



CDH1 MEN



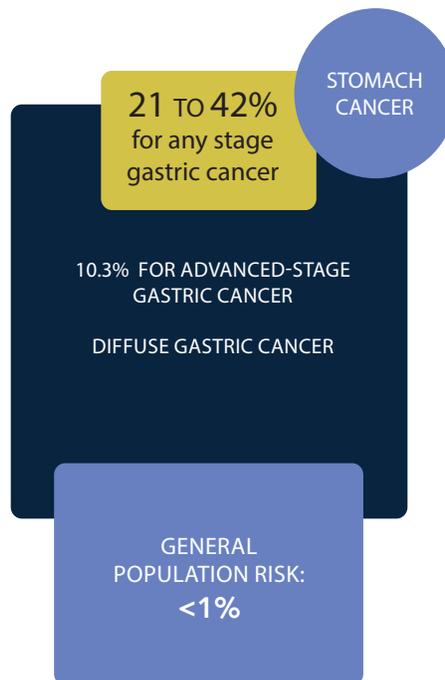
The medical management guidelines from the National Comprehensive Cancer Network (NCCN) for patients with a CDH1 pathogenic / likely pathogenic variant are listed in this document.

This overview is for informational purposes and does not constitute a personalised recommendation. **Recommended options may vary based on your personal and family history. Access to some options may also vary from one medical center to another.** The specific references should be consulted for more details before developing a treatment plan.

In addition, the information available on hereditary cancer susceptibility genes is constantly evolving and **it is recommended to check this information annually as the management guidelines may change in the future.**

LIFETIME ASSOCIATED RISKS

(UP TO AGE 75)



NOTE

Cleft lip with or without cleft palate has been associated with CDH1 pathogenic and likely pathogenic variants.

References:

Daly M et coll. NCCN Clinical Practice Guidelines in Oncology: Genetic/Familial High-Risk Assessment: Breast, Ovarian and Pancreatic. Version 3.2025-March 6, 2025. <http://www.nccn.org>

Barbosa-Matos R, Córdova L, Schrader K, et al. Diffuse Gastric and Lobular Breast Cancer Syndrome. 2002 Nov 4 [Updated 2024 Oct 10]. In: Adam MP, Feldman J, Mirzaa GM, et al., editors. GeneReviews® [Internet]. Seattle (WA): University of Washington, Seattle; 1993-2025. <https://www.ncbi.nlm.nih.gov/books/NBK1139/>

Gupta S et coll. NCCN Clinical Practice Guidelines in Oncology: Genetic/Familial High-Risk Assessment: Colorectal, Endometrial, and Gastric. Version 4.2024. April 2, 2025. <http://www.nccn.org>



STOMACH CANCER

SCREENING

STARTING AT AGE 18

IF ELECT TO NOT UNDERGO PROPHYLACTIC TOTAL GASTRECTOMY

- UPPER ENDOSCOPY EVERY 6 TO 12 MONTHS WITH MULTIPLE BIOPSIES

IMPORTANT : THE EFFECTIVENESS OF THIS SCREENING METHOD HAS NOT BEEN RECOGNIZED FOR THE DETECTION OF EARLY GASTRIC CANCER

18

STOMACH CANCER

RISK REDUCTION

SURGERY

BETWEEN AGES 18 AND 40 :
OR EARLIER ACCORDING TO FAMILY HISTORY OF GASTRIC CANCER PRIOR TO AGE 25

- PROPHYLACTIC TOTAL GASTRECTOMY (REMOVAL OF THE STOMACH BEFORE CANCER DEVELOPS) IS RECOMMENDED IF ANY OF THE FOLLOWING ARE PRESENT:
 - ESTABLISHED STAGE PT1B OR HIGHER SRCC (SIGNET-RING CELL CARCINOMA)
- PERSISTENT SIGNS AND SYMPTOMS ASSOCIATED WITH SRCC, SUCH AS : WEIGHT LOSS, EARLY SATIETY, ANEMIA AND ABDOMINAL PAIN A BASELINE ENDOSCOPY IS INDICATED PRIOR TO PROPHYLACTIC TOTAL GASTRECTOMY.
- A D2 LYMPH NODE DISSECTION IS NOT NECESSARY FOR PROPHYLACTIC TOTAL GASTRECTOMY.

IMPORTANT : THE HISTOLOGY ANALYSIS OF THE BIOPSIES SHOULD BE THOROUGH TO RULE OUT THE PRESENCE OF MACROSCOPIC TUMOR FOCI OR OTHER FACTORS THAT MAY MODIFIED THE SURGERY.

NOTE : IN THE ABSENCE OF A FAMILY HISTORY OF GASTRIC CANCER, THERE IS CONTROVERSY OVER HOW TO MANAGE GASTRIC CANCER RISK IN INDIVIDUALS WITH CDH1 PATHOGENIC AND LIKELY PATHOGENIC VARIANTS. ONE SMALL STUDY FOUND THAT >50% OF THESE INDIVIDUALS HAD GASTRIC CANCER IDENTIFIED AT THE TIME OF RISK-REDUCING TOTAL GASTRECTOMY.