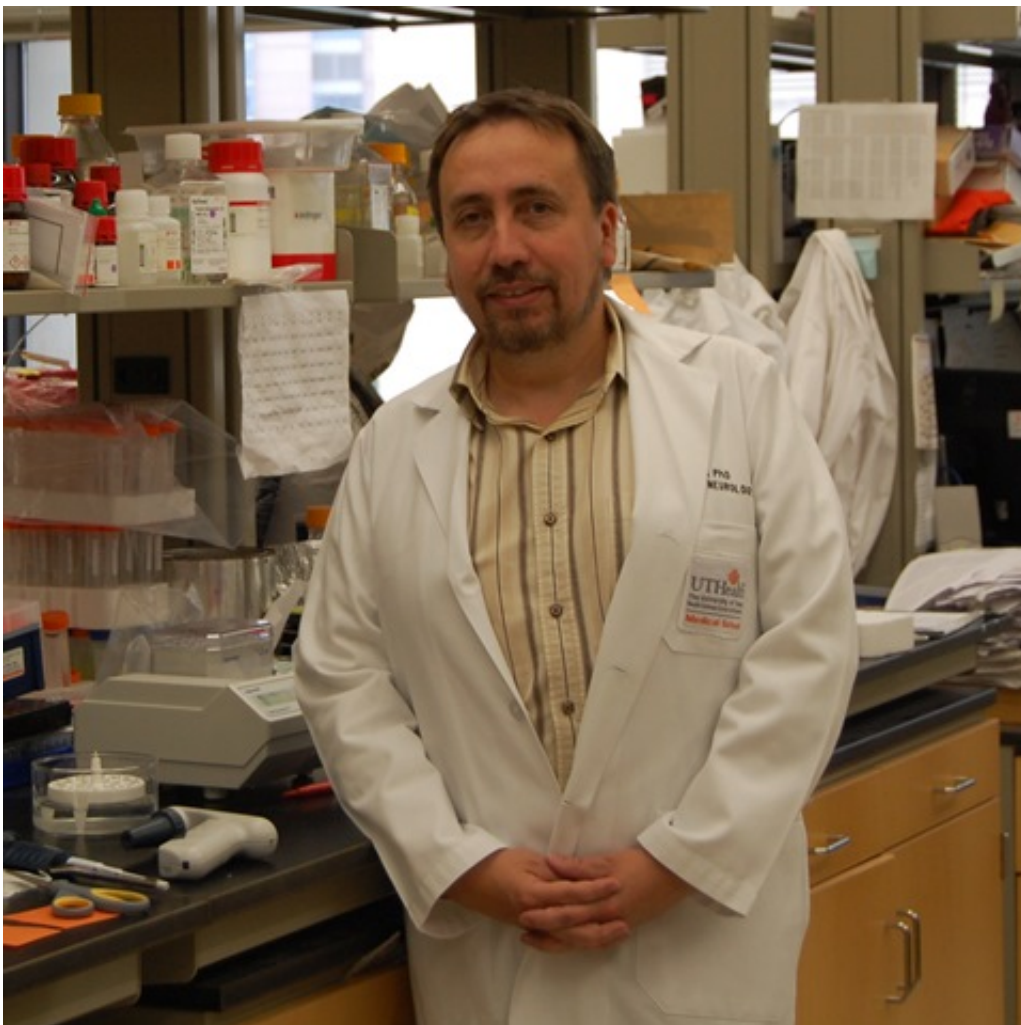


# Researchers find infectious prion protein in urine of patients with variant Creutzfeldt-Jakob disease

August 7 2014, by Deborah Mann Lake

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Claudio Soto, Ph.D., in one of his labs at The University of Texas Health Science Center at Houston. Credit: Deborah Mann Lake, UTHealth

The misfolded and infectious prion protein that is a marker for variant Creutzfeldt-Jakob disease – linked to the consumption of infected cattle meat – has been detected in the urine of patients with the disease by researchers at The University of Texas Health Science Center at Houston (UTHealth) Medical School.

The results of the international study, led by Claudio Soto, Ph.D., professor of neurology at the UTHealth Medical School, are published in the Aug. 7 issue of the *New England Journal of Medicine*.

Variant Creutzfeldt-Jakob disease in humans and [bovine spongiform encephalopathy](#) in animals – also known as Mad Cow disease – are fatal neurodegenerative disorders. There are currently no noninvasive tools available to diagnose the disease and there are no treatments.

Sporadic Creutzfeldt-Jakob disease occurs worldwide at a rate of around 1 new case per million people per year. The variant form is a new disease occurring in people who either ate the beef of cows with bovine spongiform encephalopathy or, in the case of three patients in the United Kingdom, received blood transfusions from asymptomatic infected donors.

The international team of researchers analyzed [urine](#) samples from 68 patients with sporadic Creutzfeldt-Jakob disease, 14 patients with variant Creutzfeldt-Jakob disease, four patients with genetic prion diseases, 50 patients with other neurodegenerative diseases, 50 patients with nondegenerative neurologic diseases and 52 healthy persons.

Soto's laboratory used a protein misfolding cyclic amplification assay, invented in the lab, which mimics the prion replication process in vitro that occurs in prion disease. The misfolded prion proteins were detected in the urine of 13 of 14 patients with variant Creutzfeldt-Jakob disease. The single patient whose urine was negative had been receiving an

experimental treatment of pentosan polysulfate directly into the brain. No misfolded prion proteins were detected in the urine of any the other study subjects, including the patients who had sporadic Creutzfeldt-Jakob disease.

"What could be less invasive than detecting this disease in urine? The fact that we were able to detect just the variant Creutzfeldt-Jakob disease form in the urine is very important. This could lead to the development of commercial technology for diagnosis as well as to determine the safety of donated blood and urinary products," said Soto, who is the director of The George and Cynthia Mitchell Center for Research in Alzheimer's disease and Related Brain Disorders, and founder of Amprion Inc, a biotech company developing the cyclic amplification technology for commercial application.

According to the World Health Organization (WHO), variant Creutzfeldt-Jakob disease affects younger patients, who have a median age of 28 at death, compared to sporadic Creutzfeldt-Jakob disease with a median age of 68. Most patients, after diagnosis of either form, live less than a year before death.

As of June 2, 2014, 177 of 229 people in the world with Creutzfeldt-Jakob disease were from the United Kingdom. A 2013 study published in the *British Medical Journal* has estimated that approximately 30,000 people in the United Kingdom might be carriers of the variant form of the disease.

"This study reports, for the first time, the detection of the abnormal [prion protein](#) in the urine from [patients](#) with variant Creutzfeldt-Jakob disease using the protein misfolding amplification technique pioneered by Dr. Claudio Soto," said co-author James W. Ironside, FMedSci, FRSE, professor of clinical neuropathology at the National CJD Research and Surveillance Unit at the University of Edinburgh. "This

has great potential to allow the development of a highly sensitive and specific non-invasive test that can be used for the diagnosis of variant Creutzfeldt-Jakob disease, and potentially as a screening tool for variant Creutzfeldt-Jakob disease infection in asymptomatic individuals, which is a topic of current interest in the United Kingdom."

Provided by University of Texas Health Science Center at Houston

Citation: Researchers find infectious prion protein in urine of patients with variant Creutzfeldt-Jakob disease (2014, August 7) retrieved 19 April 2024 from <https://medicalxpress.com/news/2014-08-infectious-prion-protein-urine-patients.html>

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