

Impact on the trachea in children with prenatally diagnosed vascular ring formed by a right aortic arch

Trisha Vigneswaran V¹, Christopher Oakley¹, Alice Hurn¹, Claire Suckling¹, Grace Moriarty¹, John Simpson¹, Simona Turcu¹, Jane Heraghty¹, Vita Zidere¹, and Andrew Nyman¹

¹Evelina London Children's Hospital

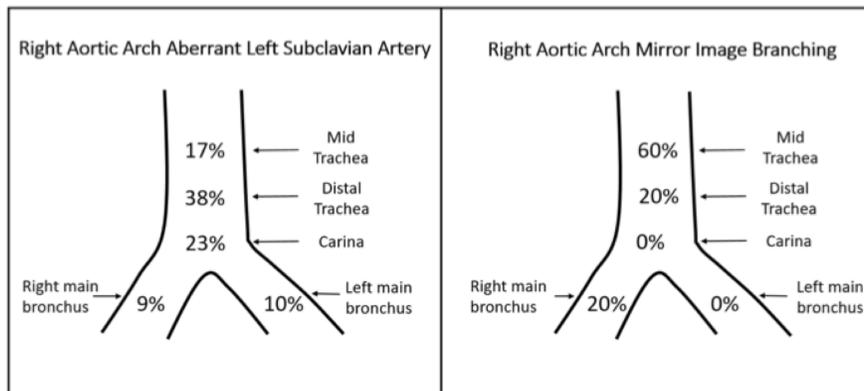
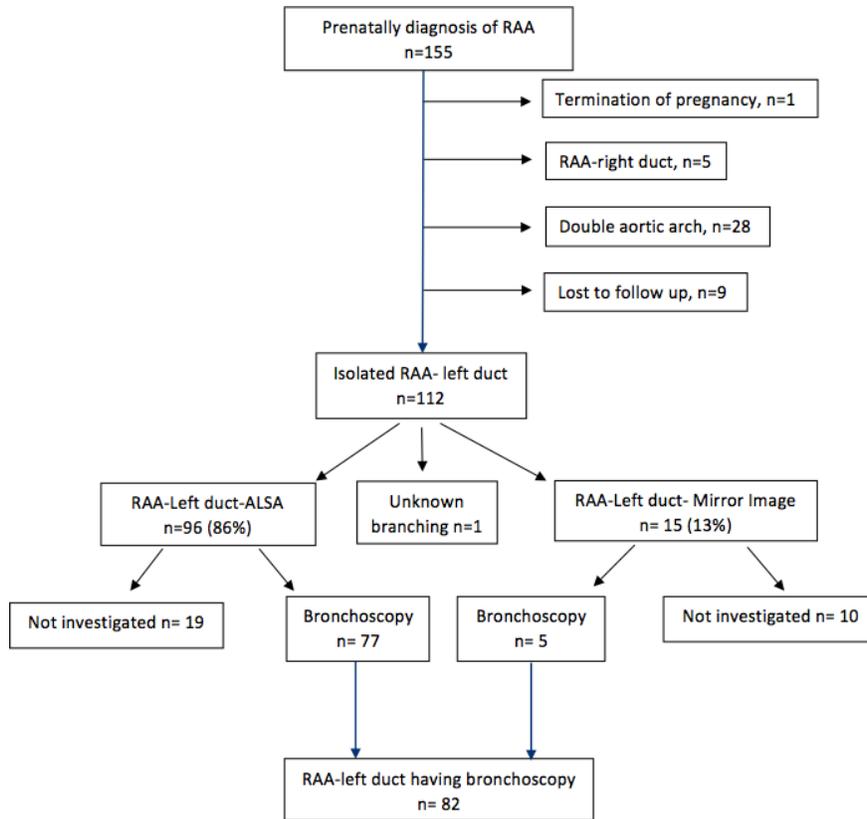
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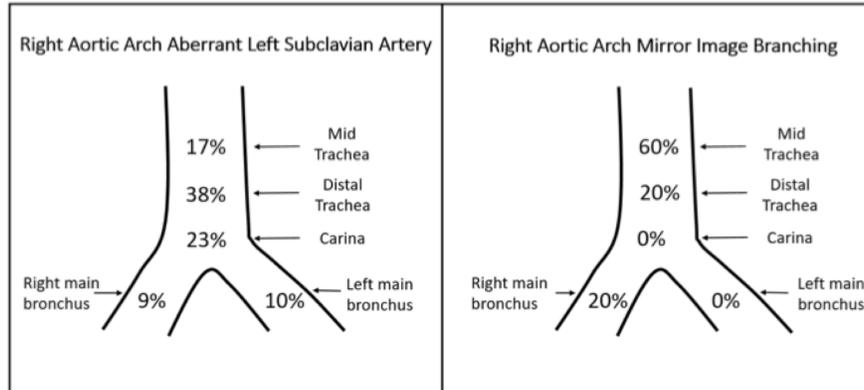
Abstract

Objective: There has been a rise in the prenatal detection of right sided aortic arch (RAA). When associated with a left-sided arterial duct (LD), this forms a vascular ring encircling the trachea. Infants may have symptoms or signs suggestive of tracheo-oesophageal compression but many are asymptomatic. The objective of this study was to investigate the relationship between symptoms and severity of tracheo-bronchial compression assessed by bronchoscopy. **Methods:** Retrospective review of all cases of prenatally diagnosed RAA-LD in the absence of associated congenital heart disease at Evelina London Children's Hospital and Kings College Hospital over a 4-year period between April 2015-2019. Clinical records, fetal echocardiograms and free-breathing flexible bronchoscopy (FB) data were reviewed. **Results:** 112 cases of isolated RAA-LD were identified of whom 82 cases (73%) underwent FB. FB was performed median age of 11 months (range 1-36 months), no complications occurred. Aberrant left subclavian artery (ALSA) was present in 86% (96/112) and mirror image branching (MIB) in 13% (15/112). 34/112 (30%) reported symptoms during follow up. 36/77 (47%) with ALSA who underwent FB showed moderate-severe compression mostly at distal tracheal and carinal level of whom 38% had parent-reported symptoms. Moderate-severe compression was seen in 3/5 (60%) with MIB mostly at mid tracheal level of whom 3 were symptomatic but only 2 with compression. In total 36% (18/50) of investigated asymptomatic patients showed moderate to severe compression. Respiratory symptoms were poorly predictive of moderate-severe compression (positive predictive value 66%). **Conclusion:** The absence of symptoms did not exclude significant tracheal compression. The anatomical effect of the vascular ring is underappreciated when symptoms alone are used as a marker of tracheal compression.

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Table 1- Findings in children with RAA-LD undergoing bronchoscopy.docx available at <https://authorea.com/users/525552/articles/595895-impact-on-the-trachea-in-children-with-prenatally-diagnosed-vascular-ring-formed-by-a-right-aortic-arch>

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Table 2- 2 x 2 table demonstrating predictive value of symptoms for tracheal compression. .docx available at <https://authorea.com/users/525552/articles/595895-impact-on-the-trachea-in-children-with-prenatally-diagnosed-vascular-ring-formed-by-a-right-aortic-arch>