Diagnosis & Management of The Nutcracker Syndrome

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DISCLOSURES

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• No relevant financial relationship reported
**Nutcracker Syndrome?**

- Nutcracker syndrome (NCS): rare clinical entity characterized by obstructed outflow from the left renal vein (LRV) into the inferior vena cava (IVC) due to extrinsic compression of the LRV.
Pathophysiology of NCS:

Pathologic compression of LRV:

- Venous hypertension
- Venous congestion of left flank and pelvis (pain)
- Diffusion of RBCs and proteins into glomerular filtrate (hematuria and orthostatic proteinuria)
There are 3 types of NCS

1. Anterior
2. Posterior
3. Anomalous
Types of NCS:

- Anterior NCS (most common type):

The LRV is compressed as it passes between the aorta and the SMA.
Types of NCS:

- Posterior NCS:

The LRV is retro-aortic and is compressed between the abdominal aorta and the vertebral column.
Types of NCS:

- **Atypical** (associated with truncular vascular malformation (left IVC))

The LRV and IVC are compressed as they pass between the aorta and the SMA.
Symptoms:

- Most common symptoms:
  - Left flank pain.
  - Either gross or microscopic hematuria (required).

- Symptoms related to PCS
  - Dyspareunia.
  - Dysuria.
  - Dysmenorrhea.
  - Vulvar and scrotal varices.
  - Gluteal and lower extremity varicosities.

- Abdominal pain and gastrointestinal symptoms have been reported in a minority of NCS patients.
Diagnostic Imaging:

- Clinical suspicion of the syndrome is confirmed by:
  - Duplex ultrasonography
  - Computerized tomography (CT)
  - Magnetic resonance imaging (MRI)
  - Contrast arterio/venography with or w/o pressure measurements (injection into artery, late phase venous filling), observe flow patterns, collaterals, reflux
**Duplex Ultrasonography:**

- Often the first diagnostic study if NCS is suspected from the history
  - Sensitivity and specificity: 78% and 100%, respectively
CT Scan (± 3D Reconstruction)
MRI:
Common CT and MRI findings:

- The LRV narrowing at the aorto-mesenteric portion.
- “Beak sign”: triangular shape of narrowing of the LRV at the aorto-mesenteric portion.
  - Sensitivity = 91.7%; Specificity = 88.9% for NCS.3

Contrast Venography with Pressure Measurements

- Considered as the gold standard for NCS diagnosis.
- Measurement of the LRV pressure gradient (across the SMA) can be helpful in diagnosis:
  - Range: 2 mmHg - 14 mmHg.
Important Venography Findings:

- Narrowing of LRV as it crosses the aorta underneath the SMA.
- Dilatation of the distal left renal vein, REFLUX into and opacifications of gonadal, ascending lumbar, adrenal and/or other collateral veins.
Indications for Treatment:

- Radiographic compression – not solely
- Collaterals - required
- Hematuria – required
- Pain in flank or pelvis – required

If you don’t choose your patients carefully, results will be poor
Treatment:

- All treatment options are directed to reduce venous hypertension
- Treatment modalities …still evolving...
  - Conservative
  - Surgical
  - Endovascular
  - Hybrid
Open Surgery

1. LRV transposition.
2. LRV transposition with patch venoplasty.
3. Patch venoplasty without LRV transposition.
4. LRV transposition with saphenous vein cuff.
5. Gonadal vein transposition.
6. Saphenous vein bypass.
7. Transposition of SMA
LRV Transposition:

- The LRV is transected from the IVC and the orifice is sutured followed by caudal transposition of the LRV

LRV Transposition w/Patch Venoplasty:

- Indicated if prolonged compression of the LRV results in permanent fibrotic distortion of the LRV
LRV Transposition w/Patch Venoplasty:

- The GSV can be used as a patch to augment the LRV–IVC confluence after transposition of the LRV
The GSV is used to form a cuff extension to the LRV to create tension-free anastomosis.
Gonadal Vein Transposition Technique:

- MINI laparotomy
- The gonadal vein is transected and reimplanted into the IVC to:
  - Decrease pelvic congestion
  - Decompress the LRV \textit{without putting the renal vein at risk}
Gonadal Vein Transposition:

- Isolation of IVC and Gonadal Vein
- Mobilization of Gonadal Vein
- Anastomosis
- Clamps Released
GSV Bypass:

- When transposition of LRV or gonadal not feasible
- GSV bypass used to bypass the compressed segment of the LRV without the need for transection and transposition of the LRV.
Endovascular Treatment:

A) CT scan shows LRV compression
B) Baseline left renal venogram
C) Stenting
D) Completion venogram
E) One year f/u CT scan
Endovascular Treatment:

- Complications:
  - In-stent restenosis
  - Stent migration.
  - Embolization to the lung.
  - Stent kinking
Comparing Treatment Outcomes for NCS: Retrospective Case Series Review

Renal vein transposition had excellent clinical results, but high incidence of bleeding complications.

Comparing Treatment Outcomes for NCS: Retrospective Case Series Review

<table>
<thead>
<tr>
<th>Author</th>
<th>No</th>
<th>M/F</th>
<th>Age (y)</th>
<th>Procedure</th>
<th>Outcome</th>
<th>Complication</th>
<th>Follow up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wang, X et al</td>
<td>30</td>
<td>28/2</td>
<td>18.2</td>
<td>Self-expanding SMART stent</td>
<td>jGradient (from 8.9 to 2.1) mmHg</td>
<td>Persistent hematuria (2), Stent migration (2)</td>
<td>12–80</td>
</tr>
<tr>
<td>Chen S, et al</td>
<td>61</td>
<td>16/45</td>
<td>26</td>
<td>1 Pasmaz, 15 Wallstents, 45</td>
<td>Symptoms resolved (61 patients in 6 months)</td>
<td>Persistent hematuria (2), Stent migration (1)</td>
<td>66</td>
</tr>
<tr>
<td>Reed N, et al</td>
<td>23</td>
<td>10/13</td>
<td>22</td>
<td>SmartControl</td>
<td>Resolution of flank pain and hematuria</td>
<td>Varicocele (2/3 patients)</td>
<td>39</td>
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<tr>
<td>Wang L, et al</td>
<td>23</td>
<td>9/14</td>
<td>28.8</td>
<td>11 patients underwent LRV transposition</td>
<td>Hematuria and proteinuria resolved</td>
<td>Paralytic ileus (2), retroperitoneal hemathoma (1), persistent pelvic pain (1)</td>
<td>14–122</td>
</tr>
</tbody>
</table>

M/F = male, female, LRV = left renal vein, SMA = superior mesenteric artery.

None of these studies evaluated GVT specifically.
Closer Look at Stent Migration:

In 2016 Wu et al. retrospectively evaluated 75 NCS patients treated with endovascular stenting:

49 males | 26 females
Age range: 16 – 43 years [mean 27]
Follow up: 6 -126 months [mean 55]
Stent migration 6.7%
Authors reported serious complications associated with stenting including:

- migration into the right ventricle (required open heart surgery w/tricuspid valve replacement)
- migration into the right atrium (required open heart surgery)
- migration into the IVC (required surgical stent extraction followed by IVC – LRV bypass)
## Treatment Outcomes: Duke Experience

<table>
<thead>
<tr>
<th>Patient</th>
<th>Technique</th>
<th>ASA</th>
<th>OR time (min)</th>
<th>Intra-op urine output (mL)</th>
<th>Estimated blood loss (mL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patient 1</td>
<td>Autotransplantation</td>
<td>3</td>
<td>318</td>
<td>1540</td>
<td>800</td>
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<tr>
<td>Patient 2</td>
<td>LRV transposition</td>
<td>2</td>
<td>122</td>
<td>560</td>
<td>100</td>
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<tr>
<td>Patient 3</td>
<td>R GVT</td>
<td>2</td>
<td>95</td>
<td>275</td>
<td>100</td>
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<tr>
<td>Patient 4</td>
<td>L GVT</td>
<td>2</td>
<td>113</td>
<td>200</td>
<td>50</td>
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<tr>
<td>Patient 5</td>
<td>L GVT</td>
<td>3</td>
<td>141</td>
<td>200</td>
<td>300</td>
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<tr>
<td>Patient 6</td>
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<td>85</td>
<td>150</td>
<td>50</td>
</tr>
<tr>
<td>Patient 7</td>
<td>L GVT</td>
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<td>80</td>
<td>200</td>
<td>100</td>
</tr>
<tr>
<td>Patient 8</td>
<td>Endovascular</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
<td>N/A</td>
</tr>
</tbody>
</table>
Treatment Outcomes: Duke Experience

- Gonadal vein transposition group:
  - OR time, mean: 107 min vs 220 min (non-GVT)
  - EBL, mean: 125 mL vs 450 mL (non-GVT)
  - No ICU stay
The benefits of GVT over other options

Gonadal vein transposition
- Is as effective as other surgical modalities in the treatment of NCS
- Does not require an incision in the leg
- Does not put left renal vein at risk
- Is durable for the life of the patient
How about in patients with complex anatomy?
27 yo female with:

- Intermittent hematuria
- Left flank pain
- Desire to join ATF
In this case LRV transposition and endovascular therapy are not an option.
Patient was treated with RIGHT GVT, remains symptom free at 24 mos, and has joined the ATF.
Posterior NCS

Compression of the LRV between the aorta and spine

Renal vein transposition much more difficult
Posterior NCS

Patient successfully treated with left GVT transposition, symptom free
Summary:

- Management of NCS continues to evolve
- Only patients who meet strict criteria for NCS should be treated
- Surgical treatment continues to be the treatment of choice and should be tailored to the individual patient’s vascular anatomy
- Improvements in technology for venous stents may change paradigm
Summary:

- GVT can be performed safely, and with excellent clinical outcomes
- It offers the benefit of shorter operative times, decreased blood loss, avoids putting the renal vein in harms way, eliminates need for vein harvest, and may be the only option for PCS and NCS associated with left sided IVC
Thank You!