Disease that killed Becky Lockhart described as 'Alzheimer's on steroids'

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Tom Smart, Deseret News

Members of the media gather at the University of Utah Medical Center for a news conference about former House Speaker Becky Lockhart's death Saturday, Jan. 17, 2015, in Salt Lake City. Lockhart, 46, died at her home Saturday from an unrecoverable and extremely rare neurodegenerative brain disease.

SALT LAKE CITY — Just before Thanksgiving, former House Speaker Becky Lockhart came down with what she thought was an inner ear infection that made her dizzy and struggle with balance.

By Jan. 5, she was hospitalized in critical condition, diagnosed with Creutzfeldt-Jakob Disease, a rare and fatal brain disorder that has no cure, no treatment and no mercy for its victims, who experience a rapid onset of symptoms before death.

The disease, commonly called CJD, is extremely rare, with only 300 new cases being diagnosed in the United States each year. Worldwide, there is one incident per every 1 million people.

At a news conference Saturday at the University of Utah Hospital, doctors detailed the strange and rare disorder that took Lockhart at the relatively young age of 46, leaving a grieving husband and three children to grapple with the inexplicable whirlwind of her death.

Dr. Jennifer Majersik, an associate professor of neurology at the U. and one of the doctors who attended to Lockhart, said CJD behaves like Alzheimer's disease on steroids.

Once Lockhart began to experience symptoms, CJD took rapid hold of her, and she succumbed quickly. Most patients can die within a few short weeks, or a couple of months from when it hits. Symptoms include failing memory, behavior changes and impaired coordination. As the disease progresses, blindness occurs, as does weakness in the extremities, involuntary movements and ultimately coma.

Sen. Curtis Bramble, R-Provo, said Lockhart was hospitalized for several days with the disorder and ultimately discharged for hospice care at her home with her family. She died shortly after noon Saturday.

Majersik said Lockhart's body will be tested by the National Prion Surveillance Center. The majority of CJD cases — 85 percent — are classified as sporadic, which means there is no known cause. Another 10 percent to 15 percent of CJD cases come from a genetic mutation that is inherited from a parent.

There is also "variant" CJD — 1 percent to 5 percent of the cases — which can come from contaminated beef or contaminated blood or plasma.

Majersik stressed that CJD is not mad cow disease, though people commonly lump the two together. Mad cow disease only affects cows, while "variant" CJD is believed to come from eating infected beef.

There have been four variant CJD cases in the United States, and all of them had origins traced to the United Kingdom, Majersik said.

Further testing on Lockhart will confirm the type of CJD she had, Majersik said. She added that Lockhart did not present symptoms or circumstances that would suggest she had the variant strain of CJD, and the doctor said she met both of Lockhart's parents, leading her to believe genetics are unlikely to be at fault.

Majersik said there's typically one to two cases of CJD at the university hospital a year. Her last patient came from Colorado.

According to the Centers for Disease Control and Prevention, the risk of getting CJD is higher in older people. The median age at death is 68 years, with the duration of the illness lasting four to five months.

In contrast, victims with variant CJD can live 13 to 14 months and the median age at death is 28.

Classic or sporadic CJD is caused by the spontaneous transformation of normal prion proteins into abnormal prions.

Normal prions are found throughout the body and in the brain. While not essential to life, prions do seem to play a role in helping neurons communicate and transport minerals. Its amino acid chains fold into a mainly helical shape.

In CJD and other prion diseases, the prion protein folds into an abnormal shape and unlike any other known protein, becomes infectious. The misfolded proteins cause other prions to misfold and infected brain cells begin to die, releasing more prions to infect normal tissue.

Dr. Stanely B. Prusiner of the University of California at San Francisco purified an infectious agent uniquely made of protein and called it a "prion." In 1997, he was awarded the Nobel Prize in Physiology or Medicine for his discovery that CJD was caused by prions.

The discovery was groundbreaking because proteins don't contain the genetic material that allow viruses and bacteria to reproduce.

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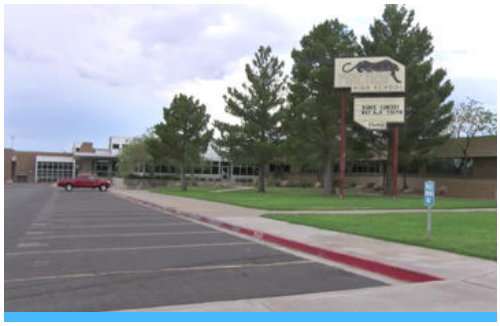

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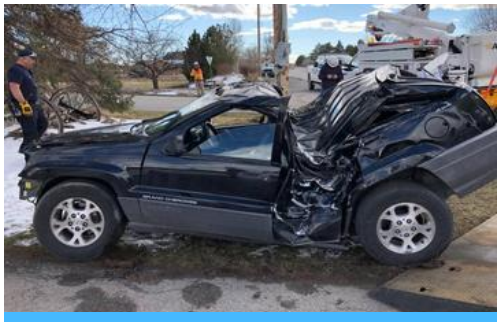

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