This is a peer-reviewed, accepted author manuscript of the following research article: Williamson, A., Young, D., & Clement, W. A. (2022). Paediatric tracheobronchomalacia: incidence, patient characteristics, and predictors of surgical intervention. *Journal of Pediatric Surgery*. https://doi.org/10.1016/j.jpedsurg.2022.05.005

Paediatric tracheobronchomalacia: Incidence, patient characteristics and predictors of surgical intervention

Abstract

Objectives: Tracheobronchomalacia (TBM), a condition where an abnormality of the tracheal walls causes collapse during the respiratory cycle, is a common cause of airway obstruction in childhood. TBM can present with a large spectrum of disease severity and underlying pathologies that may be managed medically and surgically, and it is not always clear which patients would most benefit from surgical intervention. We aim to describe the incidence, patient characteristics, and predictors of surgical intervention in a large cohort of paediatric patients.

Methods: We performed a retrospective review of all children diagnosed with TBM to a paediatric Otolaryngology unit in the west of Scotland between 2010-2020. Odds ratios for clinical predictors of surgery were calculated using logistic regression with uni- and multivariate analysis.

Results: 249 patients were identified of which 219 proceeded to data collection. Primary malacia was noted in 161 (73.5%) and secondary in 58 (26.5%). Causes of secondary malacia included compression by the innominate artery (11%) and vascular rings (7.8%). Surgical interventions were performed in 28 patients (12.8%) including division of vascular ring, aortopexy, and surgical tracheostomy. Multivariate analysis showed secondary TBM, acute life-threatening events, and difficulty weaning from mechanical ventilation were independent risk factors for surgical intervention.

Conclusions: TBM can present with a myriad of airway symptoms and is frequently associated with other airway and mediastinal pathologies necessitating multiple interventions. Children aged <1 year present with a more severe form of the disease and the presence of particular independent risk factors may indicate a need for surgical intervention.

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Keywords: Tracheomalacia; Tracheobronchomalacia; Bronchomalacia; Tracheal diseases; Vascular ring; Airway disorders

1. Introduction

Tracheobronchomalacia (TBM) is a the most common congenital tracheal pathology, where inward collapse of the tracheal cartilages and the posterior membranous wall results in a dynamic airway narrowing[1]. TBM is often described as either primary "congenital" malacia, due to an inherent abnormality of the tracheal cartilages and trachealis, or secondary "acquired" malacia resulting from external compression by mediastinal structures[2]. TBM is thought to be the most common cause of dynamic central airway collapse in children, with the primary malacia estimated to occur in 1/2,100 births[3]. The need for invasive investigations may preclude diagnosis in many children; however, its high prevalence on routine bronchoscopy suggests the true incidence may be higher[4,5].

TBM can be a diagnostic challenge, as it is a poorly defined clinical entity that is often mistaken for more common respiratory conditions. Dynamic airway assessment using flexible or rigid bronchoscopy is frequently employed to ascertain the presence of airway collapse, extraluminal compression and other culprit airway pathologies[1]. Patients with perceived external compression typically require CT or MRI imaging to confirm the presence of abnormal anatomy or masses in the mediastinum[6,7]. TBM may be treated with clinical observation and management of associated comorbidities, or through non-invasive ventilation with CPAP and BiPAP[8,9]. Surgical interventions for TBM are varied, and are often dependant on the underlying diagnosis.

Interventions can include division of a compressive vascular ring, reduction of tracheal compression through aortopexy, tracheopexy or tracheal splints and stents, and tracheostomy with or without long term ventilatory support[8,9].

The degree of malacia can be influenced by numerous factors, including the site and level of the malacic segment, rate of airflow, the biomechanical properties of tracheal cartilage, the ratio of cartilage to trachealis muscle and the presence of secondary airway pathologies[10,11]. This generates a vast spectrum of disease severity, and it can therefore often be unclear which patients may be managed conservatively and which would benefit from surgical intervention. Previous respiratory case series have largely focused on primary disease in isolation or contain few surgical patients[3,12], making it difficult to draw meaningful conclusions on the indications for surgery. Likewise, many surgical case series give a detailed overview of the indications for surgery [13–15]; but do not usually contain patients managed non-surgically. Due to the narrower focus of these past studies, it cannot be fully established how surgical patients differ clinically from the wider TBM population.

We have performed a 10-year retrospective review of all patients presenting with TBM to a tertiary referral Paediatric Otolaryngology team in Scotland. The aim of this series is to more accurately describe the patient demographics, presenting symptoms, and underlying causes of TBM and better understand their relationship to surgical management. We also aim to identify which clinical factors may predict surgical intervention, and therefore assist clinical decision making in these frequently complex patients.

2. Methods

2.1 Patient cohort & incidence

In this retrospective study, case note review was performed on all patients aged 0-16 years diagnosed with TBM on rigid bronchoscopy between 2010-2020. Patients were identified using theatre logbooks. No specific exclusion criteria were applied. Data was collected on demographics, comorbidities, prematurity of <38 weeks, previous intubations, critical care admissions, investigations, and relevant surgical procedures. In addition, information on common presenting symptoms was gathered, including stridor, acute life-threatening events (ALTEs), respiratory tract

infections (RTIs), feeding difficulties, persistent cough, dyspnoea, sleep disordered breathing, and difficulty weaning from mechanical ventilation.

Data was collected from the Yorkhill Royal Hospital for Sick Children and the Royal Hospital for Children (Glasgow, Scotland) between November 2010 to November 2020. Both hospitals are the tertiary referral centres for complex paediatric airway pathologies in Scotland (population 5.4 million) and performed an average of 387 microlaryngoscopy with rigid bronchoscopy per anum between 2011-2019. An estimated incidence was calculated using the number of cases identified per year and the mean annual birth rate of Scotland[16].

2.2 Definitions

Primary TBM was described as malacia on bronchoscopy with no cause found. Patients with oesophageal atresia and tracheoesophageal fistula (OA/ TOF) were included in this group as this is commonly termed primary malacia in previous studies[8]. Secondary TBM encompassed malacia secondary to compression by aberrant vascular anatomy (including vascular rings and innominate artery compression) or other mediastinal structures. Previous suprastomal and intrathoracic malacia due to long-term tracheostomy and ventilation was also included in this group.

“Stridor” encompasses both mono and biphasic stridor. “Feeding difficulties” was defined as reduced oral intake, coughing or choking on solids and liquids, and need for supplemental enteral feeding via nasogastric tube or gastrostomy. “Difficulty weaning from mechanical ventilation” was defined as 1 or more failed attempts at extubation or at least one episode of prolonged ventilatory weaning. “Neonatal/ paediatric intensive care (NICU/ PICU) admissions” was defined as any admission of >24 hours to NICU, PICU and special care baby units with or without intubation. “Previous intubations” were defined as one or more intubations either for a surgical case, airway or ventilatory emergency or as part of a NICU/ PICU admission. Finally, ALTEs incorporates a range of acute respiratory events requiring either hospital attendance or intubation. This includes one or

more severe episodes including apnoea, “dying spells”, cyanosis, or instances of increased work of breathing.

2.3 Statistical analysis

Variables including patient age, primary and secondary disease, cardiovascular disease, presenting symptoms (recurrent RTIs, feeding difficulties, persistent cough, dyspnoea, stridor, ALTEs), and past medical history (difficulty weaning from mechanical ventilation, prematurity, previous critical care admissions and intubations) were analysed using logistic regression. Patients with TBM secondary to tracheostomy and those lacking sufficient clinical information were excluded. Univariate analysis was first performed for multiple variables to determine if they were associated with a higher rate of surgery. Variables which were significant predictors ($p<0.05$) were used in a multivariate model to determine which were independent predictors of surgery. All analyses were done using Minitab (version 18) at a 5% significance level and stepwise selection was done to construct the multivariate model.

2.4 Bronchoscopy

Rigid bronchoscopies were performed by a consultant paediatric otolaryngologist or a supervised trainee registrar as part of the patient’s formal investigations for airway symptoms. Rigid bronchoscopy is the preferred technique in our centre as we have extensive experience in diagnosing TBM with this technique, and use of rigid endoscopy under anaesthetic facilitates evaluation and surgical management of secondary upper airway pathologies such as laryngomalacia. Bronchoscopies used an age-appropriate rigid ventilating bronchoscope (Karl Storz, Tuttlingen, Germany) and 0-degree Hopkin’s endoscope. All procedures were conducted without endotracheal intubation under general anaesthetic using inhalational or total intravenous anaesthesia. Neuromuscular blockade was not used in order to facilitate spontaneous respirations and provide a dynamic assessment of airway collapse. The vocal cords and carina were sprayed with topical anaesthesia prior to insertion of the bronchoscope.

Diagnosis of TBM was made on the basis of anterior or posterior flattening of tracheal or primary bronchi lumen or significant airway collapse during expiration, at the discretion of the consultant otolaryngologist. Diagnostic criteria such as >50% obstruction of tracheal lumen[17–19] 2:1 ratio of membranous to cartilaginous wall[17–19], were not used as they were deemed impractical for diagnostic purposes.

2.5 Ethics

In accordance with the United Kingdom 2018 data protection act, approval was sought from the local Caldicott Guardian before accessing patient's electronic medical records. Due to the retrospective design with no intervention cohort, patient consent and approval from the research ethics committee was not sought.

3. Results

3.1 Demographics and incidence

Between 2010-2020, 249 patients were diagnosed with airway malacia on rigid bronchoscopy. Indications for rigid bronchoscopy included ALTE episodes, recurrent respiratory tract infections including croup, failure to wean from ventilation, difficult perioperative intubations, biphasic stridor, and recurrent stridor in the presence of failure to thrive, cutaneous haemangiomas, and prior intubations. Thirty patients were removed insufficient clinical information (figure 1). There were 151 males and 68 females (ratio 2.2:1). Median age at presentation was 9.6 months (0-11.6 years), and at diagnosis was 12 months (range 0-11.8 years). Over half of all male (n=79, 52.3%) and female (n=34, 50%) patients presented at <1 year of age. Patient demographic data is summarised in table 1 and figure 2**.**

The mean annual birth rate in Scotland between 2010-2019 was 55,177 children per anum. By assuming a detection rate of primary and secondary malacia of 25 cases per year (249 cases over 10 years), a conservative estimate of all TBM cases in Scotland is 1 in every 2,200 new-borns.

3.2 Diagnosis

There were 151 (68.9%) cases of tracheomalacia, 56 (25.6%) tracheobronchomalacia, and 12 (5.5%) bronchomalacia. Primary malacia was noted in 161 patients (73.5%) and this was associated with cardiac anomalies in 57 (26%). Secondary TBM was demonstrated in 58 patients (26.5%), of which 11 (5%) had associated cardiac abnormalities. Nineteen (8.6%) patients with primary malacia had a history of OA/TOF.

Patient comorbidities, congenital abnormalities, and airway pathologies are summarised in table 1. The most frequently reported comorbidities were Gastro-oesophogeal reflux (GORD) diagnosed clinically or on pH impedence studies (n=52, 23.7%), developmental delay (n=21, 9.6%), and bronchopulmonary dysplasia (n=17, 7.8%). Trisomy 21 was the most common syndromic disease (n=13, 5.9%), followed by VACTERL (n=7, 3.2%) and DiGeorge syndrome (n=4, 1.8%). Laryngomalacia was the most common synchronous airway pathology in 45 patients (20.5%). Subglottic stenosis (n=25, 11.4%) and vocal cord palsy (n=7, 3.2%) were also frequently documented.

Cross sectional imaging was implemented in 120 patients (85 CT, 41 MRI). Presence of a vascular ring, innominate artery compression, or other secondary cause was noted in 41.7% of scans (CT 44.0%, MRI 31.7%). Causes of secondary TBM are listed in table 2. Innominate artery compression (n=24, 11.0%), vascular rings (n=17, 7.8%), and previous insertion of tracheostomy (n=8, 3.7%) were the most common secondary aetiologies.

3.3 Presenting symptoms

The main presenting features were stridor (n=110, 50.2%), ALTEs (n=51, 23.3%) and recurrent RTIs (n=56 25.6%). Children aged <1 year presented with more severe symptoms compared to older children, including stridor (72.1 vs 30.7%), ALTEs (35.2 vs 5.3%), and difficulty weaning from mechanical ventilation (15.6 vs 4%). Conversely, children aged >1 year more often presented with milder non-specific symptoms such as recurrent RTIs (19.7 vs 58.7%) and persistent cough (8.2 vs 22.7%). History of previous intubation (n=100, 45.7%), NICU/ PICU unit admissions

(n=116, 53.0%), and prematurity of <38 weeks (n=70, 32.0%) were common in all children. These features were more prominent in children aged <1year and in those with secondary TBM. The clinical features are summarised table 3.

3.4 Surgical interventions

Twenty-eight (12.8%) patients underwent a surgical intervention (table 4). Aortopexy was performed in 10 (4.6%) patients with a mean age of 2.2months (range 0-6months). The indications for aortopexy were primary malacia (n=3), previous division of vascular ring (n=3), innominate artery compression (n=2), and compression by complex congenital cardiac abnormalities (n=2).

Twelve (5.5%) patients underwent division of vascular rings; of which 5 had double aortic arch and 7 right aortic arch with aberrant subclavian or left ligamentum arteriosum (RAA). Mean age of patient at intervention was 15.5 months (range 0 months-6.2 years). Three patients with atypical rings and 1 with RAA and no significant evidence of compression were managed with clinical observation, and 1 patient with pulmonary artery sling is awaiting surgical intervention.

Surgical tracheostomy was performed in 11 patients (5.1%), either to facilitate long term ventilation or reduce dead space within the upper airway. Mean age at insertion was 4.1 months (range 0-18 months). Five patients underwent >1 surgical intervention, including two who had division of vascular ring followed by aortopexy, and one child who experienced division of vascular ring followed by aortopexy then surgical tracheostomy.

A further 120 airway interventions were performed in 96 patients (43.8%) at the time of or after rigid bronchoscopy. The most frequent of these included adenoidectomy +/- tonsillectomy (n=70) and supraglottoplasty (n=36).

3.5 Follow-up & outcomes

164 patients were followed up in our ENT department for a mean duration of 25.8 months, with remaining patients followed up in local centres. At time of most recent follow-up, 124 (75.6%) had reported complete resolution of airway symptoms, including 9/20 (45%) patients managed surgically, and 115/144 (79.9%) patients managed conservatively. Nine deaths occurred at a mean age of 3.7 years (range 4 months-17.5 years). All patients had complex past medical history with recurrent admissions to hospital and intensive care units. The most frequent causes of death were heart failure secondary to complex cardiac abnormalities (n=3) and multi-organ failure due to recurrent RTIs (n=2). No deaths were reported to be directly related to episodes of TBM crisis or surgical interventions.

3.6 Clinical predictors of surgical intervention

197 patients were included in the statistical analysis (tables 5 & 6). Univariate analysis demonstrated children with secondary malacia (OR=4.4176), ALTEs (OR=3.7212), difficulty weaning from mechanical ventilation (OR=5.3125), prematurity of <38 weeks (OR=2.7556), previous intubation (OR=3.8352), and previous NICU/ PICU admission (OR=3.8514) were significantly more likely to undergo surgical intervention. Those with recurrent RTIs (OR=0.2734) and persistent cough (OR=0.1586) had a significantly reduced rate of surgical intervention. Multivariate analysis identified three independent predictors of surgical intervention; secondary TBM, difficulty weaning from mechanical ventilation, and ALTEs.

4. Discussion

4.1 Summary of main results

Tracheobronchomalacia is an endoscopic diagnosis and selecting appropriate patients for surgical intervention based on clinical symptoms can be difficult. This paper is one of the largest case series of primary and secondary paediatric TBM published. We have therefore provided a more

extensive portrayal of the TBM population in children, and have highlighted several clinical features that may prompt clinicians to consider further investigations and surgery.

Our study reaffirms many known demographic features, including a significant male preponderance (151:68), and a high incidence of children aged <1year. Prior series have established males can be more prone to malacic disease, and can present at a younger age[12,20]. Why male children have a greater incidence of TBM and how this influences long term development remains unclear. The estimated incidence of 1/2,200 children is similar to the 1/2,100 reported in the Netherlands; however, it should be noted the latter only described primary disease, suggesting a possible under-estimation in our cohort.

Our patients had a high rate of GORD, which has been established as more frequent and severe in TBM patients compared to healthy controls[21]. Clinicians should therefore consider the potential need for anti-reflux medications in children with TBM. Children in this study also demonstrated high indices of prematurity, previous intubations, NICU/PICU admissions, OA/ TOF, cardiac abnormalities, and syndromic disease, all of which prominently feature in series containing children with moderate to severe TBM[22–24].

The main presenting symptoms of our series were stridor, ALTEs, and recurrent RTIs. There appears to be a variance in symptom severity in the literature, with some cases reporting milder presentations such as cough and recurrent RTIs [3,25], whilst others most frequently describe ALTEs[17,26]. Numerous reviews and case series have defined the natural history of primary TBM as one of gradual improvement with age[1,8,27]. In our cohort, children aged <1year presented with more severe symptoms such as ALTEs and exhibited a higher rate of prematurity, previous intubations, and NICU/PICU admissions. This illustrates a frequently challenging group of neonatal patients with severe comorbidities necessitating broad multidisciplinary input. Symptoms became milder in children aged >1 year, who more prominently demonstrated persistent cough and recurrent RTIs. This group can also prove to be a clinical challenge, as it contains a heterogenous cohort of infants

with multi-level airway obstruction, and older children whose symptoms may be mistaken for other lower airway disorders. Nevertheless, the lower incidence of severe symptoms may reassure the parent and patient that surgical intervention is seldom required in older children.

We noted high rates of secondary airway pathologies; in particular laryngomalacia. Resultantly, a large number of patients underwent additional airway interventions. The presence of synchronous airway lesions is documented in other endoscopic series. One report of 100 bronchoscopies in children with airway obstruction noted two or more synchronous lesions in 17.5% of cases[28], whilst another found a 47% rate of TBM in patients with severe laryngomalacia[29]. Clinicians must therefore be mindful that airway symptoms in children may be generated by multilevel pathology and the primary culprit abnormality may evolve or change as the child grows.

We did not comment on the level or extent of TBM on bronchoscopy. The reasons for this are threefold. Firstly, there are no widely accepted endoscopic diagnostic criteria for TBM, and classifications such as that proposed by the ERS task force, that grades malacia as >50% collapse of the tracheal lumen, are impractical in a diagnostic and clinical context[18]. Secondly, the appearances on endoscopy can be altered by procedural influences such as rate of airflow, depth of anaesthesia, and use of rigid or flexible bronchoscopes. *Hysinger et al* noted a discordance between tracheomalacia appearances on flexible and rigid bronchoscopy, finding a moderate correlation in the distal and poor correlation within the mid trachea, suggesting the mode of endoscopy may influence diagnosis[29]. *Okazaki et al* demonstrated in anaesthetised, paralysed children, the crosssectional area of the trachea is dependent on the airway pressure[30]. By calculating the closing pressures, they confirmed that malacic crises do not occur during normal breathing, but during episodes of elevated airway pressure such as crying and coughing. Although the children in our cohort were not paralysed, it could be concluded bronchoscopy may underestimate the degree of malacia by examining the patient in a relaxed state with low airway pressures. Thirdly, the severity of TBM on endoscopic examination correlates poorly with clinical presentation. This is reinforced in a

study comparing bronchoscopic videos with intra-tracheal pressure measurements in adult patients. The authors noted that patients with TBM had greater airflow limitation compared to healthy controls (p=0.009), but the degree of airflow limitation associated poorly with the severity of tracheal collapse[31]. It could be concluded that severe malacia on endoscopy does not necessarily indicate a worse clinical presentation. Therefore, commenting on endoscopic appearances and their relationship with surgical intervention was not deemed reliable enough to draw useful conclusions for this study.

Imaging techniques such as multi-detector CT and ultrashort echo-time MRI have been described as primary investigation modalities for TBM, but at present require a general anaesthetic or are not available outside of research institutes respectively[7,32]. CT is more frequently used as a second line investigation to assess the surrounding vessels and lung parenchyma. We found a higher than anticipated rate of scans demonstrating a secondary cause of TBM (41.7%). However, the rates of surgically appropriate disease such as vascular rings were only present 14.3% of scans. Innominate artery compression was noted on 24 scans, but only three required aortopexy due to recurrent severe ALTE's requiring intubation and ventilation. Clinicians thus need to account for not only the endoscopic appearances, but also of the severity of symptoms when investigating possible secondary causes, so as to avoid an unnecessary general anaesthetic or dose of ionising radiation.

Finally, our study is somewhat unique as univariate analysis has identified a number of clinical predictors that may indicate the likelihood of surgical intervention, including history of previous intubation, prematurity, and NICU/PICU admission. Moreover, multivariate analysis found that secondary malacia, difficulty weaning from mechanical ventilation, and ALTEs were all significant independent predictors of surgical intervention.

These features are reflected in a prior literature review of aortopexy, which noted that oesophageal atresia, ALTEs, and vascular compression were the most frequent indications for intervention[33]. Several papers have investigated predictors of successful outcome following TBM

surgery. This includes our unit's experience with 30 patients, which did not demonstrate any obvious predictors of positive post-operative outcome[23]. Others have identified varying predictors of postoperative mortality, including cardiovascular disease, pre-operative ventilation requirement, and bronchomalacia[34,35]. Combined with our own findings, children with more severe pre-operative comorbidities are more likely to undergo surgical intervention, and suffer a turbulent post-operative recovery. Children aged >1year were less likely to recieve surgery, but this did not achieve statistical significance (OR=0.4951, p=0.115). It is unclear if this reflects on the initial severity of the malacia, or natural improvement with growth. Nonetheless, symptoms seen in older children including persistent cough and recurrent RTIs were significant negative predictors of surgery, implying the majority of children aged >1year with these symptoms may be managed conservatively.

4.3 Limitations

The primary limitation of this series is its retrospective design. Additionally, presence of TBM was confirmed by the assessment of a consultant paediatric otolaryngologist and was not based on published criteria and resultantly there may be some heterogeneity in TBM diagnosis. Moreover, diagnosis was made via rigid bronchoscopy, which some authors feel can underestimate the degree of TBM due to splinting of the airway[18,29].

There are a number of sources of potential bias in this paper. Firstly, by only including children who have undergone bronchoscopy, we introduce an element of selection bias by not considering those who may have TBM but did not progress to bronchoscopy. Secondly, there is a degree of attrition throughout our cohort, with patients not progressing from initial identification to data collection and multivariate analysis, due to insufficient clinical information or follow-up occurring out with our centre.

Finally, our unit is a tertiary referral airway service, receiving complex paediatric patients from the entirety of Scotland. However, a small number of patients may be diagnosed in other centres, meaning there may be an underestimation the overall Scottish Paediatric TBM population.

4.4 Conclusions

In conclusion, TBM presents with a wide spectrum of severity and is associated with a high incidence of secondary cardiorespiratory and airway comorbidities. Older children may present with more insidious symptoms that may be mistaken for other respiratory diseases, and clinicians should therefore be mindful of the presence of TBM in those who present with otherwise unexplainable airway symptoms. Infants aged <1 year old have a greater predilection for more severe malacic symptoms, and the presence of secondary malacia, difficulty weaning from mechanical ventilation, and ALTEs should alert clinicians to the possible need for investigation and future surgical intervention.

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Paediatric tracheobronchomalacia: incidence, patient characteristics, and predictors of surgical intervention.

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Paediatric tracheobronchomalacia: incidence, patient characteristics, and predictors of surgical intervention.

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Figure 1- flow diagram summarising the investigations and subsequent diagnosis of all patients included in the study

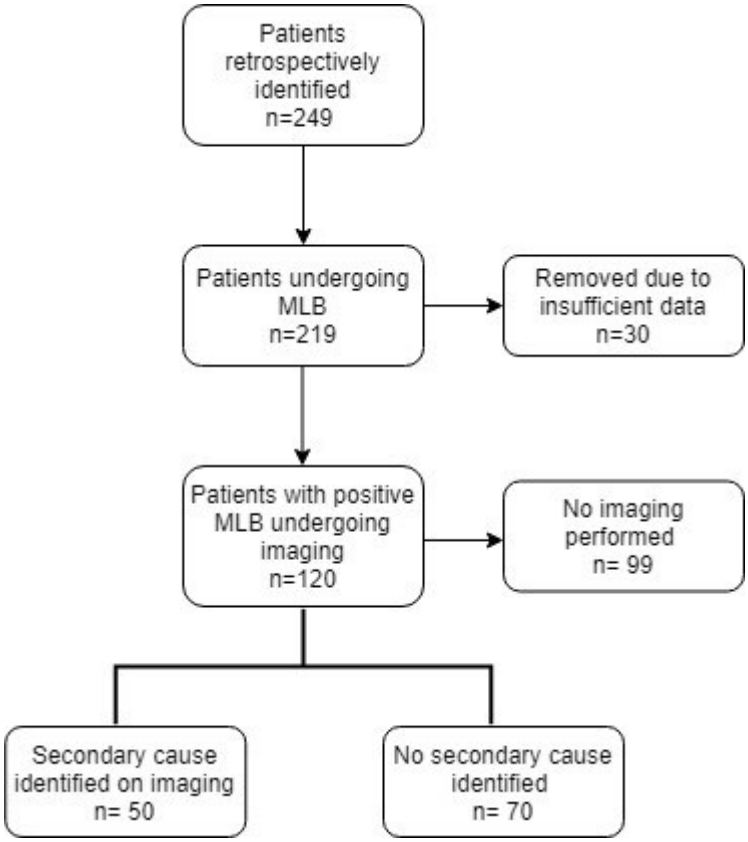

Figure 2- age at presentation of TBM, subdivided into patient gender

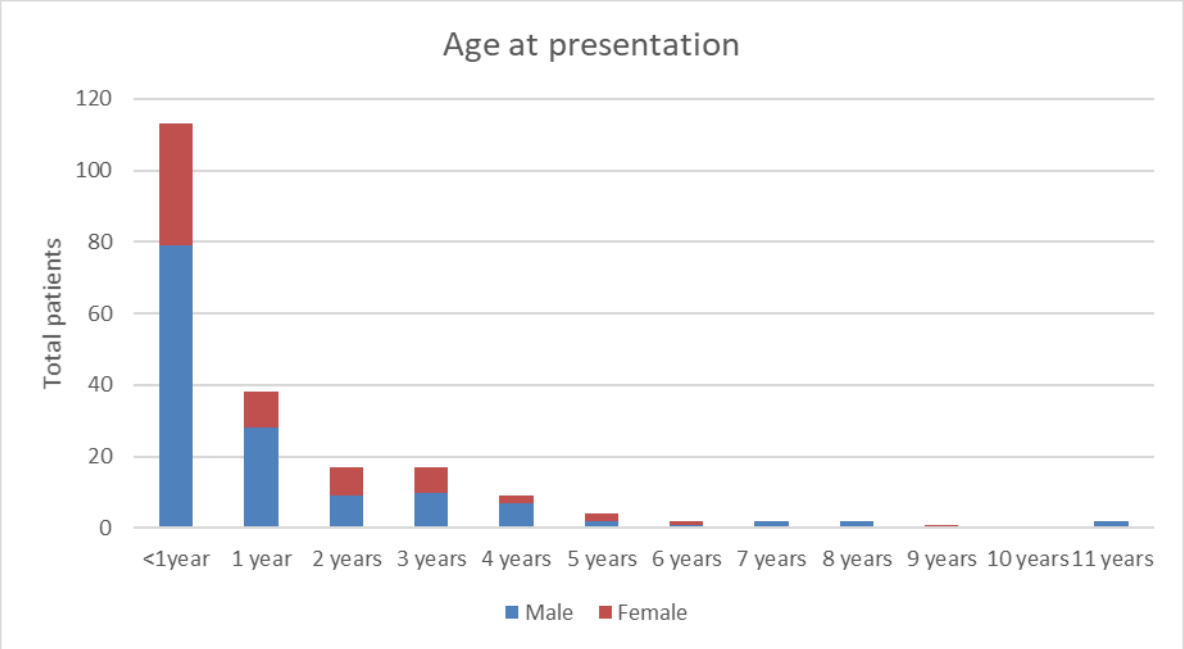

Table 1- patient demographics and the most common medical, syndromic and secondary airway comorbidities in patients presenting with tracheobronchomalacia

Demographics	**Total patients**
Male: female (ratio)	151:68 (2.2:1)
Mean age at presentation (range)	18.4 months (0-11.6 years)
Mean age at diagnosis (range)	21.5 months (0-11.8 years)
Primary: secondary malacia (ratio)	161:57 (2.8:1)
Medical diagnosis	**Total patients**
Bronchopulmonary dysplasia	17 (7.8%)
Cleft palate	7 (3.2%)
Developmental delay	21 (9.6%)
GORD	52 (23.3%)
Oesophageal atresia/ tracheoesophageal fistula	19 (8.7%)
Genetic/ syndromic diagnosis	**Total patients**
Trisomy 21 syndrome	13 (5.9%)
VACTERL association	7 (3.2%)
DiGeorge syndrome	4 (1.8%)
CHARGE syndrome	4 (1.8%)
Other	24 (11%)
Secondary airway pathology	**Total patients (%)**
Laryngomalacia	45 (20.5%)
Subglottic Stenosis	25 (11.4%)
Adenoid +/- tonsillar hypertrophy	39 (17.8%)
Granulation tissue	11 (5.0%)

Vocal cord paresis	7 (3.2%)
Vocal cord nodules	5 (2.3%)
Other	10 (4.6%)

Table 2- aetiology of secondary tracheobronchomalacia in patients based on clinical diagnosis and cross-sectional imaging.

Other causes: compression by bovine arch configuration, normal aortic arch, aberrant subclavian artery branching, tracheal diverticulum, enlarged mediastinal lymph node.

Secondary aetiology	**Total patients (%)**
Vascular Ring • Right aortic arch +/- left ligamentum arteriosum +/- aberrant subclavian • Double aortic arch • Pulmonary artery sling • Other	17 (7.8%) 9 (4.1%) 5 (2.3%) 1 (0.45%) 2 (0.9%)
Innominate artery compression	24 (11%)
Previous tracheostomy (non-tracheomalacia indication)	8 (3.7%)
Other	9 (4.1%)

Table 3- most common presenting symptoms and clinical features according to patient age (<1 and > 1 year old)

Clinical feature	**Age <1year (%)**	**Age >1year (%)**
Apparent life-threatening events	43 (35.2%)	4 (5.3%)

Difficulty weaning from mechanical ventilation	19 (15.6%)	3 (4%)
Feeding difficulties	33 (27.0%)	8 (10.7%)
Persistent cough	10 (8.2%)	17 (22.7%)
Prematurity <38 weeks gestation	39 (32.0%)	13 (17.3%)
Previous intubation	49 (40.2%)	21 (28.0%)
Previous NICU/PICU admission	63 (51.6%)	21 (28.0%)
Recurrent respiratory tract infections	24 (19.7%)	44 (58.7%)
Shortness of breath	22 (18.0%)	10 (13.3%)
Stridor	88 (72.1%)	23 (30.7%)

Table 4- most common surgical interventions performed in patients including procedures performed to correct tracheobronchomalacia (TBM interventions) and airway procedures performed after initial diagnosis and management (follow-up interventions).

Surgical intervention	**No. of patients**
Aortopexy	10 (4.6%)
Division of vascular ring	12 (5.5%)
Surgical tracheostomy	11 (5%)
Posterior tracheopexy	1 (0.45%)
Secondary interventions	
Tonsillectomy and/ or adenoidectomy	35 (16%)
Supraglottoplasty	11 (5%)
Laryngotracheal reconstruction	5 (2.3%)
Other	2 (0.9%)

Table 5- odds ratios for positive and negative predictors of surgical intervention on univariate analysis

Clinical feature	**Odds ratio**	**95% Confidence interval**	**p value**
Positive predictors			
Difficulty weaning from mechanical ventilation	5.3125	2.0985, 13.4491	p=0.001
Secondary malacia	4.4176	1.9250, 10.1377	p=0.001
Previous NICU/PICU admission	3.8514	1.5537, 9.5467	p=0.002
Previous intubation	3.8352	1.6337, 9.0035	p=0.001
ALTEs	3.7212	1.6339, 8.4747	p=0.002
<38 weeks gestation	2.7556	1.2190, 6.2287	p=0.016
Cardiac abnormalities	1.4421	0.6206, 3.3507	p=0.401
Dyspnoea	1.0072	0.3544, 2.8626	p=0.989
Negative predictors			
Persistent cough	0.1586	0.0208, 1.2106	p=0.020
Recurrent RTIs	0.2734	0.0907, 0.8241	p=0.010
Feeding difficulties	0.3746	0.1075, 1.3052	p=0.088
Age >1year	0.4951	0.1995, 1.2287	p=0.115
Stridor	0.9185	0.4045, 2.0855	p=0.839

Table 6- odds ratios for positive predictors of surgical intervention on multivariate analysis

Clinical feature	**Odds ratio**	**95% Confidence interval**	**p value**
Secondary malacia	8.2118	1.9250, 10.1377	p=<0.001
Difficulty weaning from mechanical intervention	3.8514	1.5537, 9.5467	p=<0.001
ALTEs	3.8352	1.6337, 9.0035	p=0.018