VHL Surveillance Checklist

NeuroEndocrine Cancer Australia

Surveillance means regular monitoring for people who are at risk of developing von Hippel-Lindau (vHL) syndrome related manifestations. Keeping up with routine health checks is important, as it can help detect tumours, cysts, or other health problems early, when treatment is often most effective.

Surveillance schedules may also be adjusted by specialists who are familiar with the individual and their family history. Surveillance frequency might change if symptoms have developed or vHL manifestations have occurred.

For individuals planning a pregnancy, it is recommended that surveillance assessments are completed before conception whenever possible. Ongoing eye monitoring should include a comprehensive dilated eye examination prior to pregnancy, followed by regular reviews every 6–12 months.

Routine laboratory and imaging surveillance may be discontinued for asymptomatic individuals aged 65 years who have had no vHL-related features identified on previous screening. However, annual physical examinations and eye (ophthalmological) assessments should be maintained.

Assessment	Starting age	How often	Monitoring for
Physical examination **(Age appropriate history and physical examination)**	From birth	Every year	VHL related changes and complications
Eye check with an ophthalmologist	Within the first year of life	0 – 30 years: Every 6–12 months 30+ years: Annual for those asymptomatic without active RHs	Retinal haemangioblastomas (RHs)
Vital signs and symptom Assessment **(Pulse, blood pressure and screening for signs or symptoms of catecholamine excess*)**	From 2 years of age	Every year	vHL related changes and complications, especially phaeochromocytomas and paragangliomas (PPGL).
Fasting blood test**	From 5 years of age	Every year	Phaeochromocytoma & paraganglioma (PPGL)
Hearing check (audiometry)	From 11 years of age	Every 2 years	Endolymphatic sac tumours (ELST)
MRI brain and spine	From 11 years of age	Every 2 years	Hemangioblastomas
MRI scan of the ear canal***	Between 15-20 years of age	Once (added to a planned MRI brain/ spine)	Endolymphatic sac tumours (ELST)
MRI abdomen	From 15 years of age	Every 2 years	Kidney cancer pancreatic neuroendocrine tumours and cysts.

See your doctor if you notice any new lumps, pain, numbness, or changes in your vision, hearing, or balance.

*Screening for signs or symptoms of catecholamine excess - headaches, palpitations (fast, pounding heart rate), diaphoresis (excessive sweating), hyperactivity (feeling restless or overly active), anxiety, polyuria (passing urine more often than usual), and abdominal (stomach) pain.

**Annual fasting free plasma metanephrines and if available plasma 3-methoxytyramine (if plasma unavailable 24 hour urine free metanephrines)

***A one-off baseline high-resolution MRI of the internal auditory canal should be performed once between the ages of 15 and 20 years, or at the time of VHL diagnosis if the individual is diagnosed after the age of 20 years.

Reference
Daniels, A. B., A. Tirosh, K. Huntoon, et al. (2023). "Guidelines for surveillance of patients with von Hippel-Lindau disease: Consensus statement of the international VHL surveillance guidelines consortium and VHL alliance." Cancer 129(19): 2927-2940.

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