IMAGES **in PAEDIATRIC CARDIOLOGY**

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Key Words
Double aortic arch, Dysphagia, Fallot

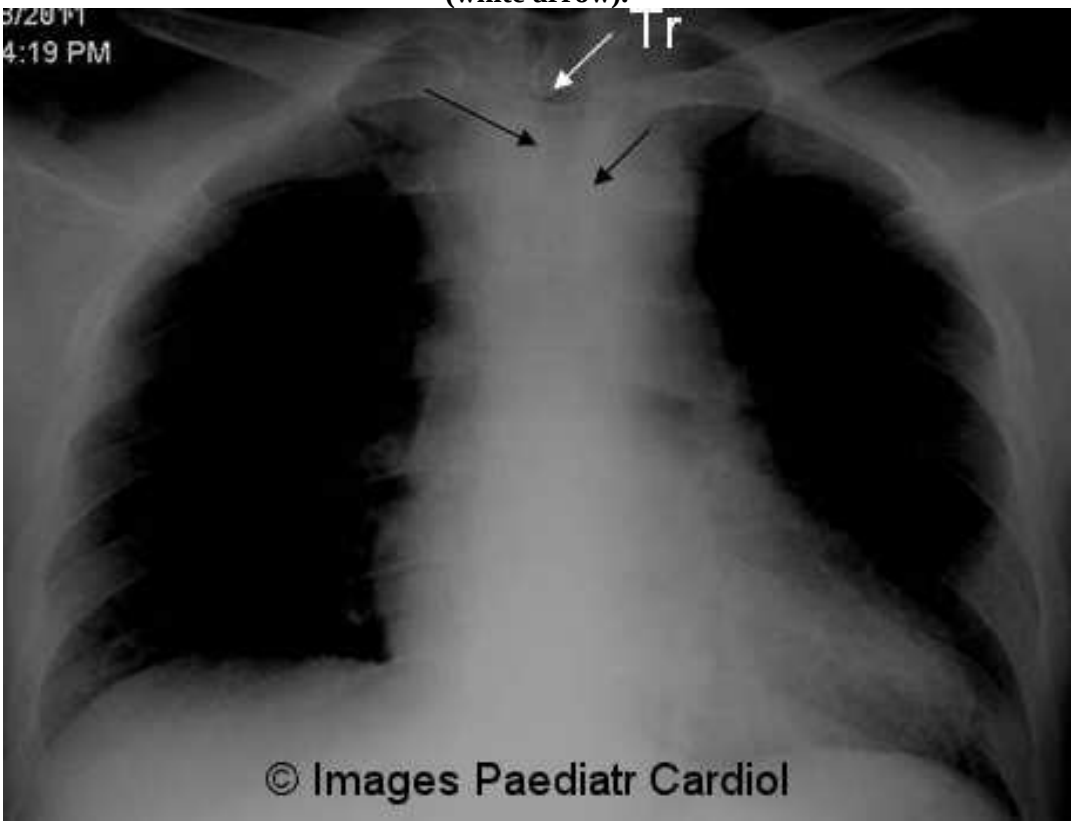
Abstract:
Double aortic arch (DAA) is a common vascular ring. It may occur in isolation or coexist with various types of congenital heart disease . The anomaly usually presents in early infancy. This reports a 23yr old male presenting with dysphagia, who was found to have a double aortic arch and tetralogy of Fallot .Both lesions were successfully corrected surgically.

Introduction:
Tetralogy of Fallot (TOF) is a common cyanotic cardiac lesion, and may be associated with aortic arch anomalies. These are more frequent with 22q11 deletions.[1,2] Of the arch abnormalities double aortic arch (DAA) is one vascular ring which completely encircles the trachea and oesophagus. The most common cyanotic congenital heart lesions associated with DAA are TOF and transposition of great arteries.[3] Most cases of DAA with or without congenital heart disease (CHD) present in infancy or early childhood. We report a patient with DAA and TOF who did not present until adulthood.

Case Report:

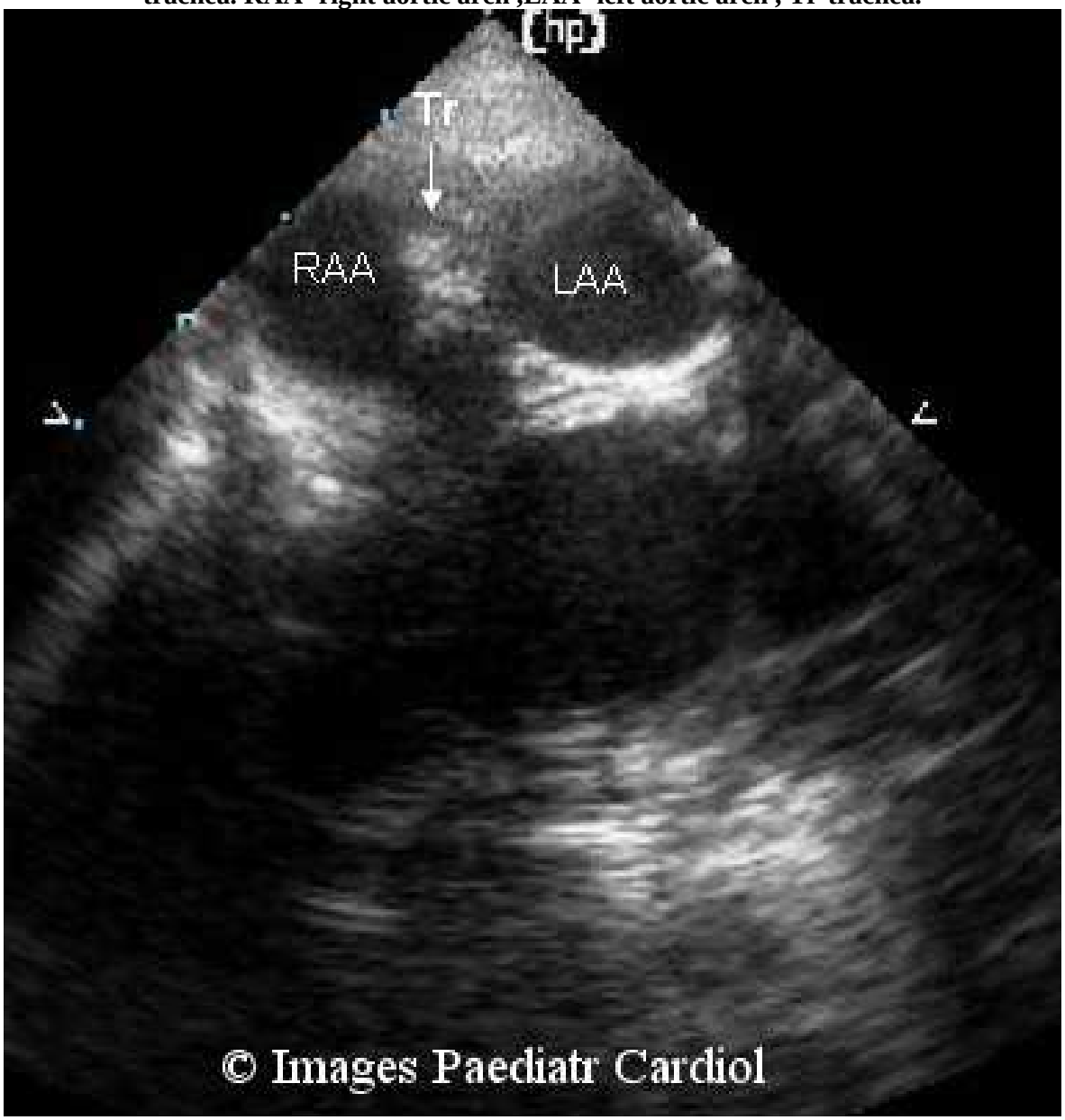
A 23yr old male presented with cyanosis and easy fatigability from early childhood. He was thinly built, deeply cyanosed [TcO2 saturation 70%] and clubbed. Further examination included a single second heart sound, and a grade 2/6 ejection systolic murmur at the upper left parasternal area. Twelve lead electrocardiography confirmed sinus rhythm, a QRS axis of +120, right ventricular dominance with early transition of the QRS complex in V2. The chest X-ray was usual for TOF, except that there was an indentation of the tracheal shadow on both sides, more prominent on the right (figure 1).

Figure 1: Chest X ray (PA view) showing no cardiomegaly, right ventricular type apex, decreased pulmonary blood flow. Black Arrows point to indentation on both side of trachea by DAA. Indentation is more prominent and higher on right side than left side. Tr-trachea (white arrow).



Echocardiography revealed the usual findings of TOF with confluent normal sized pulmonary arteries. Suprasternal short axis view demonstrated two “circles” one on each side of the trachea, suggesting a double aortic arch (figure 2).

Figure 2: Suprasternal short axis view showing two circles of both arches on both sides of trachea. RAA- right aortic arch ,LAA- left aortic arch , Tr-trachea.



To define the anatomy further, high-resolution CT, and cardiac catheterization were carried out which revealed a double aortic arch, with the right arch being smaller and more cranially situated (Figs 3-6).

Figure 3: Aortogram: Injection in ascending aorta in frontal view showing double aortic arch, dominant left arch, right arch is higher then left, descending aorta on left.

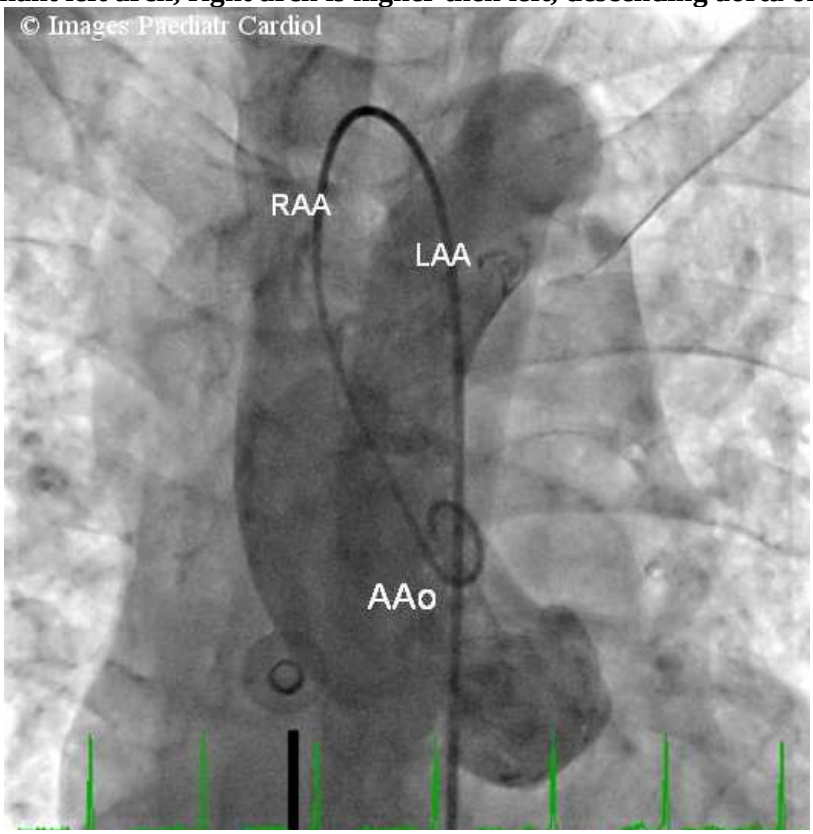


Figure 4: HRCT showing two circles of both aortic arches on both sides of trachea. Image is complimentary to figure 2. RAA- right aortic arch, LAA- left aortic arch, Tr-trachea, RPA- right pulmonary artery.

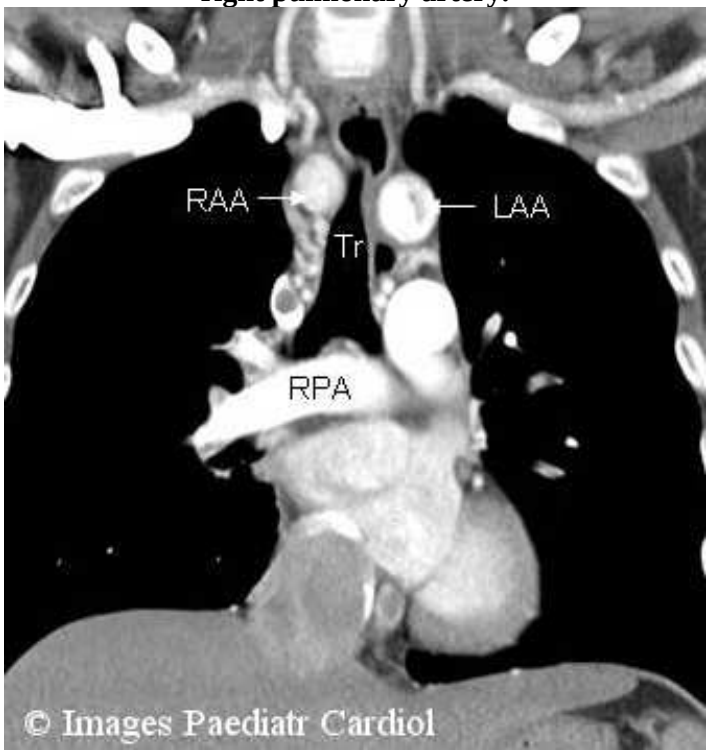


Figure 5: Three dimensional reconstruction of aortic arch showing DAA, left descending aorta, dominant left arch and superior position of right arch . RAA- right aortic arch, LAA- left aortic arch, AAo-ascending aorta, LV-left ventricle, RV-right ventricle.

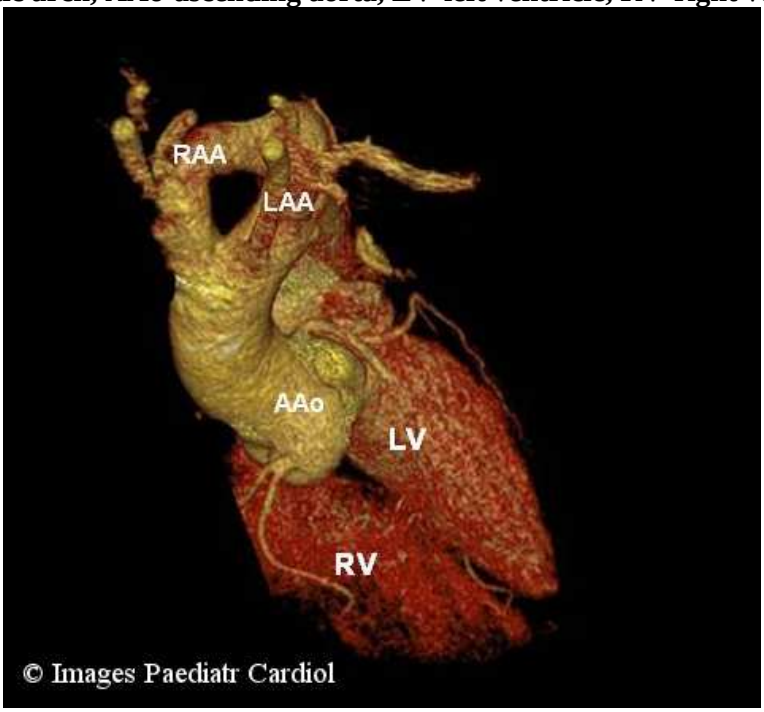
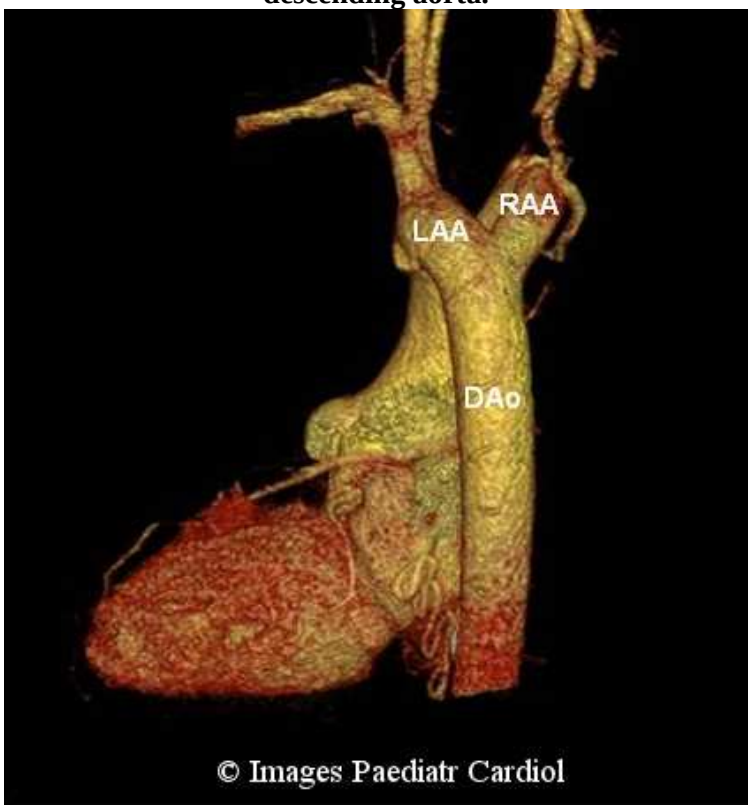


Figure 6: Three dimensional reconstruction of DAA from posterior view showing same findings as described in figure 4b . RAA- right aortic arch ,LAA- left aortic arch, DAo- descending aorta.



Two neck vessels arose from each arch, and there was no pull-back pressure gradient on either arch. The characteristic features of the TOF with adequately sized pulmonary arteries were shown. Chromosomal analysis excluded a 22q11 deletion. Having reviewed these investigations the patient admitted to direct questioning that he had suffered swallowing difficulties especially for solid foods for many years but attributed this to his cyanotic heart disease. He denied any respiratory symptoms at any time. The surgical plan was to perform single stage correction of the TOF through a midline sternotomy, together with relief of the vascular ring, by division of the minor arch. At operation, the ventricular septal defect (VSD) was closed, the infundibular muscle resected with trans annular enlargement of the right ventricular (RV) outflow, together with insertion of a pulmonary valve prosthesis (23mm Epic valve). As visualization of the aortic arches from the midline approach was difficult because of the size of the ascending aorta, relief of the DAA was deferred.

Following uneventful recovery from this initial procedure, on the 7th post operative day via a right thoracotomy, the minor (right) component of the double arch was ligated and divided distal to the origin of 2nd arch branch. This arch was non dominant, higher, and more posteriorly positioned, clearly compressing the oesophagus. Recovery continued uneventfully. At the six month postoperative review, he was doing well with complete disappearance of the dysphagia. Echocardiography confirmed no residual postoperative intracardiac issues and normal flow in the remaining left arch.

Discussion:

TOF is known to have arch anomalies especially due to its association with DiGeorge syndrome. Although very rare, several cases of TOF associated with DAA have been reported so far.[4-10]. However most of these patients belonged to the infant age group or they were detected during early childhood. Our patient was an adult with this rare combination. To the best of our knowledge probably only one adult patient has been reported previously who had TOF associated with DAA and underwent a successful surgical repair.[9] However surgical details of this case were not discussed and published.

Intracardiac anatomy in our patient was completely determined by echocardiography. DAA is very often missed on echocardiography[4,6,7,10] but we had a strong suspicion of DAA on suprasternal view. Suprasternal echocardiography can be used to diagnose a DAA.[11] Two circle appearance on suprasternal view strongly suggest the possibility of double aortic arch. In our patient, images of this view were not satisfactory due to poor quality resulting from limited acoustic windows which is not uncommon in many of adult patients.

CT and MRI are the imaging modalities of choice in a patient who is thought to have a vascular ring.[12-14] The advantage of the use of CT angiography is that the modality is a noninvasive technique that enables evaluation of vascular anomalies and the status of tracheal or esophageal compression in the same study. This information often eliminates the more traditional multistage work-up, which includes barium study, bronchoscopy, echocardiography, and angiography. In our case too, anatomy of the arch and its branches were confirmed and well profiled in relation to adjoining structures by CT scan. Three dimensional volume rendered images were of great help for the surgical team.

Catheter-directed cardiac angiography was done to rule out any additional VSD in our case of an adult with TOF. For an isolated DAA catheterization is usually not required. Compared with CT, cardiac catheterization has a higher complication rate owing to its invasive nature, typically requires a larger volume of intravascular contrast material and often imparts greater radiation dose.[14] Although echocardiography and angiography are the dominant imaging modalities in patients with

congenital heart disease, CT is an extremely valuable noninvasive adjunct which is effective in demonstrating the complex cardiovascular morphology, especially the extracardiac morphology.[12]

Compared with the CT scanners of the past, newer scanners yield images with better temporal and spatial resolution, greater anatomic coverage per rotation, and more consistent enhancement with a lesser volume of intravascular contrast material, and higher-quality two-dimensional reformation and three-dimensional reconstruction owing to the acquisition of an isotropic data set.[14] Rapid imaging with these CT scanners requires less patient sedation which makes it even superior to MRI. CT can be used to determine which arch is dominant and to establish the relations between the great vessels and each arch. This information is important because thoracotomy is typically performed on the side of the smaller arch.

Chest X-ray in our patient was not of very classical of tracheal indentation. Reterospectively we could correlate it with other better available images of CT and aortogram. Use of high kv roentgenograms or barium swallow could have shown compression of trachea and esophagus better but these additional investigations were not required in view of available CT reconstructed images.

DAA usually manifests early in the life with the symptoms of stridor, wheezing and choking episodes with feeds.[15,16] Those with loose rings can present with dysphagia during adulthood as is the case with our patient.[17] Our patient was so ignorant about this symptom that it could be elicited in the history retrospectively only.

Conclusion:

To plan effective management of congenital heart disease, a thorough clinical history is the basic need. All patients with TOF, child or adult, must be looked thoroughly for aortic arch anomalies meticulously by suprasternal echocardiography to avoid misses. A collaborative approach by cardiologists, surgeons, and radiologists in reviewing CT imaging data of DAA maximizes the diagnostic yield. A thorough preoperative understanding of complex cardiovascular anatomy in patients with congenital heart disease facilitates a directed and prepared surgical approach.

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