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Giant hydronephrosis in children: resection or preservation, what is the best approach?

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Abstract: The purpose of this study is to assess functional and morphologic recovery in children after dismembered pyeloplasty for GH caused by UPJO of children. From Jan 2005 to Dec 2010, 38 pediatric patients with GH who underwent dismembered pyeloplasty performed by a single surgeon were retrospectively analyzed. Patient follow-up was at least 3 years. The outcomes of different types of UPJO were compared. DRF and RPT assessed more than 3 years postoperatively were compared according to ages and time. Improvement of DRF and RPT was also compared. The relationship between the improvement in DRF in the first six months and renal function assessed in 3-year was also analyzed. The results showed that the renal function of functional obstruction was significantly lower than other groups ($p < 0.0001$). GH patients who underwent pyeloplasty before 36 months of age (n = 12) experienced more improvement in DRF compared with GH patients who underwent surgery after 36 months of age (n = 21) ($p < 0.0001$). DRF measured in the first six postoperative months was significantly higher than other groups (n = 12) ($p < 0.0001$). There is a positive positive correlation between DRF in the first six months and renal function assessed in 3-y ($r = 0.8193$). In conclusion, it is worthwhile to preserve the kidney in children with GH. Functional obstruction should be independent from UPJO and surgical treatment is inadvisable. The rate of improvement in DRF in the first six months is a good indicator to outcome.

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1. Introduction

Giant hydronephrosis (GH) is a common disease in children. The most common cause of this lesion is hydronephrsis due to ureteropelvic junction obstruction (UPJO) [1, 2]. The incidence of nephrectomy for GH caused by UPJO ranges from 3% to 70% in the reported literature [3-5]. In these articles, the underlying cause of the UPJO is not described. This disease process can be broadly divided into intrinsic or extrinsic causes [6]. The intrinsic causes include aperistaltic segment of the uteter, which we call functional obstruction, kinks or valves produced by infoldings of the ureteral mucosa and musculature [7]. The extrinsic causes include high insertion of the ureter, "aberrant" or "crossing"vessels. Presently, dismembered pyeloplasty [2] is the standard procedure to treat this condition. The purpose of surgery is to drain the urine and release pressure of kidney as soon as possible.

In our clinic, some patients who had severely compromised kidney function due to GH showed remarkable recovery after surgery especially in younger children. Conversely, in some children the kidney function remained poor or continued to deteriorate. The factors associated with these different outcomes are poorly understood. Until now, the reason for this phenomenon has been unclear, and there is little data referencing this topic. Moreover, there is little information about how the function and morphology of the GH kidney recovers postoperatively. The rate of recovery also has not been characterized. In this study, we assigned each case to one of three groups (intrinsic obstruction, extrinsic obstruction, or functional obstruction) and compared the outcome of pyeloplasty. We measured changes in DRF and RPT postoperatively and correlated these changes with preoperative characteristics to explore potential causes of the differing outcomes. Furthermore, we sought to identify factors associated with a good post-operative prognosis.

2. Materials and methods

We performed a retrospective review of children who underwent dismembered pyeloplasty for GH at our hospital between Jan 2005 and Dec 2010. Data collected included: age at the time of surgery, gender, laterality, presenting symptoms, preoperative imaging, surgical details, follow-up duration and postoperative course. Only cases with primary unilateral UPJO with GH were included, whereas patients with vesicoureteral reflux， bilateral UPJO, UPJO in solitary kidney and recurrent cases were excluded from the study. According to the Society for Fetal Urology (SFU) grading system, all the cases were grade IV. This study was performed in accordance with the principles of the Declaration of Helsinki.

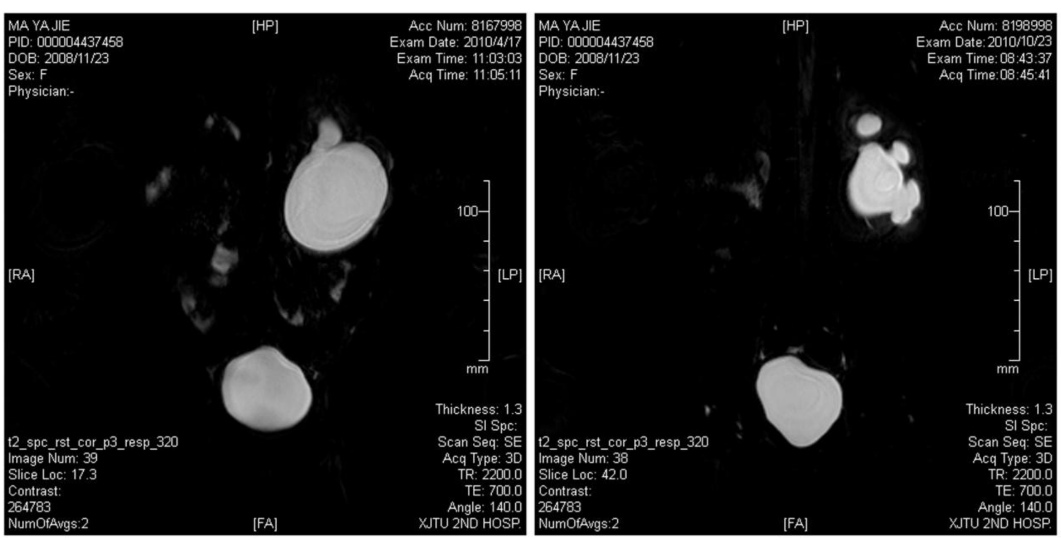
Preoperative radiology included renal ultrasound for all patients. DTPA scans were performed preoperatively and at 6, 12, 24 and 36 months after surgery. Patients were followed up for at least 3 years.

Based on the preoperative and postoperative examination, including the DTPA renal scan, diuresis renography, ultrasound, CT, MRI and the examination of post-operative specimens, patients were divided into three groups: intrinsic obstruction, extrinsic obstruction and functional obstruction. Functional obstruction included patients without symptoms, peristalsis disability indicated by diuresis renography, and the absence of identification of stenotic segments at surgery. According to the age, patients were divided into three groups: under 3, 3 to <6 and 6 to <10 years of age. According to the time after surgery, patients were divided into five groups: 0 to 6, 6 to 12, 12 to 24, and 24 to 36 months. DRF and RPT were compared between those groups. Improvement of DRF and RPT at 3 years after surgery was also compared. The relationship between the improvement of renal function recovery in the first six months and the final outcome was also analysis.

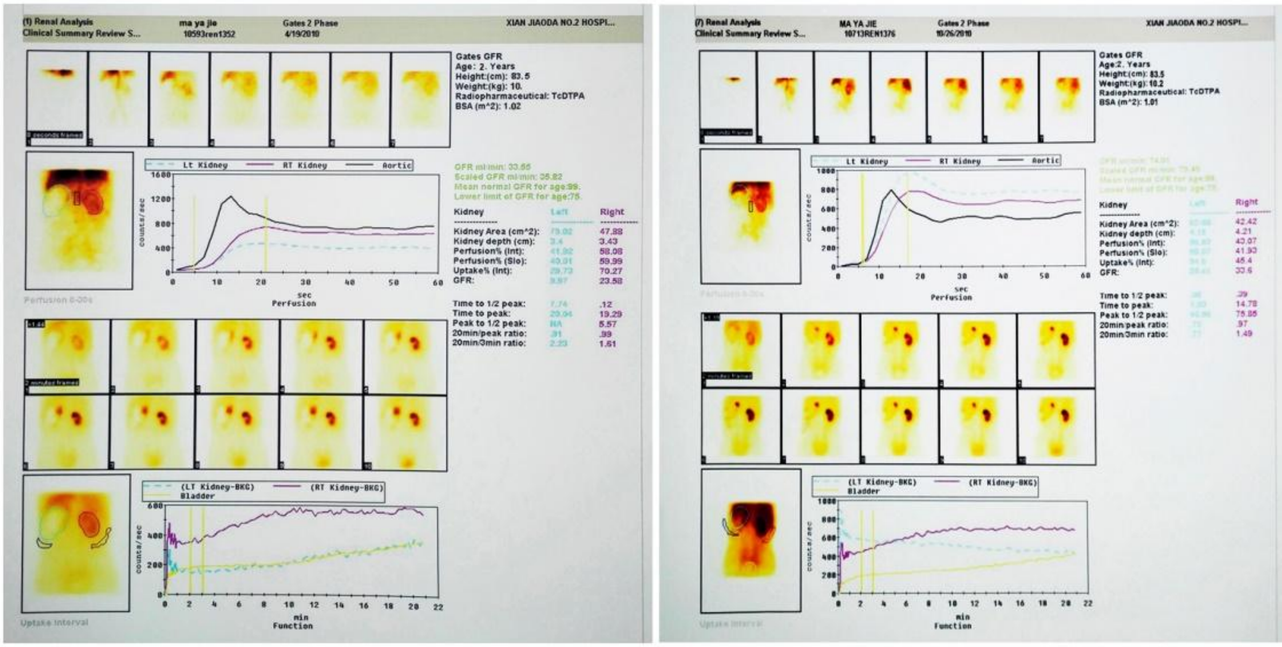
Statistical analysis was performed using SPSS 17.0 for Windows (SPSS, Inc., Chicago, IL). Statistical significance was analyzed by 1way- ANOVA among those groups. A univariate analysis was used to assess the possible prognostic factors for the success of pyeloplasty by chi-squared. Spearman r correlation was used to analyze the degree and correlation between the improvement of renal function in the first six months and final outcome. For all tests statistical significance was considered at $P \leqslant 0.05$.

3. Results

During this period, a total of 38 children had GH caused by UPJO and underwent dismembered pyeloplasty. Among the 38 GH children, 15 (39%) presented with abdominal mass, 8 (21%) with prenatal hydronephrosis, 3 with febrile urinary tract infection, and 2 with flank pain. In the total of 38 patients, 19 (49%) was caused by extraluminal factors, including the oppression of fibrous band, aberrant vessels and high ureter. 14 (37%) was caused by intraluminal factors, including stenosis, valvular, polyps. 5 (13%) was caused by functional factors. The mean age at surgery was 34.6 months (range 9-118 months). Postoperative DRF improved obviously in 32 patients (84%), increased insignificantly in 5 (13%), and deteriorated in 1 (3%). The degree of DRF improvement was in the range 9 to 41% (Pic. 1,2).



Pic.1. These dramatic before (left) and after Surgery (right) MR photos show the change of hydronephrosis.



Pic.2. These dramatic before (left) and after Surgery (right) DTPA renal scan photos show the change of Differential renal function (DRF).

Exclusive of functional group, those who underwent pyeloplasty before 36 months (n=12) had more improvement in RPT or DRF compared with patients who underwent surgery after 36 months (n= 21，$p<0.05$) (Table 1). In the first six months after surgery, the RPT or DRF improvement was higher than other period ($p<0.0001$) (Table 2).

Table 1. Pre- and postoperative clinical parameters according to the age

Age	0-3	3-6	6-10	*P*
Number of patients	12	13	8	-
Male/female (%)	8/4	9/4	5/3	-
Right/left (%)	3/9	5/8	4/4	-
RPT, preoperative USG, mm	2±0.8	2±0.8	2±0.7	0.9705
RPT, postoperative 3-y USG, mm	9±1	7±1	7±1	0.0004
Improvement, mm	6±0.8	5±1	5±0.9	0.0015
DRF, preoperative DTPA, %	13±3	12±3	13±3	0.6171
DRF, postoperative 3-y DTPA, %	45±3	40±1	35±3	<0.0001
Improvement, %	32±1	27±1	23±1	0.0003

RPT=renal parenchyma thickness; DRF=differential renal function. Values are shown as mean ± SD. Statistical significance was analyzed by 1way- ANOVA.

Table 2. Pre- and postoperative clinical parameters according to the time after surgery

	Preoperative	0-6	6-12	12-24	24-36	*p*
RPT, USG, mm	2±0.7	5±1	7±2	7±2	7±2	<0.0001
Improvement, mm	0	19±6	4±2	2±2	1±1	<0.0001
DRF, DTPA, %	12±3	31±7	35±7	36±8	38±9	<0.0001
Improvement, %	0	3±1	2±0.7	0.4±0.8	0.1±0.5	<0.0001

RPT=renal parenchyma thickness; DRF=differential renal function. Values are shown as mean ± SD. Statistical significance was analyzed by 1way- ANOVA.

Patients with functional obstruction had a worse prognosis than other groups ($p < 0.0001$) (Table 3). Among 5 children, only one's postoperative renal function (20%) improved. Three patients (60%) had no obvious changes. The other one (20%) has further deterioration of renal function and eventually received nephrectomy.

In the correlation analysis, we found, the improvement of renal function in the first six months had a positive relationship with prognosis (r=0.8193) (Table 4).

Table 3. Pre- and postoperative clinical parameters according to the cause

	Extrinsic obstruction	Intrinsic obstruction	Function obstruction	*p*
Number of patients	19	14	5	-
Male/female (%)	13/6	8/6	3/2	-
Age	23-118	17-114	9-38	-
RPT, preoperative USG, mm	3±0.9	2±0.0	2±0.0	0.0564
RPT, postoperative 3-y USG, mm	8±1	7±0.9	3±1	<0.0001
Improvement, mm	5±1	5±0.9	1±1	<0.0001
DRF, preoperative DTPA, %	13±3	12±3	11±1	0.3462
DRF, postoperative 3-y DTPA, %	42±5	39±4	20±7	<0.0001
Improvement, %	29±5	28±3	9±7	<0.0001

RPT=renal parenchyma thickness; DRF=differential renal function. Values are shown as mean ± SD. Statistical significance was analyzed by 1way- ANOVA.

Table 4. Correlation of improvement in DRF in the first six months and renal function assessed in 3-year

Time after surgery (Months)	0-6	12-24	24-36	*r*
Improve DRF, %	34±4	40±4	41±4	0.8193

DRF=differential renal function. Values are shown as mean ± SD. Statistical significance was analyzed by t test.

4. Discussion

Giant hydronephrosis was described as the massive dilatation of the kidney and renal pelvis. In 1939, Stirling defined it as the presence of more than 1,000 ml of fluid in the collecting system [8]. In children, the radiological finding used to define giant hydronephrosis is a kidney that occupies at least half the abdomen, meets or crosses the midline, or spans at least five vertebral lengths [9, 10]. According to the report, the most common reason that cases giant hydronephrosis is UPJO [11]. Shokeir *et al* [12] reported that [2-9] neonates per 1,000 births have urogenital anomaly and 50-87% of these have hydronephrosis.

At present, there are few rules or protocols for the management of a poorly functioning kidney. Urologist always use 10% as the criterion whether to remove the kidney [13, 14]. However, recent studies suggest that even if the DRF is less than 10%, the damaged kidney is worthy to be preserved, especially to children [15, 16]. In our study, patients with intrinsic or extrinsic obstruction preoperatively steadily improved after dismembered pyeloplasty, mean DRF of the 33 kidneys in long-term follow-up was 40.55%. These results imply that a giant hydronephrotic kidney with thin renal parenchyma may be salvageable in children. It is likely that the renal function could be conserved even if the hydronephrosis was severe because highly compliant collecting system alleviated the effect of pressure on the kidney parenchyma. And the other important reason is that compared the adult, the renal function of children is continue to develop until 10 years old [17]. This result was also reflected in our data. Analyzed by age at operation, the kidneys of children under 36 months increased in RPT more than that of children over 36 months. This is likely attributable to the fact that normal human renal function maturation is most rapid in infancy and then slowly decreases [18].

The surgical treatment of UPJO with a huge renal pelvis remains challenging. Successful pyeloplasty results in the relief of symptoms and an improvement in renal drainage; however, not all kidneys show improvement after surgery. In our study, the renal function keep stable or grow deteriorate in the group with functional UPJ obstruction after surgery. Most of these patients were diagnosed in fetal period and were asymptomatic. The samples of patients who receive nephrectomy were not found obviously stenotic segment. Furthermore, the data shows the split renal function of this group is worse than other groups. The potential reasons are not clear. Some researchers found congenital ureteropelvic junction obstruction always accompanied with the dysplasia of interstitial cells of cajal and smooth muscle cells [19-21]. Interstitial cells of cajal and smooth muscle cells play very important role in ureteral peristalsis [22]. In our study, the preoperative diuresis renography show that after injection of diuretics, the curve of renogram decreased significantly. This implies that the cause of blockage is not obstruction but problems with ureteral peristalsis. Onen *et al*. [23] suggested that some children detected prenatally with unilateral and bilateral hydronephrosis may not be truly obstructed and may not need early intervention. This confirmed our results. For such patients, it is essential to promote ureteral peristalsis instead of removing the potentially stenotic segment. However, at present, we have no effective means to differentiate these different types of UPJO.

We noticed that, compared with other period, postoperative renal function recovery was fastest in the first six months. Deng *et al* [24] found that after nephrostomy, parenchymal thickness increased most rapidly within the first 4 weeks. They noted that, in the first 2 weeks, parenchymal thickness increasing is a “cheating” phenomenon that results from kidney shrinkage; and the true change in thickness occurs in the subsequent 2 weeks. Our results are in accord with his study. The recovery of renal function is always ahead the parenchyma thickness increases. This phenomenon gives us an enlightenment that using parenchyma thickness as the predictor of renal function is not accurate. Sometimes, what it reflects is not the real condition of kidney.

There is debate in the literature about which factors contribute significantly to RF recovery after pyeloplasty in the pediatric population. Konda *et al* [25] holds that parenchymal thickness is a good indicator of future renal function. But Rodríguez *et al* [26] points that thin kidney parenchyma is not associated with worse function and the ratio of parenchymal-to-pelvic area is better to assess the renal function. Ahmed M *et al* [27] believed baseline DRF was the only predictor of renal functional recovery after the relief of obstruction. The group with less than 40% function was most likely to improve regardless of renal US findings. In this study, preoperative DRF of most cases was significantly improved after dismembered pyeloplasty. The result is confirmed with Ahmed. Moreover we found the improvement of renal function in the first six months had a highly positive relationship with prognosis (r =0.8193). We believe if patients’ renal function recovers quickly in the first six months after surgery they have a good prognosis.

Furthermore, we noticed the range of kidney function recovery is from 9% to 41%. But the morphology is never come back. We always tell our patients that hydronephrosis is normal after dismembered pyeloplasty, but we seldom consider the peristaltic function recovery, and there is no study focusing on this issue. Sarhan O *et al* [28] reported that there were 65% cases, after surgery, which obstruction still exited. Moreover, the renal function included in this study is not too bad. As we know, if 30% of the kidney is functional, then renal function is normal. But why there are so many postoperatively patients which renal function and morphology are not normal. Weiss *et al* [29] reaches conclusion that ureteral peristalsis is originated from the proximal portion of the urinary collecting system. We presupposed that the peristaltic center was broken in the procedure of hydronephrosis. If this peristaltic center was damaged, the function of ureteral peristalsis is affected. So after surgery, the obstruction still exit and it is real functional obstruction. However, we pay more attention to the recovery of morphology of kidney. In fact, salvaging peristaltic function after surgery is an important problem we face.

This study has some limitations. Only children were included, and the age range is narrow. Furthermore, we need more cases to reach more definitive conclusions. Nevertheless, we think this study provides some useful information for urologists to understand how the kidneys recover after obstruction is relieved, and more work is needed in dealing with ureteral functional obstruction and promoting the peristalsis of the ureter after surgery.

5. Conclusions

The most common cause for giant hydronephrosis is UPJO. However，there is no study that uses function obstruction as an independent factor to observe the outcome of GH caused by UPJO. In this study, we divided all the UPJO cases into three groups (intrinsic obstruction, extrinsic obstruction, functional obstruction) and compared the outcome after pyeloplasty. We found a statistically significant difference among the three groups and functional obstruction was associated with an unfavorable outcome. While these initial data require further validation, we believe that functional obstruction is independent from UPJO and surgical treatment is inadvisable.

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Conflict of interest statement

None declared.

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