

CHILDREN'S HOSPITAL II

POSTOPERATIVE CONGENITAL ESOPHAGEAL ATRESIA COMPLICATIONS: A REVIEW

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Background

Esophageal atresia (EA) is a congenital defect of the esophagus with or without connection to the trachea (TEF).

□ Incidence: 1 per 3500 - 4000 live births.

□ EA/TEF may occur as an isolated anomaly or may develop within the context of a known syndrome or association. (VACTERL association)

□ EA and TEF are classified according to their anatomic configuration.

Classification



Type: A Esophageal atresia without tracheoesophageal fistula (\$% of cases)

Esophageal atresia with tracheoesophageal fistula (88% of cases)



Type B: Esophageal atresia with proximal tracheoesophageal fistula (1% of cases)



Type C: Esophageal atresia with distal tracheoesophageal fistula (84% of cases)



Type D: Esophageal atresia with proximal and distal tracheoesophageal fistula (3% of cases)



Type E: Tracheoesophageal fistula without esophageal atresia (H-type) (4% of cases)

Management

□ Surgical ligation of the fistula.

- \rightarrow Primary anastomosis
- \rightarrow Staged procedures
- Methods
- \rightarrow Thoracoscopy
- \rightarrow Open thoracotomy

Author	Type of study	Level of evidence	No. of thoracoscopies vs. no. of thoracotomies	Endpoints	Results thoracosco vs. thoracotomy (%)
Borruto et al. [17] 2012	Meta-analysis	3a	69 vs. 97	leakage rate	No differences
				Stricture rate	No differences
Szavay et al. [14] 2011	Retrospective comparative study	3b	25 vs. 32	Leakage rate	4 vs. 3
				Stricture rate	0 vs. 0
				Operating rate	Longer
				pCO ₂ max value	Higher
				Ventilation time	No differences
				Complication time	No differences
Allal et al. [21] 2009	Retrospective comparative study	3b	14 vs. 14(+3)'	Leakage rate	14 vs. 19
				Stricture rate	14 vs. 50
				Operating time	Longer
				Complication rate	No differences
Al Tokhais et al. [20] 2008	Retrospective comparative study	3b	23 vs. 22	Leakage rate	17 vs. 14
				Stricture rate	8 vs. 29
				Operating time	No differences

Table 1 Comparative studies of open vs thoracoscopic TEF showing no significant difference in outcomes or complications

COMPLICATIONS

- Anastomotic leak
- Esophageal stricture
- Recurrent fistulae
- Dysphagia
- GERD with esophagitis
- GERD without esophagitis
- Respiratory tract infections
- Tracheomalacia
- □ Asthma
- Wheeze
- Persistent cough
- Barrett esophagus
- Esophageal cancer (squamous cell)

Early complications after esophageal atresia repair: analysis of a German health insurance database covering a population of 8 million

C. Dingemann et al, Center of Pediatric Surgery, Hannover Medical School, Hannover, Germany

- •The aim of this study was to evaluate the early postoperative results after repair of esophageal atresia
- •All patients who had undergone esophageal atresia repair from January 2007 to August 2012.
- •The incidences of anastomotic leak (3%) and recurrent tracheoesophageal fistula (7%)
- •Anastomotic stricture required dilatation 57%
- •93% of the patients were readmitted at least once

•A correlation between the complication rate and characteristics of the treating institutions was not identified.

Table 1 Postoperative complications in 75 patients within thefirst year after esophageal atresia/tracheoesophageal fistula repairdefined as 'any reintervention for esophageal morbidity'

Postoperative complication	Incidence of postoperative complication <i>n</i> (%)
Anastomotic stricture	43 (57)
Complicated GERD [†]	6 (8)
Recurrent tracheoesophageal fistula	5 (7)
Esophageal perforation after dilatation	4 (5)
Subglottic stenosis after long-term mechanical ventilation	3 (4)
Anastomotic leak	2 (3)
Recurrent pneumothorax requiring surgery	2 (3)
Severe tracheomalacia‡	2 (3)
Tension pneumothorax	1 (1)
Iatrogenic duodenal perforation	1 (1)

("Early complications after esophageal atresia repair: analysis of a German health insurance database covering a population of 8 million", C. Dingemann, 2015)

Table 2: Incidence of postoperative complications

Authors	Country	Number of surgical units	Number of patients	Anastomotic leak <i>n</i> (%)	Recurrent TEF n (%)	Anastomotic stricture <i>n</i> (%)
Koivusalo <i>et al</i> . ¹⁴	Finland	1	130	9 (7)	3 (2)	[102 (78)]‡
Lacher et al.5	Germany	1	111	2(2)	3 (3)	43 (39)*
Lilja and Wester ¹⁵	Sweden	1	147	10(7)	11 (7)	82 (56)
Holcomb 3rd et al. ¹⁶	United States [†]	6‡	104	8 (8)	2 (2)	33 (32)***
Konkin <i>et al</i> . ¹⁷	Canada	1	144	12 (8)	12 (8)	75 (52)
This study 2014	Germany†	37	75	2 (3)	5 (7)	43 (57)

("Early complications after esophageal atresia repair: analysis of a German health insurance database covering a population of 8 million", C. Dingemann, 2015)

Results from the French National Esophageal Atresia register: one-year outcome

Schneider et al. Orphanet Journal of Rare Diseases 2014

•The aim of this study was to assess the early morbidity of esophageal atresia.

•307 EA patients born in 2008 and 2009 of 38 multidisciplinary French centers

•The 1-year outcome: anastomotic leaks (8%), recurrent tracheoesophageal fistula (4%), and anastomotic stenosis (22%), 59% readmitted for digestive (52%) or respiratory (48%) reasons, 12% required antireflux surgery, 15% were undernourished at 12 months of age, 37% presented with respiratory symptoms and 15% had dysphagia.

•Although mortality was low, digestive and respiratory morbidities were frequent in the first year after EA repair and often required rehospitalization \rightarrow high health costs and high social and psychological effects on the relatives of these children.

Predictive factors for complications in children with esophageal atresia and tracheoesophageal fistula

R. Shah, University of New South Wales, Australia

•The aim of this study was to describe the incidence of complications in children with esophageal atresia (EA/TEF) to identify predictive factors for their occurrence.

•A retrospective chart review of 110 patients born between January 1999 and December 2010.

From univariate analysis

Early esophageal stricture was more likely in children with 'long-gap' EA ([OR] = 16.32) and easy to develop chest infections (OR = 3.33).

>Patients with severe tracheomalacia were more likely to 'cyanotic/dying' (OR = 180) and undergo aortopexy (OR = 549).

> Patients with gastroesophageal reflux disease were significantly likely to require fundoplication (OR = 10.83) and undergo aortopexy (OR = 6.417).

•From multivariate analysis,

> 'Long-gap' EA was a significant predictive factor for late esophageal stricture (P = 0.007) and for gastrostomy insertion (P = 0.001).

> Reflux was a significant predictive factor for requiring fundoplication (P = 0.007) and gastrostomy insertion (P = 0.002).

> Gastrostomy insertion (P = 0.000) was a significant predictive factor for undergoing fundoplication.

> Having fundoplication (P = 0.001) was a significant predictive factor for undergoing a subsequent aortopexy.

Outcomes at One-Year Post Anastomosis from a National Cohort of Infants with Oesophageal Atresia

Benjamin Allin et al, National Perinatal Epidemiology Unit, University of Oxford, Oxford, United Kingdom, 2014

• We aimed to provide an assessment of outcomes at oneyear post EA-TEF repair, focussing particularly on postoperative complications. We also aimed to assess the efficacy of prophylactic anti-reflux medication (PARM) in reducing stricture formation.

• A prospective, multi-centre cohort study of all infants liveborn with oesophageal atresia in the United Kingdom and Ireland in 2008- 2009 was performed. The effect of PARM on stricture formation in infants with the type-c anomaly was assessed using logistic regression analysis.



Figure 2. Flow diagram showing number of infants recruited and those who were lost to follow-up. doi:10.1371/journal.pone.0106149.g002

- 151 infants with oesophageal atresia. One-year follow-up information was returned for 105 infants (70%); the mortality rate was 8.6% (95% CI 4.7–14.3%).
- Post-operative complications included anastomotic leak (5.4%), recurrent fistula (3.3%) and oesophageal stricture (39%).
- This study provides a benchmark for current outcomes and complication rates following OA-TOF repair.

 No statistically significant difference in stricture rate between those infants who received PARM and those who did not.
 Need further investigation such as a randomised controlled trial.

Long-term esophageal and respiratory outcomes in children with esophageal atresia and tracheoesophageal fistula

Richard H. et al, *Department of Internal Medicine, Cleveland Clinic, Cleveland, OH, USA, 2015.*

- •The aim of our study was to characterize the esophageal and respiratory morbidity of EA/TEF.
- •43 patients with congenital EA/TEF evaluated from 2011 to 2014 were included.

- •Heartburn (48,8%), acid regurgitation (44.1%), dysphagia (72.1%), dysmotility (76.9%).
- •Cough and choking in 72.1% patients and dyspnea and wheezing (53.4%). Recurrent respiratory infections (44.2%).
- •Tracheomalacia (86.7%) and restrictive lung disease(54.5%).
- •Thirty patients underwent endoscopy, of which 70.0% had a stricture, and 20.0% had recurrent fistula requiring surgical intervention.
- •There is a high burden of residual esophageal and pulmonary pathology in patients with EA/TEF. Ongoing followup is required to monitor both the clinical symptoms and treatment responses.

Morbidity and Mortality in Patients with Esophageal Atresia

Jason P. Sulkowski et al, Center for Surgical Outcomes Research, The Research Institute at Nationwide Children's Hospital, Columbus, OH, 2014

•This study reports characteristics and outcomes for patients with EA/TEF.

•Patients admitted within 30 days of life who had ICD-9-CM diagnosis and procedure codes relevant to EA/TEF during 1999–2012. Post-operative outcomes were examined up to 2 years following EA/TEF repair.

• 3479 patients with EA/TEF treated at 43 children's hospitals. Within two years of discharge, 54.7% were readmitted, 5.2% had a repeat TEF ligation, 11.4% had a repeat operation for their esophageal reconstruction, and 11.7% underwent fundoplication. Mortality was 5.4%.

Treatment outcomes for eosinophilic esophagitis in children with esophageal atresia

L. J. Chan et al , Department of Paediatric Gastroenterology, Sydney Children's Hospital, University of New South Wales, Sydney, Australia

•The aim of this study is to evaluate the treatment outcomes of EoE in children with EA-TEF.

•A retrospective chart review was performed on all EA-TEF children with EoE between January 2000 and September 2013.

•Patients were treated with budesonide slurry, swallowed fluticasone, elimination diet. All patients were on proton pump inhibitors at time of diagnosis of EoE which was continued.



Fig. 3 Incidence of complications in esophageal atresia (EA) patient with and without eosinophilic esophagitis (EoE).

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Fig. 4 Need for further surgery in esophageal atresia (EA) patients with and without eosinophilic esophagitis (EoE).

•There was a significant reduction in symptoms of dysphagia and reflux post-treatment (P < 0.001). Prevalence of strictures significantly decreased (P = 0.016), as did need for dilatations (P = 0.004).

•There was also a nonsignificant trend towards improvement in weight and height 'z scores' of the patients.

•Treatment of EoE in children with EA-TEF was found to significantly reduce intraepithelial eosinophil count, symptoms, strictures and need for dilatations.

Preoperative tracheobronchoscopy in newborns with esophageal atresia: does it matter?

Pietro Atzori et al

The purpose of this study was to evaluate the influence of preoperative TBS in newborns with EA in preventing complications and improving diagnosis and outcomes.
From 1997 to 2003, 62 patients with EA underwent preoperative TBS.

•Before TBS, the Gross classification of the 62 patients was as follows: type A, 9 patients; type B, none; type C, 51 patients.

- At TBS, 3 of 9 type A patients had an unsuspected proximal fistula (type B). These 3 patients, plus the 2 with H-type fistula, were repaired through a cervical approach.
- In 4 patients, previously undetected malformations of the respiratory tree were found at TBS. Carinal fistulas in 14 type C patients were occluded by Fogarty catheter to improve ventilation during repair.
- •No complications were observed.
- •Overall, tracheobronchoscopy is a useful and safe procedure and should be recommended for babies with EA before surgical repair.

Role of preoperative tracheobronchoscopy in newborns with esophageal atresia: A review

Filippo Parolini et al

- •Tracheobronchoscopy (TBS) in newborns affected by esophageal atresia (EA) being described in 1981.
- •This review provides a detailed overview on the use of TBS in EA newborn, focus on technical and anesthesiological aspects, benefits and risks.
- •TBS is also compared with an esophageal contrast study, computed tomography scan or ultrasound.
- •Although TBS is not a routinely part of the management in many international centers, increasing evidence suggests that this procedure should be strongly recommended in the management of neonates affected by esophageal atresia.

CONCLUSION

- •Esophageal atresia (EA) with or without a tracheoesophageal fistula (TEF) is a rare congenital malformation.
- •The prognosis has improved significantly.
- •Complication rates after EA/TEF repair remain considerably high.
- •Updated information on the outcome of EA is lacking.
- •No statistically significant difference in stricture rate between those infants who received PARM and those who did not. Need further investigation such as a randomised controlled trial.
- •Increasing evidence suggests that TBS should be strongly recommended in the management of neonates affected by esophageal atresia.

THANK YOU FOR YOUR LISTENING!