

Clinician Management Resource for *CDH1* (Hereditary diffuse gastric cancer)

This overview of clinical management guidelines is based on this patient's positive test result for a *CDH1* gene mutation. Please consult the referenced guideline for complete details and further information.

Clinical correlation with the patient's past medical history, treatments, surgeries and family history may lead to changes in clinical management decisions; therefore, other management recommendations may be considered. Genetic testing results and medical society guidelines help inform medical management decisions but do not constitute formal recommendations. Discussions of medical management decisions and individualized treatment plans should be made in consultation between each patient and his or her healthcare provider, and may change over time.

SCREENING/SURGICAL CONSIDERATIONS	AGE TO START	FREQUENCY
Gastric Cancer^{1,2*}		
Prophylactic gastrectomy is recommended. <ul style="list-style-type: none"> Baseline endoscopy prior to gastrectomy Intraoperative frozen sections should be performed to ensure complete removal of gastric tissue A D2 lymph node dissection is not necessary Not recommended under 18 years of age, but may be considered for certain patients (i.e. family history of gastric cancer diagnosed under age 25) 	Between 18-40 years old	N/A
Patients who elect not to undergo prophylactic gastrectomy should be offered upper endoscopy with multiple random biopsies	Individualized	Every 6-12 months
Female Breast Cancer²		
Breast Screening <ul style="list-style-type: none"> Mammography with consideration of tomosynthesis Consider breast MRI with contrast 	30 years old, or 5-10 years before the earliest known breast cancer in the family	Every 12 months
For consideration of risk-reducing mastectomy, manage based on family history	Individualized	N/A

* There is controversy over how to manage gastric cancer risk in individuals with pathogenic/ likely pathogenic variants in *CDH1* in the absence of a family history of gastric cancer. However, one small study found that >50% of such individuals had gastric cancer at the time of risk-reducing total gastrectomy (Jacobs MF, et al. Gastroenterology 2019; 157:87-96).

1. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Gastric Cancer version 3.2020. © National Comprehensive Cancer Network, Inc. 2020. All rights reserved. Accessed September 22, 2020. To view the most recent and complete version of the guideline, go online to NCCN.org. NCCN makes no warranties of any kind whatsoever regarding their content, use or application and disclaims any responsibility for their application or use in any way.
2. Referenced with permission from the NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic. V1.2021. © National Comprehensive Cancer Network, Inc. 2020. All rights reserved. Accessed September 22, 2020. To view the most recent and complete version of the guideline, go online to NCCN.org. NCCN makes no warranties of any kind whatsoever regarding their content, use or application and disclaims any responsibility for their application or use in any way.