

Original Article

Pelvi-Ureteric Junction Obstruction at Red Cross Children's Hospital, Cape Town: a Six Year Review

Isaac E. Ocheke^{*1}, Sampson Antwi², Priya Gajjar³, Mignon I. McCulloch³ and Peter Nourse³

1. Department of Pediatrics, University of Jos, Plateau State, Nigeria.

2. Department of Child Health, Kwame Nkrumah University of Science and Technology / Komfo Anokye Teaching Hospital, Kumasi, Ghana.

3. Red Cross Children's Hospital, Cape Town, South Africa.

Abstract

Introduction: Congenital abnormalities of the kidney and urinary tract (CAKUT) are significant causes of end stage renal disease (ESRD) in children. Some of these abnormalities, when identified early, are amenable to treatment. In developing countries, very few published reports exist concerning the pattern and scope of CAKUT in childhood.

Methods: This is a retrospective review of all patients with confirmed Pelvi-Ureteric Junction (PUJ) obstruction who were diagnosed antenatally with hydronephrosis between Jan 2002 and Dec 2007 at the Red Cross Children's Hospital, Cape Town. The clinical course of every patient was reviewed for a twelve-month period after confirmation of the diagnosis.

Results: One hundred patients, 80 males and 20 females, were included in the analysis. Thirty-two children (32%) had bilateral PUJ obstruction while the rest had unilateral involvement of the left (40%) or right (28%) kidney; overall, 132 kidneys were affected. mild, moderate and severe pelvic dilatation was present in 44.7%, 36.4% and 18.9% of affected kidneys respectively. One child required nephrostomy during the first day of life while eighteen children were treated by pyeloplasty. Urinary tract infection was confirmed in eleven children. At 12 months of follow-up, 83 affected kidneys (62.9%) demonstrated spontaneous resolution of PUJ obstruction while 14 (18.5%) kidneys improved after surgical intervention. Spontaneous resolution occurred more often in kidneys with mild to moderate pelvic dilatation.

Conclusion: Spontaneous resolution of the PUJ obstruction occurred in a substantial proportion of children by twelve months of follow-up and complete resolution is more likely in mild to moderate dilatation.

* Corresponding Author; Department of Pediatrics, University of Jos, Plateau State, Nigeria. E. mail: ieocheke@yahoo.com

Keywords: Antenatal Hydronephrosis; Childhood; Pelvi-ureteric Junction Obstruction.

The authors declared no conflict of interest

Introduction

Pelvi-ureteric junction (PUJ) obstruction is the most common cause of antenatal fetal hydronephrosis [1]. It generally occurs as a sporadic anomaly, though familial inheritance has been reported with a pattern suggestive of autosomal dominance with incomplete penetrance [2]. Obstruction at the pelvi-ureteric junction usually develops as a result of intrinsic neuro-muscular abnormalities but could also be caused by external compression by, for example, an aberrant vessel. The incidence increases in the presence of other urinary tract anomalies [3, 4].

The natural history of PUJ obstruction varies considerably, but the severity of renal pelvis dilatation (RPD) usually correlates with the prognosis [5, 6]. In some cases, obstruction resolves spontaneously even in the presence of severe pelvic dilatation while others with mild or moderate RPD progress to marked hydronephrosis and progressive deterioration of renal function [7].

The presence of antenatal hydronephrosis caused by PUJ obstruction poses an important clinical challenge. Such findings often induce both parental and physician's anxiety concerning the possible outcome and the optimal approach to management. There is no consensus on management of PUJ obstruction; opinions are divided between conservative treatment with close monitoring and early intervention before significant loss of kidney function [8].

There are few reports on PUJ obstruction from developing countries [9], mainly due to lack of routine antenatal fetal screening for the detection of congenital abnormalities. However, with increasing access to improved antenatal

care, detection of congenital abnormalities such as PUJ obstruction is becoming more common, leading to questions on how to manage these infants. In this audit, we reviewed all children with antenatal hydronephrosis who had postnatal confirmation of PUJ obstruction and who were managed in our centre over a six-year period.

Methods

The study was carried out at Red Cross Children's hospital and approved by the hospitals and the University of Cape Town's ethical committees. All children with antenatal hydronephrosis and confirmed post natal diagnosis of PUJ obstruction during a six-year period from January 2002 and December 2007 were identified and their records retrieved. Each patient's clinical course was reviewed for the twelve months following confirmation of diagnosis. Using the Society of Fetal Urology (SFU) criteria, involved kidneys were classified as having mild, moderate or severe RPD by having an antero-posterior (AP) pelvis diameter of 5-9.9 mm, 10-15 mm and greater than 15 mm respectively [10]. The postnatal ultrasound examination was done after the first forty-eight hours of birth. Male neonates with bilateral hydronephrosis however had their postnatal ultrasound evaluation on the first day of life followed by a micturating urethrocytogram (MCUG) within few days to rule out other causes of congenital hydronephrosis such as posterior urethral valves (PUVs). Children with other urinary tract abnormalities such as PUVs, multicystic dysplastic kidney (MCDK), duplex systems and vesico-ureteric reflux (VUR) were excluded from the analysis. Renal scan with technetium-99-mercapto acetyl triglycine (MAG3) was done at six weeks postnatally in children with moderate to severe RPD. In few children, however, this was delayed because of logistic problems. Management included three-monthly ultrasound re-evaluation of the renal pelvis AP diameter and differential kidney function estimation by MAG3 renogram in children who showed evidence of progressive renal pelvis AP diameter dilatation. Surgical intervention (pyeloplasty) was performed in children who had progressive increase in the AP diameter, decrease in differential renal function below 30% and recurrent urinary tract infections (UTIs). None of the children received prophylactic antibiotic therapy for urinary tract infections. Resolution of the PUJ obstruction was judged to have occurred if the APD of the renal pelvis was 5 mm or less [11] in two consecutive ultrasound scans, either spontaneously or following surgery. Nephrectomy was indicated where an affected kidney contributed less than 20% of total function on renogram.

Table-1: Outcome of the studied 132 kidneys affected by pelvi-ureteric junction obstruction after 12 months of follow up

Grade of pelvic dilatation	Number of involved kidneys	PUJ obstruction resolution rate	
		Number	Percentage
Mild	59	53	89.8%
Moderate	48	41	85.4%
Severe	25	3	12.0%
Total	132	97	73.5%

Results

One hundred patients, 80 males and 20 females, were included in the analysis. Eighty four percent of the study patients had their initial postnatal ultrasound within the first week of life (range: 1 day to 26 weeks). Thirty-two infants had bilateral PUJ obstruction while 68 infants had unilateral involvement of the left (40%) or right kidney (28%); overall, 132 kidneys were affected. Mild, moderate and severe pelvic dilatation was present in 44.7%, 36.4% and 18.9% of affected kidneys, respectively.

Nineteen children overall were treated surgically. One patient had percutaneous nephrostomy on the first day of life because of massive hydronephrosis causing respiratory distress. The remaining patients underwent pyeloplasty, including 10 patients who had surgery in the first six months of life. The mean age at surgery was 10.8±8.7 months (range 1 day to 48 weeks). Among patients subjected to surgical intervention, 11 children experienced reduction of their pelvis dilatation to 5 mm or less following the initial pyeloplasty. Two patients required a repeat pyeloplasty because of progressive increase of the AP diameter above pre-operative value and evidence of further decline in their relative function on repeat MAG3 renogram. Two other children underwent unilateral nephrectomy. The child with per cutaneous nephrostomy was yet to have definitive closure at the end of the study period.

Table-1 shows the rate of PUJ obstruction resolution during the study period according to the severity of RPD. Overall, 35 of all affected kidneys (26.5%) demonstrated spontaneous resolution of PUJ obstruction at six months of diagnosis while five kidneys improved after surgical intervention. At 12 months of follow-up, 83 of all affected kidneys (62.9%) demonstrated spontaneous resolution while 14 kidneys (18.5%) improved after surgical intervention.

Urinary tract infection was documented in eleven (11%) children. *Klebsiella pneumoniae* was the most common

organism. Two patients had repeated episodes of UTI; one had three and the other had two episodes with the same organism (*E.coli*). Five of the eleven children with UTI were among the nineteen children who had surgery.

Discussion

Overall, spontaneous resolution of pelvi-ureteric junction obstruction occurred in 63% of the affected kidneys at twelve months of follow up, a finding comparable to figures previously reported in other studies [12-15]. It is important however, to note here that in some of these studies isolated RPD, a condition that could present with antenatal foetal hydronephrosis without underlying PUJ obstruction, were also included.

In our study, about two thirds of the kidneys with mild to moderate dilatation spontaneously resolved by the end of the study, while only 12% resolved in the severe group. This figure contrasts with the 33% rate of resolution of severe dilatation reported by Mani et al [15] and Ismaili et al [16] in their respective studies. The variation may be related to a longer follow up period in their study. It is possible that, the number of kidneys in the severe category with spontaneous resolution might have increased if these children were followed up longer than twelve months. Moreover, included in their study were some children with other underlying urological abnormality such as vesico-ureteric reflux without PUJ obstruction. This may have contributed to the higher values compared with our study.

One child in our study who initially had mild renal pelvis dilatation evolved later to severe dilatation in the course of follow up. Even though only one case of such condition was identified it indicates that an initially mild disease may progress to a more severe form of the condition.

The prevalence of urinary tract infections in our study was 11%. It is known that children with dilated renal pelvis are susceptible to UTI [17, 18]. The reason for this is unclear but it may be related to urine stasis. Our figure is comparable to the 14% UTI prevalence reported by Coelho et al [19] but differs from the higher values (over 25%) reported in other previous studies [20-22]. In these studies, prophylactic antibiotics were given to patients in contrast to our study. The finding of a lower UTI prevalence in our patients appears to give credence to the belief that prophylactic antibiotic therapy may have no advantage in children with dilated urinary systems and may in the long term promote antibiotic resistance.

As many as four-times more males than females were identified with PUJ obstructions in our study, with the

left kidney being more frequently involved than the right. This finding is similar to previous reports [15, 23]. There is no clear explanation for the predilection of the male sex and the more frequent affection of the left kidney but it may suggest a role for interplay of genetics.

Our study is not without limitations, being a retrospective record review. Some patient records were excluded because of incompleteness of data and some records could not be traced at all. Secondly, this review is coming from a tertiary health facility in which case most of the pregnancies screened for this abnormality were usually high risk, which could have introduced a selection bias. However, we believe that our findings are important as they provide insight into the pattern and likely outcomes of this condition.

Conclusion

Spontaneous resolution of renal pelvis dilatation due to PUJ obstruction occurs frequently in affected children. Most of these children experience complete spontaneous resolution within the first year of life and complete resolution is more likely with mild to moderate dilatation. Urinary tract infection is more likely observed in children with severe dilatation, therefore such children should be investigated for UTI when they present with symptoms.

Acknowledgement

The authors are grateful to staffs of the record department at Red Cross Children's Hospital. We are equally thankful to J Ingelfinger, Professor of Pediatrics, Harvard Medical School, Pediatric Nephrology, Massachusetts General Hospital for reviewing the manuscript.

References

1. Capello SA, Kogan BA, Giorgi LJ Jr, Kaufman RP Jr. Prenatal ultrasound has led to earlier detection and repair of ureteropelvic junction obstruction. *J Urol*. 2005 Oct; 174(4 Pt 1):1425-8.
2. Gunn TR, Mora JD, Pease P. Antenatal diagnosis of urinary tract abnormalities by ultrasonography after 28 weeks gestation: incidence and outcome. *Am J Obstet Gynecol*. 1995 Feb; 172(2 Pt 1):479-86.
3. Zhang PL, Peters CA, Rosen S. Ureteropelvic junction obstruction: morphological and clinical studies. *Pediatr Nephrol*. 2000 Aug; 14(8-9):820-6.
4. Koff SA, Mutabagani KH. Anomalies of the kidney. In: *Adult and Pediatric Urology*, 4th ed, Gillenwater JY, Grayhack JT, Howards SS, Mitchell ME (Eds), Lippincott Williams and Wilkins, Philadelphia 2002. p.2129.
5. Coplen DE, Austin PF, Yan Y, Blanco VM, Dicke JM. The magnitude of fetal renal pelvic dilatation can identify obstructive postnatal hydronephrosis, and direct

postnatal evaluation and management. *J Urol.* 2006 Aug ;176(2):724-7; discussion 727.

6. Bouzada MC, Oliveira EA, Pereira AK, Leite HV, Rodrigues AM, Fagundes LA, Gonçalves RP, Parreiras RL: Diagnostic accuracy of fetal renal pelvis anteroposterior diameter as a predictor of uropathy: A prospective study. *Ultrasound Obstet Gynecol.* 2004 Dec ;24(7):745-9.

7. Marks SD. How have the past 5 years of research changed clinical practice in paediatric nephrology? *Arch Dis Child.* 2007 Apr; 92(4):357-61.

8. DiSandro MJ, Kogan BA. Neonatal management. Role for early intervention. *Urol Clin North Am.* 1998 May; 25(2):187-97.

9. Harish J, Joshi K, Rao KL, Narasimhan KL, Samujh R, Choudhary SK, Mahajan JK. Pelviureteric junction obstruction: how much is the extent of the upper ureter with defective innervation needing resection? *J Pediatr Surg.* 2003 Aug; 38(8):1194-8.

10. Fernbach SK, Maizels M, Conway JJ. Ultrasound grading of hydronephrosis: introduction to the system used by the Society for Fetal Urology. *Pediatr Radiol.* 1993; 23(6):478-80.

11. Ismaili K, Avni FE, Wissing KM, Hall M; Brussels Free University Perinatal Nephrology Study Group. Long-term clinical outcome of infants with mild and moderate fetal pyelectasis: validation of neonatal ultrasound as a screening tool to detect significant nephrouropathies. *J Pediatr.* 2004 Jun ;144(6):759-65.

12. Cheng AM, Phan V, Geary DF, Rosenblum ND. Outcome of isolated antenatal hydronephrosis. *Arch Pediatr Adolesc Med.* 2004 Jan ;158(1):38-40.

13. Lee RS, Cendron M, Kinnamon DD, Nguyen HT. Antenatal hydronephrosis as a predictor of postnatal outcome: a meta-analysis. *Pediatrics.* 2006 Aug; 118(2):586-93.

14. Mami C, Paolata A, Palmara A, Marrone T, Berté Luca F, Marseglia L, Arena F, Manganaro R. Outcome and management of isolated moderate renal pelvis dilatation

detected at postnatal screening. *Pediatr Nephrol* 2009 Oct; 24(10):2005-8.

15. Mami C, Palmara A, Paolata A, Marrone T, Marseglia L, Berté Luca F, Arena F. Outcome and management of isolated severe renal pelvis dilatation detected at postnatal screening. *Pediatr Nephrol.* 2010 Oct; 25(10):2093-7.

16. Ismaili K, Hall M, Piepsz A, Alexander M, Schulman C, Avni FE. Insights into the pathogenesis and natural history of fetuses with renal pelvis dilatation. *Eur Urol.* 2005 Aug; 48(2):207-14.

17. Lacombe J. Urinary tract infection in children. *BMJ.* 1999 Oct 30; 319(7218):1173-5.

18. Practice parameter: the diagnosis, treatment and initial evaluation of the initial urinary tract infection in febrile infants and young children. American Academy of Pediatrics. Committee on Quality Improvement. Subcommittee on Urinary Tract Infection. *Pediatrics.* 1999 Apr; 103(4 Pt 1):843-52.

19. Coelho GM, Bouzada MC, Pereira AK, Figueiredo BF, Leite MR, Oliveira DS, Oliveira EA. Outcome of isolated antenatal hydronephrosis: a prospective cohort study. *Pediatr Nephrol.* 2007 Oct; 22(10):1727-34.

20. Lee JH, Choi HS, Kim JK, Won HS, Kim KS, Moon DH, Cho KS, Park YS. Nonrefluxing neonatal hydronephrosis and the risk of urinary tract infection. *J Urol.* 2008 Apr; 179(4):1524-8.

21. Penido Silva JM, Oliveira EA, Diniz JS, Bouzada MC, Vergara RM, Souza BC. Clinical course of prenatally detected primary vesicoureteral reflux. *Pediatr Nephrol.* 2006 Jan; 21(1):86-91.

22. de Kort EH, Bambang Oetomo S, Zegers SH. The long-term outcome of antenatal hydronephrosis up to 15 millimetres justifies a noninvasive postnatal follow-up. *Acta Paediatr.* 2008 Jun;97(6):708-13.

23. Sheu JC, Koh CC, Chang PY, Wang NL, Tsai JD, Tsai TC. Ureteropelvic junction obstruction in children: 10 year,s experience in one institution. *Pediatr Surg Int.* 2006 Jun; 22(6):519-23.