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New NCCN Clinical Practice Guidelines in Oncology focus on workup and management for 3 main adult histiocytic disorders.

New NCCN Guidelines for Histiocytosis Clarify Best Practices for Recently Defined Cancers

NCCN has published new treatment guidelines for histiocytosis. Histiocytoses are rare disorders characterized by a buildup of white blood cells called histiocytes, which leads to tissue damage. This group of disorders is diagnosed in a few thousand people every year across the United States. As a result of this low incidence rate, people are often diagnosed late and receive treatment from doctors who have limited or no prior experience with the disease. These new clinical practice guidelines from NCCN provide the latest evidence and expert-consensus for diagnosing and treating the 3 most-common forms of histiocytosis in adults: Langerhans cell histiocytosis (LCH), Erdheim-Chester disease (ECD), and Rosai-Dorfman disease.

“NCCN has assembled a panel of experts who have treated these rare cancers, enabling us to put together practice recommendations based on their collective experience supplementing the limited evidence available from published studies,” said Ronald S. Go, MD, Mayo Clinic Cancer Center, Chair of the NCCN Guidelines Panel for Histiocytic Neoplasms. “For these ‘orphan’ cancers, the main challenges are late diagnosis, uncertainty on how to stage and monitor, and difficulty getting insurance to approve beneficial off-label treatments. Many histiocytosis patients have targetable mutations that are also present in some more common cancers with FDA approvals for targeted agents.”

“Erdheim-Chester disease, a histiocytic neoplasm primarily seen in adults, was not officially recognized as a cancer by the World Health Organization until 2016. Historically, people with these disorders would receive empiric treatments from various specialists like rheumatologists and dermatologists; many wouldn’t even see an oncologist until years later,” said Gaurav Goyal, MD, O’Neal Comprehensive Cancer Center at UAB, a member of the NCCN Guidelines Panel for Histiocytic Neoplasms. “It’s crucial that patients get the full diagnostic workup to guide treatment and reverse or ameliorate the course of the disease. We’ve been able to make tremendous gains in reducing mortality through genetic testing and treatment selection. The NCCN Guidelines will help us ensure every patient is able to benefit from the panel’s collective expertise, including many with some of the broadest experience in treating people with these rare cancers.”

One of the difficulties in treating different histiocytic disorders is the fact that they vary so much from one another and present in many different ways.

“Each of these diseases is biologically different with widely variable clinical presentation, diagnostic testing, and treatments employed,” said Reem Karmali, MD, MS, Robert H. Lurie Comprehensive Cancer Center of Northwestern University, another NCCN Guidelines for Histiocytic Neoplasms panel member. “Symptoms vary as well – some patients are asymptomatic while others may develop chronic organ dysfunction or life-threatening symptoms that require immediate therapy. These NCCN Guidelines will raise awareness, educate the medical community at large, and provide a reference for commercial insurers to improve patient accessibility to diagnostic tools and potentially life-saving treatments.”

The NCCN Guidelines for Histiocytosis are one of 80 different sets of recommendations that are updated and published by NCCN. They cover rare and common cancer types along with supportive care; detection, prevention, and risk reduction; and treatment for specific populations. There are also NCCN Guidelines

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for Patients that provide the same expert content in easy-to-follow, jargon-free wording. All NCCN Guidelines are available free-of-charge for non-commercial use at NCCN.org or via the Virtual Library of NCCN Guidelines App.

“The Histiocytosis Association appreciates the dedication of all those involved at the NCCN for developing and publishing guidelines for 3 of the histiocytic disorders,” said Deanna Fournier, Executive Director, Histiocytosis Association. “As with all rare diseases, early diagnosis and awareness of the proper course of treatment has a very positive impact on patient outcomes; having access to the most up-to-date information at the ready will enable physicians around the world to continue to provide the best care to adults and children impacted by histiocytosis. We look forward to sharing the guidelines with our community.”

“The ECD Global Alliance is very thankful for the hard work of the NCCN and the involved experts from the histiocytosis medical community for the publication of these guidelines,” agreed Kathy Brewer, Global Alliance President, Erdheim-Chester Disease Global Alliance. “We are so hopeful that this will allow ECD and all histiocytosis patients to more easily gain access to the best care possible.”

NCCN Guidelines are updated at least once a year. Future versions of the NCCN Guidelines for Histiocytosis include a planned expansion into pediatric treatment. For updates, register for a free account at NCCN.org.

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Leading Health and Cancer Advocacy Groups Unite to Reduce Racial Disparities in Cancer Care

NCCN, American Cancer Society Cancer Action Network (ACS CAN), and the National Minority Quality Forum (NMQF) have presented new ideas for overcoming inequality in oncology. The recommendations—developed by a group of 17 national experts, representing patients and advocates, caregivers, healthcare providers, researchers, and industry—directly address how medical systems in the United States often disproportionately fail minority patients, particularly those who are Black and/or Indigenous, and draws on extensive polling data from a recent poll fielded by Public Opinion Strategies on behalf of the organizations to make clear the case for urgent action.

Among the notable survey findings: 63% of African American and 67% of Latinx patients, survivors, and caregivers said they had a negative experience with their oncology care team, such as having assumptions made about them or their financial situation, or trouble getting questions answered; in contrast to 43% of White respondents who reported such experiences. As for oncologists, two-thirds of those surveyed believed that non-White patients experienced worse outcomes from cancer care but only one-third felt those patient populations were receiving worse care or poorer communication during care.

“The research shows disparities in outcomes that aren’t based on biology; they result from systemic inequality and bias in access and care delivery,” said Robert W. Carlson, MD, Chief Executive Officer, NCCN. “Racism exists in the United States and impacts everyone, including the medical community. One proven method for improving equity is using guideline-concordant care. We’re grateful to be able to learn from experts about what we can do right now to make sure everyone is getting the best evidence-based care possible; the

“NCCN, ACS CAN, and NMQF present new polling data showing patient, caregiver and provider views of bias and suggest policy and practice changes to improve equity in access to high quality cancer care.”

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We can improve cancer outcomes for communities of color and rural areas by closing gaps in screening, diagnosis, treatment and survivorship, both in clinical practice and in policy.

process has been both humbling and inspiring. Now we're committed to working together to make these recommendations a reality.”

“Communities of color and other medically underserved groups continue to have higher cancer rates and are less likely to be diagnosed early or receive optimal treatment compared to other groups,” said Lisa Lacasse, President of ACS CAN. “Ensuring quality clinical practices are in place and applied equitably to all patients regardless of race, ethnicity, socioeconomic status or geographic location is essential to reducing those disparities. We cannot achieve our mission of a world without cancer until we lessen the burden of this disease across all communities; these recommendations provide important means to achieve that goal.”

“We can improve cancer outcomes for communities of color and rural areas by closing gaps in screening, diagnosis, treatment and survivorship, both in clinical practice and in policy,” said Gary A. Puckrein, PhD, NMQF President and CEO. “NMQF is pleased to collaborate with NCCN and ACS CAN to develop evidence-based recommendations using real-world data that allow us to design a system that delivers high-quality cancer care to all of America's diverse populations.”

To address the inequalities in access to cancer care, the 3 organizations convened the Elevating Cancer Equity Working Group, co-chaired by Shonta Chambers, MSW, EVP Health Equity Initiatives and Community Engagement, Patient Advocate Foundation, and Robert Winn, MD, Director, VCU Massey Cancer Center. Workgroup recommendations include a new Equity Report Card to help providers, payers, and accreditation entities advance equitable care delivery. The report card includes 17 measurable practice changes, such as having health systems provide and require annual implicit bias training for all employees, offer culturally and linguistically representative patient navigators or community health workers through internal hiring or contracting with community-based organizations, and offer flexible hours for screening and treatment appointments. The full list of practice recommendations are broken down into the following categories:

- Community Engagement
- Accessibility of Care and Social Determinants of Health
- Addressing Bias in Care Delivery
- Quality and Comprehensiveness of Care

The recommendations will be explained in greater detail during a keynote address at the NCCN Virtual 2021 Annual Conference online on March 19.

The organizations also created a series of policy change recommendations targeted toward:

- The United States Congress
- CMS and Commercial Payers
- Federal Agencies
- State and Local Policymakers

Policy examples include measures to remove barriers to clinical trial participation and requiring the FDA to consider clinical trial diversity as part of a drug approval process, providing resources to historically Black colleges and universities and other minority serving institutions with the goal of fostering a more diverse health care workforce, funding public awareness campaigns around cancer prevention that are linguistically and culturally reflective of diverse audiences, and ensuring access to and reimbursement for patient navigators to assist patients with all kinds of insurance.

The groups plan a series of ongoing engagements for providers, patients and lawmakers as a means to advance this work and improve patient care. A

deeper analysis of the Public Opinion Strategies data— which was captured in 2 surveys, one geared toward patients, survivors, and family caregivers with an oversampling to assure minority representation, and another of oncologists— will be addressed in a future, peer-reviewed article.

To learn more about the working group and next steps, visit [NCCN.org/policy](https://www.nccn.org). Join the conversation online with the hashtag #ElevatingCancerEquity.

New NCCN Guidelines Analyze Evidence for Cancer Type Found Almost Exclusively in Children

NCCN has published its first-ever treatment guidelines for children with a solid tumor cancer type in the United States. The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines) for Wilms Tumor (Nephroblastoma) contain evidence-based expert consensus recommendations for how to maximize benefits and minimize side effects for managing a rare type of kidney cancer that typically occurs in children aged ≤ 8 years. These guidelines contain crucial information, particularly for care providers who treat a full spectrum of pediatric tumors, hematologic malignancies, and other blood disorders (such as sickle cell), due to the relative infrequency of all of these conditions. The guidelines also include important diagnostic information for primary care pediatricians to help ensure this cancer is caught early enough to benefit from high curability rates, and managed in a way that reduces side effects.

“Wilms tumor is rare, but highly treatable,” says Daniel M. Green, MD, of St. Jude Children’s Research Hospital, Vice-Chair of the NCCN Guidelines Panel for Wilms Tumor. “It’s important that we make sure these guidelines and knowledge of the signs and symptoms of Wilms tumor reach primary care physicians and pediatricians so they can help catch it early and save lives.”

“Keeping up with all of the current literature for the broad array of children’s oncologic and hematologic malignancies can be daunting. The NCCN Guidelines for Wilms Tumor provide compiled, annotated, and vetted recommendations for diagnosis, treatment, and follow up—in keeping with NCCN’s goals for streamlining and improving care so that patients can live better lives,” said Elizabeth Mullen, MD, of Dana-Farber/Boston Children’s Cancer and Blood Disorders Center, another Member of the NCCN Guidelines Panel for Wilms Tumor. “People say Wilms is a ‘good cancer’ because it has a 90% cure rate but that still means too many children don’t survive. I won’t feel okay until we get that cure rate to 100%—with optimization of long-term wellness, fertility, and psycho-social wellbeing.”

“The treatment of Wilms tumor is guided by the results of a series of sequential clinical trials that started back in the 1960s,” explained Frank M. Balis, MD, Professor of Pediatrics, Louis and Amelia Canuso Family Endowed Chair for Clinical Research in Oncology, Children’s Hospital of Philadelphia/Abramson Cancer Center at the University of Pennsylvania, who chairs the NCCN Guidelines Panel for Wilms Tumor. “Treatment approaches have been refined through these clinical trials, making treatment selection more and more complex and nuanced. Management should be determined by clinical and biological risk factors, the presence of unilateral (single kidney) versus bilateral (both kidneys) disease, and patient response to treatment. The NCCN Guidelines for Wilms Tumor present clinicians with the latest evidence for the management of children with Wilms tumor based on their individual clinical and molecular profile.”

“NCCN Guidelines for Wilms Tumor shares information for earlier, safer diagnosis and treatment in effort to reduce disparities; doctors warn of late diagnosis as a possible impact from the COVID-19 pandemic.”

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The NCCN Guidelines for Wilms Tumor present clinicians with the latest evidence for the management of children with Wilms tumor based on their individual clinical and molecular profile.

Wilms tumor accounts for 5% of all childhood cancers and typically occurs between ages 1 and 5 years. Children often have abdominal swelling and/or a mass with or without other symptoms; however, they often appear healthy, which can delay diagnosis. Wilms tends to be more common among African-American children and is often caught in more advanced disease stages for this population.^{1,2} In rare cases, children with a number of genetic predisposition syndromes may get Wilms tumor; therefore, these children should be screened for Wilms tumor.

“One of the most common ways that Wilms tumor is diagnosed in a child is through detection of an otherwise asymptomatic abdominal mass during a routine well child care exam with their pediatrician,” explained Dr. Mullen. “Now that some parents and guardians have had to delay these appointments, or hold them virtually due to the COVID-19 pandemic, there is concern about the potential of an increase in higher-stage diagnoses.”

The guidelines also contain a warning for primary care physicians to be gentle if they feel a mass in the kidney area, especially if the children appear to be otherwise healthy. Wilms tumor(s) can rupture if touched with too much force, causing the cancer to spread within the abdomen or elsewhere.

NCCN Guidelines are considered the gold standard for clinical direction and policy in cancer management and are the most thorough and frequently updated clinical practice guidelines for any area of medicine.

All NCCN Guidelines can be downloaded and printed for free at [NCCN.org](https://www.nccn.org) or via the Virtual Library of NCCN Guidelines App for mobile or tablet.

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Cancer puts individuals at high risk for severe illness and death from COVID-19.

More than 125 Organizations Send Letter to President Biden Urging Priority Access to the COVID-19 Vaccines for Cancer Patients and Survivors

On February 17, 2021, 127 organizations and cancer centers sent a letter to President Joseph R. Biden, key members of his administration, and leading public health officials at state health departments to underscore the importance of prioritizing patients with active cancer and survivors of cancer when administering the lifesaving COVID-19 vaccines. The broad spectrum of organizations that signed this letter represent laboratory, translational, and clinical researchers; health care professionals; millions of patients with cancer and survivors of cancer; and patient advocates from across the United States.

Recent research has shown that patients with cancer are at increased risk of severe illness and death if infected with the virus. Moreover, patients with cancer often receive frequent in-person care, which increases their risk of exposure to the virus. Certain survivors of cancer also have a higher probability of infection and COVID-related death compared to the general population.

While the Centers for Disease Control and Prevention recognizes that cancer places individuals at higher risk for severe COVID-19, many states are broadening and simplifying the groups eligible for COVID-19 vaccination to speed up distribution of the vaccines. The letter notes that while mass, rapid

vaccination is a worthy goal, the currently limited supply of vaccines means that many who are at high risk for severe illness and death from COVID-19, including patients with cancer and survivors of cancer, may continue to wait in line for many months if high-risk groups are not provided with priority access.

The letter, initiated by the American Association for Cancer Research (AACR), urges the Biden administration and public health officials at the state level to continue to prioritize patients with cancer and survivors of cancer in statewide vaccination plans.

Patrick Delaney Named New Executive Director of NCCN Foundation

NCCN has announced the appointment of Patrick Delaney as incoming Executive Director for the NCCN Foundation. Delaney has previously held leadership roles with the Juvenile Diabetes Research Foundation, American Red Cross, and American Cancer Society. His new position with NCCN will involve fundraising for the NCCN Guidelines for Patients and other resources to empower people with cancer and their caregivers with unbiased expert guidance from the world's leading cancer experts. Delaney will also work with NCCN Foundation Young Investigator Awards Program by funding some of the nation's top next-generation cancer researchers, guiding the course of treatment innovation and advancement.

"Pat brings a wealth of experience with many of the country's biggest health nonprofits," said Robert W. Carlson, MD, Chief Executive Officer, NCCN. "His years of dedication to various health causes make him the perfect choice to lead our important work to empower patients and caregivers with the educational support they need at each step of their cancer journey, as well as fostering the future of cancer research. We are excited to welcome him to the team."

Delaney attended Albright College in Reading, Pennsylvania, before working in the financial services industry. He entered the nonprofit sector in 2000. He has also held numerous board positions and volunteer roles over the years.

"I am thrilled to join the team at NCCN as Executive Director of the NCCN Foundation," said Delaney. "I look forward to supporting initiatives that deliver true insight for people living with the challenges and complexities of a cancer diagnosis so they can take an active and informed role in their care and the care of their loved ones."

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Incoming leader to oversee fundraising to share trustworthy information about cancer for patients and caregivers as well as supporting promising early-career cancer researchers.