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Aortopexy for the management of paediatric tracheomalacia – the Alder Hey experience

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Main Article

Mr S P Williams takes responsibility for the integrity of the content of the paper

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Abstract

Background. Whilst aortopexy is an accepted and established procedure, there remains considerable heterogeneity within the literature, with inconsistencies in both the approach taken and the technique employed. Furthermore, limited data exist on both patient selection and long-term outcomes. This study aimed to report the experience of managing severe tracheomalacia by way of aortopexy in a large UK paediatric centre.

Method. A retrospective case note review was conducted. Mean follow up was five years.

Results. Twenty-five patients underwent aortopexy for severe tracheomalacia caused by external vascular compression. Acute life-threatening events precipitated investigation in 72 per cent of cases. Twenty-one patients initially presented to ENT services for investigation, which comprised upper airway endoscopy and imaging with computed tomography angiography. Post-operatively, the majority of patients demonstrated complete resolution of symptoms and were discharged from all associated services. Only four patients required a tracheostomy.

Conclusion. Aortopexy offers an effective method of treating severe tracheomalacia due to vascular compression.

Introduction

Tracheomalacia refers to a localised or generalised flaccidity of the tracheal wall that causes reduced luminal calibre by at least 50 per cent and so a degree of airway obstruction. It can be classified as primary, occurring as a result of an intrinsic abnormality within the trachea itself, or secondary, a result of tracheal compression from a surrounding structure. Clinically, there is a spectrum of severity, ranging from intermittent stridor on exertion through to acute life-threatening episodes (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. First described in the 1940s,[2] aortopexy involves the suspension of the compressive vascular structures from the posterior surface of the sternum, thereby alleviating the external vascular compression from the anterior trachea wall. The nomenclature used suggests direct involvement of the aorta itself; however, other vessels are frequently involved, with the innominate artery being the most notable example.[3]

Aortopexy is, as a concept, far from novel. Nevertheless, there remains considerable heterogeneity within the literature, with inconsistencies in both the approach taken and the techniques employed. More fundamentally, limited data exist on patient selection and the long-term outcomes of children who have undergone this procedure.[4]

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 aimed to report the experience of managing severe tracheomalacia by way of aortopexy in a large UK paediatric centre.

Materials and methods

Hospital records of all patients who underwent aortopexy at Alder Hey Children's Hospital over an 11-year period (2007–2017) were reviewed retrospectively. As well as patient demographics, data collected included the original presenting symptoms, pre-operative investigation(s) and the operative method employed in each case. The post-operative course and duration of follow up were also recorded.

This study was approved by our institution's Clinical Audit Department, and all data were kept anonymous and encrypted throughout. Data collation and the calculation of all descriptive statistics were performed using Microsoft Excel spreadsheet software for Mac (version 15.35, 2017; Microsoft, Redmond, Washington, USA).

Results

Patients

Twenty-five 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 per cent). Mean gestational age at birth was 35.2 weeks (range, 24–40 weeks) and mean body weight at operation date was 6.3 kg (range, 3.2–12.3 kg).

Acute life-threatening episodes precipitated investigation in 72 per cent of cases ($n = 18$). Other symptoms and signs noted at presentation included stridor (80 per cent, $n = 20$), recurrent episodes of oxygen desaturation (76 per cent, $n = 19$), breathing difficulties whilst feeding (24 per cent, $n = 6$) and failure to extubate (16 per cent, $n = 4$). Twelve patients (48 per cent) had required a period of mechanical ventilation at some point pre-operatively. The most frequently reported co-morbidity was gastroesophageal reflux (56 per cent, $n = 14$). The full range of co-morbidities is listed in Table 1.

Table 1. Documented co-morbidities diagnosed pre-operatively in patient group

Co-morbidity	Cases (*n*)
Gastroesophageal reflux	14
Oesophageal atresia	6
Tracheoesophageal 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

Other co-morbidities (all $n = 1$) included: asthma, 'CHARGE' syndrome (i.e. coloboma, heart defects, atresia choanae (also known as choanal atresia), growth retardation, genital abnormalities and ear abnormalities), cerebral palsy, chromosome 15q13.3 microdeletion syndrome, cleft lip and palate, developmental delay, dextrocardia, dystonia, Goldenhar syndrome, hypoxic ischaemic encephalopathy, laryngeal cleft, Smith–Lemli–Opitz syndrome, pulmonary atresia, spina bifida occulta, and ventricular septal defect. VACTERL = vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies and limb abnormalities

Diagnostic method

Twenty-one patients (84 per cent) initially presented to ENT services and underwent diagnostic microlaryngotracheobronchoscopic examination. The remaining four patients (16 per cent) underwent investigation as part of an ongoing admission to intensive care, and had tracheomalacia diagnosed by way of flexible bronchoscopic examination. The distal segment of the trachea was the portion affected in the majority of cases (60 per cent, $n = 15$), with the mid-segment affected in 12 per cent ($n = 3$), the upper-segment in 4 per cent ($n = 1$) and a more diffuse narrowing found in the remaining patients on examination.

Computed tomography (CT) angiography was the imaging methodology of choice for delineating the surrounding vasculature. Ten patients (40 per cent) were found to have tracheomalacia secondary to innominate artery compression. Tracheal compression from a vascular ring was present in four cases (16 per cent).

Operative method

It is our practice to perform aortopexy through a mini-manubriotomy approach in a combined procedure involving both ENT and cardiothoracic surgeons. A thymectomy is performed to allow access to the anterior mediastinum. Once identified, the compressive vasculature is mobilised and size 4-0 Prolene® sutures are used to suspend these vessels from the trachea, so they can be distracted from the anterior tracheal wall and thus widen its internal diameter. Intra-operative bronchoscopy was used routinely to facilitate adjustment of the aortopexy sutures as the compressive vasculature is lifted from the anterior tracheal wall.

The four patients noted to have compression from a vascular ring required more heterogeneous procedures, with ligation and division of the compressive structures prior to aortopexy suspension. On imaging with CT angiography, all of these patients were found to have a right-sided aortic arch with an aberrant left-sided subclavian artery. Two of these cases were approached as above, by way of mini-manubriotomy, with the other two requiring access by way of a left lateral thoracotomy.

Post-operative course

In the initial post-operative period, the vast majority of patients were extubated (92 per cent, $n = 23$) and had short stays in intensive care (mean = 3.6 days; range, 1–12 days). Post-operative complications included respiratory tract infection (12 per cent, $n = 3$), vocal fold palsy (8 per cent, $n = 2$) and wound infection (8 per cent, $n = 2$).

Two patients (8 per cent) failed multiple attempts at extubation and required a tracheostomy to be placed during the same admission. Three further patients (12 per cent) were initially discharged, but remained symptomatic with stridor and recurrent desaturations. Of these, one underwent a revision aortopexy with an excellent result. The remaining two cases required tracheostomy. Considering the four patients who required post-operative tracheostomy: two remain dependent on tracheostomy-delivered long-term ventilation (at five and six years post-operatively), one remains tracheostomy dependent (at six years post-operatively) and one patient died following a subsequent cardiothoracic procedure performed at a separate institution for complex congenital cardiac disease (tetralogy of Fallot with major aortopulmonary collateral arteries).

With respect to the entire patient group, mean follow-up duration was 5.2 years (range, 1.2–8.5 years). Notably, 17 patients (68 per cent) were found, at post-operative out-patient review, to have had a full remission of symptoms; they were subsequently discharged from all associated services. The remaining four patients (without tracheostomy) continue to receive ongoing follow up with ENT for monitoring with interval microlaryngotracheobronchoscopic examination as required. These patients are clinically improved but continue to experience intermittent episodes of stridor. No patients in this group have required further intervention.

Discussion

Tracheomalacia is a condition which, when severe, can precipitate acute life-threatening episodes, and can lead to a fatal outcome with an implication, in some cases, of sudden infant

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death syndrome.[5] Children with tracheomalacia display a range of different and often complex co-morbidities (Table 1 shows 63 co-morbidities from the 25 patients in this series), with a strong association between this condition and previous surgery for oesophageal atresia.[6] The majority of these cases present initially to ENT (84 per cent in this series) with stridor and recurrent episodes of oxygen desaturation being the most common presenting features. That acute life-threatening episodes were noted in as many as 72 per cent of patients reflects the potential risks of failing to diagnose and manage this condition in a prompt manner.

Whilst a reduction of greater than half of the sagittal diameter of the trachea during expiration has been suggested as diagnostic for tracheomalacia,[7] there are no specific criteria to determine what classifies tracheomalacia as severe or, as an extension to this, what might be considered a definitive indication for aortopexy. Indeed, the same severity of tracheomalacia can result in a different severity of clinical manifestation. These uncertainties make it difficult to compare and contrast existing case series alongside the data presented here. Indeed, results presented from a large systematic review of aortopexy case series (n = 758) report a considerably lower number of acute life-threatening episodes in the pooled patient group to those presented here (43 per cent *vs* 72 per cent in this series), but this may well reflect differing criteria for surgical intervention in our centre.[4]

- Acute life-threatening events precipitated investigation in 72 per cent of tracheomalacia cases; other symptoms and signs included stridor and recurrent oxygen desaturation
- There is a strong association between tracheomalacia and previous surgery for oesophageal atresia
- Most patients underwent diagnostic microlaryngotracheobronchoscopy: the distal tracheal segment was affected in 60 per cent of cases
- We perform aortopexy through a mini-manubriotomy approach in a combined procedure involving cardiothoracic and ENT surgeons
- Intra-operative bronchoscopy was used to facilitate adjustment of aortopexy sutures as compressive vasculature is suspended from the anterior tracheal wall
- In this series, 84 per cent of patients avoided tracheostomy, with clinical improvement in all these cases

We favour decision-making based on clinical impact and take a multi-disciplinary approach to this process incorporating clinicians from a range of different specialties, including not only cardiothoracic and ENT surgeons, but also radiologists, paediatric cardiologists, paediatric intensivists and paediatric respiratory physicians. The results of these discussions are shared with the patient's family, in a collaborative manner, to facilitate and enable informed consent. In this series, all cases were pre-operatively planned in this manner.

Whilst other treatments have been proposed for tracheomalacia, the relative mortality and morbidity of procedures such as stenting[8] have led most centres to consider aortopexy in the first instance, with other treatments considered if suspension of the compressive vasculature is unsuccessful.[5] The use of internal stenting or a covering tracheostomy is not within our standard practice, and, where possible, we look to perform aortopexy as a stand-alone treatment.

The operative approach chosen when performing aortopexy varies between reported series, largely due to the subjective preference of the surgeons involved in each case.[4] We favour a midline approach using a mini-manubriotomy; this maximises access to both sides of the chest, whilst reducing the necessary patient tilt required when access is via lateral thoracotomy or thoracoscopy.[3] Intra-operative direct visualisation of the tracheal lumen using bronchoscopy allows greater accuracy with the placement and directional pull of the aortopexy sutures, and is performed routinely in our practice.

In this series, the majority of patients avoided tracheostomy (84 per cent), with a clinical improvement noted in all these cases. Improvements were such that 64 per cent have been subsequently discharged from associated services to date. These results sit comfortably alongside those presented elsewhere and support the efficacy of aortopexy for the management of severe tracheomalacia due to vascular compression.[4,9]

Limitations

The results presented here are limited in that they reflect the experience of a single centre and the study is retrospective in nature. Follow-up data could also be affected by a changing patient population, as it is not possible to account for patients who post-operatively may have been initially asymptomatic before developing symptom recurrence and presenting to services elsewhere. This is unlikely, however, given that our institution is a tier one paediatric cardiac surgery centre with a large catchment area covering the northwest of England. Patients would not only have had to relocate a considerable distance away but, given their surgical history, their case would likely be re-discussed with our institution if difficulties were to arise at another National Health Service centre.

As a final point, and to return to the subject of patient selection, aortopexy is something of an indirect solution to tracheomalacia, as it removes only the external compression without treatment to the trachea itself.[3] As such, the results of all series are, to a certain extent, reliant upon the patients chosen within them and their relative suitability to treatment with this method. This study aimed not only to present our post-operative outcomes, but also to draw attention to the need for both rigorous investigation (using both airway endoscopy and CT angiography) and thorough multi-disciplinary pre-operative planning, to ensure candidates for surgery are chosen in the most appropriate manner.

Conclusion

Aortopexy offers an effective method of treating severe tracheomalacia due to vascular compression. It has been suggested that, given the ever-improving survival of premature babies, tracheomalacia will become an increasing phenomenon in clinical practice.[5] As such, the recognition of this condition will become increasingly pertinent to clinicians involved in the care of these patients, particularly those working in paediatric cardiothoracic and ENT surgery. The reportage of case series such as this is therefore of great importance if we are to gain an increased shared understanding of its optimal management.

Competing interests. None declared

References

1 Goyal V, Masters IB, Chang AB. Interventions for primary (intrinsic) tracheomalacia in children. *Cochrane Database Syst Rev* 2012;(10):CD005304

https://doi.org/10.1017/S0022215120000031 Published online by Cambridge University Press

2 Gross RE, Newhauser EBD. Compression of the trachea by an anomalous innominate artery – an operation for its relief. *Am J Dis Child* 1948;**75**:570–4
3 Jennings RW, Hamilton TE, Smithers CJ, Ngerncham M, Feins N, Foker JE. Surgical approaches to aortopexy for severe tracheomalacia. *J Pediatr Surg* 2014;**49**:66–72
4 Torre M, Carlucci M, Speggiorin S, Elliott MJ. Aortopexy for the management of tracheomalacia in children: review of the literature. *Ital J Pediatr* 2012;**38**:62
5 Weber TR, Keller MS, Fiore A. Aortic suspension (aortopexy) for severe tracheomalacia in infants and children. *Am J Surg* 2002;**184**:573–7
6 Corbally MT, Spitz L, Kiely E, Brereton RJ, Drake DP. Aortopexy for tracheomalacia in oesophageal abnormalities. *Eur J Pediatr Surg* 1993;**3**:264–6
7 Wittenborg MH, Gyepes MT, Crocker D. Tracheal dynamics in infants with respiratory distress, stridor and collapsing trachea. *Radiology* 1967;**88**:653–62
8 Vinograd I, Filler RM, Bahoric A. Long-term functional results of prosthetic airway splinting in tracheomalacia and bronchomalacia. *J Pediatr Surg* 1987;**22**:441–2
9 Calkoen EE, Gabra HOS, Roebuck DJ, Kiely E, Elliott MJ. Aortopexy as treatment for tracheo-bronchomalacia in children: an 18-year single-centre experience. *Paediatr Crit Care Med* 2011;**12**:545–51

https://doi.org/10.1017/S0022215120000031 Published online by Cambridge University Press