

Surveillance Guidelines: 2020

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Purpose: von Hippel-Lindau (VHL) affects >10 organ systems, and active surveillance of each organ system has been shown to decrease morbidity and mortality. The majority of previous guidelines have been based on expert opinion, rather than systematic review of the literature, and many are lacking in detailed guidance. Beginning in late 2018, the VHL Alliance spearheaded an effort that assembled an international consortium of 50+ VHL experts to develop new surveillance guidelines for each organ system. This presentation describes the new methodology and approach utilized by the international consortium, and we report the new multisystem screening guidelines for each affected organ and VHL manifestation.

Methods: A separate subcommittee was convened for each organ system, consisting of multiple experts in the relevant manifestation, along with experts in radiology, anesthesia, guideline writing, and patient representatives. These new guidelines are based on systematic review of the literature, rather than only expert opinion as with previous guidelines. Expert consensus was used where only low-level evidence was available. The evidence supporting each guidelines was graded using the method of Shekelle et al. (BMJ, 1999), and the strength of each recommendation was weighted according to the method of the National Comprehensive Cancer Network.

Results: Briefly, dilated eye examinations are recommended to begin within the first year of life, and occur every 6-12 months until age 30, then yearly. A history and physical geared towards assessing signs and symptoms of the various VHL manifestations should occur yearly beginning at age 1, with blood pressure and pulse included from age 2. These continue throughout life. Metanephrines are recommended yearly beginning at age 5, until age 65. Magnetic resonance imaging (MRI) with gadolinium of the entire neuroaxis is recommended every 2 years beginning at age 11, until age 65. Audiograms are recommended every 2 years beginning at age 11, until age 65. A baseline MRI of the internal auditory canal is recommended at some point after age 15 (and ideally before age 21). MRI of the abdomen to assess the kidneys, but also the pancreas and adrenals, is recommended beginning at age 15 at least every 2 years (assuming no positive findings), stopping at age 65. MRI of the neuroaxis and abdomen can be combined into a single session, but separate organ-specific imaging protocols should be employed. All indications to stop at age 65 assume that the particular patient has never shown a tumor in that organ previously. Prior to a planned pregnancy, all systems should be screened, but additional testing is not required once already pregnant. Specific details of the testing, including modalities, how to combine imaging of various systems, interpretation of findings, and algorithms for treatment for specific tumors in specific organ systems, are all included.

Conclusions: The new 2020 VHL surveillance guidelines assembled by an international consortium of >50 experts resulted in detailed, evidence-based guidelines that are different from previous iterations. An effort was also made to harmonize intervals and screening initiation and cessation ages to simplify implementation and compliance for patients, resulting in the "5-11-15-20 rule".