Scientists Weigh Risks of Beef; Muscle Alone Found Unlikely to Carry Mad Cow Disease; [FINAL Edition]
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The repeated assertion by government officials that American beef is safe to eat -- despite the discovery of the first U.S. case of mad cow disease -- is based in large part on painstakingly acquired evidence that pure muscle from mad cows simply does not contain enough of the strange infectious material to give the disease to a human being.

Muscle meat contaminated with brain or spinal tissue from infected cows is another matter. Eating it can be deadly. Scientists believe that is how about 150 people worldwide have contracted the human version of mad cow disease, which first appeared in Britain in 1996. But muscle meat alone -- beef, in short -- appears safe.

The evidence for this is strong and convincing. But as with all assessment of biological risk, it is not absolute and unqualified.

"I'd like to say for sure that muscle is safe. I'm reasonably sure that muscle is safe. But like everything else in science, the answer is incomplete," said Paul Brown, a physician and neuroscientist at the National Institutes of Health. He is a leading authority on bovine spongiform encephalopathy (BSE), the formal name of mad cow disease.

In addition to the brain and spinal cord, other infectious parts of a mad cow's carcass include a specific stretch of the small intestine, the tonsils, eyes and various other parts of the nervous system. As those "hot" tissues were identified in the years after the disease was discovered in cows in Britain in 1986, officials there and elsewhere worked to get them out of human food.

They also worked to get them out of animal food. That is because Britain's BSE epidemic arose when healthy cattle were fed protein supplements made unwittingly with tissue from infected cows -- a practice that caused an ever-expanding cycle of infection.

In the United States, some nervous system material that is known to be sometimes infectious if an animal has BSE was allowed into the meat supply as recently as last year. (The material, called "dorsal root ganglia," sticks out between the bones of the spine. It can get into meat when mechanical devices are used to remove muscle from bone. But until last month's detection of BSE in an American cow, government experts did not view that small amount of contamination as especially risky.

With a single case of mad cow disease in Washington state, the Department of Agriculture invoked rules last week that it hopes will keep all potentially infectious material, including the ganglia, out of meat. Officials believe this will make beef even safer -- should there be more than one American cow with BSE.

The move was taken, in part, because of the strange and chilling nature of the infection.

Mad cow disease is transmitted by a prion, which is an abnormally shaped version of a specific protein, called PrP, made routinely by billions of cells in many species of animals. For reasons that are not known, prions are able to force PrP molecules to abandon their normal shape and "refold" into the prion shape, or conformation.

Those new prions go on to convert other PrP molecules in a microscopic version of "Invasion of the Body Snatchers." Over time, the accumulation of prions destroys the brain.

It is difficult to inactivate prions -- much more difficult than it is to kill viruses, bacteria or single-cell parasites. (Because prions are not alive, "killing" them is not quite the right term.) They can survive relatively high temperatures and caustic chemicals that leave other pathogens in pieces.

Many mammals are susceptible to diseases caused by misshapen PrP proteins. The diseases go by various names -- BSE in cattle, scrapie in sheep, chronic wasting disease in deer and elk. In humans, it is called Creutzfeldt-Jakob disease (CJD), a rare and invariably fatal ailment that usually arises spontaneously. When it is caused by eating BSE-infected cow tissue, the disease follows a somewhat different, though equally deadly, course and is called variant
Creutzfeldt-Jakob disease (vCJD).

The incubation period for vCJD is long. Researchers believe the unlucky few with the disease in all likelihood consumed ground meat contaminated with bits of brain or spinal cord 15 years, on average, before showing any signs of illness.

The evidence that muscle from a BSE-infected cow cannot transmit the disease comes from a laborious experiment performed in Britain over the past 13 years.

The researchers began by feeding brain matter from BSE-infected animals to 30 calves -- the same route that led to more than 182,000 reported cases of the cow disease in Britain between 1986 and 2002. Whereas only 5 percent of the British herd had BSE at any one time - - a fact that suggests the disease is fairly hard to contract -- the experimental animals were given enough brain tissue to ensure all would be infected.

Over the next 40 months, the researchers periodically killed a sample of the animals and removed organs and anatomical structures. Each organ type was pooled -- spleens with spleens, spinal cords with spinal cords, etc. Each type was then ground up to form an extract.

The researchers injected the extracts into groups of uninfecte sed calves, a total of 325 animals in all. The injections were into the brain -- a surefire way of producing the disease if the injected material is infectious. Each group of calves got an extract from only one organ. Those animals were then periodically sacrificed and examined to see whether they had contracted BSE.

The experiment was designed to answer two questions: What parts of a BSE-infected cow can transmit the disease? When do those parts become infectious?

The first organ to become infectious was the far end of the small intestine. It turned six months after a calf ate BSE-infected brain. The tonsils were second; they had enough prions at 10 months to be infectious. The brainstem did not become infectious until 32 months into the experiment -- almost three years after a calf ate the infected brains.

Nearly six years after the injections, no calf that received muscle extract from BSE-infected cows has ever become infected. Evidence from other studies found that blood and milk from infected cows also are not infectious. These are the crucial findings that lead experts to proclaim that muscle meat is safe to eat.

This experiment has been done only once. Most scientists prefer that an experiment be done at least twice before putting confidence in the results. But that is unlikely to happen with this one.

"The interest in the UK in funding research on BSE is on the wane now," said Gerald A. H. Wells, the 63-year-old veterinarian who headed the study and who 18 years ago described the first case of the disease.

Although the calf study is considered the best available evidence, two recent studies have raised small red flags about the safety of muscle.

Stanley B. Prusiner, the scientist who won a Nobel Prize in 1997 for describing how prions work, did an experiment in which he took muscle from the hindquarters of mice infected with scrapie, the sheep disease. He injected muscle extract into the brains of healthy mice. They became infected.

A second study, published in November in the New England Journal of Medicine, examined tissue from people who had died of spontaneous CJD. A team of Swiss researchers found that PrP prions were detectable in the muscle tissue of one-third of them.

Although neither of the studies involved beef, together they make clear that muscle tissue in some species can carry enough PrP prions to transmit disease. This raises the possibility that it might occasionally happen in cattle muscle, too.

While the infectiousness of various parts of the cow is the biggest factor in determining whether a person gets the disease, it is not the only factor. Some people may be genetically resistant to becoming infected by BSE-contaminated meat. Whole sectors of the population may have varying risks for the disease.

The PrP proteins in human cells can also vary slightly. Some people have PrP proteins that contain a chemical compound called methionine at one particular spot -- position 129 -- on the long, necklace-like protein. Other people have at that spot a different compound, called valine. Some people have a mixture of PrP molecules, half containing methionine and half containing valine at that spot, depending on inheritance.
To date, every person who has become infected by BSE-contaminated meat has the "methionine-only" version of the PrP protein. As it happens, about 40 percent of white Northern Europeans have this variation of PrP. The remaining 60 percent have either a mixture or the valine-only version. (The percentage in other races and ethnic groups is not known.)

What makes this group of people so vulnerable? Are their proteins more easily persuaded to refold into the abnormal shape? Is everyone else resistant to infection by BSE prions? Or does it simply take longer for people with the other PrP variations to show signs of illness? Will there be a second, even more-delayed wave of cases decades in the future?

Like so much about these strange diseases, those questions will only be answered by time.