New NCCN Guidelines Are First to Address Small Intestine Cancers; Updates Highlight Developments in Genetic Testing and Hereditary Risk for Colon and Other Intestinal Cancers

NCCN published of a new set of recommendations focused on cancer in the small intestine. The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Small Bowel Adenocarcinoma (SBA) are the first treatment guidelines in the United States (and second worldwide) to address this rare cancer type that is increasing in incidence. Small bowel cancers (of which adenocarcinomas are the most common) are responsible for approximately 3% of all digestive system cancers, with an estimated 10,590 new cases expected in 2019.[1]

"Historically, small bowel adenocarcinoma has been managed in the same way as colorectal cancer (CRC), because SBA is rare and, therefore, difficult to study," said Katrina Pedersen, MD, MS, Siteman Cancer Center at Barnes-Jewish Hospital and Washington University School of Medicine, an SBA expert and member of the NCCN Guidelines Panel for CRC. "The new NCCN Guidelines for SBA were created to reflect new research and biologic insights over the past several years that show optimal SBA management can differ from CRC treatments. For example, we're incorporating a different class of drugs called taxanes into SBA treatment and deemphasizing the use of EGFR inhibitors that do not show a clear benefit. Our goal is to improve outcomes by standardizing the diagnosis, staging, and multimodal treatment possibilities for patients with SBA."

"Our panels work very hard to make sure the NCCN Guidelines are up to date and inclusive for any therapeutic advances," said Al B. Benson III, MD, Robert H. Lurie Comprehensive Cancer Center of Northwestern University and Chair, NCCN Guidelines Panel for Colon/Rectal/Anal Cancers. "We do extensive review and discussion of all existing evidence, and are constantly exploring ways to provide a more comprehensive approach for patients. In the case of intestinal cancers, which include colon, rectal, anal, and now small intestine, we're expanding the guidelines to cover rare subsets. These efforts are also reflected in personalized treatment recommendations for patients with tumors that have certain genetic mutations or characteristics, such as microsatellite instability-high (MSI-H), *BRAF* mutation, or HER2 overexpression. Hereditary risk assessment and screening are also essential components in caring for patients with intestinal cancers, since many are potentially preventable."

Rates for many other gastrointestinal malignancies have been on the decline in recent years, in part as a result of increases in screening. At the same time, rates for small bowel cancers have increased 1.8% between 2006 and 2015.[2] There is not currently a consensus on screening for SBA, because it's located higher on the gastrointestinal tract and therefore is not detected during routine colonoscopy. However, it has been shown to be associated with Lynch syndrome (among other familial syndromes), Crohn's disease, and colitis.

NCCN recently released updates to the NCCN Guidelines for Genetic/Familial Risk Assessment: CRC, reflecting the commitment to enhancing surveillance strategies that would increase early CRC detection rates, particularly as they relate to individuals with a personal or family history. The extensive update includes a clarified step-by-step process for the assessment of hereditary CRC syndromes, and a greater emphasis on the importance of genetic counseling.

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"Many of these changes reflect the widespread adoption of multigene panel testing for hereditary cancer syndromes, which allows assessment for even rare causes of polyposis and other cancer syndromes," explained Heather Hampel, MS, LGC, The Ohio State University Comprehensive Cancer Center - James Cancer Hospital and Solove Research Institute and member of the NCCN Guidelines Panel for Genetic/Familial Risk Assessment: Colorectal. "We've added a section on the principles of cancer risk assessment and counseling, and also refined the initial approach to assessing hereditary CRC syndromes, included rare genetic causes for multiple adenomatous polyps, and updated the criteria for evaluation of Lynch syndrome."

The NCCN Guidelines for CRC Screening were also recently updated. The panel of experts behind those guidelines is closely following any emerging studies regarding new CRC diagnoses in younger people.

"The panel has reviewed the recent data for initiating screening of average-risk individuals before age 50," said Dawn Provenzale, MD, MS, Duke Cancer Institute and Chair of NCCN Guidelines Panel for CRC Screening. "Based on those data, the panel continues to endorse screening of average risk individuals at age 50. The panel will continue to review this strategy and monitor data as they emerge."

The NCCN Guidelines for SBA, CRC Screening, and Genetic/Familial High-Risk Assessment: Colorectal are all available free-of-charge for non-commercial use at NCCN.org, or via the Virtual Library of NCCN Guidelines App for smartphone and tablet.

References

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2. Noone AM, Howlader N, Krapcho M, et al. SEER Cancer Statistics Review, 1975–2015, based on November 2017 SEER data submission, posted to the SEER website, April 2018. Bethesda, MD: National Cancer Institute; 2018. Available at: https://seer.cancer.gov/csr/1975_2015/.