Emma Children's Hospital
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The prevalence and clinical impact of transition zone anastomosis in Hirschsprung disease: a systematic review and meta-analysis

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Introduction

- Transition zone (TZ): histopathological zone between affected and unaffected bowel in Hirschsprung disease
- Histopathological characteristics of TZ: hypoganglions, hypodense distribution of ganglions, ectopic ganglions and hypertrophic nerve fibers [1,2]
- In case proximal anastomosis site contains TZ: transition zone anastomosis
- Prevalence of TZA in a previous meta-analysis: 35 % [3]
- No previous study directly compares functional outcome of patients with and without TZA

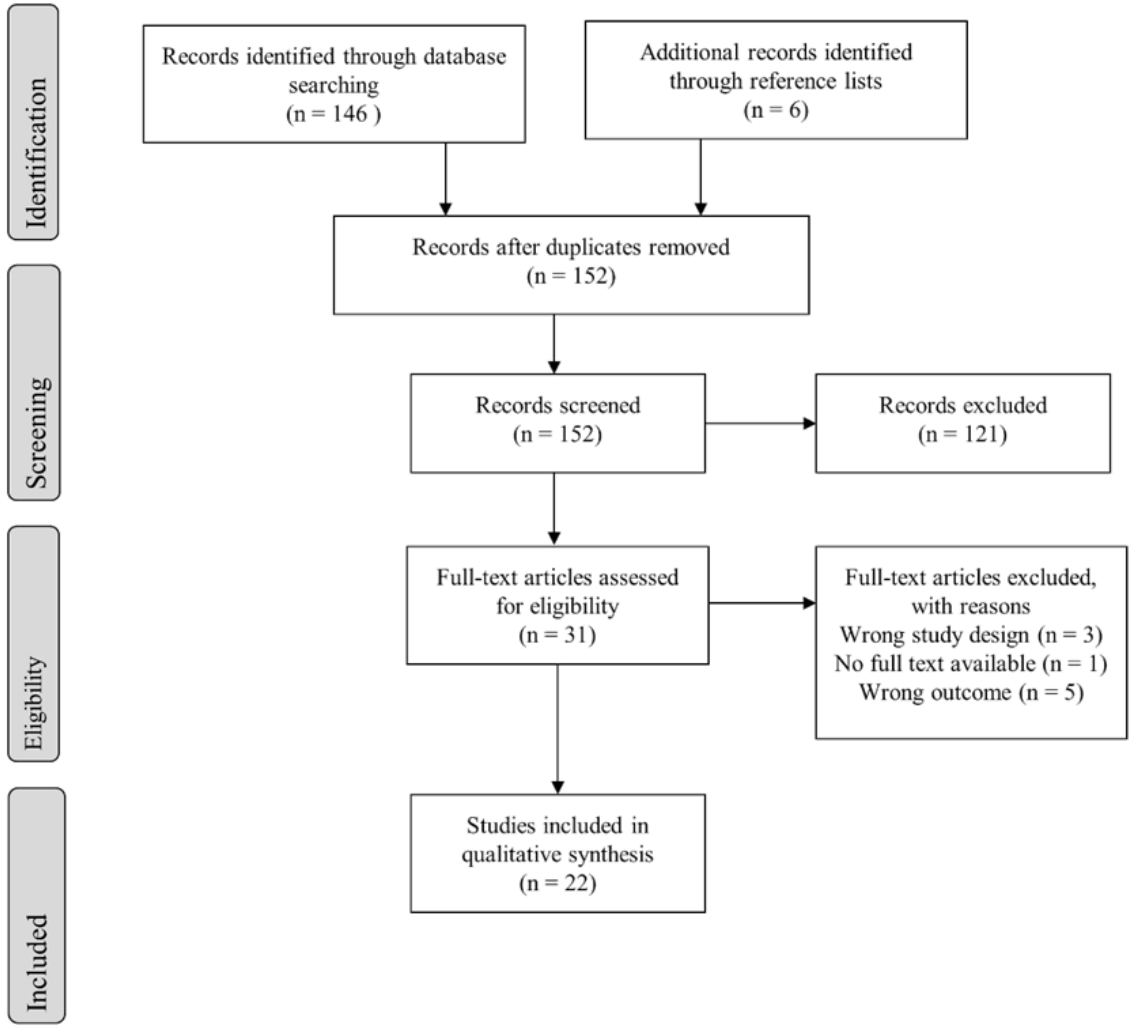
1) Kapur et al., *Pediatr Dev Pathol*, 2013; 2) White et al., *Pediatr Dev Pathol*, 2000; 3) Friedmacher et al., *Pediatr Surg Int*, 2011

Methods

- In accordance with PRISMA Guidelines
- Search: Pubmed, Embase, Web of Science and referencelists
- Double-blind selection (Rayyan)
- Quality Evaluation: Newcastle-Ottawa Scale (NOS)

Inclusion criteria

- Histopathological confirmation of absence/presence of TZA
- Observational or case-control studies



Statistics

Prevalence

- Event rate of TZA per study
- Sensitivity-analysis: primary PT vs redo surgery cohorts
- Sensitivity-analysis: PT with pouch vs PT with straight anastomosis

Clinical impact

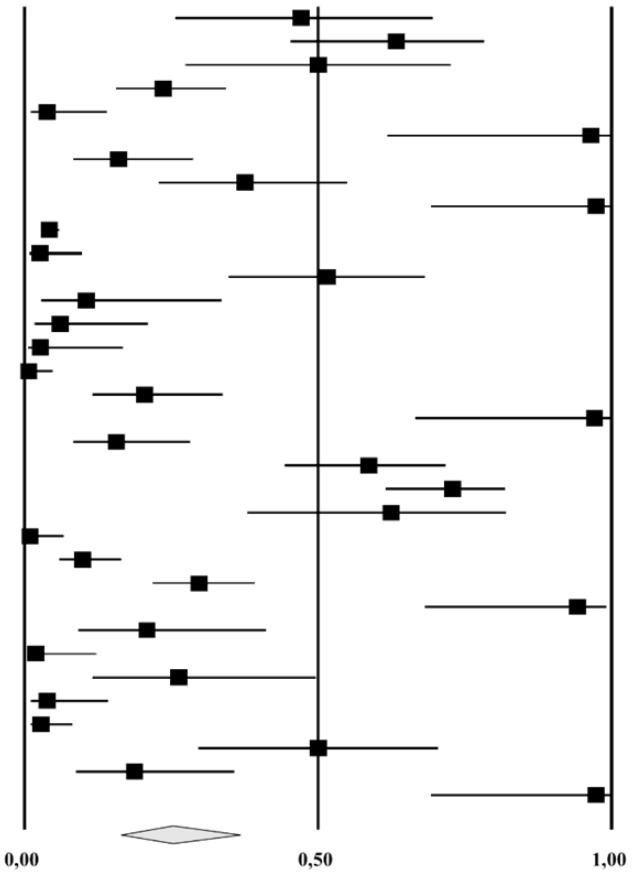
- Prevalence of obstructive defecation, enterocolitis, soiling and incontinence
- Comparison of obstructive defecation, enterocolitis, soiling and incontinence in patients with and without TZA

Results

25%

ER = 0.250 [0.162 – 0.365], p<0.001

Study name	Event rate	Lower limit	Upper limit	p-Value
Chatoorgoon, 2011	0,471	0,255	0,697	0,808
Coe, 2012	0,633	0,451	0,784	0,149
Dingemans, 2017	0,500	0,273	0,727	1,000
Farrugia, 2003	0,237	0,155	0,345	0,000
Gad El-Hak, 2010	0,038	0,010	0,141	0,000
Ghose, 2000	0,964	0,616	0,998	0,022
Ghosh, 2017	0,160	0,082	0,289	0,000
Gupta, 2019	0,375	0,227	0,551	0,162
Hadidi, 2007	0,974	0,690	0,998	0,012
Han, 2019	0,043	0,030	0,061	0,000
Imvised, 2016	0,026	0,007	0,099	0,000
Jiang, 2019	0,516	0,345	0,683	0,857
Keshtgar, 2003	0,105	0,026	0,337	0,004
Kobayashi, 1995	0,061	0,015	0,212	0,000
Langer, 2000	0,027	0,004	0,168	0,000
Langer, 2003	0,007	0,001	0,049	0,000
Langer, 2004	0,204	0,113	0,339	0,000
Lawal, 2011	0,971	0,664	0,998	0,015
Pena, 2007	0,157	0,080	0,284	0,000
Peng, 2020	0,587	0,441	0,719	0,241
Pini-Prato, 2010	0,729	0,613	0,820	0,000
Pini-Prato, 2020	0,625	0,377	0,821	0,323
Polley, 1986	0,010	0,001	0,068	0,000
Ralls, 2014	0,099	0,057	0,167	0,000
Schulten, 2000	0,297	0,216	0,393	0,000
Schweizer, 2007	0,941	0,680	0,992	0,007
Sheng, 2012	0,208	0,089	0,413	0,008
Stensrud, 2010	0,019	0,003	0,124	0,000
van Leeuwen, 2000	0,263	0,114	0,498	0,048
Vu, 2010	0,039	0,010	0,144	0,000
Weber, 1999	0,028	0,009	0,083	0,000
Wilcox, 1998	0,500	0,294	0,706	1,000
Wildhaber, 2004	0,188	0,087	0,359	0,001
Xia, 2016	0,974	0,690	0,998	0,012
	0,250	**0,162**	**0,365**	**0,000**



Results

ER after primary PT vs ER after all PT

9 % vs 59 %, Q=49.9, p<0.001

ER after PT with pouch vs ER after PT with straight anastomosis:

10 % vs 18 %, X^2= 19.21, p<0.001

- 62% obstructive defecation problems
  - 27% with TZA vs 12% without TZA, X^2=7.26, p=0.007
- 38% enterocolitis
  - 25% with TZA vs 18% without TZA, X^2=1.71, p=0.191
- 28% soiling
- 24% incontinence
- Follow-up data ranged from 6 months to 13 years of follow-up

Discussion

- Previous studies may have overestimated prevalence due to a bias caused by higher prevalence in patients undergoing redo surgery [1]
- Obstructive defecation problems occured more often in patients with TZA
- Higher prevalence of enterocolitis, soiling and incontinence were reported compared to the general literature on functional outcome after pullthrough surgery [2,3]
- High rates of TZA may be related to the lack of insight in histopathologic features of transition zone bowel [4]
- Intraoperative strategies to detect and thus prevent TZA need improvement

1) Friedmacher et al., *Pediatr Surg Int*, 2011; 2) Zimmer et al., *Pediatr Surg Int*, 2016 3) Seo et al., *Eur J Pediatr Surg*, 2018; 4) Collin et al., *Pediatr Dev Pathol*, 2013

Take Home Message

Transition zone anastomosis occurs in 10-25% of patients after pull-through surgery, resulting in more obstructive defecation problems, although the clinical impact remains to be further elucidated

Thank you for your attention!

Questions?

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