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Tracheal reconstruction in patients with aortic arch anomalies and tracheomalacia

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Introduction:
Developmental anomalies of the aortic arch, like double or right-sided aortic arch, may result in compression of the trachea and esophagus. Patients may present with neonatal stridor, respiratory distress and feeding difficulties. The treatment of choice is vascular decompression surgery.
We report our experience in the management of aortic arch anomalies using an adaptive approach of vascular decompression and tracheal reconstruction.

Methods:
A retrospective analysis of data from 24 pediatric patients with aortic arch anomalies who underwent tracheal decompression surgery in our institution between 2008 and 2013 was conducted. Age, tracheal pathology, surgical technique, associated diseases, and follow-up data were analyzed.

Results:
The patients were aged between 9 days and 16 years at the time of the surgery (median 3.2 yr), weight range was 2.3 to 64 kg (median 12.8 kg). Vascular pathologies included double aortic arch (complete vascular ring formation) in 12 patients, right-sided aortic arch in 10 patients and an aberrant subclavian artery in 2 patients. Tracheomalacia was diagnosed by tracheobronchoscopy in 8 patients. Accompanying heart defects comprised ASD, VSD and valvular anomalies. One patient was diagnosed with 22q11.2 microdeletion syndrome.
All patients underwent vascular decompression surgery and, in the case of a congenital heart defect, corrective heart surgery. Based upon the findings of severe tracheomalacia during intraoperative tracheobronchoscopy, four patients underwent resection of the malacic segment and tracheal reconstruction during the same surgery.
Follow-up data were available for 17 patients for up to 5 years (median 17 months) and proved a clinically stable outcome. One patient died 15 months after the surgery due to complications of a congenital heart defect.

Conclusions:
Although vascular decompression surgery is the current standard of care in patients with aortic arch anomalies and tracheal compression, this approach might not be sufficient in patients with concomitant tracheomalacia. We therefore recommend an intraoperative evaluation of the tracheal pathology. Patients with localized tracheomalacia may benefit from resection of the malacic tracheal segments with subsequent tracheal reconstruction during the same surgery.