Delores Hightower, 56, had been forced to use a walker and then a wheelchair to get around. The
wheelchair, she said, was almost a relief. Hightower has Friedreich's ataxia, a genetic disease that
appears 2 1/2 times more frequently among people of Acadian ancestry than in the population at large.

New money, new hope pour into La. gene research

Editor's Note: This is the first of a two-part series about Friedreich's ataxia and Usher's syndrome, gene-linked diseases that appear with higher than average frequency among people of Acadian ancestry.

Bill Decker
Staff Writer

LAFAYETTE — Delores Hightower first noticed the problem when she was 17.
A tall, slender teenager who lived in Vermillion Parish and liked to draw, she started suffering through episodes of extreme fatigue and began having trouble walking up stairs.

"I used to be able to draw well. Now I do well to write my name," said Hightower, who lives in Lafayette.

"All the little episodes started coming closer and closer together, until it got to where it was all the time."

By the time she was out of her 30s, Hightower had been forced to use a walker and then a wheelchair to get around. The wheelchair, she said, was almost a relief.

"That walker was a killer," Hightower said.

"Now at least I can do things. The walker was so cumbersome."

Yet for all the difficulties Hightower, now 56, has lived through, she counts herself lucky.

Hightower — a widow who was originally Delores Mouton — has Friedreich's ataxia.
Friedreich's is a genetic disease that most often appears in children at age 8 or 9, eventually crippling them and ending their lives before age 40.

The disease appears 2 1/2 times more frequently among people of Acadian ancestry than in the population at large. Hightower, a spokeswoman for the National Ataxia Foundation, said about 50 Friedreich's suffers live in Acadiana.

"For the younger people, it's harder to handle because they have to put up with what the other kids say," Hightower said. "And it's devasting for a family. It just wrecks them. The parents blame each other even though they had nothing to do with it."

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Cure

But help may be on the way. Louisiana governments, businesses, universities and medical schools are mounting new efforts to study genetic diseases, and they're backing up their efforts with at least $9 million in research funding.

Of that money, $1 million is devoted specifically to LSU Medical Center's efforts to study Friedreich's and Usher's syndrome, another genetic disease whose sufferers are often born deaf and eventually become blind.

Dr. Bronya Keats, who directs the LSU Medical Center's molecular and human genetics program, said the focus will be on studying the chemical interactions that bring about the diseases with hopes of finding a way to slow their progress or, in the case of Friedreich's, preventing the onset of symptoms.

Officials also hope a new concentration on biomedical and genetics research in Louisiana will create high-tech jobs here.

Genetic puzzles

Keats, who grew up in southern Australia, arrived at the LSU Medical Center in 1982 and immediately became involved with the study of Friedreich's ataxia. A Louisiana woman called her children about the disease, and three of the six children in her family were diagnosed with Friedreich's.

"I told her she'd probably see several people with Friedreich's in the same neighborhood," Keats said.

The woman eventually found five other families in which at least one member had Friedreich's and who lived nearby.

Since then, Baylor University researchers have identified the recessive gene that causes Friedreich's.

The gene linked to Usher's syndrome is more elusive. Keats said scientists have found the gene that makes the disease more prevalent in Acadian descendants, but "we know there are eight different genes that cause Usher's syndrome. Why that many genes would cause that particular clinical phenotype is unknown."

The $1 million federal grant will further the research effort by allowing Keats to use the "telemedicine" telecommunications network to research sites doing relevant work, to expand the research facility, expand education programs and attract researchers to the LSU Medical Center.

"Without your support," Keats told the House Subcommittee on Appropriations in 1997, "we will be back where we started on the Hill."