Management of ureteropelvic junction obstruction associated with congenital upper urinary tract anomalies in children: single-center experience

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Purpose **We report our experience in the management of ureteropelvic junction obstruction in children with congenital upper urinary tract anomalies.**

Materials and methods **A retrospective review was performed for all children with primary ureteropelvic junction obstruction associated with upper urinary tract anomalies between 1999 and 2011 at our hospital. Medical records were reviewed for patient age, sex, type of urinary tract anomalies, affected side, indication of surgery, and operative details. Clinical and radiological outcomes were assessed. Success was defined as both symptomatic relief and radiographic resolution of obstruction at final follow-up.**

Results **There were 12 children (10 boys and two girls) with mean age of 5.3 years (range 1–13 years). Anomalies included ectopic pelvic kidney in six patients, crossed fused ectopia in two, a horseshoe kidney in two, and duplex renal units in two. The left kidney was affected in seven and the right kidney in five children. All patients were managed by open dismembered pyeloplasty. No significant perioperative complications were encountered in the study group. The mean follow-up was 30 months (range 15–45 months). Overall success rate was 84%. Relief of obstruction was evident in 10 patients as documented by intravenous urography or diuretic renography. Secondary nephrectomy was necessitated in two patients; one had severely impaired ipsilateral renal function and the other had recurrent pyelonephritis without radiological improvement.**

Conclusion **Despite the anomalous renal anatomy and the challenging surgical exposure, dismembered pyeloplasty in children with upper urinary tract anomalies has a high success rate with excellent functional results.** ***Ann Pediatr Surg*** **11:21–24 © 2015 Annals of Pediatric Surgery.**

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Introduction

Malformations of the urinary tract account for around 30% of all prenatally diagnosed congenital anomalies and are the predominant etiology of chronic kidney disease in children [1]. Among all these, ureteropelvic junction obstruction (UPJO) is the commonest pathological entity found. Association of UPJO with other upper tract anomalies such as renal ectopia, fusion, and duplication could happen with an incidence between 20 and 35% [2–4].

Open dismembered pyeloplasty has been considered the gold standard for surgical treatment of UPJO with a high success rate exceeding 90% [5]. In adult population with upper tract anomalies, both open and laparoscopic dismembered pyeloplasty have been performed successfully with good functional results [6]. In addition, robotic surgery has gained increasing popularity and acceptance in urologic practice, providing improved operative performance and simplified suturing [7]. There are limited data on surgical management of UPJO in the pediatric age group; in addition, it is not clear which surgical technique to use. Thus, the current study was undertaken to report our experience in surgical management of UPJO in context with kidney anomalies in children.

Materials and methods

After obtaining ethical committee approval, a retrospective review of the medical records of 290 children who had undergone open dismembered pyeloplasty for primary UPJO at our institute between 1999 and 2011 was carried out. UPJO was associated with congenital upper urinary tract anomalies in 12 patients (4%). Anomalies included ectopic pelvic kidney in six patients, crossed fused ectopia in two, a horseshoe kidney in two, and a duplication anomaly in two. The left kidney was affected in seven and the right kidney in five children. Major presenting symptom included abdominal mass in five patients (42%), abdominal pain in four (33%), and incidentally discovered in three (25%). The two patients with duplex kidneys were diagnosed antenatally. Patients who underwent previous surgery were excluded from the study.

Routine abdominal ultrasound was performed in all patients as an initial step of diagnosis. Further evaluation was carried out by either excretory urography, magnetic resonance urography, or computerized tomography (Figs 1–3). Diuretic renography was required in all cases to evaluate the obstruction and as a baseline for follow-up. Diethylene triamine penta-acetic acid scans were performed preoperatively and 6 months after pyeloplasty to evaluate renal functional drainage. Drainage was classified as excellent if T-half was less than 20 min; delayed if T-half was greater than 20 min with a descending drainage curve; or poor if T-half could not be counted and there was a plateau/rising drainage curve. The kidney function was classified as good if split renal

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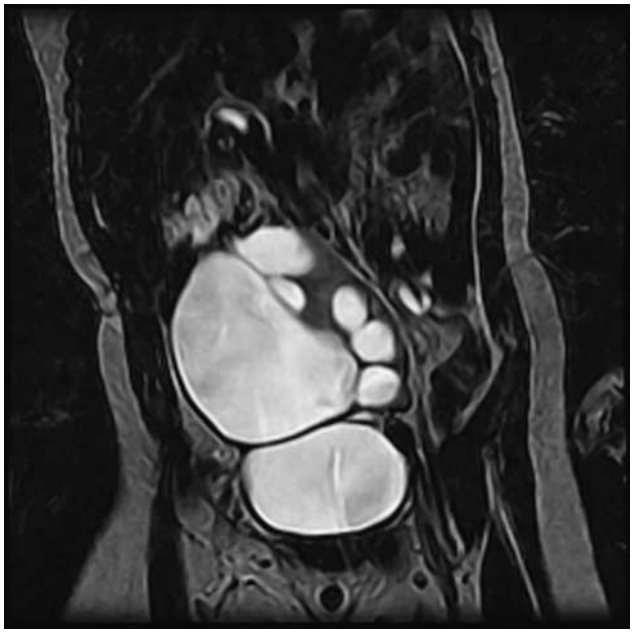
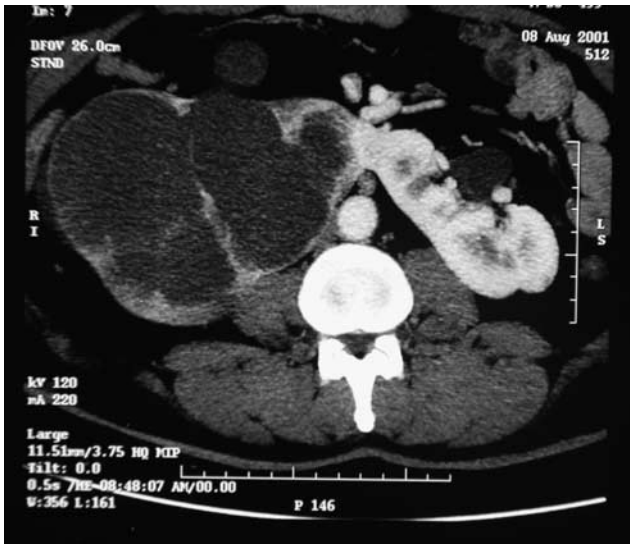
Fig. 1



Coronal magnetic resonance urography (MRU) image in a 4-year-old girl with ectopic pelvic right kidney showing advanced hydronephrosis and hugely dilated renal pelvis.

Fig. 2

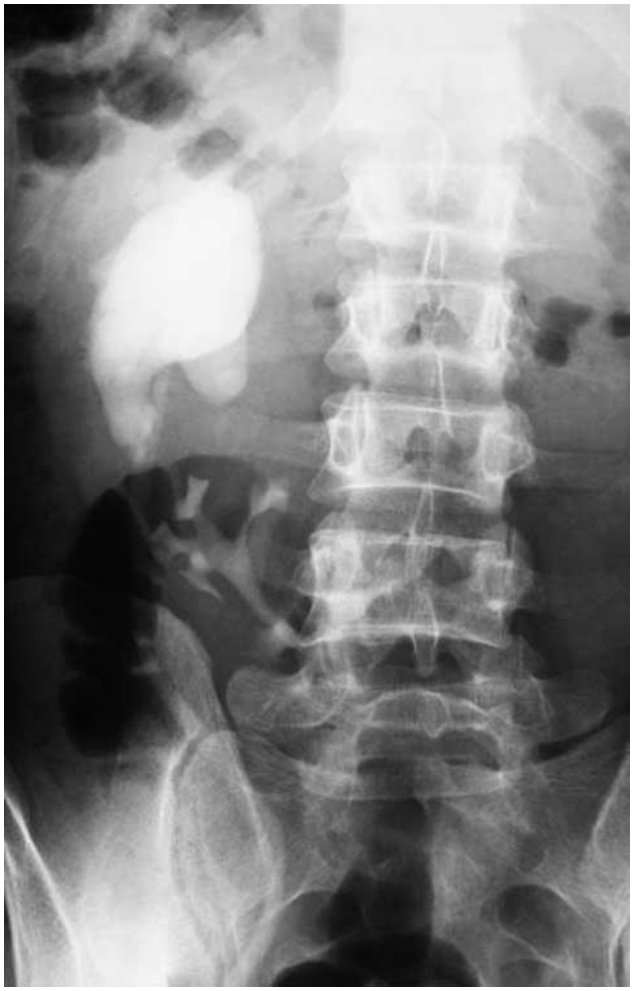


Axial computerized tomography (CT) image of the abdomen in a 10-year-old boy showing horseshoe kidney with marked hydronephrosis of the right compartment due to ureteropelvic junction obstruction.

function was more than or equal to 40% or impaired if less than 40%. Two patients with an ectopic pelvic kidney had severe hydronephrosis and impaired renal function before surgical correction.

All cases underwent surgery once the diagnosis is made without delay. Open reconstruction of the UPJ using the Anderson–Hynes dismembered pyeloplasty technique was performed in all patients. Anterior lumbar incision was adopted in cases of crossed ectopia, duplex renal units,

Fig. 3



Intravenous pyelography (IVP) image of a 13-year-old boy showing crossed fused ectopic left kidney with perfect collecting system and moderate hydronephrosis of the right kidney due to ureteropelvic junction obstruction.

horseshoe kidneys, and bifid renal pelvis, whereas extended parainguinal incision was adopted in cases of ectopic pelvic kidneys. Crossing vessel was present in one patient with ectopic pelvic kidney; the ureter and the renal pelvis were transposed to the opposite side of the vessel before completion of the anastomosis. In one of the two cases of horseshoe kidneys, dismembering of the fibrous isthmus was required to allow funnel-shaped and dependent anastomosis. High ureteral insertion was present in the case of crossed ectopic kidney, whereas the two cases of UPJ in the lower moiety of duplex renal unit were managed by dismembered pyeloplasty. Ureteropelvic anastomosis was carried out using either 6-0 polyglycolic acid or polydioxanone sutures. At the end of the anastomosis, a nephroureterostomy stent was introduced through the renal pelvis and came out to the skin by stab incision. Following completion of the anastomosis, a perinephric drain was placed by another stab incision.

Patients were scheduled for stent removal 7–10 days after pyeloplasty. Renal ultrasonography was performed 1 month after stent removal, then diuretic renography

was performed 6 months postoperatively and clinical evaluation annually thereafter. Success was defined as both clinical improvement and radiographic resolution of obstruction at final follow-up together with stability of the differential renal function.

Results

A total number of 12 children (10 boys and two girls) with mean age 5.3 years (range 1–13 years) with congenital upper urinary tract anomalies underwent dismembered pyeloplasty for UPJO. No significant perioperative complications were encountered in any patient.

The mean follow-up was 30 months (range 15–54 months). Overall success rate was 84%. Relief of obstruction was evident in 10 patients as documented by ultrasonography and diuretic renography in addition to the relief of symptoms (Table 1). One patient developed recurrent attacks of pyelonephritis with impaired ipsilateral renal function (12% differential function); hence, secondary nephrectomy was performed. However, the other patient with ectopic pelvic kidney showed stable differential function with no resolution of the hydronephrosis on ultrasonography over 24 months. Moreover, this patient developed two attacks of pyelonephritis and underwent secondary nephrectomy.

Discussion

UPJO in association with other upper urinary tract anomalies is a rare occasion. Nevertheless; open pyeloplasty is considered the gold standard surgical management in normal kidneys with UPJO, pyeloplasty technique in anomalous kidneys is not standardized. Kidney abnormalities may include abnormal position and shape of the renal pelvis, abnormal insertion of the ureter, or malrotation of the calyces, all of which may alter the technique used in surgical correction of associated UPJO. In this cohort of patients, we reviewed our data to see whether standard dismembered Anderson–Hynes pyeloplasty is still considered the appropriate choice for such cases or not.

Pelvic kidney presents a special surgical entity because of its anomalous position and vasculature with high risk for injuring abnormal vessels or nearby abdominal viscera and structures [4,8]. Both antegrade and retrograde endopyelotomy were utilized for treatment of UPJO in ectopic kidneys [9,10]. In contrast, laparoscopic dismembered pyeloplasty proved efficient in some cases [5]. Simone *et al.* [11] tried to create a tubularized renal pelvic flap with pelvic reduction to correct the obstruction. It does bear mention that these kidneys have some degree of pyelocaliectasis that can be mistaken for UPJO. Hence, it is prudent to exclude obstruction by diuretic renogram before surgery [6].

Horseshoe kidney is the commonest fusion anomaly. The success rate for open pyeloplasty ranges from 55 to 80% [2,7,12]. In adult patients with abnormally located and malrotated kidneys associated with UPJO, laparoscopic pyeloplasty is described with a success rate of 91%, which is better than that of open surgery for UPJO in horseshoe kidneys [5,6]. Some advocate division of the isthmus and lateral fixation of the lower pole of the affected kidney that allow more medial and dorsal orientation of the ureter and a dependent postpyeloplasty funnel [13]. Others suggest no division assuming that it is unnecessary in view of normal peristalsis of the ureter [7]. In our series, a dismembered pyeloplasty was performed for two children with horseshoe kidneys with division of the isthmus, as we believe that ureteral angulation and obstruction at the level of the isthmus should be appreciated, and the two children in this series showed good clinical and radiological results.

Despite duplex kidney is a common anomaly, association of lower pole with UPJO is rarely encountered in pediatric urologic practice. In complete ureteral duplication, treatment of lower pole UPJO can be accomplished with a dismembered pyeloplasty as in kidneys with a single ureter. In contrast, incomplete ureteral duplication can pose a technical challenge, especially when the junction of the upper and lower poles ureters is proximally located and in close proximity to the lower pole UPJ [14]. VanderBrink *et al.* [14] in their review of eight patients with lower pole UPJ obstruction documented that ureteral length between the UPJ and junction of the lower pole and upper pole ureter was the major determinant of which reconstructive technique they used. In case of a short lower pole ureter, pyeloureterostomy was an attractive option, whereas if long, standard pyeloplasty would be sufficient. Similarly, in our series, we performed standard pyeloplasty in two patients.

Over the last few years, laparoscopic and robotic techniques have become more popular showing high success rates. There is a large series from Nayyar *et al.* [15] who presented an overall success rate of 96.6% in 29 children using robotic pyeloplasty. Unfortunately, we did not have such experience in this technique in patients with abnormal anatomy.

Table 1 **The study group characteristics**

Age	Sex	Side	Chief complaint	Anomaly	Preoperative split function %	Postoperative split function %	Follow-up (months)
13	Male	Left	Mass	Crossed ectopic	18	22	35
1	Male	Right	Mass	Ectopic pelvic	26	29	38
8	Female	Left	Pain	Ectopic pelvic	22	19	48
10	Male	Right	Pain	Horseshoe kidney	38	41	19
3	Male	Left	Mass	Ectopic pelvic	24	27	46
5	Male	Right	Pain	Duplex kidney	25	32	26
4	Male	Left	Mass	Ectopic pelvic	16	12	54
3	Male	Left	Incidental	Crossed ectopic	22	20	17
6	Male	Left	Pain	Ectopic pelvic	17	23	24
7	Male	Right	Incidental	Horseshoe kidney	18	22	21
4	Female	Left	Mass	Ectopic pelvic	15	13	18
2	Male	Right	Incidental	Duplex kidney	36	38	15

The overall success rate of our series was 84% (100% in horseshoe kidneys, 100% in duplicated anomalies, and 75% in ectopic kidneys), and we thought that our results cope favorably with the results in the literature concerning the adult series. In comparison with our results of pyeloplasty in cases with primary UPJO without upper tract anomalies, the success rate was low (84 vs. 97%), and this is logic [16]. This is in agreement with the recent report by Helmy *et al.* [4] who showed an overall success rate of 82.6% for pyeloplasty in ectopic pelvic kidneys.

To our knowledge, there are only few reports about surgical management of UPJO in children with upper tract anomalies [4,6–9]. We believe that this series may serve as a guide to consider standard Anderson–Hynes pyeloplasty as the most accepted procedure in treatment of such cases even with some modifications of the technique. There are some inherent weaknesses in our study. First, it was a retrospective study, and therefore subject to shortcomings of nonprospective design. In addition, there was a small number of cases; it would be better if a multicenter study design was performed. Moreover, the Anderson–Hynes technique was used as the sole surgical procedure and we did not use any other technique to compare with, which may give a better outcome. In addition, our study was conducted over a long period of time in which an anticipated difference in experience and training level of the operating surgeons would affect our results.

Conclusion

Despite the complex renal anatomy and the challenging surgical dissection, dismembered pyeloplasty in children with upper urinary tract anomalies has a high success rate with excellent functional results. Ectopic pelvic kidneys should be evaluated carefully before surgical intervention to confirm obstruction. In patients with horseshoe kidneys, division of the renal isthmus needs to be considered, if it is the cause of ureteral angulation and obstruction. In duplicate renal anomalies associated with UPJO, precise knowledge of the anatomy of both ureters is mandatory for planning successful surgical reconstruction.

Acknowledgements

Conflicts of interest

There is no conflicts of interest.

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