

NOT JUST SKIN DEEP

Awareness of pediatric melanoma may improve prognosis for young patients

Guadalupe Garcia, 13, had a mole on his wrist for as long as he could remember. Then, two years ago, the mole began to grow, and a whitish line appeared around the circumference. Fortunately, a physician at an Atlanta clinic was aware that melanoma can be diagnosed in children as well as adults.

After a biopsy confirmed Guadalupe's diagnosis of melanoma, pediatric surgical oncologist Kenneth W. Gow, M.D., at Children's Healthcare of Atlanta, surgically removed the skin cancer. He also performed a sentinel lymph node biopsy (see sidebar on pg. 15) to confirm the cancer had not metastasized.

Today, Guadalupe remains cancer-free, though oncologists at the Aflac Cancer Center and Blood Disorders Service of Children's Healthcare of Atlanta will monitor his progress for at least five years. Guadalupe had never worn sunscreen until he was diagnosed with melanoma. Now, his entire family wears sunscreen whenever they go outside.

"What parents need to understand," said Louis Rapkin, M.D., a pediatric oncologist and hematologist at the Aflac Cancer Center, "is that protecting kids now provides a lifelong benefit. Parents need to make sunscreen, hats and other protective wear a normal part of their family's lifestyle."

Often viewed as an adult disease, melanoma also can occur in children. When identified early, it is almost 100 percent curable. However, not every physician is aware of the unique—and seemingly innocuous—ways melanoma can present in children. This can lead to a fatal delay in diagnosis. Physicians at Children's are working to promote awareness of the disease and encouraging other centers to modify treatment processes to meet the specific needs of children.

A Growing Threat

Melanoma is the most common form of skin cancer found in children. It is also the most dangerous of skin cancers because it is the most likely to metastasize. Left untreated, melanoma may spread to the lymph nodes, lungs, brain, gas- trointestinal tract or liver.

Melanoma affects about seven out of every million children in the U.S., according to 2002 statistics from the National Cancer Institute (NCI). That figure has risen from three per million in 1982. Recent studies have shown increases in other countries as well. In addition, melanoma prevalence has risen among adults, more than doubling in the past 30 years, according to NCI.

Dr. Rapkin said the increase could be due to a number of environmental factors, including depletion of the ozone layer, which blocks the sun's damaging ultraviolet (UV) radiation; increased sun exposure without appropriate protective measures and an increase in the use of indoor tanning facilities by young adults.

Melanoma in adults is typically attributed to a mix of family history and UV exposure. However, most children with melanoma lack a family history of the disease. Children may also lack other risk factors found in adults.

Physicians aren't certain of the risk factors for pediatric melanoma, Dr. Rapkin said. Some studies suggest an association between increased risk and Caucasian race, female gender, increasing age and exposure to UV radiation. Some experts, however, believe that skin damage from sun exposure takes a long time to develop into cancer, and usually can't be detected until after childhood.

A Different Lesion

Melanoma is a cancer of melanocytes, the skin cells found in the epidermis. Healthy melanocytes produce a protective pigment known as melanin, which gives skin its color and protects the deeper layers of skin from the sun's harmful rays. Most melanomas continue to produce melanin, which gives the lesion its characteristic brown and black appearance.

For adults, melanoma often presents as a flat lesion identified by the A-B-C-D signs: asymmetry, border irregularity, colors of mixed black and brown and diameter larger than a pencil eraser. However, according to a study published in *Pediatrics* in 2005, about half of children with melanoma do not follow this pattern. Many present with lesions that are raised, light in color or have well-defined borders.

Unfortunately, this atypical presentation in children may contribute to delays in diagnosis. Even those pediatric

melanomas with a typical appearance may be overlooked until the disease has progressed considerably.

"Changes in color, size or thickness, as well as any irregularities, itching, bleeding or new satellite lesions are all indicative of melanoma," said Dr. Rapkin. While the specific changes experienced may be different for people of different skin colors, the presence of any of these changes or irregularities demands further care by a specialist.

Top-tier Treatment

Biopsy is the primary method of diagnosis for melanoma. A small amount of skin is removed from the lesion and sent to a laboratory for further analysis. If melanoma is diagnosed, the lesion is staged and a treatment plan is developed. Staging is a standardized numerical ranking that takes into consideration the tumor's thickness, how deeply it has invaded the skin and whether the cancer has spread to lymph nodes or other organs.

Surgery is the most common form of treatment and typically involves removing the lesion as well as a 1-centimeter border of tissue around the circumference of the melanoma. If the cancer has metastasized, other treatments may be considered, including chemotherapy or immunotherapy.

The Aflac Cancer Center is one of only a few centers in the United States that offers comprehensive treatment of pediatric melanoma in a child-friendly setting. "At this point, most pediatric melanomas are treated in adult centers that can't meet the specific needs of children," Dr. Rapkin said.

Even in Atlanta, where pediatric specialists are available, many cases continue to be treated in adult cancer centers. The Aflac Cancer Center physicians are committed to a more gentle approach that is customized for children. For example, while most adults receive only a local anesthetic during biopsy, at the Aflac Cancer Center, patients typically receive general anesthesia in an effort to lessen the child's fear and pain.

Dr. Rapkin's goal for the future is to build bridges between the Aflac Cancer Center and other Atlanta centers such as Northside Hospital and the Emory Winship Cancer Institute, so that, through referrals, consultations and communication, every child can receive the benefit of the experience and knowledge at the Aflac Cancer Center. 

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On the Lookout: How to Recognize Melanoma

The first rule in screening for melanoma is to look for irregularities, referred to by some as The Ugly Duckling Method. Instructing parents to watch for moles that look completely different from other moles on their child's body may be the first and simplest step toward more timely diagnoses. While there are no clear screening guidelines for pediatric melanoma, recommendations include:

- Monitoring moles by taking photos to track growth.
- Screening for the A-B-C-D signs: asymmetry, border irregularity, colors of mixed black and brown, and diameter larger than a pencil eraser.
- Removing growths and moles that are changing.
- Performing biopsies on moles and growths that have been removed.
- Consulting an oncologist who specializes in melanoma if the condition is diagnosed or results are inconclusive.

Heading the Sentinel

Sentinel lymph node mapping is an important technique used in staging melanoma. It results in more accurate identification of potential metastases and can ultimately reduce the amount of surgery required.

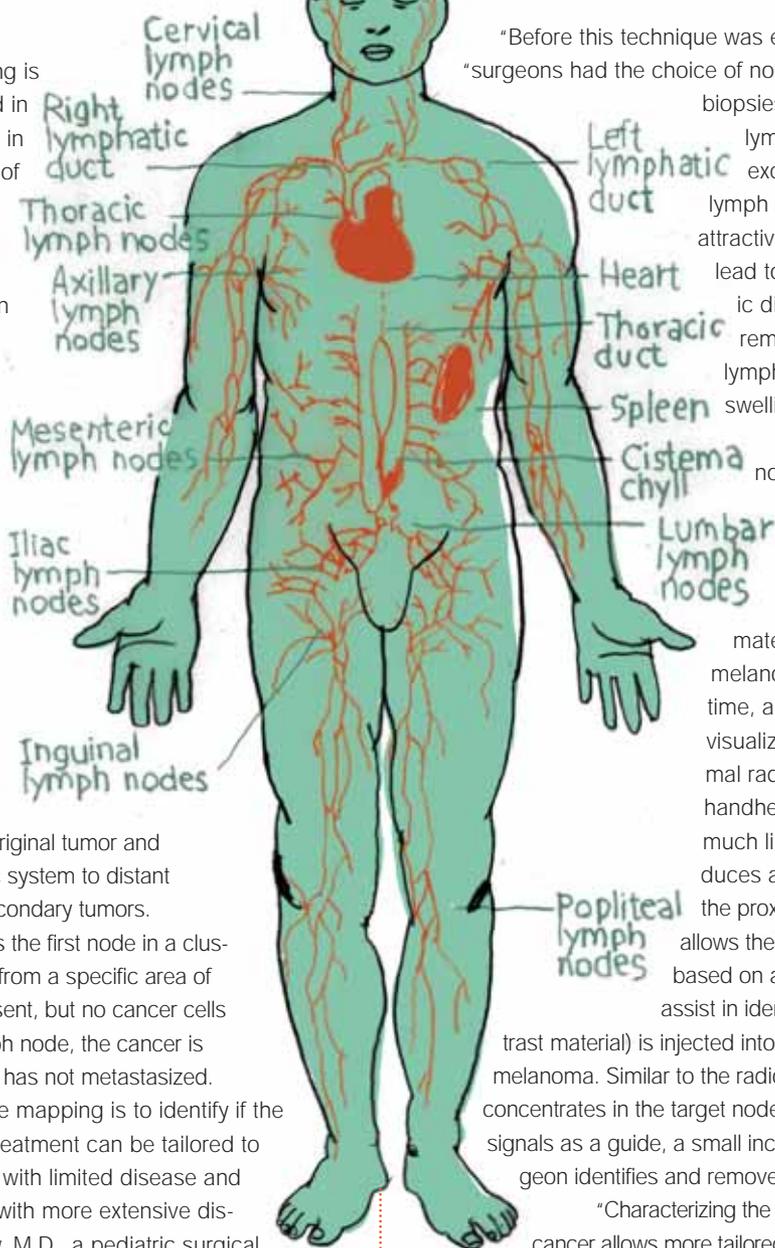
The lymphatic system is an integral part of the body's defense against infection. Its vessels contain a fluid, known as lymph, that transports cells used in the immune response as well as foreign material such as bacteria and cancer cells. Lymph nodes are small, bean-shaped nodules located in clusters along the blood vessels. The nodes produce the immune-response cells and filter out foreign material. During metastasis, cancer cells can break off from the original tumor and spread through the lymphatic system to distant parts of the body, forming secondary tumors.

The sentinel lymph node is the first node in a cluster that receives cancer cells from a specific area of the body. If melanoma is present, but no cancer cells are found in the sentinel lymph node, the cancer is deemed to be local, that is, it has not metastasized.

"The goal of sentinel node mapping is to identify if the cancer has spread so that treatment can be tailored to minimize therapy in patients with limited disease and intensify therapy in patients with more extensive disease," said Kenneth W. Gow, M.D., a pediatric surgical oncologist at Children's Healthcare of Atlanta.

For example, melanoma that is diagnosed on the arm will likely first travel to the lymph node cluster in the armpit as it attempts to metastasize. There are approximately 25 lymph nodes in the armpit, and in each patient the precise lymph node that drains the area of the body affected by the cancer is different. Microscopic assessment of the target lymph node allows the oncology team to determine if the cancer has spread.

The method allows staging without needing to remove a large section of the lymphatic system—a procedure that can cause dangerous complications such as extreme swelling or immune system deficiencies.



"Before this technique was established," said Dr. Gow, "surgeons had the choice of not performing lymph node biopsies, randomly choosing lymph nodes to remove or excising a large number of lymph nodes. None of these were attractive options as they could lead to misdiagnosis of metastatic disease. In addition, the removal of a large number of lymph nodes could cause swelling of an extremity."

During sentinel lymph node mapping, two markers are used to visualize the lymph node (or nodes) that have been affected. First, a weak radioactive material is injected around the melanoma. After a short period of time, a nuclear scan is used to visualize the node with the maximal radioactive signal. Secondly, a handheld gamma probe, which, much like a Geiger counter, produces a clicking sound based on the proximity of radioactive material, allows the surgeon to identify the node based on an audio signal. To further assist in identification, a blue dye (contrast material) is injected into the skin around the melanoma. Similar to the radioactive tracer, the blue dye concentrates in the target node. Using the audio and visual signals as a guide, a small incision is made and the surgeon identifies and removes the affected node.

"Characterizing the aggressiveness of a patient's cancer allows more tailored treatment," said Dr. Gow. "And with sentinel lymph node mapping, surgeons are able to spare children from more extensive operations." Dr. Gow would like to see sentinel lymph node mapping become the standard of care for children with melanoma.

Louis Rapkin, M.D., a pediatric oncologist and hematologist at the Aflac Cancer Center and Dr. Gow are in the process of reviewing all of the pediatric melanoma cases treated at Children's in an attempt to identify the optimal treatment procedures for children. "The ultimate goal," said Dr. Gow, "is to help everyone look after children with melanoma the best way possible. Sentinel node mapping allows us to fine-tune therapy and make treatment safer and more effective for children."