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Open Heminephrectomy in Unilateral Duplex Collecting System and Complete Ureteral Duplication with Poorly Functioning Upper Pole: A Case Report

Yohanes Adinugroho[1*], L. Bobby Nindra Nugraha[1], T. Setya Anton[2] and Syaeful Agung Wibowo[2]

[1]General Surgery Department, Sebelas Maret University, Indonesia.
[2]Urology Department, Dr. Moewardi General Hospital, Indonesia.

Authors' contributions

This work was carried out in collaboration among all authors. Author YA designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors LBNN and TSA managed the analyses of the study. Author SAW managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Aim: We report the results and experience of the Moewardi Hospital to do Physical Examination with Narcose and Heminephrectomy For Surgical Treatment Procedures Double Collecting System
Materials and Methods: Reported a 2 years old girl with double collecting system ren right with hydronephrosis upper moiety right (minimal function) + dextra hydroureter + ureterocele dextra and diagnosis of complications of chronic cystitis ureter and UK fitted. Furthermore, retrograde pyelography (RPG) was performed to obtain information on which ureteral estuary originated from superior moiety (pole) and inferior renal moiety as well as evaluation of whether or not a per-endoscopic incision was required and the boundaries of the area to be heminephrectomy. Furthermore, a heminephrectomy was planned in the upper moiety right area.
Results: Radiological examination is known to have a role in determining the presence or absence of ureteral duplication and can be done since the prenatal period. Ultrasound examination at about

**Corresponding author: E-mail: yohanestn18@gmail.com;*

20 weeks of gestation can detect the presence or absence of abnormalities in the fetal urinary tract. superior polus heminephrectomy in a system with duplicated malfunctioning, through laparoscopic ureteric ligation, or ureteral reimplantation if renal function remains good is the best management in children or patients with symptoms to resolve symptoms such as incontinence, prevent complications and maintain renal function.

Conclusion: The double collecting system is a form of congenital abnormality of the urinary system. Radiological examination is known to have a role in determining the presence or absence of duplication of the ureter and can be done since the prenatal period. Management is carried out based on the symptoms experienced by the patient and by looking at kidney function. Ureteroneocystostomy is the definitive surgical management of vesicoureteral reflux caused by ureteral duplication. If there is a case of a double collectivus system and with an ectopic ureter, then the procedure that can be given is a distal ureteroureterostomy through a laparostomy on a double J stent.

Keywords: Narcose; heminephrectomy; double collecting system.

1. INTRODUCTION

The kidney / ren is an organ that plays an important role in the urinary system. This organ is located at the level of the thoracal vertebrae 12 to lumbar 3 and is an organ that is located retroperitoneally. In the medial part there is a structure called the renal hilus which is the structure of the entry of the renal artery and veins, innervation, lymphatic system, and ureter. The kidneys are also protected by a layer of fat which form of a capsule called the renal adipose capsule and underneath it is another layer, namely the renal fibrosus capsule. The layer of the kidney consists of the so-called renal cortex, renal medulla and renal pyramid. The renal column is a structure that separates the renal pyramids. There is a structure at the end of the renal pyramid called the calyces minors, the combination is called the calyces major, then forms the renal pelvis [1].

Renal vascularization depends primarily on the renal arteries, dividing into segmental arteries and then branching into interlobar arteries. The venous vascularization of the kidneys is similar to that of arteries, but in the opposite direction. The kidneys are composed of the smallest units or structures called nephrons [2].

After the kidneys, the ureters are another structure that plays a role in the human genitourinary system. The ureters from the renal hilum to the bladder and are located retroperitoneum. Ureter functions in carrying urine from the kidneys to the vesica urinaria [2].

The vesica urinaria or urinary bladder is a bag shaped structure composed of several layers of muscularis. In men, there is a structure, namely the prostate gland that surrounds the inferior end of the vesica urinaria. Musculus detrusor is the muscularis layer which functions to regulate the function of emptying urine [2].

The urethra is a thin-walled duct-shaped structure that carries urine in the same way as the urethra, which is peristaltic [2].

The urinary tract system has a major function in the excretion process. Excretion is the process of separating substances which is metabolic waste from body fluids. This metabolic waste is a substance that is not needed or excess is produced from the body. Urine itself contains various metabolic waste, toxins, and drug metabolism products. The formation of urine in the urinary tract occurs in four stages: filtration, reabsorption, secretion, and water augmentation [1].

After the capacity of the bladder is exceeded, the afferent activity created by the pressure, volume, and nociceptors receptors is conveyed through the A□ and C fibers via the pelvic nerves and pudendus nerves to the sacral fibers of the spinal cord. This signal then goes to the mics center in the rostral pons. This signal is then processed by the cerebral cortex and other parts of the brain and the micturition process occurs through the process of delivering signals from the cerebral cortex down to the parasympathetic part of the sacral nerve and inhibition of sympathetic and somatic activity. The first stage that occurs is relaxation of the external urethral sphincter and then regular contraction of the bladder, which then causes the discharge of urine [3].

Duplication of the renal collecting system, also termed duplex kidney, is one of the few renal

anomalies more common in females. Duplex kidneys have an upper pole and a lower pole, called moieties, each drained by a ureter. Approximately one-third of the duplex kidney is drained by the upper pole ureter, with two-thirds drained by the lower pole ureter.

Many patients with duplex kidneys are asymptomatic and have no impairement of renal function. Those who are of concern before birth or after delivery (postnatal) usually have complications related to abnormal implantation of one or both ureters. The upper pole ureter can form ureterocele within the bladder causing obstruction, resulting in little or no function of the upper pole. The lower pole ureter may have short intravesical segments that produce vesicoureteral reflux (VUR). Duplex systems that have this complication also have ultrasound (US) findings that facilitate their prenatal detection. Clinically significant duplication of the collecting system may manifest prenatally with hydronephrosis of the upper pole and dilatation of the upper polar ureter, hydronephrosis of the lower pole, or ureterocele within the bladder [4].

2. CASE PRESENTATION

A girl with the initials A, age 2, came to the Urological Surgery Clinic with a referral from a pediatric colleague on November 21, 2018 with a diagnosis of chronic cystitis and right hydronephrosis. The patient was referred by a pediatric colleague with a history of recurrent UTI with chronic cystitis and findings of right hydronephrosis on imaging studies. The patient has a history of recurrent urinary tract infections. Patients are often hospitalized from 4 months of age due to recurrent UTIs. There is no history of fever. Painful stools are sometimes felt. There was no hematuria. The patient can still hold the stool when going to the bathroom. History of the right stomach enlarged since 2 months ago, no pain. From the patient's ANC history, the mother's age at delivery was 32 years; routine ANC control at doctor Sp.OG. Routine ultrasound was performed during ANC but there was no notification of antenatal hydronephrosis. The patient was born by section caesarea procedure, as indicated by DKP, birth weight 3 kg, birth length 50 cm; the patient had no history of intensive care (NICU) in the child at birth or a history of other congenital abnormalities.

From the physical examination, it was found that the patient looked moderately ill, GCS E4V5M6 compost mentis with a body weight of 13 kg, height 88 cm, heart rate 100 x / minute, respiratory rate 28 x / minute, temperature 36.7 C. normal. From the patient's abdominal examination, it was found that there was bulging in the right abdomen on inspection, normal bowel sounds on auscultation, abdomen supple, no tenderness, obtained ballottement, percussion obtained tympanic sound in all areas of the abdomen. From the urological status in the right flank region, bulging and ballottement were obtained; costovertebral knock pain was not obtained. Examination of the suprapubic region and external genitalia was within normal limits.

From blood laboratory and hormone laboratory tests, results were obtained according to Tables 1 and 2. The patient's hormone laboratory results tended to be within normal limits.

From the clinical, laboratory, and ultrasound manifestations, the patient was diagnosed with a double-collecting system of the right ren with extreme upper moiety (minimal function) hydronephrosis + extra hydroureter + right ureterocele and a diagnosis of complications of chronic cystitis.

Then planned evaluation cystoscopy on January 14, 2019 to determine the position of the ureter estuary and plug the UK. The patient was co-treated with pediatric colleague. Furthermore, Retrograde Pyelography (RPG) was performed to obtain information on which ureteral estuary originated from superior moiety (pole) and inferior renal moiety as well as evaluation of whether or not a per-endoscopic incision was required and the boundaries of the area to be heminephrectomy. Furthermore, a heminephrectomy was planned in the upper moiety right area.

Operation report dated January 14, 2019

Preoperative diagnosis: Double collecting system right ren with right upper moiety hydronephrosis (minimal function) + right hydroureter + right ureterocele.

Postoperative diagnosis: Double collecting system, right ren with right upper moiety (minimal function) hydronephrosis + right hydroureter + right ureterocele.

1. Patients with lithotomy in GA, antisepsis, then drapping.
2. Perform physical examination: vestibule: urine seepage (-) lumps (-). No visible

orificium, other than urethra externa urificium & vaginal interoitus, urine seepage (-)
3. Insertion of 10 Fr cystoscopic sheath into OUE, performed a non-hyperemic bladder mucosal cystoscopy, trabeculation (-), sacculation (-), diverticles (-), stones (-). The mouth of the right ureter is shown in its normal position in the trigone, along the ureteric ridge, and the mouth of the left ureter is in its normal position in the trigone. There is a bladder neck with ureterocele on its posterior wall until the lumen of the urethral sheath is withdrawn for urethroscopy, an extopic ureter opening is seen in the distal urethral sphincter externa at 6 o'clock, insertion of tube 3.5 Fr
4. Attach the FC 10 fl balloon to the 3 ml tube fixed to the FC
5. Operation is complete

Partial ren dextra nephrectomy (upper moiety ren dextra)

1. Lithotomy patients, field surgery antisepsis, drapping
2. Cytoscopy of the mouth of the right ureter in the trigone (+), insertion of feeding tube 3.5 Fr to NGT 5 Fr, difficult to enter, insertion of NGT 5 Fr through the mouth of the ectopic ureter at 6 o'clock distal the sphincter can enter up to the proximal, attach FC 10 Fr balloon 4 ml, fixed NGT on FC
3. The patient's position was changed to right lumbotomy, a & operative field antisepsis, XI-XII intercostal incision, penetrating cutis, subcutis, fascia MOE, MOI, MTA. Peritoneal fat is visible, fascia gerata is opened, identification of ureter from upper morety (contains NGT in it & hydroureter), sealed and released until proximal
4. Followed by partial upper morety nephrectomy & ureterectomy of the upper morety as much as possible. Simultaneous controlled bleeding abandoned spongestant & NGT drain 12 Fr
5. The operation wound was closed layer by layer
6. NGT in the ectopic ureter at aff
7. Operation over

Table 1. Laboratory results of the patient's blood (22 November 2018)

Examination	Result	Unit	Reference
Routine Hematology			
Hemoglobin	12.9	g/dL	11.5 – 13.5
Hematocrit	39	%	34 – 40
Leukocytes	9.5	Thousand /µl	5.5 – 17.0
Platelets	298	Thousand /µl	150 – 450
Erythrocytes	4.79	Mil/µl	3.90 – 5.30
Creatinine	0.4	mg/dl	0.9-1.3
Ureum	17	mg/dl	<50
Erythrocyte Index			
MCV	81.7	/um	80.0 – 96.0
MCH	**26.9**	Pg	28.0 – 33.0
MCHC	33.0	g/dl	33.0 – 36.0
RDW	**10.9**	%	11.6 – 14.6
MPV	8.1	Fl	7.2 – 11.1
PDW	**16**	%	25 – 65
Cell Count			
Eosinophils	**3.60**	%	1.00 – 2.00
Basophils	0.20	%	0.00 – 1.00
Neutrophils	**24.10**	%	29.00 – 72.00
Lymphocytes	**67.30**	%	22.00 – 44.00
Lymphocytes	4.80	%	0.00 – 6.00
Blood chloride			
Blood sodium	137	mmol/L	132 – 145
Blood potassium	4.3	mmol/L	3.1 – 5.1
Blood chloride	107	mmol/L	98 – 106
Calsium ion	1.31	mmol/L	1.17 – 1.29
Serologi			
HBsAg	Nonreactive		Nonreactive

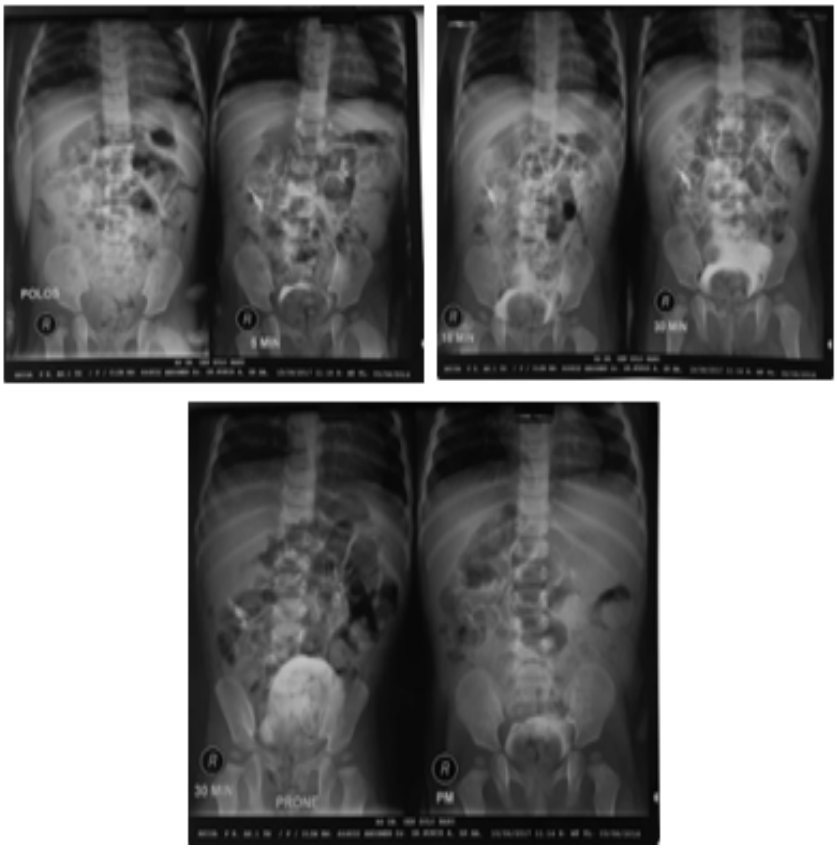

Fig. 1. BNO + IVP Patient (15 June 2017)

Impression: The function of the right and left kidneys was within normal limits. Peviocalyceal view of the right kidney is depressed inferiorly to the suspected cyst on the upper pole of the right kidney. The bladder emptying function is within normal limits

Table 2. Laboratory results of patient urine (16 October 2018)

Urine Macroscopic			
Color	Light yellow		
Chemical Urine			
Specific gravity	**1.010**		1.015-1.025
pH	7.0		4.5-8.0
Leukocytes	**Positive**		Negative
Nitrite	Negative		Negative
Protein	Negative	mg/dl	Negative
Glucose	Normal	mg/dl	Normal
Ketones	Negative	mg/dl	Negative
Urobilinogen	Normal	mg/dl	Normal
Bilirubin	Negative	mg/dl	Negative
Blood	Negative	mg/dl	Negative
Urine Microscopic			
Leukocytes	**20-25**		
Eritrocytes	**0-1**		
Bacteria	**+**		
Crystal	Negative		
Epithelium	**+**		

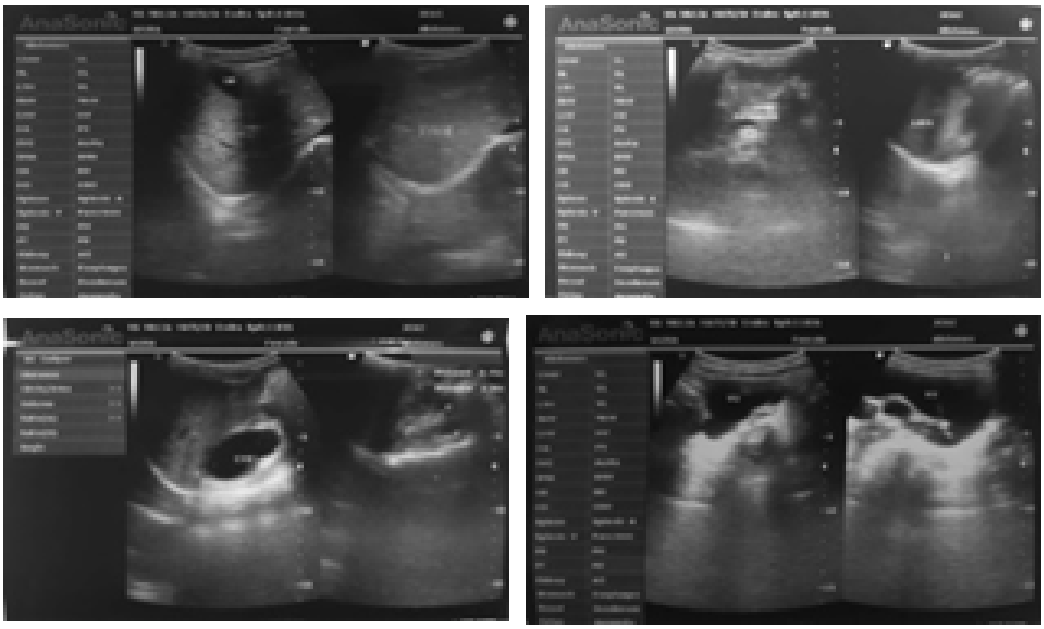

Fig. 2. Ultrasound of the patient's abdomen (20 October 2018)
Impression: right kidney cyst, diameter> 5.7 cm Multiple left kidney cysts vary in size VU diverticles with chronic cystitis Bilateral kidney multicysticism; chronic cystitis with VU diverticles

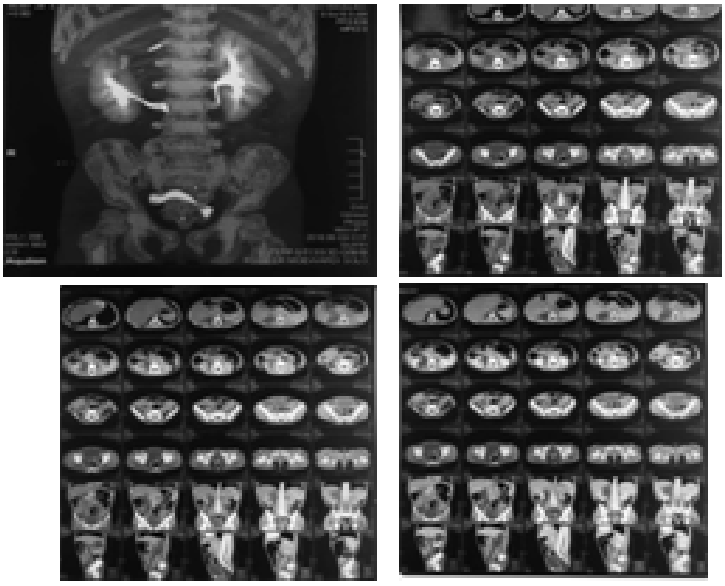

Fig. 3. CT scan of the patient's abdomen (23 November 2018)
Impression: double collecting system right ren accompanied by hydroureter 1/3 proximal to distal and uretocele in right ureter pars distalis; bilateral inguinal lymphadenopathy

Operations have been carried out in accordance with standard procedures and maintain hygiene. Performed periodic examinations by the surgical urology department and the pediatric department to maintain the condition of the postoperative patient. All image taking and publication of data were approved by the patient with evidence of signing the informed consent sheet by the patient's guardian and kept as part of the patient's medical record.

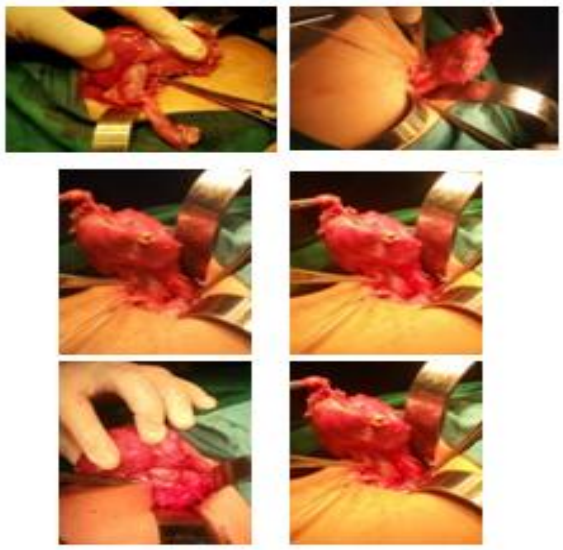

Fig. 4. Photo of cystoscopic durante

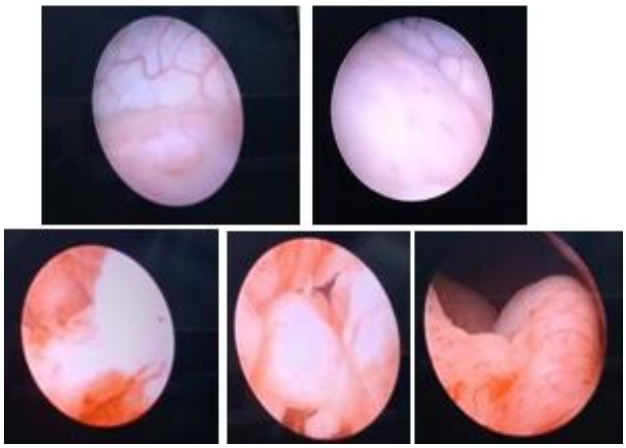

Fig. 5. Ren dextra upper pole moiety durante heminephrectomy photo

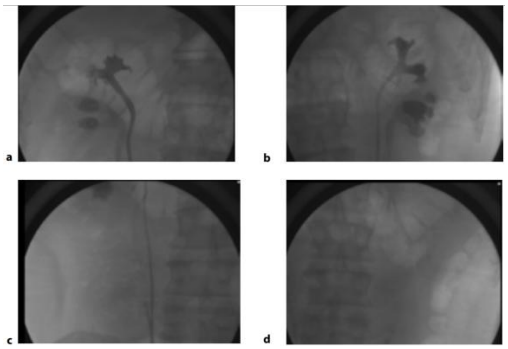

Fig. 6. Duplication of bilateral collectivus systems with duplication of complete left ureter (b, d) and duplication of partial ureters (a, c) [5]

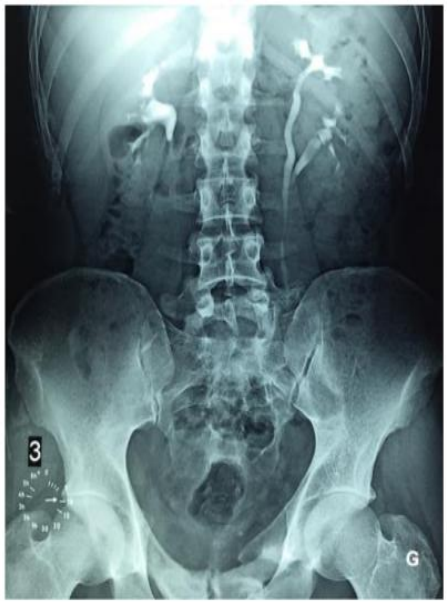

Fig. 7. Duplication of the left ureter [6]

3. DISCUSSION

It was reported that a 2-year-old girl patient entered the Urological Surgery Clinic with a referral from pediatric colleague with a diagnosis of chronic cystitis and right hydronephrosis. The patient had a history of recurrent UTI with chronic cystitis and findings of right hydronephrosis on imaging. History of the right side of the abdomen was enlarged since 2 months ago, was painless. Based on physical examination, there was a bulging of the right flank region accompanied by ballottement. In this patient, upper moiety ren dextra heminephrectomy was performed.

The urinary tract system consists of two parts: the upper and lower urinary tracts. The urinary tract consists of the kidney / ren and ureter while the lower urinary tract consists of the vesical urinary tract and the urethra. The function of the urinary tract in general is to filter waste components from the blood, regulate the balance of water, electrolytes, and acid-base in the body [1,2].

The double collecting system is a form of congenital abnormality of the urinary system [7]. This duplication occurs due to premature separation of the remaining Wolferii duct. Partial duplication occurs when the metanephron network is not completely separated, resulting in multiple lobules and overlapping collectivus tubules. Complete or incomplete duplication occurs when in the early stages of human growth there are two ureteric buds [5].

Congenital abnormalities of the kidneys and urinary tract occur in about 3-6 out of 1000 people. Duplex kidney disorders are more common in women than in men and are more common in the left ren than in the left ren. Congenital abnormalities are more common in children than adults. Bilateral duplication abnormalities (in both kidneys) are quite common, namely 20-40% of all duplication cases6. This duplication is believed to be autosomal and particularly prevalent in Caucasians [6].

The ureteric bud structure is the diverticulum of Wolfii's mesonephric duct. Overexpression of the GDNF (glial cell-derived neurotrophic factor) - RET signaling pathway induces multiple ureteral buds and results in different insertion of the pelvicocalyceal system. It is known that the absence of type II angiotensin receptors can create urinary tract anomalies and especially ureteral duplication [8].

Duplication of the ureter is known to occur most frequently in the renal pelvis, that is, in cases of incomplete duplication. Other locations for duplication are in the middle and distal third of the ureter. In these two ureters, peristaltic movements continue to occur resulting in disruption of urine flow to the bladder. In contrast to incomplete duplication, complete ureteral duplication is not interconnected, that is, there are pathways of entry in the renal pelvis and bladder that are different from one another [9].

Radiological examination is known to have a role in determining the presence or absence of duplication of the ureter and can be done since the prenatal period. Ultrasound examination at about 20 weeks of gestation can detect the presence or absence of abnormalities in the fetal urinary tract. If there is duplication of the ureter, it may reveal unequal renal size, abnormal renal parenchymal contours, and distinct dilatation of the upper and lower pelvicocalyceal poles. Another examination is voiding cystouretrography (VCUG / Voiding Cystourethrography) which is often performed on children to determine whether there is urine reflux that can be caused by ureterocele. The use of IVP and CT-Scan can be used, but is not the main test of choice to determine the presence or absence of urinary tract anomalies [8].

Management is carried out based on the symptoms experienced by the patient and by looking at kidney function. If symptoms such as urinary tract obstruction are not found and kidney function is normal, then surgical management is not necessary [5]. As for if surgery is needed, it can be through ureterectomy or superior polus heminephrectomy [10].

In children and patients with symptoms, the best management is surgery. This treatment is more useful for resolving symptoms such as incontinence, preventing complications, and maintaining kidney function and eliminating urinary tract infections. Forms of procedure that can be done are superior polus heminephrectomy in a duplicated system that does not function, through ligation of the ureter via laparoscopy, or ureteral reimplantation if kidney function remains good [10].

In kidneys that can still function normally, surgical techniques that can be performed are ureteroneocystostomy or distal and proximal ureterostomy [11].

Ureteroneocystostomy is the definitive surgical management of vesicoureteral reflux caused by ureteral duplication. This procedure can be done through a transvesicle approach, extravesicles, or a combination of both [11].

Ureteroureterostomy is a procedure that is often performed when there is either iatrogenic or penetrating ureteral trauma. Preparation before this procedure requires investigations of intravenous pyelography (IVP) or endoscopic examination. Patients who are acutely ill or who cannot be operated on by other measures can pass through a ureterorueterostomy with prior ureteric stents or percutaneous nephrostomy drainage. In this procedure, a vertical midline incision is made to allow a transperitoneal approach [12].

If there is a case of a double collectivus system and with an ectopic ureter, then the procedure that can be given is a distal ureteroureterostomy through a laparostomy on a double J stent. Another option that can be performed in children is superior polus heminephrectomy via laparoscopic retroperitenoscopy. This technique is safe to perform at any age, and can be performed on an inferior polus, requires shorter hospital stay, better cosmetic results, and does not cause complications [11].

4. CONCLUSSION

The double collecting system is a form of congenital abnormality of the urinary system. Radiological examination is known to have a role in determining the presence or absence of duplication of the ureter and can be done since the prenatal period. Management is carried out based on the symptoms experienced by the patient and by looking at kidney function. Ureteroneocystostomy is the definitive surgical management of vesicoureteral reflux caused by ureteral duplication. If there is a case of a double collectivus system and with an ectopic ureter, then the procedure that can be given is a distal ureteroureterostomy through a laparostomy on a double J stent.

CONSENT

As per international standard, parental written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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