Surgical Therapy of Obstructive Airway Disease in Patients with Intrinsic Tracheobronchial Malformations or Airway Stenosis Associated with Vascular Ring – Long Term Results


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Introduction: We analyzed the outcome of surgical airway reconstruction using cardiopulmonary bypass (CPB) as additional technique during more extensive reconstruction procedures which would have been limited by conventional ventilation.

Methods: In this retrospective single center analysis, we included all patients presenting in the period between 2008 and 2017 with severe respiratory failure due to airway obstruction/compression caused by (1) intrinsic malformations of the trachea leading to respiratory failure or (2) vascular and/or congenital heart defects associated with respiratory failure. All patients underwent bronchoscopy, additional imaging was performed by CT or MRI, and in selected cases by cardiac catheterization. Follow-up data were obtained from follow-up examinations.

Results: 114 patients, aged 0.8 years [10 days – 49 years] (Median [range]) presenting with vascular ring (78 out of 114), intrinsic tracheal malformations (n=24/114), pulmonary sling (n=12/114) were included. 33/114 patients had additional cardiac malformations including VSD (n=15/33), Fallot/DORV 4/33, aortic arch hypoplasia (1/33) or complex CHD (13/33). Surgical techniques included a) decompression of airway by transection of ligaments or vascular remnants (n=82), b) partial resection of stenotic segments of the trachea (n=19), c) slide tracheoplasty (n=9). In selected cases, external re-expansion of trachea was used (n=4). CPB was used in 75/114 patients and was a pre-requisite for all patients in whom tracheal procedures were needed or whenever additional intracardiac repair was performed. During the follow-up period, 10 syndromic patients died: 3 patients (Trisomy 21 n=1, VACTERL n=2) died due to neurological complications. Seven patients died due to multi-organ failure (one due to sepsis after leakage of tracheal anastomosis). The surviving 104 patients showed clinical improvement or – if previously ventilator dependent - could be weaned successfully.

Conclusions: Relief of vascular compression and/or reconstruction of the trachea and main bronchi can be performed safely in patients with severe airway malformations by using CPB and modern reconstructive techniques. However, pre-existing syndromatic disease and co-existing multimorbidity must be taken into account in decision making.