Paediatrica Indonesiana 18 : 279 — 286. Sept. — Oct. 1978.

CASE REPORT

Duplication of the Urinary Tract Occurring in One Family

by

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Abstract

Duplication of the urinary tract occurring in one family is reported. The penetration of this anomaly among them was not equal. The father had a bifid ureter on the right side. Among his children, one had right bifid pelvis and the other two bilateral double kidney. The smallest child had a non-functioning right upper kidney associated with an ureterocele causing trouble in micturition and recurrent infection. After surgical correction the symptoms disappeared.

Received 26th. Jan. 1978.

Introduction

Duplication of the upper urinary tract is one of the common anomalies occurring in about 1 out of 160 individuals (Campbell and Harrison, 1970). However, its occurrence in members of the same family is still rarely reported. By 1966 only 7 such families had been reported in the literature (Girsh and Kaprinski, 1956). Investigation in affected families suggested that inheritance of the anomaly was by an autosomal dominant gene (Whitaker and Danks, 1966 : Atwell et al., 1974). Since the anomaly is prone to infection than a normal kidney and frequently associated with obstructive defects (Campbell, 1951), it could lead to hydronephrosis, calculi, and destruction of the renal tissue.

In children the risk of infection is increased by twenty fold than that in adults which might be due to the higher frequency of vesico-ureteric reflux or obstruction in childhood (Campbell and Harrison, 1970). Therefore, routine screening by intravenous pyelography of sibs on index patients is recommended in order to detect a duplex urinary tract in other members of the family before the onset of complications occurs (Atwell et al., 1974). This paper reports a family in which the father and 3 siblings had double collecting systems of the urinary tract, assumed to be the first ever reported in the Indonesian literature.

Case report

Case 1

S., a three-month-old girl, was admitted to the Department of Child Health, Dr. Cipto Mangunkusumo General Hospital, on the 13th of October 1976 for the assessment of recurrent urinary tract infections. On anamnesis the baby often cried during micturition since the age of 5 days. She had also suffered from recurrent low grade fever.

Physical examination revealed the baby girl weighing 5,800 gm., with a body temperature of 36.4° C. The heart and lungs showed no abnormalities. The abdomen was supple. The liver was palpable 2 cm. below the right costal margin. The spleen and kidney were not palpable. No edema was detected.

Laboratory examination of the urine showed albumin (+), sediment: leucocyte 60 - 80/HPF, erythrocyte 4 - 5/HPF, granular cast (+). Urine culture revealed B. pyocyaneus 200,000/ml. Blood examination showed Hb. 10.7 gm.%, leucocyte 7,000/mm^3, diff. count: eos. 3%, segm. 66%, lymphocyte 29%, monocyte 2%; thrombocyte 382,000/mm^3. The blood urea was 29 mg.%, creatinine 1.08 mg.%. Intravenous pyelography showed left hydronephrosis, left double ureter, downward displacement of the right kidney and an ureterocele that was also detected (Figure 1) in the mictio-cysto-urethrogram.

The child was operated some days afterward performing excision of the ure-

terocele and resection of the upper hydronephrotic portion of the right kidney. On follow up examination the symptoms disappeared.

Case 2

F., the first sister, 8½ years old, sometimes suffered from flank pain but the urine examination showed no abnormality. Her intravenous pyelography revealed a right bifid pelvis with normal calices in both kidney (Figure 2).

Case 3

D. the fourth sister, 3½years old, often suffered from supra pubic pain, the urine showed no abnormality on repeated examination. Her intravenous pyelography revealed bilateral double kidney, left hydronephrosis. The bladder was normal, no ureterocele was detected (Figure 3).

Case 4

S., the father, 42 years old, suffered from repeated flank pain since the last 4 years. He often had abdominal colic which radiated downward to the right lower abdomen. The urine sometimes was red in color. In 1961 he was diagnosed of having nephrolithiasis. Stone analysis showed Ca oxalates. His intravenous pyelography revealed double collecting system (bifid ureter) on the right side (Figure 4).

The pedigree of the family can be seen in Fig. 5 and the urinary tract malformations are summarized in Table 1.

TABLE 1 : *Occurrence of urinary tract malformation in the family*

Case	Member of family	Age (years)	Description of malformations
1	Daughter	3/12	left hydronephrosis, double ureter both side, ureterocele, non-functioning of right upper kidney
2	Daughter	8 6/12	right bifid pelvis
3	Daughter	3 6/12	bilateral double kidney, left hydronephrosis
4	Father	42	double collecting system (bifid-ureter) on the right kidney

Discussion

Duplication of the ureter and renal pelvis occurred due to an accessory ureteric bud arising from the developing ureter to cause an incomplete duplication, or from the Wolffian duct when there was complete duplication of the collecting system (Williams, 1972). Although this anomaly occurred more often than any other urinary tract malforma-

tion, its familial occurrence has not been stressed yet. Ritter (1935), Steinlin (1936), Kares (1938), Frissel (1942), Girsh and Kaprinski (1956) reported double ureter in members of the same family. Atwell et al. (1974), investigating 30 index patients, found a duplex system in 11 (excluding a bifid renal pelvis) or 32 out of 101 relatives (including a bifid renal pelvis).

In this report upper urinary tract duplication was detected in 4 out of 7 members of the family (Figure 5). The 3rd child died at the age of 3 months and pyelography was not done in the other 2 members of the family because it was refused by the parents. Thus, the possibility that this anomaly affected all members of the family still existed. This finding is consistent with the suggested hypothesis that the inheritance is caused by a dominant gene (Whitaker and Danks, 1966; Atwell et al., 1974). Although not all members of the family were affected as mentioned in the literature (which could also happen in this family), Whitaker and Danks (1966) stated that it is still a dominant inheritance but of a low penetrance.

The anomalies found in the whole family were not equal as shown by the various types of anomaly in our cases (Table 1). The father had a bifid ureter on the right side, and among the daughters one had only right bifid pelvis and the other two bilateral double kidneys. In Case 1 the right upper kidney did not function and it was associated with an ureterocele causing trouble in micturition and recurrent infection, which necessitated surgery to relieve the symptoms. Other symptoms detected in the family were hematuria and lithiasis in the father and repeated flank pain in the other 2 siblings.

In Case 3 the left kidney had already shown early hydronephrotic changes (Fig. 3), although repeated culture revealed negative result. Regular follow up and repeated pyelography would be warranted in this case to make prompt decision of surgical intervention, if there was progression of the hydronephrosis (Belman et al., 1974). This family and the previous reports (Ritter, 1935; Girsh and Kaprinski, 1956; Atwell et al., 1974) had pointed out the occurrence of familial duplication of the upper urinary tract. We support the suggestion of Atwell et al. (1974) that routine screening by intravenous pyelography of the other members of the index patient is recommended to detect duplex urinary tract and their complications which could be potentially correctable to prevent renal disease.

Acknowledgements

The authors are very grateful to Drs. L.A. Tamaela and Widhodho T. Karjomanggolo for their help in performing the radiological examinations which made this study possible.

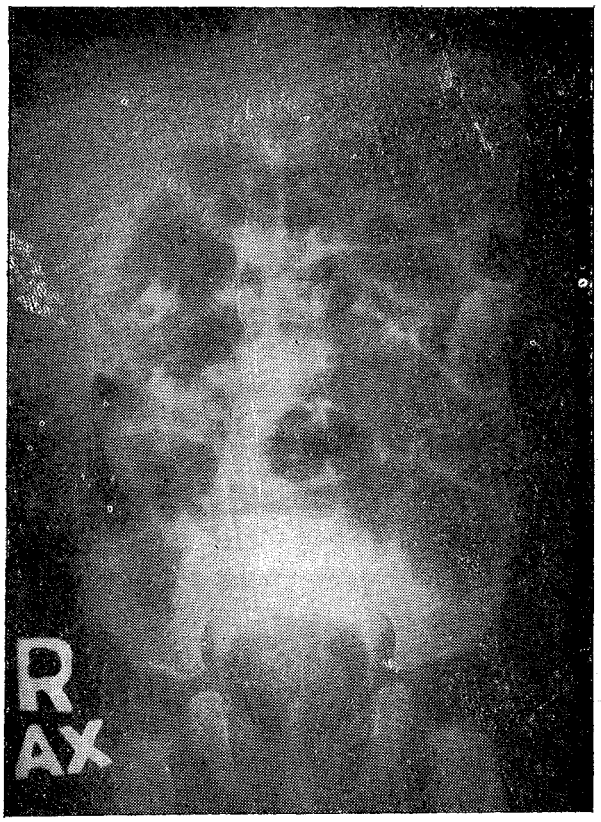

FIG 1 : *Case 1. IVP showed left hydronephrosis, left double ureter, downward displacement of the right kidney and ureterocele*

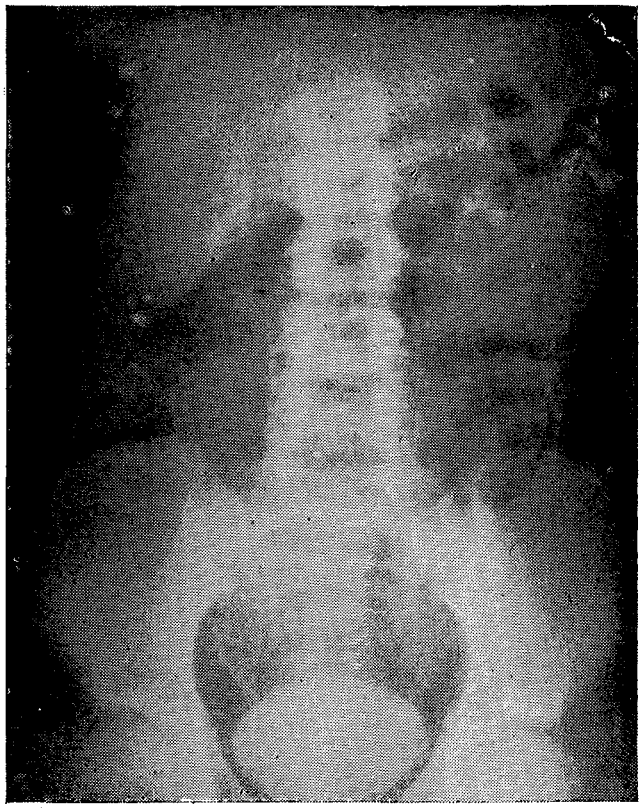

FIG. 2 : *Case 2. IVP revealed a right bifid pelvis with normal calices*

FIG. 3 :
Case 3. IVP revealed bilateral double kidney and left hydronephrosis

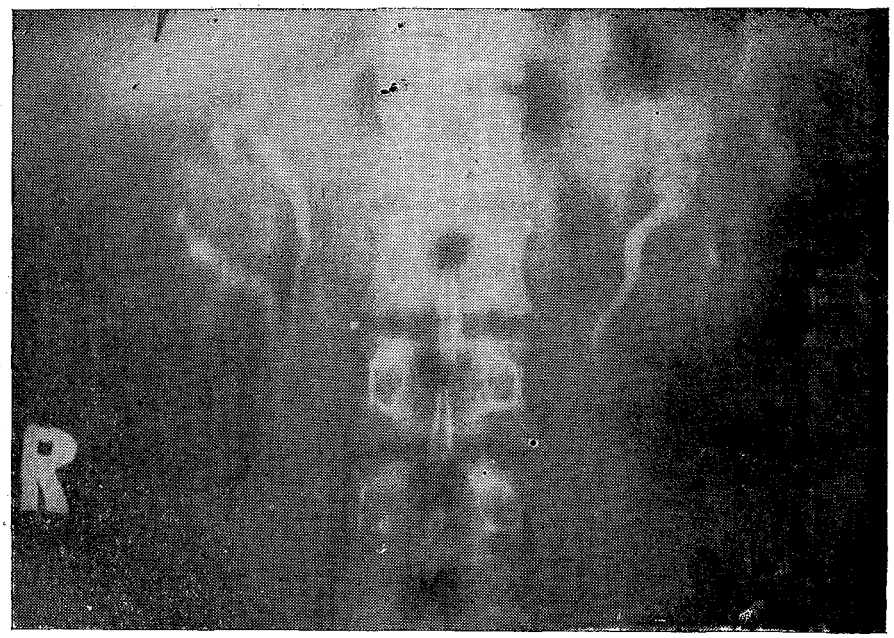

FIG. 4 : *Case 4. IVP showed double collecting system on the right side*

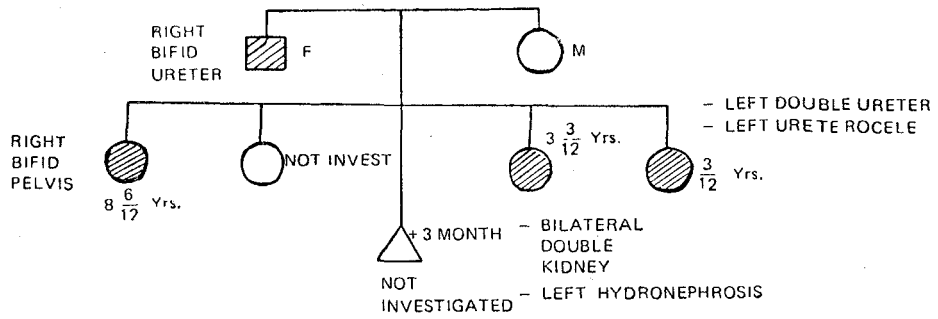

FIG. 5 *Pedegree of the family*

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