



# Bilateral En-Block Horseshoe Kidney Laparoscopic Nephrectomy

## *Nefrectomía bilateral en bloque en riñón en herradura*

Nicolas Fernandez<sup>1</sup> Mandy Rickard<sup>2</sup> Rebeca Escobar<sup>3</sup> Walid Farhat<sup>4</sup>

<sup>1</sup>Division of Urology, Seattle Children's Hospital, University of Washington, Seattle, Washington, United States

<sup>2</sup>Division of Urology, Hospital for Sick Children (SickKids), University of Toronto, Toronto, Canada

<sup>3</sup>Division of Pediatric Urology, Fundación Santa Fe de Bogotá, Bogotá, Colombia

<sup>4</sup>Department of Urology, University of Wisconsin School of Medicine and Public Health, Madison, Wisconsin, United States

**Address for correspondence** Nicolas Fernandez, MD, PhD, Division of Urology, Seattle Children's Hospital, University of Washington, 4800 Sand Point Way NE, Seattle, WA, 98105, United States (e-mail: Nicolas.fernandez@seattlechildrens.org).

Urol Colomb 2022;31(1):e28–e31.

### Abstract

Horseshoe kidney (HSK) has a prevalence of 1 in every 500 individuals. The management of patients with HSK is usually conservative, except in the presence of symptoms such as obstruction, stones, glomerulopathies, and tumors. In the following case report, we describe how a bilateral en-block transmesenteric laparoscopic nephrectomy in supine position was performed.

A 5-year-old boy, with proximal hypospadias and early onset of chronic kidney disease due to focal segmental glomerulosclerosis on biopsy, underwent a genetic molecular evaluation that confirmed a pathogenic mutation at the *WT-1* gene. Due to the increased risk of developing Wilms tumor, he underwent a bilateral transmesenteric nephrectomy.

In a five-minute video, we describe how we performed an en-block transperitoneal and transmesenteric laparoscopic nephrectomy with special attention to patient positioning, including the feasibility of performing the dissection of the left renal hilum and isthmus with the patient in supine with no need for repositioning, and then moving to the dissection of the right renal hilum and completion of the procedure.

The case herein reported enables us to describe the technical key-points to perform a bilateral en-block laparoscopic nephrectomy with shorter operative time and reduction of blood loss by preserving the entire specimen, without the need for an isthmus transection.

### Keywords

- ▶ horseshoe kidney
- ▶ laparoscopy
- ▶ nephrectomy

### Resumen

El riñón en herradura (RH) tiene una prevalencia de 1 en cada 500 individuos. El manejo del RH es usualmente conservador, excepto cuando genera síntomas como obstrucción, litiasis, glomerulopatías o tumores. Con este reporte de caso, describimos como

received  
December 20, 2020  
accepted  
August 13, 2021

DOI <https://doi.org/10.1055/s-0041-1736597>.  
ISSN 0120-789X.  
e ISSN 2027-0119.

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se realizó una nefrectomía bilateral en bloque transmesentérica en un paciente con riñón en herradura.

Un paciente de 5 años de edad, con hypospadias proximal y desarrollo temprano de enfermedad renal crónica por glomeruloesclerosis focal segmentaria, fue sometido a un estudio molecular que confirmó la presencia de una mutación en el gen *WT-1*. Dado el alto riesgo de desarrollar tumor de Wilms, se decidió realizar una nefrectomía transperitoneal laparoscópica bilateral.

En un video de cinco minutos, describimos como se realizó una nefrectomía transperitoneal y transmesentérica en bloque, con especial atención al posicionamiento del paciente, incluso la viabilidad de realizar la disección del hilio renal izquierdo y el istmo con el paciente en supino, sin necesidad de reposicionarlo, y, después, la disección del hilio renal derecho y el fin del procedimiento.

#### Palabras clave

- ▶ riñón en herradura
- ▶ laparoscopia
- ▶ nefrectomía

El caso reportado nos permite describir los puntos clave técnicos para realizar una nefrectomía laparoscópica bilateral en bloque con un tiempo operativo más corto y reducción del sangrado al preservar todo el espécimen, sin la necesidad de realizar la transección del istmo.

## Introduction

Horseshoe kidney (HSK) is the most common fusion anomaly of the kidney, and its prevalence is estimated to be of 1 in 500 individuals.<sup>1</sup> The association of HSK with other congenital anomalies has been described, as well as an increased risk of developing nephro-urological conditions, such as lithiasis and glomerulopathies. Patients with HSK have a 1.5- to 8-fold increased risk of developing Wilms tumor.<sup>2</sup> The association of Wilms tumor and hypospadias has previously been described by Köhler et al.,<sup>3</sup> who were able to identify a C to T transition at codon 362 leading to a truncated protein.<sup>3</sup> Other authors<sup>4</sup> have described the presence of *WT1* mutations in patients diagnosed with hypospadias, most of them syndromic hypospadias.

We present a five-minute video in which we describe the technical and anatomical details of an en-block transperitoneal transmesenteric nephrectomy with the intention of reducing the surgical operative time, the intraoperative bleeding, and the complications (▶ **Video 1**).

#### Video 1

Horseshoe kidney.

Online content including video sequences viewable at: <https://www.thieme-connect.com/products/ejournals/html/10.1055/s-0041-1736597>.



## Case Presentation

A 5-year-old boy was initially referred to our service for the management of proximal hypospadias. Genital surgery was performed; subsequently, at the age of 18 months, he was found to have chronic kidney disease with focal segmental

glomerulosclerosis on a kidney biopsy. He required initiation of hemodialysis with 250 ml of residual voiding volumes per day. Due to the combination of his congenital anomalies and end-stage kidney failure, a genetic workup confirmed the presence of a *WT1* gene mutation. Given the increased risk of developing Wilms tumor and the need to have hypertension under control prior to being a kidney transplant candidate, the decision to perform bilateral nephrectomies was made.

## Operative Note

The patient was placed in supine position, and was secured to the surgical table to enable extreme tilting. Since he had very low residual urinary volumes, and due to the history of proximal hypospadias, no urethral catheter was used. With the intention to complete the surgical procedure using the same ports, we placed two 5-mm trocars in the midline, one at the mesogastrium and the other at the hypogastrium, as well as a third through the belly-button. The surgical dissection was initiated by reflecting the Toldt fascia and exposing the lower pole of the left renal unit. The dissection was completed until the left ureter was identified and used as a handle for traction. With the use of LigaSure (Medtronic, Minneapolis, MN, United States), the dissection of the medial aspect of the left renal unit was completed, and the hilum was carefully ligated. This maneuver enabled us to pull traction on the kidney, which helped expose the isthmus that was completely dissected through this approach until the right renal unit was exposed. We then moved our attention to the right side, and medial reflection of the ascending colon was performed, and exposure of the right renal unit was achieved. We only changed the position of the patient by performing extreme tilting. Since the entire left renal unit and isthmus were free, traction of the right renal unit was technically easier. At this point, transection of the right ureter and careful ligation of the right hilum were

performed. The remaining proximal ureteral stump was used as a handle to pull traction on the entire HSK, which enabled a tension-free mobilization of the whole kidney underneath the mesentery and completion of the nephrectomy. Extraction of the kidney was performed through the umbilical port. The incision was extended laterally by 1 cm on each side for a total length of 3 cm wide. Closure included reconstruction of the belly button. The total operative time was of 4 hours and 22 minutes.

The patient recovered well, with acceptable pain control. He was discharged on the second postoperative day, and was submitted to continuous ambulatory peritoneal dialysis. By the time this manuscript was finished and accepted, the patient had received his renal transplant, and he is currently in stable condition 3 years after the operation.

## Discussion

We herein present the case of an en-block laparoscopic nephrectomy on a patient with end-stage renal disease who underwent an uneventful procedure. It is important to note that the entire procedure was performed without changing the patient's intraoperative position or any extra port placements. Other reports<sup>11</sup> have described a retroperitoneal approach, which might be beneficial for those cases in which an intact peritoneum is required. Nonetheless, the published operative times exceeding 10 hours until completion and the surgeons' high technical expertise on retroperitoneal surgical anatomy make this option limited and not widely applicable.<sup>5</sup> The benefit of our transperitoneal approach, in which the patient remains in supine position for the entire procedure, is the reduction in the non-surgical operative time required for patient repositioning and in the associated costs of redraping if needed. Moreover, performing the entire procedure using the same port placements is beneficial regarding the postoperative time and possible port-placement complications.

According to the surgical anatomy of HSK, both renal units and isthmus share a common fascia; this enables an en-block dissection of the entire kidney if the right surgical plane is identified.<sup>6</sup> Our patient's vascular anatomy was not as complex as it could be, considering that there were only two main arteries and one vein for each renal unit, and no main branches branching into the isthmus. Since 40% of the HSKs are located immediately below the inferior mesenteric artery, it is important to be mindful at the moment of dissecting the isthmus.<sup>1</sup> Graves' vascular classification can sometimes be very complex; we suggest a careful dissection of any structures during the entire procedure to reduce any major intraoperative bleeding complications.<sup>7</sup> At our institution, we do not perform any preoperative imaging scans prior to such cases, but we leave this to surgeon's discretion.

Some authors have reported an association of vesicoureteral reflux with HSK as high as 80%. In cases with associated vesicoureteral reflux, it may result important to consider a very distal dissection of the ureteral stump. Nonetheless, some transplant surgeons advocate leaving long ureteral stumps to be used at the moment of kidney transplantation.

The fact that this procedure was transperitoneal reduces the applicability if the peritoneum is required for dialysis. Nonetheless, careful management and dissection of the peritoneum during surgery and port closure may enable the early use of dialysis. This is supported by the fact that sometimes peritoneal dialysis catheters are made under laparoscopy. Some authors have restarted peritoneal dialysis as early as 24 hours, without any complications.<sup>8,9</sup> Depending on the clinical indications, a temporary central access can be placed prior to nephrectomy.

We acknowledge that this is the experience of a single case, and the applicability of the procedure herein described to similar clinical conditions requires further studies. Laparoscopic pediatric surgery is technically demanding, and it involves highly-technical knowledge and experience. We recommend good preoperative planning with a good understanding of the vascular anatomy prior to the case. Our results may enable the use of new robot-assisted platforms that may provide better intra-abdominal visualization in multi-quadrant surgery. Also, the use of novel technologies such as indocyanine infrared enhancement, may enable a safer and more efficient dissection of the vascular anatomy.<sup>10</sup>

## Conclusions

En-block laparoscopic nephrectomy for HSK is feasible and safe. The supine position with extreme tilting of the surgical table and midline port placement results in shorter operative times and fewer complications.

### Conflict of Interests

The authors have no conflict of interests to declare.

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