EXCEPTIONALLY LATE RECONSTRUCTION OF THE ESOPHAGUS FOR CONGENITAL ATRESIA

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In this case report, the treatment history of a woman with congenital esophageal atresia is presented. She underwent thoracotomy immediately after birth, but the distance between two atresic parts of the esophagus (7 cm) did not allow for a primary anastomosis. Gastric and salivary fistulas were constructed. A salivary fistula was made in the cervical part of the esophagus. For 21 years, the patient received nutrition only by gastrostomy. Her physical and mental conditions were normal. She underwent esophageal reconstruction of the last segment of the ileum, coecum, and ascending colon and began oral feeding when she was 21 years old.

**Key words:** congenital esophageal atresia, surgery

Congenital atresia originates from disorders of primary digestive tube division into the esophagus and the trachea between 3 and 6 weeks of life (1). There are two types of the abnormality: complete and partial atresia of the esophagus with or without concomitant fistula to the trachea (fig. 1). Clinical variants of this abnormality include esophageal atresia with the fistula to the trachea from the upper part of the esophagus, the fistula from the lower part of the esophagus, or fistulas from both parts of the esophagus. The fistula of the H type is a variant of the last abnormality without esophageal atresia.

Surgery is the only possible treatment option for congenital atresia of the esophagus and should be considered as quickly as possible (2). Type of surgical procedure depends on the kind of abnormality and general condition of the child. The most important risk factors for negative events during surgery are low body weight, inflammatory lesions in the lungs and especially other life threatening abnormalities, in particular cardiac defects (prognostic criteria according to Watson, Spitz; Montreal classification) (3). The distance between two ends of an atresic esophagus plays a key role in choosing an operative tactic.

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Fig. 1. Types of congenital esophageal atresia
A gap of 2 cm is considered to be an indication for making the primary anastomosis. A distance between 4 and 6 cm is estimated to be a limitation of possibility of the primary reconstruction of esophageal continuity. If the gap between the two ends of the atresic esophagus excludes the possibility of or increases the risks of making the primary postponed anastomosis, esophageal reconstruction remains the only option for restoration of its continuity (4).

Esophageal reconstruction is possible even in infants, although better results are achieved in older children. In many centers, the minimum weight required for patients undergoing esophageal reconstruction is 10 kilograms. Age between 6 and 12 months is considered the most suitable for reconstruction, but there are also surgeons who opt for reconstruction in children older than 18 months or between the age of 2 and 3 years (5, 6). The child is fed by gastrostomy until that time, and a salivary fistula created in the cervical portion of the esophagus provides for outflow of saliva.

CASE REPORT

I.G. (history number 1308/06), a woman born on April 6, 1985, was admitted to our clinic for the first time on November 6, 2006. She was a child born on 37-th week of gravidity with no clinical complications, in particular without hydramnios. She weighed 3300 grams and measured 54 cm. The general condition of the child immediately after labor was given the score of 9 points on the APGAR scale. Obstruction of the esophagus was confirmed during examination. The child underwent emergent right thoracotomy (fig. 2). The gap between the two ends of the esophagus measured about 7 cm, and it was not possible to make a primary anastomosis. Gastrostomy was performed after Kader method and a salivary fistula in the cervical part of the esophagus were constructed. The postoperative course was not complicated. There is no information in the documentation of the patient’s history to indicate that there was a tracheal fistula, although during nursery and school age she suffered from bronchitis several times and once had pneumonia. The physical and psychological progress of the child was normal. At the age of 4, she weighed 12.5 kg and measured 93 cm. She finished primary and than secondary school at the appropriate ages. She led a normal life in a group of peers.

Despite this, she had still not been recommended for esophageal reconstruction.

On admission to our clinic she was 21 years old, weighed 51 kg, and was 154 cm tall. She was in good general health. On physical examination, she was diagnosed with right rotation scoliosis of the thoracic part of the vertebrae that caused deformation of the thorax. Spirometry results were as follows: VC 81.3%, FVC 80.4%, FEV1 79.7%, PEF 46.7% of normal values. The salivary fistula was not big, with no inflammatory reaction of the skin, but the gastrostomy canal of 5 cm diameter was surrounded by a large inflammatory infiltration of the skin caused by constant leak of gastric juice despite the use of protective dressing around the wound. Under radiological examination with swallowed contrast medium, a portion of the esophagus reaching the level of Th2 was visualized. On this level, the contrast medium ran out of the natural esophagus. The contour of the esophagus was smooth, and its width measured 3 cm. After administration of the contrast medium via the gastrostomy site, the stomach was revealed to be in a normal location, and no lesions in its walls were found. The bulb and the duodenum loop were normal. Passage of the contrast medium through the small intestine was good, and the medium reached the coecum after 1.5 hours (fig. 3A, 3B). Other ad-
ditional examinations were normal. No other abnormalities were diagnosed.

The patient was recommended to undergo esophageal reconstruction. Her operation was on November 17, 2006. We chose a medial abdominal approach, liberated multiple adhesions, and excised the previously constructed gastrostomy in the prepyloric area. The gastrostomy was recreated in the fundus according to the Witzel method. The patient’s unfavorable vessel system did not allow for construction of the esophageal replacement of the jejunum or ileum. The new esophagus was mobilized from the end portion of the ileum, the coecum, and the ascending colon and was supplied by the medial and right colonic vessels. The esophageal replacement was located in the isoperistaltic position behind the sternum, and the side of the replacement was anastomosed with the end-side of the natural esophagus. The distal end of the reconstructed esophagus was connected with the stomach. Continuity of the digestive tract was reconstructed by anastomosing the ileum with the colon. No complications appeared during the short postoperative course.

Oral feeding was started on the 7th postoperative day and the gastrostomy was closed. The patient left hospital on the 13th postoperative day. She could eat by mouth without any difficulty. On December 12th she was examined and because she continued with oral feeding without any problems (body weight 51.7 kg) she had the drain removed out of the gastrostomy, which resulted in spontaneous closure of the canal (fig. 4). She underwent another examination in out-patient clinic in March 2007. Good functioning of the reconstructed esophagus was confirmed clinically and in radiological examinations; the oral and pharyngeal phases of swallowing were good in spite of
DISCUSSION

The most common congenital abnormality of the esophagus is esophageal atresia with concomitant distal esophago-tracheal fistula. This type of defect occurs in nearly 85% of cases. A well-developed superior part of the esophagus with good vessel supply is characteristic of this defect. In such cases, mobilization of the upper portion of the esophagus is safe. The lower portion of the esophagus, however, is significantly smaller and thinner. It runs in the direction of the trachea, becoming narrow in the area of their connection, in the location of the tracheal bifurcation (6). The blood supply of this lower portion is much poorer, and one must be careful while mobilizing it. A space between the fistula and upper segment of the esophagus in a majority of cases allows for the creation of an anastomosis of the end-to-end type, without any tension (3, 4, 5, 7).

In the case of the presented patient, atresia of the esophagus appeared without an esophago-tracheal fistula. This type of the malformation is rarer and occurs in 7% of cases. The malformation is characterized by a well-developed upper segment of the esophagus and a short, poorly developed lower part of the esophagus. The gap between atresic parts of the esophagus is so long that most often it is not possible to make either a primary or a postponed anastomosis (8).

Long gap atresia of the esophagus most commonly is associated with isolated types of atresic esophagus. It is a serious medical problem, which results in the impossibility of making a primary anastomosis, the most favorable solution from an anatomical and physiological point of view. A postponed anastomosis renders a necessity of use of techniques extending the upper part of the esophagus; however those methods may not be successful and might result in postponing the definitive operation and making the reconstruction multistage. When there is no possibility of performing either a primary or postponed anastomosis, esophageal reconstructive procedures are an option (9). These procedures include: transposition of the stomach to the posterior mediastinum (10), creating a tube flap of the greater curve (located iso- or antiperistaltically) (11), reconstruction of the entire esophagus with the small or large intestine with feeding pedicle (12), or the recently popular small bowel free graft (13).

Regardless of the surgical method chosen for reconstruction, the treatment should be finished in childhood, even if it is prolonged to nursery age. Allowing a patient to live with salivary fistula until adulthood and feeding him through a gastrostomy may cause disorders in his physical or psychological development and is also traumatic for a child’s caregiver. In the case of the presented patient, esophageal reconstruction was performed at the age of 21. It is worth mentioning that, besides the thoracic scoliosis and deformity of thorax triggered by thoracotomy on the first day of life without subsequent rehabilitation, her physical development was normal. However, there is no doubt that this disability of being able to eat orally has had a deep impact on her psyche.

We hope that the present progress that has been made in pediatric surgery and the use of classic as well as endoscopic and thoracoscopic methods (14, 15) will eliminate such long periods of waiting for esophageal reconstruction in children with congenital esophageal atresia.
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Received: 11.05.2007 r.
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