ANORECTAL MALFORMATIONS: A REVIEW OF 10-YEAR EXPERIENCE IN SULAIMANI (KURDISTAN REGION/IRAQ) PEDIATRIC SURGICAL CENTER

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ABSTRACT

Background

Anorectal Malformation (ARM) is a well recognized condition that involves congenital anomalies of the anus and rectum. It may present as a single or as a combination of abnormalities.

Objectives

The aim of this study is to describe some epidemiologic data on anorectal malformation, estimating it's prevalence in Sulaimani province, and characterizing the cases with regards to age at presentation, gender, pathological classification, associated anomalies, postoperative complication, and mortality rate.

Materials and Methods

This descriptive epidemiological study extended from Mach 2006 to March 2016, including all of patients presented with ARM in pediatric surgical unit at Sulaimani Teaching Hospital.

Results

The total cases were 194 patients (122 male and 72 females). 92% presented on the first week of life. 44.2% had associated anomalies. The prevalence rate is 6.4 cases per 10 000 live birth. Stomal complications are the most common post operative complication. The mortality rate is 8.24%. The relation between mortality and associated anomalies was statistically analyzed.

Conclusion

The associated malformations rate and types are almost the same as in the literatures; however the prevalence rate was higher. This confirms the presence of geographic variation in ARM incidence, but could not confirm that the chemical attack or environmental impact of wars in this region had effects on increasing the rate of ARM due to lack of previous studies.

Keywords: *Anorectal Malformation (ARM), Imperforate anus (IA).*

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INTRODUCTION

Anorectal malformations (ARM) are well recognized conditions since antiquity [1]. Most authors agree that the average worldwide is 1 in 4000-5000 live births [2, 3]. The cause of ARM is unknown although the condition is more common in some areas and some families have a genetic proposed autosomal dominant inheritance [4, 5].

Anorectal malformations comprise a wide spectrum of disease affecting boys and girls and can involve malformations of the distal rectum and anus, as well as the urinary and genital tracts [6, 7]. Malformations range from minor easily treated defects that have an excellent functional prognosis to complex defects that are difficult to manage, and usually associated with other anomalies, and have a poor functional prognosis [8, 9]. Early diagnosis, management of associated anomalies, and efficient meticulous surgical repair provide patients the best chance for a good functional outcome [10, 11].

The aim of this study is to describe some epidemiologic data on anorectal malformation, estimating it's prevalence in Sulaimani province, and characterizing the cases with regards to age at presentation, gender, pathological classification, associated anomalies, postoperative complication, and mortality rate.

PATIENTS AND METHOD

This descriptive epidemiological study extended from Mar 2006 to Mar 2016, including all of the patients presented with ARM in pediatric surgical unit at Sulaimani Teaching Hospital. The study population consisted of 194 patients that presented as failure to pass meconium, constipation, intestinal obstruction, or already diagnosed and referred as case of ARM.

Thorough perineal inspection done for each, this provided the most important clues about the type of the malformation, then the patients classified according to the old Wingspread classification and Krickenbeck International classification.

Rectoperineal fistula, obviating the need for further diagnostic method; we proceeded to single stage anorectoplasty.

For those babies soon after birth, without any clinical evidence, we waited the first 16-24 hours before doing radiological evaluation & making a decision about colostomy or primary surgery. The reason for this delay is that significant intra-luminal pressure is required for meconium to be forced through a fistula orifice & to reach the lower rectum.

While passing meconium with the urine in male patients means presence of recto urinary fistula (either rectovesical or rectourethral), again we didn't perform further radiological investigation for these type(rather than the important investigations to exclude associated critical anomalies), but proceeded to protective divided descending colostomy, through which later on before the final repair, we did a high-pressure distal colostogram (using low osmolar iodine containing contrast under fluoroscopy) to find the site of the fistula and to distinguish between those two types.

After 24 hours of life, and without any clinical evidence, we still depend on taking a cross-table lateral x-ray film in prone position with a coin on in the perineum, on the presumed site of the anus as we haven't yet enough experienced radiologists in the study's center to arrange for perineal ultrasound and difficulties in taking urgent MR.

If the distance between the gas in the rectum and the coin measured less than 1 cm, we proceeded to single stage anorectoplasty. If the distance was more than 1 cm, protective divided descending colostomy was opened, later on the level of fistula distinguished by the distal colostogram.

Meticulous search done for associated anomalies, especially congenital urological anomalies, abdominal ultrasound done for all of them. Echocardiography was done preoperatively to search for cardiac malformations in most of the patients. Plain radiographs of the lumbar spine and sacrum were taken to evaluate for hemivertebra and sacral anomalies, while distal colostogram, intra-venous urography and voiding cystourethrogram were done accordingly.

Data regarding the age, sex, type of the malformations and the associated congenital anomalies and post-operative complications were records prospectively. The relation between the presence of complex or life threatening congenital anomalies and the mortality was analyzed by chi-square test and the prevalence rate of this congenital anomalies per 10 000 live birth in Sulaimani was measured for.

RESULTS

During the study period of 10 years, 194 children were found to have ARM, 122 were males and 72 were females (Fig.1).

The majorities (179 patients) presented at the first week of life; 9 cases presented during infancy, and just 6 cases presented at childhood, and one at adulthood (24 years).

Mode of presentation was; (1) absence of an anal opening, (2) failure to pass mecoium or constipation, (3) signs of lower intestinal obstruction, (4) an abnormal anal opening (e.g. anal stenosis, anterior displaced anus, perineal or vestibular fistula), as shown in the figures 2 and 3. According to the old Wingspread classification 134 cases were intermediate type, were 21 high and 39 Patients with low type IA.

The cases were also classified into a more therapeutic-oriented classification (Krickenbeck International classification), as shown in Table 1 and 2.

Meticulous search done for associated anomalies, they had been recorded in 44.2 % (Fig.4), some of them had more than one associated anomaly; the organs which involved are shown in the table 3.

Postoperative complication were recorded and summarized in table 4, some had more than one complication. Most of those cases operated upon by one stage simple anoplasty were complained from different degree of constipation and responded very well to anal dilatation schedule.

Cases of vestibular anus developed mild to severe constipation and managed conservatively, and iatrogenic rectovaginal fistula was recorded in one of them.

Few cases of cloaca and IA with rectovesical fistula, are complaining from fecal incontinence ranging between grade 2 and grade 3 (soiling with or without causing social problems), it was difficult to get the exact percentage because of a short follow-up for the new patients, losing follow up for the most older patients, and secondly, toilet training was not complete in those below 4 years of age.

Revision of anorectoplasty was done for a case of an IA with rectovesical fistula, because of the partial retraction of rectal mucosa and severe anal stricture. Recurrent rectovesical fistula in one patient and rectourethral fistula in another one, revision was done. Three cases with adhesive intestinal obstruction had been operated for adhesiolysis, while treated conservatively in others.

Mortality recorded in 16 patients (8.24 %); delay diagnosis and referral, was the cause of death in a male patient with perineal fistula, and one of the females with vestibular fistula, they presented with intestinal obstruction and sepsis, both of them died within hours post operatively.

Sudden, unexplained deaths were recorded in three male patients with IA and rectovesical fistula, and one female patient with vestibular fistula, post colostomy opening operations. Associated anomalies were the cause of death either pre or postoperatively in the remainder (Fig.5). The relation between the presence of complex or life threatening congenital anomalies and the mortality was analyzed and it was significant (p= 0,008), as shown in Table 5.

The chi-square statistic was (8.3828). The *p*-value is 003788; this result is significant at $p < .05$. The prevalence rate during the last 10 year was 6.4 to 10,000 live births.

Table 1. Pathological types of ARM in the male.

Pathological type	Frequency	Percent
IA with recto perineal fistula	27	22.1
Anal stenosis	5	4
Rectal atresia	2	1.6
IA with complex defect	3	2.4
IA with Recto urethral fistula	68	55.7
IA with rectovesical fistula	13	10.6
IA without fistula	4	3.27
Total	**122**	**100**

Table 2. Pathological types of ARM in the female.

Pathological type	Frequency	Percent
IA with recto perineal fistula	4	5.5
anal stenosis	2	2.7
Anterior displaced anus	1	1.3
rectal atresia	1	1.3
IA with complex defect	1	1.3
Vestibular anus	59	81.9
Cloaca	4	5.5
Total	**72**	**100**

Table 3. Organs / systems involved and malformations found with the various types of ARM.

Associated anomalies	Frequencies	Percent
Unilateral Renal agenesis	17	8.7
Hydronephrosis	10	5.1
Renal stones	10	5.1
Rotation anomaly (kidney)	7	3.6
Vesicoureteric Reflux	10	5.1
Post Urethral valve	6	3
Hypospadius	10	5.1
Tetralogy of Fallot	1	0.5
VSD	21	10.8
ASD	53	27.3
Pure esophageal Atresia	1	0.5
VACTERL	5	2.5
Biliary atresia	2	1
Hirshsprungs disease	2	1
OEIS	3	1.5
Agenesis of internal female genitalia	2	1
Down syndrome	8	4.5
Prune belly syndrome	1	0.5
Cleft lip and palate	1	0.5
Tongue tie	3	1.5
Polydactyl	3	1.5
Total	**176**	**100**

Table 4. Post operative complications.

Post operative complications	Frequencies	Percent
Stoma complication	61	31.44
Wound infection	17	8.7
Anal stricture	7	3.6
Constipation	34	1.7
Incontinence	3	1.5
Sepsis	4	2
Retraction of rectum	2	1
Adhesive intestinal obstruction	5	2.5
Renal failure	3	1.5
Recurrent fistula	2	1

Table 5. Relation between mortality and associated anomalies.

	Mortality No.(%)	Mortality No.(%)
Patients with no anomalies **P value**	140 [0.19]	6 [2.36]
Patients associated with anomalies **P value**	54 [0.44]	10 [5.38]
Marginal Column totals	194	16

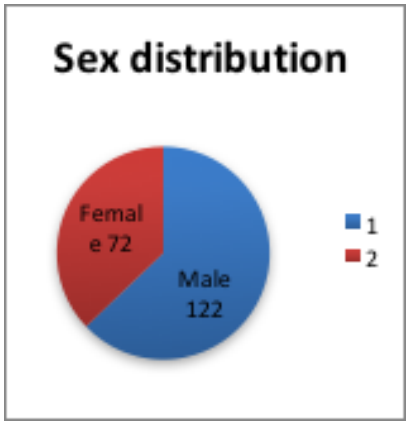

Figure 1. Shows sex incidence.

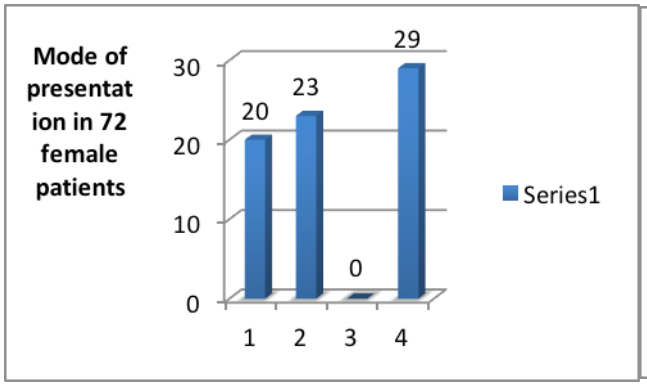

Figure 2. Mode of presentation in females.

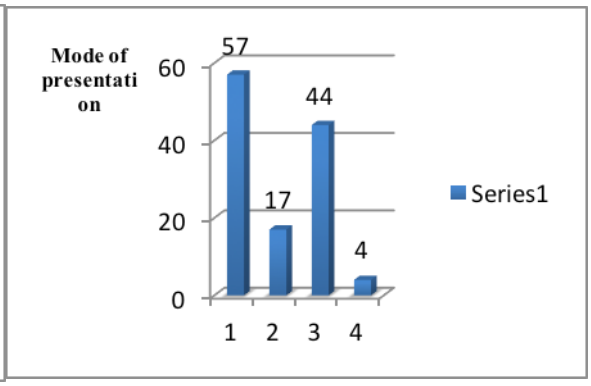

Figure 3. Mode of presentation in male.

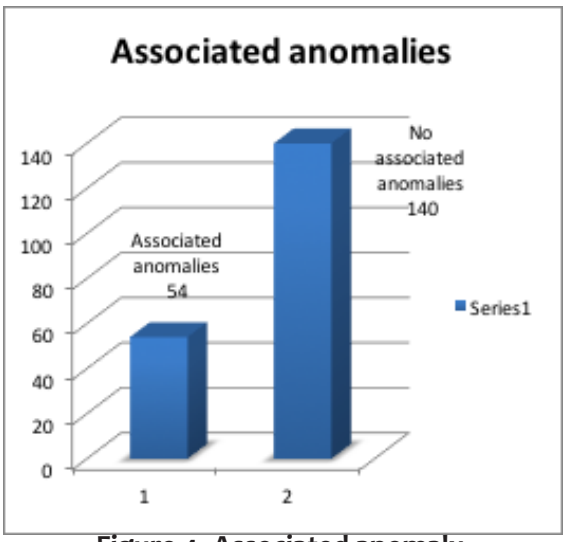

Figure 4. Associated anomaly.

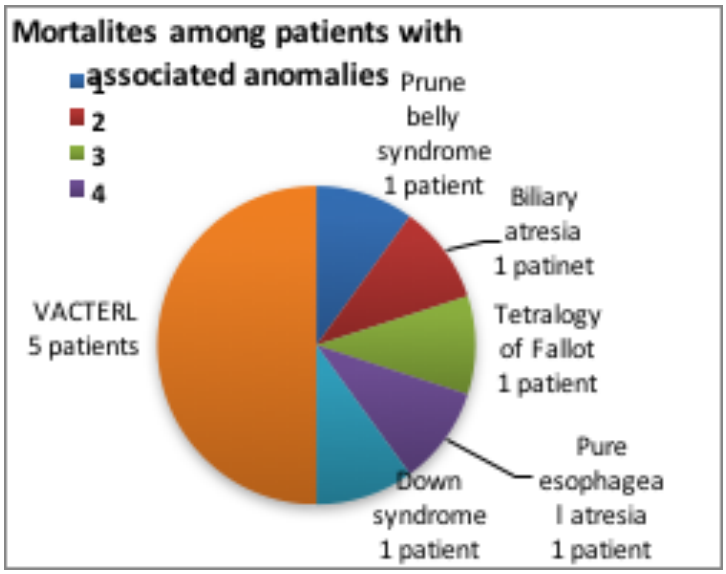

Figure 5. Relation between Associated Anomalies and Mortality.

DISCUSSION

ARMs are birth defects [1, 12]. They are somewhat more common in males than females [1, 12, 13, 14], as in this study, ARM were more common in males.

The majority of the cases (179 out of 194) were referred to the pediatric surgical unit on the first week of life. The causes of the delay presentation were improper diagnosis from the physicians' side and negligence and ignorance from the families because of socio-economic reasons as they could not afford the expenses of visiting our center from the rural areas, including cases with rectovestibular fistula, anal stenosis or anterior displaced anus, all of them were complaining from constipation and had been operated except one (16 year old) escape from the hospital.

Delay presentations for the same pathological types and for the same reasons were recorded in other theses [15].

The commonest presenting sign were absence of the anus in male patients; this was also reported in other studies [16]. The incidence of each type according to the old classification of ARM varies in the literature [13]. In our study, the intermediate type was the most frequent variety, IA with rectovesical fistula and rectourethral fistulas being the commonest abnormalities in the male cases , This result is in concordant with a study done in India in 2006 [17], While vestibular fistula was the commonest abnormality in the female cases , like other literature [1, 2, 17].

The incidence of the associated anomalies with ARM range from 30-70% according to the various studies [1, 2, 13], basically depends upon the meticulousness with which they have been searched for [18.19]. In this study the incidence is 44.2%, and genitor-urinary anomalies being the most common associated anomaly, same as in the other literature [1, 8, 13, 19].

The associated congenital anomalies in the newborns with ARM have a significant impact on the outcome of the management as the survival and prognosis depend upon the number and severity of the associated anomalies [19, 20]. In our study 10 patients among 16 mortality cases were associated with complex congenital anomalies.

Some anomalies like those of the spine, although not lethal, may have a direct bearing on the ultimate functional outcome of the case [19]. Other anomalies involving the cardiac, gastrointestinal and genitor-urinary system may lead to morbidity and mortality during the initial management of the cases [20], these also were obvious during long term follow up of those 194 patients.

Regarding post operative complication, although stoma complications were the most frequent type of complication but constipation was found to be the most common late complication, as it was reported in other studies [21].

Mortality rate was 8.24%, exactly same result of other study done at New Delhi [10], and all of them were neonate. The prevalence rate was higher than other studies done from Europe and Africa, but approximately approach to the rate at Saudi Arabia [4, 13,22].

Conclusion

The associated malformations rate and types are almost the same as in the literatures; however the prevalence rate was higher. This confirms the presence of geographic variation in ARM incidence, but could not confirm that the chemical attack or environmental impact of wars in this region had effects on increasing the rate of ARM due to lack of previous studies.

We recommend careful examination of neonate after delivery including genitopeirneal region, because still there are some cases diagnosed by the patient's families after discharging from Hospital, also we recommend further studies using advance radiological investigation and studying the gene and non genetic factors in the etiology of ARM.

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