Early And Late Results Of Surgically Managed Congenital Vascular Rings

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Objectives
Persistent respiratory symptoms or feeding problems in children may be associated with a congenital vascular ring. Increasing awareness for this problem exists with pediatricians, but the long-term outcomes are not well described. This study aims to analyze the clinical presentation and surgical treatment of a series of vascular rings, and to evaluate risk factors for mortality and late outcome.

Patients and methods
Since 1993, 58 vascular ring patients (55% male) were treated surgically at our center. Median age at operation was 1 year (range 3 days-27 years). Presenting symptoms were mainly respiratory (86%), including need for preoperative ventilation in 16%, 32% had feeding problems, 2 patients were asymptomatic. The most common diagnosis was double aortic arch (55%), followed by right arch with aberrant left subclavian artery and ductal ligament (36%), left arch with right arteria lusoria (7%), and pulmonary artery sling (2%). Associated anomalies (cardiac, gastrointestinal, chromosomal) were present in 36%. Left thoracotomy for interruption of the vascular ring was the preferred access in 90% of cases.

Results
Median time to extubation and hospital stay were 0.4 days (range 0-8) and 5 days (range 3-371) respectively. Mean follow-up was 7.8 ± 5.7 years, and was 100% complete. Mortality was 8.6% (5 patients), occurred during early follow-up (within 1.5 years postoperatively), and was associated with anatomical diagnosis (p<0.05), preoperative intubation (p<0.0001) and concomitant anomalies (p<0.05). Freedom from symptoms at 1 month and 6 months was 64% and 81% respectively. Freedom from inhalation therapy at last follow-up was 83%. Dysphagia symptoms always disappeared. A significant relationship was found between the freedom from symptoms at 1 month, at 6 months, and freedom from inhalation therapy at last follow-up, with preoperative ventilation (p<0.0001) and anatomical diagnosis (p<0.05).

Conclusion
Surgical relief of tracheo-oesophageal compression by a vascular ring is usually effective, with a swift disappearance of symptoms. In 17% of children however, respiratory symptoms persist, necessitating chronic inhalation therapy. Associated anomalies may compromise the short-term outcome. Patients with a double aortic arch, especially when ventilated before operation, are at higher risk to remain symptomatic during long-term follow-up, particularly at the occurrence of infectious exacerbations.