Role of multi-slice CT angiography in the evaluation of pulmonary venous anomalies

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Abstract  Purpose and aim: To assess the role of MSCT angiography in evaluation of extra-cardiac vascular abnormalities in pulmonary venous anomalies and compare it with echocardiography. Materials and methods: This study included 26 patients with an age range of 15 days–25 years. All of these patients underwent MSCT angiography and echocardiography. Only 10 patients underwent cardiac catheterization.

Results: Our initial experience showed that MSCT is capable of complementing echocardiography and replacing diagnostic cardiac catheterization for anatomical delineation if performed with an optimum technique.

MSCT angiography proved to be a worthy primary investigation tool in patients whom ECHO has been able to clearly identify the intracardiac anatomy, but not the extra-cardiac vascular anatomy.

Conclusion: MDCT correctly depicted the TAPVR (Total anomalous pulmonary venous return) and PAPVR (Partial anomalous pulmonary venous return) types of pulmonary venous anomalies with sensitivity 100%, and specificity 100%. The specificity of echocardiography was 50% for both findings. Inspite of the risk of ionizing radiation and contrast medium injection the adoption of our minimal invasive, low radiation, non-ECG gated protocol greatly reduces the time, radiation dose, and contrast medium volume needed to perform an optimum CT angiographic technique. Thus, paving a clear road map for pre and post operative assessment of patients with pulmonary venous anomalies.

1. Introduction

The traditional method of diagnosis of congenital heart disease has been catheter angiography. While effective in diagnosis, angiography is an invasive procedure. Evaluation of right and left sided defects may require both venous and arterial punctures (1).

Echocardiography (ECHO) is the mainstay of diagnosis of congenital heart disease. Its strength includes an absence of radiation, the ability to evaluate intra-cardiac structure and
function and the ability to perform hemodynamic assessment. However, it is limited in the evaluation of certain portions of the aorta (particularly the ascending aorta and the transverse arch), the distal pulmonary arteries, the right ventricle, and the pulmonary veins (2).

The development of multi-slice spiral computed tomography (MSCT) has increased the clinical use of cardiac CT imaging in patients with pulmonary venous anomalies. Multi-slice CT has the advantages of fast scan speed; high spatial resolution, enabling the acquisition of isotropic volume data; and simultaneous evaluation of airways and lung parenchyma, thus increasing the ability to answer most clinical questions about structural abnormalities in patients with congenital heart disease (3).

Our aim is to assess the role of MSCT angiography in evaluation of extra-cardiac vascular abnormalities in pulmonary venous anomalies and compare it with echocardiography.

2. Patients and methods

This study included 26 patients suspected or diagnosed of having pulmonary venous anomalies on examination and echocardiography. The patient population consisted of 15 males and 11 females. They ranged in age range from 15 days old to 25 years. All patients were referred from pediatric cardiologists in the period from (April-2007) to (February-2010).

Echocardiography examination was performed for all patients prior to CT.

Diagnostic cardiac catheterization was performed in 10 of our patients before performing CT and correlation between the findings was performed.

All patients were referred in order to perform further CT angiography examination of the heart and thoracic vessels and were fitting with the following inclusion and exclusion criteria.

- **Inclusion criteria:**
  1. Limited, incomplete or failure of visualization of pulmonary venous anomalies during echocardiography examination.
  2. Difficulty in demonstrating pulmonary arteries by echocardiography for determination of their presence, size and branching pattern.
  3. Difficulty in visualization of peripheral pulmonary arteries’ branches by cardiac catheter in the presence of proximal branches’ stenosis.
  4. Identify normal and pathologic pulmonary venous anatomy and drainage not adequately visualized by echocardiography especially in patients with unexplained pulmonary hypertension.
  5. Recognize the associated airway, and pulmonary parenchymal findings.

- **Exclusion criteria:**
  1. Clinically unfit patients e.g. Severe asthma.
  2. Contrast hypersensitivity or serum creatinine level > 2 mg/dl.

2.1. **CT angiography of the heart performed included**

- Study planning.
- Patient preparation.
- Technique of Examination.
- Data acquisition.
- Image reconstruction and post processing.
- Image interpretation.

2.1.1. **Study planning**

Consultation with the referring physician was attempted prior to the study to discuss the clinical background of the case and delineate specific questions raised by the physician in order to understand the reason for referral and expectations of the physician from the study. These questions usually included problems encountered with the physician during echocardiography or cardiac catheterization such as failure to delineate the pulmonary vascular tree, the effect of cardiovascular structures on the major airways or lung aeration, as well as the status of the upper abdomen to look for situs abnormalities spleen.

- Review of echocardiographic findings and cardiac catheter reports if available.
- Checking renal functions to exclude patients with a creatinine level above 2.
- History taking from the parents and patients.

2.1.2. **Patient preparation**

- Fasting for 4–6 h in all patients.
- Venous catheters (21–24-gage) were placed in an upper limb vein or the peripheral vein of the foot.
- Sedation by minor general anesthesia was used for patients. The examination was done without the need for sedation in two patients.
- Description of the Procedure to the parents with their reassurance.

2.1.3. **Technique of examination**

2.1.3.1. **Data acquisition.**

- **Scanner:**
  - All studies were performed using a dual-source CT system (SOMATOM Definition, Siemens, Forchheim, Germany).
  - The patient lies on the CT table in supine position.
  - A scanogram is obtained.
  - Scan range: from the root of the neck including the proximal aspects of the common carotid and subclavian arteries down to the level of the portal vein inferiorly. This range is important to detect associated aortic arch branch anomalies, situs inversus, situs ambiguous, abdominal aorta coarctation and infra-diaphragmatic type of total anomalous pulmonary venous drainage.
  - ECG gating: was performed in cases above 18 years of age and in case of suspected aortopulmonary windows.
    - In the rest of the cases ECG gating was not performed to avoid unnecessary excess radiation exposure by the patient.

- **Contrast injection:**
  - Non-diluted, Non-ionic contrast material was used. (Omnipaque 300–350; Nycomed Amersham, Princeton, NJ).
  - Rate of injection: 2–3 ml/kg.
Empiric scan delay between 7 and 15 s from the onset of contrast administration to start of scan acquisition is determined before start of the study.

Sterile syringe for manual injection was used in total volumes up to 20 ml in all patients.

Automatic power injector was used in higher volumes of contrast.

Scan Parameters:
- kVp: low dose protocol 80–100 kVp.
- mAs: 60 mAs.
- Slice thickness: 2 mm.
- Pitch: 1.3.
- Reconstruction increment: 1.5 mm. on- diluted, Non-ionic contrast material was used.

Multi-phase examination of the heart is then performed. Sequential series of images in the mid venous and mid arterial and delayed phases of enhancement was performed to ensure opacification of both sides of the heart and all extra-cardiac vessels. The first series is performed in the caudocranial direction to decrease contrast agent–related artifacts and to achieve homogeneous contrast enhancement while the second one is in the craniocaudal direction. The third one is performed in the caudo-cranial direction again.

Post Procedural Assessment:
- Axial images are rapidly reconstructed at 1.0 mm slice thickness and increment of 0.8 mm and reviewed to ensure satisfactory quality of the images.
- The patient is kept under observation for 15–30 min after the procedure till recovery of sedation.
- Nothing was given by mouth till complete recovery.
- The peripheral venous line is then removed.

2.1.3.2. Image reconstruction and post processing. All images were transferred to the workstation for post processing.

An image containing only the bones is created by using the appropriate threshold level while trying to keep all the data from the vascular structures out of the image to avoid being subtracted in the next step. Then this image containing only the bones subtracted from the source images to get a set of images with no bones within and only vessels is visualized.

Some views were taken before bone subtraction and others were taken afterward. Maximum intensity projections (MIP), three dimensional volume rendering (VR), multiplanar (MPR) and curved planer reformations (CPR) were created at different angles of views.

2.1.4. Image interpretation

Images were interpreted guided by the anatomical and segmental/sequential approach as follows:

I-Cardiac findings include: Situs, AV concordance, VA concordance, Great vessel relationship, Defects, Size and Effusion.

II- Extra-cardiac findings include: Aorta, Pulmonary arteries, Pulmonary venous drainage, Systemic venous drainage, Lung fields and pleural sacs, Upper abdominal cuts abnormalities.

3. Results

This study enrolled 26 patients (15 males and 11 females) with an age range of 15 days–25 years (mean age 5 months).
mality in this study. Six of the cases of PAPVR revealed to be Scimitar syndrome with CT. Echocardiography wrongly diagnosed one case of PAPVR as TAPVR. Agreement between echocardiography and CT in diagnosis of TAPVR was 50%. The findings are clearly shown in Table 2.

For better assessment of TAPVR cases the following table displays the different types of TAPVR detected by echocardiography and CT. Agreement between echocardiography and CT was approximately 50%. Echocardiography failed to demonstrate the four cases of infracardiac type of TAPVR. CT angiography was also of great benefit in diagnosing the different types of TAPVR and detecting the site of connection of the common pulmonary vein. The CT also detected associated stenosis in the common pulmonary vein in two of the infracardiac types of TAPVR as it crosses the diaphragm.

Systemic venous abnormalities detected in the study were classified into left SVC, double SVC and interrupted IVC. (see Tables 3 and 4).

4. Discussion

Congenital heart disease (CHD) occurs in approximately 8 of every 1000 live births, half of whom require surgical or other forms of treatment.

Major advancements in the surgical and medical management of cases with pulmonary venous anomalies allowed a large percentage of these cases to survive to adulthood. This required proper diagnosis and timed management to provide the child with a good chance of leading normal life with minimal if any disability.

An accurate, 3D evaluation of the cardiac and related arterial anatomy is critical for the clinical management of adult and pediatric patients with complex congenital heart disease. 3D imaging has to be able to demonstrate the shapes of, and spatial relationships between, the great arteries, proximal branch pulmonary arteries, and anomalous pulmonary venous or systemic connections. Three-dimensional information about

Fig. 1 (a–c) Represent 45 day old male Complains of shortness of breath and repeated attacks of chest infection On clinical examination Tachycardia and signs of right ventricular hypertrophy Echocardiography shows cardiomegaly with right atrio-ventricular preponderance, PDA, unexplained pulmonary hypertension, Multi-slice CT requested? Aorto-pulmonary window. Multi-slice CT Angiography shows dextroversion, AV and VA concordance, great vessel relationship: normal, Cardiomegaly with right atrio-ventricular preponderance. Anomalous right pulmonary vein (scimitar vein) draining into the IVC, anomalous systemic arterial supply from the upper abdominal aorta to the right lung base, dilated main pulmonary artery and its branches, bilateral basal areas of subsegmental pneumonitis more evident on the right side, small PDA supplying LPA. (a) Sequential thin MIP coronal images showing scimitar vein, anomalous systemic arterial supply from the upper abdominal aorta to the right lung base and right basal subsegmental pneumonitis. (b) Sequential axial images showing dextroversion, scimitar vein, and right basal subsegmental pneumonitis. (c) VRT images showing scimitar vein opening into IVC, anomalous systemic arterial supply from the upper abdominal aorta to the right lung base.
extra-cardiac morphological characteristics may determine subsequent surgical intervention, follow up the residuals of interventions, and assist with estimation of prognosis.

The advent of echocardiography (echo) in the 1970s led to a revolution in the non-invasive diagnosis of heart disease. Echo became the mainstay of diagnosis and follow-up for congenital heart disease, used to determine chamber pressures, oxygen saturation, and pulmonary vascular resistance prior to surgery (4).

In addition to echocardiographic 2-D imaging, the Doppler examination has become an essential component of the complete echocardiographic evaluation. The field of cardiac US continues to grow rapidly: recent clinical additions include 3-D imaging, harmonic imaging, and contrast echocardiography.

Cardiologists and radiologists might think ECHO is the ultimate solution to the evaluation of pulmonary venous anomalies but, according to the current literature, even in first-class cardiovascular centers, the diagnostic accuracy is only around 80%. In the remaining 20% of patients, disease is hiding in the dead spaces of echocardiography, such as the pulmonary veins, aortic arch and peripheral pulmonary arteries.

Echocardiography is a great modality for initial assessment because of its mobility and availability however, it may not be the perfect diagnostic tool because it is usually limited by the acoustic window, spatial resolution, and the subjective interpretation of the operator. An incorrect echo diagnosis might result in the wrong operation and the risk of avoidable mortality, a mistake that could potentially have been corrected by MDCT. For example, after misdiagnosing intracardiac total anomalous pulmonary venous return (TAPVR) as a primary atrial septal defect (ASD), closing the presumed ‘ASD’ could result in pulmonary vein obstruction and immediate death when the cardiopulmonary bypass is removed (5).

Cardiac catheterization has traditionally been the procedure utilized to complement echo, providing hemodynamic information and enabling visualization of extracardiac great vessels. However, the role of diagnostic cardiac catheterization in pediatric cardiology is evolving for a number of reasons.

The major advantages of digital angio-cardiography include the ability to eliminate bony structures by subtraction techniques thus reducing the risk of motion mis-registration artifacts and more importantly to use the sequential data to create quantitative analyses. One of the advantages of the technique is that it is less operator dependant functional data like intra cardiac or intra vascular pressure gradients or blood oxygenation levels are obtainable (6).

On the other hand one of the potential negative repercussions of cardiac catheterization is the invasive nature of procedure (e.g. arterial and venous compromise, stroke, bleeding) and of the exposure to radiation. In many centers, cardiac catheterization is now reserved for patients in whom hemodynamic data are essential (e.g. high risk Fontan, pulmonary hypertension), and/or in whom interventional procedures are necessary.

Krishnamurthy, 2009 (7) reported that cardiovascular magnetic resonance (CMR) imaging plays an important role in the evaluation of patients with complex CHD. It overcomes many of the limitations of echocardiography (e.g., restricted acoustic windows), computed tomography (e.g., exposure to ionizing radiation, limited functional information), and cardiac catheterization (e.g., exposure to ionizing radiation, morbidity, high cost). The direct multiplanar image capability of MR imaging allows precise depiction of the complex and often unexpected cardiac and extracardiac arterial and venous morphologies present. Intracardiac anatomy can be imaged in multiple planes, and functional assessment can be made accurately and with high reproducibility. Finally, cardiovascular MR sur-
passes catheterization, echo and MDCT in its ability to create high resolution, 3D reconstructions of complex CHD, without the use of ionizing radiation.

In 2008, Tsai et al. (5) performed SWOT (strengths, weaknesses, opportunities and threats) analysis of MDCT in a complex congenital heart disease. Strengths included excellent spatial and temporal resolution, fast acquisition time, short schedule time, cheaper than MRI, powerful post processing and good availability. Weaknesses were contrast medium administration and ionizing radiation exposure. Opportunities are that MDCT is rapidly improving and is becoming increasingly popular. It faces threats from echo due to its powerful mobility and great accessibility and from MRI due to lack of radiation.

Multidetector CT angiography (MDCTA) affords several advantages compared with other current imaging options. When compared with echocardiography, MR imaging, or conventional angiography, CT provides the most thorough evaluation of the thorax. This is especially important for the lungs and major airways such as in the setting of pulmonary rings, slings, post operative tracheal narrowing, and atelectasis, as well as in acquired lung and airway disorders. Sedation is also less frequently necessary than in the other three non radiographic cardiovascular imaging modalities. MDCTA is fast, especially with 64-slice (and greater) technology where the entire chest may be done in less than a second, or the entire heart in one rotation. Monitoring is easier than with MRA, as well. The presence of metal artifact is also less of an issue than with MRA. Cost is substantially less than conventional angiography, and procedural risks with conventional angiography must also be a part of the decision process (8).
In our study the role of multi-slice CT angiography in the evaluation of pulmonary venous anomalies was assessed. Hu et al., 2008 (9) reported that by applying the ALARA (as low as reasonably achievable) principle as far as possible in neonates and babies with CHD, can be performed by avoiding a preview scan, using 80-kV settings, adaptation of the mAs to child’s weight as well as systematic protection of non-scanned organs.

Tsai et al., 2008 (5) reported that sedation was necessary to avoid motion artifact. He used 0.2 mg per kg body weight of midazolam (Dormicum; Roche, Vienna, Austria) intravenously. This dose kept neonates sleeping even during injection of contrast agent. Because respiratory inhibition might occur after intravenous administration of midazolam, oxygen should be provided by nasal cannulae if the patient is not intubated.

ECG and blood oxygen saturation monitoring were most important during scanning.

In our study all our patients –except two- were sedated in a similar fashion by minor I.V anesthesia using a weight related protocol under direct supervision of a specialized anesthesiologist.

The two patients not sedated were 18 and 25 years of age. Only reassurance was adequate to carry on technique. No sedation related complications were encountered in the study.

Non-ECG gated acquisition was used in all examinations done to maintain a low dose radiation protocol avoiding unnecessary radiation exposure to the patient. In all studies diagnostic images were obtained allowing accurate interpretation of the extra-cardiac vascular structures.

![Fig. 2 (a, b): Represent 1.5 year old female, Complains of exertional cyanosis, no palpitation. On clinical examination no cyanosis and no clubbing , pansystolic murmur all over pericardium. Echocardiography shows dextrocardia, almost single atrium, pulmonary stenosis, AVSD, DORV, TAPVR to left SVC. Multi-slice CT Angiography shows situs ambiguous with dextrocardia, AV concordance with right atrial isomerism, great vessel relationship: DORV, cardiomegaly with right ventricular hypertrophy and hypoplastic left ventricle, aorta over-riding subaortic VSD by 30-40%, no antegrade continuity between the right ventricle and MPA, confluent hypoplastic pulmonary arteries, four pulmonary veins opening into a common pulmonary venous channel (CPVC) which ascend vertically to open into the left SVC approximately 0.9 cm below the confluence of the brachiocephalic veins, left SVC, Interrupted IVC with the hepatic veins opening directly into the right atrium, asplenia. (a). Sequential axial images showing dextrocardia, four pulmonary veins opening into a common pulmonary venous channel (CPVC) which ascends vertically to open into the left SVC. (b) Sequential coronal images (upper four images) thin VRT images (lower two images) showing dextrocardia and four pulmonary veins opening into a vertical vein opening into the left SVC.](image-url)
Tsai et al., 2008 (5) agreed with our study as he stated that the neonatal cardiac CT dose is around 1.12–1.76 mSv for the non-gated technique and 2.17–3.41 mSv for the gated technique. ECG-pulsing is probably an excellent technique for cardiac CT in adults with slow heart rates, but not for cardiac CT in neonates with very fast heart rates, because the dose reduction generated by ECG-pulsing is inversely related to the patient’s heart rate. The faster the heart rate, the less the dose saved. If only the chamber anatomy and great vessel connections are the issue, a non-gating technique will suffice. If coronary anatomy or detailed assessment of intracardiac structures is needed, gating should be applied.

Goo et al., 2003 (10) reported that CT timed acquisition of volumetric data in arterial or venous phases became possible with the ability to study different segments of the vascular system in multi-planar and three dimensional patterns.

This study included 35 patients who were injected manually by a disposable syringe when the calculated contrast dose was less than 10 cc while in the rest of the patients an automatic pump was used.

In our study we implemented a similar technique but to it we added an extra delayed phase of acquisition. The CT technique included multi-phase examination of the heart. Sequential series of images in the mid venous (right-sided opacification) and mid arterial (left-sided opacification) and delayed phases of enhancement was performed to ensure opacification of both sides of the heart and all extra-cardiac vessels. The first series is performed in the caudocranial direction to decrease contrast agent–related artifacts and to achieve homogeneous contrast enhancement while the second one is in the cranio-caudal direction. The third one is performed in the caudo-cranial direction again. The aim of the delayed phase was to confirm the data obtained in the early phases, give time for stenotic and atretic vessels to fill through their collateral system allowing excellent delineation of these collaterals, post operative assessment and provided better delineation of the pulmonary veins.

Regarding contrast administration, Tsai et al., 2008 (5) stated that for synchronization between the scan and injection of contrast medium we use a bolus tracking technique with the region of interest (ROI) set in the descending aorta. Bolus tracking is well known for its more efficient use of contrast medium and shorter procedure time than the test bolus technique. Nephrotoxicity and sedation risks are also reduced. The reason for using the descending aorta is that it is only located on either the left or right side of the thoracic spine; it is also adjacent to the lung and so the high contrast between lung and descending aorta makes bolus recognition very easy. Furthermore, it is not affected by cardiac motion, unlike the ascending aorta or cardiac chambers. Rate of injection was 3 ml/kg.
Frush and Herlong, 2005 (11) in their study stated that CTA in children is performed with 1.5 ml/kg, with a maximum dose of 3.0 ml/kg.

In our study we injected contrast at a rate of 2 ml/kg utilizing an empiric scan delay technique between 7 and 15 s from the onset of contrast administration to start of scan acquisition determined according to the weight of the patient and congenital anomaly under question. We did not perform automatic bolus tracking as in the majority of cases there was left to right shunts e.g. ASD, VSD and PDA making the process difficult and time consuming.

The scan parameters used in our study were similar to those used by Leschka et al., 2007 (12) who used slice thickness of 2 mm and increment of 1.5 mm during data acquisition while during reconstruction he utilized 1 mm slice thickness and increment of 0.8 mm.

In our study the commonest pulmonary venous anomaly was TAPVR (supracardiac type) accounting for 31% of the cases. Oh et al., 2009 (4) stated that TAPVR supracardiac type accounted for 50% of TAPVR. Oh et al. 2009 (4), stated that in all patients, MDCT correctly depicted the drainage site of the common pulmonary vein, stenosis of the vertical vein and the course of the atypical vessel into the systemic vein (sensitivity 100%, specificity 100%). The specificity of echocardiography was 100% for the three defined findings. The specificity of echocardiography, however, was 87%, 71% and 0%, respectively. An incorrect diagnosis by echocardiography might occur because of a limited acoustic window, insufficient spatial resolution or an interpretation error by the operator. These errors could potentially be reduced by MDCT. According to the recent literature, the diagnostic accuracy of echocardiography in neonatal congenital heart disease is only around 80%. The main disadvantage of CT angiography in patients with TAPVC is radiation dose.

In our study echocardiography detected eight cases of partial anomalous pulmonary venous return (PAPVR) six of which revealed to be Scimitar syndrome. However, echocardiography misdiagnosed a case of TAPVR as PAPVR. As regards the TAPVR there was 50% concordance between echocardiography and CT. Echocardiography failed to demonstrate the four cases of infracardiac type of TAPVR one of which showed stenosis as it crossed the diaphragm. CT angiography was also of great benefit in diagnosing the different types of TAPVR and detecting the site of connection of the common pulmonary vein.

Recognition of systemic venous anomalies especially left SVC is of importance if a left superior venous approach to the heart is considered in patients undergoing pacemaker or defibrillator placement, and in the use of retrograde cardioplegia for surgical procedures requiring cardiopulmonary bypass (13). In 2007 Leschka et al. (12), concluded that although echocardiography is the method of choice for diagnosing the vast majority of congenital cardiac abnormalities, CT plays an increasing complementary role by providing objective and accurate morphologic and functional information and is useful for detecting extracardiac abnormalities thus helping in pre-operative planning and post operative assessment of congenital heart disease patients (Fig. 1 and Fig. 2).

References