New Developments In The Treatment Of The Nutcracker Syndromes – Arterial (Duodenal Compression) And Venous (Renal Vein Compression)

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Disclosures
• I have nothing to disclose

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Nutcracker
Defined as a compound lever - - a rigid lever hinged and pivoted about a fulcrum.

The Nutcracker Anatomy
Nutcracker syndrome (NCS) is characterized by impeded outflow from the left renal vein (LRV) into the inferior vena cava due to extrinsic compression from the SMA.

Clinical Diagnosis of Nutcracker Syndrome (NCS)
• A high index of suspicion
• Diagnosis of exclusion.
• Caused by anatomic entrapment of the left renal vein between the aorta and SMA leading to left renal vein hypertension and pelvic venous engorgement.

Symptoms and signs at the time of presentation include:
• abdominal and left flank pain (43.4%-65.2%).
• macroscopic hematuria (30.1%-49.5%).
• microscopic hematuria (6.4%-21.5%).
• proteinuria (4.3%-26.1%).
• varicocele (8.7%-21.7%).

Pelvic Congestion
Incidence

- Prevalence of NCS is unknown,
- absence of definitive diagnostic criteria
- variability in symptomatic presentation.
- Patients can present at any age from childhood to the seventh decade.
- Peak prevalence in young (second or third decade).

Literature

Rare Condition.
Studies analyzed including two or more cases over 35 years with symptoms, diagnosis, management and follow-up.
Only 18 studies (284 patients) met the criteria for evaluation.

Investigations

- Duplex Imaging
  - Duplex Ultrasound is recommended as a first-line study for identifying LRV compression. It is easier to get such values in a semi-sitting position.
  - US has a sensitivity 69% to 90%.
  - US has a specificity of 89% to 100%, respectively.
  - PSVR of the aortomesenteric segment to the hilar portion of >4.2 to 5.0 is considered one of the diagnostic criteria of NS.

- Intravascular Ultrasound-IVUS
  - Intravascular ultrasound has a higher specificity of 90% compared with 62% with venography and may provide a more accurate representation of vessel compression and diameter than digital subtraction angiography.
  - IVUS Left Renal Vein Pullback (important for stent sizing)
Venography enables the measurement of the venous pressure gradient between the LRV and the IVC.

In healthy patients the renocaval pullback mean gradient was 0-1 mmHg

>3 mmHg gradient with venography to diagnose NCS


CT AND MRI

CT and MRI can demonstrate:
- Compression of the LRV,
- Measure diameters,
- Gonadal vein distension,
- Pelvic congestion,
- Beak angle,
- Aortomesenteric angle
  - An acute angle of <39 degrees
- Duodenal compression.

CT/MRI delineates soft tissue anatomy in the region of the compression and other anatomic anomalies.

CT Criteria for NCS

High sensitivity and specificity

CT Criteria for NCS

Management Options

Conservative Therapy

- Patients with mild to moderate symptoms.
- Patients younger than 18 years.

Nutritional support.

- Weight gain increases the retroperitoneal adipose tissue, leading to a reduction of tension on the LRV and further increase in the aortomesenteric angle.

- this approach has been shown to resolve symptoms of NS in 30% of patients.

CT Criteria for NCS

Table: CT diagnostic criteria for nutcracker phenomenon/nutcracker syndrome.

<table>
<thead>
<tr>
<th>CT Criteria</th>
<th>Sensitivity</th>
<th>Specificity</th>
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<tbody>
<tr>
<td>1. Narrowing of the LRV at the aortomesenteric portion (beak angle &lt;32°)</td>
<td>66.7% (8/12) [95.8% (10/12)</td>
<td>100% (9/9) [100% (9/9)]</td>
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<tr>
<td>2. Beak sign: acute form of narrowing of the LRV at the aortomesenteric portion</td>
<td>100% (5/5) [94.4% (4/5)]</td>
<td>94.4% (4/5) [94.4% (4/5)]</td>
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<tr>
<td>3. Left renal venous ratio (LRV to aortomesenteric) ≤ 1.5</td>
<td>100% (6/6) [100% (6/6)</td>
<td>100% (6/6) [100% (6/6)]</td>
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<td>4. Angle between the SMA and aorta ≤ 4°</td>
<td>100% (5/5) [94.4% (4/5)]</td>
<td>94.4% (4/5) [94.4% (4/5)]</td>
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<tr>
<td>5. Collateral venous circulation developed in the retroperitoneum and renal hilum</td>
<td>100% (5/5) [94.4% (4/5)]</td>
<td>94.4% (4/5) [94.4% (4/5)]</td>
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Management Options

- Non-operative Approach
- Open Surgical Approach
- Endovascular Approach
- Extravesical Sham Operation
- SMA Transposition

SMA Transposition


Endovascular Intervention

- Avoids renal ischemia and laparotomy.
- Specific designed venous stent.
- Limited number of patients and short follow-up.
- Difficult vein sizing.
- IVUS important for these endovascular cases.
- Cases of stent migration to heart reported as high as 6.6% in several series.

SMA SYNDROME

CT abdomen (with oral contrast) and barium study

Aetiological factors associated with SMA Syndrome

- Abdominal surgery
- Loss of mesenteric fat
- Postinflammatory
- Trauma
- Local pathology

- Abnormal short length of bowel
- Superior mesenteric artery with intestinal laxity
- Eating disorders
- Low-feeding-dependent patients who are underfed
- Excessive exercise e.g., military training
- Gastrointestinal surgery
- Diseases of malabsorption
- External compression (torsal syndrome)
- Spinal correction surgery
- Abdominal trauma
- Spinal cord injury
- Malperfusion
- Aortic aneurysm
- Chronic inflammation and adhesions
Intestinal Bypass Surgery
Duodenojejunostomy

Discussion
• Suspected with Clinical Features (hematuria, varicocele and left flank/abdominal pain) and Anatomic entrapment of the LRV between the aorta and SMA.
• Open Surgical and Endovascular approach offers effective therapy to alleviate symptoms.
• Results of LRV Stenting is encouraging but Durability is yet unproven.

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