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Quality of life outcomes in children after surgery for Hirschsprung disease and anorectal malformations: a systematic review and meta-analysis

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ABSTRACT

Background No systematic review and meta-analysis to date has examined multiple child and parent-reported social and physical quality of life (QoL) in pediatric populations affected by Hirschsprung's disease (HD) and anorectal malformations (ARM). The objective of this systematic review is to quantitatively summarize the parent-reported and child-reported psychosocial and physical functioning scores of such children.

Methods Records were sourced from the CENTRAL, EMBASE, and MEDLINE databases. Studies that reported child and parent reported QoL in children with HD and ARM, regardless of surgery intervention, versus children without HD and ARM, were included. The primary outcome was the psychosocial functioning scores, and the secondary outcomes were the presence of postoperative constipation, postoperative obstruction symptoms, fecal incontinence, and enterocolitis. A random effects meta-analysis was used.

Results Twenty-three studies were included in the systematic review, with 11 studies included in the meta-analysis. Totally, 1678 total pediatric patients with HD and ARM underwent surgery vs 392 healthy controls. Pooled parent-reported standardized mean (SM) scores showed better social functioning after surgery (SM 91.79, 95% CI (80.3 to 103.3), I^2=0). The pooled standardized mean difference (SMD) showed evidence for parent-reported incontinence but not for constipation in children with HD and ARM after surgery that had a lower mean QoL score compared with the normal population (SMD −1.24 (-1.79 to −0.69), I^2=76% and SMD −0.45, 95% CI (−1.12 to 0.21), I^2=75%). The pooled prevalence of child-reported constipation was 22% (95% CI (16% to 28%), I^2=0%). The pooled prevalence of parent-reported postoperative obstruction symptoms was 61% (95% CI (41% to 81%), I^2=41%).

Conclusion The results demonstrate better social functioning after surgery, lower QoL scores for incontinence versus controls, and remaining constipation and postoperative obstruction symptoms after surgery in children with HD and ARM.

INTRODUCTION

Hirschprung's disease (HD) and anorectal malformations (ARM) are congenital intestinal anomalies, typically manifesting during early infancy, and contribute to symptoms of severe constipation and intestinal obstruction.[1–3] HD and ARM affect 1 in 5000 live births with a slight male preponderance (ie, male-to-female sex ratio of 4:1).[4–6] Surgical techniques have improved the results of children with HD by decreasing operation time, blood loss, length of hospital stay, and frequency of postoperative complications at 3 years after surgery.[7–9] Despite these short-term advantages, bowel dysfunction and enterocolitis persist long-term and can adversely affect the psychosocial health of children with HD and ARM into adulthood.[10–13] Quality of life (QoL) is a construct aimed at measuring the physical and psychosocial sequelae of parents and their children afflicted by gastrointestinal diseases.[14 15] When applied to medicine, QoL refers to the 'subjective health status' of a patient and measures the effect of illness or disease, moving past physician assessment to consider the patient's well-being and progress.[16 17] The results pertaining to the social adjustment of children with ARM are mixed with certain studies reporting difficulties forming relationships,[18] elevated behavior problems,[19] absence of psychological maladjustment,[20] or the presence of emotional disturbance in children experiencing frequent soiling accidents.[21] Into older age, the major determinant of QoL continues to be fecal incontinence.[22] Multiple meta-analyses have already investigated the short-term and mid-term postoperative outcomes in patients with HD who underwent different surgical approaches, yet the rates of postoperative complications are still variable.[23–27]

There has been no systematic review and meta-analysis to date, which encompasses multiple social and physical dimensions in a pediatric population with consideration of

parent perspectives. Our aim is to conduct a systematic review of existing literature to quantitatively summarize the parent and child-reported psychosocial and physical functioning scores and frequencies of children affected by HD and ARM and in comparison to children without these diseases if data permit.[5]

METHODS

This review followed the Cochrane Methodology to identify and select the studies[28] and the Preferred Reporting Items for Systematic Reviews and Meta-Analyses.[29]

Search strategy and selection criteria

A systematic search for relevant studies was performed on November 5, 2019 and again on December 17, 2020, identifying six additional studies.[6 30–34] MEDLINE, including Epub Ahead of Print, In-Process and Other Non-Indexed Citations, were searched between 1946 to October 25, 2020, Embase from 1947 to 2019 October 25, and the CENTRAL Trials Registry of the Cochrane Collaboration (September 2019 Issue) using the Ovid interface. Searches were designed and conducted by librarian experienced in systematic reviews, using a method designed to optimize term selection.[35] Search strategies can be found in the online supplemental file 1). The study protocol has been registered in Open Science Framework (10.31219/osf.io/zqbgx). All duplicate records were removed online, and records retrieved by the electronic search were downloaded and imported into a Reference Manager database, and then uploaded to Covidence (www.covidence.org) for title and abstract screening and full text review. Five reviewers (EB, IO, MK, VG, VL) screened independently at title/abstract level and full text review stages, and citations were excluded if at least two reviewers agreed to exclude; disagreements were reviewed and resolved by the study leads, where necessary (AN). The study colead (IO) reviewed all eligible citations to confirm eligibility.

Inclusion criteria

Case-control, cohort studies, and randomized-control trials examining child and parent reported QoL in children aged less than 18 years with HD and ARM, regardless of surgery intervention (ie, Duhamel, Swenson, endorectal pullthrough, laparoscopy-assisted pullthrough and open pull through), were included.

Exclusion criteria

Case studies, literature reviews, systematic reviews, editorials, letters to the editor, conference abstracts, and commentaries were excluded. Primary studies published before 2007 that were not written in English were also excluded. A previous systematic review captured studies before 2007; hence, why we excluded studies published before 2007.

Data extraction

Three authors (IO, EB, MK) performed data abstraction using a predesigned, piloted, and modified sheet in Excel V.14.7.7, which was validated by our statisticians (LH, VB). The extracted information included the following: study details including study design; type of QoL report (parent proxy or child), length of follow-up, age of children when QoL was assessed, QoL instrument facilitated, threshold for interpreting QoL scales, and parent-reported versus child-reported QoL scores, in addition to the sample sizes of children who presented with physical complications postoperatively.

Outcome definitions

The primary outcome was the mean scores on the psychosocial functioning domains. The secondary outcomes included the followings: reported frequencies and mean scores on QoL instruments of physical symptoms for constipation, intestinal obstruction, fecal incontinence, and enterocolitis. Definitions for each outcome depended on the QoL instrument used (online supplemental file 2, table S1). We defined short-term follow-up as: results from QoL interviews or questionnaires obtained before 2 years from study onset, while long-term follow-up referred to measurements after 2 years. Parents may have reported on study outcomes if children were too young to report themselves.

Assessment of risk of bias (ROB) within studies

Two reviewers (MK and IO) independently reviewed each study. The validated Methodological Index for Non-Randomized Studies (MINORS) criteria was used to assess the quality of the studies.[36] Based on the Cochrane handbook for considering bias, reasons for disagreement were explored and resolved.[37] Items assessed included: clearly stated aims, inclusion and representativeness of patients, reliable prospective data collection, appropriate and unbiased endpoints, sufficient follow-up period, follow-up loss, adequate study size calculation, contemporary groups (to address historical bias), baseline equivalence, and adequate statistical analysis.[36] Items 1–7 apply to non-comparative studies, while items 8–12 for comparative ones. Records were given scores of zero through two. The maximum (ideal) global score is 24 for comparative studies and 16 for non-comparative studies. One study was a randomized control trial (RCT).[38] Therefore, the Cochrane Risk of Bias tool was used. The ROB tool covers six domains of bias: selection bias, performance bias, detection bias, attrition bias, reporting bias, and other bias. Within each domain, assessments are made for one or more items and support for judgment is made by providing a free text description and assigning judgment into high, low, or unclear risk of material bias for each item.[39]

Statistical analysis

All statistical analyses were performed using the R statistical programming language (V.4.0.).[40] Data were

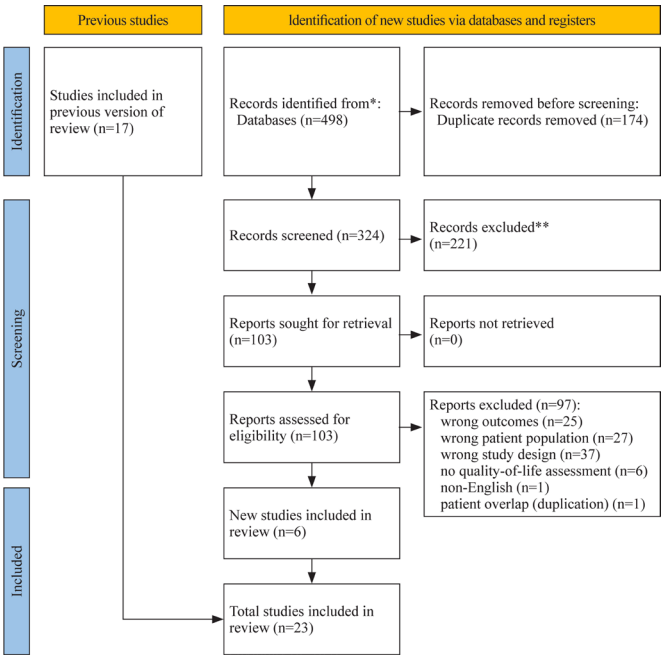

Figure 1 PRISMA flow diagram. *Consider, if feasible to do so, reporting the number of records identified from each database or register searched (rather than the total number across all databases/registers). **If automation tools were used, indicate how many records were excluded by a human and how many were excluded by automation tools.[29] PRISMA, Preferred Reporting Items for Systematic Reviews and Meta-Analyses.

meta-analyzed using a random effects model with R package 'meta'.[41] Scores of all QoL instruments were scaled to 0–100 scale using the 'min-max scaling' method (Hogg, p. 69).[42] All scores were standardized—scores closer to 100 indicated higher or better QoL. Pooled proportions and pooled QoL standardized mean scores with 95% CI were reported in cases of low heterogeneity (I^2=75%). In cases of high heterogeneity, the mean scores of individual studies without pooled estimate were presented by subgroups of potential sources of variability: duration of outcome (short 0–2 vs long 2+ years), instrument type, and scale (0–100 vs other). The results were reported separately for the child and parents.

RESULTS

Our initial search yielded 498 studies. After an update was performed, there were 6 additional studies that met our inclusion criteria and 23 (22 observational and 1 RCT) included in the systematic review, with two studies included in the meta-analysis for child-reports and nine for parent-reports (figure 1). One study was excluded, due to patient overlap.[22]

Study characteristics and individual results

In total, 1678 pediatric patients with HD and ARM who underwent surgery vs 392 healthy controls without HD and ARM were included (online supplemental file 3). Children were typically between the ages of 2 and 18 years. A mean age was not reported because the timing of symptom presentation may have differed for each outcome. There were three prospective case control studies,[43–45] two prospective case and retrospective control data studies (ie, established controls from literature),[5 46] one retrospective case control study,[33] nine retrospective cohort studies,[32 34 47–53] six prospective cohort studies,[6 15 30 54–56] one RCT,[38] and one mixed-method sequential explanatory cohort study.[31] These studies were implemented in various pediatric centers around the world, including Japan,[43] Australia,[5 44] the Netherlands,[46 48] United Kingdom,[30 32] China,[33 34 38 52] Sweden,[55] Egypt,[15] Ireland,[47] Finland,[53] and the USA.[49 51] Prevalent surgical approaches, included the trans anal endorectal pull-through, and the Duhamel methods. No study recruited patients with congenital diseases, neurological defects and other syndromes (eg, Down syndrome). The minimum follow-up was 6 months and maximum was 28 years, for QoL assessment.

Risk of bias across studies

Two reviewers (MK, IO) independently assessed RoB of the included studies. Common reasons for disagreement, included discrepancies on unbiased endpoints (ie, blinded interviewers for QoL assessments), follow-up period (2 years minimum), and loss to follow-up (<95% loss). The discrepancies were resolved by consensus (online supplemental file, table S3). Online supplemental file, tables S3 and S4 (online supplemental file 4) displays the quality appraisal results. MINORS scores for comparative studies (n=7) ranged from 14 to 20, with mean 16.4±2.1. MINORS scores for non-comparative studies (n=15) ranged from 10 to 14, with a mean of 11.7±1.0. Based on these mean scores, the MINORS criteria suggests fair study quality. Weaknesses in the comparative studies included a follow-up period of less than 2 years, greater than 5% loss to follow-up, insufficient double blinding, and no exploration of confounders or evidence of a statistical test when demographic characteristics differed between cases and controls (age and sex). Weaknesses of the non-comparative studies were: high attrition bias (>5%), no indication for Research Ethics Board (REB) approval, and potential for experimenter bias when administering QoL questionnaires (online supplemental file, table S3). The RCT published by Wang *et al*[38 38] demonstrated fair quality due to absence of reporting bias. A lack of reporting in compliance and social adjustment did not bias additional outcomes such as incontinence (online supplemental file, table S4).

Primary outcomes

Psychosocial QoL outcomes

Pooled parent-reported mean scores showed better social functioning after surgery (91.79, 95% CI (80.3 to 103.3), I^2=0) (figure 2). Due to high heterogeneity for parent-reported emotional and psychosocial domains, only a narrative synthesis is presented, suggesting high emotional and psychosocial scores after surgery (online

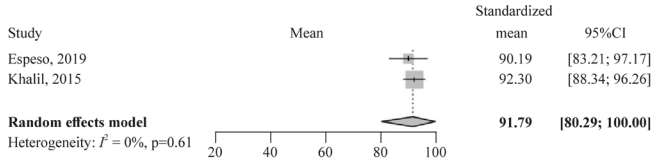

Figure 2 Pooled quality of life score mean estimate and 95% CI for parent-reported social functioning domain for Hirschsprung's disease only.

supplemental file 5). Please refer to the supplementary files that describe the altering of original scores using min-max scaling.

Secondary outcomes

Physical QoL outcomes in cases versus controls

The pooled effect for parent-reported shows evidence for incontinence but not for constipation. Children with HD and ARM after surgery have a lower mean QoL score compared with the normal population (standardized mean difference (SMD) −1.24 (−1.79 to −0.69), I^2=76% and SMD −0.45, 95% CI (−1.12 to 0.21), I^2=75%) (figure 3), respectively. For parent-reported incontinence scores, a subgroup analysis based on instrument type was performed (scale from 0 to 100 vs transformed to 0 to 100). Regardless of instrument type, incontinence scores are high or better after surgery (online supplemental file 5).

Physical complications

Pooled prevalence of child-reported constipation was 22% (95% CI (16% to 28%), I^2=0%) (figure 4). Subgroup mean score for short-term versus long-term parent-reported constipation were showed, with short-term proportions ranging from 4% to 41% and long-term from 8% to 76% (online supplemental file 5). Pooled prevalence of parent-reported obstruction symptoms was 61% (95% CI (41% to 81%), I^2=41%) (figure 5).

DISCUSSION

The objective of this systematic review was to capture the parent and child-reported psychosocial and physical functioning scores and physical complications of

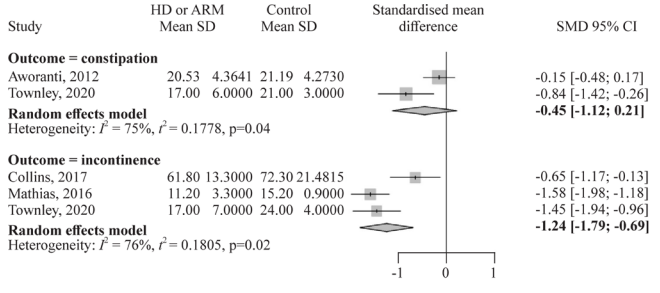

Figure 3 Pooled estimates and 95% CI for parent-reported constipation and incontinence scores. ARM, anorectal malformations; HD, Hirschsprung's disease; SMD, standardized mean difference.

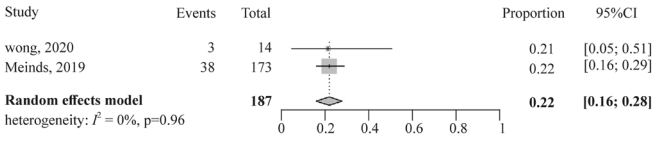

Figure 4 Pooled prevalence plot for child-reported constipation.

children affected by HD and ARM. Our main, low heterogeneity results demonstrate: (1) better social functioning after surgery; (2) lower QoL scores for incontinence versus controls; and (3) remaining constipation and obstruction symptoms after surgery in children with HD and ARM. In accordance with our findings, previous literature suggests that parents typically report better social functioning, such as good social relationships with friends in their children with HD and ARM.[5 57 58] One potential explanation for this finding is that older pediatric patients with chronic disease experience a 'response shift', in that they respond to their new reality living with the disease and adjust to functional norms. In other words, they are able to emotionally and mentally shift to living with their chronic disease, giving them feelings of greater control.

This theory might help explain why parents observed better social functioning in their children, based on the internal coping strategies the children may have employed.[58–60] However, relying on purely parent responses alone is not the only reliable method for assessing social functioning QoL in children with chronic disease. This is because discrepancies in QoL reporting persist with their children, and a combination of both is recommended.[61] In fact, children with HD may report poor psychosocial and social functioning after surgery[62 63] in comparison to their parents. Moreover, one systematic review found that children with HD and ARM experience difficulties with anxiety, peer rejection, and behavioral problems, while adolescents report low self-esteem, poor body image, and depression.[64]

Our review found that children with HD and ARM have lower QoL scores for incontinence versus published controls but no evidence were found for constipation, which is in agreement with another 2021 systematic review[65] that did not determine the significant difference of constipation rates (42%) between HD children and the general population, when strict clinical definitions for constipation were used to monitor these patients.[1 66] Therefore, when thresholds to qualify as constipation are set high, children may not meet all the

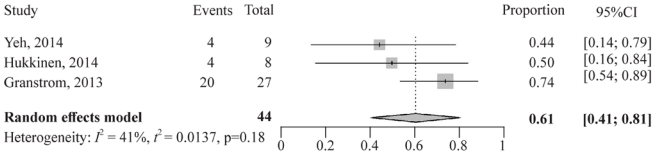

Figure 5 Pooled prevalence plot for parent-reported obstruction symptoms.

criteria at the time of assessment. Hence, constipation rates in our review may be underreported. Additionally, numerous studies have echoed that children with HD have increased or worse incontinence scores and lower physical functioning, compared with age and gender matched controls,[5 44 48 67 68] regardless of the QoL instrument used. Interestingly, incontinence scores correlate with lower QoL scores for behavior and self-esteem, unlike in normal controls.[46] Plausible reasons why children with HD and ARM suffer from fecal incontinence might be due to increasing severity of fecal incontinence over time, as children age, which consequently negatively affects their QoL.

Conversely, children without these conditions may not experience the same complications.[5] In our review, we included children between the ages of 0 and 18 years. Younger patients who have not lived long enough with HD may not cope positively to effectively manage their ongoing functional symptoms, thus reporting poorer physical QoL relative to controls.[44] Last, children with only a few functional problems are frequently discharged from follow-up and not transitioned to adolescent or adult care. As such, it is possible that this review underreported the functional problems of constipation and incontinence among cases.[48] Twenty-two per cent of children reported having constipation after surgery, while long-term parent-reported constipation was as high as 76% in our review. Parent-reported postoperative obstruction symptoms was 61%, and incontinence ranged from 13% to 68%. Comparing these findings to prior literature is difficult, given that clinical definitions for constipation, postoperative obstruction symptoms, and incontinence vary greatly across studies and may be captured differently based on the data collection method used.[65] In fact, certain studies report no HD patients experiencing constipation[69 70] vs 30%–76% after surgery.[48 66] Similarly, cohort studies document ranges of incontinence from 19% to 82%.[44 62 71–76] Long-term follow-up after surgery may increase the ability for researchers to detect the incidence of constipation and incontinence. Moreover, the pediatric population is heterogeneous in that they may present with neurological impairments and syndrome-associated diseases, which are correlated with increased constipation and incontinence scores.[1 65 77] No children in studies included in our review possessed such impairments.

Limitations

Limitations of the evidence included in this review mainly stem from the wide variability in child and parent documented psychosocial and physical QoL outcomes, which we attempted to examine via subgroups for instrument type, scale, and duration of outcome. Heterogeneity was likely due to underlying clinical differences in the pediatric population and/or discrepancies between parent and child reports. Furthermore, sample sizes in cohort studies were small, and certain case control studies captured control data from previously established literature, rather than controls recruited during their study. We also surmise that physical complications were more likely to be detected during long-term postoperative follow-up.

Future directions

Given the variability in psychosocial and physical functioning QoL instruments, there is a need to standardize child and parent-reported QoL measurements to improve the robustness and generalizability of the current evidence. Moreover, creating validated, age-specific child QoL measurements would also be beneficial to explore differences in QoL by age. In terms of the complications of constipation and incontinence, standardizing clinical definitions and treatment plans are encouraged. Prospective, multicentered, and longitudinal studies with consistent monitoring of QoL and complications of children with HD and ARM into adulthood, is an area of future research.

Clinical implications

For clinical benefits, postsurgical treatment interventions should target reducing constipation, postoperative obstruction symptoms, and incontinence scores based on consultation with patients and their health risk profile. In order to obtain an accurate perception of child social functioning, more informants (children, parents, teachers) in clinical research are recommended, to support patients and their families after surgery.[64 78 79]

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Contributors Five reviewers (EB, IO, MK, VG, VL) screened independently at title/abstract level and full text review stages, and citations were excluded if at least two reviewers agreed to exclude; disagreements were reviewed and resolved by the study leads, where necessary (AN). The study colead (IO) reviewed all eligible citations to confirm eligibility. Two analysts (LH, VB) were responsible for assisting with the analysis. IO was responsible for writing the final manuscript with assistance from VL.

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Supplementary File 1

MEDLINE

1. exp Hirschsprung Disease/ or (hirschsprung* or hirschprung* or hirsprung* or congenital megacolon or aganglionic megacolon or intestinal aganglionosis or colonic aganglionosis or (rectosigmoid adj2 aganglionosis)).ti,ab,kf.

2. Anorectal Malformations/ or (anorectal malformation* or anorectal anomal* or anorectal Atresia* or anal atresia* or anorectal stenos* or imperforate anus).ti,ab,kf.

3. Fecal Incontinence/ or Constipation/ or Defecation/ or Flatulence/ or Diarrhea/ or ((faecal or fecal or feces or faeces or defaecat* or defecat* or stool*) adj2 (problem* or symptom* or function* or incontinen* or continen* or control* or stain* or urgen* or soil* or leak*)).ti,ab,kf. or (diarrhoea or diarrhea or constipate* or flatu* or diaper* or (stool* adj2 (foul or smell or frequen*))).ti,ab,kf.

4. (Baylor Continence Scale or Baylor Social Continence Scale or BCS or HAQL or Holschneider or FII or Kelly score or LSQ or Langemeijer Stool Questionnaire or family impact questionnaire).ti,ab,kf.

5. "Quality of Life"/ or "Outcome Assessment (Health Care)"/ or exp Patient Satisfaction/ or (qol or (quality adj2 life) or patient reported or prom).ti,ab,kf. or questionnaire*.mp. or (well being or wellbeing or satisfaction or quality of life or physical function* or appearance or body image or psychological or sexual or mental or social or psychosocial).ti,ab,kf.

6. (CBCL or CBCL* or Child Behaviour Checklist or CHQ-PF50 or EuroQoL or FIL or HAQL or HOPES or Hunter Opinions Personal Expectations Scale or mos or pedsql or qqvcfca or SF36 or sf-36 or ssp or tacqol or TAIQOL).ti,ab,kf.

7. (1 or 2) and (3 or 4) and (5 or 6) and (infant* or child* or adolescences*).mp.

8. limit 7 to (english language and yr="2007 - 2020" and journal article)

Embase

1. exp Hirschsprung Disease/ or (hirschsprung* or hirschprung* or Hirsprung* or congenital megacolon or aganglionic megacolon or intestinal aganglionosis or colonic aganglionosis or (rectosigmoid adj2 aganglionosis)).ti,ab,kw.

2. exp Anorectal Malformation/ or (anorectal malformation* or anorectal anomal* or anorectal Atresia* or anal atresia* or anorectal stenos* or imperforate anus).ti,ab,kw.

3. Feces Incontinence/ or exp Constipation/ or Defecation/ or Defecation Disorder/ or Flatulence/ or exp Diarrhea/ or ((faecal or fecal or feces or faeces or defaecat* or defecat* or stool*) adj2 (problem* or symptom* or function* or incontinen* or continen* or control* or stain* or urgen* or soil* or leak*)).ti,ab,kw. or (diarrhoea or diarrhea or constipate* or flatu* or diaper* or (stool* adj2 (foul or smell or frequen*))).ti,ab,kw.

4. (Baylor Continence Scale or Baylor Social Continence Scale or BCS or HAQL or Holschneider or FII or Kelly score or LSQ or Langemeijer Stool Questionnaire or family impact questionnaire).ti,ab,kw.

5. *"Quality of Life"/ or *Outcome assessment/ or *Patient attitude/ or *Satisfaction/ or exp *Patient Satisfaction/ or (qol or (quality adj2 life) or patient reported or prom).tw. or questionnaire*.mp. or (well being or wellbeing or satisfaction or quality of life or physical function* or appearance or body image or psychological or sexual or mental or social or psychosocial).ti,ab,kw.

6. (CBCL or CBCL* or Child Behaviour Checklist or CHQ-PF50 or EuroQoL or FIL or HAQL or HOPES or Hunter Opinions Personal Expectations Scale or mos or pedsql or qqvcfca or SF36 or sf-36 or ssp or tacqol or TAIQOL).ti,ab,kw.

7. (1 or 2) and (3 or 4) and (5 or 6) and (baby* or babies* or newborn* or infan* or neonat* or preschool* or pre-school* or child* or pediatr* or paediatr* or teen* or adolescen*).mp.
8. limit 7 to (Embase)
9. limit 8 to conference abstract
10. 8 not 9
11. limit 10 to (english language and yr="2007 - 2020")

Cochrane Central Trials Registery (CENTRAL)
1. (hirschsprung* or hirschprung* or Hirsprung* or congenital megacolon or aganglionic megacolon or intestinal aganglionosis or colonic aganglionosis or (rectosigmoid adj2 aganglionosis)).ti,ab,kw.

2. (anorectal malformation* or anorectal anomal* or anorectal Atresia* or anal atresia* or anorectal stenos* or imperforate anus).ti,ab,kw.

3. (((faecal or fecal or feces or faeces or defaecat* or defecat* or stool*) adj2 (problem* or symptom* or function* or incontinen* or continen* or control* or stain* or urgen* or soil* or leak*)) or (diarrhoea or diarrhea or constipate* or flatu* or diaper* or (stool* adj2 (foul or smell or frequen*)))).ti,ab,kw.

4. (Baylor Continence Scale or Baylor Social Continence Scale or BCS or HAQL or Holschneider or FII or Kelly score or LSQ or Langemeijer Stool Questionnaire or family impact questionnaire).ti,ab,kw.

5. (qol or (quality adj2 life) or patient reported or prom).tw. or questionnaire*.mp. or (well being or wellbeing or satisfaction or quality of life or physical function* or appearance or body image or psychological or sexual or mental or social or psychosocial).ti,ab,kw.

6. (CBCL or CBCL* or Child Behaviour Checklist or CHQ-PF50 or EuroQoL or FIL or HAQL or HOPES or Hunter Opinions Personal Expectations Scale or mos or pedsql or qqvcfca or SF36 or sf-36 or ssp or tacqol or TAIQOL).ti,ab,kw.

7. (1 or 2) and (3 or 4) and (5 or 6) and (baby* or babies* or newborn* or infan* or neonat* or preschool* or pre-school* or child* or pediatr* or paediatr* or teen* or adolescen*).mp.

8. limit 7 to yr="2007 - 2020"

Supplementary File 2

Table S1. Description of Quality-of-Life Instrument used and Scoring Threshold (if appropriate) per Included Study

Author, year	Constipation	Small bowel obstruction	Fecal Incontinence	Difficulty defecating	Enterocolitis	Psychosocial problems
John, 2010			Quality of life (QOL) questionnaire The scale used ranged from 0-13 with a higher score meaning a better QOL			
Grano, 2010	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL) Scores range from 0 to 100 with higher scores indicating higher levels of functioning		Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL) Scores range from 0 to 100 with higher scores indicating higher levels of functioning			Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL) Mean scores given.
Yang, 2016			Detailed questionnaire (unspecified)		Detailed questionnaire (unspecified)	

			Proportions given.	Proprtions given.
Yeh, 2014		Medical records and standardized telephone questionnaire (unspecified) Proportions given		Medical records and standardized telephone questionnaire (unspecified) Proportions given.
Yamataka, 2009	Continence evaluation questionnaire. Questionnaire used and values for score given is % of patients, higher % is more patients experienced symptom. Postoperative constipation did not occur in surgery groups.			
Wang, 2014			Questionnaire (undefined)	

Granstrom, 2013	Patient records Proportions given, no scores	Patient records Proportions given			
Hukkinen, 2014		Bowel function score questionnaire. Proportions given. Bowel obstruction = acute intestinal obstruction.		Bowel function score questionnaire. Proportions given.	
Roorda, 2018	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL disease specific QoL) Proportions given, no scores		Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL). Proportions given.		CHQ-PF50 and CHQ-CF87. Mean scores given. Scales range from 0-100 with higher scores indicating better perceived functioning. When parent and self-report yielded the same domain score, these were pooled and compared with normative data.

Levitt, 2013	Medical/patient records and telephone/email questionnaire. Proportions given.		Medical/Patient Records and telephone/email questionnaire. Proportions given.	
Sood, 2018	Cleveland Clinic Constipation Scoring System (CCCSS) Higher scores indicate severe constipation and a global score of 15+ means patient has constipation. 30 = severe constipation.	Baylor Continence Scale. (BCS). BCS scores range from 2 to 92, where better social continence is denoted by lower scores.	REDCap colorectal database. Proportions given.	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL). Mean scores given. No SD reported for control data.
Khalil, 2015	Peds QL 4.0-Core Measurement Scale. Proportions given	Peds QL 4.0-Generic Core Scales. Proportions given.		PedsQL 4.0 Core Measurement Model. Mean scores given. A 5-point Likert scale from 0 (never) to 4 (almost always) is used. Items are then reverse scored and linearly transformed to a 0 to 100 scale. Higher

					scores indicate better QoL.
Meinds, 2019	Rome IV. Proportions given, no scores. Constipation was defined by the Rome IV criteria for functional constipation. Patients need at least 2 of the following: straining, hard or lumpy stools, incomplete evacuation, anorectal obstruction, use of manual manoeuvres to defecate, or fewer than 3 bowel movements per week. Loose stools should rarely be present.	Rome IV. Proportions given	Rome IV. Proportions given		

Mathias, 2016	Quality of life related to fecal continence in children and adolescents (QQVCFCA). The final score is obtained by summing the mean score obtained in each domain, and ranges from 4 to 16. High scores indicate good standing.	
Lane, 2016	Baylor Continence Scale (BCS). Scores range from 2 to 92, with lower scores reflecting better fecal continence/control.	PedsQL Pediatric Quality of Life Inventory. Total parent reported HRQoL scores, and psychosocial scores were the sum of emotional, social and school functioning combined. Maximum score of 100, with higher

			score corresponding to better QoL
Collins, 2017	Cleveland Clinic Constipation Scoring System (CCCSS). A score of 0 indicates normal bowel function, 15 is defined as "constipation", and 30 indicates "severe constipation".	Baylor Continence Scale (BCS). Scores range from 2 to 92, with lower scores reflecting better fecal continence/control.	PedsQL 4.0 Generic Core Scale. Maximum score of 100, with higher score corresponding to better QoL
Aworanti, 2012	Pediatric Incontinence and Constipation Scoring System questionnaire (PICSS). Mean scores presented. The maximum score is 29 for constipation (higher score means no constipation).	Pediatric Incontinence and Constipation Scoring System (PICSS). Mean scores presented. Maximum score is 32 for incontinence scale, which implies continence (closer the score is to 32 = better continence)	

Allin, 2020	Pediatric Incontinence and Constipation Scoring System (PICSS) questionnaire. Mean scores presented. The maximum score is 29 for constipation (higher score means no constipation).	Pediatric Incontinence and Constipation Scoring System (PICSS). Mean scores presented. Maximum score is 32 for incontinence scale, which implies continence (closer the score is to 32 = better continence)	Pediatric Incontinence and Constipation Scoring System (PICSS). Proportions given.	PedsQL. Items are reverse scored and linearly transformed from 0 to 100. Higher scores indicate better quality of life.
Espeso, 2020		Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL). Mean scores given. Each dimension is scored over 100; the higher the score, the better quality of life. From 0-8 years, parents completed the HAQL.	Medical records. Proportions given. Two children aged 6-11 years experienced enterocolitis versus 0 aged 12-18.	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL). Mean scores given. Each dimension is scored over 100. The higher the overall score, the better the quality of life.

Saysoo, 2020	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL-different). A score from 0 to 3 was given in response for each item and a better QoL was indicated by a higher score	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL). A score from 0 to 3 was given in response to each item and a better QoL was indicated by a higher score.	Hirschsprung/ Anorectal Malformation Quality of life Questionnaire (HAQL/modified). Mean scores are not differentiated by parent vs adolescent report. A score from 0 to 3 was given in response for each item and a better QoL was indicated by a higher score.
Townley, 2020	The Paediatric Incontinence and Constipation Score (PICS). Data are given as mean (range). Scores range from 0-29 with higher scores indicating no constipation.	Pediatric Incontinence and Constipation Scoring System (PICSS). Mean scores (with SD or range) given. Maximum score is 32 for incontinence scale, scores closer to 32 = better continence.	
Wong, 2020	Wong, 2020-A: Hirschsprung/ Anorectal	Wong, 2020-A: Hirschsprung/ Anorectal	Mean scores given. For each item, the patient was asked to

	Malformation Quality of life Questionnaire HAQL: Mean scores given. For each item, patient asked to indicate the frequency of occurrence using a 5-point scale from 1 (never) to 5 (always). Responses recoded into a linear scale of 0-100. Higher scores indicate higher functioning. Wong, 2020-B: Krickenbeck classification (grade 2): Proportions given. The Krickenbeck classification (2005) categorizes constipation into 3 types: grade 1 (manageable by	Malformation Quality of life Questionnaire (HAQL). Mean scores given. For each item, patient asked to indicate the frequency of occurrence using a 5-point scale from 1 (never) to 5 (always). Responses were then recoded into a linear scale of 0-100. Higher scores indicate higher levels of functioning. Wong, 2020-B: Kelly's score of continence: Mean scores given. An overall score of 5-6 is good, 3-4 fair, and 0-2 poor.	indicate the frequency of occurrence using a 5-point scale ranging from 1 (never) to 5 (always). The responses were recoded into a linear scale of 0-100. Higher scores indicated higher levels of functioning.

	changes in diet); 2 (requires laxatives); and 3) resistant to diet and laxatives . Grade 2 is reported. No case or control had grade 3.		
Zhuansun, 2020		Zhuansun, 2020-A: (Quality of Life) QOL scoring criteria for children with fecal incontinence: Mean scores given. Frequent is assigned a 0. Normal is assigned a 2. Good = 9 to 12 points; fair = 5 to 8; poor = 0 to 4. Zhuansun, 2020-B: Mail communications and telephone interviews: Proportions given.	Mail communications and telephone interviews. Proportions given.

Author, year. Location.	Population Studied and Mean Age	Constipation	Small Bowel Obstruction	Fecal incontinence	Enterocolitis	Social functioning	Psychosocial functioning	Emotional functioning
Cohort studies								
Khalil, 2015. Egypt.	53 children with a mean age of 5.8 years.							
Parent report (n/N)[a]		4/53	[b]	3/53				
Parent report (mean, SD)						92.3 ± 14.7		95.5± 6.5
Child report (n/N)								
Child report (mean, SD)								
Lane, 2016. USA.	325 children and youth younger than 18 years.							
Parent report (n/N)								
Parent report (mean, SD)				23			58.9± 19.2	
Child report (mean, SD)								
Child report (mean, SD)								

Grano, 2010. Italy.	62 children with a mean age of 7.3 years and 175 parents with a mean age of 24.0 years.			
Parent report (n/N)				
Parent report (mean, SD)		81.98± 24.21	79.87± 20.49	85.68± 21.69
Child report (n/N)				
Child report (mean, SD)				
John, 2010. India.	166 children with a mean age of 7.5 years.			
Parent report (n/N)				
Parent report (mean, SD)			9.1	
Child report (n/N)				
Child report (mean, SD)				
Aworanti, 2012. Ireland.	51 parents whose mean age was not reported.			
Parent report (n/N)		16/51	31/42	
Parent report (mean, SD)				

Child report (n/N)												
Child report (mean, SD)												
Granstrom, 2013. Sweden.	27 children with a mean age of 4.4 years at interview 1 and 7.4 years at interview 2.											
Parent report (n/N)		11/27		20/27								
Parent report (mean, SD)												
Child report (n/N)												
Child report (mean, SD)												
Yang, 2016. China.	31 parents whose mean age was not reported.											
Parent report (n/N)					0/31		21/31					
Parent report (mean, SD)												
Child report (n/N)												
Child report (mean, SD)												
Levitt, 2013. USA.	67 children with a mean age of 1.56 years.											
Parent report (n/N)		21/67					9/67					

Parent report (mean, SD)												
Child report (n/N)												
Child report (mean, SD)												
Hukkinen, 2014. Finland.	8 children whose mean age was not reported.											
Parent report (n/N)			4/8			5/8						
Parent report (mean, SD)												
Child report (n/N)												
Child report (mean, SD)												
Yeh, 2014. Taiwan.	9 children with a mean age of 0.49 years.											
Parent report (n/N)			4/9			5/9						
Parent report (mean, SD)												
Child report (n/N)												
Child report (mean, SD)												
Roorda, 2018. Netherlands.												
Parent report (n/N)		13/17			8/17							

Parent report (mean, SD)						95.6± 8.5
Child report (n/N)		8/26	8/17			
Child report (mean, SD)						95.6± 8.5
Zhuansun, 2020. China.	35 children whose mean age was not reported.					
Parent report (n/N)						
Parent report (mean, SD)						
Child report (n/N)			6/198	36/198		
Child report (mean, SD)						
Espeso, 2020. France.	63 child-parent dyads with a children's mean age of 11.55 years.					
Parent report (n/N)						
Parent report (mean, SD)			58± 36		88± 20	85± 20
Child report (n/N)				2/63		
Child report (mean, SD)			68± 32		94± 12	86± 26

Townley, 2020. United Kingdom.	71 children whose mean age was not reported.					
Parent report (n/N)						
Parent report (mean, SD)		17± 6	17± 7			
Child report (n/N)						
Child report (mean, SD)						
Saysoo, 2020. Indonesia.	11 children whose mean age was not reported.					
Parent report (n/N)						
Parent report (mean, SD)		2.5	2.06		2.69± 0.99	
Child report (n/N)						
Child report (mean, SD)		2.5	2.06		2.69 ± 0.99	
Allin, 2020. United Kingdom.	227 children whose mean age was not reported.					
Parent report (n/N)		27/72	29/72	108/227		
Parent report (mean, SD)						67.4± 20.96
Child report (n/N)						

Child report (mean, SD)				
Case Control Studies				
Yamataka, 2009. Japan.	24 children whose mean age was .082 years.			
Parent report (n/N)		0/14		
Parent report (mean, SD)				
Child report (n/N)				
Child report (mean, SD)				
Control (n/N, %)				
Collins, 2017. Australia.	60 parents whose mean age was not reported.			
Parent report (n/N)				
Parent report (mean, SD)		4.47± 3.97	22.2± 13.3	76± 17.9
Child report (n/N)				
Child report (mean, SD)				
Control (n/N, %)				
Meinds, 2019. Netherlands.	173 children whose mean age was not reported.			
Parent report (n/N))				

Parent report (mean, SD)								
Child report (n/N)		38/173	39/173	65/173				
Child report (mean, SD)								
Control (n/N, %)								
Wong[c], 2020. China.	21 children with a mean age of 18 years.							
Parent report (n/N)								
Parent report (mean, SD)								
Child report (n/N)								
Child report (mean, SD)		42.9		77.2		26.4		71.9
Control (n/N)								
Control (mean,SD)		60		87.5		71.7		63.6
Sood, 2018. Australia.	58 children with a mean age of 14.48 years.							
Parent report (n/N)								
Parent report (mean, SD)				16.7± 7.99				82.53± 17.61
Child report (n/N)					16/58			
Child report (mean, SD)		4.3± 2.98						82.53± 17.61

Control (n/N, %)			
Mathias, 2016. Brazil.	71 children whose mean age was not reported.		
Parent report (n/N)			
Parent report (mean, SD)		11.2± 3.3	
Child report (n/N)			
Child report (mean, SD)			
Control (n/N, %)			
Randomized Control Trial			
Wang, 2015. China.	82 children whose mean age was 2.11 years.		
Parent report (n/N)			
Parent report (mean, SD)			
Child report (n/N)			
Child report (mean, SD)			

[a]n is the sample size who experienced complication; N is the total number of patients

[b] Grey shading represents no relevant data captured in the study for that outcome

[c]LAARP patients were compared with historical controls treated with PSARP between 1996 and 2000.

Oltean I, *et al. World Jnl Ped Surgery* 2022; 5:e000447. doi: 10.1136/wjps-2022-000447

Supplementary File 3

Table S3. Methodological appraisal of observational studies

	Criteria												
	1	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**	
Studies	Clear aim	Inclusion of consecutive patients	Prospective data collection	Endpoints appropriate to the aim	Unbiased assessment of the endpoint	Follow-up period appropriate (minimum 2 years)	Follow-up loss less than 5%	Prospective calculation of the study size	Adequate control group	Contemporary groups	Baseline equivalence of groups	Adequate statistical analysis	Total
John et al 2010 [1]	2	2	1[a]	2	2	2	0	0	NA	NA	NA	NA	11
Grano et al 2010 [2]	2	2	2	2	2	1	1	0	NA	NA	NA	NA	12
Yang et al 2016 [3]	2	2	2	2	0	2	1	0	NA	NA	NA	NA	11
Yeh et al 2014 [4]	2	2	1[a]	2	0[b]	2	1	0	NA	NA	NA	NA	10
Yamataka et al 2009 [5]	2	2	2	2	2	1[c]	1	0	2	0	1[d]	2	17
Granstrom et al 2013 [6]	2	1	2	2	2	2	1	0	NA	NA	NA	NA	12

Hukkinen et al 2014 [7]	2	2	2	2	2	2	2	0	NA	NA	NA	NA	14
Roorda et al 2018 [8]	2	2	2	2	0	2	1	0	NA	NA	NA	NA	11
Levitt et al 2013 [9]	2	2	2	2	0	2	1	0	NA	NA	NA	NA	11
Sood et al 2018 [10]	2	1	2	2	2	0[e]	0	0	2	1	0[f]	2	14
Khalil et al 2015 [11]	2	2	2	2	0	2	2	0	NA	NA	NA	NA	12
Meinds et al 2019 [12]	2	2	2	2	0[g]	0	1	0	2	1	2	2	16
Mathias et al 2016 [13]	2	1	1	2	0	1	1	0	2	1	2	2	15
Lane et al 2016 [14]	2	2	2	2	0	2	2	0	NA	NA	NA	NA	12
Collins et al 2017 [15]	2	1	2	2	0	2	2	0	NA	NA	NA	NA	11
Aworanti et al 2012 [16]	2	2	2	2	0	2	2	0	NA	NA	NA	NA	12
Allin et al 2020 [17][h]	2	2	2	2	0	2	2	0	NA	NA	NA	NA	12

Espeso et al 2020 [18]	2	1	2	2	0	1[i]	0	1[j]	1	2	1[k]	2	15
Saysoo et al 2020 [19][l]	2	2	2	2	0	2	2	0	NA	NA	NA	NA	12
Townley et al 2020[20]	2	1	2	2	2	2	1	1[j]	2	1	2	2	20
Wong et al 2020[21]	2	2	2	2	0[m]	2	1	1	2	1	1[n]	2	18
Zhuansun et al 2020 [22]	2	1[o]	2	2	2	2	1	1[j]	NA	NA	0	0	13

Note: Items 1 through 7 are for non-comparative, while 8 through 12 are for comparative studies.
[a]no REB approval stated or protocol but their procedure is detailed.
[b]The staff who reviewed the charts the same as those who conducted the telephone interviews.
[c] Only 6-month follow up done but not for 2 years.
[d]Similar proportion of male vs. female in control and experimental groups.
[e]Prospective cohort study, but no follow-up.
[f]No table differentiating demographic variables or other confounders.
[g]Blinding not mentioned.
[h]No proper comparison group. They examine affected length of bowel in the same cohort.
[i]Mention that children and parents were followed-up but no median or mean follow-up value.
[j]No sample size or power calculated but detailed and appropriate statistical methods.
[k]No stat. difference in age at surgery, sex, resected length, type of surgery between participants versus non-participants but no indication if this comparison was made between children vs teens, teens vs parents, or parents vs children.
[l]No comparator group. Just divided children into surgery types.
[m]All procedures were performed by the same team of surgeons but no mention of blinding for QoL questionnaire interview.
[n]No stat. difference in gender between cases vs controls but age is very different.
[o]Patients excluded to minimize bias but the bias is unexplained.

Table S4. Cochrane Risk of Bias table for Wang et al 2015

Entry	Judgement	Support for judgement
Random sequence generation (selection bias)	Low Risk	"The patients were randomized to either control or intervention group (1:1) by using computer-generated random numbers."
Allocation concealment (selection bias)	Low Risk	"The results of the randomization were not revealed until the beginning of treatment and the group assignment was not known by the investigators who evaluated the outcome of the treatments and the nursing program."
Blinding of participants and personnel (performance bias)	Low Risk	"The results of the randomization were not revealed until the beginning of treatment and the group assignment was not known by the investigators who evaluated the outcome of the treatments and the nursing program."
Blinding of outcome assessment (detection bias) (patient-reported outcomes)	Unclear Risk	Comment: Investigators did not know the group assignment. However, it is unclear if patients did know their assignment.
Incomplete outcome data (attrition bias)	Low Risk	Comment: Intervention (n=43) and control group (n=42) were similar in sample size. All patients were followed up for 6-12 months' time. Same list of outcomes were assessed for both intervention and control groups.
Selective reporting (reporting bias)	High Risk	Comment: Outcomes such as social activities were mentioned in discussion column but not pre-specified. Parental satisfaction was pre-specified however patient emotional satisfaction was not, but it was mentioned in the discussion. "The results of this study showed that the post-operative quality of life in most cases was good, but some individuals did exhibit reduced social activities and different degrees of inferiority in peer interactions."

Threshold for converting the Cochrane Risk of Bias Tool to AHRQ Standards (Good, Fair, and Poor)[1]

Fair Quality: Selective reporting (reporting bias) was not met as it yielded "High Risk". With selective reporting domain, various outcomes were not pre-specified, however were mentioned in the discussion. As well, one or more outcomes such as social activities and patient behaviour/emotion were reported briefly thus unable to be incorporated into a meta-analysis.

[1]Higgins JPT, Altman DG, Gøtzsche PC, et al. The Cochrane Collaboration's tool for assessing risk of bias in randomised trials. *BMJ*. 2011;343(7829):1-9. doi:10.1136/bmj.d5928

Supplementary File 4

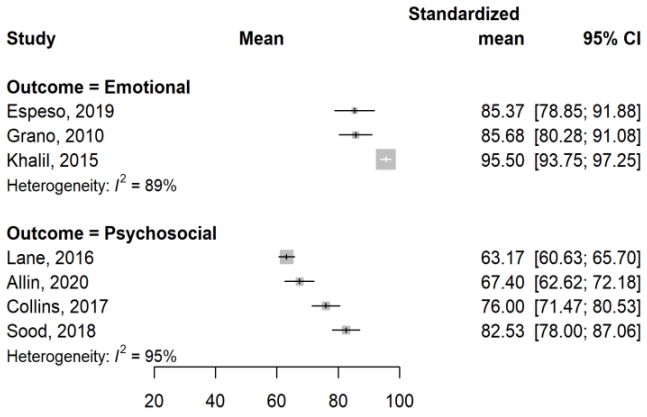

Figure S1. Standardized mean scores for parent-reported emotional and psychosocial domains only

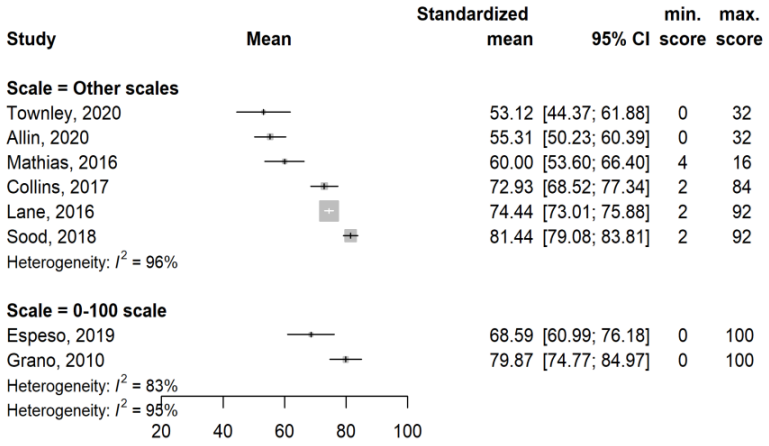

Figure S2. Subgroup analysis of instrument type (0-100 vs other) for parent-reported incontinence scores

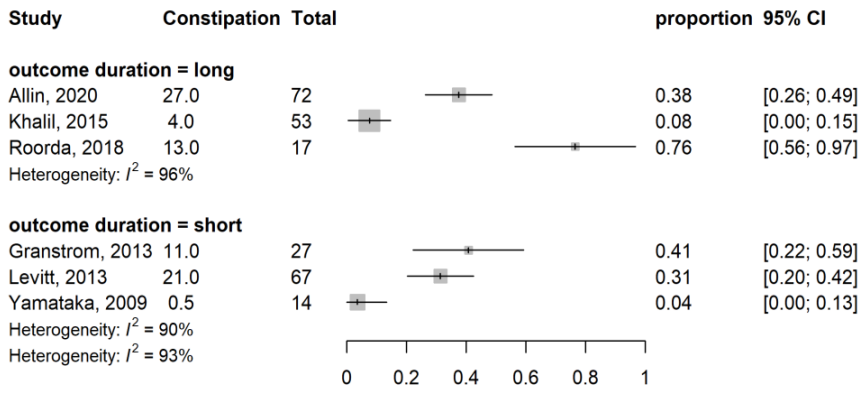

Figure S3. Subgroup analysis of duration of outcome on parent-reported constipation proportions