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Double Aortic Arch Anomalies – Anatomic Variations and Their Clinical Impact (15 Years Single Centre Experience)

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Introduction:
Edwards´ hypothetic double aortic arch (AA) can describe all existing morphological variations: with persistence of both complete AA or with an atretic segment or abnormal interruption of one arch. Clinically dominant are ring-forming subtypes, surrounding trachea/oesophagus and causing symptoms from compression; but appreciating other AA anomalies is important as well.

Methods:
Retrospective analysis of all consecutive patients with double AA variations managed during 15 years at our institution (one national centre with 1.2 million paediatric population). This long-term study, similarly like a registry, reflected epidemiology of these rare anomalies.
Analyzed were 27 patients (16F/11M); defining double AA morphological subtypes: Group 1 (ring-forming), Group 2 (w/o vascular ring), Group 3 (with persistent 5^{th} AA). Analyzed were associated congenital heart defects (CHD) and non-cardiac diseases as well.

Results:
In Group 1 were 17 patients (6 with complete double AA, 5 with right AA + dorsal left AA atresia, 6 with right AA and aberrant left subclavian artery + left arterial duct/ligament) with median age at diagnosis 5 months. Only 2 patients (11.8%) in this group had an associated CHD but in 5 patients (29.4%) also non-cardiac genetic syndromes were present. Fifteen patients (88.2%) manifested with compression symptoms (stridor and/or dysphagea) and underwent surgical transsection of the minor AA or ligament.
In Group 2 were7 patients with AA anomalies w/o a vascular ring (with arterial duct from arterial trunk, contralateral to AA side). All presented as newborns and were associated with cyanotic duct-dependent CHD. No non-cardiac pathology was found in this group.
In Group 3 were 3 patients with persistent 5^{th} AA; with median age at diagnosis 5 months. All patients had other CHD, as well as non-cardiac diseases; and AA anomaly was only an accessory finding.

Conclusions:
Our study showed population occurrence-rate of all double AA anomalies 0.023% per thousand children; ring-forming subtypes 0.014%. Understanding the double AA concept is essential to define exactly vessel structures. Although rare, it is of immense clinical importance not only in patients with vascular ring but also when differentiating arterial duct from other collateral pulmonary blood supply in newborns with cyanotic (especially duct-dependent) CHD.