

Pediatric Urology

# Management of Severe Bilateral Ureteropelvic Junction Obstruction in Neonates with Prenatally Diagnosed Bilateral Hydronephrosis

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**Purpose:** The management of prenatally detected bilateral ureteropelvic junction obstruction (UPJO) remains controversial. We attempted to develop a treatment plan for patients with severe bilateral UPJO.

**Materials and Methods:** We evaluated the records of 13 patients with prenatally diagnosed grade 3 or more bilateral hydronephrosis that led to the postnatal diagnosis of severe bilateral UPJO. Ultrasonography and <sup>99m</sup>Tc-mercaptoacetyltriglycine (<sup>99m</sup>Tc-MAG3) renal scans were performed within 1 month. Four renal units had grade 3 and 22 had grade 4 hydronephrosis. All 13 patients were managed by unilateral pyeloplasty, and the patients' mean age was 3 months. At 1 month postoperatively, we decided whether delayed surgery in the opposite renal unit was necessary according to the findings of ultrasonography and <sup>99m</sup>Tc-MAG3 scans.

**Results:** Of 13 patients, 11 underwent initial pyeloplasty on renal units with more severe hydronephrosis or lower relative renal function (RRF) on <sup>99m</sup>Tc-MAG3 scans. The remaining 2 patients simultaneously underwent percutaneous nephrostomy on renal units with a lower RRF and initial pyeloplasty on renal units with a higher RRF. In 5 patients, contralateral hydronephrosis had spontaneously improved at 1 month postoperatively, and 8 patients underwent delayed contralateral pyeloplasty at 2 months postoperatively.

**Conclusions:** In children with severe bilateral UPJO, the non-operated renal units with grade 3 and some with grade 4 hydronephrosis improved spontaneously after unilateral pyeloplasty. Therefore, delayed pyeloplasty of the opposite side should be considered at 1 month following initial pyeloplasty.

**Key Words:** Hydronephrosis; Ultrasonography; Ureteral obstruction

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## INTRODUCTION

The introduction and widespread use of antenatal ultrasonography has increased the diagnosis of fetal hydronephrosis in utero [1-3]. Antenatal ultrasound screening detects fetal hydronephrosis in around 1% of fetuses [1,4]. One of the most common causes of antenatally diagnosed hydronephrosis is ureteropelvic junction obstruction (UPJO) [3]. Many reports deal with the unilateral idiopathic UPJO with a functionally, ultrasonographically, and renographi-

cally normal contralateral renal unit and many researchers manage their patients by initially conservative approaches with subsequent ultrasonographic or renographic follow-up [5-7]. Compared with unilateral UPJO, however, few published papers deal with the management protocol for neonatal patients with prenatally diagnosed bilateral hydronephrosis that leads to the postnatal diagnosis of severe bilateral UPJO.

Factors to be considered in planning the surgical treatment of neonatal severe bilateral UPJO are as follows.

Would we operate urgently? If yes, would we operate on both renal units simultaneously or separately? If separately, which renal unit should be corrected first? How long can we wait to correct the later-operated renal unit after the initial pyeloplasty? Is it necessary to place percutaneous nephrostomy (PCN) tube drainage to the later-operated renal unit for the preservation of renal function during the waiting period? These questions have been on the minds of many pediatric urologists for a long time. But it is true that there is a paucity of clear recommendations regarding the management of severe bilateral UPJO. Therefore, we evaluated the clinical outcome of treatment in a prenatally diagnosed group with severe bilateral UPJO at our institution to offer suggestions for the management of these clinical situations.

## MATERIALS AND METHODS

From January 2000 to March 2009, 12 boys and 1 girl (26 renal units) with antenatally diagnosed grade 3 or higher bilateral hydronephrosis that led to the diagnosis of severe bilateral UPJO on postnatal ultrasonography at 2 days after birth were selected. Infants with grade 2 or less hydronephrosis in both renal units and those who had other urinary tract anomalies were excluded. The medical records of the selected 13 patients were reviewed for their hospital course and their postoperative follow-up imaging studies by using ultrasonographic and renographic assessment were reviewed on the Picture Archiving & Communications System (PACS). All neonates underwent voiding cystourethrography (VCUG) at 2 weeks after birth to rule out reflux diseases, but no reflux was found on VCUG.

Within 4 weeks after birth, ultrasonography was repeated for the reassessment of hydronephrosis and a  $^{99m}\text{Tc}$ -mercaptoacetyltriglycine ( $^{99m}\text{Tc}$ -MAG3) renal scan using the standard intravenous furosemide and 20-minute

delay technique was performed for evaluating preoperative relative renal function (RRF) and diuretic drainage characteristics. The hydronephrosis of each renal unit with UPJO was graded according to the Society for Fetal Urology (SFU) grading guidelines from 1 to 4 [8]. Among the renal units with hydronephrosis of SFU grade 4 in particular, we defined SFU grade 5 as hydronephrosis with 'paper thin' renal parenchyme. All neonates had SFU grade 3 or higher bilateral hydronephrosis and SFU grade 4 or 5 hydronephrosis was involved in all of the cases.

All cases of dismembered Anderson-Hynes open pyeloplasty through a flank incision were performed by a single surgeon (K.S.K.). Subsequent ultrasonography and  $^{99m}\text{Tc}$ -MAG3 renal scans were performed at 1 month postoperatively and we then decided whether delayed pyeloplasty in the opposite renal unit was necessary according to the findings of the ultrasonography and  $^{99m}\text{Tc}$ -MAG3 renal scan.

## RESULTS

Of 26 renal units, 4 renal units had hydronephrosis of SFU grade 3 and 22 had hydronephrosis of SFU grade 4 or more on postnatal ultrasonography (Table 1). The hydronephrosis grade and RRF for both renal units in each neonate at the preoperative, the postoperative, and the most recent follow-up are given in Table 1. A total of 13 patients were managed surgically by initial unilateral dismembered open pyeloplasty, and the mean age of the patients at the time of surgery was 3 months (range, 1-11 months). The mean follow-up time after the initial pyeloplasty was 37 months (range, 5-84 months).

Of 13 patients, 9 (cases 1-9) had bilateral UPJO with different degrees of hydronephrosis of both renal units on ultrasonography at 2 weeks after birth. These 9 patients underwent initial unilateral dismembered open pyeloplasty on the renal unit with a higher SFU hydronephrosis grade

**TABLE 1.** Changes in hydronephrosis and renal function in patients with bilateral hydronephrosis undergoing pyeloplasty during early infancy

|    | 2 weeks  |           | 1st OP<br>(PCN) | Post 1st OP |        | Post 2nd OP |           |        | Last f/u     |          |
|----|----------|-----------|-----------------|-------------|--------|-------------|-----------|--------|--------------|----------|
|    | HN (R:L) | RRF (R:L) |                 | HN (R:L)    | 2nd OP | HN (R:L)    | RRF (R:L) | 3rd OP | Age (months) | HN (R:L) |
| 1  | 4:3      | 17:83     | R               | 2:2         | -      | -           | -         | -      | 59           | 1:0      |
| 2  | 4:3      | 51:49     | R               | 3:2         | -      | -           | -         | -      | 55           | 1:2      |
| 3  | 3:5      | 53:47     | L               | 1:1         | -      | -           | -         | -      | 12           | 1:1      |
| 4  | 4:3      | 66:34     | R               | 2:2         | -      | -           | -         | -      | 88           | 2:1      |
| 5  | 5:4      | 47:53     | R               | 0:3         | -      | -           | -         | -      | 13           | 2:3      |
| 6  | 5:4      | 51:49     | R               | 0:4         | L      | 0:0         | 51:49     | -      | 9            | 0:0      |
| 7  | 4:5      | 57:43     | L               | 4:4         | R      | 4:4         | 49:51     | -      | 8            | 2:2      |
| 8  | 4:5      | 59:41     | L               | 4:4         | R      | 1:5         | 56:44     | L      | 37           | 0:4      |
| 9  | 4:5      | 48:52     | L               | 4:4         | R      | 4:4         | 47:53     | -      | 5            | 3:3      |
| 10 | 4:4      | 48:52     | R               | 0:4         | L      | 0:2         | 51:49     | -      | 64           | 0:2      |
| 11 | 5:5      | 53:47     | L               | 4:4         | R      | 4:3         | 53:47     | -      | 43           | 1:1      |
| 12 | 5:5      | 44:56     | L(R)            | 5:2         | R      | 2:1         | 24:76     | -      | 37           | 0:1      |
| 13 | 5:5      | 64:36     | R(L)            | 5:5         | L      | 3:5         | 63:37     | L      | 84           | 2:4      |

HN: hydronephrosis, RRF: relative renal function, OP: operation, f/u: follow-up, PCN: percutaneous nephrostomy, R: right, L: left

on ultrasonography. Two patients (cases 10 and 11) had bilateral UPJO with the same degree of hydronephrosis of both renal units on ultrasonography and their  $^{99m}\text{Tc-MAG3}$  renal scan demonstrated that the difference in RRF between each renal unit was not significant. These 2 patients underwent initial unilateral dismembered open pyeloplasty on the renal unit with a lower RRF on the  $^{99m}\text{Tc-MAG3}$  renal scan. The remaining 2 patients (cases 12 and 13) had bilateral UPJO with the same degree of hydronephrosis of both renal units on ultrasonography but the difference in RRF on the  $^{99m}\text{Tc-MAG3}$  renal scan between the renal units was 12% and 28%, respectively. These 2 patients underwent PCN under general anesthesia on the worse renal unit with a significantly lower RRF and simultaneous initial unilateral dismembered open pyeloplasty on the better renal unit with a higher RRF.

At 1 month after the initial pyeloplasty, the hydronephrosis of the non-operated renal unit resolved or improved spontaneously in 5 patients (cases 1-5; 38.5%). In the remaining 8 patients (cases 6-13; 61.5%), the high-grade hydronephrosis on ultrasonography and the obstructive drainage pattern on the  $^{99m}\text{Tc-MAG3}$  renal scan persisted. At 2 months after the initial pyeloplasty, these 8 patients underwent delayed contralateral dismembered open pyeloplasty. Six patients showed improvement of the hydronephrosis on ultrasonography and a good drainage pattern on the  $^{99m}\text{Tc-MAG3}$  renal scan after consecutive bilateral pyeloplasty. Two patients (cases 8 and 13) required redo-pyeloplasty because of persistent grade 5 hydronephrosis and a decrease in RRF.

## DISCUSSION

The management of neonates with unilateral UPJO has remained a controversial issue for a long time, and this issue has stayed on the minds of many pediatric urologists. King et al reported that early correction of UPJO is advocated as soon as the diagnosis is established and that young infants show more rapid improvement in RRF than do older children [9]. Tapia and Gonzalez reported that pyeloplasty in children younger than 1 year with grades 3 or 4 hydronephrosis secondary to UPJO is effective at improving renal function and they recommended early pyeloplasty for children with reduced function of the involved kidney [10]. By contrast, some authors have questioned the necessity of early pyeloplasty. Ransley et al reported that deterioration of renal function occurred in only 23% of renal units with good function as ascertained by renography initially, and they proposed that conservative observation could be possible in infants who demonstrate relatively good renal function postnatally [5]. Cartwright et al proposed that in the kidney with apparent UPJO and good function, an initial conservative approach with sequential renal scan follow-up and delayed pyeloplasty as needed is a reasonable management [6].

Recently, many institutes around the world have followed these trends in reserving pyeloplasty with con-

servative observation in the management of unilateral hydronephrosis with apparent UPJO. Compared with unilateral UPJO, however, few published management protocols exist for neonates with prenatally diagnosed bilateral hydronephrosis that leads to the postnatal diagnosis of severe bilateral UPJO. Theoretically, there are two possible approaches to these patients with severe bilateral UPJO: (1) initial conservative management followed by intervention when renal function deteriorates or progression of hydronephrosis occurs, and (2) early intervention independent of initial renal function.

There are many reports to suggest delayed surgical intervention followed by close, conservative observation focused on surveillance. Onen et al reported that only 35% of total renal units with prenatally diagnosed primary grade 3 to 4 bilateral hydronephrosis required unilateral or metachronous bilateral pyeloplasty, and the remaining 65% of renal units that were followed nonoperatively showed resolution or improvement of the hydronephrosis for a mean 54 month follow-up. Those authors thus suggested conservative management with close follow-up during the first 2 years to be a safe and recommended approach for neonates with primary bilateral ureteropelvic junction type hydronephrosis [7]. Otherwise, there are many reports emphasizing the necessity of early surgical intervention. Kim et al reported that patients who underwent early surgical correction were expected to show faster and reliable improvements in hydronephrosis, although not only the patients who underwent early surgical correction but also those treated by the conservative method could yield acceptable improvements. They concluded that early surgical intervention should be considered in patients with bilateral UPJO [11].

In many studies, patients with bilateral UPJO had hydronephrosis of SFU grade 3 or less, which is a mild form and not an emergent case requiring surgery. However, all of the cases recruited for this study had a single or two renal units with hydronephrosis of SFU grade 4 or more, which is a severe grade and thus an emergent case requiring surgery. Some authors have managed such cases by concurrent bilateral pyeloplasties. Eckstein and Drake reported the feasibility of concurrent bilateral open pyeloplasties [12], and Schwab and Casale performed successful concurrent bilateral laparoscopic pyeloplasties with favorable results [13]. But traditionally staged pyeloplasties have been recommended as a safe surgical modality. Therefore, we performed early unilateral pyeloplasty in all cases without initial conservative management. The non-operated renal units with mild hydronephrosis of SFU grade 3 (cases 1-4) showed resolution of the hydronephrosis (SFU grade 0 or 1) or improvement of the hydronephrosis (SFU grade 2 or 3) after unilateral pyeloplasty for the worse renal unit at 1 month postoperatively. However, the non-operated renal units with severe hydronephrosis of SFU grade 4 (cases 5-10) showed improvement of the hydronephrosis after unilateral pyeloplasty in only one case (case 5). The remaining 4 cases (cases 6-10) showed persistent hydronephrosis following the initial pyeloplasty and consequently delayed

pyeloplasty was performed after 1 month. After delayed pyeloplasty, one patient (case 6) showed resolution of the hydronephrosis and 3 patients (cases 7, 9, and 10) showed improvement of the hydronephrosis at the last follow-up. We observed that all of the renal units with SFU grade 3 hydronephrosis and some of the renal units with SFU grade 4 hydronephrosis improved or resolved after unilateral pyeloplasty. Therefore, we suggest that delayed pyeloplasty for the renal unit that has relatively mild hydronephrosis should be considered at 1 month following the initial unilateral pyeloplasty for the worse renal unit.

Preservation of the optimal renal function of both units is a main goal of follow-up of a patient with bilateral UPJO. Previously published papers reported that although there was no consensus regarding such a cutoff value for the recommendation of early surgical correction, an initial RRF < 30-40% can serve as an indication for early pyeloplasty [14,15]. However, these well-organized studies included mild hydronephrosis such as SFU grade 1 or 2 and enrolled cases of unilateral UPJO. In our series, we planned the initial pyeloplasty depending on the severity of hydronephrosis on renal ultrasound rather than the RRF on the  $^{99m}\text{Tc}$ -MAG3 renal scan. We performed initial pyeloplasty of the renal unit with a higher grade of hydronephrosis on renal ultrasound irrespective of the difference in RRF on the  $^{99m}\text{Tc}$ -MAG3 between the two renal units in one patient. If two renal units had the same degree of hydronephrosis on renal ultrasound, our indication for initial pyeloplasty depended on the RRF on the  $^{99m}\text{Tc}$ -MAG3 renal scan. Two patients (cases 12 and 13) had the same degree of hydronephrosis of both renal units on ultrasonography but the difference in RRF on the  $^{99m}\text{Tc}$ -MAG3 renal scan between the renal units was more than 10%. We thought PCN could protect against deterioration of renal function, and these 2 patients therefore underwent PCN on the worse renal unit with a significantly lower RRF and simultaneous initial pyeloplasty on the better renal unit with a higher RRF. However, PCN tube placement followed by delayed pyeloplasty did not preserve the renal function as we expected. Although the number of cases was small, our results suggest that preoperative PCN tube placement before delayed pyeloplasty is not a feasible management protocol.

Our study has some limitations. This was a retrospective and nonrandomized study with a limited series of 13 patients. The number of cases was very small and the follow-up period was uneven in some patients. Strictly speaking, our results could be specific to our patients only and may not be generalizable to all bilateral UPJO cases.

## CONCLUSIONS

In children younger than 1 year of age with severe bilateral UPJO, improvement or resolution of the hydronephrosis following unilateral pyeloplasty for the worse renal unit did occur in the non-operated renal units with SFU grade 3 hydronephrosis and some with SFU grade 4 hydroneph-

rosis. Therefore, delayed pyeloplasty for the opposite renal unit should be considered at 1 month following initial unilateral pyeloplasty for the worse renal unit. Preoperative nephrostomy tube placement seems to not be effective for renal function preservation. A greater number of cases and a longer follow-up in the setting of a prospective and randomized study could help to clarify the optimal management guidelines for severe bilateral UPJO.

## Conflicts of Interest

The authors have nothing to disclose.

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