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Pattern of Anorectal malformations in Gzira National Center for Pediatric Surgery (GNCPS)

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Abstract:

Background: Anorectal malformations (ARM) are one of the common clinically and socially demanding problems in the world especially in the poorly resourced countries.

Patients and Methods: It is a cross-sectional descriptive hospital based study, in the period from June 2009 to December 2011 of 80 patients of ARM in Gezira National Center for Pediatric Surgery (GNCPS). History and examination in addition to invertogram and distal loopogram were the tools of assessment.

Results: Eighty children with ARM were studied, 64 males and 16 females. Sixty six patients have delayed presentation. More than 61% presented from rural areas. Twenty-one (26%) patients had one or more associated congenital anomalies, with cardiac and limb anomalies being the most common associated malformation. High anomaly was found in 48.75% of patients, intermediate type was found in 5%, while 43.75% of patients had low anomaly.

Conclusion: Delayed presentation entails suboptimal training of midwives and resident doctors in obstetric hospitals and nurseries to evaluate congenital defects in the newborn.

ملخص:

خلفية: التشوهات الشرجية هي واحدة من المشاكل الولادية الشائعة والمكلفة سريريا وإجتماعيا لاسيما في البلدان الفيرة.

المرضى والطرق: هذه دراسة مستعرضة وصفية قائمة على المستشفى، في الفترة من يونيو 2009 الى ديسمبر 2011. إحتوت على 80 مريضا من المصابين الذين حضروا إلى المركز القومي لجراحة الأطفال (GNCPS). كان الإعتماد على التاريخ المرضي والكشف السريري والتصوير السيني .

النتائج: تمت دراسة ثمانين حالة من الأطفال المصابين بتشوهات شرجية ولادية. مثل الذكور 64 مريضا والإناث 16مريضة. لقد تأخر 6 6% من المرضى عن الحضور للمستشفى. أكثر من 61٪ قدموا من المناطق الريفية. كان واحد وعشرون (26٪) من المرضى يعانون من أحد التشوهات الخلقية المرتبطة بها، مع التشوهات القلبية وتشوهات الأطراف كونها الأكثر شيوعا. وقد عثر على الشذوذات الع الية في 48,75٪ من المرضى، والوسيطة في 5٪، في حين 43,75٪ من المرضى كانت لديهم شذوذات منخفضة.

الخلاصة: تأخر الحضور للمستشفى يظهر الحوجة لتدريب القابلات والأطباء المقيمين في مستشفيات الولادة ودور الحضانة لتقييم العيوب الخلقية في الأطفال حديثي الولادة.

Key words Anorectal malformations. Delayed presentation. Associated congenital anomalies.

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Introduction:

Anorectal malformations (ARM) continue to present a therapeutic challenge to the paediatric sur geon. This is even more so in developing countries like Sudan where there is a lack of facilities a nd personnel experienced in dealing with these anomalies. ARM occurs as one in 5000 [(1)].

An absent or abnormal anus frequently is obvious enough to allow delivery room detection. How ever, in developing countries delayed presentation is reported to be a real phenomenon that has a negative impact on the outcome of these anomalies [(2)].

This is a review of the experience with anorectal malformations in (GNCPS) of Wad Madani, Su dan.

Patients and Methods:

The data of 80 patients presented with ARM to Gzira National Centr for Pediatric Surgery (GNC PS), between June 2009 and December 2011 was collected and analyzed.

The workup of these patients includes history taking, proper clinical examination and imaging in form of invertogram done for 53 patients and distal loopogram done for 14 children.

Results:

Eighty children with ARM were studied, 64 boys (80%) and 16 girls (20%). Neonates constituted the majority of patients (74%). Forty percent of the neonate patients presented after 2 days. The i nfants, toddlers and preschool aged children represented 12%, 6% and 8% respectively.

Sixty eight percent presented to the center with failure to pass meconium without other symptom s, 12 patients (15%) presented with features of intestinal obstruction. The remaining 14 patients presented with abnormal passage of meconium. The birth weight of these patients ranges from 1.7 to 5.4 with a mean of 3.18+/-0.5 kg. On examination 36 patients (45%) were found to ha ve flat perineum, while in 24 patients (31.25%), signs of low anomaly like round buttocks, anal dimple and bucket handle were obvious. Leaking fistula had been found in 18 patients (22.5%). O ne patient has an ectopic anus.

In this study 21 (26%) of the patients have associated congenital malformations, while 59 (74%) of them proved to be an isolated anomaly. Table (1) shows the frequency of each congenital d efect.

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Table (1) shows the frequencies and percentages associated anomalies in 80 patients of AR M seen at GNCPS:

Association	Frequency	Percent
No anomalies	59	73.75
Vertebral anomaly	1	1.25
Tracheoesophageal anomalies	1	1.25
Cardiac anomalies	4	5
More than 1 of VACTERL	4	5
limb anomaly	4	5
Down's syndrome	2	2.5
OEIS (Lower midline syndrome)	2	2.5
Others	3	3.75
Total	80	100

On the base of these clinical findings and imaging results (invertogram and distal loopogram) the diagnosis was found to be High anomaly in 48.75% of patients, intermediate type in 5% and low anomaly in 43.75% of patients.

Discussion

In this study 59 patients (74%) were neonates, 10 (12%) were infants, 5 (6%) were toddlers an d 6 patients (8%) were pre-school aged children at time of presentation. 40% of the neonate pati ents presented after 2 days. Internationally delayed presentation is defined in term of days, as an y presentation after 2 days is considered as delayed presentation [3]. According to this definition 6 6% fulfilled the criteria of delayed presentation as they presented from 3 days to preschool age This is explained by the lack of routine neonatal examination and that some of the deliveries occ urs outside the hospital setting by midwives who are reluctant to assess neonates for anomalie s.

This study showed a male to female ratio of 4:1. This ratio is higher than the highest ratio found in the literature review. The sex ratio varies from 55% to 70% in favour of boys [4, 5]. In Ni geria it is 55.8% in favour of boys also [1]. We could not find a reasonable explanation for this ma le predominance. Further study is required to elucidate this phenomenon.

The study showed that more than 75% were from rural areas while the remaining were fro m urban sites. It is well known that most of people in Sudan reside in rural areas. Being the only s pecialized center of pediatric surgery outside Khartoum, GNCPS gives the service to patients c oming from other states (Gadarif, kassala, Portsudan, Sinnar and South of Blue Nile). These patie nts represented 33.75% of the study patients.

Sixty eight of the patients were observed by parents, midwives or doctors to have an abse nt anus after a variable period of the newborn failing to pass meconium . In this group no other s

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ymptoms were reported. Fifteen percent presented with features of intestinal obstruction. Six perc ent female patients passed meconium from the vagina, one male passed meconium with urine, an d 9% have perineal passage of meconium while one male baby passed meconium from an openin g just near the scrotum. The percentage summation of children who presented with abn ormal passage of meconium was 17%. O. Adejuyigbe found that 61% presented with failure or d elayed passage of meconium while 86% presented with abdominal distension or vomiting [(1)].

Seventeen percent of patients presented with symptoms of leaking fistula, while on exami nation a leaking fistula was an objective finding in 22.5% of patients. This reflects the difficulty o f detecting the fistula which sometime looks like a pin point and entails careful examination unde r good light and magnification. Some patients required an examination under anaesthesia to clarif y the anomalous perineal anatomy.

Other associated congenital malformations were discovered in 26% of the studied patients . O.Adejuyigbe found associated malformations in 27.9% of the 86 cases study in Nigeria [(1)]. Our result and this study contrast with 149 (60.6%) out of 246 patients by Stephens and Smith [(6)]. Bot h Sudan and Nigeria are poor resourced countries and the medical system lacks advanced diagn ostic prenatal and postnatal tools. This leads to under diagnosing minor associated anomalies and account for the low figures compared to those from western countries.

By referring to Table (1) we find that associated limb defects were 5%. Cardiac anomalies were encountered in 5%, Down's syndrome in 2.5%, tracheoesophageal anomalies in 1.25%, Low er midline syndrome 2.5%, more than 1 of VACTERL in 5%. Other anomalies like cleft lip and p alate and hypospadius were found in 3.75%.

Mirza B of The Institute of Child Health, Lahore stated that the common associated anom alies were urogenital (10%), cardiovascular (8%), and gastrointestinal (6%). Down's syndrome w as present in 8 (8%) patients. OEIS was present in 1%. Craniofacial in 4% and hypospadius in 1% [(7)]. It appears that the cardiac, Down's syndrome and tracheoesophageal anomalies were less freq uent in our patients than in Mirza's. Limb anomalies and lower midline syndrome are were frequ ent in our study. The reported incidence of OEIS complex is 1 of 200,000 to 400,000 live births. However the literature is flooded with other rare associated malformationInvertogram was done i n 50 patients in whom high or intermediate type is suspected and in those with equivocal perineal findings of low type. Distal loopgram was done in 14 patients in whom colostomy was done.

The diagnosis in this study was based on clinical features and radiological findings. High type without fistula is the most frequent diagnosis and represented 40% of the patients. Low type without fistula represented 25%, low type with perineal fistula represented 18.75%, intermediate t ype with rectourethral fistula represented 1.25%, intermediate type with rectovestibuler fistula re presented 3.75%, high with rectovesical Fistula 5%, high with rectovaginal fistula 3.75%. Ectopi c anus represented 1.25% and low imperforate anus with Hirschsprung's disease was found in 1.2 5%.

High anomaly was found in 48.75% of patients, intermediate type was found in 5% while 43.75% of patients have low anomaly. Internationally the high type frequencies are 20 to 18%, in termediate type is 11 to 54% and low type represents 18 to 57% [(8, 9, 10, and 11)]. The intermediate ty pe has the lowest frequency in the Japanese study [(11)] as it represents 11%. But the intermediate ty

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pe in our study is less frequent. Stephens FD series [12] showed a similar high type frequency to o ur result (46% and 49% respectively).

Conclusion: Anorectal malformations are apparent health problems in GNCPS. Delayed presentation is onr of feature, the thing that might dictate the need for more trainging among mid wives/

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