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Antenatal diagnosis of Double Aortic arch: Associated chromosomal and extra cardiac abnormalities, differential growth of arches during gestation and post-natal outcome.

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Introduction
Double aortic arch (DAO) is a rare congenital malformation that results from failure of regression of both arches during embryogenesis creating complete vascular ring that causes variable degree of compression of the trachea and/or oesophagus. In the absence of large antenatal series, counselling regarding risk of associated chromosomal and extra-cardiac abnormalities (ECA) and postnatal need for surgery remains challenging. From personal observations we also hypothesised that there is differential growth of the arches during pregnancy and initially, a patent left arch may present as atretic postnatally.

Methods
Retrospective, case series review. From computerised databases, we identified all fetuses with DAO seen in four tertiary centres between 01/2014 and 11/2017. We reviewed prenatal findings to assess patency of both arches, associated ECA and chromosomal abnormalities and reviewed postnatal outcome data.

Results
Twenty cases of DAO were identified; three of them diagnosed antenatally as right aortic arch (RAO) and postnatally as DAO. In the majority (60%), indication for cardiac scan was suspected RAO. Mean gestational age at diagnosis was 21+3 weeks (range 16+0 to 28+4). One (10%) of 10 cases that had genetic testing had 22q11 deletion. In the majority (84%) right arch was dominant, in all cases arterial duct was left-sided and in all antenatally diagnosed as DAO, the left arch was patent. In three cases (15%) there were minor additional cardiac abnormalities and one (5%) had ECA. Postnatally, computerised tomography scan (CT) showed atretic left segment in six patients (54%). In the majority (73%) of cases that have been assessed by CT, surgical or catheter intervention has been either performed (mean age = 5.1 months) or planned.

Conclusions
Antenatal diagnosis of DAO is feasible and essential for prompt postnatal management. From our study it appears that risk of chromosomal abnormalities is similar and risk of ECA less when compared with RAO and majority of patients require intervention during infancy. These provisional data also support the theory that during gestation there is preferential growth of the dominant right arch as postnatally some of the patients presented as DAO with atretic left segment.