USHER'S SYNDROME

A rare genetic defect that causes deafness and a gradual loss of vision is much more prevalent in Louisiana than the rest of the nation.

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Features writer

It took a series of car accidents and several other incidents before Linda admitted something was wrong with her vision.

Bill knew she couldn't see at night but thought other people simply had memorized where things were, and he couldn't remember as well.

Both Linda and Will have Usher's Syndrome, a relatively rare genetic defect that causes deafness and loss of vision. For deaf people, one important sense already is gone and the dependence on other senses — particularly sight — increases. But people with Usher's Syndrome have lost their hearing and slowly are being robbed of their sight.

The incidence of Usher's Syndrome in Louisiana is higher than the national average. Among deaf people in Louisiana, 15 percent have Usher's Syndrome as compared to the national average of 3-6 percent, researchers say.

There are some theories as to why it's high here. The Acadiana area, with its French settlers, was a closed community. There was the Gulf on one side, a swamp on the other and no interstate highway to link them to other areas. People married, not realizing that they probably had some common ancestors. The gene spread through the community, with increasingly large numbers of people carrying it. The same situation exists in Finland, which also has a high rate of Usher's Syndrome.

For a person to have Usher's Syndrome, someone in the family must pass on the recessive gene that causes it. A person can be a carrier without having the symptoms.

A person with Usher's Syndrome is born deaf or hearing-impaired and also has retinitis pigmentosa — a degenerative eye disease. The retinitis pigmentosa — or RP — eventually causes the person to lose peripheral vision. "Tunnel vision" is the description generally used to describe how a person with Usher's can see. In some extreme cases, the person may lose all sight.

In young children, the first sign of Usher's usually is night blindness. "Parents notice kids groping down hallways to go to the bathroom," one teacher said.

Later, the "tunnel vision" becomes apparent. "Where you or I can see all around us, without turning our head, the person with Usher's can only see directly in front of them," said Marcia Brown, a teacher at Louisiana School for the Deaf. "A person with Usher's may only be able to see like this," she said, using her hands to create a box.

That causes the person with Usher's Syndrome a lot of trouble in getting around and communicating with other people. If you're constantly looking down to see where you're walking, you can't look ahead.

"We had a little boy who used to love to walk back and forth across the bleachers when the PE room was in this building. They built a new PE building, and there were bleachers in it, but they built it so there was a wall jutting out, and he ran smack into it. He never saw it," she said.

Teachers have to be careful not to move things around in the classroom or place things in pathways. "You'll see them falling over things — huge things. Someone will be kneeling, and they'll fall over that person, and you say to yourself, 'Uh, oh, they've got it,'" one teacher said.

Richard Hoffmeyer at Louisiana School for the Deaf said studies have shown that the most rapid deterioration seems to occur in the adolescent and young-adult years. That's particularly hard because it's a time when young peo...
Students converse as Linda Amala teaches.