Outcome of Oesophageal Atresia Beyond Childhood

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Survivors of oesophageal atresia are reaching their adulthood in large numbers for the first time, thus allowing assessment of true long-term outcome among these patients. Long-term outcomes in terms of oesophageal function and respiratory complications as well as quality of life have been reported in children and adolescents (1,2), These studies have mainly been based on institutional patient series. There is no previous population-based long-term follow-up on oesophageal atresia; therefore, the long-term sequelae are unclear.

In 2003, we initiated a study project to evaluate late outcomes in adult patients who had undergone neonatal repair of oesophageal atresia. We aimed to study the incidence of oesophageal cancer, oesophageal function and morbidity, respiratory morbidity, and clinical characteristics of musculoskeletal anomalies, especially spinal defects in adulthood.

PATIENTS AND METHODS

The original study population consisted of 588 patients treated for oesophageal atresia in the Children's Hospital, University of Helsinki, from 1947 to 1985. A total of 235 were alive with their native oesophagus. Of the 235 contacted patients, 169 (72%) responded. The first 101 (median age 36 years) who replied and agreed to participate made up the clinical study group. The clinical and demographical characteristics of the study group were statistically similar to those of nonparticipants. The study group patients and also the patients not attending the clinical study responded to a symptom questionnaire that included questions on oesophageal, respiratory, musculoskeletal symptoms, and quality of life (SF-32, gastrointestinal quality of life index, respiratory symptom–related quality of life index). Age- and sex-matched general population–derived healthy controls (n = 287) randomly chosen from the Population Register Centre of Finland filled out the same questionnaire. The study patients underwent upper gastrointestinal endoscopy with biopsies, oesophageal manometry, pulmonary function tests, and full orthopaedic evaluation with radiographs. The incidence of cancer among patients with oesophageal atresia was evaluated from a population-based countrywide cancer registry that covers practically 100% of all cancer cases.

RESULTS

Symptomatic gastro-oesophageal reflux (GOR) occurred in 34% and dysphagia in 85% of the patients (8% and 2% in controls, $P < 0.001$). The endoscopic findings included hiatal hernia (28%), Barrett oesophagus (11%), oesophagitis (8%), and anastomotic stenosis (8%). Histology showed oesophagitis in 25% and epithelial metaplasia in 21%, which was gastric in 15% and intestinal in 6%. Oesophageal metaplasia was associated with oesophagitis in 7 of the 21 patients. The occurrence of pathological findings at endoscopy or histology had no correlation with symptoms of GOR or dysphagia.

Oesophageal manometry demonstrated nonpropagating peristalsis in most patients and ineffective distal oesophageal pressure in all. Manometrical abnormalities were significantly more common in those with epithelial metaplasia ($P < 0.02$). Anastomotic complications, age, low distal oesophageal pressure, and defective peristalsis predicted the development of epithelial oesophageal metaplasia (3).

Eleven percent of the patients and 2% had current respiratory symptoms ($P < 0.001$). Fifty-six percent and 70% of the patients had a history of pneumonia and bronchitis (controls 20% and 50%, $P < 0.001$); 16% of the patients and 6% of the controls had doctor-diagnosed asthma ($P < 0.001$). Respiratory-related impaired quality of life was reported by 11% of the patients but only in 6% of the controls ($P < 0.001$). Pulmonary function tests showed obstruction in 21%, restriction in 21%, and both in 36% of the patients. Bronchial hyperresponsiveness was detected in 41 patients in the histamine challenge test; 15 others had asthma-like response (4).

Thoracotomy-induced rib fusion and epithelial oesophageal metaplasia were the most significant risk factors for restrictive ventilatory defect.

Vertebral anomalies were detected in 45% of the patients, most commonly in the cervical spine (38% of the patients). The most significant risk factor for vertebral anomalies was any additional anomaly. Clinical and radiographical scoliosis was found in 56% of the patients; the risk of significant scoliosis was 13-fold when compared with the healthy population. Thoracotomy-induced rib fusion and other associated anomalies were the strongest predictors for scoliosis. In most patients the clinical course of scoliosis was mild and did not require bracing or spinal surgery. Radial ray anomalies were found in 25% of the patients. In the majority of the patients the vertebral anomalies and radial ray anomalies were not detected or recorded during the initial management period and follow-up during childhood (5).

The patients had more frequently impaired gastrointestinal (gastrointestinal quality of life index <105 in 23% of the patients vs 8% of controls) and respiratory-related (respiratory symptom–related quality of life index <45 in 12% of the patients vs 2% of controls) quality of life in relation to controls ($P < 0.001$). There was no difference in the overall health-related quality of life (SF-36) between patients and controls (6).

Despite the high incidence of oesophageal metaplasia, none of the Finnish patients with oesophageal atresia had oesophageal cancer. Three patients had cancer in other organ systems: 1 lymphoma in the small intestine, 1 leukaemia, and 1 uterine carcinoma. The overall cancer incidence was similar to that in the general population (7).

CONCLUSIONS

Morbidity associated with oesophageal atresia is significant in adults. Oesophageal symptoms such as dysphagia and GOR were

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common as was abnormal oesophageal histology. There was no association between oesophageal symptoms and histological findings. Surgical complications, increasing age, and impaired motility predicted the occurrence of epithelial metaplasia. Oesophageal anastomotic complications appear to further impair oesophageal motility and GOR; these predispose patients to the development of epithelial metaplasia. Our studies have shown that the statistical risk of oesophageal cancer is not higher than 500-fold that of the general population. The overall cancer risk of adults with repaired oesophageal atresia is similar to that of the general population. The study population was relatively young; therefore, continuing follow-up and further studies are required to clarify the risk of oesophageal cancer and also define guidelines for long-term endoscopic surveillance of adult oesophageal atresia patients.

Respiratory symptoms, asthma, and infections were more common in patients with oesophageal atresia than in controls. More than half of the patients with repaired oesophageal atresia develop scoliosis. Vertebral abnormalities, especially cervical anomalies, and radial ray anomalies were also common; most of these had not been detected earlier. The overall quality of life of the patients is comparable with healthy controls.

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