Vascular Rings: prenatal diagnosis and implication on postnatal treatment.

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Objectives: to describe prenatal diagnosis and management of isolated vascular rings (VRs) at a single tertiary care institution.

Background: VRs refer to a group of congenital anomalies that interfere with and may compress the esophagus and trachea, leading to respiratory distress shortly after birth or to tracheoesophageal compression later in life. Prompt diagnosis and treatment of these congenital abnormalities can be lifesaving.

Methods: the department registry was retrospectively searched for all patients born between January 2002 and December 2012 with a prenatal diagnosis of congenital isolated VR. The variables assessed were indications for referral, gestational age at diagnosis, and postnatal outcome. Karyotyping and prenatal testing for 22q11 microdeletions were offered to all parents. Magnetic resonance imaging or computed tomography was performed immediately in symptomatic patients or electively in the first month of life in those with complete vascular rings, for excluding esophageal or tracheal compression.

Results: 53 fetuses referred to our centre for increased risk or suspicion of congenital heart disease had diagnosis of isolated VR at a mean gestational age of 24 weeks. The most common type of VR was left aortic arch (LAA) with aberrant right subclavian artery (ARSA) seen in 31 patients of whom 2 showed trisomy 21 at karyotyping and parents opted for termination of pregnancy (TOP). The second most common type was right aortic arch (RAA) with left aberrant subclavian artery (ARSA) arising from the Kommerell’s diverticulum, seen in 15 patients. In 7 of them a circumflex retroesophageal aortic arch was found. Mild stenosis of ALSA was suspected in one newborn patient but he still didn’t need of any treatment. Seven fetuses had double aortic arch with RAA larger than LAA. One of them had associated a 22q11 deletion and parents chose TOP. Three patients with DAA and 2 with RAA and ALSA developed symptoms from tracheal compression and immediately underwent successful surgical correction. In one of them tracheoplasty was performed as well.

Conclusions: Prenatal diagnosis of these vascular anomalies may be useful for improving postnatal management of affected children, who should receive a prompt surgical correction as soon as symptoms appear, avoiding tracheomalacia.