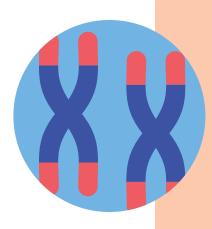
## What Is Hereditary Diffuse Gastric Cancer?



Hereditary Diffuse Gastric Cancer (HDGC) is a rare cancer syndrome that leads to a higher-than-average risk of developing two types of cancer: diffuse gastric cancer (DGC) and lobular breast cancer (LBC).



Diffuse gastric cancer is a type of cancer that spreads within the lining of the stomach. Because it does not typically cause a bulky tumor to develop, it is more difficult to find and more likely to be found at a later stage when it can be very difficult to treat successfully.

#### **How Does a Person Get HDGC?**

HDGC is inherited, meaning that the condition is passed down from a parent to a child. Most families with HDGC have a mutation\* (a change in the genetic code) in a gene called *CDH1*.

When you have a parent with HDGC caused by a mutation\* in the *CDH1* gene, you have a 50 percent chance of also having that same mutation\*.

In a small number of cases, HDGC is caused by a mutation\* in a different gene called *CTNNA1*.

For more information on genes and genetic testing, see "**Genetic Testing and HDGC**."

# CDH1 CTNNA1

**50%** chance to pass mutation\* to children

### What Should I Do If I Suspect My Family Has HDGC?

If you can, try to gather information on the types of cancer your family has experienced, pathology reports from doctors, the age of the family member at diagnosis and, if it applies, the age that they died. Tell this information to your doctor and request a referral to a genetic counselor.

#### **FAMILY HEALTH REPORT**









TYPES OF

PATHOLOGY REPORTS

AGE AT DIAGNOSIS

VISIT YOUR DOCTOR

<sup>\*</sup>A mutation is also known as a pathogenic or likely pathogenic (PLP) variant.



#### What Does It Mean to Have HDGC?

If you are diagnosed with HDGC syndrome, it does not necessarily mean that you have or will develop cancer — you just have a **higher risk**.

DIFFUSE GASTRIC CANCER 30-40% Risk

LOBULAR BREAST CANCER 40-55% Risk

#### Though It Can Be Difficult to Estimate Your Exact Risk of Cancer:

With a *CDH1* mutation\*, it is generally accepted that the lifetime risk for **diffuse gastric cancer** is in the 30% to 40% range.

Women with a *CDH1* mutation\* have an estimated 40 to 55 percent risk of having **lobular breast cancer** in their lifetime.

People with a *CDH1* mutation\* and little or no family history of stomach cancer may have a lower risk of diffuse gastric cancer, but family history may not be a completely reliable factor in determining cancer risk.

1

2

3

These cancers can affect adults of all age groups, with an average age of 47.



#### **Know Your Risk and Management Options**

Knowing that you are at a higher risk for certain cancers could save your life.



If You Are at Risk of Gastric Cancer Due to a **CDH1** Mutation\*, Consider Your Options.



FIND A TEAM WITH HDGC EXPERTISE



**SCREENING** 



TOTAL GASTRECTOMY

\*A mutation is also known as a pathogenic or likely pathogenic (PLP) variant.

#### Find a team with *CDH1* and gastric cancer expertise:

No matter what option you choose, a team with genetic counselors, gastroenterologists, surgical oncologists, pathologists and registered dietitians will be indispensable to help develop the care plan that is right for you.

#### Screening:

If you are diagnosed with a *CDH1* mutation\*, you should have an upper endoscopy (a procedure done to visually examine your upper digestive system) and stomach biopsies (the removal of samples of stomach tissue for examination).

#### **Total Gastrectomy:**

It is recommended that individuals who have a *CDH1* mutation\* and a family history of gastric cancer have a total gastrectomy, or removal of the stomach, to eliminate the risk of developing an advanced gastric cancer. This surgery is also referred to as a *risk reducing total gastrectomy*. Some people with a *CDH1* mutation\* are not able to have a gastrectomy or choose not to have the surgery. For these people, upper endoscopy every 6- 12 months is advised.





#### **BREAST CANCER**

If you are at risk of lobular breast cancer (LBC) due to a *CDH1* mutation\*, consider your options.







FIND A TEAM
FAMILIAR WITH
HDGC AND LBC

,

BILATERAL MASTECTOMY

#### Find a team with CDH1 and LBC expertise:

This may include breast oncologists, surgical oncologists and pathologists who can help you develop a care plan that is right for you.

#### **Screening:**

Recommended screening procedures include mammograms and MRIs of the breasts on a regular basis starting at age 30.

#### **Bilateral Mastectomy:**

Having a bilateral mastectomy (surgery to remove both breasts) to decrease breast cancer risk may be an option.



#### **Other Risk Factors**

Many factors can influence the risk of cance.

Diet and nutrition guidelines that may reduce the risk of stomach cancer in the absence of a *CDH1* mutation\* include the following:

Keep in mind that diet and lifestyle recommedations to reduce the risk of stomach cancer do not guarantee cancer prevention.



Maintain a healthy weight



Keep physically active



Eat a variety of whole grains, vegetables, fruit (especially citrus fruits) and beans



Avoid salt-preserved and high-salt (high sodium) foods



Avoid sugar-sweetened beverages



Limit processed and red meat consumption



Limit your alcohol intake



Avoid tobacco

#### A note about CTNNA1:

Our knowledge about *CTNNA1* cancer risk is limited. Therefore, individuals with a *CTNNA1* mutation\* are recommended to have yearly screening for gastric cancer and breast cancer. Gastrectomy and mastectomy may be options on a case-by-case basis.

\*A mutation is also known as a pathogenic or likely pathogenic (PLP) variant.