Beverly LeBlanc visits with her grandchildren, Natalie Thompson, 5, left, and Alanina Thompson, 2, as they have their lunch. LeBlanc has inherited ataxia, a neurological disorder that is transmitted genetically.

The ‘Cajun disease’

Inherited ataxia is common among people of Acadian descent

Beverly LeBlanc considers herself fortunate. In 1997, when her speech became slurred and she kept losing her balance, her daughter, a registered nurse, thought she was having a stroke and insisted she see a neurologist. "He ordered an MRI and from the MRI, he determined that it was an inherited ataxia. I was lucky. Most people go from doctor to doctor," said the 57-year-old Abbeville resident.

Mary Romero, now 58, was diagnosed in 1974. "When I got pregnant, I was wobbling from side to side. They kept telling me I had inner ear problems," said the 58-year-old New Iberia resident. "A doctor finally sent me to a neurologist in New Orleans."

There are dozens of types of inherited ataxia, a neurodegenerative condition that involve the brain, and often, the spinal cord, said Dr. Michael Wilensky, a New Orleans neurologist who specializes in ataxia. The word "ataxia" simply means "unbalanced," Wilensky said, and the term used by itself can refer to symptoms of a host of other neurological diseases, such as muscular dystrophy and multiple sclerosis.

Inherited ataxia is rare, but there is an inordinate concentration of it in southwest Louisiana because it is most common among people of northern European descent, especially those of Acadian ancestry.

“We’ll see balance problems, coordination problems," Wilensky said. "Patients can have double vision, blurred vision and whole host of problems, depending on what type they have. There are also associated symptoms. They may have neuropathy, slurred speech."

Associated problems for some types of ataxia can be difficulty breathing and heart problems.

The condition is difficult to diagnose because there is no specific test for inherited ataxia, save for DNA testing. Most of the time, it’s a matter of eliminating other neurological diseases, such as multiple sclerosis, that have similar symptoms.

There is no cure — not even treatment for the symptoms, as yet, although a few rare ataxias may respond to Vitamin E therapy, Wilensky said.

See DISEASE on Page 4C

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ONLINE
For more information on ataxia, log on to the National Ataxia Foundation's Web site at www.ataxia.org or the Louisiana Chapter at www.angelfire.com/le/ataxia/chapter/
Disease

LeBlanc's ataxia has progressed to the point that she needs the assistance of a walker — or her husband's arm, in public — to get around.

"I'm still cooking," she said, "I clean house. I'm still walking — just a little slower."

Romero's ataxia has progressed further. "I wash a little bit of dishes and can do my washing," she said, "but that's about it. I'm afraid to cook, because my hand jerks and I'm afraid stuff will fall on me.

"I can't walk anymore," she added. "I don't drive. I'm sure you can tell by my speech it's not like it used to be."

Although the disease is debilitating, it does not cause pain, Wilensky added.

What has changed over the past 10 years is the ability to identify the disorder, Wilensky said. "We now have about 15 or 20 actual genes that have been localized — specific genes for specific ataxia syndromes."

This is important, Wilensky said, so families can get genetic counseling to determine whether their children or grandchildren are likely to get the disease.

"In the recessive type, both parents carry an abnormal gene, but neither one has it. It's more common in population groups that have intermarriage. The child has a 25 percent chance of getting it."

"In the dominant form, usually one parent has it and the child has a 50 percent chance of getting it."

Friedrich's ataxia, a recessive type, is most common in the Cajun population, Wilensky said.

The diagnosis is often missed because patients are unaware the condition exists in their families, sometimes, because the symptoms may have been milder in previous generations.

About 10 years ago, Wilensky noticed a pattern. In some forms, the symptoms may not have manifested themselves until the parent was advanced in age, when shakiness or difficulty getting around could be expected.

"They think it's just old age, but (ataxia) may come on earlier and earlier in successive generations than in the parent generation. Six months later, geneticists proved the theory," Wilensky said.

While it's important to pinpoint the gene, genetic testing is expensive, said LeBlanc, who is active with the Louisiana Chapter of the National Ataxia Foundation.

"We do clinics every year," she said, adding that the clinics provide free testing for the trait to Acadiana residents. "We're trying to get a grant to hold another free testing clinic."