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Review

Esophageal atresia and transitional care—step 1: a systematic review and meta-analysis of the literature to define the prevalence of chronic long-term problems

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Review

Abstract

BACKGROUND: Esophageal atresia (EA) is a rare congenital anomaly with high infantile survival rates. The aim of this study was to outline the prevalence of common long-term problems associated with EA repair in patients older than 10 years of age.

DATA SOURCES: Original papers were identified by systematic searching of MEDLINE and EMBASE databases from January 1993 to July 2014. Fifteen articles (907 EA patients) met inclusion criteria.

CONCLUSIONS: This is the first systematic review aiming to quantify the prevalence of the long-term problems associated with EA. The main active medical conditions (pooled estimated prevalence) identified were the following: dysphagia (50.3%), gastroesophageal reflux disease with (40.2%) or without (56.5%) histological esophagitis, recurrent respiratory tract infections (24.1%), doctor-diagnosed asthma (22.3%), persistent cough (14.6%), and wheeze (34.7%). The prevalence of Barrett's esophagus (6.4%) was 4 and 26 times higher than the adult (1.6%) and pediatric (.25%) general populations. Adult and pediatric practitioners should focus on how to develop effective long-term follow-up and transitional care for these patients.

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is a rare congenital anomaly characterized by discontinuity of the esophagus and a possible connection with the trachea.[1] The reported incidence is between 1 in 2,500 to 1 in 4,500 live births.[2] The advancement of surgical techniques and neonatal intensive care management has led to postnatal survival rates of more than 90%.[3] A large number of EA repair patients are now reaching adulthood and transitioning into adult healthcare services.[3] Generally, the standard of pediatric care is good, and as shown in the literature, the overall quality of life (QoL) has been reported as favorable.[2,4,5] However, there is an undefined proportion of patients suffering from chronic long-term problems, which

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would benefit from a planned well-structured follow-up from newborn to adult life followed by pathways of transitional care.[6] At present, there is little knowledge on the exact prevalence and management regarding the chronic active problems these patients are experiencing decades after the neonatal repair. Moreover, international guidelines on the optimal esophageal and respiratory follow-up do not exist at present and patients with problems often raise this as an issue.[7]

The aims of this systematic review and meta-analysis were to quantify the prevalence of common long-term problems associated with EA repair. Management options and future research gaps to be developed in the near future will be discussed. This is the first step toward a rationale method of transition for children with EA-associated morbidity into the adult healthcare system.

Methods

Search strategy

MEDLINE and EMBASE databases were accessed using the OVID search platform on July 8, 2014. The search was performed using the key terms "Esophageal Atresia" OR "Esophageal Atresia" AND "Long term" and limited to the period January 1993 to July 2014 and English language. The search was performed by 2 independent reviewers (M.J.C. and L.R.S.). In addition, a manual search was conducted evaluating review articles' references. Results were screened independently in a systematic fashion by the same reviewers (M.J.C. and L.R.S.) for title and abstract content relevant to EA. Publications were selected based on predetermined exclusion and inclusion criteria. Conflicts in screening and selection were resolved through consensus between 4 authors (S.G., V.V.K., M.J.C., and L.R.S.). The modified PRISMA flowchart[8] showing study selection is presented in Fig. 1.

Inclusion criteria

Original articles describing medical and surgical long-term problems present in children over 10 years old and adults operated for EA at birth.

Exclusion criteria

The following exclusion criteria were applied:

- Papers published pre-1993: To ensure the most up-to-date literature was reviewed.
- Esophageal replacement: This is a small and nonhomogeneous subgroup of complicated patients with a highly variable long-term outcome.
- Age less than 10 years: Long-term defined by this review as follow-up of patients greater than or equal to 10 years.
- Review articles: These publications did not add novel evidence to the body of existing data.

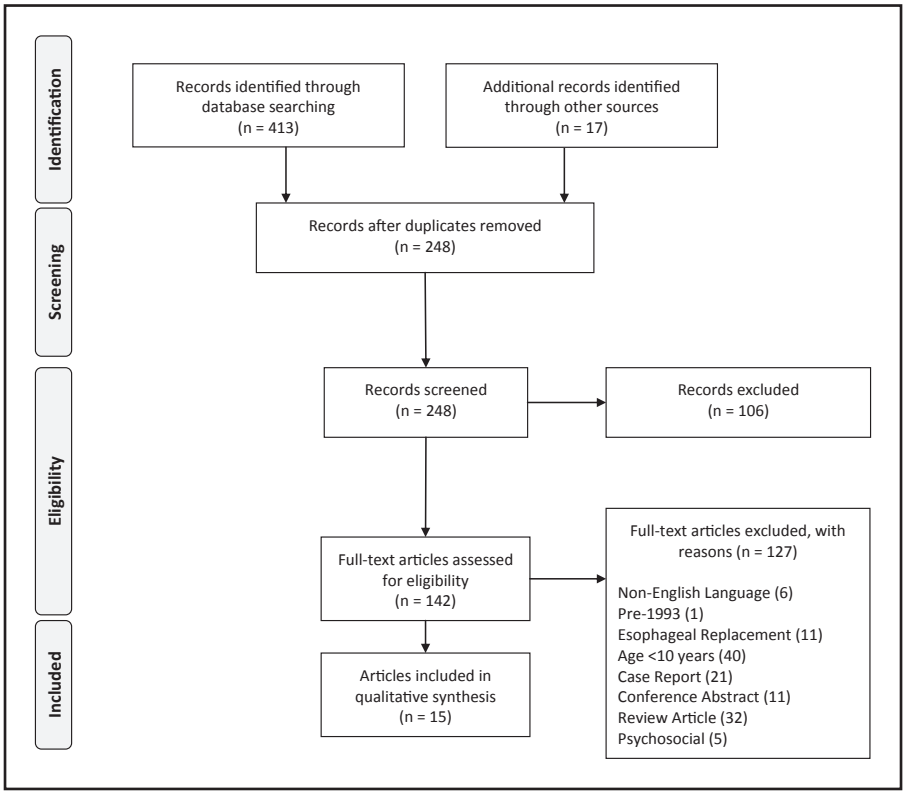

Figure 1 Algorithm for study selection modified from PRISMA flowchart.