

HDGC HEREDITARY DIFFUSE GASTRIC CANCER

Understand your risk. Make informed decisions. Advocate for yourself. Support the research.

WHAT IS HEREDITARY DIFFUSE GASTRIC CANCER (HDGC)?

HDGC is an inherited cancer syndrome that leads to a higher than average risk of developing two types of cancer:

Gastric (stomach) cancer:

- The type of stomach cancer that is associated with HDGC is called diffuse-type stomach cancer. This type of cancer tends to grow and spread through the lining of the stomach, making it difficult to find because it does not cause a tumor to develop in the stomach. Another type of stomach cancer, called intestinal gastric cancer, is not associated with HDGC.
- Doctors sometimes refer to the stomach cancer as *signet ring cell carcinoma* or *linitis plastica*.
- Diffuse gastric cancer is difficult to detect and is usually diagnosed at a late stage when it cannot be cured.

Breast cancer:

- The type of breast cancer that is associated with HDGC is called lobular breast cancer. Another type of breast cancer, called ductal breast cancer, is not associated with HDGC.

**Know your risk.
Know your
family history.**



How does a person get HDGC?

- Many families with HDGC have a mutation in a gene called *CDH1*, a tumor suppressor gene on chromosome 16.
 - This gene mutation was first discovered in a family from New Zealand's Māori population in 1998.
 - Over 150 different mutations in the *CDH1* gene have been identified.
 - *CDH1* gene mutations are passed down from one parent .
 - The inheritance pattern is called autosomal dominant, meaning that you only need one mutated copy to be affected by HDGC.
 - Since everyone has two copies of each gene, each person has a 50% random chance of inheriting the mutated copy from their affected parent. This does not mean that 50% of the family members will inherit it – each person's risk is 50%.
 - Not all families who have HDGC syndrome are found to have a mutation in the *CDH1* gene. In these families, it is not yet known how the cancer risk is passed on.

How do I know if my family has HDGC?

- Learn about the health of your family members, especially any diagnosis of cancer in your family members.
- Tell your doctor about your family health history, and update them at every well visit. If you think that your family might have HDGC syndrome, ask your doctor to send you to a genetic counselor to discuss this and consider genetic testing.
- 1-3% of gastric cancers result from inherited cancer syndromes.
- If you or your family fits into any of these categories, you may benefit from genetic counseling:
 - Two cases of gastric cancer in your family, with at least one confirmed diffuse gastric cancer
 - Diffuse gastric cancer in a family member under age 40
 - Personal or family history of diffuse gastric cancer and lobular breast cancer, one diagnosed under age 50
 - Lobular breast cancer in both breasts
 - Family history of two or more cases of lobular breast cancer under age 50
 - Personal or family history of cleft lip or cleft palate in a patient with diffuse gastric cancer
 - Anyone with signet ring cells on a gastric biopsy
- Genetic testing is performed on a blood or sputum sample to look for a mutation in the *CDH1* gene. Sometimes, a mutation in the *CDH1* gene is found when someone has panel testing, which is when multiple genes are tested at once.

For more information about HDGC visit: HereditaryDiffuseGastricCancer.org

What does having HDGC syndrome mean?

- Men with a *CDH1* gene mutation have up to a 70% risk of having gastric cancer in their lifetime.
- Women with a *CDH1* gene mutation have up to a 56% risk of having gastric cancer and up to a 42% of having breast cancer in their lifetime.
- These cancers tend to develop at a much younger age than most people would develop cancer – sometimes as early as the teens or twenties. The average age that someone with HDGC develops gastric cancer is 38.
- Having the HDGC syndrome does not necessarily mean that a person has cancer – they may just have a higher risk.
 - Not everyone with HDGC syndrome will develop cancer.
 - If you have a mutation in the *CDH1* gene, you have a 50% chance of passing the mutation down to each of your children.

What should I do about my risk of gastric cancer?

- Know the symptoms:
 - Abdominal pain, decreased appetite, weight loss, indigestion, heartburn, excessive burping, feeling full or bloated easily, difficulty swallowing, nausea, vomiting, changes in bowel movements, blood in the stool
- Consider your options:
 - Some people choose surveillance with upper endoscopy and stomach biopsies. Unfortunately, this method is not always able to detect stomach cancer and it is likely that it will miss cancers hiding in the stomach lining.
 - It is recommended that people with HDGC have a total gastrectomy, or removal of the stomach, to prevent diffuse gastric cancer. The timing of this surgery depends on many factors and should be discussed with your healthcare providers.
- Find a team of specialists who are familiar with HDGC:
 - Gastroenterologists, surgical oncologists, pathologists, and nutritionists can help you develop a care plan that is right for you.
 - Sometimes it is helpful to talk to a psychologist or therapist about the difficult decisions and how to manage the emotions that come with a diagnosis of HDGC.
- Treatment protocols continue to evolve for HDGC. Ask your provider to review current guidelines and recommendations with you.

What should I do about my risk of breast cancer?

- Know the symptoms:
 - A breast lump, skin changes or dimpling of the breast, breast asymmetry, nipple pain or discharge, breast pain
- Consider your options:
 - Some people choose surveillance with clinical breast exams, mammograms, and MRIs of the breasts on a regular basis. It is recommended to start breast surveillance at age 30.
 - Taking risk reducing drugs or having a mastectomy to prevent breast cancer may be an option. The decision depends on many factors and should be discussed with your healthcare providers.
- Find a team of specialists who are familiar with HDGC:
 - Breast oncologists, surgical oncologists, and pathologists can help you develop a care plan that is right for you.
 - Sometimes it is helpful to talk to a psychologist or therapist about the difficult decisions and how to manage the emotions that come with a diagnosis of HDGC.
- Treatment protocols continue to evolve for HDGC. Ask your provider to review current guidelines and recommendations with you.

Find current HDGC Clinical Guidelines at:
HereditaryDiffuseGastricCancer.org/hdgc-clinical-guidelines

Thanksgiving is Family Health History Day!

Create a
Family Health Portrait
familyhistory.hhs.gov

What else can I do?

- Stay healthy by eating right, exercising daily, avoiding smoking, and not drinking too much alcohol.
- Advocate for yourself and your family, and find a provider who knows (or is willing to learn) about HDGC.
- Find a genetic counselor in your area who can guide you and answer your questions about HDGC.
- Talk to all of your family members about what you learn and how they can stay healthy.
- Become involved in support groups.

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