Pediatric Case Reports

In Situ Gonadal Vein Valvulotomy and Side-to-side Gonado-iliac Bypass for the Management of Nutcracker Syndrome in an Adolescent With a Solitary Kidney and Absence of Pelvic Congestion

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Herein we present the first case report of laparoscopic exploration, endovascular valvulotomy, and open side-to-side bypass of a gonadal vein into the ipsilateral external iliac vein for Nutcracker syndrome in an adolescent patient with solitary kidney without pelvic congestion. Though prior management has included various large open surgeries or endovascular or extravascular approaches with the use of stents and shields, this case offers an expanded novel approach to weigh the unique challenges of the patient’s age and solitary renal status. If reproducible, this technique may gain value in the management of patients with this rare, incapacitating condition. UROLOGY 126: 200–203, 2019. © 2019 Elsevier Inc.

Nutcracker syndrome (NS) is a rare condition that can be associated with debilitating symptomatology. Compression of the renal vein between the aorta and superior mesenteric artery (SMA) causes venous hypertension, and can be accompanied by flank pain, hematuria, orthostatic proteinuria, and varicocele or ovarian plexus congestion with dyspareunia, dysuria, and dysmenorrhea. The reported symptoms suggest a disease spectrum based on the presence and degree of valvular insufficiency in renal vein tributaries.

Previous reports have suggested the benefit of gonadal bypass in the setting of a dilated system, but not in patients with competent valves.\(^2,3\) This case report develops the concept of addressing renal venous hypertension by facilitating flow away from the area of extrinsic obstruction toward a natural tributary—the gonadal vein—with in situ valvulotomy and runoff into the iliac system through side-to-side venous anastomosis.

CASE NARRATIVE

A 16-year-old female with a history of prenatal right multicystic dysplastic kidney presented with 6 months of left flank pain and intermittent microscopic hematuria. Her pain progressed to interfere significantly with school and sports. Initial assessment included a normal physical and gynecologic exam. Renal ultrasound and CT scan were unremarkable except for her now solitary left kidney.

Evaluation for NS with Doppler renal ultrasound showed a distended left renal vein (Anteroposterior (AP) diameter = 1.4 cm) tapering to 0.2 cm at the level of the SMA/aorta crossing. Peak flow velocity was 88 cm/s at the aortomesenteric segment and 20 cm/s in the hilum; yielding a ratio >4.\(^1\) MRI with gadolinium demonstrated an acute aorto-SMA angle with preferential drainage via the lumbar and hemizygos system (Fig. 1). The gonadal vein was not dilated on either exam. A venogram failed to opacify the gonadal vein but confirmed prominent collaterals in the left paravertebral plexus and hemizygos system. Supine pressure measurements were 10-12 mm Hg at the renal vein vs 8-9 mm Hg at the vena cava, approaching the diagnostic gradient range for hypertension.

The patient’s solitary kidney posed concerns due to the risk of renal injury and the potential for renal insufficiency. Interventions discussed balanced this with the need for symptomatic improvement.

Based on previous reports employing dilated tributaries of the renal vein as conduits for outflow, we proposed a novel intervention to redirect flow down a competent gonadal vein by selectively transecting its valves, analogous to in situ bypass in vascular surgery. A multidisciplinary approach was offered (Figs. 2 and 3).
Diagnostic Laparoscopy and Dissection
In the interventional radiology suite, the patient was positioned supine with left flank and pelvis elevation. Umbilical laparoscopic camera access was obtained, and 2 ports triangulated toward the left iliac vessels. We visualized a normal left ovary. The descending colon was mobilized and a nondilated left gonadal vein was isolated.

A Gibson-type incision was made. Small venous collateral branches were ligated to gain mobility. Following systemic heparin, the gonadal vein was isolated with bulldog clamps.

Retrograde Venogram and Valvulotomy
A 16-gauge angiocatheter was inserted through a venotomy, and venography performed to identify branch points, collaterals, and entry points into the renal vein. The catheter was exchanged for a 1.5 mm Le Maitre valvulotome (Le Maitre Vascular, Inc., Burlington, MA) and advanced fluoroscopically along the gonadal vein. It was deployed and retrieved from proximal to distal, taking care to avoid branch points, and repeated after a 45° rotation of the valvulotome. Venogram demonstrated no extravasation.

Gonado-iliac Bypass
The gonadal venotomy was extended to 1 cm. A Satinsky clamp was placed onto the left external iliac vein and a corresponding incision made. A side-to-side anastomosis was fashioned with 8-0 running Polypropylene suture. Clamps were removed, hemostasis ensured, and flow documented with intraoperative Doppler.

The Postoperative Course was Uneventful
The patient was discharged on aspirin prophylaxis to reduce risk of venous thrombosis. At 6 months, she
reported symptom resolution and had resumed activities. Doppler ultrasound demonstrated caudal flow in the gonadal vein with emptying into the external iliac vein.

DISCUSSION

Surgical treatment options for NS vary in invasiveness, morbidity, and effectiveness. The most studied procedures relocate the renal vein away from the extrinsic impingement through renal vein transposition or autotransplant. These interventions still typically necessitate a large abdominal incision. Moreover, the associated morbidity, occasional need for reintervention, and potential for renal damage or injury to the SMA are worrisome.

Modern strategies have addressed the obstruction with endovascular stents or extravascular shields. Although encouraging reports exist, there is potential for migration or thrombosis, along with the need for anticoagulation and unknown long-term implications. These issues are particularly worrisome in patients who may become pregnant. Placement of an extravascular shield can be performed minimally invasively, yet still demands delicate dissection around the SMA and renal vein.

There has been growing experience with redirecting venous flow away from the high-pressure area via bypass, that is saphenous reno-caval bypass, spleno-renal bypass, and inferior mesenteric-gonadal bypass. The basic principle is to decrease pressure by providing an alternative pathway for flow from the renal vein. There has been a clear progression toward using existing dilated tributaries, with drainage provided by a single anastomosis between anatomically close veins. The technical ease and lower morbidity without the introduction of a foreign body makes this an appealing strategy.

The basic principle followed in our case report appears to have been developed contemporaneously with Park et al. Our case expands their intervention as we did not transect the gonadal vein and performed it in an adolescent with a nondilated vein with intact valves and a solitary kidney. This approach was chosen over aforementioned alternatives to minimize risk to the solitary renal unit and for minimally invasive, optimal aesthetic considerations in this young female. The following key differences deserve mention:

- The gonadal vein was preserved, unaltering ovarian drainage. Flow was directed toward the iliac system through a side-to-side anastomosis, allowing for a wider anastomosis in a nondilated system.
- The solitary kidney and patient’s age were novel indications for this intervention due to significant implications of renal perfusion disruption.
- The use of laparoscopy for identifying the gonadal vein and determining feasibility of the procedure was a critical step, as the gonadal vein was not dilated and therefore difficult to identify on preoperative imaging.
- The gonadal system did not have evidence of dilation or retrograde flow on venogram, suggesting a competent valvular system. We used a valvulotome with a normal gonadal vein, following approaches employed for in situ extremity revascularization.

Although case reports represent one of the lowest levels of evidence, it is important to highlight that NS is a rare
condition lacking an ideal solution. Management strategies are limited with a paucity of comparative studies. Definitive correction should consider invasiveness, morbidity, and long-term consequences. Ideally, it should also avoid foreign body placement, prolonged anticoagulation, or instrumentation of the SMA, aorta, and renal vein. This is particularly important in younger patients who may experience progressive improvement, resolution purportedly related to tissue changes at the SMA origin which release the entrapped renal vein. The presented technique expands our armamentarium and offers a minimally invasive intervention with little disruption of vascular structures.

CONCLUSION
This is the first case report of valvulotomy and side-to-side bypass of a gonadal vein into the ipsilateral iliac vein for NS in a patient without pelvic congestion. If reproducible, this technique may help patients with this condition.

REFERENCES