INCIDENCE, MANAGEMENT OPTIONS AND OUTCOMES OF VESTIBULAR ANORECTAL MALFORMATIONS IN PAEDIATRIC FEMALE PATIENTS AT DR GEORGE MUKHARI ACADEMIC HOSPITAL

By

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Declaration

I, Mahumaneng Esther Mokaila, hereby declare that this research is my work. It is being submitted for the degree of Master of Medicine in the branch of Paediatric Surgery at the Sefako Makgatho Health Sciences University. It has not been submitted before for any degree or examination at any other University.

____________________
Signed

17 October 2018
Acknowledgement

I would like to express my gratitude to the following individuals for assisting me with the protocol, data collection and putting the research together. Your invaluable support is much appreciated.

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3. Professor O. Towobola, for assisting with protocol and the statistical analysis.
4. Mr Matlou Mabitsela – our research assistant – for helping to ensure quality work.

My family for their support and sacrifices during this period.
TABLE OF CONTENTS

Declaration ........................................................................................................................................... i

Acknowledgement ............................................................................................................................... ii

List of Figures ....................................................................................................................................... iv

List of Tables ......................................................................................................................................... v

Abstract ............................................................................................................................................... vi

Chapter 1: Introduction and Background ........................................................................................... vii

Chapter 2: Literature Review .............................................................................................................. 3

Chapter 3: Methods .............................................................................................................................. 7

Chapter 4: Results ............................................................................................................................... 15

4.1. Introduction ................................................................................................................................. 15

4.2. Age distribution of the children seen during the study period ................................................. 15

4.3. Symptoms .................................................................................................................................... 17

4.4. Complications ............................................................................................................................. 18

Chapter 5: Discussion & Conclusion ................................................................................................. 24

References ............................................................................................................................................ 33

Appendices A: Data Collection Sheet ................................................................................................. 37

Appendices B: Ethics Approval Certificate ......................................................................................... 38
List of Figures

Figure 1: Age distribution of the children at the time of presentation in DGMAH… 15

Figure 2: Ages of the children at first procedure to correct the malformation……. 16

Figure 3 types of procedures performed for the children in this series……… 18
List of Tables

Table 1: Symptoms recorded for the children at the time of presentation….. 17

Table 2a: Anastomotic Breakdown…………. 18

Table 2b: Bleeding as a complication…………… 19

Table 3: Anal Stricture (late complication – 4-week follow-up)…………… 19

Table 4a: Three-month follow-up: Anal Stricture…………… 20

Table 4b: Three-month follow-up: Closure of colostomy…………… 20

Table 5: Six-month follow-up: Correction of malformation…………… 21

Table 6: One-year follow-up…………… 22
Abstract

**Study Objective:** To determine the incidence, management options and outcome of Vestibular Fistula anorectal malformation in paediatric female patients presenting at Dr George Mukhari Academic Hospital.

**Design:** Retrospective review for 5-year period

**Participants:** During the study period, 54 patients who met the inclusion criteria were identified from the hospital record, however, 18 patients were excluded from the study due to incomplete information in the files. The final number of patients included in the study was 36.

**Methods:** The data was retrieved from our hospital Records department. We looked at the patients demographics, presenting symptoms and signs and management. All patients had VACTREL workup including an abdominal X-ray, to rule out a dilated recto-sigmoid. The children were initially resuscitated and fistulas were dilated to decompress the bowel. They were then offered different surgical options. These options were 1-Stage group (anoplasty), 2-Stages group (anoplasty and stoma initially and later-on stoma closure) and 3-Stages group (stoma, followed by anoplasty at later date and finally stoma closure at even later date). We looked at the surgical option groups offered and their outcome.

**Results:** The incidence rate during this time period was 8.23% (82 cases per 1000 patients seen in the unit at Dr George Mukhari Academic Hospital). The majority of our patients were early presenters at 69.4% (n=25). Most patients (63.9%; n=24) presented with abnormal perineum and functional fistula and 16.7% (n=6) presented for constipation. Four patients (11.2%) presented with bowel obstruction. One patient (2.8%) had associated duodenal atresia. The majority of our patients (69.4%) were
offered a 3-stage procedure, 22.2% was offered 2-stage procedure and 8.3% was offered 1-stage. Post-operative complications differed among the various option groups offered to our patients. In 3-Stage group, one patient (4%) had an anastomotic breakdown as an immediate complication. On post-anoplasty procedure 4 week follow-up, 6 patients had anal stricture, 3 defaulted follow-up, 1 had recurrent fistula, and 1 died unrelated to malformation. During the six month follow-up, assessing correction of malformation, 1 patient from the 1-stage group had no correction of the malformation and was converted to a 3 stage procedure. In 3-stage group, 2 patients had recurrent fistula. At 1-year follow-up, majority of the patients from all the groups where ready for toilet training. In 1-stage group, 1 patient defaulted clinic. In 3-stage group there were numerous complications (1 patient defaulted, 2 patients had persistent fistula, 1 patient reappeared with anal stenosis after defaulting from hospital for long time and 4 patients had anal strictures).

**Conclusion:** In our practice the incidence of vestibular fistula was high compared to literature. The majority of patients were early presenters, were offered 3-stage procedure and 2Stage group had the best outcome. The practice of 1Stage was associated with more severe complications to the girl child.
Chapter 1: Introduction and Background

Anorectal malformation (ARM) is one of the most common congenital anomalies seen at Dr George Mukhari Academic Hospital (DGMAH). There was a wide spectrum of presentation, ranging from low anomalies with perineal fistula with simple management to high anomalies with complex management (1). In malformation occurring in females, ARM (vestibular fistula) is the most common form of the condition and it presents with the best prognosis (2).

At DGMAH, neonates are often detected while in hospital but when diagnosis is missed during the neonatal period, the infants are bought to the hospital as late presenters. Possibility of late presentation of the condition was often due to cases of ARMs being asymptomatic at the time the neonate was discharged from hospital after birth, or no proper examination of perineum was undertaken during the birth of the neonate (3).

Studies indicated that early detection and management of the vestibular fistula condition was the key to better outcomes for two reasons (4):

- Surgery done in infant pelvis was found to be more convincing in ensuring good quality of life because at this stage the sphincter muscles are soft and easily manageable.
- The cortical integration of the somatosensory input was usually lost with advancing age.

The study was conducted among female patients because male patients tend to present early with either distal bowel obstruction, dehydration or sepsis (2). Early
management was therefore crucial (1). Patients would require an emergency diverting colostomy and those male paediatric patients would all undergo the 3-stage procedure (2). Male neonates presenting with perineal fistula would undergo primary anoplasty if there are no contraindications. There are fewer early complications post-surgery.

At DGMAH it is presently unclear how to subject each case of this malformation to either a single or staged procedure, and how this could influence the degree of clinical success as well as length of hospital stay. However, in our institution it was suggested that every case be managed through a 3-staged procedure involving 1) colostomy, 2) definitive procedure, Anterior Sagittal Anorectoplasty (ASARP), and 3) closure of colostomy (5).

In addition, the study aimed at determining the incidence of the condition, and also at assessing the pattern of management of vestibular fistula at DGMAH, the single-versus multiple-stages procedure performed and clinical outcomes of management.
Chapter 2: Literature Review

ARMs are among the more frequent congenital anomalies encountered in paediatric surgery, with an estimated incidence ranging between 1 in 2000 and 1 in 5000 live births (1).

In females, the vestibular fistula represents the most common type of anorectal defect with associated malformation (2). Vestibular fistula was described by Bryndon and Mardsen in 1960 as a fistulous tract between the bowel and the low female genital tract (6). Sharma et al. (7), in a series from Ireland, reported that despite routine physical examination postpartum, one in five neonates with imperforated anus had a delayed diagnosis. The same authors defined delayed presentation after 3 months of age in cases of low ARM in females in developing countries.

From the reports of both Sharma et al. (7) and Bokhari et al. (4), delayed presentation of ARM, usually in developing countries, was defined to be due to either:

- Inadequate treatment
- Lack of awareness
- Illiteracy
- Wrong treatment advice and social factors such as lack of money or social support
- Lack of basic healthcare facilities

In terms of clinical presentations, Kim et al. (8) reported that there is a consistent finding in terms of clinical presentations and the age of diagnosis. These authors reported that most children suffer from increased chronic constipation and/or being refractory to medical treatment around the time of introduction to solid foods. Bokhari
et al. (4) further elaborated that due to the effects of constipation or delay in alteration of surgical management, clinical outcome could lead to recurrent genital and urinary tract infection and may even lead to infertility. Delayed presenters also suffer from inadequate weight gain and increased parental anxiety for a greater period of time.

It is clear that this condition of ARM, despite its simplicity, was likely to have the highest chance of failure, leading to severe sequelae if proper reconstruction was not done at the first attempt. The reported disadvantages of primary repair include wound infection and wound dehiscence and to avoid those complications, a diverting colostomy was traditionally advised to obtain the best results (8). Nevertheless, colostomies are not without complication, and the cost of a 3-staged operation, especially in developing countries, is immense. Furthermore, the possibility exists of some patients’ being lost to follow-up after the colostomy (9). As a result, a single-stage procedure has been advocated (10). The growing trend, therefore, was to do a single-stage surgery which reduces the total cost of treatment and hospital stay (2,9) and has greater benefit for both the parents and the child. The other arguments favouring single-stage procedure are (1; 11):

- Cerebral cortical fibres develop in the first year of life and sensations of rectal fullness are essential for these fibres to develop fully so that continence can be achieved to its maximal potential.
- Easier dissection in the neonatal period due to virgin tissue planes with no fibrosis due to pouchitis.
- Done in neonatal stages; no need for bowel preparation due to the liquid consistency of the stool and lack of colonisation of the stool.
In terms of pre-operative management, the following protocol was adopted by different study groups: 1) Menon et al. (9) and Sinha et al. (10) initially excluded individuals with pouch colon and rectal ectasia. Mechanical bowel preparation was started on the evening before surgery and involved normal saline irrigation 25ml/kg/hr until rectal effluent was clear. 2) Sharma et al. (4) used mechanical bowel preparation, retention enemas, laxatives and hegar dilators for 3–11 days before theatre. This was followed by the use of a well-lubricated hegar dilator of an appropriate size to break the fecoliths and dilate the fistulous tract to facilitate bowel evacuation (7). Kumar et al. (10) started total irrigation one day before surgery, followed by the use of polyethylene glycol-electrolyte solution (25ml/kg/hr) until rectal effluent was clear.

Single-stage definitive repair of primary posterior sagittal or anterior sagittal anorectoplasty (PSARP/ASARP) is done in most cases as described by Pena et al. in 1982 (12). Post-operative management of the patients includes avoiding per oral feeding; Total Parenteral Nutrition (TPN) is given for 3 to 6 days while patients are being given broad-spectrum antibiotics during this period.

For late presenters beyond five months of age, Sharma et al. (7) have advocated for PSARP to allow for proper tissue dissection and appropriate placement of the non-decompressed rectal pouch within the sphincter complex, as this would allow the perineal body to be repaired satisfactorily with a good cosmetic outcome.

Irrespective of the approach adopted for the single-stage repair, the complications encountered post-operatively have been reported to be similar. Menon and Rao (9) reported the occurrence of anal stenosis due to non-compliance to the dilatation programme by the parents and due to wound infection (4). The other authors have also emphasised the possibility of anterior migration of the neo-anus as a complication.
associated with wound infection and dehiscence in the post-operative period (10). Kumar et al. (5) pointed out that complications occur because of inadequate mobilisation and separation of the rectum from the posterior wall of the vagina, an observation which was reported in 1982 by Pena et al. (12) Tense anastomosis and suspected devascularisation may aggravate the problems (9;5).

Short et al. (11) concluded that a good outcome can be expected following a delayed primary repair of vestibular fistula beyond the neonatal period. The approach of a single-stage procedure safely reduces the need for multiple operations and avoids the potential morbidity of a diverting colostomy. Having a protocol-based management approach provides a greater chance for normal quality of life (9;5).
Chapter 3: Methods

3.1. Study setting

The study was a retrospective audit of cases of paediatric female patients who had been admitted and treated for ARM (vestibular fistula) at DGMAH. It aimed to evaluate the impact of the specific management procedures that were implemented on these patients.

3.2. Study sample and sample size

All paediatric female patients who were admitted between 1 January 2011 and 30 June 2016 constituted the sample population. From departmental estimates of admission for this condition, the total number of cases in the five and a half year review period was approximately 54 paediatric patients. From this number and the projected female proportion of 60%, the expected sample size of this review was 35 patients.

3.3. Inclusion and exclusion criteria

Only female paediatric patients managed during this review period were included in the study. Patients with high ARM, namely cloacal malformation, were excluded from the review.

There was no discrimination regarding the age of the patients to be able to stratify data analysis for early diagnosis and late presenters among the patients. Ages were 1 day of life up to 12 years of age.

3.4. Procedure Performed

- Preoperatively
  
  All the patients had undergone radiological investigation for VACTREL workup and assessment of colon.
In neonatal stage: baby gram and an echocardiogram to exclude other anomalies.

Late presenters: rule out a dilated sigmoid colon.

There were no pre-operative bowel wash-outs as the late presenters had either undergone a 2-stage or 3-stage procedure. Many of these late presenters had suffered from refractory constipation with faecal incontinence, which did not respond to medical treatment.

All fistulas were diluted preoperatively with hegar dilators to help decompress the distal bowel.

All the paediatric female patients had either one of the following surgical management procedures.

- **1-Stage Procedure: Primary ASARP (without protective colostomy)**
  - Done under general anaesthesia. Supine position. Legs in a lithotomy position.
  - Urinary catheter inserted.
  - Given antibiotic cover intra-operatively.
  - The proposed anal site was found using an electric-neurostimulator. Marked the site of the neo-anus with silk suture.
  - Traction sutures used around the fistula site. Circular incision made with the traction suture within it to establish a plane of dissection.
  - This incision extended to the new site of the marked neo-anus, meticulously cutting the external sphincter muscles.
- Recto-vestibular fistula separated from the posterior vaginal wall by meticulous dissection.
- Rectum was pulled through the centre of the sphincter muscles.
- The perineal body was reconstructed.
- The rectum anocutaneous was sutured using vicryl 3/0 circumferential at the site of the muscle complex, with the help of muscle stimulator.

- **Post-operative**
  - Patient kept Nil per Os (NPO), given TPN for 5 days.
  - Intravenous antibiotics given during this duration.
  - Perineal irrigation with normal saline.
  - Anal dilatation resumed only after 4 weeks post-surgery.

- **2-Stage Procedure**
  - **Stage 1**
    - Covering diverting colostomy
    - Anterior sagittal anorectoplasty
  - **Procedure of diverting colostomy**
    - Done under general anaesthesia. Supine position.
    - Given a cover of antibiotics.
    - Cleaned and draped the area of work.
    - Left iliac fossa incision made, oblique.
    - Identification of the sigmoid colon. Proximal part of the sigmoid was divided, stoma is created.
    - Distal part of the sigmoid was flushed and suctioned using normal saline.
    - It was closed and attached to the fascia.
  - **Anterior sagittal anorectoplasty was done in the same setting.**
• **Post-operatively**
  - Covered with antibiotics while in hospital.
  - Once the stoma was functional, patients were started on a normal diet.
  - After 4 weeks post-operative, the neo-anus was dilated.
  
  o **Stage 2**
    - Closure of colostomy was done at least 6 weeks post anoplasty and the anal dilatation programme satisfactory.

• **3-Stage Procedure**
  
  o **Stage 1**
    - Creation of diverting colostomy, sigmoid was used.
  
  o **Stage 2**
    - Anterior sagittal anorectoplasty.
    - This was done at least six weeks post the diverting colostomy.
  
  o **Post-operative**
    - Given a cover of antibiotics.
    - Can feed immediately post operation.
    - Anal dilatation programme resumed only four weeks post ASARP.
  
  o **Stage 3**
    - Closure of colostomy at least six weeks after anoplasty and anal dilatation programme satisfactory.

Final assessment of outcomes of the surgical management was noted after one year post surgery.

3.5. **Outcome Variables**

- Age at diagnosis
- Presenting symptoms
• Age at first procedure
• Type of surgical management
• Complications early (post-operation) and late (4-week follow-up)
• Outcomes at 3-month follow-up / 6-month and 1-year follow-ups

3.6. Data Collection
Data for analysis of this study were extracted from DGMAH records of the patients and entered into a data collection form. The data collection form was designed for this purpose and contained all the variables listed in the outcome measures.

3.7. Data Analysis
Analysis of data generated from the study involved descriptive statistics to establish the indexes of mean values, standard deviation and proportions expressed as percentages. The incidence of the recto-vestibular fistula among the paediatric female population of DGMAH was calculated for this reviewed period. A profile of complications was determined and expressed as percentages. The relative risk as calculated by Odds Ratio and its 95% Confidence Interval was analysed for complications as well as surgical success in relation to the procedure performed.

3.8. Ethical Aspects For The Study
Because the study was designed as a retrospective study, no consent was required from the parents of the paediatric patients. The study was approved by the Clinical Manager at DGMAH who authorised the usage of the patients’ files without infringing on their confidentiality. The study had also undergone institutional review by the School of Medicine Research and Ethics Committee of Sefako Makgatho Health Science University and approval to conduct the study was given. All information
relating to patients’ medical records as well as identity of the patients was treated with strict anonymity and confidentiality.

3.9. Reliability For The Study

Reliability of any study is defined as reproducibility and consistency of information or data and the degree to which a method gives the same results when used on more than one occasion under the same condition. (13) In this study, reliability was ensured through the following steps:

- Selection of medical records which meet the criteria of the clinical condition to be evaluated was carried out by only the researcher.

- Because of the retrospective nature of the study design, all efforts were made to ensure at least 90% to 95% retrieval of records (retrieval rate was assured by the researcher working closely with the official filing room to retrieve the appropriate records).

- Systemic sampling of all records within the review period.
3.10. Validity Of The Study

Validity of a study refers to the concept of accuracy and it was also the degree to which the measurements reflect the true values of the variables. (14) In the proposed study, validity was ensured through:

- Ensuring that the instrument of data collection (data collection form) was developed to take into account all variables that are directly related to the objectives of the study.
- All the clinical variables that define the medical condition, the surgical management and outcomes of procedures are constantly assessed for accuracy.
- Medical records of patients with insufficient or illegible information were excluded from the study analysis.

3.11. Bias

Bias is any effect at any stage of a research process that tends to produce results that depart systematically from the true values. (15) The researcher avoided any possible bias in the study by:

- Ensuring proper selection of cases for the study in terms of age and gender.
- Statistical analysis of data generated from the study was performed by an independent statistician who would not have been involved in the conduct of the study and the compilation of data.
- The researcher avoided making inferences from the study findings that were not in keeping with the objectives of the study.
3.12. Aim

The aim of the study was to determine the incidence, management and outcomes of vestibular fistula in paediatric female patients presenting at DGMAH.

Objectives of the Study

- Determine the number of paediatric patients seen with vestibular fistula.
- Determine the proportion of patients that are late presenters and establish the predominant clinical symptoms.
- Review the surgical procedures that are being offered to these patients in terms of a choice between a single-stage versus multiple-stage procedure.
- Determine the outcome, early and late complications and the effect on future continence outcome.

3.13. Study Question

What was the frequency of occurrence, the management and the outcome of anorectal malformation (vestibular fistula) among paediatric female patients at DGMAH?
Chapter 4: Results

4.1. Introduction

Fifty-four (54) cases of the ARM (vestibular fistula) occurred during the study period stretching from 1 January 2011 to 30 June 2016 (5.5 years) among a total paediatric population of six hundred and fifty-six (656) treated in the unit at DGMAH. This figure translates to an incidence rate of 8.23% (i.e. 82 cases per 1000 patients seen in the unit at DGMAH). Of these patients, 36 medical files were retrievable for the present audit. Of note was the fact that all the cases seen during the review period presented as low malformation.

4.2. Age distribution of the children seen during the study period

Figure 1 shows the age distribution of the children at the time of presentation in DGMAH.

![Figure 1: Age distribution of the children seen during the study period](image)

Figure 1: Age distribution of the children seen during the study period
The majority of the patients (16; 44.4%) were less than one week old at the time of presentation, followed by children who were between 1 and 10 weeks old (9; 25%). Six of the children (16.7%) were in the age range 11–20 weeks, 1 child was seen at between 21 and 30 weeks of age and a further 4 children were seen at an age older than 30 weeks.

Figure 2 illustrates the ages of the patients at the time when the first procedure was performed. Similar to the ages of the children at presentation, the first procedure was performed for 41.7% (n=15) of the children at ages less than one week. A total of 25% (n=9) had had their first procedure when they were between 1 and 10 weeks old. Another group of the children (n=7; 19.4%) had their first procedure performed at ages ranging from 11 to 20 weeks. One case (2.8%) had the first procedure at an age between 21 and 30 weeks and 4 children (11.1%) were older than 30 weeks at the time of first procedure.

Figure 2: Ages of the children at first procedure to correct the malformation
4.3. Symptoms

Table 1 below shows the variety of symptoms recorded at the time the children were seen at DGMAH. The majority of the children presented with functional fistula (n=23; 63,9%), while constipation (n=4; 11,1%) and abdominal distension plus non-functional fistula (n=2; 5,6%) comprised two other significant symptoms at presentation in hospital. Other symptoms found during the study period were one case each (2,8%) of functional fistula with duodenal atresia; late presentation plus functional fistula; non-functional fistula; non-functional fistula plus bowel obstruction; perineal excoriation; constipation with perineal excoriation and one case of chronic constipation which was refractory to stool softener.

**Table 1: Symptoms recorded for the children at the time of presentation in DGMAH**

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Functional fistula</td>
<td>23</td>
<td>63,9%</td>
</tr>
<tr>
<td>Constipation</td>
<td>4</td>
<td>11,1%</td>
</tr>
<tr>
<td>Abdominal distension + non-functional fistula</td>
<td>2</td>
<td>5,6%</td>
</tr>
<tr>
<td>Functional fistula + duodenal atresia</td>
<td>1</td>
<td>2,8%</td>
</tr>
<tr>
<td>Late presentation + functional fistula</td>
<td>1</td>
<td>2,8%</td>
</tr>
<tr>
<td>Non-functional fistula</td>
<td>1</td>
<td>2,8%</td>
</tr>
<tr>
<td>Non-functional fistula + bowel obstruction</td>
<td>1</td>
<td>2,8%</td>
</tr>
<tr>
<td>Perineal excoriation</td>
<td>1</td>
<td>2,8%</td>
</tr>
<tr>
<td>Constipation + perineal excoriation</td>
<td>1</td>
<td>2,8%</td>
</tr>
</tbody>
</table>
Figure 3 shows the types of procedures performed for the children in this series.

![Pie chart showing types of procedures performed for children]

**Figure 3: Types of procedures performed for the children**

More than two-thirds of the patients in this series (n=25; 69.4%) underwent a 3-stage procedure; a 2-stage procedure was performed for 3 patients (8.3%) while 8 of the children (22.2%) underwent a 1-stage procedure.

### 4.4. Complications

The complications arising from management of the condition in these children at DGMAH were assessed in four phases. Tables 2a and 2b offer a breakdown of the immediate complications.

<table>
<thead>
<tr>
<th>Table 2a: Anastomotic Breakdown</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-Stage procedure (n=8)</td>
</tr>
<tr>
<td>None</td>
</tr>
</tbody>
</table>
Table 2b: Bleeding as a complication

<table>
<thead>
<tr>
<th></th>
<th>1-Stage procedure (n=8)</th>
<th>1-Stage procedure (n=3)</th>
<th>3-Stage procedure (n=25)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
</tbody>
</table>

In the first phase, complications were evaluated immediately after surgery, as well as anastomotic breakdown and bleeding. Immediately after surgery, there was only one case (4%) of anastomotic breakdown in the 3-stage group.

Table 3: Anal Stricture (late complication – 4-week follow-up)

<table>
<thead>
<tr>
<th></th>
<th>1-Stage procedure (n=8)</th>
<th>2-Stage procedure (n=3)</th>
<th>3-Stage procedure (n=25)</th>
</tr>
</thead>
<tbody>
<tr>
<td>4 (50%) had anal stricture</td>
<td>None</td>
<td>18 (72%) without anal stricture</td>
<td></td>
</tr>
<tr>
<td>3 (12%) defaulted</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2 (8%) with anal stricture</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 (4%) had recurrent fistula</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>1 (4%) died – unrelated to malformation</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

At the 4-week follow-up post-surgery, from the 1-stage group, 50% presented with anal stricture; from the 3-stage group, 8% had anal stricture. However, in the 3-stage group, we had numerous other problems such as 12% defaulting follow-up, 4% had...
a recurrent fistula and another 4% died, unrelated to the malformation. The patient had severe gastroenteritis.

The second phase of complication was performed as a long-term complication after three months following surgery (Table 4a & 4b). During this assessment, the 2-stage procedure group had closure of colostomy. In the 3-stage group, 90,4% had colostomy closure and 9,5% defaulted. The anal stricture evaluation revealed that 37,5% had stricture in the 1-stage group and 12,5% defaulted from the same group. In the 3-stage group, 23,5% had anal stricture.

**Table 4a: Three-month follow-up: Anal Stricture**

<table>
<thead>
<tr>
<th></th>
<th>1-Stage procedure (n=8)</th>
<th>2-Stage procedure (n=3)</th>
<th>3-Stage procedure (n=21)</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 (37,5%) had anal stricture</td>
<td>None</td>
<td></td>
<td>5 (23,8%) had anal stricture</td>
</tr>
<tr>
<td>1 defaulted (12,5%)</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

**Table 4b: Three-month follow-up: Closure of colostomy**

<table>
<thead>
<tr>
<th></th>
<th>1-Stage procedure (n=8)</th>
<th>2-Stage procedure (n=3)</th>
<th>3-Stage procedure (n=21)</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>All had colostomy closure</td>
<td></td>
<td>19 (90,4%) had closure of colostomy</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>2 (9,5%) defaulted</td>
</tr>
</tbody>
</table>
Table 5: Six-month follow-up: Correction of malformation

<table>
<thead>
<tr>
<th></th>
<th>1-Stage procedure (n=7)</th>
<th>2-Stage procedure (n=3)</th>
<th>3-Stage procedure (n=20)</th>
</tr>
</thead>
<tbody>
<tr>
<td>6 (85,7%) had correction of malformation</td>
<td>All had correction of malformation</td>
<td>17 (85%) had correction of malformation</td>
<td></td>
</tr>
<tr>
<td>1 (14,2%) had no correction, converted to 3-stage procedure</td>
<td>1 (5%) defaulted previously, came back for closure of colostomy</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>2 (10%) had recurrent fistula</td>
<td></td>
</tr>
</tbody>
</table>

The third phase of the assessment of the management of these children was a 6-month post-surgical assessment to determine the completeness of correcting the malformation (Table 5). In the 1-stage group, 85,7% had achieved complete correction of the malformation and 14,2% did not achieve correction of the malformation and converted to a 3-stage procedure. In the 2-stage group, all patients achieved correction of the malformation.

In the 3-stage group, 85% had achieved correction of the malformation. Five per cent (5%) of the previous defaulters came back for closure colostomy and 10% of the malformation persisted still at this stage of the follow-up.
The fourth phase of the assessment of the management of these children was a 1-year follow-up post-surgery.

From the 1-stage group, 83.3% were ready for toilet training while 16.6% defaulted follow-up. In the 2-stage group, all were ready for toilet training, while 61.9% of patients in the 3-stage group were ready for toilet training. We still had numerous problems at

### Table 6. One-year follow-up

<table>
<thead>
<tr>
<th>1-Stage procedure (n=6)</th>
<th>2-Stage procedure (n=3)</th>
<th>3-Stage procedure (n=21)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5 (83.3%) were ready for toilet training</td>
<td>All ready for toilet training</td>
<td>13 (61.9%) were ready for toilet training</td>
</tr>
<tr>
<td>1 (16.6%) defaulted</td>
<td></td>
<td>1 defaulter (4.8%) reappeared with anal stenosis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>1 (4.8 %) defaulted</td>
</tr>
<tr>
<td></td>
<td>2 (9.5%) had persistent fistula</td>
<td>4 (19.0%) had anal stricture</td>
</tr>
</tbody>
</table>
this follow-up as well. After defaulting for a whole year, 4.8% reappeared with anal stenosis. Another 4.8% defaulted follow-up. In 9.5% of patients the malformation still persisted and 19.0% presented with anal stricture.
5.1 Introduction
From the retrospective study that was done over the 5.5-year period, we had an incidence rate of 8.23%, which translates to 82 cases per 1000 patients seen in our unit. This translates to high incidence.

Dayang et al. (16) reported that ARM has an incidence of 1 in 5000 neonates internationally. Their study commented on the incidence of ARM in girls and reported the figures to be 2.0–2.5 per 10000 live births. There are variations in the incidence between geographical areas in the world. Louw (17) reported an incidence of 5.5 per 10000 live births in Cape Town, South Africa.

5.2 Age of distribution
Our age of distribution for the vestibular fistula ranged between less than 1 week of age up to more than 30 weeks of age. From our group younger than 1 week up to age of 10 weeks, early presenters were seen, which constituted the majority of the patients at 66.7%. Late presenters were those beyond the age of 11 weeks, which constituted 33.1% of the patients. Our reasons for the late presentation were the same found in the literature. According to literature (6;4), delayed presentation of ARM could be due to a number of possible reasons. Some common reported causes in developing countries included inadequate neonatal assessment, low parity, young mothers, misdiagnosis, wrong treatment advice as well as social factors such as lack of money or social support. Sharma et al. (7) also reiterated that 1 in 5 neonates had delayed presentation despite proper neonatal exam.

In our group, the presenting symptoms for the late presenters were as follows:
• Abdominal distension with functional fistula in the majority of our patients;
• Chronic constipation in 5 patients and 1 patient being refractory to stool softeners;
• Perineal excoriation in 2 patients and 1 patient with chronic constipation; and
• Bowel obstruction.

Bokhari et al. (4) reported similar findings in terms of the presenting symptoms in their study in the form of constipation usually associated with introduction of solid foods and inadequate weight gain.

In terms of the other associated anomalies such as renal defects, heart defects and intestinal atresia, only 1 of our patients had a duodenal atresia which was repaired during the diverting colostomy of the 3-stage operations. Most of our patients had an echocardiogram and renal ultrasound during their first presentation, during which no abnormalities were detected. In the literature, associated anomalies (VACTERL) account for 10% of the population (2), with renal anomalies and cardiac defects being the most common.

5.3 Types of procedure performed on the children
Most of our patients underwent the 3-stage procedure; 69.40% and 22.20% had a 2-stage procedure. Only 8.30% had a single-stage procedure. Our practice of 3 stages is attributed to the fear of sepsis, failure of wound healing and subsequent loss of the anal sphincter complex with the risk of faecal incontinence in the future. Those who were offered a single-stage procedure were in the group less than 1 week old. Menon et al. (9) supported this by stating that meconium is relatively
sterile; there was therefore less risk for surgical site infection and the dissection of the distal rectum was less bloody unlike in the infant stage. Patients with non-functional fistula with abdominal distension were all offered a colon diversion. All late presenters were given either a 2-stage procedure or a 3-stage procedure. This was done to avoid faecal contamination at the neo-anus, which would promote surgical site infection. Diverting colostomy was beneficial in reducing the size of the distal sigmoid, and if not done it would lead to poor peristalsis of the distal colon in the future.

Omid and Ketachian (18), in reviewing single-stage versus 3-stage repair in ARM with recto-vestibular fistula, stated that selecting a single-step or 3-step surgical procedure to treat this anomaly has been a subject of debate for many years. A reasoning for choosing the 3-stage repair is that there was lesser risk of surgical site infection because of faecal diversion by colostomy. It might be an easier surgical technique because of the delay in definitive repair and weight gain by the infant. The reasons for choosing one-stage repair are multiple, including:

- Avoidance of multistage operations;
- Saving time and costs;
- Less stress and insults for the children and their parents;
- Less psychosomatic trauma for the children; and
- Avoidance of colostomy related complication.

Another argument in favour of early and immediate use of the neo-anus is the so-called brain-defecation reflex that remains intact following the biological “use it or lose it” principle. Through early usage, the perineal musculature is trained early
and synapses of neuronal network may be formed, which may enhance the chances of normal or near normal function.

In developing countries (3), where colostomy is socially unacceptable, colostomy bags are difficult to come by. Parents’ not being able to manage the colostomy bag is another reason for favouring single-stage repair. Omid and Ketabchian (18) commented that in Egypt they showed that 2-stage repair of low ARM in girls is truly beneficial. They could perform a successful operation and achieve continence in their children. A single-stage procedure has a disadvantage of high-rate surgical site infection, as the faecal contamination was high.

Most authors are nowadays for single-stage procedures, and would only recommend or advise for a diversion in a neonate who cannot defecate via fistula because of a very thin and narrow fistula tract that does not respond to dilatation and bougienage and has obstructive symptoms. Another absolute contraindication for single-stage repair is they have severe life-threatening other congenital malformations, sepsis or necrotising enterocolitis with pneumoperitoneum, extreme prematurity and cloacae (1). It should be borne in mind that colostomy complications are up to 40%. Of these, peristomal excoriation, prolapse stoma, parastomal hernias, leakage, intra-abdominal adhesion or, even worse, bowel obstruction are the most common. (19) Meticulous construction of the colostomy was needed to reduce the significant morbidity associated with colostomies in those patients.
5.4 Immediate complication

In comparing immediate complication post-operatively between the single-stage repair versus the 3-stage repair, only 1 patient (2.8%) had an anastomotic breakdown post-colostomy. We attribute this complication to the stoma bag not being positioned properly, leading to faecal contamination of the wound. There was no one with post-operative bleeding complications. Many studies have commented on complications of stomas. One of the early studies attributed wound dehiscence to faecal contamination, when the post-operative management is insufficient.

5.4.2 Late complication (4-week follow-up)

On the review at 4 weeks, all the patients’ post-ASARP procedure were started on an anal dilatation programme. During the review, we would try and dilate to a hegar 12 in size. From the 1-stage group, 50% presented with anal stricture versus 8% who presented with anal stricture in the 3-stage group. They were dilated to an adequate size in theatre. All parents of the patients were shown and taught how to dilate at home on a daily basis. Our 3-stage group also had numerous problems; 4% presented with recurrent fistula, 12% defaulted follow-up and 1 patient (4%) died, which was unrelated to the malformation. From one of the studies, Menon et al. (4) also stated that, conventionally, dilatation of neo-anus for varying periods is advised after surgery. In the study by Dayang et al. (16), their 3-stage group were subjected to a longer period of daily anal dilatation prior to closure of colostomy. As for the single-stage repair, in most cases anal dilatation was not required.
5.5 Three-month assessment

At the 3-month assessment of our patients, all our 2-stage group patients had colostomy closure. Our 3-stage group had only 90% colostomy closure and 2 patients defaulted follow-up. There were no documented post-operative complications, such as surgical site infection or fistula formation post closure of the colostomy. From the literature, closure of colostomy is not without morbidity. Three of the patients in the study by Dayang et al. (16) with closure colostomy had complications. One patient had anastomotic leak and 2 patients had complete wound dehiscence, which all translated to a longer stay in hospital, and 2 of those patients were managed in the ward via wound care. From their conclusion, single-stage repair will avoid repeated hospitalisation, which could translate to reduction of total cost of treatment and avoidance of major complications associated with colostomy formation and closure.

In the 1-stage group, 37,5% presented with anal stricture and 23,8% was seen in the 3-stage group due to the non-compliance of the parents on the anal dilatation programme. Menon et al. (9) reported some non-compliance to the dilatation programme as the reason for post-operative anal stenosis. Defaulting follow-up could be due to low socio-economic status of the parents.

5.6 Six-month assessment

During the 6-month review of our patients, the 1-stage group had 85,7% correction of malformation; 14,2% had no correction of malformation and converted to the 3-stage procedure. The reason for the conversion was not clearly documented in the patients’ files. The 2-stage group all had correction of the malformation. The 3-stage group had 85% correction of the malformation. One patient who initially
defaulted the 3-month follow-up came back for closure of colostomy and 10% had a recurrent fistula. Our reasoning for the recurrence was due to improper mobilisation of the fistula during the ASARP. Kumar et al. (5) agree with Pena et al. that recurrence of fistula occurs because of inadequate mobilisation and separation of the rectum from the posterior wall of the vagina. A tense anastomosis and suspected rectal devascularisation may aggravate these problems. Thus, a meticulous surgical technique (ASARP) was necessary for better outcome.

1.7 Twelve-month assessment

During the twelve-month assessment post-surgery, the majority of the patients were ready for toilet training or already trained. We looked at these patients using the Kelly scoring method of incontinence, which considers at the following parameters: 1) continence, 2) staining and 3) sphincter. They were all evaluated as doing well. During our 12-month assessment, 19,0% from the 3-stage group had anal stricture which had dilated in theatre. Other complications occurring in the 3-stage group, with 4,8% presenting with anal stenosis after defaulting a whole year. A further 4,8% defaulted follow-up and 9,5% had recurrent vestibular fistula. The continence results in most studies were evaluated by using the clinical scoring method devised by Kelly. This method assesses continence and strength of puborectalis action on digital examination as well. The above results of our 12-month assessment post-surgery have shown us that our decision to do the 3-stage procedure has not really worked in our favour. We had a lot of anal stricture that was suspected to be psychosocial hurt that we imposed on our patients to be dilated by their parents at home. Most were defaulting follow-up due to non-compliance of dilating at home. Recurrence of fistula is due to the surgical
technique that was not adequately done in the number of patients that presented back to the institution with fistula. This translates to other operations, costs for the family and emotional burden on the family as well as a lengthy hospital stay.

1.8 Conclusion

Our study was a retrospective study, where the retrieval of the files delivered only 72% of those cases that were represented during the specific period. Medical record-keeping was also insufficient in some areas; thus, selection bias in some areas of the results could not be ruled out.

In our practice the incidence of vestibular fistula was high compared to literature. The majority of patients were early presenters. The surgeons favoured 3-stages even with more early presenters. This could be related to surgeon’s bias and preferences. 2-Stages group had the best outcome. The practice of 1-Stage was associated with more severe complications to the child.

The outcomes favoured multiple staged protocols even at a higher cost to the health system. The patient numbers in 1-Stage group or 2-Stages group were limited to validate their advantage.

The 1Stage option is now favoured and recommended practice in the literature. As most studies written over the last 20-year period have shown, a single-stage repair is preferred and is a reliable method for treating ARM with recto-vestibular fistula. It has an obvious advantage for both the patient and the parents.

In our setting of limited resources, it might be cost effective in the long run. However, our findings seem to show that such practice could come with high cost to the girl child. It would seem that a move toward a 2-Stages has the best of two
extremes and more cost effective to the health system and the patient. With experience and improving perinatal care, our own morbidity, operation time and hospital cost will improve, regardless of options chosen.

We also saw high rate of defaulting parents, probably due to monetary issues pertaining to coming to hospital and the psychosocial impact of the malformation on the dynamics of the families’ lives. Psychosocially, the impacts on the patient and the parent, makes it hard to dilate their own children. The 2-stage procedure would seem also to decrease our number of anal stricture that we see and improve the compliance of the parents who are dilating.

Lastly, the continence problem will dictate that we should aim to repair vestibular fistula as early as possible, due the theory that neuronal framework for normal bladder and bowel function exist at birth and that there is a learning or training period in which long-lasting, activity-driven, neuronal changes take place during neuronal circuitry development in the early life (3). By delaying the repair of ARM, critical time may be lost in which neuronal networks and synapses would have formed, resulting in normal or near normal rectal function (6).

In our setting the 2-Stage option is an obvious choice due to its best outcome. The practice of 1-Stage might be detrimental due to its associated more severe complications to the girl child.
References


### DATA COLLECTION FORM

**Title: Incidence, management and outcomes of recto-vestibular fistula and anorectal malformations in paediatric female patients presenting at BGMAH**

<table>
<thead>
<tr>
<th>Study No.</th>
<th>Age</th>
</tr>
</thead>
</table>

#### Type of Procedure
- High
- Low

<table>
<thead>
<tr>
<th>Age of 1st Presentation</th>
<th>Age of 1st Procedure</th>
</tr>
</thead>
</table>

#### Presentation Symptoms

#### Type of Procedure
- 1-stage
- 2-stage
- 3-stage

#### Complications

<table>
<thead>
<tr>
<th>Early (Immediate post-surgery)</th>
<th>Late (4-week follow-up)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bleeding</td>
<td>Anastomotic breakdown</td>
</tr>
<tr>
<td>Anastomotic breakdown</td>
<td>Anal Stricture</td>
</tr>
<tr>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>No</td>
<td>No</td>
</tr>
</tbody>
</table>

#### Outcome at 3 Months Follow-Up

<table>
<thead>
<tr>
<th>Closure of Colostomy</th>
<th>Anal Stricture</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>No</td>
<td>No</td>
</tr>
</tbody>
</table>

#### At 6 Months Follow-Up

<table>
<thead>
<tr>
<th>Complete correction of malformation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yes</td>
</tr>
<tr>
<td>No</td>
</tr>
</tbody>
</table>

#### At 1 Year Follow-Up

**Any Comment**
Appendices B: Ethics Approval Certificate

Sefako Makgatho Health Sciences University
Research & Postgraduate Studies Directorate
Sefako Makgatho University Research Ethics Committee (SMUREC)

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02 June 2016

D: ME Mokola
Department of Paediatric Surgery
P.O Box 231
Medunsa, 0204

MEETINGS:

SMUREC Ethics Reference Number: SMUREC/M/1132016: PG

The New Application received on 05 May 2016 was reviewed by members of Sefako Makgatho University Research Ethics Committee 02 June 2016 and was approved on 02 June 2016.

Title: Incidence and management outcomes of arteriovenous malformations in Paediatric - Female patients presenting at Dr George Mukhari Academic Hospital

Researcher: Dr ME Mokola
Supervisor: Prof L Mercier
Co-supervisor: Dr N Shields
Hospital Superintendent: Dr A Steyn (DGMAH)
Department: Paediatric Surgery
School: Medicine
Degree: MPH Paediatric Surgery

Please note the following information about your approved research protocol:

Protocol Approval Period: 02 June 2016 – 02 June 2017

Please remember to use your protocol number (SMUREC/M/1132016: PG) on any documents or correspondence with the REC concerning your research protocol.

Please note that the REC has the prerogative and authority to seek further questions, seek additional information, require further modification, or terminate the conduct of your research and the consent process.

After Ethical Review: Please note a synopsis of the progress report is submitted to the Research Office and should be submitted to the Committee before the year has expired. The Committee will then consider the continuation of the project for a further year (if necessary). An annual number of projects may be assessed randomly for an external audit. Translation of the consent document in the language applicable to the study participants should be submitted.

International Organisation (ROC00003691), Institutional Review Board (RBER001358) Expiry date: 06 December 2015, Federal HIE Assurance (FW400002639G) Expiry date: 31 August 2017, SMUREC No.: RER 2016/000082

Sincerely

PROF DA COLUMBAO
CHAIRPERSON SMURES