Nutcracker syndrome is caused by compression of the left renal vein between the aorta and the superior mesenteric artery where it passes in the fork formed at the bifurcation of these arteries. The phenomenon results in left renal venous hypertension. The syndrome is manifested by left flank and abdominal pain, with or without unilateral haematuria. Other common presentation is as ‘pelvic congestion syndrome’ characterized by symptoms of dysmenorrhea, dyspareunia, postcoital ache, lower abdominal pain, dysuria, pelvic, vulvar, gluteal or thigh varices and emotional disturbances. Likewise compression of the left renal vein can cause left renal-to-gonadal vein reflux resulting in lower limb varices and varicoceles in males. Its diagnosis is based on history and physical examination, basic lab tests to exclude other causes of haematuria, cystoscopy and ureteroscopy to confirm unilateral haematuria and exclude other causes of this sinister symptom. Sequence of imaging has more or less been rationalised to USS with Doppler studies, CT or MR angiography and finally phlebography with renal vein and IVC manometry to confirm the diagnosis.

Introduction

Left renal vein entrapment syndrome, characterized by the compression of the left renal vein (LRV) between the superior mesenteric artery (SMA) and the abdominal aorta was first described in 1950.1 Chait et al. described the abdominal aorta and the SMA as the two arms of a ‘nutcracker’ that can potentially compress the left renal vein.2 This description prompted the Belgian physician De Schepper to name this phenomenon as ‘Nutcracker Syndrome’ (NCS).3

The normal angle between the SMA and the abdominal aorta is approximately 90°. A normally positioned left renal vein (LRV) passes anterior to the aorta through the fork formed by the abdominal aorta and the SMA. Sagittal images on magnetic resonance angiography reveal that superior mesenteric artery originates from the aorta in almost a rectangular configuration so that SMA has a 4–5 mm course in the ventral direction before beginning a caudal descent thus resulting in an inverted J configuration. This anatomical arrangement normally prevents compression of the left renal vein by the SMA. In contrast superior mesenteric artery in cases of NCS has been noted to branch from the aorta at an acute angle (less that 90°) with initial steep caudal descent causing compression of the left renal vein in the narrow slit between the aorta and the superior mesenteric artery resulting in left renal venous hypertension. This is termed as anterior nutcracker syndrome.4

The nutcracker syndrome has also been reported in association with a retro-aortic course of the left renal vein. Retro-aortic position of the left renal vein facilitates compression of the left renal vein between the aorta and the vertebral column leading to left renal venous hypertension. This is termed as posterior nutcracker syndrome.5

A case of what can be regarded as combined (anterior and posterior) nutcracker syndrome has also been reported in association with the duplication of the left renal vein where the anterior tributary of the left renal vein was compressed between the aorta and the SMA while the posterior tributary between the aorta and the vertebral column.6
Both arterial (SMA) and venous (left renal vein) anomalies have been incriminated in the causation of NCS. Possible factors in relation to the superior mesenteric artery include abnormally low or lateral origin from the aorta, abnormal configuration of its origin or abnormal branching. Venous anomalies noted in cases of NCS include abnormal course of the left renal vein behind the aorta or bifurcation of the left renal vein with tributaries coursing in front and behind the aorta. Some investigators have also suggested other contributory or possibly exclusive factors that may cause this phenomenon. These include abnormal posterior ptosis of the left kidney with secondary stretching of the left renal vein over the aorta and abnormally high course of the left renal vein. In addition, excessive fibrous tissue have been found at the origin of the SMA, which can contribute to the compression of the left renal vein. If present the fibrous tissue needs to be excised for adequate decompression of the left renal vein.

Normal pressure gradient between LRV and inferior vena cava is less than 1 mmHg. The pressure gradient between the left renal vein and vena cava may rise up to 3 mmHg due to compression by the SMA leading to rupture of thin walled septum between the small veins and the collecting system in the renal fornix leading to haematuria. It has also been postulated that haematuria may be the result of communication between dilated venous sinuses and adjacent renal calices.

Manifestations

Review of the reported cases suggests that this condition is relatively more common in women. Most cases present in the 3rd or 4th decades of their lives although few patients have presented in adolescence and others in later years. Observation has been made that many sufferers are of above average height and tend to have an asthenic built. Buschi et al. having observed distended left renal vein on CT and sonography in 72% of patients undergoing these examination hypothesised that asymptomatic mesoaortic compression of the left renal vein is quite prevalent. However, the exact incidence of the symptomatic nutcracker syndrome is not known. Most cases in the literature have been reported from the Far-East.

The syndrome is manifested by left flank and abdominal pain, with or without unilateral macroscopic or microscopic haematuria. However, it should be noted that haematuria is not always present.

The other common mode of presentation is a symptom complex called, ‘pelvic congestion syndrome’ characterized by symptoms of dysmenorrhea, dyspareunia, post-coital ache, lower abdominal pain, dysuria, pelvic, vulvar, gluteal or thigh varices and emotional disturbances. Likewise compression of the left renal vein can cause left renal-to-gonadal vein reflux resulting in lower limb varices and varicoceles in males.

Although primarily a vascular disorder its manifestations are predominantly urological or gynaecological. Hence, most cases present either to the urologists or gynaecologists rather than vascular surgeons except when patients have lower limb varices as their main presenting complaint. Therefore, vascular and general surgeons should keep this condition in mind in patients presenting with lower limb varices or recurrent flank pain. Systemic manifestations have also been reported in adolescents including headache, abdominal pain fainting and tachycardia mimicking clinical symptoms of an orthostatic disturbance.

Investigations

Ali-el-Dein et al. proposed sequential diagnostic tests for the confirmation of classical cases of nutcracker syndrome where patients present with loin pain and unilateral haematuria. These include phase contrast microscopy, abdominal ultrasound and intravenous urography, coagulation profiles, tests for urinary bilharziasis, cystoscopic localisation of the haematuria, selective urine cytology, flexible uretero-renoscopy, renal biopsy, CT and/or MR angiography, renal vein and IVC phlebography and venous pressure manometry (Fig. 1).

The sequence of diagnostic tests, however, should depend on the mode of presentation. Patients presenting with classical symptoms of flank pain and unilateral haematuria would require investigations directed to establish the cause of haematuria. The diagnosis of vascular malformation usually comes to light as a clinical surprise. On the contrary patients with no haematuria or non-urological manifestations will require tests specifically targeted to diagnose the vascular anomaly. We present an algorithm for the sequence of diagnostic tests depending on the mode of presentation.

Real time ultrasonic imaging and Doppler flow scanning (duplex scanning) may be employed as the initial diagnostic test in patients with suspected nutcracker syndrome. Measurements should be made of the antero-posterior diameter and peak velocity at two points in the transverse plain of

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the left renal vein; (1) at the level of the renal hilum (2) at the point where left renal vein crosses between the aorta and SMA. Diagnosis of nutcracker phenomenon should be suspected when antero-posterior diameter and peak velocity at these two points exceeds by a factor of five.\textsuperscript{18} The sensitivity and specificity of colour Doppler sonography for diagnosing nutcracker syndrome have been reported as 78 and 100\%, respectively.\textsuperscript{19}

Following an initial Doppler ultrasound, patients suspected of having NCS should undergo computerized angiographic tomography of the abdomen to delineate the anatomical relation of LRV with aorta and SMA. CT angiography is non-invasive and has excellent diagnostic yield although has the disadvantage of radiation exposure. In recent years it has become the investigation of choice in many centres. It is recommended that angiographic CT should be performed before phlebography and LRV pressure measurement in patients with left renal haematuria who are suspected of having nutcracker syndrome.\textsuperscript{20}

Magnetic resonance angiography offers the advantage of multi-planer imaging in transverse, coronal and sagittal planes of the body thus, allowing an excellent anatomical definition. MRA also has the added advantage of being radiation free. Abnormalities noted on MR angiography in cases of NCS include dorso-lateral torsion of the left kidney, abnormally high course of the left renal vein, abnormally low or lateral origin of SMA, and abnormal configuration of origin of SMA. Thus, MR angiography provides a non-invasive radiation free alternative to CT angiography in the diagnostic work up of this patient population.

Retrograde phlebography and cine video-angiography with reno-caval pressure gradient determination is accepted as the gold standard in establishing the final diagnosis of NCS. Cine video-angiography allows visualisation of the point of compression of the left renal vein at the meso-aortic crossing and also demonstrates peri-renal and peri-ureteral venous collaterals with reflux into the adrenal and gonadal veins and stagnation of the contrast in the renal vein.\textsuperscript{5} The normal reno-caval pullback mean gradient pressure ranges from 0 to 1 mmHg. The pressure gradient between the left renal vein and IVC should be $>3$ mmHg to clinch the diagnosis of NCS.\textsuperscript{21}

In an interesting study, Andrianne et al. reported\textsuperscript{22} an intra-operative technique of confirming the diagnosis of NCS associated with gross haematuria. They demonstrated that clamping of the renal vein is followed by the immediate appearance of massive and reversible haematuria. Authors also suggested that a pre-operative percutaneous haematuria provocation test can be performed during venography by temporarily obstructing the left renal vein. This would allow formal diagnosis of this syndrome when the condition is highly suspected.

**Management**

Management of the renal nutcracker syndrome has been evolving over the last four decades. The available options can be sub-classified into four groups (Table 1).

a. Surveillance.
b. Open surgical procedures.
c. Intra- or extra vascular stents.
d. Intra-pelvic chemical cauterisation.

**Surveillance**

Patients with microscopic haematuria or intermittent short periods of painless gross haematuria, insignificant pain and a normal haemogram may be followed up closely without treatment. In particular conservative approach should be adopted in cases of
 puberty of the renal vein, the renal artery and the inferior vena cava are performed simultaneously.

**Open Surgical Correction**

Pastershank was the first to report the surgical treatment of a patient with nutcracker syndrome in 1974. This procedure entailed division of a fibrous tunnel between the aorta and the superior mesenteric artery to release the left renal vein. The patient was asymptomatic at 2 years follow up.24 Ever since, different open surgical procedures of varying complexity have been performed to rectify the problem. One of these was anterior nephropexy with excision of renal varicosities.7

Aim of this procedure was to correct posterior ptosis, (assumed to be the cause of NCS) and excise the varices, which were deemed to be the consequence of renal mal-positioning. However, simple nephropexy with excision of varicosities is no more recommended as it fails to address the primary pathology.

Other open surgical procedures employed included left renal vein transposition, transposition of the superior mesenteric artery, renal auto-transplantation and gonado-caval bypass.

Left renal vein transposition involves division of the left renal vein at its junction with the inferior vena cava, repair of the vena caval defect and re-anastomosis of the left renal vein to the inferior vena cava at a lower level away from the superior mesenteric artery.24

Transposition of the superior mesenteric artery is based on similar surgical principles and entails transection of the SMA at its junction with the abdominal aorta and re-anastomosis to the abdominal aorta at a lower position away from the left renal vein.18

Vessel transpositions although successful entail major surgical procedures, are associated with risks of bleeding, thrombosis and paralytic ileus. Consequences of superior mesenteric arterial thrombosis would be devastating.

Renal auto-transplantation involves nephrectomy as in live donors and transplantation of the kidney into either ipsi-lateral or contra-lateral iliac fossa.20,25

Gonado-caval bypass has been employed in patients with NCS associated with pelvic varices but has not gained wide popularity.14 Renal vein transposition and auto-transplantation have been associated with good outcomes and relatively less morbidity while SMA transposition has fallen into disfavour because of higher post-operative complication rate.

**Stenting**

Intra-vascular stenting

Intra-vascular stenting in the treatment of nutcracker syndrome using expandable metallic stents was first reported by Neste et al. The implantation of the stent is performed under digital subtraction angiography (DSA) guidance under local anaesthesia. A 8F delivery system with an endograft is introduced via the femoral vein and advanced up to the left renal vein. It is deployed in the narrow portion of the left renal vein while its medial edge is placed in the inferior vena cava. Follow up of the treated patients has ranged from 4 to 54 months. All patients were reported to be asymptomatic. Ever since, sporadic cases of intra-vascular stenting have been reported using a variety of stents but with relatively short follow up.26–29

Zhang et al.25 compared the outcomes of three patients who underwent transposition of SMA to three who underwent endograft stent (Wallstent) placement into LRV under digital subtraction angiography (DSA). Follow up ranged from 4 to 54 months. All patients were free of symptoms of loin pain, haematuria and dizziness. Pre-operatively noted proteinuria also resolved in all cases.

It is interesting to note that in spite of all stents being in the right place, with narrow portion of the left renal vein well expanded there was no change in the venous pressure at follow up in some patients. Similar observation was made by Sharper et al.6 After treating five patients by open surgery involving renal vein transposition (n=4) and gortex graft renal vein interposition (n=1) no drop in venous pressure was noted at follow up. Interestingly haematuria resolved in four and improved in one. Thus, it is a strange paradox that the main criterion on which the diagnosis of the condition is based remains unaltered in some patients even after what appears to be an effective treatment, yet symptoms resolved in almost all reported cases. It, therefore, remains a mystery what causes symptoms of pain and haematuria in this population of patients. However, others have reported drop in the venous pressure after open surgery and endo or extra-vascular grafting.14

Although endovascular stenting is a simple and attractive option its future role in the management of this condition remains to be established. Stents in the venous system can cause fibromuscular hyperplasia, which may result in venous occlusion. Another possible complication is proximal migration or embolization. Thus, we should await the long-term outcomes before deciding on the place of endovascular
Table 1. Treatment and follow up of various cases

<table>
<thead>
<tr>
<th>No.</th>
<th>Year</th>
<th>Author</th>
<th>Treatment methods</th>
<th>Follow up</th>
<th>Outcome</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>2004</td>
<td>Koyama A. et al.</td>
<td>Conservative</td>
<td>14 months</td>
<td>Hematuria and proteinuria settled</td>
<td>1</td>
</tr>
<tr>
<td>2</td>
<td>2004</td>
<td>Waga S. et al.</td>
<td>Conservative</td>
<td>7 years</td>
<td>Hematuria settled</td>
<td>1</td>
</tr>
<tr>
<td>3</td>
<td>2004</td>
<td>Moll F.L. et al.</td>
<td>Endovascular stent</td>
<td>15 months</td>
<td>Symptoms subsided</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>2003</td>
<td>Song Bo et al.</td>
<td>0.1% gm/ml silver nitrate delivered to pelvic membrane via URS</td>
<td>16 months</td>
<td>Hematuria resolved</td>
<td>1</td>
</tr>
<tr>
<td>5</td>
<td>2003</td>
<td>Min Zhou et al.</td>
<td>Venous endoprosthesis stent via Lt transfemoral vein + antiPLT therapy</td>
<td>3 months</td>
<td>Symptom free</td>
<td>1</td>
</tr>
<tr>
<td>6</td>
<td>2003</td>
<td>Zhou et al.</td>
<td>Superior mesenteric artery transposition (n=3)</td>
<td>50 months</td>
<td>n=5—hematuria settled</td>
<td>6</td>
</tr>
<tr>
<td>7</td>
<td>2003</td>
<td>Ghoneim et al.</td>
<td>Renal autotransplantation (n=6)</td>
<td>Time not mentioned</td>
<td>n=10—pain free</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Transposition of LRV (n=1)</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Venolysis, widening of window behind aorta + omental packing in window (n=1)</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Conservative management (n=3)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>2003</td>
<td>Feng Hua et al.</td>
<td>SMA transposition (n=2)</td>
<td>4-54 months</td>
<td>Symptoms disappeared</td>
<td>6</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Transposition of SMA via extraperitoneal approach (n=3)</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Endovascular stent</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Implantation of endograft stent in LRV (n=3)</td>
<td>9 months</td>
<td>Pain free, HTN resolved</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Conservative management (n=3)</td>
<td></td>
<td>90% success in stent outcome</td>
<td>9</td>
</tr>
<tr>
<td>9</td>
<td>2003</td>
<td>M. Kohno et al.</td>
<td>ES (external stent) (n=2) one of the stents inserted through laproscopic approach</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>IS (Internal stent) (n=1)</td>
<td>Mean follow up 36 months</td>
<td>12–72 months</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>GCBB (Gonado caval bypass) (n=3)</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Conservative management (n=3)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>2001</td>
<td>Scultetus et al.</td>
<td>SMA transposition (n=2)</td>
<td>4 months</td>
<td>No hematuria or renal hypertension</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Conservative management (n=3)</td>
<td></td>
<td>Venography showed restoration of blood flow</td>
<td></td>
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<td></td>
<td></td>
<td></td>
<td>Expandable metallic stent inserted</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>2000</td>
<td>S.H. Yu et al.</td>
<td>SMA transposition (n=2)</td>
<td>4 months</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>12</td>
<td>1999</td>
<td>Katsuoka et al.</td>
<td>Hemostatic agents and injection of silver nitrate—unsuccesful</td>
<td>6 months</td>
<td></td>
<td>1</td>
</tr>
<tr>
<td>13</td>
<td>1997</td>
<td>Noaki Mitsuhashi et al.</td>
<td>Expandable metallic stent inserted Conservative management</td>
<td>Resolved after pregnancy</td>
<td>Symptons settled</td>
<td>1</td>
</tr>
<tr>
<td>14</td>
<td>1997</td>
<td>Ping-Chin et al.</td>
<td>Exploratory laparotomy angle between aorta and SMA 15 degree</td>
<td>Time not mentioned</td>
<td>symptom free</td>
<td>1</td>
</tr>
<tr>
<td>15</td>
<td>1994</td>
<td>G. Williams et al.</td>
<td>Auto transplantation of left kidney</td>
<td>7–36 months</td>
<td>n=4—no hematuria, n=1—occasional hematuria</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Anterior transposition of LRV (n=3)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Venolysis of anterior vein + transposition of posterior vein anteriorly with anastomosis to IVC</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>n=1 with duplicated vein</td>
<td></td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>n=1 with posterior vein</td>
<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>LRV anterior transposition (n=2)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Autotransplantation</td>
<td>4-6 weeks</td>
<td>Hematuria settled post-operatively</td>
<td>3</td>
</tr>
<tr>
<td>16</td>
<td>1994</td>
<td>Ghoneim et al.</td>
<td>n=1 Refused surgery</td>
<td></td>
<td>Hematuria resolved</td>
<td>3</td>
</tr>
<tr>
<td>17</td>
<td>1991</td>
<td>Hohenfellner et al.</td>
<td>n=1 Nephrectomy</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>n=2 Transposition of LRV through midline approach</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>1988</td>
<td>Ernest et al.</td>
<td>Open external stent of reinforced PTFE</td>
<td>9 months</td>
<td>Asymptomatic</td>
<td>3</td>
</tr>
<tr>
<td>19</td>
<td>1982</td>
<td>Reiman et al.</td>
<td>Direct left renaloc transplanted + resection of collateral vessels</td>
<td>1–4 years</td>
<td>Asymptomatic</td>
<td>3</td>
</tr>
</tbody>
</table>

LRV, left renal vein.
stents in the management of nutcracker syndrome (Table 1).

**Extravascular stent**

Extra vascular stent using a ring reinforced PTFE graft was first reported by Barnes et al. in 1988. This procedure was performed by open surgery. After mobilisation of the duodenum left renal vein was mobilised and a 14F ring reinforced poly-tetra-fluoroethylene (PTFE) graft was applied around the left renal vein. The length of the graft was chosen to cover the segment of left renal vein medial to the point of entry of left adrenal and gondal veins to its junction with the inferior vena cava. Scultetus et al. subsequently reported laparoscopic extravascular stenting with excellent outcome at short-term follow up. It is recommended that patients undergoing any of the stenting procedure should be commenced on long-term anti-platelet medication.

**Intra-pelvic Chemical Acauterization**

A single case was reported from China in 2003 by Gong Yu and colleague, who treated a patient presenting with intermittent gross haematuria and left loin pain with twice weekly instillations of 0.1% silver nitrate solution into the renal pelvis through a ureteroscope. Haematuria resolved and did not recur during 16 months follow up. It is difficult to draw any meaningful conclusion from this single case report about the value of silver nitrate in managing this condition as in a subsequent report this treatment was found to be of no benefit.

**Summary**

From the number of cases reported in the world literature it is obvious that this condition is not very common. Its classical manifestations include loin pain, unilateral micro or macroscopic haematuria with pelvic or genital varices. Its diagnosis is based on history and physical examination, basic lab tests to exclude other causes of haematuria, cystoscopy and ureteroscopy to confirm unilateral haematuria and exclude other causes of this sinister symptom. Sequence of imaging has more or less been rationalised to USS with Doppler studies, CT or MR angiography and finally phlebography with renal vein and IVC manometry to confirm the diagnosis.

Surveillance is appropriate in cases of mild symptoms and either microscopic or insignificant macroscopic haematuria with no evidence of significant blood loss. Open surgical interventions although effective are associated with higher surgical morbidity. The alternative option of intra-vascular or laparoscopic application of extravascular stents is very appealing because of the minimal invasive nature of the procedures. If the long term outcomes remain as good as the short term results reported so far then stenting (extra-vascular/intra-vascular) may become the treatment of choice in this syndrome.

**References**


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