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CASE REPORT

Ureteropelvic junction obstruction and calyceal diverticulum in a child with Turner syndrome and horseshoe kidney

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Abstract Laparoscopic dismembered pyeloplasty for ureteropelvic junction (UPJ) obstruction is considered to be a routine procedure in many pediatric surgical centers. UPJ obstruction is known to be associated with horseshoe kidney and several reports on successful laparoscopic repair in such cases exist.

The case of a 9-month-old girl with Turner syndrome is reported. A horseshoe kidney with grade 4 hydronephrosis on the left side was diagnosed by ultrasound during the neonatal period. MAG3 diuretic renography and dynamic magnetic resonance imaging nephrography revealed a differential renal function of 31% and 69% on the left and right side, respectively. No drainage from the left renal pelvis could be demonstrated.

Laparoscopy showed a combined UPJ obstruction and a calyceal diverticulum with a narrow infundibulum of the upper pole calices on the left side of the horseshoe kidney. Laparoscopic dismembered pyeloplasty and an additional infundibulopelvic anastomosis was performed. No intraoperative complications occurred. The immediate postoperative course was uneventful. Unobstructed drainage and stable differential renal function on the left side could be demonstrated on MAG3 diuretic renography 6 weeks postoperatively.

In conclusion, laparoscopic repair of complex malformations of the upper urinary tract is feasible and leads to good functional outcome in selected cases.

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Introduction

Infundibulopelvic dysgenesis results in a wide variety of disorders. Depending on the localization and degree of the lesion, they range from focal forms, like calyceal diverticulum, pyelogenous cyst and ureteropelvic junction (UPJ) obstruction, to infundibulopelvic stenosis and multicystic kidney [1].

Turner syndrome is known to be associated with malformations of the urinary tract in 30—40% of patients. By ultrasound examination collecting system malformations are found most frequently, followed by horseshoe kidney and other positional abnormalities [2]. In addition, anomalies of structure and of the collecting system are thought to be linked to the 45, X Turner syndrome and mosaic/structural X karyotype, respectively. The case and management of an infant with Turner syndrome, horseshoe kidney and left-sided grade 4 hydronephrosis, due to the combination of an UPJ obstruction and a calyceal diverticulum, is reported.

Case report

A 9-month-old girl with Turner syndrome presented with a horseshoe kidney and left-sided grade 4 hydronephrosis that had been diagnosed during the neonatal period. MAG3 diuretic renography and MRI nephrography (Fig. 1A) showed a differential renal function of 31% and 69% on the left and right side, respectively. No drainage could be demonstrated from the left renal pelvis. Under ultrasound and MRI examination a duplicated collecting system was suspected on the left moiety of the horseshoe kidney (Fig. 1B). VCUG showed grade II reflux into the right part of the kidney. No VUR could be demonstrated on the left side. After diagnostic work up, the patient was prepared for laparoscopic dismembered pyeloplasty for UPJ obstruction.

With the patient under general anesthesia, a subumbilical mini-laparotomy was performed in the midline and a 5-mm trocar was used as a camera port. Pneumoperitoneum was established using a pressure of 10 mmHg and a flow rate of 2.5 L/min of carbon dioxide. Two additional 3-mm trocars were placed in the right lower and left upper abdomen. After mobilization of the descending colon the left moiety of the kidney was identified, but no duplicated collecting system could be demonstrated. In contrast, further exploration revealed a large calyceal diverticulum of the upper moiety, with the left renal artery crossing the narrow infundibulum. Additionally, the renal pelvis of the lower moiety was massively dilated due to a UPJ obstruction (Fig. 1C; Video 1). The narrow infundibulum of the upper

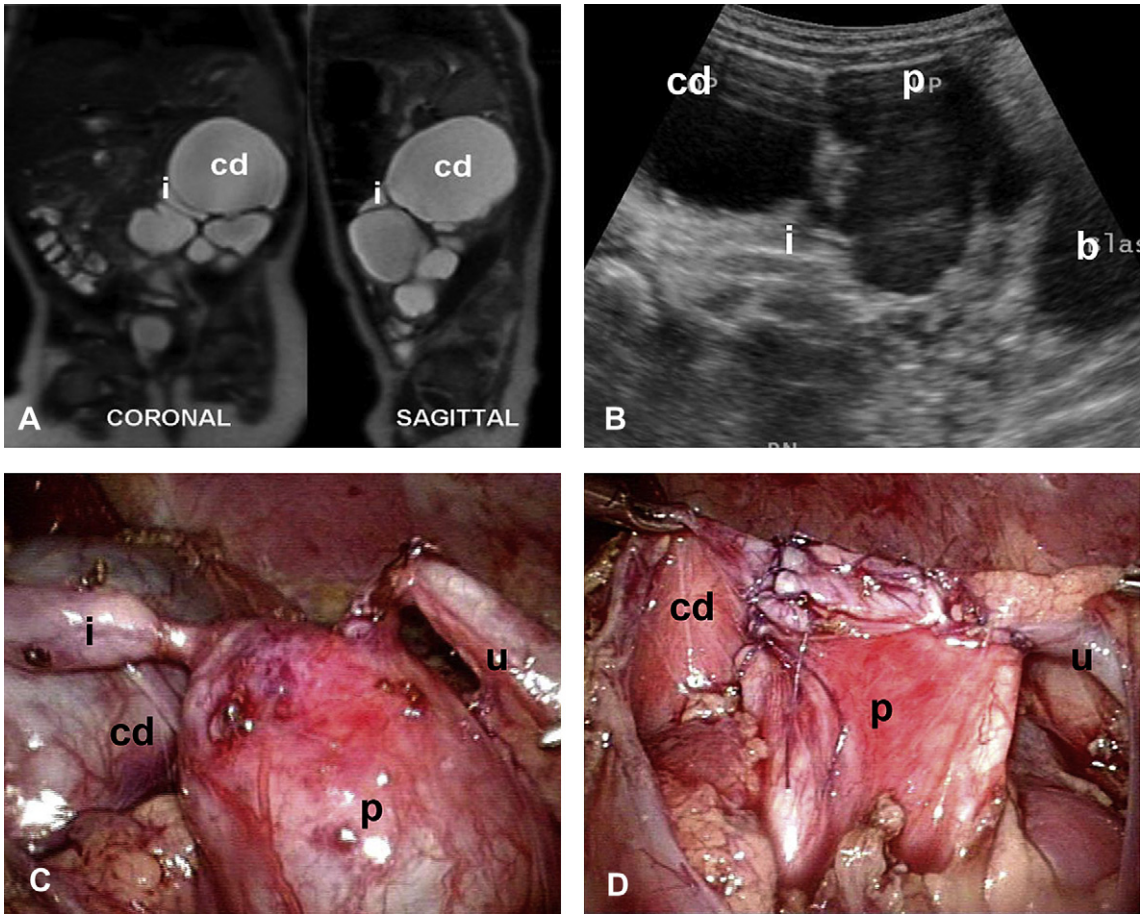

Figure 1 Preoperative MRI nephrography (A) and ultrasound examination (B) showed dilation of the left renal pelvis in a horseshoe kidney. A duplicated collecting system was suspected, but intraoperative exploration (C) revealed a large calyceal diverticulum (cd) with a narrow infundibulum (i) and dilation of the renal pelvis due to an UPJ obstruction. (D) Anastomosis of the dismembered pyeloplasty (u: ureter) as well as between the calyceal diverticulum (cd) and the renal pelvis (p).