POSTERIOR SAGITTAL REPAIR OF ANORECTAL MALFORMATIONS IN KENYATTA NATIONAL HOSPITAL

TOPIC:

BOWEL FUNCTION FOLLOWING POSTERIOR SAGGITAL REPAIR OF ANORECTAL MALFORMATIONS

(A TEN-YEAR RETROSPECTIVE STUDY)

BY

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DECLARATION

This dissertation is my original work and has not been presented for a degree in any other university

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DEDICATIONS

This thesis is dedicated to God for His excess grace for me during the entire course of M. Med. Surgery and to my beloved parents Francis and Evah Kigo who have always been a source of strength, encouragement and advice from the time I was a small boy till now.

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"In general, atresia of the rectum is more poorly handled than any other congenital anomaly in the newborn. A properly functioning rectum is an unappreciated gift of the greatest price. A child so unfortunate as to be born with an imperforate anus may be saved a lifetime of misery and social seclusion by the surgeon who with skill, diligence, and judgment, performs the first operation on the malformed rectum."

Potts (1959) description of repair of infant born with anorectal malformations

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LIST OF ABBREVIATIONS

KNH	Kenyatta National Hospital
ARM	Anorectal Malformations
CS	Currarino Syndrome
PSARP	Posterior Sagittal Anorectoplasty
PSARVURP	Posterior sagittal Anorectovaginourethroplasty
PSAP	Posterior Sagittal Anoplasty
NVD	Neurovesical dysfunction
SHH	Sonic hedgehog
VBM	Voluntary Bowel Movement
GIT	Gastrointestinal tract
GUT	Genitourinary tract
MRI	Magnetic Resonance Imaging
Prep	Preparation
Post-op	Post-operative
Pre-op	Pre-operative
IAS	Internal Anal Sphincter

1. SUMMARY

Posterior sagittal repair was used to repair 352 patients with anorectal malformations between 1st January 1990 and 31st December 1999. We evaluated 193 patients who were over 3 years of age and with an adaptation period of 6 months after the colostomy closure for bowel control.

Males were 80 (41.5%) and females were 113 (58.5%), low anomalies were 86.5% and high anomalies were 13.5%, and while sacral defects were present in 14.0%. Colostomy was fashioned in 92.2% of the patients before definitive surgery, while only 7.8% were managed without a colostomy. More than half of the patients had their colostomies fashioned before the age of one month. Majority of males (97.1%) had colostomy fashioned before the age of one month, while 66.7% of females had their colostomy fashioned after the age of one month. The overall mortality was 1.0%.

The type of colostomies fashioned was divided sigmoid in 75.8% and transverse loop colostomy in 24.2% of the patients. Approximately 17.0% of these colostomies were revised. Transverse loop colostomy was revised in 58.1%of the patients compared to only 3.7% in divided sigmoid colostomy. Majority of the patients (90.5% of the patients) had to wait longer than 6 months for definitive surgery after colostomy fashioning.

Majority of the patients in this series had PSARP in 76.7%of the patients, while 17.6% and 3.1% had anoplasty and a combined PSARP and abdominal approach respectively. By the age 5 years 93.3% of the patients have had definitive surgery while only 10.9% by age of 6 months. Only 5.6% wait for less than 6 months colostomy closure after definitive surgery.

Overall voluntary bowel movement (VBM) is achieved in 71.5% of the patients. Patients who had colostomy fashioned before the age of one month achieve VBM in 79.1% of the patients while those fashioned after one year in 61.1%. Overall females achieve VBM in 77.0% of the patients compared to 63.8% in males. Patients with perineal fistula achieve

VBM in 79.1% in males and 75.0% in females, 76.0% in rectovestibular fistula, 73.9% in rectourethral fistula, 56.0% in anorectal malformations without a fistula, 25.0% in vaginal fistula and 12.5% in rectovesical fistula.

Overall where anoplasty was the definitive surgery VBM was achieved in 82.4% of the patients and 74.3% in PSARP of the patients. The patients who had definitive surgery before the age of 6 months achieved VBM in 81.1% compared to 61.5% when done after 5years. Overall patients with sacral defects achieve VBM in 25.9% compared to 78.9% in the patients with normal sacrum. The patients with low anomalies achieved VBM in 75.4% compared to 46.1% in high anomalies.

Soiling was present in 21.2% of the patients. Patients with high anomalies and sacral defects e.g. rectovesical fistula and rectovaginal fistula have high incidence of soiling. Constipation is present in 7.3% of the patients and low (simple) anomalies have the highest incidence.

In this series 24 (12.4%) of the 193 patients underwent secondary operations. Patients who had one redo operation were 21(87.5%) while 3(12.5%) of the patients had more than one redo operations. Indications for reoperations were severe fecal incontinence and severe constipation among others. All patients for redo operations due to severe constipation had normal sacrum. The patients with normal sacrum and low anomalies required one redo operation while some of the patients with high anomalies and sacral defects had more than one redo operations.

All the patients who had one reoperation had good to fair results while all the patients who had more than one redo operations had fair to poor bowel control results. All patients with severe constipation had normal sacrum and had significant improvement after one redo operation. Sacral defects in the patients with severe fecal incontinence were present in 91.7% of the patients. Where severe fecal incontinence was the indication for redo operation, significant improvement was in 50.0% of the patients while 33.3% and 16.7% of the patients had fair and poor results respectively.

2. INTRODUCTION

The posterior sagittal approach for the correction of anorectal malformations was first described 19 years ago and has since become the gold standard for the surgical repair of anorectal anomalies[1]. The exposure facilitates accurate placement of the rectum within the pelvic muscle complex and allows the precise division and closure of the rectourinary or rectovaginal fistula. In all patients except those with perineal fistula a preliminary diverting colostomy is necessary. This approach has been adapted for the repair of cloacal anomalies.

The functional results depend on the location of the associated fistula. More than 90% of the infants with fistulas extending to the perineum or distal urinary tract have voluntary bowel movements, as compared with only 15% of those with defects extending to the bladder neck[1]. Fecal incontinence is a serious problem that provokes[2-4] social segregation and psychological sequelae. Patients with ARM frequently suffer fecal incontinence despite the efforts of pediatric surgeons. At least 25-30%[5, 6] of them will suffer from fecal incontinence. In addition another 30% will suffer from other functional defeacation disorders such as constipation, occasional soiling, and fecal incontinence during periods of diarrhea[1-4].

Posterior sagittal repair is now the definitive management of children with ARM in Kenyatta National Hospital since 1987[7]. It has provided a new hope for the children with this congenital anomaly. The control of the bowel function previously provided the greatest challenge. Prior to 1987,anoplasty, cutback anoplasty and translocation anoplasty were the treatment of choice for the low anomalies, and abdominosacroperineal pull-through or sacroperineal anorectoplasty (and their modifications) were the preferred choice for intermediate and high anomalies. Since 1987 posterior sagittal repair has been adapted exclusively as the procedure of choice for correction of the entire spectrum of ARM[7].

3. LITERATURE REVIEW

HISTORY OF REPAIR OF ANORECTAL MALFORMATIONS

Anorectal malformations have been recognized and managed since antiquity (e.g. by Aeginata of Greece). In 1835, Ammussat was the first person to open then the rectal pouch and suture it to the skin (possibly a low anomaly). Chassaignac, in 1856 performed the first colostomy for the treatment of anorectal malformation. Hydra, in 1886, performed the first abdominoperineal procedure. The first part of this century, most surgeons used a preliminary colostomy and an abdominoperineal pull-through for the treatment of high anomalies, and a perineal approach without a colostomy for the 'low anomalies'. In 1933, Wangensteen and Rice described the invertogram and used as the criterion for determining the height of the anomaly. In 1948, Rhoad and colleagues reintroduced a neonatal abdominoperineal procedure[7, 8].

In 1953, Stephens proposed an initial sacral approach followed by an abdominal operation preserving puborectalis for fecal incontinence. Stephens', approach, resulted in considerable improvement over results obtained by the Rhoad's abdominoperineal operation. However, there has remained a disturbingly high incidence of both childhood incontinence and postoperative prolapse following these procedures. Exposure through the Stephens' sacrococcygeal approach is quite limited, the presacral plexus are at some risk, particularly in cases of congenitally ectatic terminal bowel, meticulous dissection of the posterior capsule of the prostate, genital, and nerve structures is difficult if not precluded. In 1967, Kiesewetter continued the Stephens procedure, and performed sacroabdominoperineal procedure[7, 8].

In the past, exploration for a blind pouch and performance of an anoplasty through limited perineal incision resulted in persistence of a recto-genito-urinary tract fistula and poor results from damage to the nerve and muscle structures. Therefore, in the past few

decades, a perineal approach to the correction of all but the lowest anorectal anomalies had been abandoned. Stephens' view that the puborectalis portion of the levator musculature constituted the only potential sphincter available for the continence following pull-through operations, led to his supralevator, sacrococcygeal approach for the mobilization of the terminal bowel and transsection of a fistula. It allowed for the blunt creation of a small rectourethral translevator space through which the bowel could be pulled to a perineal incision for the creation of an anus.

In 1980, Pena suggested that these malformations could be approached through posterior sagittal incision, using an electrical stimulator to identify the striated muscle structures. This is now the most preferred method of treatment, with the use of a protective colostomy (excepting the low anomalies); this technique revealed important pelvic anatomy that previously was a matter of speculation[7-9].

Pena's subcoccygeal sagittal exposure, like Kiesewetter's technique, split the dorsal portion of the levators and allowed Pena to accomplish a pull-through procedure in a large majority of cases without resorting to a secondary abdominal approach for colonic mobilization. Nonetheless, a blind pulling of the bowel through a blindly and bluntly created retrourethral space, inherent in all of the above procedures, precludes any precise identification of infralevator musculature[7-9].

NORMAL ANATOMY AND PHYSIOLOGY

A. Continence

Continence is maintained partly under voluntary control by the striated muscles of external sphincter and levator ani (under somatic nerve supply) and partly by the autonomic nervous system by the smooth muscles of internal sphincter[4, 8, 10]. In normal children anal canal is exquisitely sensitive to pain, touch, cold, pressure or tension[4]. The rectum (above the anal canal) is not sensitive to all these[4]. It is sensitive to distension.

Internal sphincter is maintained in a state of near maximal contraction at all times; its major reflex response to rectal distension is relaxation. It controls 85% of the resting tone of the anal canal; its inappropriate relaxation is responsible for most cases of leakage and soiling[4, 8, 10].

External sphincter (para-sagittal muscle fibers) enables voluntary control of continence. Levator ani and external sphincter blend to become indistinguishable, forming a funnel-shaped continuum of muscle. Levator ani and puborectalis forms a muscular diaphragm in the pelvic floor. The external sphincter and levator ani are integrated with the vertical fibers that run parallel with the rectum, called the muscle complex[4, 8]. The muscle complex, levator ani, and external sphincter are indivisible structures working in concert. The contraction of the levator ani is coordinated with that of external sphincter resulting in an effective closure of the anal canal[4, 8].

The reflex response to stimuli (produced by voluntary effort, posture change, rectal distension, increased intra-abdominal pressure, and anal dilatation) of external sphincter is contraction. This response involves several different neural pathways[4, 8]. The nerve supply to the internal sphincter (motor) is hypogastric nerves (sympathetic outflow) and inhibitory supply from parasympathetic outflow. The somatic pudendal nerve S_2 supplies external sphincter. Multiple direct branches recruited from the sacral S_{2-4} supply the levator ani and the puborectalis muscles[8].

B. Defeacation

Distal rectum is normally maintained empty and in a collapsed state. The maintenance of continence and the mechanism of defeacation depend on interaction of neurosensory and neuromotor impulses. As feces accumulate in the rectum, bowel wall muscles relax, allowing distension and accommodation of the enlarging fecal mass. Sensory receptors within the anal canal determine the nature of luminal contents i.e. flatus, liquid, solid stool[8].

Distension of the rectum is the stimuli for the defeacation. As the stools enters the rectum, the internal sphincter relaxes and the external sphincter contracts. With voluntary inhibition of external sphincter contraction during mass peristaltic propulsion, defeacation occurs without effort or gas will be passed selectively. Contraction of the external sphincter removes the urgency to defeacate unless volume is large or individual has impaired sphincter mechanism[8].

If a voluntary effort is required to defeacate, intra-abdominal pressure is increased by closure of the glottis and by contraction of muscles of pelvic floor. The resultant relaxation of the pelvic muscles produces straightening of previously angulated rectum. Closure of the anal canal by the sphincters allows an increase of the pressure within the rectum so that subsequent sphincter inhibition results in expulsion of stool. At the end of defeacation, when straining is discontinued, the pelvic floor rises to normal position and obliterates the lumen[8].

ANATOMIC AND PHYSIOLOGIC ABNORMALITIES IN ANORECTAL MALFORMATIONS

Children with ARM possess a heterogeneous quantity of anorectal sphincteric muscle, sensory nerves and proprioceptive fibers, and abnormal rectosigmoid motility. Varying degrees of the external sphincter muscle and the levator musculature development add to the heterogeneity of the anomalies[3].

The sensation is impaired in most children who are born with ARM (except for cases of rectal atresia), because, in normal individuals, exquisite sensation resides in the area of the anal canal. Those children born with ARM, a feeling of fullness (proprioception) is associated with a spectrum of exquisite to no sensation at all[3-5]. The heterogeneity of the nerve endings for both perineal sensation and proprioception is associated with the spectrum of voluntary muscle present. Symmetrical relocation of neoanus between whatever striated muscle exists is important for this distension sensation[3].

Rectosigmoid dysmotility is variably present in children who have ARM[3, 11, 12]. Post-operative constipation if untreated, leads to megasigmoid colon. In extreme cases, in which the constipation has been left untreated, fecal imparction develops as well as encopresis or overflow pseudoincontinence[1, 3, 10]. At the other end of the spectrum, the child who has lost rectosigmoid has no reservoir capacity and suffers from severe fecal incontinence[3].

INCIDENCE AND PATTERN OF OCCURENCE

According to estimated figures (mainly Caucasian) anorectal malformations occur in 1/4000 newborns babies[5, 7, 8]. The most common defects seen in males and females are anorectal anomalies with a recto-urethral fistula and recto-vestibular fistula respectively. The estimated risk of a couple having a second child with an anorectal malformation is approximately 1%[5]. Males suffer this condition more frequently than females[5]. Children with Down's syndrome have increased incidence of ARM, most frequently without a fistula[1, 3, 5]. Anorectal malformations without a fistula occurs in 5% of all patients with ARM and in 95% of these also have Down's syndrome. The prognosis is good, with one series reporting 80-96% of patients having voluntary bowel movement[13].

Children with ARM have associated vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies. The incidence of associated genitourinary anomalies is directly associated with the severity of ARM as defined by the level of the rectal fistula. The close association of development (embryological) of GIT and GUT systems explains the coexistence of abnormalities in both systems.

Overall incidence of lumbosacral anomalies in the patients with ARM ranges from 30% to 44% but more common in high (48-54%) than low anomalies (15-18%) [14, 16]. Patients may present with obvious spinal anomalies at birth or neurological deficit after infancy. Important associated defects include tethered cord, and presacral masses (e.g., anterior meningocele, lipoma, dermoid, teratoma). Tethered cord occurs more frequently in

patients with severe anorectal defects, sacral hypodevelopment, myelodysplasia, presacral mass, sacral hemivertebrae or a single kidney or in those with an anorectal defect with poor functional prognosis[15]. The continence problems associated with tethered cord in ARM could be due to an associated sacral anomaly. The tension induced in the terminal portion of the cord results in vascular insufficiency leading to neurologically mediated problems of bowel[16]. Experienced pediatric radiologists are able to diagnose these associated defects by the use of spinal ultrasound provided that it is performed during the first three months of life[17]. Routine CT-scan of spine for all newborns with ARM is recommended to avoid missing this condition. In the advent of MRI early detection and treatment of these anomalies is possible.

Spinal dysraphisms and neurovesical dysfunction (NVD) is a common association with ARM[18]. NVD is common despite normal spinal cord and is commoner in the patients with high anomalies and sacral anomalies. Bladder dysfunction does not appear to be sequelae of a properly performed PSARP[19]. NVD incidence is increased in the patients with high anomalies and in the patients with sacral abnormalities[20]. However presence of low anomalies or absence of sacral abnormalities does not eliminate NVD. Urodynamic investigations should be done where NVD is suspected[20]. Cases of Currarino syndrome (CS) present an association of ARM, hemisacrum, and presacral mass[21]. A gene responsible for CS has recently been mapped in the 7q36 region[21-24]. Among the genes localised in this critical region, sonic hedgehog (SHH) thought to represent a candidate gene for CS as well as for ARM with different levels of hypodevelopment according to its role in the differentiation of midline mesoderm[21]. One series report 38% incidence of the triad when there is anorectal stenosis[21].

Japanese Study Group to determine the relative incidence of specific types of anorectal anomaly in Japan studied 1,992 patients (male to female ratio 1.46: 1)[25]. High type of anomaly accounted for 26.0% of cases, intermediate10.7%, low 57.2%, miscellaneous 4.5% and unclassified 1.8%. The most frequent deformity was male anocutaneous fistula

(18.2%), followed by male rectourethral fistula (16.7%), and female anovestibular fistula (12.1%). The overall incidence of patients having one or more associated anomalies was 45.2%: 70.6% in high deformity, 60.7% in intermediate, and 31.3% in low. The rate of association of Down's syndrome with deformities without fistula (40.3%) was significantly higher than with deformities with fistula (0.3%).

Okumu in 1995 studied the pattern of occurrence of anorectal malformations in KNH[7]. In the 8-year study period 141 patients were seen and male to female ratio was approximately 1:1; the commonest malformations in boys and girls were anorectal malformations without fistula and rectovestibular fistula respectively. Posterior sagittal repair of anorectal anomalies as the definitive form of management was carried out in 75.8% of the patients before the second birthday. The overall complication rate following the repair is 16.2%. Seventeen percent of the patients had associated anomalies with cardiac, vertebral, and skeletal anomalies being the commonest. Alimentary tract anomalies that include tracheoesophageal atresia with fistula were the anomalies associated with the highest mortality. Only 1.2% had poor bowel control.

ANATOMIC CLASSIFICATIONS OF ANORECTAL MALFORMATIONS

Several classifications have been used in ARM. Gans and Colleagues proposed a classification based on the fact that the end of the bowel is almost always open and simply terminate in the wrong position[4]. Ladd and Gross classification has been used for many years. However, in 1970, an International classification was proposed, it was complex and rather impractical and was rarely used. In 1984, Stephens and Smith, proposed 'Wingspread classification' (appendix 1), it has embryological implications and was more practical. This classification placed groups of malformations within specified categories as well as 'high, intermediate, and low'[4]. This classification has now been proven to be arbitrary and inadequate[4].

Since posterior sagittal repair of ARM, it has become easier to directly expose the anatomy of each of these defects, a better definition of the complex arrangements between the rectum and the genitourinary tract and better and more objective way to reconstruct these defects. Pena proposed a classification (appendix 2) that solved the problem of placing groups of malformations within specified categories that confused the issue of risk for fecal incontinence. This classification is based on therapeutic and prognostic implications[1, 4]. It is easy to discuss results of surgical treatment, as well as compare results of anomalies with similar potential for bowel control. In this study, this is the classification that is used.

DEFINITION OF EACH DEFECT

Perineal fistulas in both males and females patients the rectum opens in a small orifice, usually stenotic and always located anterior to the center of the sphincter. A normal sphincter does not surround this abnormal opening. This defect can be treated during the newborn period with an anoplasty and without a colostomy. Sphincteric mechanism and the sacrum are usually good. The incidence in KNH is 5%[7].

Anorectal anomalies with rectourethral fistula are present in 50% of cases in males (10.3% in KNH[7]). The fistula opens in the lower posterior urethra (bulbar) or in the upper posterior urethra (prostatic). Patients with rectourethral bulbar fistula usually have good sacrums and sphincters. The patients with prostatic fistula have a higher incidence of sacral hypoplasia and poor sphincters.

The patients with rectobladder neck fistulas represent 10% of all males (4.3% in KNH[7]). This is the only defect in males that require a laparatomy in addition to posterior sagittal approach. These patients usually have both sacrum and Sphincteric mechanism poor. Anorectal anomalies without a fistula in both males and females patients represents approximately 5% of cases; the rectum is located approximately 2cm above the skin of perineum. These patients have good sacrum and muscles. This defect is particularly

associated (in 95% of cases) with Down's syndrome[13]. The incidence in KNH 22.7%, and the association with Down's syndrome have not been studied[7].

Rectal atresia (or rectal stenosis) in both males and female patients, are born with an intact, although rather small, anal canal, with a normal sacrum and sphincter mechanism. There is an area of atresia located between the anus and the rectum. The incidence is about 1% of all cases.

Rectovestibular fistula is the most frequent defect seen in females. The incidence is approximately 25% (41.8% in KNH[7]). In these patients, the rectum opens in the vestibule, the space between hymen and perineal skin. Most of these patients good sacrum and a well developed sphincter mechanism.

In persistent cloaca, the rectum, vagina, and urethra are fused together into a common channel opening in the perineum. About 90% of the patients have an associated urological defect that include hydronephrosis, megaureters, reflux, renal dysplasia, renal agenesis, and neurogenic bladder. The degree of sacral anomalies and muscle deficiencies vary depending on the height of the defect. About 40% to 50% require abdominal approach in addition to the posterior repair to mobilise a high rectum or vagina (common channel more than 3cm)[1]. More than 50% have hydrocolpos compressing the trigone and interfering with emptying of the ureters[7, 26]. Those that have a common channel less than 3cm can be repaired posterior sagittal approach only[26]. This latter group have functionally better prognosis. The overall incidence in KNH is 1.4%[7].

INITIAL MANAGEMENT

Males

In approximately 80-90% of patients, clinical evaluation and urinalysis (rules out rectourinary fistula) provide sufficient information to decide whether a colostomy is necessary[3-5, 8]. The presence of a perineal fistula, "bucket-handle" anomaly, sub-epithelial

midline raphe fistula, anal membrane, and anal stenosis indicates that the anomaly (low anomaly) can be repaired with an anoplasty, without a protective colostomy[8].

If none of the above clinical signs become evident in 24 hours, an invertogram is indicated[5]. If the rectal-skin distance is more than 1cm represents an indication of a colostomy, otherwise anoplasty is enough. A flat perineum or presence of meconium in urine is considered indications of a colostomy. It must be assumed that these patients have high anomalies or rectourinary fistula[5, 8].

Females

A meticulous perineal inspection is important to make the correct diagnosis. The presence of single orifice with a palpable pelvic mass is indicative of a persistent cloaca. These children have a high chance of an associated urological defect and an emergency urological evaluation is required. A colostomy, urinary diversion (vesicostomy) and a vaginostomy are done at the same operation[8]. The definitive management is done at least 6 months thereafter[8].

The finding of a perineal fistula, indicate that the anomaly can be repaired with an anoplasty without a protective colostomy. Careful perineal inspection may reveal vestibular or a vaginal fistula. These patients require a protective colostomy in the neonatal period (a delay to fashion a colostomy provokes magarectum due to constipation)[5,8].

After 24 hours of observations, a baby's abdomen becomes distended, yet there is no evidence of meconium passing through the genitalia, such a baby requires an invertogram and is managed as in males[8].

DEFINITIVE PRIMARY MANAGEMENT

The posterior sagittal repair is performed as part of three-stage procedure in management of ARM, the colostomy is fashioned in the neonatal period, posterior sagittal repair done at least three months later and at least three months thereafter the colostomy is closed[1].

Colostomy

The descending colon colostomy is recommended in primary management of ARM, fashioned with two separate stomas for complete diversion of stools, rarely does it permit the passage of stools from the proximal stoma to the distal bowels which provokes potential urinary tract infections and impaction stools in the distal rectal pouch, it also has low prolapse rate compared to the other types of diversions[4, 5, 8].

The mechanical preparation for distal colon prior to definitive repair is easier. It should not be created too distal where mobilization of the rectum during the main repair is difficult. The more distal the colostomy is fashioned the more easily it allows the urine to escape into the colon in case of a large recto-urinary fistulas[4, 5,8]. When urine is absorbed leads to metabolic hyperchloraemic acidosis[4].

The colostomies in children are easily managed without colostomy bags in KNH[7] and a simple change of diapers is sufficient. Patients operated without a protective colostomy, many of them suffer from wound dehiscence (post-operative sepsis) and recurrence of the fistula[1].

Colostography: Prior to definitive repair, distal pressure colostogram is desirable[1, 4, 5]. Contrast under hydrostatic pressure is pushed through distal part of bowel. The lower the defect the more the hydrostatic pressure applied[4, 8] to fill up colon. The contrast material fills the proximal urethra and the urinary bladder through the fistula. The location of fistula, length of distal colon available for pull-through, state of urinary bladder and urethra, vesico-ureteric reflux (if present) can be demonstrated[1, 5, 8].

MAIN POSTERIOR SAGITTAL REPAIRS

A. Minimal posterior sagittal anoplasty (PSAP)

Perineal fistulas are repaired with a small anoplasty, without a protective colostomy[5]. The aim of the operation is to dissect the rectum sufficiently (usually 1-2cm) to move it posterior and place it within the limits of the external sphincter. Functional prognosis is excellent; problems occur only when the operation is inadequate or patient does not receive adequate medical management for constipation[4, 5].

B. Posterior sagittal anorectoplasty (PSARP)[9]

Pena and de Vries popularized the procedure in 1982. The fundamental principle underlying this operation is obtaining fullest possible exposure of all structures while keeping to the midline. All patients should have a functioning completely diverting divided sigmoid colostomy. The pelvic musculature and the neurovascular supply do not cross the midline and therefore a midline incision does not damage these structures. The operation is done under general anaesthesia with the patient first catheterized in the supine position. The patient is then placed in prone jackknife position and the electrical stimulation of the perineum is then done which allows the surgeon to identify the appropriate anal site and map it. This is determined by observation of contractions of sphincteric musculature.

Following prep and draping, the midline skin incision is made from the mid sacrum through the anal site (center of external sphincter) to the perineum 1-2 cm anterior to it. In order to incise the sphincteric musculature in the midline and to identify clearly the components of the sphincteric musculature, the electrostimulator is used in the course of dissection to the supralevator terminal bowel. High intensity stimulation (100-240mA) may be necessary in the cases of older patients with significant amounts of scar tissue. Much lower intensity (20-40mA) suffices in primary operations, particularly when stimulation is directed to the muscle. The coccyx is sagittally split after dividing the insertion of the deep external sphincter into lateral halves (this is no longer necessary).

The levator ani and its insertion on the ventral aspect of the coccyx and /or sacrum, is split in the midline and the incision carried down to the longitudinal smooth muscle coat of the terminal bowel. The bowel is mobilized by sharp dissection from the adherent levator muscle fibres in order to preserve any puborectalis fibres and avoid damage to the surrounding nerves and ganglia. If a vesicle, prostatic, or bulbar fistula is present or suspected, the terminal bowel is opened just dorsal to the fistula. In the prostatic area, only the submucosa is mobilized leaving the muscular coats on the prostatic capsule to avoid damage to the nerves and genital structures. Following transsection of a fistula to the urethra, bladder, or urogenital sinus, the mucosa and muscles are closed. The full thickness of the bowel wall is mobilized to a position above the level of the pelvic peritoneum so as to obtain a satisfactory length of bowel. When opened, the peritoneum should be closed before the operation is finished[27]. Enough rectal length must be gained for comfortable, tension-free anastomosis between rectum and skin.

If the terminal bowel is dilated and hypertrophic (congenital ectasia), the bowel is tapered (tailored) by wedge resection. This is done to accommodate the rectum within the limits of the muscle structures (muscle complex and the external sphincter). The mucosa-submucosa and the smooth muscle coat of the narrowed bowel tube are approximated with 5-0 suture. The sagittally split complex of striated sphincteric musculature immediately dorsal to the urethra, composed largely of external sphincter and puborectalis portion of the levator, is first sutured laterally and dorsally to the bowel tube with interrupted 5-0 sutures. Further dorsally, the split striated muscle complex and external sphincter layers as well as split coccyx are re-approximated. The muscle coat of the bowel is sutured circumferentially to the surrounding external sphincter portion of the striated muscle complex and the mucosa-submucosa is sutured to the skin. The reconstruction is done to allow the rectum to pass in front of the levator, and is located immediately behind the urethra. The anoplasty is created with a small amount of rectal-skin tension that will help to retract the mucosa and prevent prolapse. The anoplasty is started by placing the four main cardinal sutures, and then with interrupted sutures to

complete 16 stitches. Neither excess circumference nor length of the bowel is left so as to prevent or minimize mucosal prolapse. The anus and anal canal are reconstructed so as to allow for initial passage of at least Hegar's dilator size 10. The new anus must be created in the center of the external sphincter, with the caliber no greater than 5-12mm.[27]

The technique described is used to repair rectourethral fistula (prostatic fistula has a higher chance of underdeveloped muscles), vestibular fistula and low vaginal fistula. The rectobladder neck fistula and high rectovaginal fistula, the posterior sagittal approach includes a laparatomy. In cases of rectal atresia, sphincteric mechanisms are divided in the midline as above; the upper rectal pouch and the short distal anal canal are opened. An end-to-end anastomosis performed under direct vision. In the cases of rectovestibular fistula in females, the incision continues in a "racket-like" fashion, the most delicate part of dissection being the separation of the rectum from the vagina, as these structures share a common often-thin wall. Once the dissection is complete the rectum is mobilized and placed within the limits of the sphincteric mechanisms

C. Posterior Sagittal Anorectovaginourethroplasty (PSARVUP)

The management of a persistent cloaca is the most serious technical challenge in pelvic pediatric surgery[4, 5, 26]. The surgeon must be prepared to find bizarre anatomical arrangement of rectum, vagina and urethra. The main aims of the operation are: To separate the rectum from the vagina and the vagina from the urinary tract, to place the rectum within the limits of the sphincter, and to open the vagina in its normal location and reconstruct the old common channel as a neo-urethra. How many of these aims are achieved depends on the degree of the muscle and nerve development.

Separation of the vagina from the urinary tract is the most important challenge. If the common channel is longer than 3cm, the surgeon will usually have to open the abdomen to mobilize the vagina and the very highly located rectum. If the common channel is shorter than 3cm, however the entire vagina and rectum may usually be mobilized

without opening the abdomen. Once the rectum has been separated from the vagina and the vagina from the urinary tract, the urethra is reconstructed using the old common channel. The vagina is then sutured to the perineum, creating a new introitus immediately behind the urethra. The perineal body is reconstructed and the rectum is pulled down and placed within the limits of the sphincteric mechanism. A suprapubic cystostomy catheter is left in place after a cloaca repair, except in cases with a very short common channel for a week[5].

Post-operative care

The anus and constructed anal canal are calibrated with Hegar's dilators and skin sutures are removed 2 weeks post-operatively. In all tapered cases, gradual gentle dilations are started around 2-3 weeks postoperatively, and must be performed twice daily by the parent. The size of the dilator used, which depends on patient's age, is increased until the rectum reaches the desired size, and continued at least 2 months with the idea of promoting circumferential growth leading to gradual enlargement of the constructed anal canal without tearing of the surrounding musculature. The colostomy is closed 3 months thereafter and the frequency of dilations reduced to: at least once a day for 1 month, every third day for a 1 month, twice a week for 1 month, once a week for 1 month, once a week for 3 months.

In the posterior sagittal approach, and particularly in patients with good sphincter, the neoanus is fashioned in the center of the external sphincter and, following surgery it (the neoanus) remains closed rather than patulous and open. If the anal orifice is not dilated it will heal closed, and the patient will suffer a severe, intractable stricture.

Moore[28]suggests performance of PSARP at birth, at this time the operation relieves the alimentary canal, eliminates urinary tract contamination at birth, establishes anorectal continuity and maximal potential for 'normal' defeacation reflexes at birth and achieves all this in one operation than three operations.

ASSESSMENT OF BOWEL FUNCTION

Restoring of normal bowel function is the goal of anorectal reconstruction in patients with ARM. The long-term fecal continence outcome in patients with these anomalies is good or socially acceptable in majority of patients. Approximately 25% of children suffer fecal in continence; another 25-30% will have other forms of defeacation disorders such as constipation, soiling and incontinence associated with episodes of diarrhea. Fecal incontinence depends on type of ARM, and the original reconstructive procedure. Several criteria have been used to assess bowel function. Kelly, Templeton, Kieseweter and Chang, Stephens and Smith, Wingspread, have advised clinical scoring methods, according to the degree of continence and the quality of life after management[29]. Kelly's method added the strength of puborectalis action on digital examination. Most have been used prior to the advent of PSARP, and did not use symptoms other than soiling or skin rash for evaluation.

Pena proposed a protocol (appendix 3) that includes post-op clinical situations, VBM, soiling and constipation. VBM is the most valuable sign of fecal control. It is therefore easy to compare malformations with similar potential for bowel control. This is the method for assessing bowel control used in the study.

CLINICAL RESULTS

The clinical results vary depending on the type of the ARM[1], age of patients[1, 30, 31], the length of period before fashioning of colostomy[7], type of sacrum[1, 3-5], and the type of primary definitive surgery[1].

The following are determinants of bowel function: -

Age of the patient at definitive surgery: Early definitive surgery, at least theoretically by placing the rectum in the right position early in life provide better functional results, new synapses are developed which provide better sensation and better function of the

sphincteric mechanism[1, 3, 31]. Pena[8] prefers to do PSARP at 4 weeks of age, and has reported better results.

Length of pre-colostomy period[1]: Many of those patients whose colostomy is not fashioned early in life (i.e. in cases of vestibular fistula), suffer severe constipation associated with rectal ectasia before the main repair. Chronic dilatation of any kind of a hollow viscus eventually produces some degree of permanent damage that results in primary hypomotility disorder and therefore poor peristalsis.

The type of anorectal anomaly[1, 5, 8]: High defects have severe under-development, abnormal sacrum and poor muscle development, and poor innervation of the pelvic structures, and therefore poor functional results. Low and simpler anomalies are associated with almost normal muscles and better prognosis for bowel control and have more of a tendency to have constipation than high defects that have poor prognosis for bowel control.

The type of sacrum: Sacral ratio[1, 4] is useful evaluation in terms of functional prognosis. Sacral ratio of 0.76 is the average ratio in normal children. Children with ARM show a spectrum of values; lower ratios represent different degrees of sacral hypodevelopment and are associated with defects that have poor functional prognosis.

Diarrhea is seen in children who had resection of their remaining rectum and a portion of their sigmoid colon. Constipation is a common sequela following repair[1, 4, 5, 32]. In the patients with ARM, constipation seems to be secondary to a primary hypomotility disorder of the rectosigmoid, possibly due to abnormal innervation of the rectal pouch and fistula used in the reconstruction[11, 12]. Long period before colostomy fashioning produces megasigmoid due to a primary hypomotility disorder leading to constipation. Abnormal distribution of colonic interstitial cells of Cajal (intestinal pacemaker cells) in children with ARM has been identified and leads to constipation[33]. Very severe

constipation may provoke encopresis and constant soiling, secondary to overflow pseudo-incontinence[1, 8].

Perineal fistula[1, 5, 8]: Repair gives excellent results, but patients usually throughout their lives if not adequately treated, they tend to develop megarectum and mega sigmoid, which may eventually provoke symptoms of overflow pseudo-incontinence. A good prophylactic treatment for constipation will prevent this; otherwise the patients have normal bowel function.

Recto-urethral fistula[1, 5, 8]: Patients achieve voluntary bowel movement (VBM) by 3 years of age in 60-70% of cases; approximately 50% of these patients suffer grade 1 soiling. Patients with this anomaly and with abnormal sacrum seldom achieve VBM and suffer grade 3 soiling.

Recto-bladder neck fistula[1, 5, 8]: Patients achieve VBM alone in approximately 20% of the patients, provided that the sacrum is normal. All patients with abnormal sacrum are incontinent.

ARM without a fistula[1, 5,8]: Approximately 70% achieve VBM; most have a normal sacrum, those exceptional cases with abnormal sacrum are incontinent, 50% have grade 1 soiling that is related to constipation.

Rectal atresia[1, 5,8]: Patients will experience100% control.

Rectovestibular fistula[1, 5, 8]: Over 90% have VBM provided the sacrum is normal and 20% suffer soiling usually associated with constipation.

Persistent cloaca[1, 5, 8, 26]: Prognosis varies depending on the type of sacrum, the length of the common channel and quality of muscles. More than 80% of patients with a channel

shorter than 3 cm and a good sacrum achieve VBM. Those with an abnormal sacrum or a common channel is longer than 3 cm suffered varying degrees of incontinence depending on the degree of sacral dysplasia.

POST-OPERATIVE EVALUATION OF FECAL INCONTINENCE

The evaluation should begin with a review of the type of defect and operation performed. A rectal examination should be performed to evaluate sphincter tone and the position of neoanus and the contour of the buttock folds. Previous X-rays should be re-evaluated, specifically searching for lumbosacral spine defects and calculating the sacral ratio. A pathologist should review specimens of bowel for the presence of ganglion cells. Associated anomalies should be documented. A contrast study of the remaining pulled-through bowel is helpful because it defines the anatomy, and also the magnitude of dilated bowel[3].

Magnetic resonance imaging (MRI) defines the position of the neoanus in relation to the striated muscle (both para-saggital fibers of the external sphincter and the more vertical fibers of the muscle complex). Tethered cord should be ruled out (presence of myelodysplasia, abnormal sacral ratio, complex defects, cloacal extrophy are highly indicative) in patients with ARM, in cases of incontinence associated with bladder dysfunction and orthopedic disorders[3].

SECONDARY OPERATIONS

The colostomy for secondary operations is fashioned on the transverse colon on the right side of the middle colic vessels [1].

Secondary operations[1-3, 8] are performed in patients who fall into one of the following categories: Patients suffering from fecal incontinence subsequent to a repaired anorectal malformation (e.g. patients with mislocated rectum with good muscles and sacrum), and patients who underwent an attempted repair of an anorectal defect that failed and suffer

post-operative complication that include complete dehiscence of the pulled through rectum, stricture or recurrence of a fistula.

The ideal candidates for secondary operations are those who have very clear clinical or radiologic evidence of a mislocated rectum with evidence of good muscles and a good sacrum and patients with good prognosis type of defect[1, 4-7, 9]. The reconstruction consists of: Tapering the rectum as much as possible to make it fit into the levator mechanism, muscle complex and the external sphincter, and repair the lower portion of the levator muscle behind the rectum, secondly, relocation of the rectum into muscle complex and suturing this muscle structure around it, thirdly, repair of the perineum at the site where the rectum was originally placed and relocation anus at the center of external sphincter, and then, in laterally placed anus with the destruction of the lateral portion of external sphincter, the creation of neo-anus at the center of preserved part of external sphincter making a tunnel through it, is done.

Functional results: In terms of bowel function, the results of the patients subjected to secondary repair as a sequelae of complication suffered in primary operation are slightly inferior to the ones achieved in the same defect with a primary operation[4, 5]. Patients with normal sacrum, who underwent secondary operations for the treatment of fecal incontinence, achieved marked improvement in 45% of the cases, some improvement in 37%, and no improvement in 18%. In contrast to those with abnormal sacrum who have 20,30 and 50%, respectively[4, 5].

INVESTIGATIONS

X-rays studies are sometimes useful when the clinical impression is unclear. The upside down lateral film of the pelvis with the baby inverted is an inaccurate method of establishing the lower extent of the rectum, because the swallowed air may not have completely displaced the meconium from the rectum, or the striated muscle complex may be contracted, which obliterates the lumen and makes it look as if the gas in the rectum

ends high in the pelvis. With crying or straining, the puborectalis muscle and the may actually descend below the ischium, giving a falsely low estimate of rectal height. Gas in the bladder clearly indicates a rectourinary fistula.

Ultrasound: Trans-anal ultrasound shows the state and site of internal and external sphincter for post-operative evaluation of the patients with fecal incontinence and helps to select those who need reoperation. Endo-anal ultrasound[34] is able to identify internal and external anal sphincter muscles. However it has limited resolution and therefore difficult to reconstruct 3-D images.

CT-scan[7]**:** Pelvic CT-scan is useful in assessing the prognosis pre-operatively of continence. It shows the state of sphincteric muscular structures and the state of sacrum. Post-operative CT-scan demonstrates the location of the pulled-through rectum within the levator ani and the sphincteric muscular complex, the cases of incontinence associated with sacral anomalies and hypoplasia of the muscles have a poor prognosis. Therefore post-operative CT-scan is essential in selecting those incontinent patients who will need secondary posterior sagittal repair for the treatment of fecal incontinence.

Magnetic Resonance Imaging (MRI) : MRI is very useful in assessing the level of anal atresia, the extent of development of pelvic musculature and presence of associated anomalies all these greatly influence the outcome and further treatment options. In the newborns, MRI is important in assessing the level of atresia; meconium in the colon is hyper-intense under MRI, clearly demonstrating the extent of the malformation. In older patients with high atresia, MRI is useful in planning operative strategy and predicting outcome by providing useful information about the pelvic musculature which is directly associated to the size of the patient i.e. the larger the patients, the more the details the MRI provides about the pelvic musculature. In the secondary operations MRI is useful in determining the causes of the initial failure[2, 4, 9, 35-37] i.e. lack of pelvic musculature or poor placement of the distal colon to the striated muscle complex. It is also able

demonstrate unsuspected lesions e.g. tethered cord sacral deformities, presacral masses (anterior meningocele, intra-spinal lipomas, dermoid, teratoma).[22, 23]

SUMMARY

On the whole, one can expect 75%[1, 2, 5, 6] of all patients born with ARM and subjected to posterior sagittal approach will have VBM by the age of 3 years. This compares favorably with KNH[7] of 71.5%. Lack of VBM, soiling, and constipation can be predicted in a reasonably accurate way. Posterior sagittal approach provides a useful way to repair all types of malformations, only patients with very highly placed pelvic structures requires an additional laparatomy. The functional prognosis is important to determine, first, if the patients have poor prognosis, a bowel management programme is started early. False expectations to the patients should be avoided and if a patient has a good prognosis and is not toilet trained by 3 years age, explanation must be found. The answer lies in the presence of megarectum and megasigmoid, with overflow incontinence and presence of tethered cord, which must be ruled out[1, 10].

JUSTIFICATION OF THE STUDY

In KNH, approximately thirty patients undergo posterior sagittal repair as a definitive procedure for anorectal malformations every year[7]. Posterior sagittal repair has now been exclusively used for the correction of entire spectrum of ARM since 1987. Approximately 80%undergo posterior sagittal repair before the second birthday. The outcome results are generally considered excellent in 71.4% of patients. The mortality rate of 5%(1995) compared to 29.5%(1982) following repair has been observed. Some patients required multiple operations. The re-operation was necessary in 7.1% of the patients.

Bowel control previously provided the greatest challenge. Previous studies have only looked at the pattern of occurrence of ARM[7]. The outcome of bowel function following posterior sagittal repair as a primary or secondary procedure has not been critically evaluated in KNH.

The results of this study can be used as a database for planning of any future further management, setting up strategies for improvements and planning future follow-up of the patients.

OBJECTIVES

Broad objectives

To critically evaluate the outcome of bowel function following posterior sagittal repair of anorectal malformation in Kenyatta National Hospital between 1st January 1990 and 31st December 1999

Specific objectives

1. Determine whether the bowel function outcome of posterior sagittal repair for anorectal malformations is related to,

    a. The age of the patient at definitive surgery
    b. The length of period before fashioning of colostomy
    c. The type of anorectal anomaly
    d. The type of sacrum of the patients
    e. The length of time between the repair and the closure of colostomy

2. To determine the outcome following re-operations.

4. MATERIAL AND METHODS

Study design

This was a descriptive retrospective study over a period of ten years from 1st January 1990 to 31st December 1999.

Study population.

ALL the patients who sought treatment in KNH and had a diagnosis of anorectal malformation made and had posterior sagittal repair for ARM performed in KNH during the study period were included in the study. They were required to have been operated on by a qualified surgeon and to satisfy the eligibility criteria below.

Study area

The study was carried out in Kenyatta National Hospital. It is situated Nairobi the capital city Kenya (population of 2.5 million by 1989 census). It is the main referral and teaching hospital in Kenya. Six qualified pediatric surgeons manage the pediatric surgical unit in KNH. The pediatric surgical unit is the largest in any public hospital. It is the only pediatric surgical unit in a public institution where definitive management is carried out. In Nairobi it is supported by several private hospitals. Initial management e.g. colostomy fashioning is sometimes carried out in some provincial hospitals.

Sample size

The study population determined the sample size. This was based on the number of patients who presented to KNH with anorectal malformations and who had posterior sagittal repair performed for ARM and fitted in the admission criteria below.

To obtain 95% confidence interval for this proportion using a precision of 5%, the minimum size (n) is calculated to be 180 using the formula;

$n = z\ 1\text{-}\alpha / 2^2\ p\ (p\text{-}1)/d^2$

Where $z\ 1\text{-}\alpha / 2$ is the standard normal deviation corresponding to the level of significance of $\alpha = 0.5$.

P is the estimated proportion of patients who have had posterior sagittal repair for ARM

d^2 is the width of confidence interval

n the desired sample size

z is the standard normal deviation, which corresponds to the 95% confidence level

VARIABLES

Independent variables

The independent variables were: -

1. The age at colostomy fashioning
2. The age of the patient at definitive surgery
3. The type of anorectal anomaly
4. The type of sacrum of the patients
5. The level of anomaly
6. The length of time between the definitive repair and colostomy closure

Dependent variable

1. The bowel function

ELIGIBILITY CRITERIA

Inclusions

1. All patients attended to at KNH within the period of study and had had PSARP as a primary definitive procedure or as a re-operation.

2. All patients who have had posterior sagittal re-operative repair in KNH and had primary procedure outside of the institution

3. All children who have had posterior sagittal repair, over three years of age, toilet trained and had had their colostomy closed, and an adaptation period at least for six months

Exclusions

1. All patients not operated on by a qualified surgeon.
2. All patients who had posterior sagittal repair done outside of KNH and are or were being followed-up in KNH.
3. All children under three years of age
4. All children whose colostomies are still open
5. All children whose post-colostomy period is less than six months
6. All children who are mentally retarded
7. All children with complex malformations

DATA COLLECTION AND MANAGEMENT

Information about bowel function in anorectal anomalies was collected by the use of a pretested questionnaire. The principal investigator under the guidance of the supervisor collected all the data in this study. The case records of all the patients with ARM treated within the study period were retrieved from the department of records of KNH. Then information obtained in these records including details of clinical examination; radiological and operative findings were reviewed to determine the structural abnormalities, complications and outcome in these patients.

All patients with anorectal anomalies were grouped according to Pena's classification (Appendix 2). Continence was graded using the Pena's grading system (Appendix 3) by the investigator according to signs and symptoms recorded on the first visit six months after closure of the colostomy. Only patients who were over 3 years of age and toilet trained were evaluated. Patients with inability to control defecation voluntarily were considered incontinent, and those with various degrees of soiling including those with associated constipation were also considered incontinent.

After the data collection exercise, coding was done for all the questionnaires, data entry was then done into SPSS data sheet. Analysis of the data was done using SPSS computer program.

MINIMIZATION OF ERRORS

1. The data collection exercise was done by the principal investigator after a thorough pre-testing of the questionnaire

2. Guidance, clarification, and advice were sought from the supervisor throughout the study

3. The data analysis and management and typing of this manuscript was done by the principal investigator

ETHICAL CONSIDERATIONS

1. The proposal was submitted to the Ethical and Research committee of KNH and clearance was given after the standards required were met before data collection

2. All the information obtained was treated with utmost confidentiality and ONLY used for the intended purposes

3. All questionnaires and entry tables did not bear neither the name nor the ethnicity of the patients

5. RESULTS

This chapter presents the results of the study. The study was carried out between1st January 1990 and 31st December 1999 in KNH. Posterior sagittal repair was carried out on 352 patients and 193 of these patients were evaluated.

I. THE EXCLUSION CRITERIA

The 149 patients excluded from the study are shown in the table below. Records of 45 patients could not be traced, 52 patients were lost to follow-up, and 23 patients were less than six months after the closure of colostomy. Two patients died within the study period.

		Frequency
Evaluated		193
Excluded	< 6 months post-op	23
	Lost to follow-up	52
	Colostomy still open	27
	Files missing	45
	Mortality	2
Total excluded		149

Table 1: Exclusion criteria

II. CHARACTERISTICS OF THE PATIENTS

1. SEX DISTRIBUTION (n=193)

The table below (figure 1) shows the sex distribution of the patients evaluated. Males were 41.5% and females were 58.5% of the total.

	Frequency	Valid percent
Male	80	41.5
Female	113	58.5
Total	193	100

Table 2: Sex distribution

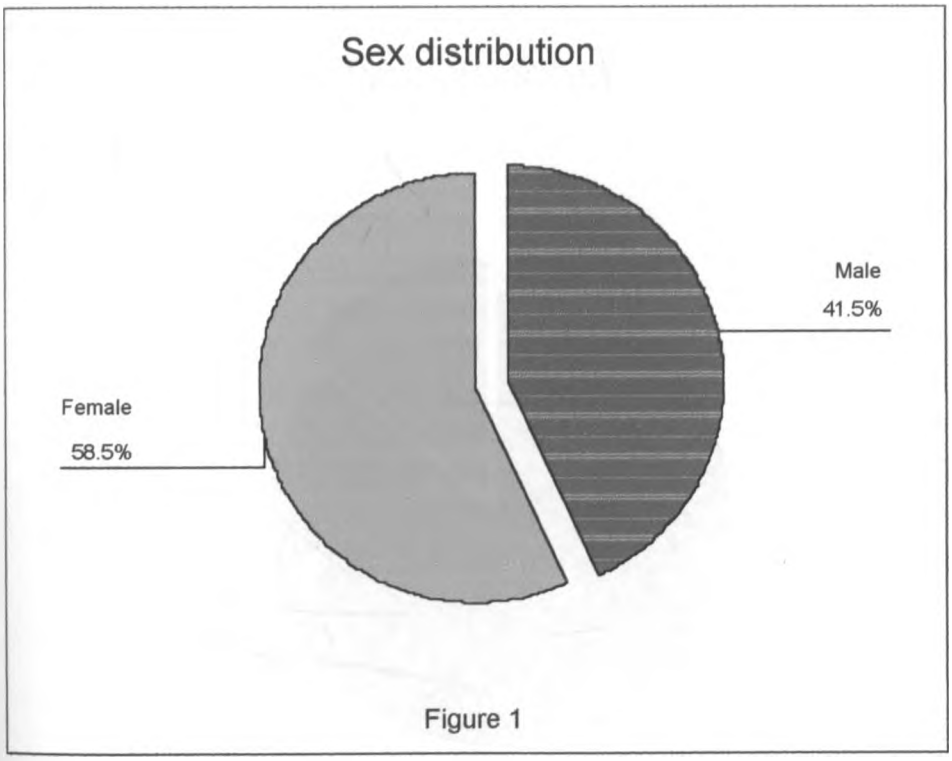

Figure 1

2. THE TYPE OF SACRUM (n=193)

Table below (figure2) shows the type of sacrum of the patients evaluated. Normal sacrum was found in 86.0% and sacral abnormalities in 14.0%of the total.

	Frequency	Valid percent
Normal	166	86.0
Sacral abnormalities	27	14.0
Total	193	100

Table 3: The type of Sacrum

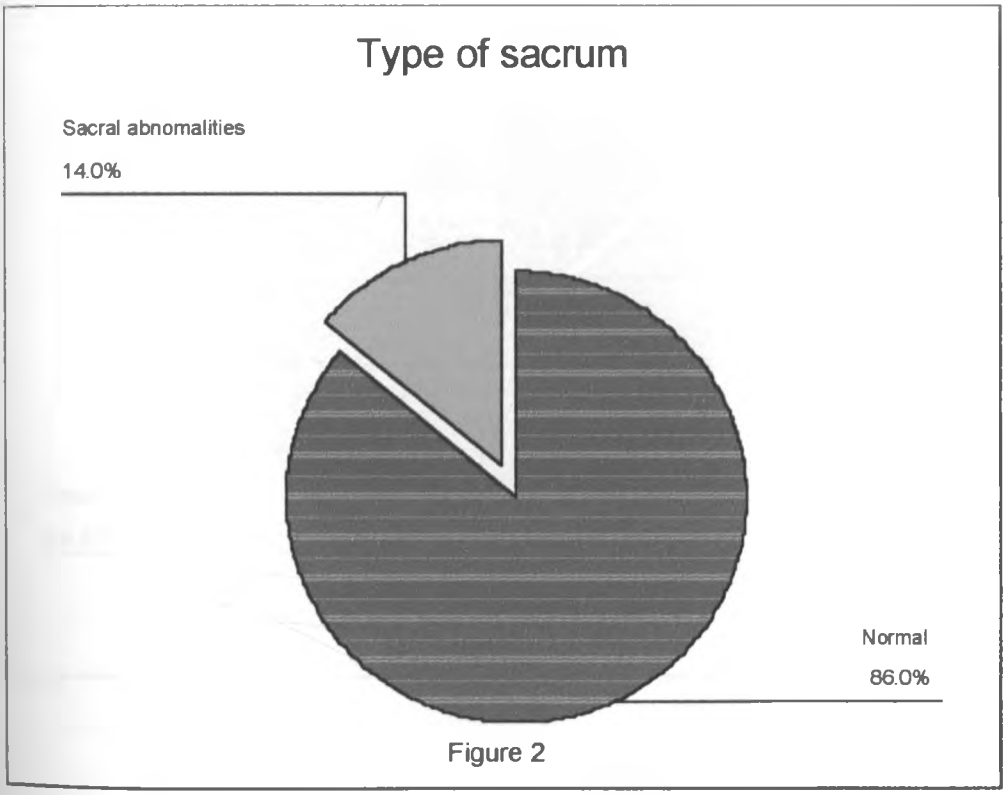

Figure 2

3. LEVEL OF ANOMALY (n=193)

Table 4 (figure 3) shows the level of anomalies of the patients evaluated, 86.5%of patients had low anomalies and 13.5% had high anomalies.

	Frequency	Valid percent
High anomalies	26	13.5
Low anomalies	167	86.5
Total	193	100

Table 4: Level of anomaly

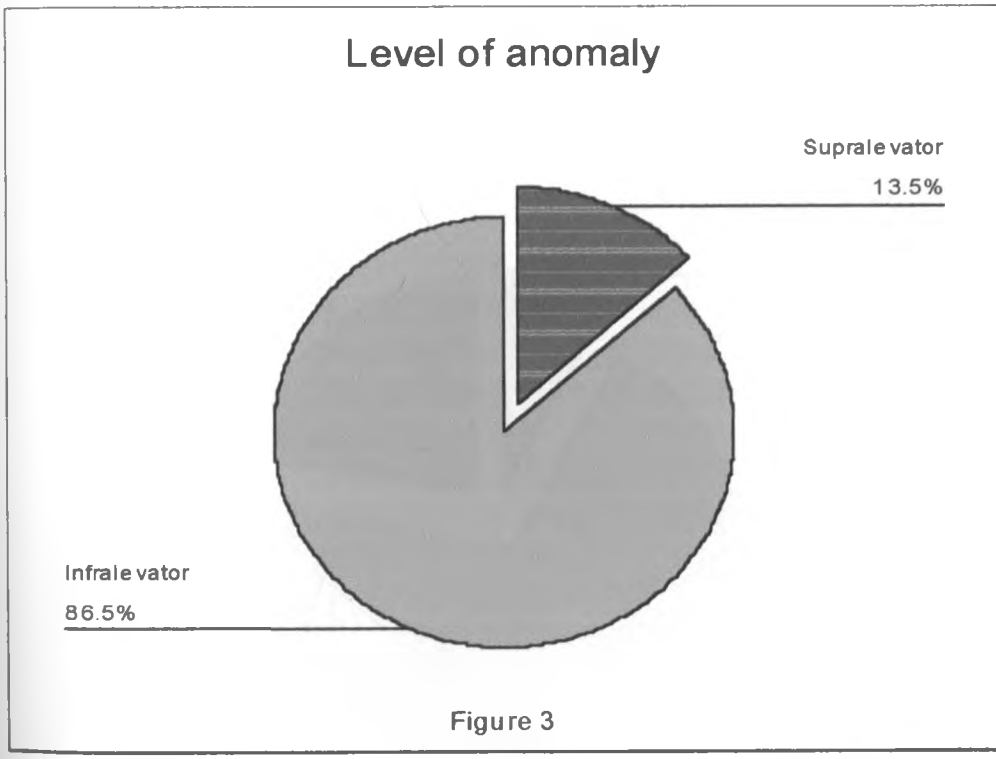

Figure 3

4. COLOSTOMY FASHIONING (n=193)

Table 5 (figure 4) shows the patients who had colostomy fashioned (92.2%) and those without colostomy (7.8%) prior to the definitive primary surgery.

	Frequency	Valid percent
Fashioned	178	92.2
No colostomy	15	7.8
Total	193	100

Table 5: Colostomy fashioning

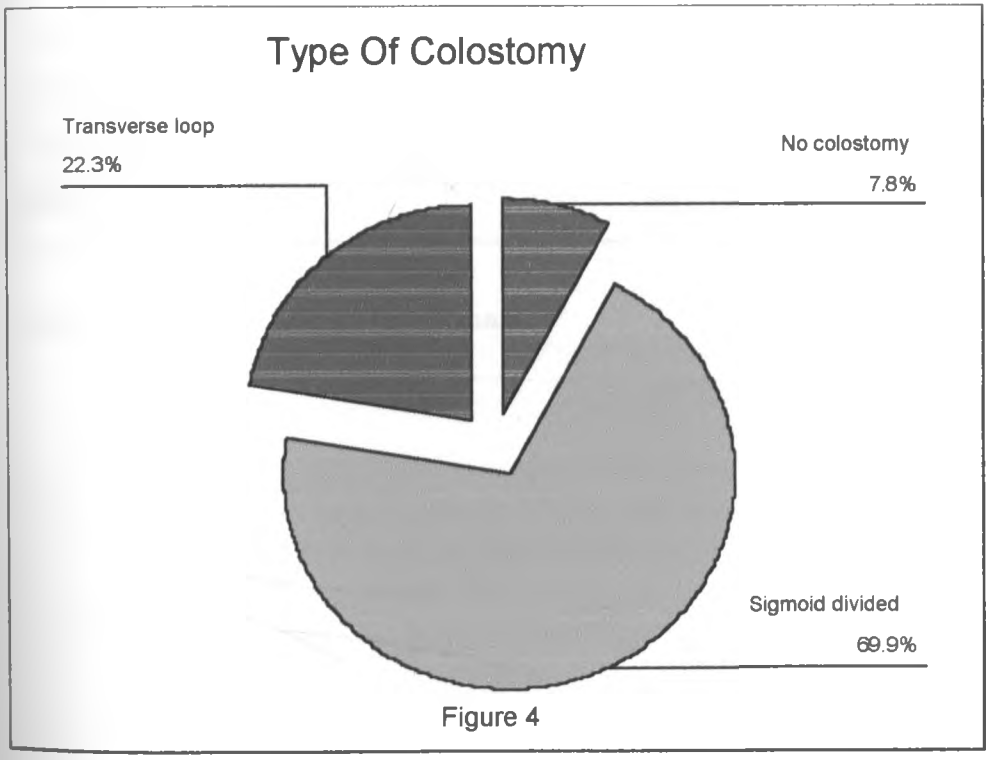

Figure 4

5. COLOSTOMY FASHIONING IN MALES (n=80)

Table 6 shows colostomy fashioning in males. Of the 80 male patients studied, 31.2% had anorectal malformations without a fistula, 30.0% had perineal fistula, 28.8% had rectourethral fistula, 10.0% had rectovesical fistula, and there was no rectal atresia.

All the patients (100%) with rectourethral fistula, rectovesical fistula, and anorectal malformations without fistula had colostomy fashioned prior to primary definitive surgery. No colostomy was required in 41.7% of patients with perineal fistula.

	No. of cases	Colostomy			
		Fashioned		No colostomy	
		No.	%	No.	%
Perineal fistula	24	14	58.3	10	41.7
Rectourethral fistula	23	23	100	0	0
Rectovesical fistula	8	8	100	0	0
Anorectal malformations without a fistula	25	25	100	0	0
Total	80	70	87.5	10	12.5

Table 6: Colostomy fashioning in males

6. COLOSTOMY IN FEMALES (n=113)

Table 7 shows the colostomy fashioning in females. Of the 113 female patients studied, 84.1% had vestibular fistula, 7.1% had perineal and vaginal fistula, and 1.7% had persistent cloaca with common channel less than 3cm.

All female patients (100%) with vaginal fistula, and persistent cloaca with < 3cm common channel had definitive surgery with a colostomy. No colostomy was required in 25.0% and 3.2% of perineal fistula and vestibular fistula respectively before definitive surgery was performed.

	No. of cases	Colostomy			
		Fashioned		Not fashioned	
		No.	%	No.	%
Perineal fistula	8	6	75.0	2	25.0
Vestibular fistula	95	92	96.8	3	3.2
Vaginal fistula	8	8	100	0	0
Persistent cloaca <3cm channel	2	2	100	0	0
Persistent cloaca >3cm channel	0	0	0	0	0
Total	113	108	95.6	5	4.4

Table 7: Colostomy in females

7. AGE AT COLOSTOMY FASHIONING (n=178)

Table 8 shows age at colostomy fashioning, 58.4% of the patients had colostomy fashioned before the age of one month, 28.1% between one and six months, 3.4% between seven and twelve months, and 10.1% after one year.

	Frequency	Valid percent
0-1 month	104	58.4
1-6 months	50	28.1
7-12 months	6	3.4
>1year	18	10.1
Total	178	100

Table 8: Age at colostomy fashioning

8. AGE AT COLOSTOMY FASHIONING Vs SEX DISTRIBUTION (n=178)

Table 9 shows the age at colostomy fashioning by sex of the patients. Males who had colostomy fashioned were 41.0% of the total. By the age of one month 97.1% of males (65.4% of the total) had their colostomy fashioned.

Females who had colostomy fashioned before definitive surgery were 59.0% of the total. Majority of females (66.7%) had their colostomy fashioned after age of one month.

	No. of cases	Sex distribution			
		Males	%	Females	%
0-1 months	104	68	65.4	36	36.6
1-6 months	50	2	4.0	48	96.0
7-12months	6	0	0	6	100
>1year	18	0	0	18	100
Total	178	70	41.0	108	59.0

Table 9: Age at colostomy fashioning Vs sex distribution

9. TYPE OF COLOSTOMY (n=178)

Table 10 shows the type of colostomy fashioned. Divided sigmoid colostomy was fashioned in 75.8% and transverse colostomy in 24.2% of the patients.

Type of colostomy	Frequency	Valid percent
Divided sigmoid	135	75.8
Transverse loop	43	24.2
Total	178	100

Table 10: Type of colostomy

10. REVISION OF COLOSTOMY (n=178)

Of the patients who had colostomy before definitive surgery, table below shows whether their colostomies were revised (16.9%) or not (83.1%).

	Frequency	Valid percent
Yes	30	16.9
No	148	83.1
Total	178	100

Table 11: Revision of colostomy

11. RELATIONSHIP BETWEEN TYPE AND REVISION OF COLOSTOMY (n=178)

Type of colostomy	No. of cases	Colostomy revised			
		Yes	%	No	%
Divided sigmoid	135	5	3.7	130	96.3
Transverse loop	43	25	58.1	18	41.9
Total	178	30	16.9	148	83.1

Table 12: Relationship between type and revision of colostomy

Divided sigmoid colostomy had only 3.7% of patients' requiring colostomy revision. Transverse loop colostomy is prone to prolapse and was revised in58.1% of the patients.

12. TIME BETWEEN COLOSTOMY FASHIONING AND DEFINITIVE SURGERY (n=178)

	Frequency	Valid percent
0-1 month	5	2.8
1- 6 months	12	6.7
6-12 months	43	24.2
>12 months	118	66.3
Total	178	100

Table 13: Time between colostomy fashioning and definitive surgery

Table 13 shows the length of time between colostomy fashioning and definitive surgery. Majority of the patients (90.5%) had this duration longer than six months and only 2.8% of the patients shorter than one month.

13. DEFINITIVE PRIMARY SURGERY (n=193)

Table 14 shows the type of definitive surgery the patients had. Majority of the patients had PSARP (76.7%) while 17.6% and 3.1% of the patients had anoplasty and PSARP with abdominal approach respectively.

	Frequency	Valid percent
Dilatation	1	0.5
Anoplasty	34	17.7
PSARP	148	76.7
PSARP+ abdominal approach	6	3.1
PSARVURP	2	1.
Abdominosacroperineal	1	0.5
Sacroperineal	1	0.5
Total	193	100

Table 14: Definitive primary surgery

14. AGE AT DEFINITIVE SURGERY (n=193)

	Frequency	Valid percent
0-6 months	21	10.9
7-12 months	31	16.1
1-2 years	78	40.4
2-5 years	50	25.9
>5 years	13	6.7
Total	193	100

Table 15: Age at definitive surgery

Table 15 shows the age at definitive surgery. By the age of 5 years majority of the patients (93.3%) have had definitive surgery. Only 10.9% have had definitive surgery by 6 months.

15. DURATION BETWEEN DEFINITIVE SURGERY AND COLOSTOMY CLOSURE (n=178)

Table 16 shows the duration between definitive surgery and colostomy closure. Majority of the patients (86.0%) have duration that is less than 2 years. Only 5.6% have duration that is less than 6 months

	Frequency	Valid percent
0-6 months	10	5.6
6-12 months	93	52.2
1-2 years	50	28.2
>2 years	25	14.0
Total	178	100

Table 16: Duration between definitive surgery and colostomy closure

III. BOWEL FUNCTION FOR PRIMARY SURGERY

1. BOWEL FUNCTION

	Frequency	Valid percent
VBM	138	71.5
Soiling grade 1	19	9.8
Soiling grade 2	14	7.3
Soiling grade 3	8	4.1
Constipation grade 1	4	2.1
Constipation grade 2	3	1.6
Constipation grade 3	7	3.6
Total	193	100

VBM= Voluntary Bowel Movement

Table 17: Bowel function

Table 17 shows the type of bowel function of the patients evaluated. Overall, 71.5% of the patients had VBM (the most valuable sign of fecal control). Involuntary leak of small amount of stool (soiling) was seen in 21.2%. Incapacity to empty the rectum spontaneously (constipation) occurs in 7.3%.

2. RELATIONSHIP BETWEEN AGE AT COLOSTOMY FASHIONING AND BOWEL FUNCTION

	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade 1	Grade >1	Grade 1	Grade >1
0-1month	104	79(76.0)	7(6.7)	12(11.5)	4(3.8)	2(1.9)
1-6 months	50	37(74.0)	4(8.0)	3(6.0)	6(12.0)	0
7-12 months	6	4(66.7)	1(16.7)	1(16.7)	0	0
>1year	18	11(61.1)	4(22.2)	3(16.7)	0	0
Total	178	131(73.5)	18(10.1)	18(10.6)	10(5.6)	2(1.1)

Table 18: Relationship between age at colostomy fashioning and bowel function

Table above shows the relationship between age at colostomy fashioning and bowel function. Colostomies fashioned before the age of one month had the best results. VBM was achieved in 76.0% of the cases. Those patients whose colostomies were fashioned after one year achieved VBM in 61.1%of the patients.

Soiling was present 33.3% of the patients whose colostomies were fashioned after one year compared to 18.2% in the patients whose colostomies were fashioned before one month. Constipation occurs in 6.7% of the patients.

3. BOWEL FUNCTION IN MALES

Male anomalies	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade1	Grade >1	Grade 1	Grade >1
Perineal fistula	24	19(79.1)	0	0	3(12.4)	3(12.4)
Rectourethral fistula	23	17(73.9)	6(20.1)	1(4.3)	0	0
Rectovesical fistula	8	1(12.5)	0	7(87.5)	0	0
Anorectal malformations without a fistula	25	14(56.0)	8(32.0)	3(12.0)	0	0
Rectal ~~abnormalities~~ Atresia	0	0	0	0	0	0
Total	80	51(63.8)	4(5.0)	11(13.8)	2(2.5)	3(3.8)

Table 19: Bowel function in males

Table above shows the bowel function in males. Overall males achieved VBM in 63.8%. Patients with perineal fistula achieved VBM in 79.1% of males, those with rectourethral fistula in 73.9%, anorectal malformations without a fistula in 56.0%, and those with rectovesical fistula in 12.5%.

Soiling was present in 18.8% of females. Soiling was present in 87.5% rectovesical fistula, 44.0% in ARM without a fistula, and 24.4% in rectourethral fistula.

Constipation was present in 6.3%of females. Constipation was present in 24.8% of those patients with perineal fistula.

4. BOWEL FUNCTION IN FEMALES

Female anomalies	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade1	Grade >1	Grade1	Grade >1
Perineal fistula	8	6(75.0)	1(12.5)	0	0	1(12.5)
Vestibular fistula	96	73(76.0)	11(11.5)	8(8.3)	1(1.0)	3(3.1)
Vaginal fistula	8	2(25.0)	2(25.0)	3(37.5)	0	1(12.5
Persistent cloaca <3cm channel	1	0	1(100)	0	0	0
Persistent cloaca >3cm channel	0	0	0	0	0	0
Total	113	87(77.0)	15(13.3)	11(9.7)	1(0.8)	5(4.4)

Table 20: Bowel function in Females

Table 9.4 shows bowel function by diagnosis in females. VBM (patients totally continent) was present among 77.0% of female anomalies, perineal fistula with 75.0%, and vestibular fistula with 76.0%, and vaginal fistula with 25.0%.

Soiling was present among 23.0% of females. Poor prognosis was seen in vaginal fistula where soiling was seen in 62.5% of cases.

Constipation was seen in 5.2% of females. Constipation was seen in 12.5% of perineal fistula and vaginal fistula.

5. BOWEL FUNCTION BY THE TYPE OF PRIMARY DEFINITIVE SURGERY

Definitive primary surgery	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade 1	Grade >1	Grade 1	Grade >1
Dilatation	1	0	0	0	0	1(100)
Anoplasty	34	28(82.4)	0	0	3(8.8)	3(8.8)
PSARP	148	110(74.3)	17(11.5)	14(9.5)	1(0.7)	6(4.1)
PSARP+ Abdominal approach	6	0	1(16.7)	5(83.3)	0	0
PSARVUP	1	0	1(100)	0	0	0
Abdominosacroperineal	2	0	0	2(100)	0	0
Sacroperineal	1	0	0	1(100)	0	0
Total	193	138(71.5)	19(9.8)	22(11.4)	4 (2.1)	10(5.2)

Table 21: Bowel function by the type of primary definitive surgery

Table above shows the bowel function by the type of primary definitive surgery. Patients who underwent anoplasty as the primary definitive surgery achieved VBM in 82.4% of the patients, and PSARP in 74.3% of the patients.

Patients who underwent abdominosacroperineal and sacroperineal had 100% severe soiling. Patients who had dilatation as a definitive surgery had 100% constipation (the number considered was small).

6. BOWEL FUNCTION BY AGE AT DEFINITIVE SURGERY

Age at definitive surgery	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade 1	Grade >1	Grade 1	Grade>1
0-6 months	21	17(81.1)	0	0	0	4(19.0)
7-12 months	31	24(77.4)	0	1(3.2)	4(12.9)	2(6.4)
1-2 years	78	54(69.2)	8(10.2)	14(17.9)	0	2(1.3)
2-5 years	50	35(70.0)	8(16.0)	5(10.0)	0	2(4.0)
> 5 years	13	8(61.5)	3(23.0)	2(15.4)	0	0
Total	193	138(71.5)	19(9.8)	22(11.4)	4(2.1)	10(5.2)

Table22: Bowel function by age at definitive surgery

Table above shows the bowel function by age at definitive surgery. Patients who had primary definitive surgery before age of six months achieved VBM in 81.1 % of the patients, and those operated on after 5 years only 61.5%.

7. BOWEL FUNCTION BY DURATION BETWEEN DEFINITIVE SURGERY AND COLOSTOMY CLOSURE (n=178)

	No. of cases	VBM	
		Frequency	Valid percent
0-6 months	10	7	70.0
6-12 months	93	59	63.4
1-2 years	50	30	60.0
>2 years	25	16	64.0
Total	178	112	62.9

Table 23: Bowel function by duration between definitive surgery and colostomy closure

The duration definitive surgery and colostomy closure ranges 2 months and 5 years. Patients with duration that is shorter than 6 months achieve VBM in 70.0% of the patients, 6-12 months in 63.4%, 1-2 years in 60.0%, and over 2 years in 64.0% of the patients.

8. BOWEL FUNCTION BY THE TYPE OF SACRUM

(n=193)

	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade 1	Grade>1	Grade 1	Grade >1
Normal sacrum	166	131(78.9)	18(10.8)	4(2.4)	4(2.4)	9(5.4)
Sacral abnormalities	27	7(25.9)	1(3.7)	18(66.7)	0	1(3.7)
Total	193	138(71.4)	19(9.8)	22(11.4)	4(2.1)	10(5.2)

Table 24: Bowel function by the type of sacrum

Table above shows bowel function by the type of sacrum. The patients with normal sacrum attained VBM in 78.9%of the patients, 12.4% soiling, and 7.8% constipation. The patients with sacral abnormalities attained VBM in 25.9% of the patients, 71.4% soiling, and constipation in 3.7%.

9. LEVEL OF ANOMALIES (n=193)

Level of anomaly	No. of cases	Bowel function				
		VBM	Soiling		Constipation	
			Grade1	Grade >1	Grade 1	Grade >1
High anomalies	26	12(46.1)	0	13(50.0)	0	1(3.8)
Low anomalies	167	126(75.4)	19(11.3)	9(5.4)	4(2.4)	9(5.4)
Total	193	138(71.4)	19(9.8)	22(11.4)	4(2.1)	10(5.2)

Table 25: Bowel function by the level of anomalies

Table above shows the bowel function by the level of anomalies. In this series low anomalies achieved VBM in 75.4% of the patients, soiling in 16.7% and constipation in 7.8%.

High anomalies overall achieved VBM in 46.1%, soiling in 50.0% of the patients, and constipation in 3.8%.

IV. REOPERATIONS (SECONDARY OPERATIONS)

In this series 24(12.4%) of 193 patients evaluated underwent secondary reoperations.

1. NUMBER OF REOPERATIONS (n=24)

Table 27 shows the number of reoperations. Patients who had one reoperation were 87.5% and more than one12.5%.

	Frequency	Valid percent
1	21	87.5
>1	3	12.5
Total	24	100

Table 26: Number of reoperations

2. NUMBER OF REOPERATIONS BY INDICATIONS (n=24)

No. of reoperations	No. of cases	Indications of reoperations		
		Fecal incontinence	Severe constipation	others
1	21	9(42.9)	6(28.6)	6(28.6)
>1	3	3(100.0)	0	0
Total	24	12(50.0)	6(25.0)	6(25.0)

Table 27: Number of reoperations by indications.

The table above shows the number of reoperations by indications. Fecal incontinence was the indication in 50.0% of the patients for reoperations, severe constipation in 25.0%, and other indications in 25.0%. All the patients who underwent reoperations more than once

had severe fecal incontinence. No patient whose indication for reoperations was constipation had more than one operation

3. THE TYPE SACRUM BY THE NUMBER OF REOPERATIONS (n=24)

Number of reoperations	No. of cases	Type of sacrum			
		Normal		Sacral abnormalities	
		frequency	%	frequency	%
1	21	10	45.5	11	54.5
>1	3	0	0	3	100.0
Total	24	10	41.7	14	58.3

Table 28: The type of sacrum by the number of reoperations

Table above shows the type of sacrum by the number of reoperations. Sacral abnormalities were present in 58.3% of the patients, while those with normal sacrum were 41.7%. The patients who had one reoperation with sacral abnormalities were 54.5% and 45.5% had normal sacrum.

All the patients who had more than one reoperation had sacral abnormalities.

4. LEVEL OF ANOMALIES AND NUMBER OF REOPERATIONS (n=24)

Number of reoperations	No. of cases	Level of anomalies			
		High anomalies		Low anomalies	
		Frequency	%	Frequency	%
1	21	13	61.9	8	38.1
>1	3	3	100.0	0	0
Total	24	16	66.7	8	33.3

Table 29: Level of anomalies and number of reoperations

Table above shows the level of anomalies by the number of reoperations. High and low anomalies were present in 66.7% and 33.3% of the reoperations respectively. The incidence of low and high anomalies when the patient had one reoperation was 61.9% and 38.1% respectively.

All the patients who had more than one reoperations had high anomalies.

5. TYPE OF SACRUM AND INDICATION OF REOPERATIONS

Indication of reoperations	No. of cases	Type of sacrum			
		Normal		Sacral abnormalities	
		Frequency	%	Frequency	%
Fecal incontinence	12	1	8.3	11	91.7
Severe constipation	6	6	100.0	0	0
Others	6	3	50.0	3	50.0
Total	24	10	41.7	14	58.3

Table 30: Type of sacrum and indications of reoperations

Table above shows the indication of reoperations by the type of reoperations. Fecal incontinence was the indication for reoperation in 91.7% of patients with sacral abnormalities and only 8.3% in those with normal sacrum. All the patients with severe constipation had normal sacra.

V. OUTCOME OF REOPERATIONS

1. INDICATIONS OF REOPERATIONS AND OUTCOME

Table below shows the outcome of reoperations by indications. The outcome was good in 75.0%, fair in 16.7%, and poor in 8.3%. Where the indication for reoperation was severe incontinence, good results were obtained in 50.0% of cases, fair in 33.3%, and poor in16.7% of the patients. All patients whose indication for reoperation was severe constipation had good results.

Indications of reoperations	No. of cases	Outcome of reoperations		
		Good	Fair	Poor
Fecal incontinence	12	6(50.0)	4(33.3)	2(16.7)
Severe constipation	6	6(100.0)	0	0
Others	6	6(100.0)	0	0
Total	24	18(75.0)	4(16.7)	2(8.3)

Table 31: Outcome of reoperations by indications

2. THE NUMBER OF REOPERATIONS AND OUTCOME

Number of reoperations	No. of cases	Outcome of reoperations		
		Good	Fair	Poor
1	21	18(85.7)	2(9.5)	1(4.8)
>1	3	0	2(66.7)	1(33.3)
Total	24	18(75.0)	4(16.1)	2(8.3)

Table 32: Number of reoperations and outcome

Table above shows the number of reoperations by the outcome. In the category patients who had one reoperation, good results were obtained in 85.7% of cases, fair in 9.5%and poor in 4.8% of the patients. the patients who had more than one reoperations had fair results in 66.7%and poor results in 33.7% of the patients.

3. THE OUTCOME OF REOPERATIONS BY THE TYPE OF SACRUM

	No. of cases	Outcome of reoperations		
		Good	Fair	poor
Normal	11	11(100)	0	0
Sacral abnormalities	13	7(53.8)	4(30.8)	2(15.4)
Total	24	18(75.0)	4(16.7)	2(8.3)

Table 33: The outcome of reoperations by the type of sacrum

Table above shows the outcome of reoperations by the type of sacrum. All the patients with normal sacrum had good results. Those patients with sacral abnormalities, 53.8% good results, 30.8% fair results and 15.4% had poor results.

4. OUTCOME OF REOPERATIONS BY LEVEL OF ANOMALIES

	No. of cases	Outcome of reoperations		
		Good	Fair	Poor
High anomalies	10	5(50.0)	4(40.0)	1(10.0)
Low anomalies	14	13(92.9)	0	1(7.1)
Total	24	18(75.0)	4(16.7)	2(8.3)

Table 34: Outcome of reoperations by Level of anomalies

Table above shows the outcome of reoperations by the level of anomalies. Low anomalies have good results in 92.9% of the patients and poor in 7.1% of the patients.

High anomalies have good results in 50%, fair in 40.0% and poor in 10.0% of the patients.

LIMITATIONS OF THE STUDY

1. The records of some of the patients were incomplete with some of the clinical details missing i.e. physical findings, investigations, findings at operations, and complications

2. There are a significant number of patients whose files were missing

3. Some of the patients' records may not have represented the correct findings of the patients due to incorrect interpretation of patients' signs and symptoms by the doctors during the follow-up

4. It was difficult to determine the patients who were able to achieve voluntary bowel movement and occasional soiling. This might have skewed results, and a prospective study is necessary.

ROLE OF THE FUNDING SOURCES

The funding sources had no role in the collection, analysis, or interpretation of the data or in decision to submit this dissertation.

6. DISCUSSION

Anorectal malformation is a common problem in Kenya. In Kenyatta National hospital (KNH), the main referral hospital, approximately 30 patients undergo definitive repair per year. A good repair saves a child too unfortunate to be born with this anomaly a lifetime of misery and social seclusion.

We evaluated children over 3 years old, toilet-trained and 6 months after closure of colostomy since after colostomy is closed patients have an adaptation period during which they have irregular bowel movements. There were 2 deaths in this study, the cause of deaths was severe peritonitis secondary to anastomotic leak after colostomy closure.

Of the 193 patients evaluated, males were 41.5% and females were 58.5%. Eighty-six percent of patients had normal sacrum, and 14.0% had sacral abnormalities. There were more patients with low anomalies compared to the high anomalies (86.5% had low anomalies). This is the old classification and has been used in most of the case records.

BOWEL FUNCTION OF PRIMARY OPERATIONS

Voluntary bowel movement (VBM) is the most valuable sign of fecal control. Overall 71.5% of all patients achieved VBM. These results are comparable with results elsewhere. Pena achieved VBM in 74.3% of all the patients in his series[1]. In our series there is a high proportion of patients with low anomalies and normal sacrum. These above factors and long duration in which posterior sagittal repair has been the standard definitive procedure for ARM may explain the results.

Fecal continence is dependent on the factors that include; intact sensation, lack of scarring, normal peristalsis, intact neural pathways, psychological factors, and anatomical factors(functioning internal and external sphincters). Overall soiling was present in 21.2% of all the patients. Historical data shows that approximately 25% of patients suffer some form of fecal incontinence[3]. Pena[1] reports fecal incontinence in 57% of the patients. The patients in the study had a higher incidence of high anomalies. In our series we had

high incidence of patients with low anomalies and patients with normal sacrum (good sphincteric musculature). High anomalies e.g. rectovesical fistula in males show the highest incidence of fecal incontinence (table 19).

Constipation was present in 7.3% of the patients. It was present in 7.8% of the patients with low anomalies compared to 3.8% of high anomalies. In this series constipation was not a serious problem. Most series report high incidence of constipation in simple (low) anomalies[1]. Constipation is a major complication of internal anal sphincter (IAS) saving procedures like posterior sagittal repair. Some series do not advocate saving the most distal rectal muscle during the repair[38]; other series have proved through in-vitro studies that saving the IAS is not important for continence clinically[39]. Dysganglionosis of distal rectal pouch and the fistula has been implicated[11, 12].

COLOSTOMY

Divided sigmoid colostomy is the standard colostomy fashioned prior to definitive surgery in KNH and all the patients in this series evaluated with transverse loop colostomy had their colostomy fashioned in the peripheral hospitals prior to referral for definitive surgery to KNH. Transverse loop colostomy is not totally diverting and allows some stools to pass to distal stoma. Majority of the patients (92.2%) had a colostomy prior to definitive repair. There was a significant number of the patients with low anomalies e.g. perineal fistula who were managed with colostomy (75.0% in males and 58.3% in females). Anoplasty in the neonatal period would have been adequate instead of three operations.

Before the age of one month 58.4% of the total patients had colostomy fashioned. Majority of males have their colostomy early (97.1% before one month) compared to females who have colostomy fashioned later (66.7% after one month). A high proportion of males had no fistula this could explain this phenomenon. Overall children who had colostomy fashioned before the age of one month had better bowel control (VBM of 79.1%) compared to those fashioned thereafter (VBM was 61.1% after one year). Early colostomy reduces the incidence of rectal ectasia developing before the main repair.

Rectal ectasia results from chronic dilatation of the rectum leading to poor peristalsis and severe constipation.

Indications for the revision of colostomy were prolapse of the colostomy stomas, stenosis, and gangrene of the stomas. In this series 83.3% of transverse loop colostomies were revised (87.3% of divided sigmoid were not revised). Transverse loop colostomies are prone to prolapse, it is inadequately diverting leading to fecal imparction in the distal rectum and severe megarectum, and promotes infections. The revision of colostomy increases the number of operations these patients have to undergo and increases the age at definitive surgery. Loss of portions of colon leads to reduced water absorption capability. Inability to form solid stools in a child with ARM usually means fecal incontinence even if the child was born with good prognosis type of defect. This leads to increased agony to patients and parents.

Majority of the patients (90.5%) waits longer than six months for definitive surgery after colostomy fashioning. Majority of the patients have low anomalies and some do not need a colostomy i.e. perineal fistula. With the rising number of patients requiring definitive surgery, these patients should undergo one operation (anoplasty) and minimize the long wait. In these series the duration between colostomy fashioning and definitive surgery does not influence the outcome of bowel function. Theoretically, the patients who waits for short time should have better results, since the parents are cooperative and they are not exposed to the traumatizing colostomy for long duration.

THE TYPE OF SACRUM

The sacrum was evaluated by a combination of invertogram and sacral ratio findings. Sacral ratio was recorded in only 2% of patients case-records evaluated. The measurement of sacrum is very easy (using sacral ratio) and eliminates the difficulties frequently experienced in trying to count the number of vertebra of the sacrum. Sacral ratios of 0.76 are the average values in normal children. Lower values are associated with poor function results. The spectrum of spine and sacral defects found included fused

sacrum, hemivertebra and kyphoscoliosis in upper spine. No cases of hemisacrum were found in this series.

In this series normal sacrum is associated with good prognosis for bowel control (table 24). Patients with normal sacrum achieved VBM in 78.9% of the cases; they also have a higher incidence of constipation. Sacral abnormalities (sacral ratio <0.76) have very poor prognosis, with 66.7% having severe soiling. VBM was achieved in only 25.9% of the patients with sacral abnormalities.

LEVEL OF ANOMALIES

The level of anomalies was evaluated using invertogram and operative findings. Majority of the patients evaluated had low anomalies (table 3 and 25). High anomalies were associated with poor prognosis for bowel control and high incidence of sacral hypodevelopment. Half of the patients with high anomalies had severe soiling (table 25).

DEFINITIVE PRIMARY SURGERY

Posterior sagittal repair is now the standard procedure for the repair of anorectal malformations. Two patients who had abdominosacroperineal and sacroperineal as the definitive procedure prior posterior sagittal repair were included in this study. These patients were included in this series since they developed severe fecal incontinence had posterior sagittal redo operations during the study period. Posterior sagittal repair for ARM is the standard definitive in KNH since 1987. The patients with low anomalies (defined above) had normal or near-normal sphincteric musculature underwent anoplasty as a definitive repair, while those with rectobladder neck fistula and high vaginal fistula (had reduced residual sphincteric muscle) underwent PSARP that included a laparotomy. The rest underwent PSARP alone. VBM was achieved in 82.4% of the patients who underwent anoplasty, 74.5% who underwent PSARP while all the patients who underwent combined PSARP and laparotomy had various grades of soiling. All patients who underwent abdominosacroperineal and sacroperineal had severe soiling. Their numbers is considered small; these patients had primary definite surgery prior to introduction of posterior sagittal repair in KNH.

By the age of 5 years 93.3%of all patients have had definitive repair (table 15). Only 10.9% have had definitive surgery by six months. Patients who had primary definitive repair before the age of one month achieved VBM in 81.1% and those operated on after 5 years only 61.5% of the patients had VBM (table 22). Patients operated before 6 months had better bowel control. By placing the rectum in the right position early in life, early restoration of anorectal continuity (for brain-defecation reflex development), new synapses are established early, provide better sensation and better function of sphincteric mechanism. Posterior sagittal anoplasty should be encouraged at birth for patients with perineal fistula. It relieves the alimentary canal, eliminates urinary tract contamination at birth; all these are achieved in one instead of three operations[28].

Majority of the patients (86.0%) have the duration between definitive surgery and colostomy closure that was less than 2 years and only 5.6% have duration that was less than 6 months (table16). When this duration was less than 6 months, patients have a better functional prognosis for bowel control (table 23). The parents are cooperative, anal dilations are for a shorter period and are not exposed to the traumatizing colostomy for long duration.

TYPE OF ANOMALIES

Overall 63.8% of males and 77.0% in females achieved VBM. Females achieved better results of bowel control than males. In this series males tended to have higher anomalies and sacral defects leading to poorer results obtained. This study noted that sex of the patients is relevant in determination of prognosis. Another study has shown that females tend to have better functional results[41].

Patients with perineal fistulas achieve VBM 79.1% in males and 75.0% in females. This type of anomaly also has the highest number of patients with constipation. The repair gives excellent results. These patients should be treated properly, early in life to prevent primary hypomotility disorder leading to development of megasigmoid, that provokes

overflow pseudoincontinence. A technique that preserves the internal anal sphincter in rectum e.g. posterior sagittal repair has a high incidence of constipation[38].

Rectourethral fistula is the second commonest malformation in males and 73.9% of the patients achieves VBM. In this study it was difficult to separate this anomaly into rectourethral prostatic and rectourethral bulbar fistula. The patients with good sacrum and have had early definitive surgery, have better results.

ARMs without a fistula are the commonest anomalies in males. Some of the patients might have had a fistula but were never discovered due to inadequate pressure colostography. A previous study shows that 33.3% of the fistula were missed by colostography and only discovered at definitive surgery[7]. In this series these patients achieve VBM in 56.0% of the patients. The possible misclassification of some of the patients might have contributed to the low figure obtained; otherwise this anomaly has the same prognosis as those with rectourethral fistula[1]. In our series there is no strong relationship with Downs' syndrome; we found only 2(8%) patients who had this syndrome. Downs' syndrome has a good prognosis one series reports 80-96% of the patients have VBM[13].

Rectovestibular is the commonest anomaly in females and achieve VBM in 76.0% of the patients. Those with good sacrum have better results. Approximately 3.2% of the patients with this anomaly were operated without a colostomy and many of these patients suffered from wound dehiscence and recurrence of fistula. These patients have good potential for bowel control. Therefore operation without a colostomy should be considered unacceptable, since it jeopardizes good results.

Rectovesical fistula has the worst prognosis for bowel control. In our series 12.5% of patients achieve VBM, and 87.5% of patients have severe soiling. The poor results should not discourage definitive surgery. A good bowel management program should be offered to these patients, so that they lead a good quality life, instead of a permanent colostomy.

REDO OPERATIONS

In this series 24 (12.4%) of 193 patients underwent posterior sagittal redo operations at least once. The redo operations have poorer results compared to primary definitive surgery. The indications for redo operations were severe fecal incontinence and severe constipation. Other indications were anal stenosis, rectal prolapse, and anal stricture among other indications. The results of redo operations were categorized good, fair, and poor. The patients were considered to have very significant improvement (good) when they developed VBM with no or minimal soiling. They were considered to have mild improvement (fair) when they declared that that there was some improvement after the operation and no improvement (poor) if no change in bowel function was observed. Patients who had severe fecal incontinence as the indication for reoperation were 50.0% of the total and majority (91.7%) had sacral abnormalities. This shows that most had inadequate residual sphincteric muscle. Twenty five percent had more than one redo operation (table 27). Patients who had redo operations due to severe constipation were 25.0% of the total and all had normal sacrum. None of the patients required more than one redo operations.

Those patients who had one redo operation had better results compared to those with multiple reoperations. Multiple dissection and resection of part of the rectum affects the rectosigmoid motility, the sensitivity of the rectum, increase the amount of scarring, increases psychological stress, increases damage to residual sphincteric muscle, and hence the associated with poor results.

The children with potential for bowel control are the best candidates for reoperation. These are patients who possess the presence of good sphincters, a near-normal or normal sacrum, and anorectal malformation with a good prognosis (e.g. perineal fistula, vestibular fistula). Therefore a careful selection of the patients for reoperations is very important, to be assured of good results.

7. CONCLUSIONS

Posterior sagittal approach is a useful way to repair all types of anorectal malformations, with a totally diverting divided sigmoid colostomy; there is accurate placement of the rectum within the muscle complex. This procedure has been used effectively to establish continence as a primary procedure and secondary operations.

On the whole, in KNH one can expect a child born with anorectal malformation and subjected to posterior sagittal repair, to achieve VBM in approximately 72% of all the patients. Good results of bowel control are associated with early age at functioning of colostomy and early definitive surgery, female sex, absence of sacral defects, and low or simple anomalies.

The children in this series suffered post-operative fecal incontinence as well as other forms of defeacation disorders e.g. constipation, soiling, and incontinence associated with diarrhea. Constipation and soiling can be predicted in a reasonably and accurately, low anomalies achieve good bowel control but have higher incidence of constipation. High anomalies have less satisfactory results and have severe soiling.

Patients with good sacrum and low anomalies have significant improvement following redo operations. Patients with severe fecal incontinence and sacral agenesis do not benefit from multiple redo operations; a good bowel management program should be started early.

8. RECOMMENDATIONS

Based on the results of this study, some recommendations can be made regarding the various aspects of management of anorectal malformations

1. Early age of colostomy fashioning (before age of one month) is associated with good prognosis for bowel control. Patients with no fistula should have a completely diverting colostomy as an emergency within 12 to 48 hours. The patients with low anomalies e.g. perineal fistula, no colostomy is required, anoplasty in neonatal period is decompressing and has good bowel function results.

2. Females with vestibular fistula should have a colostomy fashioned since these patients suffer severe constipation and rectal ectasia before the main repair. Also, they should have a colostomy fashioned prior to definitive repair otherwise disastrous consequence of infection will occur which include wound dehiscence and refistularisation[17, 40]. These patients have good prognosis for bowel control (as shown in this series) and therefore the risk of infection, wound dehiscence, and fibrosis subsequent to healing process is unacceptable. We strongly recommend sigmoid colostomy in neonatal period and limited PSARP at the age of three months.

3. The prolapse of colostomies should and can be avoided, a colostomy prolapses when the stoma is located in a mobile portion of colon[17]. The surgeons should fix the colon to the abdominal wall every time they need to fashion a colostomy in a mobile part of colon[17]. Prolapse of colostomy is a dangerous complication because it provokes ischaemia of the bowel and leads to loss of portions of colon important for water absorption. Liquid stools in a child with ARM usually mean fecal incontinence even if the child was born with a good-prognosis type of defect.

4. Divided sigmoid colostomy; a completely diverting colostomy should be done to all patients except those with low anomalies. Surgeons in peripheral hospitals should be advised to avoid fashioning transverse colostomy to patients with anorectal malformations. Transverse loop colostomy as shown in this study has a higher

incidence of prolapse, stenosis, and is not completely diverting leading to fecal imparction and has a higher incidence of revision of colostomy. Therefore transverse loop colostomy should not be fashioned before definitive management of anorectal malformations.

5. Posterior sagittal approach is a useful way to repair all types of anorectal malformations. Patients with high anomalies will require an additional laparotomy to mobilize highly located rectal pouch. The correct choice of operation is important. Early definitive surgery before the age of six months is recommended, and with a completely diverting colostomy (excepting low anomalies) good results will be achieved.

6. It is easy to predict with a significant degree of accuracy degree of bowel control the patients will have. For the patients with poor functional prognosis, we recommend a discussion with the parents concerning the future. A patient with high anomalies and poor sphincteric muscle and whom poor results expected, posterior sagittal as a primary repair is advised but good bowel management program is started early before the school going age to keep them clean and socially acceptable. The bowel management program consists of teaching parents or patients (when older than 12 years of age) to clean the colon once a day with the use suppositories, enemas, or high colonic irrigations in addition to constipating diet. No child should go to school with diapers when the rest of their classmates are using underwear.

7. Patients for reoperations should be carefully selected. The children with good potential for bowel control are the best candidates for reoperations. Multiple reoperations do not improve the prognosis. Aggressive post-operative bowel management program should be anticipated and started early for the patients who otherwise achieve less satisfactory results e.g. patients with high anomalies and with sacral defects.

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10. QUESTIONNAIRE

Data sheet.
PSARP in KNH
Serial number.....IP NO.....

I. Sex [__]
   A. Males
   B. Females

II. Age at colostomy fashioning [__]
   A. 1-2 days
   B. 3-4 days
   C. 5-6 days
   D. 1-2 weeks
   E. 3-4 weeks
   F. 1-6 months
   G. 7-12 months
   H. > 1 year (specify).....

III. Colostomy
   A. Fashioned
   B. Not fashioned

IV. Type of colostomy [__]
   A. Sigmoid divided colostomy
   B. Transverse Loop colostomy
   C. Others (specify).....

V. Colostomy revised
   A. Yes
   B. No

VI. Length of time between colostomy fashioning and definitive surgery [__]
   A. 0-1 months
   B. 1-6 months
   C. 6-12 months
   D. >12 months

VII. Classification of anorectal anomaly (appendix 2) [__]
   A. Males
      A. Perineal fistula
      B. Rectourethral fistula
         i. Bulbar
         ii. Prostatic
      C. Rectovesical fistula (bladder neck)

D. Imperforate anus without a fistula
E. Rectal agenesis

B. Females [__]
A. Perineal fistula
B. Vestibular fistula
C. vaginal fistula
D. Persistent cloaca
i. < 3cm common channel
ii. >3cm common channel
E. Imperforate anus without fistula
F. Rectal agenesis

VIII. Definitive primary management [__]
A. Dilatation
B. Anoplasty
C. PSARP
D. PSARP+ laparatomy
E. Abdominosacroperineal
F. Sacroperineal
G. Others (specify).....

IX. Age at definitive management [__]
A. 1-4 weeks
B. 1-6 months
C. 7-12 months
D. 1-2 years
E. 2-5 years
F. > 5 years (specify).....

X. Duration between definitive surgery and colostomy closure [__]
A. 1-4months
B. 4-12 months
C. 1-2 years
D. >2 years (specify).....

XI. Evaluation of bowel function (appendix 3) [__]
A. Voluntary bowel movement
B. Soiling
A. Grade 1
B. Grade 2
C. Grade 3
C. Constipation
A. Grade 1
B. Grade 2
C. Grade 3

XII. Number of times for posterior sagittal re-operations done [__]

A. 1
B. 2
C. 3
D. >3

XIII. Indications for re-operations [__]

A. For fecal incontinence
B. Severe constipation
C. For other reasons (specify).....

XIV. Outcome of above [__]

A. Good (very significant improvement)
B. Fair (mild improvement)
C. Poor (no improvement)

XV. Outcome of re-operation where PSARP was not the primary procedure [__]

A. Good (very significant improvement)
B. Fair (mild improvement)
C. Poor (no improvement)

XVI. State of the sacrum radiologically [__]

A. Normal (sacral ratio >0.76)
B. Sacral agenesis (sacral ratio <0.76)
C. Other (specify).....

XVII. Invertogram finding [__]

A. Supralevator
B. Infralevator

11. APPENDIX 1

Wingspread Classification of Anorectal Malformations (1984) [10]

Males

High
- Anorectal agenesis
  - With rectoprostatic urethral fistula
  - Without fistula

Intermediate
- Rectobulbar urethral fistula
- Anal agenesis without fistula

Low
- Anocutaneous fistula
- Anal stenosis

Females

High
- Anorectal agenesis
  - With rectovaginal fistula
  - Without fistula
- Rectal atresia

Intermediate
- Rectovestibular fistula
- Rectovaginal fistula
- Anal agenesis without fistula

Low
- Anovestibular fistula
- Anocutaneous fistula
- Anal stenosis

Cloaca

Rare malformations

APPENDIX 2

Specific Anomalies of the Spectrum of Anorectal Malformations[10]

Males

Cutaneous fistula Anal stenosis Anal membrane	No colostomy required
Rectourethral fistula Bulbar Prostate Rectovesical fistula Anorectal agenesis without a fistula Rectal atresia	Colostomy required

Females

Cutaneous (perineal) fistula	No colostomy required
Vestibular fistula Vaginal fistula Anorectal agenesis without a fistula Rectal atresia Persistent cloaca Complex malformations	Colostomy required

APPENDIX 3

Pena's classification of bowel function[7]

Parameters used for evaluating primary operations include the following:

1. Voluntary bowel movement, which is defined as the act of feeling the urge to use the toilet to have a bowel movement, the capacity to verbalize it, and to hold the bowel movement until the patient reaches the bathroom, this is considered the most valuable sign of fecal control.

2. Soiling is defined as the involuntary leaking of small amounts of stool, which provokes smearing of underwear. This may be present with or without voluntary bowel movements.
    a. *Grade 1 soiling* occurs occasionally (once or twice per week) in minimal amounts and gives no social problems to the patients.
    b. *Grade 2 soiling* refers to soiling that occurs every day but does not cause any social problems.
    c. *Grade 3 soiling* refers to soiling that is constant and represents a social problem to the patient because other people around him perceive the problem.

3. Constipation is defined as the incapacity to empty the rectum spontaneously (without help) every day.
    a. *Grade 1:* Constipation is when the patient is manageable by changes in diet.
    b. *Grade 2:* Constipation is when the patients require laxatives.
    c. *Grade 3*: Constipation is when the patients require enemas.

Re-operations were assessed using the following criteria,

1. *Very significant improvement*: patients who are able to stop medical management that keep them clean i.e. enemas. These patients develop voluntary bowel movements with no or minimal soiling.

2. *Mild improvement*: patients who note some degree of improvement.

3. *No improvement*: no change is noted