

**QUALITY OF LIFE IN CHILDREN AFTER CORRECTIVE
SURGERY FOR ANORECTAL MALFORMATION AT
KENYATTA NATIONAL HOSPITAL.**

**ASSESSMENT OF QUALITY OF LIFE IN CHILDREN AFTER
CORRECTIVE SURGERY FOR ANORECTAL MALFORMATION AT
KENYATTA NATIONAL HOSPITAL.**

BY

DR. SWALEH H. SHAHBAL

M.B.Ch.B (U.O.N)

**A DISSERTATION SUBMITTED AS PART FULFILMENT FOR THE
AWARD OF MASTER OF MEDICINE IN GENERAL SURGERY**

UNIVERSITY OF NAIROBI

©2016

STUDENTS' DECLARATION

I declare that this dissertation is my original work and has not been presented for a degree in any other university

Dr. Swaleh H. Shahbal

Signed _____ Date _____

SUPERVISORS' APPROVAL

This dissertation has been submitted for examination with our approval as university supervisors.

Mr Francis Osawa

MBChB, M.MED General Surgery (U.O.N)

Lecturer, Department Of Surgery,

University Of Nairobi

Signed _____ Date _____

Mr. James Ndung'u

MBChB, M.MED General Surgery (U.O.N), Fellow of Paediatric Surgery(UK)

Senior Lecturer, Department of surgery,

University Of Nairobi

Signed _____ Date _____

Prof. Peter L.W. Ndaguatha

MBChB, Mmed (UON), FCS (ECSA), Fellow Of Urology (UK)

Consultant Urologist and Associate Professor

Department of Surgery (Urology), University Of Nairobi.

Signed _____ Date _____

DEPARTMENTAL APPROVAL

The research proposal has been presented at the surgical departmental meeting and is hereby approved for presentation to the Kenyatta National Hospital Ethics and Research Committee.

Signed _____ Date _____

Chairman,

Department of Surgery,

University of Nairobi.

DECLARATION FORM FOR STUDENTS

UNIVERSITY OF NAIROBI

Declaration of Originality Form

This form must be completed and signed for all works submitted to the University for examination.

Name of Student _____

Registration Number _____

College _____

Faculty/School/Institute _____

Department _____

Course Name _____

Title of the work _____

DECLARATION

1. I understand what Plagiarism is and I am aware of the University's policy in this regard
2. I declare that this _____ (Thesis, project, essay, assignment, paper, report, etc) is my original work and has not been submitted elsewhere for examination, award of a degree or publication. Where other people's work, or my own work has been used, this has properly been acknowledged and referenced in accordance with the University of Nairobi's requirements.
3. I have not sought or used the services of any professional agencies to produce this work
4. I have not allowed, and shall not allow anyone to copy my work with the intention of passing it off as his/her own work
5. I understand that any false claim in respect of this work shall result in disciplinary action, in accordance with University Plagiarism Policy.

Signature _____

Date _____

DEDICATION

This work is dedicated to my beloved late brother Dr. Clive Kibwage. He pushed me into completing this work while on his death bed and I will always be grateful to him. May God rest his soul in eternal peace.

ACKNOWLEDGEMENT

I wish to sincerely thank my supervisors, Dr. F. Osawa, Mr. J. Ndungu and Prof. P. Ndaguatha, who have pushed me tirelessly into completing this work. I would also like to thank all those from the Department of surgery who walked with me through this long journey. I am also grateful to the Principal, college of Health Sciences Prof Isaac Kibwage and Dean of Medicine, Professor Were for standing by me and believing in me.

The Paediatric Surgical Unit, Prof. Kyambi, Mr. Kambuni, Mr. Lessan and all the staff in the unit who assisted me directly or indirectly in achieving this target.

Last but not least, I wish to sincerely thank my wife, Dr. Maryam Badawy for guiding me, for pushing me and for being with me through the hard times.

My parents have been a great pillar of support throughout my life and for that there are no words that can express my gratitude.

Finally to all my colleagues, Kenyatta staff and the Department's non- teaching staff like Madam Naomi, Rose and Kagendo who always told me to complete this hurdle.

May God grant you all a healthy and prosperous life.

TABLE OF CONTENTS

STUDENTS' DECLARATION	iii
SUPERVISORS' APPROVAL	iv
DEPARTMENTAL APPROVAL	v
DEDICATION	vii
ACKNOWLEDGEMENT	viii
TABLE OF CONTENTS.....	ix
LIST OF TABLES	xi
LIST OF FIGURES	xii
ABBREVIATIONS	xiii
SUMMARY	xiv
1.0 CHAPTER ONE: INTRODUCTION.....	1
1.1 Anorectal malformations.....	1
1.1 Literature Review	3
2.0 CHAPTER TWO: STUDY JUSTIFICATION.....	6
2.1 Broad Objective.....	6
2.1.1 Specific Objectives	6
3.0 CHAPTER THREE: MATERIALS AND METHODS	7
3.1 Study design	7
3.2 Study setting	7
3.3 Study population.....	7
3.4 Inclusion criteria.....	7
3.5 Exclusion criteria.....	7
3.6 Sampling technique	7
3.7 Sample size calculation	7
3.8 Data collection.....	8

3.9 Data analysis	8
3.10 Ethical consideration	9
4.0 CHAPTER FOUR : RESULTS	10
4.1 Age and Gender Distribution	10
4.3 Pattern of presentation.....	11
4.4 Associated congenital anomalies	12
4.5 Type of anomaly.....	12
4.6 Type of Surgery and Outcome	13
5.0 CHAPTER FIVE: DISCUSSION.....	18
5.1 Conclusion.....	19
5.2 Recommendations	20
REFERENCES	21
APPENDICES	23
Appendix I: Questionnaire	24
Appendix II: KNH/ERC Letter of Approval.....	27

LIST OF TABLES

Table 1: Classification of Non-syndromic ARMs	2
Table 2 : Decriptive Statistics on age	10
Table 3: Association between symptoms and outcome	12
Table 4: Anatomical anomalies encountered	12
Table 5 : Association between type of anomaly and outcome.....	13
Table 6 : Association between type of surgery and outcome	15
Table 7: Measures of Association between type of Surgery and outcome	16
Table 8 : Observed Grades of soiling	16
Table 9 : Observed Grades of Constipation.....	17
Table 10 : Overall observed functional outcome.....	17
Table 11: Comparison of Outcomes in various Anomalies.....	19

LIST OF FIGURES

Figure 1: Male to Female ratio.....	10
Figure 2 : Symptoms at presentation	11
Figure 3: Type of Anomaly.....	13
Figure 4: Types of Surgical approaches.....	14

ABBREVIATIONS

RM-	Anorectal Malformation
HD-	Hirschsprung's Disease
PSARP-	Posterior sagittal anorectoplasty
UON-	University Of Nairobi
KNH-	Kenyatta National Hospital
SPSS-	Statistical Package for Social Sciences
MACE-	Malone Antegrade Colonic Enema
PSOPC-	paediatric surgical outpatient clinic

SUMMARY

Twenty five patients with Anorectal Malformation were enrolled in the study for assessment of Quality of Life after corrective surgery for Anorectal Malformation.

Seventy two percent of the participants had good functional outcome , 12 percent had a fair outcome and 16% percent had a poor outcome.

Low malformations have a better prognosis compared to high malformations i.e. 92% vs. 33% respectively.

Redo surgery is the biggest cause of unfavourable outcomes. Only 37.5% achieved normal bowel control after redo procedures. Eight participants had redo surgeries.

Females had better outcomes than males probably because more females had lower malformations compared to males. All females enrolled in the study had recto vestibular fistulae.

Vertebral anomalies were found in two participants but this however did not affect their outcome. This is a rare finding because vertebral anomalies are known to have poor outcomes due to associated poor innervation of the anal sphincter and poor development of the sphincter muscles.

The Paediatric surgical department at KNH is evolving and adapting standards as practiced elsewhere worldwide. The old patients had abdominal perineal surgery, later Posterior sagittal anorectoplasty was adapted and now more surgeons are practising Anterior sagittal anorectoplasty with very good results.

1.0 CHAPTER ONE: INTRODUCTION

Anorectal Malformations is the most common congenital colorectal defect in the newborn. The success rates in corrective surgery for Anorectal Malformations has improved significantly because of better understanding of the pathological anatomy and physiology of these defects. Modern surgical techniques have also played a significant role in the improved outcomes¹.

However, a significant population of patients continues to suffer from defective bowel control way into their adulthood. Some children have problems with urinary control as well as sexual dysfunction. The functional problems are pronounced in patients with high Anorectal Malformations¹.

Compared with normal children, patients with Anorectal Malformations have limitations in their quality of life¹.

Corrective surgeries for Anorectal Malformations are procedures that are routinely performed at the Kenyatta National Hospital. Despite these procedures being done routinely, the quality of life of these patients in the local population is not known. Approximately 35-50 closures of colostomy are done for Anorectal Malformation per year in Kenyatta National Hospital (this is information from the theatre records).

1.1 Anorectal malformations

Anorectal Malformations comprise of a wide spectrum of diseases, which can affect boys as well as girls. They may involve the distal anus and rectum as well as the urinary and genital tracts. They occur in approximately 1 in 5000 live births².

Defects range from the very minor and easily treated anomalies, with an excellent functional prognosis, to those that are complex, difficult to manage and often associated with other anomalies. These, as expected, tend to have a poor functional prognosis. The current classification by Pena attempts to group together defects that have common diagnostic, therapeutic and prognostic features².

Table 1: Classification of Non-syndromic ARMs

MALES	FEMALES	COMPLEX AND UNUSUAL DEFECTS
Recto-perineal fistula	Recto-perineal fistula	Cloacalextrrophy, covered cloacal extra
Recto-urethral-bulbar fistula	Recto-vestibular fistula	Posterior cloaca
Recto-urethral-prostatic fistula	Cloaca with short common channel (< 3 cm)	Associated to presacral mass
Recto-bladder neck fistula	Cloaca with long common channel (> 3 cm)	Rectal atresia
Imperforated anus without fistula	Imperforated anus without fistula	
Complex and unusual defects		

The surgical approach to repairing these defects changed dramatically in 1980 with the introduction of the posterior saggital approach, which not only allowed surgeons to view the anatomy of these defects clearly, but also to repair them under direct vision². Improved imaging techniques, as well as better knowledge of the anatomy and physiology of the pelvic structures at birth have refined the diagnosis and initial management of these patients.

The main concerns for the surgeon in correcting these anomalies are bowel control, urinary control, and sexual function. With early diagnosis, management of associated anomalies as well as efficient and meticulous surgical repair, patients have the best chance for a good functional outcome².

Complications post anoplasty and posterior saggital anorectoplasty (PSARP) do occur. Wound dehiscence may occur in the early post-operative period and it directly impacts on the functional outcome of the surgery². Constipation is the most frequent functional disorder after repair of anorectal malformation^{1,2,3}.

Interestingly, patients with lower defects, and therefore with better prognosis for bowel control, suffer a higher incidence of constipation and vice versa³. Origin of Constipation is unknown but it appears to be a hypomotility disorder secondary to chronic bowel dilatation^{2,3}.

Constipation correlates directly with the degree of rectosigmoid dilation at the time of colostomy closure. Therefore, every effort should be made to try to keep the rectosigmoid empty and decompressed from day 1 in these patients^{2,3}. Persistent constipation after definitive surgery may result into megarectum and megasigmoid and can lead to fecal impaction and overflow incontinence².

Soiling is less frequently seen as compared to constipation. In a patient with good prognosis, there may be overflow incontinence. However, true fecal incontinence may occur in cases of very high Anorectal Malformation, poor muscle tone or an abnormal sacrum². Wound dehiscence affects outcome even in low malformations and this is common as has been noted anecdotally at the Kenyatta National Hospital (KNH).

Fecal and urinary incontinence can occur even with an excellent anatomic repair, reasons behind this include; a poorly developed sacrum, deficient nerve supply, and spinal cord anomalies. For these patients, an effective bowel management program including enema and dietary restrictions has been devised to improve their quality of life^{1,2}.

1.1 Literature Review

The outcomes of Anorectal Malformations have improved significantly because of better understanding of the pathological anatomy and physiology of these defects. Modern surgical techniques have also played a significant role in the improved outcomes.

However, a significant population of patients continue to suffer from defective bowel control despite corrective surgery¹. Postoperative bowel dysfunction is evaluated by taking a detailed history noting the patient's bowel habits, use of anti-motility drugs or laxatives, need for dilation or irrigation, and the type of previous surgery. The functional problems are more pronounced in patients with high Anorectal Malformations¹.

The optimal long-term outcome for patients after surgery for Anorectal Malformations is to be faecally continent and to have normal bowel movements. Usually, this outcome is achieved, however, there is a small subset of patients who have difficulty after their primary operation. Most controlled follow-up studies extending to adulthood have demonstrated impaired bowel function of variable degree, including increased incidence of fecal incontinence in relation to age-matched controls^{9,10,11}.

Today, the gold standard of anorectal repairing Anorectal Malformations is the PSARP, initially proposed by de Vries and Pena^{3,13}. Rintala et al found normal or good continence (no

social restrictions) in 64% of adolescents who had undergone PSARP during early childhood¹⁴. The same authors reviewed the functional outcome in 69 patients who were aged >15 years at the time of assessment. Normal continence was found in 43% of the patients. A total of 25 patients (36%) had minor problems, such as constipation or occasional soiling. These patients had no social restrictions. Fourteen patients (25%) had significant continence problems causing frequent soiling and used protective pads or changed underwear, or had undergone a Malone Antegrade Colonic Enema (MACE) procedure. Of 14 patients, 5 were mentally retarded. There are only a few functional outcome studies of low ARM with a follow-up extending to adulthood¹⁴. Most of the earlier series report favourable functional outcome in most patients with low anomalies. Karkowski reported good continence in 12 (80%) of his 15 patients with low malformations¹⁵. Nixon et al found entirely normal bowel control in 23 (74%) of their 31 adult or adolescent patients. The remaining patients had occasional or frequent soiling¹⁶. Constipation is the most common early functional problem in patients with low anomalies, occurring in about 40% of the children^{17,18,19}. Chronic constipation is also the main functional complication following repair of high anomalies by PSARP^{20,21,22}. The incidence of constipation following PSARP procedure has varied between less than 10% and 73%²³. Constipation seems to be more common when internal sphincter-preserving techniques have been used^{14,24}. The cause of constipation is unclear; the extensive mobilization of the anorectum may cause partial sensory denervation of the rectum and impair the awareness of rectal fullness. Also, rectosigmoid hypomotility and generalized colonic motility disturbance have been suggested²⁵.

Locally Kigo et al in 2000 found voluntary bowel movement was achieved in 71.5% of patients. Females had better voluntary bowel movement 77% compared to males 63.8%⁴.

The level of the anomaly is an important prognostic factor in terms of bowel function²⁰. Others include the presence of severe sacral abnormalities which is associated with hypoplastic sphincters^{20,23,25}, preserved functional internal sphincter^{27,28}, colonic motility disorders usually presenting as constipation^{22,23,27,28} and the surgical method of anorectal reconstruction particularly in high malformations²⁸.

In Kigo's study, 76% of patients with rectovesical fistula, 73.9% with recto-urethral fistula, 56% without a fistula, 25% with vaginal fistula and 12.5% with recto-vesical fistula achieved voluntary bowel control. He also showed that the more re-operations done on the patient resulted in poor outcomes⁴.

Scoring methods based on subjective parameters with or without clinical examination have been designed to get quantitative information about the bowel function. The most commonly used classifications are the Kelly-score, the Templeton score²⁹, Holschneider score^{30,31} and Rintala score²³.

All these classifications ultimately categorize the outcomes as good, fair, or poor. Descriptive, nonscoring methods to assess and describe functional outcome were advocated by the Wingspread group of paediatric surgeons. This method and the descriptive, nonscoring outcome classification of Peña, based on his extensive series²⁰, are the basis of the new Krickenbeck outcome classification method which shall be used in this study.

Despite improvements in understanding the pathology and developments in surgical techniques, ARM is associated with significant morbidity during childhood and adolescence. This is reflected by the psychological and social problems of both patients and their parents. Other factors that may affect psychosocial well-being of patients include protracted hospitalisations and repeated surgical procedures.¹

Impaired functional outcome after corrective surgery for ARM may have a major negative impact on the quality of life of these patients. This has been confirmed by a few existing studies on quality of life^{32,33}.

A number of factors including physical, psychosocial and mental health among others do influence the quality of life. Some studies have used social or sexual problems related to impaired functional outcomes as parameters for poor quality of life⁸.

Others have adapted medical outcome measures to assess quality of life⁵.

The continence related social problems are more common in patients with high lesions^{8,31}. The main problem is fecal soiling that restricts social activities¹.

In Rintala's series³¹, 85% of the adult patients with high ARM reported social disability related to soiling. Other problems disturbing occupational life, was inability to hold flatus and fecal urgency.

Adult patients had significantly lower education level than expected⁵.

2.0 CHAPTER TWO: STUDY JUSTIFICATION

The results of treatment of ARMs are not uniformly successful, because the surgeon's interpretation of success may differ from those of the parent's and the child. In light of the above, there has been no study conducted locally to determine the success rates and the assessment of the quality of life for patients born with ARMs after undergoing their corrective surgery.

The surgical technique practiced in the past for ARM has been PSARP, however ASARP has gained popularity in the recent past in KNH. The outcomes of this procedure has not been previously analysed and documented locally.

The data obtained from this study will therefore provide useful indicators and parameters needed by the multidisciplinary team involved in the bowel management program for children with functional defecation disorders after surgery for ARMs so as to improve their QOL.

2.1 Broad Objective

To assess the quality of life of children after corrective surgery for Anorectal malformations at the KNH.

2.1.1 Specific Objectives

- 1.To determine the rate of fecal incontinence in children after corrective surgery for ARMs at KNH.
- 2.To determine the rate of constipation in children after corrective surgery for ARM's
3. To determine the objective functional score after corrective surgery for ARM's

3.0 CHAPTER THREE: MATERIALS AND METHODS

3.1 Study design

Cross sectional study.

3.2 Study setting

Kenyatta National Hospital Paediatric surgical outpatient clinic

3.3 Study population

Children above 4 years with ARMs and have had corrective surgery and closure of colostomy at least 8weeks prior to recruitment.

3.4 Inclusion criteria

All children above 4 years of age with ARM who have had corrective surgery and closure of colostomy at least 6 weeks prior to recruitment into the study.

3.5 Exclusion criteria

Children with associated CNS anomalies e.g. mental retardation, cerebral palsy.

3.6 Sampling technique

All children above 4yrs with ARM who have undergone closure of colostomy will be called to the PSOPC. The participants will be chosen from existing registries. The parent/guardian contacts will be gotten from the patients file and will be called and asked to come to the paediatric SOPC for follow up. Data will not be collected on the phone. The calls will be made by the principal investigator or research assistant. Consecutive enrolment of patients who meet the inclusion criteria will be recruited into the study and the questionnaire will be administered to determine their bowel function score.

3.7 Sample size calculation

The sample size for this study is calculated with the formula;

$$n = N / (1+N(e^2)),$$

Where n is the sample size, N is the population size and e is the degree of precision.

The degree of precision typically appears in published surveys as "margin of error," and can range from 0 to 100 percent, but values between 1 and 5 percent are more typical, depending on the nature of the study.

Data will be disseminated to the Pediatric Surgical Unit staff, consultants, Senior house Officers, nutritionists and nurses, involved in the care. This data will be able to inform them on how well or badly they are doing in the care of their patients. Data will also be disseminated to the participants and their parents/guardians as they come for their next follow-up visits, as this is routinely done for life.

3.10 Ethical consideration

Institutional consent will be sought from the Department of surgery, University of Nairobi (UON) and Ethics and Research Committee of KNH. Informed consent shall be sought from parents/guardian of patients. Confidentiality and privacy shall be observed.

4.0 CHAPTER FOUR : RESULTS

A total of 25 patients were recruited as per approved proposal by the KNH-UON ethics and research committee.

4.1 Age and Gender Distribution

Out of the 25 recruited patients 14(56%) were males and 11(44%) were females. The mean age of our participants was 8years and 3 months with a range between 3 years and 35years.

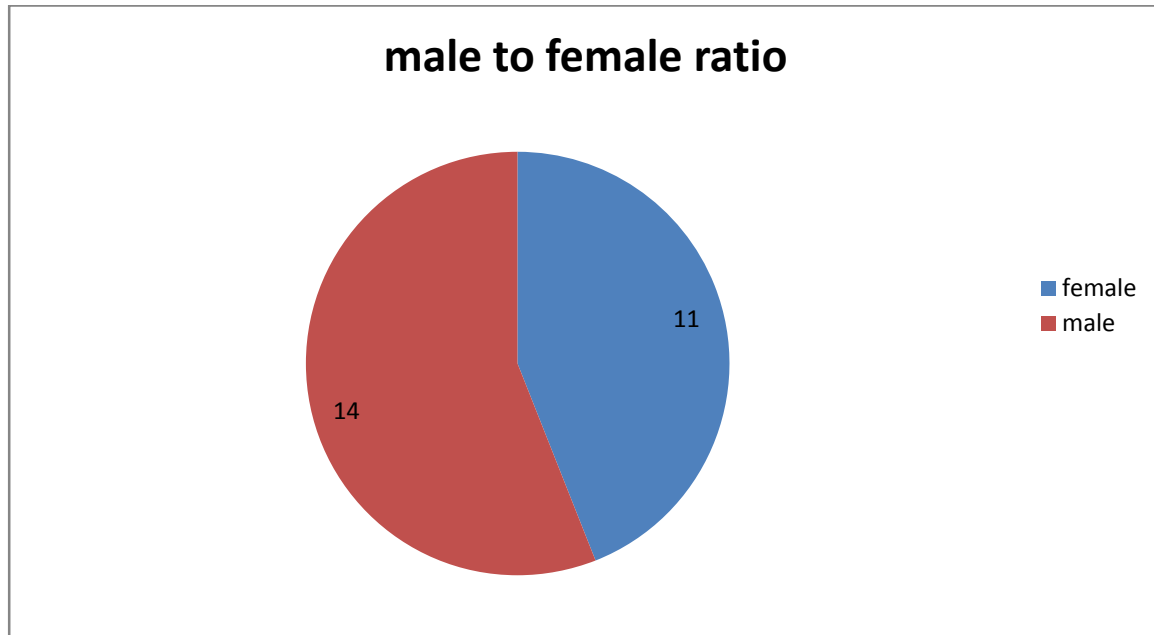


Figure 1: Male to Female ratio

Table 2 : Descriptive Statistics on age

	N	Minimum	Maximum	Mean	Std. Deviation	Skewness	
	Statistic	Statistic	Statistic	Statistic	Statistic	Statistic	Std. Error
Current age in years	25	2.50	35.00	8.2716	8.39046	2.443	.464
Valid N (list wise)	25						

The distribution based on current age is positively skewed with skewness of 2.443 and standard error of skewness of 0.464.

4.3 Pattern of presentation

In this study more males presented earlier than females. Sixty four percent of the males (7 out of 11) presented within two days of life and average time at presentation being 36 days of life. One male patient presented at 1 year of age with a recto-perineal fistula. There was one female patient who presented to KNH at two years with a recto-vestibular fistula. Fifty four percent of cases presented initially with acute intestinal obstruction the rest had passage of stool through a fistulous opening. Forty four percent of those who presented with an abnormal opening were females with recto-vestibular fistulae while 12% were males with recto-perineal fistula.

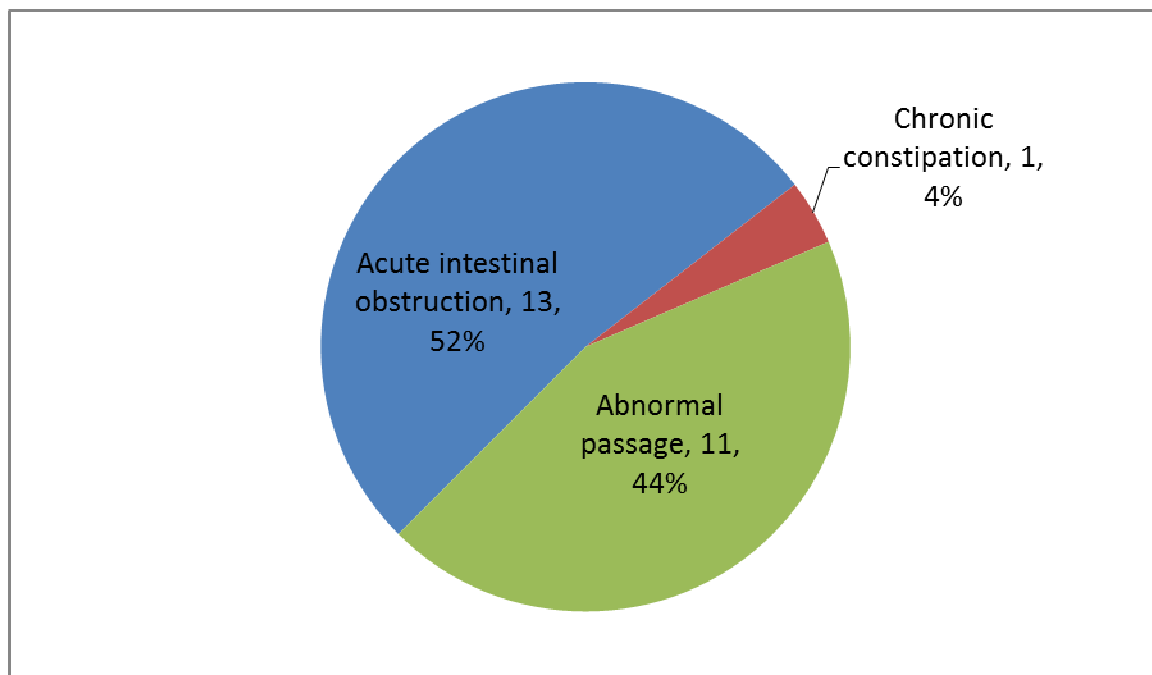


Figure 2 : Symptoms at presentation

Table 3: Association between symptoms and outcome

Symptoms at presentation	Quality of life			p-value
	poor	fair	Good	
Acute intestinal obstruction	3(23.1%)	3(23.1%)	7(53.8%)	0.320
Chronic constipation	0(0.0%)	0(0.0%)	1(9.1%)	
Abnormal passage	1(9.1%)	0(0.0%)	10(90.9%)	

4.4 Associated congenital anomalies

Five patients (20%) had other associated congenital anomalies. Vertebral anomalies were found in 2 participants. The deformities included hemi-vertebrae at T8 and lumbar lordosis. They both have a good functional outcome. The child with hemi-vertebrae also had bilateral undescended testis. Three participants had genitourinary anomalies namely renal agenesis, bladder extrophy and a solitary kidney with a duplex system. None of the participants had a cardiac anomaly.

4.5 Type of anomaly

The anatomical anomalies found in the study population is as tabled below:-

Table 4: Anatomical anomalies encountered

ANOMALY	FREQUENCY	PERCENT
Rectovestibular fistula	11	44
Recto-urethral Bulbar fistula	4	16
Recto-urethral Prostatic fistula	4	16
Recto-perineal fistula	3	12
No fistula	2	8
Rectovesical fistula	1	4

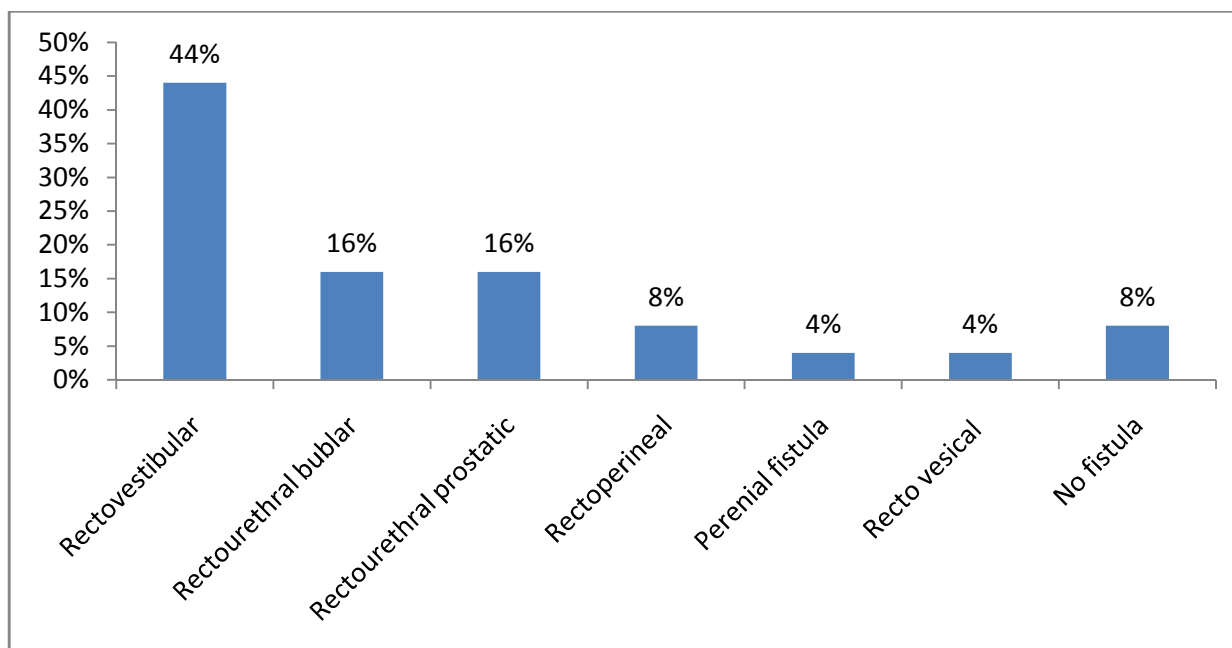


Figure 3: Type of Anomaly

Table 5 : Association between type of anomaly and outcome

Type anomalies	Quality of life			p-value
	poor	fair	Good	
Rectovestibular	1(2.5%)	1(2.5%)	2(50%)	0.103
Rectourethral bulbar	1(25%)	2(50%)	1(25%)	
Rectourethral prostatic	0(0.0%)	0(0.0%)	2(100%)	
No fistula	0(0.0%)	0(0.0%)	1(100%)	
Rectovesical	1(100%)	0(0.0%)	0(0.0%)	
Rectoperineal	0(0.0%)	0(0.0%)	2(100)	

Statistically there is no significant association between the type of anomaly and outcome ,p value 0.103. This could be explained by the skewness of the data and many different types of anomalies shared in the small sample size.

4.6 Type of Surgery and Outcome

The procedure for ARM patients in this study before the introduction of Posterior Saggittal Anorectoplasty (PSARP) involved an abdominal perineal approach. One patient had

undergone this procedure. The rest had a Posterior or Anterior approach and of these 15(60%) participants had PSARP done while 8 (32%) had ASARP . Anoplasty was done in one patient with perineal fistula.

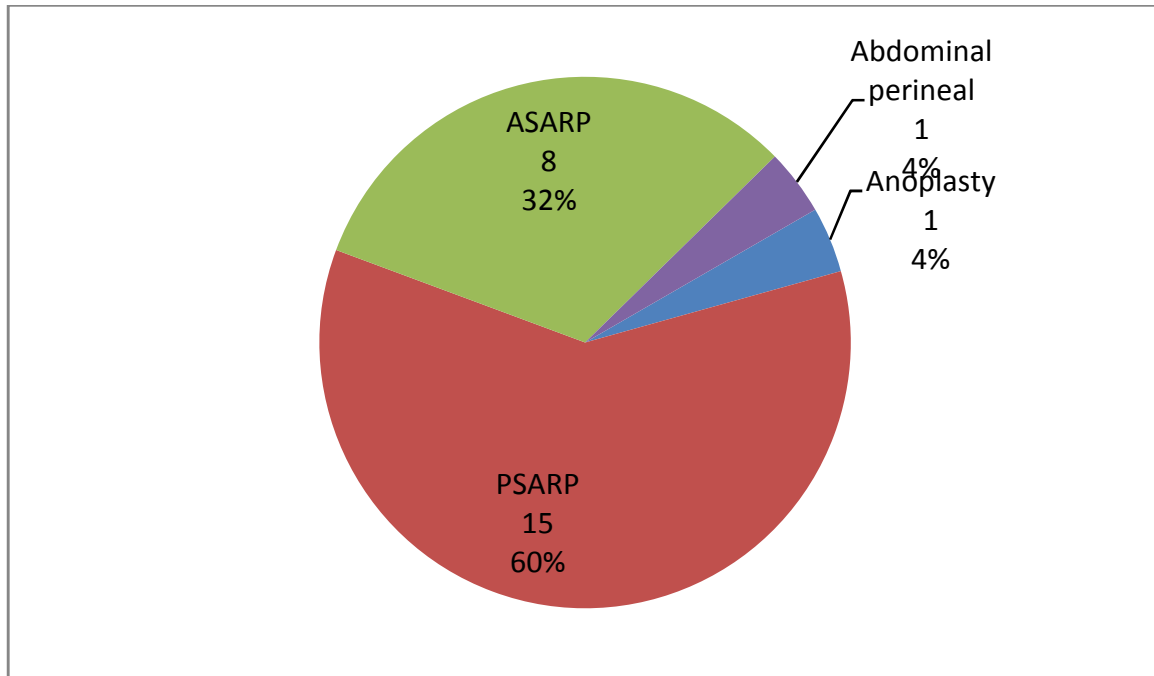


Figure 4: Types of Surgical approaches

All participants who underwent ASARP had a good functional outcome. Those who had PSARP done had a varied outcome shown in the table below. ASARP was only performed in patients with recto-vestibular fistulae while PSARP was applied in all varieties of anomalies. Three patients with recto-vestibular fistula underwent PSARP, two had a good functional outcome and one with a poor outcome. The rest are presented in the table below

Table 6 : Association between type of surgery and outcome

Type of corrective surgery		Quality of life score		
		Poor	Fair	Good
Anoplasty	n=1	0	0	1
	%	0.0	0.0	100.0
PSARP	n=15	3	3	9
	%	20.0	20.0	60.0
ASARP	n=8	0	0	8
	%	0.0	0.0	100
Abdominal perineal	n=1	0	0	1
	%	0.0	0.0	100.0

The association between type of surgery and outcome was not significant with a p Value of 0.608.

The patient who had an AP approach is 35 years old currently and has a 5 year old daughter, who is also a participant in this study, with ARM with a recto-vestibular fistula.

Redo surgeries were done in 8(32%) participants and of these 87.5% were due to ectopic neo anus. This was confirmed at nerve stimulation prior to colostomy closure. One child had incontinence of stool after closure of colostomy and was found to have an ectopic anus. One other patient had redo surgery due to stenosis of the neo anus. This occurred as a result of inadequate dilatation upon discharge after corrective surgery.

One participant had redo surgery twice ,the first due to an ectopic neo anus and later due to mucosal prolapse. Three out of the eight (37.5%)of patients who underwent redo surgery had a good functional outcome as noted using both the Kreckenbeck scoring and objective functional scoring. Three had a fair outcome and two had a poor outcome. The association

between redo surgery and final objective outcome did not show statistical significance, P value 0.105.

At examination two participants (8%) were found to have mucosal prolapse and two others(8%)an absent anal wink, while twenty one had a normal perineal exam. A total of 21 (84 %) participants had good voluntary bowel movement according to the Kreckenbeck score. The 3 participants with poor voluntary bowel movement had grade 3 soiling and a poor objective functional score. Of the 44% who had varied degrees of soiling, 28% had grade 1 and 2 soiling and were managing with bowel management program. Participants with grade 3 soiling were offered further management by referral to the paediatric surgical clinic.

Table 7: Measures of Association between type of Surgery and outcome

Type of corrective surgery	Quality of life			p-value
	poor	fair	good	
Anoplasty	0(0.0%)	0(0.0%)	2(100%)	0.608
PSARP	3(23.1%)	3(23.1%)	7(53.8%)	
ASARP	1(11.1%)	0(0.0%)	8(88.9%)	
Abdomino-perineal	0(0.0%)	0(0.0%)	1(100%)	

Table 8 : Observed Grades of soiling

Soiling grade (n=11)	Frequency
Grade 1	5 (20.0)
Grade 2	2 (8.0)
Grade 3	4 (16.0)

Constipation was experienced in 6 participants (24%)

Constipation grade (n=6)	
Grade 1	4 (66.7)
Grade 2	1 (16.7)
Grade 3	1 (16.7)

Table 9 : Observed Grades of Constipation

Participants with grade one and two constipation managed well by diet and occasional medication, those with grade 3 were dependent on stool softeners.

In the final objective functional scoring system the participants had encouraging results with 18 (72%) showing good functional outcome. Four participants (16%) had fair outcome. They experienced occasional soiling but had no social problems as they managed this mainly with a stooling program. 3 participants (12%) had a poor outcome with constant soiling. Of these only two were recommended for further examination and intervention. One of them had a MACE done and the other was to undergo nerve stimulation with anal mapping then possible Redo PSARP.

Table 10 : Overall observed functional outcome

Variable	Frequency (%)
Poor	3(16.0)
Fair	4 (12.0)
Good	18 (72.0)

All participants with a good bowel control had corrective surgery and closure of colostomy by the age of two years in this study.

The four patients with fair outcome had closure of colostomy at an average age of four years.

From this study the outcomes for our patients was noted to be affected by the age at colostomy closure, type of anomaly and whether redo surgery was done or not.

5.0 CHAPTER FIVE: DISCUSSION

Quality Of Life post corrective surgery for Anorectal Malformation is a good marker to determine the quality of surgical care offered to this group of patients. The long term goal for these patients is to be faecally continent and to have normal bowel movement.

This study reviewed 25 patients aged between 4 and 35yrs. The overall quality of life was found to be good in 72 percent of the participants which is comparable to previous studies by Rintala et al who found normal control with no social restrictions in 64% of his patients. Kigo et al in 2000 also described normal bowel movement was achieved in 71.5% of his participants.

Alberto Pena in his several studies observed 71-77% good bowel control in patients with a normal sacrum.

Nixon found normal bowel control in 74% of his patients which still compares with these results.

Most of the earlier studies found favourable outcomes in patients with low anomalies and this is confirmed in this study where 92% of those with low anomalies (recto-vestibular and recto-perineal fistulae) had good functional outcomes. Kaworski reported 80% continence in patients with low malformations.

Constipation was described as the most common early complication seen in 40% of those with low anomalies^{17,18,19}. This study observed constipation in 24% of the participants.

Kigo et al described females having had better bowel control after corrective surgery, 77% compared to 63% in males. This study found 90.9% of females and 53% of males as having good bowel control.

The level of anomaly is an important prognostic factor in terms of bowel control.

Table 11: Comparison of Outcomes in various Anomalies

Level of anomaly	Kigo et al	This study	Alberto Pena
Rectovestibular	76%	90.9%	93%
Recto urethral	73.9%	37.5%	60-80%
Recto vesical	25%	0%	15%
Without fistula	25%	100%	80%

Re operation is a major cause undesired outcomes. In this study only 37.5% had normal bowel function amongst patients who had undergone any form of redo surgery.

Rintala found 83% of patients with high Anorectal Malformation reported social disability while this study noted 67% had fair to poor outcomes. These patients had recto urethral bulbar, prostatic or vesical fistulas in this study.

The objective functional scoring system was noted to be a good follow up tool in our set up with comparable results to the Krickenbeck scoring system.

We cannot conclusively say the association between age and other related variables to outcome is not statistically significant due to the Skewness of our sample data.

5.1 Conclusion

From this study quality of life for patients after surgery for anorectal malformation is comparable to other studies. Proper newborn management and early definitive surgery followed by closure of colostomy before the age of two years will improve the outcomes of ARM in KNH.

Meticulous sphincter dissection and anatomical reconstruction is vital in the achievement of voluntary bowel movement. Redo surgeries were mainly done due to ectopic neo anus which directly relates to the surgical technique.

ASARP is a good alternative for recto-vestibular fistula and seems to have better outcomes.

Seventy two percent of the participants had good bowel control. It is therefore possible for children with Anorectal Malformation to be clean of stool either from having continence or

by being subjected to a bowel management program as seen in the children with a fair outcome.

5.2 Recommendations

A good follow up system has to be put in place in our paediatric surgery outpatient clinic so as to standardise the follow up care and enable us to pick the functional problems our patients experience. Therefore a standard tool should be adapted for follow-up.

ASARP should be embraced by all surgeons for correction of recto vesical fistula. It has better outcomes and is a good alternative to PSARP.

Most of the patients in this study had been lost to follow up due to poor communication. There is need to emphasize to patients the importance of follow up visits and encourage them to attend.

A bowel management team should be established in KNH to assist in the follow up management of these patients hence improve their Quality of life.

REFERENCES

- 1.) Rintala RJ, Pakarinen MP. Outcome of anorectal malformations and Hirschsprung's disease beyond childhood. *Semin Pediatr Surg* 2010 May;19(2):160-7 .
- 2.) Levitt MA, Peña A. Anorectal malformations. *Orphanet J Rare Dis.* 2007; 2: 33. Published online 2007 July 26.
- 3.) Pena A, Levitt MA. Anorectal Malformations . Springer surgery atlas series, , printed 2006, chapter 27 .
- 4.) Kigo CN. Bowel function following Posterior Saggital Repair of Anorectal Malformations in Kenyatta National Hospital 2001
- 5.) Hasink EA, Rieu PN, Brugman AT et al. Quality of Life after operatively corrected high Anorectal Malformation: a long term follow up study in patients 18 of age and older. *J. Paediatric Surgery* 1994; 29: 773-6
- 6.) Rintala RJ, Pakarinen MP. Long term outcomes of Hirshsprung's Disease. *Seminars inn pediatric Surgery* (2012) volume 21, 336-343
- 7.) Langer JC. Principle and Practice of Pediatric Surgery. [ed.] Colombani PM, Foglio RP, Skinner MA Oldham KT. 4th. Philadelphia : Lippincott, Williams and Wilkins, 2005. pp. 1348-63. Vol. 2.
- 8.) Rintala R, Mildh L, Lindahl H. Fecal continence and quality of life in adult patients with an operated high or intermediate anorectal malformation. *J. Pediatric Surg* 1994;29:777-780
- 9.) Heikkinen M, Rintala RJ, Louhimo I . Bowel function and quality of life in adult patients with operated Hirschsprung's disease. *Pediatr Surg Int*, 10 (1995), pp. 342–344.
- 10.) Diseth TH, Bjornland K, Novik TS *et al.* Bowel function, mental health, and psychosocial function in adolescents with Hirschsprung's disease .*Arch Di Child*, 76 (1997), pp. 100–106.
- 11.) Bai Y, Chen H, Hao J *et al.* Long-term outcome and quality of life after the Swenson procedure for Hirschsprung's disease. *J Pediatr Surg*, 37 (2002), pp. 639–642.

- 12.) Drossman D, Sandler R, McKeem D . Bowel patterns among subjects not seeking health care. *Gastroenterology*, 83 (1982), pp. 529–534.
- 13.) DeVries PA, Peña A. Posterior sagittal anorectoplasty. *J Pediatr Surg*, 17 (1982), pp. 638–643)
- 14.) Rintala RJ, Lindahl H. Is normal bowel function possible after repair of intermediate and high anorectal malformations. *J Pediatr Surg*, 30 (1995), pp. 491–494)
- 15.) Karkowski J, Pollock WF, Landon CW. Imperforate anus Eighteen to thirty year follow-up study. *Am J Surg*, 126 (1973), pp. 141–147.
- 16.) Nixon HH, Puri P. The results of treatment of anorectal anomalies: a thirteen to twenty year follow-up. *J Pediatr Surg*, 12 (1977), pp. 27–37.
- 17.) Pakarinen MP, Goyal A, Koivusalo A *et al.* Functional outcome in correction of perineal fistula in boys with anoplasty versus posterior sagittal anorectoplasty. *Pediatr Surg Int*, 22 (2006), pp. 961–965.
- 18.) Pakarinen MP, Koivusalo A, Lindahl H *et al.* Prospective controlled long-term follow-up for functional outcome after anoplasty in boys with perineal fistula. *J Pediatr Gastroenterol Nutr*, 44 (2007), pp. 436–439.
- 19.) Rintala RJ, Lindahl HG, Rasanen M. Do children with repaired low anorectal malformations have normal bowel function?. *J Pediatr Surg*, 32 (1997), pp. 823–826.
- 20.) Peña A. Anorectal malformations. *Semin Pediatr Surg*, 4 (1995), pp. 35–47.
- 21.) Rintala R, Lindahl H, Marttinen E *et al.* Constipation is a major functional complication after internal sphincter-saving posterior sagittal anorectoplasty for high and intermediate anorectal malformations. *J Pediatr Surg*, 28 (1993), pp. 1054–1058.
- 22.) Holschneider AM, Pfrommer W, Gerresheim B. Results in the treatment of anorectal malformations with special regard to the histology of the rectal pouch. *Eur J Pediatr Surg*, 4 (1994), pp. 303–309.
- 23.) Langemeijer R.A.T.M, Molenaar JC. Continence after posterior sagittal anorectoplasty. *J Pediatr Surg*, 26 (1991), pp. 587–590.

- 24.) Husberg B, Lindahl H, Rintala R *et al.* High and intermediate imperforate anus: Results after surgical correction with special respect to internal sphincter function. *J Pediatr Surg*, 27 (1992), pp. 185–189.
- 25.) Rintala R, Marttinen E, Virkola K *et al.* Segmental colonic motility in patients with anorectal malformations. *J Pediatr Surg*, 32 (1997), pp. 453–456.
- 26.) Mollard P, Meunier P, Mouriquand P *et al.* High and intermediate imperforate anus: functional results and postoperative manometric assessment. *Eur J Pediatr Surg*, 1 (1991), pp. 282–286.
- 27.) Rintala R, Lindahl H, Louhimo I. Anorectal malformations - results of treatment and long term follow-up of 208 patients. *Pediatr Surg Int*, 6 (1991), pp36-
- 28.) Holschneider AM, Frommer WP, Gerresheim B. Results in the treatment of Anorectal Malformations with regard to histology of the rectal pouch. *Eur. J paediatric Surg*, 4 (1994), pp 303-309
- 29.) Templeton JM, Ditesheim JA. High imperforate anus - quantitative result of long-term fecal continence. *J Pediatr Surg*, 20 (1985), pp. 645–652.
- 30.) Holschneider AM. *Elektromanometrie des Enddarms.* Urban & Schwarzenberg, Munich (1983), pp. 213–218.
- 31.) Rintala R, Mildh L, Lindahl H. Fecal continence and quality of life in adult patients with an operated low anorectal malformation. *J Pediatr Surg*, 27 (1992), pp. 902–905.
- 32.) Ditesheim JA, Templeton Jr JM. Short-term vs. long-term quality of life in children following repair of high imperforate anus. *J Pediatr Surg* 1987;22:581 - 587.
- 33.) Bai Y, Yuan Z, Wang W, et al. Quality of life for children with fecal incontinence after surgically corrected anorectal malformation. *J Pediatr Surg* 2000;35:462 - 4.

APPENDICES

Appendix I: Questionnaire

QUESTIONNAIRE FOR THE QUALITY OF LIFE AFTER CORRECTIVE SURGERY FOR ARM

Questionnaire No.

1. Demographic data

a) residence

b) Sex

c) Current age

d) Age at diagnosis.....

e) Age at colostomy fashioning if applicable

f) Age colostomy was closed

g) Associated anomalies (Y/N)

- Sacral/vertebral anomalies
- Cardiac anomalies
- Kidney , ureter and bladder anomalies.....
- Any other anomalies

2 .Anorectal malformation

a) Symptoms at presentation (tick one)

i) Acute intestinal Obstruction

ii) Chronic constipation

iii) Abnormal Opening

b) Type of anomaly (tick one)

- Rectovestibular no fistula
- Recto urethral bulbar Cloaca
- Rectourethral prostatic
- Rectoperineal

c) Type of corrective surgery (tick one)

- i) Anoplasty iv) ASARP.....
- ii) Minimal PSARP v) PSARVUP
- iii) PSARP

d) Was any Re-Do surgery done (yes/No)

if Yes what was done

e) Assessment of outcome (Krickenbeck)

- 1. Voluntary bowel movements yes/no
Feeling of urge
Capacity to verbalize
Hold the bowel movement
- 2. Soiling yes/no
Grade 1: occasionally (1 to 2/week)
Grade 2: every day
Grade 3: constant, social problem
- 3. Constipation yes/no
Grade 1: manageable with diet
Grade 2: requires laxatives
Grade 3: resistant to diet and laxatives

3. Perineal examination findings (yes/no)

- 1) Mucosal prolapse
- 2) Anal wink
 - a. Present or absent
 - b. If present, Circumferential or ectopic

4. Scoring system for objective functional outcome

Score	1	2	3		
1.Recurrent abdominal distension		Mild	Moderate	Severe	
2.Frequency of defecation		1-2/d	3-5/d	>5/d	
3.Stool consistency	Normal	Loose	Liquid		
4.Soiling	None	Occasionally	Permanently		
5.Urgency period	Normal	Short	Absent		
6.Diaper required		None	Occasionally	Permanently	
7.Long term use of medication		None	Antibiotics	Antidiaroheal	
8.Diet	Normal	Restricted	TPN		

Range of score

Poor----- above 16

Fair ----- 9 to 16

Good----- 0 to 8

Participants final score _____

Appendix II: KNH/ERC Letter of Approval



UNIVERSITY OF NAIROBI
COLLEGE OF HEALTH SCIENCES
P O BOX 19676 Code 00202
Tel: 726300-9
(254-020) 2726300 Ext 44355

KNH/UON-ERC

Email: uonknh_erc@uonbi.ac.ke
Website: <http://www.erc.uonbi.ac.ke>
Facebook: <https://www.facebook.com/uonknh.erc>
Twitter: @UONKNH_ERC https://twitter.com/UONKNH_ERC



KENYATTA NATIONAL HOSPITAL
P O BOX 20723 Code 00202
Tel: 726300-9
Fax: 725272

0454290

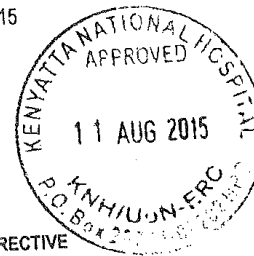
Bill No: 590890

Ref: KNH-ERC/A/345

Dr. Swaleh H. Shahbal
Dept. of Surgery
School of Medicine
University of Nairobi

Dear Dr. Swaleh

11th August 2015



RESEARCH PROPOSAL – ASSESSMENT OF QUALITY OF LIFE IN CHILDREN AFTER CORRECTIVE SURGERY FOR ANORECTAL MALFORMATION AT KENYATTA NATIONAL HOSPITAL (P77/02/2015)

This is to inform you that the KNH/UoN-Ethics & Research Committee (KNH/UoN-ERC) has reviewed and **approved** your above proposal. The approval periods are 11th August 2015 – 10th August 2016.

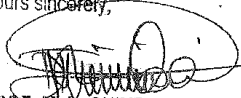
This approval is subject to compliance with the following requirements:

- Only approved documents (informed consents, study instruments, advertising materials etc) will be used.
- All changes (amendments, deviations, violations etc) are submitted for review and approval by KNH/UoN ERC before implementation.
- Death and life threatening problems and serious adverse events (SAEs) or unexpected adverse events whether related or unrelated to the study must be reported to the KNH/UoN ERC within 72 hours of notification.
- Any changes, anticipated or otherwise that may increase the risks or affect safety or welfare of study participants and others or affect the integrity of the research must be reported to KNH/UoN ERC within 72 hours.
- Submission of a request for renewal of approval at least 60 days prior to expiry of the approval period. (Attach a comprehensive progress report to support the renewal.)
- Clearance for export of biological specimens must be obtained from KNH/UoN-Ethics & Research Committee for each batch of shipment.
- Submission of an executive summary report within 90 days upon completion of the study. This information will form part of the data base that will be consulted in future when processing related research to assess scientific merit, eliminate duplication and/or plagiarism.

For more details consult the KNH/UoN ERC website <http://www.erc.uonbi.ac.ke>

Protect to discover

Yours sincerely,



PROF. W. L. CHINDIA
SECRETARY, KNH/UoN-ERC

- c.c. The Principal, College of Health Sciences, UoN
The Deputy Director CS, KNH
The Chair, KNH/UoN-ERC
The Dean, School of Medicine, UoN
The Chairman, Dept. of Surgery, UoN
Supervisors: Mr. Francis Osawa, Mr. J. Ndung'u