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Open and thoracoscopic aortopexy for airway malacia in children: 15 year single centre experience

Liam Sutton[a], Elizabeth Maughan[a], Kiersten Pianosi[a], Guled Jama [a], Maral J Rouhani [a,b], Richard Hewitt[a,c], Nagarajan Muthialu[c,d], Colin Butler[a,c,e], Paolo De Coppi[c,e,f,*]

a Department of Otolaryngology, Great Ormond Street Hospital, London, UK
b Lungs for Living Research Centre, UCL Respiratory, University College London, London, UK
c Tracheal Team, Great Ormond Street Hospital, London, UK
d Department of Cardiothoracic Surgery, Great Ormond Street Hospital, London, UK
e Department of Academic Surgery, Institute of Child Health, UCL, London, UK
f Department of Paediatric Surgery, Great Ormond Street Hospital, London, UK

*Senior and Corresponding Author:
Paolo De Coppi – Paolo.DeCoppi@gosh.nhs.uk

First Author:
Liam Sutton liammsutton@gmail.com

Abstract

Objectives:
The objective was to report and analyse the characteristics and results of open aortopexy and thoroscopic aortopexy for the treatment of airway malacia in a paediatric population.

Methods:
We report a retrospective consecutive case series of paediatric patients undergoing aortopexy for the treatment of airway malacia at a quaternary referral centre between December 2006 and January 2021. Outcome measures included days to extubation, continued need for non-invasive ventilation, further intervention in the form of tracheostomy and death.

Results:
169 patients underwent aortopexy: 147 had open procedures (135 via median/limited median sternotomy and 12 thoracotomy) and 22 thoracoscopic. Mean follow up was 8.46 yrs (range 1-20 yrs). Most common site of airway malacia was the trachea (n=106, 62.7%), and 48(28.4%) had additional involvement at the bronchi with tracheobronchomalacia(TBM). 15(8.9%) had bronchomalacia(BM) only. Incidence of bronchial disease was lower in the thoracoscopic than open group (13.6% vs 40.82%; $p<0.0001$). Mean time to extubation was 1.45 days, 2.59 days, 5.23 days in tracheomalacia, TBM and BM groups, respectively ($p=0.0047$). Mean time to extubation was 1.35 days, 2 days, 3.67 days, and 5 days in patients with external vascular compression, TOF/OA, primary airway malacia, and laryngeal reconstruction, respectively ($p=0.0002$). There were 21 deaths across the cohort, and all were in the open group. 71.4% (n=15) had bronchial involvement of their airway malacia.

Conclusions:
Open and thoroscopic aortopexy are effective treatments for airway malacia in children. We have identified that involvement of the bronchi is a risk factor for adverse outcomes, and the optimum treatment for this patient cohort is still debatable.

Keywords: Aortopexy; Tracheomalacia; Tracheopexy; Tracheobronchomalacia; Bronchomalacia

Level of Evidence: IV

Type of Study: Retrospective Study

Introduction:

Tracheomalacia (TM) is a weakness in the tracheal wall causing anteroposterior collapse on expiration or coughing [1]. It presents with a spectrum of clinical severity, not always related to the degree of collapse. Milder forms can resolve spontaneously with growth while more severe life-threatening forms often require surgical intervention. TM can involve the whole trachea or can be confined to one segment only [2]. If both the trachea and mainstem bronchi are involved, the term tracheobronchomalacia (TBM) is used. The term bronchomalacia (BM) is used to describe an isolated weakness of one or both of the main bronchi in the absence of malacia of the trachea, and is a much rarer form of the condition [3].

TM can be as a result of primary airway malacia but is more commonly seen in conjunction with tracheoesophageal fistula/ oesophageal atresia (TOF/OA) or external vascular compression. The patients are often comorbid and therefore can frequently present both anaesthetic and surgical complexities [4].

Patients present with a myriad of symptoms based both the severity and site of the malacia. Presentations can include brassy cough, fixed wheeze, recurrent infections early-onset stridor or even cyanotic and acute life threatening events [4].

Conservative medical treatments that are utilised include anti-muscarinic agents, mucolytics, bronchodilators and antibiotics, as well as chest physiotherapy. However, the evidence base is poor [5]. For severe disease, surgical options include aortopexy or posterior tracheopexy, tracheal resection of short affected segments, internal stents and external airway splinting. Aortopexy was described by Gross in 1947 [6]. From its inception it has been found to be an effective treatment for TM with the efficacy of aortopexy reported as being high in the majority of cases series (greater than 80%) [4]. However, determining which patients should undergo the procedure and the optimal approach for those with bronchial involvement remains contentious.

Many of the existing case series have small patient numbers. As such, we describe our experience with aortopexy over a 15 year period in an attempt to elucidate preoperative and operative factors affecting patient outcomes.

Methods:

This was a retrospective consecutive case series of all aortopexy procedures performed for the management of TM, TBM or BM at Great Ormond Street Hospital for Children, London, between December 2006 and January 2021. All of these patients had been assessed by a multidisciplinary team including a paediatric surgeon, a cardiothoracic surgeon and an otolaryngologist. The site of malacia was classified from direct endoscopy [microlaryngotracheobronchoscopy or flexible bronchoscopy], bronchography or computed tomography into TM, TBM and BM. Malacia was defined as >50% collapse of the airway [5]. Data was sourced from case notes, operative records and intensive care information, and included demographics, comorbidities, ex-prematurity, diagnosis and surgical approach. Outcome measures included days to extubation, further intervention in the form of stenting, tracheostomy, redo aortopexy and death.

Statistical analysis was performed using GraphPad Prism version 9.5.1. Categorical data was analysed using a Chi-square test. Normality could not be assumed in our continuous data sets, therefore Kruskal-Wallis test was used to evaluate time to extubation across groups based on level of malacia and aetiology.

Results:

A total of 169 patients underwent aortopexy; 147 were open procedures (135 via median/ limited upper partial sternotomy and 12 thoracotomy) and 22 were thoracoscopic. Mean follow up was 8.46 years (range 1-20 years). In the open group, 61.9% were male with a median age at time of operation of 232 days (range 7-6982 days). In the thoracoscopic group, 63.6% were male with a median age at time of operation of 157 days (range 63-817 days). 29 (17%) patients also underwent a posterior tracheopexy at the time of their procedure; all of these were performed in the open group.

34.9% (n=59) of the total cohort were ex-premature (born at <37 weeks). There was a non-significant tendency towards a higher proportion of the open group to be born prematurely compared to the thoracoscopic group (36.1% vs 27.3%; p=0.42).

The most common site of airway malacia across the total cohort was at the trachea (n=106, 62.7%); 48 patients (28.4%) had additional involvement at the bronchi with TBM (Figure 1A). 15 patients (8.9%) had BM only. The incidence of bronchial disease was lower in the thoracoscopic group than the open group (Figure 1B: 13.6% vs 40.82%; p<0.0001).

The most common indication for surgery across the total cohort was external vascular compression (Figure 2: n=116; 68.6%) followed by TOF/OA repair (n=59; 34.9%), previous laryngeal reconstruction (n=12; 7.1%) and primary airway malacia (n=6; 3.5%). 24 patients had more than one indication for surgery. The most common indication for surgery between the open and thoracoscopic groups differed with external vascular compression representing 73% of the open group and tracheoesophageal fistula repair representing 77% of the thoracoscopic group.

149/159 patients (93.7%) were successfully extubated in our unit. The mean time to extubation post-operatively was 2.17 days (s.d $\pm$ 3.71) Of the remaining 10 patients, eight patients were discharged to their local hospital intubated, for whom we do not have time to extubation data, and two patients had withdrawal of care and therefore were not extubated.

The mean time to extubation post-operatively was 1.45 days in the tracheomalacia group, 2.59 days in the TBM group and 5.23 days in the bronchomalacia group (p=0.0047).

The mean time to extubation post-operatively was 1.35 days in patients with external vascular compression, 2 days in patients undergoing tracheo-oesophageal fistula repair, 3.67 days in patients with primary airway malacia and 5 days in those undergoing laryngeal reconstruction (p=0.0002).

10 patients (5.9%) had a tracheostomy at the time of surgery; all of these patients then underwent open aortopexy. Of these, nine patients were decannulated successfully post-operatively (90%); one patient remains with a tracheostomy at last follow up and is noted to have TBM. Of the 159 patients who did not have a pre-operative tracheostomy, eight patients required tracheostomy insertion following their initial procedure. 62.5% (n=5) of these patients had bronchial involvement of their airway malacia.

There were 21 deaths across the cohort at last follow up; all were in the open group with none in the thoracoscopic group. 71.4% (n=15) had bronchial involvement of their airway malacia; nine had TBM whilst six had bronchomalacia only. In terms of indication for surgery, external vascular compression represented 61.9% (n=13) of these deaths. There were no deaths in patients undergoing aortopexy for primary airway malacia.

The complication rates for thoracoscopic aortopexy and open aortopexy were 18.2% and 25.8 % respectively with pneumothorax being the most common complication. Mean operative time was longer in the open group as was length of post operative stay and time to extubation (Table 1). No patients in the thoracoscopic group however underwent concurrent repair of cardiac lesions or vascular rings which may account for the shorter times. Reintervention rates in the form of tracheostomy, stenting or redo pexy procedure were comparable between the two groups (13.6% thoracoscopic and 10.9% open).

Discussion:

Whilst the incidence of TM is estimated to be 1 in 2,100 in the European population [7], the management of the condition is still contentious [5]. Although there is no unified consensus on the classification of TM, the European Respiratory Society task force define primary TM as those conditions with an intrinsic abnormality of airway cartilage, and secondary TM as those conditions where the cartilage was embryologically normal but developed abnormally due to external pressure [5]. In a systematic review of 758 patients with TM, primary malacia was identified in 9% of patients in contrast to 3.5% in our study [4]. We had a much higher rate secondary to external vascular compression (68.6%) than that of this review (34%). This may represent the referral pathways we have in place, with patients with aberrant vasculature being more commonly managed at our quaternary centre, whereas those with normal vascular anatomy being more likely to be managed locally. We also had a lower rate of post TOF/OA repair (34.9%) in our aortopexy population than that of the review (44%); again, this is likely related to referral bias. Open surgery was performed using a (limited) upper partial sternotomy in the majority of cases (n=135). We found that this approach allows for good exposure with comparable access to the vessels over both the right and left main stem bronchi as well as the trachea and a more specific elevation of the arteries while allowing for cardiovascular surgery to be performed at the same operation [8]. Thoracoscopy approach at our institution includes a 3-port 3mm technique from the left side of the patient who is positioned supine [9]. Briefly, after removing or mobilising part of the left lobe of the thymus, the pericardium is opened and 3 transsternal non-absorbable sutures are passed through the adventitia of the aorta which is mobilised anteriorly [10].

There have been no randomised controlled trials comparing surgical versus non-surgical management of malacia [11] nor have there been studies comparing the outcomes of open versus thoracoscopic surgery. We found that there was both a shorter time to extubation and a lower mortality rate in those who underwent thoracoscopic surgery. However, the patient populations and indications for surgery differed between the open and thoracoscopic groups. Open aortopexy was the approach of choice for those patients requiring concurrent repair of aberrant vasculature or cardiac lesions, providing excellent access to the mediastinal structures. The thoracoscopic group had a lower rate of prematurity, a lower rate of bronchial involvement of disease, and had no patients with preoperative tracheostomy. On the contrary, the thoracoscopic group had a higher rate of surgery for the indication of TOF/OA and short segment malacia when compared to open. While the results from thoracoscopic surgery were positive, conclusive data when compared to open surgery could not be made due to the heterogeneity of the two groups. While the procedure was only performed thoracoscopically in 22 cases, 77.3% of these were for the indication of TOF/OA with good outcomes. Further research is required to ascertain for which specific patient population this is the optimal approach.

While aortopexy is an accepted treatment for tracheomalacia [12] the management of bronchial involvement is less clear. In our series, mortality rate in those with isolated tracheal disease was 4.7% whereas mortality rate was 22.8% in those with bronchial involvement. Time to extubation in those with bronchial disease was also longer, with mean number of days to extubation of 3.69 days compared to 1.45 days in those with malacia limited to the trachea. 10.4% of those with malacia affecting their bronchi required a tracheostomy postoperatively compared to 2.8% of those without bronchial disease. Those with bronchial disease also had a higher rate of concurrent posterior tracheopexy (22.2%) compared to those without (14.2%). Posterior tracheopexy was also more commonly used in those with TOF/OA (41.4%), with the rationale that it provides support to the posterior wall, than any other indication (33.6%). This patient cohort size was not large enough to establish whether combining posterior tracheopexy with aortopexy conferred a benefit to those with malacia of the bronchi.

Following surgery, we achieved successful tracheostomy decannulation in 90% of patients with a preoperative tracheostomy. Decannulation rates vary in the literature with one small study from a UK paediatric centre reporting post-aortopexy tracheostomy rates of 6.25% [13]; a further UK study with a sample size of 25 patients reported 16% [14], and in a small-volume USA case series post-aortopexy tracheostomy rates were 40% [15]. Eight patients in our case series required a postoperative tracheostomy with one patient remaining with their preoperative tracheostomy, bringing the total postoperative tracheostomy rate to 5.33%.

Interestingly, we found that the shortest mean time to extubation was in the patients with external vascular compression. Unsurprisingly, aortopexy required for laryngeal cleft surgery had the longest length of intubation as five of the nine surgeries were performed for grade 3 or 4 laryngeal cleft surgery and standard practice is for these patients to remain intubated for five days following the laryngeal repair.

Limitations:

While including a large number of total patients we acknowledge that the study is retrospective in design and single centre. Careful patient selection was required for each of the intervention groups. Significantly different patient numbers and heterogeneity between the two groups, made statistical comparisons difficult.

Conclusions:

Our case series demonstrates that aortopexy is a safe and effective treatment for large airway malacia. We have identified that involvement of the bronchi is a risk factor for adverse outcomes and the optimal treatment for this patient cohort is still debatable. Thoracoscopic approach has excellent outcomes for a select patient group and should be considered in those with favourable anatomy. Randomised controlled trials comparing the open to thoracoscopic approach in this patient population may help delineate which children would benefit most from a minimally invasive approach.

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Figure Legend:

Figure 1: A; Site of airway malacia in the total, open and thoracoscopic groups. B; Bronchial involvement per group. **** denotes $p<0.0001$

Figure 2: Indication for surgery in the total, open and thoracoscopic groups

Table 1: Comparison of open and thoracoscopic aortopexy

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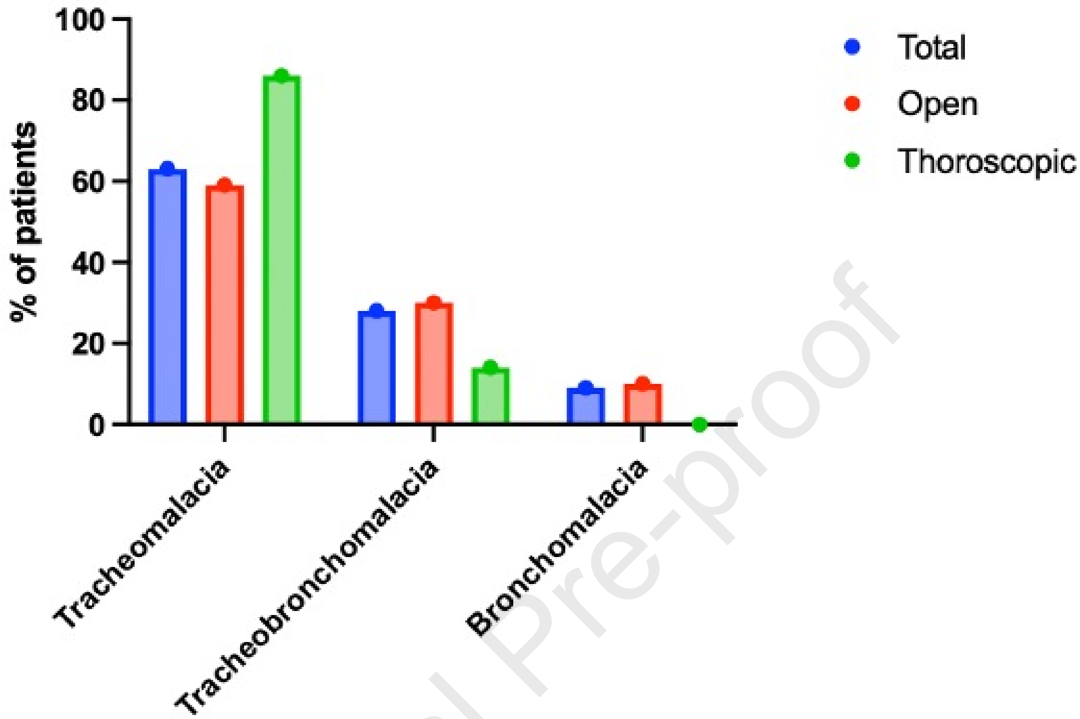
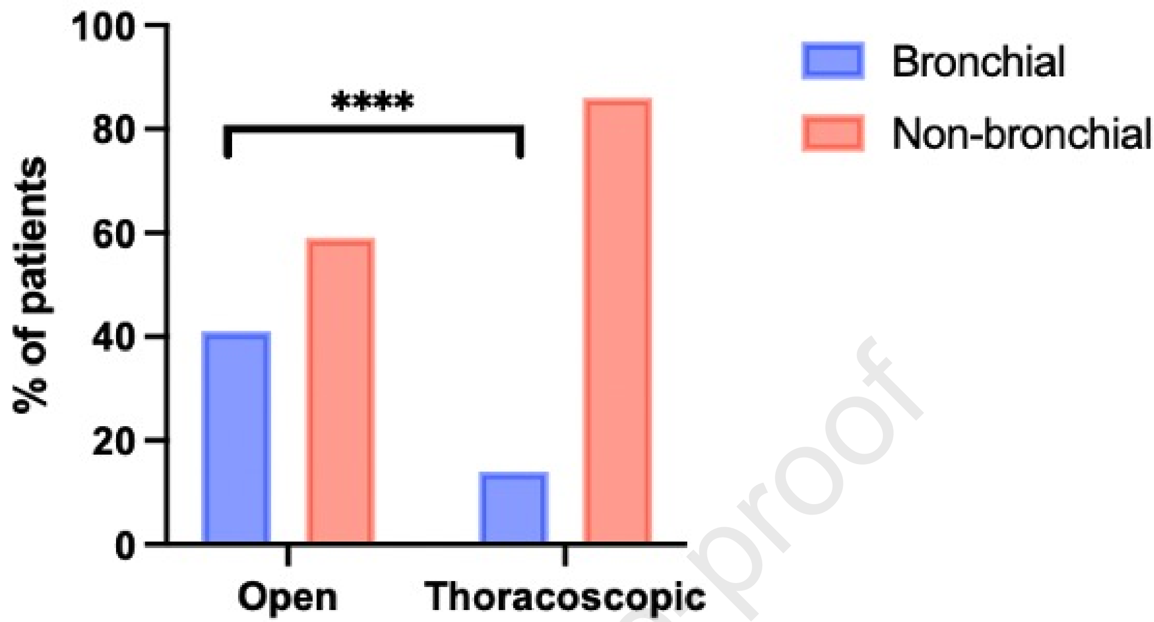
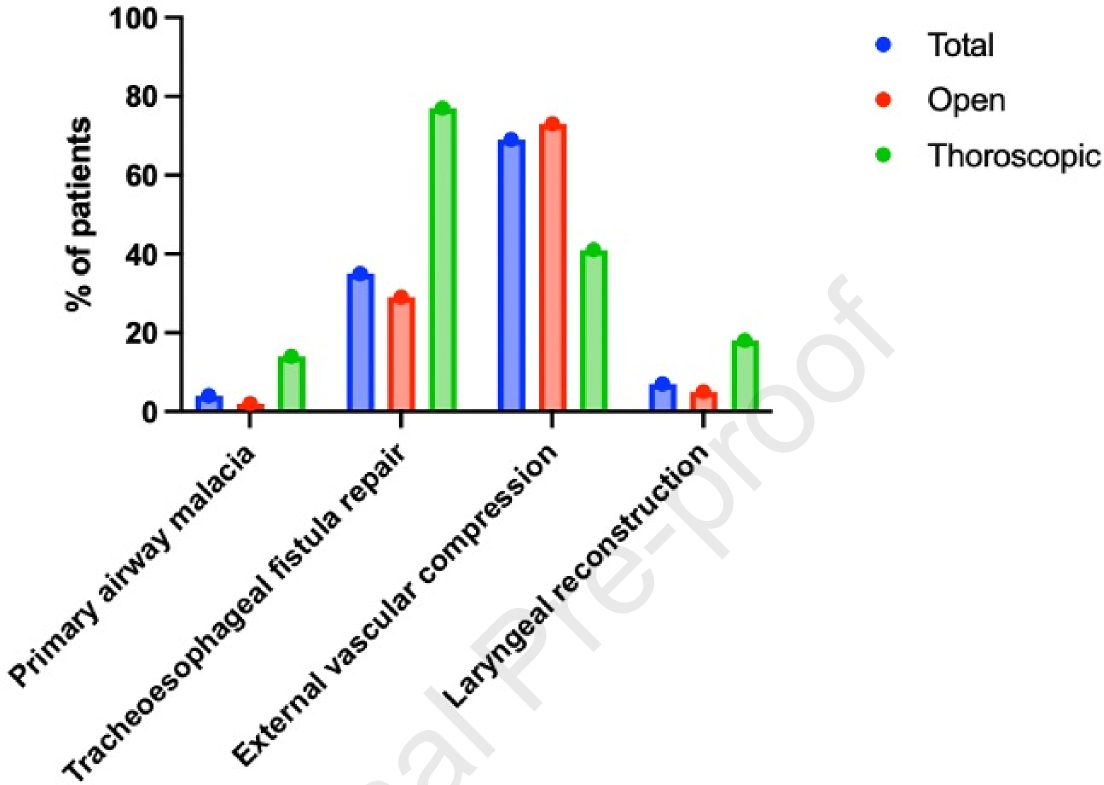
TABLE 1

	Thorocoscopic	Open
Complication rate	4/22 (18.2%)	38/147 (25.8%)
Operative time (mins)*	110 (40-180)	144 (42-280)
Length of postoperative stay (days)*	26 (1-139)	44 (1-363)
Time to extubation (days)*	1.41 (0-7)	2.29 (0-21)
Reintervention	3/22 (13.6%)	16/147 (10.9%)

* Median (range)


100
80
60
40
20
0
% of patients
Tracheomalacia
Tracheobronchomalacia
Bronchomalacia
Total
Open
Thoroscopic


100
80
60
40
20
0
% of patients
****
Open
Thoracoscopic
Bronchial
Non-bronchial



100
80
60
40
20
0
% of patients
Primary airway malacia
Tracheoesophageal fistula repair
External vascular compression
Laryngeal reconstruction
Total
Open
Thoroscopic

Highlights:

- Aortopexy is an effective treatment for large airway malacia in a paediatric population.
- Bronchial involvement is a risk factor for prolonged intubation, tracheostomy and mortality.
- Thoracoscopic approach has excellent outcomes for select patients and has a role for those with favourable anatomy.
- Optimum treatment for bronchial involvement requires further studies.