

VHL The Moffitt Cancer Center Experience

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When you think of VHL you must think about ten possible locations

• Initial Evaluation

- Making or confirming the diagnosis
 - Family history
 - Prior consultations
 - Laboratory Studies
 - Radiologic studies
 - Physical findings
 - Symptoms



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Initial Evaluation

- Eye/retinal examination
- MRI scan of abdomen
- Physical examination
- Laboratory tests for metanephrines
- MRI scan of brain and spine



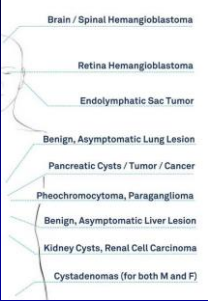
Initial Consultation

- Complete workup by our multi-discipline team
- History and family tree
- Genetic counseling and testing to confirm diagnosis
 - Insurance issues
 - Impact on other health related issues and
 - Impact on family members
- GU evaluation with a physical exam and imaging studies
- GI evaluation with a physical exam and imaging studies
- Neuro-surgical assessment with brain and spinal cord examination and radiologic studies
- Endocrine evaluation including history and physical exam with laboratory studies and imaging studies
- Ophthalmology evaluation with a through exam

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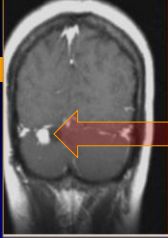
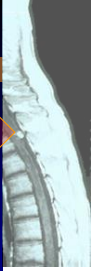
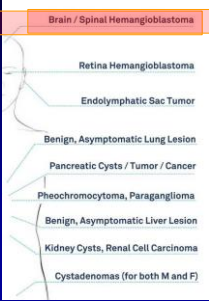
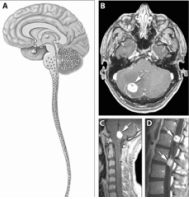
Making the diagnosis

- If you have a family history
 - you need only a single VHL associated tumor.

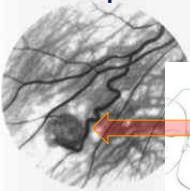
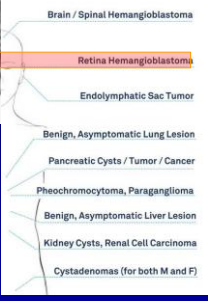


- Without a family history you need at least two VHL associated tumors to make a clinical diagnosis or:
 - One hemangioblastoma
 - PLUS
 - Visceral tumors

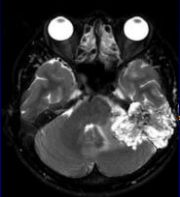
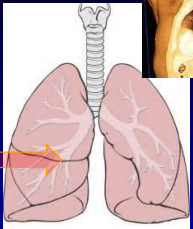
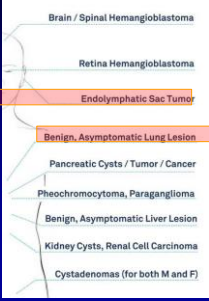

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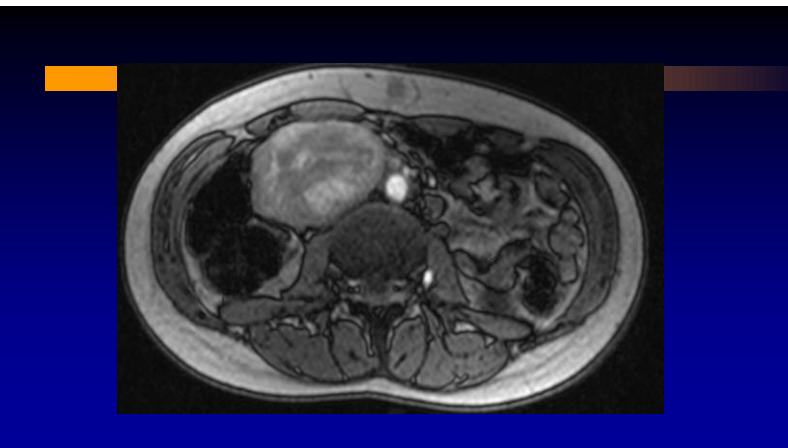







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Anatomical diagrams showing the pancreas (HEAD, BODY, TAIL, Lobules, Pancreatic duct, Common bile duct, Accessory pancreatic duct, Duodenal papilla, Duodenum) and the spine. A CT scan of the abdomen shows a lesion in the adrenal gland, indicated by a purple arrow.

- Brain / Spinal Hemangioblastoma
- Retina Hemangioblastoma
- Endolymphatic Sac Tumor
- Benign, Asymptomatic Lung Lesion
- Pancreatic Cysts / Tumor / Cancer
- Pheochromocytoma, Paraganglioma
- Benign, Asymptomatic Liver Lesion
- Kidney Cysts, Renal Cell Carcinoma
- Cystadenomas (for both M and F)



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A photograph of a pheochromocytoma/paraganglioma specimen, which is a reddish, lobulated mass. A purple arrow points from this image to the corresponding item in the list below.

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When you think of VHL you must think about ten possible locations

Adrenal gland

Tumor in adrenal gland

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- Retina Hemangioblastoma
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	Average age of onset (age range of diagnosis), Years	Frequency in Patients, %
Central Nervous System		
Retinal Hemangioblastomas	25 (0-68)	25-60
Endolymphatic sac tumors	22 (12-50)	10-25
Cerebellar hemangioblastomas	33 (9-78)	44-72
Brain stem hemangioblastomas	32 (12-46)	10-25
Spinal cord hemangioblastomas	33 (11-66)	13-50
Visceral		
Renal cell carcinoma or cysts	39 (13-70)	25-75
Pheochromocytomas <small>*Excludes the 20% of these tumors that occur outside the adrenal gland, i.e. paragangliomas. Frequency depends on VHL subtype.</small>	27 (5-58)	10-25
Pancreatic Neuroendocrine Tumor (PNET)	36 (5-70)	11-17
Pancreatic cyst	38 (5-70)	<=75
Epididymal cystadenomas	Unknown (17-43)	25-60 (of males)
APMO or broad ligament cystadenomas	Unknown (16-46)	10 (estimated, of males)

Screening

- Eye/retinal examination **Annually**
- Ultrasound alternating with MRI scan of abdomen **Alternating every other year**
- Physical examination **Annually**
- Laboratory tests for metanephrines **Annually**
- MRI scan of brain and spine **Every other year**

Adrenal gland

Tumor in adrenal gland

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The Moffitt Cancer Center VHL clinics

- International Plaza
 - Central location
 - Easy access and parking
 - Full laboratory and radiologic services
 - Devoted clinical space for our VHL patients

**This is what we do at the Moffitt Cancer Center
VHL Clinic**

Summary

- Thorough and extensive initial evaluation
- Initial diagnostic criteria is applied utilizing historic, radiologic, physical and genetic data
- Baseline and surveillance monitoring
- Multidisciplinary approach to care
- Specialists trained in the nuances of VHL

Total care of our VHL patients in a supportive well coordinated manner