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Vascular ring surgery: future trends and challenges in diagnosis and treatment

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Vascular rings are congenital aortic arch malformations in which the arch and its branching vessels encircle the oesophagus and trachea. The extent of symptoms and the age at initial presentation depend upon the exact arch configuration. Patients with anatomically complete vascular rings generally present earlier in life than their counterparts with incomplete ones. The latter may not necessarily lead to signs of disease and often remain undiagnosed for a long time. In contrast, the complete rings produce compression symptoms in almost all affected patients [1, 2]. Two main variants of true vascular rings can be distinguished: double aortic arch (DAA) and right aortic arch with left ligamentum (RAA-LD). According to large surgical series, they combined represent the majority of all symptomatic vascular rings [3–5]. Since the first surgical division of a vascular ring by Robert E. Gross at Boston Children's Hospital in 1945, care for children with these types of vascular abnormalities has evolved dramatically. Nowadays, CT or MRI complements echocardiography in the preoperative work-up, and for surgical planning, the barium oesophagram is needed only in exceptional cases. In addition, the surgeon can accurately assess the success of the measures utilizing pre- and perioperative flexible bronchoscopy. Operative mortality does rarely occur in contemporary series. Furthermore, the operative morbidity following the surgical management of DAA or RAA-LD is comparatively low, and complications like chylothorax, pneumothorax, pleural effusion and bleeding rarely have a lasting negative effect on the course of the patient [4]. Nevertheless, a subset of patients suffers from residual symptoms and signs of tracheo-oesophageal compression, and some even need reintervention or reoperation. Prenatal, early detection of vascular rings could play an essential role in detecting the symptoms of tracheo-oesophageal compression promptly after birth and immediately initiating imaging and treatment.

The manuscript of Swarnkar *et al.* [6] describes the effect of prenatal diagnosis on surgical outcome and symptomatic relief after vascular ring surgery in children without associated major congenital heart disease over a 14-year period. Even though there are limitations related to this single-centre report (retrospective methodology, relatively short follow-up, lack of a validated grading system for symptoms and signs of vascular rings after birth), the most striking finding of this elegant study is that prenatal detection of the vascular ring is beneficial to achieve good outcomes. The hypothesis that prenatal awareness of the problem leads parents and caregivers to earlier identification of symptoms and consequently earlier initiation of surgical therapy is appealing. A total of 132 children underwent surgery for DAA or RAA-LD, and a prenatal diagnosis was made in an astonishingly high proportion (76%) of patients. There was no operative mortality, and the 6% of the patients suffering from postoperative complications like vocal cord palsy, prolonged chylothorax or wound healing difficulties are consistent with recent results from other studies. Over the whole cohort, persistent symptoms were more frequently seen in patients with DAA compared to those with RAA-LD. Comparing the pre- and postnatally diagnosed patients after 1 year revealed a remarkably higher number of patients with residual symptoms or signs of tracheo-oesophageal compression in the postnatally diagnosed group (64% vs 31%). Finally, the authors conclude that a multicentre study is needed to assess the surgical approaches in prenatally detected and asymptomatic cases. This summary is undoubtedly correct, as prospective multicentre studies on the treatment of congenital vascular rings are unfortunately lacking [7].

The community of paediatric medical providers continues to improve the understanding and management of congenital vascular rings, but several significant challenges remain:

1. Increasing the accuracy of prenatal and postnatal imaging to diagnose tracheo-oesophageal compression in the cases with no or mild symptoms early, consequently reducing the impact of secondary tracheobronchomalacia.
2. Creating evidence on indications for surgical release of vascular rings in asymptomatic patients and those with partial vascular rings and a Kommerell's diverticulum.

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3. Improving surgical techniques to reduce the number of patients with complications in the short term and residual symptoms or signs of tracheo-oesophageal (re-)compression after releasing a vascular ring in the mid-to-long-term follow-up.

To address these issues before guideline-quality studies are available, a consensus group could conduct a systematic review of data on diagnosing and treating patients with congenital vascular rings to guide medical caregivers of all relevant specializations in managing this challenging patient group.

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