

The VHL gene provides instructions for making a [protein](#) that functions as part of a complex (a group of proteins that work together) called the VCB-CUL2 complex. This complex targets other proteins to be broken down (degraded) by the cell when they are no longer needed.

[Hemangioblastomas](#) may arise sporadically in a solitary form or associated with [Von Hippel-Lindau disease](#) with multiple tumors. [VHL protein](#) is defective in both forms of [hemangioblastoma](#), leading to the accumulation of hypoxia-inducible factor, stimulating [angiogenesis](#) via [VEGF](#) and [PDGF](#) mainly ¹⁾.

1)

Migliorini D, Haller S, Merkler D, Pugliesi-Rinaldi A, Koka A, Schaller K, Leemann B, Dietrich PY. Recurrent multiple CNS hemangioblastomas with VHL disease treated with pazopanib: a case report and literature review. *CNS Oncol.* 2015;4(6):387-92. doi: 10.2217/cns.15.22. Epub 2015 Oct 26. PMID: 26497655; PMCID: PMC6083944.

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Last update: **2024/06/07 02:50**

