Right aortic arch with vascular ring and aberrant left subclavian artery

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Objective
To present ultrasonographic (US) findings of right aortic arch (RAA) with vascular ring and aberrant left subclavian artery (ALSA).

Methods
This is a case report.

Results
Right sidedness of the aortic arch that affects the position of the aortic branching can occur in association with other congenital heart disease. There is a recognised association with 22q 11 microdeletion, and occasionally with trisomy 21 and other aneuploidies. The prevalence of aortic arch anomalies is estimated to be around 0.1% in the adult population. A 31-year-old woman G2P1 with an unremarkable history, in the 26th week of gestation was referred to our hospital after an isolated finding of hyperechogenic intracardiac focus. The results of the first and second trimester screening were negative. On cardiac assessment, the aortic arch was right sided, and the pulmonary trunk and ductus arteriosus were seen to the left of the trachea. In the three vessel –trachea view, the typical V shape of the pulmonary and aortic arteries could not be obtained. By Color Doppler imaging the the U-sign was visualised and a loose vascular ring was considered to be seen around the trachea. There was no cardiac and extracardiac associated abnormalities. The initial diagnosis was a RAA with ALSA. Fetal karyotyping was offered to rule out chromosomal aberrations primarily 22q 11 microdeletion. Amniocentesis was performed, and the result was normal. The pregnancy is ongoing at 32 weeks gestation.

Conclusion
RAA with a vascular ring is asymptomatic but it can cause some clinical symptoms such as persistent stridor or upper airway obstruction and swallowing difficulties due to compression of trachea and oesophagus, secondary to vascular ring formation. RAA can be associated with other cardiac anomalies such as Tetralogy of Fallot or conotruncal anomalies. Targeted morphologic US examination, fetal chromosomal karyotyping especially evaluation of 22q 11 microdeletion should be offered. Prognosis is very good in isolated cases.