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Duplex Kidney and Ureteral Duplications Associated Congenital Anomalies and Complications in Two Private Hospitals, Aden

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Abstract

Introduction: Congenital anomaly of the upper urinary tract is common anomaly in the superior urinary tract. It is more often asymptomatic. However, can associate with other anomalies and resulted in high morbidity in adults. The early detection of the anomalies is helpful to avoid complications and preserve renal function**.** The objective of the study is to describe the associated congenital anomalies, complications and treatment in two Private Hospitals.

Methods: A descriptive study was carried out on adult patients with duplex renal and ureteral system in two Private Hospitals in Aden Governorate during the period May 2012 to November 2018. Records of adult patients with unspecific clinical manifestations were assessed. Diagnosis was carried out with ultrasound, contrasted urography and computerized tomography describing the associated anomalies, complications, and treatment.

Result: Ninety patients were included; 46% males and 54% females with ages ranged from 16 to 61 years (mean age = 33 years). Associated anomalies were found in 33% of patients. The most common anomalies were adult dominant polycystic kidneys (63.3%) and ureteroceles (23%). Complications occurred in 40% of patients. Of them, 64% of patients had urinary tract infections (UTIs), 14% stones, 14% ureteroureteral reflux, 8% non-functional moiety and 3% transitional cell carcinoma in left lower pole of incomplete ureteral duplication. Renal function was normal in 74%, delayed in 22% and non-functional renal moieties in 4% of patients. Medical treatment was carried out in 75.6% and surgical intervened in 24.4% of patients

Conclusion: The most frequent complications were recurrent UTIs. The commonest associated anomaly was adult dominant polycystic kidneys. Computerized tomography is the effective imaging study, followed by contrasted urography. Medical treatment is the commonest type of treatment.

Keywords: Adults, Duplex System, Complications, Aden.

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التشوهات الخلقية في الكلى والحالبين المثني التشوهات المصاحبة والمضاعفات في إثنتين من المستشفيات الخاصة في عدن

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ملخص الدراسة

المقدمة: التشوهات الخلقية في الكلى والحالبين المثنى خلقياً شائعة في الجهاز البولي العلوي. غالبا لا تظهر الأعراض، لكن يمكن أن تصاحبها تشوهات خلقية أخرى وتسبب مضاعفات عديدة في البالغين. التشخيص المبكر مهم لتجنب المضاعفات والحفاظ على وظائف الكلى. هدفت هذه الدراسة وصف التشوهات الخلقية المصاحبة، المضاعفات، والعلاج الطبي الأكثر استخداما.

المنهجية: تم إجراء دراسة استرجاعيه لملفات المرضى البالغين الذين أصيبوا بمضاعفة الكلى والحالبين دون ظهور أعراض محددة في إثنتين من المستشفيات الخاصة في محافظة عدن، خلال الفترة مايو 2012 حتى نوفمبر 2018. تم التشخيص بالسونار وأشعة الكلى الملونة والأشعة المقطعية لتحديد التشوهات الخلقية المصاحبة لها والمضاعفات ونوع العلاج.

النتائج: شملت الدراسة تسعون مريضا، 46% كانوا من الذكور و54% من الإناث بأعمار 18 وحتى 61 عاما (متوسط العمر 33 عاما). التشوهات الخلقية المصاحبة حدثت في33% من المرضى، وأكثرها حدوثاً هي الأكياس الخلقية الكلوية المتعددة في البالغين (63%) و التكيس الحالبي (23%). المضاعفات حدثت في 40% من المرضى وهي التهابات المسالك البولية المتكررة (64%)، الحصوات (14%)، ارتجاع البول (14%)، وفقدان وظيفة الكلى في الجزء المثني للكلية (8%)، وسرطان الجزء الأسفل من الحالب الأيسر المثنى خلقيا (3%). وظيفة الكلى كانت طبيعية عند 74% من المرضى و ظهر تأخر الإخراج في الكلى عند 22%، وفقدان الوظيفة في الجزء المتأثر عند 4%. تم تقديم العلاج الطبي لـ 75.6% والتدخل الجراحي لـ 24.4% من المرضى.

الاستنتاج: الأكياس الخلقية الكلوية المتعددة في البالغين هي التشوهات المصاحبة الأكثر حدوثا أما المضاعفات الأكثر شيوعا فهي التهابات المسالك البولية. الأشعة المقطعية هي الوسيلة الأساس للتشخيص تليها أشعة الكلى الملونة. العلاج الطبي هو الأكثر استخداماً.

الكلمات المفتاحية: البالغون، التشوهات الخلقية للكلى المثني، المضاعفات، عدن.

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Introduction

Duplicated ureter renal and duplex system is a common congenital anomaly in the superior urinary tract. It occurs in 1% of the population and usually diagnosed incidentally during imaging studies and occasionally found in abdominal examination or during abdominal surgery. It is classified as a partial or complete duplication, which may have either single or bifid ureter joins together and inserted into the bladder (partial duplication) or double separated ureters draining a single renal parenchyma and distally inserted into the urinary bladder (rather than split). Complete duplication occurs when the ureteral bud from the mesonephric duct arises twice. Complete duplications have two ureters that inserted separately into the bladder. Incomplete unilateral duplication occurs in 81% of patients and bilateral in 19% of patients while complete duplication is less frequent [1,2].

Complete duplication of the upper moiety ureter may associate with ureteroceles, ectopic ureter and segmental dysplastic kidney, and the lower moiety ureter may associate with reflux [3,4]. They present with unspecific clinical manifestations such as recurrent urinary tract infections, flank pain, incontinence hematuria, and palpable kidney [5,6].

Duplex system and ureteral duplication in adults, if not diagnosed early can lead to a significant morbidity. Furthermore, they can present with obstruction, stones, recurrent infection, hypertension, renal parenchymal atrophy of the affected moiety, or reduced renal function [7,8]. Duplex kidney usually diagnosed in children and adolescents, however, can present in adults producing significant complications which require surgical interventions. An imaging study plays an important role in early diagnosis and proper management and follow-up. New imaging methods such as computed tomography urography, MRI, and nuclear radioisotopes facilitate the diagnosis, and reveals the associated abnormalities and complications [9,10].

In rare instances duplex system appears in adults, leading to diagnostic challenges. Traditionally, they are operated by open surgery or by minimally invasive endoscopic and laparoscopic methods [11]. The objective of this study is to describe the congenital duplex renal system and ureters in adults, the associated anomalies, complications, and type of treatment.

Methods

A descriptive study carried out on patients diagnosed as duplex pelvicalyceal system and ureteral duplication in adults incidentally diagnosed by an imaging study in Alnaquib and Almansorah Private Hospitals in Aden Governorate during the period May 2012 to November 2018. Patients presented unspecific clinical manifestations. Those less than 18 years were excluded.

A detailed history and physical examination were carried out with proper laboratory investigations. Imaging studies included abdominal and pelvic ultrasound performed in all patients, contrasted urography,

abdominal and pelvic computerized tomography scan, describing the type of renal duplex system and ureteral duplication whether incomplete or complete, unilateral or bilateral, the associated urinary anomalies, complications, and type of treatment.

Data were processed and analyzed using SPSS Version 21. All data were summarized using the descriptive statistics including the mean for quantitative variables and frequency and percentages for qualitative variables. Statistical test was carried out for variables including the associated congenital anomalies, and complications to determine the *p*-value for significance using the Pearson significant Coefficient and Spearman's rho test for associated anomalies and complications and their effect on renal function. Patients identifiers were not included and organs exposure not revealed. Ethical approval and consent for participants Not applicable.

Results

Ninety patients were diagnosed as having duplex pelvicalyceal system and ureteral duplication. Incomplete unilateral duplication occurred in 62 patients (68.9%), and bilaterally in 20 (22.2%) with a total of 82 patients (91.1%). Complete duplications occurred in eight patients (9%), unilaterally localized six (75%), and bilaterally in two patients (25%) as demonstrated in Figure 1.

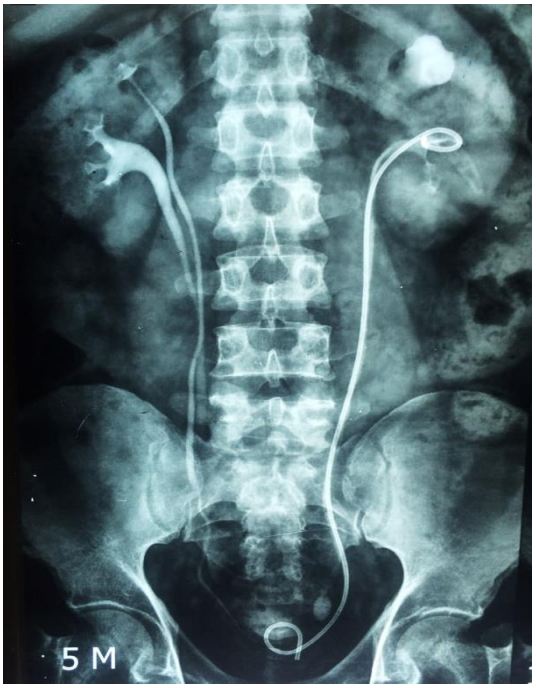

Figure 1: Contrasted Urography Reveals Complete Bilateral Renal and Ureteral Duplication Associated and Bilateral Ueteroceles.

The associated congenital anomalies (Table 1) are presented in 30 patients (33%). The highest percentage (63%) were adult dominant polycystic kidney disease (ADPKD). Ureteroceles was found in seven (23%), four of them with were complete unilateral duplication, and three with complete bilateral duplication. Other associated rare anomalies were found at smaller percentages.

Table 1: Anomalies associated with Adult congenital duplex renal system with (n=30)

Associated congenital anomalies	No	%
Adult polycystic kidneys	19	63.3
Ureteroceles	7	23.0
Renal agenesis	1	3.3
Segmental renal dysplasia	1	3.3
Horseshoe kidneys	2	7.0

The anomalies included right renal agenesis and incomplete left duplex system and lower pole hydronephrosis it associated with ADPKD and left lower pole segmental renal dysplasia in one patient in one patient (Figure 2).

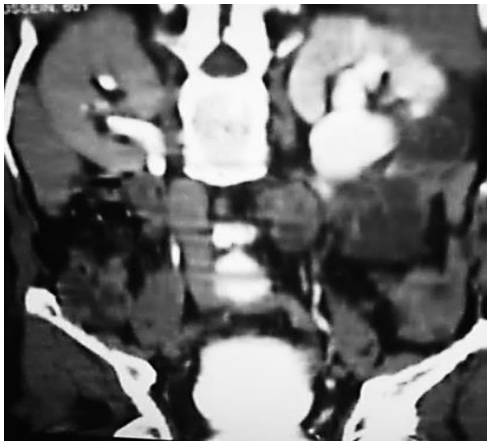

Figure 2: CT Urography: Incomplete Left Renal Duplication Associated with Lower Pole Segmental Cystic Dysplasia.

Incomplete bilateral collecting renal systems associated with bilateral horseshoe kidneys in one patient (Figure 3) and cystic mass in right moiety of horseshoe kidney and abnormal ventromedial malrotation in one patient (Figure 4).

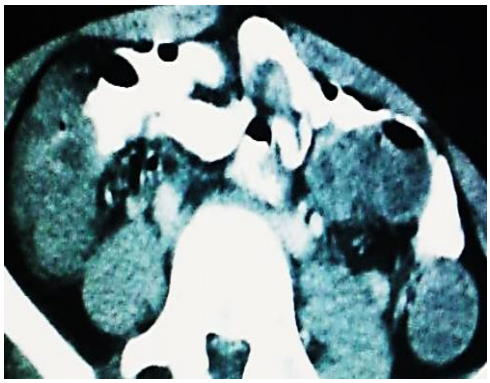

Figure 3: Contrasted CT Scan: Shows Incomplete Duplex Collecting Renal System Associated with Horseshoe Kidney, and Ventromedial Abnormal Malrotation.

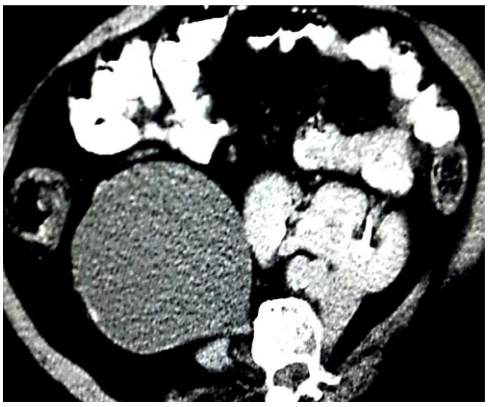

Figure 4: CT Scan Shows Right Renal Cystic Mass in the Right Component of Horseshoe Kidney Associated with Incomplete Duplex System with Left Kidney Ventromedial Malrotation.

Complications (Table 2), occurred in four patients of complete duplication and 32 patients with incomplete duplications with a total of 36 patients. These included urinary tract infections (64%), urinary stones (14%) mainly seen in incomplete duplications (Figure 5), vesicoureteral ureteroureteral reflux (14%), non-functional moieties (8%), and transitional cell carcinoma in left lower pole of incomplete ureteral duplication (3%) (Figure 6).

Table 2: Complications Associated with Congenital Adult Duplex System (n=36)

Complications	No	%
Urinary tract infection	23	64
Stones	5	14
Vesicoureteral reflux	5	14
Nonfunctional renal moieties	3	8

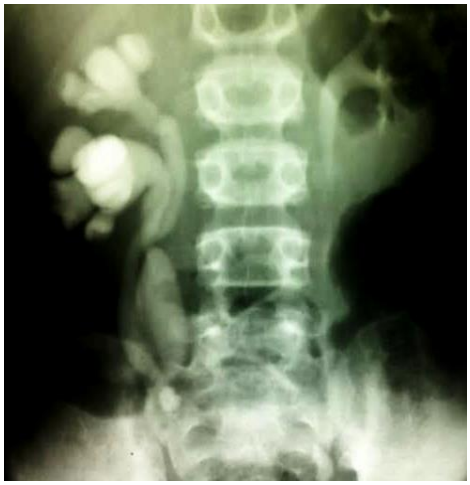

Figure 5: Contrasted Urography Demonstrates Incomplete Duplex System and Stone in Right Iliac Region.

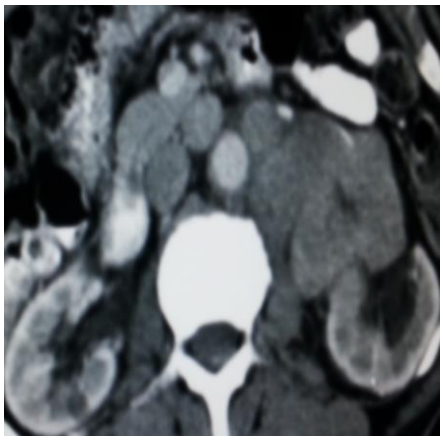

Figure 6: Contrasted CT reveals left incomplete duplex system with lower pole transitional cell carcinoma of ureter.

Renal function was normal in 67 patients (74.5%), whereas delayed renal excretion was found in 20 patients (22.2%), and non-functional renal moieties in three (0.3%).

Sixty-eight patients (75.6%) were managed by medical treatment whereas 22 (24.4%) by surgical intervention. Types of surgical intervention are shown in Table 3. These included detachment of ureteroceles (31.8%), stones removal and heminephrectomy (22.7% each), pyeloplasty (9.09%), and reimplantation of ureter with anti-reflux technique (13.8%).

Table 3: Surgical procedures in adult congenital duplex renal system (n=22)

Surgical procedures	No.	%
Ureteroceles detachment	7	31.8
Stones removal	5	22.7
Heminephrecomy	5	22.7
Reimplantation of ureter	3	13.8
Pyeloplasty	2	9.09

Discussion

In the present study, incomplete unilateral ureteral duplication was the most frequent type of duplex renal system and ureteral duplication. It is more commonly seen in the ureteropelvic region, followed by iliac region whereas complete bilateral duplex system was less frequent finding. This is consistent with what was reported by Stunnell *et al* in 2007 [12]

Recurrent UTIs were the most frequent complications followed by ureteroureteral reflux and stones which mainly occurred in incomplete duplications, and ureteroceles were most frequent in complete duplex system corresponding with described by others reported [13-15]. Complications in the upper moiety included, multicystic dysplastic moiety caused by ureteroceles and vesicoureteral obstruction, and lower moiety complicated by Vesicoureteral reflux and renal scarring as reported by Zissin *et al*; (2001); and Jaram *et al*; 2011 [1‹16].

This study presented rare associated congenital anomalies including adult ADPKD. Ureteral transitional cell carcinoma in the lower duplicated left ureter found in one patient. It is a rare finding. Kao *et al*. (2013) also reported one case. [17]. Right renal agenesis with left lower pole duplex system with hydronephrosis was seen in one patient and segmental cystic dysplastic kidney of the lower pole in another one. This is uncommon finding. The associated Horseshoe kidney with incomplete left duplex system found in one patient. They are extremely rare congenital anomalies. In 2009, Keskin *et al*. reported one case [18] whereas in 2011 Ongeti *et al*, reported another case [19]. Five cases of duplex collecting system in horseshoe kidneys were only reported by Singh *et al* in 2015 [20].

Ultrasound is the first imaging study for diagnosis of ureteroceles and demonstrated the non-function upper pole moiety but was not effective for the diagnosis of associated anomalies and complex anatomy. Ultrasound is shown to be ineffective study for diagnosis of associated ureteroceles in the duplex kidneys, and complex anatomy diagnosed by contrasted computerized tomographic (CT) urography. Doery *et al* in 2015 [21] considered ultrasound as the most common imaging study for early detection of complications.

Contrasted excretory urography is the first line imaging study, demonstrated the tortuous ureters and level of obstruction, difficulty arise in poor or absent renal function in one of the two moieties, consistent with reported. Antegrade pyelography is useful in patients with hydronephrosis; it demonstrated a second ureter and the level of ureteric termination [22, 23].

CT urography and excretory urography were the most effective imaging studies, revealed abnormalities of the collecting systems, and ureteoceles. CT scan helps in resolving the complex anatomy which could be not detected by ultrasound and is effective for detection of complications and associated anomalies [24]. The association of horseshoe kidney with incomplete duplex system is very rare finding [25]. Cystic renal cancer in an associated horseshoe kidney is an extremely rare finding. It represents 1% of renal cell carcinoma [26].

Conservative treatment carried out in the majority of patients in the present study. Presence of complications such as obstructive ureteroceles, stones, vesicoroureteral reflux, and nonfictional renal moiety should intervene surgically consistent with reported [27]. Upper pole heminephrectomy should consider as a surgical procedure for nonfunctional renal moieties was described by Clement *et al*; in his systematic review of literature in 2018 [28]. Duplex system in adults is a diagnostic challenge in the performed surgical interventions. The early detection of congenital anomalies and their associated anomalies is important to avoid the complications because delayed diagnosis have an increased risk of significant morbidity.

Conclusion

The study concluded that he most frequent complications are recurrent UTIs and the commonly associated congenital anomaly was adult dominant polycystic kidney disease.

Contrasted CT is the most effective imaging, followed by contrasted urography.

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