

Safe To Eat?

PETER MANSBRIDGE (HOST):

Safe To Eat? She thought her son died of a random illness, one with no known cause, no cure. Now some scientists are questioning whether there's a new link to mad cow disease. Kelly Crowe reports. -

For years, scientists have known that B.S.E. causes variant Creutzfeldt-Jakob disease. What some people call the human form of Mad Cow Disease. But another form of C.J.D., Sporadic C.J.D., has always confounded them. The most common theory has been that the deadly illness occurs spontaneously with no known cause. Now some of the world's leading scientists in the field are having another look at sporadic C.J.D. and the possibility that it too is linked to mad cow. Here's Kelly Crowe with a feature report.

KELLY CROWE (REPORTER):

It starts out slowly. At first, the victims don't know what's wrong with them. They can't put their finger on it. They keep making little mistakes. The spelling mistakes that just look sloppy... numbers become easily confused... the words on the page become blurry. It's the beginning of a terrifying slide in to dementia and death. That's what happened to Jeff Schwan. Little by little, his brain was ravaged by a disease so rare, his mother had never even heard of it.

UNIDENTIFIED WOMAN (MOTHER OF JEFF SCHWAN):

And finally on August 11th, we received words you'll never want to hear. He has a disease that has no treatment and no cure, and we just tried to take that in.

NORMAN FOSTER (DOCTOR):

Check eye movements, look right here at my finger.

KELLY CROWE (REPORTER):

It was Dr. Norman Foster who discovered the terrible truth. He specializes in diseases of the brain, and he's the one who finally figured out what was wrong with Jeff Schwan.

NORMAN FOSTER (DOCTOR):

The major problem was a seizure disorder, and that was investigated and treated, but despite treatment, he became progressively worse. And it was unclear at the time what was causing these seizures, and it was suspected that this was due to an encephalitis or meningitis or some other problem, and I strongly advocated that everything be done to identify what the cause of this is, and that led to a brain biopsy. When we examined the tissue under the microscope, we were somewhat surprised to find evidence of Creutzfeldt-Jakob disease.

KELLY CROWE (REPORTER):

Jeff Schwan died from a mysterious illness called Sporadic Creutzfeldt-Jakob disease or Sporadic C.J.D. It's a rare and fatal brain-wasting disease that seems to strike out of nowhere. But was it really just bad luck that killed this healthy 26-year-old man, or is there another explanation? Sporadic Creutzfeldt-Jakob disease is a type of prion disease. Prions are proteins found in humans and other mammals, but when an altered form of the prion appears, it somehow starts a critical chain reaction turning healthy prions in to deadly ones that ultimately destroy the brain. Prion diseases are always fatal. There is no cure, no treatment. And science now knows that prion diseases can be spread by eating meat. It was first realized here in Papua New Guinea back in 1957 where a brain-wasting disease called Curu was sweeping through a tribe of cannibals.

Scientists realized these people were developing Curu after feasting on the remains of their dead who were already infected. It was the first evidence that prion diseases could be spread through eating infected material. 30 years later, British cattle began developing a prion disease. It was called B.S.E. or Mad Cow Disease. It was spread when the animals ate feed made from diseased cattle. Then young Britons began dying, and scientists realized humans could catch a prion disease from eating infected beef. That disease is called Variant C.J.D. So far, it has claimed 170 victims. But none of that seems to explain what happened to Jeff Schwan. He had the other prion disease, the one that is thought to strike at random, so-called Sporadic C.J.D. Like the other prion diseases, the brain fills with sponge-like holes, but scientists don't know what causes sporadic C.J.D., and some are asking, could it also be caused by eating meat? Here's one possible clue: The disease sometimes shows up in clusters in people who live near each other. It happens too often to be explained only by chance. British epidemiologist Simon Cousins worked on two scientific studies that documented clusters of the disease in both England and France. The studies concluded that it must be caused by something on the outside, that it can't be just a spontaneous disease.

SIMON COUSINS (EPIDEMIOLOGIST):

The spontaneous event would be, doesn't really explain why they might be clustering. So the clustering is pointing one in the direction of thinking from time to time people are exposed to a common external source.

KELLY CROWE (REPORTER):

One common external source, infected meat. It's a frightening possibility that is being actively investigated by some European scientists. Here in Switzerland, there was an epidemic of Mad Cow Disease in the 1990s, and now they're seeing a more than doubling in the rate of Sporadic C.J.D. Swiss scientists think there might be a connection.

ADRIANO AGUTZY (SCIENTIST):

What we need to find out is whether the whole problem of the enhanced Creutzfeldt-Jakob does, indeed, have something to do with mad cow's disease, but we certainly did have our definite window of exposure, and so if we now, if we had B.S.E. ten years ago and now we have enhanced Creutzfeldt-Jakob disease, it's unavoidable to ask the question whether the two phenomena are related.

KELLY CROWE (REPORTER):

DR. Adriano Agutzy is one of the world's experts on prion diseases. He says he has ruled out most of the other explanations, and now his main working hypothesis is that at least some Sporadic C.J.D. in Switzerland could be another form of human Mad Cow Disease.

ADRIANO AGUTZY (SCIENTIST):

But this by no means excludes that B.S.E. may manifest itself in humans with different characteristics, and maybe B.S.E. in Switzerland is also different from B.S.E. in the U.K., and then variant C.J.D. will also be different. So I think from the U.K. experience, it's impossible to draw the conclusion that B.S.E. will only give rise to what we know as variant C.J.D.

KELLY CROWE (REPORTER):

What it means is cattle infected with prion disease might be causing Sporadic C.J.D. To find out, the scientists are trying to match the tissue from infected Swiss cattle with the tissue from Swiss victims. If the tissue looks the same on the lab tests, then it will be strong evidence the two are connected. Right next door in Italy, they might have already made that connection. This doctor was studying the brains from B.S.E. infected cattle when he noticed a form of the disease he'd never seen before. He realized it was a new form of Mad Cow Disease. It was the first time scientists realized there could be more than one kind of prion disease in cattle.

UNIDENTIFIED MAN:

So our assumption is that since there is this biochemical similarities together with this, we think there might be a connection between the two.

KELLY CROWE (REPORTER):

He got a second shock when he ran the new cattle prion through the lab tests. It looked exactly like Sporadic C.J.D. He could hardly believe his eyes. He was looking at possible proof that humans could catch a second prion disease from cattle.

UNIDENTIFIED MAN:

Yes, this is only a biochemical evidence because at the beginning when B.S.E. has been discovered and the Variant C.J.D. had been discovered in the U.K., They found the single cell. I mean, the biochemical part of both B.S.E. and variant were similar. We found the same biochemical. There are pathological similarities and ecological similarities.

KELLY CROWE (REPORTER):

There is still one more step to prove the link. This new strain of B.S.E. will be injected into mice that have human genes. It will take two years, but the results will bring scientists a little bit closer to the answer of whether Sporadic C.J.D. is caused by meat.

ADRIANO AGUTZY (SCIENTIST):

I think that that is a possibility. Personally, I would say that it's possible that some of the cases are caused, I mean, maybe what would cause Sporadic C.J.D. is a collection of different diseases, and some of it could actually be transmission of B.S.E.

UNIDENTIFIED MAN:

It might be a possibility because it is an infectious disease. I mean, if you get a piece of brain of this with classic C.J.D. and you inoculate the animal, you transmit the disease, but on the other side, it's not clear whether the animal transmit the disease to man.

KELLY CROWE (REPORTER):

Scientists agree that cattle prion disease must be kept out of the human food supply. The problem is finding it before the animal is slaughtered. Bitter experience has already proven that the disease can be lurking even as officials boast that their herd is B.S.E.-free. After years of making those claims, the U.S. has had to admit to a homegrown case of B.S.E. in a Texas cow this past June. And that case was discovered only after testing was dramatically increased. And yet the U.S. and Canada are still only testing cattle that look sick. And with that kind of testing and millions of animals slaughtered every year, the disease is easy to miss. A lesson the Italians already learned. In Italy, they didn't know for sure that they had Mad Cow Disease until they started testing, not just the animals that looked sick, but every single animal over 24 months that goes into the food chain. Now four years later, they've only had one animal that looks sick, but they've found more than 130 cases of Mad Cow Disease in healthy looking animals. In other words, if they hadn't been testing those 130 animals would have gone into the food chain. In North America, they're only looking at symptomatic cases, zero so they could be missing a lot of disease.

UNIDENTIFIED MAN:

Sure, they take only neurological cattle, cattle with neurological symptoms or downer cattle. So this is the so-called passive surveillance. When you have neurological, you make the potential diagnosis of C.J.D. And the only way to get rid of this is that all the animals should be tested, and people at least buy a piece of meat to say this is B.S.E.-free.

KELLY CROWE (REPORTER):

Dr. Agutzy says the United States isn't trying hard enough to find prion disease in its cattle.

ADRIANO AGUTZY (SCIENTIST):

I think that in North America, there is a huge problem because North America has refused for many years to put in place any kind of serious surveillance of cattle, and I think this is a disgrace, and it really cries to Heaven about the fact that particularly the U.S. has just refused to even take into consideration the possibility that B.S.E. may be prevalent. It's a huge problem.

KELLY CROWE (REPORTER):

It's also not clear how widespread Sporadic C.J.D. is. The final diagnosis can only be made after death by examining the brain in an autopsy, but today autopsies are rarely done. And although Sporadic C.J.D. is a rare condition, estimated at one in a million, Dr. Norman Foster suspects the rate is higher.

NORMAN FOSTER (DOCTOR):

In many states in the United States, if a physician suspects a case, they can report it as a public health problem, but there's no mechanism to investigate or no way to pursue this in a systematic way.

KELLY CROWE (REPORTER):

So you could be missing lots of cases?

NORMAN FOSTER (DOCTOR):

So we are missing lots of cases.

KELLY CROWE (REPORTER):

Jeff Schwan is on the official record as just another unfortunate victim of a rare disease that seems to strike for no reason. And for his mother, it is the unanswered questions that weigh most heavily on her shoulders.

UNIDENTIFIED WOMAN (MOTHER OF JEFF SCHWAN):

So all of these people who are dying of Sporadic C.J.D., they don't have an answer for. It's very, very frustrating because we want answers.

KELLY CROWE (REPORTER):

To get those answers, she must wait for the slow deliberate pace of science. Kelly Crowe, CBC News, Sterling Heights, Michigan.