IMAGES **in PAEDIATRIC CARDIOLOGY**

Patel B,[1,5] Coyle JC,[1,2,5] Poe E,[1,2,5] Rosenbloom C,[3,4,5] Stevens RM,[3,4,5,6] Coren JS,[7] Ge S,[1,2,5,6], Mesia IC,[1,2,5,6] Moulick A,[3,4,5,6] Toib A.[1,2,5,6] Three Dimensional Rotational Angiography Imaging of Double Aortic Arch Vascular Ring. Images Paediatr Cardiol 2013;15(1):1-6.
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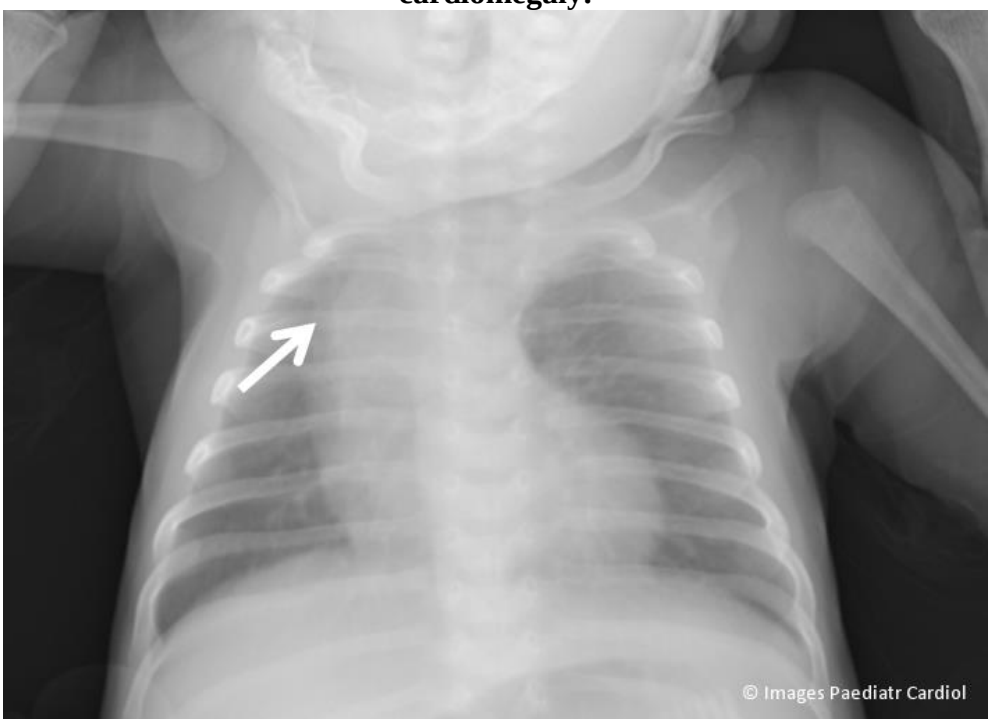
MeSH

Imaging, Three dimensional, Angiography, Aorta, Thoracic

Abstract
Background: Three dimensional (3D) rotational angiography is a technique used increasingly for imaging in congenital heart disease. Results: We describe the use of this technique for imaging of double aortic arch vascular ring and discuss the advantages of this modality. Conclusions: 3D rotational angiography is an excellent tool for imaging of various vascular anomalies. It provides high quality accurate images through a quick and safe procedure.

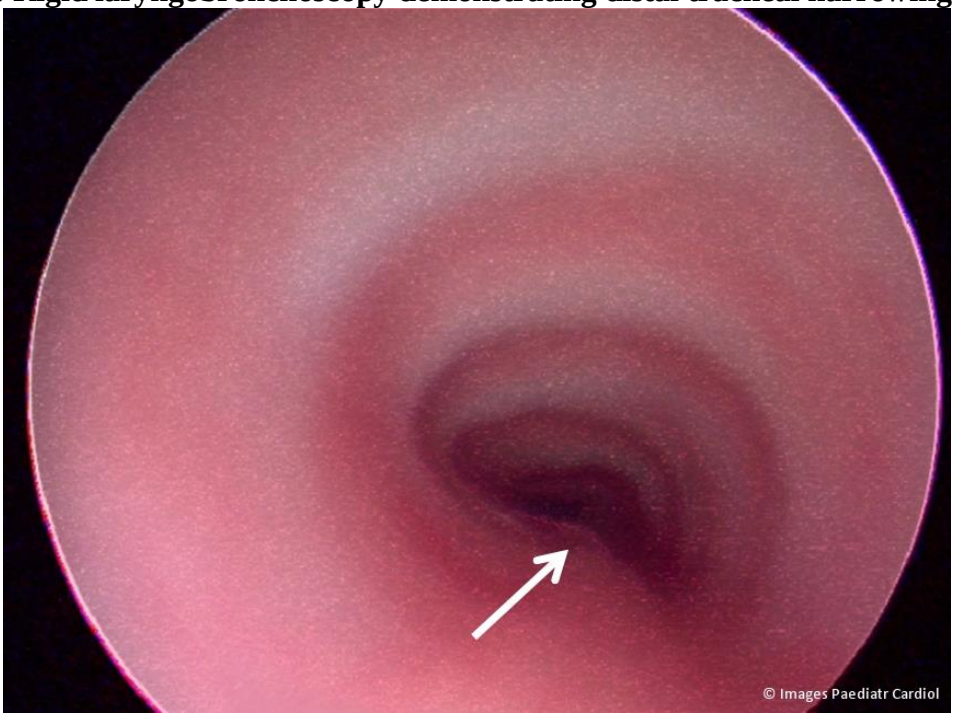
Care report
An 18 days old male, full term product of a non complicated pregnancy, presented to the emergency room with stridor since birth. Initial assessment revealed mild respiratory distress with retractions and abdominal breathing. Chest X ray demonstrated right aortic arch, clear lung fields without cardiomegaly (Fig 1).

Figure 1: AP Chest X ray demonstrating right aortic arch (arrow), clear lung fields without cardiomegaly.



Prior to the emergency room visit he was diagnosed with laryngomalacia according to flexible laryngoscopy at 11 days of life. Because of worsening symptoms, he had a rigid laryngobronchoscopy performed, which revealed distal tracheal narrowing, as well as extrinsic, pulsatile compression of the distal left bronchus (Fig 2).

Figure 2: Rigid laryngobronchoscopy demonstrating distal tracheal narrowing (arrow).



Echocardiogram demonstrated a double aortic arch with a dominant right aortic arch, giving rise to the right common carotid and the right subclavian arteries (Fig 3) and a smaller left aortic arch giving rise to the left common carotid and left subclavian arteries (Fig 4), creating a vascular ring encircling the trachea and the esophagus (Fig 5).

Figure 3: Supra sternal long axis view of an echocardiogram demonstrating a double aortic arch with a dominant right aortic arch

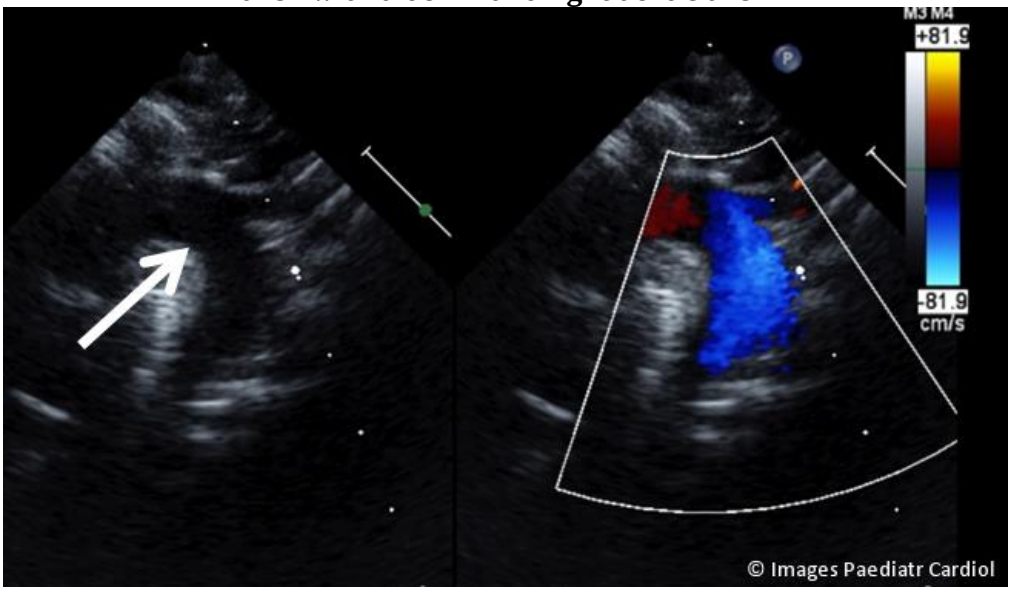


Figure 4: A smaller left aortic arch connecting to the descending aorta.

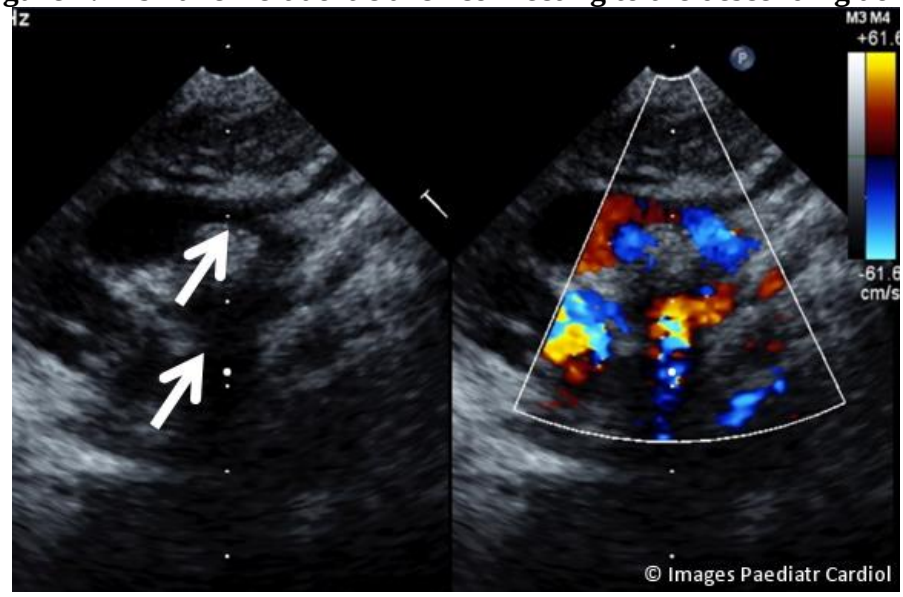
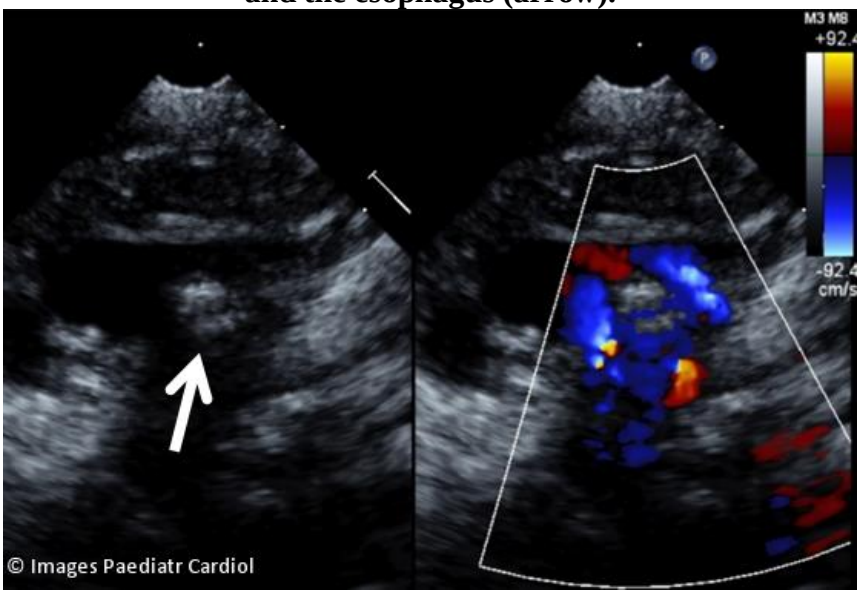


Figure 5: Supra sternal short axis view demonstrating the vascular ring encircling the trachea and the esophagus (arrow).



As a definitive diagnostic imaging prior to surgery, a cardiac catheterization was performed including 3D rotational angiography (Toshiba, Vitrea 3D Enterprise suite). The angiography was performed, while the patient was rapidly paced using transesophageal pacing catheter (Fig 6, highlighted in red), in order to reduce artifact and optimize images quality. The rotational angiogram and 3D reconstruction demonstrated a double aortic arch with a hypoplastic left aortic arch and significant hypoplasia (narrow lumen) of the left isthmus, connecting the left subclavian artery with the ductal ampulla (Fig 6-8).

Figure 6: 3D Rotational angiography demonstrating a double aortic arch with a hypoplastic left aortic arch and significant hypoplasia of the left isthmus (dashed line), connecting the left subclavian artery (full line) and a dominant right aortic arch (dotted line). A: anterior view.

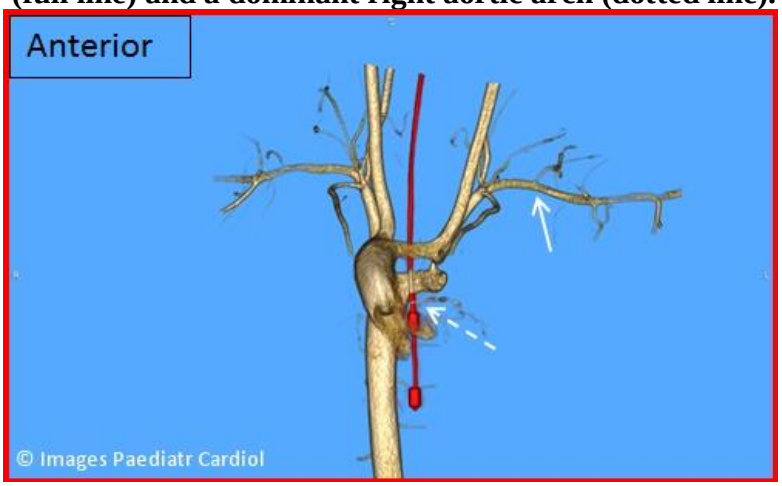


Figure 7: 3D Rotational angiography demonstrating a double aortic arch with a hypoplastic left aortic arch and significant hypoplasia of the left isthmus (dashed line), connecting the left subclavian artery (full line) and a dominant right aortic arch (dotted line). B: Posterior view.

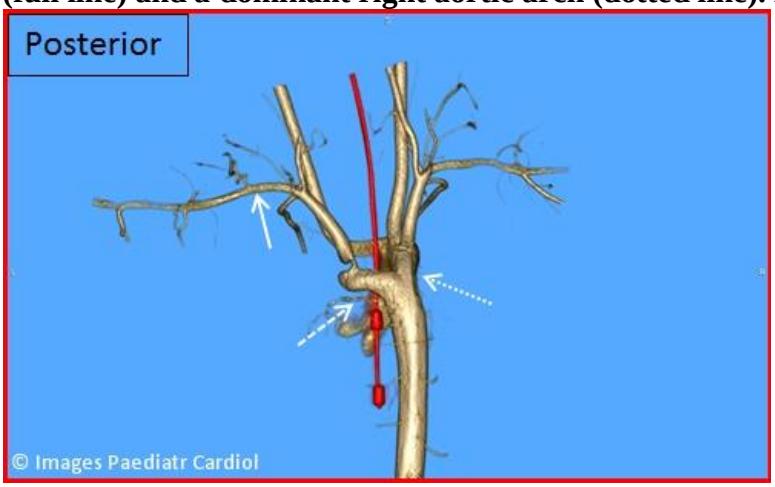
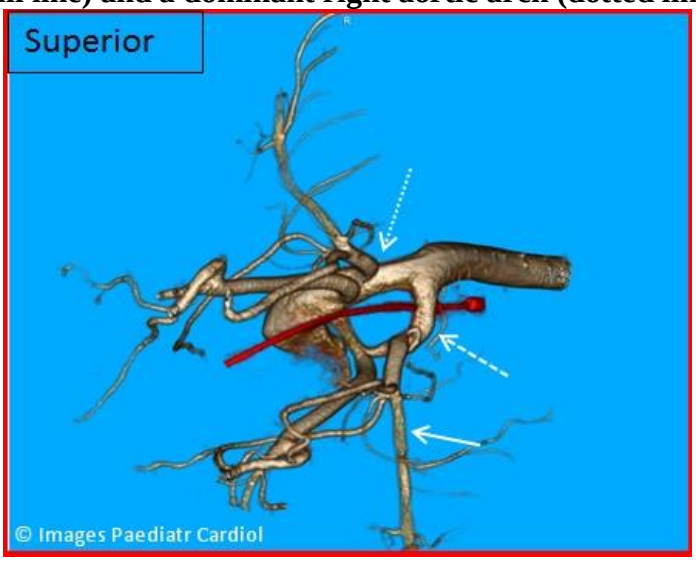
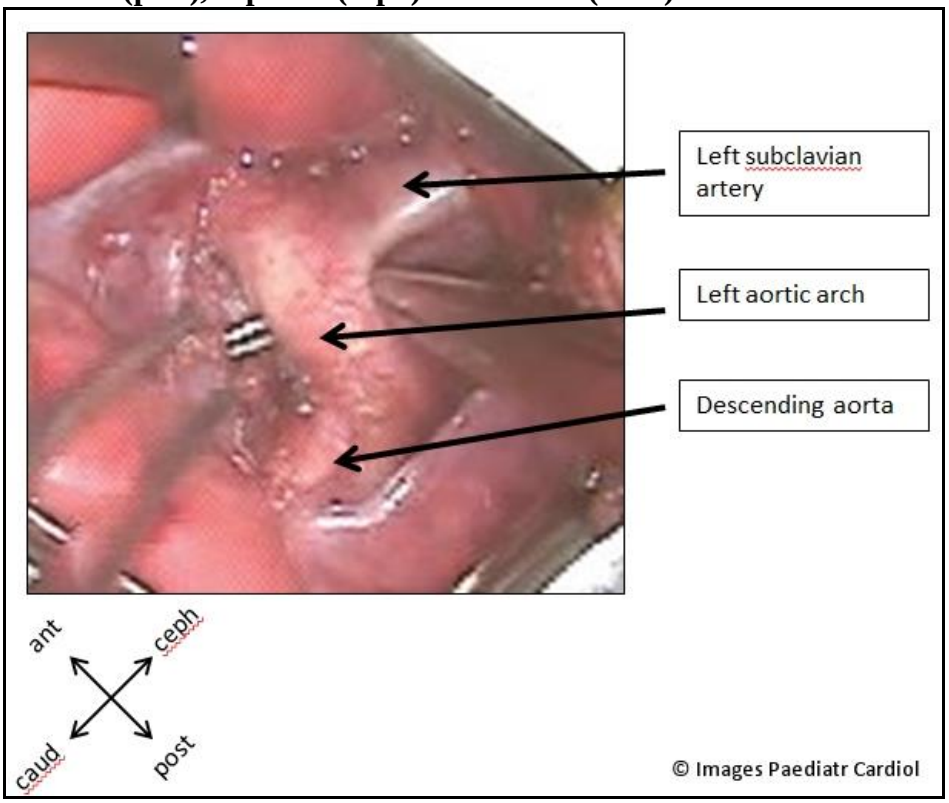


Figure 8: 3D Rotational angiography demonstrating a double aortic arch with a hypoplastic left aortic arch and significant hypoplasia of the left isthmus (dashed line), connecting the left subclavian artery (full line) and a dominant right aortic arch (dotted line). C: superior view.



Surgical repair (Fig 9) was performed through left posterolateral thoracotomy and included ligation, division and oversewing of the two ends of the left aortic arch. Of note, the external appearance of the left aortic arch during surgery did not reveal the hypolastic intra-luminal cavity, as identified by angiography.

Figure 9: Surgical view of the double aortic arch demonstrating the dissected left aortic arch giving rise to the left subclavian artery and connecting to the descending aorta. Anterior (ant), posterior (post), cephalic (ceph) and caudal (caud) orientation is noted.



Post-operative flexible laryngobronchoscopy demonstrated resolution of the airway narrowing, with clinical improvement of the patient's stridor and respiratory distress.

3D rotational angiography is a technique used increasingly for imaging in congenital heart disease.[1,2] To the best of our knowledge, this is the first description of using this modality for imaging of vascular rings in general, and double aortic arch specifically. This modality enables maneuvers such as Adenosine induced ventricular asystole or rapid ventricular pacing, which avoids cardiac contraction movement artifacts.[3] The procedure can be performed under sedation or general anesthesia. In the latter, ventilator manipulation can further reduce respiratory variation artifacts. These factors are especially important in imaging infants and young children, who have high respiratory and heart rates. Although limited data has been published, studies suggest that this technique exposes the patient to lower radiation dose compared to CT angiography,[4,5] which is currently the most commonly used modality for imaging of vascular rings in children. Data acquisition and reconstructions are generated quickly (minutes),[4] and the image quality exceeds that of a classical cardiac CT.[5] Of note, the major limitations of this technique include the need for an invasive procedure and lack of imaging of the surrounding tissues (airways and esophagus).

In conclusion, 3D rotational angiography is an imaging technique that provides clear, accurate and high quality images in preparation for surgery. This technique can be applied for pre-surgical imaging of various vascular anomalies.

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