

## What you should know about Hereditary Diffuse Gastric Cancer (HDGC)

HDGC accounts for less than 1-3% of all gastric cancer. Diffuse Gastric cancer is a specific type of invasive stomach cancer that thickens the wall of the stomach wall without forming a distinct tumor. Diffuse gastric cancer is also called signet ring carcinoma or isolated cell-type carcinoma. Women with HDGC have a significantly increased risk to develop lobular breast cancer. Individuals with HDGC also have an increased risk for certain other types of breast cancer, as well as colon cancer and pancreatic cancer.

## The risk for cancer associated with Hereditary Diffuse Gastric Cancer (HDGC)

It is estimated that about 80% (4 out of 5) individuals who have HDGC will develop gastric cancer. The majority of patients develop diffuse gastric cancer by the age of 40. Women with HDGC have a 40-50% lifetime risk of developing lobular breast cancer. The average age for breast cancer diagnosis in women with HDGC is 53.

## The risks to family members

HDGC is diagnosed based on a patient's personal and family history of cancer. About 30-50% (1/3 to 1/2) of people with HDGC have a mutation in the CDH1 gene that can be identified by blood testing. In some hereditary families a genetic mutation may not be detected by the current technology, therefore close relatives will still need to be treated as high risk. Any child, brother, sister, or parent of an individual who has CDH1 mutation has a 50% chance of also having the mutation.

## Managing the Risk

Once an individual has been diagnosed with HDGC, endoscopies should be done annually, usually after age 16. Women with HDGC should have annual mammograms and breast MRIs, as well as biannual clinical breast exams, to evaluate for breast cancer beginning at age 35. A colonoscopy every 3-5 years beginning at age 40 may be recommended to screen for colon cancer. Individuals with HDGC may also be tested for H. pylori, which is a bacteria associated with an increased risk of gastric cancer.

Evidence suggests removal of the stomach (gastrectomy) is significantly better at preventing gastric cancer rather than endoscopic surveillance. Gastrectomy is generally not recommended for individuals with HDGC younger than 20 years old. Females with HDGC may consider prophylactic mastectomy to significantly reduce the risk of breast cancer.

Cancer screening should begin earlier if family members have been diagnosed with cancer at young ages.