Case Series

Pyeloplasty for the treatment of ectopic renal hydronephrosis in adults: a case series of six patients

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Background: Ectopic renal hydronephrosis is a relatively rare condition, with literature primarily consisting of scattered case studies. The ectopic kidney poses technical challenges to surgeons due to its structural and anatomical abnormalities. This study aims to share our initial experiences with minimally invasive pyeloplasty as a management approach for ectopic renal hydronephrosis in adult patients.

Case Description: Between August 2018 and January 2023, six patients underwent minimally invasive pyeloplasty for ectopic kidneys. The patient cohort consisted of four individuals with pelvic kidneys, one with an iliac kidney, and one with an abdominal kidney. Among these, three cases were left-sided, two were right-sided, and one was isolated. The median patient age was 27 (range, 18–45) years. Four patients underwent robot-assisted laparoscopic pyeloplasty, while two underwent laparoscopic pyeloplasty. The median operative time was 134 (range, 63–240) minutes. After a median follow-up duration of 28.5 (range, 6–59) months, two patients required nephrectomy, with vesicoureteral reflux (VUR) and malrotation potentially contributing to surgical failure in these cases. The remaining four patients showed either stable or improved hydronephrosis and renal function. The overall operative success rate was 66.7%.

Conclusions: The rarity and anatomical variations of ectopic kidneys hinder the standardization of surgical approaches. The integration of three-dimensional (3D) virtual reconstruction and tailored surgical techniques contributes to enhanced outcomes in ectopic renal hydronephrosis cases. Furthermore, continued research and refinement of surgical approaches are needed.

Keywords: Ectopic kidney hydronephrosis; pyeloplasty; three-dimensional virtual reconstruction (3D virtual reconstruction); case series

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Introduction

Ectopic kidney, also known as renal ectopia, is a congenital anomaly characterized by the failure of one or both kidneys to migrate to the retroperitoneal renal fossa during embryonic development between 6 and 9 weeks (1). The reported incidence ranges from 2 to 10 per 10,000 depending on the method of detection (2,3). Ectopic kidneys can be located in the pelvic, iliac, abdominal, or thoracic in order of frequency, and in rare cases, crossed or even fused ectopias may occur (1). In addition to positional anomalies, the ectopic kidney may be small, irregularly shaped, malrotated, functionally impaired, and may be associated with other malformations such as hypospadias, vaginal atresia, cryptorchidism, tetralogy of Fallot, vertebral agenesis, or cleft palate (4,5).

Ectopic kidney is usually asymptomatic and discovered incidentally, but it may be associated with higher risk in pathological conditions such as nephrolithiasis, urinary tract infection (UTI), and hydronephrosis (6,7). Asymptomatic cases require monitoring, while in patients with recurrent symptoms and impaired renal function, timely surgical intervention is indicated. Surgical procedures, such as pyeloplasty, can be used to treat ureteropelvic junction obstruction (UPJO) and improve urinary drainage in cases where treatment is required. Traditional two-dimensional imaging techniques like computed tomography (CT) and magnetic resonance imaging (MRI) may fall short in complex anatomical cases, whereas three-dimensional (3D) virtual reconstruction offers more detailed visualization, thereby enhancing the precision of preoperative planning and improving surgical outcomes.

Although the managing of UPJO in renal, robotic-assisted laparoscopic pyeloplasty as proven effective for adult patients with concomitant UPJO and renal anomalies (8). This study presents our initial experience in managing hydronephrosis in adult patients with ectopic kidney using laparoscopic or robot-assisted laparoscopic pyeloplasty, guided by 3D virtual reconstruction. We present this article in accordance with the AME Case Series reporting checklist (available at https://tau.amegroups.com/article/view/10.21037/tau-24-142/rc).

Highlight box

Key findings

- Three-dimensional (3D) virtual reconstruction is important for guiding pyeloplasty surgery for ectopic renal hydronephrosis.
- Vesicoureteral reflux (VUR) and renal malrotation were causes of failure in two cases.

What is known and what is new?

- Ectopic renal hydronephrosis can be caused by obstruction, VUR, malrotation, or a combination of factors.
- A comprehensive preoperative assessment and meticulous postoperative follow-up are crucial for the success of pyeloplasty in ectopic kidneys.

What is the implication, and what should change now?

- The routine integration of 3D virtual reconstruction in preoperative evaluations is recommended for improving surgical planning. Surgical approaches must be tailored to the specific characteristics of each patient with ectopic renal hydronephrosis.

Case presentation

Clinical materials

Between August 2018 and January 2023, six patients were admitted to the hospital with a diagnosis of ectopic kidney treated with minimally invasive pyeloplasty surgery, which was performed by the same surgeon who had performed more than 100 robotic surgeries. We retrospectively analyzed the demographics, perioperative variables, and follow-up data of the six patients collected in our Reconstruction of Urinary Tract: Technology, Epidemiology and Result (RECUTTER) database, as shown in *Table 1*. All procedures performed in this case series were in accordance with the ethical standards of the Ethics Committee of Peking University First Hospital (No. 2020-SR-346) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patients for publication of this case series and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

Preoperative preparation

Renal ultrasonography (US) was routinely performed preoperatively, and pelvis dilatation diameter and parenchymal thickness were estimated in all patients. Computed tomography urography (CTU) was conducted in all patients, including those with significantly decreased renal function, after making a shared decision with the patients, to ensure accurate assessment of the urinary tract anatomy. The acquired images were then used to create 3D reconstructions of the renal anatomy. This process involved segmenting the kidney and surrounding vasculature from the CTU data to generate a detailed 3D model, which was then used to access the spatial relationships between the renal structures and the

Table 1 Patient characteristics and postoperative results

Case	Age (years)/ sex	Location	Side	Presentation	Pelvis diameter (mm)	Renal function GFR (mL/min)/ratio	Surgical approach	OT (min)	Follows-up (months)	Outcomes pelvic dilatation/ renal function
1	18/F	Pelvic	Single	Asymptomatic	36	78.58/100%	LS	63	59	Improved/ improved
2	44/F	Pelvic	Left	UTI	14.9	18/17%	LS	240	31	Nephrectomy at 30 months
3	32/F	Pelvic	Left	Abdominal distension	63	20/25%	RA	139	29	Nephrectomy at 11 months
4	40/F	Iliac	Right	UTI	20.8	32/40%	RA	129	28	Improved/stable
5	22/M	Abdominal	Left	Asymptomatic	29.1	40/63%*	RA	116	18	Improved/stable
6	19/F	Pelvic	Right	Abdominal distension	22	30/34%	RA	215	6	Stable/ improved

*, GFR may be overestimated due to overlapping left renal imaging with the spleen. GFR, glomerular filtration rate; OT, operative time; F, female; LS, laparoscopy; UTI, urinary tract infection; RA, robotic-assisted; M, male.

blood vessels (*Figure 1*). 99m technetium diethylenetriamine pentaacetic acid (^{99m}Tc-DTPA) with diuretic administration was used to evaluate drainage and assess renal function. In case 3, nephrostomy was performed 2 months before surgery. Preoperative double J stent (D-J stent) placement under cystoscopic guidance was performed in three patients (cases 1, 4, 6) and retrograde pyelography was performed in two patients (cases 1, 6). These preoperative interventions were necessary to alleviate hydronephrosis, improve renal function, and optimize the outcomes of the subsequent pyeloplasty. The performance of a laparoscopic versus robotic procedure depended on patient's preference and availability of equipment.

Surgical technique

Indication for surgery included persistent clinical symptoms, renal function deterioration, and obstruction evidenced by radiographic results. All surgeries were performed via a transperitoneal approach. Following general anesthesia, a sterile Foley catheter was inserted into the bladder. The patient was positioned obliquely with the lesion side up. After creating pneumoperitoneum, trocar was arranged in a fan shape with the ectopic kidney as the central reference point, the camera port at the umbilicus, and working ports laterally. Placement was optimized for the surgical to ensure effective instrument access and maneuverability.

The surgical approach of laparoscopy and robotic surgery is similar. Different types of reconstructive procedures were used when a dilated renal pelvis was detected and isolated. Patient 2 underwent pyelovesicostomy for complete ureteral obstruction caused by tuberculosis infection, while patient 3 underwent pelvic flap pyeloplasty (9). In case 5, dismembered pyeloplasty was performed due to the presence of crossing vessels, and a flexible cystoscope was used to remove the caliceal stones through the pyelotomy incision. The remaining three patients underwent Y-V pyeloplasty to minimize surgical trauma and reduce operative time (OT). Before completing the running anastomosis, a D-J stent was advanced through the renal pelvis to the bladder. After the completion of the anastomosis, a drain was placed through a trocar into the pararenal space (*Figure 2*). The OT was recorded from the skin incision to skin closure, including the docking process for robot-assisted surgery.

Postoperative treatment and follow-up

Routine kidney-ureter-bladder (KUB) plain films were performed on the first postoperative day to confirm the appropriate positioning of the D-J stent. The drain was usually removed when the output was <50 mL after the operation. Foley catheters were removed within one week postoperatively to ensure adequate bladder drainage and minimize the risk of urinary retention, while the D-J stent was removed 2–3 months after surgery. Patients were followed up for 3 months, 6 months for the first year, and annually after surgery through telephone interviews or outpatient visits. During the follow-up visits, physical

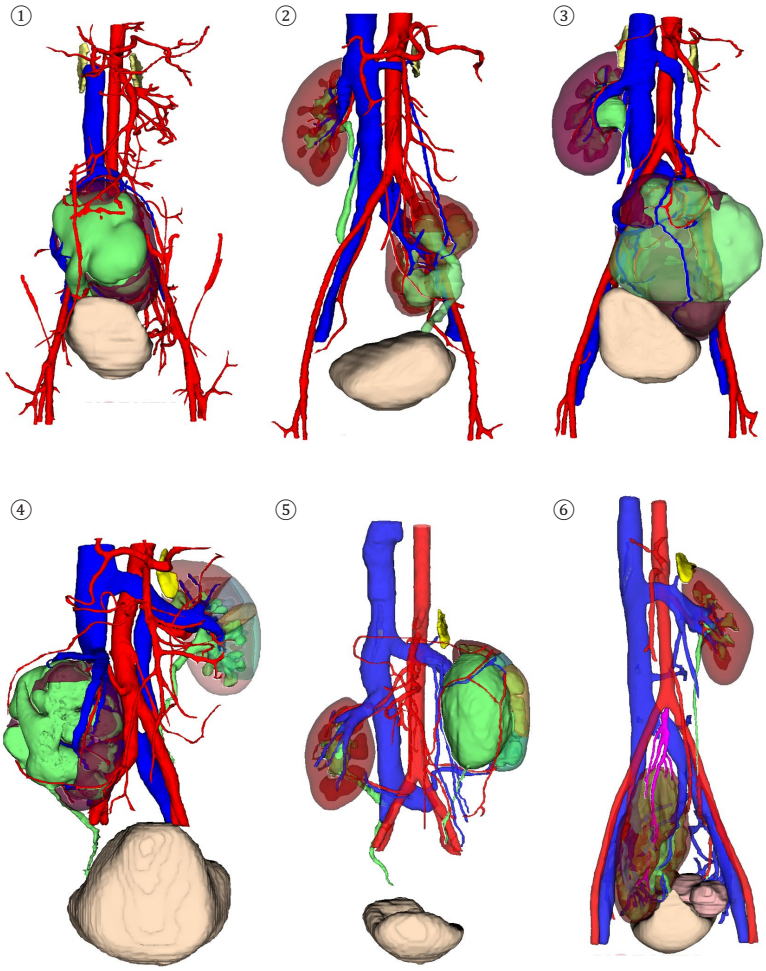

Figure 1 Preoperative three-dimensional image reconstruction of six patients. Case 1: isolated pelvic kidney with bilateral CIA supply and IVC return. Case 2: left pelvic reverse rotation kidney with AA supply, and CIV/IVC return. Case 3: left pelvic kidney with CIA/AB supply, CIV/IVC return, and contralateral UPJO. Case 4: right iliac reverse rotation kidney with AA/CIA supply, and IVC return. Case 5: left abdominal kidney with AA supply, CIV/IVC return, and contralateral abdominal kidney. Case 6: right pelvic kidney with AB supply and contralateral CIV return. CIA, common iliac artery; IVC, inferior vena cava; CIV, common iliac vein; AA, abdominal aorta; AB, aortic bifurcation; UPJO, ureteropelvic junction obstruction.

examinations, complete blood count (CBC), and urinalysis (UA) were performed. Renal function was evaluated by serum creatinine at each visit, and diuretic renal dynamic imaging was conducted at the six-month and one-year marks. Radiographic examinations consisted of magnetic resonance urography (MRU) at the 3rd month, CTU at the 6th month, and ultrasound every 3 months in the first year, followed by semiannual ultrasound thereafter. Subjective success was defined as the relief of symptoms, while objective success was determined by stable or improved radiographic evidence of hydronephrosis and renal function. Operative success was defined as the combination of both subjective and objective success.

Results

The patients' characteristics are shown in *Table 1*. The median age of the patients was 27 (range, 18–45) years, and the median body mass index (BMI) was 22.38 (range, 19.36–25.15) kg/m^2. There were four patients with pelvic kidney, one patient with iliac kidney and one patient with abdominal kidney. Three cases were left-sided, two were right-sided, and one was isolated. Four patients presented with symptoms such as abdominal distension

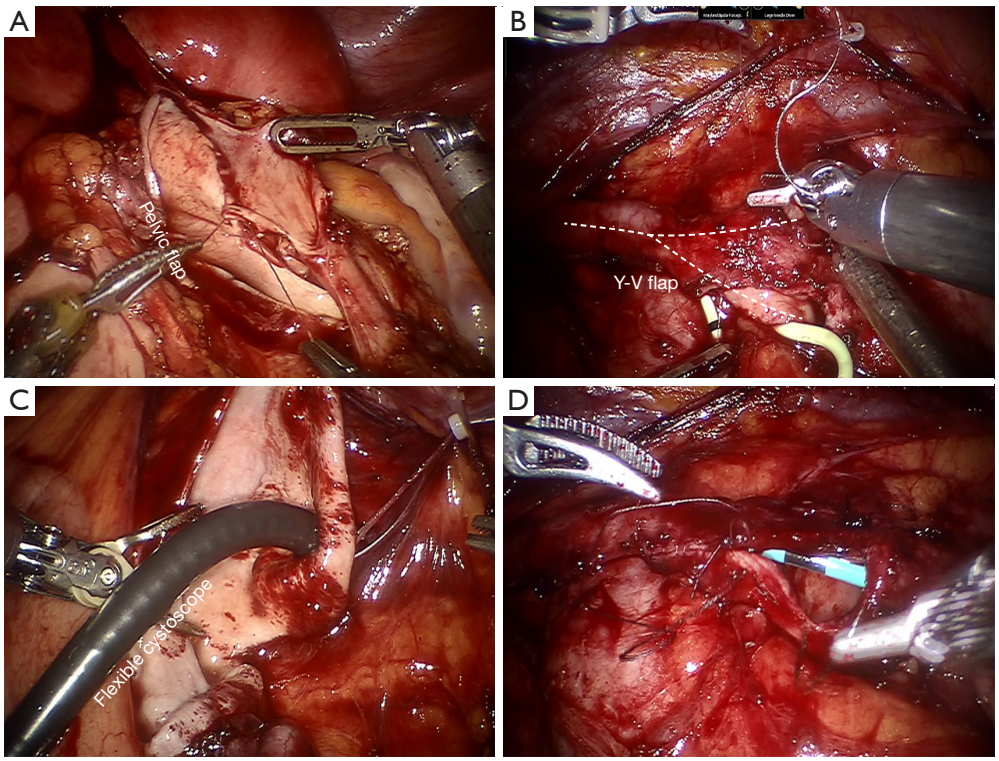

Figure 2 The pictures of operation. (A) Pelvic flap pyeloplasty in case 2. (B) Y-V pyeloplasty in case 4. (C) Flexible cystoscope was passed through the pyelotomy incision to extract the caliceal stones in case 5. (D) A D-J stent was inserted prior to anastomosis. D-J, double J.

or urinary tract infections, while the remaining two were asymptomatic, with their ectopic kidneys incidentally discovered during health check-ups. For other anomalies, patient 3 exhibited a contralateral UPJO in addition to the presence of scoliosis and autoimmune type 1 renal tubular acidosis (RTA). In case 5, the contralateral kidney was also positioned lower than normal. In case 6, the patient presented with concurrent conditions of inguinal hernia and uterine developmental abnormalities. Four patients underwent robotic-assisted laparoscopic pyeloplasty, and two underwent laparoscopic pyeloplasty. The median OT was 134 (range, 63–240) min. The median estimated blood loss (EBL) was 15 (range, 10–50) mL. All surgeries were effectively completed, and no case required open conversion. The median postoperative hospital time was 4.5 (range, 3–21) days. In case 2, the patient experienced postoperative fever, and a urine culture indicated the presence of *Enterococcus faecalis*. Intravenous administration of anti-infective therapy led to an extended hospitalization period of 21 days. No postoperative complications of high grade (grade III and IV) occurred within 1 month of surgery according to the Clavien-Dindo classification system. After median follow-up duration of 28.5 (range, 6–59) months, symptoms were completely alleviated in cases 4 and 6. Four patients showed either stable or improved hydronephrosis and renal function. In cases 1, 4, and 5, the pelvic diameter decreased by an average of 0.92 (range, 0.85–0.99) cm, as measured on ultrasound scans. Renal function increased in case 6, with an increase of 13 mL/min. In case 3, there was recurrence of UTI in the postoperative period, and renal function deteriorated after the reintroduction of the D-J tube and balloon dilatation. Ultimately, nephrectomy was performed 11 months after pyeloplasty. In case 2, the left ovarian chocolate cyst exhibited a notable increase in size, measuring 0.7 cm in the 15th postoperative month and expanding to 7.2 cm in the 27th postoperative month. The cyst pushed forward against the pyelovesical junction, exacerbating the outflow obstruction and ultimately leading to a decline in renal function, necessitating nephrectomy. Therefore, the operative success rate was 66.7% (4/6).

Discussion

Performing a minimally invasive procedure on a hydronephrotic

ectopic kidney, especially in the case of a pelvic ectopic kidney, presents technical challenges to the surgeon due to structural and anatomical abnormalities (10). Firstly, the ectopic pelvic kidney lacks a perirenal fatty capsule, and its stability depends on retroperitoneal abnormal adhesions or the presence of multiple sets of vessels entering the hilum from different directions. The narrow pelvic space, presence of viscera, vessels, and nerve plexus increase the risk of damage to these structures (11). Secondly, the aberrant vasculature supplying the kidneys, which represents transient arteries that normally disappear as the kidney migrates to its position in the lumbar region, becomes a permanent blood supply in the case of ectopic kidneys (11). These vessels may arise from the iliac arteries (external, internal or common), the distal aorta, the middle sacral artery, or the inferior mesenteric artery, and the pattern of the vascular network of ectopic kidneys is entirely anomalous (12,13). Thirdly, the malrotation of the kidney, the wide variation in the position and orientation of the renal pelvis outlet, and the presence of a short and tortuous ureter, complicate surgical intervention. It is important to note that the ectopic renal hydronephrosis extends beyond being merely a UPJO issue. These combined factors can create a complex treatment dilemma for the surgeon.

The complexity of ectopic kidneys often renders traditional two-dimensional imaging techniques, such as CT or MRI, insufficient for conveying the detailed anatomical information required for effective surgical planning. Recognizing this limitation, our center has adopted 3D virtual reconstruction technology as a critical tool in managing urinary reconstruction, encompassing preoperative planning, intraoperative navigation, and postoperative outcome evaluation (14).

In this case series, all patients underwent preoperative 3D virtual reconstruction, which generated detailed visualizations of anatomical morphology and spatial relationships, particularly concerning anomalous blood vessels. Hydronephrosis was present in all six cases, with varying degrees of severity, necessitating meticulous dissection. The 3D reconstruction provided crucial guidance by clearly delineating the dilated renal pelvis and its relationship to the surrounding aberrant vasculature. Furthermore, the 3D reconstruction was instrumental in understanding the atypical orientation of the renal pelvis, enabling a tailored surgical approach that avoided kinking of the ureter during pyeloplasty.

For all patients, the use of contrast-enhanced imaging for 3D reconstruction was carefully weighed against the potential risks and thoroughly discussed with them. The benefits of obtaining detailed anatomical were deemed to outweigh the risk of further renal impairment, leading to improved surgical outcomes. These 3D models provided the surgical team with a comprehensive, stereoscopic understanding of the surgical scenarios, enabling more accurate decision-making. The use of 3D reconstruction significantly enhanced the identification and avoidance of aberrant vasculature, optimized trocar placement, and facilitated precise surgical maneuvers, thereby reducing the risk of intraoperative complications. Additionally, 3D virtual reconstruction allowed for real-time adjustments during surgery, crucial for navigating the complex anatomy of ectopic kidneys. This adaptability contributed to the successful completion of all procedures without injury to surrounding structures, vascular complications, or the need for open conversion.

Helmy and colleagues reported a study involving 43 cases of open dismembered pyeloplasty for ectopic pelvic kidneys in children. After a follow-up period of 42 months, the overall success rate was 82%. However, the postoperative improvement rates of hydronephrosis and renal function were only 52.6% and 31.6%, respectively (15). Yin *et al.* retrospectively analyzed 16 cases that underwent laparoscopy and flexible ureteroscopy combined pyeloplasty for the treatment of ectopic pelvic kidney with stone and UPJO. The mean OT was 171 min. After a median follow-up of 29.3 months, no stone remnants were found and there was an alleviation of pelvicalyceal dilatation after surgery (16). Esposito *et al.* gathered the largest series of 48 complex UPJO cases in a multicentric study treated with robot assisted laparoscopic pyeloplasty, including 9 patients with pelvic ectopic kidneys. The median OT was 178.5 minutes. After a follow-up of 18.2 months, the overall success rate was 95.8%. However, it is unclear whether the two cases of failure were among those with pelvic ectopic kidneys (17). Overall, laparoscopic and robotic-assisted laparoscopic pyeloplasty were deemed safe and feasible for the treatment of ectopic kidney.

In our case series, all procedures were performed by an experienced surgeon without conversion to laparotomy, and in case 5, simultaneous pyelolithotomy was performed. Our minimally invasive approach provided excellent surgical exposure, with an OT comparable to that reported by other centers (16,17). Furthermore, concomitant pyelolithotomy could be performed without the need for additional trocars, enabling intact stone removal within a single operative session. However, from long-term follow-up, the success rate is 66.7%, and two patients ultimately underwent

nephrectomy, leaving us to ponder the reasons for surgical failure. In previous literature reports, Helmy *et al.* attributed the lower success rate of pelvic ectopic pyeloplasty compared to primary UPJO without upper tract anomalies to pyelocaliectasis, malrotation or anomalous vasculature. Additionally, preoperative renal function and surgeon's experience are predictors of renal function improvement after pyeloplasty (15).

Approximately 56% of ectopic kidneys have hydronephrosis, Gleason *et al.* reported that hydronephrosis in renal ectopia was due to primary obstruction in 52% of cases, either at the ureteropelvic or ureterovesical junction (37% and 15%, respectively), vesicoureteral reflux (VUR) grade III or greater in 26% and extrarenal collecting systems with malrotation in 25%. Notably, hydronephrosis was present in 26% of non-ectopic contralateral kidneys, with VUR being the most common cause in 60% of cases (4). Arena *et al.* found that out of 36 patients with simple renal ectopia identified by neonatal ultrasound, 16.6% had VUR, all of which occurred in patients with pelvic ectopia (18). In 16 cases of renal pelvis in pediatric patients, UPJO due to high insertion of the ureter into the renal pelvis was the commonest anomaly requiring surgical correction, accounting for 31%, followed by 3 cases of ureteral ectopia and reflux due to aberrant migration of the lower ureter (19). The incidence of VUR in ectopic kidney is estimated to be 11% to 30% according to the reported articles. Therefore, the diagnosis of VUR plays a crucial role in determining the appropriate surgical procedure for patients with ectopic renal hydronephrosis (20-22).

The prevalence of VUR in patients with UPJO has been reported to be 8.2% based on a meta-analysis (23). In patients with coexisting VUR and UPJO, particularly in the context of ectopic kidneys, the cause of hydronephrosis must be clearly identified, as obstruction may be secondary to severe reflux, which causes kinking and tortuosity of the ureter an the ureteropelvic junction and fixation by inflammatory adhesions. Patients with high grade reflux often exhibit dilated and tortuous ureters, increasing the risk of obstruction fivefold (24). This was evident in case 2, where tuberculosis infection led to complete ureteral obstruction, as confirmed preoperatively by ureteroscopy and retrograde pyelogram. In patients with primary UPJO, mild VUR may resolve spontaneously following pyeloplasty. However, in cases where UPJO is secondary to severe reflux, pyeloplasty remains the preferred treatment, as anti-reflux surgery may pose a risk of acute ureteropelvic junction decompensation. Conversely, in pseudo-UPJO, where the pelvis is dilated but urinary drainage is unobstructed, ureteral reimplantation should be considered the first-line treatment, avoiding pyeloplasty (25). VUR increases the risk of acute pyelonephritis and subsequent kidney scarring, leading to further renal function deterioration (26). The lower anatomical position of ectopic kidneys potentially increases susceptibility to severe infection due to reduced gravitational resistance to reflux, though this assertion is based on mechanistic inference rather than direct evidence. Therefore, ensuring sterile urine and promoting regular urination to prevent retention are essential components of preoperative and postoperative management strategies for patients with ectopic kidneys.

Another factor to take into account during pyeloplasty for ectopic kidney is the issue of malrotation. Renal malrotation encompasses nonrotation or incomplete rotation, reverse rotation, hyperrotation, and sagittal rotation (1,27). The most common types are incomplete rotation and nonrotation, wherein the hilum occupies an anterior position or lies between the anterior and the standard medial position. Reverse rotation is another commonly observed type, characterized by the orientation of the renal hilum posterior to the spine, with renal vessels crossing the kidney anteriorly to reach the hilum (1). In both of these cases, the ureter is situated laterally (27). In the two patients whose surgeries were not successful, case 2 involved a reverse rotation type, and case 3 showed an incompletely rotated kidney. Jayakumar *et al.* reported four cases of UPJO associated with laterally malrotated kidneys (28). The modified anterior dismembered pelvi-ureteric anastomosis approach resulted in good drainage. However, the inferior approach failed to improve the ureteropelvic junction drainage in both patients. After undergoing redo pyeloplasty with anterior anastomosis, one patient exhibited adequate restoration of drainage. Divjak *et al.* reported a case of a hyperrotated (270°) kidney where laparoscopic ureterocalicostomy was performed, involving inverted funnel-shaped parenchymal resection and reimplantation of the ureter antero-inferiorly on the inferior calyx (29). This surgical approach allows for optimal drainage and helps avoid secondary stenosis. A conventional pyeloplasty is less likely to succeed because the renal pelvis may become recurrently obstructed due to ureteropelvic junction kinking upon its return to the original position (27). Therefore, for malrotated ectopic kidneys, ureterocalicostomy—where the ureter is attached to the inferior calyces—may be the best option to improve urine drainage. The decision to undergo nephropexy remains a contentious issue (28,30).

Our study has certain limitations that should be acknowledged. Firstly, the retrospective design and small sample size of only six cases make it inadequate to draw definitive conclusions. However, the rarity of ectopic renal hydronephrosis makes it challenging to gather a large number of cases. Furthermore, the absence of examinations such as voiding cystourethrogram (VCUG) to identify VUR early in the diagnostic process, particularly in patients with ureteral dilatation and recurrent UTI, limits our ability to provide comprehensive guidance for treatment selection and postoperative management in these cases. As a result, we can only hypothesize that VUR was one of the reasons for failure. We believe that even a small case series can provide valuable insights and contribute to the existing literature.

Conclusions

The management of ectopic renal hydronephrosis requires special consideration due to its complex etiology, including obstruction, VUR, and malrotation. Although our study is limited by a small sample size, it underscores the clinical value of 3D virtual reconstruction in improving surgical planning and outcomes. This technology enables more precise and individualized treatment approaches. Our findings suggest that minimally invasive pyeloplasty, guided by 3D virtual reconstruction, is a viable option for patients with ectopic kidneys. Future studies with larger sample sizes are needed to further validate the role of 3D virtual reconstruction in the surgical management of ectopic kidneys.

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Footnote

Reporting Checklist: The authors have completed the AME Case Series reporting checklist. Available at https://tau.amegroups.com/article/view/10.21037/tau-24-142/rc

Peer Review File: Available at https://tau.amegroups.com/article/view/10.21037/tau-24-142/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tau.amegroups.com/article/view/10.21037/tau-24-142/coif). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All procedures performed in this case series were in accordance with the ethical standards of the Ethics Committee of Peking University First Hospital (No. 2020-SR-346) and with the Helsinki Declaration (as revised in 2013). Written informed consent was obtained from the patients for publication of this case series and accompanying images. A copy of the written consent is available for review by the editorial office of this journal.

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