Overview of Imaging Modalities in the Management of VHL Lifecycle

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Disclosures-VHL Alliance meeting

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- and -

I will not discuss off label use and/or investigational use in my presentation.

Current recommendations, image-based screening:

<table>
<thead>
<tr>
<th></th>
<th>Pediatric</th>
<th>Adults</th>
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<tbody>
<tr>
<td>Retina</td>
<td>Annual visualization</td>
<td>Annual visualization</td>
</tr>
<tr>
<td>Brain/cerebellum (Internal auditory canal)</td>
<td>If symptomatic (if recurrent ear inf.)</td>
<td>MRI every 2-3 years or more as needed</td>
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<tr>
<td>CTLS Spine</td>
<td>As above</td>
<td>As above</td>
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<tr>
<td>Adrenal/Paragangioma</td>
<td>MRI/MIBG if blood/urine test +</td>
<td>MRI/MIBG if blood/urine test +</td>
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<tr>
<td>Kidney</td>
<td>Annual ultrasound (MRI if findings)</td>
<td>Annual US or MRI, MRI at least q2y</td>
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<tr>
<td>Pancreas</td>
<td>No recommendation</td>
<td>Annual US or MRI, MRI at least q2y</td>
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</tbody>
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Retinal imaging
Imaging Studies for RCH

- Fundus photography (wide angle)
- Fluorescein angiography
- Fundus autofluorescence
- Ultrasound
- Optical coherence tomography (OCT)

What is the ophthalmologist looking at?

Regular vs. Widefield Retinal Photography (both images on same scale)

VHL Associated Retinal Capillary Hemangio-blastoma (RCH)
A.K.A. Retinal Angioma

Fluorescein Angiography

Early filling Late
Retinal ultrasound

Optical Coherence Tomography

CNS Imaging

MRI imaging of cerebellar hemangioblastoma
Internal Auditory Canal MRI

MRI imaging of spinal hemangio-blastoma

Needs to be a: “total spine” -or- CTLS spine

Body Imaging

Why MRI over CT?

- Airplane flight at 35,000 ft = 0.003 mSv/hr
- 10 hr flight to Japan = CXR

Chernobyl: Currently 0.001 mSv/hr

*1 day at Chernobyl today = CXR

*New low dose procedures can reduce exposure 50%
CT—when do we get it?

- Planning for some procedures
- Need a scan fast
- Monitoring therapy

US or MRI? Quality vs Value

MRI Imaging for pheochromocytoma

$^{131}$I or $^{123}$I MIBG scan

Pheo or paraganglioma
Imaging pancreatic lesions

Summary

• Ophthalmology imaging of the retina: Direct visualization, recording of findings over time. Fluorescent imaging, US, Tomography, reveals defects that can be harder to see on photography, or characterize lesions.
• Imaging of the brain and spine is with MRI, and includes specialty scans to image specific regions.
• Imaging of the abdomen for kidney, adrenal, or pancreas tumors:
  • US is easy, non-invasive, fast, cheap, but has low sensitivity
  • CT is easier to read, relatively inexpensive, easy to get, but exposes to cumulative lifetime exposure to radiation risk.
  • MRI is the highest quality image, hard to read, hard to schedule, expensive, but high sensitivity and specificity.
• Specialty scans for endocrine cells can identify hormonally active lesions.

Thank you!