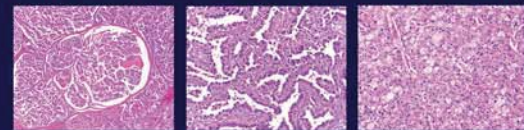


When to think about hereditary RCC

Patients with any renal tumour (benign or malignant) AND any one of the following:

- Bilaterality or multifocality
- Onset of ≤ 50 years of age
- 1st or 2nd degree relative with any renal tumour
- Pneumothorax (BHD/FLCN)
- Dermatologic findings:
 - Skin leiomyomas (HLRCC/FH)
 - Skin fibrofolliculomas/trichodiscomas (BHD/FLCN)
- Associated tumours:
 - Pheochromocytoma/paraganglioma (VHL, SDH)
 - Hemangioblastoma of the retina, brainstem, cerebellum or spinal cord (VHL)
 - Early onset of multiple uterine fibroids (< 30 years of age) (HLRCC/FH)
- Lymphangiomyomatosis (TSC)
- Childhood seizure disorder (TSC)

Patients with non-clear cell RCC with unusual associated features, such as:



papillary type I

papillary type II

chromophobe, oncocytoma, oncocytic hybrid

Patients, with or without RCC, who report a family member with a known clinical or genetic diagnosis of any one of the syndromes in the table below;

Patients with RCC and history of a second cancer suggestive of any one of the syndromes below:

ASSOCIATED CLINICAL FINDINGS	SYNDROME / GENE / (RCC HISTOLOGY)	SUGGESTED SURVEILLANCE IN ADULTS
	Von Hippel-Lindau <i>VHL</i> (ccRCC)	<ul style="list-style-type: none"> - Annual imaging alternating between US and MRI - Annual ophthalmologic examination - Annual 24-hour urine catecholamines/metanephrines, plasma metanephrines - Annual audiometry - MRI of brain and spine every 2y
	Tuberous sclerosis complex <i>TSC1/TSC2</i> (Angiomyolipoma, RCC)	<ul style="list-style-type: none"> - MRI of abdomen every 1-3y - MRI of brain every 1-3y (age ≥ 25 y) \pm EEG - CT of chest for women every 5-10y or symptomatic males - Annual dermatologic examination - Dental exam every 6mo - Annual ophthalmologic examination - ECG every 1-3y
	PTEN hamartoma syndrome/ Cowden syndrome <i>PTEN</i> (various)	All patients: <ul style="list-style-type: none"> - US of abdomen every 1-2y (age ≥ 40y) - Annual US of thyroid - Colonoscopy every 5y (age ≥ 35y), - Dermatologic examination Women: <ul style="list-style-type: none"> - Clinical breast exam every 6-12mo (age ≥ 25y) - Annual mammography and MRI of breast (age ≥ 30y) - Annual random endometrial biopsies and/or transvaginal US (age ≥ 30y)
	Hereditary papillary RCC (HPRCC) <i>MET</i> (Papillary type I)	<ul style="list-style-type: none"> - Annual MRI of abdomen
	Hereditary leiomyomatosis and RCC (HLRCC) <i>FH</i> (Papillary type II)	<ul style="list-style-type: none"> - Annual MRI of abdomen - Dermatologic examinations every 1-2y - Annual gynecologic assessment and transvaginal US
Paraganglioma, pheochromocytoma, GI stromal tumors	SDH-associated kidney cancer <i>SDH</i> (ccRCC, chromophobe, oncocytoma)	<ul style="list-style-type: none"> - Annual MRI of abdomen and MRI of head and neck - Annual 24-hour urine catecholamines/metanephrines, and plasma metanephrines
	Birt-Hogg-Dubé (BHD) <i>FLCN</i> (Oncocytoma, mixed oncocytic, chromophobe)	<ul style="list-style-type: none"> - Annual CT or MRI of abdomen or every 3y in persons without renal lesions - Baseline CT of chest - Annual dermatologic examination
Uveal melanoma, melanoma, mesothelioma	BAP1 Hereditary Cancer Syndrome <i>BAP1</i> (ccRCC)	<ul style="list-style-type: none"> - Not established; consider annual abdominal imaging - Annual dermatologic assessment - Annual ophthalmologic assessment
Multiple cancers: <ul style="list-style-type: none"> - Sarcoma - breast cancer - brain tumours - adrenocortical carcinoma - leukemia, lymphoma 	Li-Fraumeni Syndrome <i>TP53</i> (various)	<ul style="list-style-type: none"> - US of abdomen every 6mo - Annual rapid full body MRI - Annual MRI of brain - CBC, LDH, ESR every 6mo - Annual mammography and MRI of breast - Colonoscopy every 2-5y - Annual dermatologic examinations