



Evaluation of Aortic Arch Anomalies by Echocardiography and CT Angiography, Could CT be the Primary Method of Diagnosis?

H. M. Seif El Dien¹, L. A. Ibrahim^{2*} and R. H. Hashem¹

¹Radiology Department, Faculty of Medicine, Cairo University, Egypt.

²Department of Pediatrics, Faculty of Medicine, Cairo University, Egypt.

Authors' contributions

This work was carried out in collaboration between all authors. Author HMSED designed the study, wrote the protocol, do the CTA and wrote the first draft of the manuscript. Author LAI revised the manuscript, do the echocardiography, managed the literature searches. Author RHH helped in CTA revised the manuscript. All authors read and approved the final manuscript.

Original Research Article

Received 5th January 2014
Accepted 23rd February 2014
Published 19th March 2014

ABSTRACT

Aims: In our study we highlight the importance of noninvasive imaging, stressing the role of CTA over echocardiography in the diagnosis of congenital aortic arch anomalies.

Study Design: Prospective design.

Place and Duration of Study: Radiology and pediatric department, in Cairo University children's hospital, over the period of 15 months from February 2011 to April 2012.

Methodology: Forty five patients (23 males and 22 females) were examined in this study; they were referred to the pediatric cardiology outpatient clinic in Cairo university children's hospital, by history or clinical examination suspicious of aortic arch anomalies. For each patient full history and clinical examination were obtained including: age, sex, residence, consanguinity, cardiac manifestations weak or absent femoral pulsations, abnormal gradient in blood pressure between upper and lower limbs (>20 mmhg). Echocardiography was done and if aortic arch anomalies were diagnosed; the patient referred to do CT angiography of the heart after taking clear consent from the patient's parents.

Results: Our study included 45 patients, their age range (5 days -11 years), they were 23 males and 22 females, with the following diagnosis: 15 had hypoplastic aortic arch by

*Corresponding author: Email: lamyazambely@yahoo.com;

echocardiography, 6 of them proved to have multiple levels of narrowing by CT angiography, 11 patients had aortic coarctation by echocardiography, 3 of them were proved to have more than one segmental narrowing by CT angiography and one patient had hypoplasia of the whole aorta. Six patients had tetralogy of Fallot with right sided aortic arch, mirror imaging branches or aberrant left subclavian artery and hypoplastic pulmonary trunk. Four patients had interrupted aortic arch. One patient had aneurysmal dilatation of ascending aorta and aortic arch. One patient had aortopulmonary window. Six patients had persistent truncus arteriosus.

Keywords: *Echocardiography; MDCT; children; aortic arch anomalies.*

ABBREVIATIONS

CTA: Computerized tomographic angiography; MDCT: Multidetector computerized tomography; PDA: Patent ductus arteriosus; CCA: Common carotid artery; ECA: External carotid artery; IAA: Interrupted aortic arch; SCA: Subclavian artery; CHD: Congenital heart diseases; VSD: Ventricular septal defect; ASD: Atrial septal defect; MIP: Maximum intensity projection.

1. INTRODUCTION

Cardiovascular disease is the first leading cause of death. Importantly, it remains the foremost cause of preventable death globally [1]. Aortic abnormalities are common cardiovascular malformations, accounting for 15%- 20% of all congenital cardiovascular diseases [2]. Diseases of the aorta are for many years chest radiography (CXR) and echocardiography (ECHO) were the first-line imaging modalities used in diagnosing congenital aortic arch anomalies, particularly in children, however echocardiography in addition to being highly operator dependent, it may not be sufficient for adequate evaluation of the great vessels due to acoustic window limitations [3,4]. CT angiography (CTA) is now considered the preferred method of imaging arch anomalies [5]. High-quality 2D reformatted and 3D reconstructed CT images complement axial images and assist in the understanding of complex cardiovascular anomalies [6]. Therefore, CTA is considered an important tool helpful in establishing the primary diagnosis, defining anatomic landmarks and their relationships, and identifying associated cardiovascular anomalies. It is also an adjunct in the evaluation of complications during follow-up [7]. In our study we highlight the importance of noninvasive imaging, stressing the role of CTA over echocardiography in the diagnosis of congenital aortic arch anomalies.

1.1 Embryology

The aortic arch and its branches develop from the branchial apparatus, which begins during the second week of gestation and is completed by the seventh week. The apparatus consists of 6 branchial arches in the wall of the foregut; they are numbered from cephalad to caudad [2]. Truncus arteriosus forms the ascending aorta; the ventral aorta forms most of the arch, the common carotid and external carotid arteries. The 3rd arch forms the internal carotids. The left 4th arch forms the distal arch; dorsal aortic root segments form the isthmus. Finally the descending aorta is formed by the left dorsal aorta (Fig. 1) [8].

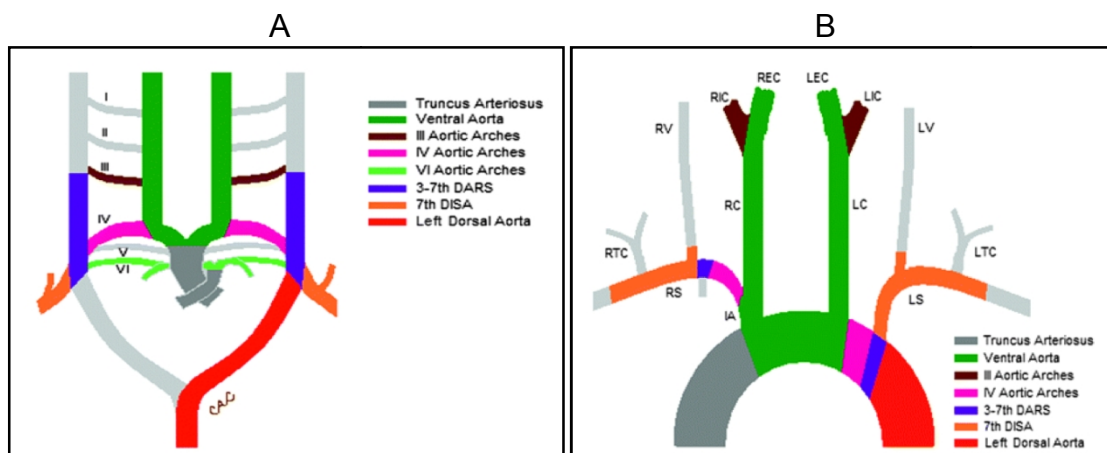


Fig. 1A and B: Schematic drawing that shows the primitive ventral, dorsal aortae, embryonic arches, DARS (dorsal aortic root segments), DISA (distal intersegmental arteries), RC (right common carotid), LC (left common carotid), REC (right external carotid), LEC (left external carotid), RV (right vertebral), LV (left vertebral), IA (innominate artery), RS (right subclavian), LS (left subclavian), RTC (right transverse cervical), LTC (left transverse cervical) (8)

Congenital aortic arch anomalies result from errors in involution or migration of primitive arches, usually aberrations occur at the level of interruption of the primitive arches [2]. The coarctation, therefore, is a manifestation of abnormal interruption of this arch system at different locations, if a longer segment is involved, hypoplastic arch will be the result [9]. The persistence of right fourth arch with regression of the left arch is associated with a right aortic arch and when both fourth arches persist, a double aortic arch is present [7].

2. MATERIALS AND METHODS

Forty five infants and children (23 males and 22 females) were examined in this study, they were referred to the pediatric cardiology outpatient clinic in Cairo University children's hospital over the period of 15 months from February 2011 to April 2012, all patients with history or clinical examination suspicious of aortic arch anomalies were included in the study, for each patient full history and clinical examination were obtained including: age, sex, residence, consanguinity, cardiac manifestations including: cyanosis, heart failure (poor feeding, sweating, failure to thrive, tachypnea, tachycardia, enlarged tender liver), manifestation of respiratory compression, low cardiac output symptoms, weak or absent femoral pulsations, abnormal gradient in blood pressure between upper and lower limbs (>20 mmHg) and unequal pulsation on both right and left side e.g. in aberrant right subclavian artery. Echocardiography was done and if aortic arch anomalies were diagnosed; the patient referred to do CT angiography of the heart after taking signed consent from the patient's parents.

2.1 Echocardiography Examination

An echocardiography examination was done for each patient by machine (vivid 7, GE, Vingmed, ultrasound AS, Hortin, Norway, probe 5S) after sedation by oral chloral hydrate

(1mg/kg) for patients less than 4 years, the patients were placed in a reclining position in comfortable environment in a dark room, with appropriate pillows and blankets, the sonographic gel was warmed to body temperature before the start.

2.2 CTA Imaging Technique

2.2.1 Patients' preparation

Fasting for at least 2-3 hours was required prior to the examination; proper hydration was advised for at least 4 hours before contrast injection.

2.2.2 Patients' sedation

Pediatric aortic MDCTA typically requires mild or no sedation. General anesthesia is infrequently needed for MDCTA because quiet breathing rarely causes appreciable artifacts. Oral and rectal chloral hydrate, were used if necessary [10,11]. Sometimes, IV diazepam was used with a dose of 1ml/kg only when needed.

2.2.3 CTA protocol

Only low-osmolar or iso-osmolar intravascular iodinated contrast agents (300 mg I/mL or greater) should be used for CTA of children. The volume of contrast material injected is usually weight based, ranging from 1.5 to 3.0 mL/kg [12], with a typical maximum volume of 90–125 mL, [12,13,14]. Power injector administration of IV contrast material is preferred to hand injection because homogeneous intravascular enhancement can be achieved [12,16]. Injection rates for pediatric thoracic CTA range from 0.4 to 1.0 mL/s through smaller-gauge angiocatheters for younger children, to 3.0–4.0 mL/s through larger-gauge for older children [15].

The patients included in this study were examined by (GE, Phillips, Malaysia) bright speed 8 detectors CT machine, the following parameters were use: helical scan type, 0.8 sec. rotational time, slice thickness 1.25 mm, pitch 1.675, speed 16.75, interval 0.625 cm, gantry tilt 0, small FOV, 100KV, 29 mA, total exposure time 6, 21 sec. the scan started at the level of thoracic inlet to include the proximal parts of the carotid and subclavian arteries and ended at the level of renal arteries.

With advances in CT scanners and software, a variety of high quality 2D reformatted and 3D reconstructed images are generated that aid in the understanding of complex cardiovascular anatomy. A combination of 2D multiplanar and 3D maximum-intensity-projection and volume-rendered images is most commonly used in the evaluation of congenital heart diseases (CHD) [4,16,17]. Two-dimensional multiplanar reformations can be generated with resolution comparable with that of axial images. Coronal and sagittal images provide information about cardiovascular structures, particularly structures that traverse the z-axis and may not be apparent on axial images [17,18]. Three-dimensional maximum-intensity-projection and volume-rendered images are used to display a portion or all of a data set.

Approval from the research ethics committee, Faculty of Medicine, Cairo University, was taken, according to university and research ethics committee guidelines.

3. RESULTS

Our study included 45 patients, their age ranges from 5 days to 11 years, they were 23 males and 22 females, with the following diagnosis: 15 had hypoplastic aortic arch by echocardiography, 6 of them proved to have multiple levels of narrowing by CT angiography, 11 patients had aortic coarctation by echocardiography, 3 of them were proved to have more than one segmental narrowing by CT angiography and one patient had hypoplasia of the whole aorta. Six patients had tetralogy of Fallot with right sided aortic arch, mirror imaging branches or aberrant left subclavian artery and hypoplastic pulmonary trunk. Four patients had interrupted aortic arch. One patient had aneurismal dilatation of ascending aorta and aortic arch secondary to congenital valvular aortic stenosis. One patient had aortopulmonary window. Six patients had persistent truncus arteriosus (4 patients type IV, 2 patient type II). The echocardiographic and CTA findings of our patients are illustrated in Table 1.

Table 1. New findings detected by CT over echocardiography in our patients

Echocardiographic findings	No	CT angiography findings	No
Hypoplastic arch	15	Multiple levels	4 (26.7%)
Coarctation of the aorta	11	Multiple levels	3 (27.3%)
		Diffuse hypoplasia	1 (9.1%)
Tetralogy of Fallot with right sided arch, mirror imaging branches, pulmonary hypoplasia, VSD	6	Tiny PDA	4 (66.7%)
Interrupted aortic arch	4	Accurate diagnosis instead of severe coarctation with hypoplastic arch	2 (50%)
valvular aortic stenosis with aneurysmal dilatation of the ascending aorta, aortic arch with dilated tortuous innominate artery	1	Assess extent, severity, involvement of main branches	
Aortopulmonary widow	1	Exact delineation of the site, length of the aortopulmonary defect as well as the associated anomalies	
Persistent truncus arteriosus	6	Detection of major aortopulmonary connections	2 (30%)

4. DISCUSSION OF CTA FINDINGS

4.1 Aortic Hypoplasia

Gradual tapering of the distal aortic arch and isthmus is a normal finding in the first 3 months of life. Persistence of this stricture later on is pathologic [19]. The proximal arch segment is defined as hypoplastic when its external diameter is less than 60% of that of the ascending aorta. The corresponding limit for hypoplasia in the distal arch is 50% and for the isthmus the limit is 40% [20,21]. The term tubular hypoplasia refers to a combination of abnormal small diameter and increased length (>5 mm in infants) between the segments of the aortic arch.

In the current study, 15 patients had aortic arch hypoplasia, 6 of them had associated multiple concentric narrowed segments that weren't detected by echocardiography in 4 of them (Fig. 2).

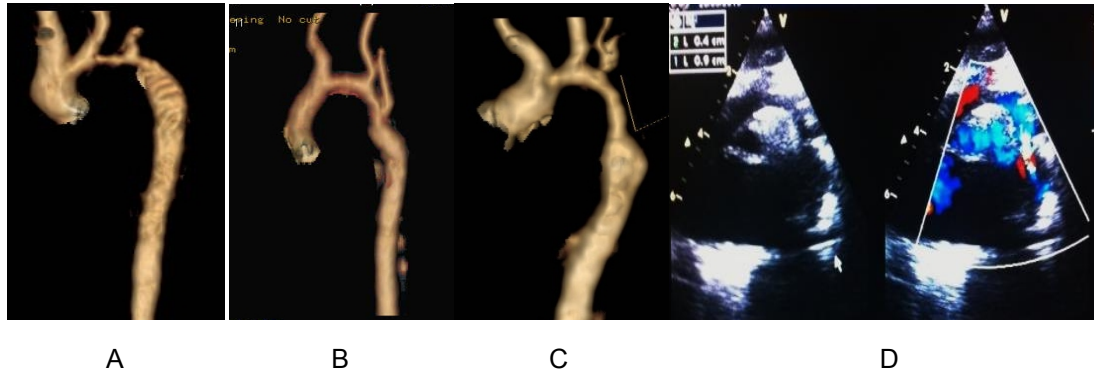


Fig. 2. 3D reconstruction image showing (A) hypoplastic aortic arch with multiple concentric narrowing segments. (B) Diffuse arch hypoplasia involving the origin of the left SCA as well as the proximal dorsal aorta in (C) for 15mm length, (D) the echocardiography picture of the same patient in image C, cannot delineate the lower limit of the hypoplastic segment

Echocardiography fails to establish the lower limit of hypoplastic segment especially if long. CTA was greatly helpful in detecting the location, extent and length of the hypoplastic segment, delineating the associated narrowed segments as well as the presence of PDA and collaterals [18] (Fig. 3).

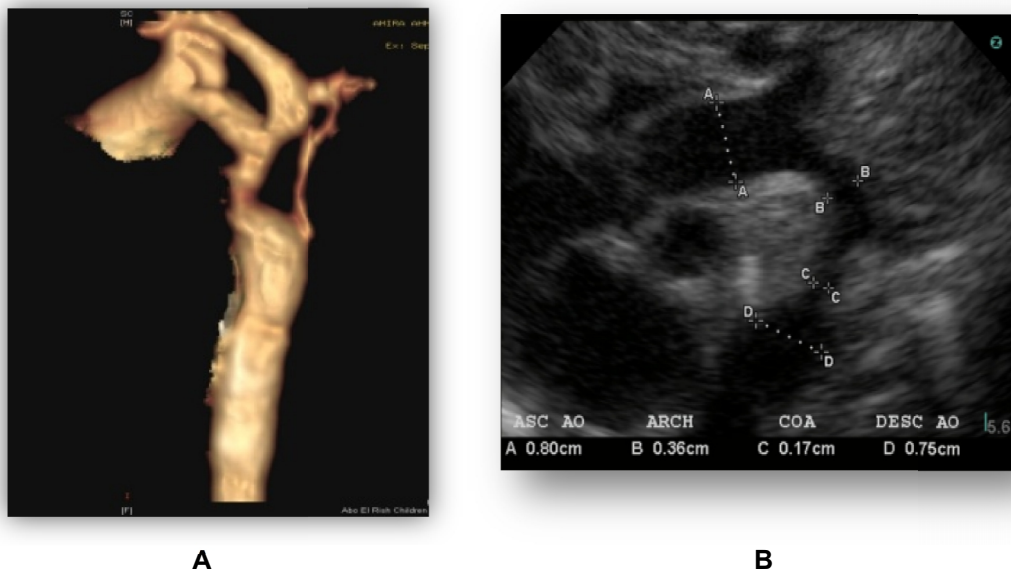


Fig. 3. A- 3D reconstructed image of hypoplastic aortic arch with few segments of narrowing showing collaterals between left SCA and upper dorsal aorta. B- Echocardiography picture of the same patient showing the hypoplastic arch with the distal narrowed segment

Whole aortic hypoplasia was seen in one female patient whose age was 6 months with associated hypoplastic left sided heart, PDA and relatively small sized branches (Fig. 4). the ascending aorta measures 8.3mm, the diameter reaches 6mm at the isthmus, then 5.7mm at the dorsal aorta, the abdominal aorta measures 6mm, the patient also had associated DTGA and bronchogenic cyst.

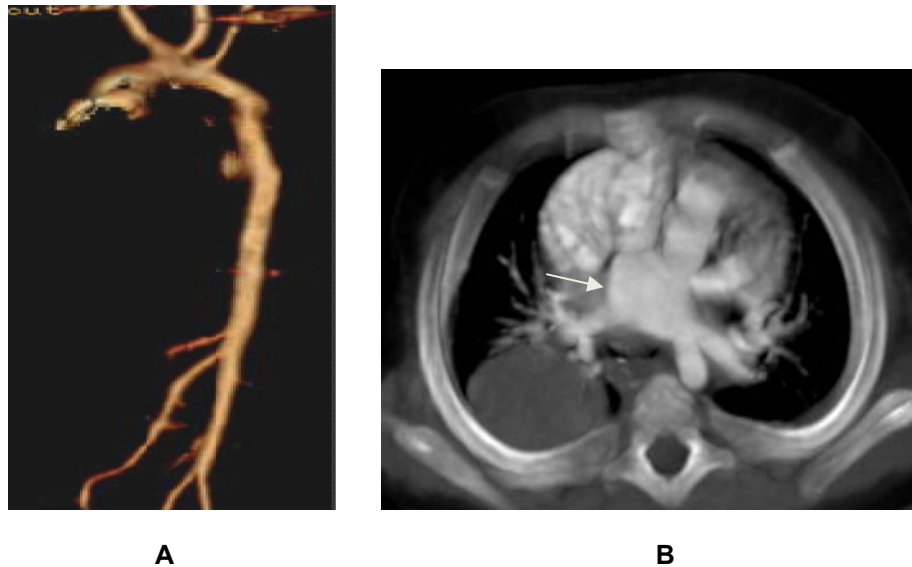


Fig. 4A. 3D reconstruction image of the globally hypoplastic aorta with relatively smaller branches. B. Axial image showing dilated pulmonary trunk, PDA (arrow) and right posterior bronchogenic cyst

4.2 Aortic Coarctation

Aortic coarctation involves aortic narrowing in the region of the ligamentum arteriosum just distal to the left subclavian artery; aortic coarctation represents 7% of congenital heart disease [22]. The stenotic segment frequently develops in a juxtaductal location but may show extension into the aortic arch and isthmus, the previous classification into preductal (infantile) and postductal (adult) is less currently used because aortic coarctation is always periductal [23]. The exact mechanism by which aortic coarctation is produced is unknown, but a hemodynamic hypothesis (i.e. abnormal preductal flow) and a ductal hypothesis (i.e. ectopic ductal tissue that extends into the aorta) have been proposed [24]. In the current study beside the tight stenosis found in 11 patients, 4 of them had more than one level of narrowing, mild narrowing frequently involving the distal arch or the isthmus was also observed (Fig. 5), echocardiography could not establish the diagnoses in these cases, it only revealed mild pressure gradient across this segment which needed to be confirmed by CT angiography. By viewing the reformatted multiplanar or 3D volume-rendered images in oblique and sagittal planes, the narrowed segment can be clearly visualized. CTA allows the assessment of the number, length, site and the extent of the narrowed segments as well as the associated collaterals and PDA (Fig. 6).



Fig. 5. 3D reconstructed image of mild narrowing at the level of aortic arch isthmus

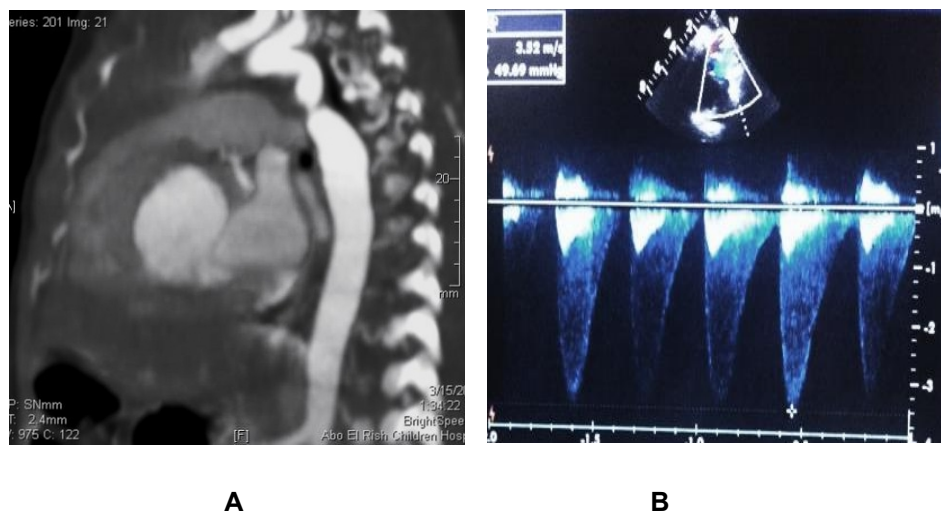


Fig. 6. MIP images of two cases of aortic coarctation: (A) Single leveled concentric aortic narrowing with dilated intercostals and anterior chest wall collaterals, (B) echocardiography picture of the same patient showing high pressure gradient across the narrowed segment

Multidetector CT (MDCT) imaging can also demonstrate associated findings such as bicuspid aortic valve, left ventricular hypertrophy and VSD. Nevertheless, in the current study CT is limited in providing functional information because no gradient can be calculated, unlike echocardiography and magnetic resonance imaging (7).

4.3 Patent Ductus Arteriosus

In the normal heart with a left-sided aortic arch, the ductus arteriosus connects the left pulmonary artery near its origin to the descending aorta just distal to the left subclavian

artery [25]. In full-term newborns, the ductus arteriosus typically closes functionally 18–24 hours after birth and anatomically at 1 month. If it remains permeable 3 months after birth, it is considered a PDA [7]. The ductus arteriosus may persist in a wide variety of sizes and configurations. Usually, the aortic end of the patent ductus is larger than the pulmonary artery end, which results in a somewhat conical configuration [25]. A morphologic classification described by Krichenko et al. [26] has been used: Type A (conical) is the most common type and is characterized by an ampulla at the aortic end. Type B (window) is a ductus that narrows at the aortic end. Type C (tubular) is a ductus with no narrowing or constrictions. Type D (complex) is a ductus with multiple constrictions. Type E (elongated) is a ductus with a bizarre configuration and with constriction away from the pulmonary artery end. This classification is useful in selecting the type of closure and the correct device. A detailed description of the ductus (i.e. length, diameter at the aortic and pulmonary insertion sites, diameter and location of the narrowest portion of the ductus, morphologic type, presence of calcification or aneurysm, and angle of the ductus to the descending aorta) is important before attempting percutaneous closure of the PDA and during selection of closure hardware [7]. PDA could be seen with other congenital cardiac and extra cardiac anomalies. In the current study tiny PDAs have been seen associating tetralogy of Fallot in 4 patients and were not diagnosed by echocardiography (Fig. 7). These patients had right sided aortic arch (2 had mirror imaging branches, 2 had aberrant left subclavian artery), hypoplastic pulmonary trunk and PDA. PDA has been also noted in other patients with aortic hypoplasia, coarctation or as an isolated anomaly. The manually generated sagittal and oblique reformatted images as well as the 3D reconstructed images greatly helped in demonstrating the site, width and course of the PDAs especially those having oblique tortuous courses.

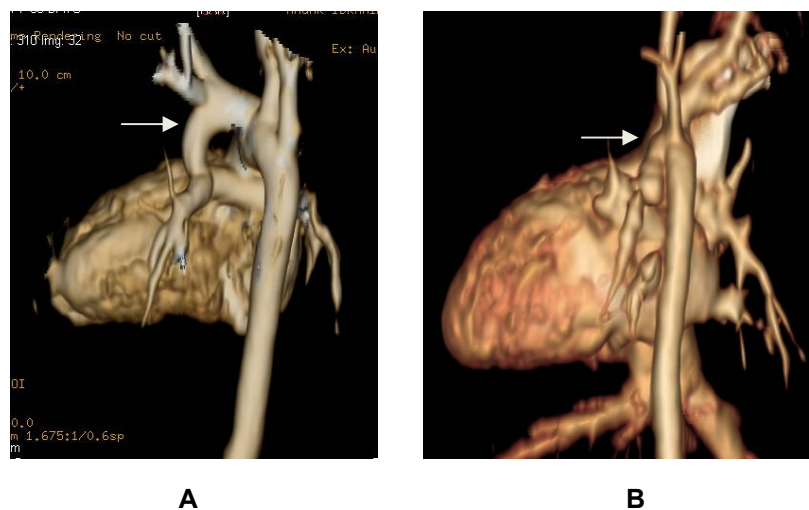


Fig. 7A. 3D reconstructed image of a patient with atretic pulmonary trunk, narrowed both pulmonary branches, right sided aortic arch, aberrant left SCA and a large curved PDA (arrow) that joins the origin of LPA with narrower end and gives arise to left CCA and left SCA. B. Another 3D reconstructed image of a patient with large tortuous PDA that joins the left main pulmonary artery (arrow)

4.4 Interrupted Aortic Arch

Interrupted aortic arch (IAA) is defined as complete luminal and anatomic discontinuity between the ascending and descending aorta, as described by Steidele [27] (Fig. 8). Altered hemodynamics through the fourth aortic arch and teratogenic exposure during the intrauterine period has been proposed as its potential causes. It may occur as a simple or complex anomaly [7]. During the preoperative evaluation, echocardiography did not allow differentiation of IAA from severe aortic coarctation with a hypoplastic arch. In these cases, the assessment is complemented by CT or MR imaging [29, 30]. These techniques easily demonstrate the morphologic features of IAA and the potential complex associated findings because of their multiplanar capabilities, which facilitate understanding of the anomaly and its anatomic relationships.

In the current study, we diagnosed 4 patients with interrupted arch; they were mostly of type B in which the descending aorta is in direct continuity with the pulmonary trunk. A narrow vascular connection was noted between the ascending and descending aorta that gives arise to left subclavian artery which appear hypoplastic as well in two cases, in the other two cases the left subclavian artery arises from the junction between the descending aorta and the pulmonary trunk (Fig. 9). This vascular connecting segment is similar to that described by Dillman et al. [30] and was preliminary diagnosed as severely hypoplastic aortic arch by echocardiography, and needed MDCT angiography to confirm the presence of aortic arch interruption.

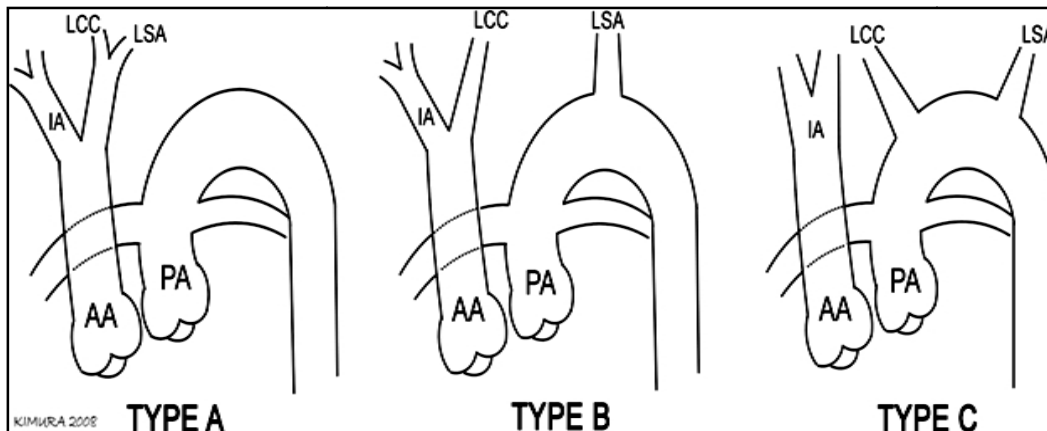


Fig. 8. Celoria and Patton classification of IAA [28]. Type A is defined as an interruption distal to the left subclavian artery (LSA). In type B, the absent segment is between the left common carotid artery (LCC) and left subclavian artery. Type C is defined as an interruption distal to the innominate artery (IA). Note that the descending thoracic aorta reconstitutes from the pulmonary artery (PA) through a ductus arteriosus. AA = ascending aorta

4.5 Aneurysmal Dilatation of the Ascending Aorta Secondary to Valvular Aortic Stenosis

Congenital aortic valve abnormalities are common in the population, a bicuspid aortic valve occurs in 1% of the population with aortic valve stenosis presenting clinically in 3% to 8% of

all CHD patients [31]. With significant obstruction, LV after load increases, the myocardium hypertrophies, and chamber compliance is reduced, which results in higher left atrial filling pressures. Patients with bicuspid valves have been found to have abnormalities of the connective tissues of the aorta and are prone to marked dilation of the ascending aorta even in the absence of significant obstruction or insufficiency. These patients should be followed up noninvasively to rule out rapid aortic growth, because root replacement may be required to prevent dissection [32]. Currently, no clear consensus exists for this population as to the size at which prophylactic aortic root replacement should be performed. In the current study, one male patient 11 years old with valvular aortic stenosis was diagnosed showing aneurysmal dilatation of the ascending aorta, aortic arch with dilated tortuous innominate artery as well (Fig. 10), the diameter of the ascending aorta reaches 25mm and exceeds the acoustic window of the echocardiography. CTA was important to accurately visualize the extent and severity of the dilatation as well as involvement of the main branches.

4.6 Aortopulmonary Window

Aortopulmonary window, described by Elliotson in 1830, is defined as a communication between the ascending aorta and the pulmonary trunk or right pulmonary artery, according to the site of communication aortopulmonary window is classified into three types (Fig. 11) [33].

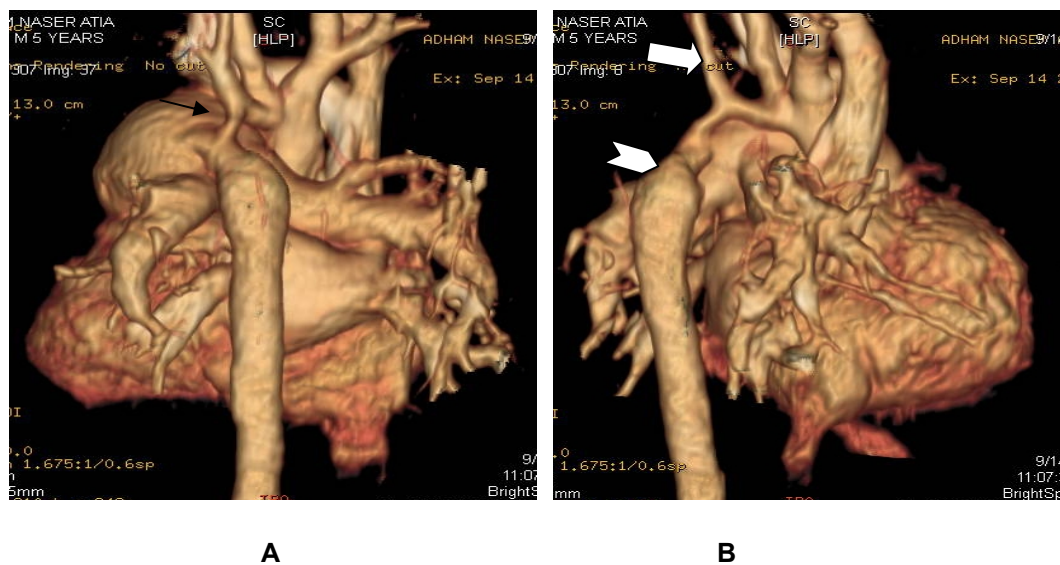


Fig. 9A and B: 3D reconstructed images showing the ascending aorta continues as the right innominate and the left CCA, the descending aorta is in direct continuity with the pulmonary trunk through large PDA (small arrow), Hypoplastic connecting segment (arrow head) is noted between the ascending and descending aorta that gives origin to the left SCA that appears also hypoplastic (arrow)

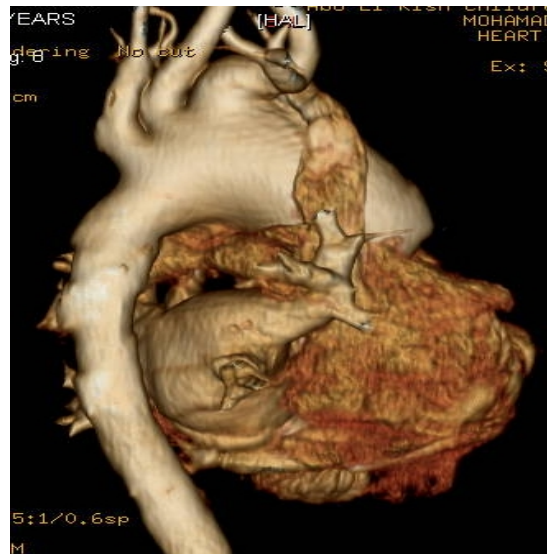


Fig. 10. A reconstructed image of a patient with valvular aortic stenosis with aneurismal dilatation of the ascending aorta, aortic arch as well as the right innominate artery, marked concentric muscular hypertrophy of the left ventricle is also noted

In the current study, we diagnosed one patient with aortopulmonary window proximal type (type 1) (Fig. 12), that was also seen satisfactory by echocardiography. Other anomalies were also noted as right sided aortic arch with mirror imaging branches and atretic left pulmonary artery. CTA is important in exact delineation of the site, length of the aortopulmonary defect as well as the associated anomalies.

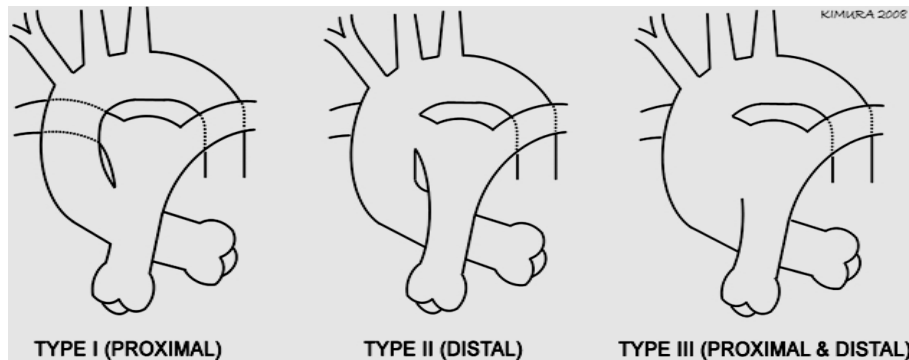


Fig. 11. Mori classification of aortopulmonary window [31]. Type I or proximal communication occurs near the semilunar valves. Type II or distal communication involves the pulmonary bifurcation at the level of the right pulmonary artery. In type III, there is a wide communication between the aorta and pulmonary artery owing to total absence of the aortopulmonary septum

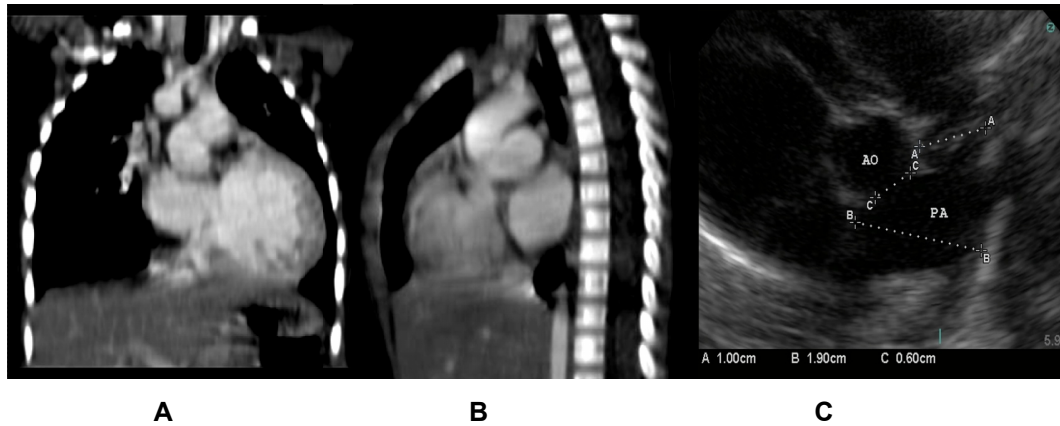


Fig. 12 A and B: coronal and sagittal reconstructed images for a patient with aortic pulmonary window proximal type (type 1), C. Echocardiography picture for the same patient that shows the aortic pulmonary connection which is being measured by the cardiologist

4.7 Persistent Truncus Arteriosus

In this condition, a common arterial trunk arises from common conotruncal valve and gives origin to systemic, pulmonary and coronary circulation and associated with VSD [6]. This anomaly is due to septation defects involving the aortic pulmonary, truncal and infundibular septa, four types have been described according to Collett and Edwards [34] (Fig. 13).

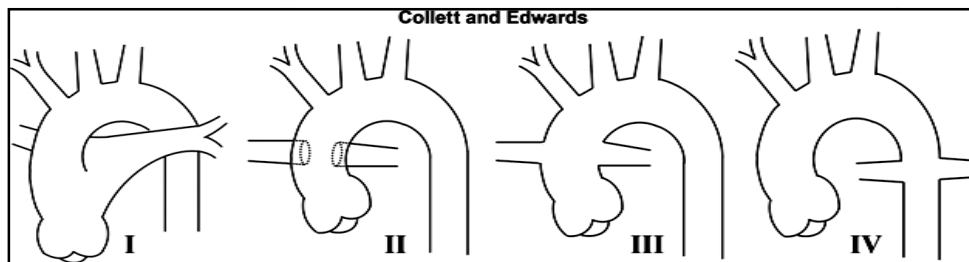


Fig. 13. Classification of truncus arteriosus according to Collett and Edwards [33].

The Collett and Edwards classification is the most commonly used. In type I, the pulmonary artery trunk arises from the proximal portion of the truncus arteriosus. In types II and III, there is no pulmonary artery trunk and the pulmonary branches arise from the posterior and lateral midsegments of the truncus, respectively. Finally, in type IV, the pulmonary circulation is dependent on major aortic pulmonary collateral arteries

The presence of the truncal valve allows its differentiation from aortic pulmonary window in which two separate valves are present [33,35]. Authors prefer to classify Type IV embryologically with those having pulmonary atresia and major aortic pulmonary collaterals [36]. Surgical operation must be done early to avoid the risk of development of pulmonary obstructive disease. Echocardiography can efficiently demonstrate types I, II and III, but

accurate display by MDCT is mandatory, however major aortopulmonary collaterals in type IV are poorly seen by echo and need to be visualized by MDCT (Fig. 14).

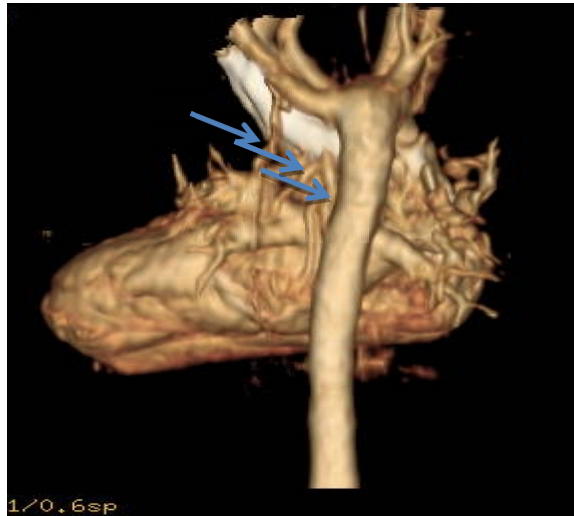


Fig. 14. Type IV truncus arteriosus, 3D reconstructed image showing atretic pulmonary trunk and both hilar pulmonary arteries are reconstructed by major aortopulmonary collaterals that arise from dorsal aorta and subclavian arteries (arrows), with associated right sided aortic arch and mirror imaging branches

5. CONCLUSION

Echocardiography is commonly utilized for preliminary evaluation of aortic arch anomalies; however MDCTA provides exquisite anatomical details for good evaluation. In the current study, echocardiography was limited in detection of multiple narrowing segments, abnormal vascular connection and collaterals in cases of aortic coarctation or hypoplasia especially if located in the distal arch, also echocardiography might not allow differentiation of IAA from severe aortic coarctation with a hypoplastic arch, which highlights the role of CTA in demonstration of these anomalies. The multiplanar reformatted and 3D reconstructed images were essential to visualize the tortuous PDAs and detection of their exact type, course and diameters. Markedly dilated arteries that exceeded the limits of echocardiography acoustic windows were accurately assessed by CTA. Major Aortopulmonary collaterals can hardly be seen by echo. The major disadvantages of CTA were the hazards of ionizing radiation as well as contrast injection. Recently new techniques are used to lower the radiation doses to minimum. The alternative non invasive studies also have inherent limitations. Echocardiography may be limited in its ability to define extra cardiac vessels, but still is the most suitable for screening and establishing the primary diagnosis. MRI, although not studied in this paper, may be as good as CT, reducing radiation that can be significant particularly in children. Furthermore, MRI may be repeated without the radiation accumulative risk. But MRI is time-consuming, may require prolonged patient sedation, and may be difficult to perform in seriously ill patients.

CONSENT

All authors declare that 'written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

All authors hereby declare that all experiments have been examined and approved by the appropriate ethics committee and have therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Santulli G. Epidemiology of cardiovascular disease in the 21st century: Updated numbers and updated facts. *JCVD*. 2013;1(1):1-2.
2. Stojanovska J, Cascade P, Chong S, Quint L, Sundaram B. Embryology and imaging review of aortic arch anomalies. *Journal of Thoracic Imaging*. 2012;27:73-84.
3. 2-Tsai IC, Chen MC, Jan SL, Wang CC, Fu YC, Lin PC, et al. Neonatal cardiac multidetector row CT: Why and how we do it? *Pediatr Radiol*. 2008;38:438-451.
4. 3- Leschka S, Oechslin E, Husmann L, Desbiolles L, Marincek B, Genoni M, et al. Pre- and postoperative evaluation of congenital heart disease in children and adults with 64-section CT. *Radio Graphics*. 2007;27:829-846.
5. 4- Obregon C, Fogel M. Imaging of aortic arch anomalies and associated findings. *Expert Rev Cardiovasc Ther*. 2012;10(12):1497-1516.
6. 5- Dillman R, Hernandez R. Role of CT in the evaluation of congenital cardiovascular disease in children. *AJR*. 2009;192:1219-1231.
7. Kimura-Hayama E, Meléndez GL, Mendizábal A, Meave-González AB, Zambrana GF, Corona-Villalobos C. Uncommon Congenital and acquired aortic diseases: Role of multidetector CT angiography. *Radio Graphics*. 2010;30:79-98.
8. Satti SR, Cerniglia CA, Koenigsberg RA. Cervical vertebral artery variations: An anatomic study. *AJNR Am J Neuroradiol*. 2007;28:976-80.
9. Edwards JE. Anomalies of the derivatives of the aortic arch system. *Med Clin North Am*. 1948;32:925-949.
10. Lee T, Tsai IC, Fu YC, Jan SL, Wang CC, Chang Y, et al. Using multidetector-row CT in neonates with complex congenital heart disease to replace diagnostic cardiac catheterization for anatomical investigation: initial experiences in technical and clinical feasibility. *Pediatr Radiol*. 2006;36:1273-1282.
11. Choo KS, Lee HD, Ban JE, Sung SC, Chang YH, Kim CW, et al. Evaluation of obstructive airway lesions in complex congenital heart disease using composite volume-rendered images from multislice CT. *Pediatr Radiol*. 2006;36:219-223.
12. Siegel MJ. Multiplanar and three-dimensional multi-detector row CT of thoracic vessels and airways in the pediatric population. *Radiology*. 2003;229:641-650.
13. Lee EY, Siegel MJ, Hildebolt CF, Gutierrez FR, Bhalla S, Fallah JH. MDCT evaluation of thoracic aortic anomalies in pediatric patients and young adults: Comparison of axial, multiplanar and 3D images. *AJR*. 2004;182:777-78.

14. Kim TH, Kim YM, Suh CH, Park IS, Kim WH, Lee YT. Helical CT angiography and three-dimensional reconstruction of total anomalous pulmonary venous connections in neonates and infants. *AJR*. 2000;175:1381-1386.
15. Ley S, Zaporozhan J, Arnold R, et al. Preoperative assessment and follow-up of congenital abnormalities of the pulmonary arteries using CT and MRI. *Eur Radiol*. 2007;17:151-162.
16. Frush DP, Herlong JR. Pediatric thoracic CT angiography. *Pediatr Radiol*. 2005;35:11-25.
17. Frush DP, Yoshizumi T. Conventional and CT angiography in children: Dosimetry and dose comparisons. *Pediatr Radiol*. 2006;36(14):154-158.
18. Bean MJ, Pannu H, Fishman EK. Three-dimensional computed tomographic imaging of complex congenital cardiovascular abnormalities. *J Comput Assist Tomogr*. 2005;29:721-724.
19. Ho SY, Anderson RH. Coarctation, tubular hypoplasia and the ductus arteriosus: Histological study of 35 cases. *Br Heart J*. 1979;41:268-274.
20. Moolaert AJ, Bruins CC, Oppenheimer-Dekker A. Anomalies of the aortic arch and ventricular septal defects. *Circulation*. 1976;53:1011-1015.
21. Matsui H, Adachi I, Uemura H, Gardiner H, Ho YS. Anatomy of coarctation, hypoplastic and interrupted aortic arch: Relevance to interventional/surgical treatment. *Expert Rev Cardiovasc Ther*. 2007;5:871-880.
22. Aboulhosn J, Child JS. Left ventricular outflow obstruction: Subaortic stenosis, bicuspid aortic valve, supravalvular aortic stenosis and coarctation of the aorta. *Circulation*. 2006;114:2412-2422.
23. Abbruzzese PA, Aidala E. Aortic coarctation: An overview. *J Cardiovasc Med (Hagerstown)*. 2007;8:123-128.
24. Rao PS. Coarctation of the aorta. *Curr Cardiol Rep*. 2005;7:425-434.
25. Schneider JD, Moore WJ. Patent ductus arteriosus. *Circulation*. 2006;114:1873-1882.
26. Krichenko A, Benson L, Burrows P, Mões C, McLaughlin P, Freedom R. Angiographic classification of the isolated, persistently patent ductus arteriosus and implications for percutaneous catheter occlusion. *Am J Cardiol*. 1989;63:877-880.
27. Steidele RJ. Sammlung verschiedener in der chirurg. Prakt Lehschule Gemachten Beobbb (Viena). 1778;2:114.
28. Celoria GC, Patton RB. Congenital absence of the aortic arch. *Am Heart J*. 1959;58:407-413.
29. Yang DH, Goo HW, Seo DM, et al. Multislice CT angiography of interrupted aortic arch. *Pediatr Radiol*. 2008;38:89-100.
30. Dillman JR, Yarram SG, D'Amico AR, Hernandez RJ. Interrupted aortic arch: Spectrum of MRI findings. *AJR*. 2008;190:1467-1474.
31. Hoffman JI, Christianson R. Congenital heart disease in a cohort of 19,502 births with long-term follow-up. *Am J Cardiol*. 1978;42:641-647.
32. Bauer M, Pasic M, Meyer R, Goetze N, Bauer U, Siniawski H, et al. Morphometric analysis of aortic media in patients with bicuspid and tricuspid aortic valve. *Ann Thorac Surg*. 2002;74:58-62.
33. Soares AM, Atik E, Cortêz TM, et al. Aortopulmonary window: Clinical and surgical assessment of 18 cases. *Arq Bras Cardiol*. 1999;73:59-74.
34. Collett RW, Edwards JE. Persistent truncus arteriosus: A classification according to anatomic types. *Surg Clin North Am*. 1949;29:1245-1270.
35. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital heart surgery nomenclature and database project: Aortopulmonary window. *Ann Thorac Surg*. 2000;69(4):44-49.

36. Mair DD, Ritter DG, Davis GD, Wallace RB, Danielson GK, Mcgoon DC. Selection of patients with truncus arteriosus for surgical correction: Anatomic and hemodynamic considerations. *Circulation*. 1974;49:144-151

© 2014 Dien et al.; This is an Open Access article distributed under the terms of the Creative Commons Attribution License (<http://creativecommons.org/licenses/by/3.0>), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Peer-review history:

The peer review history for this paper can be accessed here:
<http://www.sciencedomain.org/review-history.php?iid=461&id=12&aid=4036>