HLRCC Foundation

HLRCC: hereditary leiomyomatosis and renal cell cancer

Information for patients and families



What is HLRCC?

HLRCC is short for hereditary leiomyomatosis and renal cell cancer, and HLRCC is the preferred name. It is sometimes referred to as FH tumor predisposition syndrome. [1]

People with HLRCC are born with a harmful change in one copy of their *FH* gene.[1, 10]

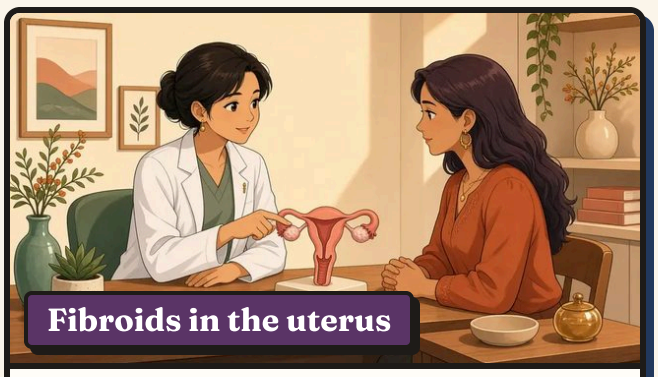
Having HLRCC does not mean you have cancer or that you will get it.



Skin bumps (leiomyomas)

See a skin doctor (dermatologist) every 1 to 2 years for a full skin check.[1]

These skin bumps are non-cancerous (benign).[1, 10]



Fibroids in the uterus

Fibroids are common in women with HLRCC. [1, 2]

Most women with HLRCC-related fibroids have heavy or irregular periods, and many have pelvic pain.[1, 2, 9]

See a gynecologist who knows about HLRCC. Ask how often you should be checked.[1, 2]

Kidney cancer risk



15 to 30

out of every 100

Estimates vary widely. NCI's summary: most estimates 15 to 30 in 100[2]; a 2020 study of large genetic databases suggests lower[6].

Most people with HLRCC do not get kidney cancer.

If a scan finds a tumor, it should be removed by surgery soon, even if it is small.



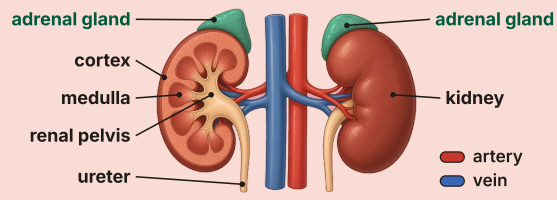
A kidney MRI every year

From age 8, a yearly kidney MRI, for life. Your urologic oncologist or medical oncologist will decide the exact timing.[1, 2]

Ask for a kidney MRI with 1 to 3 mm slices through the kidneys.[1, 4]

An ultrasound alone is never enough to check the kidneys in HLRCC.

Rare: paraganglioma or pheochromocytoma



A few specific *FH* changes raise the chance of a rare tumor in an adrenal gland (pheochromocytoma) or along nerves elsewhere (paraganglioma).[7, 8]

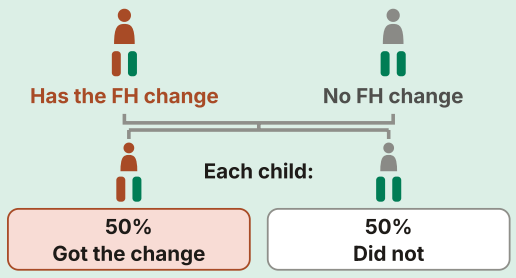
Pheochromocytoma/paraganglioma: rarely occurs in some individuals with an *FH* mutation.[1]



Genetic testing

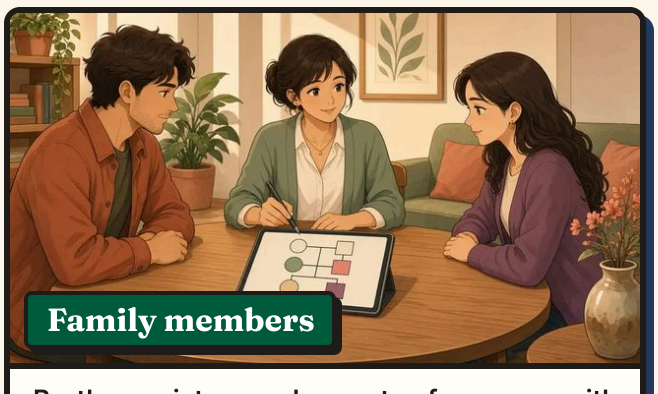
A genetic test on a blood or saliva sample looks for a change in the *FH* gene.

Passing it on



changed *FH* copy working copy

Each child of a parent with HLRCC has a 50% chance of inheriting the *FH* change, like a coin toss.[1]

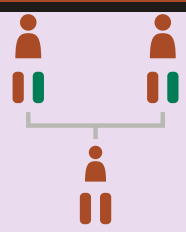


Family members

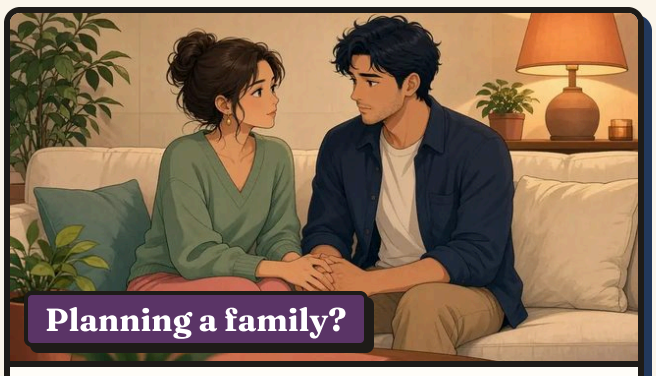
Brothers, sisters and parents of a person with HLRCC may also have the same change.

A genetic counselor can help you decide about testing and explain what results mean.

If both parents carry an FH change



If both parents carry an *FH* change, a baby can be born with two changed copies. This causes fumarase deficiency, a very rare and severe condition that starts in newborns.[2, 11]



Planning a family?

Talk with your gynecologist early about timing.

A genetic counselor can explain your options, including testing during pregnancy or IVF with embryo testing.[1]

Where to get help in the United States

Find an HLRCC specialist

hlrcc.org/hlrcc-map

Ask your doctor for a referral to a genetic counselor

findageneticcounselor.nsgc.org

Learn more and connect with other families

hlrcc.org

For education only; it does not replace advice from your own doctor. Physician review pending (Dean Elhag, MD).

Sources for this infographic

Numbers in brackets on page 1 refer to this list. HLRCC Foundation, hlrcc.org. Sources checked October 30, 2026.

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