

A retrospective and observational study of urological anomalies in anorectal malformation in children at a tertiary care center in Telangana



Srinivas Srirampur¹, Kavimozhy Ilakkiya Poyyamozy², Nagarjuna Kumbha³

¹Associate Professor, ²Senior Resident, ³Professor and Head, Department of Paediatric Surgery, Gandhi Medical College and Hospital, Secunderabad, Telangana, India

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ABSTRACT

Background: Anorectal malformations (ARM) are one of the common congenital abnormalities in a pediatric surgical unit. Urological anomalies occur frequently with ARM and the incidence increases with increasing complexity of ARM having dramatic impact on quality of life. **Aims and Objectives:** This study is done with an objective to know the incidence of various urological anomalies in ARM and comparing our observation with existing literature. **Materials and Methods:** This is a retrospective study conducted in Pediatric Surgical unit, Gandhi Medical College and Hospital, Secunderabad, from January 2017 to December 2021 based on the records of 45 patients who were diagnosed to have ARM with urological anomalies. Incidence of urological anomalies in ARM is studied with respect to demographic data and pattern of coexistence. **Results:** Total number of ARM cases noted during the period of study was 101, of which 45 cases had associated urological anomalies and these 45 cases were taken in this study. The overall incidence of urological anomalies was 42.5%. The incidence of urological anomalies was significantly swelled up in high ARM when compared with low lesion. Vesicoureteric reflux and hydronephrosis were the most common anomalies observed in our study. **Conclusion:** The high incidence of associated urological anomalies necessitates standardized protocol of investigating all ARM patients as it is an important cause of morbidity and mortality. Early diagnosis warrants timely intervention and prevents upper urinary tract deterioration.

Key words: Anorectal malformation; Associated urological anomalies; Micturating cystourethrography; Standardized protocol

INTRODUCTION

Anorectal malformation (ARM) is one of the common congenital abnormalities in pediatric population.¹ ARM is associated with multisystem abnormalities, of which urological anomalies are by far the most common association² with incidence of 20–50%.^{3,4}

This frequent association of urological anomalies and ARM can be better understood due to the common embryological development. Urological anomalies include both structural and functional abnormalities. Its proven to

have high morbidity and mortality if not addressed early.^{5,6} Vesico ureteric reflux (VUR) and hydronephrosis are the common abnormalities associated with ARM.^{1,7} Renal anomalies have been observed in 50–60% of patients with high or intermediate forms and 15–20% of low ARM.⁸ The incidence of urological anomaly swells up with complexity of ARM.

Aims and objectives

This study is done with an objective to know the incidence of various urological anomalies in ARM and comparing our observation with existing literature.

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Address for Correspondence:

Dr. Kavimozhy Ilakkiya Poyyamozy, Senior Resident, Department of Paediatric Surgery, Gandhi Medical College and Hospital, Secunderabad - 500 025, Telangana, India. **Mobile:** +91-8828499660. **E-mail:** kavimozhy86@gmail.com

MATERIALS AND METHODS

From January 2017 to December 2021, a total of 101 ARM cases were treated at Gandhi Hospital, Secunderabad, Telangana. Of which 45 cases of ARM had associated urological anomalies and these cases were analyzed retrospectively in our study. The study was undertaken after hospital ethical committee clearance. Case records were procured from medical record department to collect demographic data, clinical data, and relevant investigations. Additional data were taken from department archives. All patients had cross table prone lateral X-ray, invertogram, X-ray (spine and Limb), echocardiography, and ultrasound abdomen (Renal, abdominal, and Spine in neonates). All patients with ARM in our study had undergone micturating urethrography (MCU) on follow-up. ARM patients with VACTERL anomaly were also studied to find their association with respect to urological anomalies. MCU was performed on follow-up under antibiotic cover with strict aseptic precautions. MCU results graded the severity of VUR as per international reflux grading system. Pressure augmented distal colograms that was performed before definitive procedure were also studied as they revealed urological anomalies in few patients. Magnetic resonance urography (MRU) was performed in those children who were diagnosed to have complex urological abnormalities in ultrasonography (USG) KUB and MCU. Patients presented to pediatric surgery outpatient department late after seeking consultation from other specialties were included in the study. Moreover, referrals from elsewhere with definitive diagnosis for further management were also included. The subjects with urological anomalies include both structural and functional abnormalities of urinary tract.

The data collected were compiled, categorized, and tabulated. The incidence of urological anomalies associated with ARM was studied with respect to demographic design, the pattern of coexistence of various structural and functional urological anomalies, and compared with existing literatures. Patients with incomplete data and those lost to follow-up were excluded from the study.

Statistical analysis

Descriptive statistics were applied to the study.

RESULTS

A total of 101 cases of ARM were treated at Gandhi hospital, Secunderabad, during the study period. This study evaluated 45 cases of ARM with associated urological anomalies. Of the 45 cases, 23 (51%) were female and 22 (48.9%) were male with no significant sex preponderance. Age of presentation varied from day 1

to 9 years with mean age at the time of diagnosis being 2 years. Weight ranged from 1.2 kg to 25 kg with mean of 8 kg (Table 1). Follow-up period ranged from 14 months to 58 months. Based on the Wingspread classification, types of ARM were studied during the evaluation for associated urological abnormality. Nineteen (42%) children with high ARM had urological anomalies, of which 15 (33.4%) were low and 11 (24.4%) were intermediate, respectively (Figure 1). Overall the incidence of urological anomalies escalates with increasing level complexity of ARM.

Among ARM with VACTERL anomaly, urological abnormality was the second most common association contributing to 19 (43.3%) cases.

USG KUB was performed in all cases of ARM as a screening investigation for urological anomalies. Hydronephrosis was the most common urological anomaly diagnosed in 22 children and eight children had hydronephrosis. Structural abnormalities such as renal agenesis, cystic kidney disease, duplex kidney, and crossed-fused ectopic kidney were also noted. USG revealed renal agenesis in nine children with ARM.

MCU revealed cases of VUR, duplex systems, and posterior urethral valve (PUV). MCU helped in diagnosing low-grade reflux too (Figure 2a). PUV was diagnosed in one with high-grade reflux (Figure 2b). Two duplex systems cases

Table 1: Demographic data of ARM patients with urological anomalies

Demographics	Number of patients
Male	22 (48.9%)
Female	23 (51.1%)
Age	1 day–9 years
Weight	1.2–25 kg
Total	45

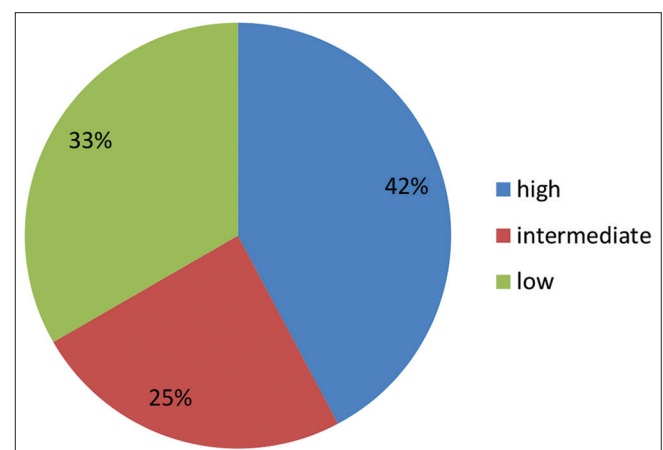


Figure 1: Depicts the type of anorectal malformations with associated urological anomalies in our study

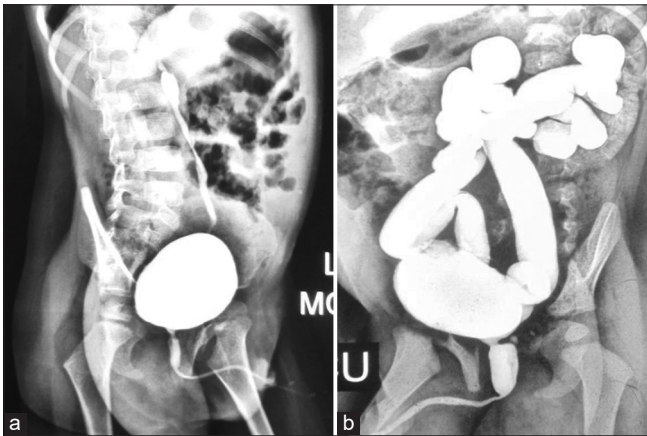


Figure 2: (a) Micturating urethrography depicting low-grade reflux and (b) high-grade reflux with posterior urethral valve

were detected with Grade 3 VUR and Grade V VUR. MRU confirmed one case with complete and other as partial duplex system (Figure 3).

DISCUSSION

Of all the anomalies associated with ARM, urological anomalies are the most frequent ones.² Overall, 40% of ARM patients have associated urinary tract anomaly.^{3,4} It has dramatic impact on the length and quality of life.² The importance of investigating and treating urinary tract anomalies in patients born with ARM has been under emphasized. The incidence of urological anomalies in ARM varies between 11 and 54% in various studies.³⁻⁸ In the present study, incidence of urological anomalies with ARM was 42.5% (Table 2).

Table 2 shows that studies^{9,11-13} have varying incidence of urological anomalies ranging from 34 to 49% due to inclusion of the genital abnormalities along with urological abnormalities. Our study included only urological abnormalities. Whereas Indian studies, Shenoy et al., and Mittal et al., reported higher incidence of urological anomaly with ARM.^{13,14} Srivastava et al.,¹⁰ did not include MCU for all their cases and the overall incidence was quite less (11.3%). In our study, MCU was a standard protocol of investigation for all ARM cases. No gender preponderance was noted in our study whereas Ratan et al., stated that boys were more prone to have associated urological abnormalities in ARM children especially with increasing complexity.

Age of children at the time of diagnosis ranged from day 1 to 9 years in the study. Certain referred cases presented tardy as urological anomaly diagnosis was made only after evaluation of symptoms and few failed to undergo screening investigation as a part of work up to rule

Table 2: Comparison of associated urological anomalies in various studies (UG: urogenital)

Studies	Incidence of urological anomalies in ARM (%)
Present study	42.5
Endo et al., ⁹ 1999	49.12 (UG)
Ratan et al., 2004	37.4
Sangkhathat et al., 2002	24.54
Srivastava et al., ¹⁰ 2005	11.3
Mirshemirani et al., ¹¹ 2008	48.6 (UG)
Islam et al., 2015	27.8
Sejdi et al., ¹² 2015	49.12 (UG)
Shenoy et al., ¹³ 2019	34.2 (UG)

out associated anomalies with ARM background. Early diagnosis helps in instituting prompt treatment in timely manner decreasing the morbidity thereby improving the quality of life to a greater extent. Hence, it is very important to have a standardized protocol for screening all ARM patients for associated anomalies universally. In our observation, incidence of associated anomalies (42.5%) escalated with increasing complexity of the ARM akin to various studies¹⁵⁻¹⁷ emphasizing the necessity for urological anomaly work up. Urological anomalies were the second frequent association among VACTREL associated patients.

The most common urological abnormality noted on screening USG was hydroureteronephrosis in the present study. Minneci et al., reported, 89.7% of renal anomalies on screening USG and Sangkhathat et al., had similar observations. In our study, USG detected 9 cases (20%) of renal agenesis. USG also detected other structural anomalies such as cystic kidney disease, renal ectasia, and duplex systems. USG screening for such anomalies in ARM patients is an accurate tool in detecting upper tract abnormalities¹⁵ and should be the customary care.¹⁸

All patients with ARM underwent MCU as a standard protocol in our study. Most frequently encountered anomaly on MCU in the present study was VUR analogous to other studies.^{11,16,17,19} MCU is one of the key investigations in diagnosing urological abnormalities. USG KUB and MCU are two different modalities to identify urological anomalies. USG is a non-invasive screening method of investigation with relative ease of availability in most of the centers. USG KUB was superior in identifying structural abnormalities of urinary system and upper urinary tract involvements. Functional urological abnormalities such as VUR and PUV are best diagnosed with MCU and are the gold standard investigation. Sangkhathat et al., made similar observation in their study spanning for 13 years that functional and lower urinary tract anomalies can be missed if only USG was instituted for investigating associated urological anomalies in ARM patients. In our study, MCU detected VUR in 22 children, of which eight children had low-grade reflux. On contrary, USG KUB could

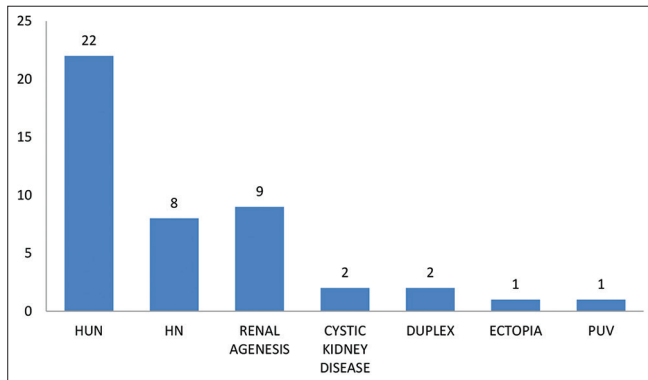


Figure 3: Depiction of various urological abnormalities in anorectal malformations in this study

only show total of 14 children with hydroureteronephrosis (Figure 3). Hence, we conclude that MCU is more sensitive in detecting even low-grade reflux (Figure 2a). Since it detects primarily refluxing urinary system, chemoprophylaxis can be initiated. This simple preventive measure helps in protecting upper renal tract from deterioration when compared to patients in whom reflux was addressed late or in undiagnosed cases. Therefore, it is necessary to highlight the importance of systematic screening to detect such clinically significant associated urological abnormalities influencing morbidity and mortality in patients of ARM. Minneci et al., performed a multicentric study that included almost ten children hospitals with significant numbers and emphasized on following standardized screening protocol to diagnose associated anomalies in cases of ARM.¹⁸

One interesting case of PUV associated with ARM was noted in our study which is quite a rare association.²¹ MR urography was done in two cases of duplex renal system in our study. In diagnosing complex structural urological anomaly, MR urography is superior to conventional intravenous urography and CT urography.²²

Majority of the urological abnormalities observed in our study represent important source of upper urinary tract deterioration which is preventable. Even though the association of urological abnormalities in ARM is well recognized, the lack of systematic approach in evaluating these patients leads to missed urological anomalies causing falsely negative cases. Therefore, standard algorithm needs to be followed in all cases of ARM.

Limitations of the study

Small sample size of patients.

CONCLUSION

Every ARM patient should be screened for associated urological anomalies at the earliest with standardized

protocol which includes MCU. Including MCU in screening protocol increases the chances of detecting low-grade VUR patients. Such proactive screening and prompt management of urological anomalies in all ARM children significantly reduces the morbidity and mortality improving the quality of life.

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Authors' Contributions:

SS- Concept and study design, coordination, and prepared first draft of manuscript statistical analysis, and interpretation; **KIP-** Interpretation results, reviewed the literature, and manuscript preparation; and **NK-** Concept and revision of manuscript.

Work attributed to:

Gandhi Medical College and Hospital, Secunderabad - 500 025, Telangana, India.

Orcid ID:

Dr. Srinivas Srirampur - <https://orcid.org/0000-0003-1581-9679>

Dr. Kavimozhy Ilakkiya Poyyamozhy - <https://orcid.org/0000-0001-6978-2993>

Dr. Nagarjuna Kumbha - <https://orcid.org/0000-0001-5912-6132>

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