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Patterns and Treatment Outcomes of Anorectal Malformations in Mbarara Regional Referral Hospital, Uganda

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ABSTRACT

Background/Purpose: Anorectal malformation (ARM) is a common condition. Due to scarcity of pediatric surgery services in Uganda, however, relatively little is known about this condition.

Methods: This was a retrospective review of medical records of all patients admitted to Mbarara Regional Referral Hospital (MRRH) from January 2014 to May 2016. MRRH serves 3-8 million people in southwest Uganda. We also enrolled patients prospectively from June 2016 to December 2016.

Results: 78 patients were enrolled in the study. 63.38% had delayed diagnosis (presenting > 48 hours after birth), and most of these were self-referrals from home. The most common malformation was a vestibular fistula. Associated congenital anomalies were seen in 20% of patients, and this was associated with increased mortality. These anomalies included limb, eye, ear and genitourinary anomalies as well as ventricular septal defects, patent ductus arteriosus, spina bifida and trachea-esophageal fistula. Posterior sagittal anorectoplasty (PSARP) was the definitive surgery used. It was performed in 58.97% of the patients. Median age of patients who underwent PSARP was 11 months. 73.91% of PSARP patients had their colostomies closed and 62.5% of these who were ≥ three years old had good continence outcomes. Overall mortality rate was 8.97%.

Conclusion: The majority of patients are diagnosed late. Vestibular fistula is the overall most common type of ARM. In males, however, rectourethral fistula is the most common type. Definitive surgery at MRRH is performed late. Continence outcomes are good and comparable to other countries with more resources.

Key Words: Anorectal malformations; Early diagnosis; Continence; Uganda

Level of Evidence: III

Abbreviations: Mbarara Regional Referral Hospital (MRRH), Anorectal Malformation (ARM), Posterior Sagittal Anorectoplasty (PASRP)

1. INTRODUCTION[1]

Anorectal malformations (ARM) include a wide spectrum of defects in the development of the lowest portion of the intestinal and genital tract [1]. There is poor development of the anus and the rectum with associated fistulae to the genitourinary system or perineum [1, 2]. ARMs are common major congenital anomalies in pediatric surgery patients throughout Africa [3]. Incidence ranges from 1:2000-1:5000 live births worldwide [4, 5]. In 2001, prevalence of ARM among the total congenital anomalies admitted to Mulago Hospital (the national referral hospital in the Ugandan capital, Kampala) was 24.7% [6].

[1] ARM: anorectal malformation. MRRH: Mbarara Reginal Referal Hospital. PSARP: posterior sagittal anorectoplasty. RDS: respiratory distress syndrome

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ARMs can occur as isolated congenital birth defects, as part of a syndrome or associated with other congenital anomalies [7]. Associated congenital anomalies are found in 20-80% of ARM worldwide [4, 7]. Associated congenital anomalies, together with delay in seeking treatment, contribute significantly to morbidity and mortality [8, 9]. Management has improved in the past 20 years but some patients suffer from fecal and urinary incontinence, and sexual inadequacy after surgery [10]. Management is delayed in Uganda due to scarcity of pediatric surgery services [9, 11-13].

The pattern and treatment outcome of ARM in western Uganda is not known. The absence of data makes it hard for health service providers to set up policies and protocols for early diagnosis and management. In this study, we document the pattern and treatment outcomes of ARM at Mbarara Regional Referral Hospital (MRRH) with the aim of providing baseline data on nature and extent of the problem in Mbarara.

2. MATERIALS AND METHODS

The study was carried out at Mbarara Regional Referral Hospital (MRRH) between June 2016 and March 2017. MRRH is a 425-bed, regional referral hospital in southwest Uganda that serves a catchment area of over 3 million people, and it is a specialty referral center for a region of 8 million people [14]. It is a government funded facility through the Ministry of Health offering free health care services covering 10 districts. It is also associated with a university, medical school and nursing school, with residents training in various specialties. Adequate diagnostics are available in the hospital and in private laboratories and imaging centers near the hospital. The largest hospital in Uganda is the national hospital, Mulago, in the capital of Kampala. Below the level of the national hospitals there are EIGHT regional hospitals that serve as regional referral centers distributed around the country. The referral hospital is the first place that has specialty surgery. MRRH is one of the larger referral hospitals

The pediatric surgery unit has a pediatric surgeon and trains pediatric surgery fellows. The inpatient section has 17 beds with a total of 6 nurses who work in three shifts every day. The morning shift has 2 nurses and 1 nurse for the evening and night shifts. The hospital has an adult ICU (Intensive Care Unit) with one bed allocated for pediatric patients below 5 years. Oxygen is

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available in the hospital all the time. The medical supplies are provided by the ministry of health and donors from other countries, and are available almost all the time.

This is a hospital-based descriptive study with retrospective and prospective components. We included all patients admitted to MRRH with anorectal malformations between January 1, 2014 and December 31, 2016. We reviewed inpatient and outpatient medical records and also recruited new patients as they were admitted during the study period. Each patient was followed up for a minimum of one month after each operation at MRRH.

Variables included age at diagnosis, gender, age of the mother, delivery provider, type of referral, type of anorectal malformation, type of temporary colostomy, type of definitive surgery, age at the time of definitive surgery, continence to stool after definitive surgery, home district of the patients and associated congenital anomalies. Associated congenital anomalies were screened through physical examination, abdominal ultrasonography, X-ray of the vertebrae and limbs for the patients with obvious deformities on physical exam, and echocardiogram for the patients with a murmur on auscultation of the precardium. Weeks of gestation at birth is always documented on the medical forms. Retrospective review demonstrated similar work up. Mortality and causes of mortality were also assessed.

Data collected was entered into Epi Info 7 and later exported to STATA 12 for cleaning and analysis. Descriptive statistics were used in the analysis, including frequency and proportions, Median and range. A Chi Square Statistic was used to measure associations. The home districts of the patients were mapped in Google maps to describe their geographical distribution.

Ethical approval for this study was obtained from the Institutional Review Committee at Mbarara University of Science and Technology, Faculty Research Ethical Committee, and Institutional Review Board at MRRH.

3. RESULTS

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3.1 General characteristics of the patients

During the three-year study period, 78 patients with anorectal malformations were admitted at MRRH and all were recruited for this study. Table 1 shows their general characteristics and their association with mortality.

Male to female ratio was 1:1.2. Seven patients (8.97%) were unavailable for follow up. We considered all patients who were diagnosed within 48 hours after birth to have early diagnosis, the rest were considered delayed. Twenty-seven patients (36%) were diagnosed in their first two days of life. Median age of diagnosis was 4 days, with a range of 1 day to 12.67 years. Data on obstetric provider was available for 71 patients. A formally trained obstetric provider (person with a medical background) delivered 58 patients (81.69%). The remainder were delivered by personnel without any formal medical training. Of these 71 patients, 41 (57.75%) were referred to MRRH from a health facility, and the remainder were self-referred from home. On univariate analysis, there was no association between age at diagnosis, gender of patient, age of mother, delivery provider, referral location and mortality.

3.2 Type of anorectal malformations and their associated congenital anomalies

The type of anorectal malformation was classified according to the Krickenbeck classification system. As shown in Figure 1, the most common anorectal malformation in females was vestibular fistula, accounting for 32.05% (25 patients). The least common was cloaca, accounting for 1.28% (1 patient). The most common anorectal malformation in male patients was rectourethral fistula, accounting for 19.23% (15 patients) of the patients. The bulbar urethra was more involved (12 patients) than prostatic urethra (3 patients). On univariate analysis, there was no association between type of anorectal malformation and mortality.

As shown in Table 1, data on associated congenital anomalies was only available from 65 patients. Thirteen patients (20%) had associated congenital anomalies. There was a relationship between associated congenital anomalies and mortality (p=0.005). There was no association between sex and associated congenital anomalies. Of the patients with associated congenital anomalies, 31% had more than one anomaly (Figure 5). The most common congenital anomalies were genitourinary and included: hypospadias (seen in three patients); bifid scrotum (two patients); ectopic kidney (two patients); cystic kidney disease (one patient); and cryptochidism

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(one patient). Other congenital anomalies included: ventricular septal defect (three patients); patent ductus arteriosus (one patient); tracheo-esophageal fistula (one patient); radial aplasia (one patient); ectrodactyly (one patient); unilateral anophthalmia (one patient); spina bifida occulta (two patients); and periauricular remnant (one patient).

3.3 Time of diagnosis

Table 2 shows how the place of referral is related to the time of diagnosis and obstetric provider. Patients who were diagnosed after 2 days (63.38% of the total) had delayed diagnosis. There was an association between delayed diagnosis and form of referral (p= 0.003). Of the patients who presented late, 55.56% were self-referrals from home. This means that most patients who were diagnosed early were referred from a health facility, and most patients who were diagnosed late were self-referrals from home. There was also an association between obstetric provider and form of referral (p=0.001). The majority of patients (84.62%) who were delivered by an obstetric provider who had no formal training in obstetric care were self-referrals from home. The majority of the patients who were delivered by a formally trained obstetric provider were referred from a health facility. In brief, this means that most of the patients who were self-referral from home were delivered by a person with no medical background.

The home districts for the patients were mapped in Google maps. As shown in Figure 6, the patients were evenly distributed throughout southwestern Uganda, a few from central and southeastern Uganda, and even some from Rwanda and Tanzania.

3.4 Treatment of ARM at MRRH

As shown in Table 1, data on type of colostomy created for temporary fecal diversion was available for 68 patients. Forty-two of the patients had their colostomies inserted at MRRH. Sixteen of them were inserted by the attending pediatric surgeon and the rest by surgical residents. The majority of patients, 25 (36.76%), had a double barrel colostomy. 100% of the patients received a colostomy at the time of diagnosis. Overall, 46 patients (58.97%) had posterior sagittal anorectoplasty (PSARP) done as a definitive management, and the median age at definitive surgery was 11 months (range 1-144 months). The definitive treatment for the remainder of the patients was a colostomy. All PSARPs were performed by a pediatric surgeon.

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Thirty-five of the PSARPs were performed by the attending pediatric surgeon, and the other 11 were performed by three visiting pediatric surgeons. Of the 46 patients who had PSARP, 34 (73.91%) had their colostomies closed at MRRH after assessing patency of the neo-anus.

3.5 Treatment outcome of ARM at MRRH

Fecal continence was assessed using the Kelly's score [2]. This was performed for patients who had definitive surgery (PSARP), whose colostomies were reversed, and who were three years and older. No tool is available to test the fecal continence among children less than three years of age. As shown in Table 3, overall, 13 patients (17%) met the above criteria for assessment of fecal continence. The majority of the patients had good results (as defined by the Kelly score) in terms of continence. The type of ARM in patients with good results included: vestibular fistula (4 patients); rectovaginal fistula (2 patients, figure 2); perineal fistula (1 patient, figure 3); and recto-urethral fistula (1 patient). The type of ARM in patients with fair results included: vestibular fistula; perineal fistula; and no fistula (figure 4). Patients who were lost to follow up had rectovaginal fistula and recto-urethral fistula.

A total of 7 patients (8.97%) died. Mortality was significantly increased among patients with associated congenital anomalies. Two of the patients died of heart failure secondary to congenital heart disease; and others from severe prematurity, severe protein energy malnutrition, respiratory distress syndrome (RDS), and intestinal obstruction with severe dehydration and electrolyte imbalance. One patient died of cardiac arrest on operating table.

Apart from mortality, other complications after surgery were seen in 12 patients (12.82%). These included: dehiscence of PSARP wound requiring wound revision in theatre (five patients); entero-cutaneous fistula following colostomy reversal (one patient); malposition of the neo-anus intra-operatively (one patient); stoma sepsis (two patients); colostomy prolapse (one patient); and incisional hernia (two patients). There was no significant relationship between time of diagnosis and mortality or other complications.

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4. DISCUSSION

4.1

The purpose of this study was to describe the pattern and treatment outcome of patients admitted with ARM at MRRH over a 3-year period from 2014 to 2016. Overall, 78 patients were admitted over the three years. We found a slight female predominance, which was not statistically significant in our study, with a male: female ratio of 1:1.2. This is similar to what was found in another study from Uganda [6]. Different studies from different countries, however, have shown a variation, with some showing male and others female predominance [8, 9, 11, 15-19].

4.2 Type of anorectal malformations

The most common ARM among females was vestibular fistula and the most common ARM in males was recto-urethral fistula. These both support earlier studies in Africa [3, 9]. Anorectal malformations are frequently associated with other congenital anomalies. Our study found a 20% prevalence of associated congenital anomalies among patients admitted with ARM at MRRH. Comparing with studies from other countries, associated congenital anomalies among patients with ARM was seen in 71% in the USA, 89% in Netherlands, 28% in Pakistan, 33% in India, 68.4% in South Africa, 27.9% in Nigeria, and 16.9% in Sudan [8, 18-23] . The most common associated anomalies in our study were with the genito-urinary system. The presence of these associated congenital anomalies was associated with increased risk of mortality (p= 0.005). For this reason, we think that the incidence of associated congenital anomalies among patients with ARM admitted at MRRH may be higher than what we found in our study. Due to our high rate of delayed presentations, the rate of ARM might also be higher if these patients never make it to hospital.

4.3 Time of diagnosis

Delayed diagnosis of anorectal malformations is common and can lead to severe complications, including intestinal obstruction and perforations, sepsis, and death [24, 25]. Delayed diagnosis (diagnosis after 48 hours of birth) occurred in 63.38% of all the patients seen in our study. A study in Ireland found 21.6% delay in diagnosis after 48 hours [16]. Other studies found delay in diagnosis to be 42% in United Kingdom, 57.9% in South Africa, 86.1% in Nigeria and 47.4% in

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Sudan. In these studies, delay in diagnosis was after 24 hours of birth [18, 19, 25, 26]. This is similar to what is reported in many African countries [9, 18, 26]. This is comparably higher than what is observed in the developed countries [24, 25]. The delay in diagnosis in our study is attributed to the significant number of patients who had delayed diagnosis because they were self-referrals from home. This means that they were either delivered at home by an obstetric provider with no formal medical training or delivered by a formally trained obstetric provider at a health facility, but the diagnosis of ARM was not made at birth. The parents discovered the anomalies late and delayed seeking health care. Also, the delay in diagnosis was caused by the long distances that patients had to travel to seek health care at MRRH, as shown in figure 7. Most of the patients are poor, making it difficult to afford transport from their homes to Mbarara and to sustain themselves during the days they are admitted in hospital.

4.4 Treatment

Babies in Uganda often receive very little prenatal care and the birth attendants often have minimal formal training. Babies with ARM's, therefore present obstructed, dilated, dehydrated and malnourished. There are only five pediatric surgeons in a country of 40 million people. The Ugandan babies that survive to see a surgeon universally require diversion as the first treatment. For many, this will also be their only treatment.

We evaluated the surgical procedures performed on the patients in our study. All patients were treated with a temporary colostomy to divert stool before the definitive surgery. A double barrel colostomy was the most commonly used colostomy at MRRH. For all the patients who underwent a definitive surgery, a PSARP was performed. The exception was the child with cloaca. In this case total urogenital mobilization was performed, in addition to the PSARP. The mean age at definitive surgery was 11 months, with a range of 1 month to 144 months (12 years). This range of up to 12 years is large and implies that some children live with ARMs for a long time before definitive reconstruction. This delay has been shown to affect the child's quality of life and social development [15, 17]. If a child has an anorectal malformation and needs a colostomy, it impacts psychosocial and financial implications on the parents as well. A study performed in India found an increased psychosocial burden and poor quality of life among parents of children with ARM [17]. In our study, we found that many children with ARM, especially from rural areas, live with this deformity and a colostomy for up to 12 years because

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of limited access to a pediatric surgeon. There are only four pediatric surgeons in Uganda, and three of those are based in the capital city of Kampala [12, 13].

4.5 Treatment outcome

Overall, 12.82% of the patients experienced post-operative complications and 8.97% died. Definitive management is aimed at reconstruction of the anorectal region with normal continence of stool. This however, depends on the type of ARM. The lower the ARM, the better the expected results in terms of continence [10]. In our study, the majority of the patients above three years (62.5%) had good results in terms of fecal continence; and no patient had poor results. This is comparable to the study in Finland where 68% of females and all males attained good results, and in USA where 75% of the patients had good results [27, 28]. This means that anorectal malformations can be treated in MRRH with good outcomes with respect to continence.

Mortality rate in our study was 8.97% and was significantly associated with presence of associated congenital anomalies. This mortality rate is comparable to the 7% that was found in South Africa, 16% in India, 15% in Pakistan, and 4% in Leicester, United Kingdom [8, 29-31].

5. CONCLUSION

5.1

The pattern of anorectal malformations at MRRH found in our study shows a slight but not statistically significant female predominance, and the majority of patients were diagnosed late. Vestibular fistula was the overall most common type of ARM. In males, however, rectourethral fistula was the most common type. Mortality was significantly increased by the presence of associated congenital anomalies. Mortality rate was also comparable to other countries in Africa and South Asia. Definitive surgery was performed late, but outcome in terms of continence to stool was good and comparable to studies in the USA and Finland.

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Figure 1: Breakdown of anorectal malformations using Krickenbeck classification at Mbarara Regional Referral Hospital (MRRH), stratified by sex.
Figure 2: Rectovestibular fistula in a female patient with ARM at MRRH
Figure 3: Rectoperineal fistula in a female patient with ARM at MRRH
Figure 4: Rectal atresia without fistula in a male patient with ARM at MRRH
Figure 5: Breakdown of anomalies associated with anorectal malformations in pediatric patients at Mbarara Regional Referral Hospital (MRRH)
Figure 6: Geographical distribution of patients with anorectal malformation at Mbarara Regional Referral Hospital (MRRH)

Table 1: Characteristics of patients with anorectal malformations at Mbarara Regional Referral Hospital (MRRH) and their association with mortality
Table 2: Form of referral associated with time of diagnosis and delivery provider.
Table 3: Assessment of patient's fecal continence based on Kelly's score after surgery for anorectal malformations at MRRH.

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Table 1

Characteristics	All patients (% of total)	Alive (%)	Died (%)	Chi^2, P-value
Age at diagnosis in days				
<3	27 (36)	22 (81.48)	4 (14.81)	3.9645, 0.681
3-10	20 (26.67)	16 (80.00)	2 (10)	
11-60	18 (24.00)	16 (88.89)	1 (5.56)	
>60	10 (13.33)	10 (100)	0 (0.00)	
Median (Range)	4 days (1 day-12.67 years)	4 days (1 day-12.67 years)	2 days (1 days-21 days)	
sex				
Female	42 (53.85)	37 (88.10)	2 (4.76)	2.5446, 0.28
Male	36 (46.15)	27 (75.00)	5 (13.89)	
Maternal age in years				
<21	11 (16.92)	11 (100)	0 (0.00)	2.9355, 0.402
21-25	31 (47.69)	27 (87.10)	4 (12.9)	
26-30	13 (20.00)	12 (92.31)	1 (7.69)	
> 30	10 (15.38)	10 (100)	0 (0.00)	
Median (Range)	24 (16-40)	24.5 (16-40)	24 (23-30)	
Obstetric provider				
Formally trained	58 (81.69)	53 (91.38)	5 (8.62)	0.5467, 0.46
No formal medical training	13 (18.31)	11 (84.62)	2 (18.38)	
Form of referral				
Self referral	30 (42.25)	29 (96.67)	6 (14.63)	2.4895, 0.115
Referred from a health facility	41 (57.75)	35 (85.37)	1 (3.33)	
Associated with other congenital anomalies				
Yes	13 (20.00)	10 (76.92)	3 (23.08)	0.0584, 0.005
No	52 (80.00)	51 (98.08)	1 (1.92)	
Temporizing colostomy type				
Loop	22 (32.35)	24 (96.00)	0 (0.00)	4.9949, 0.288
Divided	21 (30.88)	19 (86.36)	3 (13.64)	
Double barrel	25 (36.76)	19 (90.48)	2 (9.52)	
Definitive management done (PSARP)				
Yes	46 (58.97)	44 (97.78)	1 (2.22)	
No	32 (41.03)	20 (76.92)	6 (23.08)	
Median age at definitive management in months (Range)	11 (1-144)	11 (1-144)	N/A	

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Table 2

Characteristics	All patients (% of total)	Referral from health facility	Self-referrals	Chi^2, P-value
Age group in days				
0-2	27 (36.62)	21 (80.77)	5 (19.23)	8.9114, 0.003
>2	45 (63.38)	20 (44.44)	25 (55.56)	
Obstetric provider				
With formal medical training	13 (18.31)	2 (15.38)	11 (84.62)	11.7040, 0.001
No formal medical training	58 (81.69)	39 (67.24)	19 (32.76)	

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Table 3

Evaluation	Total number (%)	Mean age (years)	Type of ARM
Good	8 (62.5%)	6.22	Vestibular fistula (4), perineal fistula (2), rectovaginal fistula, Rectourethral fistula.
Fair	3 (23.1%)	6.37	Rectourethral fistula, vestibular fistula, and ARM with no fistula
Poor	0 (0%)	N/A	N/A
Lost to follow-up	2 (15.4%)	4.02	Vestibular fistula and perineal fistula

ACCEPTED MANUSCRIPT

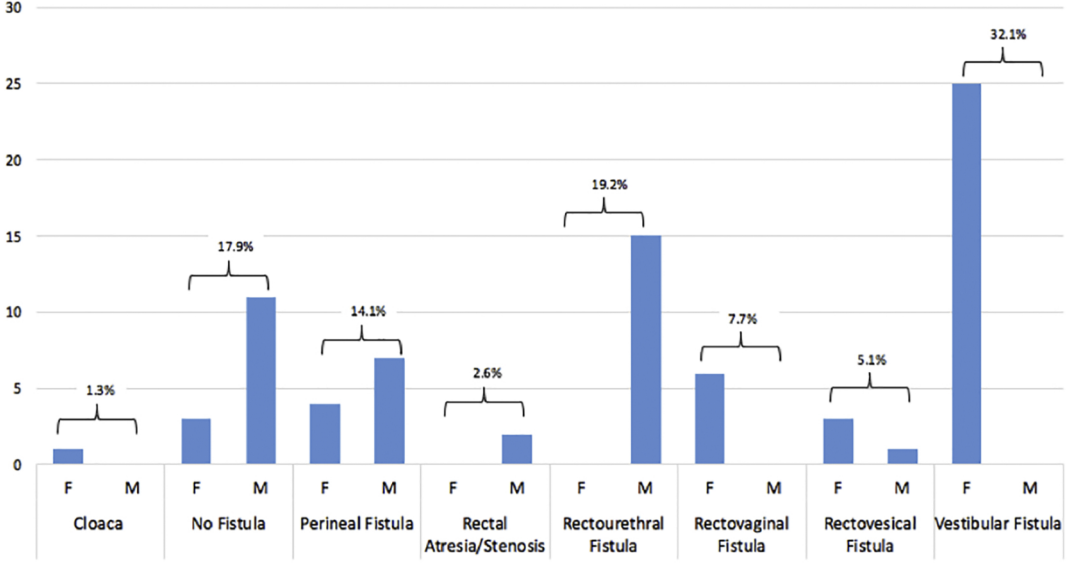

30
25
20
15
10
5
0
1.3%
17.9%
14.1%
2.6%
19.2%
7.7%
5.1%
32.1%
F
M
Cloaca
No Fistula
Perineal Fistula
Rectal Atresia/Stenosis
Rectourethral Fistula
Rectovaginal Fistula
Rectovesical Fistula
Vestibular Fistula

Figure 1



Figure 2



Figure 3



Figure 4

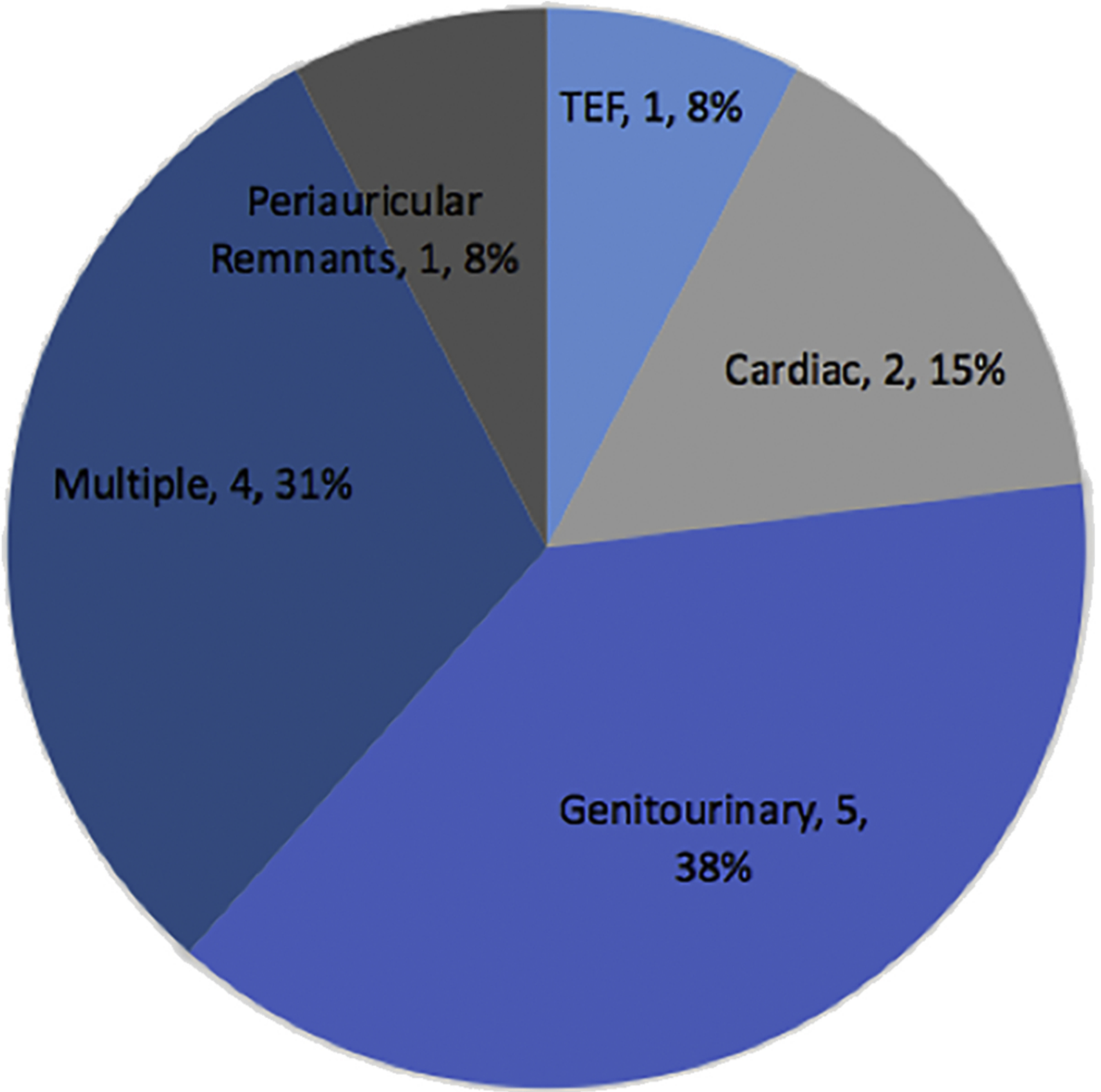

TEF, 1, 8%
Cardiac, 2, 15%
Genitourinary, 5, 38%
Multiple, 4, 31%
Periauricular Remnants, 1, 8%

Figure 5

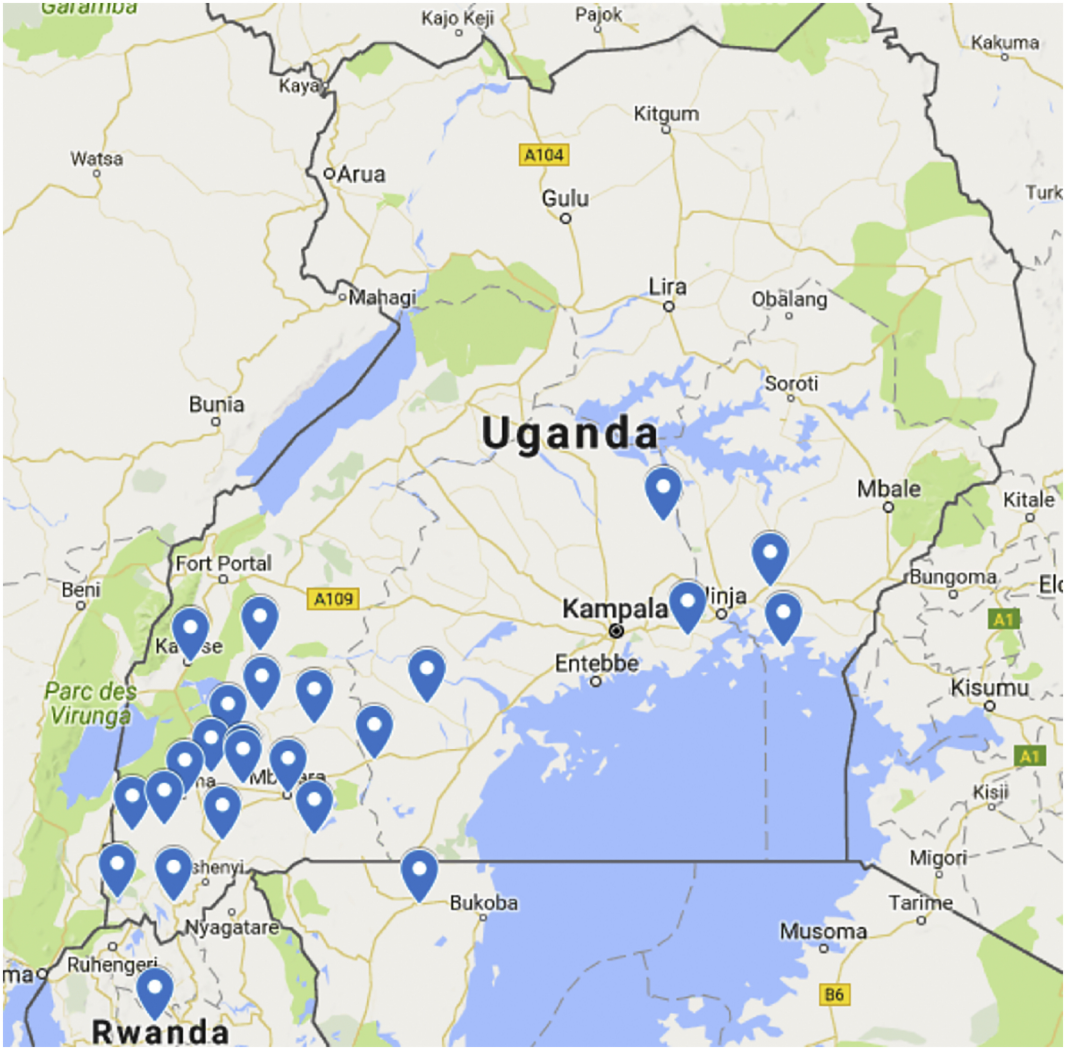

Kajo Keji
Pajok
Kakuma
Kaya
Kitgum
Watsa
Arua
A104
Gulu
Lira
Mahagi
Obalang
Soroti
Bunia
Uganda
Mbale
Kitale
Fort Portal
Beni
A109
Bungoma
Kampala
Entebbe
Parc des Virunga
Kisumu
A1
Kisii
Migori
Tarime
Bukoba
Musoma
Nyagatare
Ruhengeri
B6
Rwanda

Figure 6