Seminars in Pediatric Surgery (2009) 18, 50-56

ELSEVIER

SEMINARS IN PEDIATRIC SURGERY

Outcome of esophageal atresia beyond childhood

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KEYWORDS
Esophageal atresia;
Esophageal dysmotility;
Esophagitis;
Gastresophageal reflux;
Long-term outcome;
Metaplasia

Survivors of esophageal atresia are reaching their adulthood in large numbers for the first time enabling assessment of true long-term outcome among this group of patients. This review summarizes the current knowledge on the subject focusing on late symptoms and complications, esophageal pathology and pulmonary function. Relationships between esophageal dysmotility, gastroesophageal reflux, esophagitis and epithelial metaplastic changes including esophageal cancer are outlined. In addition to pertinent literature, institutional experience, and follow-up of patients with esophageal atresia for more than 60 years is included.

The overall survival rate of patients with esophageal atresia (OA) has increased significantly during the last decades, and today exceeds 90% of all patients.[1] Today, practically all patients without concomitant severe malformations survive, and the very high mortality of very low birth weight patients and patients with severe cardiac malformation has decreased significantly. Many surviving patients, however, continue having functional problems, especially from their gastrointestinal tract and respiratory system after the initial postoperative period.[2] The overall impact of these problems on the long-term outcome of OA patients is not fully understood. Moreover, it is unclear what impact postnatal postoperative, usually relatively long hospitalization time, and repeated anesthesias for anastomotic dilations in many patients may have on later psychosocial well-being of the patients.

There are a limited number of reports concerning long-term outcome of patients with OA. The eventual quality of life appears to be good in most adults.[3,4] This assumption is based on some recent studies that have evaluated late sequelae of OA mainly by validated questionnaires. These questionnaires fail to detect subtle functional differences and may ignore the fact that OA patients have had abnormal esophageal function from birth and may not experience functional aberrations as symptoms at all.

This review tries to summarize the outcome of OA beyond childhood. The focus will be in late esophageal and pulmonary function, late surgical complications, and complications such as persistent gastroesophageal reflux that may predispose to later malignancy. In addition to data from literature, preliminary results of institutional late follow-up of nearly 100 adult survivors following OA repair are used in this communication.

Late clinical symptoms

Dysphagia is one of the most common symptoms that OA patients in all age groups suffer from. It is likely that dysphagia is associated with innate esophageal dysmotility that is associated with OA.[5,6] The role of surgical dissection and anastomosis in the development of dysphagia is unclear but may be significant. Most patients with dysphagia have no anastomotic strictures, although these have to be ruled out in patients with significant symptoms. Dysphagia is commonly associated with food impaction requiring endoscopy during childhood; the peak incidence is between 1 to 5 years.[7] Food impaction requiring surgical measures is

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1055-8586/$ -see front matter © 2009 Elsevier Inc. All rights reserved.
doi:10.1053/j.sempedsurg.2008.10.010

much less common in adults despite persistence of dysphagia. The reported incidences of dysphagia are between 10% and 60%.[8,9] This variance is caused by definition of dysphagia. If mild symptoms, such as the need to drink fluids to facilitate swallowing of meals and occasional swallowing problems, are included, almost two-thirds of adult patients can be considered to have dysphagia.[9,10] In our institutional review, more that 80% of adults with OA had to use excessive liquids during eating or restrict their diet to foods that are less prone to get stuck in the esophagus.

Regurgitation and other symptoms suggesting gastroesophageal reflux (GER) are more or less as common as dysphagia. The reported incidences of reflux symptoms in preadolescent children and adults are between 27% and 75%.[8,10-13] However, reflux symptoms are also common in the general population. Some authors have found that the incidence of reflux symptoms in adults with repaired OA is only marginally more frequent than in the general population and the difference is not statistically different.[14] On the other hand, it is likely that patients with repaired OA have had reflux from birth and may not recognize milder symptoms as pathologic. Either way, it appears that subjective symptoms of reflux are often mild enough not to interfere with daily life.

Chest wall deformities occur in a significant number of patients with repaired OA.[15] Typical findings include scapular winging, anterior chest wall deformity, and scoliosis. All of these can be caused by surgical complication, scoliosis also by congenital vertebral anomies. The incidence of scapular winging has been reported to be around 20%.[16] The incidence of scapular winging could be diminished by careful preservation of long thoracic nerve during thoracotomy. Thoracotomy and especially multiple thoracotomies are the leading cause for chest wall asymmetry in OA patients. Damage to the innervation of serratus anterior muscle may contribute to the chest wall deformity. The incidence of chest wall asymmetry has been reported to be as high as 25%.[15,17,18] In our adult series, the overall incidence of secondary deformities following neonatal right thoracotomy was high: over 75% of patients had shoulder height differences, one-fifth had limited motion of right upper extremity, and 14% had asymmetric thoracic wall. In addition to chest wall asymmetry, female patients may develop significant breast asymmetry that may require later plastic surgical repair.[17,18] Chest wall deformities are less common in patients who have undergone only one thoracotomy and no rib resection. At this time, it is still unclear whether chest wall deformities can be avoided if esophageal repair is performed by mini-invasive thoracoscopic approach.

Scoliosis may occur after thoracotomy for OA but is more common and more severe in patients with associated vertebral anomalies. The incidence of scoliosis in patients with no vertebral abnormalities has been reported to be between 14% and 47% in those with vertebral anomalies.[19] In our own adult review, the overall incidence of scoliosis was around 50%. Moreover, vertebral anomalies were common, occurring most commonly in the cervical spine (over one-third of the patients had cervical vertebral anomalies). Scoliosis that is associated with thoracotomy is usually mild and does not cause symptoms. In patients with vertebral anomalies, scoliosis may become symptomatic, and some patients require spinal surgery to correct the defect and stop its progression.

Late complications of esophageal anastomosis

Esophageal stricture is a common early sequel following repair of OA. Need for dilation of the strictured anastomosis has been reported in recent large series to occur in up to 80% of the patients.[20-22] Strictures are more common in patients with a long-gap atresia.[2,23] Anastomotic technique also plays a role; patients with one layer end-to-end anastomosis have less frequently strictures than those who have had end-to-side anastomosis or sleeve anastomosis.[7] Patients who have undergone staged repair have usually had long-gap atresia and, therefore, a high incidence of anastomotic strictures. Persistent strictures are usually associated with GER, and the management of GER is an essential part of treatment of recalcitrant esophageal strictures.[23,24] Late esophageal strictures are, on the other hand, very uncommon; esophageal dilatations are rarely required in older children and adults (Biller). Late strictures are invariably associated with severe GER.

Recurrent tracheoesophageal fistula occurs on average in 8% to 10% of OA patients. The typical age of presentation is 2 to 18 months after primary repair.[2,7] Recurrent fistula is more common in patients with initial end-to-side anastomosis.[7] Recurrent fistula is best diagnosed by tracheobronchoscopy or cineradiographic esophagography with the patient prone. Late presentation of recurrent fistula is possible. Typical symptoms of late-presenting fistula include recurrent pneumonias and pulmonary infections. Cough or choking associated with meals is less common symptoms in older patients. During the last 15 years, three adolescent and adult patients have been treated for late-presenting recurrent tracheoesophageal fistula in our institution. In two patients, the fistula was closed endoscopically by laser coagulation of the mucous lining of the fistula accompanied by fibrin glue injection; in one patient, operative closure of the fistula was required following unsuccessful attempts of endoscopic therapy.

Esophageal motility

Various abnormalities of esophageal motor function have been repeatedly demonstrated among long-term survivors of OA.[5,6,8,25] Three separate studies of esophageal manometry (46 individuals total) 7 to 31 years after repair of OA