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Pattern of anorectal malformations in Sudan

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ABSTRACT

Anorectal malformations are a congenital malformation in which the terminal portion of the hindgut lies partially or completely outside the sphincter mechanism. To determine the pattern of anorectal malformations among Sudanese children a multicentric cross sectional study included all newborn babies admitted with imperforated anus during the period from Jan 2012 to Dec 2013. Data were collected using a predesigned questionnaire. The study included 59newborn babies their mean age was 2.6 ± 1.2 days (range, 1- 6 days), with male predilection; male to female ratio was 1.7:1. The defect was high in 61%. Male patients had a tendency to develop high imperforated anus (31/37 (83.8%)), whereas females had a tendency to develop low variety (13/22 (59.1%)), p=0.000. Associated fistula was seen in 18 (30.5%) patients, and 5.1% had a family history of imperforated anus. Co-existent anomalies were seen in 16.9%, and the commonest were genital anomalies and Down syndrome that observed in 6.8% and 3.3% respectively. The presence of co-existent anomalies was not affected by pattern of the condition, maternal age or gender (p> 0.05). In conclusion malesare affected morethan females and theyhad high defects more frequently. Females were more likely to have low lesions. The most common associated defects were genital anomalies and Down syndrome.

Keywords: Anorectal malformations;Imperforated anus (IA); Pattern; Fistula; Co-existent anomalies.

Introduction

Imperforated anus (IA) has been described in literature since ancient times [1]. Most authors have written that the average incidence worldwide is 1:4000 — 5000 live births [2,3], although the condition is more common in some areas such as Africa because of high fertility rate and associated malnutrition and poor antenatal care. Anorectal malformations are slightly more common in boys, and boys are twice as likely as girls to have higher anomalies. Some families have a genetic predisposition, with anorectal malformations being diagnosed in succeeding generations [4]. Anorectal malformations represent a spectrum of abnormalities ranging from mild anal anomalies to complex cloacal malformations. The etiology of such malformations remains unclear [3,5] and is likely multifactorial [3].

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It occur as a result of failure of development of the cloacal membrane or failure of recanalization of the secondarily closed anal canal during embryonic life [5]. There are however reasons to believe there is a genetic component [3], as imperforated anus might presents in association with several syndromes [6]. Although imperforated anus may occur as an isolated malformation, it coexists with duodenal atresia, tracheoesophageal fistula, vertebral and renal anomalies, Down syndrome, and congenital heart disease. Patients with Down syndrome usually have a unique anomaly – imperforated anus with no fistula [7]. Approximately 60% of patients have some form of associated urologic malformation [8]. The position of the distal rectal pouch with respect to the puborectalis sling of the levator ani muscle is a critical factor when considering whether an imperforated anus is of the high-type, intermediate-type or low type, i.e., above the sling (high or supralevator), at the sling (intermediate), and through the sling (low or infralevater) [1,9]. Various radiological modalities have been used to determine the level of the distal pouch in infants with IA. The modalities include inverted lateral radiography (invertography), distal colostography (loopography),

US, CT and MRI [10-13]. Of the various diagnostic modalities, transperineal ultrasonography (US) has been used to determine the IA type and to search for an associated internal fistula. Previously, the differentiation of a low-type from a high-type IA had been performed indirectly by measuring the distance from the distal rectal pouch to the perineum [10,14,15].However, recent improvement in US resolution has facilitated the identification and determination of location of internal fistulas with the use of transperineal US [14].To our knowledge, a comprehensive account focusing specifically on congenital anomalies of the alimentary tract in Sudan has not yet been documented. This study is vital in determining the surgical anatomy and pattern of imperforated anus in our local population, so the appropriate definitive repair can be planned.

Patients and methods

A multicentric cross sectional study was carried out in Khartoum, National Ribat University, Soba, Omdurman and Khartoum north teaching hospitals, during the period from Jan 2012 to Dec 2013. It included all newborn babies from both sex with anorectal malformations.

Anorectal malformationsare classified according to the International Classification of 1970 and the proposed "Wingspread" modification of 1986 as a high-type, intermediate-type and low-type. These classifications are based on the level of the distal rectal pouch relative to the puborectalis sling of the levator ani muscle [1,9].The required data were collected using a predesigned questionnaire, spread in master sheet, and entered computer. The statistical analysis was performed using Statistical Package for Social Science (SPSS) version 21. The results were provided as number (percentage) of patients. Categorical data was expressed as percentage and compared using Pearson Chi-Square test. The confidence level was set at 95% CI and p values less than 0.05 were statistically considered significant. Hospital administration ethical approval was obtained prior of conduction of the study and the confidentiality of the units and operators were kept.

Results

The study included 59 newborn babies (37(62.7%) were males and 22 (37.3%) were females) who underwent surgical management due to anorectal anomalies with male to female ratio of 1.7:1. Their mean age was 2.6 ± 1.2 days (range, 1- 6 days).Forty babies (67.8%) were born in hospital, whereas the remainder 19 (32.2%) were born at home. The discovery of the anomaly in the 1st day was in 75% and 52.6% among the group of patients who were born at hospital and at home respectively (Table 1). The observed difference in delayed diagnosis between the groups was statistically insignificant (p= 0.17).

Table 1: Comparison between the place of born and the time when the anorectal malformations were discovered (n=59)

Place of born	Day of discovery			Total
	1st day	2nd—3rd day	> 3rd day	
Hospital	30 (75.0%)	5 (12.5%)	5 (12.5%)	40 (67.8%)
Home	10 (52.6%)	6 (31.6%)	3 (15.8%)	19 (32.2%)
Total	40 (67.8%)	11 (18.6%)	8 (13.6%)	59 (100.0%)

Pearson Chi-Square= 3.568, p= 0.17

The pattern of anorectal malformations were high, low and intermediate as seen in 36 (61%), 17 (28.8%) and 6 (10.2%) respectively. Study found that male patients had a tendency to develop high imperforated anus as it was seen in 31/37 (83.8%), whereas females had a tendency to develop low variety as it was seen in 13/22 (59.1%), the difference was statistically significant (p=0.000), (Table 2).

Table 2: The pattern of Anorectal malformations in the study group (n=59)

Gender	Pattern			Total
	High	Intermediate	Low	
Male	31/37 (83.8%)[a]	2/37 (5.4%)	4/37 (10.8%)	**37 (62.7%)**
Female	5/22 (22.7%)	4/22 (18.2%)	13/22 (59.1%) [a]	**22 (37.3%)**
Total	**36/59 (61%)** [a]	**6/59 (10.2%)**	**17/59 (28.8%)**	**59 (100.0%)**

[a]The highest incidence

Associated fistula was seen in 18 (30.5%) patients (in 7/37 (18.9%) males and 11/22 (50%) females), the difference was statistically significant (p=0.012). Fistulae were seen in patients with low, high and intermediate imperforated anus in 10/17 (58.8%), 7/36 (19.4%), and 1/6 (16.7%) respectively, the difference was statistically significant (p=0.011). In 3 (5.1%) patients there was a family history of imperforated anus.Co-existent anomalies were seen in 10 (16.9%) patients, and the commonest were genital anomalies and Down syndrome that observed in four (6.8%) and two patients (3.3%) respectively (Table 3). The presence of co-existent anomalies was not affected by pattern of the condition, maternal age or gender as p values were p=0.08, p=0.37 and p=0.47 respectively.

Table 3: Anorectal malformations associated with other abnormalities

Co-existent anomaly	No.	Percentage
Genital anomalies	4 (2 abnormal genital and 2 undescended testis)	6.8
Down syndrome	2	3.3
Cardiac	1	1.7
Spina bifida	1	1.7
Single kidney and right ureter	1	1.7
Inguinal hernia	1	1.7
Total	**10**	**16.9**

Mean maternal age was 28.7 ± 4.5 years (Range, 19 to 42 years). Pattern of imperforated anus, presence of fistula and presence of coexistence anomaly were not affected by maternal age as p values were p=0.9, p=0.3 and p=0.4 respectively.

Discussion

Since unawareness of the variations of the anorectal malformations may be hazardous for the surgeon and patient, our intention in this study was to learn more about our patient population regarding the surgical anatomical variations of anorectal malformations in Sudanese patients. The study revealed a slight predilection for male infants that in keeping with literature findings [9,14].This study was conducted in the five major hospitals in the country, where the departments for Pediatric Surgery were established, and receives all cases of Pediatric Surgery converted from all other hospitals distributed in the country. This reflects the extent of the shortage in the departments of Pediatric surgery in this country. Consequently the delay in diagnosis observed in this study can be attributed to this shortage. Boys seem to be at a slightly higher risk than girls, this in accordance with the earlier study [16].In males, high defects were more common (83.8%), whereas low defects were commonest in females (59.1%), this almost similar to the results obtained by Bhargava P. et al [17]. Imperforated anus is a condition that is often found in conjunction with other serious anomalies, leading to significant morbidity and mortality [18]. In this study, additional anomalies were present in 16.9% of all cases with imperforated anus. The rate of additional anomalies was lower than in previous postnatal reports [19,20], but the rates of various types of associated anomalies were in accordance and in line with earlier study [21]. Of interest the study found that the associated malformations are not affected by the pattern of the imperforated anus. Whereas, previous studies have shown that associated malformations are more frequent in “high” defects that are complex and difficult to manage with a poor functional prognosis than in “low” defects that are less complex and easily treated with an excellent functional prognosis [22].Regarding family history of imperforated anus, %94.9 of the patients has no family history of imperforated anus. This result is in support of Spouge et al. [23] who stated that 96.8% had no family history and 3.2% had family history. Similar results were given by Stoll et al. [24] who mentioned that 92.6% had no family history and 7.4 % had no family history of imperforated anus, more report was given by Rittler et al. [25].The mean maternal age of 28.7 years did not differ from the mean maternal age of the non-selected population in Norway [21].The association between anorectal malformations and maternal age has been reported to be increase with increasing maternal age [26]. This study found that there is no apparent association between maternal age pattern of imperforated anus, and presence of fistula or other

associated anomalies, this was in accordance with earlier study [24].

Conclusion

When performing surgery for anorectal malformations, we must acknowledge the possible variations in its anatomy to ensure a safe method of management to be used. Anorectal malformations occurred mainly in males and they had high defects more frequently. Females were more likely to have low lesions. The most common associated defects seen were genital anomalies and Down syndrome. The surgical approach to repairing these defects mandate surgeons to view the anatomy of these defects clearly, to help in repair them, and to learn about the complex anatomic arrangement of the region. Surgeons with lacked objective anatomic guidelines unfortunately might leave many patients with various unwanted preventable complications.

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