

Twin Reno girls treated for rare disease

April 16 2009

(AP) -- Twin 5-year-old girls living with a rare disease are among the first in the country being treated for their condition at a Reno hospital after the U.S. Food and Drug Administration granted special permission to use an experimental drug.

Addi and Cassi Hempel of Reno were admitted to Renown Regional Medical Center on Monday to get the experimental treatment for Niemann-Pick Type C disease.

The FDA recently cleared them for intravenous infusions of the drug cyclodextrin, a sugar compound that has been successful in laboratory studies.

The [girls](#) were diagnosed in October 2007 with NP-C. There's currently no known cure for the disease, which also is referred to as childhood Alzheimer's.

It causes cholesterol to build up and get trapped inside the cells, which leads to debilitating neurological and physical problems.

The process eventually slows down cells and leads to cell death. Most patients with NP-C don't live past their teens, according to the National Niemann-Pick [Disease](#) Foundation.

The girls' parents, Hugh and Chris Hempel, said they're optimistic about the use of cyclodextrin - an additive in cholesterol-free foods that studies show also might help combat the HIV virus.

"I'm fairly confident, but the question is about the dosing. They're getting half the recommended dose," Hugh Hempel told the Reno Gazette-Journal.

"The FDA asked us to start slowly. So, there will be proving to the FDA that it's safe," he said.

Dr. Ron Browne of Sun Valley, Idaho has been a liaison between the FDA and some families with children living with NP-C. He said the Hempels got approved for cyclodextrin through the FDA "compassionate use" program. Their condition will be evaluated every few weeks.

"The FDA is usually cautious," Browne said. "There needs to be life-threatening situations where no other option is available."

The girls, who wear diapers, began losing their ability to speak several months ago, Chris Hempel said.

"They used to be able to sing nursery rhymes and say their names but they've regressed, like any neurological condition," Chris Hempel said. "We can understand what they want, and they're still taking in what we're saying."

Chris Hempel said staying positive is the only choice available.

"You have to be positive when you have kids in this situation," she said.

Information from: Reno Gazette-Journal, <http://www.rgj.com>

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Citation: Twin Reno girls treated for rare disease (2009, April 16) retrieved 6 May 2024 from <https://medicalxpress.com/news/2009-04-twin-reno-girls-rare-disease.html>

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