

Congenital Anomalies of the Urinary System among Sudanese on Hemodialysis at Gezira Hospital for Renal Diseases and Surgery, Gezira State, Sudan; 2018: Sonographic Study

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Abstract: Congenital anomalies of urinary system are of wide spectrum ranging from mild, asymptomatic malformations to severe fatal conditions. To investigate occurrence of congenital anomalies in urinary system. It was a cross-sectional study. Study populations were Sudanese patients with ESRD on regular hemodialysis attending Gezira Hospital for Renal Diseases & Surgery, Gezira State, Sudan. Independent variables were horseshoe kidney, polycystic kidney disease, double ureter or ureteral duplication, ureteropelvic junction obstruction, posterior urethral valves, hypogenesis, agenesis, and kidney position. Dependent variable was hemodialysis. Investigation was done using abdominal ultrasound. Personal data was obtained using a questionnaire. Data was analyzed using SPSS version 21. *P-value* ≤ 0.05 was considered statistically significant. Results: Thirty one (62%) of respondents was male. Minimum age was 19 years while maximum age was 70 years. Maximum affected age group was age group (36-45) years; followed by age groups (46-55) and (56-65) years. Minimum age at which disease was diagnosed was 13 years. Overall prevalence was 12%. Four types of congenital anomalies were encountered and these are: polycystic kidney disease, uretero-pelvic junction obstruction, hypogenesis, and agenesis. Minimum disease duration was 1 year while maximum duration was 16 years. Mean duration was 6.42 years. Gender distribution of these types was as follows: Male (Two cases of polycystic kidney disease, one case of ureteropelvic junction obstruction, and one case of agenesis); female (one case of polycystic kidney disease, and one case of hypogenesis). Duration of disease is higher among male. Conclusion and recommendation: The current study highlighted a significant statistical relationship of the congenital anomalies in the urinary system among respondents and duration of ESRF. Screening for presence of congenital anomalies of renal system among general population of Central State may be of great help.

Keywords: Congenital Anomalies of Renal System, ESRD, Sudanese Patients

Introduction: Congenital anomalies of the urinary system are of wide spectrum ranging from mild, asymptomatic malformations (e.g. double ureter or minimal pelvic obstruction) to severe fatal conditions as bilateral renal agenesis (Rodriguez M. 2014). Consideration of the congenital abnormalities of the urinary system is very important in order to be discovered early so as to minimize the morbidity that related to the urinary tract system diseases. Congenital anomalies of urinary system that associated with hemodialysis are: Horseshoe kidney, polycystic kidney disease, double ureter or ureteral duplication, ureteropelvic junction (UPJ) obstruction, posterior urethral valves, hypogenesis, agenesis, and kidney position [Elsharif, (2011)].

The incidence rate of congenital anomalies of the kidney and urinary tract (CAKUTs) in USA is 3–6 per 1000 live births; and constitute 34–59% of chronic kidney disease (CKD) and 31% of end-stage kidney disease (ESKD) in children. It is a cause for the most pediatric end-stage renal disease (ESRD) [Yosypiv, (2012)].

Ultrasound study is very important for diagnosis and treatment of the kidney abnormalities. Ultrasound is an accessible, inexpensive and fast aid for decision-making in patients with renal symptoms. It is safe, readily available, easily performed at the bedside or in the office, and free of radiation. The size and shape, the echogenicity, the urinary space (including the lower urinary tract), the presence of masses, and

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the vasculature are evaluated using ultrasound. It is the first-line imaging modality for evaluation of urinary obstruction and renal size. It is also valuable for differentiating congenital variants and simple cystic lesions from renal masses. The complex internal architecture of the kidneys has a highly variable appearance on ultrasound. However, renal ultrasound has certain limitations, and other modalities, such as CT and MRI, should be considered as supplementary imaging modalities in the assessment of renal disease [Hansen, et al. (2016)].

Sonographic findings in the normal kidney: The shape of the kidney, in the longitudinal scan plane, is similar to an oval bean. The perirenal fat is echogenic and it is separated from the kidney by a thin linear capsule that surrounded the kidney. Kidney is composed of two parts: (i) parenchyma: it is more hypoechoic and homogenous and divided into cortex externally and medullary pyramids (less echogenic) internally (ii) renal sinus and it is components (calyces, the renal pelvis, fat and the major intrarenal vessels) are hyperechoic. There is cortical infoldings (columns of Berin) spreading between the medullary pyramids. In pediatrics, hypoechoic medullary pyramids can be differentiated easily from the more echogenic cortex [Hansen, et al. (2016)].

Size and Shape of the kidney: Size of the kidney is a key parameter that should be measured carefully. The length of adult kidney usually between 10–12 cm and the right one is longer than the left; while its size varies according to body height and age; and, unfortunately, there are no nomograms for normal kidneys based on large population studies. Nomograms are available for children. Cortical thickness is 7-10 mm, measured from the base of the medullary pyramid to the edge of the kidney; and varies within a kidney, being thicker at the poles. Parenchymal thickness is 15-20 mm and can be used when it is difficult to differentiate pyramids. The echogenicity of the cortex decreases with age and is less echogenic than or equal to the liver and spleen at the same depth in individuals older than six months; while it is more echogenic than the liver and spleen when compared at the same depth in neonates and children up to six months of age [Hansen, et al. (2016)].

Horseshoe kidney: The identification of horseshoe kidneys is by demonstrating fused functional renal masses on both sides of the vertebral column. Their ureters are not crossed, on their way to urinary bladder. The connecting isthmus may be located in the center or deviated laterally. The lateral deviation of the isthmus leads to asymmetric horseshoe kidney.

The rate of occurrence of left deviation is about 70%. The site of fusion is mainly at the lower pole (90% of cases). In healthy population, incidence rate is about 1 in 500; with male to female ratio of 2:1. In urological patients may reach 1:304 and associated with chromosomal disorders. It is twice as common in males. Horseshoe kidneys are asymptomatic in about one third of patients. It is benign in nonsyndromic case. It may be associated with ureteropelvic junction (UPJ) obstruction and blood vessels crossing anteriorly. Other associated complications include vesicoureteral reflux in 50% of patients, injury in blunt trauma to abdomen, and different renal cancers. It is usually seen in trisomy 18 [Rodriguez, (2014)]. The rate of occurrence of horseshoe kidney in the general population is 0.25% [Briones, et al, 1998].

Polycystic kidney disease (PKD): It is composed of large number of growing fluid filled cysts within the kidneys resulting in renal damage that ending in renal failure (Polycystic Kidney Disease Foundation). Polycystic kidney disease is genetically determined; both autosomal dominant and recessive [Halvorson, et al, (2010)]. The most common genetic type is ADPKD [Hansen, et al. (2016)]. Characteristics of ADPKD is slow progressive enlargement of the kidneys ending in renal failure at fifth to sixth decade of life; with a rate of occurrence of 1:800 – 1:1000 patients. ADPKD contributed by about 2.5% of all cases of end-stage renal disease. While ARPKD occurs in a younger people, with an incidence of 1:20000 live births [Halvorson, et al, (2010)]. Polycystic kidney disease and renal artery stenosis are among the renal causes of secondary hypertension which may end in ESRD (Ilyas M). ADPKD is asymptomatic in most of patients at the time of diagnosis. Hepatic cysts are the most common extrarenal manifestations of ADPKD. Intracranial aneurysm is the most dangerous extrarenal manifestations of ADPKD [Halvorson, et al, (2010)]. The prevalence of nephrolithiasis among patients with autosomal dominant polycystic kidney disease (ADPKD) is greater than in the general population (Grampsas, et al. (2000)]. ARPKD mostly diagnosed in utero or early after birth. ARPKD characterized by echogenic very large kidney and oligohydramnios (Halvorson CR, Bremmer MS & Jacobs SC 2010). Having a parent with the disease is the major risk factor for PKD. Gene mutation plays a role in about 10% of patients. Both, men and women are affected equally by PKD (The Kidney Foundation of Canada). The overall rate of polycystic kidney disease among ESRD Sudanese patients is 0.89% [Elsharif. and Elsharif, (2011)]. Autosomal dominant polycystic kidney disease (ADPKD) is the main hereditary cause of ESRF in Sudan (4.7%); while in Europe

represent 9.8% of causes among patients receiving renal replacement therapy. ADPKD is the fourth cause of ESRD among Sudanese patients [Banaga, et al. (2015)].

Double ureter and duplex system: Complications of double ureter and duplex system include obstruction of the collecting system, lithiasis, ureterocele, and vesicoureteral reflux; and these may end into end-stage renal failure. So, early detection of them is highly needed for better management [Prakash, et al, (2011)]. Certain congenital renal and urinary tract abnormalities show family relationship. These abnormalities include incomplete and variable penetrance [(Alpay, et al, (2013)]. The global rate of occurrence of ureteral duplication is 1%. It can be complete or incomplete duplication; the first type is mostly insignificant while the second type may cause problems. Complete duplication is associated with vesicoureteral reflux, ectopic ureterocele, or ectopic ureteral insertion, all of which are more common in girls than in boys; and ureteropelvic junction obstruction which occurs only in the lower pole and is seen in boys more than girls. Depending on the effective management, the final outcome of duplicated ureter, ureteral ectopia, or ureterocele is very good [Gatti and Cendron, (2017)]. In a study done in USA, 20 families (66%) had affected first-degree relatives. Conjoint ureter is more common than complete ureteral duplication [Rodriguez, (2014)]. There is an association between duplex system and ureteropelvic obstruction, reflux, and infection [Rodriguez, (2014)]. Duplication of ureter may lead to urinary tract infections and/or renal stones with its complications [Ojha and Prakash. (2016)]. Insertion anomalies of ureters, ureteral muscular hypertrophy, peripelvicalyceal fibrosis, and abnormal blood vessels crossing over the ureter or renal pelvis are the main causes of ureteropelvic junction (UPJ) obstruction [Rodriguez, (2014)].

Obstructive nephropathy: The most common cause of lower urinary tract obstruction in male infants and fetuses is the posterior urethral valve [Rodriguez, (2014)]. About 40% of ESRD in Arab World is due to obstructive uropathy [Shaheen, and Al-khader, (2005)]. Obstructive uropathy, which is one of the important preventable non-communicable diseases that lead to ESRD, is a cause of about 40% of ESRD among patients in some Arab countries [Elamin, et al, (2012)].

Rate of obstructive uropathy among ESRD Sudanese patients is 9.6%. The incidence rate of ESRD in Sudan range 70-140/million/year. Studies done in 1987 and 2009 indicated that obstructive nephropathy is the second cause of CKD in Sudan. A cause of

about 11.6% of ESRD among Sudanese patients is due to obstructive nephropathy. Obstructive uropathy (9.6 %) is the third cause of ESRD among Sudanese patients. The disease affects the productive age group [Banaga, et al. (2015)]. The overall rate of obstruction, and reno-vascular is 11.61%, and 0.45% respectively. The outcome of slowly progressive kidney damage is End-stage renal disease (ESRD). So, early detection and intervention may improve the situation and prevent the development of ESRD. The incidence rate of ESRD in the Middle East Countries is 93/million population/year; while in Sudan is about 70–140/million population/year. Obstructive nephropathy is the second cause of ESRD among Sudanese patients [Elsharif. and Elsharif, (2011)].

Renal dysplasias and agenesis: They are a spectrum of severe malformations that represent the primary diseases requiring dialysis and transplantation in the first years of life. There is failure of the development of nephrons as well as failure of the branching of the ureteric bud; leading to complete failure of formation of collecting ducts. Sometimes these abnormalities result in involution of the kidneys and renal agenesis. Renal agenesis may also result from failure of the ureteric bud to contact and/or induce the metanephric mesoderm. The frequency of bilateral renal agenesis is 1/10,000 births, and it leads to renal failure [Sadler, (2012)].

Knowledge about the causes of end-stage renal disease is highly needed by nephrologists for planning preventive measures. About 53.57% of causes of end-stage renal diseases among patients, attending Gezira Hospital for renal disease, for hemodialysis are unknown. Usually end-stage renal disease result from progressive slowly damage to kidney [Elsharif, and Elsharif, (2011)].

Renal disease is asymptomatic, so, it is detected late where medical interventions are ineffective. The course of such disease can be changed or stopped by early therapeutic interventions [Elsharif, and Elsharif, (2011)]; [Rodriguez, (2014)].

Problem identification and Justification: There is a gap of knowledge regarding congenital anomalies of urinary system among Sudanese Patients on hemodialysis attending Gezira Hospital for Renal Diseases & Surgery, Gezira State, Sudan. So, this study was conducted using ultrasound; to determine the prevalence of these anomalies and their relation to hemodialysis; in order to help physicians, patients and their families for planning to obtain better outcome of interventions. It was done during the period from February to April 2018.

Aim of the study: To investigate the occurrence of congenital anomalies in the urinary system among Sudanese Patients with ESRD on regular hemodialysis attending Gezira Hospital for Renal Diseases & Surgery, Gezira State, Sudan; using ultrasound; during the period from February – April 2018.

Methods:

Study design: It was an observational cross-sectional facility base study.

Study area and setting: This study was conducted at Gezira Hospital for Renal Diseases & Surgery, Gezira State, Sudan. It serves the Central States (Gezira, Sinnar, Blue Nile, and White Nile) and Eastern States (Elgedarif and Kassala) of Sudan. So, the total population served by this hospital is about 7,000,000. The hospital consists of six wards divided among two departments: Department of Urology and Surgery and Department of Nephrology (Medical ward - Surgical ward - Intensive care ward) in addition to a department of hemodialysis and peritoneal dialysis. Also, the hospital has a department of operations consisting of two sections for minor and major operations, part for kidney transplantation and another for other operations and operations performed by Gastrointestinal Endoscope. The capacity of the hospital is 80 beds divided equally between urology and nephrology departments. It has an intensive care for kidney transplant patients. Number of dialysis machine is 29; out of which the working machines are only 24. Five of the working machines are isolated for patients infected with HBV and HCV.

Study populations and sampling: They were Sudanese patients with ESRD on regular hemodialysis (HD) at the Gezira Hospital for Renal Diseases and Surgery. Fifty respondents were selected.

Data collection methods and techniques: Presence of the congenital anomalies in the urinary system was determined using abdominal ultrasound. Personal

data (age, residence, occupation, age of onset and duration of dialysis) was obtained by direct interview using a questionnaire.

Data management and analysis: Data was managed and analyzed using SPSS version 21. *P-value* ≤ 0.05 was considered statistically significant.

Ethical Concern: Ethical approval was obtained from Gezira University Ethical Board and from Gezira Hospital for Renal Diseases and Surgery, and then patients consent was taken.

Results:

Gender composition of the study population was 31 (62%) male and 19 (38%) female. The minimum age was 19 years while the maximum age was 70 years. The mean age was 45.10 years.

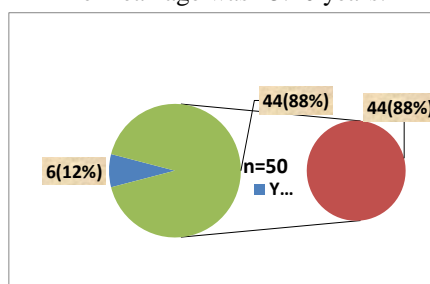


Figure 1: Presence of congenital anomalies of the renal system among patients on Hemodialysis at Gezira Hospital for Renal Diseases and Surgery, Sonographic Study; Gezira State, Sudan; 2018. (n = 50)

The percentage of the presence of the congenital anomalies among the study population was 8% (4 cases) (Figure 1).

Table 1: Types of congenital anomalies of the renal system among patients on Hemodialysis at Gezira Hospital for Renal Diseases and Surgery, Sonographic Study; Gezira State, Sudan; 2018 (n = 50)

| Types of congenital anomalies | Frequency | Percent |
|--|-----------|------------|
| Polycystic Kidney Disease | 3 | 6 |
| Uretero-pelvic junction (UPJ) obstruction – right kidney | 1 | 2 |
| Hypogenesis – left kidney | 1 | 2 |
| Agenesis – right kidney | 1 | 2 |
| Not applicable | 44 | 88 |
| Total | 50 | 100 |

The type and the percentage of the present congenital anomalies of the renal system among the respondents were found to be as follows: polycystic kidney disease 3 (6%), uretero-pelvic junction (UPJ) obstruction – right kidney 1 (2%), hypogenesis of the left kidney 1 (2%), and agenesis of the right kidney 1 (2%) (Table 1)

Table 2: Age of respondents with congenital anomalies of the renal system when first diagnosed, among patients on Hemodialysis at Gezira Hospital for Renal Diseases and Surgery, Sonographic Study; Gezira State, Sudan; 2018; (n = 50)

| Types of congenital anomalies | Age at 1 st diagnosis (years) | Duration of the disease (years) | Age at the study time (Present) |
|---|--|---------------------------------|---------------------------------|
| Polycystic kidney disease | 40 | 5 | 45 |
| | 13 | 16 | 29 |
| | 13 | 16 | 29 |
| Uretero-pelvic junction (UPJ) obstruction | 31 | 11 | 42 |
| Hypogenesis | 20.5 | 2.5 | 23 |
| Agenesis | 51 | 10 | 61 |

Age of patient with congenital anomalies of the renal system when diagnosed for the first time was found to be: 40 years and 13 years for polycystic kidney disease, 31 years for Uretero-pelvic junction (UPJ) obstruction, 20.5 years for hypogenesis and 51 years for agenesis (Table 2).

Table 3: The relation between gender and presence of congenital anomalies of the renal system among patients on Hemodialysis at Gezira Hospital for Renal Diseases and Surgery, Sonographic Study; Gezira State, Sudan; 2018; (n = 50)

| Gender | Gender and presence of congenital anomalies of the renal system | | Total | Chi-square test |
|--------|---|----|-------|-----------------|
| | Yes | No | | |
| Male | 4 | 27 | 31 | 0.802 |
| Female | 2 | 17 | 19 | |
| Total | 4 | 46 | 50 | |

The overall distribution of the presence of the congenital anomalies of the renal system among different genders of the respondents is four for male and two for female. *Chi-square* test (0.802) was insignificant at 0.05 level. So, there is no relation between the presence of the congenital anomalies of the renal system and genders of the respondents (Table 3).

Table 4: The relation between gender and types of congenital anomalies of the renal system among patients on Hemodialysis at Gezira Hospital for Renal Diseases and Surgery, Sonographic Study; Gezira State, Sudan; 2018; (n = 50)

| Gender | Gender * Types Of Congenital Anomalies Of Renal System | | | | | Total | Chi-square |
|--------|--|--|-------------|----------|----------------|-------|------------|
| | Polycystic Kidney Disease | Ureteropelvic junction (UPJ) obstruction | Hypogenesis | Agenesis | Not applicable | | |
| Male | 2 | 1 | 0 | 1 | 27 | 31 | 0.576 |
| Female | 1 | 0 | 1 | 0 | 17 | 19 | |
| Total | 1 | 1 | 1 | 1 | 46 | 50 | |

The distribution of the types of congenital anomalies of the renal system among genders of the respondents was found to be as follows: Male (Polycystic kidney disease, Ureteropelvic junction (UPJ) obstruction, and agenesis); female (Polycystic kidney disease, and hypogenesis). *Chi-square* test (0.576) was insignificant at the level of 0.05. So, there is no relation between the type of the congenital anomalies of the renal system and gender of the respondents (Table 4).

Discussion: A cross sectional facility base study was done among Sudanese patients with ESRD on regular

hemodialysis at Gezira Hospital for Renal Diseases and Surgery. Fifty respondents were selected. About 62% of the respondents were male. This was similar to what is reported by EBSCO (2016) that male is more prone to hemodialysis than female.

The overall prevalence of the congenital anomalies of the renal system among the respondents was 12%. Four types of congenital anomalies of the renal system were encountered and these are: polycystic kidney disease, uretero-pelvic junction obstruction, hypogenesis, and agenesis. The contribution of

polycystic kidney disease to ESRD among our study population (2%) is nearly similar to what is reported (2.5%) by Halvorson, et al, (2012). The rate of polycystic kidney disease among ESRD Sudanese patients is reported by Banaga, et al, (2015) to be 4.7%.

Gender distribution of types was as follows: Male (Two cases of polycystic kidney disease, one case of Ureteropelvic junction obstruction, and one case of agenesis); female (One case of Polycystic kidney disease, and one case of hypogenesis). There is no relation between the presence and types of the congenital anomalies of the renal system and genders of the respondents. Nationally, it is reported by Elsharif, and Elsharif, (2011) that the overall rate of obstruction, and polycystic kidney disease, among ESRD Sudanese patients is 0.89%, and 0.45% respectively. Internationally ureteropelvic junction obstruction which occurs only in the lower pole is seen in boys more than girls [Fernbach, et al, (1997)]. In the current study the UPJO was observed only among male and this was consistent with that of Fernbach, et al, (1997). The observed discrepancy between our findings and that of Elsharif, and Elsharif, (2011) may be due to the sampling and sample size together with the study populations and methods of diagnosis.

Congenital anomalies of the renal system were observed only among respondents from the Central State. This may be due to the location of the hospital together with the increased number of respondents from the Central State.

Strikingly there was strong statistical relationship between the duration of the disease and both of presence and type of congenital anomalies of renal system among respondents.

Conclusion: Four types of congenital anomalies of the renal system were encountered among the respondents and these are: polycystic kidney disease, uretero-pelvic junction obstruction, hypogenesis, and agenesis. The overall prevalence of the congenital anomalies of the renal system among the respondents was 12%.

Recommendations: Screening for the presence of the congenital anomalies of the renal system among the general population of the Central State may be of great help. Another research among patients on hemodialysis, with good coverage may help to give a clear picture about the situation.

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