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The hidden mortality of imperforate anus

Nikki Beudeker, Emily Broadis[1], Eric Borgstein[1], Hugo A. Heij

ABSTRACT

Background: Anorectal malformations (ARMs) affect 1 in 4000-5000 births and are a big challenge in western countries. However, little is known about ARMs in Africa. The aim of this study is to evaluate the incidence, treatment and outcome of ARMs in Malawi. **Materials and Methods:** Over a 4-year period (2006-2009), data was extracted from patients up to and including the age of 5 years or less who underwent a colostomy, posterior sagittal anorectoplasty or colostomy closure. **Results:** Of the data that could be retrieved 46 patients met the criteria of congenital ARMs; 65.2% were female (*N* = 30) and 34.8% were male (*N* = 16). The median distance from patient to the hospital was 79 km and the median age at presentation was 24 days. In female patients: The most common ARM was the vestibular fistula (*N* = 21; 70%), a recto-vaginal fistula was found four times, a cloaca was found three times and a perineal fistula or no fistula were both found once each. The most common ARM among boys was the recto-urethral fistula (*N* = 10). Two boys had no fistula. A perineal fistula and a recto-vesical fistula were both found once each. Nearly, half of the patients (*N* = 22) had complications. Complications occurred less often in the group, which lived closest to the Surgical Unit (25%). Associated anomalies were found in one patient. **Conclusion:** This study shows a skewed distribution of age at presentation and type of ARM. The most likely explanations are (1) the distance to the hospital: Because none of the male patients presented after 4 weeks and many may have passed away before arriving at the tertiary care centre; (2) lack of knowledge among primary caregivers since very few patients with rectoperineal fistulas were seen. The rate of complications was high, probably also related to advance age at presentation. Therefore, Malawi needs more awareness for earlier detection and quicker intervention.

Key words: Anorectal malformation, fistula, imperforate anus, Krickenbeck, Malawi

INTRODUCTION

Congenital anorectal malformation (ARM) is a condition of the digestive tract in which the anus and rectum are not normally developed. This anomaly can also involve the urinary and genital tract. ARMs occurs in 1 in 4000-5000 births and affects boys more often than girls.[1] Furthermore, ethnic differences appear to exist both in incidence and type of ARM.[2] When it comes to incidence and type of ARMs in people of African descent as opposed to Caucasians, various studies differ.[3] Ameh found that ARMs represented 67% of neonatal emergency surgical procedures in developing countries.[4] Therefore ARM constitutes a significant clinical and surgical load in developing countries, not only as emergency procedures, but also in terms of definitive corrective surgery and long-term follow-up. The aim of this study is to evaluate congenital ARMs among children in Malawi and to reflect on the presentation, the distance between hospital and patients' place of residence, associated anomalies and possible reasons for complications.

MATERIALS AND METHODS

Data were obtained from the paediatric surgical ward in the Queen Elizabeth Central Hospital in Blantyre, Malawi. Using the theatre record book, we registered children up to and including the age of five who underwent one of the following operations: Colostomy, posterior sagittal anorectoplasty (PSARP) or colostomy closure. Data was collected for the years 2006-2009 inclusive. A retrospective case

Department of Pediatric Surgery, Pediatric Surgical Center of Amsterdam (AMC & VUMC), Amsterdam, The Netherlands, [1]Department of Pediatric Surgery, Queen Elizabeth Central Hospital, Blantyre, Malawi

Address for correspondence:
Prof. Hugo A. Heij,
Meibergdreef 9, 1105 AZ, Amsterdam, The Netherlands.
E-mail: h.a.heij@amc.uva.nl

note analysis was performed, recording information about patient demographics, type of ARM, distance from patient residence to the hospital and clinical outcomes. We used the Krickenbeck Classification for recording the type of ARM.[1] A database was created with the use of Epidata. Statistical analysis was performed using the statistical package for the social sciences statistics (SPSS) version 19.0 (IBM Corp., Armonk, NY, USA) frequencies and descriptives.

To calculate the distance between the place of residence and Blantyre, the Macmillan Malawi Traveller's Map was consulted.[5] This map has a schedule, which shows the road distances between Malawi's main centres. If a location was not in the schedule, the distance was identified using Google maps.

RESULTS

In total, 131 children underwent formation of colostomy, PSARP or a colostomy closure for an ARM between 2006 and 2009. 57 medical records were retrieved from the medical records office. Of these 52 were ARMs. 46 met the criteria of congenital ARMs. Six patients with acquired ARMs were excluded.

Demographics

Of 46 children with ARMs, 65.2% were female (N = 30) and 34.8% were male (N = 16).

The median age at presentation was 24 days. The median age among boys was 3 days (range: 1-24) and girls presented after a median age of 90 days (range: 1-1185).

One male and seven female patients presented in the hospital during the 1st week after birth. Three female patients presented after more than 1 year [Figure 1]. Figure 2 shows the patients who presented after more than 1 year lived at the greatest distance more than 200 km outside of Blantyre. Patients who resided within a 5 km radius from Blantyre always presented within the first 4 weeks after birth (N = 3).

The majority of the patients lived within a 200 km radius from Blantyre city centre (N = 22; 47.8%). Only four patients were living within a 5 km radius from Blantyre [Figure 3]. The median distance from patient to the hospital was 79 km.

As shown in Figure 4, the most common ARM among females (N = 30) was the vestibular fistula (N = 21; 70%). A perineal fistula (N = 1) and no fistula (N = 1) were the least common ARMs in females. Three patients (10%) had a cloacal anomaly and four patients had a recto-vaginal fistula (13.3%).

The most common ARM among boys (N = 16) was the recto-urethral fistula (N = 10; 71.4%). The ARMs, which

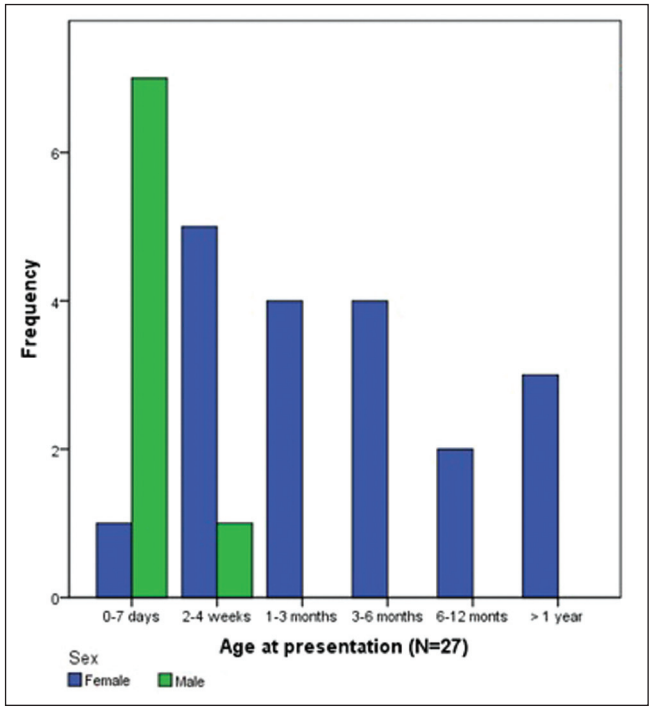


Figure 1: Age at presentation of study population

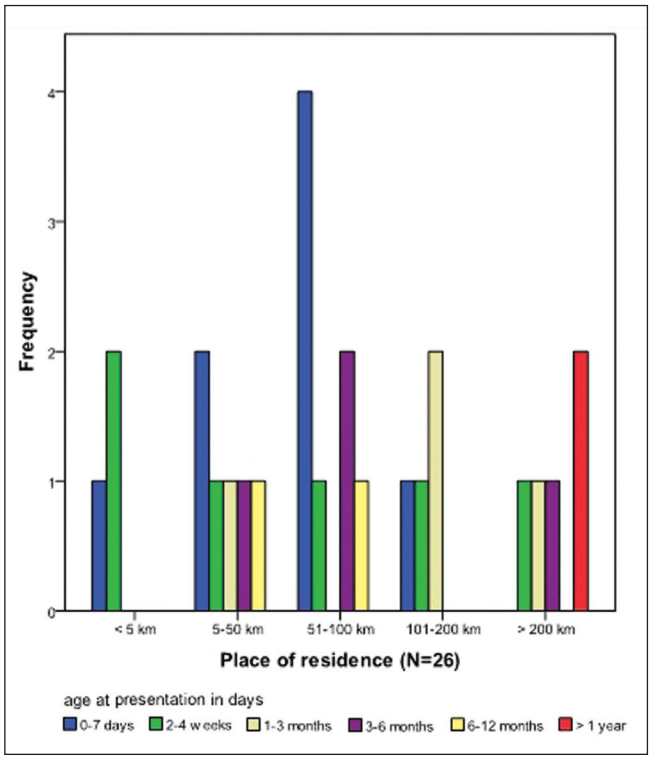


Figure 2: Age at presentation versus place of residence

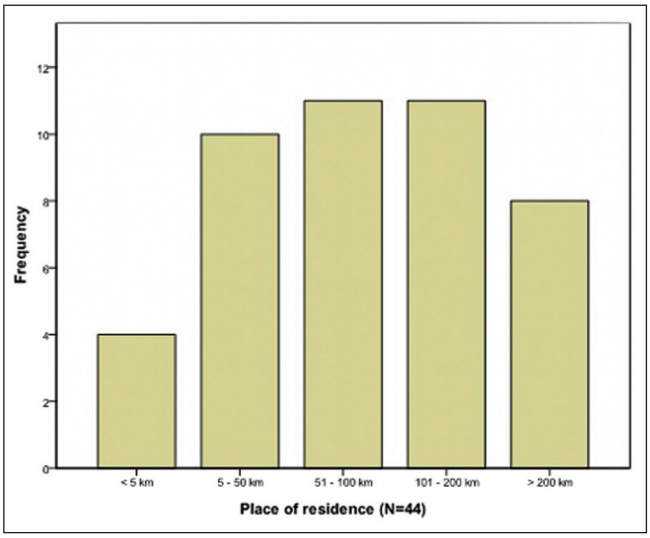


Figure 3: Place of residence of study population

were the least common in this group, were the perineal fistulae and the recto-vesical fistulae. These ARMs were both found once each. In two patients no fistula was found. Documentation was incomplete in two patients and they could not be classified. Anal atresia, rectal atresia/stenosis, pouch colon and H-fistula were not found in our cohort of patients.

There were few associated anomalies. One patient was noted to have an underdeveloped right leg, but in no other patient were other anomalies documented.

Outcome

There were 12 early complications in 9 patients: wound infection or dehiscence,[6] pneumonia,[2] abdominal wound dehiscence after closure of colostomy.[1]

Late complications were seen in 17 patients, four of whom had early complications that extended into the long-term follow-up. Six children developed an anal stricture, five had a partial breakdown of the PSARP repair, two patients had mucosal prolapse of the neo-anus and one patient developed a urethral stricture. In the 41 patients who initially underwent a colostomy formation [6 missing data] three patients had a colostomy prolapsed. The response to dilatation of anal strictures and the outcome in terms of incontinence were not reported in the files. There was one death in the cohort. A 5 day old boy died of sepsis secondary to meconium aspiration, 4 days after the formation of a colostomy.

Figure 5 shows the relationship between distance to the hospital and the frequency of complications. Patients living in the direct vicinity had 25% risk of

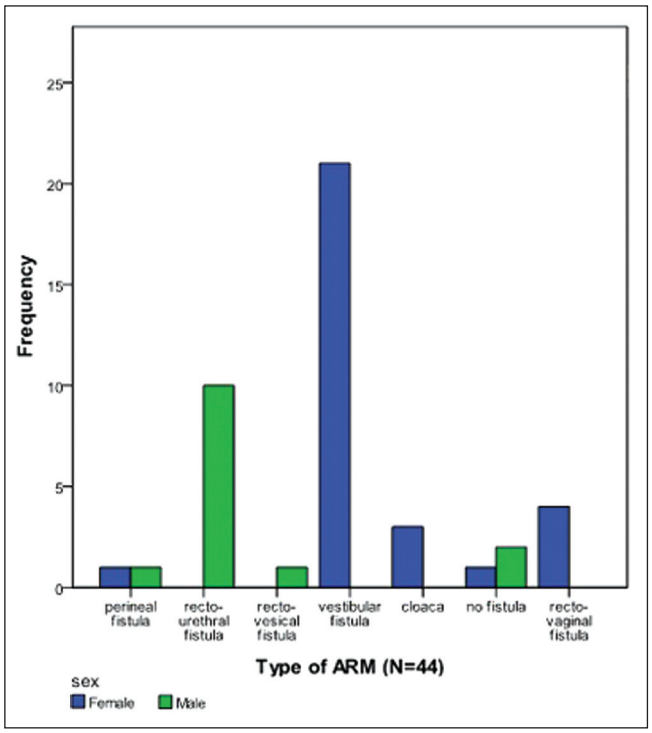


Figure 4: Type of anorectal malformation versus sex

complications, whereas this risk in children living more than 50 km from the hospital rose to 63%.

DISCUSSION

The number of patients in this study is small and we suspect a gross underestimate of the true number of patients aged less than 5 years with the ARM in Malawi during 2006-2009. Malawi has a population of around 15 million, with an estimated 600,000 live births per year.[7] Although the exact incidence of ARMs in Africa remains unclear, the expected incidence of ARM in a Black African population ranges from 120 to 265 per year, with a male predominance.[1-3,8]

There are several factors which contribute to our low pick up. Firstly, case notes were often missing from storage therefore only 46 case notes were analysed, out of 131 patients identified.

Secondly, we suspect that patients with a perineal fistula may not be recognised as an anomaly by the parent or health-care worker (HCW), particularly if it is not causing a complete obstruction. The high residue diet in Malawi may contribute to the low incidence of defecation disorders.

Third, our study has found that the distance from the hospital is an important factor in clinical outcome.

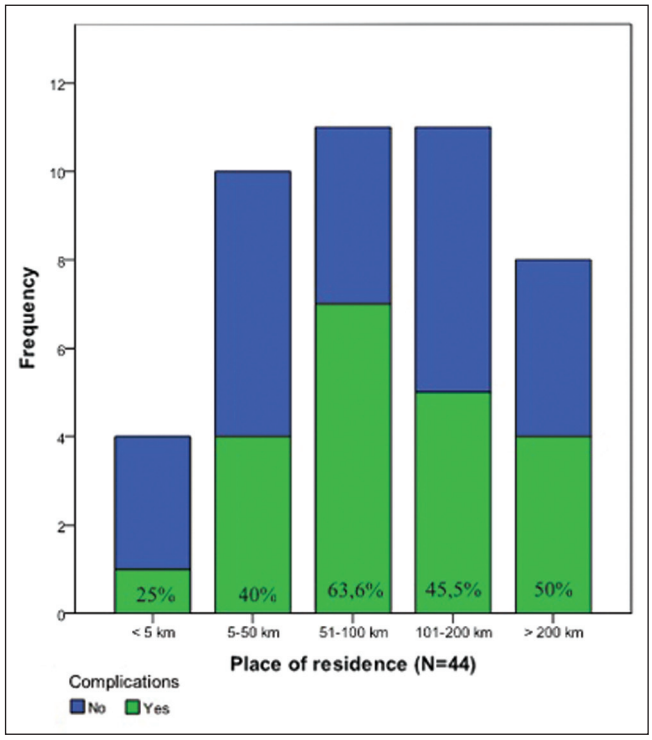


Figure 5: Place of residence versus complications

Social and financial impediments for travelling long distances play a role. As shown in Figure 3, only a small minority lived within a 5 km radius from Blantyre. The median distance from patient to the hospital was 79 km, which is a more than 1 h drive by minibus. Colostomies are only performed in the central hospitals in Malawi and definitive surgery for the ARM is only available in one hospital in the entire country.

Delayed presentation of ARM is widely described in the literature. The prevalence of delayed presenters of ARMs in developing countries reached 25%.[9] When using the same criteria as Sinha *et al.*[9] described, one-third (33%) of our patients can be classified as delayed presenters. Delayed presentation could be the result of many factors. There are few qualified HCW in Malawi. Most women deliver at home and are seen only by a traditional birth assistant, therefore there may be a lack of knowledge or understanding of ARMs and surgical treatment may not even be known about. As mentioned previously, distance from the hospital will also add to delay in presenting.

Delayed presentation contributes in turn, to higher morbidity and mortality rates.[6,10-12]

In our study, there is a large difference in age at presentation between male and female patients. A reason could be that ARMs in boys are more obvious than the types of ARMs girls could suffer from. For example, a recto-urethral fistula is noticed sooner than a vestibular fistula because of absent anus and faecal contamination of urine. The common female variant of ARM does not usually lead to complete obstruction, whereas most male variants do. As a result, female lesions, such as vestibular fistulae, are likely to be missed initially. A Canadian study agreed with our findings.[12]

The most plausible reason why the boys all presented in the 1st month would be that the boys who were older than 4 weeks died before reaching hospital. The types of ARMs in male patients are not only more obvious than those of female patients, but if left untreated, are life-threatening. The inability to pass stool leads to serious complications such as bowel perforation and sepsis, thus mortality rates are higher in boys.[6,10,11] Coupled with the long distance patients had to travel to the hospital, it is possible that many boys may have been too late to get treatment.

The far most common ARM among girls is the vestibular fistula. Among boys the recto-urethral fistula is the most common, which both supports an earlier ARM study in Africa.[3] However, it is hard to compare the outcome of this study in Malawi with other studies in western countries since there is no uniformity in classification used. Nevertheless, there is a study done in the UK where the Krickenbeck classification is used and which also showed that the recto-urethral fistula is the most common ARM among boys.[1] A striking difference between the Western world and this study in Malawi is the low incidence of perineal fistulae. The Krickenbeck classification shows that this is a common fistula and Western studies support this.[1] However, this study in Malawi has only reported two patients with this type of fistula. This could be due to the fact that there are many more hospital deliveries in Western countries than there are in Malawi, so abnormalities are more likely to be diagnosed in the Western world. However, in Malawi there is a shortage of doctors in district hospitals. HCW may not have much knowledge of ARMs, especially when it comes to perineal fistulae, which are not really obvious at first sight, therefore as discussed above, the diagnosis may be missed.

Figure 5 showed that patients who lived closest to the hospital had the least post-operative complications. Patients who lived close to the hospital all presented within the first 4 weeks after birth. Therefore, living close to the hospital contributes to early presentation and thus fewer complications.

ARMs are frequently associated with other kinds of anomalies. Although the incidence of associated anomalies varies widely in the literature, it could be stated that incidence ranging from 45% to 78% is high.[13-16] Owing to the fact that the incidence in African countries is comparable to those internationally,[2] these associated anomalies are a major cause of high morbidity and mortality rates in children with ARMs.[15,17] This may lead us to expect a high incidence of associated anomalies in Malawi as well. However, other kinds of anomalies were rarely found in our population. It seems reasonable to expect that this is due to two reasons. Firstly, there is a lack of a thorough examination. Less knowledge of HCW and a lack of diagnostic imaging may result in anomalies that remain unnoticed. Secondly, as associated anomalies account for a higher mortality rate and are more frequently present in boys,[14] the low number of associated anomalies in our study population corresponds to the low number of male patients presenting at a later stage, supporting our theory that these patients may die prior to arriving at a tertiary care centre.

Summary

The findings in this study are based on hospital records. These are indubitably deficient and have led to biased results. Another cause of bias is the selection of patients who reached the hospital. It can only be guessed how many patients with ARM did not present at the tertiary care centre. For a crude birth rate of 44 children per thousand inhabitants,[7] one would expect that annually 200 children with ARM are born in Malawi. The 46 patients analysed in this study, therefore represent the "ears of the hippo." Likely explanations for this are: (1) Lack of recognition of minor anomalies such as rectoperineal fistulae; (2) logistic problems in travelling long distances in rural Africa, so that many patients who need specialised paediatric surgical care may not arrive at the centre. This may lead to an increased mortality of patients with types of ARM that, unless treated early, are not compatible with life, especially higher anomalies in boys. Thus, girls with rectovestibular fistula have the best chances for survival.

CONCLUSION

Many patients with ARM in Malawi never reach specialised paediatric surgical care. Some of them have low anomalies and probably do well and others die before arriving at the hospital. This represents the hidden mortality of ARM in rural Africa. Improvement of health-care services, but also of logistics is required to turn this tide.

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