

# The urological anomalies associated with anorectal malformations

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## Summary:

**Background:** Urological anomalies are frequently associated with anorectal malformations which are a common source of significant morbidity.

**Objective:** Is to evaluate the incidence and nature of the urological anomalies with patients of anorectal malformations (ARM).

**Patients & methods:** The data from 95 patients with ARM were studied from January 2009 to January 2012 in this cross-sectional study. All patients underwent sonography of urinary tract. Voiding cystourethrogram (VCUG) was done in patient with abnormal sonography & to all male patients with ARM who underwent colostomy. Other imaging studies were done in selected cases.

**Results:** Males significantly constitute the majority 64/95 (67.4%) of ARM cases, while females constitute 31/95 (32.6%) of patients, male to female ratio is 2:1. High type anomaly significantly constitutes the majority of the patients 79/95 (83.2%) in both genders. Urologic malformations were found in 24/95 (25.3%) patients with a significantly high frequency in males 21/64 (32.8%) while 3 of them were females 3/31 (9.7%). Urological anomalies occur more with high type ARM (27%) versus (12%) in low type ARM. Vesicoureteral reflux (VUR), renal agenesis, and hydronephrosis were the most common urologic anomalies respectively.

**Conclusions:** The high incidence of associated urogenital anomalies necessitates a careful investigation of all patients with ARM and continued long term follow up of these patients is mandatory to avoid deterioration of renal function in future.

**Keywords:** Urological anomalies, anorectal malformations.

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## Introduction:

Urinary tract malformations have been described in up to 50% of patients with anorectal malformations (ARM) in several large series<sup>(1,2)</sup>. Upper urinary tract anomalies present in 50% of boys and 30% of girls. The risk for both sets of problems increased with the level of the anorectal lesion<sup>(3)</sup>. ARM may be classified as either "high" or "low" anomalies based on their relationship to the levator muscle complex<sup>(4)</sup>. Low lesions such as perineal cutaneous fistula have less than 10% chance of having urinary anomaly, while high lesions such as rectovesical fistulas have as high as 90% risk<sup>(5)</sup>. The mortality and morbidity of ARM are influenced by the associated anomalies as the anorectal lesion itself<sup>(1)</sup>. The earlier series reported, that the mortality from many of the associated anomalies was higher than it is at present time<sup>(3)</sup>. A wide range of urinary tract abnormalities, including neuropathic bladder, vesico-ureteric reflux (VUR), duplication of the ureter and ureterocele are common with ARM and may increase the long-term morbidity<sup>(6)</sup>. Hydronephrosis, urosepsis and metabolic acidosis from poor renal function represent the main sources of morbidity in infants with ARM. Urogenital malformations may cause renal damage and may lead to chronic renal failure if not detected in time. Early diagnosis of urogenital malformations in neonatal period is essential in preventing future complications<sup>(5)</sup>.

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Usually the management of ARM gets priority & the urological evaluation is not always given its due importance, though in many cases the urinary tract disorder is the primary cause of morbidity and mortality<sup>(7)</sup>. The objective of this study is to evaluate the incidence and nature of the urological anomalies associated with anorectal malformations (ARM).

## Patients & methods:

In this cross sectional study, the records of 95 patients with ARM were reviewed who were admitted to Children Welfare Teaching Hospital between January 2009 and January 2011. The variables included: gender, the level of ARM (High or low), the findings of physical examinations and the results of radiological and imaging studies. The level of the anomaly was detected by radiography and surgeon's report on operation. All patients underwent ultrasound evaluation of the urinary tract, AP and lateral X-ray of lumbosacral spine, intravenous urography were done in selected cases with abnormal ultrasonography. Voiding cystourethrogram (VCUG) was done in patient with abnormal sonography and in all male patients with high type ARM who underwent colostomy. Urinary tract anomalies were defined as any renal, ureteral, bladder or urethral malformations, excluding fistula<sup>(5)</sup>. Statistical Package for Social Sciences-version 18 (SPSS 18) was used for data analysis. Chi square test for goodness of fit used to

test the significance of observed distributions. Chi square test for independence used to test the significance of association between discrete variables. Findings with P value less than 0.05 considered significant.

### Results:

Of the 95 neonates with imperforated anus, 64 patients (67.4%) were males and 31 patients (32.6%) were females. Males significantly constitute the majority (67%) of ARM cases ( $P < 0.05$ ). Male to female ratio is 2:1. Totally, 79/95 (83.2%) patients have high type ARM, that's include 52 males and 27 females, and 16/95 (16.8%) patients have low type ARM, which includes 12 males and 4 females. High type anomaly significantly constitutes the majority of the cases in both genders ( $P < 0.05$ ), (table 1). The associated urinary anomalies were detected in 24 (25.3%) patients; twenty one of them were males 21/64 (32.8%) and three of them were females 3/31 (9.7%). It is significant to find males to have more associated urinary anomalies than females ( $P < 0.05$ ), (table 2). The incidence of associated urological anomalies were found to be higher with "high" type ARM patients 22/95 (23.2%) than the "low" type ARM 2/95 (2.1%), but this is not considered statistically significant ( $P > 0.05$ ), (table 3). Multiple congenital urinary anomalies were seen in 11 patients (11.6%) from the whole study sample, eight were males (8.4%) & three were females (3.2%). The most frequently encountered lesion was Vesicoureteral reflux (VUR) (Fig.1) which affect 8 patients (8.4%), while Solitary kidney was seen in 6 patients (6.3%). Hydronephrosis was seen in 5 patients (5.3%). Ectopic kidney, renal stone and hypospadias all were seen in 4 patients of each (4.2%), Multicystic kidney was found in 2 patients (2.1%) and both ectopia vesica and megaureter was found in one patient (1.1%) of each, (table 4).

### Discussion:

In the present series, male patients were significantly constitute the majority 64/95 (67.4%) of the ARM patients ( $p < 0.05$ ), this was consistent with Nah SA et al<sup>(8)</sup> in Singapore from 2002 to 2011 & Metts et al<sup>(9)</sup>, in Tennessee (USA) from 1974 to 1995, while in Tabriz (Iran) between 2003 to 2005 Fakhrossadat<sup>(10)</sup> found no gender difference in anorectal malformations. In the present series. High type ARM significantly constitutes the majority of patients 79/95 (83.2%), ( $p < 0.05$ ), this was consistent with Alireza Mirshemirani et al<sup>(11)</sup> in Tehran (Iran) between 2002 to 2003 while Fakhrossadat<sup>(10)</sup> found no significant association in the level of deformity. It was significant to find males (21/64, 32.8%) to have more associated urinary anomalies than females, this was consistent with Metts et al<sup>(9)</sup>, but it was not consistent with Fakhrossadat<sup>(10)</sup> who found no gender difference in urinary anomalies but he found male predominance in genital anomalies only. The associated urinary anomalies were detected in 24 (25.3%) patients out of 95 patients with anorectal malformations, this was higher than Vaishali et al<sup>(7)</sup> in Kalkata (India) from 2002 to 2003, who found that the associated urological anomalies is only (11.36%), and is lower than Mark A. Rich et al<sup>(12)</sup> in New

Hyde Park (USA) from 1981 to 1988 and W. J. H. Goossens et al<sup>(13)</sup>, in Netherlands from 1983 to 2003 who found (48-52) % associated genitourinary anomalies respectively, but it is consistent with the thirteen years review (1988-2001) in Thailand by Sangkhathat et al<sup>(14)</sup> who found 25.6% incidence and Nah SA<sup>(8)</sup> who found 28.28% incidence.

The incidence of associated urological anomalies were found to be higher with "high" type ARM patients 22/95 (23.2%) than the "low" type ARM 2/95 (2.1%), although this was not considered statistically significant ( $P > 0.05$ ) but it is consistent with Sangkhathat et al<sup>(14)</sup> who reported higher incidence of urologic anomalies in high ARM. Mark A. Rich et al<sup>(12)</sup> found that the incidence and severity of associated genitourinary anomalies was directly related to the level of the fistula between the blind ending rectum and the genitourinary tract, high level fistulae to the bladder neck in males and the cloaca in females demonstrated 90% incidence of associated genitourinary anomalies, in contrast the lower level fistulae to the perineum which revealed only 14% incidence of associated genitourinary anomalies<sup>(12)</sup>. In the present series, VUR was the most common urological anomaly which is seen in 8.4 % of patients, while Vaishali et al<sup>(7)</sup> found 5.45% of their patients had VUR. Mark A. Rich<sup>(12)</sup> recognized 14.4% incidence of VUR, while others recognized higher percentage of VUR such as Fakhrossadat Mortazavi et al<sup>(10)</sup> and Alireza Mirshemirani et al<sup>(11)</sup> who recognized 42% and 52.9% respectively. This wide variation in incidence of VUR is related to the different methods of the studies, in some studies VCUG was performed only when ultrasonographic findings were abnormal. The ultrasound is an accurate tool in the examination of the upper urinary tract, but it is not sensitive enough to detect lower tract anomalies, especially VUR<sup>(14)</sup>. Ideally, the ARGUS protocol devised by Boemer et al<sup>(15)</sup> should be used for evaluating the new born with ARM. However, the poor socioeconomic condition of most of our patients precludes the use of all investigations suggested by Boemer et al<sup>(15)</sup> to be mandatory in all patients with ARM. Voiding cystourinarygram is mandatory even in those with normal sonography and prophylaxis antibiotics for urinary infection should be initiated until VCUG is performed, because the incidence of VUR is really high<sup>(10, 11)</sup> (42% and 52.9) respectively. Thus, the actual incidence of reflux is probably higher than the diagnosed in the present study, VUR was found in only 8 patients in the present series but only a half of patients underwent voiding cystourethrogram, so the true incidence of reflux unrelated to other problems may be higher. Even after definitive management of ARM, continued long term follow up of these patients is mandatory to exclude the possibility of urinary tract infection or neurogenic bladder<sup>(12)</sup>. Hydronephrosis and renal agenesis are the most common anomalies of the upper urinary tract in this study which found in 5 & 6 patients respectively. Considering that hydronephrosis is secondary to other anomalies such as VUR and bladder dysfunction, so renal agenesis may be considered as the most common primary anomaly of upper urinary tract which is seen in (6.3%) of patients and this percentage is consistent with Vaishali et al<sup>(7)</sup> and Fakhrossadat Mortazavi et

al<sup>(10)</sup> who found renal agenesis (5.45%-8.6%) of their patients respectively, while Mark A. Rich et al<sup>(12)</sup> found renal agenesis in 18% of patients.

In the present study, renal ectopia found in (2.27%) of the patients which is consistent with Fakhrossadat Mortazavi et al<sup>(10)</sup> who found 3.8% unilateral renal ectopia, while Vaishali Srivastava et al<sup>(7)</sup> found (9.09%). In the present series, hypospadias was found in (4.2%) of patients which is similar to Fakhrossadat Mortazavi et al<sup>(10)</sup> who reported (5.7%) while Vaishali Srivastava et al<sup>(7)</sup> reported (16.36%) incidence.

**Conclusions:** The high incidence of associated urogenital anomalies necessitates a careful investigation of all patients with ARM during neonatal and early infantile period, VCUG is essential even with normal sonographic findings to avoid

deterioration of renal function in future, continued long term follow up of these patients is mandatory to exclude urinary tract infection or neurogenic bladder.

**Table 1: Distribution of study sample according to level of ARM and to gender.**

	Level of ARM						Chi square	P value
	High		Low		Total			
Gender	N	%	N	%	N	%		
Male	52	81.2	12	18.8	64	100.0	25.000	< 0.001
Female	27	87.1	4	12.9	31	100.0	17.065	< 0.001
Total Sample	79	83.2	16	16.8	95	100.0	41.779	< 0.001

**Table 2: Distribution of study sample according to the presence of associated urinary anomaly and to gender.**

Presence of Associated Urinary Anomaly	Gender				Total		Chi-square	P value
	Male		Female		N	%		
	N	%	N	%			N	%
Yes	21	32.8	3	9.7	24	25.3		
No	43	67.2	28	90.3	71	74.7		
Total	64	100.0	31	100.0	95	100.0	5.920	0.015

**Table 3: Distribution of cases with renal anomalies according to the level of ano-rectal malformation and to gender.**

Presence of Associated Urinary Anomaly		Level of ARM						Chi square	P value
		High		Low		Total			
		N	%	N	%	N	%		
Total sample	Yes	22	23.2	2	2.1	24	25.3	1.660	0.198
	No	57	60.0	14	14.7	71	74.7		
	Total	79	83.2	16	16.8	95	100.0		
Male	Yes	19	29.7	2	3.1	21	32.8	1.746	0.186
	No	33	51.6	10	15.6	43	67.2		
	Total	52	81.3	12	18.8	64	100.0		
Female	Yes	3	9.7	0	0.0	3	9.7	---	1.000*
	No	24	77.4	4	12.9	28	90.3		
	Total	27	87.1	4	12.9	31	100.0		

\*Fisher Exact Test

**Table 4: Distribution of study sample according to type of anorectal malformation and to gender**

Associated Urinary Anomaly	All		Male		Female	
	(% out of 95)		(% out of 64)		(% out of 31)	
	N	%	N	%	N	%
Vesico-ureteric Reflux	8	8.4	6	9.4	2	6.5
Multicystic Kidney	2	2.1	1	1.6	1	3.2
Ectopic Kidney	4	4.2	4	6.3	0	0.0
Renal Stone	4	4.2	3	4.7	1	3.2
Solitary Kidney	6	6.3	5	7.8	1	3.2
Hypospadias	4	4.2	4	6.3	0	0.0
Ectopia Vesica	1	1.1	1	1.6	0	0.0
Megaureter	1	1.1	1	1.6	0	0.0
Hydronephrosis	5	5.3	3	4.7	2	6.5



Fig:1: Grade IV Vesicoureteric Reflux

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