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Oesophageal atresia with no distal tracheoesophageal fistula: Management and outcomes from a population-based cohort☆

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ABSTRACT

Purpose: To describe the incidence and outcomes to one-year in infants born with oesophageal atresia (OA) with no distal tracheoesophageal fistula within a population cohort.
Methods: A subgroup analysis of a prospective multicentre population cohort study was undertaken describing the outcomes of infants with OA and no tracheoesophageal fistula, (type A) and those with only an upper pouch fistula, (type B).
Main results: Twenty-one of 151 infants in the whole cohort were diagnosed with type A or B oesophageal atresia (14%). Fifteen were type A (71%) and six type B (29%). Infants with type B had a shorter gap length than those with type A: 2.5 vertebral bodies (2–3) vs. 5 (4–6) ($p = 0.008$). All infants with type B OA underwent oesophageal anastomosis, 83% ($n = 5$) as the primary procedure. All infants with type A, underwent staged management. Six (40%) had delayed primary anastomosis and eight required oesophageal replacement (53%). One infant died prior to reconstruction. The median time to delayed primary anastomosis in infants with type A or B OA was 82 days (75–89 days) ($n = 7$). The median time to oesophageal replacement was 94 days (89–147 days) ($n = 8$). Median length of stay for infants with type A or B OA from first operation to first discharge was 101 days (31–123 days).
Conclusions: Infants with type B OA had a shorter gap length and all were managed with oesophageal anastomosis. OA with no distal tracheoesophageal fistula is uncommon at a population level and frequently has a complex course.
Level of Evidence: Rating: II.

Oesophageal atresia (OA) and tracheoesophageal fistula (TOF) are congenital anomalies affecting 2.6–2.9 per 10,000 live-born infants [1,2]. The Gross Classification (Fig. 1) describes the common anatomical variants [3]. Population-based outcomes in infants with type C oesophageal atresia from the United Kingdom have previously been reported up to one year of age [4,5]. Infants with no distal fistulous connection (types A and B) are born less frequently [6]. Evidence from previous work suggests that these infants may be born earlier and at a lower gestational age than those with type C oesophageal atresia, although no statistically significant differences have been observed, possibly because of limitations of study power [4]. Infants without distal fistula frequently have a 'long-gap' between the two ends of the esophagus. The lack of attachment of the distal esophagus to the airway is thought to be responsible for its relative underdevelopment. Where primary anastomosis is not possible, surgical options include formation of an oesophagostomy and gastrostomy or performing gastrostomy alone and draining the upper pouch with continuous sump suction [7,8]. Reflux from the stomach into the lower esophagus combined with somatic growth means that delayed primary repair is often possible [8–10]. Infants with OA and no distal fistula therefore present a significant challenge to the health professionals caring for them with regard to surgical decision-making and technique as well as supportive respiratory, nutritional and developmental care.

Infants with type A and B variants may readily be detected pre-operatively because of a lack of distal intestinal gas. This pathognomonic feature combined with the absence of a connection between the airway and the lower gastro-intestinal tract mean that there is seldom a need to proceed to theater urgently. Thus surgery may be approached in a planned manner with an appropriate level of support and expertise available.

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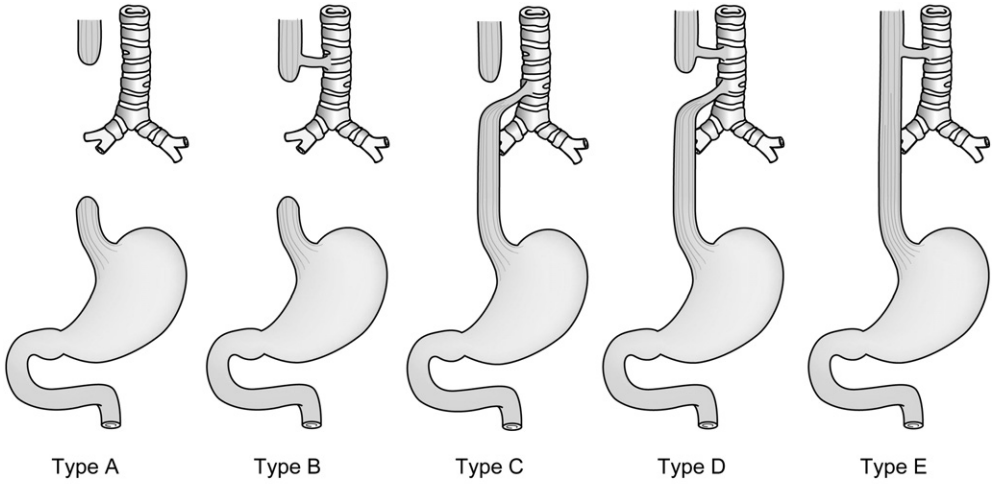

Fig. 1. The gross classification of oesophageal atresia subtypes.

The majority of the published literature on these cases reports on only small numbers of infants, or are retrospective studies covering a long period of time, during which, treatment, and subsequently outcomes, may have evolved. The present study aimed to gather data prospectively from all infants in the UK and Ireland over a one-year period to establish the current incidence and outcomes of cases of OA with no distal fistula. The data presented here reflect management choices and contemporary outcomes on a population basis in this challenging group of infants.

1. Methods

We have previously reported the results of a prospective multicentre cohort study undertaken in the UK and Ireland which included all live born infants with oesophageal atresia and tracheoesophageal fistula born between April 1st 2008 and March 31st 2009 referred for surgical care in one of 28 pediatric surgical centers [4,5]. Ethical approval was granted by the London Research Ethics Committee (reference 07/H0718/92).

All eligible infants were identified by nominated clinicians who reported the cases using the British Association of Pediatric Surgeons Congenital Anomalies Surveillance System (BAPS-CASS) as described by Owen et al. [11]. Anonymised data were collected using detailed clinical data collection forms; one in the first few months after birth and a second requesting one-year outcomes. These data were coded and double-entered into a database checking for duplicates and for missing information. If inconsistencies were identified or answers were missing, the reporting clinicians were contacted to clarify.

Outcomes in infants with type C oesophageal atresia have previously been reported up to one year of age [4,5]. Within this analysis, we focus on reporting the management and outcomes of a subgroup of the population with type A and B oesophageal atresia.

Data analysis was undertaken in *Stata*, version 13. Data are quoted as median and interquartile range. Continuous data were compared using the Mann–Whitney test. Categorical data were compared using a chi^2 or Fisher's Exact Test as appropriate. 95% confidence intervals (c.i.) were calculated and $P < 0.05$ was assumed to represent statistical significance.

2. Results

2.1. Characteristics of infants with no distal fistula within the cohort

184 cases of OA were notified from centers in the UK and Ireland during the data collection period, (2008–2009). Completed data collection forms were received for 160 infants (88%). 9 of these were excluded as they were either duplicates or reported incorrectly. The characteristics of the 151 remaining infants have been previously been described [4,5]. Table 1 shows the number of infants with each subtype of OA [4,5]. The estimated live-birth prevalence of OA from this study (1.7 per 10,000; 95% c.i. 1.5–2.0) is compatible with estimation from other robust UK population-based data sources, including congenital anomaly register data (1.97 per 10,000; 95% c.i. 1.47–2.6) thus, there is unlikely to have been significant under-ascertainment [4,12]. Twenty-one infants had OA with no distal fistula, of these 15 (71%) had no upper pouch fistula (type A) and six, (29%) had an upper pouch fistula (type B). Only 32 infants in the cohort had the diagnosis of OA suspected antenatally (21%); 41% of these infants ($n = 13$) were type A or B. In 119 infants the diagnosis was not suspected antenatally (79%); 7% of these infants ($n = 8$) were type A or B. The absence of a distal connection, (leading to a smaller stomach) was strongly associated with OA being suspected antenatally ($p < 0.0005$). The median gestational age at birth for infants with type A and B OA was 37 weeks (33.5–38 weeks) and the median birthweight was 2.7 (1.6–3.0) kg. The median gestational age of those infants with type C and type E OA was 38 (36–39) weeks and their median birthweight 2.6 (2.1–3.0) kg. Neither of them was statistically significantly different from those with type A or B ($P = 0.06$) and ($P = 0.5$) respectively.

Infants with type A OA had a median gap length of 5 vertebral bodies (4–6) compared with a median gap length of 2.5 vertebral bodies (2–3) in infants with type B OA ($P = 0.008$).

2.2. Management

Fig. 2 shows the management pathways of the infants with type A and B OA within the cohort.

Table 1
Distribution of Anatomical Types of OA Within the Cohort.

OA classification	Whole cohort n (%)
Type A	15 (10)
Type B	6 (4)
Type C	126 (83)
Type D	0 (0)
Type E	4 (3)
Total	151