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Vascular Rings - Aortic Arch Abnormalities. Single Center 31 years of Experience

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OBJECTIVE: To retrospectively analyze our center's experience in the diagnosis and management of this rare cardiac malformation (< 1% of congenital heart diseases) and it's complications

METHODS Between January 1986 and December 2017, 34 congenital vascular rings were submitted to our Department for surgical correction. There were 26 infants (age: 12 days-12 months, median: 4.5 months; weight: 1.9-8.9 kg, median: 7.4 kg), and 8 children (age: 1-6 years, median: 3.5 years; weight: 7.5-24 kg, median: 15.5 kg). Patients were divided into five groups: 1) Double aortic arch with both arches patent and left ligamentum arteriosum (12 cases), 2) Double aortic arch with atresia mainly in the distal part of the left arch and left ligamentum arteriosum (5 cases), 3) Right aortic arch with left ligamentum arteriosum (11 cases), 4) Left aortic arch with aberrant right subclavian artery in combination with CoA and/or PDA (4 cases), and 5) Pulmonary artery sling (2 cases). Main diagnostic tools were 1) cardiac echo 2) oesophagography 3) bronchoscopy and lately 4) CT angiogram with 3D reconstructions and 5) MRI angiogram

RESULTS We used the left posterolateral thoracotomy for the majority of the cases except 2 cases with pulmonary artery sling where median sternotomy was performed. In all cases we did extended mobilization of the compressed structures from surrounding soft fibrous bands. We also had to perform tracheal plasty reconstruction to one infant with pulmonary artery sling due to severe tracheomalachia. We had no intraoperative mortality but in the postoperative period, 1 neonate died of respiratory failure and sepsis. The children have been followed up every 6 months for the first three years and then once every year. All are clinically well and mostly asymptomatic.

CONCLUSIONS High suspicion, early diagnosis and prompt surgical treatment are necessary to relief symptoms and avoid future severe complications, especially tracheomalachia.