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A patient led, international study of long term outcomes of esophageal atresia: EAT 1

Evelyn Svoboda [a], JoAnne Fruithof [a], Anke Widenmann-Grolig [a], Graham Slater [a], Frederic Armand [a], Bernhard Warner [a], Simon Eaton [b], Paolo De Coppi [b], Edward Hannon [b,*]

[a] The Federation of Esophageal Atresia and Tracheo-Esophageal Fistula Support Groups, Sommerrainstrasse 61, 70374 Stuttgart, Germany.
[b] UCL Great Ormond Street Institute of Child Health, 30 Guilford St, London WC1N 1EH, UK

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ABSTRACT

Introduction: Long term outcomes of esophageal atresia (OA) are poorly understood. The Federation of Esophageal Atresia and Tracheo-Esophageal Fistula support groups (EAT), a collaboration of patient support groups aimed to define patient reported long term outcomes and quality of life (QoL) in a large international cohort of OA patients.

Methods: Questionnaires were designed focusing on patient/parent reported outcomes including surgical history, current symptomatology and quality of life. Members of support groups within EAT were invited to complete questionnaires electronically via SurveyMonkey®.

Results: 1100 patients from 25 countries responded to the questionnaire and 928 were analyzed. 80% had type C anatomy, 19% type A and 1% type E. Patient ages were <5 years (42%), 5–10 years (26%), 11–17 years (16%) and 18 years and older (16%). 49% of all patients reported previous dilatations which was similar across age groups. Reflux symptoms affected 58% of patients and persisted into adulthood. Dysphagia also persisted in the adult population with 50% reporting sometimes or often getting food stuck. Reflux was significantly more frequent in 'long gap' versus 'standard gap' patients ($p < 0.005$). Respiratory symptoms and chest infections decreased in frequency with age. In children median SDS for height was −0.41 (IQR −1.4 to 0.67) and that for weight was −0.63 (−1.6 to 0.67). BMI in adults was 21.5. Quality of life was described as *significantly* affected by OA in 18% of patients while 25% reported no effect on QoL.

Conclusions: These results highlight the significant long term morbidity suffered by OA patients as children and into adulthood and suggest the need for quality transitional care. The patient designed and reported nature of the study gives a unique perspective to the results and emphasizes the benefits of collaboration.

1. Background

Outcomes in esophageal atresia (OA) have improved in the last 50 years with survival now of more than 90% [1–3] leading to an increasing population of long term survivors. While our understanding of short term outcomes in infants with OA is good [4–7], longer term functional outcomes are more poorly understood with a limited research base [8–10]. There is increasing interest therefore in longer term outcome studies, and results of such studies potentially impact current practice in management of OA. This may be especially important for transitional care as patients leave the care of the pediatric surgeon for the adult world. The current literature tends to be limited to small national [4,9] and institutional datasets or meta-analysis of such studies [8]. Patient reported outcomes (PROMS), as an adjunct to reporting of outcomes by physicians and surgeons are becoming increasingly recognized as very important for improving care in a variety of conditions [11,12].

The Federation of Esophageal Atresia and Tracheo-Esophageal Fistula support groups (EAT) is an international federation of patient support groups (Table 1), led by patients and parents aiming to share the international knowledge and experience of OA, promote awareness of the condition across the world and support worldwide research and collation of information concerning the treatment and care of people born with esophageal atresia. EAT therefore has a keen interest in the short and long term outcomes of OA and has first-hand experience of the difficulties faced by many patients and carers at all times in their journey from diagnosis through transition and on into adult care. In order to gain an idea of the patient-reported outcomes in OA, EAT has conducted a patient and carer designed and led study. This paper aims to report the data from this unique study in order to provide insight into patient-reported short and long term outcomes of OA in a large international cohort of patients with OA.

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* Corresponding author. Tel.: +44 207 9052158.
E-mail address: edward.hannon@ucl.ac.uk (E. Hannon).

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Table 1
Member organizations of EAT.

Organization		Country
AFAO	Association Francaise de l'Atresie de l'Oesophage	France
ABeFAO	Association Belge Francophone de l'Atrésie de l'Oesophage	Belgium
FATE	Famiglie con Atresia dell'Esofago	Italy
KEKS	Kinder und Erwachsene mit kranker Speiseröhre	Germany
KEKS	Kinder und Erwachsene mit kranker Speiseröhre	Austria
OARA	Oesophageal Atresia Research Association	Australia
OA-Switzerland	OA-Switzerland	Switzerland
TOFS	Tracheo-Oesophageal Fistula Support	United Kingdom
TROAD	Türkiye Özofagus Atrezisi Çocuk ve Aile Destek Derneği	Turkey
VOKS	Vereniging voor Ouderen en Kinderen met een Slokdarmafsluiting	The Netherlands
AQAO	Association Quebecoise de l'Atresie de l'Oesophage	Canada

2. Methods

This study was conceptualized as a questionnaire which was then designed by the board of EAT (see author list) with support from a Professor of Quality of Care (MvD). It was designed to be a patient/carer focused study looking at outcomes of specific interest to those groups. The questionnaire was therefore designed to assess the following keys areas of interest:

1. Current symptomatology
2. Patient/carer satisfaction
3. Quality of life (QoL)

Demographics and operative details were also obtained as part of the questionnaire in order to describe the OA population surveyed. A questionnaire was then piloted within the EAT board, prepared in multiple languages (English, Dutch, German, French and Italian) and put in an electronic format using SurveyMonkey®. Invitations to participate were sent electronically to all members of 11 different European patient support groups (Table 1). This included patients and carers in 24 different countries, as members of the support groups sometimes resided outside of Europe.

Where possible, patients completed the questionnaire; otherwise this was performed by carers. Completed questionnaires were collected centrally, collated and analyzed anonymously. Initial data were presented by the Chairman of EAT at British Association of Paediatric Surgeons Congress (Cardiff, 2015). Following this the organization approached UCL Institute of Child Health for assistance in data analysis and interpretation. Formal ethics approval was not initially sought as this was a patient group led study. Approval was obtained for anonymized data analysis from ICH/GOSH Research & Development office at (project number 16DS21). Initial data were cleaned and duplicate responses excluded, together with responses where date of birth was not completed. Responses with missing or duplicate data for individual questions were included for those questions answered and percentages calculated from only completed answers.

Patient height and weight were analyzed using standard deviation scores (SDS) using LMS Growth Excel Plug In version 2.69 and UK-WHO Growth Charts in respondents younger than 18 years. Body mass index was calculated for those 18 years and older and compared to the WHO International Classification. Data were compared by Fisher's exact test or Mann–Whitney, regarding $p = 0.05$ as the cut-off for significance.

3. Results

3.1. Demographics

1100 responses were received from an estimated 2500 members of EAT organizations invited to participate. Following exclusions, 928 responses (56% male and 44% female) were analyzed with a patient age range of 1 month to 60 years. For analysis, patients were divided into the following age groups, <5 years (42% of responses), 5–10 years (26%), 11–17 years (16%) and 18 years and older (16%).

Responses were received from 25 different countries (Fig. 1). Smaller numbers of responses were also received from Canada, USA, Ireland, South Africa, New Zealand, Spain, Guadeloupe, Tunisia, Sweden, Malta, Lithuania, Israel, Hungary, Hong Kong, Greece and Denmark in decreasing order of number of responses. 121 (18%) of responses were completed by patients and 761 (82%) by a parent or carer.

OA with tracheoesophageal fistula (OA-TOF) was the most common defect, reported in 80% of cases, 19% were pure OA and 1% an H type fistula. Participants reported their defect as 'long gap' either as supplementary to reporting the anatomy of their defect or as an isolated diagnosis in 18% of cases. Similar distribution of type of defect was seen in all age groups although there were a significantly higher proportion of pure OA and long gap participants in the adult age group with 13% of cases being pure OA in participants younger than 18 years versus 23% in the adult cohort ($p = 0.004$).

3.2. Surgical care

Overall, the median length of hospital stay following initial surgery was 6 weeks (range 1 to 174 weeks). However, considering only the respondents older than 18 years, the median length of stay was 8 weeks (IQR = 16). When compared by diagnosis, those with OA-TOF had a median LoS of 6 weeks (IQR) versus 8 weeks for those with pure OA (IQR). Long gap cases reported that median LoS was 18 weeks (IQR) versus 5 weeks (IQR) for standard gap OA. Fifty-three percent (0–5 years), 46% (5–10 years) and 52% (11–17 years) of participants in the younger age groups respectively only required 1 operation to repair their defect, whereas in the 18 year and above group only 36% were repaired with a single operation and 45% needed 2–5 procedures.

The proportion of patients receiving dilatation following repair is summarized in Fig. 2. Forty-nine percent of all respondents had esophageal dilatation and the number of dilatations received was broadly similar across each age group. If dilatation was performed the median number was 4 dilatations and was the same for all age groups.

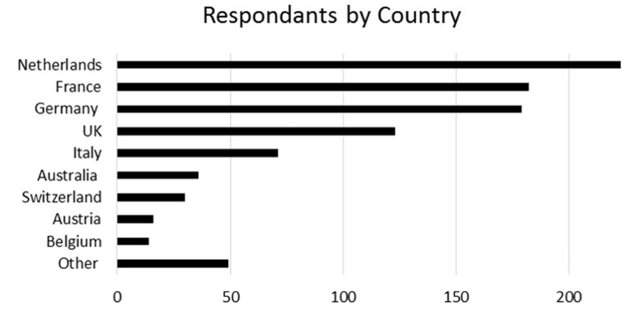

Fig. 1. Respondents by country.