

New Pediatric Cancer Guidelines Published by NCCN Focus on Improving Long-Term Outcomes Worldwide

NCCN published new guidelines for treating children, adolescents, and young adults with pediatric aggressive mature B-cell lymphomas, including Burkitt lymphoma and diffuse large B-cell lymphoma. NCCN published the first pediatric NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) earlier this year that addressed Acute Lymphoblastic Leukemia. These new pediatric guidelines—with more to follow—further NCCN's efforts to improve quality of care and outcomes for children with cancer worldwide.

Historically, most children with cancer in the United States have been included on active clinical trials, with clear protocols in place to standardize treatment and maximize response, while addressing potential toxicities. However, improved cure rates and decreased research funding have resulted in more patients being treated outside of clinical trials, where treatment guidelines are lacking. The new NCCN Guidelines are intended to fill the void and make sure management is provided in the best possible manner to improve both short- and long-term outcomes. Furthermore, globally (especially in resource-constrained settings), many patients do not have access to oncologists who specialize in pediatric cancers, making guidelines highly relevant for these situations.

Non-Hodgkin lymphoma is responsible for approximately 5% of all cancers in children aged ≤ 14 years and 7% of all cancers for adolescents between ages 15 and 19 years.¹ Major supportive care concerns include disease-associated infections, renal dysfunction, bowel obstruction, treatment-related tumor lysis syndrome, neurotoxicity, and mucositis.²⁻⁵

“Over the course of my career, one of the successes I have witnessed is the improvement in treatment outcomes for children, adolescents, and young adults with aggressive mature B-cell lymphomas. These previously fatal diseases can now be cured in almost all cases, thanks to decades of cooperative international research into how to optimize the way we approach treatment and supportive care,” explained Kimberly J. Davies, MD, Medical Director of Dana-Farber's Pediatric Oncology Clinic at Dana-Farber/Boston Children's Cancer and Blood Disorders Center and Chair of the NCCN Guidelines Panel for Pediatric Aggressive Mature B-Cell Lymphomas. “Treatments for these lymphomas are intensive and complicated, but when applied appropriately and with good supportive care, they have high curative potential. The NCCN Guidelines have been developed by experts in the field to share decision-making recommendations with all pediatric oncologists for the best treatment of these lymphomas.”

“These new recommendations from NCCN represent one of the first guidelines developed specifically for the care of pediatric patients with cancer,” said Matthew Barth, MD, Roswell Park Comprehensive Cancer Center, Vice-Chair of the NCCN Guidelines Panel for Pediatric Aggressive Mature B-Cell Lymphomas. “Based on evidence from recent clinical trials, we are now achieving unprecedented success in treating children with aggressive mature B-cell non-Hodgkin lymphomas, including Burkitt lymphoma and diffuse large B-cell lymphoma, and we want to make sure that providers around the world have access to these best practices. This resource will help guide treatment decisions for providers treating pediatric, adolescent, and select young adult patients—for whom recommended regimens depart significantly from the

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recommendations for older adults—while also providing guidance on appropriate supportive care, which has also played a major role in improving survival outcomes in childhood B-cell lymphomas.”

“Together with the new NCCN Guidelines for Pediatric Acute Lymphoblastic Leukemia, these NCCN Guidelines for Pediatric Aggressive Mature B-Cell Lymphomas begin the exciting expansion of the widely used NCCN Guidelines into pediatric diseases,” added Alexandra E. Kovach, MD, Vanderbilt-Ingram Cancer Center, Member of the NCCN Guidelines Panel for Pediatric Aggressive Mature B-Cell Lymphomas. “The goal of these guidelines is to provide oncologists, pathologists, general practitioners, and students with a comprehensive yet concise and user-friendly guide to modern diagnosis, staging, treatment and management of pediatric Burkitt lymphoma and diffuse large B-cell lymphoma, and to illustrate how management differs from how these diseases are handled in adult patients.”

Up next, NCCN plans to adapt these guidelines into NCCN Harmonized Guidelines for Sub-Saharan Africa, which take into account specific regional concerns and potential resource limitations. The organization will also continue expanding into pediatric recommendations with upcoming NCCN Guidelines for Wilms Tumor and Pediatric Hodgkin Lymphoma. Recently, NCCN also published new supportive care guidelines detailing best practices in the lead-up to and after-effects of stem-cell transplantation, and significantly updated the NCCN Guidelines for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic.

All NCCN Guidelines can be viewed and downloaded free-of-charge for noncommercial use at NCCN.org or via the NCCN Virtual Library of NCCN Guidelines app.

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Closing Gaps in Perspective of Value Between Patients With Cancer and Their Healthcare Providers

NCCN hosted an annual NCCN Patient Advocacy Summit: Delivering Value for Patients across the Oncology Ecosystem in Washington, DC, on December 11, 2019. The summit brought together patients, advocates, clinicians, policy-makers, and others to share diverse perspectives on the meaning of value in cancer care. The event also featured a keynote address on incorporating the patient voice into evidence-based care from Paul G. Kluetz, MD, Deputy Director, Oncology Center of Excellence, FDA.

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“We must always keep in mind that no two individuals will define value in exactly the same way,” explained Ilana Feuchter, MA, Senior Manager for National Advocacy, National Ovarian Cancer Coalition. “Therefore, patients must be included in any discussion of value at the beginning of their diagnosis and throughout each step of the process as that definition may change following treatment or if there’s a change in prognosis.”

Representatives from some of NCCN’s Member Institutions shared their experience from the clinical and institutional perspective, with lessons learned from their high volume of patients with cancer.

“Improving patient experience is an essential task for clinicians and hospitals,” said Justin E. Bekelman, MD, Associate Professor and Director of the Penn Center for Cancer Care Innovation, Abramson Cancer Center of the University of Pennsylvania, who delivered a keynote address titled ‘High-Value Cancer Care: What’s in It for Patients, Clinicians, Hospitals, Industry, and Payers?’ “It’s part of our mission, right up there with improving treatment outcomes and affordability, to ensure that each patient is approached with humanity and humility and feels the care they received was the best it could possibly be.”

“Personalized care for people with cancer not only is driven by clinical algorithms, but also includes a care design for each unique patient and their families, starting with access, finances, social service support, and one-on-one relationships with their clinical support team. Personal care design allows every patient’s voice to be heard so their goals for care can be achieved,” said Kimberly Bell, BSN, MBA, Administrator of Cancer Services, Cleveland Clinic Taussig Cancer Institute. “The value-based payment models being developed by payers and providers must consider the patient’s perspective on quality, care continuum, and finances. As we evolve in this area, this perspective needs to be integrated so patients feel the positive impact on care.”

Financial toxicity was a recurring topic throughout the summit, including issues like fee transparency, lost wages, and auxiliary costs of care. “We know that behind the words ‘patient,’ ‘sick,’ and ‘cancer,’ there is a family,” said Carla Tardif, Chief Executive Officer, Family Reach. “That family’s only worry should be getting to the other side of cancer, but when financial barriers get in the way of treatment, that is simply not the case.”

Shame and silence around a type of cancer, or as a member of a marginalized community, can pose additional barriers to high-quality care. “LGBT people fear homophobia and heteronormative values will crowd out inclusive consideration of the healthcare needs for our communities regarding value-based care,” said Darryl Mitteldorf, LCSW, Executive Director, The National LGBT Cancer Project. “LGBT people can have different configurations of what a family is, and our families are rarely considered by the healthcare system in the United States. The National LGBT Cancer Project, along with our partner, Malecare, facilitates the world’s largest anxiety support group, with over 43,000 active members, because LGBT people present with higher incidence rates of anxiety than most groups.”

“You have to be your own best advocate. If you believe something is wrong, press on, because you are probably right,” said Lillian Kreppel, Patient Ambassador, The Anal Cancer Foundation. “Don’t ignore symptoms like bleeding, lumps, bumps, or hemorrhoids. Don’t be so embarrassed that you won’t seek medical help. And don’t necessarily think that doctors have all the answers or know everything, especially when you are the one who lives in your body. Get a second and third opinion, and get educated on the HPV vaccine.”

Sexual health was also a key topic of conversation. “One big issue for patients with colorectal cancer regardless of age, but especially younger patients, is that physicians need to better address intimacy, fertility preservation, and overall sexuality,” said Ronit Yarden, PhD, MHSA, Senior Director of Medical Affairs, Colorectal Cancer Alliance. “Treatments for colorectal cancer often come with debilitating side effects that can reduce a patient’s quality of life. We not only want patients to survive, but we want them to thrive. They need open and honest conversations about their different options and the impact that their treatment will have on their everyday lives. Having these conversations before treatment—not afterwards, when patients are often surprised and overwhelmed—will help ensure that survivors can live their best lives.”

“Maintaining quality-of-life specific to sexual health and intimacy is important to patients with prostate cancer posttreatment,” agreed Chuck Strand, Chief Executive Officer, Us TOO Prostate Cancer Education & Support. “Erectile dysfunction and urinary incontinence can be temporary or ongoing treatment side effects from surgery or radiation. It’s important to patients and their partners to address this with their healthcare team during the process of making a shared decision on a treatment pathway, and be prepared to address management of these issues, if necessary, while recognizing the difference between intimacy and sex.”

Several panelists reiterated that new laws and policies can serve as an important motivation for improvement, as long as they are implemented with the end goal of improving the patient experience, and not simply a checklist to cross off. Increasing use of shared decision-making and patient-reported outcomes can result in care that better meets the needs of the people receiving it.

The panel discussions were moderated by Clifford Goodman, PhD, Senior Vice President, The Lewin Group, and included the following additional participants:

- Alan Balch, PhD, CEO, National Patient Advocate Foundation
- Kristin L. Carman, PhD, MA, Director of Public and Patient Engagement, Patient-Centered Outcomes Research Institute (PCORI)
- Brian Connell, Executive Director, Federal Affairs, The Leukemia & Lymphoma Society (LLS)
- Andrea E. Ferris, MBA, President and Chief Executive Officer, Lungevity
- Elizabeth Franklin, LGSW, ACSW, Executive Director, Cancer Policy Institute, Cancer Support Community
- Miranda Goff, LICSW, Support Services Manager, GO2 Foundation for Lung Cancer
- Edward Kim, MD, Chair, NCI Central Institutional Review Board, Adult CIRB - Early Phase Emphasis (EPE) Board
- Molly McDonnell, Director of Advocacy, Fight Colorectal Cancer

The NCCN Patient Advocacy Summit follows the recent NCCN Policy Summit on Defining, Measuring, and Applying Quality in an Evolving Health Policy Landscape and the Implications for Cancer Care. In 2020, NCCN Policy will host summits on how to accelerate advances in cancer care research, drive down healthcare costs, and improve patient access to quality cancer care across the lifespan. For dates and other information, visit [NCCN.org/policy](https://www.nccn.org/policy), and join the conversation online with the hashtag #NCCNPolicy.

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NCCN Announces Projects Funded for Metastatic Breast Cancer Grant Program, in Collaboration With Pfizer

The NCCN Oncology Research Program (ORP) recently announced the proposals that were selected to receive funding, in collaboration with Pfizer Global Medical Grants, to improve the quality of care and outcomes for people with metastatic breast cancer (MBC). Up to \$1.4 million in funding will be provided by NCCN through a grant from Pfizer for a period of 3 years. The NCCN ORP oversaw project application and selection and will provide administrative oversight for the life of the projects.

This project builds on the success of a program that began at the Dana-Farber Cancer Institute, called EMBRACE (Ending Metastatic Breast Cancer for Everyone), which was first funded in 2015 under a previous NCCN and Pfizer collaboration. The EMBRACE program developed a consistent intake process, multifaceted engagement, coordination, and follow-up approach for patients with MBC that has made a tangible improvement in care.

“The EMBRACE program at Dana-Farber enabled us to build a comprehensive, multipronged initiative designed to meet the needs of patients, community-based physicians, and Dana-Farber-based care teams, and to deliver cutting-edge care that leverages the strength of collaboration,” said Nancy U. Lin, MD, Director of the Metastatic Breast Cancer Program at Dana-Farber Cancer Institute. “Since 2015, we have reached over 2,500 patients with MBC and interacted directly with more than 500 referring physicians through the EMBRACE program. I’m thrilled that other institutions will now have the opportunity to develop their own initiatives, and look forward to their success.”

“We’re glad to expand on this program to find new ways to harness the combined strengths of academic cancer centers and community hospitals in order to provide coordinated care that improves outcomes for patients with MBC,” said Wui-Jin Koh, MD, Chief Medical Officer, NCCN. “We hope these projects will transform the delivery of care to people with this disease, and perhaps other cancer types as well. By encouraging collaboration between community-based physicians and their academic counterparts, we also hope to increase clinical trial enrollment, which could lead us to the cures of tomorrow.”

The chosen initiatives will explore ways to leverage existing academic-community partnerships in order to develop and implement methods for improving radiation treatment delivery; create a personalized care delivery model for patients with MBC and central nervous system metastases; launch a multidisciplinary clinic for treatment recommendations, supportive care, and clinical trial screening; and increase access to genomic testing through a virtual molecular clinic. All projects will highlight the importance of patient education and the necessity of a coordinator to support patient navigation throughout the complex treatment landscape. These efforts are also expected to promote evidence-based care, be sustainable after the funding period, collect data and report outcomes, be adaptable to patient variability, and promote system-wide efficiency.

The following investigators and initiatives have been selected for funding:

- Allison Lipitz-Snyderman, PhD, and Erin Gillespie, MD, Memorial Sloan Kettering Cancer Center
 - Leveraging an Academic-Community Partnership Model to Improve the Quality of Radiation Treatment for Metastatic Breast Cancer Patients
- Aki Morikawa, MD, PhD, University of Michigan Rogel Cancer Center

- Personalized Multi-Care: A Tailored Approach to Multidisciplinary Care Coordination Delivery for Metastatic Breast Cancer Patients With Central Nervous System Metastases
- Karen Lisa Smith, MD, MPH, The Sidney Kimmel Comprehensive Cancer Center at Johns Hopkins
 - The Johns Hopkins Metastatic Breast Cancer Partners Program: Collaborating to Improve Metastatic Breast Cancer Care
- Laura Spring, MD, Massachusetts General Hospital Cancer Center
 - Expanding Precision Medicine for Patients With Metastatic Breast Cancer in the Community: Leveraging Academic Strength and Community Partnership

“We believe true advancements in cancer care come from listening to people living with the disease,” said Dany Habr, MD, Chief Medical Officer, Pfizer Oncology. “Our continued collaboration with NCCN is founded on metastatic breast cancer patient education and encourages the oncology community to work together to optimize patient care. We look forward to seeing the impact of these projects as they are implemented.”

The NCCN ORP fosters innovation and knowledge discovery to improve the lives of patients with cancer. Visit [NCCN.org/GRP](https://www.nccn.org/GRP) to learn more about these projects and other ways the NCCN ORP is developing insights into cancer care delivery outcomes and quality and identifying solutions to barriers affecting optimal cancer management.

Updated Genetic Screening Guidelines Published by NCCN Feature Emerging Evidence on Personalized Medicine

NCCN recently announced the publication of the newest genetic risk assessment recommendations for breast, ovarian, and pancreatic cancers. The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic, Version 1.2020, contains several updates—including new and expanded sections on risk assessment and management related to the 3 major cancer types—while also maintaining a more conservative approach toward testing practices where the evidence is still lacking. In the rapidly moving field of cancer care and genetics, NCCN Guidelines synthesize the latest evidence and expert consensus to ensure recommendations are firmly supported by quality research, in order to establish best management paradigms that improve outcomes for people with cancer.

“These guidelines are as inclusive as possible, wherever there’s strong, unbiased evidence to back up our recommendations,” said Mary B. Daly, MD, PhD, Fox Chase Cancer Center, Chair of the NCCN Guidelines Panel for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic. “The guidelines include genes that have been found to increase cancer-susceptibility. These NCCN Guidelines still have a strong focus on *BRCA1* and *BRCA2* mutations, but also now include other high and moderate penetrance genes associated with breast, ovarian, and pancreatic cancers. We continuously review any new data on genes that might increase a person’s risk of getting cancer or impact the effectiveness of their treatment.”

The updated guidelines are concentrated around simplified criteria to clarify the genetic testing process. For example, in a newly added guide for individuals

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of Ashkenazi Jewish ancestry who have not been diagnosed with cancer, genetic testing may be offered for the 3 Ashkenazi Jewish founder mutations in the context of a long-term research study, regardless of family history. These individuals should be encouraged to consult with a cancer genetics professional.

The NCCN Guidelines for Genetic/Familial High-Risk Assessment are organized by both disease and syndrome type, and also now include streamlined information on appropriate subsequent steps for persons who meet criteria for genetic testing. The panel acknowledges that genetic mutations can impact the approach to cancer treatment, and the guidelines now state that testing may be clinically indicated if it will aid in systemic therapy decision-making.

“Genetic testing is becoming increasingly utilized in oncology because of its potential to impact surgical decisions and chemotherapy,” explained Robert Pilarski, MS, LGC, MSW, Professor, Clinical Internal Medicine, The Ohio State University Comprehensive Cancer Center, Vice-Chair of the NCCN Guidelines Panel for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic. “At the same time, the complexity of this testing is increasing, with a growing number of genes and tests available, a limited understanding of the management implications of some of the newer genes, and even uncertainty over the implications of mutations in well-established genes in some situations (for example in a condition known as ‘mosaicism,’ in which the mutation is not present in all of the cells of the body). Because of this, the NCCN Guidelines continue to highlight the critical importance of genetic counseling for patients prior to undergoing genetic testing to ensure that patients are fully informed of the test implications.”

Mr. Pilarski also offered an important word of caution about the potential risks from direct-to-consumer genetic testing: “More and more patients are presenting to clinic having already had themselves tested through direct-to-consumer labs. Providers need to be aware that the tests offered by many of these labs are not equivalent to traditional genetic testing, and the results may need to be confirmed in another laboratory before being used for clinical care.”

The guidelines recommend that all patients with pancreatic cancer get genetic testing, and the recent update now includes more information about which genes are associated with pancreatic cancer recommendations. Genetic testing in pancreatic cancer can help determine which treatments would be most effective (eg, PARP inhibitors) and whether family members would benefit from screening and preventive action.

“There’s been an explosion of recent data showing that roughly 4% to 10% of individuals with pancreatic cancer harbor inherited genetic mutations, including *BRCA1*, *BRCA2*, *ATM*, the Lynch syndrome genes, and others,” said Matthew B. Yurgelun, MD, Dana-Farber/Brigham and Women’s Cancer Center, Member of the NCCN Guidelines Panel for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic. “Such data have, surprisingly, shown that classic ‘high-risk’ features of inherited cancer risk (eg, young age at diagnosis, strong family histories of cancer) are often absent in individuals with pancreatic cancer who carry these mutations. Based off of these data, there is now a compelling reason for all individuals with pancreatic cancer to be offered genetic counseling and germline testing for such variants—particularly given the possibility that their at-risk family members could greatly benefit from known, effective cancer risk-reducing interventions (eg, surgical removal of the ovaries for female *BRCA1/2* mutation carriers). Emerging data have also begun to suggest possible benefits to pancreatic cancer screening in select high-risk individuals who harbor such mutations. These new guidelines address many of the important nuances and limitations of this exciting and rapidly evolving body of literature.”

The NCCN Guidelines for Genetic/Familial High-Risk Assessment are created and maintained by an interdisciplinary panel of experts from the alliance of 28 leading cancer centers that comprise NCCN. NCCN panels also include patients and advocates to ensure treatment recommendations meet the needs of people with cancer and their caregivers.

“Participating on the NCCN panel allows FORCE to share the real-world experiences of patients making complex and often agonizing medical decisions about hereditary cancer treatment and risk management,” said Sue Friedman, DVM, Executive Director, Facing Our Risk of Cancer Empowered (FORCE), Member of the NCCN Guidelines Panel for Genetic/Familial High-Risk Assessment: Breast, Ovarian, and Pancreatic. “As an advocacy organization for people and families affected by hereditary cancer, we see the importance of having standardized guidelines. These guidelines are a critical piece of informed decision-making; we frequently direct our community to NCCN for up-to-date, clear, and credible information developed by experts in the field.”

NCCN Guidelines are the recognized standard for clinical policy in cancer care and are the most thorough and frequently updated clinical practice guidelines available in any area of medicine. The intent of the NCCN Guidelines is to assist in the decision-making process of individuals involved in cancer care—including physicians, nurses, pharmacists, payers, patients and their families—with the ultimate goal of improving patient care and outcomes. In addition to covering at least 97% of cancers affecting patients in the United States, there are also NCCN Guidelines for cancer detection, prevention, risk-reduction (including smoking cessation), and supportive care (including the management of pain, distress, and fatigue), and for specific populations (including children and young adults).

NCCN Guidelines are available free-of-charge for noncommercial use at [NCCN.org](https://www.nccn.org), or via the Virtual Library of NCCN Guidelines App.

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