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Patient-driven healthcare recommendations for adults with esophageal atresia and their families

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ABSTRACT

Background: Adults with esophageal atresia (EA) require a multidisciplinary follow-up approach, taking into account gastroesophageal problems, respiratory problems and psychosocial wellbeing. Too little is known about the full scope of these individuals' healthcare needs. We aimed to map all medical and psychosocial needs of adults with EA and their family members, and to formulate healthcare recommendations for daily practice.

Methods: A qualitative study was performed, using data from recorded semi-structured interviews with two focus groups, one consisting of adult patients with EA (n = 15) and one of their family members (n = 13). After verbatim transcription and computerized thematic analysis, results were organized according to the International Classification of Functioning, Disability and Health. Ethical approval had been obtained.

Results: Healthcare needs were described through 74 codes, classified into 20 themes. Most important findings for patients included the impact of gastrointestinal and pulmonary problems on daily life, long-term emotional distress of patients and parents and the need of a standardized multidisciplinary follow-up program during both child- and adulthood.

Conclusion: The focus groups revealed numerous physical and mental health problems, as well as social difficulties, that require attention from different healthcare providers. We have formulated several healthcare recommendations that physicians may use in long-term follow-up.

Abbreviations

COREQ	= COnsolidated criteria for REporting Qualitative studies
EA	= esophageal atresia
GER	= gastroesophgeal reflux
GP	= general practitioner
ICF	= International Classification of Functioning, Disability and Health
ISCED	= International Standard Classification of Education
PTSD	= posttraumatic stress disorder
QoL	= quality of life

Type of Study: Treatment Study
Level of Evidence: Level III

1. Introduction

Advancements in treatment strategies have led to increased survival rates of newborns with esophageal atresia (EA). Subsequently, more children with this rare congenital malformation nowadays reach adulthood. Many adults with EA, however, still experience sequelae: gastrointestinal symptoms such as dysphagia or gastroesophageal reflux (GER) [1,2]; respiratory problems such as wheezing, coughing or lung function abnormalities; or impaired exercise capacity due to pulmonary problems [3]. Therefore, we have ex-

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tended our hospital's longitudinal multidisciplinary follow-up program for these children with transition to adult healthcare [4].

An important element of follow-up is psychosocial wellbeing. In the past, the quality of life (QoL) of adults with EA has been assessed with different questionnaires. Overall, a normal health-related QoL was reported on the general SF-36 questionnaire [5–7]. However, when focusing on gastrointestinal symptoms with the Gastrointestinal Quality of Life Index, or on pulmonary complaints with the Respiratory Symptoms-Related Quality of Life Index, impaired QoL was reported [7–10].

The currently available literature does not sufficiently describe the healthcare needs of adults with EA. Questionnaires are either too broad or too specific, and do not address the specific problems encountered in daily life. Moreover, certain aspects have never been studied adequately such as mental problems or the impact on relationships. Recently started endoscopic surveillance programs for Barrett's esophagus and esophageal cancer [11] have brought new burdens, adult relationships could raise concerns about heredity, and parent-child relationships could be affected by events in the past. The impact of having a child with EA on parents has only been described for school-aged children [12]. The long-term effects of EA on mental health and family relationships have never been published.

It is recommended that patients with EA are followed up by physicians with expertise in EA care [13]. Thus far, too little is known about the full scope of healthcare needs of EA adults. Patient experience data are considered meaningful for healthcare improvement in terms of safety and effectiveness [14]. In focus groups, thoughts and feelings can be elicited by promoting self-disclosure among its participants [15]. The aim of this study was to map all medical and psychosocial healthcare needs of adult EA patients and their family members, resulting in the formulation of recommendations to be used in daily practice.

2. Patient and methods

This is a qualitative study using in-depth data from semi-structured focus group interviews addressing the worries, needs and preferences of EA adults and their family members. Approval from the institutional ethics review board had been obtained (MEC-2019–0160). The COREQ (COnsolidated criteria for REporting Qualitative studies) checklist was used as a reporting framework [16].

2.1. Participants

Two focus groups were composed, aiming at 10–15 participants as recommended [15]; one consisting of adult patients with EA, the other of their family members and/or partners. Patients were randomly selected from all adults with EA currently participating in our follow-up program [11], using an online tool (www.randomizer.org) [17]. Patients were invited to participate through a personal letter, and were asked to invite their partner or a family member of their choice as well. Inclusion criteria were age $\geq$18 years, born with EA or a partner/family member of a patient with EA and Dutch-speaking. Written informed consent for interview recording was obtained from all participants.

2.2. Data collection

An expert team was formed for this study, consisting of a gastroenterologist (MS) and a pulmonologist (LK), both specialized in treating adults with EA; a pediatrician (SG), who is the coordinator of the standardized longitudinal follow-up program for children with congenital anomalies in our hospital and involved in the transition of children with EA to adult healthcare; a neuropsychologist (AR) with experience in moderating focus group interviews; an adult patient with EA; a representative of the Dutch patient association (JF) and a researcher (CtK). Prior to the focus group interviews, the expert team met to determine the interview topics, based on literature research and clinical experience.

Both focus group interviews were conducted on June 20th 2019, within the framework of a national symposium for adults with EA. The interviews were moderated by a male neuropsychologist (AR). After a brief introduction, the topics were introduced and with open questions participants were stimulated to discuss their worries and needs (see Supplementary File 1). The interviews were recorded audio-visually and transcribed verbatim. Additionally, participants were asked to fill out a questionnaire on baseline characteristics and, if applicable, physical complaints.

2.3. Data analysis

Transcripts were imported into the qualitative software ATLAS.ti 8.3.20 (Scientific Software Development GmbH, Berlin, Germany). Following the steps of thematic analysis [18], the transcripts were reviewed and coded by two members of the research team (CtK and DL) independently and systematically. Initial codes covered the basic element of a text fragment, and were modified or merged during the analysis. Codes from both transcripts were combined into overarching themes. All codes and themes were reviewed by a third investigator (AR) and discussed until consensus was reached. The expert team reviewed the themes and supplemented clinically relevant subjects where necessary.

Next, the themes were structured according to the International Classification of Functioning, Disability and Health (ICF) [19], which describes five health-related domains: body functions and structures, activity, participation, personal factors and environmental factors. Results are described in a qualitative manner and illustrated by quotes extracted from the interviews (see Table 1). Quotes were translated from Dutch to English by forward-backward translation by a native speaker to validate consistency of the translation.

Educational levels were classified according to the International Standard Classification of Education (ISCED) [20]. Descriptive data for the baseline characteristics were generated using SPSS V.24.0 (IBM, Chicago, Illinois, USA).

3. Results

3.1. Participants

In June 2019, our follow-up program contained 195 adults with EA, of whom 55 had been randomly selected. Three patients were excluded because of intellectual disability. Therefore, 52 were invited for this study. Thirteen invitees did not respond, and 24 invitees refused because of lack of time or transportation. Thus, fifteen patients consented to participate. Eleven of them brought a family member; one brought two family members. In total, 28 participants were included: 15 patients and 13 family members (10 parents, 2 children and 1 partner). Based on the distribution of the baseline characteristics and physical complaints, we considered this sample representative for the EA population (see Table 2 and Supplementary Table 1).

The interview with the patient group lasted 62 min; that with the family member group 67 min. Thematic analysis identified 74 codes which could be classified into 20 themes (see Table 3). Eventually, no new codes could be identified, suggesting data saturation.

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Table 1
Extracted quotes from the focus group interviews. ICF = International Classification of Functioning, Disability and Health, GP = general practitioner.

ICF classification	Theme	Quote
Body functions and structures	Physical problems	*"My mother was raised very protected. As a child, she was told that it was better not to exercise. Nowadays, she has a worse exercise capacity than her peers."* – daughter, 43 years old
	Mental health problems	*"My father is what I would call traumatized. Whenever I talk about it, he starts crying."* – patient, 32 years old
Activity & participation	Obstacles in daily life	*"Going to a restaurant is different for me than for others. I have to be careful with my choices."* – patient, 54 years old
	Social difficulties	*"With my first girlfriend, I pretended to have already eaten at home. I did not want to offend her parents by not being able to eat their food."* – patient, 21 years old
	Limitations in employment	*"One time, there was not enough time to eat, after which I started vomiting. Since then, they give me the time I need."* – patient, 21 years old
Personal factors	Feeling guilty	*"I'm an only child because I needed a lot of care. My father would have liked more children. Now that I have two healthy sons of my own, I feel like he has finally gotten the sons he always wished for."* – patient, 37 years old
Environmental factors	Specialized healthcare	*"Our GP told us that we should call the hospital if we had questions. He did not want anything to do with it."* – mother, 62 years old
	Follow-up and transition to adult healthcare	*"The transition to the adult hospital was awful. I could no longer stay with my child all day."* – mother, 64 years old
	Impact on family relationships	*"You go through a very intensive period together. For us, we grew as a couple. Whatever will come, we can handle it."* – father, 55 years old

3.2. Patient perspectives

3.2.1. Physical and psychosocial problems

Childhood was characterized by frequent hospitalizations due to pulmonary infections or upper endoscopies for esophageal dilatations or stuck food boluses. The main problem in adulthood was dysphagia. All patients could eat solid food but had to drink water with every meal. Other major physical complaints were pain, postprandial bloating, coughing, impaired lung capacity and poor exercise capacity. Anti-reflux surgery at childhood because of GER has led to new forms of discomfort such as an inability to vomit. Coughing caused a poor night's sleep for some patients. Some patients are still afraid to visit the hospital or to undergo medical procedures.

3.2.2. Worries about the future

Pregnancy was stressful for patients due to concerns about heredity and the unborn baby's health. Endoscopic screening of the esophagus raised concerns about Barrett's esophagus. Some patients had not visited the hospital for 40 years. Older patients never had a proper explanation about their condition, and were happy to be finally informed.

3.2.3. Obstacles in daily life

Eating had the greatest impact on patients' everyday life. They needed more time to finish a meal than their peers, and always had to consider the type of food they ate. This made certain activities challenging, e.g. restaurant visits, a quick meal on the street or buffet meals. Some patients could not eat or drink anything in the evening, because this led to severe heartburn overnight. Some older patients were limited in their daily activities due to a poor exercise capacity. As a child, they were advised not to go outside in the winter or play sports, due to increased susceptibility to respiratory infections.

3.2.4. Social difficulties

Patients received negative comments about EA-related situations, e.g. when a stuck food bolus needed to be pushed through. Patients with coughing complaints were often unfairly criticized for smoking.Some older patients had large scars, whereas most of the younger patients had hardly visible scars. A possible coping strategy at school age was giving a talk about EA to their classmates.

The extent to which patients were open about having EA varied. In partner relationships, some patients had not informed their partner until having children was discussed. In friendships, some patients did not want to bother friends with their story. In general, patients kept their explanation short when informing people.

At work, the main problem was the mealtime. Patients needed more time to finish their lunch than co-workers. Some did not get enough time from their boss; others found it hard to take the time they needed, feeling guilty for letting their work pile up.

3.2.5. Effects on personal life

Overall, patients became more resilient. Some still had trouble letting people get emotionally close due to hurtful comments or experiences from their youth. Others were full of fighting spirit, and would not give up easily. Patients felt guilty towards their siblings and parents as they grew up, for being born with EA and receiving so much attention.

Patients clearly stated that they no longer feel like a patient, or even found it annoying to be called a patient. Some started feeling like a patient again when they received the endoscopic surveillance invitation. Patients wished for medical identification to carry with them in case of emergency, e.g. a choking incident or a stuck food bolus.

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Table 2
Baseline characteristics of the focus group participants. Data are presented as median (range) or n (%). One patient and two family members have not filled out a questionnaire. EA = esophageal atresia, ISCED = International Standard Classification of Education, VACTERL = vertebral, anorectal, cardiac, tracheoesophageal, renal or limb anomalies. [A] According to Gross classification (2). [B] Birth weight <10th centile (3). [C] According to Solomon criteria (4). * Five patients were 20–30 years old, six patients were 30–40 years old, three patients were 50–60 years old and one patient was 71 years old.

	Patients (n = 15)	Family members (n = 13)
Age (years)	32.6 (20.8–71.0) *	63.5 (22.7–67.1)
Male	10 (66.7)	5 (38.5)
Relationship to EA patient		
Parent of EA patient		10 (76.9)
Child of EA patient		2 (15.4)
Partner of EA patient		1 (7.7)
Educational level		
Low (ISCED 0–2)	4 (28.6)	4 (36.4)
Middle (ISCED 3–4)	2 (14.3)	3 (27.3)
High (ISCED 5–8)	8 (57.1)	4 (36.4)
Type of EA [A]		
Type A	1 (7.7)	
Type C	11 (84.6)	
Type E	1 (7.7)	
Gestational age in weeks	38.72 (32.0–43.0)	
Birth weight in grams	3100 (1465–3600)	
Preterm birth	4 (26.7)	
Small for gestational age [B]	4 (26.7)	
Staged repair	2 (13.3)	
VACTERL association [C]	2 (13.3)	
Dysphagia score (1)		
Grade 0	11 (78.6)	
Grade 1	3 (21.4)	

3.2.6. Transition to adult healthcare

Patients remembered the pedagogical staff guiding them through unpleasant procedures or preparing them for surgery. Younger patients often had multiple check-ups, but older patients had not received any follow-up after their first year of life. They were surprised being invited for a check-up after several decades. Most patients did not know what EA entailed until this check-up. Patients appreciated the possibility of follow-up. It resulted in a better understanding, for example their pulmonary complaints finally fell into place.

Patients notice that general practitioners (GPs) lack medical expertise on EA. They expressed the desirability of designating one coordinating physician. Usually, the gastroenterologist was in charge, with which patients were satisfied.

3.3. Family perspectives

3.3.1. Psychosocial problems

All parents could vividly describe memories from the first years of their child's life. Parental anxiety was widespread during the perinatal period and remained over the years. Some parents had been told that the child would not survive. Parents were especially worried during pulmonary infections or choking incidents. Most parents still suffered from emotional problems, some parents even were severely traumatized by such experiences.

3.3.2. Social difficulties

Parents received negative comments as well, e.g. when they gave their child medication in public. Finding a babysitter was difficult for parents. People were afraid to look after their child and to administer medicines, for example.

3.3.3. Effects on personal life

Some parents found their child more sensitive or emotional than peers. Parents felt guilty towards their other children because it was difficult to divide attention. They also felt guilty towards themselves for neglecting their own health sometimes when their child was sick.

3.3.4. Specialized healthcare

Most parents had negative experiences with giving birth at regional hospitals due to the lack of medical expertise. At the academic hospital they felt reassured and got a satisfying explanation about the diagnosis, surgery and what to expect. Most common frustrations during hospitalizations were limited visiting hours or postponement of surgery. Another frustration was the lack of knowledge of the GP, who for example did not take the pulmonary problems of their child seriously. Parents could call a direct phone number from the hospital if they had any questions.

Parents emphasized the importance of receiving timely and proper information. When their newborn was transported to an academic hospital, often the mother stayed behind and remained uninformed about the condition or prognosis of her child for several days.

Parents experienced insufficient support from the hospital in the first period. They missed a professional to talk to. Although they turned to other sources of support – such as keeping a journal, or talking to family, friends or other parents on the ward – this was felt not enough compensation. For parents of younger patients, transition to adult healthcare was a major step.

3.3.5. Impact on family relationships

For parents, especially the first years were hard. Early parent-child bonding was difficult. Due to travel distances and lack of transportation, parents sometimes could not visit their child for several weeks. After a long and intensive hospital period, discharge home was an enormous transition. Regular maternity care that new parents usually receive was no longer available. Later, parents struggled between their roles as parent and caregiver because they had to give their child medication, tube feeding, or parenteral feeding.

The sick child represented also a large burden on the relationship between parents; sometimes resulting in divorce, sometimes strengthening the relationship. Parents felt it was difficult to remain strict in the child's upbringing. Parents had a hard time letting go of the child when it left home at adult age.

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Table 3
Overview of identified themes and corresponding codes. ICF = International Classification of Functioning, Disability and Health, EA = esophageal atresia, GP = general practitioner.

ICF classification	Themes	Codes
Body functions and structures	Physical problems (past)	Dilatations, eating disorder, food getting stuck, fundoplication, tracheoesophageal fistula, variable course of treatment, chocking incidents
	Physical problems (present)	Barrett's esophagus, vomiting, comorbidities, exercise capacity, food getting stuck, no complaints at all, general health, pulmonary problems, pain when eating, frequent hospital visits
	Mental problems	Anxiety of parents, anxiety of patients, worries, memories, optimism, home birth
	Worries	Heredity, reassurance
Activity & participation	Obstacles in daily life	Alcohol, exercise capacity, pulmonary problems, gastroesophageal reflux
	Sleeping	Sleeping
	Eating	Frustrations about eating, restaurant visit
	Illness	Pulmonary problems, frequent hospital visits
	Exercise	Sports
	Appearance	Scars, looks
	Social contacts	Insecurities of environment
	Maintaining relationships	Impact on relationship and family, relationships
Personal factors	Coping	Coping, resilience
	Feeling a patient	Feeling a patients, wish for medical identification
	Guilt	Feeling guilty as a child, feeling guilty as a parent
Environmental factors	Expertise in healthcare	Relationship with treating physician, need for specialized care, frustrations about healthcare, satisfied about healthcare, guidance in regional hospital, lack of expertise in regional hospital, positive experience with GP, negative experience with GP, late diagnosis, home birth, explanation by parents
	Support	Somebody to talk to, contact with fellow peers, pedagogical healthcare, support network, trauma processing
	Screening, follow-up and transition	Prenatal screening, follow-up childhood, transition to adult healthcare, surveillance program
	Family relationships	Parents involved during adulthood, parent-child bonding, parents as caregivers
	Interaction with environment	Frustrations about work, telling people about EA, getting attention from environment

4. Discussion

This study is the first to provide an overview of the medical and psychosocial health status of adults with EA and their family members. The patient-driven data from focus group interviews with patients and family members of different ages gives insight in the impact of the disease over time, which may help to optimize medical care and psychological guidance.

4.1. Body functions and structures

Various symptoms have been described in adolescents and adults with EA, such as regurgitation, heartburn, aspiration and dysphagia [21]. In our study, dysphagia was the main complaint. Dysphagia is often caused by delayed esophageal clearance due to disturbed motility [22]. Surprisingly, GER was not addressed as a problem. A likely explanation is that all patients in this study are followed by a gastroenterologist, and have been given anti-reflux medication if needed. . From our own experience, we know that patients often will not bring up GER complaints themselves as they do not experience these as symptoms [11]. Hence, it is important to actively ask patients about this during follow-up.

In line with the literature, patients reported pulmonary complaints and poor exercise capacity. Previous studies described significantly more respiratory symptoms and infections in EA adults compared to controls ($p \leq 0.002$). Pulmonary function tests showed both obstructive, restrictive and combined lung disease [21, 23, 24]. Unfortunately, recent data on large cohorts is lacking. Nevertheless, considering the patients' experiences revealed in our study, we recommend that every adult with EA should be referred to a pulmonologist specialized in EA to optimize lung condition.

The poor exercise capacity of older patients (>50 years old) was striking. We know from previous studies that children with EA are at risk for decreased exercise tolerance [25–28], possibly influenced by diminished physical activity as a child, which may be partially due to parental anxiety. A standardized follow-up program – which was not yet available during the childhood of these older patients – will allow intervention at an early stage. Adults with EA suffer from impaired performance capacity as well [29]. Still, pulmonary rehabilitation may improve exercise tolerance [30]. This emphasizes the importance of extending the multidisciplinary care approach for EA into adulthood.

Despite increasing pedagogical guidance in the last decades, hospital anxiety remained present even among younger patients. The relationship between preoperative anxiety and postoperative anxiety and sleeping problems in children is well known [31], but the long-term effects of undergoing multiple procedures – like in EA – have not been studied.

Likewise, in adults with EA anxiety associated with the endoscopic surveillance program should be considered. Given the relatively new nature of this program, this has not yet been investigated. In other surveillance programs, endoscopies were reported as burdensome, with elevated anxiety levels beforehand [32]. Participating in a follow-up program might cause problems in getting mortgages or insurance since it emphasizes the chronicity of EA. The attitudes of banks and insurance companies in this respect are still unknown.

Patients worried more around the fertile age and throughout pregnancy. Currently, non-syndromic EA is considered to have a multifactorial cause [33] with a recurrence risk of 2–4% for offspring [34]. However, many pathophysiological mechanisms still remain unclear. In our hospital, a geneticist is involved in the transition to adult healthcare. We strongly suggest personalized genetic counselling when there is an active child wish, preferably before pregnancy.

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Table 4
Healthcare recommendations to be used in daily practice when addressing adults with EA or their parents during childhood. EA = esophageal atresia, GER = gastroesophageal reflux, GP = general practitioner, PTSD = post-traumatic stress disorder.

	Problem	Screen for	Provide
Adult patients	Gastrointestinal problems	Dysphagia, coping strategies, pain or discomfort during/after the meal, GER	Inform about EA related problems Explain esophageal motility rather than stenosis Prescribe anti-reflux medication if necessary
		Risk for Barrett's esophagus and esophageal cancer	Inform about the risk and surveillance possibilities Refer to gastroenterologist for endoscopic surveillance program
	Pulmonary problems	Coughing, pulmonary infections, lung capacity, asthma	Inform about EA related problems Refer to a specialized pulmonologist (computed tomography scan, lung function tests)
		Exercise capacity	Start pulmonary rehabilitation, advise about sports/exercise
	Mental health problems	Anxiety for the hospital or medical procedures	Refer to psychologist if necessary Point out possibility of peer support through patients association
		Pregnancy-related worries	Refer to a clinical geneticist
	Social participation	Problems with telling people about having EA	Help patients find the right explanation Provide information material Provide medical identification (see Fig. 1)
		Problem with emotions of connecting to people	Refer to psychologist if necessary Point out possibility of peer support through patients association
	Economic participation	Problems longer lunch breaks or taking a personal day, concerns about career	Help patients find the right explanation Provide information material Provide medical identification (see Fig. 1)
		Concerns about mortgage or insurance	Provide information material Contact with patients association
	Specialized healthcare	Coordination of multidisciplinary follow-up Lack of knowledge GP	Appoint one coordinating physician, preferably the gastroenterologist Inform healthcare provides about EA and EA related problems through folders, symposia or scientific journals.
Parents	Mental health problems	PTSD, feeling guilty	Be aware of mental complaints during hospitalization and follow-up Offer professional support during hospitalizations Refer to psychologist if suspicion of PTSD Point out possibility of peer support through patients association
		Disturbed parent-child interaction	Possibility of staying with their child around-the-clock
	Social participation	Reactions/fears from environment, finding a babysitter, getting comments when giving medication	Offer help of a social worker Point out possibility of peer support through patients association
	Specialized healthcare	Understanding diagnosis and prognosis Coordination of multidisciplinary follow-up Lack of knowledge GP or regional hospital	Explain condition as soon as suspected Appoint one coordinating physician, preferably the pediatric surgeon Inform healthcare provides about EA and EA related problems through folders, symposia or scientific journals.

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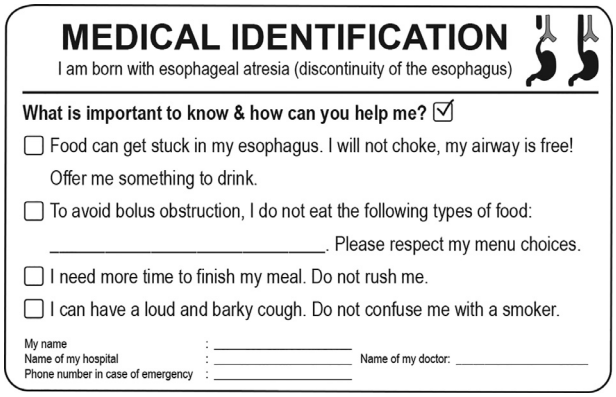
MEDICAL IDENTIFICATION

I am born with esophageal atresia (discontinuity of the esophagus)

What is important to know & how can you help me? ☑

☐ Food can get stuck in my esophagus. I will not choke, my airway is free!
Offer me something to drink.

☐ To avoid bolus obstruction, I do not eat the following types of food:
_____. Please respect my menu choices.

☐ I need more time to finish my meal. Do not rush me.

☐ I can have a loud and barky cough. Do not confuse me with a smoker.

My name : _____
Name of my hospital : _____ Name of my doctor: _____
Phone number in case of emergency : _____

Fig. 1. Example of a medical identification for patients with esophageal atresia. Patients can adjust, fill out and print this card themselves.

4.2. Activities and participation

Although all patients functioned autonomously, certain food-related activities remained difficult. Coping strategies (e.g. drinking water) prevent major limitations in daily life. EA could potentially influence one' working career. Sick leave, longer lunch breaks or taking days off for hospital visits may lead to potential career limitations [35]. Proper explanation and educational material such as brochures can be supportive in explaining EA to other people. Herein lies an imported role for healthcare providers and patient associations.

It is noteworthy that some patients found it offensive to be labelled as patients because they feel healthy. Healthcare providers should keep this in mind when addressing this population. Interestingly, patients expressed a wish for medical identification. A credit card-sized pass (see Fig. 1) that can be adjusted, filled out and printed by each individual patient might fulfill this need.

4.3. Personal and environmental factors

Lack of medical expertise is the main frustration for both patients and parents. Parents must be informed as soon as possible about their newborn's condition, even when the baby is not born at an academic hospital. Standardized follow-up programs might better ensure that patients and parents adequately understand all aspects and consequences of EA. This should be verified – and if necessary clarified - at transition to adult healthcare.

The follow-up for different specialties should by coordinated by one designated physician. A recent patient-led survey study found that half of the surveyed EA adults had no current healthcare provider [36]. In our opinion, the gastroenterologist would be best qualified as coordinating physician for adult patients. It is his responsibility to inform the GP about what EA implies and how GPs can anticipate to specific problems that patients can encounter.

Being separated from their child due to travel distances or lack of transportation was traumatizing for parents. Previous research acknowledged that hospitalization of a newborn can disturb the parent-infant relationship and attachment [37]. Today, in our hospital parents can stay with their child around-the-clock, in line with the family-centered care strategy that has been associated with improved outcomes [38]. We suggest all centers to offer this in order to promote parent-child bonding.

Although the results of this study are not sufficient to draw a conclusion on posttraumatic stress disorder (PTSD), parents showed multiple symptoms: re-experiencing traumatic events, avoiding certain situations and getting overly emotional [39]. A study among parents of school-aged children with EA reported PTSD in more than half of the parents, and increased levels of anxiety [40]. Similar results were found for parents of children with other congenital anomalies [41]. Feeling guilty – as parents in our study described – could be a possible risk factor for long-term PTSD [42].

One could wonder if enough attention is paid to trauma stressors during hospitalization [43]. It is recommended to provide sufficient information about support resources at discharge, including contact details of patient associations and primary care providers [44]. Also, professional psychological support may be offered to parents during initial hospitalization and follow-up, with awareness of the strain parents might experience because of their dual role as parent and caregiver.

4.4. Strengths and limitations

To our knowledge, this is the first qualitative study addressing the needs and worries of both adults with EA and their family members. Nowadays, patient-centered care and patients' perspectives become more and more important. Patient-driven data provides new insights that quantitative research cannot provide, such as persistent hospital anxiety or how to address these patients. The widespread age of the participants can be considered as both a strength and limitation. It represents the population on one hand, but complicates the interpretation of the results on the other hand. Moreover, using the ICF classification to measure health and disability enabled us to identify the consequences for daily life. Despite the small sample size, data saturation suggests sufficient quality of the data. However, given the nature of this study, a quantitative analysis of the data was not possible.

Some limitations should be addressed. First, since patients were selected from our follow-up program in a tertiary hospital with a response rate of 29% (15 out of 52 patients), results may be influenced by a selection bias of well-informed and assertive participants. Second, although the focus groups were characterized by a safe atmosphere, some patients could have been reluctant to share particular feelings or concerns. Third, topics about childhood relied on memories, which could be less accurate for parents of older patients. Next to this, the Netherlands is a small and high-income country with well-organized healthcare. In contrast to many countries, home birth is common. Last, qualitative research is explorative and does not aim to represent the entire population. These are all facts that should be taken into account when extrapolating our results worldwide.

5. Conclusions

This qualitative study gives a unique insight into the healthcare needs of adults with EA and their families. The focus groups revealed numerous physical and mental health problems and social difficulties, that require attention from different healthcare providers. Our findings therefore emphasize the importance of a structured, long-term, multidisciplinary follow-up program for these patients. We have formulated several healthcare recommendations that physicians may use (see Table 4).

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Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.jpedsurg.2020.12.024.

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