

Tumors associated with Von Hippel-Lindau disease

Cerebellar hemangioblastoma

see [Cerebellar hemangioblastoma associated with Von Hippel-Lindau disease](#).

Spinal cord hemangioblastomas

see [Spinal cord hemangioblastoma](#).

Brainstem hemangioblastomas

see [Brainstem hemangioblastoma](#).

Pheochromocytomas

see [Pheochromocytoma](#)

Endolymphatic sac tumors

see [Endolymphatic sac tumor](#).

Retinal hemangioblastomas

see [Retinal hemangioblastoma](#).

Renal cell carcinoma

see [Renal cell carcinoma associated with Von Hippel-Lindau disease](#).

Renal cysts

- a) 50–70% of [Von Hippel-Lindau disease](#) patients have bilateral and multiple renal cysts
- b) rarely cause profound renal impairment
- c) chronic renal failure or renal hypertension not as common as with polycystic kidney disease

Epididymal cystadenomas

- a) benign lesions that arise from the epididymal duct
- b) found in 10–60% of male VHL patients
- c) typically appear in the teenage years
- d) may cause infertility if bilateral

e) may be multiple

Broad ligament cystadenomas

a) arise from the embryonic mesonephric duct

b) true incidence unknown

c) rarely reported and usually not recognized in women with VHL

Pancreatic neuroendocrine tumors and cysts

a) 35 to 70% of patients with VHL develop an endocrine tumor or cyst

b) pancreatic cysts are generally asymptomatic and often multiple

c) pancreatic neuroendocrine tumors are usually non-functional and 8% of them are malignant

d) differential diagnosis: pancreatic islet cell tumors, MEN2

Sinonasal renal cell-like adenocarcinoma

[Sinonasal renal cell-like adenocarcinoma](#) is an emerging tumor associated with VHL syndrome and it is hoped that future studies shed light on the underlying biology of this unique tumor ¹⁾.

¹⁾

Maharaj S, Seegobin K, Wakeman K, Chang S, Potts K, Williams B, Redman R. Sinonasal renal cell-like adenocarcinoma arising in von Hippel Lindau (VHL) syndrome. *Oral Oncol.* 2022 Jan 5;125:105705. doi: 10.1016/j.oraloncology.2021.105705. Epub ahead of print. PMID: 34998175.

From: <https://operativeneurosurgery.com/> - **Operative Neurosurgery**

Permanent link: https://operativeneurosurgery.com/doku.php?id=tumors_associated_with_von_hippel-lindau_disease

Last update: **2022/01/09 14:33**

