

About Genes

Genes are in every cell in our bodies. Genes are made of DNA, which gives instructions to cells about how to grow and work together. We have two copies of each gene in every cell—one from our mother and one from our father. When they work properly, some genes help keep cancer from developing. If one copy of a gene has a mutation, it cannot work as it should. This raises the risk for certain tumors and cancers.

The *BAP1* gene controls cell growth. When the *BAP1* gene is not working right, cells can grow and divide without stopping. Tumors, and sometimes cancer, can form. Having a mutation in the *BAP1* gene can make you more likely than average to get certain cancers and tumors that are not cancer.

The *BAP1* gene was discovered a short time ago. We are still learning about what can happen when a person has a *BAP1* mutation. We do know that having a *BAP1* mutation does not mean you are certain to get a tumor or cancer.

BAP1 Mutations and Cancer Risks

In the general population, about 33 people in 100 will get some form of cancer during their entire lifetime. For people with *BAP1* mutations, about 85 in 100 will get cancer by age 65. This is early in life compared to the general population.

BAP1 mutations also raise the risk to get several rare cancer types. This table compares the lifetime risk for getting these cancers among those with *BAP1* mutations and those without.

Cancer Type	With <i>BAP1</i> Mutation	General Population
Melanoma of the eye	30 in 100	5 in 1 million
Melanoma of the skin	13 in 100	3 in 100
Mesothelioma	27 in 100	1 in 100,000
Kidney cancer	22 in 100	16 in 100,000

Melanoma is a serious type of skin cancer. It can also happen in the eye. It often spreads to other parts of the

body. Melanoma of the eye has happened in teens with *BAP1* mutations.

Mesothelioma is cancer in the lining of the lung, abdomen, and other organs. Exposure to tobacco smoke, asbestos, and arc welding are risk factors for this type of cancer.

Kidney cancer may happen early in life for people with a *BAP1* mutation. It can also grow and spread more quickly than in people without the mutation.

Recommendations

Starting at age 11

- Yearly eye exam with eye drops to make pupils larger

Starting at age 20

- Yearly full-body skin exam by dermatologist

All ages

- Follow sun safety recommendations whenever you are outside. Use SPF30 or higher sunscreens. Wear long sleeves, long pants, and wide-brim hats.
- Avoid smoking. Avoid exposure to asbestos and arc welding.
- Talk with your doctor about yearly MRI or ultrasound screening of the abdomen.

KIDS AND SIBLINGS

Siblings and children of people with a *BAP1* mutation have a 1 in 2 chance of also having the mutation. We recommend genetic testing and counseling for children and young adults, because some *BAP1*-related cancers have happened in teenagers.

People in the family who test negative for the *BAP1* mutation are thought to have the same risk for cancer as the general population, if they have no other risk factors. They should follow general cancer screening guidelines.

Resources

If you have a personal or family history of any of these tumor types, you may be eligible for genetic testing and counseling. If you already know you or a family member has a *BAP1* or other gene mutation, our team of doctors and genetic counselors can help find the screening plan you need.

Call Huntsman Cancer Institute's Family Cancer Assessment Clinic to learn more: 801-587-9555.