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THE MANAGEMENT OF POST-APPENDECTOMY ABSCESS IN CHILDREN; A MULTICENTER HISTORICAL COHORT STUDY

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Aim of the study: To compare the outcomes of non-invasive and invasive treatment strategies for children with post-appendectomy abscess (PAA).

Methods: A historical cohort study was performed, including all children (<18 years) treated for a radiologically confirmed PAA between 2014-2021 in a tertiary referral center and two large peripheral centers. Non-invasive treatment strategies were preferred in one of the peripheral centers, while the other preferred aggressive treatment strategies with drainage procedures. Medical charts were reviewed to compare non-invasive (i.e. antibiotics) and invasive (i.e. drainage procedures) treatment strategies. Primary outcome was the immediate success rate of treatment, defined as the absence of complications and no need for further interventions related to PAA.

Results: During the study period, 70 children were treated for PAA, of which 29 were treated non-invasively and 41 invasively. PAA resolved in all patients, but In the non-invasive group treatment was immediately successful in 21/29 patients compared to 25/41 patients in the invasive group. Non-invasive treatment was immediately succesful in 7/7 cases of unifocal small (<3cm) PAA and 12/15 patients with unifocal medium size PAA (3-6cm), but not for multiple abscesses. Invasive treatment was preferred for PAA >6cm and was immediately successful in 8/13 patients with PAA <6cm.

Conclusions: Non-invasive treatment of small and medium size PAA in children seems to be safe and equally successful as invasive treatment. Based on these results, a standardized treatment protocol was developed, in which a step-up approach is recommended for PAA <6cm. Prospective validation of this treatment protocol is recommended.

Table 1. Immediate success rate of treatment

	Non-invasive treatment (n=29)	Invasive treatment (n=41)
Immediate success rate		
< 3cm	7/7	0/1
3 - 6cm	12/15	8/12
> 6cm	2/3	9/16
Multiple	0/3	7/11
Unknown size	0/1	1/1

Data is displayed as count

MANAGEMENT AND OUTCOME OF HIGH-RISK NEUROENDOCRINE TUMORS OF THE APPENDIX IN CHILDREN; A SYSTEMATIC REVIEW OF THE LITERATURE

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Aim of the study: To systematically review the literature regarding the management and outcomes of children with a high-risk neuro-endocrine tumor (NET) of the appendix.

Methods: A systematic search was performed in PubMed, Embase and Web of Science. All randomized controlled trials, cohort studies, and case series, reporting on the management and outcomes of patients (<20 years) with a histopathologically proven NET of the appendix were eligible for inclusion. Two authors independently selected eligible articles, assessed risk of bias, and extracted data. High-risk (pT2 with risk factors and pT3) tumors were analyzed, in which the outcomes of complementary surgery were compared to appendectomy without complementary surgery. Primary outcomes were recurrence rate, disease-free and overall survival.

Results: The literature search yielded 607 articles, of which 26 were included. These studies reported on 796 patients, of which 121 with high-risk NET. Heterogeneity between studies was large and risk of bias was serious in 24 and moderate in two studies. Complementary surgery after primary appendectomy was performed in 48/121 patients. Length of follow-up ranged between 1-612 months. In both treatment groups no recurrences were reported, and thus disease-free survival was 100%. Overall survival was 100% in the complementary surgery group and 98.4% in the appendectomy group.

Conclusion: Based on current literature, the value of complementary surgery for pediatric high-risk NET of the appendix can be questioned. However, evidence is scarce, of low-quality, and heterogeneity between studies is large. An EUPSA endorsed international study is planned to generate high-quality evidence on this topic.

OUTCOMES AFTER SURGICAL TREATMENT FOR RECTAL ATRESIA IN CHILDREN: IS THERE A PREFERRED APPROACH? A SYSTEMATIC REVIEW.

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Background: Rectal atresia (RA) affects 1-2% of all children with anorectal malformations. No consensus on optimal treatment strategy is yet achieved. Therefore, the aim of this systematic review is to summarise all surgical interventions for RA and outcomes described in the current literature.

Methods: A literature search was conducted in Pubmed, Embase, Web of Science and Cochrane Library on January 24th 2022. All studies describing treatment for RA in children (<18 years) were included. Operation technique and postoperative complications were listed. Only descriptive analysis was anticipated. Quality of the studies was assessed using JBI critical appraisal checklist for case reports and series.

Results: The search yielded 6716 studies, of which 4028 were excluded based on title and abstract screening. After full-text assessment, 22/90 studies were included, yielding 70 patients. Posterior sagittal anorectoplasty (PSARP) and pull-through were most performed (43/70 and 18/70 patients, respectively). Four patients experienced postoperative complications: anal stenosis (n=1), anastomotic stenosis (n=2), and death due to a pulmonary complication (n=1). All of the included studies had moderate to high methodological quality.

Conclusions: RA is extremely rare and little is known about the various treatment options. The majority of patients (61/70) were treated with posterior sagittal or pull-through approach. Postoperative complication rates were low, but often not reported. Larger cohort studies should be performed to determine optimal treatment strategy for children with RA, taking into account accurate definition of RA and surgical intervention, and registration of postoperative complications as well as long-term functional outcomes.

HISTOPATHOLOGICAL CHARACTERISTICS OF HIRSCHSPRUNG DISEASE REDEFINED

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Background: To determine proximal resection plane during definitive surgery for Hirschsprung disease (HD) we need to correctly recognize transition zone (TZ). Therefore, we studied histopathology of healthy bowels in comparison to HD bowels in order to identify characteristics and cut-off values for TZ.

Methods: We compared autopsy-tissue from babies without intestinal disease with TZ and ganglionated bowel from HD-resection-specimen (circumferentially sampled with 1-cm-margins) on submucosal and myenteric (ectopic/hypocellular) ganglia presence, myenteric ganglion-density and submucosal nerve diameter. ROC-curves were used determining cut-off values for ganglion-density and nerve diameter, expressed as area under curve (AUC). Correlation with length TZ was tested using Spearman's correlation.

Results: We compared histopathology of 16 control-slides with TZ from 17 HD patients (157 slides) with mean age of 10 (range 2-155) weeks and 15 (range 9-306) weeks respectively. We found myenteric ganglion-density <13.9 ganglion/cm/area distinctive (AUC .865) and nerve diameter > 29.6 µm not distinctive for TZ (AUC .530). We identified following TZ-characteristics: myenteric/submucosal aganglionosis of ≥1/8th circumference, myenteric ectopic/hypocellular ganglion(s) and myenteric ganglion hypodensity (<13.9 ganglion/cm/area). HD patients had mean length of 5.3 (range 0-20) cm aganglionated bowel and 6.0 (range 0-16.5cm TZ). Length of TZ did not correlate with length aganglionated bowel (-.455; p=.138) and age at resection (0.063, p=.846).

Conclusions: Characteristics of TZ are myenteric/submucosal aganglionosis of ≥1/8th circumference, myenteric ectopic/hypocellular ganglion(s) and myenteric ganglion density <13.9 ganglion/cm/area. Length of TZ cannot be determined based on length of aganglionated bowel in patients with HD.

RISK FACTORS FOR COMPLICATIONS IN PATIENTS WITH HIRSCHSPRUNG DISEASE WHILE AWAITING SURGERY: BEWARE OF BOWEL PERFORATION

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Background: Patients with Hirschsprung disease (HD) mostly undergo surgery around the age of three to six months. While awaiting surgery, therapy to treat the obstruction such as transanal irrigation (TAI) or laxatives is applied. The aim of this study was to gain insight in the prevalence and severity of complications occurring while awaiting surgery and to identify patient characteristics associated with the development of these complications.

Methods: This study retrospectively analyzed data of patients with HD operated in our center between 2000-2021. Complications emerging while awaiting surgery were graded using Clavien-Dindo (CD). Patient characteristics as predictor of a complication were tested using logistic regression analysis.

Results: Twenty-two of 132 (17%) included patients (preoperative treatment: 94% TAI; 2% laxatives; 2% other therapy) developed 45 complications while awaiting surgery, including predominantly major complications (91%). Bowel perforation occurred most frequently (n=9, 7%) wherefrom six caused by TAI (5%), including three patients with total colon aganglionosis (TCA) (2%) counting one life-threatening and one lethal perforation. The other perforations were caused by meconium ileus (n=2) and Hirschsprung associated enterocolitis (HAEC) (n=1). Other frequent complications were: sepsis (5%), ileus (4%) and persistent obstruction (4%). Predictive factor for developing complication was TCA (OR 9.905, CI 2.994-32.772, $p<0.001$).

Conclusions: We found a complication rate of 17% in patients while awaiting surgery, reporting bowel perforation most frequently. We found this complication in patients with TCA being highly dangerous causing one life-threatening and one lethal perforation. Therefore, we advise in patients with (suspected) TCA to limit the time awaiting surgery.

DIAGNOSING HIRSCHSPRUNG DISEASE: INSIGHTS IN INCIDENCE OF COMPLICATIONS OF RECTAL SUCTION BIOPSY, FINAL DIAGNOSIS AND PREDICTIVE FACTORS FOR DIAGNOSIS

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Background: Gold standard for diagnosing Hirschsprung disease (HD) is pathological examination on rectal suction biopsy (RSB). The aim of this study was to gain insight into complications following RSB, final diagnosis of patients undergoing RSB and predictive factors for HD.

Methods: Neonates suspected of HD undergoing RSB at our tertiary centre between 2000-2021 were analyzed retrospectively. Severity of complications of RSB was assessed using Clavien-Dindo (CD) grading. Predictors for HD were tested using multivariate regression.

Results: HD was diagnosed in 151 of 371 included patients (40.7%) with median age of 44 days (2-175). Three patients developed complications following RSB (0.8%) including ongoing rectal bleeding (n=3), all graded CD-1. Functional constipation was diagnosed in 100 patients (26.9%) followed by treatment resistant constipation in thirteen patients (3.5%). Predictors for HD were: male sex (OR 3.19, CI 1,56-6,53), presence of syndrome (OR 7.18, CI 1,63-31.69), lower age at RSB (OR 0.98, CI 0.98-0.85) meconium passage > 48 hours (OR 3.15, CI 1.51-6.56), distended abdomen (OR 2.09, CI 1.07-4.07), bilious vomiting (OR 6.39, CI 3.28-12.47) and failure to thrive (OR 8.46, 2.11-34.02) (model R^2=0.566).

Conclusions: RSB is safe procedure with few and only minor complications. Therefore, we advise to continue current clinical practice: obtaining low-threshold RSB. In case of negative RSB, functional constipation should be considered.We recommend to start adequate treatment for HD without awaiting the results of the RSB, in patients with two or more classical symptoms (>48 hours meconium passage, bilious vomiting and distended abdomen), to prevent complications from delayed bowel decompression.

TRANSITION ZONE PULL-THROUGH IN PATIENTS WITH HIRSCHSPRUNG DISEASE: IS PERFORMING REDO SURGERY BENEFICIAL FOR THE LONG-TERM QUALITY OF LIFE?

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Background: The definitive treatment of patients with Hirschsprung Disease (HD) is surgery, whereby in case of transition zone pull-through (TZPT) the surgeon faces dilemma of performing redo surgery. Therefore, we aimed to assess long-term quality of life, short-term postoperative complications after redo-surgery, long-term functional outcomes and the requirement of interventions in patients with TZPT treated conservatively or with redo surgery, compared to HD patients without TZPT.

Methods: We retrospectively included patients with TZPT operated between 2000 and 2021 in our center. Each TZPT patient was matched with two patients who underwent successful surgery based on length of resected bowel and age at resection. Included patients received the Hirschsprung/Anorectal Malformation Quality of Life questionnaire and items of Groningen Defecation & Continence, and we assessed the requirement of interventions and occurrence of hirschsprung associated enterolocitis (HAEC) comparing scores between conservative-group, redo-group and control-group using One-Way ANOVA.

Results: Fifteen TZPT-patients, of which six treated conservatively and nine received redo surgery, were matched with 30 control patients. Mean follow-up length was 90 months (SD = 59). No significant differences between groups were found in quality of life (p=0.63), functional outcomes (0.67), use laxatives (p= 0.33), use rectal irrigation (p=0.11) and given Botulinum Toxin injections (p= 0.06). HAEC was developed in three control-patients (10%), one redo patient (11%) and zero conservative patients (0%).

Conclusions: Our findings suggest that conservative treatment of TZPT should be considered, because of similar long-term quality of life, functional outcomes and the requirement of interventions compared to patients undergoing redo surgery.

DETERMINING THE CORRECT RESECTION LEVEL IN PATIENTS WITH HIRSCHSPRUNG DISEASE USING CONTRAST ENEMA AND FULL THICKNESS BIOPSIES: CAN THE DIAGNOSTIC ACCURACY BE IMPROVED BY EXAMINING SUBMUCOSAL NERVE FIBER THICKNESS?

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Background: Intraoperative resection level in patients with Hirschsprung disease (HD) is determined by contrast enema, surgeon's intraoperative judgement and full thickness biopsy (FTB) identifying ganglia. This study aims to evaluate diagnostic accuracy of contrast enema and FTB in determination of resection level and whether this can be improved by measuring submucosal nerve fiber diameter.

Methods: We retrospectively analyzed contrast enema and intraoperative FTBs obtained in our center, determining diagnostic accuracy for level of resection. Gold standard was pathological examination of resection specimen. Secondly, we matched transition zone pull-through (TZPT) patients with non-TZPT patients, based on age and length of resected bowel, to blindly compare nerve fibers diameters between two groups using group comparison.

Results: From 2000-2021, 209 patients underwent HD surgery of whom 180 patients (138 males; median age at surgery: 13 weeks) with 18 TZPTs (10%) were included. Positive predictive value of contrast enema was 65.1%. No caliber change was found in patients with total colon aganglionosis (TCA). Negative predictive value of surgeon's intraoperative judgement and FTB in determining resection level was 79.0% and 90.0% (91.2% single-stage, 84.4% two-stage surgery) respectively. Mean nerve fiber diameter in TZPT was 25.01 µm (SD= 5.63) and in non-TZPT 24.35 µm (SD= 6.75) (p=0.813).

Conclusions: Determination of resection level with combination of contrast enema, surgeon's intraoperative judgement and FTB results in sufficient diagnostic accuracy in patients with HD. If no caliber change is seen with contrast enema, TCA should be considered. Resection level or transition zone cannot be determined by assessment of submucosal nerve fiber diameter in FTB.

DIAGNOSTIC ACCURACY OF CALRETININ AND ACETYLCHOLINESTERASE STAINING OF RECTAL SUCTION BIOPSIES IN HIRSCHSPRUNG DISEASE EXAMINED BY UNEXPERIENCED PATHOLOGISTS

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Background: Rectal suction biopsy (RSB) is gold standard for diagnosing Hirschsprung disease (HD). Calretinin staining of RSB is increasingly used by experienced pathologists due to non-complex examination and comparable diagnostic accuracy with acetylcholinesterase (AChE). However, the diagnostic accuracy of calretinin examined by unexperienced pathologists remains to be elucidated. Therefore, we aim to compare diagnostic accuracy of calretinin with AChE on RSB for diagnosing HD when examined by unexperienced pathologists.

Methods: We prospectively analyzed sections from RSB stained with AChE+HE and calretinin. Blinded examination was done by five unexperienced pathologists (pathology residents) and three experienced pathologists (senior pediatric gastro-enterology pathologists) assessing for the presence of HD. Cases for the study included ones proven to be HD on resection specimens and cases without HD. Diagnostic accuracy was determined calculating area under the curve (AUC), sensitivity, specificity, likelihood ratio and posttest probability. Fleiss' kappa analysis was performed to assess interobserver agreement between reviewers.

Results: Eleven of 18 included patients (61%) were diagnosed with HD. Comparing the diagnostic accuracy of unexperienced pathologists, calretinin versus AChE+HE showed sensitivity of 80.0% versus 74.5% and specificity of 100% versus 65.4%, AUC of 0.87 (0.78 – 0.96) versus 0.59 (0.45-0.72). Unexperienced pathologists showed substantial agreement with calretinin (kappa 0.72 [0.61-0.84]) and fair agreement with AChE+HE (kappa 0.34 [0.23-0.44]).

Conclusions: We found calretinin having higher diagnostic accuracy in diagnosing HD compared to AChE+HE when examined by unexperienced pathologists. Therefore, we recommend to use calretinin as the standard technique for staining RSB in diagnosing HD.

OUTCOME OF ROUTINE CYSTOSCOPY AND VAGINOSCOPY PRIOR TO SURGICAL TREATMENT OF CHILDREN WITH ANORECTAL MALFORMATION

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Background: In +/- 40% of children with anorectal malformations (ARM) the urogenital tract is also affected. Early identification of these anatomical and functional anomalies is important for an optimal outcome and helpful in counselling of the parents. The aim of this study is to evaluate the results and usefulness of routine cystoscopy and vaginoscopy, prior to surgical treatment of children with ARM, in identifying additional urological and gynecological anomalies.

Methods: All patients born between January 2019 and December 2021 who were scheduled for a PSARP or ASARP were eligible for inclusion. They underwent a routine cystoscopy and in girls also a vaginoscopy. Preoperative symptoms, VACTERL documentation, findings during cysto- vaginoscopy and postoperative complications were analysed.

Results: In total, 31 patients underwent a ASARP/PSARP in this time period; in 17 patients cysto- and/or vaginoscopy was done: 13 males, 4 females (Table 1).
In 4 patients a fistula could be visualized. In 1 boy the orifice of the fistula was found in the urethra instead of the bladderneck, as suggested by radiological investigations, and a laparoscopy was thus prevented.
Preoperatively 3 boys suffered from febrile urinary tract infections (UTI) and all 3 had urological anomalies. In the girls, no vaginal anomalies were seen. No complications related to cystoscopy were encountered. Median follow up was 18 months.

Conclusions: Although few abnormalities were identified by routine cystoscopy and vaginoscopy, it is a safe and quick method identifying potential harmful anomalies and thus prevent urological sequelae in future. Further evaluation in larger numbers is needed.

General characteristics		Outcomes				
Gender	**Type of fistula**	**Abnormalities cystoscopy**	**UTI*/LUTS****	**Abnormal US# kidney/spine**	**Reflux**	**Level of fistula know preoperative**
Male	Recto-perineal n=6	2 / 6 syringocele PUV^ (n=1), mild PUV^ (n=1)	0	0	0	NA##
	Recto-urethral n=1	1 / 1 fistula urethral not bladderneck	0	1 /1 caudal regression syndrome	0	1 / 1
	Recto-prostatic n=2	2 / 2 fistula	1 / 2 UTI and LUTS	2 / 2 hydronephrosis (n=1), thick bladder wall (n=1)	0	2 / 2
	Recto-vesical n=2	1 / 2 PUV^	1 / 2 UTI	1 / 2 low conus medullaris	0	1 / 2
	Anal stenosis n=2	1 / 2 syringocele	1 / 2 UTI	1 / 2 ectopic kidney	1 / 2 bilateral vesico-ureteral reflux	NA##
Female	Recto-perineal n=2	1 / 2 bladder trabeculation	0	0	0	NA##
	Recto-vestibular n=2	0	0	0	0	NA##

Table 1. *UTI = urinarytract infection; **LUTS = lower urinarytract symptoms; ^PUV = posterior urethral valves; #US = ultrasound; ##NA = not applicable

DIAGNOSTIC ACCURACY OF PALPATION AND ULTRASONOGRAPHY FOR DIAGNOSING HYPERTROPHIC PYLORIC STENOSIS: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background: Although palpation was used to diagnose infantile hypertrophic pyloric stenosis (IHPS) earlier, nowadays ultrasonography is generally used. However, many different parameters are used and a systematic review regarding the optimal diagnostic strategy is lacking. We conducted a systematic review and meta-analysis to obtain diagnostic accuracy of methods to diagnose IHPS.

Methods: According to the PRISMA guidelines we searched MEDLINE and Embase to identify studies reporting sensitivity and specificity of all methods to diagnose IHPS. Inclusion criteria were infants with suspicion of/or diagnosed with IHPS who underwent pyloromyotomy or had clinical follow-up. A random-effects model was used to obtain pooled estimates of sensitivity, specificity and area under the receiver operating characteristic curve (AUC).

Results: After screening 4493 studies, we included 42 studies with 5801 infants (n=4099 IHPS; n=1702 controls)(Figure 1). The diagnostic sensitivity of palpation ranged from 10.0%–93.4% and decreased over time. Different parameters for ultrasonography were found with sensitivity and specificity ranging from 51.7%-100% and 0%-100%, depending on patient population, parameter and cut-off level chosen. Most used parameters were pyloric muscle thickness (PMT) ≥3 mm (pooled sensitivity 97.5%, specificity 99.6%, AUC 0.999) or a combination of PMT ≥4 mm and/or pyloric canal length (PCL) ≥16 mm (pooled sensitivity 94.0%, specificity 91.7%, AUC 0.981). Insufficient evidence was found for the use of other diagnostic methods.

Conclusions: Palpation has limited sensitivity in diagnosing IHPS. We showed that ultrasonography has highest diagnostic accuracy to diagnose IHPS and advise to use PMT ≥3 mm as cut-off.

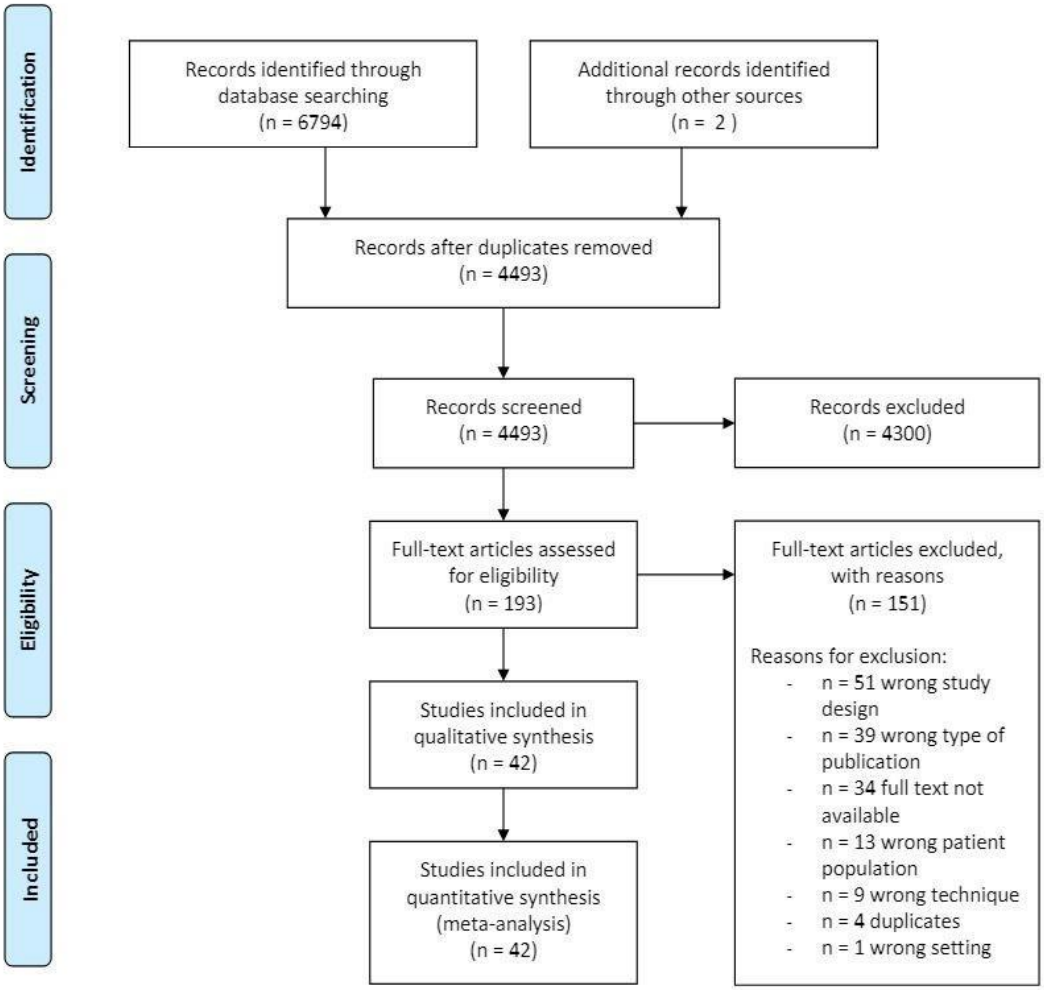

Figure 1. PRISMA flowchart

AGE-SPECIFIC AND FAMILY-CENTERED APPROACH TO PREPARE CHILDREN AT HOME FOR DAY-CARE SURGERY

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Background: Surgery induced stress and anxiety can be reduced by providing preoperative information adapted to the needs of children and parents. This study aimed to investigate the efficacy of a coloring page, mobile application and videos to prepare children and their parents for day-care surgery in terms of child's preoperative anxiety and postoperative pain, and preoperative anxiety and satisfaction of parents.

Methods: Prospective observational study including children and parents that were offered specifically developed information modalities to prepare for day-care surgery. Results were compared between children that used none (i.e., control group) versus one or more information modalities (i.e., intervention group). Primary outcomes were preoperative anxiety measured using PROMIS v2.0 Anxiety and postoperative pain in children. Secondary outcomes were preoperative parental anxiety (STAI questionnaire) and family satisfaction with information and communication (modified PedsQL Healthcare Satisfaction questionnaire). Subgroup analyses were performed between preschoolers (0-5 years) and school-aged (≥5 years) children.

Results: 93 patients (male 53%) were included in the intervention (n=56) and control group (n=37). Levels of children's preoperative anxiety and postoperative pain, and parental anxiety were equivalent between both groups. Families of prepared children were more satisfied with information and communication about preoperative surgical information (8 vs. 6.6, p=0.004) and satisfaction with how parents (7 vs 8, p=0.019) and children (8 vs 6, p=0.018) were prepared for surgery.

Conclusions: Preoperative anxiety was similar between prepared and unprepared children. The use of specifically developed family-centered and age-appropriate information modalities to prepare children for day-care surgery at home improves family satisfaction.

CENTRAL VENOUS CATHETER ASSOCIATED BLOODSTREEM INFECTIONS AND THROMBOSIS IN PATIENTS TREATED FOR GASTROSCHISIS AND INTESTINAL ATRESIA. A RETROSPECTIVE COHORT STUDY OF 238 PATIENTS.

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Background: Central venous catheter related complications, mainly central line associated bloodstream infections (CLABSI) and thrombosis, can complicate treatment of intestinal atresia or gastroschisis. Incidences and risk factors, e.g., location (central versus peripheral inserted) or mode of insertion (tunnelled versus non-tunnelled), of these complications are widely unknown. Therefore, we aim to identify the incidence of and risk factors for CLABSI and thrombosis in these patients.

Methods: Children treated for intestinal atresia or gastroschisis between 1998-2021 at our tertiary referral center were retrospectively included. Risk factors for CLABSI were identified using logistic regression and the correlation between thrombosis and location/mode of insertion was evaluated using chi^2-tests.

Results: In 238 patients, 35% developed CLABSI and 7% thrombosis. Treatment by enterostomy (OR:3.1;95%-CI:1.5–6.4) and non-tunnelled catheters (OR:2.0;95%-CI:1.3–4.6) significantly increased the CLABSI-risk whilst patient's sex, catheter placement directly into central vein, preterm birth, trisomy 21, experiencing a major postoperative complication (Clavien-Dindo grade ≥III) and birthweight didn't. Catheter dwell time was shorter ($p<0.01$) and CLABSIs developed faster ($p=0.02$) in non-tunnelled compared to tunnelled catheters. Catheter related thrombosis occurred more often in non-tunnelled catheters ($p=0.03$), whilst no correlation ($p=0.34$) was found between location of insertion.

Conclusion: CLABSI occurs in approximately one third of the children treated for intestinal atresia or gastroschisis and a catheter related thrombosis occurs in one in fourteen. When in doubt which catheter to use in these patients, a tunnelled catheter is preferred over a non-tunnelled, specifically in case of enterostomy formation, since a tunnelled catheter is comparatively less at risk of catheter related infections and thrombosis.

ROUTINE CONTRAST ENEMA PRIOR TO STOMA REVERSAL SEEMS ONLY REQUIRED FOLLOWING TREATMENT FOR NECROTIZING ENTEROCOLITIS: AN EVALUATION OF THE DIAGNOSTIC ACCURACY OF THE CONTRAST ENEMA

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Background: Contrast enemas are often made prior to stoma reversal in order to detect distal intestinal strictures distal of the stoma. If untreated these strictures can cause obstruction which might necessitate redo-surgery. However, the value of contrast enemas is unclear. Therefore, we aim to evaluate the contrast enema's diagnostic accuracy in detecting strictures in children with a stoma.

Methods: Young children (≤3 years) treated with a stoma between 1998-2018 were retrospectively included. The STARD criteria were followed. Patients treated for anorectal malformations and those that died before stoma reversal were excluded. Surgical identification of strictures during reversal or redo-surgery within three months was used as gold standard. Sensitivity, specificity, positive predictive value (PPV), negative predictive value (NPV) and area under the curve (AUC) reflected diagnostic accuracy.

Results: In 224 included children, strictures were found during reversal in 10% of which 95% in patients treated for necrotizing enterocolitis. Contrast enema was performed in 68% of all patients and detected 92% of the strictures. In the overall cohort, the sensitivity was 100%, specificity 98%, PPV 88% and NPV 100% whilst the AUC was 0.98. In patients treated for NEC, the sensitivity was 100%, specificity 97%, PPV 88% and NPV 100% whilst the AUC was 0.98.

Conclusion: Strictures prior to stoma reversal seem to be mainly identified in patients treated for NEC and not in other diseases necessitating a stoma. Moreover, the contrast enema shows excellent diagnostic accuracy in detecting these strictures. For this reason we advise to only perform contrast enemas in patients treated for NEC.

INSIGHTS INTO THE EARLY STAGES OF COLORECTAL ANASTOMOTIC HEALING AND LEAKAGE IN RATS

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Background: Anastomotic leakage is a dreaded complication after colorectal surgery. The pathophysiology is largely unknown, especially the first stages of healing and leakage. The aim of this study was to assess healing and disturbed healing in the first 24 hours after anastomosis creation in rats.

Methods: Wistar rats were divided into a healing model (AH) with sufficient anastomosis or leakage model (AL) with insufficient anastomosis. Rats were sacrificed at 6, 12 and 24 hours. Abdominal cavity and anastomosis were assessed macroscopically with anastomotic complication score (ACS). For histological assessment, modified Ehrlich-Hunt Score (mEHS) was used. Transcriptome analysis was performed to assess differences between AL and AH at the 3 different time points to find affected genes and biological processes.

Results: In both models, 15 rats were operated (n=5 per time point). In the AL group, mean (±SD) ACS showed an increase at 24 hours after surgery relative to AH (4.0 (±0) vs 1.8 (±0.45), $p<0.001$). Edema was more present in AH, there was no difference in mEHS for inflammation. Principal component analysis of the transcriptome revealed differences between AH and AL at 6 and 24 hours. Gene set enrichment analysis indicated differences in processes in immune response and cell metabolism.

Conclusions: Histological assessment revealed no difference between AH and AL in the first 24 hours. However, at transcriptional level early after the creation of the anastomosis, AH and AL can be differentiated. These early differences will provide new leads for future research.

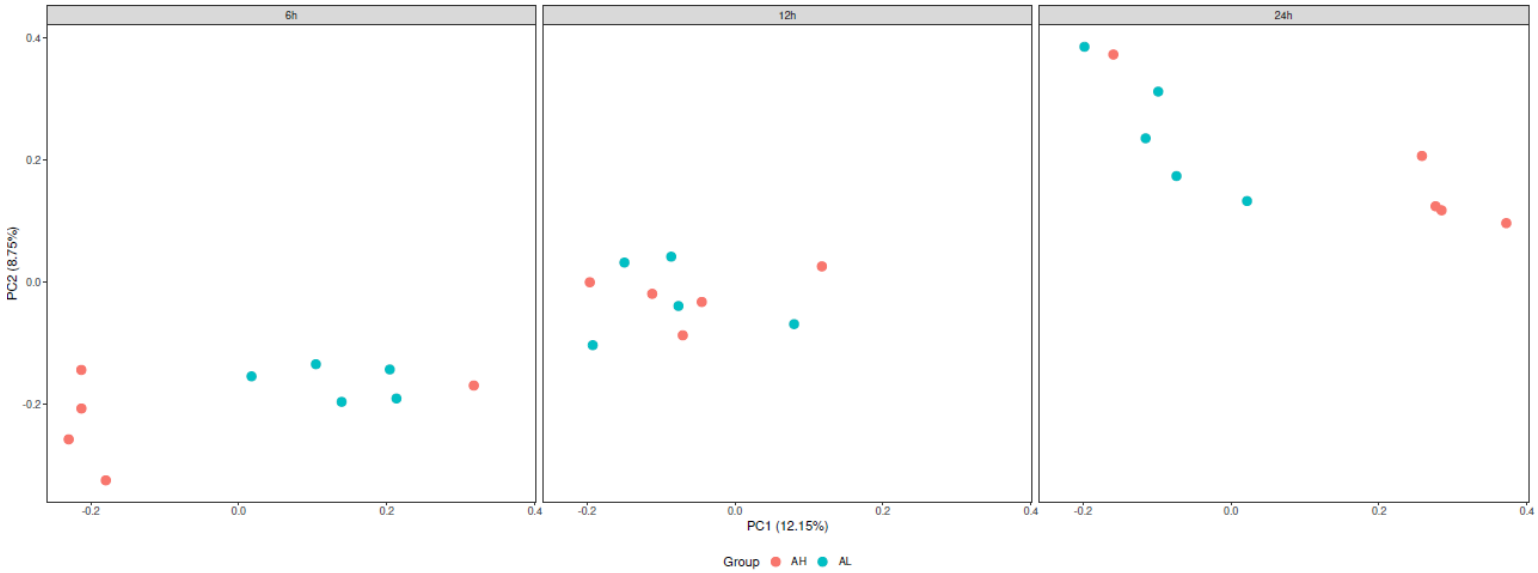

Figure 1. Principal component analysis at time 6 hours, 12 hours and 24 hours postoperative, where red is AH and blue is AL. Principal component 1 (PC1) is driven by AH/AL and principal component 2 (PC2) by time

MORBIDITY & OVERALL MORTALITY IN VERY LOW AND EXTREMELY LOW BIRTH WEIGHT INFANTS TREATED FOR ESOPHAGEAL ATRESIA: NATIONAL MULTI-CENTER COHORT STUDY.

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Background: Mortality and morbidity rates in very and extreme low birth weight (VLWB and ELBW) infants range between 15%-61% and 3%-93%. Esophageal atresia (EA) might increase these rates due to the high risk of peri-operative complications in this specific population and associated congenital anomalies. The aim of this study is to evaluate the mortality and surgery-related morbidity rates of children born with VLBW or ELBW after EA repair.

Methods: A multi-center national retrospective cohort study was performed in five academic hospitals in the Netherlands. Data was extracted from the medical record of patients born with EA and birthweight <1500 grams (VLBW) or <1000 grams (ELBW) between 2000-2019. The main outcomes were overall mortality and specific surgery-related morbidity 90 days postoperative. Only descriptive analyses were performed.

Results: In total, 44 children were born with EA and VLBW/ELBW, of which 39 (89%) underwent EA repair, 10 ELBW and 29 VLBW. In two patients repair was technically not possible and three deceased before repair. Overall mortality after repair was 23%, 5 VLBW and 3 ELBW. Anastomotic leakage occurred in 17% and 0%, anastomotic stricture 41% and 10%, pneumothorax 17% and 0% and wound infection 3% and 10%, in VLBW and ELBW respectively. For an overview see Table 1.

Conclusion: Mortality after EA repair was 23% and ranged from 17% in VBLW to 30% in ELBW. Surgery-related complications occurred in 49% of the children. Future research is needed to study the effect on long term outcomes and quality of life.

Table 1.

	Total N=39 (100%)	**VLBW N=29**	**ELBW N=10**
Birth weight (grams)	1200 (980-1294)	1220 (1144-1340)	882 (816-949)
Gestational age (weeks)	31 (29-32)	31 (29-32)	28 (27-30)
Sex (female)	16 (41%)	11 (38%)	5 (50%)
Atresia type			
Type A	2 (5%)	-	2 (20%)
Type C	37 (95%)	29 (100%)	8 (80%)
Other anomalies			
Yes	26 (67%)	18 (62%)	8 (80%)
No	13 (33%)	11 (38%)	2 (20%)
VACTERL	12 (31%)	7 (24%)	5 (50%)
Overall mortality	9 (23%)	6 (17%)	3 (30%)
Specific surgery related complications	19 (49%)	17 (59%)	2 (20%)
Anastomotic leakage	5 (13%)	5 (17%)	-
Anastomotic stenosis	13 (33%)	12 (41%)	1 (10%)
Pneumothorax	5 (13%)	5 (17%)	-
Surgical site infection	2 (5%)	1 (3%)	1 (10%)
Other complications			
Cerebral	13 (33%)	9 (31%)	4 (40%)
Pulmonary	22 (56%)	15 (52%)	7 (70%)
Cardiac	26 (67%)	18 (62%)	8 (80%)
Infectious	9 (23%)	7 (24%)	2 (20%)

Values presented as n (%) or median (IQR)

THE VALUE OF SERUM ALPHA-FETOPROTEIN LEVELS IN DIAGNOSING RECURRENT SACROCOCCYGEAL TERATOMA

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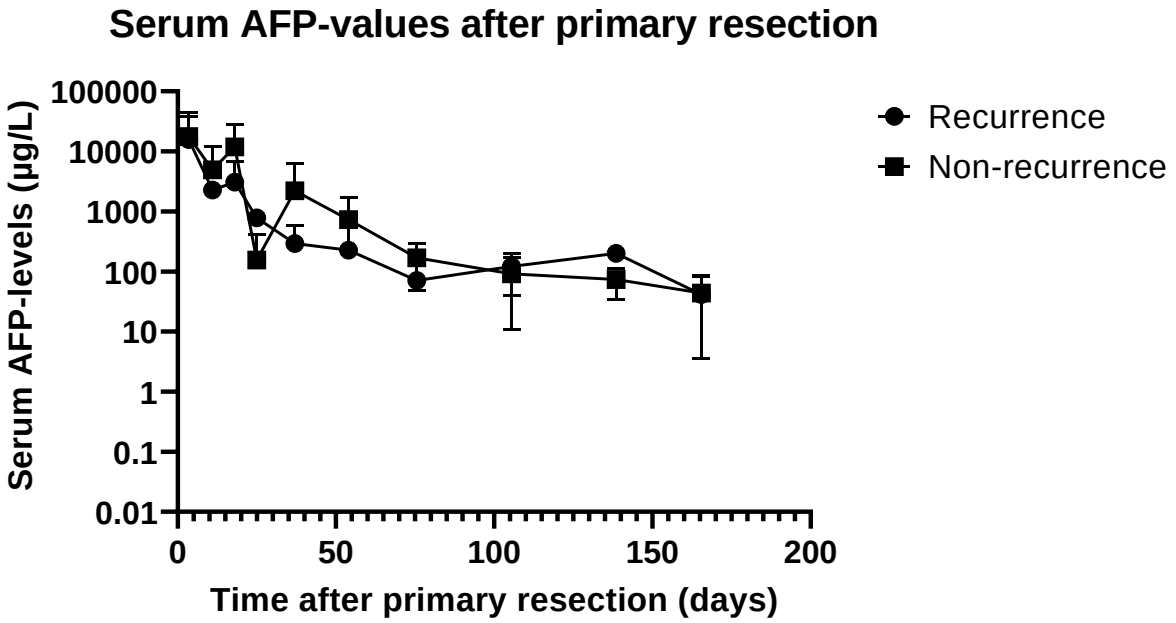
Background: Serum alpha-fetoprotein (AFP) is often used as tumor marker for recurrent sacrococcygeal teratoma (SCT), especially malignant recurrences. However, the diagnostic accuracy of serum AFP levels in detecting recurrent SCT has not been well established. We aimed to assess the normal dynamics of serum AFP-levels after resection and serum AFP-levels in detecting recurrent SCT.

Methods: This retrospective study included patients with SCT treated between January 1980 to December 2018 at the paediatric surgical centres in the Netherlands. Differences in serum AFP-levels between SCT patients with recurrence and age-matched SCT patients without recurrence were evaluated. Normal AFP dynamics were analyzed with linear regression and AFP increase prior to recurrence with Mann-Whitney U and Wilcoxon test.

Results: 57 patients with 427 AFP samples were included in the study. No significant difference was found in the serum AFP level dynamics between the recurrence and non-recurrence group after primary resection (p=0.950) (Figure 1). Nineteen children developed 20 recurrences at a median of 447 days after resection. Serum AFP-levels did not significantly increase before recurrence (p=0.850) compared to serum AFP-levels of children without recurrence at the same time. However, serum AFP levels did significantly increase in malignant recurrences (p=0.009) compared to mature and immature recurrences.

Conclusions: Dynamics of serum AFP is not different between children with and without recurrence after primary resection. Serum AFP-levels are not predictive for mature or immature recurrent SCT. However, serum AFP-levels can be used to detect malignant recurrences.


Serum AFP-values after primary resection
Serum AFP-levels (µg/L)
100000
10000
1000
100
10
1
0.1
0.01
0
50
100
150
200
Time after primary resection (days)
Recurrence
Non-recurrence

RISK OF MALIGNANT TRANSFORMATION AND TUMOUR RECURRENCE OF SACROCOCCYGEAL TERATOMA: RESULTS OF 'THE SCT STUDY'

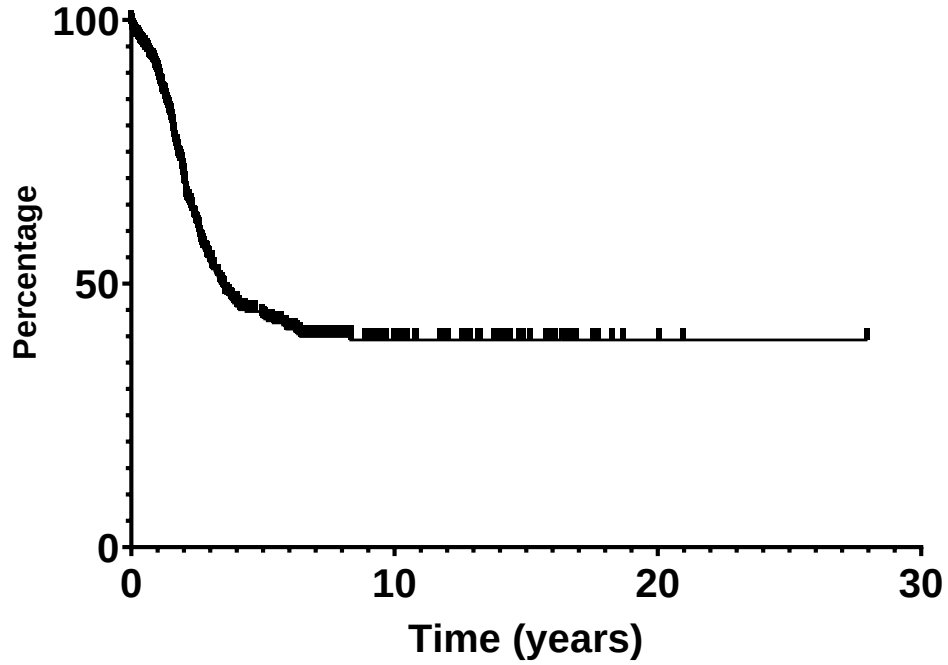
Background: Sacrococcygeal teratoma (SCT) has the risk of malignant transformation which increases with age and a risk of recurrence. Because these risks are relatively unknown and the results of previous relatively small studies differ, we aimed to examined these risks in a large cohort of SCT patients.

Methods: Retrospective SCT data were collected in an international retrospective study (The SCT Study) including patients from 150 institutes from 65 countries. Risk factors were analyzed in univariate and multivariate analysis.

Results: 3593 patients entered analysis. Freedom from malignancy at primary resection (censored for resection and death) was found in 96.8%, 94.9%, 90.2% and 69.9% of patients at three months, six months, one year and two years, respectively (Figure 1). Recurrent SCT developed in 421 (11.7%), of which 114 (27.1%) were malignant after a median interval of 9.7 months (range 11 days to 265 months). Risk factors for recurrence were Altman type II (odds ratio (OR) 1.6, 95% confidence interval (CI) 1.1-2.1), Altman type III (OR 1.7, 95% CI 1.2-2.4), initial immature histology (OR 2.2, 95% CI 1.7-2.9) and initial malignant histology (OR 4.9, 95% CI 3.7-6.6). Ten years patient survival was 94%.

Conclusions: The SCT study shows that malignant transformation in SCT increases with age with malignancy rate of 30.1% after two years, but remains relatively stable at 60% after 6 years. Recurrence after resection was found in 11.7% of patients. Altman type II or type III, and immature or malignant histology were risk factors for recurrence.

Figure 1. Freedom of malignancy at primary resection



THE RISK OF MALIGNANT TRANSFORMATION OF SACROCOCCYGEAL TERATOMA VERSUS PRESACRAL TERATOMA IN CURRARINO SYNDROME: RESULTS OF 'THE SCT-STUDY'

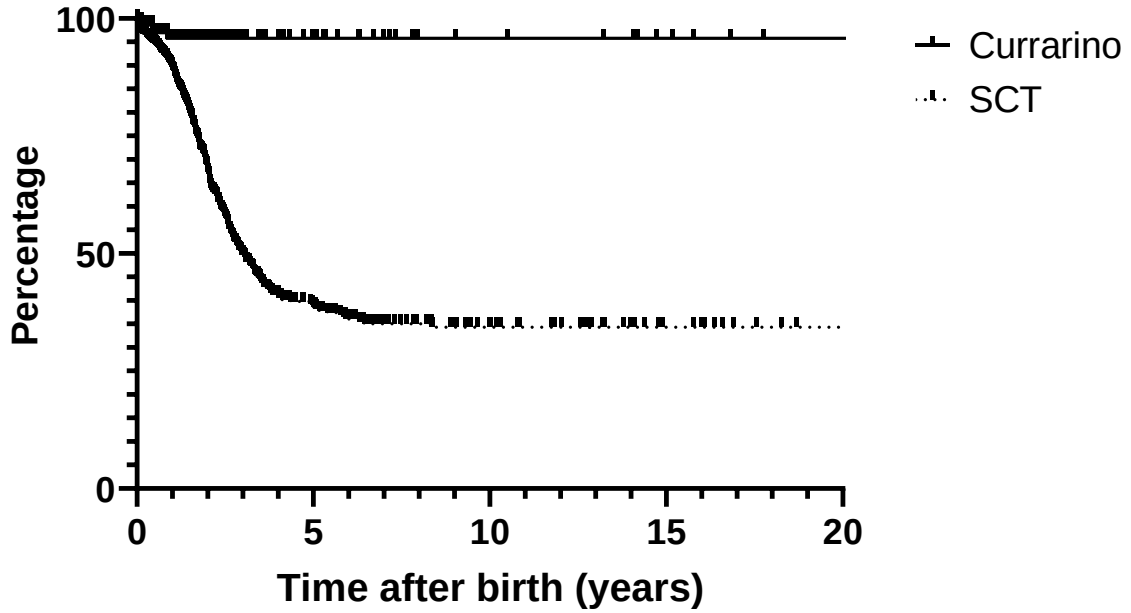
Background: The risk of malignant transformation and recurrence of Sacrococcygeal Teratoma (SCT) is relatively high. It has been suggested that the risk of malignant transformation of presacral teratoma associated with Currarino Syndrome (CS) would be lower, however the existing literature gives contradictory results. We aimed to examine the risk of malignant transformation between SCT and CS patients.

Methods: Malignant transformation and recurrence in SCT and CS patients were compared in an international retrospective cohort study (The SCT-Study). We included SCT and CS patients treated in one of 150 participating institutes from 65 countries. Risk of malignant transformation was analyzed with log rank test.

Results: Included in the analyses were 3188 SCT and 205 CS patients. Malignant transformation at primary resection increased with age in SCT patients; 10.4% and 32.1% after one and two years versus 4.2 % in CS patients after two years ($p<0.001$) (Figure 1). Recurrence was found in 376 (11.8%) SCT and 19 (9.3%) CS patients ($p=0.423$). Recurrence histology was malignant in 34.0% (128/376) of SCT patients and 10.5% (2/19) of CS patients ($p=0.075$). Overall survival was equivalent; 97.7% and 97.2% after one and two years in SCT patients versus 98.3% and 96.9% in CS patients ($p=0.343$).

Conclusions: The SCT-study shows that malignancy is more often present in SCT patients with a risk of malignant transformation increasing with age compared to CS patients in whom malignancy is rare. Risk of malignant recurrence did not differ between groups. Overall survival is equal between groups.

Figure 1. Freedom from Malignancy at Primary Resection



PREDICTING EARLY MOTOR DEVELOPMENT AFTER INFANT SURGERY UNDER GENERAL ANESTHESIA BASED ON INTRAOPERATIVE VITAL FUNCTION MONITORING: A MACHINE LEARNING APPROACH

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Background: Infants with congenital malformations undergoing surgery and anesthesia are at risk for impaired early motor development. This study aims to study the relation between intraoperative vital function parameters and early motor outcome, and to compare linear regression with machine learning support vector regression for outcome prediction.

Methods: Patients with congenital malformations undergoing surgical correction at infant age (0-15 months of age) under anesthesia who participated in our standardized follow-up program were included. Clinical variables and intraoperative vital function parameter time series were extracted from the patients' electronic medical record. Linear regression (LR) and Support Vector Machine regression (SVR) were used to predict early motor outcome. Performance of the models was evaluated and compared using R2 and Root Mean Squared Error (RMSE).

Results: Early motor development outcome in the 85 included patients was impaired compared with normative data (t=-2.393, Cohen's d=-0.271, p=0.019). Reference models including only clinical background variables had significant yet modest predictive value for early motor outcome (LR: R2=0.03 [95%CI: 0.02-0.04]; SVR: 0.12 [95%CI: 0.08-0.15]. Adding static features of intraoperative vital function parameters increased the predictive value significantly (LR: R2=0.05; SVR: R2=0.19). SVR models had significantly higher predictive value (R2s = 0.12-0.19) and lower prediction error (0.78-0.83) as compared to LR models (R2s = 0.03-0.06; prediction errors: 98-1.71).

Conclusions: Our findings suggest that impaired early motor outcome in patients with surgical congenital malformations may be related to clinical characteristics and static features of intraoperative vital function parameters. SVM models had better predictive performance compared to LR models.

DID AGE AT SURGERY INFLUENCE OUTCOME IN PATIENTS WITH HIRSCHSPRUNG DISEASE? A NATIONWIDE COHORT STUDY IN THE NETHERLANDS

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Background: Hirschsprung disease (HD) requires surgical resection of affected bowel, but the current evidence is inconclusive regarding the optimal age for resection. The aim of this study was to assess whether age at resection of the aganglionic segment is a determinant for surgical outcomes.

Methods: A cross-sectional cohort study was done including all consecutive patients with HD between 1957 and 2015, aged eight years or older (n=830), who were treated in one of the six pediatric surgical centers in the Netherlands. Outcome measures were mortality, postoperative complications, ostomy rate and redo surgery rate, retrieved from the medical records. Additionally, constipation and fecal incontinence rate in long term were assessed with the Defecation and Continence Questionnaire (DeFeC and P-DeFeC).

Results: The medical records of 830 patients were reviewed, and 346 of the 619 eligible patients responded to the follow-up questionnaires (56%). There was a small increase in the risk of a permanent stoma (Odds Ratio (OR) 1.01 [95% CI: 1.00 -1.02], p=0.019) and a temporary stoma (OR 1.01, [95% CI: 1.00 -1.01], p=0.022) with increasing age at surgery, regardless of the length of the aganglionic segment and operation technique. Both adjusted and unadjusted for operation technique, length of disease, and temporary stoma, age at surgery was not associated with the probability and the severity of constipation and fecal incontinence in long term.

Conclusions: In this study we found no evidence that the age at surgery influences surgical outcomes, thus no optimal timing for surgery for HD could be determined.

APPENDICITIS AND ITS ASSOCIATED MORBIDITY AND MORTALITY IN INFANTS UP TO THREE MONTHS OF AGE: A SYSTEMATIC REVIEW

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Background: We recently encountered the diagnostic dilemma and treatment considerations of appendicitis in two surviving young infants. Although appendicitis is rare in this population, the previously reported mortality rates are high. We aimed to elucidate the current associated morbidity rate (post-operative complications scored as Clavien-Dindo grade I-IV, fatal outcome not included) and mortality rate of appendicitis in infants ≤ 3 months of age.

Methods: We performed a systematic review according to the PRISMA Statement and a search using Pubmed and Embase (February 8th 2022). Studies published after 1980, including patients ≤ 3 months of age with abdominal manifestations of appendicitis (AA) and those with herniated appendicitis (such as Amyand's hernia) (HA) were considered. Patients were divided into newborns (0-28 days) and infants (29 days-3 months).

Results: In total, 118 articles were included after screening of 2358 articles. Including our own two cases, 183 patients were encompassed. Overall, 70% of the patients had AA and 30% had HA. Eighty percent of patients were newborns and the overall perforation rate was 69%. We found a post-operative complication rate (CD I-IV) of 10% and mortality rate of 10%. All fatal cases concerned patients with AA, and a higher rate of fatal cases was found among newborns, term-born patients, patients with non-perforated appendicitis and patients with significant comorbidity.

Conclusions: Awareness of appendicitis remains critical in infants ≤ 3 months of age due to its associated high morbidity and mortality rates. Several patient characteristics were identified that may be associated with poor outcome.