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Contemporary study of fetal right aortic arch: change in detection rate, status of associated anomalies and perinatal outcomes

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Objective: To evaluate the prenatal findings of right aortic arch (RAA), its associated anomalies, and the outcome.
Methods: We reviewed all fetuses with RAA diagnosed between 2010 and 2018 in two centres, from the University Hospital of Wales and Kocaeli University Hospital. Prenatal findings of all fetuses with aortic arch anomalies, intracardiac, extracardiac and genetic abnormalities were studied.
Results: A right aortic arch was identified in 52 fetuses. Mean gestational age at diagnosis was 23 weeks (range, 19-33 weeks). There were 23 cases of isolated aortic arch. Associations with additional intracardiac malformations were found in 29 cases: Tetralogy of Fallot in 10, heterotaxy syndromes in four, double outlet right ventricle in four, VSD in four, crossed pulmonary artery in two and others in five fetuses. The detection rate of RAA increased over the study period from occasional encounter to frequent findings after adopting a three-vessels and trachea view in the screening planes. However, in the three quarters of the study period the majority of patients were referred for a suspicion of congenital heart disease on obstetric scanning. The RAA was suspected with the aid of the three-vessels and trachea view in 24 fetuses by obstetricians or sonographers. The RAA was more frequently detected by non-paediatric/fetal cardiologist in isolated cases (16/23) then the RAA with cardiac abnormalities (8/26).). The karyotype and 22q11 status were checked in 22/52 cases: six had confirmed chromosomal anomalies, two 22q11, three trisomy 18, and one 46XX inv-(9)-(p11q12)(20). The rates of chromosomal abnormalities and 22q11 deletion were 2/11 (18%) in fetuses with isolated RAA, 4/11 (36%) in fetuses with intracardiac anomaly. An extracardiac anomaly was observed in in 3/23 (13%) fetuses with isolated RAA, 6/29 (20%) in fetuses with intracardiac anomaly (total 10 fetuses, 19%). There were six pregnancy interruptions; two intrauterine deaths, 44 live births, two neonatal deaths and two patients were lost to follow-up. The RAA caused symptoms of vascular ring in one patient in the postnatal period.
Conclusion: The RAA has become more noticeable during fetal scans in recent years in spite of almost 50% of these being isolated cardiac lesions. Chromosomal and extracardiac anomalies are lower in isolated RAA but not negligible. The diagnosis of a right aortic arch can be made easily by non-paediatric/fetal echocardiographer whether it occurs as an isolated lesion or in association with other cardiac malformations. If the three vessels-trachea view is used as a routine screening method, the chances of picking up the right aortic arch in the fetus will be higher.