

Children's Mercy Kansas City

## SHARE @ Children's Mercy

---

Research Days

GME Research Days 2024

---

May 17th, 11:30 AM - 1:30 PM

### 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

Mohamed Aashiq Abdul Ghayum  
*Children's Mercy Kansas City*

Anmol Goyal  
*Children's Mercy Kansas City*

Aliessa P. Barnes  
*Children's Mercy Kansas City*

Sanket Shah  
*Children's Mercy Kansas City*

Let us know how access to this publication benefits you

Follow this and additional works at: <https://scholarlyexchange.childrensmc.org/researchdays>

 Part of the [Cardiology Commons](#), and the [Pediatrics Commons](#)

---

Abdul Ghayum, Mohamed Aashiq; Goyal, Anmol; Barnes, Aliessa P.; and Shah, Sanket, "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" (2024). *Research Days*. 10.  
[https://scholarlyexchange.childrensmc.org/researchdays/GME\\_Research\\_Days\\_2024/ResearchDay5/10](https://scholarlyexchange.childrensmc.org/researchdays/GME_Research_Days_2024/ResearchDay5/10)

This Poster Presentation is brought to you for free and open access by the Conferences and Events at SHARE @ Children's Mercy. It has been accepted for inclusion in Research Days by an authorized administrator of SHARE @ Children's Mercy. For more information, please contact [hlsteel@cmh.edu](mailto:hlsteel@cmh.edu).

# 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

Mohamed Aashiq Abdul Ghayum, MD<sup>1</sup>; Anmol Goyal, MD<sup>1</sup>; Aliessa P. Barnes, MD<sup>1</sup>; Sanket Shah, MD<sup>1</sup>

<sup>1</sup>Pediatric Cardiology, Ward Family Heart Center, Children's Mercy, Kansas City, MO

## 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, RCCA: Right subclavian artery.

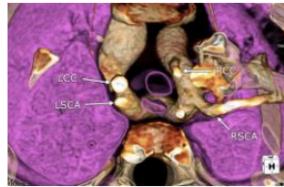


Figure 2: The presumed atretic proximal left aortic arch ligament between the right and left aortic arches is shown as a blue bar. The left-sided ductal ligament connects the dorsal aorta with the distal pulmonary trunk as evidenced by the presence of a ductal dimple under the left common carotid artery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

