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# Double Aortic Arch with Atresia of the Left Aortic Arch Proximal to the Left Common Carotid Artery in a Patient with PHACE Syndrome – A Management Conundrum

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## Background

- Complex arch anomalies including Double Aortic Arch (DAA) has been associated with PHACE syndrome.
- Atresia of the proximal segment of the left arch in DAA is extremely rare and can pose a diagnostic and management conundrum due to formation of a loose vascular ring.

## Case Presentation

- A 5-year-old male with PHACE syndrome and an incidental finding on head imaging of a right aortic arch (RAA) with an aberrant left subclavian artery at an outside hospital.
- Clinically, he was asymptomatic, growing well with no respiratory symptoms or dysphagia.
- Cross-sectional imaging with gated CT angiogram was performed which aided in the diagnosis of this rare DAA variant.

## Diagnostic Images

Figure 1(A) and Figure 1(B): 3-D reconstruction using multiplanar format showing the double aortic arch with ipsilateral head and neck vessels and an atretic proximal left aortic arch. Asc Ao: Ascending Aorta, LCC: Left common carotid, R CC: Right common carotid, LSCA: Left subclavian artery, RSCA: Right subclavian artery.



Figure 2: The presumed atretic proximal left aortic arch ligament between the right and left arches is shown as a blue bar. The left-sided ductal ligament connects the dorsal aorta with the distal pulmonary trunk as evidenced by the presence of a ductal dimple under the left common carotid artery (yellow bar).



## Discussion

- Diagnostic clues include:
  - Proximity of the RCC and LCC arteries.
  - Symmetric four vessel sign at the thoracic inlet.
  - Posterior course of a patent segment of the atretic left arch.
  - Presence of the ductal dimple under the LCC artery.
- Surgical intervention has been performed in limited reported cases when patients had symptoms of airway compression or associated left to right shunt related to PDA.

## Conclusion

- Given the “loose vascular ring” and asymptomatic nature, patient specific discussion should be considered with the family to assess optimal management strategy.
- Complementing cross sectional imaging with baseline echocardiography even in asymptomatic cases can be useful in the diagnosis of this rare DAA variant.