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Clinical profile of duplex kidneys in children and its association with vesicoureteric reflux

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Abstract

Background Duplex kidney is a relatively common renal anomaly with a wide array of associations. With routine antenatal screening it is increasingly diagnosed these days. With this study, we aim to assess the clinical profile, prevalence of vesicoureteric reflux and highlighting the need for aggressive workup and management in selected cases to prevent further renal damage.

Results Fifty-three children with duplex kidneys who attended the department of pediatric surgery at our institution from December 2016 to December 2021 were included in the study. The most common mode of presentation was urinary tract infection (58%). Fifty-three percent had an antenatally detected renal anomaly (out of these 46% were asymptomatic, 33% had UTI and 11% had other symptoms like straining, incontinence and abdominal mass). Sixty-one percent of patients were conservatively managed and 19 patients needed surgical intervention. Ureterocele was the most common condition necessitating surgery. Prevalence of vesicoureteric reflux was 60% and was comparable with other studies. Thirty-six percent of patients had renal scarring. Among the patients with scarring, 84% had VUR and 63% had febrile UTI.

Conclusion Duplex kidney though a common congenital anomaly, requires a systematic evaluation to detect its various associations and a structured management protocol according to the associations. It has a high incidence of VUR which is a potentially hazardous condition which can affect the renal function if not evaluated and managed appropriately. Duplex can also be associated with obstructive conditions like PUJO, ureterocele, and ectopic megaureter which can cause rapid deterioration of renal function if not managed early enough. Though majority of the patients need only follow-up and antibiotic prophylaxis, there may be a certain sub group of patients who develop early scarring and need aggressive management. Our study aims to highlight the need to detect such patients early enough to reduce the morbidity.

Keywords Duplex kidneys, Vesicoureteric reflux, Ureterocele, Micturating cystourethrogram, Clinical profile

Background

Duplex collecting system is a common anomaly of the urinary tract and can either be complete or incomplete [1]. A duplex (duplicated) system is a kidney with two pelvicalyceal systems. Anomalies like ureterocoele and ectopic ureter mostly affects the upper moiety whereas anomalies like vesicoureteric reflux and pelviureteric junction obstruction mostly affects the lower moiety [2, 3]. Vesicoureteric reflux (VUR) is the most common association.

Duplications can be incomplete with the confluence of ureters above the ureteric orifice ranging from bifid pelvis to Y duplication or complete. Anthony Caldamon et al. describes incomplete duplication as a kidney with two ureters that fuse into a unit proximal to the bladder and then drains into the bladder through a single orifice [4]. Complete duplication refers to a kidney with two ureters that drain separately into or below the bladder.

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Duplex system arises due to an additional ureteral bud arising from the mesonephric duct and meeting the renal blastema at a separate site from the original bud [5]. In cases of complete duplication, the orifice of the upper moiety is characteristically located medial and caudal than the orifice of the lower moiety that is located cranial and lateral. This relationship is so consistent that it is called Weigert-Meyer rule, which is seen to be obeyed in more than 90% of the cases [6]. According to Sam D.

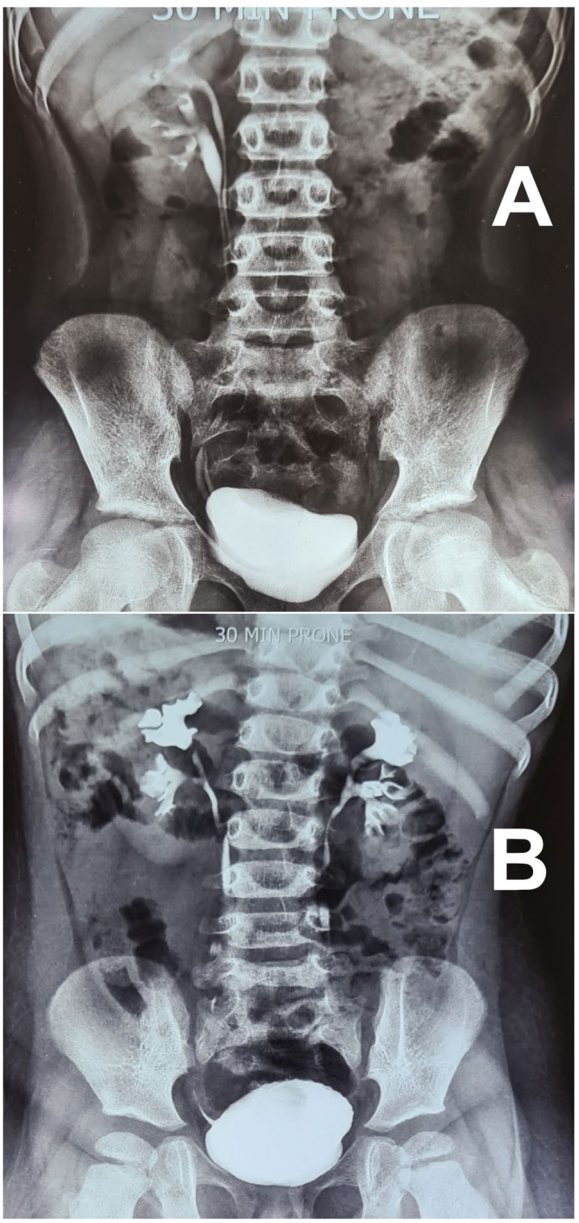

Fig. 1 **A** IVP film showing unilateral complete duplication on one side and non-functioning kidney on opposite side. **B** IVP film showing bilateral incomplete duplication

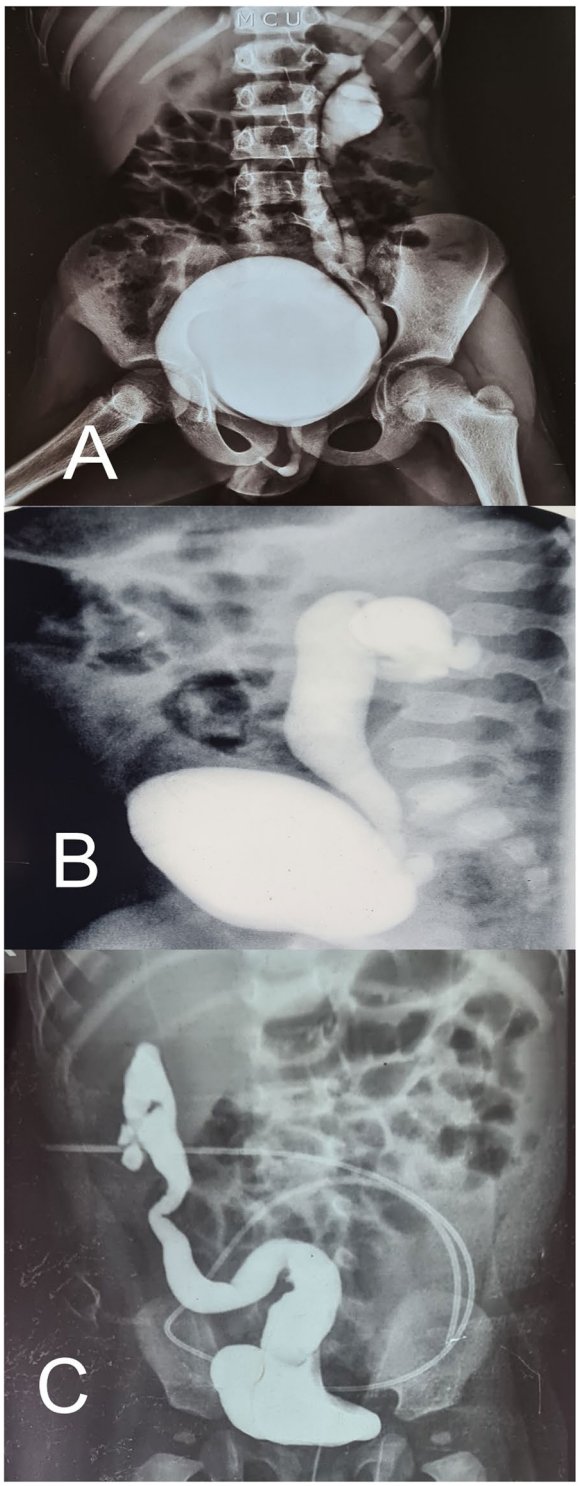

Fig. 2 **A** MCU showing reflux into left kidney with "Y" duplication. **B** MCU showing reflux into the lower moiety with "Drooping lily" sign. **C** Reflux into the upper moiety ureter with cystic dilatation of lower end

Graham Jr et al., ureteroceles are congenital cystic dilatations of the intravesical submucosal ureter [7].

Many of the duplicated units show congenital dysplasia (scarring) and hydronephrosis. There is an increased incidence of infection because both VUR and obstruction are much more common in duplicated systems [8] as stated by Mackie GG et al. in a study in 1975.

Duplex renal systems are being diagnosed with increased accuracy and at an early age these days due to the advent of newer and more sophisticated imaging modalities. Although a group of them are asymptomatic and incidentally detected, the symptomatic patients present right from the newborn age and pose a significant challenge in management. Delay in diagnosis and management can lead to renal dysfunction, recurrent infections and compromise the quality of life. So, it is important to identify this subgroup of patients who need early intervention. Since there are no clear cut guidelines for management, it should be individualised according to each patient.

Management can be challenging and is influenced by patient's age, presence of infection or incontinence, renal function, moiety involved, type of duplication, and surgeon's preference.

The aim of our study is to determine the clinical profile (age and mode of presentation, natural history, and severity) of duplex kidneys in children, identify the associated urological anomalies and to estimate the prevalence of VUR in children with duplex kidneys.

Methods

The primary objective of the study was to determine the clinical profile of Duplex kidneys in children below 12 years who presented between December 2016 and December 2021. The secondary objective was to estimate the prevalence of vesicoureteric reflux in children with duplex.and the tertiary objective was to identify the other anomalies in the duplex spectrum (ureterocoele, PUJO, ectopic ureter etc.).

Our study was a descriptive study on patients with duplex kidneys. All children from day 1 to 12 years of age who presented to the Department of Paediatric Surgery, at our institution from December 2016 to December 2021 with a diagnosis of duplex kidney in ultrasound or micturating cystourethrogram (MCU), were included in the study.

Patients with single system ureterocoeles and ectopic ureter, asymptomatic newborns with suspicion of duplex but have not completed investigations, patients with associated neurogenic bladder and other causes of lower urinary tract obstruction were excluded from the study.

Data collection was started after Institutional Review Board approval (IRB no.: 97/2021 dated 28/01/2022). Data was collected by reviewing the hospital inpatient

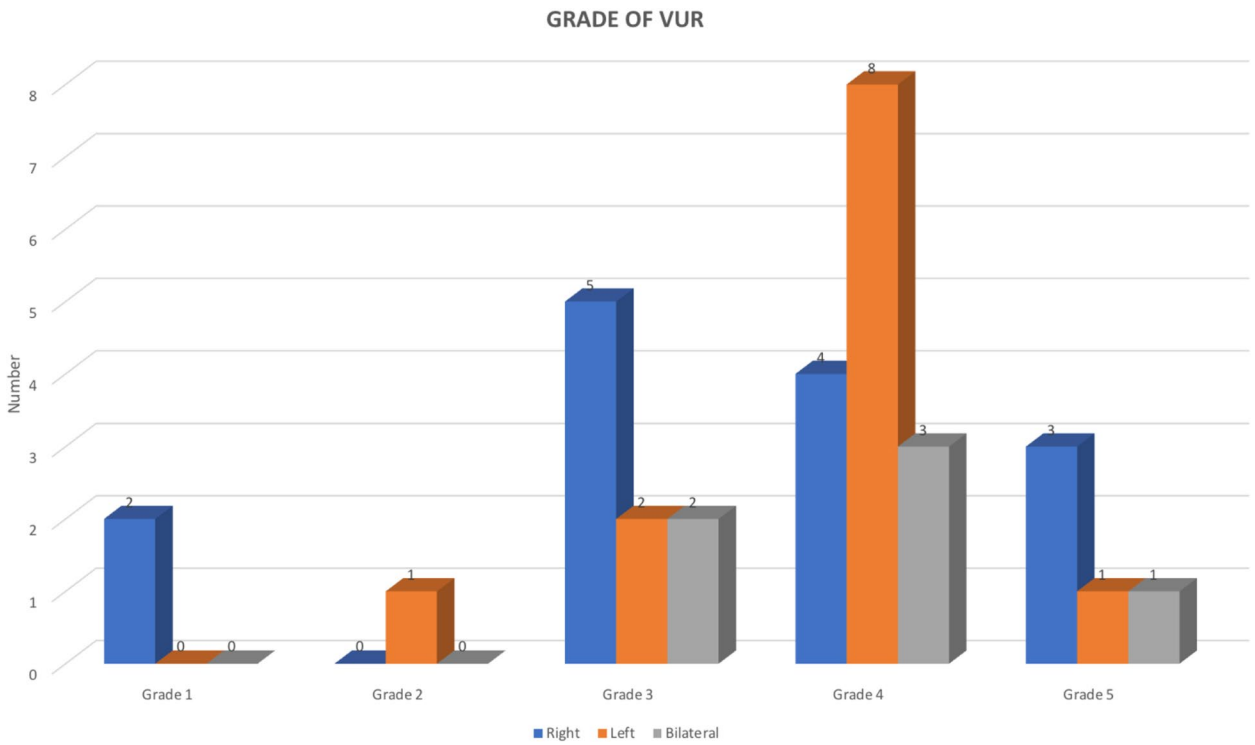

Fig. 3 Graph depicting the frequency distribution of different grades of VUR in our study

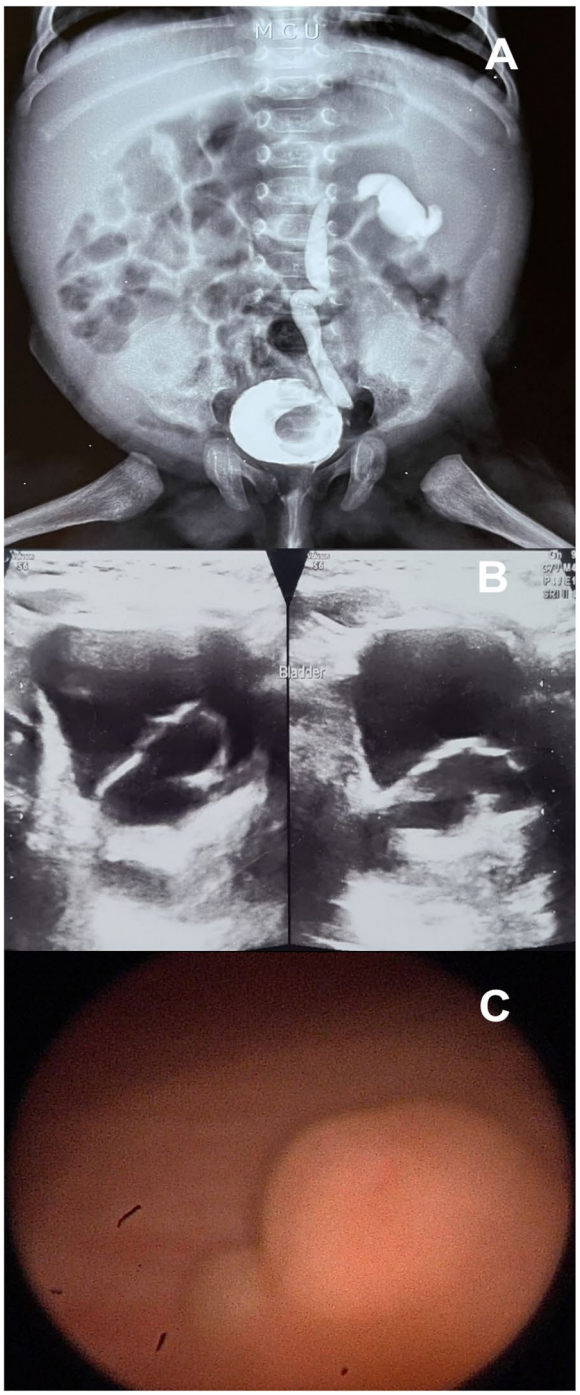

Fig. 4 **A** MCU showing ureterocele (*seen as filling defect in the bladder*) of the upper moiety with reflux into the lower moiety. **B** USG showing a ureterocele within the bladder. **C** Cystoscopy finding of a left sided ureterocele within the bladder

and out-patient records. Age, demographics, symptoms, clinical features, relevant urine and blood reports, ultrasound KUB, MCU, intravenous urogram, and treatment details were collected.

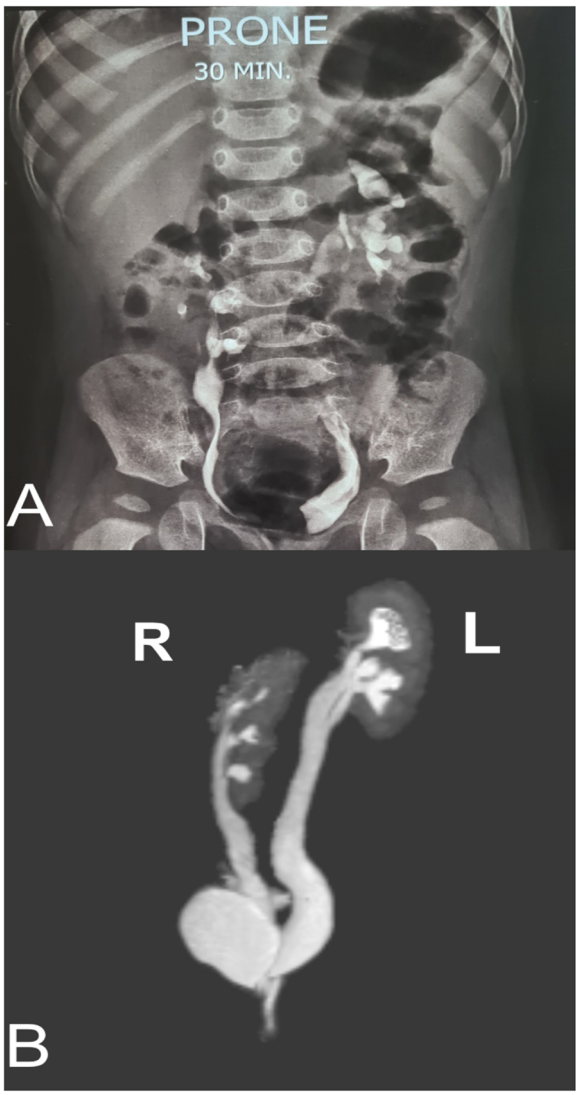

Fig. 5 **A** IVP image showing bilateral duplex with bilateral ectopic ureters draining outside the bladder.Bladder not opacified even after 30 min. **B** MRU image showing bilateral incomplete duplication with ectopic insertion of the ureters

The data was analyzed systematically and entered into Microsoft Excel v2007 for Windows. Descriptive analysis of the data was done. Since the data was predominantly categorical in nature, findings are presented using frequency and percentage.

Results

From December 2016 to December 2021 we encountered 53 cases of duplex kidney, out of which 45% ($n=24$) were males and 55% ($n=29$) were females. Fifty-seven percent ($n=30$) of patients presented before one year of age, 32% ($n=17$) belonged to the 1- to 5-year age group, 11% ($n=6$) of patients presented

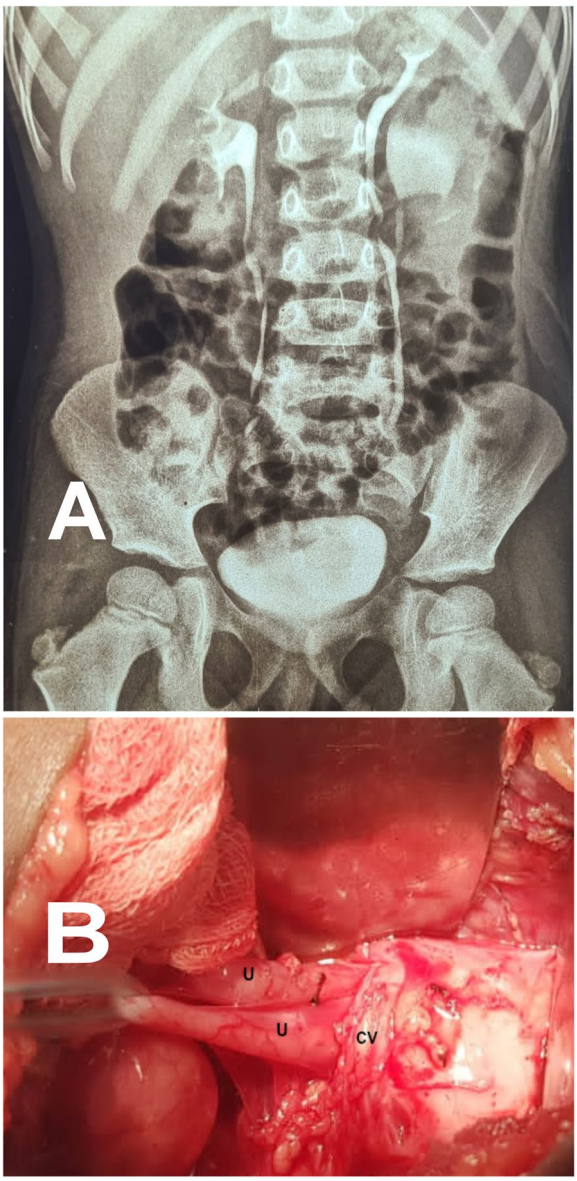

Fig. 6 **A** IVP image showing lower moiety hydronephrosis with PUJO. **B** Intraoperative image of the same patient demonstrating the crossing vessel causing lower moiety PUJO. Legend: U—ureters, CV—crossing vessel

between 5 and 12 years of age (mean 20 months, median 6 months).

The most common mode of presentation was urinary tract infection 58% ($n=31$). Fifty-three percent ($n=28$) of the anomalies were detected antenatally. Among the antenatal detected 28 cases, 46% ($n=13$) were asymptomatic, 33% ($n=9$) had evidence of UTI.

Other modes of presentation were abdominal mass (19%, $n=10$) straining during micturition (11%, $n=6$), introital mass (7%, $n=4$), dribbling (4%, $n=2$), retention (4%, $n=2$), and incontinence (4%, $n=2$). One patient presented with an abnormal perineal opening. Eleven percent ($n=6$) antenatally detected patients also had other symptoms like abdominal mass, introital mass, urinary retention, and incontinence.

Urine culture was positive in 55% ($n=29$) of cases, most common organism being *Escherichia coli*. One patient with a large cyst at the lower end of upper moiety ureter presented with urosepsis, abdominal mass, and abnormal renal function.

In our study, 42% ($n=22$) of duplex were left sided, 34% ($n=18$) were right-sided and 24% ($n=13$) were bilateral. Seventy-six percent ($n=40$) of cases were unilateral. 47.5% ($n=19$) of the unilateral duplications were complete type. Out of the bilateral duplications, 62% ($n=8$) were bilateral incomplete, 23% ($n=3$) were a combination of complete on one side and incomplete on the other side. The remaining 15% ($n=2$) were bilateral complete (Fig. 1).

Vesicoureteric reflux (VUR) was seen to be associated with 60% ($n=32$) of cases (Fig. 2). 94% ($n=30$) of the refluxing patients had VUR to the duplex side whereas 6% ($n=2$) of cases had VUR to the opposite normal kidney. Right-sided VUR was more common (43%, $n=13$), left-sided VUR was seen in 38% ($n=11$) and bilateral in 19% ($n=6$).

The commonest grade was grade 3 (53%). Higher grades of VUR were also encountered grade 4 (38%) and grade 5 (18%) (Fig. 3).

Renal scarring on DMSA was seen in 36% ($n=19$) of the total cases. Out these 84% ($n=16$) were refluxing and 3 were non-refluxing. Among the patients with scarring, 63% ($n=12$) of patients had febrile UTI.

The second most common anomaly was ureterocele (Fig. 4) which was seen in 30% ($n=16$) of patients. In this group with ureterocele, VUR to lower moiety was seen in 8 patients and VUR into opposite kidney in 1 patient. Nine patients with ureterocele underwent ureterocele incision whereas the rest had no evidence of obstruction and hence conservatively managed. Three patients developed upper moiety VUR after ureterocele incision and were managed according to upper moiety function (Fig. 4). These 3 patients did not have pre-existing VUR.

Other associations were ectopic ureter 15% ($n=8$) (Fig. 5), pelviureteric junction obstruction 11% ($n=6$) (Fig. 6) and multi-cystic dysplastic kidney of the opposite side 8% ($n=4$). We also encountered few rare associations like horse shoe kidney (2 patients), ectopic kidney (1 patient), crossed fused ectopia (1 patient), anorectal malformation (1 patient), and retrocaval ureter in 1 patient (Fig. 7).

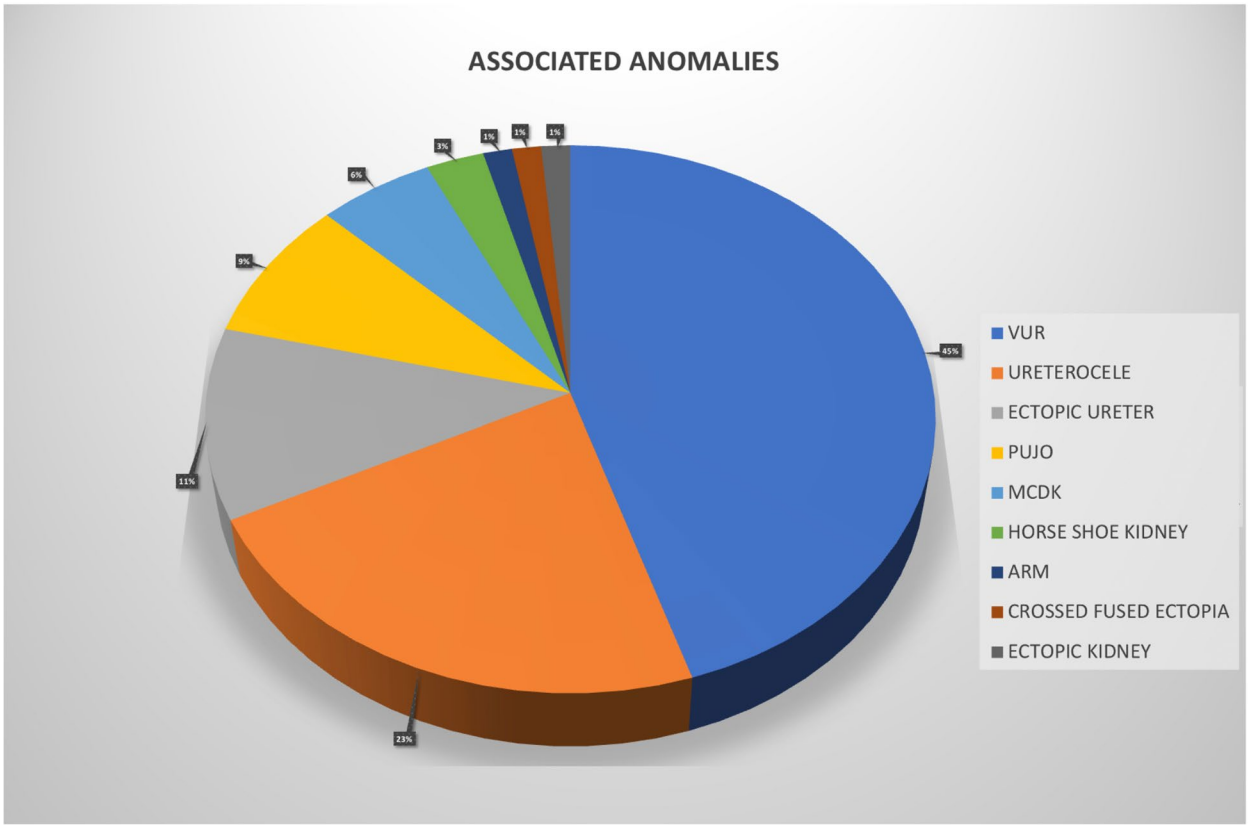

Fig. 7 Pie diagram depicting the associated anomalies of duplex in our study

Sixty-one percent ($n=34$) of these patients were managed conservatively with or without antibiotic prophylaxis. Nineteen patients underwent surgery. Out of 16 patients who had a ureterocele, 9 underwent ureterocele incision. Three of them developed VUR into upper moiety after ureterocele incision. Among the operated group, 5 patients underwent Re-implantation, 2 underwent heminephrectomy, 3 underwent uretero-ureterostomy, 1 underwent pyeloplasty, and 1 underwent endoscopic treatment of VUR using detranomer hyaluronic acid injection. Details of the patients who were surgically managed have been summarised in the table below (Table 1). The algorithm for managing duplex cases in our institution are represented in the chart given below (Fig. 8).

Discussion

Duplication is a common congenital anomaly of the urinary tract. An incidence of 0.7% was found in one series of more than 50,000 autopsies done by Kelalis PP et al. in 1976 [9]. Privett et al. observed that the duplication anomalies were present in 1.8% of the general population, and females were twice as commonly involved as males [10]. In our study, 45% were males and 55% were females.

Various modes of presentation include antenatal detection (Antenatal USG), UTI, bladder outlet obstruction, incontinence, mass at the introitus (prolapsing ureterocele), Hydronephrosis, abdominal mass, and renal failure [11]. These have been classified by Mandell et al. and Jee et al. and has been described by Kelalis et al. In our center, urinary tract infection was the most common mode of presentation (58%) whereas dribbling, retention, and incontinence were seen in 4% each. The routine use of antenatal ultrasound has considerably increased the detection of asymptomatic duplex anomalies. We found that 53% were detected during antenatal screening.

According to the existing literature, vesicoureteric reflux (VUR) was found to be the most common anomaly and is present in 70% of patients who present with UTI [12]. Ureterocele is the second most common anomaly with a reported incidence of 20% [13]. In females with ectopic ureters, 80% drain duplicated systems, thus incontinence being another mode of presentation in females [14]. Pelvi-ureteric junction obstruction (PUJO) is also seen in association with duplex, more commonly seen in the lower moiety with an incidence of 2 to 7% [15]. Very rare associations like

Table 1 List of patients with duplex kidneys who underwent surgical intervention

Number	Age at surgery	Sex	Laterality	Type	Diagnosis	Procedure	Remarks
1	1.5 years	M	Bilateral	Incomplete	Right obstructed megaureter	Right ureteric Cohen's reimplantation	Preoperative Nephrostomy with AGP
2	3 years	M	Right	Complete	Cystic megaureter right	Uretero-ureterostomy and cystic lower ureter excision	Ureterostomy at 3 months for pyonephrosis
3	1 years	F	Right	Complete	Right ureterocele	Ureterocele incision	Repeat MCU normal
4	5.5 years	F	Left	Incomplete	Left lower moiety PUJO	Left lower moiety Anderson Hyne's pyeloplasty	Crossing vessel present
5	3 years	F	Bilateral	Right complete Left incomplete	Right ureterocele	Ureterocele incision	Repeat MCU normal
6	5 years	M	Left	Incomplete	Bilateral VUR with left bifid pelvis	Dextranomer hyaluronic acid injection bilateral	Repeat MCU normal
7	8 years	M	Left	Incomplete	Left VUR	Left ureteric reimplantation (Cohen)	Follow-up normal
8	5 months	M	Right	Complete	Right ureterocele	Ureterocele incision	Follow-up normal
9	2 years	F	Right	Complete	Right ureterocele	Ureterocele incision	Follow-up normal
10	1 year	F	Right	Complete	Right ureterocele	Ureterocele incision	Follow-up normal
11	1.75 year	F	Left	Complete	Left non-functioning Upper moiety	Left upper moiety heminephrectomy	Follow-up normal
12	10 year	F	Bilateral	Left complete Right incomplete	Left lower moiety non-functioning and obstructed	Left lower moiety hemineprectomy	Small residual cyst present
13	1 year	F	Left	Complete	Left ureterocele	Ureterocele incision followed by left uretero-ureterostomy	Post-incision VUR into functioning Left upper moiety
14	3 year	F	Bilateral	Complete	Bilateral duplex with ectopic ureters	Bilateral reimplantation	Follow-up normal
15	1.5 year	M	Bilateral	Right incomplete Left complete	Left ureterocele	Ureterocele incision	Non-functional left kidney with VUR
16	1 year	F	Bilateral	Right complete Left incomplete	Right upper moiety ureterocele and lower moiety VUR	Right common sheath reimplantation	Follow-up normal
17	2 year	F	Right	Complete	Right ureterocele	Ureterocele incision	Post-incision VUR into non-functioning right upper moiety
18	5 month	M	Right	Complete	Right ureterocele	Ureterocele incision followed by right uretero-ureterostomy	Post-incision VUR into functioning right upper moiety
19	10 years	F	Left	Complete	Left HUN with VUJ calculus	Left common sheath reimplantation	Follow-up normal

congenital giant megaureter was also seen associated with Duplex [16]. In our study, VUR was seen in 60% of cases, ureterocele in 30%, ectopic ureters in 15%, PUJO in 11%, and giant megaureter in 2%.

VUR though the most common association in our series, was of low grade in 63% of cases. High-grade VUR cases which led to renal scarring were managed surgically depending on the function of the moiety.

Management can be challenging and is influenced by patient age, presence of infection or incontinence, renal function, moiety involved, type of duplication and surgeon's preference. Management of ureterocele depends on whether they are asymptomatic or symptomatic. The former needs only follow-up whereas symptomatic ones are treated by endoscopic incision. In our study 56% of the ureteroceles required treatment as they were symptomatic.

Heminephrectomy was the treatment offered by us to all non-functioning moieties. It is a procedure which has its own set of complications like mild decrease in function of remaining moiety [17], injury to the good ureter during complete ureterectomy [18] and vascular damage to the renal pedicle [19]. In our series, we left behind distal most ureteral stump in all the 3 cases to avoid injury

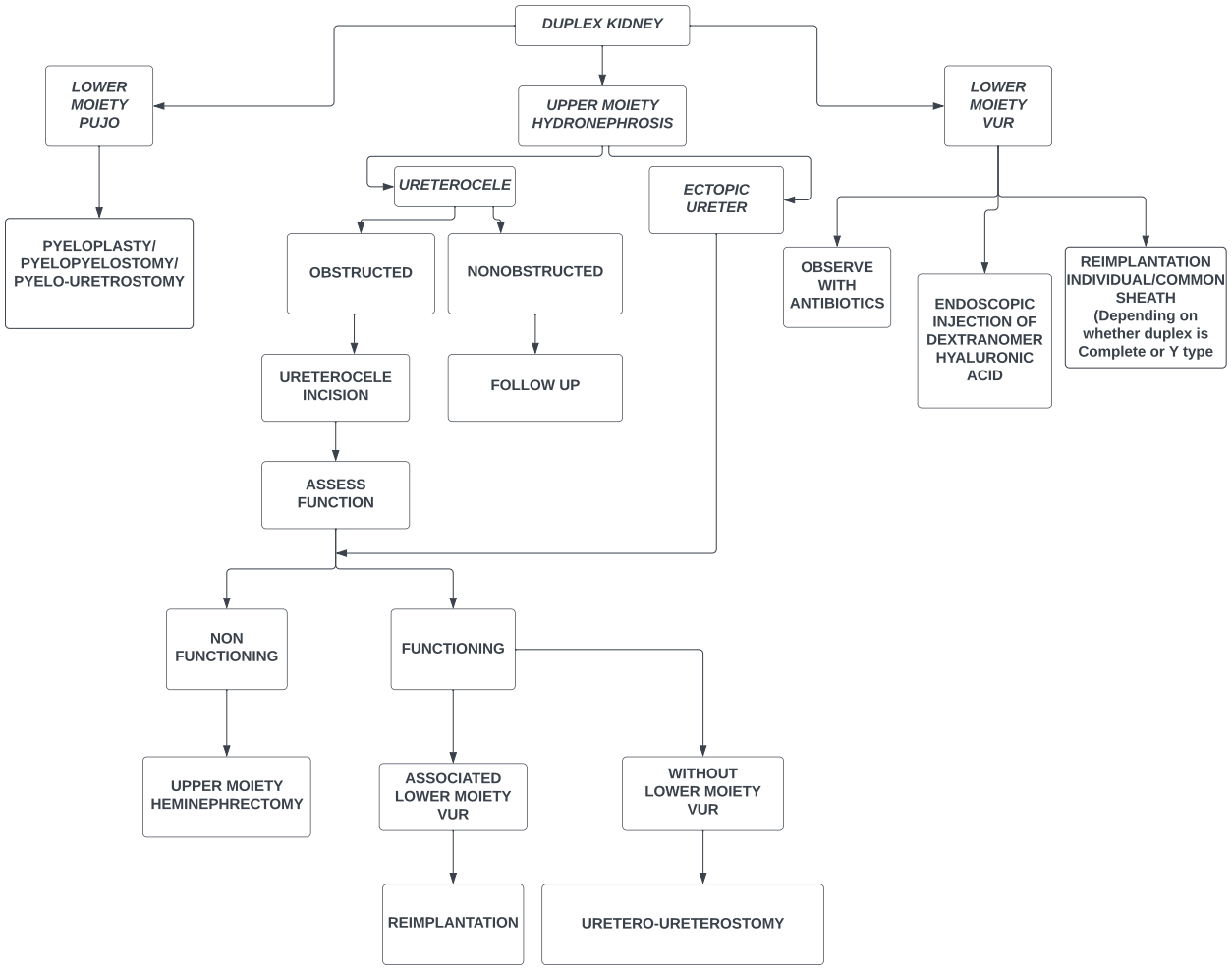

Fig. 8 Institutional Algorithm for treatment of VUR

to the adjacent normal ureter and none of them had any complications as a result of this.

Uretero-ureterostomy and common sheath reimplanation are feasible options for patients with functioning moieties. In our series, 3 patients underwent uretero-ureterostomy and 5 patients underwent reimplantation.

Duplex kidneys, though a common anomaly are usually asymptomatic and need only follow-up. Indications for intervention include recurrent urinary infections, progression of hydronephrosis, or deterioration of renal function. In our study, 61% of the subjects needed follow-up only with or without antibiotic prophylaxis. Only 39% needed intervention in the form of surgery.

There were some limitations to our study. Since this study was conducted in a single referral center, the data may not be applicable to the general population. The study being retrospective in nature, some subjects had to be excluded from the study due to in-availability of proper records or lapse in follow-up.

Conclusion

Duplex kidney, though a common congenital anomaly, requires a systematic evaluation to detect its various associations and a structured management protocol according to the associations. Our study revealed that most of the symptomatic patients presented with urinary tract infections. Vesicoureteric reflux was seen in majority of patients with ureterocele being the second common association. The incidence of these associations are similar to that in existing literature. Nearly half of the patients had renal scarring on radionuclide imaging (DMSA).

It has a high incidence of VUR which is a potentially hazardous condition which can affect the renal function if not evaluated and managed appropriately. Duplex can also be associated with obstructive conditions like PUJO, Ureterocele and ectopic megaureter which can cause rapid deterioration of renal function if not managed early enough. Though majority of the patients need only follow-up and antibiotic prophylaxis, there may be a

certain subgroup of patients who develop early scarring and need aggressive management.

Abbreviations

VUR	Vesicoureteric reflux
MCU	Micturating cystourethrogram
DTPA	Di ethylene penta acetic acid
DMSA	Di mercapto succinic acid
PUJO	Pelviureteric junction obstruction
E.coli	Escherichia coli
IVP	Intravenous pyelogram
KUB	Kidney ureter bladder
USG	Ultrasonogram
MRU	Magnetic resonance urogram

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Authors' contributions
AP collected, analyzed, and interpreted the results. SJ helped in data analysis and was the major contributor in writing the manuscript. TV also participated in the analysis and interpreted the results. All the authors read and approved the final manuscript.

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Availability of data and materials
The data used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Declarations

Ethics approval and consent to participate
Ethics approval was obtained from the Institutional Review Board, Government Medical College Kottayam on 27/05/2022 (IRB number: 37/2022). Consent not applicable as it is a retrospective study. Consent not applicable as it is purely a record based study

Consent for publication
Not applicable.

Competing interest
The authors declare that they have no competing interests.

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