Transposition of the Left Renal Vein for the Treatment of Nutcracker Syndrome in Children: A Short-term Experience

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Background: Nutcracker syndrome is caused by compression of the left renal vein between the superior mesenteric artery and the aorta. Invasive surgical intervention for this pathologic entity is controversial, particularly in the pediatric population. We aim to describe our early clinical and operative experience with such patients.

Methods: We report 3 cases of pediatric patients undergoing successful left renal vein transposition for the treatment of nutcracker syndrome.

Results: All 3 patients were female (age 9–17 years) and presented with a mean of 11.7 months of abdominal or left flank pain requiring chronic narcotic analgesia. Initial clinical presentations were associated with either hematuria or proteinuria. Diagnosis of nutcracker syndrome was supported in each case by an elevated renocaval pressure gradient and/or axial imaging demonstrating mesoaortic compression of the left renal vein. All patients underwent open surgical repair, which included left renal vein transposition, liberation of the ligament of Treitz and associated adhesions, as well as excision of periaortic nodal tissue (mean hospital length of stay 5.7 days). After mean follow-up of 13 months, all patients report complete resolution of symptoms and hematuria/proteinuria.

Conclusions: Transposition of the left renal vein is a safe and effective treatment for nutcracker syndrome in appropriately selected pediatric patients. Further experience and long-term follow-up are warranted to better evaluate the sustained efficacy of this procedure in this unique patient population.

Nutcracker phenomenon, or left renal vein entrapment syndrome, is caused by compression of the left renal vein between the superior mesenteric artery (SMA) and aorta. The normal angle between the SMA and the aorta is approximately 90°; however, more acute angulation of the takeoff of the SMA from the aorta predisposes to compression of the left renal vein. Both arterial and venous anomalies have been cited as potential contributors to this pathologic process, including the presence of a low or lateral origin of the SMA off the aorta and aberrant left renal vein branches or tributaries coursing in front of the aorta.

The nutcracker phenomenon is usually an incidental finding and not associated with symptoms. Infrequently, mesoaortic compression results in variable degrees of venous hypertension, which may manifest clinically as abdominal pain, flank pain, micro- or macroscopic hematuria, or proteinuria. In severe cases, patients may develop pelvic congestive syndrome characterized by dysmenorrhea, dyspareunia, dysuria, or varices of the pelvis, thigh, or vulva. Although the terms nutcracker phenomenon and nutcracker syndrome are frequently used interchangeably in the literature, the term nutcracker syndrome is reserved for patients with the aforementioned clinical presentation and associated morphologic features of left

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Table I. Clinical presentation of 3 pediatric patients with nutcracker syndrome

	Patient 1	Patient 2	Patient 3
Age (years), gender	14, female	17, female	9, female
Height (cm)/weight (kg)	166/65.5	160/49.9	142/30.9
Body mass index (kg/m^2)	23.8	19.5	15.3
Flank/abdominal pain	Yes	Yes	Yes
Proteinuria	No	No	Yes
Hematuria	Yes	Yes	No
Renocaval pressure gradient (mm Hg)	6	3	Not performed
Follow-up (months)	6	29	16

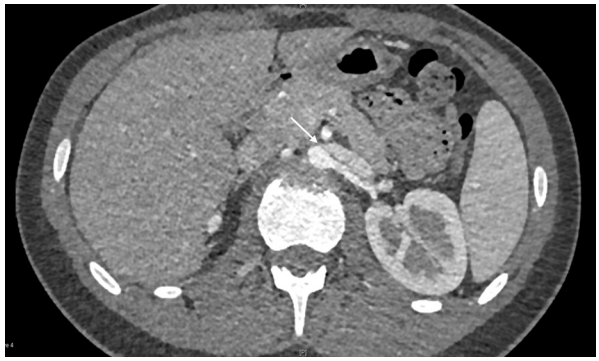

Fig. 1. Contrast-enhanced computed tomography of the abdomen demonstrating left renal vein compression between the aorta and the superior mesenteric artery (*arrow*).

renal vein compression. Diagnosis of nutcracker syndrome is often difficult and delayed, with patients frequently presenting for renal, gynecologic, or urologic consultation before ever being evaluated by a vascular surgeon. The optimal management of nutcracker syndrome, particularly when pertaining to the pediatric population, is not well recognized. We describe our experience with the diagnosis and surgical treatment of nutcracker syndrome in 3 children with refractory symptomatology (Table I).

CASE REPORTS

Case 1

A 14-year-old female was referred to vascular surgery clinic with a 4-month history of recurrent gross hematuria and associated flank pain. Her symptoms were aggravated by physical activity and deep breathing. Renal function was normal and there was no evidence of proteinuria. Use of chronic narcotics was required to ultimately achieve adequate analgesia. Initial work-up included a renal ultrasound, cystoscopy, and kidney biopsy, all of which were unrevealing. Cross-sectional imaging showed compression of the left renal vein by the SMA (Fig. 1). Subsequent venography demonstrated effacement of the renal vein just distal to the inferior vena cava (IVC) and a 6 mm Hg renocaval gradient (Fig. 2).

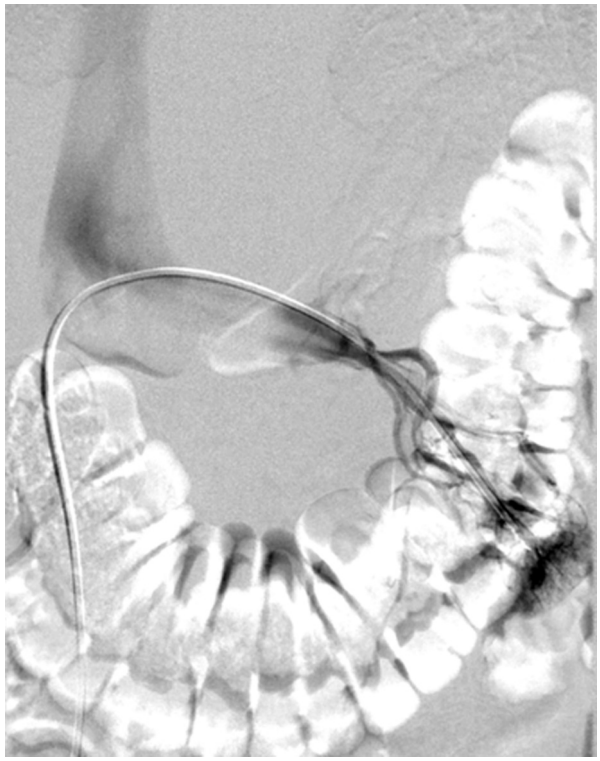

Fig. 2. Retrograde venography demonstrating compression of the left renal vein by the superior mesenteric artery. A left renal vein/inferior vena cava (renocaval) gradient of 6 mm Hg was observed.

Given clinical, physiologic, and radiographic evidence of nutcracker syndrome, operative intervention was recommended. An upper midline abdominal incision with mobilization of the transverse colon provided sufficient exposure to the retroperitoneum. The left adrenal and gonadal veins were ligated and perivascular fibrous bands were resected to completely mobilize the left renal vein. Following systemic heparinization, a side-biting vascular clamp was placed on the IVC across the left renal vein confluence. The left renal vein was transected at the caval

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