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Prevalence, Management and Outcome of Anorectal Malformations in Children Aged 14 Years or Less: A 7 Years Retrospective Study in Three Hospitals of Douala

Prévalence, prise en charge et résultats des malformations anorectales chez les enfants de 0 à 14 ans : une étude rétrospective de 7 ans dans trois hôpitaux de Douala

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ABSTRACT

Background: Anorectal malformations (ARM) consist of a wide spectrum of congenital malformations involving the anus and rectum. Their incidence varies from 1 in 2000 to 1 in 5000 live births. The aim of our study was to determine the prevalence, assess the management and outcomes of anorectal malformations in Douala. **Materials and methods**: This was a hospital based retrospective and descriptive study at the paediatric and surgical units in three hospitals in Douala (Laquintinie hospital, Protestant Hospital and the Obstetric and Gynaecologic and Paediatric Hospital). All medical records of patients aged 0-14years hospitalized for Anorectal Malformations from January 2013 to December 2019 were reviewed. Data was collected using a structured data collection tool. Data analysis was done using the statistical package for social sciences (SPSS) version 25.0. **Results**: We had a total of 68 patients with ARM, 42 were males and 26 females with a male to female ratio of 1.6:1. The children's age ranged from 1 day to 270 days (average of 2.5days). There was delayed presentation in 50% of patients. 41(60.3%) had low ARM, 16(23.5%) had intermediate ARM and 11(16.2%) had high ARM. The most common in males was imperforate anus without fistula while in females was the rectovestibular fistula. Associated malformations were found in 8(11.8%) with genitourinary anomalies being the most common. Diagnosis was made following results of physical examination, invertogram and colostogram. Low ARM were managed by YV anoplasty with or without protective colostomy. High and intermediate ARM were managed by PSARP following colostomy. The mortality rate was 17.6%. **Conclusion**: The prevalence of anorectal malformations is low in Doualabut difficulties still arise during the management. Low anorectal malformations are the most common and males are more affected than females.Late presentations coupled with inadequate peri-operational reanimation services adversely influence the outcome.

RÉSUMÉ

Contexte. Les malformations anorectales (MAR) consistent en un large éventail de malformations congénitales impliquant l'anus et le rectum. Leur incidence variant de 1 sur 2000 à 1 sur 5000 naissances vivantes. Le but de notre étude était de déterminer la prévalence, d'évaluer la prise en charge et les résultats des malformations anorectales à Douala. **Matériels et méthodes.** Nous avons mené une étude rétrospective descriptive dans les unités pédiatriques et chirurgicales de trois hôpitaux de Douala (l'hôpital Laquintinie, l'hôpital protestant et l'hôpital obstétrique, gynécologique et pédiatrique). Tous les dossiers médicaux des patients âgés de 0 à 14 ans hospitalisés pour des malformations anorectales de janvier 2013 à décembre 2019 ont été examinés. Les données ont été recueillies à l'aide d'un outil de collecte de données structuré. L'analyse des données a été effectuée à l'aide du progiciel statistique pour les sciences sociales (SPSS) version 25.0. **Résultats.** Nous avons eu un total de 68 patients atteints de MAR, 42 étaient des hommes et 26 des femmes avec un ratio homme/femme de 1,6/1. L'âge des enfants allait de 1 jour à 270 jours (moyenne de 2,5 jours). La présentation a été retardée chez 50 % des patients. 41(60,3 %) avaient une MAR faible, 16(23,5 %) une MAR intermédiaire et 11(16,2 %) une MAR élevée. L'anus imperforé sans fistule était le plus fréquent chez les hommes, tandis que la fistule rectovestibulaire était la plus fréquente chez les femmes. Des malformations associées ont été trouvées dans 8 cas (11,8 %), les anomalies génito-urinaires étant les plus fréquentes. Le diagnostic a été posé en fonction des résultats de l'examen physique, de l'invertogramme et du colostogramme. Les MAR faibles ont été prises en charge par une anoplastie YV avec ou sans colostomie de protection. Les MAR élevés et intermédiaires ont été pris en charge par PSARP après colostomie. Le taux de mortalité était de 17,6 %. **Conclusion.** La prévalence des malformations anorectales est faible à Douala. Les malformations anorectales basses sont les plus fréquentes et les hommes sont plus touchés que les femmes. Les présentations tardives couplées à des services de réanimation péri-opératoire inadéquats influencent négativement le résultat.

INTRODUCTION

Anorectal malformations (ARMs) consist of a wide spectrum of congenital abnormalities which involve the anus and rectum. These malformations range from skin level defects such as rectoperineal fistulas to complex lesions such as persistent cloaca [1]. ARMs are among the frequent congenital anomalies encountered in paediatric surgery, with an estimated incidence ranging between 1 in 2000 and 1 in 5000 live births [2]. A study done in South Africa revealed an overall prevalence of 5.5 / 10,000 live births which is slightly higher than the 4.05 per 10,000 live births calculated in a survey of 4.6 million births in Europe between 1980 – 1994[3, 4]. Other studies further support the suggestion that ARM lesions are common in Black African populations and make a significant contribution to the burden of surgical disease in Africa [5, 6]. ARMs are the most common major structural congenital malformations presenting to general paediatric surgeons on the continent [7] and also the leading congenital cause of intestinal obstruction in African children [8,9]. The aetiology of ARM remains unclear but is likely to be multifactorial and include genetic and environmental factors [10, 11]. Anorectal malformations can also occur in association with other anomalies. A study carried out in 2012 showed that more than 75% of children with anorectal malformations have other associated malformations. The most frequent malformations seen were genitourinary (28%) and spinal anomalies (26%) [12]. Diagnosis of most ARMs can be suspected prenatally during the prenatal imaging study which shows findings of multiple systems with abnormalities (digestive, vertebral and genitourinary are most common) [13]. Postnatal diagnosis is made by physical examination. Imaging modalities which include ultrasound, cardiac echocardiography and spine MRI are used to rule out other associated anomalies [14]. Multidisciplinary approach in the management has been shown in more optimal care of patients with ARM [15]. In general, the child with ARM will need three surgeries to correct the malformation as determined by the anatomy. Some children will only need one surgery to place the rectum through the anal sphincter [15, 16]. Various complications arise during some of which require re-operation [17]. There is limited data on the prevalence, management and outcome of anorectal malformations in Douala. Our study aims to evaluate the prevalence, management and outcome of anorectal malformations in children aged 0-14years in three hospitals in Douala.

MATERIALS AND METHODS

It was a hospital based retrospective study in the surgical and paediatric units at the Laquintinie hospital of Douala, Protestant Hospital of Douala and the Obstetric and Gynaecologic and Paediatric Hospital of Douala. Case files of patients admitted for ARMs from January 2013 to December 2019 were reviewed using a designed data collection sheet. This was a 7 years retrospective study from January 2013 to December 2019. We included in the study, all available and complete files of patients <15years admitted for ARM. Data was collected from clinical records of neonates and from operation registers which included; Socio-demographic, clinical and paraclinical data, surgical management, complications and final outcomes. The analysis of the variables was carried out using the Statistical Package for Social Sciences (SPSS) software, version 25.

RESULTS

A total of 68 patients were treated for ARM in the three hospitals in Douala over a 7year period from 2013-2019. The total number of paediatric admissions was 26,952 and the total number of paediatric cases operated was 4,941. The prevalence of anorectal malformations amongst the paediatric admissions in the three hospitals was 0.3% and amongst the paediatric surgical cases -1.4%.

In this study, we had a total of 68 cases of ARM in the hospital of Douala. 42 cases (61.8%), and 26 cases (38, 2%) were females. The age at consultation varied between a minimum of 1 day and a maximum of 270 days with a median age of 2.5 days. We found birth weights between 1700g and 4000g with a median of 2950g.

As shown in table 1, 37.3% of patients presented with signs and symptoms of intestinal obstruction at the time of diagnosis.

Table 1: Clinical Features of ARM

Type	Low ARM	Intermediate & High ARM	Total	%
Feature				
Absence of anal opening	10	4	14	20.6
Fistula	10	11	21	30.9
Meconuria	0	5	5	7.3
Abdominal distention	21	6	27	39.7
Cloaca	0	1	1	1.5
Total	41	27	68	100

We found, according to the Wingspread classification, three types of malformations: Low – 60, 3% (n=41), Intermediate - 23, 5% (n=16) and High ARM – 16, 2% (n=11). The types of ARMs were also classified according to the Krickenbeck. From the 68 cases studied, the most common type of ARM was imperforate anus without fistula in males and vestibular fistula in females as represented in figure 1 below.

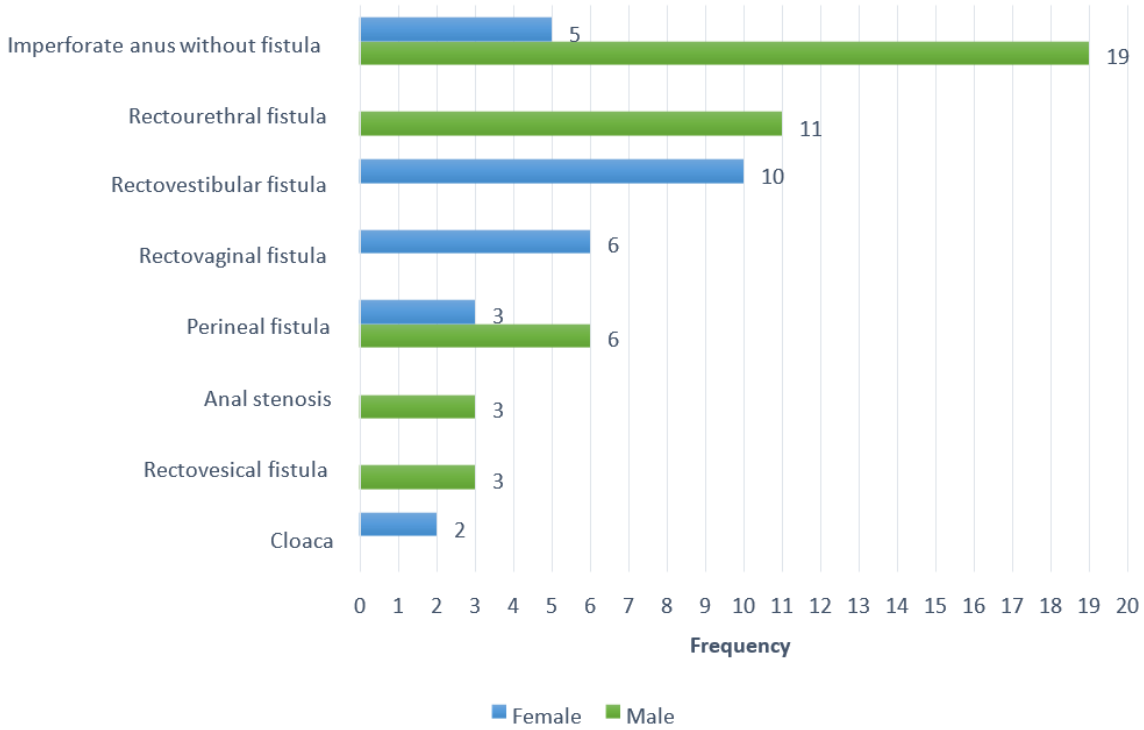

Figure 1: Type of Malformations according to Krickenbeck Classification

We found out that the invertogram was the most common investigation used to diagnose ARM (42.6%, 29 cases), followed by the colostography (15 cases, 22, 1%). Twenty -four patients (35, 3%) didn't pass any investigation. Concerning associated anomalies, abdominal ultrasound was the most common investigation used (93, 9%)
Associated malformations were reported in 8 (11.8%) cases, as showed in the table II.

Table II: Associated Malformations in Patients with ARM

Associated Malformation	Frequency	Percentage (%)
Omphalocele	2	2.9
Cryptorchidism	2	2.9
Duodenal atresia	1	1.5
Persistent urachus	1	1.5
Club foot	1	1.5
Anophthalmia	1	1.5
TOTAL	8	11.8

Concerning surgical management, prior to surgery, patients with obstruction were resuscitated, orotracheal intubation was done for all patients and they were placed under general anaesthesia (table 3).

Table 3: Distribution of Surgical Management in Relation to the Study population

Type	Low ARM	Intermediate and High ARM	Total
Surgery			
Anoplasty	23	0	23
Colostomy only	5	11	16
Colostomy and PSARP	13	16	29
Colostomy closure	9	11	20

PSARP :Posterior Sagittal Anorectoplasty

In this study we noted 8 patients with complications (4 with low ARM and 4 with high and intermediate ARM), that means a morbidity of 11, 7% (table 4).

Table 4: Distribution of Post-Operative Complications In Relation to Patients with ARM

Type	Low arm	Intermediate high arm	Total	Percentage (%)
Complication				
Local Infection	1	1	2	25,0
Anal atresia	2	1	3	37.5
Wound dehiscence	1	0	1	12.5
Haemorrhage	0	1	1	12.5
Colostomy prolapse	0	1	1	12.5
Total	4	4	8	11.8

A total of 12 patients (17.64%) died. Mortality was higher in patients who presented late and those who had intermediate or high ARMs. We didn't fine any difference between the mortality depending on the type of malformation (p=0.313).

DISCUSSION

This study showed a male predominance with 42 males (61.8%) and 26 (38.2%) females presenting with anorectal malformations giving a male to female ratio of 1.6:1. We found no explication for this predominance but this is similar to results reported by Pena et al in the United States of America (60% for males and 40% for females) [1], Makanga et al in Rwanda (63% for males and 37% for females) [18], Elmarie et al in South Africa (57.1% for males and 42.7% for females)[19]. Up to 50% (n=34) of patients presented late (after 2 days of life).This delay in diagnosis could be explaind by the lack of systematic perineal examinations of the newborns in the delivery room. Males with ARM generally presented early (67, 6%) because the most common type of ARM in males was imperforate anus without fistulas in which signs and symptoms of intestinal obstruction occurred early, prompting early presentation. The fistula in males usually has a small opening which leads to early obstruction. Females presented late because they had fistulas with wide openings and could allow passage of meconium without getting obstructed. This is similar to that reported by Marieme et al in Morocco [62]. Delay in presentation was attributable to the poor knowledge of the parents about ARMs, long distances they had to travel to reach the hospital and their low socio-economic status which made it difficult to afford transport and sustain themselves during their stay in the hospital. A majority of patients had a normal birth weight which indicates that birthweight had no influence on the occurrence of ARM.

At the time of diagnosis, 39.7% of patients had intestinal obstruction. This is similar to 38% reported by Marieme et al in Morocco [20]. Our results are lower compared to 63.7% by Mouafo et al in Côte-d'Ivoire [21]. This could be due to the fact that the number of patients with fistula was higher (44 cases) in our study compared to 28 cases in their study.

Concerning the type of ARM, a majority of cases (60.3%) had low Anorectal Malformations which is similar to the results reported by Makanga et al in Rwanda [22], and Luhiriri et al in Congo [23]. Imperforate anus without fistula was the most common type in males 19(45.3%) and in females, recto vestibular fistula was the most common 10(38.5%). This is similar to results reported by Kuradusenge et al in Kenya [24]. In contrast to studies done by Adejuyigbe et al in Nigeria [25] and Elmarie et al in South Africa [25] who reported recto urethral fistula as the most common in males. In this study, Invertogram was the most common investigation used to diagnose ARMs (42.6 %%). Ultrasound was not used for diagnosis in this study. Ultrasound was frequently used to check for associated malformations (93.8%) in patients who were available for screening. This frequency of ultrasonography was high since the investigation is non-invasive, was readily available and a preoperative investigation in all patients.

Associated malformations were reported in 8 (11.8%) cases in our study with genitourinary malformations being the most common. This is similar to 16.5% reported by Mfinanga et al in Tanzania [26]. We had a lower frequency of associated malformations because para-clinical examinations were not systematic in our patients. Some patients died before they could be adequately screened for associated malformations while others were lost to follow up.

In our study treatment for low ARM was single staged (Y-V anoplasty, cutback anoplasty) in a majority (23/41) of patients with low ARM. Anoplasty with a protective colostomy was performed in 13/41 patients. 5 patients had only colostomy because they presented late in acute intestinal obstruction and when the paediatric surgeon was unavailable. Some surgeons have proposed primary repair of all ARMs during the neonatal period. The advantages of this single stage surgical procedure are that the patient will have no memory of the procedure, colostomy and urinary tract infections can be avoided, dilatations are easier in young babies and the long term results are comparable with three staged surgical procedures [27]. However, management for intermediate and high ARMs in our study was in three stages: colostomy (descending colostomy), followed by the definitive surgery then colostomy closure. The advantages of this three staged procedure included the relieve of the intestinal obstruction through the colostomy, later permitting the determination of the type of ARM by colostography and diversion of faces following definitive surgery thus giving enough time for the perineal wound to heal without infections. The preferred method for the definitive surgery was Posterior Sagittal Anorectoplasty (PSARP) performed at the age of 5 months as the anatomy was clearer while separating the fistula. In this study we noted that the age of definitive surgery (PSARP) following colostomy ranged from 5 months to 5 years which implied that some children lived with colostomies and the ARMs for a long time before the definitive surgery. This delay was attributed to the low economic status of the parents and limited accessibility of paediatric surgeons. Following colostomy only 16/27 patients with intermediate and high ARM returned for PSARP. This could be due to the fact that some patients died before the surgery, and low socioeconomic status of the parents. PSARP with abdominal approach was performed in 3 patients with high ARM (rectovesical fistula). Serial anal dilatations were done as from two weeks following definitive surgery and colostomy closure performed 2 to 3 months following the definitive surgery. In our study, 66.2% (45/68) of patients were managed by colostomy at their first admission. Following colostomy, only 29/45 of the patients had definitive surgery and by the end of the study only 20 patients had their colostomy closed. This decrease in the number of patients may be due to the fact that some patients died before definitive surgery could be performed while others were lost to follow up.

We recorded complications which were noted during hospitalization. These complications were noted in 8 (11.8%) cases. This is similar to 11.8% reported by

Kayima et al in Uganda. Anal stenosis was the most common complication (3 out of 8). This was because some parents did not respect the protocol for serial anal dilatations which led to this complication.

We recorded 12 deaths (17.6%) which shows that patients can still die despite excellent surgery. This mortality is similar to 16% reported by Mfinanga et al in Tanzania [26]. Though the mortality rate was higher in patients with high and intermediate ARM compared to patients with low ARM (22.2% versus 14.6%), this difference was not statistically significant (p-value= 0.313). The mortality rate in children who presented late was higher than in children who presented early (20.6% versus 14.7% respectively), this difference was not statistically significant (P value= 0.376).These deaths could be due to late presentation of the patients with acute intestinal obstruction and sepsis which made it difficult for them to support the surgery and also due to lack of pre and postoperative intensive care facilities for these babies.

CONCLUSION

The prevalence of anorectal malformations is low (0.3%) in Douala but there still exist some challenges in the management of this malformations. Low anorectal malformations are the most common types seen and males are more affected than females. Physical examination and invertogram are sufficient for the diagnosis of the different types of anorectal malformations. Management of anorectal malformations is sorely by surgery. Late presentation coupled with the inadequate peri-operational facilities adversely influence the outcome of treatment for ARM.

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