Aortopexy for the management of paediatric tracheomalacia – the Alder Hey experience 2007-2017

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Introduction

Tracheomalacia refers to a localised or generalised flaccidity of the tracheal wall causing reduced luminal calibre and so a degree of airway obstruction. Clinically, there is a spectrum of severity ranging from intermittent stridor on exertion through to acute life threatening episodes (ATLEs; also referred to as “dying spells”). Whilst many different treatment strategies have been proposed for the management of tracheomalacia,¹ most series report on the utilisation of aortopexy as their treatment of choice for tracheomalacia secondary to vascular compression. There remains considerable heterogeneity within the literature with inconsistencies in both the approach taken and the techniques employed. Furthermore, limited data exists on patient selection and the long term outcomes of children who have undergone this procedure.² The relatively small number of case series found in the literature likely reflects both the rarity of severe tracheomalacia as a condition and the specialisation of aortopexy as a procedure. This study aims to report the experience of managing severe tracheomalacia by way of aortopexy in a large UK paediatric centre.

Method

Retrospective review of case notes of children undergoing aortopexy at our institution. This study was approved by our institution’s Clinical Audit Department, and all data was kept anonymous and encrypted throughout. Data collation and the calculation of all descriptive statistics were performed using Microsoft Excel for Mac (version 15.35, 2017, Microsoft, Redmond, WA).

Results

25 patients (mean age 9.4 months, range 2 weeks–2.9 years) underwent aortopexy for tracheomalacia caused by external vascular compression. The gender split of the patients was unequal with an apparent male preponderance (19 males; 76%). Mean gestational age at birth was 35.2 weeks (range 24-40) and mean body weight at operation date was 6.3 kg (range 3.2-12.3 kg).

ATLEs precipitated investigation in 72% of cases (n=18). Other symptoms and signs noted at presentation included stridor (80%, n=20), recurrent episodes of oxygen desaturation (76%, n=19), breathing difficulties whilst feeding (24%, n=6) and failure to extubate (16%, n=4). Twelve patients (48%) had required a period of mechanical ventilation at some point pre-operatively.

The full range of co-morbidities is listed below:

Comorbidity	n
Gastro-oesophageal reflux	14
Oesophageal atresia	6
Trache-oesophageal fistula	6
Laryngomalacia	5
Atrial septal defect	4
Complex congenital heart disease	3
Trisomy-21	3
VACTERL association	3
Patent ductus arteriosus	2
Patent foramen ovale	2
Others (all n=1): asthma, CHARGE syndrome, cerebral palsy, chromosome microdeletion 15q13.3, cleft lip and palate, developmental delay, dextrocardia, dystonia, Goldenhar syndrome, hypoxic ischaemic encephalopathy, laryngeal cleft, Smith-Lemli-Opitz syndrome, pulmonary atresia, spina bifida occulta, ventricular septal defect	

Documented co-morbidities diagnosed preoperatively in the patient group

Computer tomography angiography was the imaging methodology of choice for delineating the surrounding vasculature. Ten patients (40%) were found to have tracheomalacia secondary to innominate artery compression

CTA (axial section) showing mid-tracheal stenosis caused by external compression from the innominate artery (MLTB showed pulsatile mid-tracheomalacia)

It is our practice to perform aortopexy through a mini-manubriotomy approach in a combined procedure involving both ENT and cardiothoracic surgeons. Intraoperative bronchoscopy was used routinely to facilitate adjustment of the aortopexy sutures.

Average follow up was 4 years. Notably, 17 patients (68%) were found to have had a full remission of symptoms postoperatively and have been discharged from all associated services. One patient required a repeat procedure and only 4 patients required a tracheostomy to be performed after the original aortopexy.

Conclusion

Aortopexy offers an effective method of treating severe tracheomalacia due to vascular compression. The majority of these cases present initially to ENT. Our figures show that aortopexy is an effective treatment of tracheomalacia due to vascular compression and avoids tracheostomy in the majority of cases.

References

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