Current management and outcome of tracheobronchial malacia and stenosis presenting to the paediatric intensive care unit

Abstract  Objective: To identify factors associated with mortality and prolonged ventilatory requirements in patients admitted to our paediatric intensive care unit (PICU) with tracheobronchial malacia and stenosis diagnosed by dynamic contrast bronchograms.

Design: Retrospective review.

Setting: Tertiary paediatric intensive care unit.

Patients: Forty-eight cases admitted to our PICU over a 5-year period in whom a diagnosis of tracheobronchial malacia or stenosis was made by dynamic contrast bronchography (1994–1999).

Interventions: Conservative management, tracheostomy and long-term ventilation, surgical correction, internal or external airway stenting.

Measurements and results: Recording of clinical details, length of invasive ventilation and appearance at contrast bronchography. Five groups of patients were defined: isolated primary airway pathology \((n=7)\), ex-premature infants \((n=11)\), vascular rings \((n=9)\), complex cardiac and/or syndromic pathology \((n=17)\) and tracheoesophageal fistulae \((n=4)\). The overall mortality was 29%. Median length of invasive ventilation in survivors was 38 days and in patients who died 45. Mortality was highest in the patients with complex cardiac and/or syndromic pathology \((p=0.039\) Cox regression analysis\) but was not related to any other factor. Patients with stenosis required a significantly longer period of ventilatory support (median length of ventilation 59 days) than patients with malacia (39 days).

Conclusions: Length of ventilation and bronchographic diagnosis did not predict survival. The only factor found to contribute significantly to mortality was the presence of complex cardiac and/or syndromic pathology. However, patients with stenosis required longer ventilatory support than patients with malacia.

Key words  Tracheal stenosis · Bronchomalacia · Tracheomalacia · Tracheobronchomalacia · Bronchogram · Ventilation · Mortality
Introduction

Tracheobronchomalacia is a condition of dynamic airway collapse during expiration. In tracheal stenosis, fixed airway narrowing causes airway obstruction in inspiration and in expiration. These conditions, first described in 1952 [1], may be primary, when cartilaginous rings are congenitally malformed, or secondary, due to degeneration of previously normal cartilage. In neonatal chronic lung disease, development of the tracheobronchial tree may be abnormal and the cartilage may be weak [2]. Extrinsic airway compression by a vascular ring or by enlarged great vessels in the presence of a large shunt can result in permanent airway narrowing [3]. Tracheo-oesophageal fistulae are often associated with malacia of a portion of the trachea at the point of insertion of the fistula. Airway abnormalities may develop in recurrent aspiration syndromes and in acquired cartilaginous disorders such as relapsing polychondritis. The severity of disease depends on the length, location and degree of narrowing in the affected airway segments.

Although primary isolated tracheobronchial malacia rarely requires ventilatory support and is often self-limiting, infants with tracheobronchial malacia or stenosis with abnormal bronchograms who require ventilatory support have been reported to have a high morbidity and mortality, often related to airway obstruction and respiratory failure [4]. This has not been our experience. We therefore reviewed children admitted to our intensive care unit with tracheobronchial malacia and stenosis, diagnosed at contrast bronchography, to determine which factors affected outcome.

Materials and methods

Following local ethics committee approval, the notes of 48 patients, who had been admitted to our intensive care unit between 1994–1999 and in whom a diagnosis of tracheobronchial malacia or stenosis had been made at contrast bronchography, were reviewed. Information collected included underlying diagnosis, length of invasive ventilation around the time of diagnosis, surgical interventions, mortality, time between diagnosis and the present day for survivors and time between diagnosis and death for those who died. Underlying diagnostic categories were defined as isolated primary airway pathology or airway pathology associated with neonatal chronic lung disease, complex cardiac and/or syndromic pathology, vascular rings or tracheo-oesophageal fistula.

Radiographic data were also reviewed. Bronchography was performed as previously described [5]. The patients were lightly sedated, intubated and were breathing spontaneously. The endotracheal tube was maintained in a high position with the tip in the subglottic region. Opacity was obtained by bolus injections of 0.5–2 ml of contrast medium (Omnipaque, Nycomed, Birmingham, UK), injected into the airway via the endotracheal tube through a 3.7 Fr angiography catheter with a single end-hole. Contrast dispersal was obtained by hand ventilation. Up to five affected sites were identified (trachea, left main bronchus, right main bronchus, bronchus intermedius and peripheral bronchi). A pressure monitor was connected to the airway circuit via a Y connector and when collapse was found positive end-expiratory pressure (PEEP) was applied and opening pressures noted. Airways were defined as stenosed when the narrowing was fixed and unaffected by PEEP up to 25 cmH₂O and malacic when the narrowing was dynamic and improved with positive airway pressure. Infants with tracheal stenosis with distal malacia were classified as tracheal stenosis for the purposes of this analysis since the numbers of patients with both forms of pathology was small (n = 10). Our bronchograms are not stored as cine-films or videos but as static images, so no attempt was made at retrospective blinded severity scoring as this was felt to be unlikely to yield any useful information.

Data were analysed using SPSS 8.0 for Windows (SPSS, UK). Analysis of survival and length of invasive ventilation was performed using Cox regression analysis. The Kruskal-Wallis test was used to determine whether differences in the length of follow-up in survivors were significant.

Results

Of the 48 patients, 19 were female. The median age at diagnosis was 137 days (interquartile range 62–213 days). The number of patients in each group was as follows: isolated primary airway pathology (7), ex-premature infants (11), vascular rings (9), complex cardiac and/or syndromic pathology (17) and tracheo-oesophageal fistulae (4). The overall mortality was 14/48 (29%). Median length of follow-up in survivors was 439 days and was not significantly different between any of the groups (p = 0.635, Kruskal-Wallis test). Outcome data is presented related to diagnosis and to operative procedure (Tables 1 and 2).

Isolated primary airway pathology

Seven patients with primary airway pathology were identified. The median age at diagnosis was 66 days. One term infant was referred for surgical management of congenital subglottic stenosis, tracheobronchomalacia being diagnosed when weaning difficulties were encountered following surgery. Invasive ventilation was defined in this group as the time from first intubation for airway obstruction to death or weaning to either extubation or continuous positive airway pressure (CPAP) or biphasic positive airway pressure (BiPAP) via a tracheostomy. The median length of invasive ventilation of survivors was 60 days. Five patients had isolated tracheobronchomalacia and two had tracheal stenosis with airway malacia distal to the stenotic segment. Two patients with mild tracheobronchomalacia were extubated, one after a cricoid split procedure for congenital subglottic stenosis, the other requiring no intervention. The latter patient was excluded from the statistical analysis as he had severe combined immune deficiency and died from graft versus host disease following bone