Results after surgery for vascular rings; a study of 40 consecutive cases operated 1994-2012 in a single institution

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Objective
Vascular rings are embryological malformations of the aorta and the great vessels that can give rise to complications from the encircled organs, mainly the trachea and the esophagus. Diagnosis is commonly delayed after onset of symptoms. Treatment is surgical.

Methods
Our institution serves about half of Sweden’s 9 million inhabitants. From 1994-2012, 40 patients (mean age 5.7 years, range 1 month to 16 years, 24 girls) underwent surgery for right aortic arch (16 cases), double aortic arch (13), aberrant right subclavian artery (9) and pulmonary sling (2). Symptoms were breathing problems, feeding/swallowing difficulties, or both. Three patients showed acute respiratory failure prior to operation. Patients had no other congenital heart defect except for PDA (2 cases). Co-morbidities were Down-syndrome (2), Turner-syndrome (1), diabetes type I (1), Behcet (1) and aberrant bronchal anatomy (2).

Treatment consisted of division of one of the double aortic arches, division of the ligamentum arteriosum, division and re-implantation of the aberrant pulmonary artery and division with or without re-implantation of the aberrant subclavian artery. A prominent or aneurysmal Kommerell diverticulum was resected or oversewn when found necessary. Aortopexy was performed as indicated.

Results
No short or long-term mortality. Complications were seen in four patients (10%) consisting of incomplete Horner syndrome (2 cases) and transient post-thoracotomy neuralgia (1). One patient required three operations for complications: ligature of lymphatic vessel for chylothorax, aortopexy for continued tracheal impression and puncture of pericardial effusions. This patient also suffered paresis of the recurrent nerve. One patient suffered tracheal fibrosis as a consequence of long-standing impression, leading to resection and end-to-end anastomosis.

Conclusions
The long time to diagnosis, severity of symptoms as well as the good short- and long-term prognosis after surgery show that these congenital malformations require a high index of suspicion in pediatricians for children with wheezing, stridor, respiratory distress and dysphagia. Surgical treatment almost completely relieving symptoms is performed without mortality and with a low complication rate.