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Original Research Article

Urogenital anomalies associated with anorectal malformations

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ABSTRACT

Background: Anorectal malformations are one of the common congenital anomalies with an incidence of 1-3000 to 5000. Up to 70% of the patients have associated anomalies. Urogenital anomalies are the most common associated anomalies.

Aim: This study was done to determine the type and frequency of urogenital anomalies associated with different variants of ARMs according to the Krickenbeck classification.

Materials and Methods: 150 patients were included in the study. A full physical examination of the child was conducted followed by an infantogram, echocardiogram, spinal ultrasound scan, and KUB ultrasound was done to investigate different associated anomalies. MRI was done if further clarity was needed. The patients were classified according to the Krickenbeck classification.

Result: ARM with perineal fistula was the common type of ARM. 73% had associated anomalies, and 32% of patients had urogenital (UG) anomalies. 70.8% of patients had urinary anomalies. 52.1% of patients had genital anomalies. The most common type of urinary anomaly found in the present study was hydrourteronephrosis (HUN) (41.2%). 32.4% of the patients with urinary anomalies had vesicoureteral reflux (VUR). Renal agenesis was found in 32.4%, neurogenic bladder 5.9%, hydronephrotic kidney 5.9%, ectopic ureter 5.9%, cross fused ectopic kidney 5.9%.

Conclusion: Urogenital anomalies associated with different subtypes of anorectal malformation vary drastically from patient to patient. The knowledge of these associated anomalies will help the surgeons in planning the course of treatment which can determine the prognosis and quality of life of the patients.

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1. Introduction

Anorectal malformations (ARM) are one of the frequent congenital anomalies encountered in pediatric surgery with an estimated incidence ranging between 1 in 2000 to 1 in 5000 live births.¹ Congenital anomalies of the urogenital tract are the leading associated anomalies in patients with ARM. Previous studies show an incidence of 20-30%.² Most of the genital anomalies can be identified by routine clinical examination, and ultrasound sonography test (USG) of kidney, ureter, and bladder (KUB) is done to determine

urological anomalies. In many cases management of ARM is given priority and urological evaluation is not given due importance even though urinary tract disorders may be the primary cause of morbidity and mortality.³ The purpose of the present study was to estimate the type and frequency of urogenital anomalies among different variants of ARM.

2. Materials and Methods

This study was conducted in the Department of Paediatric Surgery, Bangalore Medical College and Research Institute from Jan 2014 to Dec 2016. All children with ARM treated and followed up in the department were included in the study. Written informed consent was obtained from the

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parents of the patients. The study was cleared by the institutional ethical committee.

A full physical examination of the child was conducted for evaluation of ARM and a detailed radiological investigation was carried out to confirm the type and extent of ARM. Ultrasound KUB was done to determine urogenital anomalies. Micturating cystourethrogram was also done when necessary.

3. Results (Tables 1, 2 and 3)

150 patients were included in the present study. The ages of the patients varied from one day to 13 years. These patients were admitted to the Paediatric ward for different stages of surgery. Out of 150 patients, 101 (67.3%) were males and 49 (32.7%) were females. 110 (73.3%) had one or more associated anomalies. 40 (26.6%) patients had isolated ARM.

48 (32%) patients had urogenital (UG) anomalies. 34 (70.8%) patients had urinary anomalies of which 24 were males and 10 were females. 25 (52.1%) patients had genital anomalies which included 17 males and 8 females. 11 (7 males and 4 females) patients had both urinary and genital anomalies.

The most common type of urinary anomaly found in the present study was hydroureteronephrosis (HUN) (41.2%). 32.4% of the patients with urinary anomalies had vesicoureteral reflux (VUR). Renal agenesis was found in 32.4%, neurogenic bladder 5.9%, hydronephrotic kidney 5.9%, ectopic ureter 5.9%, cross fused ectopic kidney 5.9%.

Table 1: Urinary anomalies

Urinary Anomalies	Number	Percentage (%)
Hydroureteronephrosis	14	41.2
VUR	11	32.4
Left renal agenesis	6	17.6
Right renal agenesis	5	14.7
Cross-fused ectopic Kidney	2	5.9
Dilated tortuous ureter	2	5.9
Ectopic ureter	2	5.9
Hydronephrotic Kidney	2	5.9
Neurogenic bladder	2	5.9
Cyst at the Lower pole of the kidney	1	2.9
Cystitis	1	2.9
Pelvic Ectasia	1	2.9
Urinary bladder diverticulum	1	2.9

Hypospadias (16%) and non-palpable undescended testis (NPUdT) (16%) were the frequently encountered genital anomalies. Bifid uterus and ventral chordee were found in 12% respectively. Absent left ovary, bifid scrotum, hydrocele, penoscrotal hypospadias, and septate vagina was found in 8% each.

The most frequently encountered form of ARM was perineal fistula (20%), but only 3 patients with perineal fistula had urinary anomalies and only 2 had genital anomalies. Rectobulbar fistula was found in 18.7% of patients and no fistula in 18.7%. HUN was frequent along with rectobulbar fistula, while no fistula had VUR. Rectoprostatic fistula was found in 14% of patients and 14.7% had vestibular fistula.

Table 2: Genital anomalies

Genital Anomalies	Number	Percentage (%)
Hypospadias	4	16
NPUdT	4	16
Bifid Uterus	3	12
Ventral Chordee	3	12
Absent left ovary	2	8
Bifid Scrotum	2	8
Hydrocele	2	8
Penoscrotal hypospadias	2	8
Septate Vagina	2	8
Aphalia	1	4
Dorsal Hood	1	4
Epididymitis	1	4
Hematometrocolpos	1	4
Hemi scrotum	1	4
Ill developed scrotum	1	4
Micro penis	1	4
Penoscrotal transposition	1	4
Right ovarian cyst	1	4
Uterus didelphys	1	4
Vaginal duplication	1	4

4.7% of patients had cloaca. 85.7% of patients with cloaca had urinary anomalies and 57.1% had genital anomalies. Pouch colon was present in 4% of patients, 50% had urinary anomalies and 50% had genital anomalies. Rectovaginal fistula and anorectal agenesis did not have any urogenital anomalies.

4. Discussion

Anorectal malformation includes a wide spectrum of congenital malformations of anorectum frequently associated with other anomalies. ARM has an incidence ranging from 1 in 2000 to 1 in 5000. The present study has an incidence of 1 in 3000 similar to published literature.⁴ One or more anomalies are associated with ARM. 71.3% of the patients in the current study had one or more anomalies which is higher than that reported by Lowery et al⁵ (66.6%) and Balanescu et al.²

According to Bhargava et al,⁶ forty to seventy percent of ARM patients have one or more associated anomalies, and urogenital defects are the most common anomaly associated with ARM followed by defects of the spine, gastrointestinal, craniofacial, extremities, and cardiovascular system. Nah et al¹ reported that UG anomalies were present in 28% of

Table 3: Type of ARM: Frequency of urogenital anomalies

Type of ARM	Number	Urinary	%	Genital	%
Cloaca	7	6	85.7	4	57.1
No Fistula	28	3	10.7	4	14.3
Rectobulbar Fistula	28	8	28.6	6	21.4
Rectoprostatic Fistula	21	9	42.9	3	14.3
Vestibular Fistula	22	1	4.5	2	9.1
Pouch Colon	6	3	50.0	3	50.0
Rectal Atresia	4	0	0.0	1	25.0
Rectovaginal Fistula	2	0	0.0	0	0.0
Anorectal Agenesis	2	0	0.0	0	0.0
Perineal Fistula	30	3	10.0	2	6.7

patients with VUR being the most common type of UG anomaly. In the present study, 31.3% of the patients had UG anomaly with HUN (41.2%) being the most common UG anomaly followed by VUR (32.4%) and left renal agenesis (17.6%). Vander Brink et al⁷ reported that 85% of ARM patients who had epididymitis also had VUR, but in our study, the patients with epididymitis had right renal agenesis. Srivastava et al⁸ reported 5.45% had unilateral renal agenesis. Sanchez et al⁹ reported 22% of ARM patients had VUR which is lower than the findings in our study. Boemers et al¹⁰ reported 24% of patients in his study had neurogenic bladder which is way more than our findings of 8.7%. 30% (17% left and 13% right) had renal agenesis compared to 9% reported by Cho et al¹¹ and 17% by Blaauw et al.¹² Daradka¹³ in his study found that HUN and unilateral agenesis were common (39.4%).

Genital anomalies were found in 53.2% of the patients. The incidence of genital anomalies was found to be 36% and 56% by Sabzehia et al¹⁴ and Kella et al¹⁵ respectively. NPUDT (16%) and hypospadias (16%) were the most common type of genital anomaly in our study which is less than the findings of Mirshemirani et al¹⁶ of 54%. Hamadi HA et al¹⁷ reported the incidence of hypospadias in 21.9% of patients which is higher than our findings of 16%. Kumar et al¹⁸ in their study found NPUDT in 10% of patients and bifid scrotum in 8%. In the present study, bifid scrotum was found in 8% of patients.

5. Conclusion

Urogenital anomalies are the most common type of associated anomaly in patients with ARM. Most of the genital anomalies can be detected on physical examination but many other anomalies may not be evident which may be a major cause of morbidity or even mortality. Even though the present study shows that the incidence of urogenital associated anomaly decreases with the level of lesion a thorough clinical evaluation and systemic investigation of all ARM patients should be done to avoid future complications.

6. Sources of Funding

None.

7. Conflict of Interest

None.

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