



- **Analysis based on the highest quality data**
- **Report** at EUPSA Congress (dissemination and discussion)



Esophageal atresia and TEF

1. Diagnostic and operative technique

1. Routine preoperative bronchoscopy
2. Open vs thoracoscopic repair
3. Magnetic anastomosis

Huang Yingying, Haitao Zhu
Chen Young and Stella Sabbatini
Lisandro Luques, Stella Sabbatini and Naho Fujiwara

2. Postoperative strategy

1. Trans-anastomotic tube
2. Ventilation, muscle paralysis and neck flex
3. Antacid treatment

Elke Rutenstock
Naho Fujiwara and Mashriq Alganabi
Nigel Hall

3. Management of long gap

1. Growth by traction (open or thoracoscopic)
2. Delayed anastomosis
3. Kimura procedure

Tomas Wester, Carmen Mesas Burgos + Simon Eaton
Fabian Doktor and Augusto Zani
Naho Fujiwara

4. Esophageal replacement

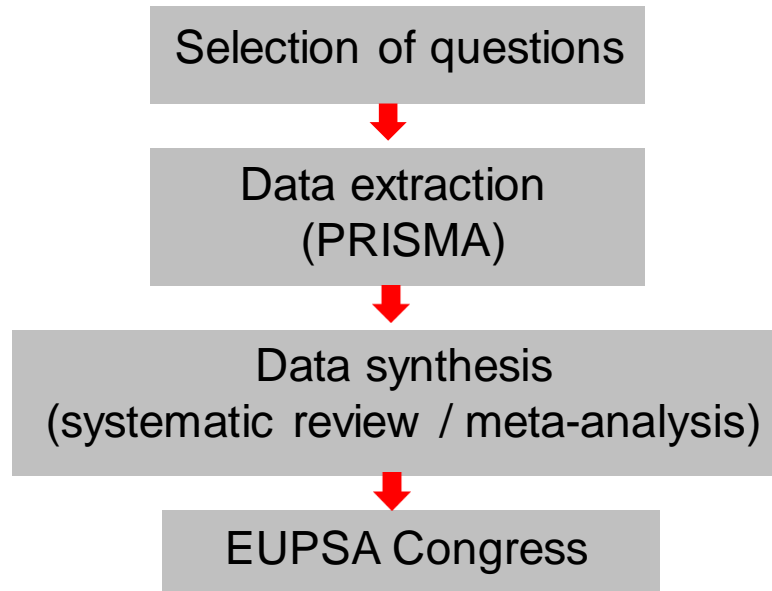
1. Gastric transposition
2. Gastric tube
3. Colonic interposition
4. Jejunum interposition

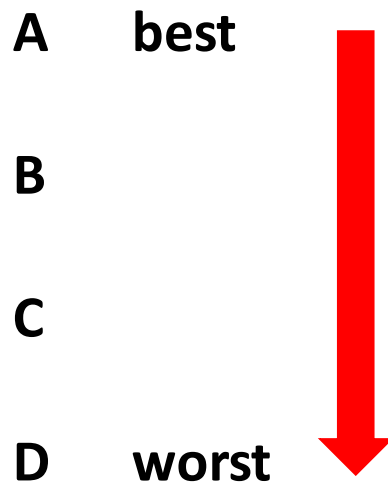
Giuseppe Lauriti, Maria Enrica Miscia, and Francesco Morini
Francesco Morini, Maria Enrica Miscia and Giuseppe Lauriti
Reto Baertschiger and Lisandro Luques - Annika Mutanen
Annika Mutanen

5. Tracheomalacia

1. Aortopexy or posterior tracheopexy

Ramon Gorter, Paul van Amstel and Stefaan Tytgat





Adrian Baker et al. Clin Med 2010;10:358-363

Is a routine pre-operative bronchoscopy beneficial to children with esophageal atresia(EA) and/or tracheoesophageal fistula (TEF)?

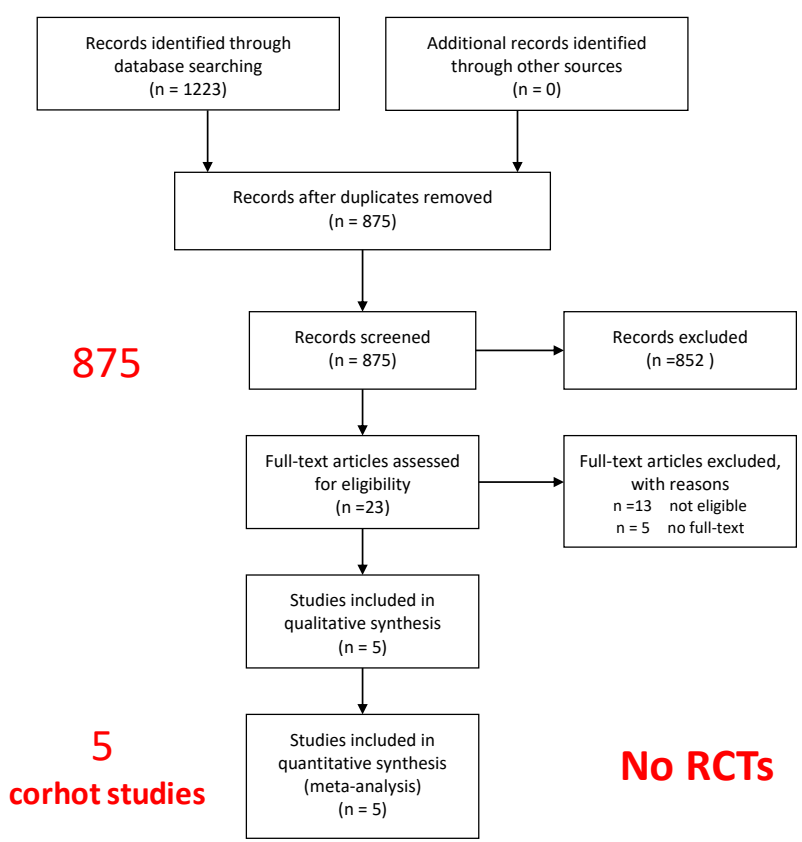
Yingying Huang & Haitao Zhu

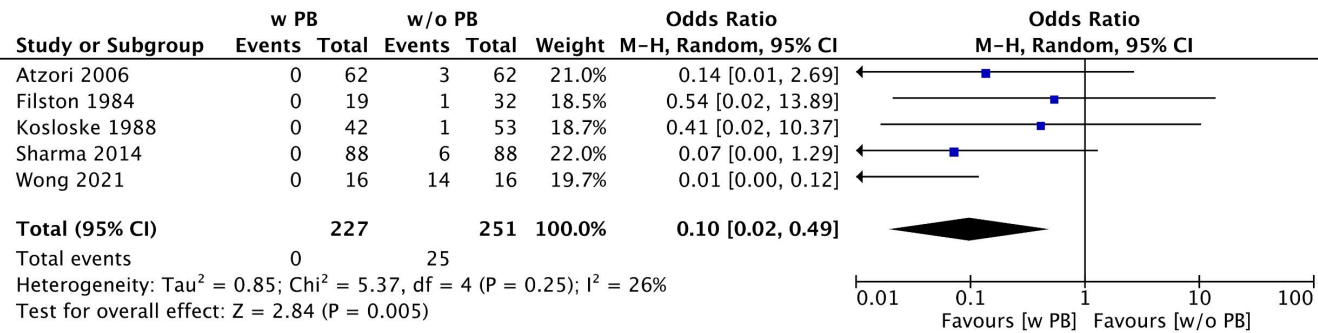
- **Population:** Children clinically diagnosed with EA and/or TEF
- **Intervention:** Bronchoscopy before primary EA and/or TEF repair
- **Comparison:** No bronchoscopy
- **Outcome:**
 - **Primary:**
 - Fistula identification
 - **Secondary**
 - Surgical management variations
 - Additional findings of associated anomalies
 - Procedure-related complications



PRSIMA Flowchart

Identification
Screening
Eligibility
Included



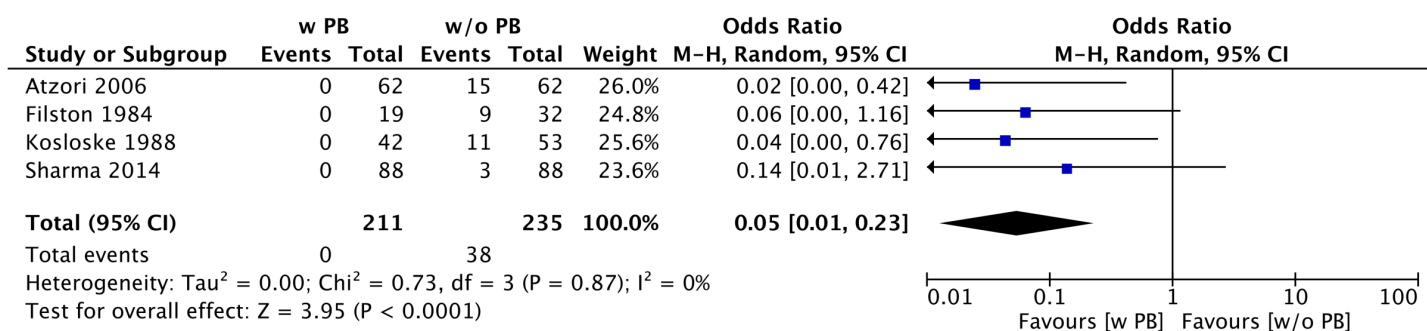


Reduced misdiagnosis rate of EA types in routine pre-op bronchoscopy vs no pre-op bronchoscopy (OR 0.1, P=0.005)

Especially diagnosis of upper pouch fistula and H type EA



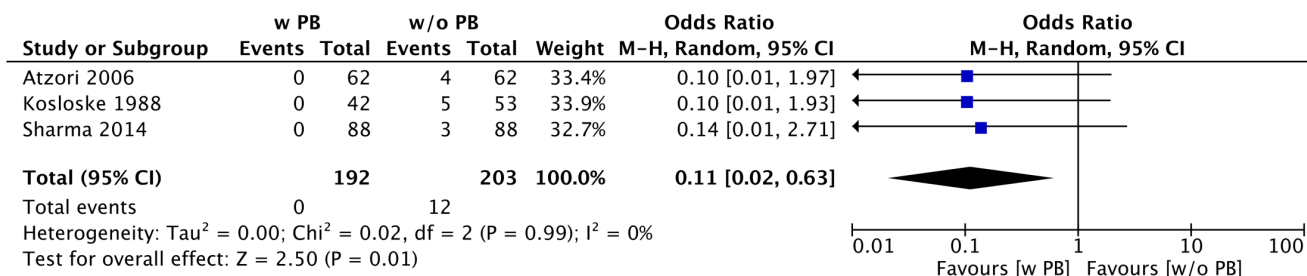
Secondary Outcome----Surgical Management Variation



Reduced surgical management variation in routine pre-op bronchoscopy vs no pre-op bronchoscopy (OR 0.05, P<0.00001)

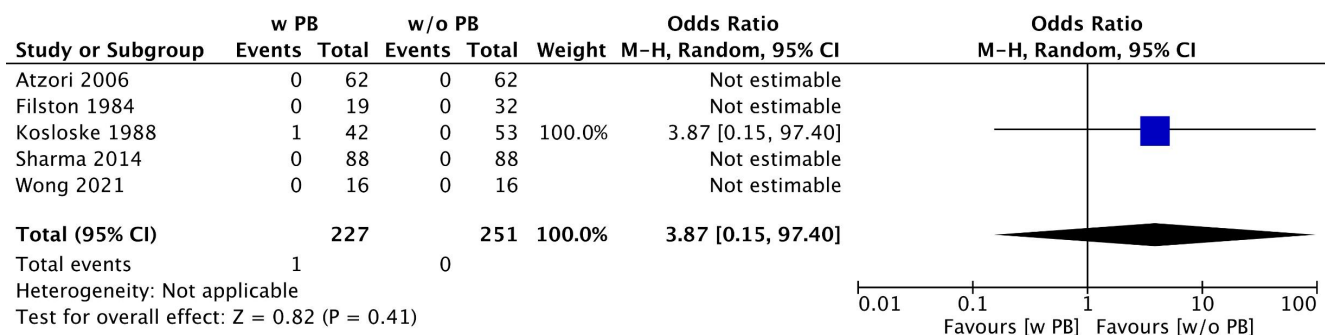


Secondary Outcome----Misdiagnosis Rate of Associated Anomalies



Reduced misdiagnosis rate of associated anomalies in routine pre-op bronchoscopy vs no pre-op bronchoscopy (OR 0.11, P=0.01)

Especially co-existing airway anomalies



No statistically difference between groups
(OR 3.87, P=0.41)

Conclusions

- A routine pre-operative bronchoscopy may be beneficial to children with EA/TEF
- Reduced misdiagnosis rate of EA types and/or associated anomaly before primary EA repair
- Reduced surgical management variation before definite repair
- Without additional intra-op/post-op complications
- Lack of high quality studies

Recommendation **GRADE B**

- A routine pre-operative bronchoscopy is recommended in children with EA/TEF

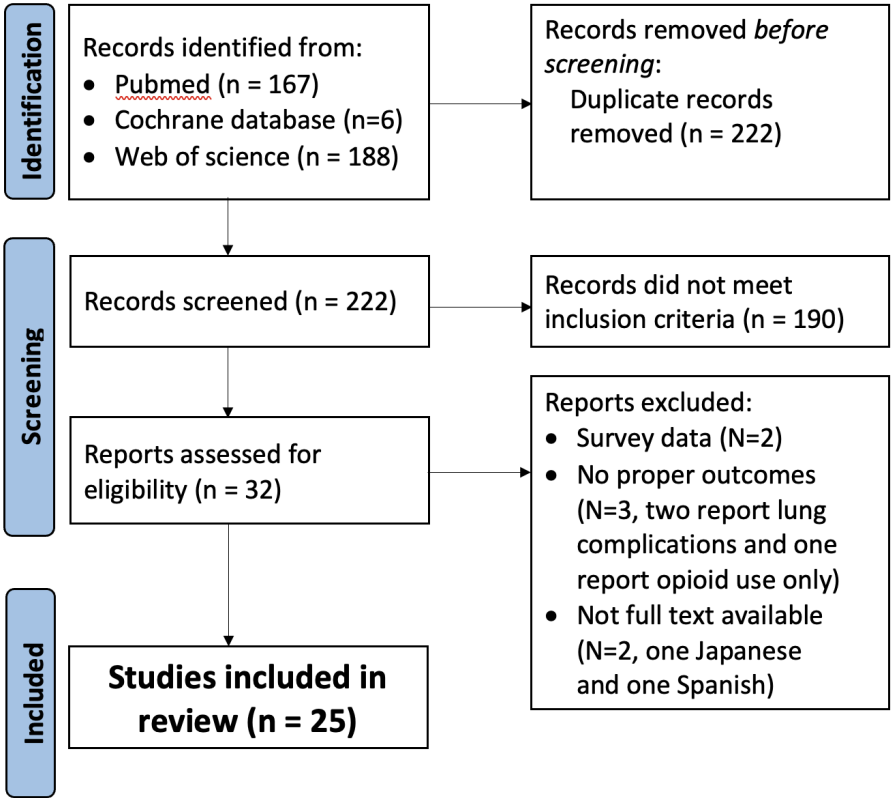


Thoracoscopic versus open repair for esophageal atresia

Chen Yong, Stella Sabbatini, Pierro Agostino

What are the advantages and risks of thoracoscopic versus open repair for esophageal atresia?

Identification of studies via databases and registers



Included studies (N=25)

- 2 RCT
- 2 case control
- 21 cohort studies

Sample size:

- Thoracoscopic: 755 cases (20 in RCTs)
- Open repair: 2215 cases (20 in RCTs)

**PRISMA
Flowchart**

Total 25 studies

Included for meta-analysis (N=14)

- 2 RCTs
- 1 prospective case-control
- 1 retrospective case-control
- 10 retrospective cohorts

Sample Size:

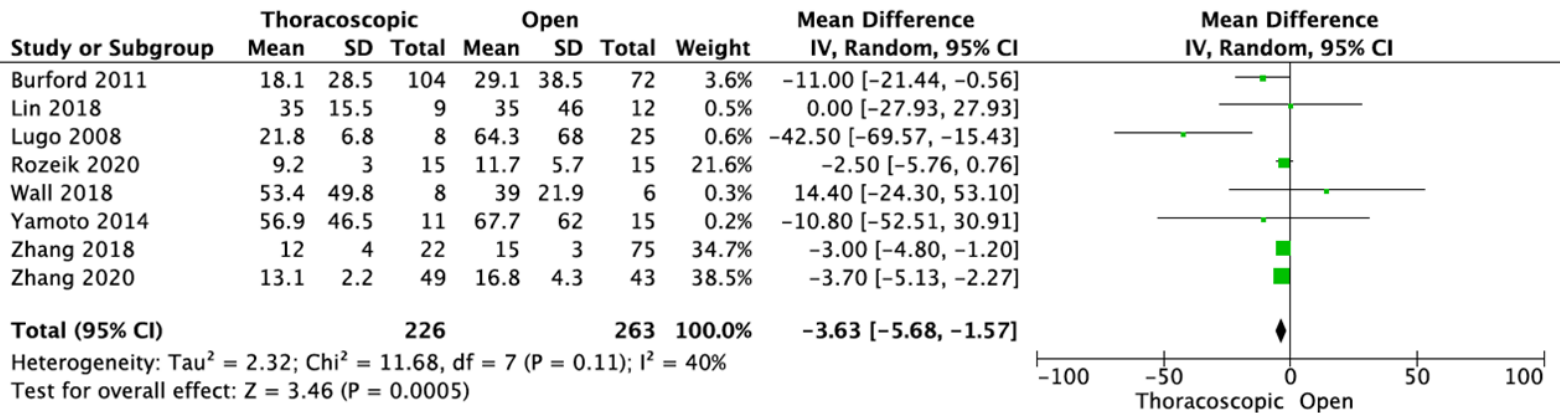
- Thoracoscopy: 309
- Open repair: 452

Excluded for significant selection bias (N=11)

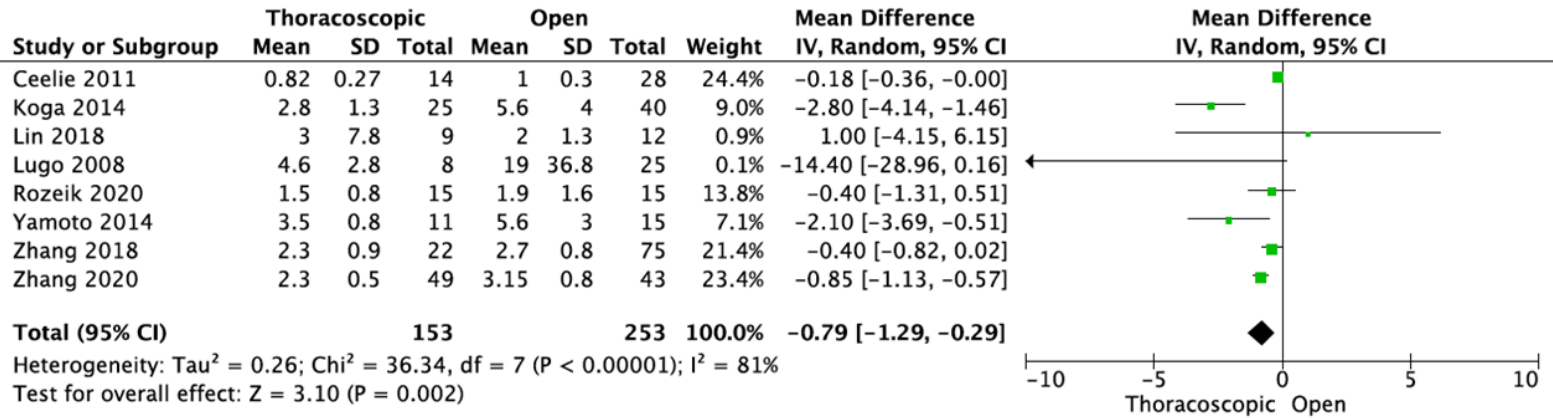
- gestational age
- weight
- comorbidities

Type of atresia:

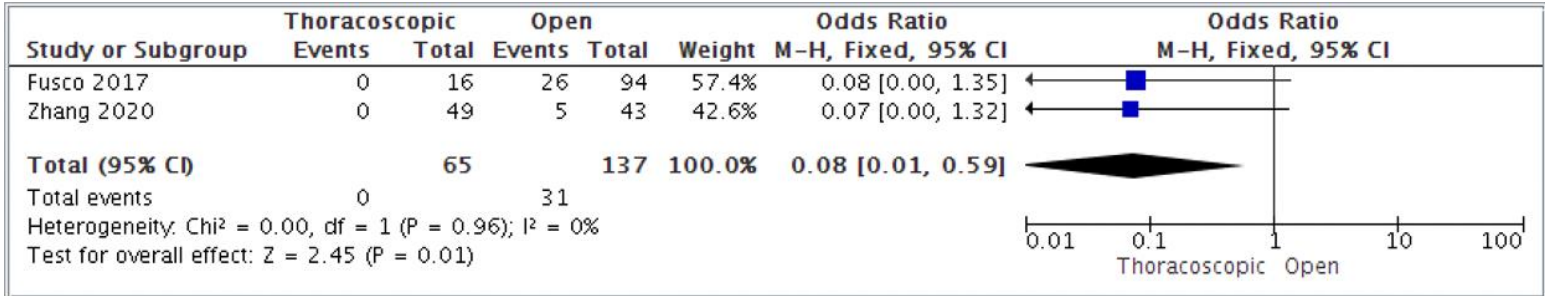
- Type A (4)
- **Type C (388)**
- Type D (1)
- Unknown (368)



Length of stay shorter in thoracoscopy vs open repair
 (MD - 3.63 days , $P=0.0005$)



Ventilation time shorter in thoracoscopic vs open repair
 (MD -0.79 days, P=0.002)



Less musculoskeletal sequelae in thoracoscopic (0%) vs open (22.6%) (P=0.01)

Outcomes	Included studies	Thoracoscopy	Open repair	P value
Operative time	11	176 min	156 min	0.06
Time to start feeding	6	10 days	14 days	0.37
Anastomotic leakage	9	12%	12%	0.99
Anastomotic stricture	10	13%	23%	0.31
Recurrent TEF	4	2%	4%	0.66
Fundoplication	4	21%	13%	0.24
Mortality	5	2%	3%	0.56

- Thoracoscopy appears superior to open repair for esophageal atresia with fewer musculoskeletal sequelae, shorter ventilation time and length of hospital stay.
- Mortality, time to first feeding, operative time, recurrent TEF, fundoplication rate, anastomotic leak, and stricture are comparable between two approaches.

Recommendation: **Grade B**

Evidence Based Guidelines – EA/TEF

Magnetic Anastomosis

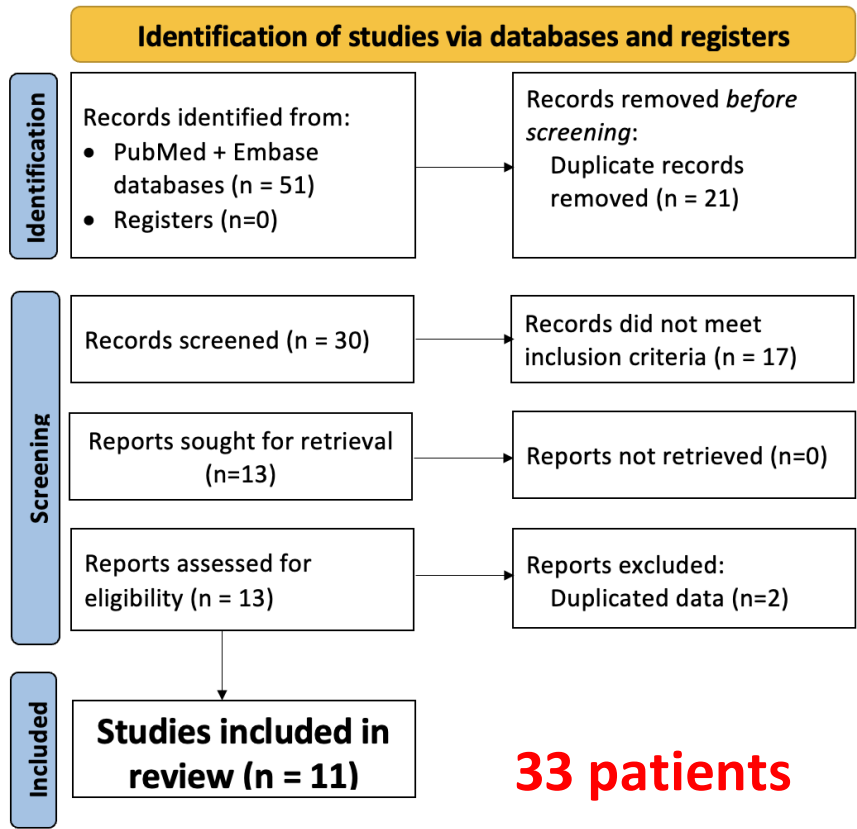
Luques L., Sabbatini S.

The Hospital for Sick Children (SickKids), Toronto, ON, Canada

Background

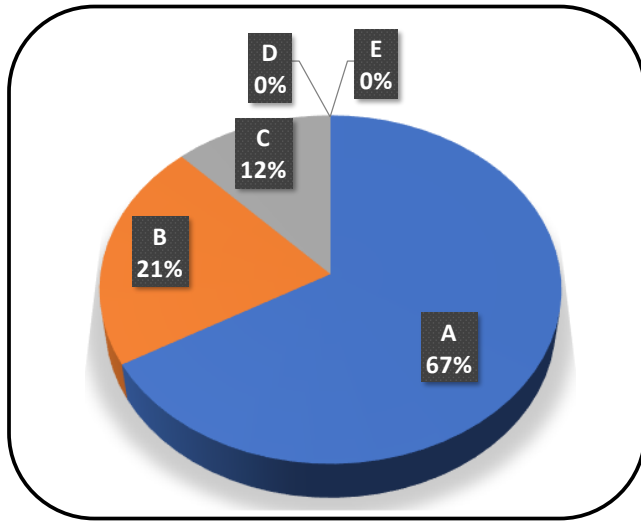
- First reported for treatment of EA in humans in 2009 (Zaritzky *et al.*)
- Few publications: single cases or short series.
- Possible publication bias towards successful treatment.
- Variable indications – salvage procedure, unsuitable patients for surgery or primary repair
- Variable dispositive design and procedural preparation

PRISMA Flowchart

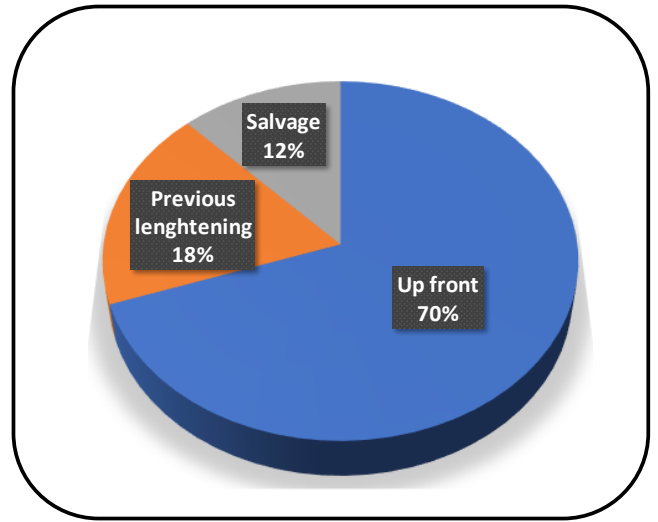


Results – Indications

Type of EA/TEF (Gross Classification)



Surgical history



Results – Outcomes

- Follow up: 25 months (7-112)
- Success rate: 73% (24/33 patients)
- Re-operation rate: 12% (4/33 patients)
- Mortality rate: 0%
- Complications:
 - Leak 12% (4/33)
 - Stricture 91% (30/33)
 - Others 12% (4/33)

Summary and Recommendations

- Promising non-invasive solution.
- Patient selection.
- Success bias should be addressed with prospective studies.
- **No evidence-based recommendations** can be done regarding indications and technic with the available evidence.
- Magnetic anastomosis should be reserved for patients participating in prospective studies (**Strength of recommendation: weak**)

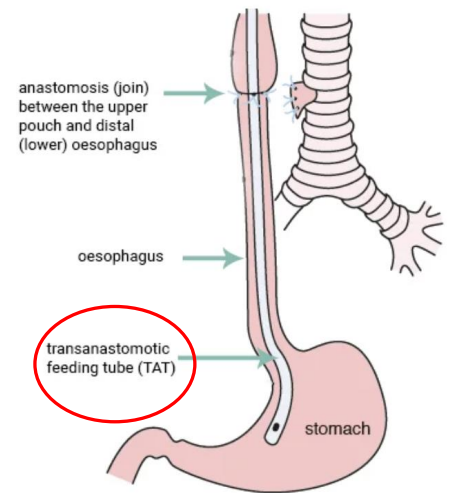
***Esophageal Atresia and
Tracheoesophageal Fistula***

Evidence for treatment and recommendations

Elke Zani-Rittenstock

Post-operative trans-anastomotic tube:

Does positioning of a trans-anastomotic tube increase postoperative complications?



RESEARCH ARTICLE

Open Access



What is the impact of the use of transanastomotic feeding tube on patients with esophageal atresia: a systematic review and meta-analysis

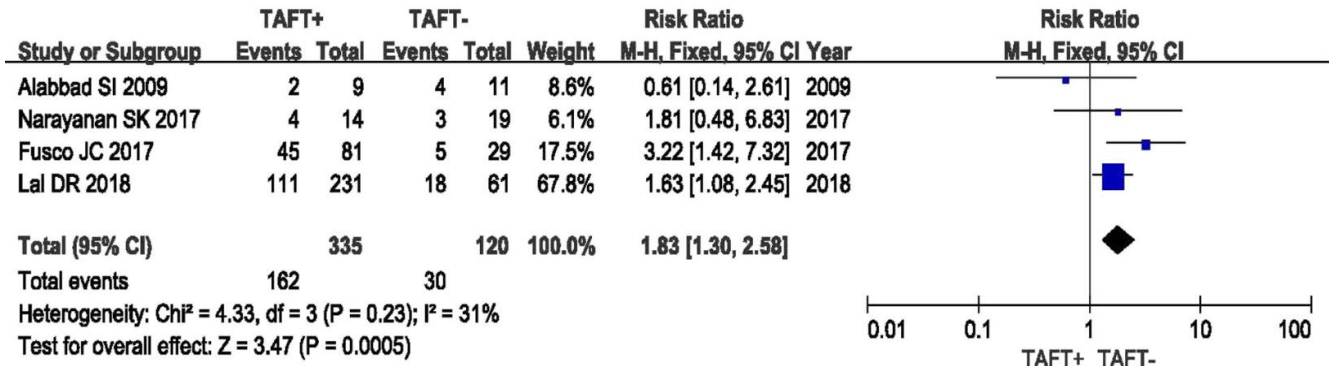
Chuan Wang^{1†}, Liwei Feng^{2†}, Yanan Li³ and Yi Ji^{4*} 

Table 1 Characteristics of included studies

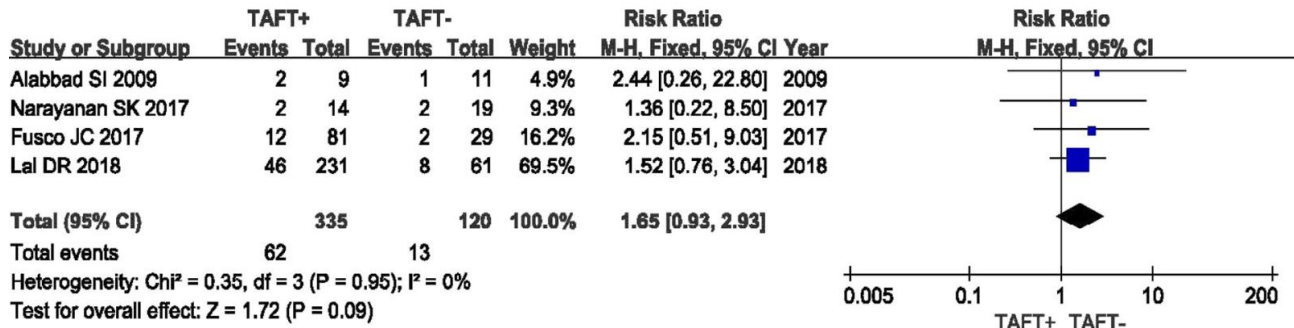
Study	Study type	Sample size	Age at surgery (day)	Gestational age (week)	Birth weight (kg)	weight (kg)	NOS
Alabbad SI 2009	OCS (retrospective)	TAFT+:9	NA	39.00 ± 2.1	3.13 ± 0.55	NA	7
		TAFT-:11	NA	37.64 ± 2.5	2.82 ± 0.69	NA	
Fusco JC 2017	OCS (retrospective)	TAFT+:81	2.4	NA	NA	2.69	7
		TAFT-:29	2.3	NA	NA	2.71	
Narayanan SK 2017	OCS (retrospective)	TAFT+:14	NA	35.64 ± 2.60	2.30 ± 0.23	NA	7
		TAFT-:19	NA	36.52 ± 2.20	2.50 ± 0.32	NA	
Lal DR 2018	OCS (retrospective)	TAFT+:231	NA	NA	NA	NA	6
		TAFT-:61	NA	NA	NA	NA	

TAFT ransanastomotic feeding tube, OCS observational clinical study, NOS Newcastle-Ottawa scale, NA not available

Total of 455 patients

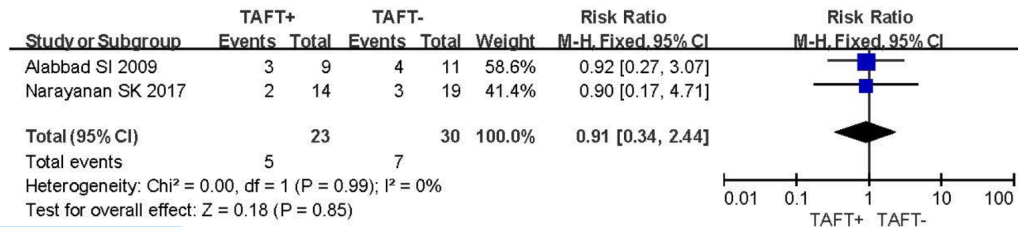


Use of TAT tube significantly increases esophageal stricture rate

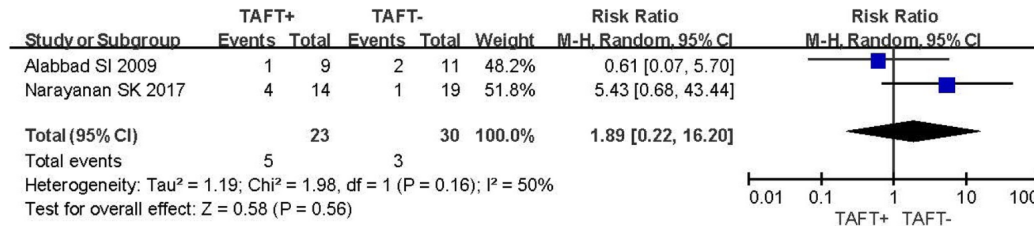


Use of TAT tube is not associated with an increase in anastomotic leakage rate

3.1 Sepsis

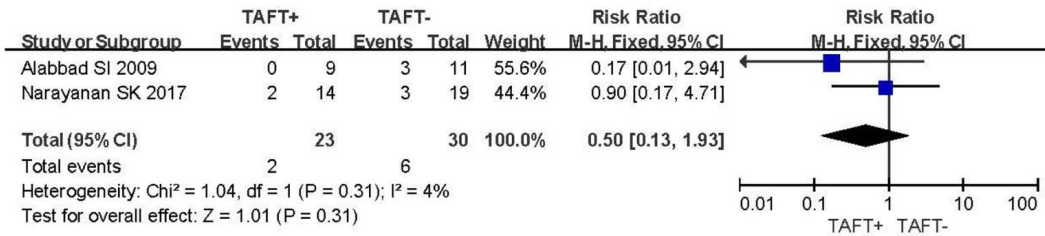


4.1 Tracheomalacia



No association between TAT tube use and sepsis or tracheomalacia

5.1 Gastroesophageal reflux.

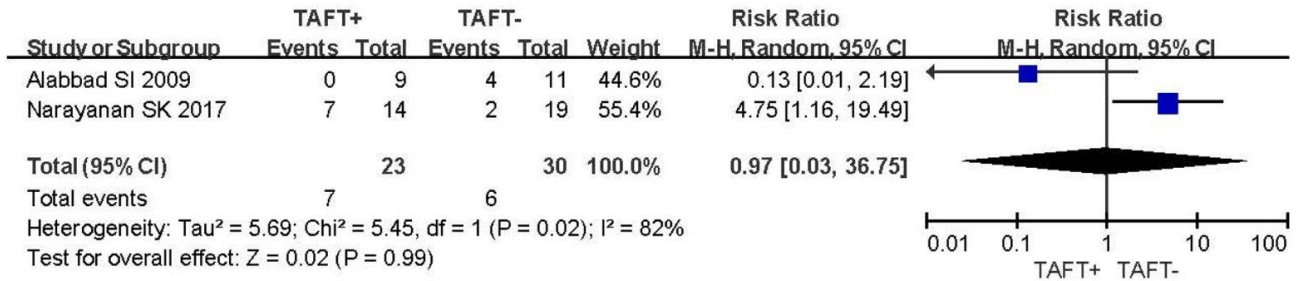


6.1 Wound infection



No association between TAT tube use and GERD or wound infection

7.1 Pneumonia



No association between TAT tube and pneumonia



ERNICA (2018)



ERNICA

Original Article

ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative, and Postoperative Management

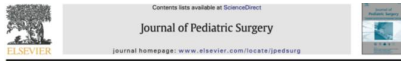
Carmen Dingemann¹ Simon Eaton² Gunnar Aksnes³ Pietro Bagolan⁴ Kate M. Cross⁵
Paolo De Coppi^{2,5} JoAnne Fruithof⁶ Piergiorgio Gamba⁷ Steffen Husby⁸ Antti Koivusalo⁹
Lars Rasmussen¹⁰ Rony Sfeir¹¹ Graham Slater¹² Jan F. Svensson¹³ David C. Van der Zee¹⁴
Lucas M. Wessel¹⁵ Anke Widenmann-Grolig¹⁶ Rene Wijnen¹⁷ Benno M. Ure¹

Eur J Pediatr Surg 2020;30:326–336.

ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative, and Postoperative Management

24	In cases with suspected right descending aorta, a right-sided thoracic approach is the first option	+	76.9	10/13	8 (1–9)
25	The azygos vein should be preserved whenever possible	–	71.4	10/14	6.5 (2–9)
26	The tracheoesophageal fistula should preferably be closed by transfixing suture	+	100	14/14	9 (6–9)
27	The esophageal anastomosis should be preferably performed with absorbable sutures	+	85.7	12/14	8 (1–9)
28	The esophageal anastomosis should be preferably performed with interrupted sutures	+	100	14/14	9 (6–9)
29	A transanastomotic tube should be routinely inserted	+	80	12/15	8 (1–9)
30	A chest drain should be routinely placed	–	21.4	3/14	1 (1–9)
31	The thoracoscopic approach is a viable option	+	87.5	14/16	9 (5–9)

2018



Challenging surgical dogma in the management of proximal esophageal atresia with distal tracheoesophageal fistula: Outcomes from the Midwest Pediatric Surgery Consortium

Dave R. Lal^{1*}, Samir K. Gadepalli², Cynthia D. Dowward³, Daniel J. Ostlie⁴, Peter C. Minneci⁵, Ruth M. Swedler⁶, Thomas H. Chelius⁷, Laura Cassidy⁸, Cooper T. Rapp⁹, Deborah Billime¹⁰, Steven Bruch¹¹, R. Carland Burns¹², Katherine J. Deans¹³, Mary E. Fallat¹⁴, Jason D. Fraser¹⁵, Julia Grabowski¹⁶, Ferdynand Hebel¹⁷, Michael A. Helmuth¹⁸, Ronald B. Hirschi¹⁹, Rashmi Kabre²⁰, Jonathan Kohler²¹, Matthew P. Landman²², Charles M. Leys²³, Grace Z. Mak²⁴, Jessica Raque²⁵, Beth Rymeski²⁶, Jacqueline M. Saito²⁷, Shawn D. St. Peter²⁸, Daniel von Allmen²⁹, Brad W. Warner³⁰, Thomas T. Sato³¹, on behalf of the Midwest Pediatric Surgery Consortium

- Multi-centre, retrospective study
- 2009-2014
- 292 patients

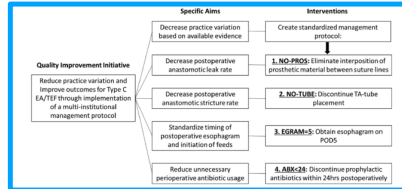
- Stricture rate 48%
- TAT tube is associated with increased risk for anastomotic stricture

2021



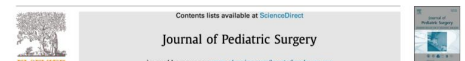
Clinical outcomes following implementation of a management bundle for esophageal atresia with distal tracheoesophageal fistula*

Christina M. Bence¹, Beth Rymeski², Samir Gadepalli³, Thomas T. Sato⁴, Peter C. Minneci⁵, Cynthia Dowward⁶, Ronald B. Hirschi⁷, Richa A. Amin⁸, R. Carland Burns⁹, Linda Cherny-Spafford¹⁰, Catherine M. Courtney¹¹, Katherine J. Deans¹², Mary E. Fallat¹³, Jason D. Fraser¹⁴, Julia E. Grabowski¹⁵, Michael A. Helmuth¹⁶, Rashmi D. Kabre¹⁷, Jonathan E. Kohler¹⁸, Matthew P. Landman¹⁹, Amy E. Lawrence²⁰, Charles M. Leys²¹, Grace Z. Mak²², Elissa Fort²³, Jacqueline M. Saito²⁴, Jared Silverberg²⁵, Mark B. Slidell²⁶, Shawn D. St. Peter²⁷, Misty Trout²⁸, Sarah Walker²⁹, Tiffany Wright³⁰, Dave R. Lal^{31*}, on behalf of the Midwest Pediatric Surgery Consortium



- Bundle implementation including no TAT tube use
- Significant reductions in postoperative strictures when TA-tubes are not used

2022



Acid suppression duration does not alter anastomotic stricture rates after esophageal atresia with distal tracheoesophageal fistula repair: A prospective multi-institutional cohort study

Alexis N Bowder^{1*}, Christina M. Bence², Beth A Rymeski³, Samir K. Gadepalli⁴, Thomas T. Sato⁵, Aniko Szabo⁶, Kyle Van Arendonk⁷, Peter C. Minneci⁸, Cynthia D. Dowward⁹, Ronald B. Hirschi¹⁰, Troy Markel¹¹, Cathleen M. Courtney¹², Katherine J. Deans¹³, Mary E. Fallat¹⁴, Jason D. Fraser¹⁵, Julia E. Grabowski¹⁶, Michael A. Helmuth¹⁷, Rashmi D. Kabre¹⁸, Jonathan E. Kohler¹⁹, Matthew P. Landman²⁰, Amy E. Lawrence²¹, Charles M. Leys²², Grace Mak²³, Elissa Fort²⁴, Jacqueline Saito²⁵, Jared Silverberg²⁶, Mark B. Slidell²⁷, Shawn D. St. Peter²⁸, Misty Trout²⁹, Tiffany N. Wright³⁰, Dave R. Lal³¹, on behalf of the Midwest Pediatric Consortium

- Prospective, multi-centre study (156 pts)
- Acid suppression

- Acid suppression did not decrease stricture rate, but no TAT tube does!

Findings do not support routine use of TAT tube

- Based on the evidence currently available in the literature (low, no RCT's), positioning of a trans-anastomotic tube post TEF repair seems to increase the risk for anastomotic stricture.
- Routine placement of a trans-anastomotic tube is therefore NOT recommended.
- Grade of recommendation: Grade B

Postoperative strategy - Ventilation, muscle paralysis and neck flexion

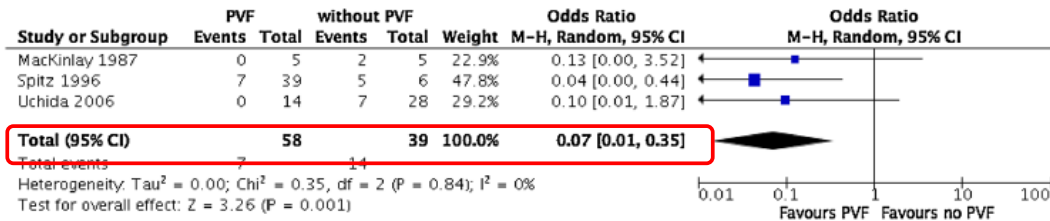
Naho Fujiwara and Mashriq Alganabi

The Hospital for Sick Children (SickKids),

Toronto, Canada

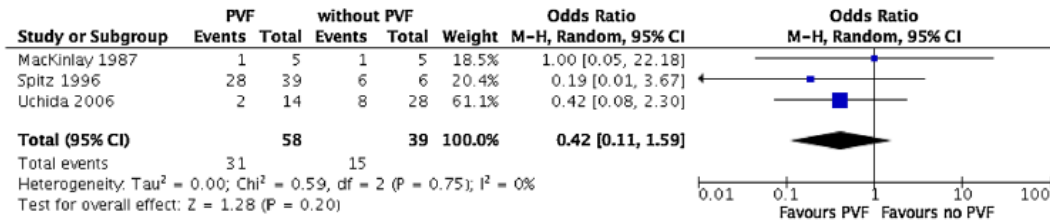
Previous Systematic Review Data

A Association of PVF with the occurrence of "anastomotic leak"



Abbreviations: PVF = elective post-operative muscle paralysis, positive-pressure ventilation, and head flexion.

B Association of PVF with the occurrence of "anastomotic stricture"

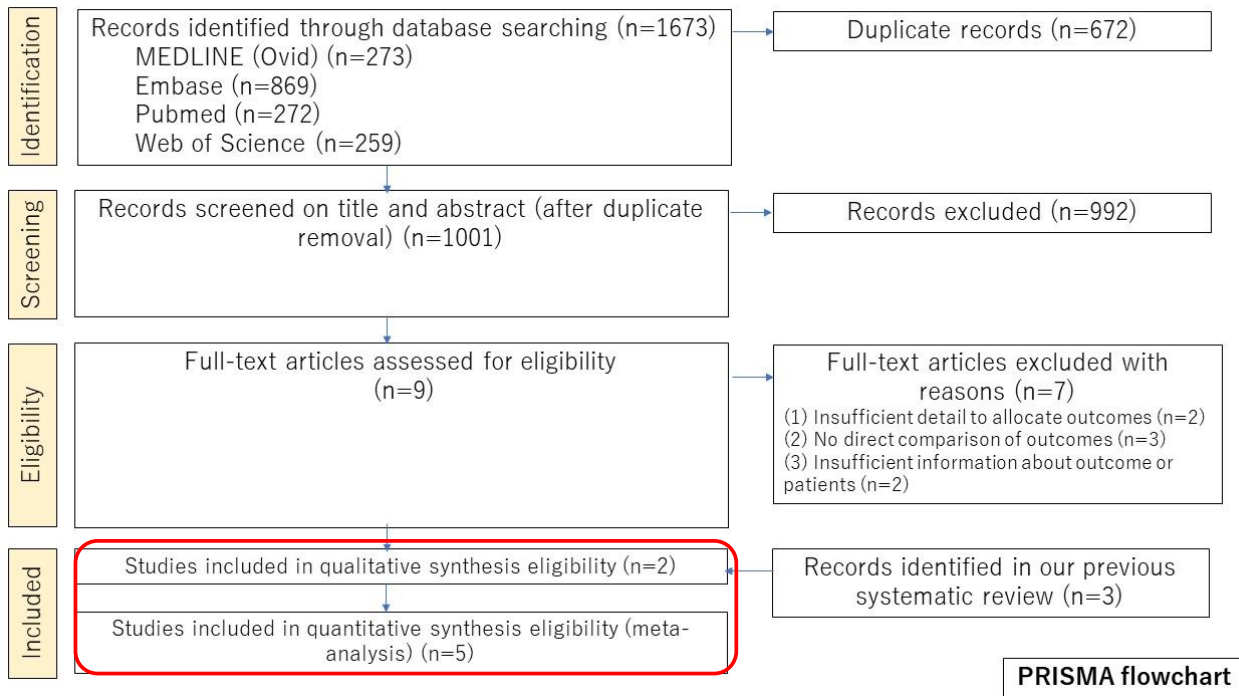


Abbreviations: PVF = elective post-operative muscle paralysis, positive-pressure ventilation, and head flexion.

O'Connell JS, et al. 2018

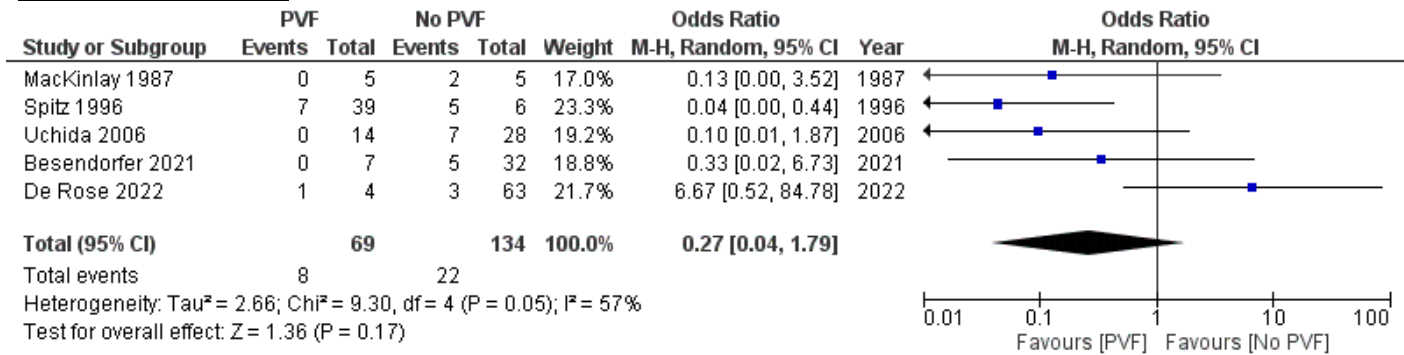
Now we are updating this data

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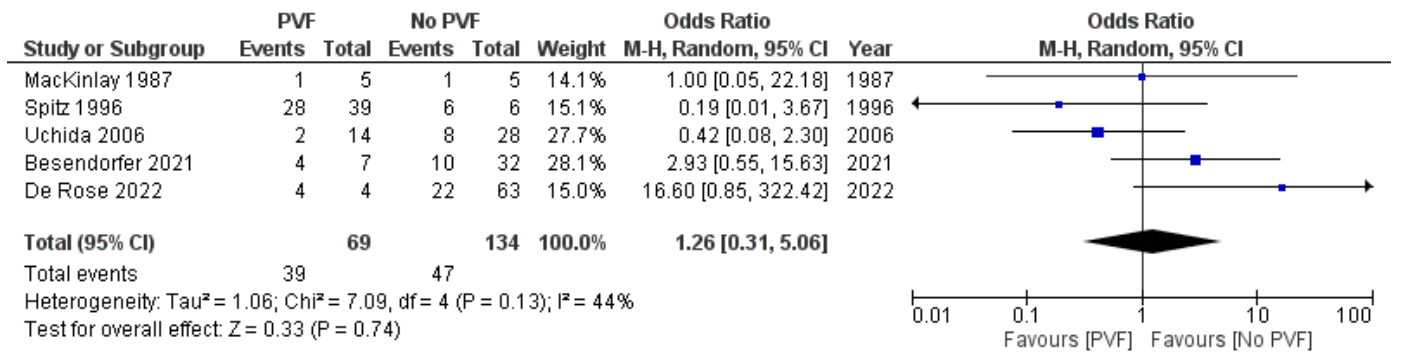


Results

Anastomotic Leaks



Anastomotic Strictures



Risk of Bias Assessment (ROBINS-I)

Author/Year	Bias due to confounding	Bias in the selection of participants into the study	Bias in classification of interventions	Bias due to deviations from intended interventions	Bias due to missing data	Bias in measurement of outcome	Bias in selection of the reported results	Overall Bias
MacKinlay 1987	?	?	+	+	+	+	?	?
Spitz 1996	?	?	+	+	+	+	?	?
Uchida 2006	?	?	+	+	+	+	+	?
Besendorfer 2021	?	?	?	+	+	+	+	?
De Rose 2022	?	?	?	+	+	x	+	?

 low risk of bias
  moderate risk of bias
  serious risk of bias
  critical risk of bias

Conclusions

- On the basis of the existing evidence and our analysis, elective post-operative PVF did **not significantly change** the incidence of anastomotic leaks or strictures.
- **The level of evidence is low**, also the level of risk was assessed to be of moderate risk due to the groups compared being retrospective without necessarily matched baseline characteristics.

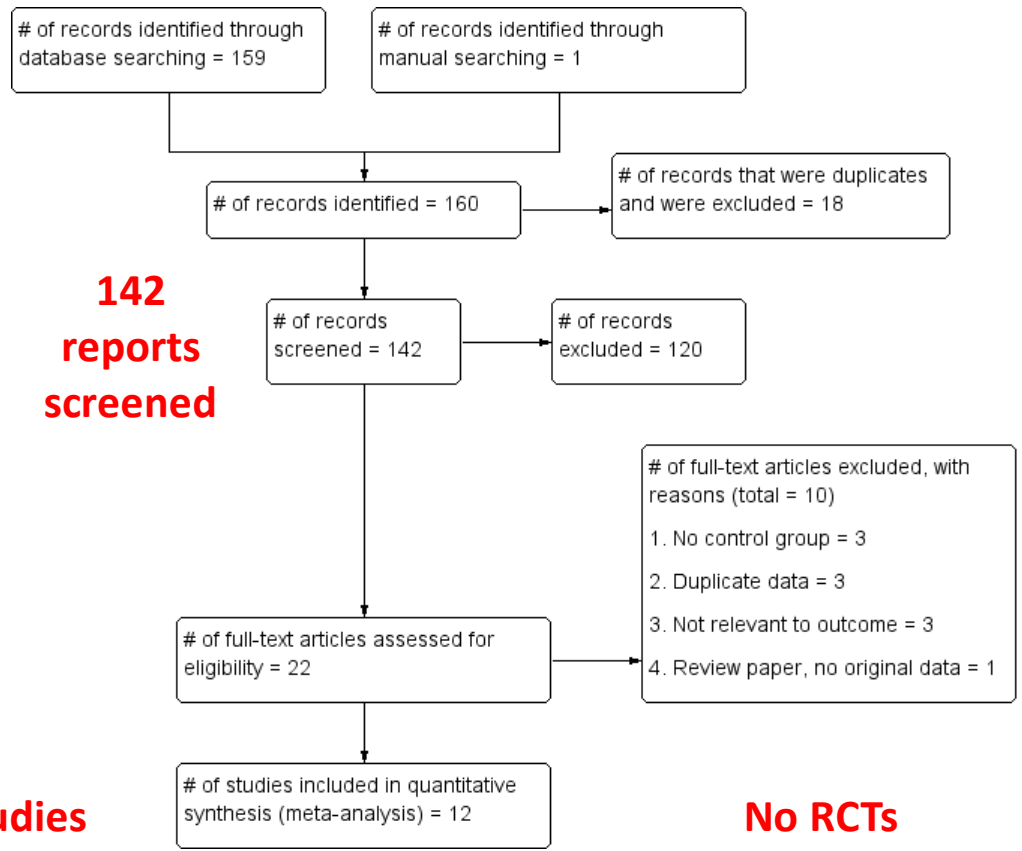
Does routine use of antacid medication reduce incidence of anastomotic stricture follow EA/TEF repair?

Nigel Hall

- **Population:** Children undergoing repair of EA +/- TEF
- **Intervention:** Routine antacid medication
- **Comparison:** None or symptomatic treatment only
- **Outcome:**
 - **Primary:**
 - Anastomotic stricture
 - **Secondary**
 - GERD
 - Anastomotic leak
 - Esophagitis



**PRSIMA
Flowchart**

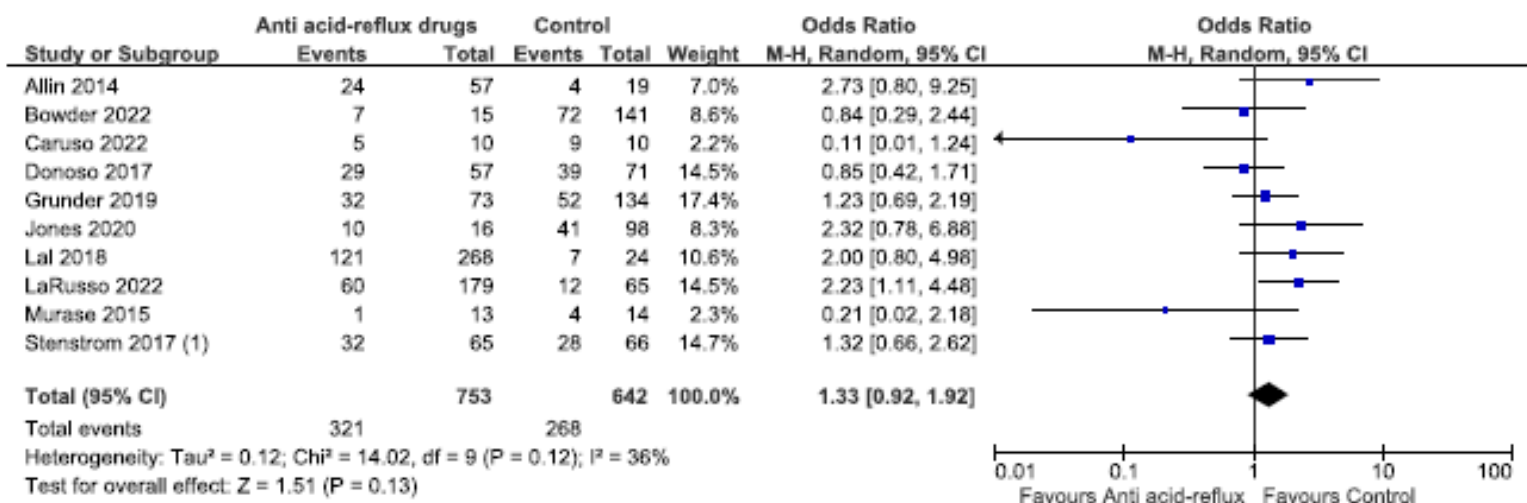


High risk of bias in majority

Retrospective
Poorly defined primary outcome

Overall grade of recommendations

	Confounding	Measurement of exposure	Selection of Participants	Post-exposure Interventions	Missing Data	Measurement of Outcome	Selection of Reported Result	Overall risk of Bias
Allin 2014	+	+	+	?	+	-	+	-
Bowder 2022	+	?	+	+	?	-	+	-
Caruso 2022	-	+	?	?	?	-	+	-
Donoso 2017	+	-	+	?	+	-	+	-
Grunder 2019	+	+	+	?	+	+	+	+
Hagander 2012	-	+	+	+	+	+	+	+
Jones 2020	+	+	+	+	?	-	+	-
Lal 2018	+	+	+	+	+	-	+	-
LaRusso 2022	+	+	+	+	+	-	+	-
Murase 2015	-	+	+	+	+	-	+	-
Stenstrom 2017 (1)	-	+	+	+	+	+	+	+
Yasuda 2019	-	+	+	+	+	+	+	+



No difference in incidence of stricture
 (OR 1.33 (95%CI 0.92-1.92), P=0.13)

Outcome	Studies	Participants	Effect Estimate
GORD	3	395	0.52 [0.24, 1.13]
Anastomotic leak	4	674	0.84 [0.46, 1.55]
Oesophagitis/Oesophageal erosion	1	573	1.16 [0.40, 3.38]

No statistically significant association between routine use of antacids and GERD, Anastomotic leak or Esophagitis

Conclusions

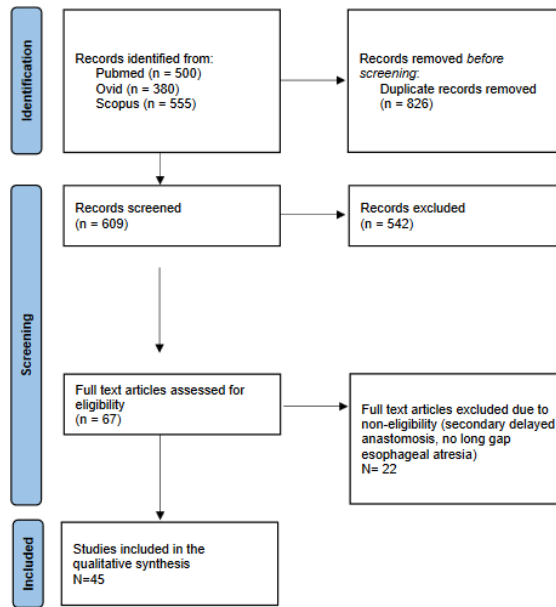
- No evidence to support or refute the routine use of antacid medication following EA repair to reduce incidence of anastomotic stricture
- Some evidence of potential side effects of antacid medication
- Lack of high quality studies

Recommendation **GRADE B**

- **Routine use of antacid medication not recommended based on existing data**

PRISMA 2020 flow diagram for new systematic reviews which included searches of databases and registers only

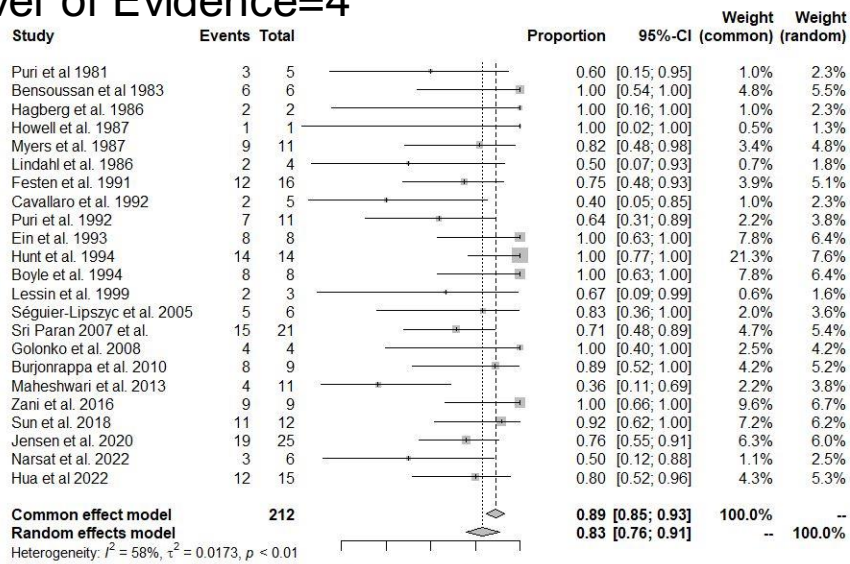
PRISMA 2009 Flow Diagram





Question 3: What is the success rate of delayed primary anastomosis (patients on full enteral feeds)?

- 22 articles, Level of Evidence=4



Overall, 83% of patients were on full enteral feeds (95%CI: 76-91%, $I^2=58\%$; $p<0.01$)



Question 3: How long should the surgery be postponed?

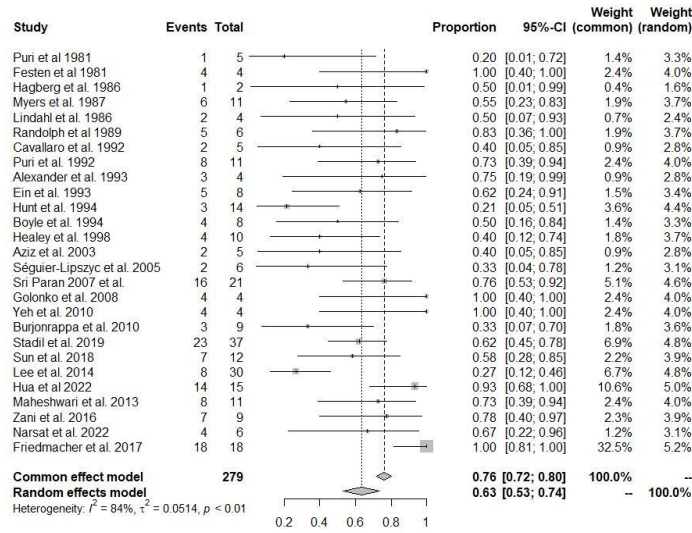
- 26 articles
- Range: several days up to 34 weeks
- Median time until repair:
 - 11.6 weeks

An evaluation of the maximum duration or the period of time surgeons should wait before esophageal replacement is taken into consideration, has not been conducted



Question 3: What are the complications of delayed primary anastomosis?

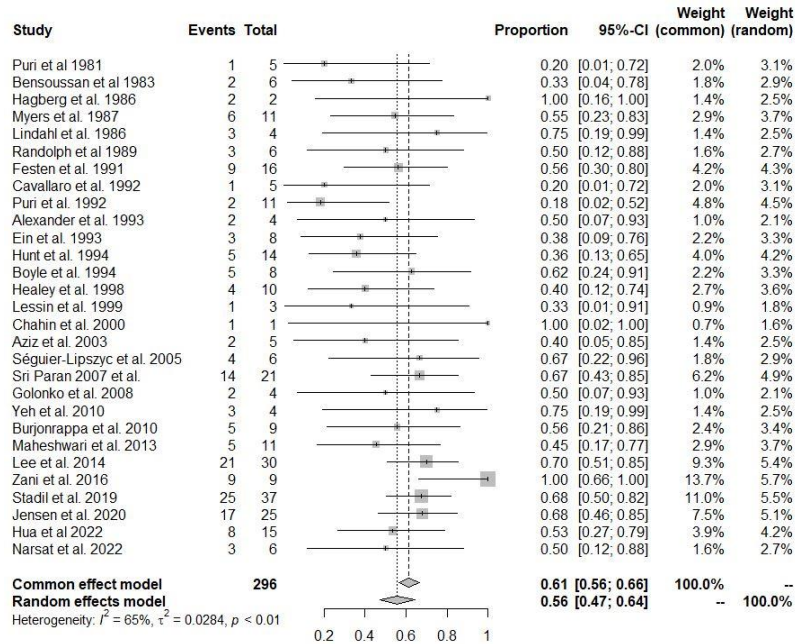
Overall: **523 complications in 468 patients**



Strictures are to be expected in 63% (95%CI: 53-74%; $I^2=84\%$; $p<0.01$)



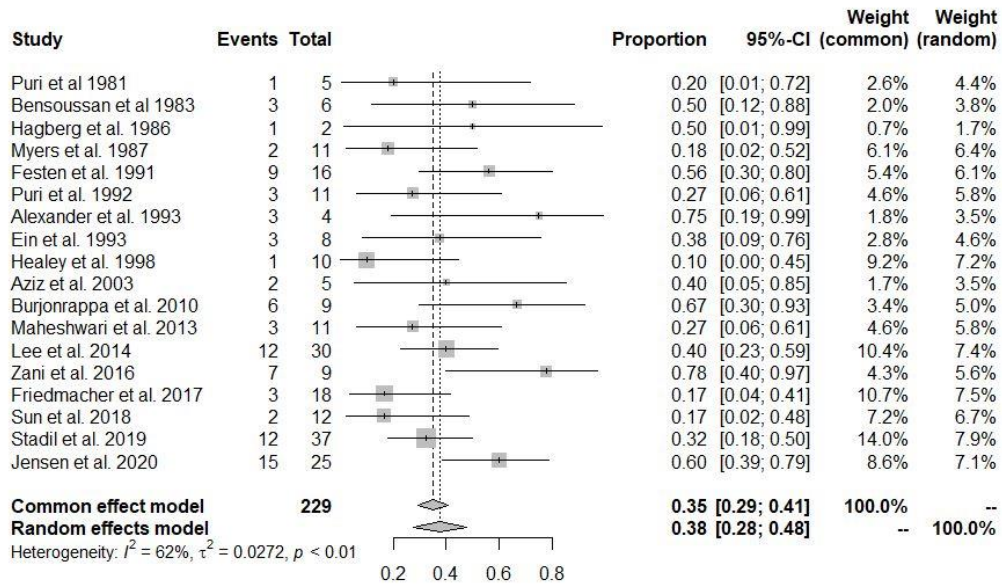
Question 3: What are the complications of delayed primary anastomosis?



GERD is to be expected in 56% (95%CI: 47-64%; $I^2=65\%$; $p<0.01$)



Question 3: What are the complications of delayed primary anastomosis?



Postoperative anastomotic leakage in 38%
 (95%CI: 28-48%; $I^2=62\%$; $p<0.01$)



Recommendations:

- DPA may be offered as an option for long-gap esophageal atresia
- No recommendation can be drawn for the time until DPA can/should be performed
- **Short- and long term complications are common** demonstrating the necessity of long-term follow-up in this patient population

Grade D

Topic: Management of long gap c. Kimura procedure

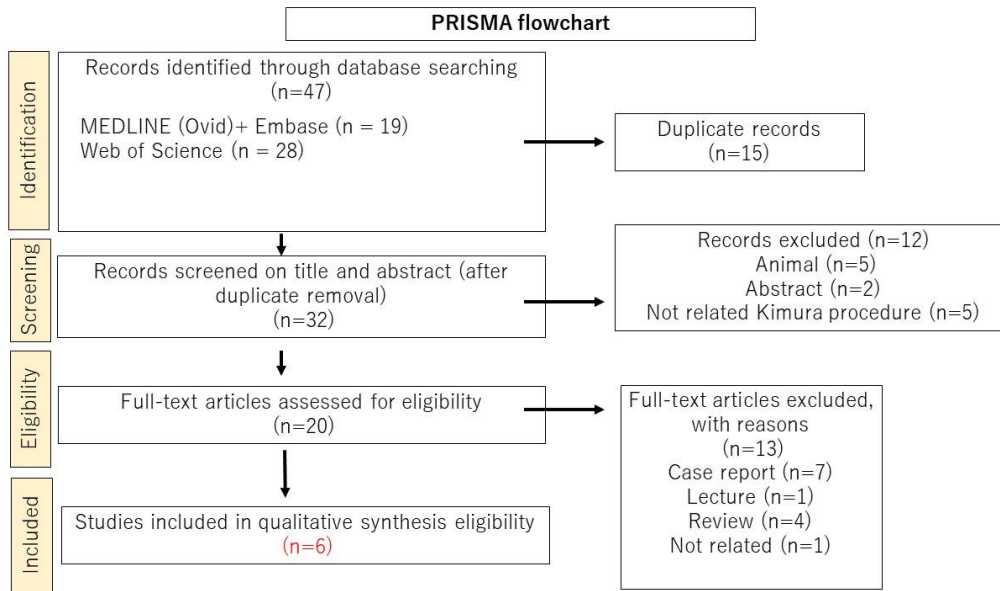
Naho Fujiwara

The Hospital for Sick Children (SickKids),

Toronto, Canada

Topic: Management of long gap c. Kimura procedure

- Q
1. What is the success rate of “Kimura procedure”?
 2. Which are the complications of “Kimura procedure”?



6

Literature on Kimura procedure

	N of pts	Complications	
		Leaks	Stenosis
Kimura, 2001	12	3	12
Takamizawa, 2005	7	2	7
Tamburri, 2009	12	3	6
Miyano, 2013	4	2	4
Sroka, 2013	6	6	5
Oliver, 2021	3	0	2

Kimura Procedure

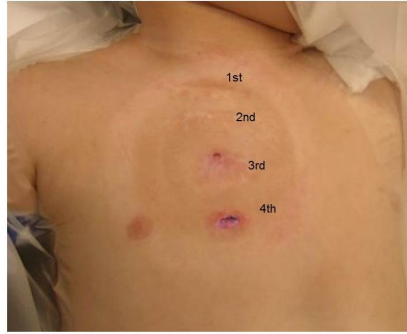


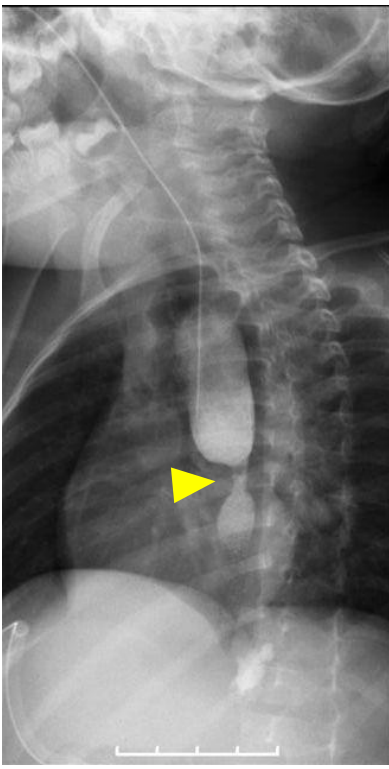
Photo kindly provided by Prof. Yamataka

Outcome

Patient	Age (mths)	BW (kg)	Gap (vert)	Op time (hours)	Open	Complications Post-Op	Dilations	Stenting (days)	Extubation (days)	Feeding (days)
1	27	13	2	9	(-)	stenosis	> 10 times	8	2	7
2	25	9	1.5	8	(-)	stenosis	5	3	2	9
3	27	10	2	10	(+)	leakage/stenosis	2	56	5	31
4	10	7.2	0	10	(-)	leakage/stenosis	3	24	5	24

Open: conversion to open, Stenting: postoperative duration until stent tube removal, Extubation: extubation, Feeding: eating was recommenced

Prof. Yamataka's comment



Postoperative Barium Meal

- Recently we are not using Kimura procedure
- All patients require multiple dilation
- The tip of proximal esophagus is severely fibrotic and full thickness. So it is the cause of postoperative stenosis.

Evidence Based Guidelines – EA/TEF

Esophageal Replacement

Luques L.^{1,2}, Baertschiger R.M.¹, Lauriti G.³, Miscia M.E.³,
Morini F.⁴, Mutanen A.⁵, Pierro A.¹

¹ *The Hospital for Sick Children (SickKids), Toronto, ON, Canada;* ² *Hadassah Medical Center, Jerusalem, Israel;*

³ *University of Chieti-Pescara, Pescara, Italy;* ⁴ *Azienda Ospedaliero-Universitaria Meyer, Firenze, Italy;*

⁵ *Helsinki University Central Hospital, Helsinki, Finland.*

Background

- Conservation of the native esophagus is always the best choice.
- When not possible, different techniques for esophageal replacement have been proposed.
- Four main techniques are currently available: **gastric transposition, gastric tube, jejunal interposition** and **colonic interposition**.
- Each technique has its own pros and cons.
- Reports are variable and well conducted comparative studies are lacking.

Outcomes analyzed

1. First outcomes for analysis

- a. **Success rate** defined as full oral feeding within 6 months from surgery.
- b. **Overall complication rate** during three different periods (early, within 30d from surgery; late, between 31d and 1 year and; long-term, after 1 year)
- c. **Overall mortality** within the follow-up period

2. Secondary outcomes

- a. Rate of **specific complications** at the three analyzed periods.

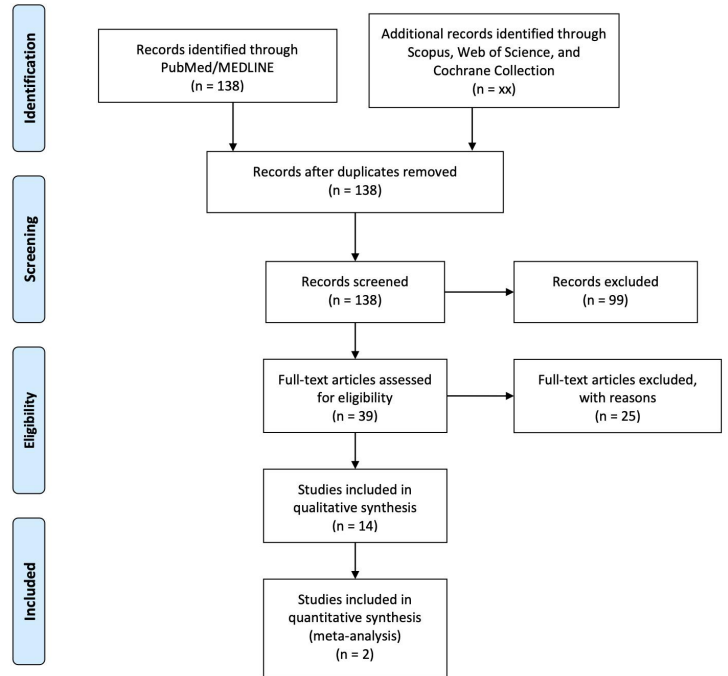
3. Data collection included demographics and mean follow up.

Evidence Based Guidelines – EA/TEF

Esophageal Replacement – Gastric transposition

Lauriti G. , Miscia M.E. , Morini F.

- 17 articles identified (422 patients)
- **14 meet inclusion/exclusion criteria (375 patients)**
- 1 excluded because pooled data with cases of caustic strictures
- 2 excluded because EA undergoing gastric pull-up were <10 patients
- **2 comparative study (1 colon; 1 gastric tube)**



- 375 patients included for analysis
 - 97 EA Gross Type A - 26%
 - 116 EA Gross Type C - 31%
 - 162 N/A - 43%

- Length follow up: 9.3 ± 2.1 years (5/14 studies)

Success Rate	Overall mortality	Complications		
		Early	Late	Long term
72.9% ±19.0% (157/216 patients, 9/14 studies)	7.6% ±6.6% (16/209 patients, 9/14 studies)	Leakage 17±20.9% (54/304 patients)	Strictures 15.7±19.4% (46/292 patients)	N/A
		Pleural effusion 20±11.9% (12/60 patients)	Respiratory 35.1±19.3% (52/148 patients)	N/A
		Re-fistula 12.2±11.2% (7/57 patients)	Dysphagia 28.8±24.7% (21/73 patients)	N/A
			DGR 24.1±25.9% (14/58 patients)	N/A
		Overall 65.6±38.2% (124/189 patients)		

- **Graft failure** was addressed in 2/14 studies and reported in **0/59 patients (0%)**
- Need for **endoscopic dilatation** was addressed in 8/14 studies and reported in **47/200 patients (23.5±30.4%)**

Evidence Based Guidelines – EA/TEF

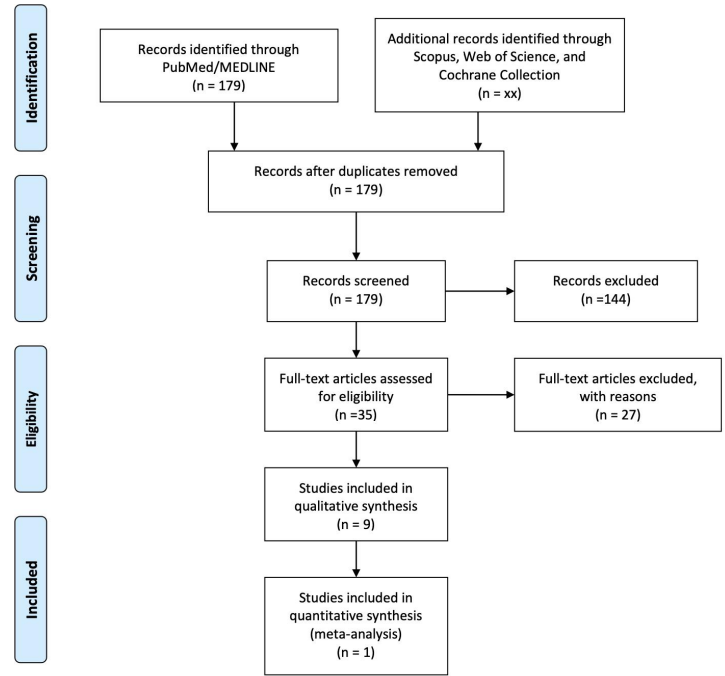
Esophageal Replacement – Gastric Tube

Miscia M.E. , Morini F., Lauriti G.

Gastric Tube - PRISMA

- 9 articles identified (143 patients)
- **9 meet inclusion/exclusion criteria**
- **1 comparative study**

- Only **1/9 papers** published in **current decade**



- 143 patients included for analysis
 - 53 EA Gross Type A - 37%
 - 26 EA Gross Type C - 18%
 - 64 N/A - 45%

- Length follow up: 5.5 ± 2.2 years (4/9 studies)

Gastric Tube - Outcomes

Success Rate	Overall mortality	Complications		
		Early	Late	Long term
100% (56/56 patients, 3/9 studies)	10% ±10.6% (6/60 patients, 4/9 studies)	Leakage 42.5±21.6% (34/80 patients)	Strictures 33.9±19.0% (35/103 patients)	N/A
		Respiratory 35.7% (5/14 patients)	Respiratory 27.5±27.6% (11/40 patients)	N/A
		Re-fistula 40.7±30.6% (11/27 patients)	Dysphagia 24.4±15.4% (11/45 patients)	N/A
		SSI 10.7±5.0% (3/28 patients)	DGR 42.4±36.3% (28/66 patients)	N/A
		Overall N/A		

- **Graft failure** was addressed in 2/9 studies and reported in **3/26 patients (11.5±8.1%)**
- Need for **endoscopic dilatation** was addressed in 7/9 studies and reported in **32/110 patients (29.5±26.0%)**

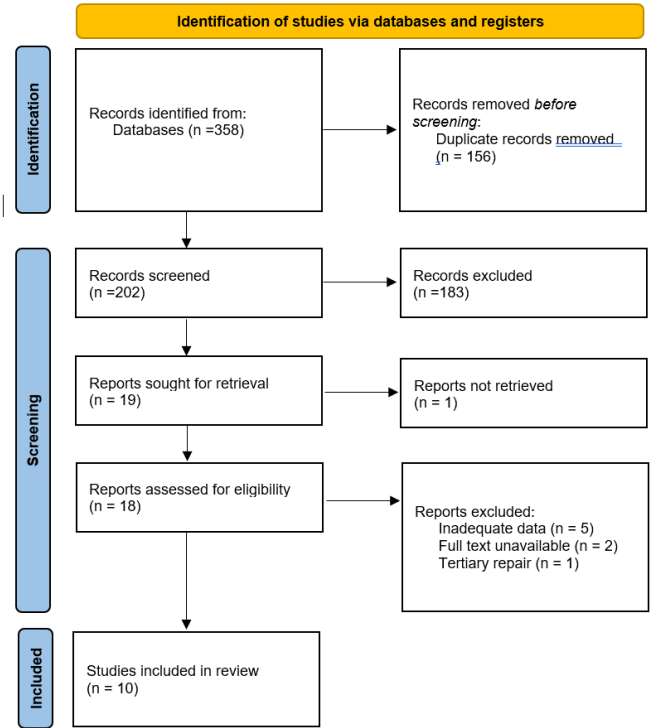
Evidence Based Guidelines – EA/TEF

Esophageal Replacement – Colonic Interposition

Luques L., Mutanen A., Baertschiger R.M.¹

Colonic Interposition - PRISMA

- 19 articles identified, 18 retrieved for review
- 8 articles excluded
 - 5 inadequate data
 - 2 full text unavailable
 - 1 Tertiary repair
- **10 meet inclusion/exclusion criteria (318 patients)**



- 318 patients included for analysis
 - 125 EA Gross Type A - 39%
 - 2 EA Gross Type B - 0.5%
 - 9 EA Gross Type C - 3%
 - 25 EA Gross Type D - 8%
 - 157 EA N/A - 49.5%
- Length follow up: 5.9 ± 2.6 years (7/10 studies)

Colonic Interposition - Outcomes

Success Rate	Overall mortality	Complications		
		Early	Late	Long term
97% (75-100%) (191/197 patients)	4% (0-10%) (14/318 patients)	Leakage 19% (46/238 patients)	Strictures 14% (34/238 patients)	Strictures 5% (11/229 patients)
		Respiratory 6% (14/238 patients)	Re-operation 2% (5/238 patients)	Redundancy 5% (11/229 patients)
		Sepsis 2.5% (6/238 patients)	Others 18% (43/238 patients)	Bowel obstruction 9% (21/229 patients)
		Others 11.34% (27/238 patients)		Others 16% (37/229 patients)
		Overall 37% (19-70%) (118/318 patients)	Overall 26% (3-63%) (83/318 patients)	Overall 27% (4-61%) (83/310 patients)

- **Graft failure** was addressed in 10/10 studies and reported in **13/318 patients (4% - range 0-20%)**
- Need for **endoscopic dilatation** was addressed in 8/10 studies and reported in **30/232 patients (13% - range 0-60%)**

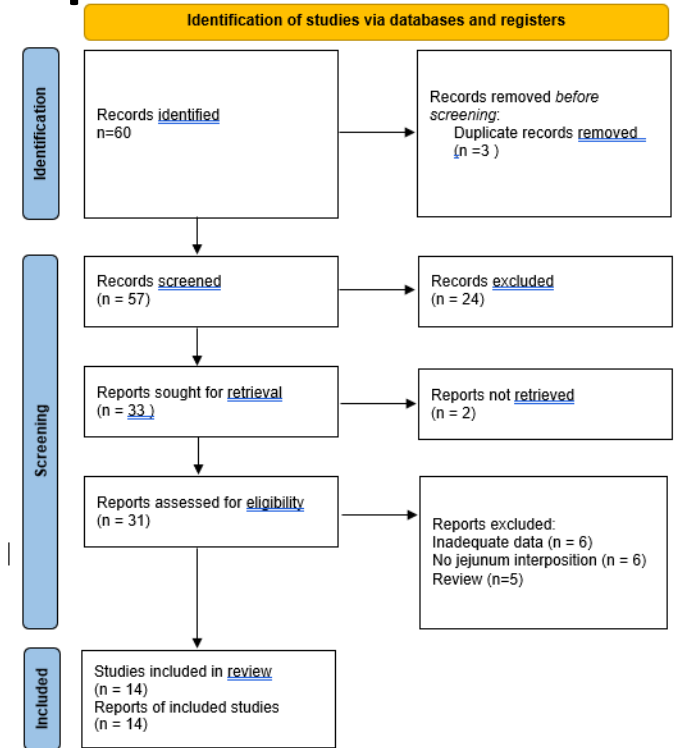
Evidence Based Guidelines – EA/TEF

Esophageal Replacement – Jejunum Interposition

Mutanen A.

Jejunum Interposition - PRISMA

- 33 articles identified, 31 retrieved for review
- 17 articles excluded
 - 6 inadequate data
 - 6 no jejunum interposition
 - 5 review articles
- **14 articles included (176 patients)**



Jejunum Interposition

- Patients and Follow-up

- 176 patients included for analysis
 - 46 EA Gross Type A - 26%
 - 33 EA Gross Type B - 19%
 - 39 EA Gross Type C - 22%
 - 58 N/A - 33%
- With microvascular anastomosis in 51/176 (29%), 4/14 studies
- Length follow up: median 2.7 years (9/14 studies)

Success Rate	Overall mortality	Complications		
		Early	Late	Long term
58% (33-100%) (12/14 studies)	8% (33-100%) (11/14 studies)	Leakage 18% (0-60%) (33/188 patients, 11/14 studies)	Strictures 24% (9-50%) (34/142 patients, 8/14 studies)	Strictures 34% (11-53%) (20/59 patients, 4/14 studies)
		Respiratory 12% (0-17%) (12/104 patients, 5/14 studies)	Re-op 22% (10-29%) (28/125 patients, 5/14 studies)	Redundancy 7% (5-10%) (4/58 patients, 3/14 studies)
		Sepsis 12% (3/25 patients, 2/14 studies)		GI symptoms 39% (5-87%) (17/44 patients, 3/14 studies)
		Overall 45% (5-93%) (73/161 patients, 9/14 studies)	Overall 50% (15-60%) (45/90 patients, 3/14 studies)	N/A

- **Graft failure** was addressed in 10/14 studies and reported in the **range of 0-33%**
- Need for **endoscopic dilatation** was addressed in 6/14 studies and reported in **26/78 patients (33%)**

- Well conducted **comparative studies are lacking**
- The type and rate of **complications vary widely** between the different techniques and the analyzed periods of time
- Despite that some medium and long-term studies are available, the comparison is difficult due to **high heterogeneity of the analyzed outcomes**
- Grade of recommendation: **Grade C/D**



Esophageal Atresia: Tracheomalacia

Evidence for treatment and recommendations

Ramon Gorter, Paul van Amstel and
Stefaan Tytgat



Esophageal Atresia

Questions → Evidence Based Guidelines

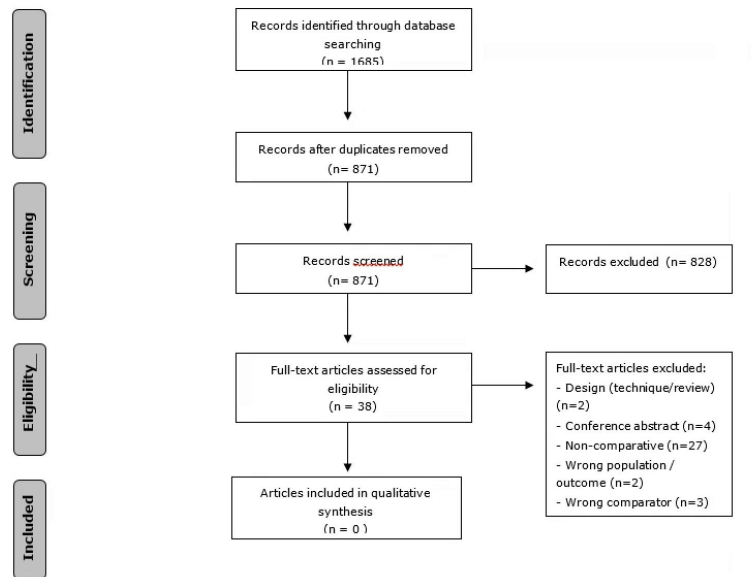
1. What is the preferred surgical procedure for tracheomalacia in children born with esophageal atresia?

Aortopexy vs tracheopexy

2. Is primary tracheopexy during esophageal repair beneficial?

Primary tracheopexy vs no primary tracheopexy

Figure 1. Flowchart of the search and selection procedure of studies.



Conclusion:

No studies are identified comparing aortopexy with (posterior) tracheopexy in children with EA.

Recommendation:

No recommendation can be made regarding the preferred surgical procedure for tracheomalacia in children with EA.

Further studies should focus on this omittance in current literature

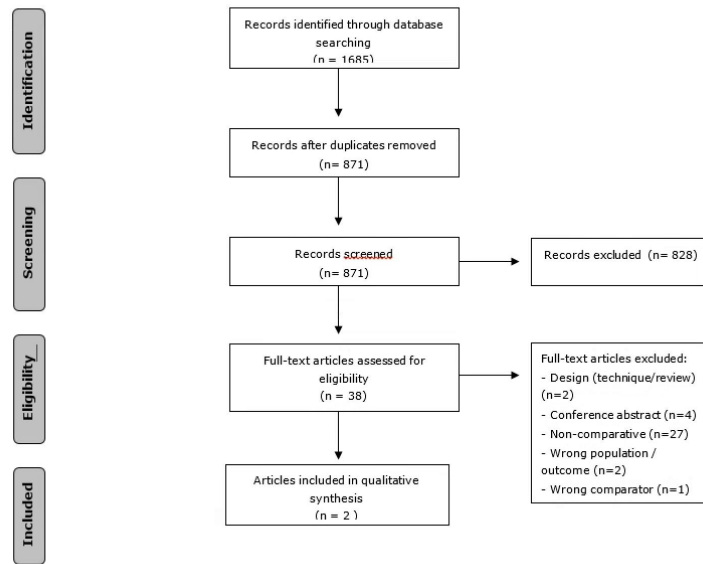
Level of evidence: -

Level of recommendation: Grade D

Is primary tracheopexy during esophageal repair beneficial?

Primary tracheopexy vs no primary tracheopexy

Figure 1. Flowchart of the search and selection procedure of studies.



One excluded study

Retrospective comparative cohort studies

Shieh (2018)

Reason: Compared primary versus secondary PT

Two included studies

Retrospective comparative cohort studies

- Hinoki (2022)
- Van Tuyll Serooskerken (2021)

- Risk of Bias (cohort studies, ROBINS-I)

Author	Bias due to confounding	Bias in selection of participants into the study	Bias in classification of interventions	Bias due to deviation from intended interventions	Bias due to missing data	Bias in measurement of outcomes	Bias in selection of the reported results	Overall Risk of Bias
Hinoki (2022)	Serious	Serious	Serious	Low	Low	Serious	No information	Serious
van Tuyl van Serooskerken (2021)	Moderate	Moderate	Low	Low	Moderate	Serious	Moderate	Serious

Hinoki et al (2022)

Mortality and complications

- Anastomotic leakage 0/8 PPT vs 1/14 no PPT ($p=1.0$)
- Chylothorax 1/8 PPT vs 0/14 no PPT ($p=0.36$)
- Anastomotic stricture 1/8 PPT vs 3/14 no PPT ($p=0.53$)

Additional surgical interventions within 60 days

- PPT: 1/8 (1 tracheostomy) vs no PPT: 8/14 (5 tracheostomy and 3 aortopexy) ($p=0.07$)

Improvement of TM symptoms

- Respiratory dependence rate at 30 days postoperative 2/8 PPT vs 11/14 no PPT ($p=0.03$)
- Intubation 0/8 PPT vs 1/14 no PPT ($p=1.0$)
- CPAP 2/8 PPT vs 10/14 no PPT ($p=0.07$)

Van Tuijll van Serooskerken et al (2021)

Mortality

Group 1 vs Group 2: 1/28 vs 0/36

Cause of death: Accidental decannulation tracheostomy

Complications

- Group 1 vs Group 2 anastomotic leakage: 3/28 vs 6/36 (p=0.72)
- PPT vs no PPT (only group 2) anastomotic leakage: 3/22 vs 3/14 (p=0.66)

Improvement of TM symptoms

- *Brief Resolved Unexplained Events*
 - Group 1 vs Group 2: 11/28 vs 7/36 (p=0.09)
 - PPT vs non PPT in group 2: 1/14 vs 6/22 (p=0.21)
- *Respiratory tract infection*
 - Group 1 vs Group 2: 17/28 vs 9/36 (p=0.004)
 - PPT vs non PPT (group 2): 3/14 vs 6/22 (p=1.0)

Conclusion:

Very limited data suggest that primary tracheopexy is safe and feasible (no increase in mortality and complications) and might improve respiratory outcomes although hard evidence is not available.

Recommendation:

A formal recommendation regarding whether or not primary tracheopexy should be done can't be made based upon the available evidence.

We recommend that an international study will be initiated with clear definitions and outcomes to answer this question.

Level of evidence: *Very low*

Level of recommendation: *Grade D*