Deaf students confront sight-threatening ailment

By TOM GUARISSCO
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Michelle Eaton is trying to finish her degree in fine arts and design at Delgado Community College in New Orleans and then hone her skills before she goes blind.

Eaton is 20 years old, a graduate of the Louisiana School for the Deaf and a victim of Usher syndrome. The genetic flaw rendered her deaf since birth, and one day will rob her of her sight.

“I’m not going to worry about it,” Eaton said. “I think it will happen many years later. I don’t worry about it — it could mess up my eyes if I have too much pressure.”

After graduating, Eaton hopes to attend the National Technical Institute for the Deaf in New York, and then open her own design business.

“I hope I will be cured of Usher syndrome,” she said.

People with Usher syndrome are born deaf, and their vision gradually erodes. Its root cause is closely related family members generations ago having children together, which was a common occurrence in many places years ago, including south Louisiana, when people lived in remote areas.

A group of Swedish students and a Finnish student spent last week visiting with pen pals at the Louisiana School for the Deaf in Baton Rouge. The young people live thousands of miles apart, but they share the same genetic abnormality.

The descendants of parents who were closely related may be carriers of the flawed gene. The gene remains dormant unless they have children with another carrier of the Usher syndrome gene. In that case, each child has a one in four chance of being born with Usher.

At present, scientists can’t pinpoint how closely related those parents had to be — or even if there are generations back — to pass on the gene.

People can research their family history for anyone who was born deaf and became blind, which is a strong indicator of Usher syndrome. At present, there are no simple tests. However, thanks to research in Louisiana and elsewhere, Eaton and other sufferers might one day be cured. LSU Medical Center in New Orleans is playing a key role.

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Helen Davidsson of Sweden enjoys Bluebonnet Swamp last week as Josh Broussard, right, of Louisiana School for the Deaf peers skyward.

Davidsson was in a visiting group of Swedish deaf students who share a rare genetic cause of deafness with some students at the Baton Rouge school. Behind them, from left, are Swedish students Helen Lindberg and Maria Nyman, and interpreter Desiree Wilkund.
A year ago I was saying that within a year we might find the gene, but we don't have it yet," said Bronya Keats, an LSU Medical Center geneticist. "It's a very tricky one to find."

Keats is an expert on Usher syndrome, and studies it under a grant from the Foundation Fighting Blindness.

LSU is involved because Usher is common in Louisianians of Acadian, or Cajun, descent. Researchers believe one in 70 Cajuns is a carrier of the gene.

"We're trying to find this gene that causes the more severe form in the Acadian population," Keats said.

They have narrowed their search to a small area of genetic material, but have not yet identified the specific Usher syndrome gene.

The work is tedious.

"It's a matter of working through all the DNA in the region and looking for a mutation that might be responsible, then checking it out and ensuring it's only in (Usher syndrome) patients," Keats said.

"Once we identify it we have no idea what kind of protein will be encoded by this gene, so then we need to characterize the protein and see what it's doing," Keats said.

"This is likely to be many more years' research."

Researchers hope that eventually a simple blood test will identify the Usher syndrome gene, and genetic therapies may be developed to curb its effects.

Preventing deafness will be extremely difficult, Keats said, but researchers may one day figure out a way to reduce the visual impairment Usher syndrome causes.

 Luck is how educators at Louisiana School for the Deaf first found Usher syndrome among their students in 1977.

Some of the students persisted in describing in sign language - with a symbol they invented - that some of them suffered from tunnel vision. This lack of peripheral vision is a common first phase of blindness in many who suffer from Usher syndrome.

Michelle Eaton, a 1998 graduate of Louisiana School for the Deaf, talks with LSD graduate Jose Perez over lunch at Mulatte's last week. Eaton, who attends community college in New Orleans, spent time with a group of deaf students from Sweden.

drome.

Faye Melancon, a recently retired counselor from the school, remembers how staff stumbled onto the finding after an administrator suggested Usher might be the cause of blindness among students. The administrator had seen the same syndrome in an isolated community in Florida where he had worked previously.

"None of us knew but it had been going on all these years - people knew they would eventually go blind but they didn't know why," Melancon said.

We felt so helpless. As a counselor working with the kids I had to explain to them life is going to be real tough, and you won't be able to see, able to drive," Melancon said.

"You had to tell them terrible things about their lives - try not to marry someone too close to you."

After a schoolwide screening, they learned that one in 10 suffered from Usher. For the first time, educators knew what was causing the deaf students' blindness.

Since the discovery of Usher syndrome at the Baton Rouge school 22 years ago, educators have been helping the students and their families who carry the gene. A special support group has met since then, helping the students cope with their complex of challenges in daily life.

The 10 percent of the school's students who have Usher today meet regularly. A few years ago, one class dubbed themselves Dice 12 describing the dozen who felt lucky to get out of regular classes for their own gatherings. Later classes have named themselves Lucky 13 and the Bravehearts.

The school also counsels families of those carrying the genes. The incidence of Usher has dropped somewhat, but it is still a threat.

Last week was a rare treat for the Swedish visitors and the Usher students at the School for the Deaf. Between the Cajun food and cultural visits, they hardly spoke about the ugly truth that links them - that eventually they will all probably go blind. Usher syndrome sufferers usually lose their sight by middle age.

Connie Lindfors knows several languages but he has never heard a word in any of them.

He is a 20-year-old Swedish student studying humanities in that country's only high school for the deaf.

"Before I came here, I thought the school here and life for deaf students would not be as good as in Sweden," Lindfors said through interpreters. "Now I have changed my mind."

Thanks to interpreters and translators, the Swedish-signing students got along well with their U.S.-signing friends at the Louisiana school.

Lindfors made new friends and polished up his American sign language, one of several he is learning.

His sister, Helen Davidsson, especially liked the kitchen at the Louisiana School for the Deaf, and its custom tools and utensils for deaf and blind students. She plans to pursue a career in catering and management.

Maria Nyman of Finland spent a morning at the State Fair looking after a first-grader from Louisiana School for the Deaf. Nyman has a knack for caring for young children, and so the 20-year-old plans to be a first-grade teacher when she finishes college.

The students rarely dwell on or worry about the fact they have Usher.

"I don't think about it today," said Nyman.

Lindfors agreed.

"I don't worry about the future, I worry about what I can do now," Lindfors said.

Perhaps the most important part of the visit was simply seeing the United States.

"When they lose their sight they'll still be able to picture it - it will give them memories," said Catharina Bjuhr, an adviser to the Swedish students. "It puts a memory in their backpack."