Esophageal atresia: Long-term morbidities in adolescence and adulthood

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Abstract

Survival rates in esophageal atresia patients have reached 90%. In long-term follow up studies the focus has shifted from purely surgical or gastrointestinal evaluation to a multidisciplinary approach. We evaluated the long-term morbidity in adolescent and adult esophageal atresia patients and discussed mainly non-surgical issues. Dysphagia is common and reported in up till 85% of patients. In young adults gastro-esophageal reflux disease occurs frequently with development of Barrett esophagus in 6% reported in different series. It is difficult to estimate respiratory morbidity from the literature because many different definitions, questionnaires, and study designs have been used. However, many patients seem to suffer from respiratory problems even into adulthood. In conclusion, morbidity is not only restricted to surgical problems but many different domains are involved. These are all related and together determine to a large extent the quality of life of EA-patients and also of their families. We assume that a multidisciplinary care approach seems best to address their special needs.

Keywords: esophageal atresia, long-term morbidity, follow up studies, adolescent, young adult

Introduction

Improved intensive care treatment and surgical techniques have raised survival rates in esophageal atresia (EA) to 90%[1,2]. For many years, EA outcome research mainly focused on evaluation of surgical techniques and functional gastrointestinal morbidity, i.e. esophageal motility, dysphagia, and gastrointestinal reflux disease (GERD). Long-term follow up studies are mainly cross-sectional and in small study populations. The past few years have seen a shift in focus to multidisciplinary evaluation of EA patients. This review addresses the long-term morbidity in adolescent and adult EA patients with an emphasis on non-surgical issues. Based on current knowledge recommendations will be made for follow-up beyond infancy and childhood.

Gastrointestinal morbidity

Dysphagia is the most reported gastrointestinal problem in older EA patients. The pathogenesis depends on the type of EA correction and remaining anatomy. For example, an end-to-end anastomosis may be a source of swallowing problems as a result of abnormal motility with coordination problems between the proximal and distal parts of the esophagus. Furthermore, intestinal (e.g. colonic) interposition may lead to stasis of food in the “neo-esophagus” but is otherwise usually favorable for these patients[3,4]. Most dysphagia studies concern groups of patients

with different type of esophageal atresia, operative correction, and age at follow-up. Besides, dysphagia is assessed in different ways: Some studies administer questionnaires on feeding behavior[5-7]; others apply esophageal manometry[8-11]. Recent long-term follow up studies in EA-patients reported dysphagia complaints in 30-85% of adolescents and adults[5,6,8,11,12].

GERD is another frequently reported symptom in EA beyond childhood. Several cross-sectional studies have reported either subjective complaints[3,5,7] or outcomes of objective assessment (pH-monitoring, endoscopy, histological examination of esophageal biopsies)[8,11,13]. Rintala and coworkers summarized the results of 6 different studies on long-term morbidity in EA patients[2]. Overall, symtoms of GERD were reported in 45% of adolescent and adult patients; esophagitis in 53%; and Barrett esophagus in 6%[2]. In a recent study 5 out of 37 adolescents (13.5%) reported GERD symptoms in a reflux questionnaire[7]. This was 35% in the series of 57 adolescent type III EA patients[6]. Sistonen and coworkers studied 101 EA survivors aged 21 to 57 years: esophagitis was present in 25% and Barrett esophagus in 6%[11]. A relatively new phenomenon reported in case series is the presence of eosinophilic esophagitis in EA patients and its possible association with dysphagia and GERD[14].

Respiratory morbidity

Respiratory tract infections (RTI) are common within the first years of life[15] but respiratory problems at adolescent and adult age have been reported as well, especially in patients with corrected tracheo-esophageal fistulas. In addition, anomalies of the tracheobronchial tree, shown in EA patients, may have a role[16]. It is, however, difficult to obtain reliable figures on the prevalence of respiratory symptoms from the current literature. Study populations are small and different methods are used to assess respiratory morbidity. RTI are usually thought to have occurred if antibiotics were prescribed. However, objective information such as chest X-rays is usually lacking and respiratory symptoms might well have reflected viral infections with barky cough and mucociliary clearing difficulties as a result of airway malacia. Bias may occur in retrospective study designs. While recurrent RTI at adolescent and adult age have been reported in 32-52% of patients[6,15,17,18], pneumonia was diagnosed on X-ray in no more than 5%[15].

In questionnaires, 40-60% of adolescent EA patients reported respiratory symptoms[6,12]; 12-40% of adolescent and adult patients reported recurrent wheezing episodes or docter-diagnosed asthma[12,15,17-19].

Lung function abnormalities include obstructive, restrictive, or combined obstructive and restrictive patterns[6,18,20]. Thoracotomy-induced rib fusion and reflux-associated problems are considered major

risk factors for restrictive ventilator defects[18]. Future studies comparing lung function in EA patients following thoracoscopic repair are needed to elucidate whether restrictive lung function occurs less frequently following thoracoscopy. A first study in infants showed similar lung volume after thoracoscopic and after thoracotomic repair of EA[21].

From the present data it is unclear whether airflow obstruction is caused by small airway disease, by proximal obstruction due to airway malacia, or by epithelial damage caused by GERD and recurrent infections. Sistonen and coworkers reported a prevalence of atopy in adults following EA repair that was comparable with that of the Finnish general population[18]. From previous studies in younger children it is known that EA-patients are at risk for decreased exercise tolerance[22-24]. However, data on exercise tolerance in adolescent and adult EA survivors have – to our knowledge – never been published.

Physical growth

Limited data on physical growth are available, especially in adolescent and adult EA patients. Somppi and coworkers described normal physical growth in the majority of patients aged from 3.5 to 30 years (mean 12.6 years); height was below -2 SD in 5% of patients[17]. Mild growth retardation has been reported in school-age children with EA[23,24]. In 15 of 22 adolescents aged 14-18 years we found normal physical growth (68%) (unpublished data). Sistonen reported normal body mass index in adult EA patients[11].

Psychosocial well-being and school performance

Several studies have evaluated quality of life in adolescent and adult EA-patients. Legrand and coworkers showed that children with EA have a lower health-related quality of life than healthy peers. Their scores were, however, higher than those of children with chronic disease. Patients who were born prematurely, or had symptoms of GERD or barky cough had the lowest scores[6]. Adolescents studied by Peetsold and coworkers scored similar as the reference population in most domains but significantly lower in the domain of general health perception[7]. This group had significantly higher scores on the family activities scale. General health perception was negatively associated with GERD-symptoms[7]. A small group of adolescent EA-patients reported a significantly higher total competence score compared with the norm population as they scored better on self-esteem, social acceptance, and physical appearance[25]. Birth weight and dilatations of the esophagus were significant predictors of mental health and psychosocial well-being[25]. In adult patients only those with dysphagia complaints and disturbed esophageal motility had impaired quality of life, measured with a generic instrument (SF-36). Patients with GERD symptoms, however, had normal

quality of life scores. It might well be that EA patients who grew up with GERD may have become accustomed to these symptoms[8]. Health-related quality of life is generally normal in adult EA survivors; in 15% of them morbidity from gastrointestinal or respiratory disorder may impair quality of life[26]. Stam and coworkers used a questionnaire to retrospectively assess risk behavior and the achievement of developmental milestones. They concluded that the course of life of young adults grown up with esophageal atresia was not delayed compared with that of peers[27].

Data on school performance in EA patients are scarce and contradictory. In small groups of Dutch[28] and Norwegian[25] children and adolescents 22% and 33%, respectively, followed special education. Unfortunately, the indication for special education is unclear. Members of a parent support group in Germany reported attendance of a special-needs school in 11%[29]. In the study of Peetsold and coworkers more than half of the adolescent EA patients attended secondary school on the highest educational level[7]. We have a similar experience in a cohort of 14-18 year old EA-patients (unpublished data). In the study of Deurloo and coworkers 48% of adult EA patients had completed advanced high school or obtained a university degree and only one patient had finished education at a primary school level[8].

Conclusions

Increased survival rates in EA-patients result in more long-term morbidity. Morbidity is not only restricted to surgical problems but many different domains are involved, e.g. gastrointestinal morbidity, respiratory problems, and physical growth. These are all related and together determine to a large extent the quality of life of EA-patients and also of their families. Although some symptoms, such as respiratory problems, seem to decrease with age[20] other problems persist beyond childhood and have even been reported in adulthood. Besides, new problems may arise in adulthood: Barrett's esophagus and esophageal cancer are serious medical problems that may call for lifelong monitoring of EA-patients[11,30,31].

Recommendations and future perspectives

For EA-patients a multidisciplinary care approach seems best to address their special needs. This approach should aim for optimal treatment of GERD, prevention and adequate treatment of respiratory problems, as well as appropriate dietary management to manage feeding difficulties and to provide sufficient caloric intake for optimal physical growth. Early recognition of motor function problems and decreased exercise tolerance is important to prevent further deterioration that may lead to physical inactivity. All these factors may contribute to an increased health performance and quality of life. The available data on long-term morbidity strongly indicate that multidisciplinary care

should be continued during adolescence and even into adulthood. The advantages of multidisciplinary teams have been outlined by Levesque[32] and include not only advantages for the patients but for their families as well. In our institution we offer a multidisciplinary follow-up program until 18 years of age (Figure) evaluating physical health, development, quality of life and parental wellbeing. At the age of 18 years the adolescent is transferred to a dedicated adult gastroenterologist for repeated endoscopies at least every five years.

Since current knowledge on the long-term morbidity in EA-patients is mainly based on cross-sectional studies in (usually) small series, it is important to initiate a prospective registration of data from longitudinal standardized evaluations by multidisciplinary care teams. In addition, clear definitions are needed (e.g. to describe recurrent RTI). The use of standardized questionnaires including scoring systems for symptoms such as dysphagia or GERD may be helpful. However, we have to keep in mind that EA-patients may get used to symptoms (acid reflux, barky cough) and consider these as normal. Therefore, follow-up should also include objective evaluation of morbidities by medical professionals.

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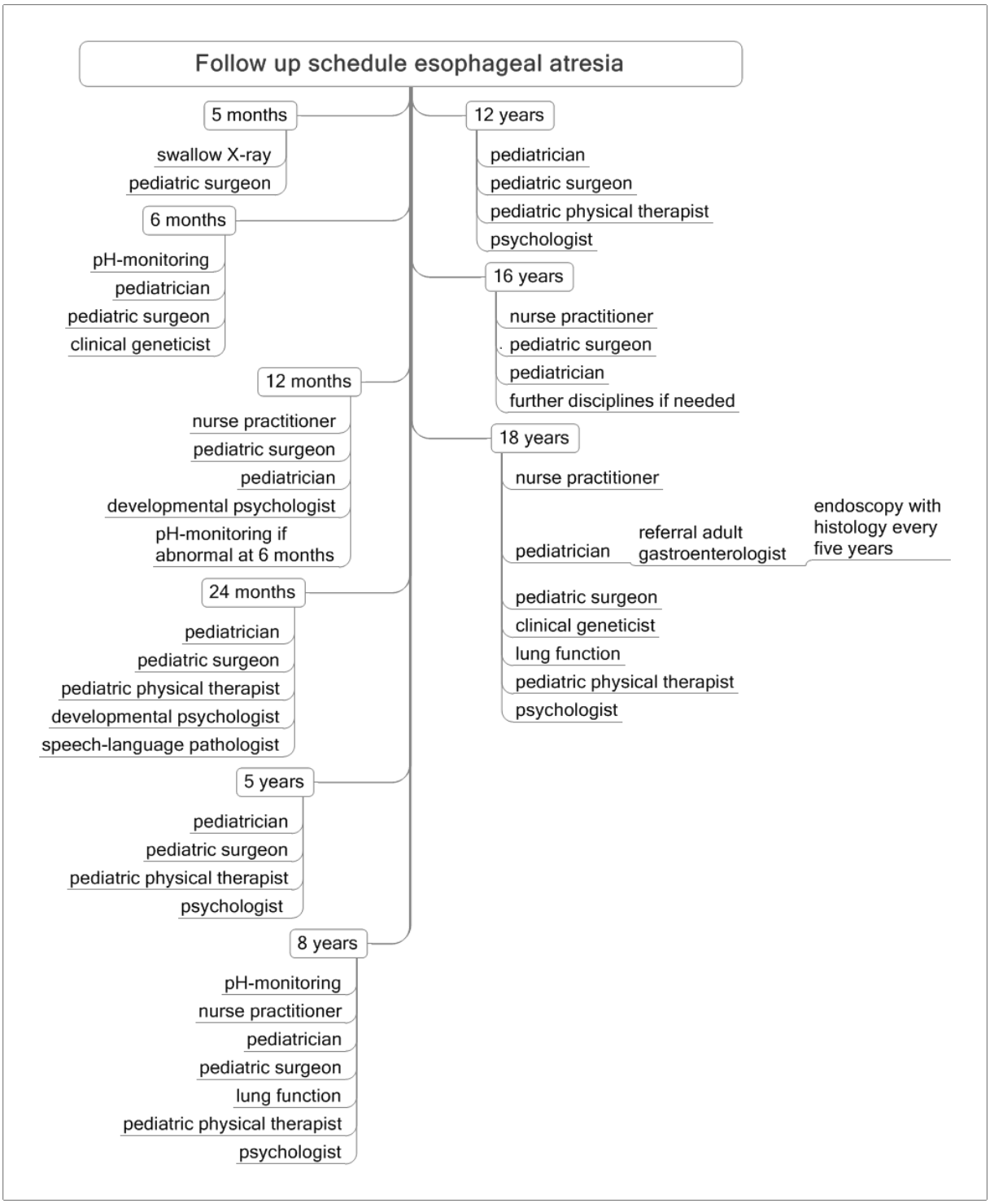

Figure Legend

Standardized multidisciplinary follow-up schedule of EA-patients treated in Erasmus MC – Sophia Children's Hospital in Rotterdam, The Netherlands.