"QUALITY OF LIFE OF PATIENTS WITH HIRSCHSPRUNG'S DISEASE AND ANORECTAL MALFORMATION AFTER PULL THROUGH SURGERY": OBSERVATIONAL STUDY"

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With reference to the above, we wish to inform you that your proposed research project titled **"QUALITY OF LIFE OF PATIENTS WITH HIRSCHSPRUNG'S DISEASE AND ANORECTAL MALFORMATION AFTER PULL THROUGH SURGERY: OBSERVATIONAL STUDY FOR PERIOD OF 1 YEAR AT KAHER'S DR. PRABHAKAR KORE CHARITABLE HOSPITAL AND MEDICAL RESEARCH CENTRE "**, is ethical and justifiable. The proposed research project has been cleared by the JNMC Institutional Ethics Committee on Human Subjects Research.

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LIST OF ABBREVIATIONS

KAHER	KLE Academy of Higher Education and Research
KLES	Karnataka Lingayat Education Society
HD	Hirschsprung's Disease
ARM	Anorectal malformations
QOL	Quality of life
HAQL	Hirschsprung's disease/anorectal malformation quality of life questionnaire
PSARP	Posterior sagittal anorectoplasty
ASARP	Anterior sagittal anorectoplasty
NOS	Nitric oxide synthetase
HAEC	Hirschsprung's-associated enterocolitis
FTB	Full-thickness rectal biopsy
CE	contrast enema
RSB	rectal suction biopsy
LATEP	laparoscopic-assisted trans anal endorectal pull-through
TEP	trans anal endorectal pull-through
HRQoL	health-related quality of life
VACTERL	vertebral, anorectal, cardiac, tracheoesophageal, radial, and limb anomalies
LAARP	laparoscopic-assisted anorectal pull-through
BFS	bowel function score
SD	Standard deviation

ABSTRACT

BACKGROUND:

Hirschsprung's disease and Anorectal malformations are the most common conditions for which pediatric surgery outpatients come to the hospital. Most common procedures performed for Hirschsprung's disease are Swenson procedure, Duhamel procedure, Soave procedure where aganglionic segment is removed and anastomosis is done. Posterior sagittal anorectoplasty/Anterior sagittal anorectoplasty is the most common procedures performed for Anorectal malformations. But irrespective of the type of surgical procedure patient undergone for the anatomical correction quality of life of the patient post-surgery is still unclear. As post operatively these children experience wide range of complications such as fecal incontinence, constipation, enterocolitis etc., A disease-specific tool measuring the quality of life (QoL) of children and adolescents with fecal incontinence was developed in the Netherlands and is called the Hirschsprung's Disease/Anorectal Malformation Quality of life Questionnaire (HAQL) which helped to determine the quality of life of the subjectively.

AIM:

To study the midterm quality of life in the patients of Hirschsprung's disease and Anorectal malformation after pull through surgery.

METHODOLOGY:

A Hospital based one-year observational study was conducted in the Department of Pediatric surgery, KLE'S Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi from 1st January 2021 to 31st December 2021. Patients who

were attended to the outpatient and inpatient department of pediatric surgery for follow-up after minimum of 6 months of pull through surgery for ARM and HD and Age matched patients who are attending the pediatrics outpatient department who present with non-gastrointestinal symptoms were included as controls in the study where demographic characteristics and presenting symptoms, the principal investigator assessed their Quality of Life using HSCR/Anorectal malformation quality of life questionnaire (HAQL).

RESULTS:

A total of 35 cases and 16 controls where 60% of the cases are male and 40% are females with history of consanguinity in 25.71% patients in the cases were assessed. In the cases sample it was subdivided into Hirschsprung's disease(n=16) and anorectal malformations(n=19). In the comparative analysis between Hirschsprung's disease and controls where there is significant difference in the laxative diet, presence of diarrhea, fecal continence, physical symptoms and overall HAQL scores. When comparative analysis done between Duhamel's and soave's procedure there is no significant difference between the both groups in parameters and overall quality of life.

When comparative analysis done between ARM and controls there is significant difference in the laxative diet, presence of diarrhea, presence of constipation, fecal continence and physical symptoms. But there is not significant difference in comparison between high and intermediate arm.

CONCLUSION:

Here by using modified Hirschsprung's disease/Anorectal Malformation Quality of Life questionnaire (HAQL) provides us an opportunity to access these children with respective intermediate outcomes, regularity of bowel habits, parental acceptance, and the problems faced by children and parents after surgery. During regular follow-up appointments, merely noting the bowel habits is insufficient. It is crucial to incorporate the idea of QOL.

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INTRODUCTION

The most prevalent congenital colorectal anomalies are Hirschsprung's disease (HD) and anorectal malformation (ARM). Alive births account for 1.0 to 1.5 in 5,000 cases of ARM and 1.0 in 5,000 cases of HD, respectively. [1–3]

Clinical manifestations of anorectal malformations (ARMs) span a broad spectrum. These abnormalities range from simple, readily treatable problems with a good functional outlook to complex, challenging to cure disorders[4]. Faecal incontinence and constipation continue to be serious postoperative problems that hinder social and psychological development in individuals with high imperforate anus, despite surgical advancements[5]. Compared to their counterparts, children with ARMs have much lower gastrointestinal function. The severity of the ARM correlates with the level of functional impairment, and some patients with high ARM may experience late functional sequelae such as faecal incontinence and/or constipation, which may frequently persist into adulthood[6,7]. Infants with HD have functional blockage because of absence of ganglion cells in the distal gut that extend over a range of intestinal lengths proximally[8]. HD is a complex hereditary condition.

Hirschsprung's disease is an intestinal motility condition that is indicated by the absence of myenteric (Auerbach plexus) or submucosal ganglion cells (Meissner plexus) in the distal portion of the large intestine. Functional intestinal obstruction is one problem that might result from the dysmotility of the afflicted segment[9]. One of the most often utilized therapy modalities for kids is surgery[10,11].

Many theories have been put forth in an attempt to explain its pathogenesis, which is not fully known. The most widely recognized theory about the aetiology of HD is that there is a malfunction in neuroblasts deriving from the neural crest's migration along the craniocaudal axis, a process that takes place between the fourth

and twelfth weeks of pregnancy[12]. The rectosigmoid area is where the short-segment HD is most frequently found. The entire colon may be impacted by long-segment HD[13].

Affected people have some genetic defects in about 50% of cases[14]. Boys are more likely than girls to develop the condition[15]. The condition might present clinically as intestinal blockage in an infant or as gradual persistent constipation as people age. In the early years, malnutrition, intestinal motility issues, and growing abdominal distension affect about 80% of patients[15]. Clinical evaluation, radiographic findings, and histological analysis of biopsy samples all contribute to the diagnosis[15].

Surgical therapy aims to remove the aganglionic bowel and reconstruct the digestive system by bringing the normally innervated colon down to the anus and maintaining normal sphincter function.[17]. According to Swenson and Bill[18], Duhamel[19], or Soave, this has traditionally been done in three stages with diverting colostomy before reconstructive surgery[20].

In the 1980s, the surgical technique evolved gradually from a three-stage pull-through to a one-stage pull-through, and in the most recent ten years, it has been replaced by a minimally invasive technique that uses laparoscopic pull-through[21] and trans anal anorectal pull-through[22] without laparotomy.

The most frequent late side effects following the pull-through procedure in children with HD are chronic obstructive symptoms, incontinence, and enterocolitis[17]. The majorities of long-term follow-up studies are restricted to 5 to 15 years after surgery and show that gastrointestinal symptoms have improved by the time the kids reach adolescence[23,24]. Additionally, patients with HD have seen an improvement in their overall quality of life (QoL) up until adolescence[25,26]. It is unknown if people with HD will continue to see this improvement[27].

Functional results are thought to get better as patients age, especially after they hit adolescence[21,22]. The long-term effects of HD in adulthood, however, are not at all satisfying, according to other studies[18,19]. Defecation issues did, in fact, get worse after the individuals reached maturity, according to one study[18].

Constipation and faecal incontinence are examples of persistent defecation issues that may negatively affect QoL[16,28]. Generic QoL and health-related QoL are generally distinguished from one another, with the latter emphasizing areas of life that are directly impacted by an individual's health.

The association between functional complaints and QoL in HD patients has been investigated in the past, although these investigations tended to employ health-related QoL questionnaires and very rarely generic QoL questionnaires[29]. Furthermore, it's still not apparent how functional outcomes affect quality of life once a person enters adolescent stage and adulthood stage[29].

There are now several clinical grading methods in use to evaluate the functional and stools of children with ARM[30]. For ARM patients, the idea of QoL has also been investigated. It is important to achieve functional continence as a prerequisite for social adjustment and acceptability, according to certain authors who have noticed the socially devastating effects of stooling and functional problems[31,32].

The Hirschsprung's disease/anorectal malformation quality of life questionnaire (HAQL) is acknowledged as valid and effective for evaluating the QoL of patients after definitive surgery in a wide range of languages.[33-25].

With these in view this study was conducted to assess the midterm quality of life in the patients of Hirschsprung's disease and Anorectal malformation after pull through surgery.

OBJECTIVES

- To study the midterm quality of life in the patients of Hirschsprung's disease and Anorectal malformation after pull through surgery.

REVIEW OF LITERATURE

ANORECTAL MALFORMATIONS

Anorectal malformations (ARMs), with an estimated prevalence ranging between 1 in 2000 and 1 in 5000 live births, are among the more frequent congenital defects encountered in pediatric surgery[36]. Rare is the antenatal diagnosis of an isolated disease of ARM. The majority of instances are discovered in the early neonatal stage. From modest anomalies with simple therapy of a perineal fistula to high abnormalities with sophisticated management, there is a wide range of presentation.

Etiology

Such abnormalities have an unknown origin that is probably complex. Families seem to associate at a low rate, but some seem to have an autosomal dominant inheritance pattern with a higher prevalence to 1 in 100—than others. SHH, EN2, and HLXB9 are three important loci on chromosome 7q39 that are linked to the development of ARM. Several HLXB9 mutations were found in multiple studies to be related to ARM[37]. A select few syndromes, including Townes-Broks, Currarino's, and Pallister-Hall syndromes, have an autosomal dominant type of inheritance[38].

Classification

It has always been necessary to categorize these anomalies in order to choose management and forecast results. The terminal bowel's relationship to the levator ani or pelvic floor was the basis for the early classification of ARM[39]. The level of the cessation of rectal descent and the patient's sex were used to classify by pediatric surgeons meet at Wingspread[40]. This classification was well-liked and was used since a long time.

Level of anomaly	Male	Female
High	Anorectal agenesis Rectovesical fistula Without fistula Rectal atresia	Anorectal agenesis Rectovaginal fistula Without fistula Rectal atresia
Intermediate	Rectourethral fistula Anal agenesis without fistula	Rectovestibular fistula Rectovaginal fistula Anal agenesis without fistula
Low	Anocutaneous (perineal) fistula Anal stenosis (perineal) fistula	Anovestibular (perineal) fistula Anocutaneous Anal stenosis
Miscellaneous	Rare malformations	Persistent cloacal anomaly Rare malformations

Figure 1: Wingspread classification (1984)

It was discovered, following the development of the posterior-sagittal technique by Pea et al., that the location of the fistula had a significant impact on the prognosis of these patients[41]. On the basis of the presence and location of the fistula, Pena developed a classification in 1995 as a consequence of his experience with the posterior sagittal anorectoplasty (PSARP) [42].

Males	Female
Perineal fistula	Perineal fistula
Rectourethral fistula Bulbar Prostatic	Vestibular fistula
Rectovesical fistula	Persistent cloaca <3 cm common channel >3 cm common channel
Imperforate anus without fistula	Imperforate anus without fistula
Rectal atresia	Rectal atresia

Figure 2: Pena Classification (1995)

Later, the Krickenbeck group developed its classification in an effort to standardize the process for evaluating the results of patients with ARM[43]. Wingspread and Pena's categorization criteria were incorporated into the Krickenbeck classification scheme. A diagnostic category, a surgical technique category, and a category describing functional outcome criteria made up its three separate components.

Major clinical groups	Rare/regional variants
Perineal (cutaneous) fistula	Pouch colon
Rectourethral fistula	Rectal atresia/stenosis
Bulbar	Rectovaginal fistula
Prostatic	H-type fistula
Rectovesical fistula	Others
Vestibular fistula	
No fistula	
Anal stenosis	

Figure 3: Krickenbeck classification (2005)

Management

A perineal examination must be done on new-borns when a practitioner suspects anorectal abnormalities. Because significant intraluminal pressure is necessary for a meconium to be forced through a fistula orifice, it is crucial to wait until the patient is 24 hours old before making a decision about the colostomy or the primary surgery. If radiological investigations are performed before 24 hours, they are more likely to reveal the presence of a very high rectum and produce a false diagnosis because a significant amount of intraluminal pressure is needed to overcome the muscle tone of the sphincter that surrounds the lower part of the rectum. Baby should receive fluid resuscitation, antibiotics, and intestinal decompression to reduce risk

of aspiration throughout the first 24 hours. By employing an echocardiography, plain radiography of the lumbar spine, sacral and abdominal ultrasound, the clinician should use these hours to thoroughly rule out the presence of related defects such as cardiovascular malformations, sacral anomalies, and urological abnormalities.

If a perineal fistula is noticed during a perineal examination suggest a low arm therefore anoplasty is indicated. If the rectal gas is below the coccyx without any associated problems, PSARP/ASARP with or without a colostomy is indicated. If rectal gas is above the coccyx and there are accompanying abnormalities, a colostomy is suggested.

Posterior Sagittal Anorectoplasty

Surgery starts with the patient being placed in prone position with slightly raised pelvis in Trendelenburg position. External Sphincter is mapped by using the electrical stimulator pre and perioperatively. Depending on how much exposure is required to properly treat the particular deformity, the incision's length varies.

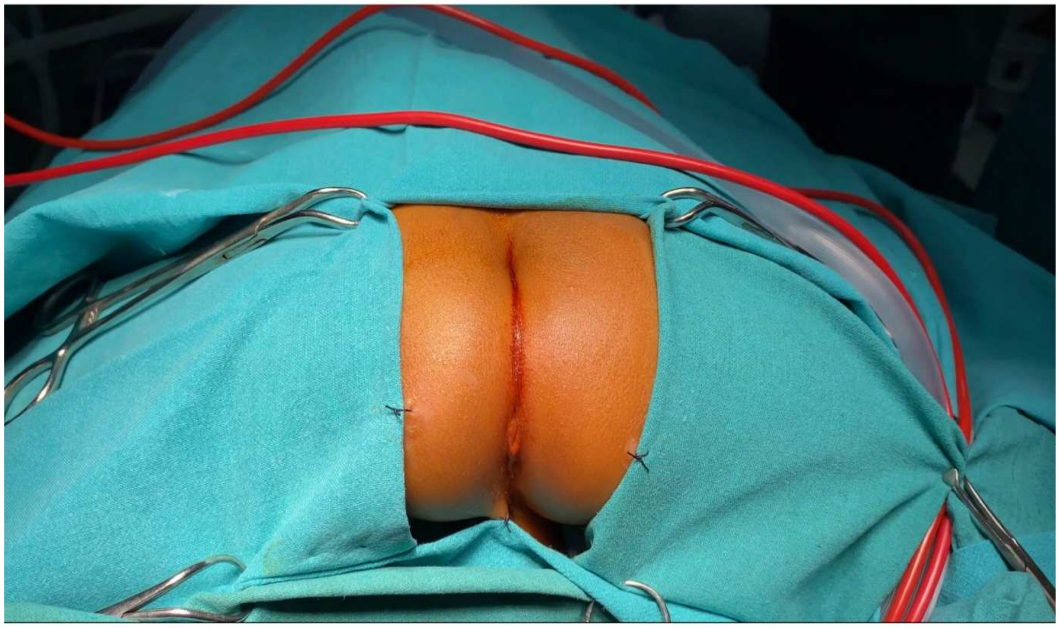


Figure:4 Showing absence of the anal opening preoperatively.

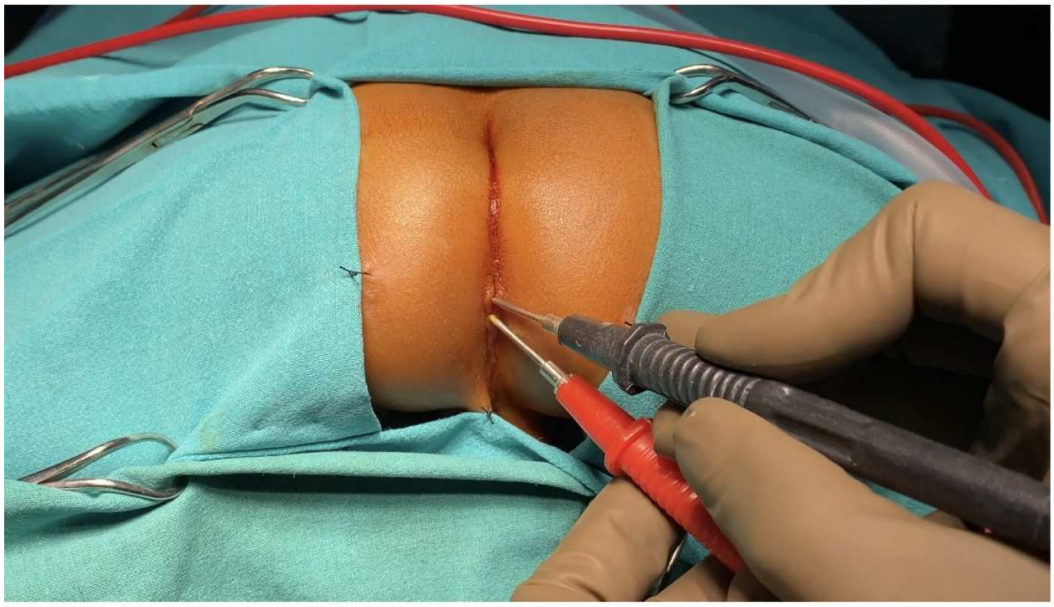


Figure: 5 Sphincter is mapped by using the electrical stimulator Intra operatively.

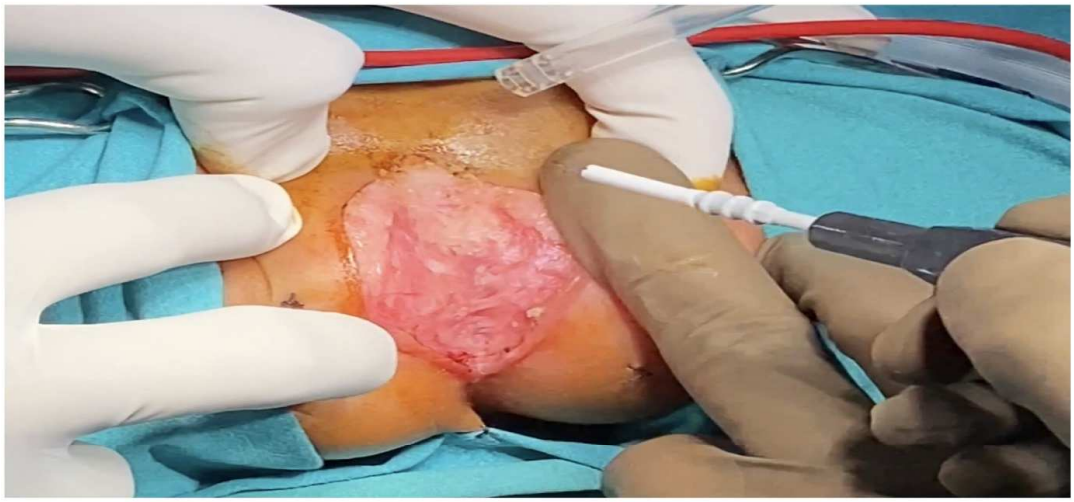


Figure:6 Parasagittal fibres, muscular complex, and levator muscles are separated in the midline by the incision, which also involves the skin and subcutaneous tissue.

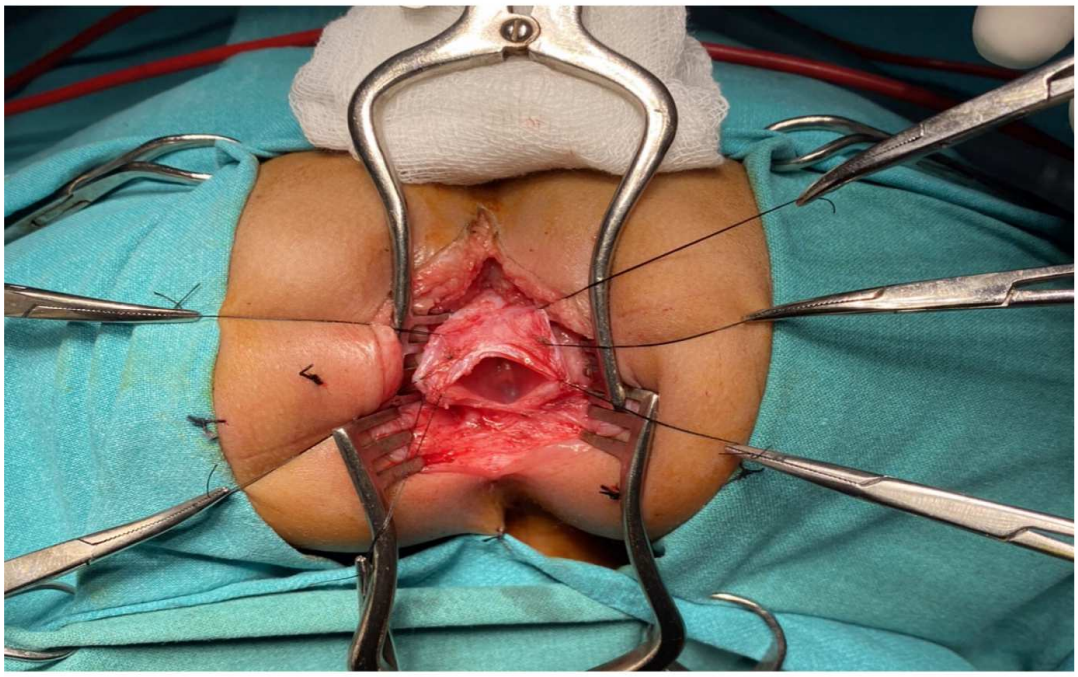


Figure:7 The rectum is instantly noticeable and several silk sutures are applied to the rectum to apply uniform traction and aid the safe separation from surrounding structures.

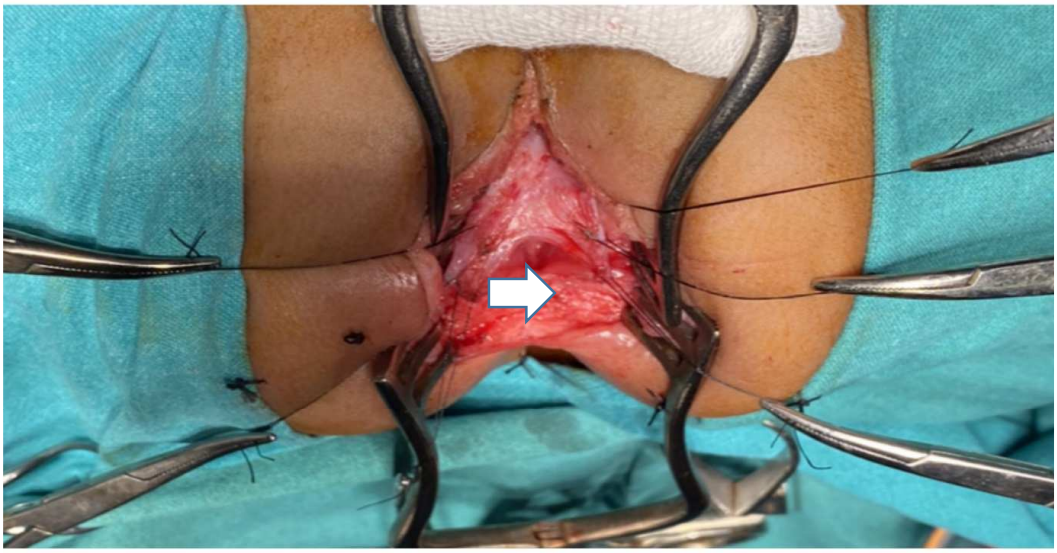


Figure:8 Rectobular urethral fistula being dissected off rectum with submucosal dissection. Arrow showing Rectobular urethral fistula.

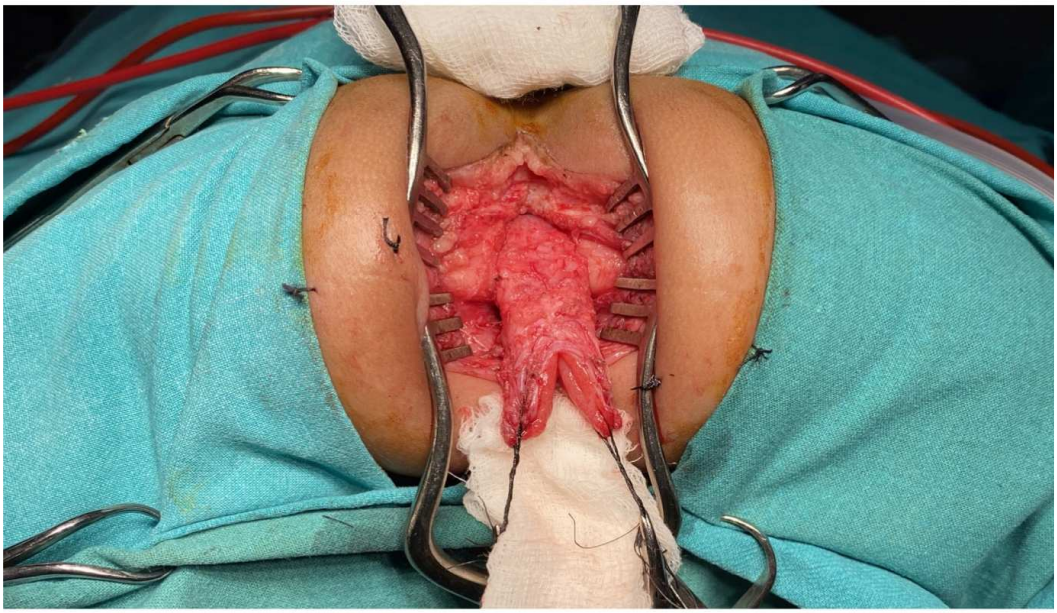


Figure 9:Rectum being mobilised from surrounding structures.

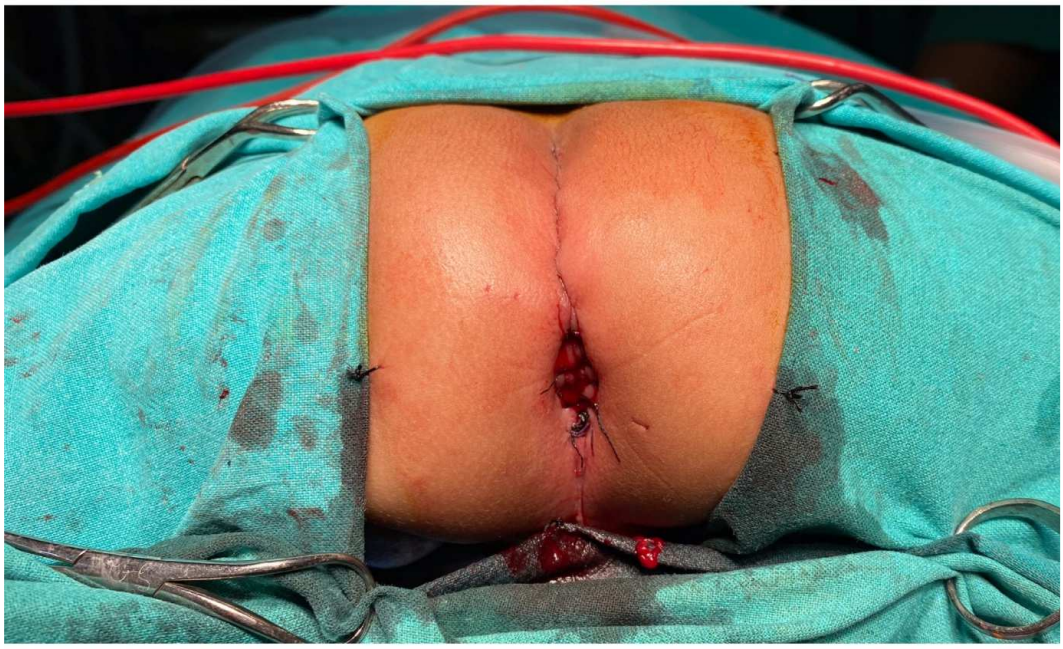


Figure:10 Completed PSARP

The parasagittal fibres, muscular complex, and levator muscles are separated in the midline by the incision, which also involves the skin and subcutaneous tissue. Only the parasagittal fibres and the midline muscle complex are separated by the incision. Levator muscle is not visible in this dissection. The crucial phase is the separation of the rectum from the urogenital tissues after the sphincter mechanism has been split. The rectum is instantly noticeable in perineal and vestibular fistulas, and to provide uniform traction and facilitate the rectum's safe separation from surrounding structures, numerous silk sutures are placed on it. Even in these less severe anomalies, care must be taken to completely separate the rectum from its anterior attachments to the urethra or vagina. The anoplasty will be under tension if this is not done, which leads to numerous unsuccessful repairs. 7 The rectum is positioned in the preoperatively marked location which has been precisely mapped using an electrical

stimulator indicating the centre of the muscle complex. To fix the perineal body, interrupted long-term absorbable sutures are employed. The posterior rectal wall is secured by suturing to the posterior margin of the muscle complex. Because it surrounds the rectum, thus preventing rectal prolapse. 26 The repair is subsequently finished by performing an anoplasty, closing the posterior incision, and reapproximating the parasagittal fibres.

Anterior Sagittal Anorectoplasty

Anal dimples were used to pinpoint the location of the new anus, and electrical muscle stimulation was also used to confirm it. After that, a short-term Vicryl 5/0 suture was placed here.

A raquet incision was carried out extending from the fistula to the centre of muscle complex. The incision divided the vertical as well as the circular muscle fibres. Puborectalis is not divided as incision didn't extended to it. At the mucocutaneous junction of the anal orifice, circumferential 5/0 silk stay sutures were then applied. By sealing the anus with sterile gauze, the field was protected against faeces soiling. Circumferential incision is made around the anal opening to facilitate the separation of rectal wall from the posterior vaginal wall by using blunt or sharp dissection, beginning at the posterolateral points and ending at the anterior point, where the common wall was carefully divided into two walls until complete separation was achieved. Rectal pouch should be freed from the surrounding structure's till it reaches the native anal site without tension. Using 5/0 polyglactin sutures, the rectum was positioned in the centre of the muscle complex and secured.

The perineal body was repaired from the inside out, as was the posterior fourchette. To mimic a natural perineum, the perineal skin was closed.

HIRSCHSPRUNG DISEASE

Harald Hirschsprung first correctly identified Hirschsprung disease (HD) in older children in 1886. Until the distal aganglionosis was discovered in the late 1940s, the pathogenesis remained a mystery for many years[44]. In the years that followed, it was understood that a significant element in the progression of the disease was the colon's failure to empty due to functional blockage. Swenson and Bill, Duhamel, and Soave created a number of operative techniques for the surgical treatment of HD, typically comprising two or three steps. Single-stage pull-through was described by So HB et al[45]., has since become more popular. Although surgical care of HD has significantly improved recently, the fundamental pathophysiology is still not fully understood.

Etiology

The basic etiology of HD is anomalies at the cellular and molecular levels throughout the ENS's formation and the migration of neural crest cells into the developing gastro intestinal tracts[46]. By 5 weeks of gestation, in the esophagus of the growing fetus neural crest-derived neuroblasts start to form. From 5 to 12 weeks of gestation, these cells move into the rest of the developing intestine in a craniocaudal manner.[47]Given the wide variety of potential ENS development abnormalities and the various timeframes in which the migration of neural crest-derived cells can be halted, the HD phenotype is diverse[47,48]. A long section of aganglionosis results from early migration arrest in the growing foetus. Other elements have also been proposed to have a role in HD development, including altered extracellular matrix elements, autoimmune destruction of ganglion cells, neurotrophic factor anomalies, and neural cell adhesion molecules[47].

Genetic factors

Evidence of underlying genetic causes for HD can be seen in the higher incidence of HD in siblings of HD patients, the mismatched sex ratio, the association of HD with other chromosomal abnormalities and congenital disorders[49,50]. The occurrence of HD is attributed to mutations in 10 separate genes, according to genetic studies[50]. RET gene, EDNRB gene, and END3 gene mutations are among the more prevalent ones[49,50]. The RET protooncogene has been connected with more than 20 distinct mutations, and some variants in this gene are linked to specific HD symptoms[51].

Even though HD is a unique trait, 5–32% of HD children have congenital defects and related disorders. Trisomy 21 related with HD has been found in about 7% of HD youngsters, according to reports[49,50]. The most frequent disorders among the organ systems are gastrointestinal, followed by nervous system and genitourinary[49]. The penetrance of HD varies based on the sex of the affected person. According to recent studies, proband's sibling has a 4% chance of developing HD again, which equals a relative risk of 200[50,52].

Pathophysiology

The underlying pathophysiological aspect of HD is functional blockage, which is brought on by a constricted colon and prevents peristaltic waves from spreading because parasympathetic intrinsic ganglion cells are not present[53]. Despite much investigation, the reason for the tonic contraction of the aganglionic gut is still not fully understood. Nitric oxide synthetase (NOS), aganglionosis, cholinergic hyperinnervation, anomalies of the interstitial cells of Cajal, and defects in nerve

distribution have all been attributed to HD's aetiology.[54], But a thorough understanding of the causes of the anomalies seen in HD is still unclear.

Classification

Often these patients can be classified as having rectosigmoid HD, long-segment HD, or total colonic aganglionosis depending on how far aganglionosis extends proximally from the internal anal sphincter.[55].

Types of HSCR	Typical level of aganglionosis	Frequency (%)
Rectosigmoid	Sigmoid	74-80
Long-segment	Splenic flexure or transverse colon	12-22
Total colonic aganglionosis	Terminal ileum	4-13

Figure 11: Types of HD

Clinical features

A neonate with HD often presents as a full-term infant with a meconium passage delay[56]. In the first 24-48 hours after birth, nearly all healthy full-term new-borns pass meconium[57]. However, between 60 and 90 percent of kids with HD are unable to pass meconium during that time[58]. Any youngster who has trouble scrounging up during the new born stage should also be suspected of having HD.

Digital rectal examination can sometimes identify a tight anus and relieve an acute intestinal obstruction by allowing meconium to pass through. In 63-91% of new-borns with HD, the abdomen is distended, and bilious vomiting occurs in 19-

37% of kids with HD[59]. Between 5%[60] and 44%[61] of kids may have Hirschsprung-associated enterocolitis (HAEC) at presentation.

Emergence of severe unpleasant-smelling diarrhoea, pyrexia, and abdominal distension indicates signs of HAEC, which if left untreated might progress to a toxic megacolon which could be fatal. To reduce the risk of death from HAEC, it is critical to diagnose the condition quickly and Rectal irrigation, antibiotics, and intravenous fluids should be used to treat it. Children with delayed HD diagnoses may experience HAEC more frequently, which emphasizes the value of an early diagnosis.

Diagnosis

When a neonate exhibits the aforementioned clinical appearance, HD should be suspected. Full-thickness rectal biopsy (FTB), contrast enema (CE), anorectal manometry, and rectal suction biopsy (RSB) are among the tests that can be used to diagnose HD. Initial test for diagnosing HD varies between different medical facilities. The range of CE, ARM, and RSB sensitivity and specificity reported in studies including neonates in comparison to the gold standard of FTB[62].

Test	Sensitivity (%)	Specificity (%)
Contrast enema (CE)	65-80	66-100
Anorectal manometry (ARM)	75-100	85-97
Rectal suction biopsy (RSB–AChE)*	91-100	97-100
Rectal suction biopsy (RSB–H&E)†	97-100	99-100

*RSB–AChE indicates studies using acetyl cholinesterase staining for RSB.

†RSB–H&E indicates studies using hematoxylin and eosin for staining for RSB.

Figure 12: Diagnosis of HD with Sensitivity /Specificity of different investigations

Management

Plans for surgical management are created if the diagnosis is confirmed. When a neonate has HAEC, the enterocolitis is initially treated with rectal irrigation, aggressive resuscitation, and intravenous antibiotics.

A two or three stage operational repair is generally carried out. After performing "levelling" colonic biopsies to gauge the severity of aganglionosis, the first stage is a faecal diversion through stoma creation. In the second stage aganglionic bowel Resection and coloanal anastomosis are performed, which is carried out later, often between 3 months and 1 year of age. Either during this operation or a third-stage procedure, the previous stoma is closed. For the administration of HD, a variety of operations have been discussed. The most often done operations are the rectosigmoidectomy described by Swenson and Bill, the retrorectal-transanal technique described by Duhamel, and the endorectal operation described by Soave. Laparoscopic techniques have also been used to carry out these surgeries[63,64].

Duhamel Procedure

Preoperative bowel preparation was done. The extent of the aganglionosis was noted preoperatively by using serial seromuscular biopsy from rectum, recto sigmoid, 5cms from the rectosigmoid and 10cms from the rectosigmoid.

Anteriorly peritoneal reflection and posteriorly pelvic floor are the limits for the dissection. Proximal bowel Dissection was done till the ganglionic segment of the bowel which was identified form the biopsy. Rectum was ligated at the peritoneal reflection and the divided. Transverse incision was made 0.5cms above the dentate line retro rectally and posterior rectal space was entered to meet the space dissected

from above. Ganglionic normal colon was pulled through and anastomosed circumferentially with the created opening in the posterior rectum.

Anteriorly aganglionic and posterior ganglionic segment are anastomosed using an EndoGIA (blue cartridge 3.5cm long) under visual control. Usually one cartridge needed to be fired, and occasionally two cartridges are needed. Finally, the upper rectum was closed with a running 3 · 0 Vicryl suture. Outcome measures included postoperative complications, hospital stay, and long-term outcome such as constipation, enterocolitis, faecal incontinence, enuresis, stenosis, and adhesive obstructing bowel.

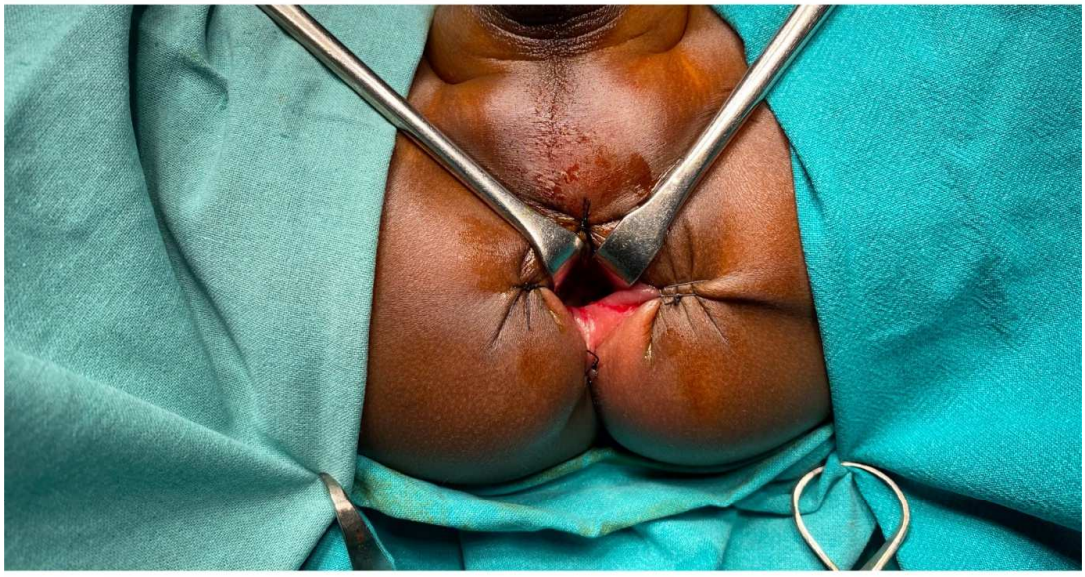


Figure:13 Position for full thickness rectal biopsy, with adequate retracting sutures and langenbeck retractor Insitu

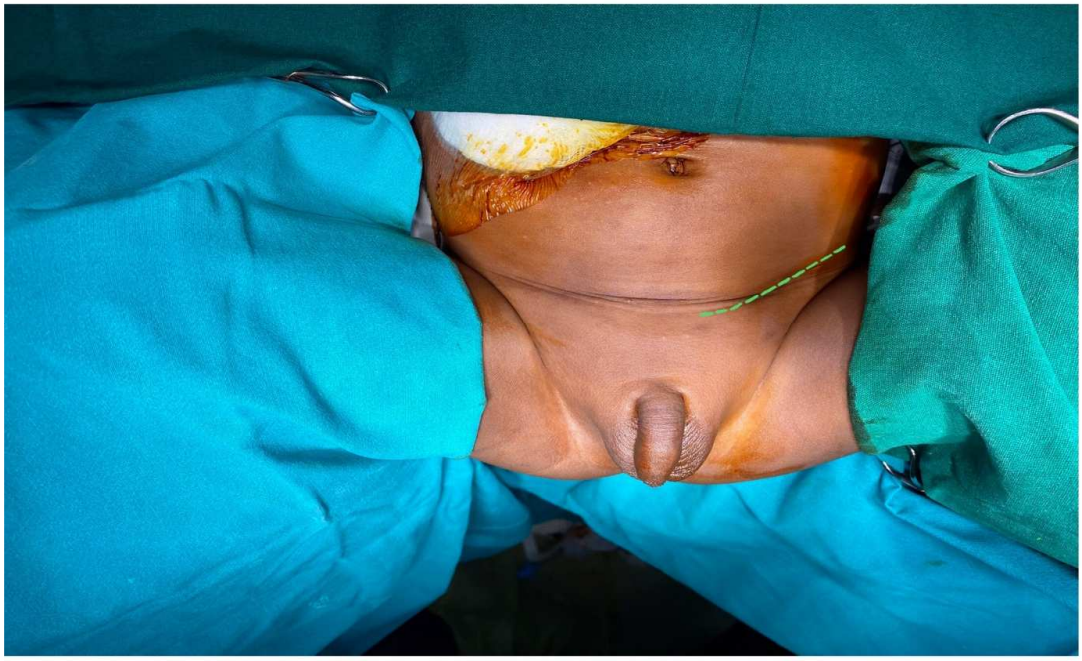


Figure:14 left iliac fossa skin crease incision

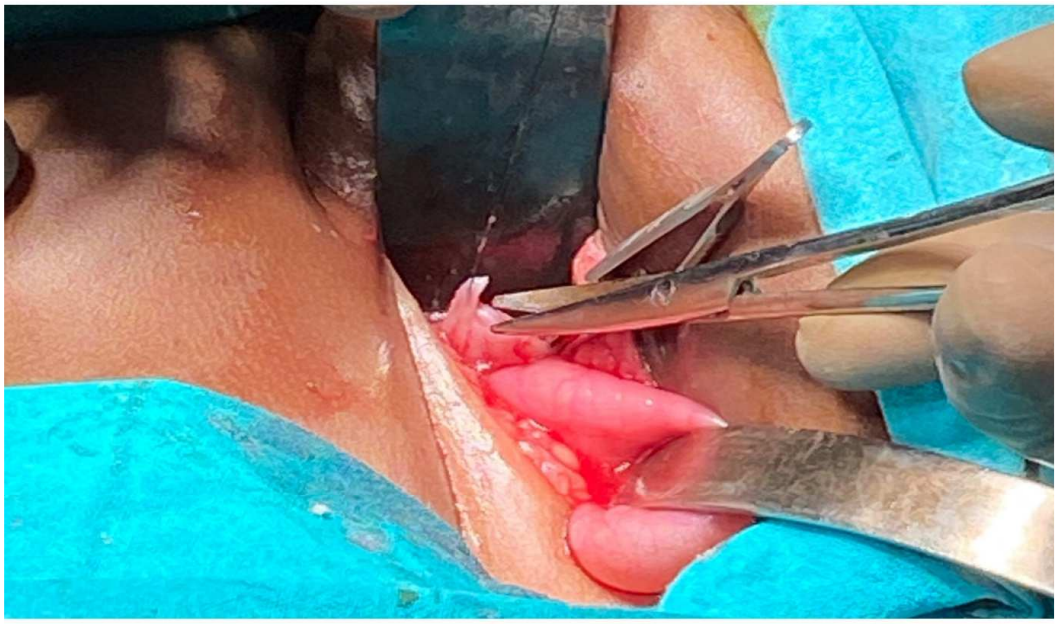


Figure:15 seromuscular (extramucosal) colonic biopsy being done

Figure:16 biopsy samples being transported in formalin medium

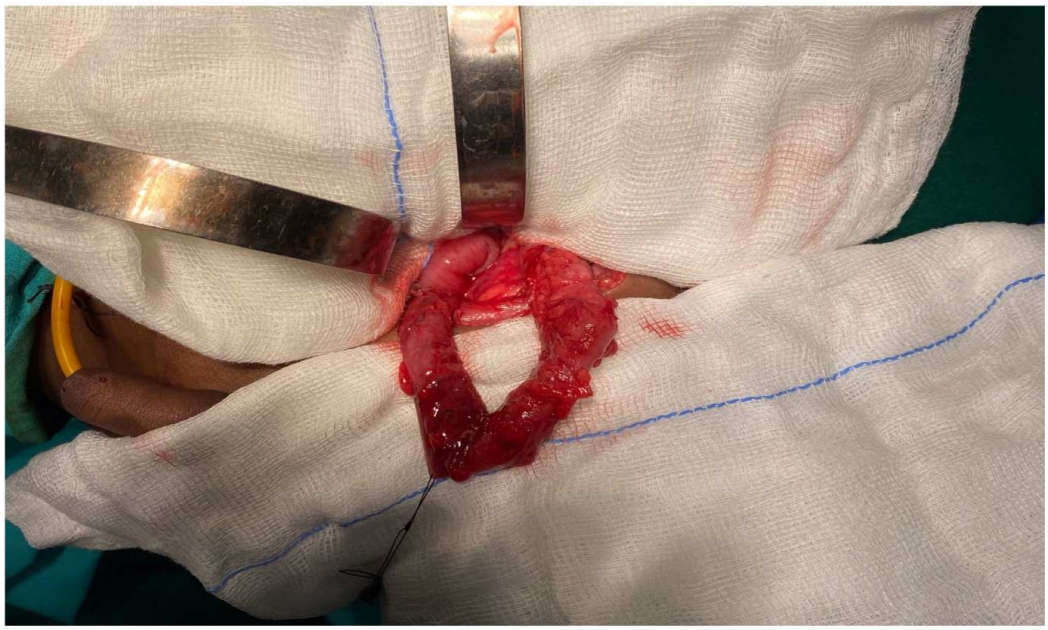


Figure:17 aganglionic segment being resected

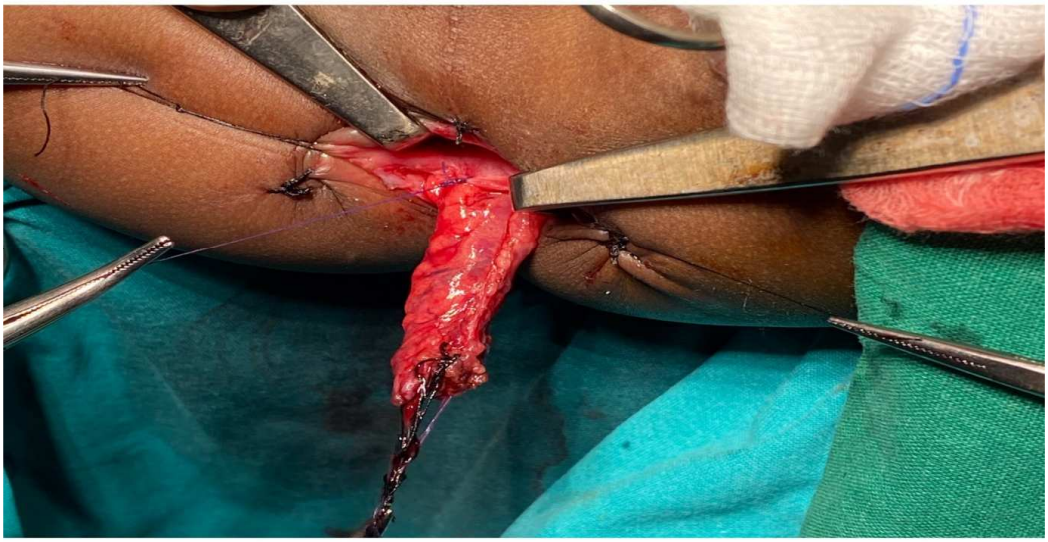


Figure:18 retrorectal pull through and anastomoses with native rectum posteriorlybelow dentate line

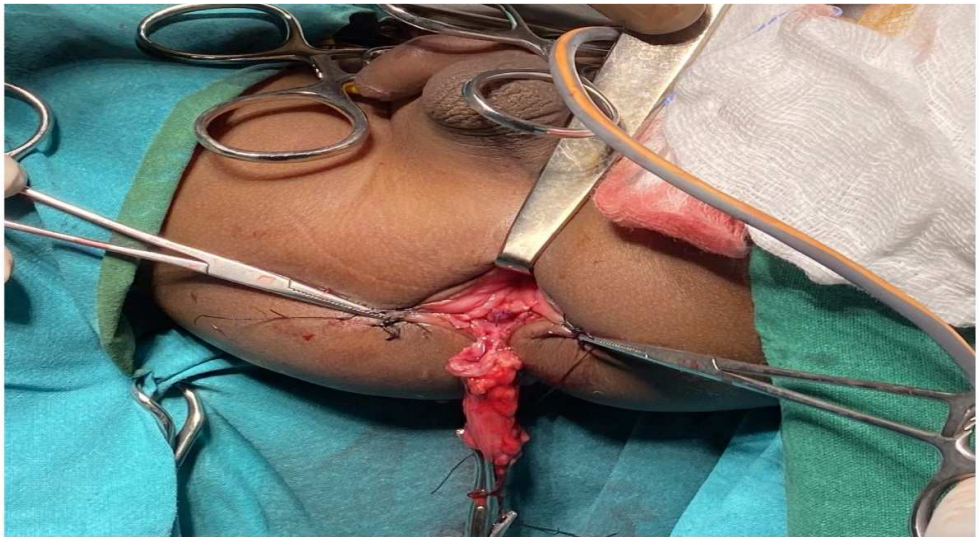


Figure: 19 Anastomosis being done

Figure:20 linear cutter with stapler

SOAVE'S PROCEDURE:

In 1964 endorectal approach was first described by Soave but first used by Yancey in 1952 for the management of two adult Hirschsprung's disease patients. The rectum's mucosa and submucosa are removed during the procedure, and ganglionic intestine is pulled through the aganglionic muscular cuff. The procedure will involve a coloanal anastomosis and a distal colectomy[86]. Important sensory fibres and the structural integrity of the sphincter are kept intact by staying in the muscular cuff of aganglionic segment.

The paediatric surgeons may now treat HD in one step thanks to advancements in paediatric nursing, anaesthetics, and intensive care during the previous ten years. Since the initial study by So BH et al[45], considering that the majority of patients diagnosed during the neonatal period, several institutions have adopted single-stage repair with encouraging results. A number of major contraindications to the main pull-through exist, such as concurrent life-threatening abnormalities, severe enterocolitis, severe proximal intestinal dilatation, and poor general health.

When new-borns with left-sided HD appear without any medical conditions that would preclude a primary repair, laparoscopic-assisted trans anal endorectal pull-through (LATEP) is also suggested.

The steps in trans anal endorectal pull-through (TEP) are, in brief, as follows. Laparoscopically taken seromuscular samples are sent for frozen section examination at various levels. Once the level of normal bowel has been established, an ultrasonic scalpel or hook electrocautery is used to separate the mesocolon close to the aganglionic gut. A trans anal circumferential incision is made 5 mm above the dentate

line, and endorectal dissection is then carried out in the rectum's submucosal plane until the level of the peritoneal cavity is reached and the muscular cuff of the rectal wall intussuscepts freely. The intraabdominal colon is freed from the muscle sleeve by continuing to split the muscular rectal wall circumferentially. The muscle cuff divides posteriorly all the way to the level of the intended anastomosis. The aganglionic colon is brought out onto the anus and resected there using the divided muscular sleeve. The anus and the normal ganglionic colon are anastomosed. Laparoscopic examination is carried out to close any potential hernia gaps and to stop the neorectum from twisting when it enters the pelvis. The portion of the bowel that was removed is sent for pathological analysis.

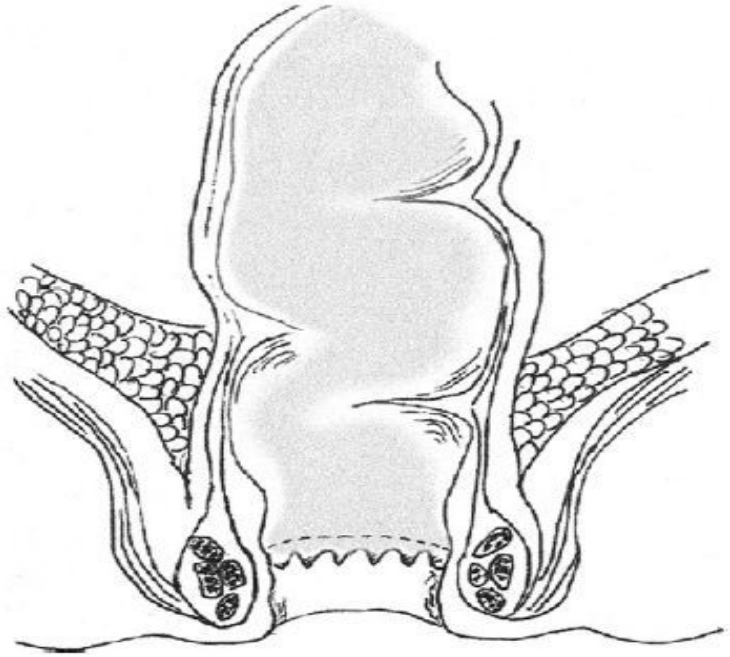


Figure 21: Trans anal circumferential incision at the intended location (dotted line)

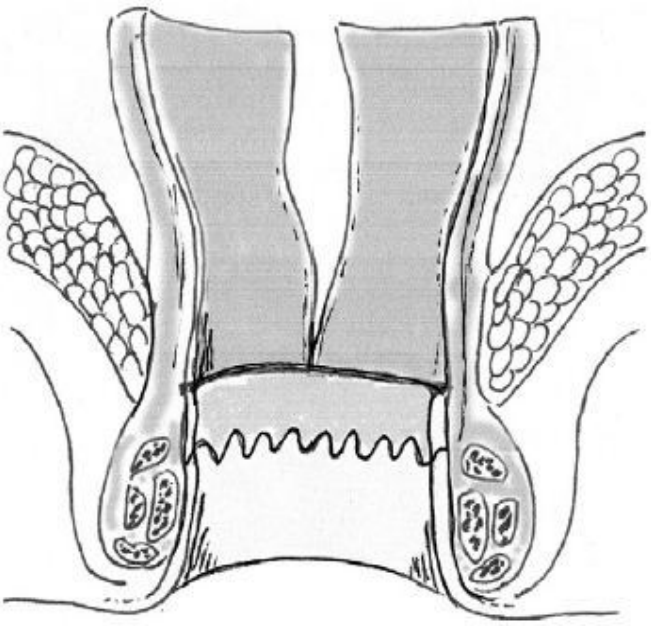


Figure 22: At the level of the intended anastomosis, the rectal cuff is split posteriorly.

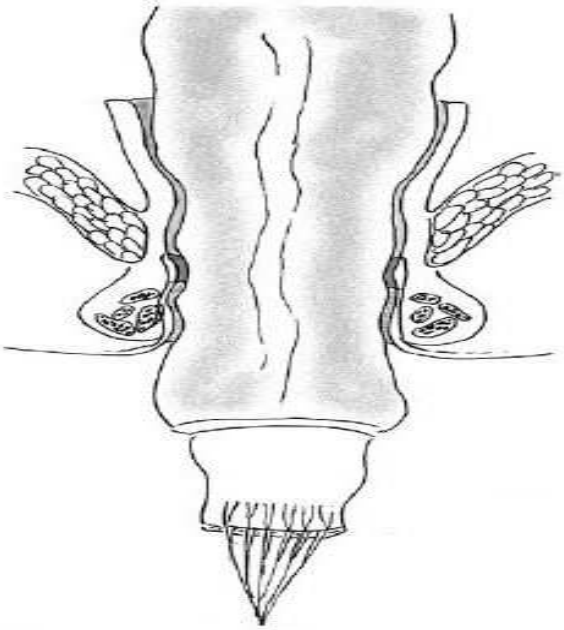


Figure 23: Before the resection, the mobilised colon was pulled through the anus to a level proximal to the transition zone.

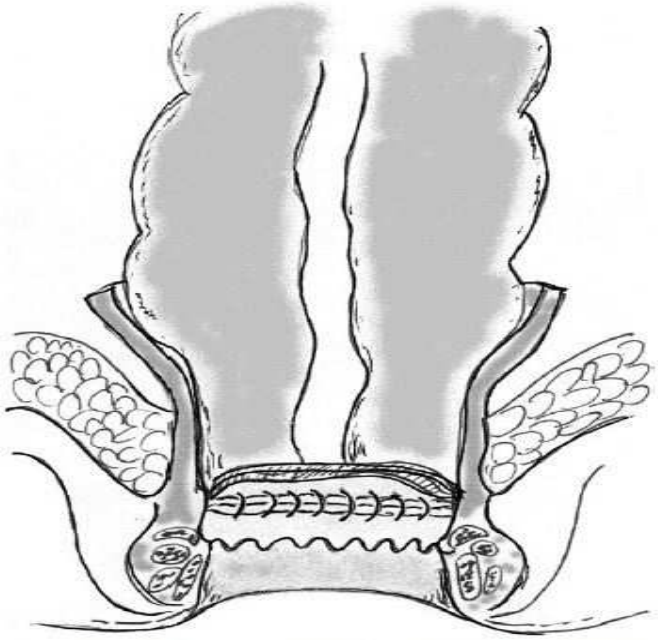


Figure 24: With the interrupted absorbable sutures anastomosis completed.

Post-operative complications

Early	Late
Anastomotic leak and cuff abscess	Bowel obstruction
Bowel obstruction	Constipation
Perineal excoriation	Enterocolitis
Stomal complications	Incontinence
Wound infection	Stricture
Wound dehiscence	

Figure 25: Post-operative complications

Heikkinen M et al[66] (1995) in their cohort study the post-operative follow-up time span was between 15 and 39 years. The study included 100 post cases and 81 healthy controls. Good faecal continence scores were achieved by all 91 patients and controls. Three of the nine patients with fair ratings were mentally disabled and experienced issues from their postoperative anastomosis. There was no significant difference in faecal continence between both patients and controls after excluding these 6 individuals with an identifiable explanation for poor anal function. All surgical techniques produced similarly acceptable continence results. Only 1 subject experienced chronic constipation as an adult. However, among 40 patients who acquired a recurring recto colonic septum after the Duhamel procedure, constipation during childhood was the predominant issue. Following the septum's recrushing, constipation disappeared. Urinary incontinence didn't exist in any of the patients. There were no restrictions on any patient's work, social life, or physical activities who had a positive continence outcome. They come to the conclusion that most patients with operated HD have similar adult faecal continence and QoL to healthy adults.

Mills J L A et al[67] (2008) in their study as HD children mature, their QOL and bowel function are measured. They stated that 51 volunteers, who had kids ranging in age from 3 to 21, were a part of their study. Faecal continence, out of all the factors in our study, had the highest correlation with QOL scores as people aged. Although there may be a clinically relevant QOL impairment, particularly in psychosocial scores, The QOL scores of children with HD and control subjects did not differ statistically.

Mattioli G et al[68] (2008) conducted research comparing the long-term results of people with classic HD who underwent the Georgeson surgery with laparoscopic assistance to those of patients who underwent the Soave-Boley treatment. They

claimed that the overall outcomes had been comparable. Long-term bowel function did not reveal any appreciable variations either.

In order to increase patients' comfort, perception of their general health, and psychologic acceptance, it is apparent that the minimum invasive technique should be favored, if possible, while hospitalization and cosmetic outcomes are taken into account.

Hashish M et al[69] (2010) conducted a study to evaluate the QoL (QOL) of patients with high imperforate anus and to quantify patient-based functional stooling outcomes. They stated that the survey's mean age was 6.5 years. The QOL and stooling score had a clear relationship. It's interesting to note that functional stooling scores declined with age. A high percentage of patients with concomitant congenital abnormalities had poor QOL. Stooling ratings drastically declined as the ARM's intensity and intricacy increased. They came to the conclusion that functional stooling issues are widespread in children and are directly related to low QOL.

Ieiri S et al[70] (2010) reported that in their study bowel movements were good. The proportion of patients who had utterly normal bowel function, however, was small. After the final procedure to treat Hirschsprung disease, paediatric surgeons should continue to strive for optimal bowel function because incontinence and soiling lower quality of life.

Gunnarsdo´ttir A et al[71] (2010) in their study examined the long-term QoL in adults following surgery for juvenile Hirschsprung disease. They claimed that individuals who had Hirschsprung disease treated as children had good long-term QoL. In comparison to the matched control group, female scores for both general and mental

health were lower. According to their research, the influence of an aganglionic segment on QoL in old age increases with segment length.

Jarvi K et al[72] (2010) in their study evaluated the gastrointestinal health and bowel function of persons with HD who had had surgery. According to their findings, patients' mean total gut function scores were considerably lower than those of controls. They noted significant differences in all parameters and reported higher incidences of inability to hold back defecation (40 percent vs 17 percent), faecal soiling (48 percent vs 22 percent), constipation (30 percent vs 9 percent), and social problems related to bowel function (29 percent vs 11 percent). Patients' gastrointestinal QoL was only slightly worse, primarily as a result of questions assessing disease-specific aspects like continence and stool function receiving much lower values. Poor bowel function could only be predicted by age, and this only provided a indication of gastrointestinal QoL.

Mustafawi A et al[73] (2012) performed a study to compare the trans anal and Duhamel pull-through surgical outcomes, parental satisfaction, and children's QoL. They noted that compared to 60.7% of patients in DPT, only 27% of TEP patients required postoperative medication. With the TEP, 22.5% of parents reported being moderately content, and 69% of parents reported being satisfied, but in the DPT, 31.8% of parents reported being poorly satisfied, 40.9% reported being fairly satisfied, and 27.3% reported being satisfied. Patients were above 3 years old; no patients showed below par results with TEP compared to 33.3% with DPT.

In comparison to Duhamel pull-through, they claimed that TEPT exhibited a lower chance of postoperative enterocolitis, failure to thrive, need for additional

surgery, and requirement for anticonstipation drugs. They came to the conclusion that both groups had significantly higher parent satisfaction and QoL.

Hondel D V D et al[74] (2015) reported that Self-reported psychosexual issues were not correlated with life satisfaction or the kind of procedure or deformity. They came to the conclusion that 50% of female ARM and HD patients experienced sexual dysfunction unrelated to QoL or type of malformation, compared to 13% of male ARM and HD patients who reported erectile dysfunction. Patients with ARM and HD thought that sexual issues needed to be better addressed while receiving medical care.

Granström A L et al[75] (2015) performed a study to evaluate bowel function and general well-being in individuals who had undergone HD surgery as children. According to their research, the median age of the patients who were included was 28 years old among the 48 participants. At the time of follow-up, two patients had stomas, and the majority of patients (73.4 percent) had had Soave's procedure. When compared to controls, the HD group's bowel function was impaired, particularly with regard to flatulence and the requirement to strain during bowel emptying, and the requirement for much defecation to clear the bowels. Additionally, faecal incontinence issues were much worse for individuals in the HD group than for controls. The GIQLI score revealed a considerably worse outcome in the HD group compared to the controls, although QoL did not significantly differ between patients and controls. They came to the conclusion that controls and adults treated for HD in childhood had similar overall QoL. They do, however, have poor gastrointestinal QoL and reduced bowel function in lax term.

Nah S A et al[76] (2015) stated that 87 participants were controls, 62 were ARM cases, and 44 were HD cases out of the 193 participants. Bowel function has an effect on

QoL, they claimed. Based on patient and caregiver self-reported outcomes, those with ARM and HD can experience a decent QoL that is equivalent to that of controls.

Khalil M et al[77] (2015) in their study, after trans anal pull-through surgery for HD, the health-related QoL for kids and parents were evaluated. In their report, they found that trans anal pull-through surgery resulted in improved postoperative long-term HRQoL. All facets of children's HRQoL were significantly negatively impacted by overflow incontinence and age during surgery. High parental satisfaction could be increased by incorporating the entire family more.

Tannuri ACA et al[78] (2016) performed a study to evaluate the presence of faecal continence and the children's QoL, both high and low, in late surgical follow-up after ARM correction. They included 63 cases and 59 controls in their investigation. In the control group, they reported that 42.4% of patients had good continence and 57.6% had normal continence. They discovered that, in comparison to controls, children with ARM have lower global QoL across all domains and on the ICF questionnaire. Patients with high and low faults showed no differences. Other related defects were present in 50.8 percent of individuals. They came to the conclusion that both QoL and ICF were negatively impacted in ARM correction patients, and there was no variation between patients with high- and low-type illness. Notably, half of the cases involved additional oddities.

Raman V S et al[79] (2016) evaluated the functional results and QoL of children treated with ARM. They claimed that there was no discernible difference in stooling outcomes and QOL in their comparison. The type of abnormality, age at evaluation, or type of surgical operation had no effect on QOL.

When compared to children without VACTERL relationship, children with associated vertebral, anorectal, cardiac, tracheoesophageal, radial, and limb anomalies had a worse QoL. Children who underwent the abdominal-perineal pull-through operation had poor stools. They came to the conclusion that the majority of children with ARM who had surgery appeared to have satisfactory QOL regardless of the stooling results.

Neuvonen M I et al[80] (2016) stated that after TEPT, HD individuals experience substantial faecal control impairment during childhood, but symptoms get better as they get older. Although overall QoL seems equivalent to controls, adults may see decline in the emotional and sexual domains.

Collins L et al[81] (2017) examined the QoL results in HD kids. In their research, 60 HD patients' parents were questioned. They stated that rectosigmoid illness affected 78% of the population. Psychosocial QoL significantly decreased when compared to youngsters in good health. Ageing, faecal incontinence, constipation, and improper elimination all have an impact on psychosocial functioning. Incontinence related to faeces also decreased physical functioning QoL. Faecal incontinence was substantially more common in children with HD. They came to the conclusion that functional results and psychosocial QoL are much lower in HD children.

Tran V Q et al[82] (2017) compared HD patients' long-term impacts and QoL to controls. According to their findings, rectosigmoid aganglionosis accounted for 72% of all HD cases. 75.5 percent of cases required open surgery, while 24.5 percent required minimally invasive surgery. Faecal incontinence and constipation were both prevalent at the time of the inquiry, at 22.6 and 13.2 percent, respectively. In the QoL survey, 180 controls and 45 patients were included; 8 patients with neurological impairment were not. Six patients had a fair QoL, two had a bad QoL, and 82.2

percent of the patients were rated as having a good QoL. A low QoL was substantially connected with the long aganglionosis variant of HD. In subscale analyses, operated patients had significantly greater prevalence rates of each dimension, such as fecal continence, school absence, dissatisfaction or anxiety, food restriction, and peer rejection, compared to controls.

Kyrklund K et al[83] (2017) stated that despite modern surgical therapies, patients with severe ARMs and HD run the danger of social limitations due to bowel function impairment. However, rather than disease-specific characteristics, self-perceptions of sickness developed in infancy may have a greater impact on adult QoL outcomes.

Sood S et al[84] (2018) in their study evaluated the long-term QoL results in HD adolescents. In their study, 58 patients were said to have been involved. In terms of general QoL scores, there were no appreciable differences between patients and healthy controls. Reduced QoL was discovered in patients and families through disease-specific questionnaires, with 17 percent of parents reporting that their child's bowel dysfunction prevented them from letting them socialize and 47 percent of parents reporting some level of anxiety or depression related to their child's bowel condition. Constipation, dysfunctional elimination, and fecal incontinence all had a negative correlation with QoL ratings.

Meinds R J et al[85] (2019) in their study assessed the QoL and long-term functional outcomes for patients with Hirschsprung's illness. According to their findings, among 346 instances, constipation was prevalent in both patients and controls as well as paediatric and adult patients. Adults with HD experienced straining and insufficient evacuation noticeably more frequently than controls. Hirschsprung's illness affected both adults and children with a greater proportion of faecal incontinence, most

frequently soiling, than controls. In a number of QoL domains, patients with poor functional results performed significantly worse. They came to the conclusion that functional outcomes are superior in adults than in children, although a sizable portion of individuals with Hirschsprung's illness continue to have constipation and soiling.

Ghorbanpour M et al[86] (2019) studied both immediate and long-term side effects, and assessed HD Soave surgical results. According to their analysis, the average age of the 55 infants who underwent surgery was 38 10 days old, with 56.4 percent of the patients being female. The typical length of stay was 7.3 days. The average birth weight of children was 2970±447 gr. The majority of the patients (52.7 percent) and term births (74.5 percent). Congenital cardiac disease was the comorbidity that was most prevalent. Intestinal obstruction was the most prevalent short-term problem (25.5%) and the most frequent long-term event (constipation, 27.3% each case), with a mortality rate of 14.5 percent. They came to the conclusion that a single stage of surgery combined with appropriate postoperative care is a safe and successful treatment for Hirschsprung's illness.

Drissi F et al[87] (2019) in their study evaluated the QoL and social outcomes for adult patients who underwent childhood Hirschsprung disease surgery. They enrolled 43 patients with Hirschsprung disease in their study. The average QoL score for HD and anorectal malformations was 611 out of 800.

Physical discomfort and diarrhoea were the 2 most negatively affected aspects. The degree of faecal continence was little altered. Hirschsprung disease patients outperformed the French populace in terms of educational attainment. They came to the conclusion that adult patients with Hirschsprung disease sequelae have slightly

worse QoL. Despite the negative effects of this congenital anomaly, the final condition can be regarded as good.

Townley OG et al[88] (2019) conducted a study to evaluate the bowel habits and general well-being of people with HD. The average follow-up was 5.4 years among 71 patients. 24 of the 38 eligible individuals had their gastrointestinal function evaluated. The average score for incontinence was 17 and the average score for constipation was 17. The scores for incontinence and constipation were noticeably worse than those of healthy controls and did not get better with age. A QoL assessment was done on 56 patients, and there was no difference between their group and healthy controls.

Wong CWY et al[89] (2019) compared the effectiveness of laparoscopic-assisted anorectal pull-through (LAARP) with posterior sagittal anorectoplasty in terms of QoL and long-term dysfunction (PSARP). They asserted that the two groups' QOL ratings obtained using the HAQL questionnaire were comparable, with the exception of social functioning, where the LAARP patients had significantly lower mean scores. The 10-year results for patients with LAARP and PSARP were found to be comparable in terms of QOL and impaired function.

Zheng H et al[90] (2019) evaluated the QoL and mid-term bowel functions in kids with low-type ARMs. Children with low-type ARM were shown to have adequate bowel control and QOL. Despite the fact that ARMs are benign, many children who have them experience issues with anal function that lower their QoL. The iatrogenic causes of fecal incontinence include repeated surgeries, misaligned anuses, and inadequate constipation care.

Fosby M V et al[91] (2020) conducted a study to see whether the bowel's functionality improves with age after receiving TEP for HD. Two interviews were reportedly conducted seven years apart. The two interviews' median ages were 8.1 and 15.4 years, respectively. The percentage of soiling was the same in the first (52%) and second (52%) follow-ups. At the first follow-up, 20% of patients and 24% at the second reported having constipation. At the first and second interviews, bowel management was utilized by 30% and 32%, respectively. They came to the conclusion that HD patients frequently have constipation and soiling years after surgery and that there was no evidence of bowel function improving with age.

Saysoo M R et al[92] (2020) in their study HQAL was used to evaluate QoL after surgical care of HD/ARM. They stated that for the Duhamel and Soave groups, the mean HAQL scores were 2.5 and 2.8, respectively. Parents of patients who were questioned for the qualitative study discussed how their child's life has improved as a result of the procedure. However, after Soave surgery, often bloating was a prominent complaint, whereas after Duhamel surgery, firm stools were still a major issue. They came to the conclusion that, in comparison to the Duhamel group, Patients who underwent Soave tended to score higher overall on QoL measures.

Espeso L et al[93] (2020) examined the children with HD's QoL. Only 50% of children aged 6 to 11 and 68% of adolescents were found to have acquired good voluntary bowel movements among the 63 individuals included in the study, according to the researchers. 55 percent of teenagers and 70 percent of children had soiling problems. The total HAQL proxy 6-11 score was 528/700; the highest results were in the categories of faecal continence, social functioning, and urinary continence, while the lowest results were in the categories of general well-being and diurnal faecal

continence. The aggregate HAQL proxy 12–16 score was 607/700, with the strongest ratings coming from social and urine continence. Additionally, they came to the conclusion that the sole factor strongly linked to low QOL was soiling.

Byström C et al[94] (2020) conducted a study to assess bowel function, lower urinary tract symptoms, and QoL in patients treated for HD with TEP in comparison to healthy controls in order to identify any short-term problems and evaluate long-term effects. According to their findings, HD patients' median bowel function scores were considerably poorer than those of controls. In addition, more HD patients than controls reported having poor gastrointestinal function. Compared to controls, HD patients reported a somewhat poorer QoL in the KIDSCREEN domain financial resources.

Dai Y et al[95] (2020) in their systematic review evaluated the frequency of long-term effects and the standard of living for HD patients who had surgery. According to their findings, the combined prevalence of symptoms related to faecal incontinence, constipation, and bladder dysfunction was 0.20, 0.14, 0.07, and 0.95, respectively, while the combined mean score for gastrointestinal-related QoL was 118. They came to the conclusion that HD patients over the age of ten have a significant prevalence of faecal incontinence and poor QoL overall.

Saysoo M R et al[96] (2020) indicated that the mean HAQL scores of Duhamel group was 2.50 and mean HAQL scores of Soave group was 2.79. Interviewed parents of patients spoke on how their child's life has changed as a result of surgery. But following the Soave procedure, recurrent bloating was a common complaint, but after the Duhamel procedure, hard stools were a significant problem.

Loganathan A K et al[97] (2021) in their study, HD children's overall QoL, influence on family, and functional bowel status were assessed. 86 youngsters took part in the study, and they reported that 67.4% of them had rectosigmoid illness and underwent Soave's endoanal pull through (74.4 percent). Low Rintala scores in 21% of individuals indicate poor functional bowel outcomes. Only 11% of kids had subpar QoL ratings. The outcomes of family functioning in the same patient subgroup were likewise adversely impacted. The connection between Rintala scores and QoL scores was statistically significant. Disease severity, surgical technique, and length of follow-up did not statistically significantly affect QoL.

Gunadi et al[98] (2022) evaluated how HD was treated using Soave (n = 8), Duhamel (n = 4), and TEP (n = 13) surgical procedures in their study. The median age for HD diagnosis was 10 months, while the median age for pull-through surgery was 17 months. The median BFS level follow-up for HSCR patients was 72 months after pull-through. Ten individuals had BFS levels that were normal, and 11 patients had levels that were good. Additionally, 54% of TEP patients had normal BFS levels, as opposed to 50% of Soave patients and 50% of Duhamel patients, who had good BFS levels and poor BFS levels, respectively. Accordingly, 38.5 percent of TEP group had fewer than one stain weekly and didn't need to keep changing undergarments, 50% of Soave patients had no signs of soiling, and up to 50% of Duhamel patients needed protective aids owing to regular soiling.

In addition, whereas 46.2 percent of TEP patients and 75% of Soave patients experienced an accident, 75% of Duhamel patients did not. Particularly, there may be certain advantages of the TEP technique over the Soave and Duhamel procedures,

they concluded, suggesting that the type of definitive surgery may have an impact on the long-term bowel functional outcome.

Huizer V et al[99] (2022) carried out a meta-analysis to assess the influence of various potential factors on the HRQoL of HD patients after surgery. They discovered that HD patients' general HRQoL was not noticeably poorer as compared to healthy control subjects. Physical, psychological, and social HRQoL scores were not substantially worse than those of healthy controls. Health-related outcomes were unaffected by the gender of the patients or whether generic HRQoL was measured by parental proxy or self-report. HD patients' physical HRQoL was decreased by disease-specific symptoms, but not by either psychosocial or social HRQoL.

MATERIALS AND METHODS

Study Design:

Prospective cross-sectional study was carried out to assess the Quality of life (QoL) of patients with Hirschsprung's disease and anorectal malformation after pull through surgery.

Study Area:

In the department of Pediatric Surgery of Kaher's Dr Prabhakar Kore Hospital and Medical Research Centre this study was conducted.

Study population:

During the study period of 1st January 2021 to 31st December 2021, patients who were attended to the outpatient and inpatient department of pediatric surgery for follow-up after minimum of 6 months of pull through surgery for ARM and HD and Age matched patients who are attending the pediatrics outpatient department who present with non-gastrointestinal symptoms were included as controls in the study.

Study period:

The study was carried out from the first of January 2021 to the last day of December 2021.

Inclusion criteria:

- Age 3 year and beyond
- Both sexes
- All cases of Hirschsprung's disease
- All cases of Anorectal malformation
- Completed all stages of ARM/Hirschsprung's surgery and 6 months beyond last stage of surgery

were included in this study.

Exclusion criteria:

- Major associated Gastro intestinal anomalies requiring independent surgery
- More than two definitive surgeries (redo-cases)
- Sacral vertebral defects
- Presence of neural tube defects
- Mentally retarded children

were excluded from the study.

Sample size:

Formula used for sample size calculation was

$$n = \frac{\sigma^2 z^2}{E^2}$$

n is the sample size required,

σ^2 is the variance,

E is the precision,

Z is the value corresponding to level of confidence required, for 95% confidence level $Z\alpha_{/2}$ is 1.96.

Physical health component summary scale of patients with ARM observed to be 45. 3±15.This standard deviation was used for sample size calculation.

From above formula**,**

$$n = \frac{15^2 \times (1.96)^2}{5^2}$$

$$n = 34.57 \approx 35$$

Hence a total of 35 cases those who underwent pull through procedure for the management of ARM and HD before 6 months.

Ethical committee approval:

The institutional human ethics committee granted consent for this study to evaluate the quality of life of patients with Hirschsprung's disease and anorectal malformation following pull-through surgery among cases attending the outpatient and inpatient department of pediatric surgery.

Data Collection:

Patients attending the outpatient and inpatient department of paediatric surgery with past history of surgical correction for HD and ARM using pull through procedures were included in the study along with patients who are attending the paediatrics outpatient department who present with non-gastrointestinal symptoms were included in the study. Each participant was given information about the study and given the assurance that their identity would be kept completely private. They

were also given the option to withdraw from the study at any point of time of the study.

Prior to the interview, the study participants gave written informed consent. Participants underwent a thorough history and examination by the lead investigator using a pre-structured proforma after obtaining written informed consent. Along with demographic characteristics and presenting symptoms, the principal investigator assessed their Quality of Life using HSCR/Anorectal malformation quality of life questionnaire (HAQL). The following dimensions were used to categorize these items: laxative diet, constipating diet, presence of diarrhoea, presence of constipation, faecal continence, urine continence, social functioning, emotional functioning, body image, and physical functioning. Each item received a value between 0, 1, 2, 3, and 4 which indicates response as never, almost never, sometimes, often, almost always respectively using PedsQL^{TM} Quickview^{SM} Scoring with a higher score indicating a higher quality of life. The scores were determined for each dimension by dividing the total number of items answered in that dimension by the sum of the item scores. By computing the sum of the all-dimension, the overall QoL score was calculated[100,101].

The same proforma was used for both groups' findings, and the primary investigator entered the clinical presentation.

DATA ANALYSIS AND STATISTICAL METHODS:

Data was entered in excel sheet. Descriptive analysis was carried out by mean and standard deviation for quantitative variables, frequency, and proportion for categorical variables. Non normally distributed quantitative variables were summarized by median and interquartile range (IQR). Data was also displayed using the relevant diagrams, such as box plots, pie charts, and bar charts.

By visually inspecting histograms and normality Q-Q plots, all quantitative variables were examined for normal distribution within each category of explanatory variable. Additionally, the Shapiro-Wilk test was used to evaluate the normal distribution. When the Shapiro-Wilk test's p value was >0.05, the distribution was regarded as normal.

Using the Chi square test or Fisher's Exact test (or both, if the total sample size was less than 20 or the anticipated value in any cell was less than 5), categorical outcomes were compared between study groups. For normally distributed Quantitative parameters the mean values were compared between study groups using independent sample t-test (2 groups)

P value < 0.05 was considered statistically significant. IBM SPSS version 22 was used for statistical analysis. (1)

1. IBM Corp. Released 2013. IBM SPSS Statistics for Windows, Version 22.0. Armonk, NY: IBM Corp

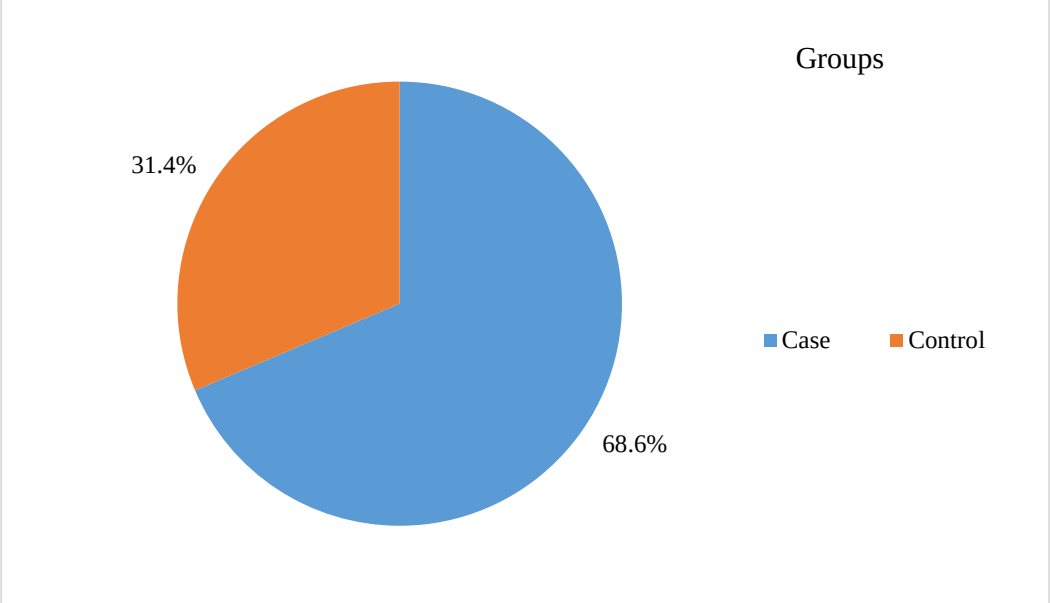
RESULTS

Table 1: Descriptive analysis of groups in the study population (N=51)

Groups	Frequency	Percentages
Case	35	68.63%
Control	16	31.37%

According to the data above, there are 68.63% cases and 31.37% controls in the study participants.

Graph 1: Pie chart of groups in the study participants (N=51)



The above pie chart shows, there are 68.63% cases and 31.37% controls in the study population.

Table 2: Comparison of mean of age between groups(N=51)

Parameter	Groups (Mean± SD)		P value
	Case (N=35)	Control (N=16)	
Age	6.66 ± 3.32	4.92 ± 1.43	0.051

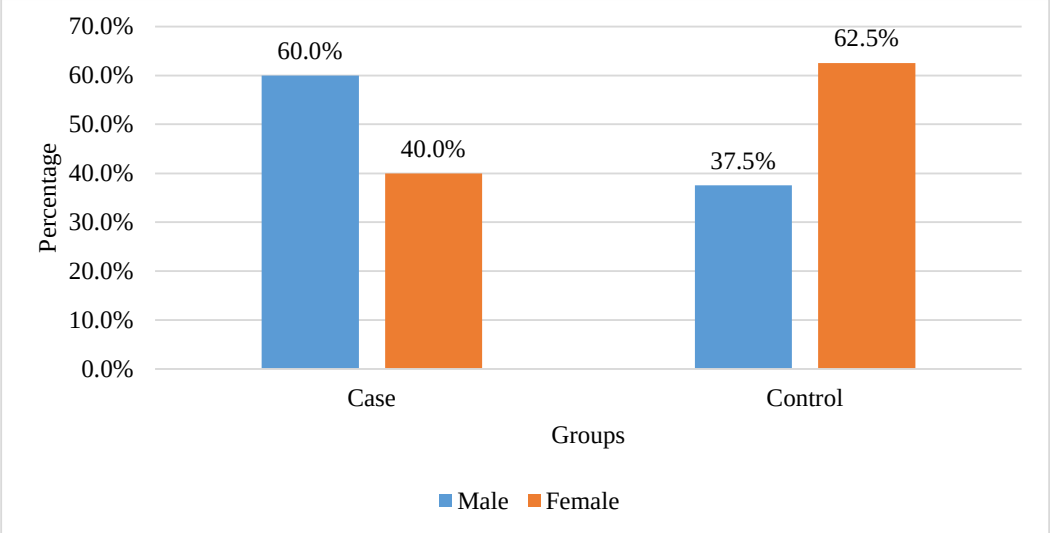
Above table shows that Mean± SD of case is 6.66 ± 3.32 and control is 4.92 ± 1.43 with a p value showing significance (p value: 0.05).

Table 3: Comparison of gender between groups (N=51)

Gender	Groups		Chi square	P value
	Case (N=35)	Control (N=16)		
Male	21 (60%)	6 (37.5%)	2.231	0.135
Female	14 (40%)	10 (62.5%)		

According to the data above, 60% are male and 40% are females in cases and 37.5% are male and 62.5% are female in controls in the study population. P value is not significant.

Graph 2: Cluster bar chart of comparison of gender between groups (N=51)



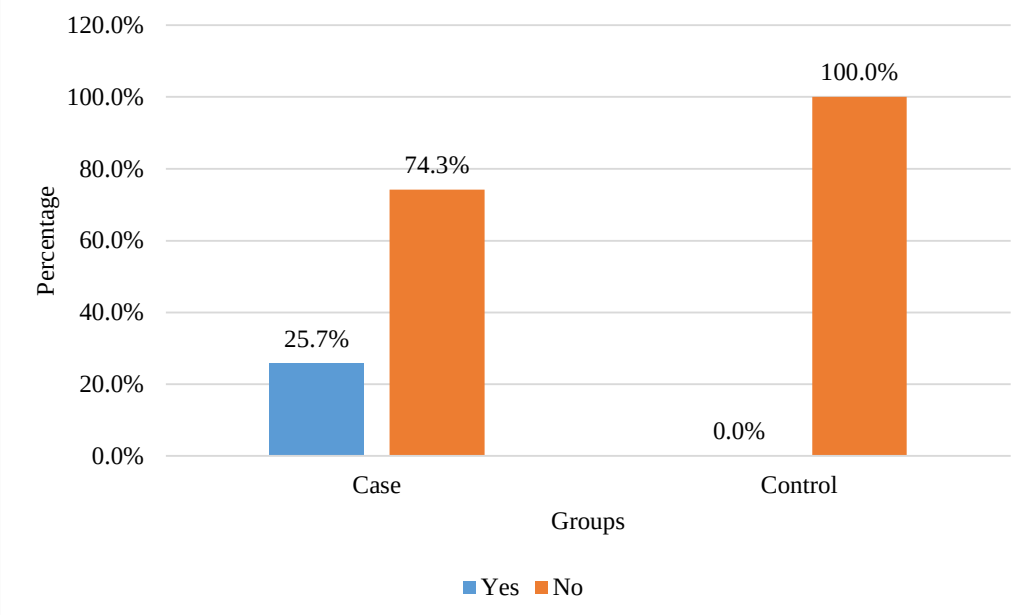
Above bar chart shows gender comparison between both groups.

Table 4: Comparison of consanguinity between groups (N=51)

Consanguinity	Groups		Chi square	P value
	Case (N=35)	Control (N=16)		
Yes	9 (25.71%)	0 (0%)	4.99	0.025
No	26 (74.29%)	16 (100%)		

According to the data above, 25.71% patients in cases have history of consanguinity whereas controls have 0%. P value (0.02) is significant.

Graph 3: Cluster bar chart of comparison of consanguinity between groups (N=51)



Above bar chart shows comparison of history of consanguinity between both groups

Table 5: Comparison of degree of consanguinity between groups (N=9)

Degree Of Consanguinity	Groups	
	Case (N=9)	Case (N=0)
1st Degree	1 (11.11%)	-
2nd Degree	7 (77.78%)	-
3rd Degree	1 (11.11%)	-

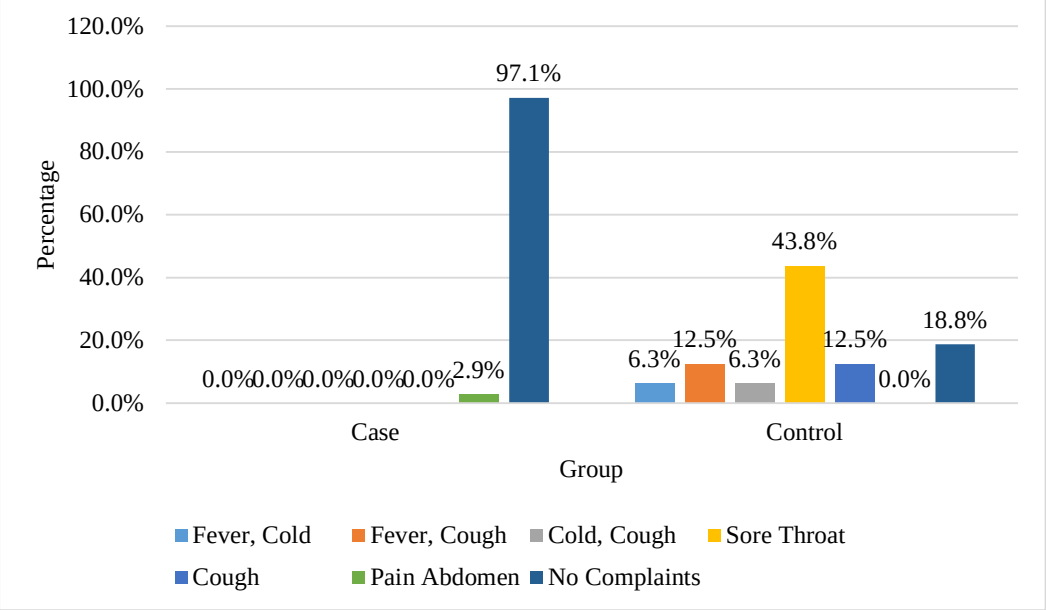
Above table shows the comparison of degree of consanguinity between both groups.

Table 6: Comparison of presenting illness between groups (N=51)

Presenting Illness	Groups	
	Case (N=35)	Control (N=16)
Fever, Cold	0 (0%)	1 (6.25%)
Fever, Cough	0 (0%)	2 (12.5%)
Cold, Cough	0 (0%)	1 (6.25%)
Sore Throat	0 (0%)	7 (43.75%)
Cough	0 (0%)	2 (12.5%)
Pain Abdomen	1 (2.86%)	0 (0%)
No Complaints	34 (97.14%)	3 (18.75%)

Above table shows comparison between the cases and controls by which they presented to hospital. As we see all the patients in control group are presented with non-gastrointestinal complaints unlike in cases group one patient presented with pain abdomen.

Graph 4: Cluster bar chart of comparison of presenting illness between groups (N=51)



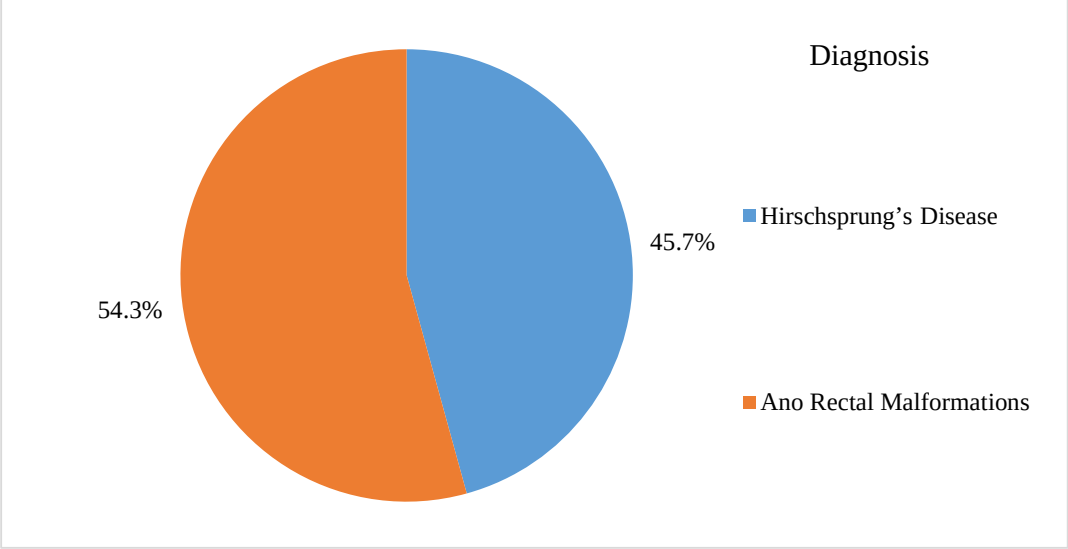
The above bar chart shows comparison of presenting illness between both groups.

Table 7: Descriptive analysis of diagnosis in the study population (N=35)

Diagnosis	Frequency	Percentages
Hirschsprung's Disease	16	45.71%
ANO Rectal Malformations	19	54.29%

According to the data above, there are 45.71% Hirschsprung's Disease and 54.29 % ANO Rectal Malformations in the cases group of the study population.

Graph 5: Pie chart of diagnosis in the study participants (N=35)



Above shows that in the study population 45.71% are Hirschsprung's Disease and 54.29 % are ANO Rectal Malformations in the cases group.

Table 8: Types of Hirschsprung's disease in the study participants: a descriptive analysis (N=16)

Type of Hirschsprung's Disease	Frequency	Percentages
Ultra-short segment	0	0%
Short Segment	16	100.00%
Long segment	0	0%
Total colonic agangliosis segment	0	0%
Extended intestinal agangliosis segment	0	0%

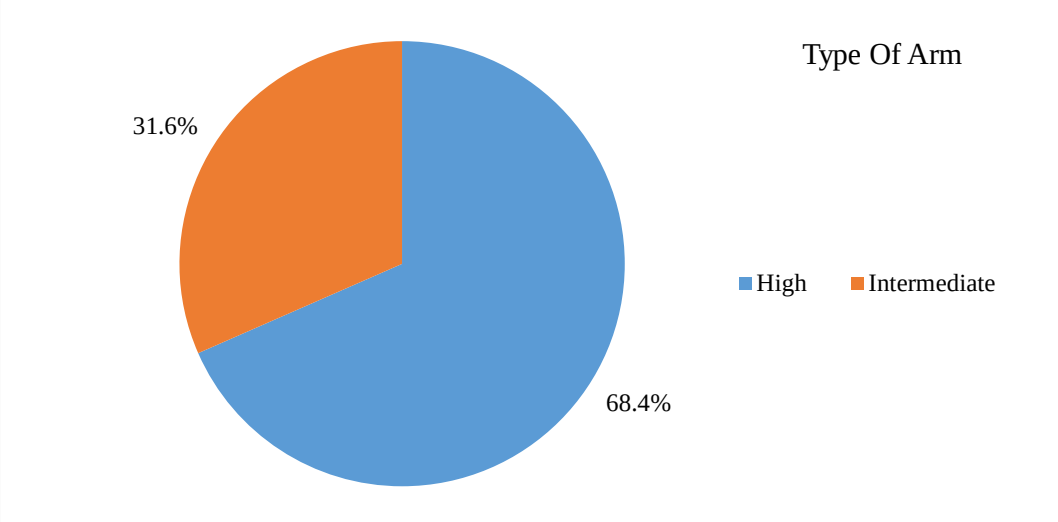
Above table shows the frequency of the types of the Hirschsprung's disease in the study population.

Table 9: Type of arm in the study participants: a descriptive analysis (N=19)

Type of ARM	Frequency	Percentages
High	13	68.42%
Intermediate	6	31.58%
Low	0	0%

Above table shows the frequency of the types of the ARM in the study participants.

Graph 6: Pie chart of type of in the study participants (N=19)



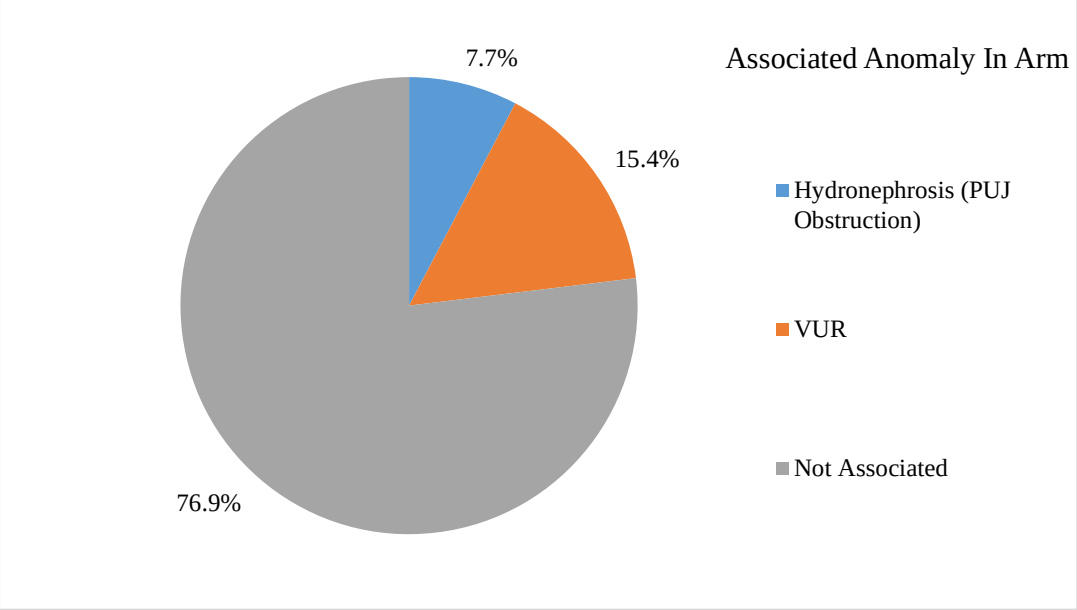
Above pie chart shows the frequency of the types of the ARM in the study population.

Table 10: Comparison of associated anomaly in arm between type of arm (N=19)

Associated Anomaly In Arm	Type Of Arm	
	High (N=13)	Intermediate (N=6)
Not Associated	10 (76.92%)	6 (100%)
VUR	2 (15.38%)	0 (0%)
Hydronephrosis (PUJ Obstruction)	1 (7.69%)	0 (0%)

Above table shows the associated anomalies of the ARM in the ARM group.

Graph 7: Pie chart of associated anomaly in High arm in the Associated Anomaly In Arm study population (N=13)



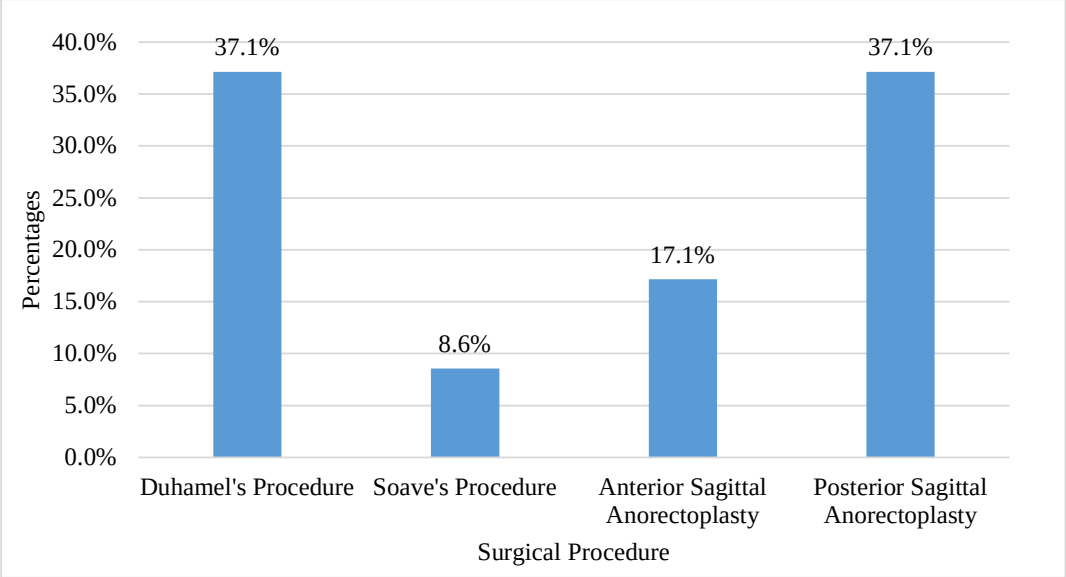
Above pie chart shows the associated anomalies of the ARM in the ARM group.

Table 11: Surgical procedure in the study participants: a descriptive analysis (N=35)

Surgical Procedure	Frequency	Percentages
Duhamel's Procedure	13	37.14%
Soave's Procedure	3	8.57%
Anterior Sagittal Anorectoplasty	6	17.14%
Posterior Sagittal Anorectoplasty	13	37.14%

Above table shows the frequency of surgical procedures that patients of Hirschsprung's disease and anorectal malformation had undergone i.e., Duhamel and Posterior Sagittal Anorectoplasty each has percentage of 37.14%, soaves procedures percentage is 8.57% and anterior Sagittal Anorectoplasty percentage is 17.14%.

Graph 8: Bar chart of surgical procedure in the study participants (N=35)



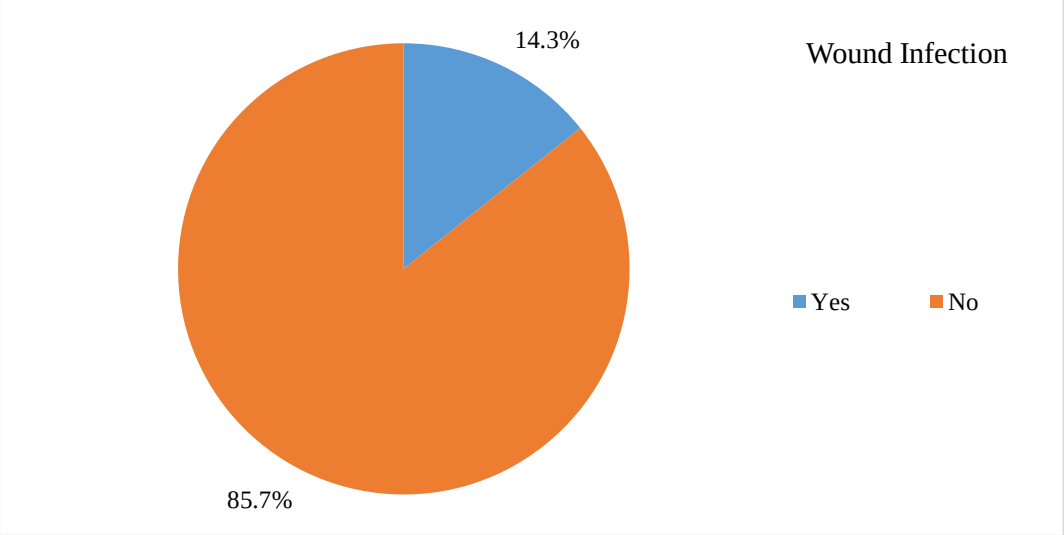
Above bar chart shows the frequency of surgical procedures that patients of Hirschsprung's disease and anorectal malformation had undergone.

Table 12: wound Infection in the study participants: a descriptive analysis (N=35)

Wound Infection	Frequency	Percentages
Yes	5	14.29%
No	30	85.71%

Above table shows the incidence of surgical site infection in the study population i.e. 14.29%.

Graph 9: Pie chart of wound infection in the study participants (N=35)



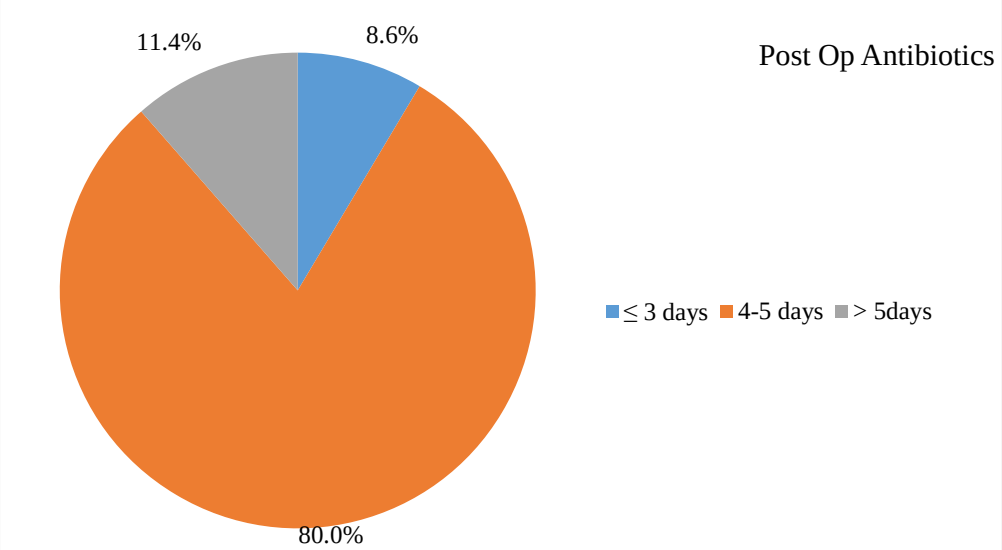
Above pie diagram shows the incidence of surgical site infection in the study population i.e. 14.29%.

Table 13: Descriptive analysis of post op antibiotics in the study population (N=35)

Post Op Antibiotics	Frequency	Percentages
≤ 3 days	3	8.57%
4-5 days	28	80.00%
> 5days	4	11.43%

Above table shows the duration of the post-operative antibiotics given in the study population i.e. ≤ 3 days, 4-5 days, > 5days are 8.57%,80%,11.43% respectively.

Graph 10: Pie chart of post op antibiotics in the study population (N=35)



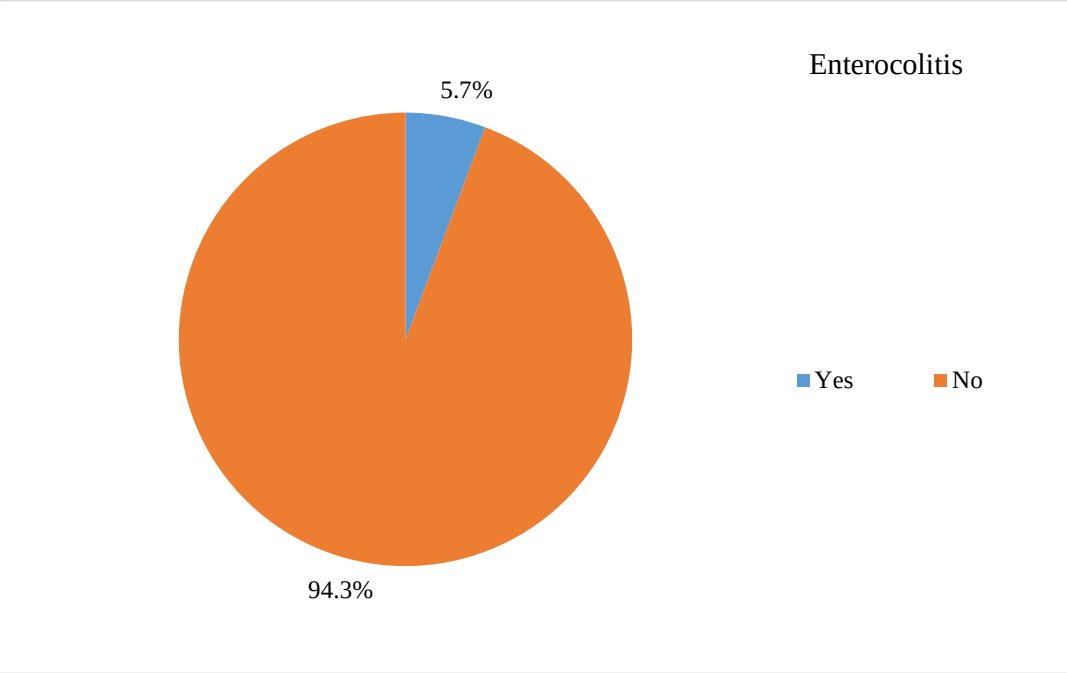
Above pie diagram shows the duration of the post-operative antibiotics given in the study population.

Table 14: Descriptive analysis of enterocolitis in the study population (N=35)

Enterocolitis	Frequency	Percentages
Yes	2	5.71%
No	33	94.29%

Above table shows the history of enterocolitis in the study population i.e. 5.71%.

Graph 11: Pie chart of enterocolitis in the study population (N=35)



Above pie diagram shows the history of enterocolitis in the study participants.

Table 15: Descriptive analysis of the number of days that the study population spent in hospitals following surgery (N=35)

Parameter	Mean ± SD	Median	Minimum	Maximum
Hospital Stay Duration Post Surgery in Days	4.8 ± 1.35	5.00	3.00	8.00

Above table shows the duration of the hospital stay in the study population with a range of 3days to 8days.

Table 16: Comparison of mean of mean of HAQL scores between HD & Controls (N=32)

Parameter	Groups (Mean± SD)		P value
	Hirschsprung's Disease (N=16)	Control (N=16)	
LAXATIVE DIET	89.06 ± 18.19	100 ± 0	0.023
CONSTIPATING DIET	93.75 ± 14.43	100 ± 0	0.094
PRESENCE OF DIARRHEA	96.09 ± 5.98	100 ± 0	0.014
PRESENCE OF CONSTIPATION	87.5 ± 22.36	98.44 ± 6.25	0.069
FECAL CONTIENCE	95.12 ± 6.14	100 ± 0	0.003
URINARY CONTIENCE	100 ± 0	100 ± 0	*
SOCIAL FUNCTIONING	94.27 ± 22.92	100 ± 0	0.325
EMOTIONAL FUNCTIONING	98.96 ± 4.17	100 ± 0	0.325
BODY IMAGE	100 ± 0	100 ± 0	*
PHYSICAL SYMPTOMS	94.44 ± 7.79	100 ± 0	0.008
Total Mean of The HAQL Score	94.92 ± 4.7	99.84 ± 0.63	<0.001

Above table shows the comparison of the HAQL scores between Hirschsprung's disease and controls. Where there is significant difference in the laxative diet, presence of diarrhoea, faecal continence, physical symptoms and overall HAQL scores between the both groups with control having the better HAQL scores in comparison to Hirschsprung's disease with p value <0.05**.**

Table 17: Comparison of mean of mean of HAQL scores between HD surgical procedures (N=16)

Parameter	Surgical Procedure (Mean± SD)		P value
	Duhamel's Procedure (N=13)	Soave's Procedure (N=3)	
LAXATIVE DIET	88.46 ± 19.41	91.67 ± 14.43	0.794
CONSTIPATING DIET	94.23 ± 14.98	91.67 ± 14.43	0.792
PRESENCE OF DIARRHEA	97.12 ± 5.48	91.67 ± 7.22	0.162
PRESENCE OF CONSTIPATION	84.62 ± 24.02	100 ± 0	0.298
FECAL CONTIENCE	95.19 ± 6.58	94.79 ± 4.78	0.923
URINARY CONTIENCE	100 ± 0	100 ± 0	*
SOCIAL FUNCTIONING	92.95 ± 25.42	100 ± 0	0.647
EMOTIONAL FUNCTIONING	98.72 ± 4.62	100 ± 0	0.647
BODY IMAGE	100 ± 0	100 ± 0	*
PHYSICAL SYMPTOMS	94.66 ± 8.21	93.52 ± 6.99	0.828
Total Mean of The HAQL Score	94.59 ± 5.16	96.33 ± 1.42	0.581

Above table shows the comparison of the HAQL scores between Duhamel's and Soave's surgical procedures for the surgical management of Hirschsprung's disease. Where it shows there is no significant difference between the both groups in individual parameters and overall HAQL score.

Table 18: Comparison of mean of mean of HAQL scores between ARM & Controls (N=35)

Parameter	Groups (Mean± SD)		P value
	ANO Rectal Malformations (N=19)	Control (N=16)	
LAXATIVE DIET	81.58 ± 28.68	100 ± 0	0.015
CONSTIPATING DIET	92.11 ± 18.73	100 ± 0	0.102
PRESENCE OF DIARRHEA	92.11 ± 11.19	100 ± 0	0.008
PRESENCE OF CONSTIPATION	73.68 ± 35.82	98.44 ± 6.25	0.010
FECAL CONTIENCE	83.72 ± 21.67	100 ± 0	0.005
URINARY CONTIENCE	98.61 ± 5.89	100 ± 0	0.354
SOCIAL FUNCTIONING	97.8 ± 7.79	100 ± 0	0.269
EMOTIONAL FUNCTIONING	96.01 ± 11.96	100 ± 0	0.193
BODY IMAGE	100 ± 0	100 ± 0	*
PHYSICAL SYMPTOMS	91.37 ± 11.6	100 ± 0	0.006
Total Mean of The HAQL Score	95.44 ± 20.3	99.84 ± 0.63	0.393

Above table shows the comparison of the HAQL scores between Anorectal malformations and controls. Where there is significant difference in the laxative diet, presence of diarrhoea, presence of constipation, faecal continence and physical symptoms between the both groups with control having the better HAQL scores in comparison to Anorectal malformations with p value < 0.05. But over all HAQL shows no significant differences between both groups.

Table 19: Comparison of mean of mean of HAQL scores between type of ARM (N=19)

Parameter	Type Of Arm (Mean± SD)		P value
	High (N=13)	Intermediate (N=6)	
LAXATIVE DIET	80.77 ± 30.88	83.33 ± 25.82	0.862
CONSTIPATING DIET	92.31 ± 18.78	91.67 ± 20.41	0.947
PRESENCE OF DIARRHEA	92.31 ± 10.87	91.67 ± 12.91	0.911
PRESENCE OF CONSTIPATION	69.23 ± 34.09	83.33 ± 40.82	0.441
FECAL CONTIENCE	84.13 ± 24.26	82.81 ± 16.63	0.906
URINARY CONTIENCE	97.92 ± 7.22	100 ± 0	0.496
SOCIAL FUNCTIONING	96.79 ± 9.36	100 ± 0	0.420
EMOTIONAL FUNCTIONING	94.5 ± 14.33	99.31 ± 1.7	0.431
BODY IMAGE	100 ± 0	100 ± 0	*
PHYSICAL SYMPTOMS	91.02 ± 13.38	92.13 ± 7.33	0.854
Total Mean of the HAQL Score	96.83 ± 24.42	92.42 ± 6.08	0.673

Above table shows the comparison of the HAQL scores between types of ARM. Where it shows there is no significant difference between the both groups in individual parameters and overall HAQL score.

DISCUSSION

The goal of surgical treatment in HD and ARM is to ensure that the kid has regular, spontaneous bowel movements free from incontinence or constipation. Numerous studies have examined quality of life after surgery. However, these researches were conducted in the developed world, therefore it was unknown how they would apply in a developing nation. As these patients move through life, auxiliary services in developed economies are crucial to their treatment. In low- and middle-income countries, these services are badly underfunded, which should, in theory, result in a poor quality of life[97]. Using healthy controls from a developing nation as a comparison, the goal of this study was to examine the quality of life of patients after receiving definitive surgical treatment which was conducted in Belgavi, India, at the KLES Dr. Prabhakar kore hospital and medical research centre, for a period of one year in paediatric surgery department.

One of the key outcomes for all patients who have ARM and HD surgery is acceptable quality of life. A disease-specific tool measuring the quality of life (QoL) of children and adolescents with faecal incontinence was developed in the Netherlands and is called the Hirschsprung's Disease/Anorectal Malformation Quality of life Questionnaire (HAQL)[92]. In addition to questions about social and emotional functioning, body image, and physical symptoms, the HAQL includes several categories with questions about diet, laxative use, constipation, diarrhoea, urine incontinence, and faecal incontinence[92]. At this time, it's not apparent if patients of HD and ARM actually have lower QoL when compared to controls without the disease.

In our study we had observed that there is significant difference in the usage of laxative diet, presence of diarrhoea, faecal incontinence, physical symptoms

parameters of HAQL questionnaire scores of Hirschsprung's disease when compared to the healthy paediatric controls. Faecal incontinence has the negative impact on the quality of life of the patients. In this study we had observed a significant difference in the faecal incontinence with p value of 0.003 when compared with controls , where 12.5% has the need to be around the neighbourhood of a toilet, 25% complains of loss of faeces before reaching toilet, 18.75% complains of soiling in the daytime, 31.25% complains of soiling in the night, none of the patients complaints of loss of faeces during physical activity, emotional moments, coughing or sneezing which was significant with the previous studies done by Collins L et al [(81)] where they observed similar findings such as 96% of parents claimed that their children could sense bowel movements. There were zero bowel accidents reported by 19 parents (37%) each week, one to five reported by 18 parents (35%) per week, and more than five reported by 15 parents (29%) per week. The majority of parents—82% and 96%, reported not using oral or rectal medications respectively [(81)]. The majority of parents claimed to have noticed significant dietary changes in their children's bowel movements (54/60, 90%) and a sensitivity to certain foods (44/60, 73%)[(81)]. 22% of children with Hirschsprung's illness were hospitalised for constipation, and roughly 28% of them had abdominal pain [(81)]. Similar findings were seen in our study, where 12.5% of patients reported feeling abdominal pain and 31.25 % of patients avoided eating certain foods to maintain thin stools. Whereas Ieiri et al.[(10)] showed faecal incontinence in 36% of the study population[(10)]. In other study done by Diseth et al.[(23)] they reported incontinence of more than 30%[(23).]

The majority of research discovered a link between the aganglionic segment's length and poor functional and, subsequently, quality of life results. Collins L. et al., however, found that findings for quality of life were unaffected by the length of the

aganglionosis section. [81] In our study as all the reported Hirschsprung's patients are short segment aganglionosis better quality of life results are noted in comparison to other studies. [10,23]

In our study there is over all significant difference between the both groups with controls having better quality of life is seen i.e. p value of < 0.001 which is consistent with the other reported studies [10,23,81].

According to the Saysoo MR[92] study, patients who underwent the Soave operation had improved quality in terms of faecal continence and spontaneous bowel movements. In this study when HAQL scores are compared between the surgical procedures of Hirschsprung's disease i.e. Duhamel's and Soave's procedure there is no significant difference in the overall score HAQL score which is consistent with study reported by Nah SA et al [76]. The sample size of the patients operated on using the Soaves technique (n=3) is smaller than that of the patients operated on using the Duhamel's approach, which is the cause of the study's insignificant difference between the two techniques (13).

In our study when HAQL scores of ARM cases compared to healthy controls showed no significant difference in the overall quality of life which are consistent with study reported by Nah SA et al[76] where sub group analysis showed not significant difference in the quality of life. In our study we observed significant difference in the use of the laxative diet i.e around 42.1%, presence of diarrhoea was reported in 10.5% and p value of 0.008, presence of constipation was seen in 26.2% and p value of 0.01, faecal incontinence was seen in 52.4% where we had observed a significant difference with p value of 0.005 when compared with controls , where 36.8% has the need to be around the neighbourhood of a toilet, 47.36% complains of loss of faeces before reaching toilet, 52.63% complaints of soiling in the daytime,

26.3% complains of soiling in the night, 15.78% of the patients complained of loss of faeces during physical activity, emotional moments, coughing or sneezing, in physical symptoms where as 36.8% complained of being flatulent and 10.52% reported of not being able to differentiate between flatulence or faeces. Associated anomalies, type of definitive surgical procedure i.e. Posterior sagittal anorectoplasty and anterior sagittal anorectoplasty had no effect on quality of life when compared to controls which was consistent with results of Raman VS et al.[79]. When HAQL scores of high and intermediate type of arm is compared no significant difference observed similar results were reported by Tannuri AC et al[78] where the some of the cases are associated with anomalies. Because of the abnormal colonic mobility in arm patients, or as Holsehneider recently pointed out, because of discrepancies in the rectal end innervation used by surgery, 31.57% of the study population in our study experienced constipation, difficult to pass stools, or straining while passing stools. Whereas Zheng H et.[90] al investigation's made comparable observations, Approximately 70% of the patients complained of constipation[90].

In our study we observed about 25.71% of the patients has history of consanguinity which shows significant relation to the risk of development of the disease. In about 14.29% patients wound infection was observed and more than five days of antibiotics are used in about 11.43% patients. In about four patients secondary suturing was needed and two patients i.e. 5.71% had enterocolitis episodes which lead to increase in hospital stay.

In this study impairment of the bowel habits are observed in both Hirschsprung's disease and anorectal malformation patients as constipation could be managed by usage of oral laxatives daily or periodical enemas. But faecal incontinence leads to severe impairment in the quality of life of patients in regards to

frequent soiling of clothing, diarrhoea, fear of bad odour which impairs the social functioning of the patient. Similar findings are observed in the study conducted by Ieiri S et al. [10]

As we said, as one gets older, the quality of life improves. This is due to one's increased self-awareness, or a greater understanding of the disease and acceptance of one's limitations, which encourages patients and patients' families to cooperate more with the required medical management. The second is coping mechanisms, which reduce anxiety in patients and caregivers while improving patient quality of life. Conversely, unhelpful coping mechanisms result in denial and a low quality of life. It has also been demonstrated that problem-focused (getting medical help) and emotion-focused (accepting, seeking emotional support) interventions improve quality of life[102] similar findings are noted in our study. This study adds to a growing body of information showing that children with HD develop gastrointestinal problems after surgical management, primarily faecal incontinence.

Limitations of the study

There is a chance of selection bias because many children are still receiving follow-up care and are probably symptomatic as a result of having weak bowel movements.

Long term results could not be predicted, our study just provides a snapshot of the child's life at this point in time.

Due to the small sample size, we were unable to conduct some statistically significant subgroup analysis.

CONCLUSION

Hirschsprung's disease and anorectal malformation surgery is the most commonly performed procedure in paediatric surgery departments globally. Quality of life is a crucial component of medical care, particularly for Hirschsprung's disease and Ano rectal malformation children who have undergone surgery and experience impairment of bowel habits.

However, there isn't a single technique that can access their outcomes, results, follow-up requirements, or need for additional therapy objectively.

Here by using modified Hirschsprung's disease/Anorectal Malformation Quality of Life questionnaire (HAQL) provides us an opportunity to access these children with respective intermediate outcomes, regularity of bowel habits, parental acceptance, and the problems faced by children and parents after surgery. As surgery is not the end point of the treatment in children.

During regular follow-up appointments, merely noting the bowel habits is insufficient. It is crucial to incorporate the idea of QOL.

The majority of the patients in this study had a decent quality of life, and there was no correlation to the way the stooling went, the kind of anomaly, or the kind of definitive surgery.

SUMMARY

Hirschsprung's disease and anorectal malformation is the most common condition for which patients attend to the paediatric surgery outpatient department and after the definitive surgical correction the amount of improvement of the quality of life is unclear.

In this study we had conducted a cross sectional study to assess the Quality of life (QoL) of patients with Hirschsprung's disease and anorectal malformation after pull through surgery for a period of one year by comparing the patients with the health pediatric controls.

In this study we studied cases of 35 and controls of 16 where 60% of the cases are male and 40% are females with history of consanguinity in 25.71% patients in the cases. in the cases sample it was subdivided into Hirschsprung's disease(n=16) and anorectal malformations(n=19). In the comparative analysis between Hirschsprung's disease and controls Where there is significant difference in the laxative diet, presence of diarrhea, fecal continence, physical symptoms and overall HAQL scores. in comparative analysis between Duhamel's and soave's procedure there is no significant difference between the both groups in parameters and over all quality of life. When comparative analysis done between ARM and controls there is significant difference in the laxative diet, presence of diarrhea, presence of constipation, fecal continence and physical symptoms. But there is not significant difference in comparison between high and intermediate arm.

Quality of life is the ultimate goal when it comes to surgical management. in the developing countries like ours where auxiliary services are underfunded which might lead to the poorer quality of life. In regards to that the HAQL questionnaire was

developed in Netherlands for accessing the quality of life in patients of Hirschsprung's and anorectal malformations after surgical correction. Which can tailor the management in specific to the need of the patient attending to the outpatient department after the surgical correction of the anorectal malformation and Hirschsprung's disease.

Here by incorporating the idea of quality of life questionnaire in the regular follow seems beneficial to the patients, thus improving their quality of life.

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ANNEXURE II: CONSENT

INFORMED CONSENT FOR PARTICIPATION IN RESEARCH STUDY

Mr./Mrs. _____ we are requesting you to enroll yourself in study titled **"QUALITY OF LIFE OF PATIENTS WITH HIRSCHSPRUNG'S DISEASE AND ANORECTAL MALFORMATION AFTER PULL THROUGH SURGERY " : OBSERVATIONAL STUDY FOR PERIOD OF 1 YEAR"** conducted by REG NO: BH0120018, Postgraduate in M.S.General Surgery under the guidance of DR. _____ Department of General Surgery, J.N. Medical college, Belgaum under KLE university, Belagavi, Under Co-Guidance of Dr. _____ department of Pediatric Surgery, J.N. MEDICAL COLLEGE, Belgaum under KLE university, Belagavi.

Respected Sir/ Madam,

We request you to participate in our study. Your participation in the research is voluntary. Your decision to participate in the study or otherwise will not affect the relationship with KLES Prabhakar Kore hospital. If you decided not to participate, you are free to withdraw at any time.

Purpose of study:

The purpose of the study is to find out "Quality of life of patients with Hirschsprung's disease and Anorectal malformation after pull through surgery".

Procedure involved:

Procedure after counselling and proper consent the patients included in the study were subjected for proper history since birth, before and after surgery.

The patient subjected to both clinical examination and systemic examination i.e, mainly per abdominal examination, per rectal examination. Per abdominal examination involves with respective to abdominal distension, scars and their healing. Per rectal examination with respect to empty rectus / fecal matter, caliber of lumen anocutaneous reflex and bulbocavernosus reflex.

Risks and Benefits:

There are no invasive procedures, hence there is risk involved.

Type of Study:

This study is an observational study. It involves patients with pull through surgery for Hirschsprung's and anorectal malformation with HAQL questionnaire will be compared with same questionnaire with same age normal peers.

Participant selection:

It includes all patient coming for follow up with Hirschsprung's disease and anorectal malformation meeting inclusion and exclusion criteria.

Voluntary Participation:

Your participation in research is voluntary. It is your choice whether to participate or not. Your decision whether to participate in the study or not will not change present or future health care services offered to you and will not affect your relationship with J.N. Medical College. If you choose not to participate in this study, you will still be offered the routine treatment of Hirschsprung's disease and anorectal malformation that is given at our hospital. You will continue to receive the routine care at our hospital even if you decline to participate in this study. If you decide to participate you are free to withdraw at any time.

Privacy and Confidentiality:

The only people who will know that you are the research subject will be the members of the research team. No information about you or information provided by you during the research will be disclosed to others without your written permission except:

1. In emergency to protect your rights and welfare.

2. If required by law.

Authorization to Publish Results:

When the results of the research are published or discussed, in a conference, no information will be displayed that would disclose your identity. Any information that is obtained in connection with this study and that can be identified with you will remain confidential. Results of the study will be used to compare the two procedures on the points listed above.

Right to refuse or withdraw from study:

You do not have to participate in this research if you do not wish to. You can withdraw at any time from the study. There will be no penalty for withdrawal. Your treatment and care in this hospital will not change irrespective of whether you agree to participate or not. You can be removed from the study if necessary.

Alternative:

You are free to withdraw yourself from this study at any point of time. You will continue to receive the routine care even if you decline to participate in the study. You will be treated for the same even if you have declined from the study. You will be informed about any new information that may affect your decision to participate in the study.

Institutional/sponsor's policy:

In the event of any injury related to the study, treatment will be made available through KLE's Hospital & MRC, Belgaum. There is no compensation or payment for such medical treatment by law. If you are injured you may contact REG NO: BH0120018, Post graduate student, Department of General Surgery, KLE's Hospital& MRC

Consent Statement

Mode of communication of consent form: Verbal / Written

Contents: Self read /Read out by Investigator

Participant's awareness regarding voluntary withdrawal from study: Yes / No

Investigators decision to remove participants from study: Yes /No

Awareness regarding voluntary participation: Yes / No

Adequate time given to clarify any doubts about the study or rights to study participant: Yes / No

In case they have any questions related to the study, in future or in case of study related injury or illness, they can contact REG NO: BH0120018 Department of General Surgery, KLES Hospital and MRC, Belagavi, or DR. _____ Professor and Head of Dept. Of General Surgery, KLES Hospital and MRC, Belagavi or Dr. _____Professor and Head of department of Pediatric Surgery.

If they have any queries about their rights as a study subject, they may call DR.HARSHA HEGDE, Chairman, and Ethical Committee for Human Subjects Research. Professor, Department of Paediatrics, J. N. Medical College, Belagavi, Phone number-9448113403.

Signature or left thumb print of participant or legally authorized representative

_____Participant's name. _____Participant's signature/thumb print

_____Experimenters' name _____Experimenters' signature

_____Witness' name _____Witness' signature

ASSENT (<18 Years)

I have read the information in this form. After understanding all details about the study, I agree to give assent to be included as a volunteer in the study titled **"**QUALITY OF LIFE OF PATIENTS WITH HIRSCHSPRUNG'S DISEASE AND ANORECTAL MALFORMATION AFTER PULL THROUGH SURGERY": OBSERVATIONAL STUDY FOR PERIOD OF 1 YEAR**"**

Name of the Participant: Signature/Left thumb impression:

Name of the Parent: Signature of the the Parent:

Name of Principal Investigator: Signature:

Name of witness: Signature:

Date: Place:

SCREENING FORM

1. PATIENT'S UHID NO: ☐☐☐☐☐☐☐

2. AGE: ☐ YEARS ☐ MONTHS

3. GENDER: ☐

   1. MALE
   2. FEMALE

4. HEIGHT (IN CMS): ☐

5. WEIGHT (IN KGS): ☐

6. DIAGNOSIS: ☐

   1. HIRSCHSPRUNG'S DISEASE
   2. ANORECTAL MALFORMATION

7. ANY MAJOR ASSOCIATED ANOMALIES REQUIRING INDEPENDENT SURGERY: ☐

   1. YES
   2. NO

8. SURGICAL PROCEDURE DONE: ☐

   1. DUHAMEL'S PROCEDURE
   2. SOAVE'S PROCEDURE
   3. SWENSON'S PROCEDURE
   4. ANTERIOR SAGITTAL ANORECTOPLASTY
   5. POSTERIOR SAGITTAL ANORECTOPLASTY

9. DATE OF SURGERY: ☐ DD ☐ MM ☐ YY

10. NO. OF DEFINITIVE SURGERIES: ☐

    1. ≤ 2 DEFINITIVE SURGERIES
    2. > 2 DEFINITIVE SURGERIES

11. DATE OF INTERVIEW: ☐ DD ☐ MM ☐ YY

SCREENING FORM

12. ADDRESS: ☐

1. BELGAVI
2. OUTSIDE BELGAVI WITH IN KARNATAKA
3. OUTSIDE KARNATAKA

13. PHONE NO OR LANDLINE NO: ☐☐☐☐☐☐☐☐☐☐

14. INFORMANT: ☐

1. PARENTS
2. OTHERS

15. OCCUPATION : ☐

1. UNEMPLOYED
2. UNSKILLED
3. SEMI-SKILLED
4. SKILLED
5. PROFESSIONAL

16. EDUCATION : ☐

1. ILLITERATE
2. PRIMARY (1ST-7TH STD)
3. HIGH SCHOOL (8TH-10TH STD)
4. INTERMEDIATE
5. DEGREE AND ABOVE

17. SOCIO-ECONOMIC STATUS : ☐

1. LOW
2. MIDDLE
3. HIGH

18. APPLICANT IS WILLING TO GIVE CONSENT: ☐

1. YES
2. NO

19. FINAL RESULT: ☐

1. INELIGIBLE
2. ELIGIBLE BUT REFUSED
3. ELIGIBLE AND PARTICIPATING

PROFORMA

1. PATIENT'S UHID NO: ☐☐☐☐☐☐

2. AGE: ☐ YEARS ☐ MONTHS

3. GENDER: ☐

   1. MALE
   2. FEMALE

4. INFORMANT: ☐

   1. PARENTS
   2. OTHERS

5. ADDRESS: ☐

   1. BELGAVI
   2. OUTSIDE BELGAVI WITH IN KARNATAKA
   3. OUTSIDE KARNATAKA

6. PRESENT ILLNESS:

7. HISTORY:

8. FAMILY HISTORY: ☐

   1. CONSANGUITY

      A. YES
      B. NO

   2. DEGREE: ☐

      A. I DEGREE (MARRIAGE BETWEEN BROTHER AND SISTER)
      B. II DEGREE (MARRIAGE WITH THE FALHER'S OWN SISTER OR MOTHER'S OWN BROTHER)
      C. III DEGREE (MARRIAGE WITH FATHER'S SISTER'S SON OR DAUGHTER OR MOTHER'S BROTHER'S SON OR DAUGHTER)
      D. IV DEGREE (MARRIAGE IS BETWEEN DISTANT RELATIVES)

PROFORMA

9. TREATMENT HISTORY:

10. CLINICAL DIAGNOSIS: ☐

1. HIRSCHSPRUNG'S DISEASE
2. ANORECTAL MALFORMATION

11. GENERAL PHYSICAL EXAMINATION:

1. VITAL SIGNS:
   1. PULSE RATE: ☐
   2. RESPIRATORY RATE: ☐
   3. BLOOD PRESSURE: ☐
   4. TEMPERATURE: ☐

2. ANTHROPOMETRY:
   A. WEIGHT IN KGS: ☐
   B. HEIGHT IN CMS: ☐

3. PRESENCE OF PALLOR: ☐
   A. YES
   B. NO

12. INVESTIGATIONS:

1. CBC
   A. HB% ☐
   B. PCV ☐
   C. TLC ☐
   D. PLATELET COUNT ☐

PROFORMA

2. BLOOD UREA ☐

3. SERUM CREATININE ☐

4. SERUM ELECTROLYTES

   A. SERUM SODIUM ☐

   B. SERUM POTASSIUM ☐

   C. SERUM CLORIDE ☐

   D. BICRABONATE ☐

5. ERECT ABDOMEN X-RAY

   A. DILATED BOWEL LOOPS: PRESENT ☐ ☐ ABESNT

   B. PNEUMO PERITONEUM: PRESENT ☐ ☐ ABSENT

   C. INVERTOGRAM ☐

      1. HIGH ANORECTAL MALFORMATION

      2. LOW ANORECTAL MALFORMATION

   D. AFFECTED BOWEL OF SMALL CALIBER: PRESENT ☐ ☐ ABSENT

   E. VARIABLE AMOUNTS OF COLONIC DISTENSION ☐

      1. SHORT SEGMENT DISEASE

      2. LONG SEGMENT

      3. TOTAL COLONIC AGANGLIONOSIS

      4. ULTRASHORT SEGMENT DISEASE

6. USG ABDOMEN ☐

   A. LOADED COLON

   B. AIR FLUID LEVEL

   C. RECTUM FULLNESS

PROFORMA

7. OTHERS (AS PER REQUIREMENT)

   1. BARIUM ENEMA ☐
   2. ANORECTAL MANOMETRY ☐
   3. COLON TRANSIT TIME ☐
   4. FULL THICKNESS RECTAL BIOPSY ☐
   5. RECTAL PULL THROUGH SEGMENT BIOPSY ☐
   6. BOWEL TRANSIT TIME ☐

13. OPERATION DETAILS:

   1. DEFINITIVE SURGICAL PROCEDURE DONE: ☐
      A. DUHAMEL'S PROCEDURE
      B. SOAVE'S PROCEDURE
      C. SWENSON'S PROCEDURE
      D. ANTERIOR SAGITTAL ANORECTOPLASTY
      E. POSTERIOR SAGITTAL ANORECTOPLASTY
   2. DEFENITIVE OPERATIVE FINDINGS:
   3. OPERATIVE DIAGNOSIS: ☐
   4. HISTOPATHOLOGY REPORTS:

14. POST OPERATIVE:

   1. ANTIBIOTICS:
      A. DURATION: ☐
      B. SIMPLE ANTIBIOTICS: ☐
      C. ESCLATION ANTIBIOTICS: ☐

PROFORMA

2. PRESENCE OF WOUND INFECTION:

A.

1. YES ☐

2. NO

B. SITE: ☐

1. ABOMINAL WALL

2. PERINEAL WALL

C. DAY OF WOUND INFECTION: ☐

D. MANAGEMENT OF WOUND INFECTION: ☐

E. DURATION OF HEALING: ☐

3. HOSPITAL STAY: ☐

4. ENTEROCOLITIS EPISODES: ☐

A. PRESENT:

1. NUMBER OF EPISODES ☐

B. ABSENT

5. OTHERS: ☐

PROFORMA

6. FOLLOW UP:

HIRSCHSPRUNG'S DISEASE ANORECTAL MALFORMATION QUALITY OF LIFE QUESTIONNAIRE (HAQL):

1. LAXATIVE DIET:

A. EATING SPECIAL FOOD ON PURPOSE TO GET THIN STOOLS ☐

B. AVOID EATING SPECIAL FOOD 8TO MAINTAIN THIN STOOLS ☐

2. CONSTIPATING DIET:

A. EATING SPECIAL FOOD ON PURPOSE TO GET THICK STOOLS ☐

B. AVOID EATING SPECIAL FOOD TO MAINTAIN THICK STOOLS ☐

3. PRESENCE OF DIARRHEA:

A. THIN STOOLS ☐

B. MORE THAN FOUR TIMES A DAY THIN STOOL ☐

4. PRESENCE OF CONSTIPATION:

A. SOLID STOOLS / REDUCED FREQUENCY OF DEFECATION / DIFFICULTY IN DEFECATION / STRAINING AT DEFECATION. ☐

5. FAECAL CONTINENCE:

A. IMPORTANT TO BE IN NEIGHBOURHOOD OF A TOILET ☐

B. LOSS OF FAECES BEFORE REACHING THE TOILET ☐

C. SOILING AT DAYTIME ☐

D. SOILING AT NIGHT TIME ☐

E. LOSS OF FAECES AT NIGHT TIME ☐

F. LOSS OF FAECES DURING PHYSICAL ACTIVITY ☐

G. LOSS OF FAECES DURING EMOTIONAL MOMENTS ☐

H. LOSS OF FAECES DURING COUGHING OR SNEEZING ☐

PROFORMA

6. URINARY CONTINENCE:

A. LOSS OF URINE BEFORE REACHING THE TOILET ☐

B. LOSS OF URINE DURING PHYSICAL ACTIVITY ☐

C. LOSS OF URINE DURING EMOTIONAL MOMENTS ☐

D. LOSS OF URINE DURING COUGHING OR SNEEZING ☐

7. SOCIAL FUNCTIONING:

- **12-16 YEARS:**

A. AMOROUS FEELINGS ☐

B. FANTASIZE ABOUT MAKING LOVE ☐

- **ALL AGE GROUPS:**

A. DAILY ACTIVITIES ☐

B. SOCIAL ACTIVITIES ☐

C. STAYING THE NIGHT ELSEWHERE ☐

8. EMOTIONAL FUNCTIONING

- **8-11 AND 12-16 YEARS:**

A. BEING ASHAMED OF LEAVING THE CLASSROOM TO GO TO THE TOILET ☐

B. FEELING THAT YOU ARE TEASED MORE THAN OTHER KIDS. ☐

- **17 YEARS:**

A. FEELING EMBARRASSED ☐

B. FEELING UNCERTAIN ☐

C. FEELING WORRIED FOR THE FUTURE ☐

PROFORMA

- **ALL AGE GROUPS:**

A. FEELING ASHAMED ☐

B. FEELING DIFFERENT ☐

C. FEELING LESS APPRECIATED BY OTHERS ☐

D. FEELING AFRAID THAT OTHERS WILL SMELL YOUR FAECES ☐

9. BODY IMAGE:

A. FEELING LESS ATTRACTIVE ☐

B. FEELING DISSATISFIED WITH YOUR BODY ☐

10. PHYSICAL SYMPTOMS:

A. FEELING SWOLLEN (ABDOMEN) ☐

B. FEELING NO URGE WHILE BOWELS ARE FULL OF FAECES ☐

C. GOING TO THE TOILET WHILE HAVING NO URGE ☐

D. DIFFICULTY TO LOOSE FAECES ☐

E. DIFFICULTY TO DISCRIMINATE BETWEEN FLATULENCE OR FAECES ☐

F. BEING FLATULENT ☐

G. DIFFICULT TO LET A FLATUS ☐

H. HAVING ABDOMINAL PAIN ☐

I. HAVING BOWEL MOVEMENTS ☐

11. SEXUAL FUNCTIONING (>-17YEARS):

A. INTEREST IN SEX ☐

B. SEXUAL ACTIVITY ☐

PROFORMA

PedsQL™ Quickview℠ Scoring:

RESPONSE CHOICES	NEVER	ALMOST NEVER	SOMETIMES	OFTEN	ALMOST ALWAYS
RAW SCORES	0	1	2	3	4
0-100 SCALE SCORES	100	75	50	25	0

Sl no	Ip no	Age	Sex	Consanguity	degree of Consanguity	presenting illness	X ray abdomen	Rectal biopsy	type of Hirschsprung's disease	crosstable prone lateral x ray	type of ARM	associated anomaly in ARM	Diagnosis	Surgical Procedure	Post op antibiotics	wound infection	enterocolitis	duration post surgery	HAQL 11	HAQL12	HAQL21	HAQL22	HAQL31
		(in years)	1-male	1.yes	1- I st degree	1- constipation	1- dialated bowel loops	1- s/o hirschsprung's disease	1.short segment	1-high anorectal malformation	1. High	1.not associated	1-Hirschsprung's disease	1-duhamel's procedure	1- ≤ 3 days	1-yes	1-yes	(in days)	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score
			2-female	2.no	2- 2nd degree	2- fecal incontinence	2-pneumo peritoneum	2-normal	2.ultra short segment	2- intermediate anorectal malformation	2.Intermediate	2.renal anamolies	2-ano rectal malformations	2-soave's procedure	2- 4-5 days	2-no	2-no		1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)
					3- 3rd degree	3-urinary incontience	3-normal	3-not done	3.long segment	3- low anorectal malformation	3. Low	A.VUR		3-swenson's procedure	3- > 5days				2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)
					4-4rth degree	4- pain abdomen			4.total colonic agangliosis segment			B. hydronephrosis (puj obstruction)		4-anterior sagittal anorectoplasty					3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)
						5-no complaints			5.extended intestional agangliosis segment			C. agenesis of kidney		5-posterior sagittal anorectoplasty					4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)
												D. hypospadias							5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)
												E. undescended testis											
1	884934	4years 6 months	1	2		5	1	3		1	1	1	2	5	2	2	2	5	5	5	1	1	1
2	446870	11years 7months	2	1	2	5	1	3		1	1	1	2	4	2	2	2	5	1	1	1	1	1
3	968130	3years 1 month	1	2		5	3	3		1	1	2A	2	5	2	2	2	5	1	5	1	5	2
4	376259	11years5months	2	1	2	5	1	3		1	1	1	2	4	2	2	2	5	1	3	1	1	1
5	692410	17years 3 months	2	2		5	1	3		1	1	1	2	5	2	2	2	3	1	1	1	1	1
6	859280	11 years 6 months	2	2		5	1	3		1	1	1	2	4	2	2	2	3	1	1	1	1	1
7	825577	6years2months	2	2		5	1	3		1	1	1	2	4	2	2	2	5	1	5	1	5	1
8	913817	5years 3 months	1	1	1	5	1	3		1	1	1	2	5	2	1	2	7	1	3	1	1	2
9	846821	4years 6months	1	2		5	3	3		1	1	2A	2	5	2	2	2	5	1	1	1	1	2
10	866385	4years 4 months	2	2		5	3	3		1	1	1	2	4	2	2	2	4	1	1	1	1	1
11	947995	6years 7 months	1	2		5	1	3		1	1	2B	2	5	2	2	2	3	1	1	1	1	1
12	618561	7years 11 months	2	1	2	5	1	3		1	1	1	2	5	2	2	2	3	1	1	1	1	2
13	975385	3years 1month	2	2		5	1	3		1	1	1	2	5	2	2	2	5	1	1	1	1	2
14	711854	5years10months	1	2		5	1	3		2	2	1	2	5	2	2	2	5	1	5	1	1	1
15	898459	4years	2	2		4	1	3		2	2	1	2	4	2	2	2	5	1	5	5	1	1
16	870854	4years 6 months	1	2		5	3	3		2	2	1	2	5	2	2	2	3	1	1	1	1	2
17	496133	11 years 6 months	1	2		5	1	3		2	2	1	2	5	2	2	2	3	1	1	1	1	1
18	485762	11years 3months	2	1	2	5	3	3		2	2	1	2	5	3	1	2	8	1	1	1	1	1
19	602632	9years 3 months	1	2		5	1	3		2	2	1	2	5	2	2	2	5	1	1	1	1	2
1	1000156	4years 1month	1	2		5	1	1	1				1	1	1	2	2	3	1	5	1	1	1
2	886999	4 years 5 months	1	2		5	1	1	1				1	1	3	1	2	8	1	5	1	1	1
3	1038815	3years 1 month	1	1	2	5	1	1	1				1	1	1	2	2	5	1	1	1	5	1
4	906065	3years 7months	2	2		5	1	1	1				1	1	2	2	2	5	1	1	1	1	1
5	939047	3years 2months	1	1	2	5	1	1	1				1	1	2	2	2	4	1	3	1	1	1
6	887236	5years8months	2	1	3	5	1	1	1				1	1	3	1	2	7	1	1	1	1	1
7	652781	7years 11 months	1	2		5	1	1	1				1	1	2	2	1	5	1	1	1	1	1
8	661143	9years 3 months	1	2		5	1	1	1				1	1	2	2	2	5	1	1	1	3	1
9	768823	6years2months	1	2		5	1	1	1				1	1	2	2	1	4	1	1	1	1	2
10	851661	11years 7months	1	2		5	1	1	1				1	1	2	2	2	5	1	1	1	1	1
11	92821	3years 4 month	1	1	2	5	1	1	1				1	1	1	2	2	4	1	1	1	1	1
15	926143	3years 4months	1	2		5	1	1	1				1	1	2	2	2	5	1	1	1	1	1
16	883525	4years 7months	1	2		5	1	1	1				1	1	2	2	2	5	1	3	1	1	1
12	842350	7years8months	2	2		5	1	1	1				1	2	3	1	2	7	1	1	1	1	2
13	885258	7years6months	1	2		5	1	1	1				1	2	2	2	2	4	1	1	1	3	1
14	861896	5years 6months	2	2		5	1	1	1				1	2	2	2	2	5	1	3	1	1	1

HAQL32	HAQL4	HAQL51	HAQL52	HAQL53	HAQL54	HAQL55	HAQL56	HAQL57	HAQL58	HAQL61	HAQL62	HAQL63	HAQL64	HAQL71	HAQL72	HAQL73	HAQL74	HAQL75	HAQL81	HAQL82	HAQL83	HAQL84	HAQL85
rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	12-16 ages					8-16 years		17 years		
1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score
2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)
3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)
4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)
5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)
														5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)
1	5	5	5	5	5	5	1	1	1	5	1	1	1			1	1	1					
1	1	1	3	1	1	1	1	1	1	1	1	1	1			1	1	1	1	1			
2	2	5	5	3	5	5	1	3	3	1	1	1	1			1	1	5					
1	4	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1	3	4			
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1	1	1	1	1	2
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	1	5	5	5	1	1	1	1	1	1	1	1	1			1	1	1					
2	2	2	2	1	1	1	1	1	1	1	1	1	1			1	1	2					
1	2	1	1	2	2	1	2	1	1	1	1	1	1			1	1	1					
1	2	1	1	2	1	2	2	1	1	1	1	1	1			1	1	1					
1	4	1	2	2	1	1	1	1	1	1	1	1	1			1	1	1					
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
2	3	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	1	1	4	4	2	2	5	3	2	1	1	1	1			1	1	1					
1	1	5	1	1	1	3	1	1	1	1	1	1	1			1	1	1					
2	1	2	3	2	2	2	1	1	1	1	1	1	1			1	1	1					
1	1	3	3	2	1	1	1	1	1	1	1	1	1			1	1	1	1	1			
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
2	5	1	1	2	1	1	1	1	1	1	1	1	1			1	1	1	1	2			
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	3	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	1	5	2	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	3	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
2	1	1	2	1	2	2	1	1	1	1	1	1	1			1	1	1					
1	1	1	1	1	1	2	1	1	1	1	1	1	1			1	1	1					
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1	1	1			
1	1	1	1	2	2	1	1	1	1	1	1	1	1			1	1	1					
1	3	5	1	3	1	1	1	1	1	1	1	1	1			4	5	5	1	5			
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
1	3	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1					
2	1	1	2	1	2	2	1	1	1	1	1	1	1			1	1	1					
1	1	1	1	2	2	1	1	1	1	1	1	1	1			1	1	1					
1	1	1	1	1	1	1	1	1	1	1	1	1	1			1	1	1	1	1			
2	1	1	2	1	2	2	1	1	1	1	1	1	1			1	1	1					

HAQL86	HAQL87	HAQL88	HAQL89	HAQL91	HAQL92	HAQL101	HAQL102	HAQL103	HAQL104	HAQL105	HAQL106	HAQL107	HAQL108	HAQL109	HAQL111	HAQL112
rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	>17 years	
1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	rawscore/0-100 scale score	rawscore/0-100 scale score
2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	1- (0/100)	1- (0/100)
3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	2- (1/75)	2- (1/75)
4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	3- (2/50)	3- (2/50)
5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	4- (3/25)	4- (3/25)
															5- (4/0)	5- (4/0)
1	1	1	1	1	1	3	5	1	5	5	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	2	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
4	4	2	1	1	1	2	1	1	4	2	4	3	2	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	3	2		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	2	1	1	3	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	2	1	1	1		
1	1	1	1	1	1	3	4	1	1	1	1	1	1	1		
2	2	1	2	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	2	2	1	1		
1	1	1	1	1	1	1	2	1	1	1	1	1	1	1		
1	1	1	1	1	1	5	1	1	1	1	2	1	3	1		
1	1	1	1	1	1	1	2	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	4	1	2	1		
1	1	1	1	1	1	1	1	1	1	1	2	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	5	1	1	1	1	1		
1	1	1	1	1	1	3	2	1	2	1	1	1	1	1		
1	1	1	1	1	1	3	5	1	5	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	3	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	2	1	1	1	1	1	1	3	3		
1	1	1	1	1	1	1	1	1	3	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	3	1	1	1		
1	1	1	1	1	1	2	1	1	1	1	1	1	3	3		
1	1	1	1	1	1	1	1	1	1	1	1	1	1	1		
1	1	1	1	1	1	1	1	1	1	1	3	1	1	1		

Sl no	op no	Age	Sex	Consanguity	degree of Consanguity	presenting illness	diagnosis	HAQL 11	HAQL12	HAQL21	HAQL22	HAQL31	HAQL32	HAQL4	HAQL51	HAQL52
		(in years)	1-male	1.yes	1- I st degree	1- fever	1.conjunctivitis	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score
			2-female	2.no	2- 2nd degree	2- cold	2.tonssiltis	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)
					3- 3rd degree	3-sore throat	3.respiratory tract infection	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)
					4-4rth degree	4- cough	4.followup	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)
						5-no complaints		4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)
								5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)
1	6723580	3years6months	2	2		1,2	1	1	1	1	1	1	1	1	1	1
2	6587906	6years	2	2		5	4	1	1	1	1	1	1	1	1	1
3	6811726	3years	2	2		5	4	1	1	1	1	1	1	2	1	1
4	6387383	6years 2months	2	2		3	2	1	1	1	1	1	1	1	1	1
5	6389008	6years6months	2	2		3	2	1	1	1	1	1	1	1	1	1
6	6773060	6years3months	2	2		3	2	1	1	1	1	1	1	1	1	1
7	6780485	4years2months	1	2		3	2	1	1	1	1	1	1	1	1	1
8	6780786	5years 11 months	2	2		5	4	1	1	1	1	1	1	1	1	1
9	6773981	5years 9months	2	2		3	2	1	1	1	1	1	1	1	1	1
10	585558	6years1month	2	2		3	2	1	1	1	1	1	1	1	1	1
11	5857511	7years1month	1	2		1,4	3	1	1	1	1	1	1	1	1	1
12	6762658	3years1month	2	2		3	2	1	1	1	1	1	1	1	1	1
13	6761979	3years4months	1	2		4	3	1	1	1	1	1	1	1	1	1
14	6789976	4years3months	1	2		1,4	3	1	1	1	1	1	1	1	1	1
15	6790900	3years2months	1	2		2,4	3	1	1	1	1	1	1	1	1	1
16	6790288	4years6months	1	2		4	3	1	1	1	1	1	1	1	1	1

HAQL53	**HAQL54**	**HAQL55**	**HAQL56**	**HAQL57**	**HAQL58**	**HAQL61**	**HAQL62**	**HAQL63**	**HAQL64**	**HAQL71**	**HAQL72**	**HAQL73**	**HAQL74**	**HAQL75**
rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	12-16 ages				
1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score
2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)
3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)
4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)
5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)
										5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1
1	1	1	1	1	1	1	1	1	1			1	1	1

HAQL81	HAQL82	HAQL83	HAQL84	HAQL85	HAQL86	HAQL87	HAQL88	HAQL89	HAQL91	HAQL92	HAQL101	HAQL102	HAQL103	HAQL104
8-16 years		17 years			rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score
rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)
1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)
2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)
3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)
4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)
5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)										
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1
					1	1	1	1	1	1	1	1	1	1

HAQL105	HAQL106	HAQL107	HAQL108	HAQL109	HAQL111	HAQL112
rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	rawscore/0-100 scale score	>17 years	
1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	1- (0/100)	rawscore/0-100 scale score	rawscore/0-100 scale score
2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	2- (1/75)	1- (0/100)	1- (0/100)
3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	3- (2/50)	2- (1/75)	2- (1/75)
4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	4- (3/25)	3- (2/50)	3- (2/50)
5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	5- (4/0)	4- (3/25)	4- (3/25)
					5- (4/0)	5- (4/0)
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		
1	1	1	1	1		