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Original Article

Anomalies Associated with Ano Rectal Malformation: Experience in Dhaka Shishu (Children) Hospital

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Abstract:

Anorectal Malformation is one of the most common congenital anomalies. It is associated with others anomalies. But those others anomalies are sometime neglected. The purpose of this study is to review the incidence of other congenital anomalies associated with anorectal malformation (ARM). This prospective study was carried out in the department of Pediatric Surgery, Dhaka Shishu (Children) Hospital during the period of June, 2013 to July, 2014. A number of 45 cases were included and data were collected on the type of ARM and associated congenital anomalies which were categorized according to organ system. Total study population was 45. Male: Female ratio was 1.37:1. Majority (55.55%) presented with ARM without fistula, followed by recto vestibular fistula 26.67%, perineal fistula 13.33%, and 2 patients had cloacal anomaly. Thirty two (71.1%) patients had associated congenital anomaly. Among them, 42%, urogenital anomalies, 37.78% cardiovascular, 20% craniofacial anomalies, 11.11% musculoskeletal and anomalies of central nervous system 11%. Among 32 patients, 57.14% patients had multiple congenital anomalies. It is imperative that a thorough clinical evaluation and systemic investigations of all patients with anorectal malformation necessary to exclude or confirm the presence of genitourinary, cardiac and other abnormalities which are directly related with mortality and morbidity.

Key words: Anomalies , Anorectal Malformation.

Introduction:

Anorectal malformations (ARMs) are one of the most frequently presenting congenital anomalies in pediatric surgical practice[1]. They include a series of defects ranging from a slight malformation to more complex anomalies of the hindgut and urogenital organs[2]. An anorectal malformation occurs in one of every 4000 to 5000 newborns and is slightly more common in males[3]. Management of ARM in the neonatal period is essential and it focuses on the accurate classification of anorectal malformation and the best way to restore the normal intestinal anatomy and function. However surgeons must be aware of the high incidence of associated congenital anomalies in case of ARM[4]. About 20-80% patient of anorectal malformation also present one or more congenital anomalies including gastrointestinal, cardiovascular, genitourinary, central nervous system and musculoskeletal system[5,6]. These associated congenital anomalies have a very important role in the surgical management of ARM patients.

As the classification of ARM has been revised several times over the years, problems in comparing the result of different studies have been arisen. In previous studies association of congenital anomalies were shown on the basis of Pena classification or Wingspread classification (Low, Intermediate, and high)[7,8]. In May 2005 a new international classification called Krickenbeck classification was introduced which incorporates the anatomic description, of the malformation, type of surgical procedure performed and post operative assesment of bowel movement and soiling[1,8].

In our country so far our knowledge no studies has been carried out to evaluate the associated anomalies in

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ARM patients, although several studies has been done in different places of the world[2,5-10]. In this point of view we want to evaluate the incidence of other congenital anomalies in relation to the type of ARM as defined by the Krickenbeck classification.

Materials and Methods:

This was a prospective study held in department of pediatric surgery, Dhaka Shishu hospital a tertiary level paediatric hospital in Dhaka, Bangladesh. The study period was June, 2013 to July, 2014 and still continuing. Data regarding types of ARM and associated anomalies were collected and classified according to Krickenbeck criteria. Anomalies were categorized as Cardiovascular, Gastrointestinal, Urogenital, Neurological, Craniofacial, Musculo skeletal, Respiratory system.

All the patients were clinically examined for the diagnosis of the type of ARM, along with others congenital anomalies. To diagnose the patients with stoma we took help from previous medical records. Neonates with ARM who presented to us; an X-ray prone cross table lateral view with raised pelvis was performed to see the level of distal gas shadow.

An ultrasound of the abdomen was performed in all cases to identify the urogenital, gastrointestinal anomalies. Patients who had positive findings in ultrasound regarding urogenital system further evaluation were done by micturating cystourethrogram and or intravenous urogram.

Ultrasonogram of brain and spinal cord was performed in patients whose fontanelle was not closed in order to identify the central nervous system anomaly and neural tube defect. X-ray spine for elder child and a baby gram was performed for all neonates to confirm the vertebral anomalies.

If there was a positive finding on auscultation of the precordium, an echocardiogram was performed. Other investigations like ECG, X-ray Chest, MRI scanning of Spine/brain & CT scanning were performed in selective cases.

Result:

In our study we included all ARM patients even with underwent colostomy admitted in our department. All patients were pediatric age group. Total study population was 45 [Figure-1].

Majority of the patients presented with ARM without fistula. Number was 25 out of 45 patients (55.55%) followed by recto vestibular fistula 12 patients (26.67%), perianal fistula 6 patients (13.33%), 2 patients had cloacal anomaly that is (4.44%).

In our series 32 (71.1%) out of 45 patients had associated congenital anomaly rest 13 patients (28.88%) had no other congenital anomalies. The major groups of system involve urogenital anomalies (42%), cardiovascular (37.8%), and craniofacial anomalies (20%). Other system involves are Musculoskeletal (11.2%) and anomalies of central nervous system (11%). [Figure-II]

Out of 32 patients presenting with congenital anomalies 18 (57.14%) patients had multiple congenital anomalies affecting multiple system or different anomalies in the same system.

Among urogenital anomalies 7 patients had multiple and 10 patients had single disorder. Most common urogenital anomalies in our series were hypospadias, hydronephrosis and pelviureteric junction obstruction.

Regarding cardiovascular anomalies atrial septal defect was most commonly associated anomaly. Ventricular septal defect, patent ductus arteriosus, Tricuspid regurgitation of different grades and pulmonary artery hypertension is next to associated with ARM.

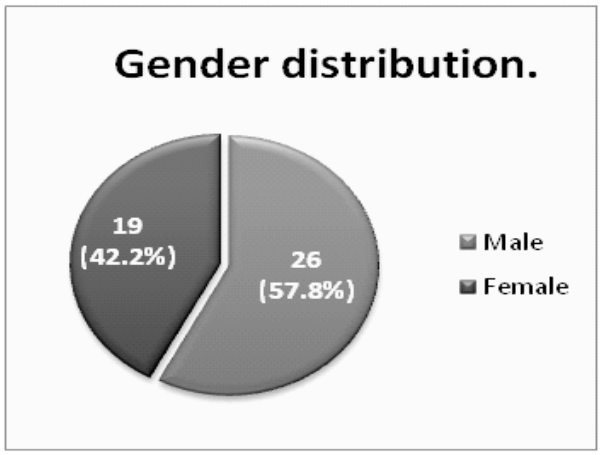

Figure-I: Gender Distribution

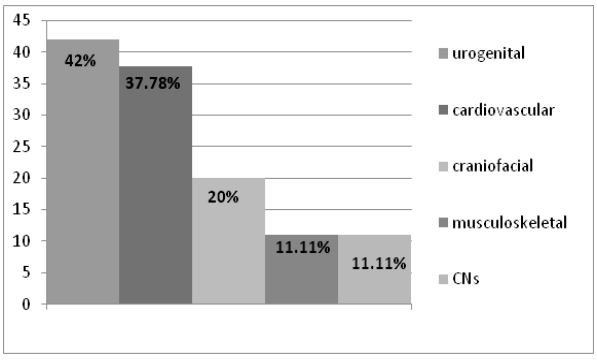

Figure-II: Congenital Anomalies Associated with ARMs

Discussion:

The aim of our study is to demonstrate the incidence of other congenital anomalies with ARM patients presented to us, by associating these anomalies to the anatomic type of malformation as described by Krikenbeck classification.

ARMs represent a frequently occurring pathology in pediatric surgical practice. However mortality and morbidity mostly depends on the wide spectrum of associated congenital anomalies rather than ARM itself.

The reported range of other congenital anomalies varies between 28-70%. In our series 71% patients had other congenital anomalies which are comparable with other published study. High anomalies are commoner in the male in our series as in other studies[11]. With very few exception most of the previous studies had been carried out on the basis of either Wingspread or Penna classification. But we did our study on the basis of Krickenbeck classification which allows us to analize the association of congenital anomalies more precisely between groups.

The most common anomalies associated with ARM reported around the world were urogenital anomalies. About 50% of ARM patient suffers form urogenital anomalies[9]. In present study 19 patients has urogenital system anomaly which is about 42%. Most common urogenital anomalies encountered in our study were hypospadias and hydronephrosis. Renal aplasia or dysplasia also noticed in our study. We found 7 patients with hypospadias. Among them 4 were coronal, 2 patients subcoronal and 1 patient with mid penile hypospadias. Six patients had hydronephrosis due to PUJ obstruction, 3 patient had meatal stenosis and 1 patient had right sided undescended testis.

In our series 17 out of 45 patients had cardiac anomalies. Among this 17 patient 12 had multiple congenital anomalies where as 5 patients had single cardiac anomaly. Nine patients among 17 patients who had fistula had cardiac anomaly which proves the higher association of cardiac anomalies with ARM with fistula. Most frequent cardiac anomaly was atrial septal defect then persistant ductus arteriosus, ventricular septal defect and tricuspid regurgitation of different grades.

Though other studies show very low associated rate of craniofacial anomalies[10] our results shows that craniofacial anomalies are 3rd most commonly associated congenital anomalies with ARM. In our study 9 patients had craniofacial anomaly. Among them patients had agenesis external ear; all had defect on right side. Three patients had both cleft lip and palates of different variety and 1 patient had only cleft lip. Another one patient presented with external angular dermoid on lateral side of right eye brow. Though other studies from different countries showed a large number of gastrointestinal anomalies as they include Fistulas to Gastrointestinal system but we didn't include fistulas in gastrointestinal system. In our series we found 4 patients with gastrointestinal anomalies, 3 of them had mescenteric cyst and 1 had omental cyst.

Regarding central nervous system, we found five patients with central nervous system anomalies. Among them three patients had spina bifida occulta and one patient with a small meningocele.

In our series five patients had TEV which was included in musculoskeletal system. 2 had right, 2 had left and 1 patient had bilateral TEV.

On summery of our study we have concluded that 71% of the newborns with anorectal malformations had associated anomalies, which lies close to the reported range. We found urogenital anomalies to be the most common associated congenital defects in our patients; the groups presenting rectovestibular fistula were most likely to present cardiac abnormalities. The incidence of craniofacial anomalies was equal in the group with fistula and without fistula.

Conclusion:

Regardless of the type of anorectal malformation, it is imperative that a thorough clinical examination and systemic investigations of all patients to be done in order to exclude or confirm the presence of, genitourinary, cardiac or any other abnormalities which can increase mortality and morbidity in the management of ARMs.

References :

1. Holschneider AM, Hutson JM. Anorectal Malformations in Children. Berlin: Springer; 2006.
2. Rintala Risto J. Congenital Anorectal Malformations: Anything New? Journal of Pediatric Gastroenterology and Nutrition 2009; 48(2):S79-S80.
3. Levitt MA, Peña A. Imperforate Anus and Cloacal Malformations. In: Holcomb GW, Murphy JP, eds. Ashcraft's Pediatric Surgery. 5th Edition. Philadelphia: Saunders Elsevier; 2010. p. 468-490.
4. Levitt MA, Peña A. Anorectal Malformations. In: Mattei P, editor. Fundamentals of Pediatric Surgery. NewYork:SpringerScience; 2011. p. 499-511.
5. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Anorectal malformations in neonates. Afr J Paediatr Surg. 2011;8(2):151- 4. doi: 10.4103/0189-6725.86051.
6. Cho S, Moore S.P, Fangman T. One hundred three consecutive patients with anorectal malformations and their associated anomalies. Arch Pediatr Adolesc Med. 2001; 155(5):587-91.
7. Holschneider A, Hutson J, Peña A, Beket E, Chatterjee S, Coran A, et al. Preliminary report on the international conference for the development of standards for the treatment of anorectal malformations. J Pediatr Surg. 2005; 40(10):1521-6.
8. Hassett S, Snell S, Hughes-Thomas A, Holmes K. 10 year outcomeof children born with anorectal malformation, treated by posterior sagittal anorectoplaty, assessed according to the Krickenbeck classification. J Pediatr Surg. 2009; 44(2):399-403. doi: 10.1016/j.jpedsurg. 2008. 10.092.6t5
9. Goossens WJ, de Blaauw I, Wijnen MH, de Gier RP, Kortmann B, Feitz WF. Urological anomalies in anorectal malformations in the Netherlands: effects of screening all patients on long-term outcome. Pediatr Surg Int. 2011; 27(10): 1091-7. doi: 10.1007/s00383-011-2959-4.
10. Bălănescu RN, Topor L, Mog A. Anomalies Associated with Anorectal Malformations. Chirurgia (2013) 108: 38-42 No. 1, January - February
11. Islam MK. Pattern and aetiology of Anorectal anomaly, Bangladesh Institute of Child Health. 1992 Thesis.