EMBJ.ORG FOR YOUNG DOCTORS

EUROMEDITERRANEAN BIOMEDICAL JOURNAL
2018,13 (11) 057–059
(*FORMERLY: CAPSULA EBURNEA*)

Case Report

ESOPHAGEAL ATRESIA AND FOOD DISORDERS: A CASE REPORT.

Manuela Capozza [1], Maria Elisabetta Baldassarre [1], Domenico Martinelli [2], Davide Ferorelli [3], Antonio Di Mauro [1], Nicola Laforgia [1].

1. Department of Biomedical Science and Human Oncology. Neonatology and Neonatal Intensive Care Unit. University of Bari – Italy.
2. Section of Neonatology and Neonatal Intensive Care. Ecclesiastical authority "Miulli" Regional General Hospital. Bari – Italy.
3. Interdisciplinary Department of Medicine. Section of Legal Medicine. University of Bari – Italy.

ARTICLE INFO

Article history:
Received 31 October 2017
Revised 06 February 2018
Accepted 29 March 2018

Keywords:
Esophageal atresia; food disorders; pediatric surgery.

ABSTRACT

Esophageal atresia is a relatively common congenital anomaly, which is often associated with other anomalies. We report a case of a newborn affected by esophageal atresia with significant post-operative and long-term complications, including feeding and swallowing disorders.

1. Introduction

The esophageal atresia (EA) encompasses a group of congenital defects of the continuity of the esophagus with or without a tracheoesophageal fistula. The etiology is not known and is probably multifactorial. This anomaly is the most common significant esophageal malformation, with an incidence of 1:2500 live births. The first successful primary repair of EA was performed by Cameron Haight in 1941. EA is frequently associated with other congenital defects, and it could be part of the CHARGE syndrome (coloboma, heart defects, atresia choanae, retarded neuro-development, genital hypoplasia, ear abnormalities) or of the VACTERL syndrome (vertebral (V), anorectal (A), cardiac (C), tracheoesophageal (TE), renal (R), and limb (L) defects), first reported by Quan and Smith. Both syndromes represent a non-random association of congenital defects with unknown etiology and pathogenesis. However, EA could be associated to many other abnormalities not visible or clinically relevant, such as the absence of the vagus nerve, an abnormal intrinsic innervation of the esophagus and tracheobronchial tree, or malformations of the lung lobes [1,2]. The EA is typically characterized by the interruption of the anatomy of the upper third of the esophagus, shaped as a blinded sac located behind the membranous trachea. A tracheo-esophageal fistula (TEF) is frequently present. Less often, the EA is not associated with TEF and, in such cases, the distal esophagus is reduced to a small pouch crossing the diaphragmatic hiatus ending in the lower mediastinum. The most widely used EA classification is Vogt classification (1929), modified by Ladd and Gross (1953), which takes into account the seat of atresia and the presence of tracheal fistula. Regarding the embryogenesis, some theories have been proposed, including vascular insufficiency, abnormal growth of the esophageal epithelium and mesenchyme, abnormal notochord, and involvement of the neural crest cell[3]. The probability of suspecting the EA in a fetus increases if a polyhydramnios is found, or when the stomach bubble is either absent or reduced. The insertion of a nasogastric tube at birth should be done in all infants born from mothers with polyhydramnios, but final diagnosis requires a standard chest and abdomen X-ray with a feeding tube in place, into which air is injected. Contrast should be avoided because of the risk of air inhalation. In cases of EA, the tube ends in the cervical or high thoracic site. The presence of air in the gastro-intestinal tract indicates the presence of a fistula (type D – Gross classification). Definitive treatment includes closure of the tracheo-esophageal fistula and primary anastomosis of the esophagus. According to the gap length between the two esophageal segments, the surgeon decides which type of surgical correction to implement. In only about 10% of all cases of EA is the gap

* *Corresponding author*: Manuela Capozza, manuela.capozza@uniba.it
DOI: 10.3269/1970-5492.2018.13.11
 ISSN: 2279-7165 - Available on-line at www.embj.org

considered too long to perform the anastomosis (long gap) and, in these situations, the surgical treatment remains controversial. Options for the reconstruction of the esophagus include the use of native esophagus or replacement of the esophagus with the stomach, colon or small intestine. However, since the extension of the native esophagus gives the best functional results, every effort must be made to preserve the native organ[4]. Thanks to advances of both pediatric surgery and neonatal intensive care, the chances of survival for neonates with EA are excellent. Infants weighing over 1.5 kg and without heart problems have a survival rate of almost 100%, whereas the survival rate decreases if other risk factors are present. Today, the presence of cardiac abnormalities is more important than low birth weight. The EA French national register found that, out of a population of 307 patients with EA born in 2008 and 2009, 34% of the cases had medical complications (8% anastomotic leaks, 4% new tracheoesophageal fistula and 22% stenosis of the region of the anastomosis, with the need of endoscopic dilation, with an average of 2 dilations/patient). A new hospitalization was required for 59% of patients (2.5 hospitalizations/patient) because of feeding (52%) or respiratory (48%) problems. About 12% of patients required surgery for gastro-esophageal reflux (GER) after about 164 days (range 33-398 days) and 1% experienced an aortopexy for tracheomalacia. About 15% of patients were malnourished at 12 months of age, 37% had respiratory symptoms, while 15% had dysphagia during follow-up. The ability to feed the patients by mouth at discharge was associated with a reduced risk of complications ($p = 0.007$)[5].

2. Case report

We report the case of a late-preterm infant (35+5 weeks of gestational age) born by caesarean section with a prenatal suspect of EA. At birth, chest x-ray confirmed the presence of type D (Gross classification) EA.

On the third day of life, a termino-terminal esophageal anastomosis with fistula repair was done. After surgery, minimal respiratory assistance was required. The baby was exclusively parenterally fed for 9 days, than he received milk by nasogastric tube and then by mouth.

During the follow-up, at one month of life, the endoscopic evaluation detected an esophageal stenosis due to a tracheal cartilaginous ring, so a second intervention was needed. Enteral feeding was initiated seven days after surgery with a nasogastric tube, and then by bottle, after 28 days, when the baby was 73 days old. Frequent episodes of desaturations, after or concurrent with the feedings, developed. Despite several changes of the milk type and of the duration and of the number of feedings, parenteral nutrition was needed for 44 days.

Endoscopy showed no evidence of re-stenosis or tracheal-esophageal fistulas. Esophageal impedance-pH monitoring was negative for gastroesophageal reflux disease. Bronchoscopy revealed a tracheomalacia secondary to a vascular ring attributable to an aortic arch, for which an aortopexy was performed.

After this third intervention, the baby did very well; he showed very good suction with proper sucking-swallowing coordination and no desaturations. He progressively tolerated increasing milk quantities and, after 4 weeks, he was sent home.

3. Discussion

Getting back to a normal feeding modality is one of the main issues in children with EA after surgical correction. Dysphagia could be due to motility disorders, anatomical lesions, obstruction and inflammation of the esophagus. Endoscopic evaluation, manometry and esophagogram could be needed for the diagnosis. Salivogram fluoroscopy, fiberoptic endoscopic evaluation of swallowing and high-resolution manometry estimate the percentage of risk of aspiration of gastric contents. This latter situation can be alleviated by the use of food thickeners and changing the frequency of meals, transpyloric feeding, or fundoplication[6].

A Canadian study investigated eating behaviors of children with EA through a questionnaire filled out by the attending physician during the follow-up. The extremely pre-term infants, and those with EA different from type 3 (Vogt classification), had the greatest difficulties in feeding, which requires a careful exploration of risk factors in order to allow for prompt intervention[7].

The possible association of EA with other gastrointestinal tract malformations, such as hypertrophic stenosis of pylorus, should also be considered. The incidence of hypertrophic stenosis of pylorus is 30 times higher in EA compared to normal populations, and children with recurrent or persistent vomiting after surgical correction of EA should be evaluated for this disease. The reason for the higher rate of this association requires further research[8].

The main goal of medical care is to prevent any abnormal feeding with a pre-emptive plan to help children at different stages of development. In order to stimulate adequate sucking and swallowing, protocols of "sham feeding" were tested on patients undergoing a delayed surgical correction and they proved to shorten the times for complete recovery and effective enteral feeding. Another very important aspect is a normal mother-infant relationship, which can be strained due to very long periods of hospitalization[9].

Discussions and meetings with families of children born with EA revealed that, regardless of the outcome of the surgical procedure and any subsequent treatment, some children may develop a sudden food refusal. At the origin of this food aversion, there could be the traumatic experience of the child related to the multitude of medical events that can affect nutrition behavior (i.e. intubation, gastroesophageal reflux, examinations)[10].

Our case report underlines the need for a complete pre-operative investigation followed by continuous post-operative vigilance, because the unusual association of “hidden” congenital anomalies should be considered.

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