Strengths and limitations of imaging techniques in congenital cardiac diseases: How to proceed?

Poster No.: C-1714
Congress: ECR 2016
Type: Educational Exhibit
Authors: A. Cherif¹, N. Mama¹, A. Houda¹, A. Ben Abdallah¹, Y. BEN CHEIKH², A. Berrich¹, K. Mrad Dali³; ¹SOUSSE/TN, ²Nimes/FR, ³Hammamfousse/TN
Keywords: Ultrasound, CT, Vascular, Arteries / Aorta, Cardiac, Imaging sequences, Catheters, Congenital
DOI: 10.1594/ecr2016/C-1714

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Learning objectives

- To consider the technical approaches to imaging congenital heart disease in neonates and infants with emphasis of changes to the general CTA techniques needed for this group of patients
- To illustrate main congenital cardiac diseases assessed by CT.
- To compare Cardiac CT versus echocardiography in the study of congenital cardiac diseases: limits and added value
Background

Transthoracic echocardiography is the first-line modality for cardiovascular imaging in children with congenital heart disease.

A detailed understanding of the spatial relationships of cardiac structures and associated vascular abnormalities, necessary to plan treatment, is rarely adequate unless proceeding to further non invasive imaging means such as Cardiac CT and MRI.

I-Epidemiology :

Congenital heart disease is common, occurring in 8 of 1000 live births. With the successes in cardiothoracic surgery over the past 3 decades and the ongoing improvements in the diagnostic, interventional, and critical care skills of pediatric cardiologists, #90% of children born with heart defects now survive to adulthood.

II-Pathophysiology :

A. Cardiac malformations

1) Atrial Septal Defect

The formation of the atrial septum is a complex process, consisting of the growth and partial reabsorption of 2 tissue membranes, septum primum and septum secundum; the fusion of these membranes to the forming endocardial cushions; and the reabsorption of the fetal sinus venosus into the structure that will ultimately become the RA.

An error in this developmental process will result in a defect in the wall separating the 2 atria, an atrial septal defect (ASD). There are a number of types of ASD including four major types distinguished according to their location within the septum, Fig. 1 on page 9

· Secundum ASD
  o 60-90% of all ASDs
  o Usually an isolated abnormality

· Primum ASD
5-20% associated with cleft anterior mitral valve leaflet (partial atrioventricular septal defect)

- **Sinus venosus**
  - 5%
  - Associated with anomalous right pulmonary venous return to the superior vena cava or right atrium.

- **Coronary sinus type ASD** ("unroofed coronary sinus") <1%

2) **Ventricular Septal Defect**

Ventricular septal defect (VSD) is the most common form of congenital heart defect in children, accounting for #20% of human cardiac malformations.

This is based on the complexity of the embryological development of the ventricular septum, which involves the fusion of multiple distinct septal components.

The membranous septum, the site at which all of these components fuse, sits behind the septal leaflet of the tricuspid valve and immediately below the aortic valve in the LV outflow tract.

The perimembranous type of VSD, which accounts for #80% of all VSDs, occurs in this location. Defects of the muscular septum are the next most frequent, are the result of excessive fetal muscular resorption, can be single or multiple, and can be located anywhere in the muscular septum.

Endocardial-cushion-type VSDs (associated frequently with primum ASD as part of a complete AV canal defect), malalignment defects, and other defects of the outlet septum are less common, this is often associated with AV valve insufficiency, and outflow defects often are associated with aortic insufficiency, which further complicate the physiological consequences of the lesion.

3) **Patent Ductus Arteriosus**

The ductus arteriosus, , is a critical component in the fetal circulation. It is a tubular arterial structure connecting the aorta and main pulmonary artery. In utero, the ductus allows blood flow from the RV to bypass the nonfunctioning lungs to return to the placenta via the descending aorta. Within 72 hours of birth, the ductus closes in most newborns through
the contraction of an arteriolar smooth muscle layer, a mechanism signaled by the rise in postnatal systemic oxygen levels.

If the lumen of the ductus is not fully obliterated, an arterial connection remains between the systemic and pulmonary circulations, a "patent" ductus arteriosus (PDA).

4) Coronary Fistulas

Communications between the coronary arteries and the cardiac chambers (coronary-cameral fistulas) or low-pressure veins (coronary arteriovenous malformations) are most often congenital in nature. They also may be acquired secondary to trauma or from invasive cardiac procedures such as pacemaker implantation, endomyocardial biopsy, coronary artery bypass grafting, or coronary angiography. Fistulas may arise from any branch of the coronary artery system. In congenital fistulas, drainage is most often to the RV, RA, or the pulmonary arteries and less frequently to the superior vena cava, coronary sinus, pulmonary veins, or LA.

5) Pulmonary Arteriovenous Malformations

Pulmonary arteriovenous malformations (AVMs) are abnormal connections between branches of the pulmonary arterial and pulmonary venous systems that bypass the small arterioles and the air-containing spaces of the lung. They occur most frequently as part of the hereditary hemorrhagic telangiectasia syndrome, and may occur as isolated or multiple defects that develop later in life.

Isolated pulmonary AVM also may be congenital in nature or secondary to trauma or infection.

6) Ebstein’s Anomaly of the Tricuspid Valve

Ebstein’s anomaly of the tricuspid valve (TV) is a well-described congenital malformation in which the septal leaflet of the TV is conjoined to the septal surface well below the valve annulus into the body of the right ventricle.

The other 2 leaflets of the valve elongate to coapt with the abnormal septal leaflet, displacing the resulting coaptation point into the RV outflow tract.

7) Transposition of the Great Arteries
D-transposition of the great arteries (D-TGA), another early embryological malformation, results in the embryological inversion of the great arteries in an otherwise normally developing heart.

The aortic root is positioned anterior and to the right of the pulmonary artery ("D" transposed) and becomes the outlet for the RV. The pulmonary artery arises from the LV.

Transposition of the great arteries (L-TGA) is the result of abnormal ventricular looping of the developing fetal heart, in which the primitive ventricle moves to the right and the bulbus cordis moves to the left. Because these structures are the precursors of the LV and RV, respectively, the morphological RV and TV end up on the left side of the heart, receiving blood from the left atrium (LA) and pumping blood to the aorta, whereas the morphological LV and mitral valve receive inflow from the RA and deliver deoxygenated blood to the lungs.

The aortic root, arising from the RV, is anterior and leftward of the pulmonary artery root.

The inverted atrioventricular (AV) connections, combined with the reversed ventriculoarterial connections, "correct" the flow pattern such that deoxygenated systemic venous blood flows to the lungs, and oxygenated pulmonary venous blood is pumped to the systemic circulation.

Although ventricular septal defects (VSDs) and pulmonary outflow obstructions are frequently associated with TGA, patients without associated lesions have nearly normal cardiac physiology and may easily be undiagnosed until adulthood.

8) Tetralogy of Fallot

Tetralogy of Fallot is the most common complex cyanotic congenital heart lesion. It also has the longest surgical history and the most intensively studied outcomes and follow-up data of all congenital cardiac anomalies. Adult patients with unrepaired tetralogy of Fallot are extremely rare; however, in areas where patients have no access to health care, particularly in developing countries, some of these patients may survive to adulthood.

*The 4 distinct components of the tetralogy, as described in 1888 by Fallot, include:

- VSD,
- subpulmonary stenosis,
- overriding aorta, and
- RV hypertrophy.
However, embryologically, the defect appears to be a single developmental error that involves the terminal portion of the spiral septum, which divides the primitive truncus arteriosus from the pulmonary artery. In tetralogy of Fallot, this muscle bundle is displaced rightward and anteriorly, which precludes correct fusion with the growing muscular ventricular septum, narrowing the pathway from RV to pulmonary artery and enlarging the aortic root such that it extends over the RV outflow tract. The RV hypertrophy is a secondary response to increased afterload. Obstructions in the branch pulmonary arteries, coronary anomalies, a right-sided aortic arch, and additional VSDs are common additional associations.

B. Vascular abnormalities:

1. Coarctation of the aorta

Coarctation of the aorta causes mechanical obstruction of blood flow from the left ventricle. High pressure proximal to the coarctation is usually seen.

development of collateral vessels to bypass the obstruction; the most common collaterals are the internal mammary and the intercostals. Finally a left ventricular hypertrophy develops in response to the elevated pressure.

Coarctation of the aorta is usually congenital, but may be acquired as in Takayasu's arteritis. It is also associated with Williams Syndrome, Turner syndrome, and with other congenital heart diseases.

2. Pulmonary atresia

Pulmonary atresia is a congenital heart defect in which the pulmonary valve does not develop normally or remains blocked after birth.

the right ventricle of the heart, the pulmonary artery, and the tricuspid valve may be underdeveloped. Some infants may also have a ventricular septal defect,

The left ventricle pumps this mixture of oxygen poor blood into the aorta and out to the body. The infant appears blue (cyanotic) because there's less oxygen in the blood. The only source of lung blood flow is the patent ductus arteriosus.

3. Anomalous pulmonary venous return
In normal circulation, blood is sent from the right ventricle to pick up oxygen in the lungs. It then returns through the pulmonary (lung) veins to the left side of the heart, which sends blood out through the aorta and around the body.

In Total Anomalous Pulmonary Venous Return, oxygen-rich blood returns from the lungs to the right atrium or to a vein flowing into the right atrium, instead of the left side of heart. In other words, blood simply circles to and from the lungs and never gets out to the body.

**III-Imaging modalities:**

- Chest RX
- Ultrasonography : ETT, EET, Doppler
- CT
- MRI

**CARDIAC CATHETERIZATION**
Fig. 1: types of ASD

Findings and procedure details

I. Main imaging modalities

A/Chest RX

Not specific imaging modality, but some signs are suggestive of particular congenital heart disease;

- **Transposition of the great arteries**: Ovoid heart configuration, narrow vascular pedicle, increased pulmonary vascularity. Fig. 2 on page 18

The heart appears globular due to abnormal convexity of the right atrial border and left atrial enlargement and therefore appears like an egg; the superior mediastinum appears narrow due to stress induced thymic atrophy and hyperinflated lungs which gives the picture of an egg suspended by a string "egg on a string").

- **Tetralogy of Fallot**: Wooden-shoe heart, pulmonary hypovascularity. Fig. 3 on page 18

- **Persistent ductus arteriosus**: pulmonary artery, left atrium, left ventricle and ascending aorta enlargement, increased pulmonary vascularity

- **Coarctation of the aorta**: Cardiomegaly, Left Ventricular hypertrophy, costal indentation

B/US:

**Transthoracic echocardiography** (TTE) is the first-line cardiovascular imaging modality in congenital heart disease patients, being particularly suitable for the real-time interrogation of relatively small, thin and mobile intracardiac structures, leaflets, septa, jets and infective vegetations, which may be less well seen on CT or MRI.
Transoesophageal echocardiography (TEE) has the advantage of access to more posterior parts of the heart, particularly for three-dimensional (3D) visualisations of the valves, and is suitable for intra-operative use.

Doppler echocardiography has the important advantage of being able to measure the velocities, and hence the presumed intercavity pressure difference, of relatively narrow jets of tricuspid or pulmonary regurgitation.

Echocardiography is limited by a small field of view, an acoustic window, and operator dependence.

**C/CT (computed tomography):**

CT plays an important role in delineating the morphologic features of the heart, great vessels, and the other vascular structures, with advantages over other imaging modalities; CT provides added value in its evaluation of the anatomy and function of the right ventricle and can provide precise information about cardiac and vascular anatomy.

CT offers excellent spatial resolution. It is well suited for imaging the epicardial coronary arteries and their relations to adjacent structures or conduits.

ECG-gated cine CT allows:

- measurements of biventricular size and function
- adequate opacification of each intraventricular blood volume.
- Visualisation of coronary and collateral arteries
- Visualization of conduits and stents
- Visualization of associated lung pathology.

CT takes less time and has fewer requirements for sedation than does MR imaging, it can be more easily performed in an unstable patient who needs intensive monitoring and care.

**Methodological limitations**

No flow measurement
Limited cine visualization

Ionizing radiation; its associated risk of cancer; This risk is dose, age and gender dependent, and makes repeat CT examinations or studies in young patients unattractive

II. Main cardiac malformations:

A. Atrial septal defect

1. US

a) Benefits:

- Excellent visualization of defects of the atrial septum. Fig. 4 on page 19
- Associated mitral valve prolapse may be identified.
- In ostium primum atrial septal defect, 2D echocardiography is considered the standard for the diagnosis.

Detecting and characterizing a double-orifice mitral valve, an association that occurs in approximately 3% of patients with ostium primum atrial defect.

b) Limitations

Echocardiography may fail to provide adequate information in patients with associated structural cardiac abnormalities.

2. CT:

Transverse tomography provides clear spatial separation of the inflow and outflow portions of the atrial and ventricular septa.

As a result of the absence of overlying structures defined on CT scans and the 3-dimensional (3D) nature of ultrafast CT acquisition, the size of the atria and ventricles can be measured.

B. Ventricular septal defect:

1. US:
Doppler color-flow and 2D mapping may be used to identify the type of defect in the ventricular septum. Fig. 5 on page 20

2. CT:
Allows direct visualisation of the defect on contrast CT

C. Tetralogy of Fallot

1. US
Echocardiography is the primary imaging method for examining a child in whom tetralogy of Fallot is suspected.

Intracardiac anomalies, including pulmonary infundibular and valvular stenosis and the position of the aortic root overriding the ventricular septal defect, are identified with 2-dimensional echocardiography. The origins of the coronary arteries can also be identified.

Doppler ultrasonographic examination of the pulmonary outflow tract can be used to measure the velocity gradient in the right ventricular outflow tract and to differentiate severe stenosis from atresia. Fig. 6 on page 21

2. CT

CT can give precise information about:
- Pulmonary stenosis Fig. 7 on page Fig. 8 on page 22
- The position of the aorta, overriding the ventricular septal defect
- Origins of the coronary arteries

This modality is useful also for the evaluation of surgical complications such as infection or pseudoaneurysm formation. Helical CT scanning can be used to identify airway compression that is caused by a large ascending aorta that is associated with tetralogy of Fallot.

The origins of the coronary arteries can also be identified.

D. Ebstein's anomaly

1. US:
Elongated tricuspid valve leaflets (especially the anterior leaflet)

Dilated right ventricular cavity, abnormal septal motion

Low-velocity tricuspid regurgitation, which reflects the usual inability to generate high pressure in the right ventricle, is common

The severity of the lesion can be estimated from the degree of right atrial and ventricular dilation and from marked apical displacement of the tricuspid valve leaflets.

Echocardiography is an excellent method for assessing Ebstein anomaly, and it may be the first and only cross-sectional imaging investigation that is required

2. CT:

On a cardiac CT scan, the right atrium and ventricle appear dilated. A variable degree of apical displacement of the tricuspid valve attachments can be seen relative to the atrioventricular junction. The atrialized and functional portions of the right ventricle may be discerned. The conspicuity of the myocardium may be greater in the functional portion of the right ventricle than in the atrialized portion. Leaflet thickening may be seen. Functional information can be obtained with retrospective gating at the cost of higher radiation dose

E. Transposition of the great arteries

1. US

The four chamber-view is normal. The great arteries arise in parallel arrangement. The aorta arise close to the anterior wall from the right ventricle and the pulmonary trunk arise posteriorely from the left ventricle

2. CT

visualization of the ventriculoarterial discordance in which the aorta arises from the morphologic right ventricle and the pulmonary artery arises from the morphologic left ventricle

Fig. 9 on page 23 Fig. 10 on page 24

F. Anomalous pulmonary venous return

US
In cases of anomalous pulmonary venous return, the echocardiogram demonstrates a large right ventricle; in addition, a pattern of abnormal pulmonary venous connections is usually seen. The demonstration of a vessel in the abdomen with Doppler venous flow away from the heart is pathognomonic of total anomalous pulmonary venous return (TAPVR) below the diaphragm.

**CT**

Utilisation of contrast-enhanced studies with MDCT technology, enables both detection and characterization of the anomalies. It is considered the imaging modality of choice.

Fast CT is useful for defining pulmonary drainage. The degree of confidence is good for anomalous pulmonary venous return. False-positive and false-negative results are rare in anomalous pulmonary venous return.

*Fig. 11 on page 25, Fig. 12 on page, Fig. 13 on page 26*

**III - Congenital aortic disease:**

**A. Persistent ductus arteriosus**

1. **US**

The findings demonstrate normal-sized right heart chambers unless pulmonary hypertension is present.

In suitable patients, especially the young, the PDA can be visualized directly between the distal arch aorta and pulmonary artery at the origin of the left pulmonary artery *Fig. 14* on page 27.

A left-to-right shunt is demonstrated using contrast echocardiography.

2. **CT**

A PDA may be visible on CT angiography, it requires the use of ionizing radiation and usually intravenous contrast agents. Occasionally, calcification of the PDA is demonstrated in a characteristic position. CT angiography may be used to image the aorta for possible aneurysm.
The degree of confidence is high in detecting complications of PDA, such as ductus arteriosus aneurysm. Fig. 15 on page 28 Fig. 24 on page 30

B. Coarctation of the aorta

1.US:

Transthoracic echocardiography is ideal for the initial assessment, since it provides comprehensive assessment of valvular and ventricular function, in addition to reliable assessment of the pressure gradient across the coarctation.

The segment of coarctation may usually be visualized by means of 2-dimensional echocardiography.

Color Doppler imaging is useful for demonstrating the specific site of the obstruction.

Associated anomalies of the mitral and aortic valve are associated with hypopulsatile descending aorta

Pulsed and continuous-wave Doppler study may be used to determine the pressure gradient directly at the area of coarctation.

US determine the pressure gradient directly at the area of coarctation

However, in the presence of a patent ductus arteriosus, the severity of the narrowing may be underestimated.

2.CT:

CT assesses pre# and postoperative assessment of the thoracic aorta. CT scan is often extremely useful for neonates with isthmic coarctation associated with hypoplasia of the aortic arch. It precisely identifies the site of the coarctation, determines the degree of narrowing and, above all, defines precisely the extent of hypoplasia of the aortic arch, thereby assisting the choice of surgical technique (simple left posterolateral thoracotomy or midline sternotomy). CT is preferable to MRI for this group of children owing to the simplicity of the examination and the rapidity of image acquisition, generally less than 2 seconds for neonates. Fig. 17 on page 29, Fig. 18 on page 31 Fig. 19 on page 32

C. Interrupted aortic arch
Interrupted aortic arch (IAA) is defined as a complete luminal and anatomic discontinuity between the ascending and descending aorta, as described by Steidele. IAA is rare, accounting for only 1% of congenital heart diseases. It is considered by some authors to be a phenotype of a heterogeneous group of causative factors. Altered hemodynamics through the fourth aortic arch and teratogenic exposure during the intrauterine period have been proposed as its potential causes. It may occur as a simple or complex anomaly. [Fig. 21](#) on page 34

In simple IAA, only ventricular septal defect (VSD) and persistent ductus arteriosus (PDA) are associated. The simple form is more common than the complex form, in which cardiovascular anomalies other than VSD and PDA, such as truncus arteriosus, transposition of the great arteries, double outlet right ventricle, aortopulmonary window, and functional single ventricle, are combined with IAA [Fig. 20](#) on page 33

**D. Aortic arch abnormalities**

A vascular ring is an aortic arch anomaly in which the trachea and esophagus are surrounded by vascular structures. Vascular rings are uncommon anomalies (<1% of all congenital cardiac defects) with a similar frequency in both sexes. They are caused by abnormal persistence or regression of one of the six embryonic aortic arches. The two most common types of complete vascular rings are a double aortic arch [Fig. 22](#) on page 34 and a right aortic arch with left ligamentum arteriosum [Fig. 23](#) on page 35

**IV-Diagnosis accuracy according to imaging modality:**

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<td>Transposition of great arteries</td>
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Images for this section:

**Fig. 2:** patient with transposition of great arteries, Ovoid heart configuration, appearing like an "EGG"

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Fig. 3: Wooden-shoe heart, patient with tetralogy of fallot

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Fig. 4: Atrial septal defect, Doppler echocardiography showing Left to right shunt

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Fig. 5: Ventricular septal defect

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Fig. 6: VSD with overriding aorta

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**Fig. 8**: pulmonary stenosis

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Fig. 9: Aorta arises from the right ventricle.

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Fig. 10: The pulmonary artery arises from the morphologic left ventricle

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Fig. 13

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Fig. 14: Persistant ductus arteriosus

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**Fig. 15:** para sagittal thin slice CT image: persistant ductus arteriosus on the lower edge of the post-isthmic aorta.

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Fig. 17

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**Fig. 24:** VR_Persistant ductus arteriosus

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Fig. 19: CT assesses collateral circulation which is correlated with the severity of the coarctation

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Fig. 20

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Fig. 21: interrupted aortic arch

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Fig. 22: Annular aortic arch

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Fig. 23: Left subclavian artery arising from 4th portion of the aorta and going behind the oesophagus

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Conclusion

Transsthoracic echocardiography is the first-line modality for cardiovascular imaging in children with congenital heart disease, however CT is a very useful imaging modality for the morphologic evaluation of CHD and the assessment of main vascular abnormalities.

Reformatted images from multisection spiral CT can accurately and systematically delineate the normal and pathologic morphologic features of the cardiovascular structures.

Although both US and CT does have some disadvantages in this setting, affected patients can be evaluated with MR imaging.
References


