

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.

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