Von Hippel-Lindau (VHL) is an autosomal dominant hereditary disorder in which a single mutated gene is transmitted to offspring from an affected parent (National Cancer Institute [NCI], 2015). Mutations in the VHL gene predispose individuals to various types of benign and malignant neoplasms in different organs (NCI, 2015). For instance, central nervous system tumors are named hemangioblastomas. Other types of VHL tumors may develop in the kidneys, the pancreas, genital tract, or other organs (NCI, 2015).

Diagnosis, treatment, and evaluation of patients with VHL require a multidisciplinary approach. Specialists and healthcare personnel who are involved in treating patients with VHL may include oncology surgeons, neurosurgeons, general surgeons, ophthalmologists, endocrinologists, neurologists, genetic counselors, nurse practitioners, and nurses (NCI, 2015). VHL is likely to be a burden for patients, families, nurses, and healthcare personnel who are not familiar with its complexity. Therefore, the multiple roles that nurses play become critical as they embrace the need to act as a liaison, educator, and coordinator in managing VHL. Because nurses are on the front line of patient care, they must have a basic understanding of VHL and current evidence regarding its diagnosis and treatment.

The term von Hippel-Lindau was first used in 1936 (Maher, Neumann, & Richard, 2011). VHL is named for two famous European physicians, Eugen von Hippel and Arvid Lindau. The German ophthalmologist Eugen von Hippel first described angiomas in the eye in 1904. Arvid Lindau, a Swedish pathologist, described the angiomas of the cerebellum, kidney, and spine in 1926 (Molino, Sepe, Anastasio, & De Santo, 2006). However, the term VHL was not in common use until the 1970s (Maher et al., 2011).