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Isolated tracheoesophageal fistula *versus* esophageal atresia – Early morbidity and short-term outcome. A single institution series

R.B. Tröbs [a, *], W. Finke [b], M. Bahr [a], C. Roll [c], M. Nissen [a], M.R. Vahdad [d], G. Cernaianu [e]

[a] Department of Pediatric Surgery, St. Mary's Hospital, St. Elisabeth Group, Ruhr-University of Bochum, D-44627, Herne, Widumer Str. 8, Germany
[b] Department of Anesthesiology and Surgical Intensive Care, St. Mary's Hospital, St. Elisabeth Group, Ruhr-University of Bochum, D-44627, Herne, Widumer Str. 8, Germany
[c] Vest Children's Hospital, University of Witten-Herdecke, Department of Neonatology and Pediatric Intensive Care, Dr. Friedrich-Steiner-Str. 5, D-45711, Datteln, Germany
[d] Department of Pediatric Surgery and Pediatric Urology, University of Marburg, Universitätsklinikum, Baldingerstrasse, D-35043, Marburg, Germany
[e] Department of Pediatrics and Adolescent Medicine, Pediatric Surgery, University of Cologne, Kerpener Str. 26, D-50937, Cologne, Germany

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ABSTRACT

Purpose: We compared the postnatal course, morbidity and early results after repair for cases of isolated or "pure" TEF with those for cases of esophageal atresia (EA) with distal tracheoesophageal fistula (TEF).
Methods: Twenty-four consecutive infants were divided into two groups: isolated TEF [TEF group] (n = 5) and EA with distal TEF [EA group] (n = 19). Results. A high rate of prematurity (29%) and major cardiac and other surgically-relevant malformations (0.8 vs. 0.7 per infant) was found in both groups. The median age at surgery was 8 days for the TEF group vs. 1 day for the EA group ($p < 0.01$). Most infants of both cohorts had stable acid-base and respiratory parameters at admission. Generally, tracheoscopy provided valuable information regarding the position of the TEF. Surgery for isolated TEF was performed via right cervicotomy in 4 cases and via thoracotomy in one. Postoperative thoracostomy tubes were inserted in 3 cases and one emergency gastrostomy was created for acute gastric overextension (exclusively in patients with EA). The duration of postoperative mechanical ventilation (49 vs. 113 h, $p = 0.045$) and the median length of stay in the pediatric surgery unit (10 vs. 20.5 days, $p = 0.003$) were shorter for the isolated TEF group. Four EA patients experienced severe events. Total mortality was 8% (0 out of 5 with TEF vs. 2 out of 19 with EA).
Conclusion: Developmental delay and a high rate of morbidity were found in both groups. More complex surgery increased perioperative morbidity in cases of EA. With early recognition of isolated TEF, a less complicated course can be expected in comparison with esophageal atresia.

1. Introduction

Esophageal atresia (EA) encompasses a spectrum of malformations comprising an interruption of the esophagus, predominately in combination with persistent communication with the trachea (tracheoesophageal fistula, TEF) [1].

The worldwide prevalence of EA and TEF is 1 in 4099–4608 newborns [2,3]. Survival rates up to 95% have been achieved in centers that provide the best neonatal care [4–6]. Newborns with EA are not able to swallow saliva and milk, and gaseous extension of the gastrointestinal tract combined with gastric reflux develops via the TEF.

"Pure" or isolated TEF without EA occurs in approximately 4–5% of infants with EA/TEF [1,6,7]. The terms "H-type" or "N-type" TEF refer to congenital fistulous tracks between the posterior wall of the trachea and the anterior circumference of the esophagus [8]. Tracheoesophageal fistulas at a high origin show a more transverse course (H-type) than N-type fistulas that are located more caudally. The prevalence of this rare subgroup is 1 per 100, 000 births [7]. Infants with isolated TEF present with coughing and cyanosis

* Corresponding author. Klinik für Kinderchirurgie, Marienhospital Herne, St. Elisabeth-Gruppe, Klinikum der Ruhr-Universität Bochum, Widumer Str. 8, 44627, Herne, Germany.
E-mail addresses: ralf-bodo.troebs@elisabethgruppe.de (R.B. Tröbs), werner.finke@elisabethgruppe.de (W. Finke), micha.bahr@elisabethgruppe.de (M. Bahr), c.roll@kinderklinik-datteln.de (C. Roll), matthias.nissen@elisabethgruppe.de (M. Nissen), rvahdad@aol.com (M.R. Vahdad), grigore.cernaianu@uk-koeln.de (G. Cernaianu).

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during feeding; recurrent severe bronchitis and pneumonia also occur. A less severe clinical picture may lead to a postponed diagnosis.

Surgery involves dissection of the TEF and anastomosis of the esophagus in cases of esophageal atresia. Either open or thoracoscopic repair has been used to surgically correct both conditions [9–11].

The main goals of treatment are timely recognition of the underlying malformation, reduction of morbidity and provision of the best possible quality of life for patients and their parents.

This report focuses on the postnatal course, congenital malformations and short-term outcome of infants with isolated TEF [TEF group] compared with those with EA and distal TEF [EA group]. We looked for perioperative problems and early life-threatening events. Data were discussed in light of recent literature.

2. Materials and methods

A total of 24 cases was included in this retrospective study. We evaluated all infants with TEF (Gross E) treated between January 2006 and August 2016. Data from 19 infants with EA and distal TEF (Gross C) treated between January 2013 and August 2014 served as the control.

We evaluated clinical records and surgical reports to extract data about perinatal course, acid-base- and respiratory status at admission, surgery, malformations and in-hospital morbidity during their first stay.

In infants with a birth weight above 1000 g, we routinely performed upper airway endoscopy to identify the vocal cords, the shape of larynx and trachea and the location of the TEF. In two newborns with a birth weight below 1000 g, we did not attempt to insert the tracheoscope.

Patients were also classified according to the risk groups defined by Spitz et al. [1]: group 1: birth weight > 1500 g, without major cardiac disease; group 2: birth weight < 1500 g or with a major cardiac disease; group 3: birth weight < 1500 g and with a major cardiac disease.

The presence of VACTERL association was diagnosed if at least 2 of the following were found in addition to the tracheoesophageal malformation: vertebral anomaly (V); anal atresia (A); and cardiovascular (C), renal (R) or limb (L) malformations, defects or anomalies [12].

Length of stay was defined as the duration of the stay in the initial pediatric surgery and in the intensive and intermediate care units.

To evaluate medium-term results, we re-evaluated our charts and conducted a telephone interview (follow-up between 6 months and 8 years).

2.1. Statistics

We calculated medians, arithmetic means ± standard deviations (SD) and ranges.

To compare the two groups, we applied the Mann-Whitney test at the 0.05 level. Since both sample sizes are very small, interpretation of the resulting p-values requires great caution.

2.2. Ethics statement

This study was approved by the ethics committee of the Ruhr University of Bochum (4951-14).

3. Results

3.1. Baseline characteristics (Table 1)

Overall, 5 infants had TEF and 19 infants had EA with TEF. Two vs. 11 infants were males. Median gestational age of both groups was 38 vs. 37 weeks (range 25–40) and the median birth weight was 2985 g vs. 2500 g (range 1930–3670 vs. 720 to 4060 [EA]). The total number of premature births was 7 (29%, all EA) and 5 (21%) newborns with EA with birth weights below 1500 g.

Birth weight and gestational age did not differ significantly between groups.

3.2. Acid-base and respiratory status at admission

The spectrum of biochemical parameters recorded included hematocrits (Hct), acid-base status (pH, partial pressure of carbon dioxide [pCO_2] and standard bicarbonate [HCO_3^-]). Data are shown in Table 2.

Infants with isolated TEF had significantly higher levels of standard bicarbonate and a tendency for higher pH. Partial pressure of carbon dioxide, the respiratory parameter, was predominately below 50 mmHg in both groups; there was no difference between TEF and EA patients.

3.3. Diagnosis and surgery

The diagnostic evaluation included chest X-rays for the initial confirmation of EA, echocardiography and ultrasound screening of the abdomen.

Isolated TEF was initially identified either by tracheoscopy performed by a neonatologist (n = 1) or an esophagogram with water soluble contrast medium (n = 2, both diagnostic for TEF). In 2 cases, clinical symptoms led to admission to the pediatric surgery unit and both tracheoscopy and fistula dissection were performed in a single anesthesia.

Upper airway endoscopy did not reveal any abnormalities of the vocal cords. In all cases, with the exception of infants below 1000 g, the TEF was identified (Fig. 1) [13].

Additional esophagoscopy was applied in three cases with isolated TEF (Fig. 2).

For the dissection of isolated TEF, we employed a low right-sided cervical approach with transversal incision (Fig. 3) in 4 cases. A thoracotomy was applied in the fifth newborn in whom the esophagogram had revealed the TEF at the level of the 6th thoracic vertebra. Using tracheoscopy, the TEF was located 3 tracheal rings above the carina (Table 2). During surgery, we temporarily inserted a 3-French catheter into the TEF. The presence of the recurrent laryngeal nerve (RLN) was always kept in mind, although we did not attempt to identify this nerve.

EA repair was performed with a right-sided thoracotomy with preservation of the intact latissimus dorsi muscle. The esophagus was accessed via a retropleural route (Fig. 4). Generally, all of the infants in both groups received a transanastomotic, size 4- to 6-French feeding tube for early postoperative feeding. With the exception of the 720 g infant, all surgeries were performed in a single step.

The levels of TEF and surgical approaches are listed in Table 3.

To check the passage and ensure the integrity of the esophageal suture [TEF group] or anastomosis [EA group], a routine postoperative esophagogram with water-soluble contrast medium was performed 5–7 days postoperatively. Routine postoperative laryngoscopy was not performed.