Anorectal malformations at University Teaching Hospital of Butare in Rwanda: A review of 46 Operative cases.

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Background*:* Anorectal malformations (ARM) are comprised of a wide spectrum of disease that involves congenital anomalies of the anus and rectum, as well as the urinary and genital tracts. They occur in approximately 1 in 5000 live births and affect males more than females. They may present as a single or as a combination of abnormalities. Survival and prognosis of anorectal malformations depends upon the severity and number of the associated anomalies. This study was aimed at establishing the incidence, types of ARM, associated anomalies and outcome of surgery among children with anorectal malformations seen at University Teaching Hospital of Butare in Rwanda.
Methods: A descriptive retrospective study was carried out on 46 children with ARM admitted to Paediatric and/or surgical departments of Butare University Teaching hospital and operated between 1st May 2002 and 31st May 2007. Information regarding age at presentation, sex, type of abnormality, associated congenital anomalies, operative findings, outcome of surgery was extracted from the patients' clinical records and operation registers. Data was analyzed using Epidata and SPSS 11.3 computer programs.
Results: Of the 2264 patients operated on for gastrointestinal conditions during the period under review, 648 (28.6%) of them were children of whom 46 (2% of all operated digestive pathologies or 7% of digestive pathologies operated in children) were for anorectal malformations. All these 46 were included in the study. 63 % were male and 37% female. The majority of them consulted in the first week of life (56.5%) and 43.5% have been operated in that first week. 60.9% of our cases had low ARM (LARM), 26.1% had intermediate ARM (IARM) and the rest (13%) had high ARM (HARM). Associated anomalies were seen in 77.4% of patients. The major associated anomalies consisted of fistulas (47.3%), gastrointestinal malformations (17.2%), skeletal malformations (10.75%), and cardiac (2.15%). The overall survival rate was 87%. It was 92.85% for LARM, 100% for IARM and 33.3% for HARM with a significant statistical difference (p=0.011). The survival rate was 91.3% and 90.5% among those who underwent colostomy and anoplasty as first intervention respectively.
Conclusion: Anorectal malformations are common in children's surgical pathologies, the incidence being in favour of LARM. The survival depends upon the type of ARM, the treatment and the severity of associated anomalies. All children with ARM merit a meticulous search for associated anomalies in order to optimize the management.

Introduction

Anorectal malformations comprise a wide spectrum of disease affecting boys and girls and can involve malformations of the distal anus and rectum, as well as the urinary and genital tracts. Malformations range from minor easily treated defects that have an excellent functional prognosis to complex defects that are difficult to manage, are often associated with other anomalies, and have a poor functional prognosis. The most common anomaly in females is a rectovestibular fistula. Perineal inspection shows a normal urethra, normal vagina, and another orifice that is the rectal fistula in the vestibule. Early diagnosis, management of associated anomalies, and efficient meticulous surgical repair provide patients the best chance for a good functional outcome. In 80-90% of newborn boys, clinical evaluation and

urinalysis provide enough information for the surgeon to decide whether the baby requires a colostomy. Performing a diverting colostomy is the safest option for a surgeon without extensive experience in anorectal anomalies when faced with a baby who has clinical evidence of a rectovestibular fistula. Colostomy before the main repair avoids the complications of infection and dehiscence. Definitive repair of this anomaly in the newborn period should be reserved for surgeons who have significant experience repairing these defects. This study was aimed at establishing the incidence, types of ARM, associated anomalies and outcome of surgery among children with anorectal malformations seen at University Teaching Hospital of Butare in Rwanda.

Patients and Methods

This was a retrospective descriptive. The study population consisted of 46 children with anorectal malformations admitted to Paediatric and/or surgical Departments of Butare University Teaching Hospital and operated between 1st May 2002 and 31st May 2007 inclusive. Case records of all these cases were retrieved and analyzed for, age at the first consultation and first intervention, types of ARM, associated anomalies, and type of the first intervention and the outcome of surgery. Data was analyzed using Epidata and SPSS 11.3 computer programs. The p-value equal to 0.05 or less was considered to statistically significant. The findings are presented.

Results

During the study period of 5 years and 1 month, 2264 patients had operations for gastrointestinal pathology. Of these, 648 (28.6%) were children. Forty-six (7.1%) of children that is 2.0% of the 2264 of cases operated for gastrointestinal conditions, had anorectal malformations. There was a predominance of males 63% with a male to female ratio of 1.7:1. The majority (56.5%) were hospitalized in the first week of life. Six (13%) consulted for the first time at more than six months of age (Table 1). The major complaint was absence of anus in 39.1% of cases. In 30.4% of cases, the patients presented with intestinal obstruction (Tables 2). Twenty patients (43.5%) were operated during the first week of their life (Table 3). Of all these 46 neonates and children, 60.9% (28 of 46) had low anorectal malformation (LARM), 26.1% (12 of 46) had intermediate anorectal malformation (IARM) and 13% (6 of 46) had low anorectal malformation (LARM) (Table 4). Associated anomalies were seen in 77.4% of patients.

The major associated anomalies consisted of fistulas (47.3%), gastrointestinal malformations (17.2%), skeletal malformations (10.75%), and cardiac anomalies (2.15%). In 10 (22.6%) of cases, there were no associated anomalies (Table 5). The majority (72.7%) of our patients were postoperatively hospitalised between 8 and 21 days with a minimum of 0 day (direct post operative death) and 42 days. The mean was 22 days (Table 6). The overall survival rate was 87%. It was 92.8% for LARM, 100% for IARM and 33.3% for HARM with a statistically significant difference (p=0.011). Thirty six (78.3%) recovered without any complications.

Postoperative complications were recorded in 14 cases. Infection has been the immediate complications in 10 cases, stool incontinence in 2 cases, while anal stenosis was reported later in 2 cases. There were 6 deaths giving a 13.0% mortality rate. Two children who underwent anoplasty died due to anaesthetic complications (Table 7).

According to the type of first intervention, the survival rate was 91.3%, 0%, 90.5% among those who underwent colostomy, ileostomy, anoplasty as first intervention respectively, with a significant difference (p=0.033). (Table 9)

Age	Number	Percentage
First week of life	26	56.5%
2-4 weeks	6	13.05%
5-24 weeks	8	17.4%
> 24weeks	6	13.05%
Total	46	100%

Table 1. Age Distribution

Table 2. Distribution according to main clinical sign at the first consultation

Signs	Number	Percentage
No anus on birth	18	39.1%
Meconiuria	10	21.7%
Constipation	4	8.7%
Occlusive syndrome	14	30.4%
Total	46	100%

Table 3. Distribution according to the age of first intervention

Age	Number	Percentage
First week of life	20	43.5%
2-4 weeks	8	17.4%
5-24 weeks	12	26.1%
> 24weeks	6	13.0%
Total	46	100%

Table 4. Distribution according to types of malformation

Type	Number	Percentage
LARM	**28**	**60.9%**
IARM	**12**	**26.1%**
HARM	**6**	**13%**
Total	**46**	**100%**

Table 5. Distribution of associated anomalies

Type of Associated Anomalies	Sub type of Associated anomalies	Number	Percentage
Gastro-intestinal malformations	-Ileocaecal atresia	2	4.3%
	-Sigmoid colon duplication	2	4.3%
	-Intestinal malrotation	4	8.6%
Fistulas	-Recto-vesical/urethral	4	8.6%
	-Recto-vaginal	8	17.2%
	-Recto-perineal	8	17.2%
	-Ano-cutaneous	2	4.3%
Skeletal malformations	-Polydactylia	2	4.3%
	-Pieds bots*	3	6.45%
Others	Situs inversus	1	2.15%
No anomaly diagnosed		10	22.6%

Table 6. Duration of *Hospitalisation*

Days	Number	Percentage
< 7 days	4	9.1%
8-14 days	20	45.4%
15-21 days	12	27.3%
> 21 days	8	18.2%
Total	44	100%

Table 7. Distribution according to outcome

Outcome	*Frequency*	*Percentage*
Still in life:		
-Healed	36	78.3%
-With complications	4	8.7%
Dead	6	13%
Total	46	100%

Table 8. Distribution according to type of ARM vs outcome

Outcome	*Type of ARM*						*P value*
	LARM		IARM		HARM		
	Freq.	Perc.	Freq.	Perc.	Freq.	Perc.	
Still in life	26	92.85%	12	100%	2	33%	
Dead	2	7.15%	0	0%	4	66.7%	0.011
Total	28	100%	12	100%	6	100%	
Overall survival rate is 87%							

Table 9. Distribution according to complications

Complications	*Frequency*	*Percentage*
Infection	10	50%
Anal stenosis	1	5%
Stool incontinence	3	15%
Death	6	30%
	20	100%

Table 10. Outcome According to Type of first intervention

Types of intervention	*Outcome*				*P value*
	Still in life		Died		
	Frequency	Percentage	Frequency	percentage	
Colostomy	21	91.3%	2	8.7%	
Ileostomy	0	0%	2	100%	0.033
Anoplasty	19	90.5%	2	9.5%	
Total	40		6		

Discussion

Anorectal malformations comprise a wide spectrum of disease affecting boys and girls and can involve malformations of the distal anus and rectum, as well as the urinary and genital tracts[1,14]. They occur in approximately 1 in 5000 live births[3,4]. It is somewhat more common in boys than girls and may include a single abnormality or a combination of abnormalities[4,7,13]. Associated congenital anomalies in neonates with anorectal malformation assume significance, as survival and prognosis depend upon the number and severity of the associated anomalies[13]. Some anomalies like those of the vertebra, though not letndhal, may have a direct bearing on the ultimate functional outcome of the case. Other anomalies involving the cardiac, gastrointestinal and genitor-urinary systems a may lead to morbidity and mortality during the initial management of neonates with ARM[7,9,13]. The incidence of associated anomalies with ARM range from 30 to 70% of cases according to various studies[7,8,9,11,13]. The incidence of associated anomalies basically depends upon the meticulousness with which they have been sought after.

Malformations range from minor easily treated defects that have an excellent functional prognosis to complex defects that are difficult to manage. They are classified into 3 types: low, intermediate and high anorectal malformations[2]. The incidence of each type varies from study to study[5,7,8,9,10,11,12,13]. In our study, low ARM was the most frequent variety.

As far as treatment is concerned, throughout the centuries, doctors have seen and have tried to treat babies born with imperforate anus. Very few patients are described, so most patients are assumed to have died without treatment. Paulus Aegineta in the 4th century wrote the earliest account of a survivor of surgery for imperforate anus. He suggested rupturing an obstructing membrane with the finger or point of a knife and then dilating the tract until healing was complete. This approach was used for many years[3].

Almost 1000 years later, in 1660, Scultet treated an infant with anal stenosis with dilatation. In 1676, Cooke used incision and dilatation and advised care of the sphincter muscles. In 1787, Bell suggested using a midline perineal incision to find the bowel. In 1783, Dubois acted on Littre's suggestion from 1710 by performing an inguinal colostomy for imperforate anus. Other surgeons followed suit, but almost all infants died, so colostomy remained unpopular and only a procedure of last resort. Formal perineal proctoplasty (ie, mobilization of the bowel through a perineal incision with suturing of it to the skin) was described by Amussat in 1835, and this technique gained rapid acceptance. Strictures were less common than was observed with earlier procedures. In addition to Amussat, Dieffenbach described anal transposition (1826); Chassaignac used a probe through a stoma to guide the perineal dissection (1856); and Leisrink (1872), McLeod (1880), and Hadra (1884) recommended opening the peritoneum if the bowel was not encountered from below[2,3].

Imaging to delineate the abnormality was first advocated by Wangensteen and Rice in 1930. Single-stage abdominoperineal procedures became widely used after reports by Rhoads, Pipes, Randall, Norris, Brophy, and Brayton (1948-1949). Stephens (1953) described this procedure and emphasized preservation of the puborectalis muscle. This surgery and its modifications were the standard approach until 1980[2]. In 1980, the surgical approach to repairing anorectal malformations changed dramatically with the introduction of the posterior sagittal approach[2,5]. This approach allowed pediatric surgeons to view the anatomy of anorectal malformations clearly and to repair them under direct vision, with better visualization and understanding of the anatomy than previous approaches. Surgeons were able to understand the complex anatomic arrangement of the junction of rectum and genitourinary tract. This is the approach that is being used in our department of surgery and is giving good results.

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