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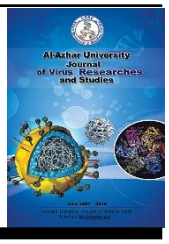


Urogenital Anomalies Associating with Undescended Testis

Asmaa Shaban Yousef*[1], M A Baky Fahmy[2], Yehia Hassan Zayed[3] and Dalia Gad[2]

[1]M.B.B.CH, Faculty of Medicine for Girls, Al-Azhar University, Cairo, Egypt

[2]Department of Pediatric Surgery, Faculty of Medicine for Girls, Al-Azhar University, Cairo, Egypt

[3]Department of General Surgery, Faculty of Medicine for Girls, Al-Azhar University, Cairo, Egypt

***E-mail: asmaashaheen883@gmail.com**

Abstract

Undescended testis is one of the most common congenital anomalies in pediatrics. The incidence of this anomaly is between 1.6% and 9.0%and up to 45% in preterm infants. Urogenital anomalies to be associated with undescended testis are renal agenesis, Ureteropelvic junction (UPJ) obstruction, hypospadias, hypoplastic phallus and others**.** Aim of the Work: The aim of the study is to evaluate the urogenital anomalies associated with undescended testis and emphasis the importance of early diagnosis and management of these cases**.** Twenty male patients were included in this analytical prospective study presented by undescended testes with associating urogenital anomalies admitted in the pediatric surgery unit at AL-Zahraa University Hospital in the period from April 2021 to October 2021**.** The range of age from 1 to 12 years old with mean (4.13 ± 2.76). Ten cases had bilateral undescended testes and ten cases had unilateral. Five cases were impalpable, and fifteen cases were palpable undescended testis. One associating urogenital anomaly was detected in 70 %. Two associating anomalies in 20 %. Three associating anomalies in 5%. Four associating anomalies in 5%. The most common urogenital anomaly was scrotal hypoplasia in 35 % and proximal hypospadias in 35%**.** Early diagnosis and management of undescended testis is important to prevent complications as fertility problems. Also, early detection and management of associating urogenital anomaly is important to improve the quality of life and to prevent psychic and medical complications.

Keywords: Undescended Testis, Associating Urogenital Anomalies.

1. Introduction

Undescended testis is one of the most common congenital anomalies in pediatrics. The incidence of this anomaly is between 1.6% and 9.0% and up to 45% in preterm infants [1]. The incidence drops to 0.9-1.8% at 3 months of age due to the spontaneous descent of the testes [1]. The embryological formation of the intermediate mesoderm derived urogenital system begins as two separate, yet

interwoven processes, development of the urinary system from the nephrogenic cord and development of the reproductive system from the gonadal ridge. The interplay between multiple genes and hormones is essential for both systems to develop properly. Mutation or misstep in any portion of the cascade of events can cause a double, absent, or malformed structure [2]. The testicle descends from the abdomen to scrotum to find a lower ambient temperature for normal spermatogenesis [3]. The testicular descent occurs in two stages with different anatomical, mechanical mechanisms and hormonal controls: the trans-abdominal phase and the inguino-scrotal phase [4]. Trans-abdominal phase lasts from the 8th to 15th week of gestation and the inguino-scrotal phase lasts from the 25th to 35th week of gestation. An interval of around 10 weeks of inactivity between the phases is not readily explained [5]. The most common congenital anomalies according to their incidence are musculoskeletal, cutaneous, and urogenital. The most frequent genitourinary anomalies are renal, testicular, and urethral, respectively. About 10 % of the population has some kind of genital or urinary system anomaly [6]. Urogenital anomalies found to be associated with undescended testis are renal agenesis, ectopic kidney, hypospadias, hypoplastic phallus, pelvi-ureteric junction (PUJ) obstruction, Extrophy of the bladder, urachal cyst and bilateral vesico ureteric reflux (VUR) [7]. Renal agenesis has been associated with genital abnormalities in 20-70% of the cases. In males, many anomalies have been reported like seminal vesicle cysts and an ectopic drainage of the ureter. In very few cases have renal agenesis have been reported in association with undescended testis [8]. The incidence of intersexual disorder in the patient population with undescended testes was 4 %, but it increased to 55% in the group of patients with both undescended testes and hypospadias [9]. Both undescended testes and hypospadias are believed to be related to androgen secretion or receptor disorders [10].

2. Patients and Methods

Twenty male patients were included in this study which is an analytical prospective study for children presented by undescended testis with associating urogenital anomalies admitted in the pediatric surgery unit at AL-Zahraa University Hospital in the period from April 2021 to October 2021. We included in this study boys whose age between 6 months to 12 years who have unilateral or bilateral, palpable or impalpable undescended testis with associating urogenital anomalies. While we excluded boys whose age above 12 years old or below 6 months, Patients who have isolated undescended testicle and Patients with syndromes. All patients were subjected for the following: Informed consent was obtained by their gardians and the study was approved by local ethical committee. Detailed history including: (Name, age, residency, preterm or term). Prenatal history whether the mother received medications, hormonal therapy or exposed to pollutants and radiations during pregnancy. Complaint includes (empty scrotum with abnormal site of meatal opening, drippling, recurrent urinary tract infection UTI). Time of discovery compliant and other manifestation suggestive for other system affection like recurrent urinary tract infection or abdominal pain. History of neonatal intensive care unit (NICU) admission, any medical diseases and history of previous operations, timing of circumcision and its relation to time of discovery. General examination: carefully done, Local examination of the genital system penis (size, meatal opening, chordee, penoscrotal angel), scrotum, both testes unilateral or bilateral undescended, palpable or impalpable), median raphe and examination of perineum. Abdominal

examination for palpation of both kidneys. Complete blood picture, liver & kidney function tests, urine analysis) and Hormonal assay for some cases, (like proximal hypospadias, microphalus) in the form of FSH, LH, free and total testosterone, and Anti-müllerian hormone. Karyotyping to define sex if the patient has penoscrotal hypospadias with undescended testis. Inguinoscrotal, pelvic and abdominal US. Selected cases needed MRI abdomen and pelvis, MRU and voiding cystourethrogram.

3. Treatment

3.1 Hormonal therapy

The dose of hormonal therapy is usually as follows: Human chorionic gonadotrophin (HCG 50-100 IU/kg body weight intramuscular once a week for 3 weeks). If testicle responding to hormonal therapy (scrotal positioning of the testis) no surgical interference and follow up. If it still undescended surgical interference to be done. palpable undescended testis treated by open orchiopexy. Impalpable undescended testis: laparoscopic orchiopexy whether in one stage if adequate long cord or two stages if short cord.

3.2 Management of associating anomalies

Absent kidney: Follow up by kidney function tests and pelvi abdominal US, MRU, or further renal scanning. Ureteropelvic junction obstruction Follow up by ultrasonography and renal scan then Pyeloplasty if needed and the case in our study did not need pyelopasty. Hypospadias: repair after management of undescended testis, distal cases with stenotic mobilized meatus operated by meatal advancement and glansplasty (MAGPI), wide deeply located meatus treated by glans approximation procedure (GAP), wide mouthed meatus with flat shallow ventral groove tabularized incised plate urethroplasty (TIP) was done. Proximal Hypospadias: two-staged approach is the most frequent technique used. First stage, chordee was corrected and foreskin flap or buccal graft, whereas the urethroplasty was performed in the second stage.

3.3 Hypo plastic Phallus

Surgical correction represents the gold standard therapy for penile chordee. Plication techniques (including excisional corporoplasty, incisional corporoplasty or plication-only)

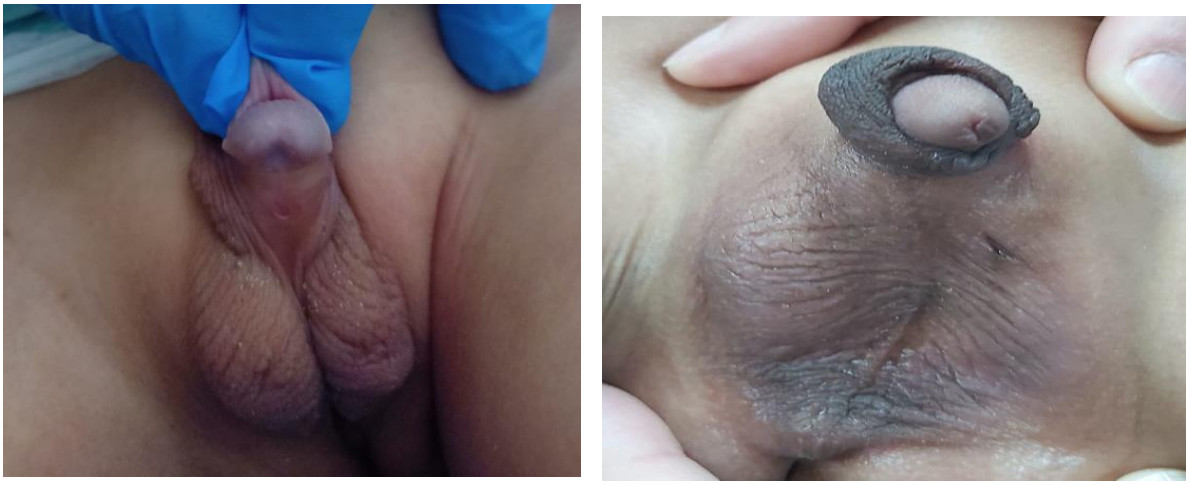
3.4 Hypo plastic Phallus

Topical application of 5% testosterone cream once per day, with review after 6–8 weeks to assess the effect. Patients with macropains who also suffer from penile dysmorphic disorder require careful and intensive psychological counseling.

3.5 Scrotal hypoplasia

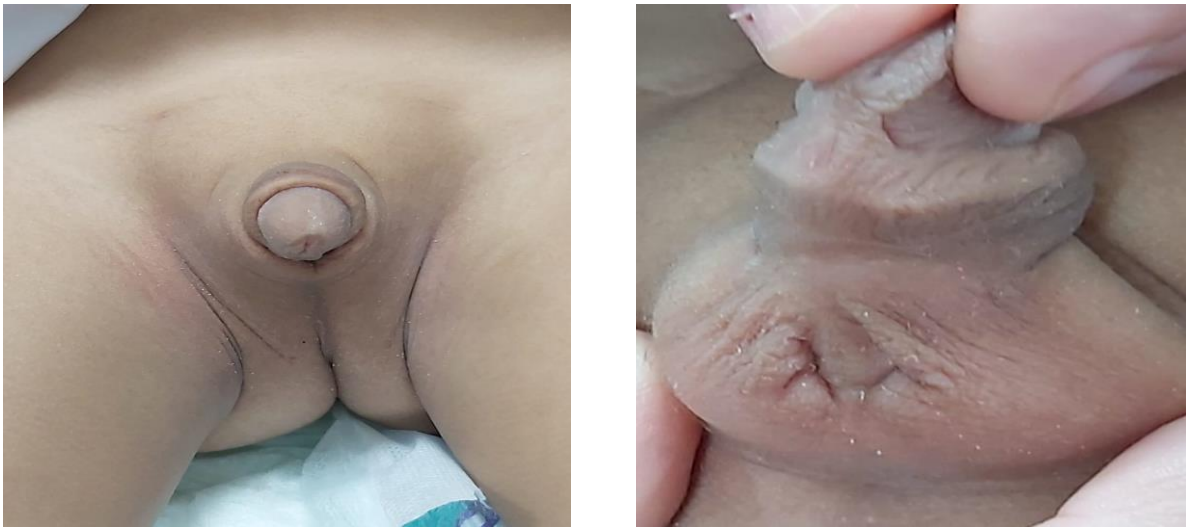
Topical testosterone for six weeks in all diagnosed cases of SH before any attempts to do orchidopexy or hypospadias repair, with a strict follow up. A promising results and minimal side effects were encountered. Follow up by kidney function tests and pelviabdominal US if the case has renal anomaly**.** Regular follow up for testis **.** This study recorded single anomaly association in (70%) of cases, two anomaly association (hypoplastic scrotum with hypospadias or penile chordee with hypospadias or hypospadias with scrotal transposition) in (20 %), three (hypoplastic scrotum, small phallus and hypospadias) in (5%) and four (hypoplastic scrotum) hypospadias, penile chordee and small phallus) in (5%**.(** The most common urogenital anomaly was scrotal hypoplasia in 7 cases (35%) and proximal hypospadias in 7 cases (35%). Small phallus in 4 cases (20%), Absent kidney (Renal agenesis) in 3 cases (15%),

Penile Chordee in 2 cases (10%), Scrotal transposition in 2 cases (10%), Ureteropelvic Junction (UPJ) Obstruction in1 case (5%), Distal hypospadias in 1 case (5%), Ectopic testis in 1 case (5%), Müllerian remnant in 1 case (5%) in which the vas was closely related to this remnant and only biopsy was taken.



(A) (B)

Figure (1): (A) bilateral Undescended teste with proximal hypospadias. (B): Undescended testis with penile chordee



(A) (B)

Figure (2): (A) Bilateral undescended testis with bilateral hypoplastic scrotums (B) post orchidopexy Postoperative follow up for any surgical complication as wound infection.

3. Results

Table .1 shows that there was statistically significant increase in the percentage of patients with distal hypospadius and müllerian remnant in maternal risk factors group than patients with no maternal risk factors while no statistically significant relation found between maternal risk factors and the other associating urogenital anomalies. Table .2 shows that there was no statistically significant relation found between presence of fetal risk factors and presence of associating urogenital anomalies.

Table (1): Relation between presence of maternal risk factors such as (hormonal therapy, usage of cosmetics and maternal obesity) and associating urogenital anomalies of the studied patients.

		No Maternal risk factor		Maternal risk factor		Test value	P-value	Sig.
		No.	%	No.	%			
Proximal hypospadias	Negative	10	62.5%	3	75.0%	0.220	0.639	NS
	Positive	6	37.5%	1	25.0%			
Distal hypospadias	Negative	16	100.0%	3	75.0%	4.211	0.040	S
	Positive	0	0.0%	1	25.0%			
Small phallus	Negative	12	75.0%	4	100.0%	1.250	0.264	NS
	Positive	4	25.0%	0	0.0%			
Chordee	Negative	15	93.8%	3	75.0%	1.250	0.264	NS
	Positive	1	6.3%	1	25.0%			
Hypoplastic scrotum	Negative	9	56.3%	4	100.0%	2.692	0.101	NS
	Positive	7	43.8%	0	0.0%			
Scrotal transposition	Negative	15	93.8%	3	75.0%	1.250	0.264	NS
	Positive	1	6.3%	1	25.0%			
Ectopic testis	Negative	15	93.8%	4	100.0%	0.263	0.608	NS
	Positive	1	6.3%	0	0.0%			
Müllerian remnant	Negative	16	100.0%	3	75.0%	4.211	0.040	S
	Positive	0	0.0%	1	25.0%			
Renal anomaly	Negative	13	81.3%	4	100.0%	0.882	0.348	NS
	Positive	3	18.8%	0	0.0%			
UPJ obstruction	Negative	15	93.8%	4	100.0%	0.263	0.608	NS
	Lt UPJ	1	6.3%	0	0.0%			
No. of anomaly finding 1		11	68.8%	3	75.0%	0.580	0.901	NS
2		3	18.8%	1	25.0%			
3		1	6.3%	0	0.0%			
4		1	6.3%	0	0.0%			

P-value >0.05: Non-significant (NS); P-value <0.05: Significant (S); P-value< 0.01: highly significant (HS) *: Chi-square test

Table (2): Relation between presence of fetal risk factors such as (prematurity and intrauterine growth restriction) and associating urogenital anomalies of the studied patients.

			No Fetal risk factor		Fetal risk factor		Test value	P-value	Sig.
			No.	%	No.	%			
Proximal hypospadias		Negative	12	66.7%	1	50.0%	0.220	0.639	NS
		Positive	6	33.3%	1	50.0%			
Distal hypospadias		Negative	17	94.4%	2	100.0%	0.117	0.732	NS
		Positive	1	5.6%	0	0.0%			
Small phallus		Negative	14	77.8%	2	100.0%	0.556	0.456	NS
		Positive	4	22.2%	0	0.0%			
Chordee		Negative	16	88.9%	2	100.0%	0.247	0.619	NS
		Positive	2	11.1%	0	0.0%			
Hypoplastic scrotum		Negative	11	61.1%	2	100.0%	1.197	0.274	NS
		Positive	7	38.9%	0	0.0%			
Scrotal transposition		Negative	16	88.9%	2	100.0%	0.247	0.619	NS
		Positive	2	11.1%	0	0.0%			
Ectopic testis		Negative	17	94.4%	2	100.0%	0.117	0.732	NS
		Positive	1	5.6%	0	0.0%			
Müllerian remnant		Negative	17	94.4%	2	100.0%	0.117	0.732	NS
		Positive	1	5.6%	0	0.0%			
Renal anomaly		Negative	16	88.9%	1	50.0%	2.135	0.144	NS
		Positive	2	11.1%	1	50.0%			
UPJ obstruction		Negative	17	94.4%	2	100.0%	0.117	0.732	NS
		Lt UPJ	1	5.6%	0	0.0%			
No. of anomaly finding	1		12	66.7%	2	100.0%	0.952	0.813	NS
	2		4	22.2%	0	0.0%			
	3		1	5.6%	0	0.0%			
	4		1	5.6%	0	0.0%			

P-value >0.05: Non-significant (NS); P-value <0.05: Significant (S); P-value< 0.01: highly significant (HS) *: Chi-square test.

Table .3 shows that there was no statistically significant relation found between laterality and associating urogenital anomalies among the studied patients.

Table (3): Relation between laterality and associating urogenital anomalies of the studied patients.

			Unilateral		Bilateral		Test value	P-value	Sig.
			No.	%	No.	%			
Proximal hypospadias		Negative	7	70.0%	6	60.0%	0.220	0.639	NS
		Positive	3	30.0%	4	40.0%			
Distal hypospadias		Negative	10	100.0%	9	90.0%	1.053	0.305	NS
		Positive	0	0.0%	1	10.0%			
Small phallus		Negative	8	80.0%	8	80.0%	0.000	1.000	NS
		Positive	2	20.0%	2	20.0%			
Chordee		Negative	10	100.0%	8	80.0%	2.222	0.136	NS
		Positive	0	0.0%	2	20.0%			
Hypoplastic scrotum		Negative	8	80.0%	5	50.0%	1.978	0.160	NS
		Positive	2	20.0%	5	50.0%			
Scrotal transposition		Negative	10	100.0%	8	80.0%	2.222	0.136	NS
		Positive	0	0.0%	2	20.0%			
Ectopic testis		Negative	9	90.0%	10	100.0%	1.053	0.305	NS
		Positive	1	10.0%	0	0.0%			
Müllerian remnant		Negative	9	90.0%	10	100.0%	1.053	0.305	NS
		Positive	1	10.0%	0	0.0%			
Renal anomaly		Negative	8	80.0%	9	90.0%	0.392	0.531	NS
		Positive	2	20.0%	1	10.0%			
UPJ obstruction		Negative	10	100.0%	9	90.0%	1.053	0.305	NS
		Lt UPJ	0	0.0%	1	10.0%			
No. of anomaly finding	1		9	90.0%	5	50.0%	4.143	0.246	NS
	2		1	10.0%	3	30.0%			
	3		0	0.0%	1	10.0%			
	4		0	0.0%	1	10.0%			

P-value >0.05: Non-significant (NS); P-value <0.05: Significant (S); P-value< 0.01: highly significant (HS) *: Chi-square test.

4. Discussion

Previous Patients with undescended testes are known to have associated anomalies. In. Cheng et al., 1996 [11] study, the incidence was 20 %. The incidence of intersexual disorder in the patient population with undescended testes was 4%, but it increased to 55 % in the group of patients with both undescended testes and hypospadias. This was very similar to Rajfer's figure of 53 %. [9]. Both undescended testes and hypospadias are believed to be related to androgen disorders [10]. Diaphragmatic hernia in their categorization was grouped with omphaloceles and umbilical hernia. It had been demonstrated that the larger the abdominal wall defect, the higher the incidence of undescended testes, suggesting decreased intra-abdominal pressure as one of the possible causes of undescended testes [12]. There were two patients with lumbosacral myelomeningocele. Hutson found that the incidence of undescended testes among patients with high lumbar myelomeningocele was nearly 40 per cent and attributed the maldescent to disruption of genitofemoral nerve-mediated androgen effect [13]. One patient had both cleft palate and cleft lip, but he was not short in stature as seen in neurofacial midline defect which involves hypothalamo-pituitary dysfunction. Undescended testis is part of Prader-Willi syndrome and Noonan syndrome. If the total number of patients was divided into two groups, one with bilateral undescended testes and one with unilateral ones, 46 % of the bilateral group were found to have at least one associated anomaly, whereas only 10% of the unilateral group had any associated anomalies. Patients with bilateral undescended testes were thus more likely to have other anomalies. When they look at multiple anomalies (i.e., patients with more than one anomaly), the bilateral group was again found to have a higher incidence of multiple associated anomalies than the unilateral group (2 %). They conclude that bilaterality of undescended testes should raise a clinician's suspicion of possible associated anomalies and the combination of hypospadias and undescended testes is likely to be associated with intersexual disorders [11,12,13]. In this study twenty patients of undescended testis have been found to have associating urogenital anomalies and properly managed according to each case**.** this study recorded single anomaly association in (70%) of cases, two anomaly association in (20 %), three in (5%) and four in (5%).

The most common urogenital anomaly was scrotal hypoplasia in 7 cases (35%) and proximal hypospadias in 7 cases (35%). Small phallus in 4 cases (20%), Absent kidney (Renal agenesis) in 3 cases (15%), Penile Chordee in 2 cases (10%), Scrotal transposition in 2 cases (10%), Ureteropelvic Junction (UPJ) Obstruction in1 case (5%), Distal hypospadias in 1 case (5%), Ectopic testis in 1 case (5%), Müllerian remnant in 1 case (%5)

There was statistically significant increase in the percentage of patients with distal hypospadias and müllerian remnant in maternal risk factors group than patients with no maternal risk factors while no statistically significant relation found between maternal risk factors and the other associating urogenital anomalies. There was no statistically significant relation found between presence of fetal risk factors and presence of associating urogenital anomalies. There was no statistically significant relation found between laterality and associating urogenital anomalies among the studied patients. There was statistically significant increase in the percentage of patients with scrotal transposition in palpable patients than impalpable patients while no statistically significant relation found between palpable/impalpable and the other associating urogenital anomalies.

5. Conclusion

The main goal of UDT treatment is to pull the testis down to the scrotum. Early diagnosis and management of any associating Urogenital anomaly is important to prevent Psychic and medical complications. Also, if the associating urogenital anomaly is surgically treatable like hypospadias, proper surgical repair to be done.

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