International Survey on the Management of Esophageal Atresia

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Abstract

Introduction  Because many aspects of the management of esophageal atresia (EA) are still controversial, we evaluated the practice patterns of this condition across Europe.

Methods  A survey was completed by 178 delegates (from 45 [27 European] countries; 88% senior respondents) at the EUPSA-BAPS 2012.

Results  Approximately 66% of respondents work in centers where more than five EA repairs are performed per year. Preoperatively, 81% of respondents request an echocardiogram, and only 43% of respondents routinely perform preoperative bronchoscopy. Approximately 94% of respondents prefer an open approach, which is extrapleural in 71% of respondents. There were no differences in use of thoracoscopy between Europeans (10%) and non-Europeans (11%, p = nonsignificant). Approximately 60% of respondents measure the gap intraoperatively. A transanastomotic tube (90%)

Keywords
► esophageal atresia
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► esophageal replacement
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and chest drain (69%) are left in situ. Elective paralysis is adopted by 56% of respondents mainly for anastomosis tension (65%). About 72% of respondents routinely request a contrast study on postoperative day 7 (2–14). Approximately 54% of respondents use parenteral nutrition, 40% of respondents start transanastomotic feeds on postoperative day 1, and 89% of respondents start oral feeds after postoperative day 5. Pure EA: 46% of respondents work in centers that repair two or more than two pure EA a year. About 60% of respondents opt for delayed primary anastomosis at 3 months (1–12 months) with gastrostomy formation without esophagostomy. Anastomosis is achieved with open approach by 85% of respondents. About 47% of respondents attempt elongation of esophageal ends via Foker technique (43%) or with serial dilations with bougies (41%). Approximately 67% of respondents always attempt an anastomosis. Gastric interposition is the commonest esophageal substitution.

Conclusion Many aspects of EA management are lacking consensus. Minimally invasive repair is still sporadic. We recommend establishment of an EA registry.

Introduction
Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) has always been considered as an index pediatric surgical condition, as the result of its treatment have often been used to portrait the standards of care provided by different institutions. However, in spite of the advances in neonatal care, anesthestics, and technologies many aspects of the management of this condition still remain controversial.

The aim of this study was to identify practice patterns in the management of EA–TEF and pure EA, to ascertain the degree of variability among pediatric surgical centers.

Methods
A questionnaire was administered to delegates attending the EUPSA-BAPS Joint Congress (Rome, June 13–16, 2012) and it was collected on the last day of the conference. Respondents were asked to fill in their position (Head of Department/Permanent Staff or Consultant/Trainee), hospital, and country of practice. The questionnaire was divided into the following two parts: (1) questions on the pre-, intra-, and postoperative management of EA–TEF (Fig. 1A); and (2) questions on controversial aspects of pure EA management (Fig. 1B).

Questionnaires were completed by 178 delegates, including 38 heads of department, 107 consultants, and 18 trainees. Of the 178 delegates, 15 did not disclose their degree.

On the 168 questionnaires, respondents reported their country of origin. A total of 131 were from 27 European countries (78%) and 37 were from 18 non-European countries (22%).

Results
Esophageal Atresia with Trachea–Esophageal Fistula
Center
Overall, 54 (31%) respondents work in centers that treat 2 to 5 EA–TEF a year, 64 (36%) respondents reported working in a center that treats 5 to 10 EA–TEF a year, and 52 (30%) respondents in a center that treats more than 10 EA–TEF a year. Only 6 (3%) respondents work in a center that treats less than 2 EA–TEF a year. A total of 119 (67%) respondents work in centers where there are only selected respondents (median, 3; range, 1–10) who operate on EA–TEF.

Preoperative Evaluation
The majority of respondents (n = 143; 81%) would request a preoperative echocardiogram. However, in case of a rightsided aortic arch, only 56% (n = 96) of respondents would opt for a left thoracotomy. The majority of respondents (n = 99; 56%) do not routinely perform a preoperative bronchoscopy, whereas 43% of respondents perform a preoperative bronchoscopy either isolated (n = 57; 32%) or in combination with an esophagoscope (n = 20; 11%).

Surgery
The preferred surgical approach for EA–TEF is open thoracotomy (n = 160; 94%), with thoracoscopy favored by a minority (n = 10; 6%). Most respondents would access the esophagus extrapleurally (n = 114; 71%); some would perform muscle-sparing (n = 70; 44%), and some would open the chest posterolaterally (n = 90; 56%). The axillary approach is performed by 24% (n = 38) of respondents.

Gap Assessment, Transanastomotic Tubes, and Chest Drains
Approximately 60% (n = 99) of respondents routinely measure the gap intraoperatively, either before (n = 49; 52%) or after (n = 30; 32%) or both before and after (n = 15; 16%) the surgical repair. The majority leave a transanastomotic tube in situ (n = 160; 90%) and routinely insert a chest drain (n = 122; 69%).

Postoperative Care and Feeding
Postoperative elective paralysis is adopted by 56% (n = 99) of respondents. Patients are maintained paralyzed for a median of 2 days (range, 1–7), to avoid tension on the esophageal
anastomosis \( (n = 57, 63\%) \), for respiratory concerns \( (n = 15; 17\%) \) or for both \( (n = 18, 20\%) \).

Before starting feeding, 72\% \( (n = 127) \) of respondents perform a routine contrast study at a median of 7 (range, 2–14) postoperative days. Only 2\% \( (n = 4) \) of respondents create a gastrostomy at the time of EA–TEF repair. Parenteral nutrition is routinely used by 54\% \( (n = 94) \) of respondents. Enteral feeding via the transanastomotic tube is started on or before postoperative day 2 by 40\% \( (n = 69) \) of respondents, on postoperative day 3 or day 4 by 35\% \( (n = 60) \) of respondents and on day 5 or after that by 25\% \( (n = 43) \) of respondents. Oral feeding is started on postoperative day 3 or day 4 by only 11\% \( (n = 19) \) of respondents, with the vast majority instead after postoperative day 5 \( (n = 156, 89\%) \). The general trend is not to perform esophagoscopy routinely \( (n = 144, 81\%) \). Postoperative follow-up of EA–TEF patients is variable: less than 1 year by 2\% \( (n = 4) \) of respondents, between 1 and 5 years by 27\% \( (n = 48) \) of respondents, between 5 and 10 years by 24\% \( (n = 43) \) of respondents, between 10 and 16 years by 26\% \( (n = 45) \) of respondents, and beyond 16 years of age by 21\% \( (n = 37) \) of respondents.

Pure Esophageal Atresia

Only 80\% \( (n = 45) \) questionnaires contained accurate data on this section of the study.

Center

About 54\% \( (n = 43) \) of respondents work in a center that treat less than 1 case of pure EA a year, 37\% \( (n = 29) \) of respondents treat between 2 and 4 cases a year, and only 9\% \( (n = 7) \) of respondents treat more than 4 cases a year.

Surgical Management

The most popular preferred management for pure EA is delayed primary anastomosis with gastrostomy formation without esophagostomy \( (n = 48, 60\%) \), followed by attempted primary anastomosis with no esophagostomy or gastrostomy \( (n = 19, 24\%) \), and delayed primary anastomosis with esophagostomy and gastrostomy formation \( (n = 13, 16\%) \). A delayed anastomosis is usually performed at a median of 3 months \( (n = 67, 86\%) \).
The favored surgical approach is open surgery \((n = 68, 85\%)\). Elongation of the ends of the esophagus is attempted by 47\% \((n = 37)\) of respondents, mainly via growth by traction (Foker technique, \(n = 16, 43\%\)), and serial dilatation with bouginage \((n = 15, 41\%)\). A minority would suture both ends to encourage spontaneous fistula formation \((n = 3, 8\%)\), fashion an esophagostomy for external elongation of the upper end \((n = 2, 5\%)\) or perform a circular myotomy of the proximal esophagus \((n = 1, 3\%)\). Approximately 67\% \((n = 51)\) of respondents always attempt an esophageal anastomosis. Approximately 23\% \((n = 16)\) of respondents perform esophageal replacement without attempt at anastomosis when the median gap is at least 5 cm \((\text{range}, 4-7)\) or four vertebral bodies.

**Esophageal Replacement**

The preferred type of esophageal replacement is gastric interposition \((n = 36, 51\%)\), followed by colonic \((n = 25, 36\%)\), jejunal \((n = 6, 9\%)\), and gastric tube \((n = 3, 4\%)\).

**Discussion**

Many aspects of the management of EA, especially in its pure form, are still lacking consensus.

Because of the rarity of the disease and the specialized assistance that it requires, once the diagnosis has been established, the infant with EA should be transferred from the birthplace to a regional pediatric surgical center. In an era of service centralization and superspecialization, our
survey reveals a trend toward selecting respondents dedicated to EA–TEF surgery, as evidenced by two-thirds of the respondents. This is in line with the recommendations by Spitz et al. to improve OA outcome, centralisation and concentration of patients are advocated particularly in case of pure OA, replacement procedures, recurrent TOF, severe tracheomalacia, and associated major cardiac defects.

Preoperative echocardiography is favored by the vast majority of respondents. This is in agreement with the UK National multicenter cohort study, where echocardiography before surgery was done in 83% of patients. However, echocardiography has been reported to correctly identify a right-sided aortic arch in only a proportion of patients. Conventional imaging in such patients is often inaccurate and leads to a thoracotomy on the same side of the aortic arch. Nonetheless, half of the respondents in our survey would opt for a left thoracotomy in case of a right-sided aortic arch. In this light, echocardiography seems to be requested more to answer preoperative anesthetic questions than to drive the surgical approach.

Although supported by some authors, the use of perioperative endoscopy still remains controversial. Our survey shows that nearly half of the respondents perform preoperative endoscopy, mainly bronchoscopy. This confirms that some respondents consider it an important procedure to detect an upper pouch fistula, whereas others do not rely on this investigation.

Although thoracoscopic repair of EA–TEF has been described more than 10 years ago, this approach is performed at only a few centers. A recent meta-analysis has confirmed the effectiveness of this approach, which seems to have outcomes at only a few centers. A recent meta-analysis has confirmed that nearly half of the respondents perform preoperative endoscopy still remains controversial. Our survey shows that nearly half of the respondents perform preoperative endoscopy, mainly bronchoscopy. This confirms that some respondents consider it an important procedure to detect an upper pouch fistula, whereas others do not rely on this investigation.

As for EA–TEF, the favorite surgical approach of the respondents for pure EA is open.

In conclusion, this survey has confirmed that there are still unsolved controversies in the management of EA, for both EA–TEF and pure EA variants.

Given the rarity of the condition and the need for homogenizing various aspects of treatment, we recommend the establishment of an EA registry. This registry would provide an accurate picture of the incidence and outcomes of EA across the participating centers, it would represent an accurate source of data to aid surveillance, and it would be a resource for research, audit, and service planning.

Conflict of Interest
None.

References
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