

CAS CLINIQUE / CASE REPORT

RAPID and SUSTAINED RECOVERY of RENAL FUNCTION with TRANSIENT PLACEMENT of an INTRAURETRAL NEPHROSTOMY CATHETER in an INFANT with URETEROPELVIC JUNCTION OBSTRUCTION and ACUTE RENAL FAILURE

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Aoun B, Ulinski T, Termos S, Kalkas G, Fakhoury H, Schmitt CP. Rétablissement rapide et durable de la fonction rénale par néphrostomie transitoire chez un enfant porteur d'un syndrome de la jonction pyélo-urétérale et souffrant d'une insuffisance rénale aiguë. *J Med Liban* 2014 ; 62 (1) : 54-56.

ABSTRACT : Ureteropelvic junction obstruction (UPJO) is a common, congenital urinary malformation in the pediatric age group. In most cases the diagnosis is made antenatally and resolves spontaneously. Postnatal diagnosis is made when symptoms of urinary tract infection or abdominal pain occur. We report a six-month-old girl with single kidney and known vesico-ureteral reflux grade IV presenting with severe acute renal failure (ARF), requiring acute peritoneal dialysis (PD). After diagnosis of decompensated UPJO, a nephrostomy was performed, and renal function restored within seven days. UPJO was subsequently treated by open pyeloplasty.

To our knowledge, this is the first case of UPJO requiring PD due to severe renal failure in a child. Children with UPJO and major morbidity of the contralateral kidney are at risk of renal failure and should therefore be followed carefully to prevent serious complications.

Keywords : Ureteropelvic junction obstruction, acute renal failure, peritoneal dialysis, children

INTRODUCTION

Ureteropelvic junction obstruction (UPJO) is a congenital narrowing or occlusion where the ureter meets the kidney. In most cases the diagnosis is made antenatally and resolves spontaneously [1-2]. Postnatal diagnosis is made when symptoms of abdominal pain, urinary tract infection (UTI), or hypertension occur [3-5] or in asymptomatic children during ultrasound investigation for other reasons. UPJO can be associated with vesico-ureteral reflux (VUR) and in 10% of children requires surgical intervention [6]. We report a child with a single kidney and UPJO who presented with severe acute renal failure (ARF) requiring transient peritoneal dialysis (PD).

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RÉSUMÉ : Le syndrome de la jonction pyélo-urétérale (JPU) est une malformation de la voie urinaire bien connue à l'âge pédiatrique. Le diagnostic est réalisé souvent en période anténatale avec une résolution spontanée dans la majorité des cas. Le diagnostic en période postnatale est souvent évoqué lors de la présence d'une infection urinaire ou d'une douleur abdominale.

Nous rapportons le cas d'un nourrisson de 6 mois de sexe féminin suivi pour rein unique avec un reflux vésico-urétéral grade IV qui a présenté une insuffisance rénale aiguë nécessitant la mise en route d'une dialyse péritonéale. À l'échographie, une majoration de l'obstruction de la JPU a été diagnostiquée et une néphrostomie percutanée a été réalisée. La fonction rénale s'est normalisée au bout de sept jours et la patiente a pu bénéficier d'une cure chirurgicale par la suite.

À notre connaissance, il s'agit du premier cas de syndrome de la JPU décrit à l'âge pédiatrique diagnostiqué à l'occasion d'une décompensation avec insuffisance rénale sévère. Le message principal de ce cas est d'insister sur la possibilité d'une coexistence de reflux vésico-urétéral massif et d'un syndrome de la JPU pouvant entraîner une insuffisance rénale aiguë s'il concerne un rein unique. Un syndrome de la JPU doit être éliminé devant toute dilatation pyélique, même en présence d'un reflux massif.

Mots-clés : syndrome de la jonction pyélo-urétérale, insuffisance rénale aiguë, dialyse péritonéale, enfants

CASE REPORT

In the female infant left side renal agenesis was diagnosed antenatally, but no regular follow-up was done postnatally until the age of two months when the infant developed febrile UTI. A voiding cystography revealed grade IV reflux. The infant was started on antimicrobial prophylaxis until the age of six months when the parents noticed a decrease in urine output, without other symptoms. At admission the patient was pale, lethargic, anuric with stable vital signs. She was transferred to the pediatric intensive care unit with an elevated serum creatinine of 5 mg/dl. Repeated biochemical investigations showed a further increase in serum creatinine to 6 mg/dl, blood

urea nitrogen 80 mg/dl, potassium 7 mmol/l, and serum phosphorus 6 mg/dl. Due to respiratory failure, the infant was intubated and ventilated. Chest X-ray showed an increase of the cardiothoracic index with bilateral pleural effusion. Based on the patient clinical status and biological investigation, peritoneal dialysis was started immediately. Abdominal ultrasound reconfirmed renal agenesis on the left side, and showed compensatory hypertrophy of the right kidney with normal echogenicity, absence of stones but dilatation of the pelvis and calyces in the anuric child (Figure 1). Vascular thrombosis was ruled out by Doppler sonography. A voiding cystography demonstrated reflux and substantial narrowing of the UPJ (Figure 2). After five days, a nephrostomy tube was inserted by an interventional radiologist. The radiologist, while inserting the tube, passed the UPJ despite some distinct resistance and left the catheter in situ. Two days after insertion of the nephrostomy, urine output via the urethra started in addition to urine output via the nephrostomy tube with recovering of renal function. Five days after insertion of the nephrostomy the girl was extubated, and the nephrostomy tube removed. Transient polyuria was compensated by IV infusions for seven days. Ten days later, the infant was discharged in unremarkable clinical condition.

On follow-up, one month after discharge, the plasma creatinine level was normalized (0.6 mg/dl), subsequent tc-99m-mag3 scintigraphy, however, still demonstrated urodynamically relevant UPJO, which was removed by open surgery.

DISCUSSION

UPJO is a common, mostly congenital urinary tract malformation in infants due to hypertrophy in the muscle that surrounds the UPJ that might lead to obstruction and ultimately ARF. Other potential causes include local inflammation, kidney stones, and vascular malformations [7-8].

In our case, the infant presented with severe, life-threatening acute renal failure requiring PD. This complication has been described in pregnant women with a single kidney but rarely in children. Intraoperatively, no



FIGURE 1. Ultrasound of the kidney demonstrating dilation of the pelvis and calyces in the anuric infant but no ureter dilatation, suggesting UPJO.

kinking of the ureter, 'oestling bands' or crossing vessels were found. One might suspect that the febrile UTI at the age of 2 months caused ureteric inflammation and stricture at the UPJ site. The excellent recovery after intrarenal pressure release via the nephrostomy suggests a rather short history of complete obstruction without irreversible damaging.

The striking feature of this case is that passing the UPJ with the nephrostomy catheter despite local resistance and leaving it in place for several days may have resulted in a relevant benefit by dilating the obstruction. Of note the catheter had apertures intraureterally and in the pelvis. This could have promoted the excellent recovery of the renal function, even after removal of the catheter and thus allowed postponing the surgical correction until the patient was in good clinical condition. Still the effect was too small to remedy the UPJO.

One may argue that immediate surgery should have been performed in face of the renal function loss and the obstruction. This option, however, was disapproved in face of the rapidly declining clinical condition of the infant. After placement of the nephrostomy tube the clinical condition rapidly improved and renal function normalized. Thus, postponing the surgical correction of the UPJO was justified. Still, a close follow-up of the infant is mandatory. The VUR reflux could not adequately be classified in association with the UPJO. If further episodes of pyelonephritis develop with antibiotic prophylaxis and the reflux into the single kidney does not spontaneously decrease, subureteral injection of a bulking agent such as dextranomer/hyaluronic acid copolymer or surgical reflux repair need to be considered.



FIGURE 2. Due to the ultrasound findings a retrograde cystography was performed. It demonstrates VU reflux but also suggests narrowing of the UPJ.

In conclusion, UPJO should be carefully monitored in children with major morbidity of the contralateral kidney, since acute renal failure and associated sequelae may develop. If renal failure has developed an interventional approach with insertion of a nephrostomy catheter may be justified to improve renal function and the clinical condition of the patient prior to surgical UPJO correction. Our case suggests that a particular good outcome can be achieved by dilating the UPJO with the nephrostomy catheter placed intraureterally. This allowed postponing the surgical repair for several weeks. Whether a more aggressive dilation approach with balloons is justified in children with UPJO requiring a nephrostomy is unknown at present, the all over benefits and risks are yet unknown.

REFERENCES

1. Belloli G, Musi L, Campobasso P, Biscuola G, Cappellari F, Benigno V. [Hydronephrosis in infancy and childhood]. *Pediatr Med Chir* 1982; 4: 237-46.
2. 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; 174: 1425-8.
3. Roth CC, Hubanks JM, Bright BC et al. Occurrence of urinary tract infection in children with significant upper urinary tract obstruction. *Urology* 2009; 73: 74-8.
4. Braren V, West JC Jr, Boerth RC, Harmon CM. Management of children with hypertension from reflux or obstructive nephropathy. *Urology* 1988; 32: 228-34.
5. Waard D, Dik P, Lilien MR, Kok ET, de Jong TP. Hypertension is an indication for surgery in children with ureteropelvic junction obstruction. *J Urol* 2008; 179: 1976-8; discussion 1978-9.
6. Estornell Moragues F, Martinez Verduch M, Dominguez Hinarejos C, Marco Macian A, Muro Velilla MD, Garcia-Ibarra F. [Pyeloureteral junction syndrome. Associated vesicoureteral reflux]. *Arch Esp Urol* 1992; 45: 455-8.
7. Park BS, Jeong TK, Ma SK et al. Hydronephrosis by an aberrant renal artery: a case report. *Korean J Intern Med* 2003; 18: 57-60.
8. Bandy LC, Anderson EE, Gall SA. Acute renal failure in pregnancy associated with solitary kidney, congenital ureteropelvic junction obstruction, and renal cortical cyst. *South Med J* 1984 Aug; 77 (8): 1056-7.
9. Perlberg S, Pfau A. Management of ureteropelvic junction obstruction associated with lower polar vessels. *Urology* 1984; 23: 13-18.