

## MP4-16

### **Aortic arch anomalies associated with vascular ring: Clinical relevance of prenatal diagnosis**

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**Introduction:** In the majority of cases aortic arch anomalies (AAA) associated with vascular rings are detected due to clinical symptoms resulting from tracheal compression. An increasing number of these anomalies are detected now by fetal echocardiography. The purpose of this study was to assess the incidence of postnatal airway obstruction in different types of prenatally detected AAA.

**Methods:** We analyzed all cases of prenatally detected AAA associated with vascular ring and compared findings of fetal echocardiography with postnatal echocardiography, MRI, CT-thorax and clinical symptoms.

**Results:** From 11/2010 –12/2015 14 cases of AAA associated with vascular ring were diagnosed by fetal echocardiography in our tertiary referral center. Only one patient had significant congenital heart disease (VSD). 3 patients had double aortic arch (DAA) with patency of both arches, while another 3 patients had DAA with atresia of the left arch between the left subclavian artery and left ductus arteriosus. The remaining 8 patients had less tight vascular rings formed by a right aortic arch (RAA), aberrant left subclavian artery (ALSA) originating from a left-sided diverticulum of Kommerell and a left sided ductus arteriosus. Chromosomal anomalies (trisomy 21) were present in 2/14 patients. The prenatal diagnosis of a vascular ring was confirmed in all cases by postnatal echocardiography, MRI (4 patients) or CT-thorax (3 patients). 4/14 patients (29%) developed tracheal obstruction and underwent surgery including one newborn and one infant with DAA, one infant with incomplete DAA and one infant with RAA, ALSA and left-sided ligamentum arteriosum. 10 patients (71%) remained asymptomatic without stridor or swallowing problems during a mean follow-up of 2.6 years.

**Conclusions:** Fetal echocardiography provides important new information on the clinical relevance and natural history of aortic arch anomalies associated with vascular ring: Patients with RAA, ALSA and left ductus arteriosus resulting in a loose vascular ring remained asymptomatic in the majority of cases during infancy and childhood. Although tracheal compression was present in half of the patients with DAA, 50% of these patients with complete vascular ring were asymptomatic. Since airway obstruction may develop later in life, patients with vascular ring should be offered long-term follow-up.