis, pulmonary artery stenosis, renal insufficiency and hypercalcaemia. Cardiovascular disease accounts for most cases of early mortality associated with Williams syndrome. Factors implicated in sudden death include aortic or pulmonary stenosis, myocardial ischemia secondary to either coronary insufficiency or biventricular outflow tract obstruction with ventricular hypertrophy.
**Fig. 14**: Volume Rendered images reveal narrowing of the proximal descending thoracic aorta; main, right and left pulmonary arteries.

**References**: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Another patient of Supravalvular Aortic Stenosis(below) (Fig. 15 on page 36 & )
Fig. 15: Axial and Sagittal Reformatted images show hypertrophy of the myocardium of the left ventricle and marked narrowing of supravalvular ascending aorta.

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN
**Fig. 16:** Volume Rendered images show marked narrowing of the supravalvular aorta. Mild ectasia of right and left coronary ostia is also noted.

**References:** Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

**Patent Ductus Arteriosus** (**Fig. 17** on page 37)

PDA is defined as persistent patency of the ductus arteriosus beyond functional closure after birth. It has a prevalence of 0.02% and 0.006%. Uncomplicated PDA connects the proximal descending aorta below the origin of the left subclavian artery with the roof of the main pulmonary artery near the orifice of the left pulmonary artery.
**Fig. 17:** Maximum Intensity Projection and Volume Rendered Oblique Sagittal images show Patent Ductus Arteriosus.

**References:** Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

**Abnormal Pulmonary Venous Connections**

**Total Anomalous Pulmonary Venous Connection (TAPVC) (Fig. 18 on page 38)**

It represents 1.5 % of all cardiovascular malformations with a prevalence of 6.8 per 1,00,000 population. It represents 1.5 % of all cardiovascular malformations with a prevalence of 6.8 per 1,00,000 population. There is a 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. The connection may be supra cardiac, cardiac, infra cardiac or mixed.
Fig. 18: Coronal and Oblique Coronal Maximum Intensity Projections and Volume Rendered images show superior and inferior pulmonary veins opening into a common vertical vein.

References: Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

Partial Anomalous Pulmonary Venous Connection (PAPVC) (Fig. 19 on page 39)

It is a rare congenital cardiac defect with prevalence of 0.4-0.7% of all cardiovascular disease. It is twice as common from the right lung as from the left lung. Its associations may include ASD (80-90% cases), complex congenital heart disease (heterotaxia),
Scimitar syndrome (right pulmonary vein to inferior vena cava with lung sequestration) or an intact atrial septum (isolated PAPVC).  

![Fig. 19: Coronal Maximum Intensity Projection and Volume Rendered images show left superior pulmonary vein draining into the innominate vein](image)

**References:** Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

**Pulmonary Veno-occlusive Disease (Fig. 20 on page 40)**

Pulmonary vein stenosis is a rare condition with a bimodal age distribution ranging from eight weeks of life to seventh decade. In pediatric patients, the disease may be due to inadequate embryological connections between the intrapulmonary venous system, that
is between the common pulmonary vein and the left atrium. Approximately 50% of the patients with primary pulmonary vein stenosis have some associated cardiac defect.\textsuperscript{9} It is therefore necessary that the protocol for evaluation of patients with congenital heart disease should specifically include the pulmonary veins.

\textbf{Fig. 20:} Coronal Volume Rendered and Maximum Intensity Projections and Axial reformatted images show marked stenosis of the left inferior pulmonary vein and narrowing of the right upper lobar artery

\textbf{References:} Radiology, Sir Ganga Ram Hospital, Sir Ganga Ram Hospital - New Delhi/IN

\textbf{Abnormal systemic venous connections}

\textbf{Abernathy malformation} (\textbf{Fig. 21} on page 41)
It is an extremely rare anomaly of the splanchnic venous system in which mesenteric & splenic venous blood bypasses the liver and drains into the renal veins or the inferior vena cava. There are two types of Abernathy malformation. In type 1 there is complete diversion of portal blood into the systemic circulation with absent intrahepatic portal branches. It may be associated with Interrupted IVC with azygous continuation, left isomerism with polysplenia, right aortic arch with double SVC. In type 2, the intrahepatic portal vein is intact, but some of the flow is diverted into a systemic vein through a side-to-side shunt.\textsuperscript{10}

**Fig. 21:** Patient referred as a case of atrioventricular discordance on Echocardiography. Oblique Coronal Maximum Intensity Projections, Volume Rendered and reformatted images show situs ambiguous with left isomerism and polysplenia, right sided aortic arch and descending thoracic aorta with mirror image branching, double superior vena cava, interrupted inferior vena cava with azygous continuation

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Conclusion

Even today Level II antepartum scans are not a routine at many places. As a result, many congenital heart diseases go undetected. Moreover, patients with these anomalies may have distorted thoracic cage anatomy and/or cardiac malposition that limit the usefulness of echocardiogram. These patients benefit immensely by MDCT.

Low dose MDCT with paediatric protocol has the advantage of less radiation in patients who are going for repeated studies for congenital heart disease.

There is better delineation of aortic and pulmonary vessels morphology by three dimensional reconstructions on MDCT. Pulmonary and systemic venous returns, measurement of coarctation of aorta, visualisation of major aortopulmonary collaterals are also accurately done on CT angiography. There is better assessment of patients after shunt procedures.

Reformatted images from MDCT can accurately and systematically delineate the normal and pathologic morphologic features of the cardiovascular structures. The capabilities of MDCT imaging for depicting this anatomy ensure an increasing role for radiologists.

Hence, Low dose high resolution MDCT images manipulated by Maximum Intensity Projections/Volume Rendered/Multiplanar reformations depict the congenital cardiac anomalies with greater ease and understanding, thus helping in institution of correct and timely interventions and treatment.


