



## Partial Nephrectomy for Multifocal, Unilateral Wilms Tumor in a Patient with Hemihypertrophy

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<b>OBJECTIVE</b>	To present the surgical approach to a multifocal Wilms tumor found on screening ultrasound in an asymptomatic 2-year-old female.
<b>MATERIALS</b>	A 2-year-old female with hemihypertrophy underwent screening imaging every 3 months with renal ultrasound. A solitary, incidental renal mass was detected. Physical exam was unremarkable except for left leg hemihypertrophy. Laboratory workup was largely normal other than an elevated lactate dehydrogenase. Staging imaging revealed multiple masses on the right kidney and a normal left kidney. There were no distant metastases. The most likely diagnosis was Wilms tumor. After receiving chemotherapy for 6 weeks, imaging revealed an excellent response to chemotherapy and surgery was performed.
<b>RESULTS</b>	The patient underwent open partial nephrectomy of the 3 tumors on the right. Prior to beginning, cystoscopy and ureteral stent placement was performed as it was felt the collecting system would likely be entered during resection. The renal hilum was not clamped throughout this resection and manual parenchymal compression was used to minimize global ischemia. Retroperitoneal lymph node dissection was also performed. The patient recovered well and was discharged home 5 days after surgery. Her stent was removed at home without complications. Final pathology revealed nephrogenic rests with all 7 lymph nodes were negative for malignancy.
<b>CONCLUSION</b>	Management of multifocal, unilateral Wilms tumor with open partial nephrectomy after neoadjuvant chemotherapy is an important part of protocol management for patients with Wilms tumor with predisposition syndromes. UROLOGY 133: 243–244, 2019. © 2019 Elsevier Inc.

### VOICEOVER TRANSCRIPT

This is a 2-year-old female with hemihypertrophy found to have an incidental right renal mass on screening imaging.

Past medical history is significant for hemihypertrophy without genetic testing.

Physical exam was unremarkable except for left lower extremity hemihypertrophy. Lab workup was normal except for an elevated lactate dehydrogenase.

Screening renal US showed a 5 cm right upper pole renal mass. This was followed by a CT chest abdomen and pelvis with intravenous contrast which revealed multiple masses on the right kidney, the largest of which was identified on the screening US. The left kidney was normal and lungs were clear.

This patient has multiple renal masses in the setting of hemihypertrophy, suspicious for a predisposition syndrome

to Wilms tumor development. She had a port placed and was then started on neoadjuvant therapy with regimen EE-4a for 6 weeks, followed by repeat imaging. This revealed an excellent response to chemotherapy with all masses decreasing in size significantly. She was then taken to the operating room for cystoscopy and placement of a right double J ureteral stent with strings, open R partial nephrectomy and retroperitoneal lymph node dissection.

After ureteral stent placement, an adequate transverse abdominal incision is marked and incised sharply. The abdomen is entered and the peritoneal and liver surfaces palpated without nodularity.

The right colon is reflected medially revealing the right retroperitoneum. The renal vein, artery, and ureter are identified and tagged with vessel loops. Note that only handheld retractors are used as fixed ring retraction does not allow for fine real time adjustment in exposure that is easily done with handheld retractors.

Gerota's fascia is opened and freed from the surface of the kidney. Upward retraction of Gerota's toward the ceiling allows for easy identification of this plane.

Once the kidney is freed it is visually inspected and palpated to identify the 3 renal masses. The Bookwalter

**Conflict of Interest:** None.

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retractor is now assembled as we begin excision of the masses. Intraoperative ultrasound is used to confirm the location and extent of each of the 3 renal masses.

Each of the masses is excised in the same way. We begin at the superior pole at the largest mass. The margins of the tumor are scored using ultrasound guidance and these margins are deepened using the Bovie dissection. Note that the renal hilum remains freely flowing and unclamped throughout this entire dissection with local manual compression for hemostasis. This removes the element of ischemia time in a complex excision and reconstruction. Additionally, the small caliber of the vessels in children has been associated with an increased risk of thrombosis, which we like to avoid. The tumor is excised sharply using Metzenbaum scissors carefully ensuring a small rim of normal tissue surrounds the tumor. Intentional local suction with a fine-tipped sucker is essential to allow proper visualization. It is imperative that the hand clamping remains of consistent pressure and location through this excision to minimize bleeding and thus improve visualization.

Once the tumor is completely excised the tumor bed is inspected for hemostasis. The manual pressure is released one had at a time to visualize bleeding vessels. Bleeding areas are oversewn using figure of 8 with 4-0 vicryl.

Once hemostasis is achieved, the tumor bed is fulgurated using the Bovie. Importantly, the kidney is no longer compressed by the assistant until the next mass is approached. The renorrhaphy is then performed using 2-0 vicryl on a CT1 needle with a HemOLock and knot on the free end. Large loops are left over the surgical bed to allow for placement of GelFoam and surgical bolster. The vicryls are then cinched down using the sliding clip technique.

The second tumor was approached in the same way except that it was much larger and irregularly shaped. The collecting system was entered during this excision as the previously placed ureteral stent was seen. This was oversewn using 4-0 vicryl with the end of the stent being tucked in prior to placement of the last stitch. Resection bed vessels were oversewn for hemostasis. The base was fulgurated and the renorrhaphy performed as previously shown. Again, GelFoam and surgical bolster are used.

The third and final tumor is approached in the same way. The tumor is scored and sharply excised, hemostasis is assessed and achieved and a renorrhaphy performed.

The kidney is returned to the orthotopic location and Gerota's fascia is closed using HemOLocks. The vessel loops are removed.

Retroperitoneal lymph node dissection is then performed. Lymphatic tissue between the ureter and inferior vena cava (IVC) is excised from where the ureter crossed the iliac artery caudally, up the lateral IVC to the renal hilum cranially. Care is taken to avoid the gonadal vein entering the IVC during this dissection as this is preserved. Lymphatic channels are tied using 4-0 silk as encountered. Both the most caudal and cranial extent of this dissection is tied with a 2-0 silk to minimize lymphatic leakage.

The wound is irrigated with sterile water and the abdomen is closed in layers. The patient already had a port in place but this was not removed until final pathology returned and confirmed no further chemotherapy was necessary.

Pathology revealed perilobar nephrogenic rests without active Wilms tumor in all 3 tumors. All 7 lymph nodes were uninvolved.

The patient recovered from surgery well and was discharged home on postoperative day 5. Her stent was removed at home on postoperative day 7. She has subsequently done well, and her nadir creatinine is 0.5. She has not had recurrence or new tumor development in the 12 months postoperatively.

Learning points from this case include prenephrectomy ureteral stent placement when there is anticipated collecting system entry. Delay setting up a fixed ring retractor and use handheld retractors to allow for fine real time repositioning during kidney mobilization. In patients with small caliber vessels and complex renal excision and reconstruction, hand clamping is preferred to minimize thrombosis and reduce the stress of ischemia time. Remember that lymph node sampling is critical during excision of any renal tumor in pediatric, adolescent and young adult patients, per protocols.

 The video related to this article can be found online at: <https://doi.org/10.1016/j.urology.2019.07.031>.