

Preoperative laryngotracheobronchoscopy in infants with esophageal atresia: why is it not routine?

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Abstract The value of laryngotracheobronchoscopy (LTB) immediately prior to repair of esophageal atresia with or without tracheo-esophageal fistula is contentious. Currently, there is a wide range of opinion on the utility of this investigation which is reflected by huge variation in clinical practice. This review is a critical analysis of the arguments for and against performing routine LTB prior to esophageal atresia repair. Reserving LTB for selected cases only is potentially disadvantageous since it limits the surgeon's and anesthetist's familiarity with the procedure, equipment, and range of potential findings. There is sufficient evidence to suggest that routine preoperative LTB becomes the standard of care.

Keywords Esophageal atresia · Bronchoscopy · Laryngoscopy · Tracheo-esophageal fistula

Introduction

The utility of laryngotracheobronchoscopy (LTB) immediately prior to repair of esophageal atresia (EA) with or

without tracheo-esophageal fistula (TEF) is contentious. Contemporary international surveys have highlighted how variably this investigation is performed in clinical practice. A recent survey of 170 IPEG members from 31 countries found that LTB was routinely undertaken by 60% of pediatric surgeons, selectively by 13%, and never by 27% [1]. Another survey of 178 delegates at the EUPSA-BAPS 2012 Congress found that 56% do not perform routine preoperative endoscopy [2]. A prospective French national registry of EA showed a preoperative LTB rate of just 22% [3] and retrospective multicenter studies from Australasia and the USA recorded figures of 41 and 66%, respectively, with huge variations in rates between individual institutions [4, 5]. Given this wide variation in clinical practice, we decided to review the arguments for and against performing routine LTB prior to repair of EA.

The case for routine preoperative laryngotracheobronchoscopy

Laryngotracheobronchoscopy preceding repair of EA ± TEF has two main functions: (1) to characterize the number and site of TEF, and (2) to identify co-existing anomalies.

1. The number and site of tracheo-esophageal fistula(e)
Upper pouch fistula The common anatomical varieties of EA ± TEF and their relative frequencies derived from six published series totaling more than 2200 cases [6–11] are shown in Fig. 1. Given this distribution, some experts have argued for selective use of LTB, reserving it for cases where the incidence of an upper pouch fistula is proportionately greater [12]. Considering Fig. 1, the number needed to 'treat' (NNT) to identify an upper pouch fistula increases from approximately 60 when

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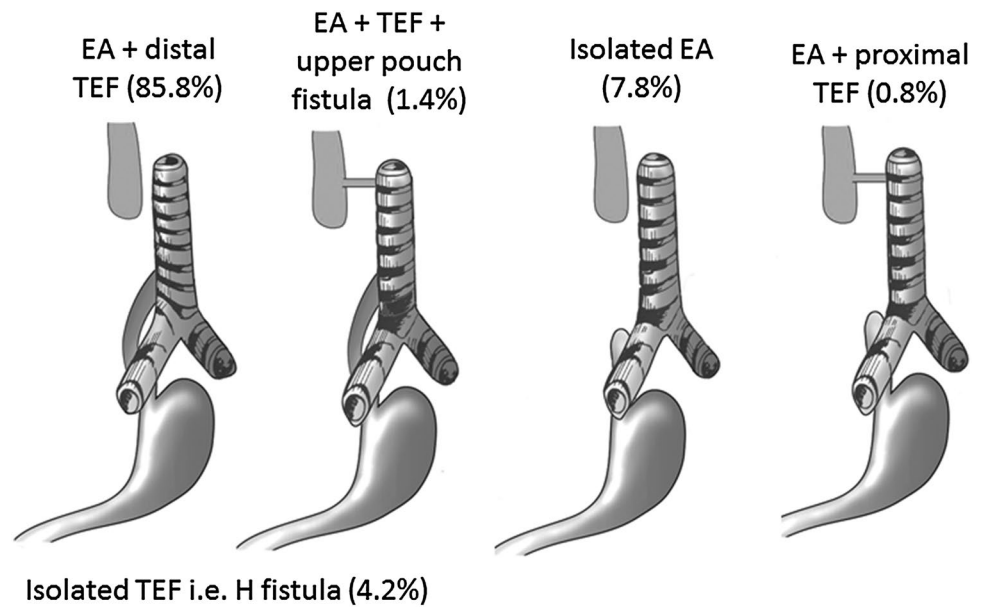
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Fig. 1 Major anatomical configurations of esophageal atresia and tracheo-esophageal fistula and their frequencies. According to these figures, if gas is visible on a plain abdominal radiograph in an infant with EA 98.4% will have EA + distal TEF and 1.6% will have EA + distal TEF + upper pouch fistula. If the abdomen is gasless 90.7% will have isolated EA and 9.3% EA + proximal TEF



distal gas is present on a plain radiograph to 11 when there is no distal gas. This logic assumes that all upper pouch fistulae in these collected series were eventually detected but we know that an upper pouch fistula may rarely be detected for the first time in adolescence or early adulthood [13, 14]. In addition, some centers have reported a higher incidence of upper pouch fistula with EA/TEF [15]. Smaller individual institutional experiences estimate that the NNT to define an upper pouch fistula in the context of absent distal gas may be less [16], even as low as 5 [17], 3 [18] or 2 [19, 20]. In one center, the detection of an upper pouch fistula was statistically significantly improved following the introduction of routine tracheobronchoscopy [20]. However, it must be acknowledged that an upper pouch fistulae may be missed on LTB. When present, it may only be visible as a small pit just distal to the vocal cords that does not necessarily open and close with ventilation; concomitant insufflation of the esophagus whilst observing the pit can be helpful [21, 22].

Distal tracheo-esophageal fistula It is commonly assumed that a gasless abdomen equates to absence of a distal fistula, whereas in reality up to 15% of such patients have a distal fistula that is either very small or occluded by a mucous plug [23, 24]. In one series, 3 of 11 patients with suspected pure EA were found to have a distal TEF at tracheobronchoscopy [16].

Establishing the site of entry of the fistula into the tracheobronchial tree is useful for both the anesthetist and surgeon. It may modify the positioning of the endotracheal tube and the surgical approach [25–27]. Awareness of the likely anatomy of the TEF prior to surgery may facilitate esophageal dissection [26] or prepare the

surgeon for dealing with a potential long gap [28]; the latter is arguably even more important when selecting patients for thoracoscopic repair. Of 149 patients with EA assessed by bronchoscopy in one series, the location of the fistula was as follows: 1% bronchial, 9% carinal (Fig. 2), 23% within 1 cm proximal to the carina, 59% mid-tracheal, and 8% cervical [14]. Some surgeons consider that the area of transmitted pulsation at LTB can predict the side of the aortic arch [26] which could influence the operative approach to repair of the EA/TEF. However, the reliability of this observation has not been critically evaluated.

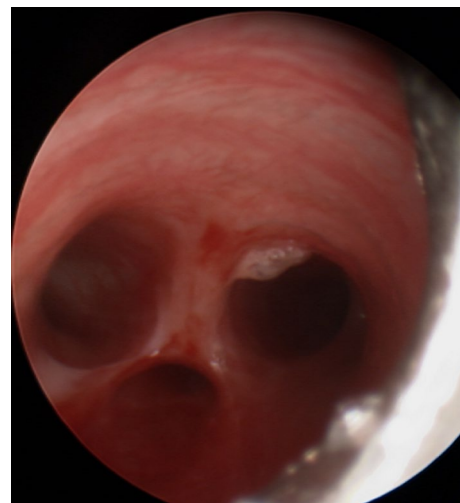


Fig. 2 Flexible tracheobronchoscopy in a neonate with esophageal atresia and a distal tracheo-esophageal fistula opening at the carina

Tracheobronchoscopy also enables the size of a distal TEF to be assessed which may assist with airway management by indicating the potential likelihood of intubating the fistula. Visualizing the TEF may also facilitate selective intubation beyond the fistula [29] or enable temporary placement of a Fogarty balloon catheter to occlude the fistula [18, 30] to control an air leak through it (especially important in infants with respiratory distress and EA/TEF) [25].

Misdiagnosis of EA/TEF There are rare reports of iatrogenic neonatal pharyngoesophageal perforation where the abnormal passage of a naso/orogastric tube causes diagnostic confusion with EA/TEF [31, 32]. There are usually clinical and radiographic clues but some infants have been subjected to needless thoracotomy for presumed EA/TEF [31, 33]. Preliminary LTB would confirm the absence of a TEF in such cases.

Multiple TEF There are rare cases of three fistulae connecting the trachea to the esophagus [34]. A detailed preliminary LTB provides no guarantee that all three fistulae would be identified at the outset of treatment but it may alert the surgeon to unusual anatomy.

2. Co-existing airway pathology

Potentially important co-existing airway anomalies can be diagnosed by preliminary LTB. In one consecutive series of 88 neonates with EA ± TEF who underwent LTB, there were 2 (2.3%) with a laryngeal cleft, one of which was significant [16]. In series where preliminary LTB was not routinely undertaken, laryngeal clefts have been subsequently detected in 0.5–1.4% of cases [8, 35, 36]. While it can be argued that type 1 clefts are often not clinically significant and failure to make the diagnosis is not harmful, some patients with EA have a more severe laryngeal cleft and are at risk of repeated pulmonary aspiration [8, 35–37].

Other rare congenital airway problems diagnosed by LTB include congenital subglottic stenosis [16, 17], valvular cyst [16], and tracheal and/or bronchial stenosis [14, 17, 18, 26, 38–40].

Tracheomalacia is a clinically significant comorbidity in 5–20% of children with EA/TEF, although it is present to a variable degree in most affected children because of a developmental deficiency in the tracheal cartilaginous rings [41]. Tracheomalacia is usually diagnosed on tracheobronchoscopy by observing anteroposterior narrowing or collapse of the tracheal lumen associated with anterior ballooning of the widened posterior membranous tracheal wall on expiration during spontaneous ventilation [42]. Tracheomalacia tends to worsen during the first few months of life and findings on preoperative LTB at the time of EA/TEF repair correlate poorly with later development of clinically significant tracheomalacia [43]. Consequently, *preoperative*

LTB adds little to the evaluation of this particular airway problem.

Unilateral or bilateral symptomatic vocal cord paresis or paralysis is reported in 3–5% of infants after EA ± TEF repair [4, 44, 45]. The true prevalence of vocal cord palsy may well be higher because symptoms can be mistakenly attributed to laryngotracheomalacia or gastroesophageal reflux [45]. Vocal cord palsy contributes to respiratory morbidity through impaired airway protection, aspiration pneumonia, and failed extubation. It is not yet clear whether vocal cord palsy associated with EA is predominantly congenital or iatrogenic but the latter is more likely since it appears to be more common after extensive upper pouch mobilization [45] and possibly after thoracoscopic EA/TEF repair [46]. However, abnormal development of the vagus nerve and recurrent laryngeal nerve branching pattern have been demonstrated in an experimental animal model of EA/TEF [47]. Prospective evaluation of vocal cord function prior to EA ± TEF repair should help determine the relative contributions of congenital and acquired pathology.

The case against routine preoperative laryngotracheobronchoscopy

1. Logistics

Although it has been stated that the additional procedural time of performing LTB is “minimal” [18] or “a few minutes” [48], this depends in part on the familiarity of the pediatric surgeon and anesthetist with the procedure. Laryngotracheobronchoscopy can be performed using a variety of techniques and equipment which must be tailored to the size and stability of the infant. A rigid ventilating neonatal bronchoscope is most often used since this allows positive pressure ventilation and, if required, can be used to pass a Fogarty catheter into the fistula. Alternatively, flexible fiberoptic bronchoscopy can be used; this is usually performed while maintaining spontaneous ventilation [18] and each endoscopic assessment is limited to about 30–45 s [49]. There are numerous techniques including passing a 2.2-mm fiberoptic bronchoscope down a 3-mm ETT whilst maintaining continuous positive pressure ventilation [48] or using a small rod lens without the outer ventilating sheath [50]. The choice of equipment is dictated by tracheal size, the aims of the procedure (diagnostic or therapeutic), anesthetic expertise, availability of equipment, and surgeon preference. Traditionally, fiberoptic scopes of 2.2 mm [51] (or 2.5 mm with a working channel) or even smaller diameter [52] have been used, but 2.8 mm and 3.7 mm videoscopes (the latter with a working chan-

nel) with improved image resolution are now available. LTB has been performed in neonates with EA \pm TEF as small as 1.3 kg [18, 48].

It is beyond the scope of this article to discuss the relative merits and limitations of other methods used to characterize the anatomy of EA \pm TEF preoperatively including contrast studies, ultrasound scans, cross-sectional imaging, and esophagoscopy. These have been reviewed elsewhere [49] and currently do not provide the diagnostic accuracy [19, 20] or versatility afforded by LTB, although they may be complementary investigations.

2. Risks

Complications of neonatal bronchoscopy are uncommon but include moderate/transient desaturation, laryngospasm, apnoea, bradycardia, inadvertent extubation, and airway trauma [51, 53, 54].

3. Other arguments

Various arguments against routine preoperative LTB have been put forward: when distal gas is present on a plain radiograph in an infant with EA up to 60 such infants need to be examined to identify an upper pouch fistula, raising questions of validity and cost-effectiveness; in the presence of a gasless abdomen and apparently isolated EA, if a distal TEF is present it will still be found at operation; prompt surgical control of the fistula rather than attempted endoscopic occlusion is the best option in infants with respiratory distress and EA/TEF [55]; an upper pouch fistula can still be missed despite careful LTB and a negative examination might provide false reassurance [21, 22]; most laryngeal clefts associated with EA are type 1 and clinically insignificant while more severe varieties will be identified because of symptoms (as will other rare major airway anomalies); tracheomalacia is an evolving condition that cannot be adequately assessed by early LTB; and vocal cord paresis is predominantly iatrogenic and therefore documenting vocal cord function preoperatively is not particularly helpful.

Conclusions

Robust arguments can be developed for and against routine preoperative LTB prior to repair of EA \pm TEF. Despite numerous publications supporting the value of this investigation, pediatric surgeons remain divided about its role in clinical practice. Reserving LTB for selected cases only is potentially disadvantageous since it limits both the surgeon's and anesthetist's familiarity with the procedure, equipment,

and range of potential findings. Perhaps it is time to recommend that routine preoperative LTB becomes the standard of care.

Compliance with ethical standards

Conflict of interest The authors declare that there is no competing interest.

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