'Recessive' genetic conditions can appear from nowhere

Editor's Note: This is the second of a two-part series about Freidreich's ataxia and Usher's syndrome.

Bill Decker
Staff Writer

LAFAYETTE — Two gene-linked diseases that appear with higher-than-average frequency among people of Acadian ancestry can have devastating consequences for victims and their families.

And they can strike without warning, sometimes in families where the diseases haven't appeared for generations.

The diseases are Freidreich's ataxia and Usher's syndrome.

Freidreich's most often appears at age 8 or 9 and eventually impairs the patient's ability to walk and talk. It also shortens their lives.

Usher's victims are most often born unable to hear and, as they grow up, they become blind because their retinas degenerate.

Both diseases occur 2 to 5 times more frequently among Acadian descendants than in the rest of the population, according to Dr. Bronya Keats, director of the Molecular and Human Genetics Center at the LSU Medical Center in New Orleans.

The seemingly unpredictable way the diseases appear comes from the fact that the diseases are caused by recessive genes.

Traits carried by recessive genes appear only when a baby inherits the recessive gene from both parents. People who inherit the recessive gene from only one parent are unaffected, but they can pass the gene on to their children.

Statistically, each child born to two carriers has a one in four chance of developing the disease, a two in four chance of being a carrier, and a one in four chance of being neither a victim nor a carrier.

Genetic diseases affecting Acadians are sometimes discussed in hushed tones, as though the conditions were the result of familial inbreeding. But that's not the case, Keats said.

The diseases' frequency has more to do with the fact that Acadian people are more likely to carry the Usher's or Freidreich's genes and that Acadians have historically been a distinct geographic, cultural and demographic community in

Please see GENETIC on Page 8A

Dr. Bronya Keats, director of the Molecular and Human Genetics Center at the LSU Medical Center in New Orleans, focuses on genetic diseases affecting Acadians.

Continued from Page 1A

Genetic

Louisiana. It's not that Acadians are marrying their first or second cousins. We're all carrying recessive genes," Keats said. "In general, it's unlikely that we'll marry somebody else with the same recessive genes.

"It's just that any two Acadians who get married have a greater chance of passing the genes on to their children."

Freidreich's and Usher's may not be the only diseases with a genetic link that appear with higher-than-usual frequency in South Louisiana.

Vivian Chen, director of the Louisiana Tumor Registry in New Orleans, said doctors identified a higher-than-expected incidence of pancreatic cancer among Acadians as early as the 1970s. But they're not sure whether the frequency is the result of a genetic link or some environmental factor like diet or smoking.

White men in Louisiana are about one-third more likely than men in the rest of the country to develop lung cancer. And black men in Acadiana have one of the highest lung cancer incidence rates in the world, Chen said.

"The predominant risk factor is smoking," Chen said. "Ninety percent of lung cancer can be attributed to smoking and out of every 10 people who smoke, one will develop lung cancer."

"What we don't know is why the one person gets lung cancer and the other nine don't. There is some discussion that it may be a genetic-environmental link." Chen said the answer may be genes that cause the body to produce enzymes that either break down chemicals into cancer-causing substances or slow down the process by which the body cleans out potential carcinogens.