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A prospective comparative evaluation of persistent respiratory morbidity in esophageal atresia and congenital diaphragmatic hernia survivors☆

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Abstract

Purpose: The aim of the study was to compare long-term respiratory morbidity in children after repair of esophageal atresia (EA) or congenital diaphragmatic hernia (CDH).

Patients and Methods: Children were seen at 6, 12, and 24 months and 5 years within a prospective longitudinal follow-up program in a tertiary children's hospital. Respiratory morbidity and physical condition were evaluated at all moments. At age 5 years, pulmonary function and maximal exercise performance were tested.

Results: In 3 of 23 atresia patients and 10 of 20 hernia patients, bronchopulmonary dysplasia was developed. Seventeen atresia and 11 hernia patients had recurrent respiratory tract infections mainly in the first years of life. At age 5, 25% of EA and CDH patients measured showed reduced forced expiratory volume in 1 second (z-score < -2). Both atresia and hernia patients showed impaired growth, with catch-up growth at 5 years in patients with EA but not in those with hernia. Maximal exercise performance was significantly below normal for both groups.

Conclusions: Esophageal atresia and CDH are associated with equal risk of long-term respiratory morbidity, growth impairment, and disturbed maximal exercise performance. Prospective follow-up of EA patients aimed at identifying respiratory problems other than tracheomalacia should be an integral part of interdisciplinary follow-up programs.

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Congenital diaphragmatic hernia (CDH) and esophageal atresia (EA) are both severe congenital anatomical anomalies requiring neonatal surgery and intensive care treatment. Follow-up for children with EA tends to focus on gastrointestinal pathologic condition [1-3]. Respiratory pathologic condition, however, seems equally important and is widely described as related to the variable amount of tracheomalacia. Abnormal development of the tracheobronchial tree may contribute to tracheomalacia and recurrent atelectasis [4]. Mild lung function abnormalities after EA repair have been described [5,6]. Several cross-sectional studies report secondary morbidity. Up to 50% of EA patients were found to have associated anomalies such as cardiac anomalies and consequently higher morbidity [7].

New treatment modalities such as high-frequency oscillation (HFO) ventilation, nitric oxide (NO) administration, and extracorporeal membrane oxygenation (ECMO) have improved survival rates in CDH patients [8-10]. Evidence is emerging, however, that better survival coincides with a great deal of morbidity [11-17]. Long-term pulmonary sequelae in CDH survivors seem to result not only from residual lung hypoplasia with persistent pulmonary hypertension but also from lung injury induced by ventilatory support [18]. Other risk factors for morbidity are large diaphragmatic defects, ECMO therapy, and patch repair [16].

We hypothesized that children after repair of EA show the same extent of respiratory pathologic condition as CDH survivors, although probably of a different nature, with different causative mechanisms. The aim of the present study was to describe respiratory morbidity in EA and CDH patients with respect to baseline characteristics, respiratory tract infections (RTIs), lung function, and maximal exercise performance. In addition, we evaluated gastrointestinal morbidity and physical growth because these factors may influence the main end points with respect to respiratory morbidity. This is the first study to prospectively evaluate respiratory morbidity in EA patients at the age of 5 years.

1. Methods

This longitudinal, observational, prospective, cohort study consists of repeated measurements at 6, 12, and 24 months and at 5 years.

1.1. Setting

The facility used for the study was the Pediatric Surgical Department of the Erasmus MC-Sophia Children's Hospital, Rotterdam, the Netherlands. This is the only tertiary academic facility in the Southwestern part of the Netherlands equipped for all major surgical specialties. The referral area has 4 million inhabitants with 44,000 newborns annually.

Since 1999, a multidisciplinary team—consisting of a consultant senior pediatric surgeon, pediatricians, psychologists, a pediatric physiotherapist, nurses, and a social worker—runs a follow-up program for children born with a major anatomical malformation and their families. A clinical geneticist was added to the team in 2004. The program aims to reduce the overall morbidity associated with these malformations, in particular, the index diagnoses as described by Ravitch [19].

1.2. Patients

All 68 patients with EA and CDH admitted to the ICU of our department within 7 days after birth from January 1999 to February 2003 were eligible for this study. For the present study, we excluded data from 4 patients having infections and growth impairment as a result of a major syndromal or

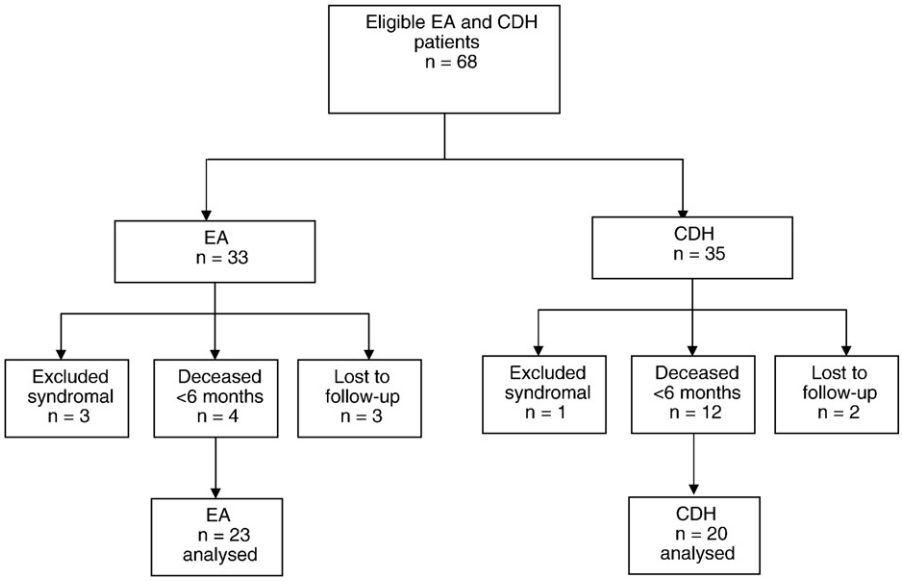

Fig. 1 Flowchart showing group composition.