

# Battle Against Nerves

As doctors try to understand neurofibromatosis, patients cope with tumors and people's unpleasant reactions

BY EUNA LHEE  
(SUN REPORTER)

**A**S A TEENAGER, JEANNINE Lancaster started to see spots like mosquito bites on her arms and back. She didn't think much of them then. A decade later, the bumps started to multiply on her face.

The bumps are neurofibromas, or tumors, which have severely disfigured her appearance over the past 18 years. Now, at age 43, Lancaster can't escape the pointing and staring when she goes out of her home. In these situations, she smiles and hands strangers a card:

"If you're curious, I have neurofibromatosis."

Commonly known as NF, the genetic disorder produces benign tumors within and around nerves. The tumors, which can be throughout the body, squeeze the nerves and interrupt signaling in the nervous system. Some people may not show visible bumps but instead suffer from internal effects. Learning disabilities or bone deformities may result through mechanisms not yet understood.

Megan Leaf, a special-education aide



Jeannine Lancaster says the bumps on her face have made going out difficult.

in Baltimore's public schools, doesn't suffer any physical disfigurement. For most of her life, she has had learning disabilities and problems with her vision and coordination.

"I don't look like as if anything is different with me, but I might have trouble saying or seeing something," said Leaf, 27. In elementary school, she had trouble seeing the blackboard and needed the help of a paraprofessional to copy notes.

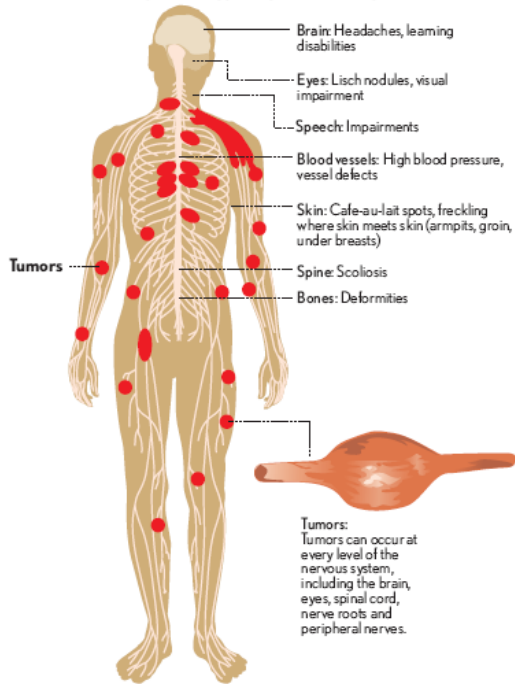
Doctors say the variability of symptoms makes NF difficult to diagnose.

"NF is a common genetic condition, which is underrecognized by both physicians and the public," said Dr. Jaishri Blakeley, director of the Comprehensive Neurofibromatosis [Please see TUMORS, 3C]

## NF CAUSES TUMORS ON NERVES

Neurofibromatosis is a genetic disorder in which tumors grow along nerves or within nerve tissue. It often causes weakness, sensory loss, pain and disfigurement.

How NF-1, the most prevalent type, may affect the body



Source: Neurofibromatosis Inc.

## FROM THE COVER

# Neurofibromatosis: a battle against nerves

**TUMORS** [From Page 1C]

Center at Johns Hopkins Hospital. "All those patients don't have a voice and often feel very self-conscious and misunderstood."

Lancaster and Leaf are among an estimated 100,000 Americans with NF, which occurs in one out of 3,000 individuals. According to the Children's Tumor Foundation, an advocacy group, this makes the condition more prevalent than cystic fibrosis, hereditary muscular dystrophy, Huntington's disease and Tay-Sachs disease combined.

There is no cure for any of the three subtypes of NF. But surgeons can remove tumors when they become painful or threaten normal bodily functions, such as vision and movement. In rare cases, tumors may become malignant and are treated as other cancers would be, with radiation or chemotherapy.

Lancaster underwent separate surgeries to remove tumors on her lung and neck, which were encroaching on her spine and becoming painful. But the operations have left her in more pain, she says.

"I sometimes feel electricity shooting down my body ... just completely unexpected," Lancaster said. "It lasts for seconds and then goes away."

Even so, Lancaster struggles more with the psychological and social aspects of NF.

"I can't deal with most people ... the rudeness people have; the outright pointing, staring, screwed up face while looking at me," said Lancaster, a retired State Department employee who lives in Suitland. "When I go out, I need to have my guard on. I'm ready for a comment or a look, and that's why I have my cards."



Jeannine Lancaster holds a photo of herself as a teen. A decade later, her spots began multiplying.

GLENN FAWCETT [SUN PHOTOGRAPHER]

Most people are unfamiliar with NF, she says, and they often ask if the bumps are mosquito bites. But from time to time, she hears grossly offensive comments.

"On Halloween, a kid asked me if I had a mask on," Lancaster said. "And that really hurt my feelings."

To improve her appearance, dermatologists removed several tumors from her face in 2006. But they just grew back, Lancaster

said.

Doctors usually discourage surgery for cosmetic reasons.

"It's not recommended procedure because there are just too many to remove," Blakeley said. "And it may not be worth the risk of sensory and nerve damage."

Doctors are trying to understand the symptoms and genetics of NF and the disorder's implications on a patient's life.

crimination," Blakeley said. "They might not have a life-threatening form, but every day is like being a cancer survivor for them."

Leaf, like Lancaster, finds it difficult to deal with people's reactions. "When I bring up NF, people are not accepting of that," she said.

She underwent surgery 14 years ago to remove a tumor growing on her cerebellum, the region of the brain that controls motor function. The Charles Village resident is unable to drive or even walk in a straight line, so she depends on public transportation. Despite these challenges, Leaf recently received a master's degree in teaching at the College of Notre Dame.

"My mother has taught me that I am no different from everyone else and has always strived to make me independent," Leaf said.

Many patients worry about the possibility of passing on the condition to their children. According to the Children's Tumor Foundation, each child of an affected parent has a 50 percent chance of inheriting the gene.

Leaf's family has no history of the disorder, but it has touched at least three generations in Lancaster's family. Her mother had the same bumps on her face. Doctors also diagnosed Lancaster's 18-year-old daughter and 13-year-old son when they showed numerous cafe-au-lait spots on their skin.

"I just think we need to find a cure for this," Lancaster said.

NF research is managed primarily by the Defense Department at the direction of Congress, who has reduced spending on NF from a peak of \$25 million in 2005 to \$8 million this year. Researchers and advocates, such as John Risner, the Children's Tumor Foundation president, express concerns. "This is money well-spent, and other programs are not seeing the same

"The researchers focus on the illness, but the patients focus on dis-

cuts," he said.

To raise awareness, the foundation plans to host the first national symposium in Washington in April, when scientists, patients and family members are expected to come together to discuss the latest research.

Current NF studies focus on the triggers of tumor growth. Scientists speculate that hormone fluctuations may be one of them. Lancaster is among the patients who saw their tumors grow and multiply during puberty and pregnancy.

Scientists add that research may shed light on other types of tumors, including malignant ones.

As a Hopkins surgeon who has done extensive work with NF, Dr. Allan Belzberg said he is frequently approached by patients who hope he can remove other types of tumors without damaging the nerves.

"If we try to understand this disease, there will undoubtedly be other spinoffs that will help us in other areas of medicine," Belzberg said.

In addition, several multicenter Phase II drug trials are under way on medications that may stop or reduce tumor growth. Development of successful drugs might give patients more control over their conditions. Treatment could lead to fewer surgeries and reduce the chance of tumors becoming malignant.

Lancaster said she hopes to attend the Washington conference, where few explanations will be needed if she meets a stranger. Until then, she said, she's not bitter when passing a card.

"I'm happy to educate," she said. "But I'm tired of explaining myself every day."

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Megan Leaf, who is unable to drive or even walk in a straight line, recently earned a master's in teaching at the College of Notre Dame.  
PHOTOS BY GLENN FAWCETT [SUN PHOTOGRAPHER]

Earlier in the summer, I reported a story on Fran Visco, a breast cancer survivor who lobbied for more research funding and became one of the pioneers of the Congressionally Directed Medical Research Programs in the Department of Defense's budget. After the story was published, I received a study from the Children's Tumor Foundation, an advocacy group, regarding the prevalence of neurofibromatosis (NF) in the American population. I had never heard of this genetic condition before and wanted to investigate further, since the Defense Department also gave millions of dollars each year to fund NF medical research.

I quickly found that NF is a complicated and poorly understood condition. Most often, it is misdiagnosed. Some patients have mosquito-bite-like bumps on their arms and faces, while some have no visible symptoms at all and, instead, suffer from learning disabilities. In addition to these physical manifestations, patients were often discriminated against in their daily lives because people assumed that the condition was highly contagious. I decided to concentrate my story on the psychological effects of NF.

I found my two main characters, Jeannine Lancaster and Megan Leaf, through the Neurofibromatosis Center at Johns Hopkins Hospital. The two women shared their lives and showed me their daily struggles through photographs and videos. I also spent time with NF specialists at the Center to understand the mechanisms of the condition and to help design the informational graphic.

After the story ran as the centerpiece in the Health Today section, I was touched to hear from my main characters that the story changed their lives for the better. Amazing... the power of medical journalism!

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