

# Rare brain disease concerns in US after death

September 5 2013, by Holly Ramer

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Public health officials believe one person in the U.S. has died of a rare, degenerative brain disease, and they say there's a remote chance up to 13 others in multiple states were exposed to the fatal illness through surgical equipment.

Dr. Joseph Pepe, president of Catholic Medical Center in New Hampshire, said officials are 95 percent certain that a patient who had [brain surgery](#) in May and died in August had sporadic Creutzfeldt-Jakob Disease.

The disease progresses rapidly once symptoms appear and is always fatal, usually within a few months. But the symptoms can take decades to show up. They include behavior changes, memory loss, impaired coordination and other [neurological problems](#).

Nearly 90 percent of cases of Creutzfeldt-Jakob disease happen spontaneously, when an agent causes [proteins in the brain](#) to fold incorrectly. Because those abnormal proteins can survive standard sterilization practices, there is a small risk of exposure for those who had surgery after the patient who died, Pepe said.

"The risk of exposure is extremely low, but it's not zero," he said.

The hospital has notified eight of its patients who may have been exposed, and hospitals in other states are working to do the same because some of the surgical equipment was rented and used elsewhere, state

public health director Dr. Jose Montero said. He would not identify the other states but said no more than five additional patients were potentially exposed.

Pepe said the potentially exposed patients range in age from mid-30s to mid-80s.

"They took it very well. I don't believe that people were angry or extremely emotionally upset," he said. "We did the best job we could in trying to alleviate their fear."

Worldwide, Creutzfeldt-Jakob affects about one person in every one million each year; in the United States, about 200 cases are recorded annually, according to the National Institutes of Health.

In fewer than 1 percent of the cases, the disease is transmitted by exposure to brain or nervous system tissue, Montero said. Only four cases of transmission via surgical instruments have been recorded, none in the United States, he said.

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