Prenatal diagnosis of right aortic arch: A Spanish multicenter study.


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Introduction: the 3-vessels trachea (3VT) view has led to a higher prenatal detection of aortic arch anomalies. Early diagnosis of right aortic arch (RAA) is important as it maybe associated with cardiac, extracardiac anomalies and chromosomopaties.

Methods: Retrospective, multicenter, echocardiographic analysis of all cases diagnosed prenatally of RAA from 1/2010-12/2011. Type of RAA and branching pattern were assessed. Different diagnosis were sought; vascular ring: RAA with anomalous left subclavian artery (ALSA) and double aortic arch (DAA), no vascular ring: RAA with mirror image branching (MIB) and RAA undetermined type (when the branching pattern was impossible to determine). Intracardiac anomalies were studied. Gestational age at diagnosis, karyotype and outcome data were assessed. Fisher’s exact test was used.

Results: 73 patients had a RAA. 33 had a RAA with ALSA, 5 DAA (all with a predominance of the RAA), 28 RAA and MIB and 6 RAA and undetermined branching. Mean gestational age at diagnosis was 22 weeks. A RAA was associated to intracardiac anomalies in 32 cases (44%). There was a significant association to other heart defects in the case of MIB (78.6%, p<0.001). Most fetuses in the groups of ALSA (84.8%, p<0.001), undetermined type (66.7%) and double aortic arch (60%) had normal hearts. Karyotype was available in 39/73 cases, 8 had anomalies, 4 were 22q11 deletions (2/33 with RAA and ALSA and 2/28 with RAA+MIB). Extracardiac malformations were detected in 6/73 cases, 66.7% of which were RAA and MIB vs 33.5% RAA with ALSA. Pregnancy was interrupted in 14 cases. The rest of on going pregnancies were assessed postnatally with a correct diagnosis in 93% of cases. 3/73 patients with a RAA were symptomatic during the first year of life, 2 of whom had a vascular ring (DAA and RAA+ALSA) and the other had a RAA and MIB. There were 2 deaths related to intracardiac anomalies.

Conclusions: Prenatal diagnosis of RAA and vascular ring is feasible. RAA and MIB is strongly associated to intracardiac anomalies and it may be associated with extracardiac and chromosomal anomalies. Most cases of RAA+ALSA and DAA are asymptomatic during the first year of life.