Horseshoe kidney and uretero-pelvic-junction obstruction in a pediatric patient. Laparoscopic vascular hitch: A valid alternative to dismembered pyeloplasty?

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Abstract

Horseshoe kidney (HSK) is a congenital defect of the urinary tract that occurs in 0.25% of the general population. Laparoscopic Vascular Hitch (LVH) according to Hellstrom-Chapman represent an alternative approach in treatment of extrinsic hydronephrosis by crossing vessels (CV) in pediatric age. In our Department from 2006 to 2016, 36 children with extrinsic-Uretero-Pelvic-Junction (UPJ)-Obstruction (UPJO) underwent laparoscopic vessels transposition. Over the last 4 years, we have treated three patients with extrinsic hydrenephrosis in HSK; two males and one female respectively of 6, 7 and 8 years. The side affected was the left in all patients; symptoms of onset: recurrent abdominal pain, vomiting with associated intermittent hydrenephrosis at ultrasonography. The preoperative examinations performed were: ultrasound/Doppler scan, MAG3-renogram, functional-magnetic-resonance-urography (IMRU). Mean operative time was 120'; median hospital stay 3–days. Intraoperative diuretic-test (DT) confirmed an extrinsic-UPJO in all patients. No JJ-stents and drain were used and there were no perioperative complications. Clinical and ultrasound follow-up (18 months–4 years) show resolution of symptoms and decrease in hydronephrosis grade in all patients. Our series is the largest in pediatric population by a revision of the literature. We believe that LVH is feasible in patients with symptomatic hydrenephrosis by CV in HSK. Intraoperative-DT and the correct selection of patients are crucial to the success of the technique. According to us, this procedure is appropriate in those cases where the UPJ-anatomy is disadvantageous to a resection/re-anastomosis between ureter and renal pelvis. Our initial results are encouraging, although long-term follow-up and a more significant patient sample are required.

Introduction

Horseshoe kidney is a known congenital anomaly involving the upper urinary tract, which takes place as result of a developmental defect occurring between 4th to 8th weeks of embryogenesis. This congenital anomaly resulted from an abnormal mediastinal fusion of the metanephric blastema, originally destined to result in anatomically normal right and left kidney causing a failure of ascent and rotation. It occurs in 1 in 400–800 individuals with an incidence of 0.25% and a male predominance (2:1 relation). The fusion area of the lower poles of both kidneys consists of parenchymal or fibrous tissue, also known as isthmus. The renal vessels thus develop an abnormal relation to the renal pelvis and ureters. Normal kidney in the course of development migrates from near the level of the second sacral vertebra to the lumbar region. Horseshoe kidneys on the contrary, are usually ectopic, low in position and sited at the aortic bifurcation. The fusion is usually found between inferior poles of kidneys (90%), but it may occur also at the upper poles.

The ureter arises from the anterior surface of the pelvis; it tends to be obstructed if the pelvi-ureteric junction is high or if the ureter crosses over the prominent lower pole or bulky isthmus. Anomalies in the blood vessels are common. About one third of all HSKs have a normal pedical blood supply, but in other two thirds of them the vascular relations are abnormal.
an abnormal supply to the isthmus is seen. There are frequently one, two or three arteries to each kidney arising from the aorta and a further one running to the upper or lower part of the isthmus.

Materials and Methods

Laparoscopic vascular hitch in horseshoe kidney included 3 patients aged 6, 7 and 8 years. All patients were admitted to our Department from 2012 to 2016, for recurrent episodes of intense abdominal pain (flank pain) associated with nausea and vomiting. Based on ad hoc ultrasonography (USG), an intermittent hydronephrosis with primarily extrarenal dilatation was discovered suspecting an extrinsic obstruction by crossing vessels in all patients (Figure 1). In two patients, an HSK was suspected at ultrasound. In one patient, it was discovered with fMRU (Figure 2). The median age at presentation was 6.5 years. None of the children had prenatal diagnosis. Preoperative diagnostic studies were: ultrasound/Doppler Scan, 99mTc-MAG3, and fMRU. All presented high-grade hydronephrosis (GIII–GIV) according to the Society of Fetal Urology classification. Preoperative diuretic renogram results invariably displayed an obstructive washout curve. Surgical indications for extrinsic UPJO in our center included two or more of the following conditions: presence of clinical symptoms, obstruction on diuretic renogram (99mTc-MAG3), decrease on relative renal function, clear or suspected image of polar vessels on functional-magnetic-resonance-urography (fMRU), worsening of intermittent hydronephrosis on follow-up. All patients qualified for laparoscopic surgical treatment. They were hospitalized 24 hours before surgery, starting with liquid diet and bowel cleansing with laxative and enemas. All underwent Hellström Vascular Hitch modified by Chapman with DT by administering a bolus of normal saline (20 mL/kg IV) starting 10’-15’ before the laparoscopic vessel mobilization, followed by furosemide 1 mg/kg IV early in the operative course.

Surgical technique

The procedure was performed with a transperitoneal approach using three trocars: an optical port of 5 mm with a 30° laparoscope and two 3-mm working ports, in the epigastrium and in the ipsilateral iliac fossa (IF) at the midclavicular line, to allow an ideal triangulation during dissection of the aberrant CV and completion of the pelvic wrap. The lower position of HSK required a lower position of the two trocars, some cm under the median line in epigastrium and under the midclavicular line in IF compare with normal kidney. The patients were placed at the edge of the surgical table in a modified lateral decubitus (45°). The technique, consolidated in our center, consisted in the exposure through a mobilization of colonic flexure or a window in the mesocolon (on the left side) of the dilated pelvis, CV dissection and mobilization off the UPJ, releasing the ureteral adhesions. Checked the full mobility of the UPJ (shoeshine manoeuvre) confirming the appropriate mobilization of the pelvis wall, inspected and excluded an UPJ intrinsic stenosis (with DT) the vessels were sutured in a tunnel formed by an invagination of the anterior pelvic wall as described in the Chapman procedure with two interrupted 4/0-polydioxanone absorbable sutures (Figure 3). With HSK in these patients the first step was the identification of the isthmus of HSK, following the lower pole of kidney and visualization of the lower pole vessels. With this malformation is very important to have a clear image of the anatomy: is possible o find different vascular bundles, and a distortion of the ureters due to isthmus.

Results

Mean operative time was 120’ (range 100’-150’). One patient was affected by Goldenar Syndrome. Two patients presented 2 vascular bundles (Figure 4) while one only two vessels (artery and vein) obstructing the junction. The intraoperative diuretic-test (DT) confirmed an extrinsic-UPJO in all patients. No JJ-stents and drain were used. There were no complications either during or after the procedure. Nutrition starts on the first/second day. On the third day, all patients were discharged from the hospital. Clinical and ultrasound follow-up (18 months-4 years) show resolution of symptoms and a decrease in the hydronephrosis grade in all patients (Table 1).

Discussion

HSK is the most common congenital fusion abnormality of the urinary tract, which combines three anatomic abnormalities: ectopia, malrotation and vascular anomalies. The HSK can be classified according to the morphological appearance of fusion. The U-shaped HSK is formed by the medial fusion of the kidneys placed in symmetrical position on either side of the vertebral spine and...
represent the majority of cases (90%). However, a lateral fusion between a horizontal and a vertical kidney (the asymmetric L-shaped HSK) with an isthmus laterally to the midline is possible. Other rarest fusion anomalies are recognized: H-shape fusion (near upper poles), inverted U-shaped (upper poles), or fusion of both poles (Krapfen-shape). All these fusion variations are associated to ureter different insertions and course. HSK site varies along the normal embryologic ascent of the kidney but more commonly it is located in a low position (against the lumbar vertebral spine), because the isthmus does not permit ascent beyond the inferior mesenteric artery. It could be discovered in all age groups from fetal life (first trimester) to adult, but is more prevalent in children.6 There is a wide variation in horseshoe-kidney vascular supply. Renal arteries can originate from the aorta, the iliac arteries, and the inferior mesenteric arteries.7,8 A classification system with six basic patterns of arterial supply for each HSK segment has been proposed.10 Furthermore, HSK is frequently accompanied by Inferior Vena Cava (IVC) abnormalities such as the double, left and pre-isthmic IVC. The HSK has been reported to have a close relationship with vascular, calyceal and ureteral abnormalities.6 One-third of all patients with a horseshoe kidney are asymptomatic, and the diagnosis is made incidentally during radiologic examination. The most common associated pathology in HSK is uretero-pelvic-junction-obstruction, which occurs in up to 35% of cases and is often the cause of problems. Stones develop in 20% to 60% of patients and may be associated with obstruction and recurrent infections. Obstruction determines urinary stasis and urolithiasis predisposing to kidney infection (27% to 41% of patients), HSK is also associated with some neoplasms is higher. The risk of Wilms' tumor is 2-fold in a horseshoe kidney accounting for 28% of malignant lesions.11

In children UPJO is the more frequent pathology associated to HSK. UPJ in fact, is located higher than normal. This situation can possibly result in UPJ obstruction of various degrees. The ureter

Table 1. Patients’ data.

<table>
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<th>Patient 1</th>
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<th>Patient 3</th>
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<td>US, MAG3, fMRU</td>
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<td>Hospital stay (days)</td>
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UPJ, uretero-pelvic-junction; US, ultrasound; MAG3, renogram; fMRU, functional-magnetic-resonance-urography.
while descending to the bladder usually crosses the anterior surface of the renal isthmus. The possible causes of obstruction of the UPJ in horseshoe kidney may be due to: high insertion of the ureter, distortion of the proximal ureter as it loops over the renal isthmus, compression from the aberrant vessels at the hilum, intrinsic pathogenesis. Many times, this has to be surgically corrected. Classic literature on the surgical management of UPJO in horseshoe kidneys includes the standard open dismembered pyeloplasty with sometimes-associated isthmoectomy.

Since first being introduced by Schussler in 1993, minimally invasive pyeloplasty has become a well-established treatment for UPJO. By a revision of literature only a limited number of reports have been published on the application of laparoscopic pyeloplasty for the treatment of UPJO associated with HSK. The first dismembered laparoscopic pyeloplasty in a horseshoe kidney was reported in 1996. Since then, fewer than 57 cases of UPJO in HSK have been reported in the literature until 2017 including among these series a small number of pediatric patients: Faddegon et al. (2012) 4 patients, Blank et al. (2014) 10 patients, Shadpour et al. (2015) 8 patients, Oderda et al. (2017) 1 patient, Moscardi et al. (2017) 1 patient.

These sparse reports may reflect the relative rarity of horseshoe UPJ obstruction, but may also relate to a perceived increase in surgical difficulty due to unfamiliar and variable anatomy. All that is rarest if we consider the pediatric population patients. In fact, by a review of literature we count only 24 pediatric patients with HSK treated for symptomatic UPJO. The treatment of choice prevalently described is dismembered pyeloplasty realized with various minimvasive approach: laparoscopic, Laparoscopodisco Single-Site(LESS), robotic. Among those patients we found only three Helstrom Vascular Hitch (2 in Faddegon series, 1 in Shadpour series).

Our cohort so added three other pediatric patients to literature, all treated with laparoscopic vascular hitch according to Hellstrom-Chapman. Highly variable anomalous blood supply to horseshoe kidneys further increases the potential for extrinsic compression of ureter. The real contribution of crossing vessels to obstruction is difficult to demonstrate and much less to quantify, but an accurate diagnostic preoperatory study as we do in our center, (US, MAG3, fRMU) consents to select the appropriate patient for LVH. Transperitoneal laparoscopy can provide a clear picture of the real anatomy in particular in horseshoe kidney and an adequate workspace to release the ureter and vessels. Intraoperatory diuretic test confirm the real extrinsic obstruction and consent to perform LVH. The majority of pediatric and adult patients with HSK and UPJO underwent dismembered pyeloplasty which is considered in different reported series the gold standard treatment despite the anatomical technical challenges due in this population to aberrant lower pole vessels, unfamiliar caudal position of the kidney, and the renal isthmus.

Conclusions

Currently, there is no consensus regarding the optimal surgical approach for the treatment of horseshoe UPJO; but following our experience, which include the largest series of children reported in literature with extrinsic-UPJO in orthotopic kidney treated with Helstrom-LVH (success rate of 97%), we believed that the technical challenges due to distorted anatomy make vessels transposition an optimal alternative solution compared with pyeloplasty. This case series is limited by its small number, thus making impossible a meaningful statistical comparison of LVH in orthotopic kidneys. Due to the rarity of horseshoe kidney, large case series will take time to occur, which limits our ability to interpret the outcomes and/or advantages of various techniques. However, intraoperative-DT and the correct selection of patients are crucial to the success of the technique. Our initial results are encouraging, but long-term follow-up and a more significant patient sample are required.

References