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# Atresia of the esophagus - thoracotomy vs thoracoscopy

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## Abstract

Esophageal atresia is a congenital medical condition associated with the disorder of the alimentary tract. It is usually associated with one or more fistulas to the trachea. Esophageal atresia is often associated with other congenital defects. The most frequent anomalies with atresia of the esophagus are those that occur in the VACTERL association. The incidence of oesophageal atresia mention in the literature varies 0.7-4.55/10000, because of the different percentage of defects other than esophageal atresia and associated with esophageal atresia changes. The etiology of esophageal atresia is mainly unknown, but it is considered

multifactorial, including genetic and environmental factors. Indeed, 6-10% of cases of esophageal atresia have been diagnosed with a chromosomal abnormality or syndrome.

For the purpose of this study, we analyzed the current papers on esophageal atresia found in the PubMed database.

Traditionally, the esophageal atresia has been operated by the right posterior thoracotomy. The first thoracoscopic repair of classic esophageal atresia was performed in 1999, and the first successful thoracoscopy of the tracheo-oesophageal fistula a year later. Together with these milestones, numerous health centers have begun adapting this surgical technique. Although thoracoscopic surgery in the case of esophageal atresia in patients with tracheo-bronchial fistula was conducted in many highly developed children's surgery centers, the safety and efficacy of this method remained controversial. The benefits of thoracoscopic surgery are obvious, including excellent visualization, less use of post-operative drugs and cosmetic effects. Jaureguiza et al. described the "scaly scapula", chest wall deformity, scoliosis and the development of the bad nipple in patients who underwent open surgery due to esophageal atresia with the accompanying tacha-oesophageal fistula. In the case of open thoracotomy, it was necessary to withdraw the lungs to expose the posterior mediastinum, resulting in lung damage and respiratory complications.

Compared to the open surgery, thoracoscopy significantly reduced the time of hospital stay and the moment of the first oral meal. However, thoracoscopy was associated with a longer time of surgery. The incidence of leaks, narrowings, pulmonary complications, and blood loss were similar in both surgical technique. It seems that the benefits of thoracoscopy are significant.

### Introduction

Esophageal atresia is often associated with other congenital defects. The incidence of 10,000 cases of esophageal atresia published in the literature varies from 0.7 in Oxford in the United Kingdom (Rankin et al., 2005) to 4.55 in Mainz, Germany (working group Pedersen, Calzolari, Husby, Garne and EUROCAT, 2012). Range of results is changing because of the percentage of defects other than esophageal atresia but associated with it.

This percentage ranged from 36.8 in Boston (Greenwood and Rosenthal, 1976) to 81.9 in Hawaii (Forrester & Merz, 2005). Differences in literature were also significant in the percentage of chromosomal and non-chromosomal as well as heterogeneous congenital anomalies associated with esophageal atresia. Anomalies associated with atresia of the esophagus include: anomalies of the cardiovascular system, musculoskeletal system, urinary tract, gastrointestinal tract and central nervous system.

The most frequent anomalies with atresia of the esophagus are those that occur in the VACTERL association (V - vertebral abnormalities, A - Anorectal malformations, C - Cardiovascular anomalies, T - tracheo-esophageal fistula, E - esophageal atresia, R - Renal (Kidney) and/or radial anomalies, L - limb defects) (de Jong et al., 2008). They may also accompany CHARGE syndrome (C - Coloboma of the eye, central nervous system anomalies) H - heart defects A - choanal atresia R - Retardation of growth and/or development G - Genital and/or urinary defects (Hypogonadism, undescended testicles, besides hypospadias.) (E) - Ear anomalies and/or deafness and abnormally bowl-shaped and concave ears, known as "lop ears". The etiology of esophageal atresia is mostly unknown, but it is considered multifactorial, including genetic and environmental factors (de Jong, Felix, et al., 2010). Indeed, between 6% and 10% of cases of esophageal atresia have been diagnosed with a chromosomal abnormality or syndrome, and in about 10% of cases of esophageal atresia, associated anomalies matched the VACTERL compound. [1,2]

Esophageal atresia is a congenital disorder associated with the disorder of the esophageal discontinuity. It is usually associated with one or more fistulas to the trachea. There are five recognized types of esophageal atresia, based on the presence and location of the fistula associated with atresia. Type A isolated atresia of the esophagus without fistula, type B atresia of the esophagus with the proximal tracheoesopharyngeal fistula, type C atresia of the esophagus with the distal tracheoesopharyngeal fistula, type D atresia of the esophagus with the duble fistula, type E tracheo-oesophageal fistula without atresia. [3] Type C is the most frequent and constitutes about 85% of cases. Esophageal atresia is a rare congenital anomaly (1 in 2,500 to 1 in 4,500 live births), with a slight predominance in men.

Most children with this condition require surgical repair in the first few days of life. Including division and ligation of esophago-tracheal fistulas, primary anastomosis or prolongation esophagus, including possible anastomosis in patients with a wide separation between the proximal and distal segment of the esophagus. In 1941, when surgical treatment was successful for the first time, the mortality rate was high.

Over the years improved surgical technique and intensive treatment have resulted in reduced mortality. As more and more children are successfully operated due to esophageal atresia and reaches adulthood, we should considerate the long-term effects and complications of esophageal atresia. Both from the disease itself and from surgical intervention. Gastrooesophageal and respiratory disorders are rare, including gastroesophageal reflux disease (GERD), intestinal metaplasia, recurrent esophago-tracheal fistulas, aspiration and recurrent lower respiratory tract infections. [4]

It's possible to begin diagnostics already in the prenatal period. During ultrasonography we can notice polyhydramnios, occurring in about 50-95% cases of congenital esophageal atresia. This symptom is the result of an abnormal circulation of the amniotic fluid, also the inability to swallow fluid through the fetus. Other symptoms in the ultrasound image of the fetus suggesting atresia of the esophagus is invisible stomach or its small size. The diagnostic value of these two prenatal ultrasound symptoms is about 20-56%. Newborns with prenatal suspicion of esophageal atresia after childbirth need to be implemented urgent examination to exclude or confirm the defect. [5,6]

Prenatal diagnosis of esophageal atresia without tracheo-esophageal fistula is difficult, with false positive results of 78%. Reports from the literature note that polyhydramnios is the most sensitive to prenatal diagnosis of esophageal atresia; however, because it also occurs in other disease entities, it is characterized by low specificity. In addition, a small or absent stomach may be absent when at the same time duodenal atresia; in this case an enlarged stomach will be detected, giving the previous two results a total of 85% of the positive predictive value. Finally, it was found that the presence of the upper pouch signifying the enlargement of the upper esophagus with a blind end has a 100% positive predictive value. [2]

Magnetic resonance imaging (MRI) and biochemistry of amniotic fluid were included in the diagnostics with the suggestion that these are second-order diagnostic tools that help confirm the diagnosis in the prenatal period. Indeed, they help to detect the esophagus pocket probably more reliably than ultrasound, and gamma-glutamyl transpeptidases (GGTP) and alpha-fetoprotein (AFP) in amniotic fluid show a higher level in the case of esophageal atresia. [7]

Traditionally, the esophageal atresia has been operated from the right posterior thoracotomy. The open surgery of the esophageal atresia with tracheo-esophageal fistula is based on the isolation of the fistula, removal of the upper capsule, activation of the lower capsule and completion of the anastomosis. The first thoracoscopic repair of pure esophageal atresia was performed in 1999, and the first successful thoracoscopy of tracheo-oesophageal fistula a year later. Together with these milestones, numerous health centers have begun operate by this surgical technique. Although thoracoscopic surgery in case of esophageal atresia in patients with tracheo-bronchial fistula was conducted in many highly developed pediatric surgery centers, the safety and efficacy of this method remained controversial. [8]

The advantages of thoracoscopic surgery are obvious, including excellent visualization and dissection of posterior mediastinal structures, less use of post-operative drugs and cosmetic

effects. In 1985, Jaureguiza et al. described chest wall deformity, scoliosis and the development of malignant nipple in 89 patients who underwent open surgery due to esophageal atresia with accompanying tracheo-oesophageal fistula and have been watching it for more than 3 years. In the case of open thoracotomy, it was necessary to displacement the lungs to expose the posterior mediastinum, resulting in lung damage and respiratory complications [9,10].

Compared to the toracotomy, thoracoscopy significantly reduced the time of hospital stay and the moment of the first oral meal. However, thoracoscopy was associated with a longer time of surgery. The incidence of leaks, narrowings, pulmonary complications, blood loss was similar in both surgical approaches. [8.11]

The main risk factors for developmental disorders include: birth weight and heart defects. Moreover, after surgery treatment of esophageal atresia, the most common complications were: GERD, as well as problems with swallowing food. [12]

Esophageal atresia is not only a disorder of infant. Symptoms also occur at a later age, mainly: reflux 58% of patients and persisting in adulthood. Also dysphagia maintain in adults. Symptoms of the respiratory system and chest infections have decreased with age. The long-term care for adults seems to be essential, in particular the supervision of Barrett's esophagus and early esophageal cancer. Further research is needed to determine which patients and at what age anti-reflux surgery is the most beneficial. [4.13]

The incidence of swallowing disorders is estimated at 82% and significantly exceeds the incidence of esophageal stenosis (39%), indicating that not all patients with swallowing disorders have a mechanical problem, but are likely to have functional dysphagia. Therefore, parents and over time, patients should be informed about the probability of dysphagia, its nature and related eating habits. They should also be calmed down and informed that swallowing disorders are often mild, mainly because of hard food and have no effect on the quality of life or daily activities. Secondly, not all patients with swallowing disorders have associated effects (Stenosis or GERD), and if appropriate, the esophageal dysfunction should be assessed. [14]

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