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Management of congenital hydronephrosis with ureteropelvic junction obstruction: The Vienna-AKH experience 1986–2001

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Summary. One of the most common causes of congenital hydronephrosis is obstruction of the ureteropelvic junction. The obstruction can be detected with prenatal ultrasonography screening and treated before renal function is reduced; the obstruction may also resolve spontaneously. Currently, there is no test for predicting the outcome of this obstruction.

Management guidelines for neonates with asymptomatic obstruction of the ureteropelvic junction are based on expert opinions, but not on evidence-based data.

In our retrospective study, we evaluated management and outcome of 26 renal units in 23 infants (15 boys, 8 girls) with congenital obstruction of the ureteropelvic junction treated in our institution between 1986 and 2001. These infants had isolated hydronephrosis on prenatal and postnatal sonography, showed an obstructive curve pattern in the postnatal diuretic nephrogram and had at least one follow-up nephrogram during a follow-up period of at least 1.5 years. Of these renal units, 16 demonstrated normal function (Group I), five moderate function (Group II) and five severely reduced function (Group III). In group I, 6 of 12 primarily conservatively managed kidneys resolved spontaneously and remained normal in function. In group II, all infants were operated and 83% improved their kidney function. In group III, all infants were operated but none demonstrated relevant improvement.

These data support the current expert opinion of the Arbeitsgemeinschaft Pädiatrische Nephrologie (APN), that ureteropelvic junction obstruction in neonates with normal renal function can be managed primarily conservatively with close monitoring. In neonates with moderately – but not with severely – reduced renal function, early surgery is effective in the prevention of deterioration.

Key words: Hydronephrosis, congenital, ureteropelvic junction obstruction, diuretic nephrogram, infant.

Introduction

Since the introduction of ultrasound (US) screening, hydronephrosis (HN) is detected in about 0.5% of neonates. One of the most common causes of HN is ureteropelvic junction obstruction (UPJO) [1–5]. Most of these hydronephrotic kidneys with UPJO have the potential to resolve spontaneously without renal deterioration, some of them have an obstruction that needs operative correction to prevent renal damage, and a few of them are damaged and do not improve with surgery. Obstruction is therefore functionally defined as a restriction of urinary outflow that, when left untreated, will cause progressive renal deterioration [1].

In this study, we evaluated outcome data of infants with congenital UPJO in our institution, where all renal units with reduced function were operated in early infancy and those with normal function were mostly primarily managed nonoperatively.

Patients and methods

We retrospectively analysed the charts of 180 infants (129 male, 51 female) with 246 prenatally diagnosed hydronephrotic renal units who were treated in our institution between 1986 and 2001.

We identified all infants with upper urinary tract dilatation, who (1) had isolated HN in the prenatal and postnatal US, (2) showed an obstructive curve pattern in the postnatal diuretic nephrogram (DNG) in the kidney with HN, (3) had at least one follow-up DNG and (4) a follow-up period of at least 1.5 years.

US HN grading was based on the Society of Fetal Urology guidelines (grade I-IV) [6]. DNG was used to identify those kidneys with an obstructive curve pattern and to evaluate the initial differential function, given in percent. Technetium-mercaptoacetyltriglycerine (MAG3) was used as a marker and furosemide was given 0.5 mg/kg after 20 minutes when there was prolonged drainage time. Drainage time of more than 20 min and maximal elimination rate of less than 7%/min after furosemide was defined as an obstruction, drainage time between 15 and 20 min and maximal elimination rate between 7 and 14%/min was defined as an equivocal obstruction, drainage time under 20 min and maximal elimination rate greater 14%/min was defined as no obstruction.

For analysis we assigned the infants to three groups, according to initial differential function on DNG. Group I demonstrated initially normal differential function (>40% function of each renal unit), group II demonstrated moderately reduced function (15–40% function) and group III had severely compromised differential function (<15% function).

Table 1. Data of infants with congenital HN who were primarily managed conservatively

	Our data	Koff [6–8]	Dhillon [4]	Palmer [5]
Kidneys (n)	11	104	36	16
Selection criteria				
DNG	obstruction	variable	variable	obstruction
Initial function	≥ 40%	variable	> 40%	≥ 40%
US	mild-severe HN	severe HN	severe HN	severe HN
Observation (months)	mean 35 (18–144)	mean 78	60	36
Threshold to delayed surgery	variable*	function loss > 10%	function loss > 10%; symptoms	function loss > 10%; symptoms
Outcome				
Delayed surgery	31%	22%	20%	25%
Permanent function loss	0	0	3%	13%

* Not prospectively defined, retrospectively: function loss, persistence of obstruction.

Results

In 63 renal units in 53 infants (37 male, 15 female), hydronephrosis was diagnosed as caused by UPJO. In 25 of those renal units there was equivocal obstruction on diuretic renogram. These infants were not routinely followed by repeat diuretic renogram. Thirty-eight kidneys in 34 infants (23 male, 11 female) showed an obstructive curve on diuretic nephrogram. In nine of those renal units available data were incomplete, two renal units were lost to follow-up, and one with reduced function also had severe vesicoureteral reflux and recurrent urinary tract infections and was excluded from further analysis.

We could therefore identify data of 23 infants (15 male, 8 female) with 26 kidneys with obstruction and in whom a follow-up DNG was obtained. All infants had US during the first postnatal week; the mean age at initial DNG was 6 weeks. In two infants DNG was done in the first and second week because of bilateral HN, in another two infants DNG was done in the third week because of massive HN, and one infant had his first DNG at the age of 23 months, because US initially improved (from grade IV to II in the first months) and only later deteriorated. Voiding cystourethrography (VCUG) was performed in 18 (80%) of 23 infants and vesicoureteral reflux (VUR) was detected in three (13%): 1 girl with bilateral VUR grade III, 1 girl with VUR grade IV on the kidney with the UPJO and VUR grade III on the contralateral kidney, and 1 girl with VUR II-III on the contralateral kidney. These infants suffered no urinary tract infections and their renal units demonstrated good function.

Based on an expected incidence of about 0.5%, our data are probably representative for the total population (between 1990 and 2001 there were 33,026 live births at our institution). Cases available for analysis increased from about 39% of expected patients identified during the early 1990s to about 100% in more recent years (see Fig. 1).

Group I: In 16 renal units in 15 infants (11 boys, 4 girls), initial differential function was normal (42% to 56% measured by DNG and/or a normal creatinine clearance, including three solitary functioning kidneys).

Five renal units underwent early pyeloplasty in the 2nd, 3rd, 8th, 10th and 13th week, respectively, based on physicians' decisions (including the three solitary kidneys). These operated renal units kept normal function postoperatively. Eleven renal units (in 11 infants) in this group were primarily conservatively managed with repeated US and DNGs to evaluate differential development and to follow up renal function and obstruction. In six renal units, obstruction spontaneously resolved at three to 36 months (mean 8 months) without loosing function. In one infant with bilateral UPJO, one kidney underwent percutaneous nephrostomy (PCN) in the 13th week because of function loss from 50% to 40%. Pyeloplasty was performed in the 16th month. Another renal unit showed function reduction in the follow-up DNG at 11 months (from 54 to 30%) and underwent pyeloplasty one week later. Sonographically, the HN had been severe but stable. Postoperatively, renal function returned to normal in both cases. In three additional renal units, delayed pyeloplasty was performed at the ages of 14, 15, and 25 months (despite a stable normal function on DNG), based on

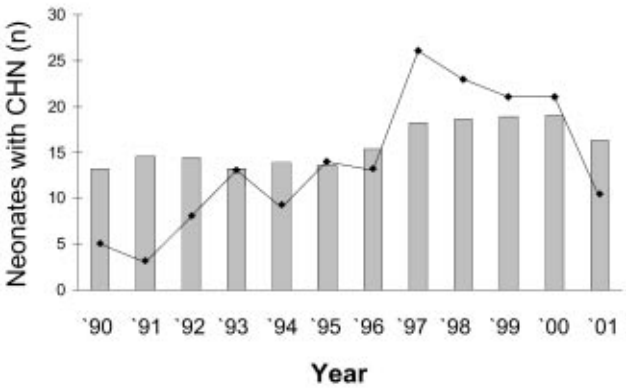

Fig. 1. Expected number of neonates with congenital HN with an incidence of 0.5% (bars) and actual number of neonates with congenital HN who were treated in our institution (line)

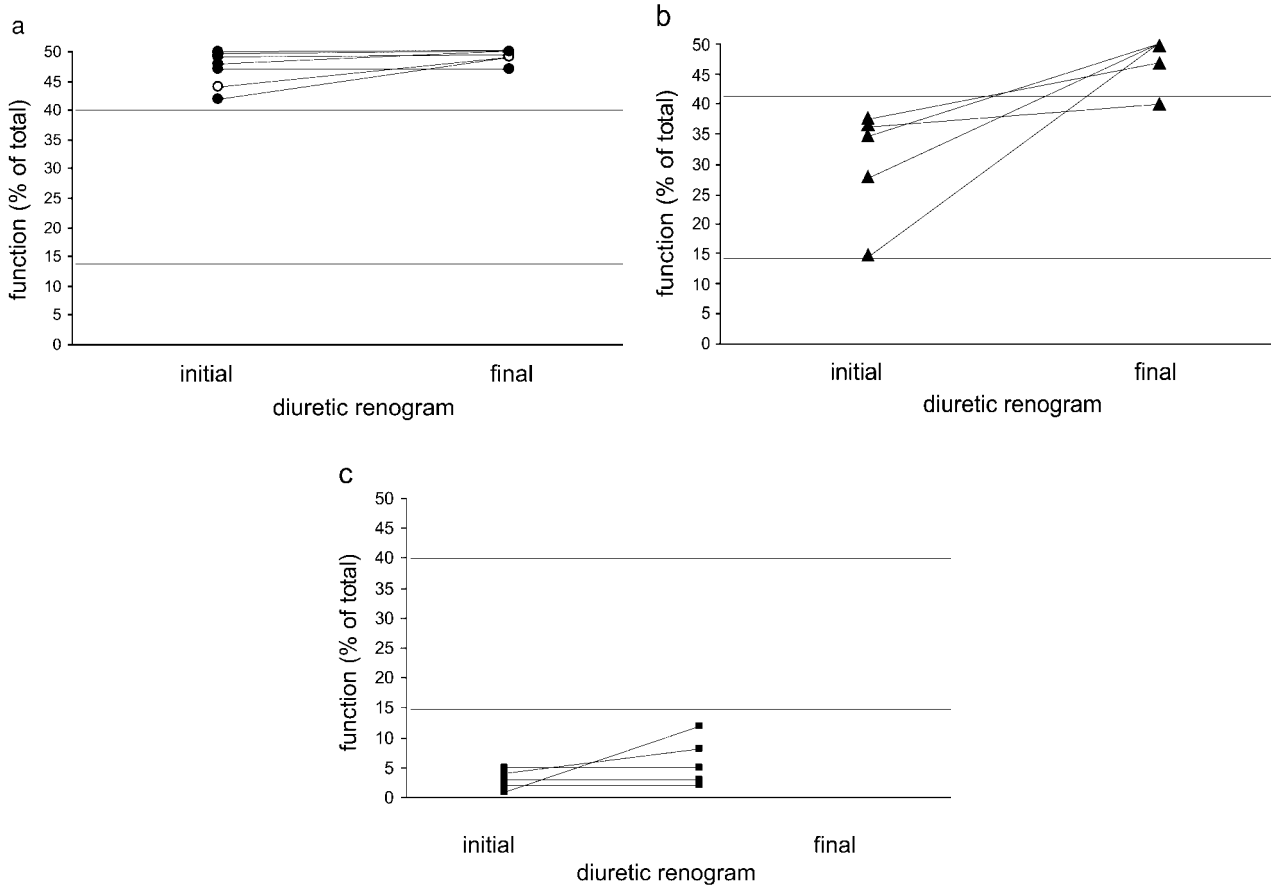

Fig. 2. Initial and final renal function measured on DNG in group I (**a**), group II (**b**) and group III (**c**). In Fig. 2a all solitary functioning kidneys are given a normal function of 50%. Full dots are the operated kidneys; empty dots were treated conservatively

physician and/or parental decision because the UPJO did not show any signs of resolving. A total of 67 DNGs were performed in these infants, corresponding to 4.5 DNG/infant (2 to 8 DNGs/infant). Follow-up time in this group was median 40 months (18–150).

Group II: In five renal units in five infants (4 boys, 1 girl) initial function was reduced to 15 to 40% (15–38% measured on DNG, mean 33%).

In this group, all infants underwent operative intervention: one kidney was drained via PCN within the third week, all the others were primarily corrected by pyeloplasty at the age of 10 to 20 weeks (mean 13 weeks). Postoperatively, four renal units improved their function to normal (preoperative 15% versus 51% postoperative, 28% versus 50%, 35% versus 50% and 38% versus 47%); one renal unit did not show relevant changes in function (37% pre- versus 40% postoperative). Follow-up time in this group was median 38 months (19–66).

Group III: In five renal units in five infants (2 boys, 3 girls) initial function was less than or equal to 10%. Two infants in this group had bilateral UPJO, the contralateral kidney functioning normally (included in group I).

In this group, all infants underwent operative intervention: surgery was performed between the 2nd and 19th weeks (mean 15 weeks). Postoperatively, differential function increased from 6% preoperatively to 12% in one kidney only. None of the other operated renal units improved their function; nephrectomy was later performed in three neonates. Follow-up time in this group was median 72 months (21–144).

Discussion

In congenital HN, early surgery may prevent deterioration when obstruction is present. However, currently there is no single investigation to discriminate between HN with relevant obstruction and those that will spontaneously improve. Therefore the dilemma of the indication and optimal timing of operative intervention in asymptomatic hydronephrotic kidneys has become a controversy. Although there is much evidence on the fate of kidneys followed by close observation or being operated upon, there has been no randomized study to prove the superiority of one concept over the other.

In 2001 the APN (Arbeitsgruppe Paediatrische Nephrologie) formed a consensus, based on an expert panel, on the management of neonates with UPJO [2]. In short, conservative management was advocated in infants with normally functioning kidneys and surgery in those with reduced function.

In this paper, we report the outcome of 15 years of similar management in a representative population of a

center. Our results showed that the majority of infants with unilateral UPJO and an initially normally functioning kidney (group I), despite the obstructed curve pattern in DNG, did not need surgical intervention because of function loss. Interestingly, in that group most infants who underwent surgical correction did so because of "physician bias", only partly explained by solitary kidneys or bilateral HN. When conservatively managed, UPJO frequently improved. Only a few infants showed function loss during follow-up, but improved to normal after operative correction. However, it was disturbing that such deterioration in function could not be predicted from symptoms or ultrasonographic changes. "Persistent obstruction" was the most frequent indication for delayed surgical repair in these infants. In that regard, we must bear in mind that each DNG exposes the infant to radiation and also requires sedation. Our infants underwent a mean of 4.5 DNGs [2–8] before surgery; in one infant surgery was performed after the eighth DNG.

Koff was one of the first proponents of conservative management in congenital HN [9–11]. He and his group described a long-term follow-up of neonates who had asymptomatic unilateral congenital HN with suspected UPJO. Surgical intervention was performed only when function loss of more than 10% or progression of HN in US became evident, regardless of absolute function in the initial DNG. Over a period of 10 years, more than two-thirds of HN cases completely resolved and 30% improved. Only about 20% of the neonates underwent delayed pyeloplasty. In no case was there permanent loss of renal function. However, only a minority (37%) of that population was comparable with our population, as we included only infants with obstructive pattern on DNG and performed early surgical intervention when renal function was less than 40%.

In our opinion, our population is better compared with that of Dhillons randomized controlled study. She included infants with prenatally diagnosed unilateral HN with >15 mm anteroposterior diameter on US, caliceal dilatation on intravenous pyelography and a differential function of >40% at three months. These infants either underwent early surgical intervention (39 infants) or were managed conservatively (36 infants). Seven (20%) of the latter group subsequently developed renal functional deterioration and underwent pyeloplasty. Not all but most (six of seven) of those renal units recovered function. In the remaining conservatively managed infants, the obstruction resolved in about 60%, while 40% remained obstructive but stable in function. Dhillon concluded that in infants with suspected unilateral UPJO and normally functioning kidneys management can be primarily conservative with close follow-up with US and isotope studies [7].

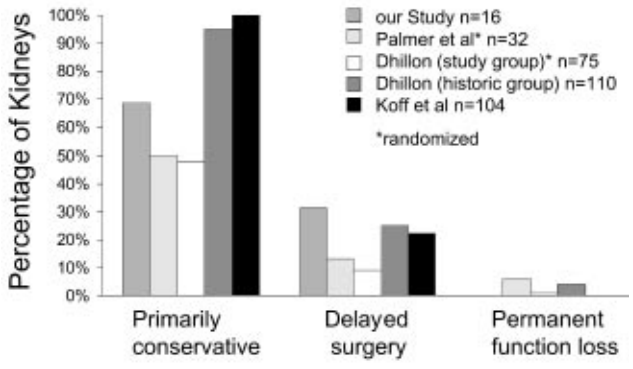

Fig. 3. Management and outcome of infants with congenital HN compared with those in the literature

Similar data were also reported by Palmer et al., whose randomized multicenter study included infants with unilateral UPJO and normal differential renal function. Infants managed conservatively were compared with those who underwent primary pyeloplasty. Crossover from the observation group to the surgical arm because of deterioration in renal function occurred in 25% (4 of 16 infants); in two of them renal function did not recover after surgical intervention [8]. Contrary to Koff and Dhillon, Palmer et al. concluded that all neonates with UPJO should have early surgical intervention, even those with initially normal renal function.

Taken together, these reports show similar results but yield different interpretations. Based on our data and on those reported in the literature, we conclude that the method of 'wait and see' is certainly not without risk in hydronephrotic kidneys with normal function, but might be safely used with close follow-up intervals for investigations.

Our management of all infants with reduced renal function (group II and III) was based on early operative interventions. Our data show that early surgery resulted in different outcomes depending on initial function. Moderately impaired renal units (group II) improved to normal function. However, as our data are uncontrolled, they do not address whether renal function might have also improved without surgery (as the Koff group demonstrated in their uncontrolled designed study [9–11]. However, we are not aware of any controlled study in infants with initially reduced renal function. In the group with poorly functioning kidneys, we could not see relevant improvement of renal function after operative intervention. Literature data for this population are inconclusive; some authors reported improved renal function after decompression [12–15]. As these reports also included older children, their data are not valid for congenital obstruction.

Similar to our findings, Mc Aleer and Capolicchio also found no improvement after pyeloplasty, regardless of patient age at time of surgery or preoperative differential renal function [16, 17]. Our data suggest that only minimal intervention, such as PCN, should be primarily performed in poorly functioning kidneys.

These data support the current expert opinion of the APN, namely that primary conservative management of UPJO with close monitoring is recommended in infants with normal renal function. In infants with moderately – but not with severely – reduced renal function, early surgery is effective in the prevention of deterioration.

Further studies incorporating risk/benefit analysis are needed to investigate the maximum acceptable duration of follow-up in persistent UPJO. As our data show, some infants required many DNGs, as obstruction persisted over more than 1.5 years. The problem of compliance

increases in cases requiring repeated invasive procedures, and this was clearly the situation for at least three of our patients who underwent surgery.

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