You, Your VHL, and Your Urologist

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National Cancer Institute
Urologist = Genito-urinary Surgeon

General
- Kidney Stones
- Erection issues
- Infertility
- Leaking Urine
- Cancer
- Prostate Issues
- Robotic Surgery
- Pediatric Urology

VHL Specific
- Kidney Cancer
- Pheochromocytoma
- Testicular Masses
VHL Related Cancers

Renal Cell Carcinoma
- Lifelong risk of hundreds of tumors
- Both kidneys are affected
- Cancers can spread when large
- Surgery affects kidney function

Pheochromocytoma
- Cancer of the adrenal gland
- Both adrenal glands can be affected
- “epinephrine secreting tumors”
- Can be life threatening at any size
von Hippel-Lindau (VHL)
Multiple Clear Cell Renal Carcinomas

Multiple Renal Cysts Containing RCC

Clear Cell RCC
J Urol  153:1995
VHL-Associated RCC
High Persistence Rate

Microscopic Tumors

- **VHL**
  - 600 clear cell RCC/kidney
  - 1300 cysts/kidney

*J Urol* 154:1995
Not all tumors are created equally: impact of histology

<table>
<thead>
<tr>
<th>Type</th>
<th>Clear Cell 75%</th>
<th>Papillary Type 1 5%</th>
<th>Papillary Type 2 10%</th>
<th>Chromophobe 5%</th>
<th>Oncocytoma 5%</th>
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</thead>
<tbody>
<tr>
<td>Gene</td>
<td>VHL</td>
<td>Met</td>
<td>FH</td>
<td>BHD</td>
<td></td>
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</tbody>
</table>

[Image of histology types]
Summary of Staging

Stage I (5-year survival: 96%)\textsuperscript{6}
Tumor $\leq 7$ cm in greatest dimension
and limited to kidney.\textsuperscript{4,5}

Stage II (5-year survival: 82%)\textsuperscript{6}
Tumor $>$ 7 cm in greatest dimension
and limited to kidney.\textsuperscript{4,5}

Stage III (5-year survival: 64%)\textsuperscript{6}
Tumor in major veins, adrenal
gland, or perinephric tissue (not
beyond Gerota’s fascia) and/or 1
regional lymph node involved.\textsuperscript{4,5}

Stage IV (5-year survival: 23%)\textsuperscript{6}
Tumor beyond Gerota’s fascia, $>$1
regional lymph node involved,
and/or $\geq$1 distant metastasis.\textsuperscript{4,5}
Fundamentals of VHL Renal Mass Management

- Early and routine surveillance with MRI
  - MRI is better than CT which is better than ultrasound
- Intervene when tumor approaches 3 centimeters
  - Minimal risk of metastasis under 3-4cm
- Maximize cancer control
- Maximize kidney function
  - Remove all tumors and cysts
- Need experienced urologic oncologist
  - VHL cancers are treated differently than sporadic
- Partial adrenalectomy whenever possible
  - Treat pheochromocytoma when less than 3cm
Understanding VHL Kidney Masses
Central Role of Imaging

Simple Cyst → Benign

Indeterminate Cyst → Look for solid component

Solid Mass → Malignant
• MRI is the best test for kidney detail
• Differentiates solids from cysts
• Gives excellent information even without contrast
• Can give contrast with lower kidney function
• No radiation!

• Cons: claustrophobia, metal implants
Management Options

- **Radical Nephrectomy**
  - Open, Laparoscopic, Robotic Approaches
  - Loss of functioning nephrons

- **Partial Nephrectomy**
  - Open, Laparoscopic, Robotic Approaches
  - Maximize renal function
  - Higher surgical morbidity (bleeding, urine leak)
  - Technically challenging

- **Ablation (cryo and radiofrequency)**
  - Percutaneous, Laparoscopic
  - No 10 year outcomes, hard to surveil after ablation

- **Active Surveillance – 3 Centimeter Rule**
Partial Nephrectomy

Open partial nephrectomy

Laparoscopic Partial Nephrectomy

Robotic/Laparoscopic Partial Nephrectomy
Utilization of Partial Nephrectomy

Renal masses 2-4cm

Graph showing percentage cases of total nephrectomy and partial nephrectomy from 1988-1989 to 2000-2001.
The Age of Robotics
Robotic Partial Nephrectomy: Where we started

Indications

- Normal Contralateral Kidney
- Tumors < 4cm
- Exophytic Tumors
- Polar Tumors
Robotic Partial Nephrectomy: Where we are now....

- T1 and T2 tumors
- Endophytic tumors
- Cystic tumors
- Hilar tumors
- Posterior tumors
- Multifocal / Bilateral tumors
- Solitary kidney
- Morbidly Obese
- Caval Thrombus
Robotic Partial Nephrectomy

**Goals:**
- Negative margins
- Minimize warm ischemia time
- Minimize parenchymal loss
- Maximize renal function
- Minimize complications

**Challenges:**
- Margins?
- Warm Ischemia time?
- Minimize parenchymal loss?
- Patient selection?
- Generalizability?
Tumor excision along the natural plane between the peritumor pseudocapsule and the renal parenchyma without removing a visible rim of normal parenchyma.
Definition of Pseudocapsule

An acquired rim of fibrous tissue adjacent to the tumor measuring 0.1mm-1mm in thickness next to a thin 1-2mm rim of compressed renal parenchyma.
## Comparison of Surgical Technique

<table>
<thead>
<tr>
<th></th>
<th>On Clamp Partial Nx</th>
<th>On Clamp Enucleation</th>
<th>Off Clamp Enucleation</th>
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<tbody>
<tr>
<td>Margin</td>
<td>Variable</td>
<td>Pseudocapsule</td>
<td>Pseudocapsule</td>
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<tr>
<td>Ischemia</td>
<td>+++</td>
<td>+++ to ++</td>
<td>None</td>
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<tr>
<td>Renorrhaphy</td>
<td>Multilayer</td>
<td>Minimal to None</td>
<td>None</td>
</tr>
<tr>
<td>Oncologic Outcome</td>
<td>Excellent</td>
<td>Excellent</td>
<td>Excellent</td>
</tr>
<tr>
<td>Preserved Parenchyma</td>
<td>Good</td>
<td>Maximal</td>
<td>Maximal</td>
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Robotic Retro ON Clamp Enucleo-resection
Case 1

3cm solid mass and a cyst

- Partial nephrectomy
  - Remove solid and cyst by enucleation
  - Robotic surgery if possible
Case 2

Bilateral solid masses

- **Left Partial nephrectomy**
  - Remove solid mass by enucleation
  - Robotic surgery if possible

- **Watch right side**
  - Intervene when approaching 3 cm

1.5cm solid

6cm solid
Take Home Points

- **Establish relationship with Urologic Oncologist**
  - Experience matters, get a second opinion always
  - Surgeons motto: Do the right thing!

- **Get Screened and undergo surveillance**
  - Use MRI whenever possible

- **Intervene when the tumor approaches 3 -4 cm**
  - Remove all solids and cysts at same time
  - If minimally invasive, great. If not don’t compromise
  - Nephrectomy is always a last option

- **Enucleation is the best approach to VHL masses**
  - Saves kidney function, doesn’t compromise cancer control
Thank you

We also treat the human spirit.®

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