Paediatric Respiratory Reviews 19 (2016) 10–15

Contents lists available at ScienceDirect

Paediatric Respiratory Reviews

Mini-symposium: Oesophageal Atresia and Tracheo-oesophageal Fistula

Surgical management of oesophageal atresia

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EDUCATIONAL AIMS

The reader will come to appreciate how:

1. To understand the rationale for preoperative work up in oesophageal atresia
2. To describe the surgical correction of oesophageal atresia
3. To review the role of thoracoscopic surgery in oesophageal atresia
4. To describe surgical options to correct long-gap oesophageal atresia
5. To review early post-operative complications.

ARTICLE INFO

Keywords:
Oesophageal atresia
Long Gap oesophageal atresia
Tracheo-oesophageal fistula

SUMMARY

There have been major advances in the surgery for oesophageal atresia (OA) and tracheo-oesophageal fistula(TOF) with survival now exceeding 90%. The standard open approach to OA and distal TOF has been well described and essentially unchanged for the last 60 years. Improved survival in recent decades is most attributable to advances in neonatal anaesthesia and perioperative care. Recent surgical advances include the use of thoracoscopic surgery for the repair of OA/TOF and in some centres isolated OA, thereby minimising the long term musculo-skeletal morbidity associated with open surgery. The introduction of growth induction by external traction (Foker procedure) for the treatment of long-gap OA has provided an important tool enabling increased preservation of the native oesophagus. Despite this, long-gap OA still poses a number of challenges, and oesophageal replacement still may be required in some cases.

INTRODUCTION

The first successful repair of oesophageal atresia (OA) and tracheo-oesophageal fistula (TOF) was performed in 1941: Cameron Haight ligated the TOF prior to an end-to-end oesophageal anastomosis through a left extrapleural approach [1]. In the subsequent 75 years the overall survival has improved to exceed 90%, with mortality now usually associated with prematurity and/or cardiac comorbidity [2,3]. Despite improved survival, OA still presents unique surgical challenges, many of which are discussed here. This review focuses on the surgical management of OA and any associated TOF, highlighting the preoperative investigations, timing of surgery, surgical approaches, surgical complications and challenges of 'long-gap' OA.

PREOPERATIVE ASSESSMENT

The two primary goals of preoperative assessment in the patient with a clinical diagnosis of OA are:

1. Confirmation of the diagnosis;
2. Identification of associated anomalies with immediate management implications for the planned oesophageal atresia surgery.

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http://dx.doi.org/10.1016/j.prrv.2016.04.003
1526-0542/

Table 1
Frequency of associated anomalies in OA (Data extracted from Burge et al. 2013 [4])

Associated Anomaly	All OA	OA with distal TOF	Other OA subtypes
Any	52%	54%	44%
Vertebral	7%	6%	16%
Anorectal	11%	6%	40%
Cardiac	34%	36%	28%
Renal	9%	8%	16%
Limb	11%	14%	3%
VACTERL	13%	12%	24%
Chromosomal	3%	-	-

As more than half of OA patients have an associated anomaly (Table 1), [4] additional systems screening for anomalies is ordinarily deferred until after primary OA surgery. Exceptions to this are outlined briefly below.

The clinical diagnosis of OA is made when a 10 French (Fr) tube arrests in the upper oesophagus approximately 10-12 cm from the lips. Tubes of narrower calibre may give the false impression of tube passage by coiling in the dilated upper oesophageal pouch, or rarely traversing the trachea and TOF to enter the stomach [5,6]. A diagnosis of OA is then confirmed with a plain chest x-ray to show the arrested 10 Fr tube within the relative lucency of the dilated upper oesophageal pouch. An accompanying distal TOF is indicated by the presence of gastrointestinal gas.

Particular attention is paid to marked stomach dilatation, which may reflect preferential ventilation of the stomach via the TOF, or concomitant duodenal atresia with a 'double bubble'. In either setting, emergency distal TOF ligation is indicated to prevent the rare but morbid complication of gastric perforation [7]. Conversely, a 'gasless abdomen' on preoperative x-ray raises the possibility of either pure OA or OA with a proximal TOF, and should prompt further investigation as detailed below. The chest x-ray should also be assessed for evidence of early complications (e.g. pulmonary aspiration, intubation) or associated anomalies (e.g. abnormalities of the cardiac silhouette, vertebrae and ribs).

A preoperative echocardiogram is undertaken to define any associated major congenital heart disease (CHD), particularly duct-dependent lesions which may necessitate particular anaesthetic management or prior cardiac surgery. In addition, an echocardiogram may demonstrate vascular anomalies relevant to operative decision-making, most notably a right-sided aortic arch (RAA) which is present in approximately 4% of OA cases [8]. The importance of *routine* preoperative echocardiography is highlighted by expert commentators, [5,6] whilst others present data to moderate this stance, [9,10] showing preoperative echocardiography findings of CHD and/or RAA seldom alter the operative plan, including the choice of right vs left thoracotomy in cases with a known RAA [8,10].

The role of routine preoperative laryngotracheobronchoscopy (LTB) in OA patients remains a matter of debate, [11] with only 43-60% of contemporary paediatric surgeons routinely using preoperative LTB in this setting [12,13]. Advocates cite preoperative LTB findings which may impact management in 21-45% of OA patients, most notably unusual fistula position (Figure 1) and tracheobronchial tree anomalies, and less commonly laryngeal clefts or subglottic stenosis [14–16]. The presence of significant tracheomalacia may alert clinicians to the need for non-invasive support following extubation. The benefit of preoperative LTB is maximal in newborns with suspected pure OA due to a 'gasless abdomen' on x-ray. In this OA subgroup, LTB reveals a proximal TOF in 20-33% with resultant change in management [14–17]. An upper pouch oesophagogram may augment LTB to identify a 'near-missed' proximal TOF, [18] but is highly dependent on local radiology expertise. Preoperative endoscopic intubation of the TOF with the aim facilitating surgical repair, Fogarty balloon TOF occlusion to aid ventilation and selective trans-tracheal gastric drainage have been described [11,15,16,19].

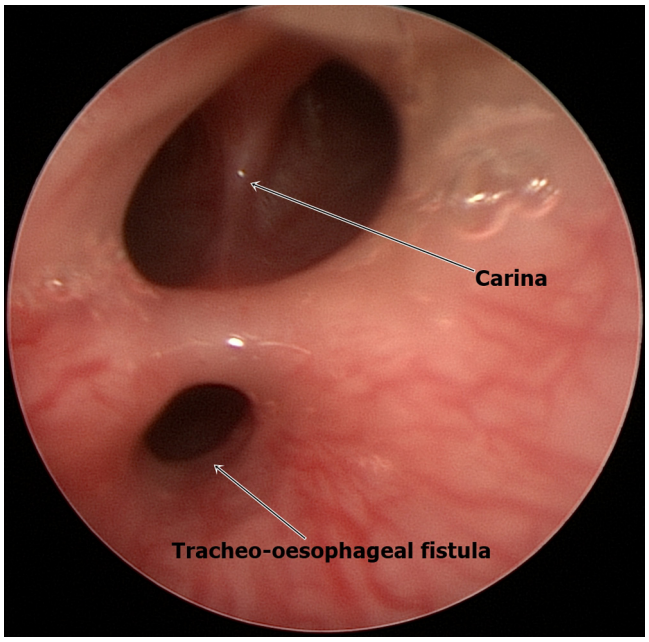

Figure 1. Bronchoscopy of a distal tracheo-oesophageal fistula.

Routine screening investigations for other associated anomalies would include renal ultrasound, spinal ultrasound and sacral x-ray. These are ordinarily not performed preoperatively, except a renal ultrasound in the newborn who has not voided (OA is uncommonly associated with renal agenesis), [6] or urgent genetic testing where an undiagnosed lethal syndrome is suspected, e.g. Trisomy 13 (Patau) or 18 (Edwards).

OPEN SURGERY

With rare exceptions, OA surgery is not an emergency and can be deferred whilst preoperative investigations are obtained and an appropriately skilled anaesthetic and surgical team assembled. Indications for emergency OA surgery are limited to a markedly dilated stomach at risk of perforation, or the premature infant with evolving Respiratory Distress Syndrome (RDS) in whom preferential TOF ventilation is a significant contributor to a deteriorating ventilatory status. Two recent studies report increased complications in OA patients undergoing surgery within 24 hours of birth or after-hours [20,21]. Selection biases aside, these studies highlight the benefits of in-hours OA surgery, which may be safely deferred beyond 24 hours of life.

The goals and key steps of the open approach have remained largely unchanged over decades, and are summarised below. Specific goals and operative strategies for other OA variants, e.g. long-gap OA, are described elsewhere in this review.

Key operative goals

1. **TOF ligation** to prevent further soiling of the tracheobronchial tree with stomach contents and restore the ventilation dynamics of the intact trachea.
2. **Restoration of oesophageal continuity**, which can be deferred if the child's status is poor at completion of TOF ligation.