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Original Research Article

Comparative Study of Various Surgical Procedures in the Treatment of Hirschsprung Disease at Different Age Groups

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Abstract

Background: Hirschsprung disease (HD) is a congenital disorder characterized by the absence of ganglion cells in the distal bowel, leading to functional intestinal obstruction. Early diagnosis and surgical intervention are crucial to prevent complications like enterocolitis and to improve long-term outcomes. The main goal of surgery is to excise the aganglionic bowel and restore gastrointestinal continuity with ganglionated bowel to the anus. Several surgical techniques have been developed, including the Swenson, Soave and Duhamel procedures.

Literature supports the notion that early intervention can reduce the risk of enterocolitis and improve bowel function, but it may also be associated with higher rates of anastomotic complications. Conversely, delayed surgery allows for better nutritional status and growth but may prolong exposure to the risks associated with the aganglionic segment.

A comparative study is warranted to evaluate the efficacy and safety of different procedures across various age groups. Such a study is likely to generate critical evidence regarding optimizing surgical management strategies for HD, tailoring interventions to individual patient needs, and improving long-term outcomes. By analyzing factors such as postoperative complications, bowel function, continence, and quality of life, this research aims to identify the most suitable surgical approaches for different age cohorts.

Aim and Objectives: To study outcomes of Duhamel, Soaves and Primary transanal pull-through in different age groups. This study objective is to assess the outcomes of various surgical procedures, suitable for age, growth outcomes/nutritional status after surgery and complications.

Materials & Methods: This is a hospital-based prospective comparative study on all patients under 12 years with a confirmed histological diagnosis were considered and treated for Hirschsprung's disease in the Department of Pediatric Surgery, King George Hospital, Visakhapatnam. All confirmed cases of Hirschsprung disease that aligned with the inclusion criteria were analysed between May 2023 and April 2025, totaling 41 cases.

Results: In the 41 participants, 7 (17.1%) were neonates, 12 (29.3%) were infants, 15 (36.6%) belong to 1-5 years group and 7 (17.1%) belong to 6-11 years group. Males were 29 (70.7%) and 12 (29.3%) were females. constipation, abdominal distension, delayed meconium was observed in all cases (100%) while bilious vomiting was observed in 8 cases (19.5%). In the study, 75.6% who underwent surgery were below 3 years and 24.4% were above 3 years. Regarding type of aganglionosis, 82.9% had classical, 12.2% had long segment and 4.9% had TCA. 36 children (87.8%) had two stages pl 3 babies (7.3%) had one stage and 2 cases had three stages. Enterocolitis was seen in 4 cases (9.8%). Out of these 4 cases, 1 presented as preop and 3 presented as postop. In the study, DUHAMEL was done in 22 cases (53.7%), SOAVE in 16 cases (39.0%) and TEPT was done in 3 cases (7.3%). Out of 41 participants regarding stoma, colostomy was done in 36 cases (87.8%) and ileostomy was done in 2 cases (4.9%) and no stoma (primary) was done in 3 cases (7.3%). Regarding stoma complications, 4 cases (9.8%) had excoriation, 2 cases (4.9%) had excoriation with diarrhoea, 3 cases (7.3%) had stomal prolapse, 2 cases (4.9%) had non-function of stoma, 1 case (2.4%) had retraction and 29 cases (70.7%) had no stoma related complications. 3 cases (7.3%) underwent colostomy revision For stoma related complications, The complications related to management, 5 cases (12.2%) had surgical site infection, 3 (7.3%) had Spur, 2 (4.9%) had wound dehiscence and 1 case (2.4%) had residual segment. For these complications, 3 (7.3%) underwent Spur excision, 2 (4.9%) had suturing and 1 case (2.4%) had myectomy. Among the study participants, constipation was seen in 3 cases (7.3%), soiling was seen in 4 cases (9.8%) and voluntary bowel movements were seen in 37 cases (90.2%). Among 4 cases who had soiling, one case was grade 1 severity and three cases were grade 2 severity. All the three cases who had constipation were of grade 3 severity. 46.3% cases were under nourished and 53.7% cases were well nourished. Among the study participants, there was a significant

improvement in mean and height between preop and postop (P value <0.05) neonate underwent TEPT. Among 4 cases which had soiling, 1 case was of grade 1 severity, 3 cases were of grade 2 severity and all of these belonged to SOAVE group only. Constipation presented in 3 babies were of grade 3 severity and belonged to DUHAMEL group only. Among the study participants, 50% each in DUHAMEL group and SOAVE group were undernourished. However, none of the cases in TEPT group had undernourishment status. On performing chi square test, this difference was not found to be statistically significant (P value >0.05).

Conclusion: The findings indicate that the age at which we perform definitive surgical procedure is a significant factor in the choice of surgical management. The TEPT procedure was the definitive management for neonates. In contrast, in our study, Duhamel and Soave procedures were the most common choices for infants and older children, with the Duhamel procedure being utilized most frequently in both age groups.

Overall, the study found no statistically significant difference in the incidence of postoperative complications between the Duhamel, Soave, and TEPT groups. The results indicate that all three procedures provide generally favourable outcomes for voluntary bowel movements and constipation, while Duhamel and TEPT procedures may offer superior continence preservation compared to the Soave procedure.

Keywords: Hirschprung's disease, surgical procedures, complications.

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Introduction

Hirschsprung disease (HD) is a congenital disorder characterized by the absence of ganglion cells in the distal bowel, leading to functional intestinal obstruction. HD remains a significant cause of neonatal and infantile bowel obstruction, with an incidence of approximately 1 in 5,000 live births and a higher prevalence in males. [1]

The exact aetiology is not known, but the basic pathophysiology involves the failure of neural crest cells to migrate completely during intestinal development, resulting in an aganglionic segment that lacks peristalsis. This leads to a functional blockage, causing symptoms such as delayed meconium passage, abdominal distension, and chronic constipation. Early diagnosis and surgical intervention are crucial to prevent complications like enterocolitis and to improve long-term outcomes. [2,3]

Plain radiograph shows dilated bowel loops. Initial diagnostic approach for Hirschsprung disease typically involves a contrast enema, which may reveal a characteristic transition zone—marked by a narrowed distal aganglionic segment juxtaposed against a dilated proximal segment with normal innervation. While this radiographic finding is suggestive, definitive diagnosis necessitates histopathological examination of a rectal biopsy. This is a gold standard diagnostic method that confirms the absence of enteric ganglion cells and often identifies hypertrophied nerve fibres. Additionally, increased acetylcholinesterase activity detected through immunohistochemical staining further substantiates the diagnosis.

Over the decades, they have developed several surgical techniques for the definitive management of HD. The main goal of surgery is to excise the aganglionic bowel and restore gastrointestinal continuity with ganglionated bowel to the anus. The definitive treatment for HD is surgical resection of the aganglionic segment followed by anastomosis of the healthy bowel to the anus. Several surgical techniques have been developed, including the Swenson, Soave, and Duhamel procedures. Every method offers distinct benefits and potential complications, and the choice often depends on the surgeon's experience and the patient's specific condition. Modern surgical techniques have further evolved to include laparoscopic-assisted and transanal approaches, that provide lower complication rates, decreased duration of hospitalization, and faster recovery.

Timing of surgery remains a subject of clinical debate. Surgical correction can be performed in the neonatal period or deferred until infancy or early childhood. Neonatal surgery may be characterized by higher technical demands due to fragile tissues and immature physiology, but may reduce the risk of enterocolitis and allow early normalization of bowel habits. Delayed surgery, conversely, allows for improved nutritional status and reduced perioperative risk, though it may prolong exposure to potential complications of the disease. [4,5]

Literature supports the notion that early intervention can reduce the risk of enterocolitis and improve bowel function, but it may also be associated with higher rates of anastomotic complications. Conversely, delayed surgery allows for better nutritional status and growth but may prolong exposure to the risks associated with the aganglionic segment. [3]

Given the variety of surgical techniques and the potential impact of age at surgery on outcomes, a

comparative study is warranted to evaluate the efficacy and safety of different procedures across various age groups. Such a study is likely to generate critical evidence regarding optimizing surgical management strategies for HD, tailoring interventions to individual patient needs, and improving long-term outcomes.

By analyzing factors such as postoperative complications, bowel function, continence, and quality of life, this research aims to identify the most suitable surgical approaches for different age cohorts. The results could guide future clinical decision-making and assist surgeons in making evidence-based decisions regarding the timing and type of surgical intervention for HD patients.

Aim and Objectives

To study outcomes of Duhamel, Soaves and Primary transanal pull-through in different age groups. This study objective is to assess the outcomes of various surgical procedures, suitablefor age, growth outcomes/nutritional status after surgery and complications.

Materials &Methods

This is a hospital-based prospective comparative study on All patients under 12 years with a confirmed histological diagnosis were considered and treated for Hirschsprung's disease in theDepartment of Pediatric Surgery, King George Hospital, Visakhapatnam. All confirmed cases of Hirschsprung disease that aligned with the inclusion criteria were analysed between May 2023 and April 2025, totalling 41 cases.

Ethical Consideration: Prior to enrolling participants, approval was granted by the Institutional Ethics Committee, of the King George Hospital. Parents of the participating pediatric patients were provided with comprehensive information regarding the study, and written informed assent was obtained before inclusion. Confidentiality of the data was maintained, ensuring that participant anonymity was preserved throughout the study.

Data analysis: Upon completion of the data acquisition phase, the collected information was systematically transcribed into a Microsoft Excel Spreadsheet. Following data entry, a detailed statistical analysis was performed using the Statistical Package for the Social Sciences (SPSS) version 25. Categorical variables were summarized using frequencies and percentages, while continuous variables were described using means and standard deviations. The Chi-Square test was applied as appropriate, with a p-value of <0.05 considered statistically significant.

Results

Th age distribution of the disease ,sex distribution, clinical presentation, age at which undergone surgery, type of aganglionosis, type of surgery, complications like enterocolitis , surgical site infections, wound dehiscence, residual spur were studied in this study. Other factors like constipation and nutritional status were also observed. In the 41 participants, 7 (17.1%) were neonates, 12 (29.3%) were infants, 15 (36.6%) belong to 1-5 years group and 7 (17.1%) belong to 6-11 years group. Males were 29 (70.7%) and 12 (29.3%) were females. constipation, abdominal distension, delayed meconium was observed in all cases (100%) while bilious vomiting was observed in 8 cases (19.5%).In the study, 75.6% who underwent surgery were below 3 years and 24.4% were above 3 years. Regarding type of aganglionosis, 82.9% had classical, 12.2% had long segment and 4.9% had TCA. 36 children (87.8%) had two stages pl 3 babies (7.3%) had one stage and 2 cases had three stages. Enterocolitis was seen in 4 cases (9.8%). Out of these 4 cases, 1 presented as preop and 3 presented as postop.

In the study, DUHAMEL was done in 22 cases (53.7%), SOAVE in 16 cases (39.0%) and TEPT was done in 3 cases (7.3%).Out of 41 participants regarding stoma, colostomy was done in 36 cases (87.8%) and ileostomy was done in 2 cases (4.9%) and no stoma (primary) was done in 3 cases (7.3%).

Regarding stoma complications, 4 cases (9.8%) had excoriation, 2 cases (4.9%) had excoriation with diarrhoea, 3 cases (7.3%) had stomal prolapse, 2 cases (4.9%) had non-function of stoma, 1 case (2.4%) had retraction and 29 cases (70.7%) had no stoma related complications. 3 cases (7.3%) underwent colostomy revision For stoma related complications,

The complications related to management, 5 cases (12.2%) had surgical site infection, 3 (7.3%) had Spur, 2 (4.9%) had wound dehiscence and 1 case (2.4%) had residual segment.For these complications, 3 (7.3%) underwent Spur excision, 2 (4.9%) had suturing and 1 case (2.4%) had myectomy.

Among the study participants, constipation was seen in 3 cases (7.3%), soiling was seen in 4 cases (9.8%) and voluntary bowel movements were seen in 37 cases (90.2%). Among 4 cases who had soiling, one case was grade 1 severity and three cases were grade 2 severity. All the three cases who had constipation were of grade 3 severity.46.3% cases were undernourished and 53.7% cases were well nourished. Among the study participants, there was a significant improvement in mean and height

between preop and postop (P value <0.05) neonate underwent TEPT.

Majority of the participants in other age groups like infant, 1-5 years and 6-11 years underwent DUHAMEL. On performing chi square test, this difference was found to be statistically significant (P value <0.05).Among the DUHAMEL group, one case which had residual segment underwent mycetoma. Three cases who had spur underwent spur excision. Two cases which had wound dehiscence underwent suturing.

Among 4 cases which had soiling, 1 case was of grade 1 severity, 3 cases were of grade 2 severity and all of these belonged to SOAVE group only Constipation presented in 3 babies were of grade 3 severity and belonged to DUHAMEL group only.

Among the study participants, 50% each in DUHAMEL group and SOAVE group were undernourished. However, none of the cases in TEPT group had undernourishment status. On performing chi square test, this difference was not found to be statistically significant (P value >0.05).

Discussion

The most prevalent congenital intestinal motility abnormality, Hirschsprung disease (HSCR), affects 1 in 5000 live infants annually. It frequently manifests as a distal bowel obstruction in the newborn stage or as persistent constipation in later life as a result of interrupted or partial neural crest migration, which results in ganglion cell absence in the intestinal myenteric and submucosal plexuses. Surgical procedure can be carried out in two steps (delayed pull-through operation after stoma creation) or in one stage (primary pull-through treatment). The duration of aganglionosis, the length of aganglionosis segment, the existence of enterocolitis, the child's sickness, comorbid diseases, and the surgeon's comfort all influence the surgery choice.

In the current study, 36.6% of the cases are under Child of age (1-5years) which is the highest proportion among all the cases, A significant portion of HD cases are diagnosed in the age range of (1 month to 1 year) i.e, 29.3% with mild or short-segment HD.

The typical time of diagnosis for classical Hirschsprung disease was the neonatal period (0–28 days), and in the present study, it constitutes around 17.1% which is a small proportion. Children in the age group (6-11 years) also presented with a small proportion of cases i.e, 17.1%. HD in older children (6-11 years) is relatively uncommon, and these children have persistent constipation since birth, and sometimes misattributed to dietary issues or behavioural problems. Arafa et al 8conducted the study among 20 patients who were above the age of 3 years, and the age range was 3 to 8 years. Allaberganov et al. [7] conducted a study among children aged 2 months to 11 years, and all of the findings are similar to our study findings.

Hirschsprung disease shows a strong male predominance in the current study, with 70.7% cases.The study conducted byAhmed Nasr et al. [5] also found that 24.7% of female participants received a diagnosis of Hirschsprung disease.

In the current study, delayed passage of meconium beyond 48 hours and constipation were observed in all the cases (100%). Abdominal distension was also seen in all the cases, which has indicated a strong diagnostic relevance. Bilious vomiting was observed in a smaller subset of patients, i.e about 19.5% cases. It is not as common as other symptoms, and it indicates the urgency for surgical evaluation, as it may be a complicated or extensive disease, and the cases require close attention. All the findings align with the classical presentation of HD in neonates and young infants. L.Senthil Kumar et al. [8] conducted a study and found that all the neonates presented with the history of delayed passage of meconium and abdominal distension. Vomiting was present in 40% of the neonates, and the commonest presenting symptoms in infants were abdominal distension and constipation (92.3%). All children above one year of age presented with abdominal distension and chronic constipation, and similar findings were observed in our study.

In the present study, Enterocolitis occurredin 9.8% of the Hirschsprung disease cases, in which 1 case was preoperative, which highlights the need for surgery earlier. 3 cases were postoperative, which reinforces that surgical treatment doesn’t eliminate the risk entirely, similar to the study conducted by V. Kumaran et al. [9] This study population shows that the majority of the cases had the classical recto-sigmoidtype, i.e, 82.9% while a smaller proportion of cases had more extensive forms like long segment and total colonic aganglionosis, i.e, 17.1%. in accordance with the study conducted by K. Jayapal et al. [6] In the present study, the Duhamel procedure was the most commonly performed technique i,e, 53.7% followed by the Soave procedure,which is 39% and 7.3% of patients were operated with TEPT. The surgical procedure was tailored to optimize the outcomes based on the clinical presentation and resources available.

The majority of patients, 29 (70.7%), had no complications following stoma creation. The most common complication was peristomal excoriation, seen in 4 patients (9.8%), followed by excoriation with diarrhoea (ileostomy) in 2 patients (4.9%). Stomal prolapse occurred in 3 patients (7.3%), non-

functioning stoma in 2 patients (4.9%), and stoma retraction in 1 patient (2.4%). Notably, there were no cases of stenosis or herniation reported. These findings indicate thatstoma-related complications were infrequent, and the majority of patients experienced an uneventful postoperative course. Among the complications observed, excoriation and stomal prolapse were the most common, though relatively mild and manageable. The absence of serious complications like stenosis or herniation suggests effective surgical technique and postoperative care in the management of stomas in Hirschsprung disease.

The bowel function outcomes in the current study showed that the majority of patients (37 cases, 90.2%) had voluntary bowel movements, indicating good postoperative bowel control. Soiling was observed in 4 cases (9.8%), of which 1 case was of grade 1 severity (mild, occasional staining) and 3 cases were of grade 2 severity (moderate, frequent episodes but manageable). Constipation was reported in 3 cases (7.3%), with grade 3 severity in the Duhamel procedure, which required spur excision.V. Kumaran et al. [9] conducted a study and found Soiling in 6 patients (27%), constipation in 14% of the patients and postoperative enterocolitis was noted in 2 patients (9%).

Overall, voluntary bowel control was well maintained in most patients, and a small proportion experienced significant postoperative bowel dysfunction with grade 3 constipation, which was surgically corrected later, compared to soiling.

The present study shows that the distribution of age at the time of definitive surgical management for Hirschsprung disease showed significant variation across the different operative procedures. In the neonatal period, definitive management was performed only in a single case, managed by the TEPT procedure, accounting for 100% of neonates in this category. Among infants, the majority underwent the Duhamel procedure (50.0%), followed by the Soave procedure (38.9%), while TEPT was performed in 11.1% of cases. In children aged 1–5 years, the Duhamel procedure continued to be the most frequently utilized (60.0%), followed by Soave (40.0%). Similarly, in children aged 6–11 years, Duhamel accounted for 57.1% of cases, while Soave was performed in 42.9%. Statistical analysis revealed a significant association between age at definitive management and the choice of surgical procedure (Chi-square = 14.834, p = 0.022), indicating that age plays an important role in determining the surgical approach. K. Jayapal et al. [15] study found that in the group of 30 Hirschsprung disease patients, the Duhamel procedure was performed in 15 cases, of which 20% were long-segment disease and 80% were short-segment disease. The Soave procedure was also performed in 15 cases, with 13.3% being long-segment and 86.7% short-segment disease. Statistical analysis (Chi-square = 0.24, p = 0.62) indicated no significant association between the type of definitive surgical procedure and the length of the aganglionic segment. Ahmed Nasr et al. [5] study found that the mean age at surgery for Hirschsprung disease cases in the study was 0.26 years (approximately 3 months) with a standard deviation of ±1.16 years, indicating that most patients underwent definitive surgical management in early infancy, although the wide standard deviation reflects variability in the age at operation, the adjusted odds ratio for the studied variable in Hirschsprung disease cases was 1.00 (95% CI: 0.72–1.39), with a p-value of 0.98. This indicates no association between the variable and the outcome, as the odds ratio is close to 1, the confidence interval includes 1, and the p-value is not statistically significant.

These findings highlight that TEPT was primarily employed in neonates, while Duhamel and Soave procedures were more commonly utilized in infants and older children, reflecting age-related surgical preferences and considerations in the management of Hirschsprung disease in the present study.

In the current study, the data outlines the distribution of postoperative complications in Hirschsprung disease patients managed with Duhamel, Soave, and TEPT procedures, along with statistical analysis results. Complication-free outcomes were most common in the Soave group (46.7%), followed closely by Duhamel (43.3%), while TEPT had the lowest proportion without complications (10%). Residual segment and spur formation were observed exclusively in the Duhamel group. Surgical site infections occurred in both Duhamel (60%) and Soave (40%) procedures, with none reported in TEPT. Wound dehiscence was also seen only in the Duhamel group. Although the Duhamel procedure demonstrated a wider range of complications compared to Soave and TEPT, statistical analysis using the Chi-square test (χ^2 = 6.927, p = 0.545) revealed no significant association between the type of surgical procedure and the occurrence of complications, indicating that the observed differences may be due to chance rather than a true procedural effect.

The present study data shows the assessment of soiling outcomes in Hirschsprung disease patients, based on age at definitive management and type of surgical procedure, shows variation across age groups and operative techniques, though without statistically significant associations. In neonates, only one patient underwent surgery (TEPT), and no soiling was reported (100%). Among infants, none of the patients operated by Duhamel 50% experienced soiling, Soave 38.9%, and TEPT 11.1%, indicating uniformly favourable continence outcomes in this age group. In children aged 1–5

years, soiling was absent in all patients who underwent Duhamel. In Soave, three cases had soiling, and another three cases had no soiling, though the difference was not statistically significant ($\chi^2 = 2.934$, $p = 0.087$). In the 6–11 years group, soiling was absent in all 66.7% patients treated with the Duhamel procedure, and one patient developed soiling following Soave, while 33.3% did not have soiling. This difference was also not significant ($\chi^2 = 0.024$, $p = 0.876$). Collectively, these findings suggest that soiling is rarely observed in neonates and infants following the procedures. However, as the statistical results indicate no significant association, neither age at definitive surgery nor surgical technique appears to be an independent determinant of postoperative soiling in this study.

`The current study data presents the evaluation of constipation in relation to age at definitive management and surgical procedure for Hirschsprung disease patients. The majority achieved satisfactory bowel function, with no statistically significant differences between groups. Among neonates, the patient who underwent definitive surgery with TEPT and did not experience constipation (100%). In infants, all patients across the three surgical techniques were free of constipation, including 50% in the Duhamel group, 38.9% in the Soave group, and 11.1% in the TEPT group.

In children aged 1–5 years, constipation was reported in only one case managed with the Duhamel procedure, while the remaining patients (57.1% Duhamel and 42.9% Soave) had no complaints, with statistical analysis showing no significant difference ($\chi^2 = 0.714$, $p = 0.398$). Similarly, in children aged 6–11 years, constipation was present in two patients following the Duhamel procedure, while all patients treated with Soave 60% remained free of constipation; the difference again was not statistically significant ($\chi^2 = 0.365$, $p = 0.546$).

Overall, these findings suggest that constipation after definitive management of Hirschsprung disease is uncommon across all age groups, occurring only sporadically in older children following the Duhamel procedure, and that neither age at surgery nor the type of procedure had a significant impact on postoperative constipation.

In the current study, the evaluation of nutritional status in relation to definitive surgical management of Hirschsprung disease patients showed a varied distribution across the different procedures, though no statistically significant differences were observed. Among patients who underwent the Duhamel procedure (n = 22), half were well nourished (11 patients, 50.0%), while the remaining half were undernourished (11 patients, 50.0%). A similar pattern was observed in the Soave group, where 8 patients (50.0%) were well nourished and 8 patients (50.0%) were undernourished. In contrast, all patients who underwent transanal endorectal pull-through (TEPT) (n = 3) were found to be well nourished (100.0%), with no cases of undernutrition. While the TEPT group showed a noticeable difference, analysis did not reveal a significant correlation between the type of definitive procedure and nutritional status (Chi-square = 2.795, $p = 0.247$). The results indicate that, although nutritional outcomes seem more favorable following TEPT, the overall influence of surgical technique on nutritional status in Hirschsprung disease patients is not statistically significant.

Limitations

The primary limitation of this study is its modest sample size of 41 patients, the short-term follow-up period (May 2023 to April 2025) means the study could not assess long-term functional and nutritional outcomes of the procedures.

Conclusion

The findings indicate that the age at which we perform definitive surgical procedure is a significant factor in the choice of surgical management. The TEPT procedure was the definitive management for neonates. In contrast, in our study, Duhamel and Soave procedures were the most common choices for infants and older children, with the Duhamel procedure being utilized most frequently in both age groups.

Overall, the study found no statistically significant difference in the incidence of postoperative complications between the Duhamel, Soave, and TEPT groups. However, a significant association was identified between the type of procedure and the occurrence of soiling. A postoperative complication, like soiling, is observed in 25% of patients who underwent the Soave procedure, but was not observed in patients treated with the Duhamel or TEPT procedures.

The results indicate that all three procedures provide generally favourable outcomes for voluntary bowel movements and constipation, while Duhamel and TEPT procedures may offer superior continence preservation compared to the Soave procedure. The findings offer insights that can support clinical guidelines and assist surgeons in making evidence-based decisions on the most effective surgical approach, considering both patient's age and the risk of specific postoperative complications like soiling.

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