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Research Article

Hospital Course and outcome for patients with Anorectal malformations (Bucket-handle deformity), a 3-year experience from Khartoum north teaching hospital (2016 -2019)

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Abstract

Introduction: Anorectal malformations are some of the most common structural congenital malformations treated by pediatric surgeons globally. A bucket-handle deformity is a subtype of it characterized by a subepithelial midline raphe fistula. The etiology has not been fully understood but it is more likely to be multifactorial; genetic and environmental factors implicated. The outcome of surgery depends on the clinical presentation, early diagnosis, other associated malformations, surgeon's expertise and a post-operative follow up care.

Objectives: To analyze and evaluate pediatric patients with Bucket-handle deformity in terms of their hospital course (insight into the pre, peri and post-operative course).

Methodology: This study was done at Khartoum North Teaching Hospital in Khartoum; Sudan. The targeted population were all pediatric patients with this deformity in our pediatric surgery department. Data was collected using a data collection sheet filled by the in-charge surgeon in the period from (March 2016 - March 2019).

Results: In 72 pediatric patients with Bucket-handle deformity included, 39 (54.2%) were males and 33 (45.8%) were females. The mean age for patients was 2.28 years and 2.3 years at the time of surgery. When it comes to their gestational age at time of delivery, almost of them (82%) were born at term and about (16.7%) were preterm and only a small fraction (1.4%) were post-term. Ten patients (13.9%) have other

anomalies that mainly cardiac and only 3 (4.2%) have an associated family history of congenital anomalies. Only 9.7% of patients were septic on admission and they had a slightly longer pre-operative and post-operatively hospital course when compared to non-septic patients. The average number of days patients stayed in hospital pre-operatively was 6.56 days and 10.72 days post-operatively. Overall, there was no post-operative complications of urinary or fecal incontinence and there was no constipation in the 6 months period following the surgery.

Conclusion: This study showed that all pediatric patients with Bucket-handle deformity in our pediatric surgery department had good surgical outcome with no post-operative complications despite having a relatively long pre-operative and post-operative hospital course.

Recommendations: Although results were excellent regarding the surgery, further studies should be carried out to expand the literature regarding evaluation of pediatric patients with Bucket-handle deformity in terms of their hospital course. More efforts need to be put to optimize pediatric patients for surgery even before referring them to a specialized pediatric surgery unit for the goal of minimizing the duration of their admission course.

Keywords: Anorectal malformations; Bucket-handle deformity; Surgery; Low resource settings and surgical complications

1. Introduction

Anorectal malformations (ARMs) represent a variety of diagnoses often known as imperforate anus. Patients with these diagnoses do not have a normal anal opening, but instead, they have a fistulous tract that opens onto the perineum anterior to the anal muscle complex or into adjacent anatomical structures. The fistula can open into the urinary system in males and in the gynecological structures in females. The distance of the opening from where the proper location of the anal opening usually determines the severity of the defect [1]. In Africa, congenital malformations account for one third to two fifths of the operative workload of pediatric surgeons [2,3]. ARM is the commonest major structural congenital malformation presenting to general pediatric surgeons in Africa [3]. The prevalence of ARM is 4/10000 with a gender distribution of around 1:1.5 females: males [4-6] and in Africa there is a slight to moderate male preponderance in cases of ARM ranging from 55 to 71% according to majority of reports [7-13] but a few had shown the reverse [14-17] and a true birth incidence of ARM is difficult to obtain because there are no formal birth registries in most parts of Africa and most reports in the literature are hospital based. The best available population based estimates are from South Africa where the incidence of ARM has been reported to range from 1.79/10,000 live births in the Western Cape Region to 3.26/10,000 live births on the West Coast [7]. These figures are about the same as the incidence of 1 in 5,000 live births reported elsewhere [18]. The presence of associated malformations in other systems is seen in 9-44% of patients in various series across Africa [8,11,14,16-19,20]. Those figures are lower than the expected and more widely documented proportion of 58-78% (21-24). The incidence of reported anomalies is variable, but most groups agree the genitourinary anomalies (40-50%) are most common followed by cardiovascular (30-35%), spinal cord tethering (25-30%), gastrointestinal anomalies 5-10%, and VACTERL (4-9%) anomalies [25]. The treatment of children with ARM is a major aspect of the work of pediatric surgeons as colostomy

for ARM and the definitive anorectoplasty or anoplasty are the commonest colorectal procedures they perform [26]. Early management is recommended in the treatment of children with ARM in order to prevent sepsis and other morbidities related to intestinal obstruction [18]. Delay in presentation of patients with ARM leads to progression of neonatal intestinal obstruction, sepsis, aspiration pneumonia, intestinal perforation, and sometimes death [12,16,27,28]. The early management is crucial with 2 main questions should be answered within the first 48 hours in life. The first question, is there any associated anomalies? And second, should the infant undergo a primary procedure and no protective colostomy or a protective colostomy and a definitive repair at a later date? Presentation outside the neonatal period is widespread as 19-85% of patients with ARM in Africa present outside the first 4 weeks of life [29,8,11,13,14,30]. Delay in diagnosis has been correlated with poorer outcome and a higher mortality [8]. Delayed diagnosis beyond 24-48 hours of birth is unusual in developed countries and furthermore, extreme delays beyond childhood are uncommon except in low resource settings such as in many developing countries. Some perineal signs that may be found in patients with ARM especially in low malformations include the presence of meconium at the perineum, a "bucket-handle" malformation (a prominent skin tag located at the anal dimple below which an instrument can be passed), and an anal membrane (through which one can see meconium) [31]. In terms of diagnostic tests done, there are a variety of imaging studies that can be done including abdominal ultrasonography to evaluate for urological anomalies and plain radiographs of the spine that can show spinal anomalies such as spina bifida and spinal hemivertebrae and also to measure the sacral ratio. If later in life, ie: after 3 months of age then MRI can be performed to better visualize the spinal cord [31]. The infant's physical examination, the appearance of the perineum, and any changes that occur over the first 24 hours of life are the factors determining the decision to go for immediate anoplasty or to perform a colostomy and delay the repair [32]. The outcome of surgery depends on the clinical presentation and early diagnosis, type of malformation, associated congenital malformations, available perioperative facilities, expertise of the surgeon, treatment options, good post-operative follow up. The overall mortality in children with ARM range from 8.97 % to 31.0% [8,13,17,33]. The mortality rate is higher in children with associated malformations [8,13], those with "higher" malformations [10] and in the neonatal period [9]. The commonest causes of death are associated malformations and sepsis [8,13]. The aim of this study is to analyze and evaluate patients with anorectal malformations (Bucket-handle deformity) in terms of their hospital course and to report a 3-year outcome and experience gained with surgical management for children diagnosed with anorectal malformation (Bucket-handle deformity).

2. Methodology

This 3-year cross-sectional study was undertaken at the pediatric surgery department in Khartoum North Teaching Hospital, Khartoum state; Sudan between March 2016 and March 2019 for all pediatric patients with Bucket-handle deformity admitted to the department. The participants were all pediatric patient with Bucket-handle deformity in the pediatric surgery department in Khartoum North Teaching Hospital the period (March 2016 - March 2019). Data was collected using data collection sheet (Appendix 1) by the in-charge surgeon. Statistical analysis was performed using the Statistical Package for Social Science (SPSS,

Version 23). Results were expressed as tables. The ethical approval was obtained from the local ethical committee at Khartoum North Teaching Hospital.

3. Results

A total of 72 children were managed for anorectal malformation (Bucket-handle deformity) during the period (2016-2019) were included, as expected we have a slightly increased number of male patients constituting 39 patients (54.2%) when compared to females, 33 patients (45.8%). The majority of patients involved in the study were born at home (76.4%) and around the same percentage (77.8%) were born vaginally. When it comes to their gestational age at time of delivery, almost (82%) were born at term and about (16.7%) were preterm and only small fractions (1.4%) were post-term (Table 1).

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	Preterm	12	16.7	16.7	16.7
	Term	59	81.9	81.9	98.6
	Post-term	1	1.4	1.4	100
	Total	72	100	100	

Table 1: Gestational age distribution

With regard to the associated anomalies in our population sample, (13.9%) have other anomalies that were mainly cardiac and only (4.2%) have an associated family history of congenital anomalies. While most patients admitted were not septic, only (9.7%) were septic on admission. Almost all patients have received antibiotics therapy and blood transfusion throughout their hospital course and none of them needed an ICU admission. Echocardiography was done for (44.4%) of patients, while chest x rays, abdominal x rays, abdominal CT, abdominal ultrasound and barium studies were not done to any patient. They were aged between one day of age and 8 years with a mean age for patients involved was 2.28 years and 2.3 years at the time of surgery with a standard deviation of 2.39 and 2.40 respectively. The average number for the days the patients stayed in hospital pre-operatively was 6.56 days and 10.72 days post-operatively with standard deviations of 6.56 and 10.72, respectively. When comparing gender with regard to the age at presentation, age at time of surgery, pre-and post-operative hospital stay in days there was significant difference between males and females as follows:

Gender		N	Mean	Std. Deviation
Male	Age	39	3.436	2.2337
	Pre-op hospital stay in days	39	7.28	5.568
	Age at time of surgery	39	3.474	2.2152
	Post-operative hospital stay in days	39	11.79	4.578
Female	Age	33	0.909	1.7961
	Pre-op hospital stay in days	33	5.7	3.283
	Age at time of surgery	33	0.909	1.7961
	Post- operative hospital stay in days	33	9.45	5.063

Table 2: Shows means and standard deviations for age, pre-operative, age at time of surgery and post-operative hospital stay in days according to being male and female

Males were older than females with regard to their age at time of presentation and time of surgery and they had a slightly longer pre-op hospital stay and post-op hospital stay (table 2) when comparing to females. When comparing septic versus non-septic patients with regard to the pre-operative hospital stay in days, we noted that the average days patients stayed in the hospital if they were septic is about 9 days (M= 9.14, SD= 1.864) and about 6 days in non-septic patients (M= 6.27, SD= 4.833). When it comes their post-operative course, septic patients stayed for a longer duration averaged an approximately 12 days (M= 11.86, SD = 5.014) and around 10 days for non-septic patients (M= 10.60, SD= 4.927). Also, septic patients were younger than non-septic at age of presentation and time of surgery (table 3).

Was the patient septic on arrival		N	Mean	Std. Deviation
Yes	Age	7	0	0
	Pre-op hospital stay in days	7	9.14	1.864
	Age at time of surgery	7	0	0
	Post-operative hospital stay in days	7	11.86	5.014
No	Age	65	2.523	2.3937
	Pre-op hospital stay in days	65	6.28	4.833
	Age at time of surgery	65	2.546	2.3926
	Post- operative hospital stay in days	65	10.6	4.927

Table 3: Shows a comparison between septic and non-septic patients with Bucket-handle deformity with regards to their age, pre-operative, age at time of surgery and post-operative hospital stay.

4. Discussion

Anorectal malformations (ARM) include a wide spectrum of congenital defects with variable clinical presentations and outcomes, which represent a variety of diagnoses in which patients do not have a normal anal opening but instead, they have a fistulous tract that opens onto the perineum anterior to the anal muscle complex or into adjacent anatomical structures (Illustration 1 and 2). It can be associated with urinary or gynecological fistulas and can often present as part of a genetic syndrome. These defects may be isolated or may present with other associated congenital anomalies. Overall, the etiology of ARM has not been fully understood but it is more likely to be multifactorial; with some implicated genetic components [18]. The outcome of surgery is dependent on the clinical presentation and early diagnosis, type of malformation, associated congenital malformations, available perioperative facilities, expertise of the surgeon, treatment options, good post-operative follow up. The treatment options depend on clinical presentation and facilities available for the perioperative care of children with complex congenital malformations. A bucket-handle deformity is a rare subtype of ARM characterized by a subepithelial midline raphe fistula.

4.1 Illustration (1) (2)

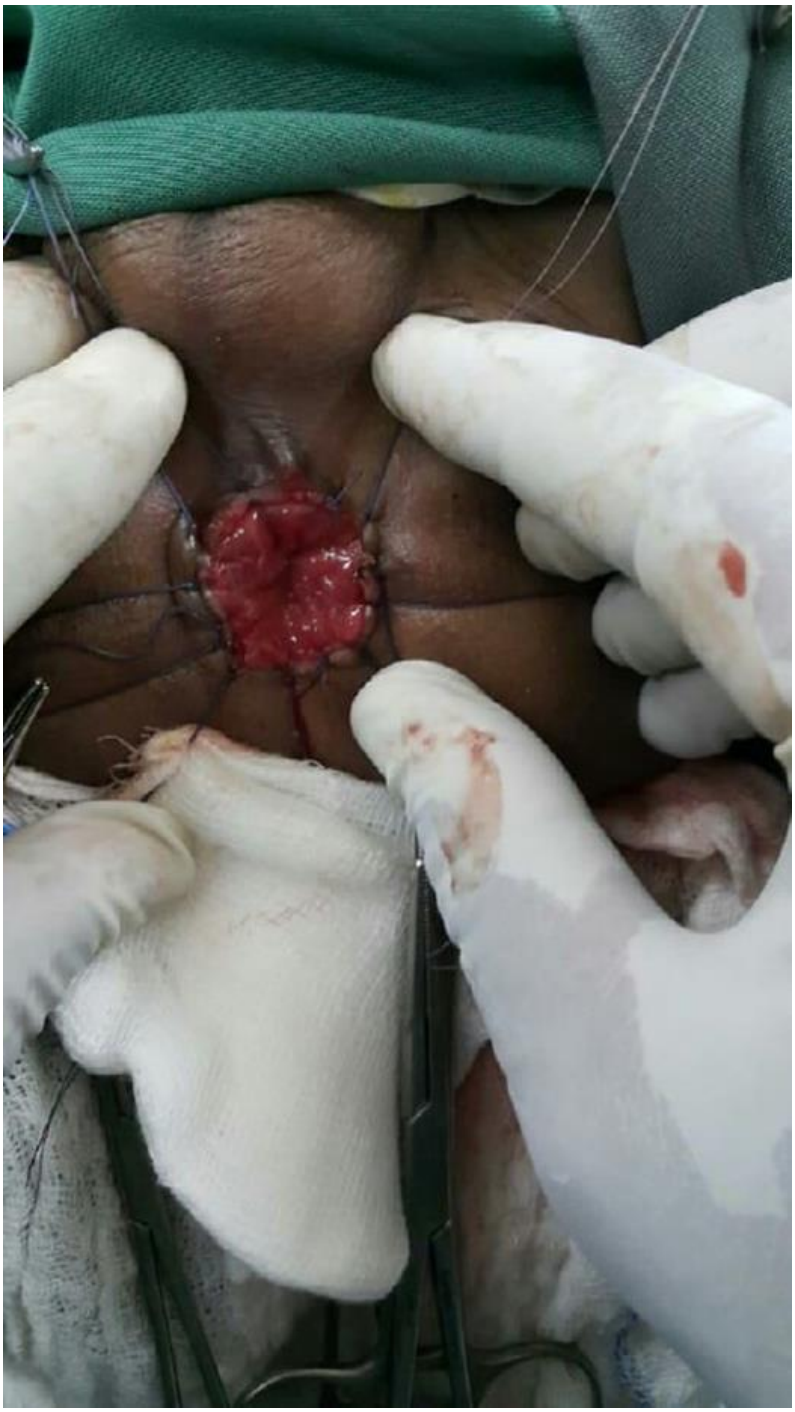

Figure 1: Shows a bucket-handle deformity after surgery

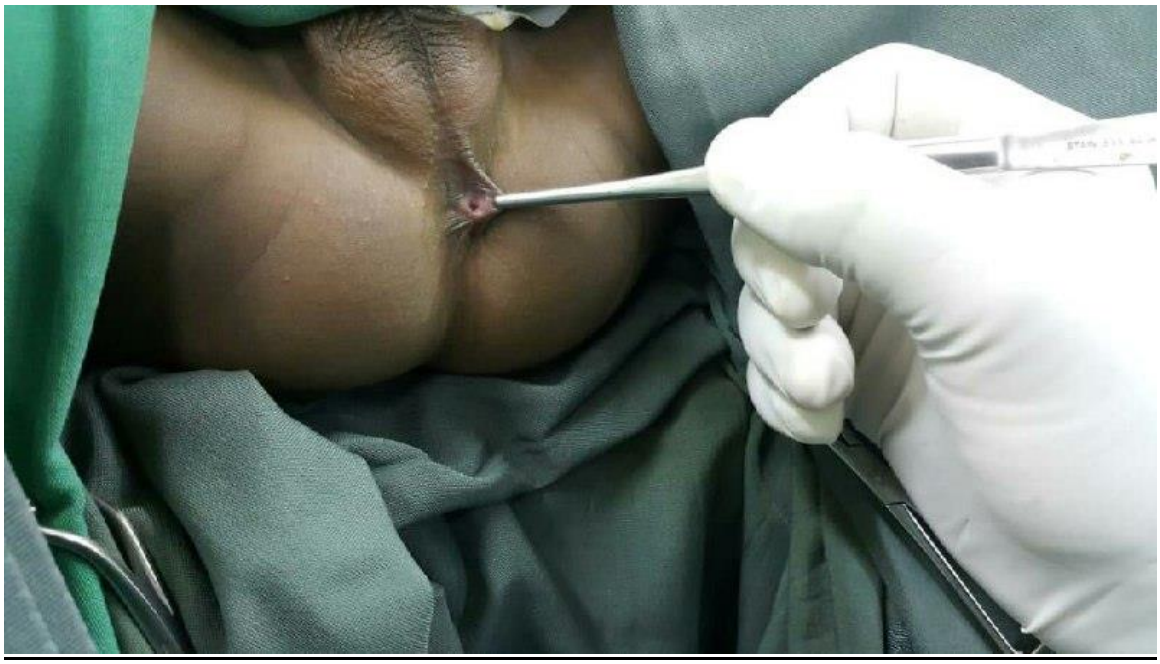

Figure 2: Shows a bucket-handle deformity before surgery

In this study, we give a view on the course of anorectal malformations (Bucket-handle deformity) patients in terms of their hospital course (pre, peri and post-operative course) and we try to know if there is any gender preference in patients with Bucket-handle deformity and also the complications following surgery. There are no studies available regarding the hospital course for patients with Bucket-handle deformity specifically, instead almost all studies published online have their focus on all types of ARM in general. Therefore, this survey is considered the first international and national survey on this deformity in terms of their hospital course (pre, peri and post-operative course) and the outcome of surgery. Of the 72 pediatric patients with Bucket-handle deformity were included, 39 (54.2%) were males and 33 (45.8%) were females. There is a slight to moderate male preponderance in cases of ARM ranging from 55- 71% according to majority of reports in Africa [7,13] but a few has shown the reverse [14,17]. We have found that 4.2% of our patients have an associated family history of congenital anomalies. This study has showed that among 72 ARM patients included in the study, almost (80%) were born at term and about (16.7%) were preterm and only a small fraction (1.4%) were post-term. It is worth mentioning that a population-based case-control study including all patients with ARM born in Sweden 1973-2014, has found that prematurity and small for gestational age (SGA) were more common among ARM patients than non-ARM patients [3]. In this study, we have found that the majority of patients were born at home (76.4%) and around the same percentage (77.8%) were born vaginally. With regard to the associated anomalies in patients included, (13.9%) have other anomalies that were mainly cardiac. Literature search showed that patients with ARM have cardiac anomalies in 10 -30% of cases. Most associated anomalies will not require urgent

treatment [34] and the initial management should include an echocardiogram, through which the next step in management will be determined. The lower incidence of associated malformations (13.9%) in this study and in most reports from Africa can be attributed to the less accurate detection and possibly because some children with more lethal associated defects would have never be seen at a hospital after birth at home and subsequent demise [8,20]. For patient with ARM in general, when discussing the complications and outcomes associated with ARMs, both short- term and long-term issues must be assessed. With regard to the functional complications following the surgery for ARMs, they are often dependent on the type of lesion with lower lesions having a better functional outcome than higher lesions. Those with lower lesions are, however, more likely to have issues with constipation that remains a significant problem following the repair of ARM. If untreated, constipation can lead to overflow incontinence and poor bowel motility secondary to megacolon. Most patients with constipation following treatment of an ARM should be managed medically with stool softeners or enemas [35]. Nonetheless, in this study and with regard to the post-operative complications, we report no fecal or urinary incontinence and there was no constipation in the 6 months period following the surgery. The delayed presentation of patients with ARM may lead to neonatal intestinal obstruction, sepsis, aspiration pneumonia, intestinal perforation, and sometimes death [12,18,27,28,35]. In this study, we identified only 9.7% of our patients as septic on admission, and almost all 72 patients have received antibiotics therapy and blood transfusion throughout their hospital course (98.6%). None of the 72 patients have needed an intensive care unit (ICU) admission. In term of the pre- and post-hospital stay, we have means for pre-operative and post-operative hospital stay in days as 6.56 and 10.72 days, respectively. This long course can be attributed to the time needed to optimize patients for surgery and deal with problems aroused from late presentation post-operatively. Early management of a newborn with an anorectal anomaly is crucial. In developing countries, presentation can be delayed and may be associated abdominal distension, dehydration and sepsis. Initial resuscitation with intravenous fluid and broad-spectrum antibiotics holds the key for the final outcome in such cases. The same is true for our patients, as almost all patients have received antibiotics therapy and intravenous fluid or blood transfusion throughout their hospital course (98.6%).

5. Conclusion

Anorectal malformations are some of the most common structural congenital malformations treated by pediatric surgeons globally. This study showed that all pediatric patients with Bucket-handle deformity in the pediatric surgery department at Khartoum North Teaching Hospital had good surgical outcome with no post-operative complications despite having a relatively long pre-operative and post-operative hospital course.

6. Recommendations

Although results were excellent regarding the surgery, further studies should be carried out to expand the literature regarding evaluation of pediatric patients with Bucket-handle deformity in terms of their hospital course. More efforts need to be put to optimize pediatric patients for surgery even before referring them to a specialized pediatric surgery unit for the goal of minimizing the duration of their admission course. Further studies should be carried out regarding the long-term complications following surgical

management of pediatric patients with anorectal malformation especially bucket-handle deformity.

References

1. Smith CA, Avansino J. Anorectal Malformations. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing (2020).

2. Ameh E, Chirdan L. Paediatric surgery in the rural setting: prospect and feasibility. West Afr J Med 20 (2001): 52-55.

3. Lawal TA, Adeleye AO, Ayede AI, et al. Congenital paediatric surgical cases in Ibadan: patterns and associated malformations. Afr J Med Med Sci 46 (2017): 49-55.

4. Holschneider A, Hutson J, Pena A et al. Preliminary report on the International Conference for the Development of Standards for the Treatment of Anorectal Malformations. Journal of Pediatric Surgery 40 (2005): 1521-1526.

5. DeVries PA, Pena A. Posterior sagittal anorectoplasty. J Pediatr Surg 17 (1982): 638-643.

6. Borg HC, Holmdahl G, Gustavsson K et al. Longitudinal study of bowel function in children with anorectal malformations. J Pediatr Surg 48 (2013): 597-606.

7. Theron A, Numanoglu A. Birth prevalence of anorectal malformations for the western cape province, South Africa, 2005 to 2012. Eur J Pediatr Surg 27 (2017): 449-54.

8. Adejuyigbe O, Abubakar AM, Sowande OA, et al. Experience with anorectal malformations in Ile-Ife, Nigeria. Pediatr Surg Int 20 (2004): 855-858.

9. Hesse A, Appeadu-Mensah W. Anorectal Anomalies in Ghana- a review of 54 Cases. Afr J Paediatr Surg 3 (2006): 4-8.

10. Makanga M, Ntirenganya F, Kakande I. Anorectal malformations at University teaching hospital of butare in Rwanda: a review of 46 operative cases. East Central Afr J Surg 12 (2007): 110-115.

11. Eltayeb AA. Delayed presentation of anorectal malformations: the possible associated morbidity and mortality. Pediatr Surg Int 26 (2010): 801-806.

12. Lukong CS, Ameh EA, Mshelbwala PM, et al. Management of anorectal malformation: changing trend over two decades in Zaria, Nigeria. Afr J Paediatr Surg 8 (2011): 19-22.

13. Kuradusenge P, Kuremu RT, Jumbi G, et al. Pattern of anorectal malformations and early outcomes of management at moi teaching and referral hospital Eldoret-Kenya. East Afr Med J 91 (2014): 430-434.

14. Archibong A, Idika I. Results of treatment in children with anorectal malformations in Calabar, Nigeria. South Afr J Surg 42 (2004): 88-90.

15. Sowande O, Adejuyigbe O, Alatise O, et al. Early results of the posterior saggital anorectoplasty in the treatment of anorectal malformations in Nigerian children. J Ind Assoc Pediatr Surg 11 (2006): 85.

16. Gama M, Tadesse A. Management of anorectal malformation: experience from ethiopia. Ann Afr Surg 15 (2012) 25-28.

17. Mfinanga RJ, Massenga A, Mashuda F, et al. Clinical profile and outcome of surgical management of anorectal malformations at a tertiary care hospital in Tanzania. Tanzania J Health Res 20 (2018): 1-11.

18. Levitt MA, Pena A. Anorectal malformations. Orphanet J Rare Dis 2 (2007): 33.

19. Moore SW, Sidler D, Hadley GP. Anorectal malformations in Africa. S Afr J Surg 43 (2005): 174-175.

20. Moore S, Alexander A, Sidler D, et al. The spectrum of anorectal malformations in Africa. Pediatr Surg Int. 24 (2008): 677-683.

21. Rintala RJ. Congenital anorectal

malformations: anything new? J Pediatr Gastroenterol Nutr 48 (2009): S79-82.

22. Nah SA, Ong CC, Lakshmi NK, et al. Anomalies associated with anorectal malformations according to the Krickenbeck anatomic classification. J Pediatr Surg 47 (2012): 2273-2278.

23. Bălănescu R, Topor L, Moga A. Anomalies associated with anorectal malformations. Chirurgia 108 (2013): 38-42.

24. Stoll C, Dott B, Alembik Y, et al. Associated anomalies in cases with anorectal anomalies. Am J Med Genet A. 176 (2018): 2646-2660.

25. Watanabe Y, Ando H, Seo T, et al. Three-dimensional image reconstruction of an anorectal malformation with multidetector-row helical computed tomography technology. Pediatr Surg Int 19 (2003): 167-171.

26. Lawal TA, Olulana DI, Ogundoyin OO. Spectrum of colorectal surgery operations performed in a single paediatric surgery unit in sub-Saharan Africa. Afr J Paediatr Surg 11 (2014): 128-131.

27. Tong WD, Ludwig KA. Neonatal colon perforation due to anorectal malformations: can it be avoided? World J Gastroenterol 19 (2013): 3915.

28. Olatunji T, Igoche M, Anyanwu P, et al. Spontaneous ileal perforation complicating low anorectal malformation. Afr J Paediatr Surg 12 (2015): 152.

29. Lawal TA, Olulana DI, Ogundoyin OO. Spectrum of colorectal surgery operations performed in a single paediatric surgery unit in sub-Saharan Africa. Afr J Paediatr Surg 11 (2014): 128-131.

30. Lindley RM, Shawis RN, Roberts JP. Delays in the diagnosis of anorectal malformations are common and significantly increase serious early complications. Acta Paediatr 95 (2006): 364-368.

31. Falcone RA Jr, Levitt MA, Peña A, et al. Increased heritability of certain types of anorectal malformations. J Pediatr Surg 42 (2007): 124-127

32. Liu G, Yuan J, Geng J, et al. The treatment of high and intermediate anorectal malformations: one stage or three procedures? J Pediatr Surg 39 (2004): 1466-1471.

33. Kayima P, Kitya D, Punchak M, et al. Patterns and treatment outcomes of anorectal malformations in Mbarara Regional Referral Hospital, Uganda. J Pediatr Surg (2018).

34. Teixeira OH, Malhotra K, Sellers J, et al. Cardiovascular anomalies with imperforate anus. Arch Dis Child 58 (1983): 747-749.

35. Levitt MA, Kant A, Pen ˜a A. The morbidity of constipation in patients with anorectal malformations. J Pediatr Surg 45 (2010): 1228-1233.