

Isolated right aortic arch prenatally detected: prevalence and post-natal outcome

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Introduction: Right aortic arch (RAA) is usually associated with congenital heart disease (CHD). In the absence of cardiac defects, the significance of RAA has not been determined.

Methods: 925 cases referred to our Fetal Cardiology Centre between January 2008 and June 2015 were retrospectively examined for RAA whether associated with CHD or isolated.

Results: Out of 361 cases with structural heart anomalies, 51 (14%) with RAA were detected. In 25 (49%) cases RAA was associated with CHD: 11 tetralogy of Fallot (TF), 7 pulmonary atresia and ventricular septal defect (VSD), 3 double outlet right ventricle, 2 tricuspid atresia, 1 VSD, 1 truncus arteriosus, 1 absent pulmonary valve with intact ventricular septum. RAA was not associated with CHD in 26 (51%) cases. The AD was left-sided in 20 (77%), 13 of these had Kommerel diverticulum and aberrant left subclavian artery. In 6 (23%) cases aortic arch and AD were both right-sided.

Postnatal outcome was obtained in all cases. During a mean follow-up of 4 years, we observed three complications. Among isolated RAA and left-sided AD, one patient had resection of the arterial ligament at 18 months of age because of progressive symptoms of upper airway obstruction. Another case had a transient and asymptomatic pulse loss of the left upper limb. Among cases with RAA and right AD, one patient had non-confluent pulmonary arteries (PAs) and the left one being perfused by of a second left-sided AD, missed at prenatal examination. After closure of this AD the newborn developed asymptomatic thrombosis of left PA. At late diagnosis the patient was considered unsuitable for surgical reconnection of the disconnected PA.

Conclusions: Prenatal diagnosis of RAA is constantly increased. Nearly half of cases are associated with CHD (most with PA obstruction) and half are isolated. The majority of isolated RAA have a left-sided AD with Kommerel diverticulum and aberrant left subclavian artery. Despite a potential vascular ring in this subgroup, symptoms are very rare and surgical resection is needed only in a minority of cases. In RAA and right-sided AD, fetal and post-natal examination must rule out bilateral AD and disconnection of PAs.