Clinical significance of prenatally diagnosed right aortic arch and double aortic arch

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Objectives
To evaluate the postnatal outcome of the fetuses with prenatally diagnosed right aortic arch (RAA) and double aortic arch (DAA) without significant cardiac malformations.

Methods
This was a retrospective study of the fetuses prenatally diagnosed with RAA and DAA between January 2010 and December 2018 at Asan Medical Center, Seoul, Korea. We included associated minor intracardiac anomalies, such as small ventricular septal defect, and persistent left superior vena cava.

Results
During the study period, 56 cases of RAA and 18 cases of DAA were prenatally diagnosed and postnatally confirmed. The mean gestational age (GA) at diagnosis was 25.6 weeks and 25.5 weeks for RAA and DAA, respectively. Among the 56 cases of RAA, 32 cases (57.1%) showed left ductal arch (LDA) and three cases (5.3%) had right ductal arch (RDA). 21 cases had closed patent ductus arteriosus before echocardiography. Among RAA patients, 34 cases (60.7%) had an aberrant left subclavian artery, which associated with six cases (10.7%) of the Kommerell’s diverticulum.

Karyotyping was done for 13 patients (23.2%) and only one patient had a Robertsonian translocation 45,XX,der(14;15)(q10;q10). In RAA cases, there were no report of any symptoms related to tracheal or esophageal compression due to vascular ring and a corrective surgery. For the 18 DAA patients, four patients (22.2%) had symptoms related to tracheal and esophageal compression. six patients (33.3%) underwent a corrective surgery and remained in good health until now. During surgery, five cases (27.7%) revealed right dominant arch, and one case (1.5%) had left dominant arch. Karyotyping was done for six patients (66.7%) and none of them had abnormal result.

Conclusions
RAA and DAA can be detected by prenatal echocardiography and may form a vascular ring that can possibly cause obstructive symptoms. However, even with vascular ring, the RAA has a good prognosis and the DAA also has good clinical outcome after the surgical release of vascular ring.