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PATTERN OF ANORECTAL MALFORMATIONS AND EARLY OUTCOMES OF MANAGEMENT AT MOI TEACHING AND REFERRAL HOSPITAL ELDORET- KENYA
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ABSTRACT

***Objectives*: To describe the anatomical sub-types of Anorectal malformations, their management and the early outcome at Moi Teaching and Referral Hospital (MTRH) over a 16 month period.**
***Design*: A prospective study.**
***Setting*: MTRH, in the neonatal Unit and paediatric surgical wards for the initial capture of patients and initial follow up. The Paediatric Surgical out- patient clinic was used for the subsequent follow ups.**
***Subjects*: All infants diagnosed with ARM (Anorectal malformations) at MTRH from November 2011 to April 2013.**
***Main outcome measures*: Sub-types of the Anorectal malformations, coexisting abnormalities morbidity and mortality rates.**
***Results*: There were 42 participants including 24 (57%) males and 18 (43%) females. Neonates presented at an average age of 4±3, three days and older children presented on average age of 152±118, three days. There were 30 (71%) neonates and 12 (29%) older infants. In males, the predominant sub-type was imperforate anus without a fistula found in ten participants (42% of males). In females, the predominant sub-type was recto-vestibular fistula found in 14 participants (78% of females). Mortality occurred in 13 (31%) participants among them ten (24%) had coexisting abnormalities. The main causes of morbidity were: colostomy complications in four (9.5%); wound infections in one (5%); and wound dehiscence in one (5%).**
***Conclusions*: Patients with Anorectal malformations presented late at MTRH. The diagnosis at birth was missed in babies born at home as well as those delivered in health institutions.**

INTRODUCTION

Anorectal malformations comprise a wide spectrum of congenital malformations affecting both boys and girls and may involve the rectum, anal canal and uro-genital tracts (1,2).

They remain a significant birth defect with geographic variation in incidence and sub-types (3). It occurs globally in approximately one out of 5000 live births. It is one of the most common surgical congenital anomalies (1,2). In Africa, ARM is aleading cause of neonatal intestinal obstruction (1,4). There is male predominance of 55-65% of all ARM5. High anomalies also occur more frequently in males. Females commonly have low anomalies and hence good outcome after definitive reconstruction (6). Reconstruction has evolved over time to the current Pena's procedureor PSARP (Posterior Sagittal Ano-rectoplasty) and its modification ASARP(Anterior Sagittal Ano-Recto Plasty) which are associated with low morbidity and low mortality (1,3,7). Factors that negatively affect outcomes in developing countries have been identified as: delayed presentation at referral institution, poor infrastructure, scarcity of expertise, parental ignorance among others (8,9). These factors could impact negatively on expected local outcomes in comparison to the developed world.

MATERIALS AND METHODS

This was a prospective study done from November 2011 to April 2013 at MTRH, in the new-born Unit and paediatric surgical wards for the initial capture of patients and initial follow up. The Paediatric Surgical out- patient Clinic was used for the subsequent follow-ups. All infants diagnosed with ARM at MTRH during study period constituted the study population,

Using a census approach, all 42 infants treated for ARM from November 2011 to April 2013 were recruited and included into the study. Anorectal malformations operated outside MTRH were excluded.

Procedure: Consecutive patients presenting with ARM were enrolled into the study following parental informed consent. Investigations were done to confirm the ARM sub-types and when positive clinical findings were suggestive of coexisting abnormalities. Anoplasty or anal dilatation was performed for low ARM in neonates. Three-staged interventions (colostomy, PSARP or ASARP, colostomy closure) were done in neonates with high, intermediate ARM and all older infants. Standard PSARP or ASARP are methods of ARM reconstruction in staged interventions at MTRH.

The main outcome measures were: demographic distribution (age, gender), sub-types of the Anorectal malformations, coexisting abnormalities, morbidity and mortality rates. These outcomes were recorded within one month after colostomy closure (three-staged operations) or primary Anoplasty (one stage).

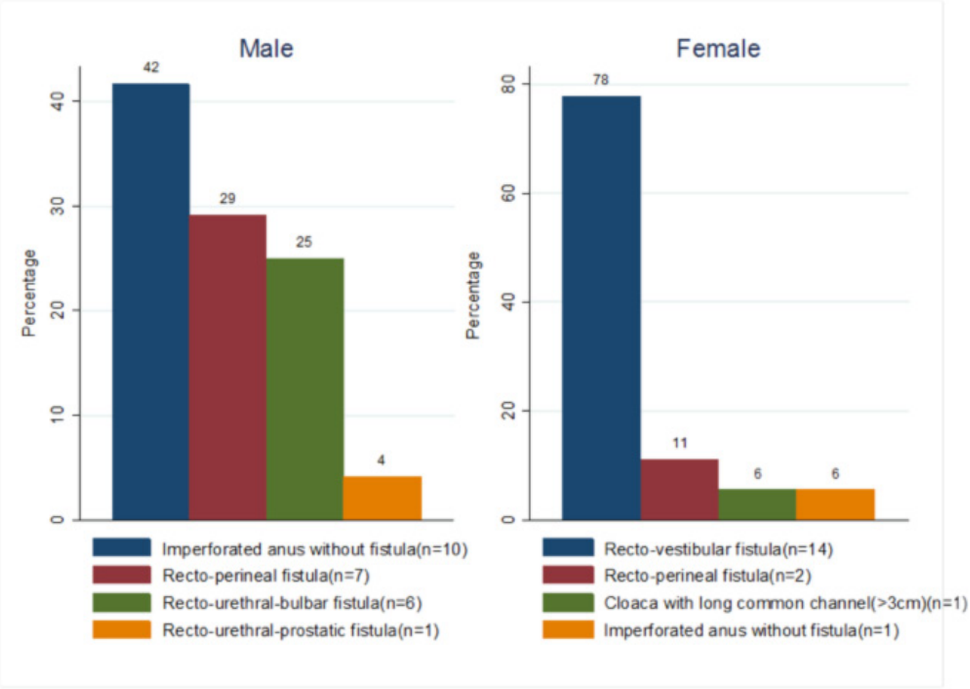
Ethical consideration: Approval of the study was obtained from the Institutional Research and Ethics Committee (IREC) before starting the study.

RESULTS

Records of 42 participants with Anorectal malformations were analysed. Male participants were 24 (57%) and females 18 (43%) hence a M:F ratio of 1.3:1.There were 30 (71%) neonates and 12(29%) infants. Only one was diagnosed at birth at MTRH out of three delivered in the same institution, 39 other patients were referred from peripheral health facilities. Most of infants had been delivered at home (62%). The neonates presented at an average age of four (std: 3.3) days with a median age of three (IQR: 2-5) days. Older children presented at 152 (std: 118.3) days on average with a median age of 105 (IQR:50-255) days.

Anorectal malformations sub-types diagnosed at MTRH: Imperforate anus without fistula dominated ARM anatomical sub-types in males and most of female participants had recto-vestibular fistula anomaly Figure 1.

Figure 1
Distribution of ARM sub-types per gender



Co-existing abnormalities: Co-existing abnormalities identified in this study were as represented in Table 1.

Table 1
Distribution of coexisting abnormalities

Characteristic	n (%)
Total Number of participants	42
CNS	6(15%)
Urogenital	7(17%)
Cardiac	3(7%)
Gastro- intestinal	3(7%)
Limbs	6(15%)

Some children were seen with more than one coexisting abnormality. Detailed CNS associated congenital abnormalities were encephalocele: one (2.4%), hydrocephalus: two (4.8%), spina bifida two (4.8%) and anostia on left ear: one (2.4%). Urogenital defects: horseshoe kidney: one (2.4%), left renal agenesis: one (2.4%), hydronephrosis: one (2.4%), hypospadias: two (4.8%), undescended testis: four (9.6), vesico-ureteric reflux that had caused chronic renal failure: one (2.4%). Cardiac abnormalities included: atrial septal defect: one (2.4%), persistent ductus arteriosus: one (2.4%) and one (2.4%) who had a significant heart murmur died before investigations were completed. Gastro intestinal abnormalities included one (2.4%) who had duodenal atresia and colonic atresia, Meckel's diverticulum: one (2.4%) and umbilical hernia: one (2.4%). Limb abnormalities were: congenital talipes-equino -varus: one (2.4%), amelia of right lower limb: one (2.4%), radial dysplasia: one (2.4%), polydactily: two (4.8%) and one (2.4%) had syndactily (only three fingers on each hand).

Investigations: A few radiologic investigations or Ultra sound were done: plain X-ray: 28 (67%), Ultrasound abdomen and pelvis: 13 (31%), Distal colostogram: 4 (9.5%), Echo (cardiac): 3 (7%).

Types of operations: Only one participant with Recto-vestibular fistula was awaiting colostomy. Among the 38 infants who were operated, there were 29 neonates and nine children. Thirty one (73.8%) underwent staged intervention: first stage colostomy, second stage: PSARP or ASARP and final third stage colostomy closure. Six (14.3%) neonates with low anorectal malformations underwent one stage definitive anoplasty.

The initial surgical intervention was performed at average age of five days for neonates and 154 days for older infants. Twelve second stage surgical interventions (ASARP or PSARP) was done at seven months average (Mean = 211 (std. 111) days). Median = 225 (IQR: 180-300) days).Third stage (colostomy closure) was performed at average age of one year (n = 5. Mean = 379 days (STD: 33.62), Median = 390 days (IQR: 365-390).

Complications: The post-operative complications included: neonatal sepsis in seven (17%) participants and hypothermia with hypoglycemia in one (2.4%) participant. Wound infection and wound dehiscence were present in three (7%) and two (5%) participants respectively. Among the 31 patients who had colostomy, four (10%) patients developed colostomy complications: 1(2.4%) had prolapse and three (7.4%) had retraction of the colostomy.

Mortality: There were 13(31%) deaths: ten of whom were neonates. The causes of death were found to be neonatal sepsis: nine (69.2%), hyperkalemia: one (7.7%), hypothermia and hypoglycemia: one (7.7%), meningitis: one (7.7%) pneumonia in severe malnutrition: one (7.7%).Among the recorded deaths, ten (77%) had one or more coexisting abnormalities. The mortality occurred after 71 days on average from the day of the first intervention (n = 13. Mean = 71 (118.3) Median = 30 (7 – 39)

Factors associated with poor outcome: The morbidity and mortality were significantly higher in the group of infants with coexisting abnormalities (p-value=0.003) .The comparisons of morbidity and mortality between neonates and older infants, and between male and female participants were not significantly different.

DISCUSSION

There was a male predominance (M:F 1.3:1) similar to global reports (Ratio M:F is 1.1:1-1.3:1) (5). Similar to finding from elsewhere in the developing world, late presentation was a significantfactor associated with high mortality recorded in this study.

Delay is due to several preventable factors; majority of births occur at home where birth attendants cannot identify ARM early enough coupled with poor referral system which is associated with inherent delay at every stage (8).In this study, the diagnosis was missed at birth even in those born in health facilities. Delayed presentation leads not only to complications of neonatal intestinal obstruction but also disrupts the standard care plans. For example investigations to identify the level of rectal pouch on the first day of life become irrelevant after 48 hours (1,2). Other consequences of delay include; sepsis, aspiration pneumonia, respiratory embarrassment occasioned by massive abdominal distension, colonic perforation, electrolyte imbalance and death (3,9,10).

Empowering pregnant women to deliver in health institutions should lead to early diagnosis, proper resuscitation and early referral of babies born with this anomaly. Systematic examination of

neonates should be done to rule out any congenital anomalies and immediate referral to specialized units should be undertaken when any anomaly is identified.

Geographical variations in distribution of ARM sub-types occur. In this study, Recto-vestibular fistula was the most predominant sub-type in females similar to what has been reported globally (1,2). Imperforate anus without fistula was found to be predominant in males. Pena *et al*. reports indicate ARM with recto-urethral fistula to predominate in males (1,2). Similarly to this study, North African and Pakistan studies reported predominance of imperforate anus without fistula in males (11,12). Other studies have reported recto-urethral fistula as the most common subtype in males (1,2).

The standard ARM surgical reconstruction is globally based on PSARP or its variants ASARP. Divided proximal sigmoid colostomy is applied at MTRH as described by Pena and is associated with low rate of complications (13). Exteriorising the stoma and mucous fistula through separate openings with an intact skin bridge has minimised breakdowns of the incision,

High mortality of 31% was recorded in this study. Morbidity was equally significantly higher than what is reported in studies from developed countries. Haider *et al*. in England reported 4% mortality in a study of 52 cases that was associated with delayed presentation (10). Available studies from Africa report significant mortality. Uba *et al*. reported 26 % mortality rate in a study of 82 cases of ARM at Jos University Teaching Hospital in Nigeria. This mortality rate is almost similar to that found in this study (14).

Coexisting abnormalities were investigated with imaging when physical examination was suggestive. Globally, abnormalities of VACTERL group are systematically investigated from the time of diagnosis. In this study, coexisting abnormalities was a factor associated with significant mortality and morbidity similar to what is reported globally (1,2,15).

Participants with one or more coexisting abnormality had higher risk of developing complications and mortality. Coexisting abnormalities are recognised globally to impact negatively on ARM management.A meticulous search for coexisting abnormalities is necessary once the diagnosis of ARM is made (1,2,15).

In conclusion, patients with Anorectal malformations presented late at MTRH. The diagnosis at birth was missed in some babies delivered in health institutions. Morbidity was high and mortality of 31% was higher than that found in western series but similar to that reported in the developing countries (10,14). Coexisting congenital abnormalities were a significant factor associated with poor early outcomes.

Recommendation: Mechanisms should be put in place to enable early diagnosis and presentation by institutionalising a comprehensive physical examination of all newborns against a checklist of congenital anomalies.

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