Using the renal pelvis flap to replace the whole hypoplastic ureter: a preliminary report

Khaled M. El-Asmar, Ehab A. El-Shafei and Sameh Abdel-Hay

Background **Hypoplastic ureter is a rare condition usually associated with hypoplastic kidney, and it ends with nephrectomy in most of the cases. Many techniques have been described as ureteric substitutes in the literature. Here, we describe a new technique using the renal pelvis flap to replace the whole hypoplastic ureter in two cases.**

Objective **The aim of this study was to describe a new surgical technique in the management of ureteric hypoplasia.**

Patients and methods **Of the two boys diagnosed antenatally, unilateral hydronephrosis was detected in one boy and a huge renal cyst was present in the other, with evidence of postnatal progressive obstruction necessitating surgical intervention. On exploration, hypoplastic ureter throughout its entire length was an accidental intraoperative finding. The renal pelvis flap was taken and tubularized to replace the entire ureter, and reimplanted into the urinary bladder. This technique was the primary procedure in one case, whereas it was the secondary procedure in the other case after failure of initial trial of pyeloplasty.**

Results **The postoperative period was uneventful with adequate drainage of the renal pelvis in the short-term follow-up (6 and 3 months consecutively).**

Conclusion **The renal pelvis flap is a new feasible alternative procedure for ureteric replacement in a hypoplastic ureter when there is preserved renal parenchyma.** ***Ann Pediatr Surg*** **10:68–71 © 2014 Annals of Pediatric Surgery.**

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Introduction

Congenital anomalies of the urinary tract system are common causes of morbidity and renal failure in children [1]. Among these anomalies, ureteric anomalies such as congenital ureteric stenosis [2] and long hypoplastic ureter are rare, and are usually associated with significant renal dysplasia with loss of excretory function on renal scan where nephrectomy is the primary treatment modality [3].

If there is still residual functioning of the renal parenchyma, ureteral replacement for these anomalies is performed classically by replacing the ureter using a segment of the ileal loop. However, ureteric replacement using an intestinal segment carries the risk of short-term and long-term morbidities, for example, anastomotic stricture, intestinal leakage, and metabolic complications in the form of hyperchloremic metabolic acidosis [4,5].

Therefore, it is better to reconstruct the ureter using the urinary tract itself. The flap technique described in these two cases using the renal pelvis is not well documented in the literature as a surgical option for ureteric replacement.

Case 1

A full-term boy presented to our unit with right-sided hydronephrosis suggesting ureteropelvic junction (UPJ) obstruction as diagnosed by routine antenatal ultrasound. Postnatal renal ultrasound was performed at 1 week of age, which confirmed the diagnosis of UPJ obstruction. A follow-up ultrasound performed at the age of 1 month revealed progressive increase in the anteroposterior diameter of the renal pelvis (Fig. 1).

Exploration of UPJ was carried out through the anterior abdominal approach, which revealed a markedly dilated renal pelvis and a hypoplastic ureter with a very small caliber that can hardly accommodate a 20-G cannula; trial of resection and primary anastomosis with the tiny ureter was performed, but it was complicated by postoperative failure of pelvic drainage that manifested on removal of the pyelostomy tube. Postoperative ultrasound showed huge dilatation of the pelvis with turbid urine suggesting infection (Fig. 2). Temporary percutaneous nephrostomy was left in place with antibiotic administration according to results of the urinary culture till resolution of infection. Re-exploration was performed 1 month later, which showed complete fibrosis and obstruction of the thinned atretic ureter; it was corrected by the renal pelvis flap procedure. The postoperative period was uneventful with adequate drainage of the renal pelvis. A renal scan was performed 1 month later and it showed a residual function of 10% on the right kidney with no evidence of obstruction. Follow-up sonography after 6 months showed good cortical thickness and preserved corticomedullary differentiation with no evidence of infection.

Case 2

A male neonate was diagnosed antenatally with a right-sided huge renal cyst; postnatal ultrasonography performed on day 7 showed a huge renal cyst with thinned out parenchyma; increase in the cyst diameter and thinning of the parenchyma was observed at the follow-up after 1 month (Fig. 3). A diuretic renal scan (DTPA) showed a split kidney function of 22% on that side with evidence of UPJ obstruction (Fig. 4).

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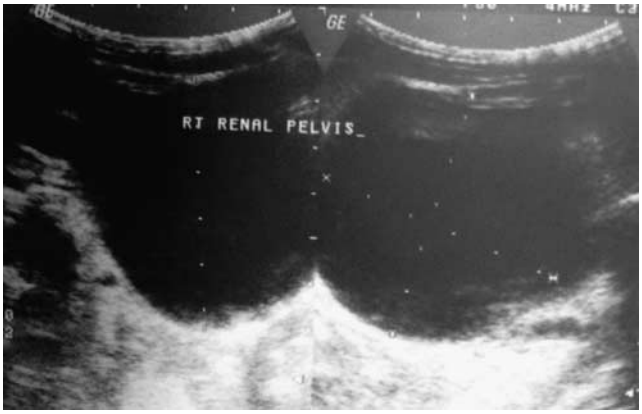
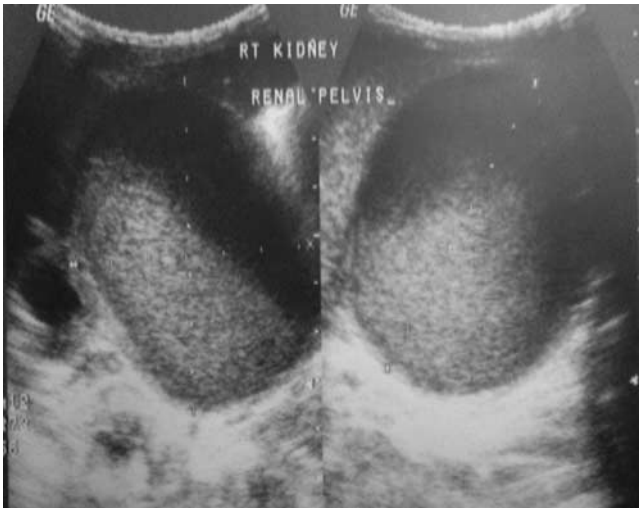
Fig. 1



An ultrasonographic image showing preoperative hydronephrotic changes in case 1.

Fig. 2



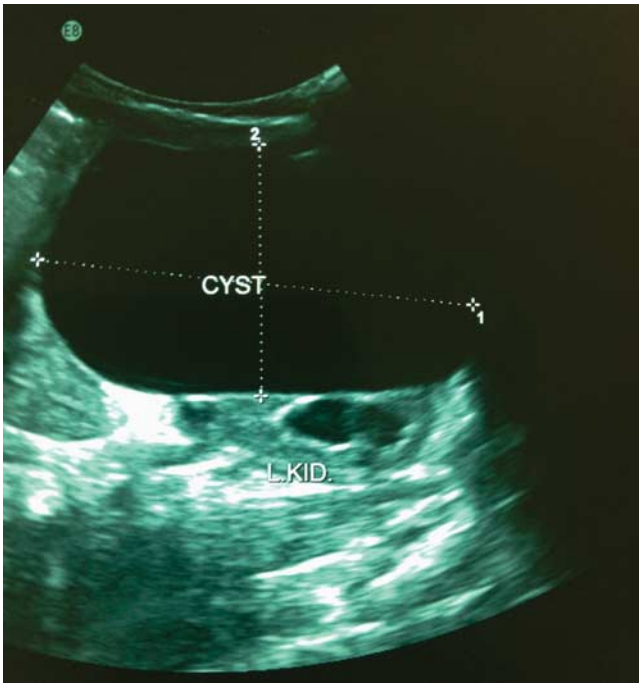
Turbid urine inside the renal pelvis after the first trial of pyeloplasty, indicating the development of pyonephrosis.

Therefore, the decision of surgical intervention was taken. On exploration, this cyst was revealed to be a dilated renal pelvis associated with a very narrow hypoplastic ureter; hence, the diagnosis of hypoplastic ureter was established only intraoperatively. The renal pelvis flap technique was the primary procedure for the correction of the ureteral anomaly in this case. The follow-up ultrasound 1 month postoperatively showed a decrease in the pelvic dimensions and patent UPJ with good thickness and echogenicity of the renal parenchyma on that side (Fig. 5). A renal scan performed 3 months postoperatively showed a split kidney function of 28% on the operated side.

The operative technique

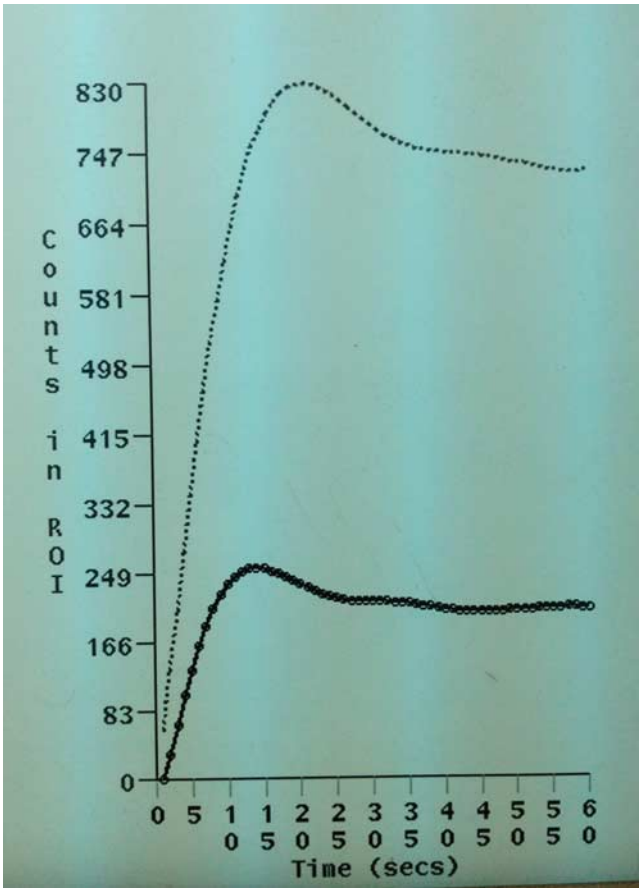
An informed consent was obtained from the parent, explaining the full operative details and possible post-operative complications.

Fig. 3



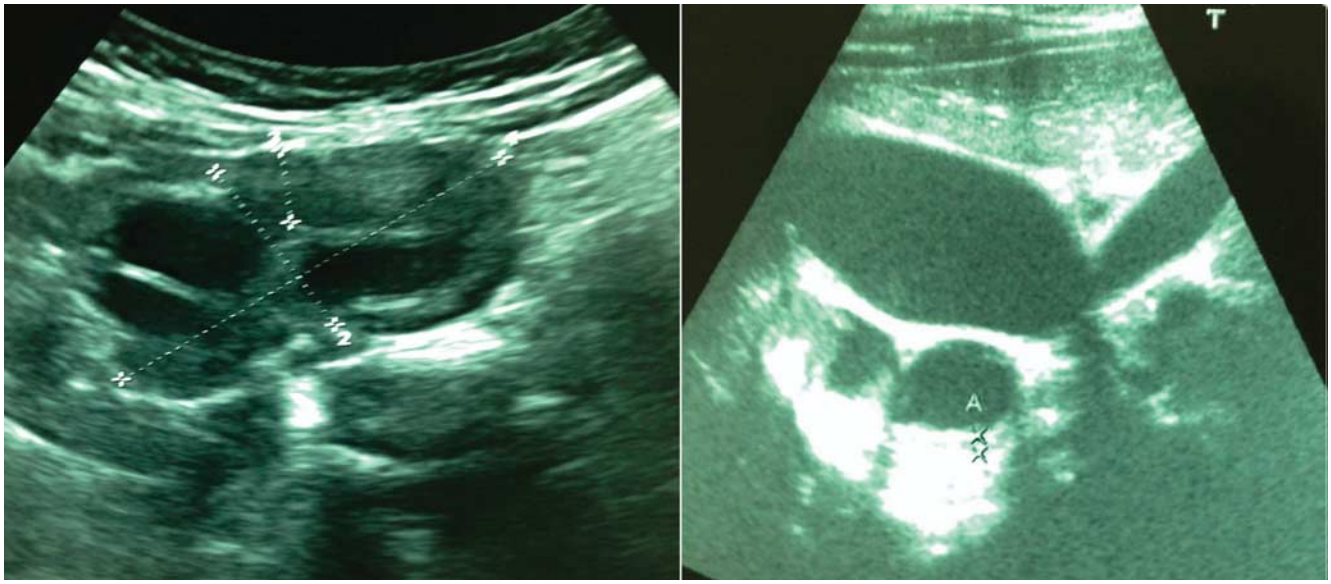
An ultrasonographic image of case 2, showing the misdiagnosis of marked hydronephrosis as a huge renal cyst.

Fig. 4



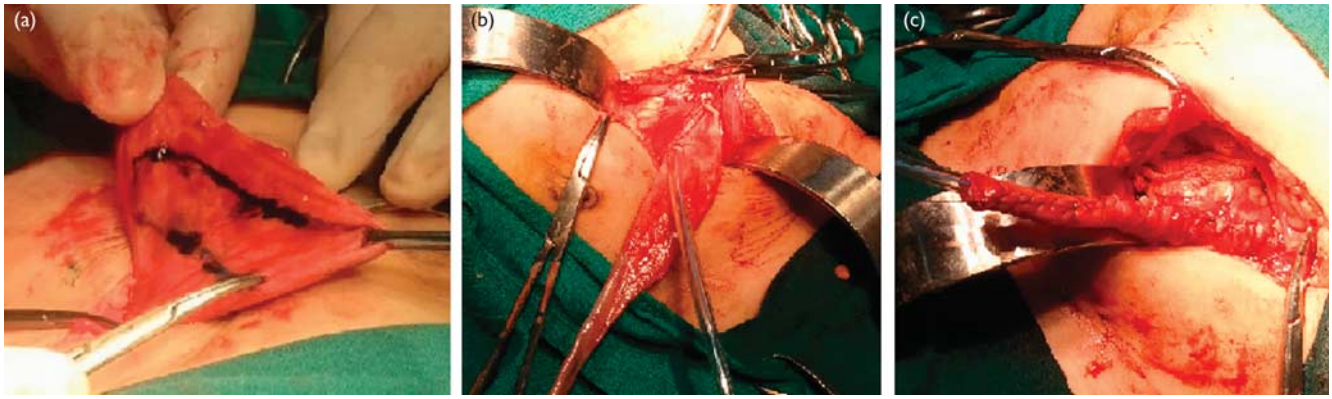
A diuretic renal scan (DTPA) showed a split kidney function of 22% on the right side with evidence of ureteropelvic junction obstruction.

Fig. 5



Follow-up ultrasound 1 month postoperatively showed a decrease in the pelvic dimensions (a) and patent ureteropelvic junction (b).

Fig. 6



The surgical technique: (a) designing the renal pelvis flap; (b) reflection of the flap based on the lower border of the renal pelvis; (c) tubularization of the flap around a 12-Fr nelaton catheter.

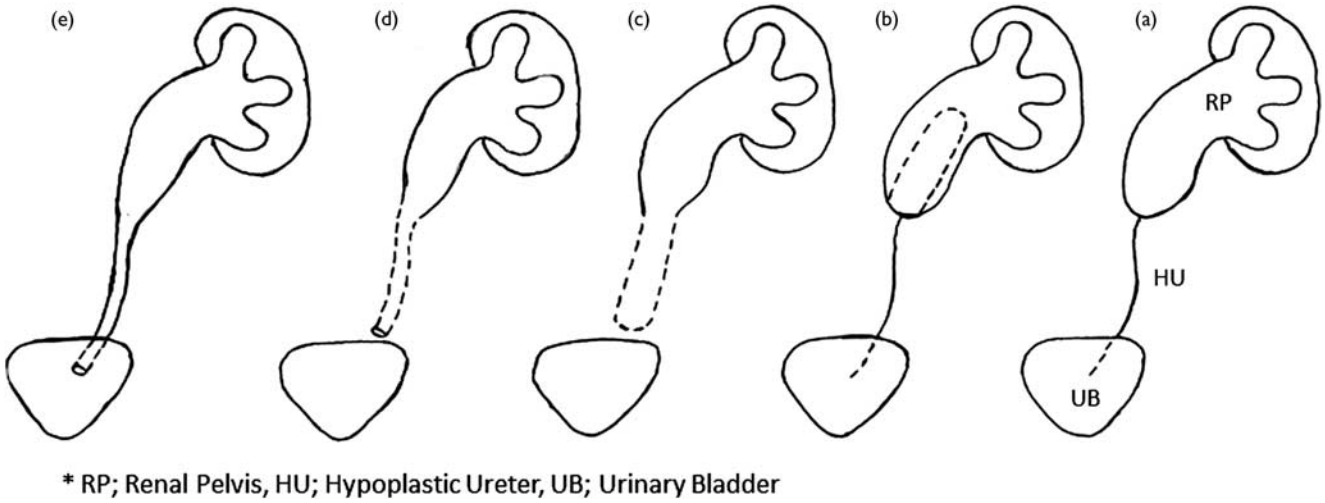
Our technique uses the hugely dilated renal pelvis as a flap to replace the entire length of the hypoplastic ureter.

(1) A designed flap is drawn on the anterior surface of the renal pelvis starting 1 cm from the kidney sinus, and 2 cm in width based on the lower edge of the pelvis (Fig. 6a). Incision of the flap is made, which is then reflected towards the urinary bladder (Fig. 6b) and tubularized over a 12-Fr catheter using a 6-0 polyglycolic absorbable suture after checking its viability. Tubulization was performed in a continuous manner for the proximal part of the flap, and then continued by interrupted sutures for the distal part; this allows discarding any excess length or a nonviable segment (Fig. 6c). The new ureter was reimplanted in the urinary bladder using the Lich Gregoir extravesical technique after resection of the native hypoplastic ureter. An illustrated diagram of the surgical technique in shown in Fig. 7.
(2) The ureteral stent, the pyelostomy tube, and the perinephric periureteric drain were left. Removal of the stent, the tube, and the drain was performed on days 7, 8, and 9, respectively.

Discussion

Ureteral hypoplasia is not well described in the English literature; it is usually associated with hypoplastic kidney and an ectopically inserted ureter, and it ends usually with unilateral nephrectomy as the renal parenchyma is usually lost at the time of diagnosis [6]. In our two cases, ureteral hypoplasia was wrongly diagnosed antenatally and postnatally as UPJ obstruction in the first case and as a huge renal cyst in the second one; correct diagnosis was established only intraoperatively, with a hugely dilated

Fig. 7



A diagram illustrating the steps of the renal pelvis flap technique for the replacement of a hypoplastic ureter. (a) The hugely dilated renal pelvis and the short hypoplastic ureter. (b) The dashed line represents the incision of the flap taken from the anterior surface of the renal pelvis. (c) The flap reflected downwards. (d) Tubularization of the flap and closure of the residual renal pelvis. (e) Reimplantation of the ureter into the urinary bladder. HU, hypoplastic ureter; RP, renal pelvis; UB, urinary bladder.

pelvis and a very narrow hypoplastic ureter. Trial of primary anastomosis was very difficult in one case and did not achieve adequate drainage of the renal pelvis, which necessitated a redo surgery using the flap technique.

Surgical correction for this anomaly is performed classically by replacement of the ureter by a segment of the ileal loop. Although it is a standard technique, it is a sophisticated technique with a long operative time and has its possible morbidity as the gastrointestinal tract is entered, and carries all risks of intestinal resection and anastomosis and a higher risk of wound infection due to breaching intestinal lumen, in addition to long-term complications: stone formation, electrolyte disturbance (hyperchloremic metabolic acidosis) that results from the absorptive surface of the graft, and the theoretical incidence of malignant transformation [4,5].

In our two cases, we used the hugely dilated pelvis to design the local pelvic flap, which tubularized as a conduit to replace the hypoplastic ureter and was then reimplanted in the urinary bladder. This technique has the advantage of the use of local flap, having the native urothelium in reconstruction, which is more physiological and avoids the potential complication of the intestinal graft. In these two cases, the hugely dilated pelvis and the young age of the patients made the ureteric length that is needed to be replaced short, making this technique feasible to replace the entire ureter.

Culp's ureteropelvioplasty, a technique using the pelvic flap in the correction of UPJ obstruction, is reported to replace the upper ureter in cases of upper congenital ureteral obstruction, which substitutes only a short segment, and not the whole ureteric length [7], and to our knowledge, there are no reported cases on the use of the renal pelvis flap to replace the entire ureter.

However, in this series, we did not perform intraoperative retrograde pyelography as it is not routinely carried out for the UPJ obstruction management protocol in our unit. Intraoperative retrograde pyelography could help in diagnosing and anticipating congenital ureteral anomaly before surgical exploration, especially in cases where there is a discrepancy between antenatal and postnatal ultrasonography [8].

Conclusion

The renal pelvis flap should be considered as an alternative procedure for ureteric replacement in cases of hypoplastic ureter to preserve the remaining renal parenchyma and to rescue the residual renal function. However, a comparative study on a larger number of cases with long-term follow-up and a comparison with other ureteric replacement techniques is needed to evaluate this technique.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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