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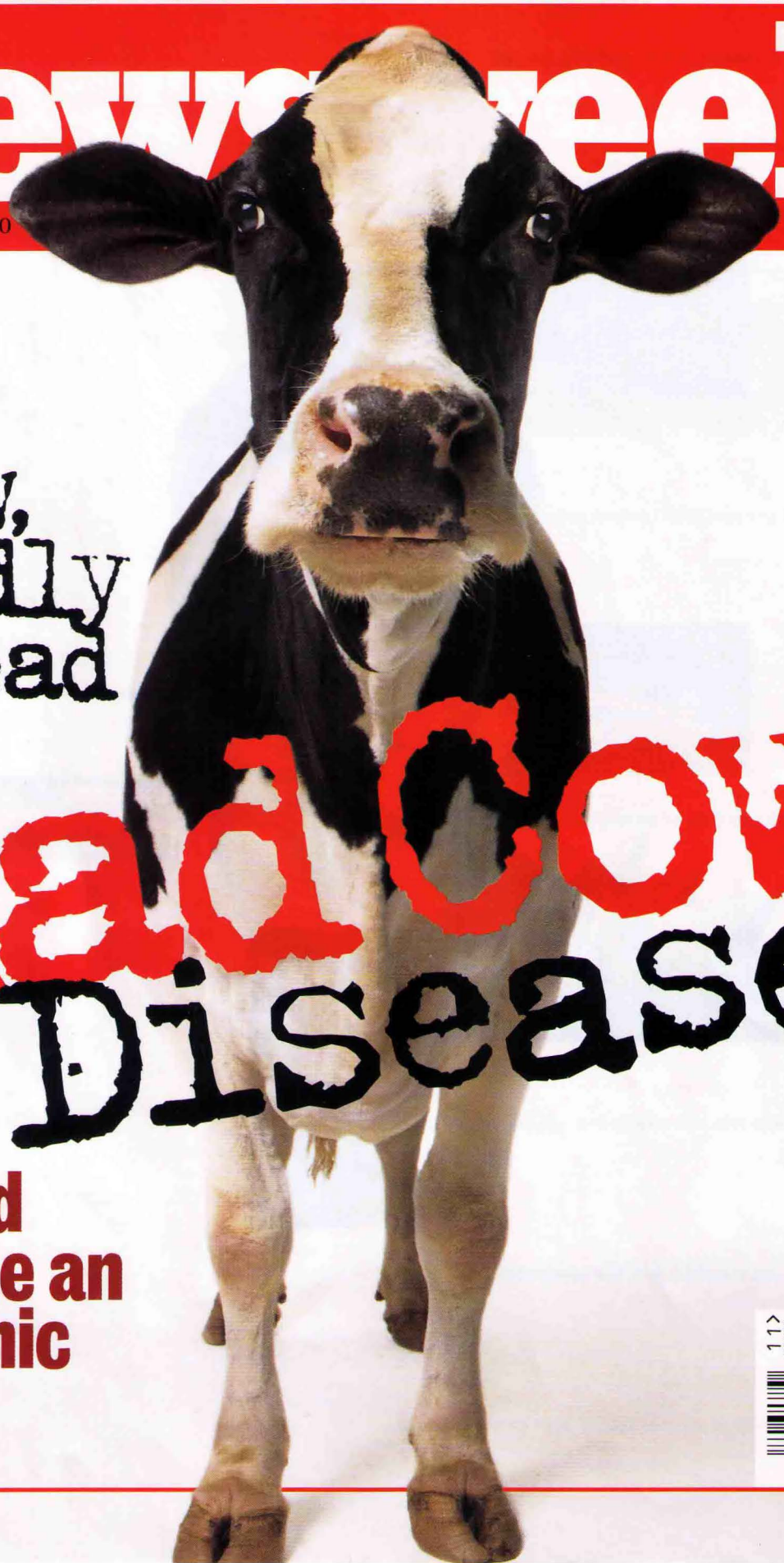

The Slow, Deadly Spread of Mad Cow Disease

How It Could Become an Epidemic

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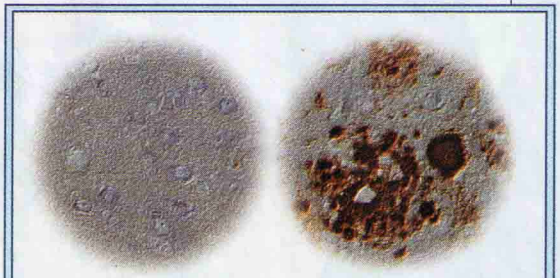
CANNIBALS TO COWS: THE PATH OF A DEADLY DISEASE

HEALTH OFFICIALS SAY THEY'VE GOT MAD COW UNDER CONTROL, BUT MILLIONS OF UNAWARE PEOPLE MAY BE INFECTED. WHY IT COULD STILL TURN INTO AN EPIDEMIC. BY GEOFFREY COWLEY

PETER STENT WAS A SEASONED DAIRYMAN, BUT HE HAD never seen anything like this. Just before Christmas, in 1984, one of his cows at Pitsham Farm in South Downs, England, started shedding weight, losing its balance and acting as skittish as a cat. When the vet came to investigate, the animal was acting completely crazy—drooling, arching its back, waving its head, threatening its peers. And by the time it died six weeks later, Stent was seeing the same symptoms in other cows. Nine were soon dead, and no one could explain why. The vet dubbed the strange malady Pitsham Farm syndrome, since it didn't seem to exist anywhere else. Little did he know.



YOUR BRAIN ON PRIONS: Normal human tissue (left), and with the spongy texture of vCJD

Alison Williams was 20 years old at the time, and living in the coastal village of Caernarfon, in north Wales. She was bright and outgoing, a business student who loved to sail and swim in the nearby mountain lakes. But her personality changed suddenly when she was 22. She lost interest in other people, her father recalls, and quit school to live at home with her parents and her brother. She still enjoyed the outdoors, but she took to sitting alone on her bed, staring out the window for hours at a time. By 1992, Alison was having what her doctors diagnosed as nervous breakdowns, and by 1995 she had grown paranoid and incontinent. "A month before she died, she went blind and lost use of her tongue," her dad recalls. "She spent her last five days in a coma."

Anyone with a television has heard such stories, maybe even sussed out the connection between them. Mad-cow disease, or bovine spongiform encephalopathy (BSE), has killed nearly 200,000 British and European cattle since it cropped up on Pitsham Farm. The human variant that Alison Williams contracted has claimed 94 lives as well. What few of us realize is that these tolls could mark the beginning of something vastly bigger. No one knows just how BSE first emerged. But once a few cattle contracted it, 20th-century farming practices guaranteed that millions more would follow. For 11 years following the Pitsham Farm episode, British exporters shipped the remains of BSE-infected cows all over the world, as cattle feed. The potentially tainted gruel reached more than 80 countries. And millions of people—not only in Europe but throughout Russia and Southeast Asia—have eaten cattle that were raised on it.

It's possible, of course, that the worst is already behind us. After dithering for a decade, governments in the United King-

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OPPOSITE PAGE: CRAIG CUTLER
ABOVE: PHOTOS COURTESY DR. JAMES IRONSIDE, NATIONAL CJD SURVEILLANCE CENTER, U.K. AND DR. PIERLUIGI GAMBETTI, NATIONAL PRION DISEASE PATHOLOGY SURVEILLANCE CENTER, CLEVELAND, OHIO

dom and Europe have lately taken bold steps to control BSE. The number of bovine cases is now falling in Britain—and the United States has yet to even report one. American officials banned British cattle feed in 1988, as soon as scientists implicated it in BSE, and later barred the recycling of domestic cows as well. The U.S. government, the cattle industry and many experts now voice confidence in the nation's fire wall and say the risk to consumers is slight. In truth, however, America's safeguards and surveillance efforts are far weaker than most people realize. And in many of the developing countries that now face the greatest risk, such efforts are nonexistent. How many of the world's cattle are now silently incubating BSE? How many people are contracting it? The truth is, we don't know. "We have no idea how many deaths we're going to see in the coming years," says Dr. Frédéric Saldmann, a French physician who has recently seen both cows and people stricken in his country. "We've been checkmated."

Mad cow is the creepiest in a family of disorders that can make Ebola look like chickenpox. Scientists are only beginning to understand these afflictions. Known as transmissible spongiform encephalopathies, or TSEs, they arise spontaneously in species as varied as sheep, cattle, mink, deer and people. And once they take hold they can spread. Some TSEs stick to a single species, while others ignore such boundaries. But each of them is fatal and untreatable, and they all ravage the brain—usually after long latency periods—causing symptoms that can range from dementia to psychosis and paralysis. If the prevailing theory is right, they're caused not by germs but by "prions"—normal protein molecules that become infectious when folded into abnormal shapes. Prions are invisible to the immune system, yet tough enough to survive harsh solvents and extreme temperatures. You can freeze them, boil them, soak them in formaldehyde or carbolic acid or chloroform, and most will emerge no less deadly than they were.

The prion story starts in the 1730s, when shepherds in Britain and Europe described the disease we call scrapie. Like Peter Stent's cows, afflicted sheep would grow ill-tempered and wobbly. Then, over three to six months, they would suffer

A KILLER IN OUR MIDST

Doctors don't know exactly what causes most cases of Creutzfeldt-Jakob. But once in a while the disease can be traced to a specific exposure. Some of these victims may have been spared if the risks had been fully understood at the time.

PARIS, FRANCE

CAUSE: TAINTED MEAT

At first, Laurence Duhamel seemed depressed. Later she developed paranoid delusions. But Duhamel wasn't having a breakdown. Before she died last year at 36, a biopsy confirmed that her brain had been destroyed by variant CJD, a condition caused by eating infected beef. Her family has sued the French government.

BETHPAGE, N.Y.

CAUSE: PITUITARY HORMONES

As a child Wendy Nofi was so short her doctor prescribed human growth hormones. "Her parents assumed everything was safe," says her husband, Michael. The treatment seemed to work; she grew to an almost normal 4 feet 10. But at 30, she started losing her balance. By the time the National Institutes of Health confirmed that she had received tainted hormones, she could not walk, dress herself or swallow. She lasted 2½ years in a comalike state, fed through a tube. Nofi left behind three small children when she died in 1998.

CLOCKWISE FROM TOP LEFT: AP, COURTESY OF BISSELL FAMILY, COURTESY OF BRENDA KOEPKE, JESSICA WYNNE

DENVER, COLO.
CAUSE: DURA-MATER GRAFT

After Karen Bissell drove from Miami to Denver to visit her parents in 1998, her legs and neck ached. At the time her folks blamed the long drive. But less than two months later Karen came home to die. She had contracted CJD during brain surgery years before. Doctors had used a patch of brain sheathing called dura mater to close the incision. The patch was infected.

FALLON, NEV.
CAUSE: UNKNOWN

Jim Koepke spent his life as a ranch hand, tending cattle and sheep. He also loved hunting, and as a child he ate elk and deer killed by his father. Looking back, his widow, Brenda, doesn't know whether he contracted CJD through his diet or his work. She knows the disease killed him fast. The 6-foot-1 cowboy shrank to less than 120 pounds before he died in 1999 at 39. "I could carry him," Brenda says.

seizures, paralysis, blindness and death. Scrapie is still common in sheep, but doesn't seem to strike people. As far as we know, no one has ever gotten sick by eating infected mutton.

Dr. Carleton Gajdusek knew nothing of scrapie when he landed in Papua New Guinea in the 1950s. But Gajdusek, an American pediatrician and virologist employed by the National Institutes of Health in Maryland, soon encountered something similar. A strange neurological disease was killing the Fore people of the country's Eastern Highlands—especially the women and children. The Fore called the condition "kuru," which means shaking or shivering, and they knew its 16-month progression well: tremors and an unsteady gait, followed by slurred speech, joyless laughter and, finally, stupor and death.

The Fore knew kuru as a curse cast by sorcerers. Like most outsiders, Gajdusek suspected it was an epidemic disease, somehow related to the tribe's eating habits. Fore men supplemented their bean-and-sweet-potato diets with small game, but women and children lacked protein. The women had recently created a ritual to fill the gap. Instead of burying dead loved ones, they ate them. As Richard Rhodes recounts in his 1997 book, "Deadly Feasts," "They did not eat lepers or those who died of diarrhea, but the flesh of women killed by [kuru] they considered clean."

The link between kuru and cannibalism seemed clear enough. But as he examined living patients, Gajdusek saw no outward signs of infection—no fever, no inflammation—and culture tests turned up nothing suspicious in their spinal fluid. By sending autopsy samples to his colleagues back in Maryland, Gajdusek did learn that the patients' brains resembled those of people with Creutzfeldt-Jakob disease (CJD), a rare and fatal brain condition that German researchers had discovered in the 1920s. Both conditions filled the brain with "vacuoles," small cavities resembling the holes in a sponge. And despite some differences, they ran essentially the same course. But that only deepened the mystery. As far as anyone knew, CJD was just a biochemical fluke, a disease that strikes randomly and infrequently all over the world. Kuru was spreading like a plague.

Gajdusek published several reports on kuru over the next couple of years, and one of them caught the attention of Dr. William Hadlow, a scrapie expert. Hadlow noticed that the vacuoles in Gajdusek's kuru brains resembled those he'd seen in sheep. The symptoms sounded familiar, as well. In a letter to The Lancet, Hadlow listed the parallels between kuru and scrapie, and posed a tantalizing question. Studies had shown that healthy animals developed scrapie when injected with a sheep's diseased brain tissue. What would happen, he wondered, if you injected a healthy animal with brain tissue from a kuru victim?

BSE IS THE CREEPIEST OF THE PRION DISEASES AND MAKES EBOLA VIRUS LOOK LIKE CHICKENPOX

Would this disease spread in the same way?

To find out, Gajdusek and a colleague started injecting chimps and monkeys with the ground brains of Fore tribeswomen. By 1965 they had shown that kuru was transmissible. Gajdusek then repeated the experiment with brain tissue from an American CJD victim and got the same result. These astonishing discoveries helped control kuru in New Guinea. They also won Gajdusek a Nobel Prize. He had shown that scrapie, kuru and CJD could all spread and kill in the same manner. Unfortunately, the responsible pathogens were still unknown. And as it turned out, eating one's relatives was not the only way to contract them.

While Gajdusek and his colleagues were investigating kuru, other scientists were pursuing the secrets of growth and maturation—and making equally thrilling discoveries. Endocrinologists had found a rich store of hormones in the pituitary, a pea-sized gland near the front of the brain. And by the early 1960s they had shown that one of these substances—human growth hormone—could help dwarves reach more normal heights. Human cadavers were the only known source of hGH, and demand was intense. So in 1963, the federal government created a National Pituitary Agency to harvest and distribute the glands. Over the next two decades, roughly 8,200 kids got hGH through the agency, and similar programs cropped up throughout Europe.

All seemed well until 1984, when a troubling pattern emerged. In its common "sporadic" form, Creutzfeldt-Jakob disease is rare in people under 50. Yet patients

who'd received growth-hormone injections were getting the disease in their 20s. By spring 1985 there were four such cases on record, and the implication was obvious: the medical establishment had created the high-tech equivalent of a Fore funerary feast. No one knew how many pituitary donors had been silently incubating CJD, or how many recipients were now set to die from it. But 27,000 of the world's children had received injections when the practice was stopped on April 20. As Dr. Paul Brown of the NIH wrote later that year, America now faced the "ominous possibility of a burgeoning [CJD] epidemic." (Twenty-two cases have now been recorded in U.S. hormone recipients, and new ones are still turning up each year.)

Britain faced an epidemic as well, and hormone recipients were not the only ones at risk. Peter Stent had just lost those nine cows when the hGH crisis came to light, and other English farmers were soon having similar experiences. In 1986, pathologists discovered that Pitsham Farm syndrome was yet another variation on scrapie, kuru and CJD—a *bovine* spongiform encephalopathy, or BSE. And when the toll continued to rise, health officials started considering defensive measures. In 1988, they mandated the destruction of stricken cows and halted the use of cows, sheep and other ruminant animals in cattle feed. Unfortunately, they underestimated the threat they faced.

BSE was by now so rampant that existing feed supplies were infecting hundreds of cattle every week. But because most of the infected cows were still healthy, the epidemic appeared small. Instead of seizing all potentially tainted feed, the government gave farmers and feedlots five weeks to use

WHAT ABOUT HAMBURGER?

A QUICK GUIDE TO ASSESSING YOUR REAL RISK

BY ANNE UNDERWOOD

THE CHANCES OF GETting Creutzfeldt-Jakob disease are very small. So far, U.S. officials believe no one here has contracted the disease from eating beef. But given CJD's deadly nature and the fact that scientists are still unraveling its mysteries, consumers may be uneasy. Here are answers to some common questions:

SHOULD I STOP EATING BEEF?

That depends on your level of risk tolerance. You can't contract variant CJD (the human form of mad-cow disease) unless you eat meat from an infected animal—and no tainted cows have been identified here so far. Critics charge that's because the government hasn't been looking hard enough.

ARE CERTAIN CUTS OF BEEF MORE RISKY THAN OTHERS?

European health agencies believe the greatest dangers come from burgers, sausages and meat still attached to the bone, such as T-bone steak. Because these products are more likely to contain nerve fibers, they're also more likely to harbor the prions involved in mad-cow disease. Flank steak and filet mignon are presumably safer.

MARIANNA DAY MASSEY—ZUMA

HOW SAFE? So far, American cows have tested disease-free

SHOULD I AVOID EATING MEAT OVERSEAS?

Even in Europe, where a dozen countries have lost cattle to mad-cow disease, the odds of contracting vCJD from a serving of beef are remote. But that hasn't stopped Europeans from jettisoning red meat faster than you can say *bistecca alla fiorentina*. The good news is that most European countries now monitor their herds. The risk may be higher in Russia and in Southeast Asia, where countries that imported feed and cattle from Britain during the 1980s and '90s have yet to mount extensive surveillance efforts.

ARE PORK, LAMB AND CHICKEN ALL SAFE TO EAT?

Chickens and pigs are unlikely to harbor mad-cow disease. Lambs can develop scrapie, but that disease isn't known to to harm people.

DO SUPPLEMENTS POSE A DANGER?

Cow hides and bones are used to make the gelatin in gel caps, hard capsule shells and glossy pill coatings. But prions have not been found in these products. A bigger worry are memory-boosting supplements that contain raw cow brains and glandular concentrates.

WHAT ABOUT VACCINES?

The risks are minimal. Vaccines are grown in cell cultures derived from humans and animals, including cows. But the FDA recommends that no cows from countries with mad-cow disease be used. Even in Britain, there is no evidence that transmission has ever occurred through vaccines.

CAN MEDICAL PROCEDURES SPREAD CJD?

Yes. Standard CJD has been transmitted by surgical instruments and by hormones and brain tissues taken from human cadavers. But these cases are exceedingly rare. In more than 20 years, the United States has confirmed only three from dura-mater grafts, three from surgical instruments, two from corneas and 22 from pituitary hormones that are no longer used.

COULD CJD INVADE THE BLOOD SUPPLY?

In theory, yes. That's why the U.S. government bans blood donations from people who spent more than six months in the United Kingdom between 1980 and 1996. Still, transfusions have apparently never spread the disease, even among hemophiliacs.

With bureau reports

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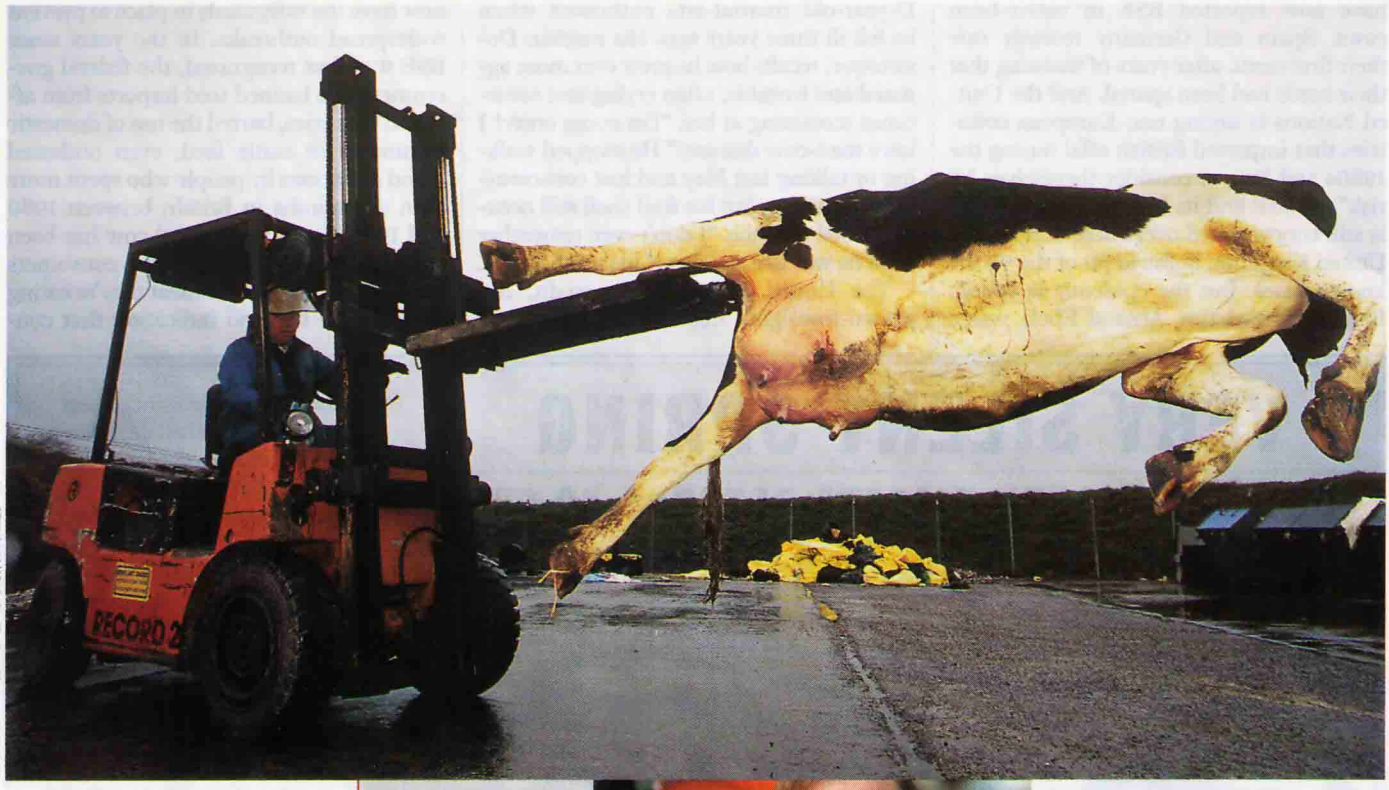


up their inventories. And instead of barring cow and sheep offal from all feed products, the new rules focused narrowly on feed intended for British cows. Beef byproducts continued to circulate in pig and chicken feed, which proved impossible to segregate from cattle feed on farms and in factories. And because the new rules said nothing about exports, Britain's banned cattle feed flooded other countries for another eight years. From 1988 to 1996, Asian nations alone bought nearly a million tons.

Meanwhile, people continued to eat as much beef as ever. The British government dismissed concerns about the food supply, but there were soon clear signs that BSE could spread beyond cattle. The disease started cropping up in zoo animals and domestic cats, which were receiving beef byproducts in their feed. Then researchers succeeded at transmitting BSE from a cow to a pig through injections of brain tissue. Each finding caused a new wave of public concern, forcing the government to adopt new precautions. By late 1990, health officials had banned a list of highly infectious cow parts—the brain, spinal cord, spleen, thymus, tonsils and intestines—from *all* food products, human or animal. Yet the government continued to insist that people had no cause to worry.

DENIAL: In 1990 British Agriculture Minister John Gummer bit into a burger to allay fears about meat; six years later, infected cows were being incinerated

Britain's chief medical officer was still denying any risk in 1993 as Alison Williams, the young Welsh woman, drifted into a stupor. But everything changed two years later, when pathologists examined the brains of Williams and several other young adults who had died of what looked like CJD. In addition to the spongiform vacuoles that are the hallmark of the disease, their brains were littered with large, flower-shaped plaques. And their lesions were concentrated not in the cerebral cortex (the usual locus of CJD damage) but in the cerebellum, a globular structure perched near the base of the brain. In short, their tissue samples had BSE written all over them. In 1996, Health Secretary Stephen Dorrell went before Parliament to announce that BSE had spawned a new human disease: a "variant of CJD," or vCJD. "Beef is one of the great unifying symbols of our culture," The Guardian lamented in an editorial. "The Roast Beef of Old England is a fetish, a household god, which has suddenly been revealed as a Trojan horse for our destruction."

Finally, almost a decade after the first cow got sick, Britain banned *any* recycling of farm animals and stopped exporting meat-based cattle feed. The country has since spent billions destroying cows more than 30 months old, regardless of their apparent health, and disposing of the remains. Some 500,000 tons of ground carcass are now stored at 13 sites around the country. Sealed tankers transport the stuff to high-temperature incinerators. The ashes are then buried. Thanks to these belated efforts, British farmers are now reporting only 30 BSE cases a week, down from 1,000 or more in the early '90s.

For the rest of the world, the worst almost surely lies ahead. A dozen European countries

ASIAN NATIONS HAVE BOUGHT NEARLY A MILLION TONS OF SUSPECT BRITISH FEED

have now reported BSE in native-born cows. Spain and Germany recently saw their first cases, after years of insisting that their herds had been spared. And the United Nations is urging non-European countries that imported British offal during the 1980s and '90s to consider themselves "at risk" for BSE and its human variant. vCJD is still concentrated overwhelmingly in the United Kingdom, home to 89 of the 94 the known cases. But the epidemic is young. France's first victim, Arnaud Eboli, was a 17-year-old martial-arts enthusiast when he fell ill three years ago. His mother, Dominique, recalls how he grew ever more agitated and irritable, often crying and sometimes screaming at her, "I'm going crazy! I have mad-cow disease!" He stopped walking or talking last May and lost consciousness in August, but his frail shell still occupies a bed at home. "I don't even remember what he was like anymore," his mother says.

The United States, to its credit, has shown foresight. Most experts believe we now have the safeguards in place to prevent widespread outbreaks. In the years since BSE was first recognized, the federal government has banned feed imports from affected countries, barred the use of domestic ruminants in cattle feed, even outlawed blood donations by people who spent more than six months in Britain between 1980 and 1996. Not a single mad cow has been reported in this country, and consumers seem confident that the meat they're eating is safe. "We have no indication that con-

A VERY SILENT SPRING

FOOT-AND-MOUTH DISEASE RETURNS TO EUROPE

HAYDN WEST—PA

ASHES: Infected pig and cattle carcasses blaze in England

BY STRYKER MCGUIRE

THE SCENE IS SICKENingly familiar: thousands of pig, sheep and cow carcasses being piled on funeral pyres. This time it's not mad-cow disease that's to blame, but the resurgence of a centuries-old virus called foot-and-mouth disease. Since the first signs of FMD came to light in February, 45,000 animals have been slaughtered in Britain. In Scotland, Dolly, the famous cloned sheep, was quarantined, and London locked the gates of Richmond Park to protect the royal deer inside. Across the channel, the Belgians, the Germans and the French are prophylactically incinerating tens of thousands of animals as well. Everywhere a sense of foreboding has settled over farm towns. "It's the silent spring here," said Paul Evans, a conservationist in Much Wenlock, England. "The woods are full of deer, but there's no one about at all. You can almost feel the tension in the land."

Unlike mad-cow disease, foot-and-mouth represents little danger to the public health, but it is a critical economic blow to European farming. It is not a prion disease. It is seldom fatal to animals and is rarely transmissible to humans. What it does, though, is cause blistering of the mouths and feet of animals with cloven hooves, as well as eventual lameness, loss of appetite and wasting. And it is so contagious—it can be carried in dust particles clinging to a car antenna—that one infected animal can easily contaminate the entire herd. "It is like a ball at the top of the hill: you know it is going to pick up speed," said James McInnes of Britain's National Farmers Union. To protect their business, European farmers have started to exterminate at-risk animals and quarantine the others. And because farmers are not shipping livestock, meat production in Britain, at least, has virtually ceased. Until the epidemic is contained, British farmers stand to lose about $86 million a week. European governments have already offered their farmers hundreds of millions of dollars in relief.

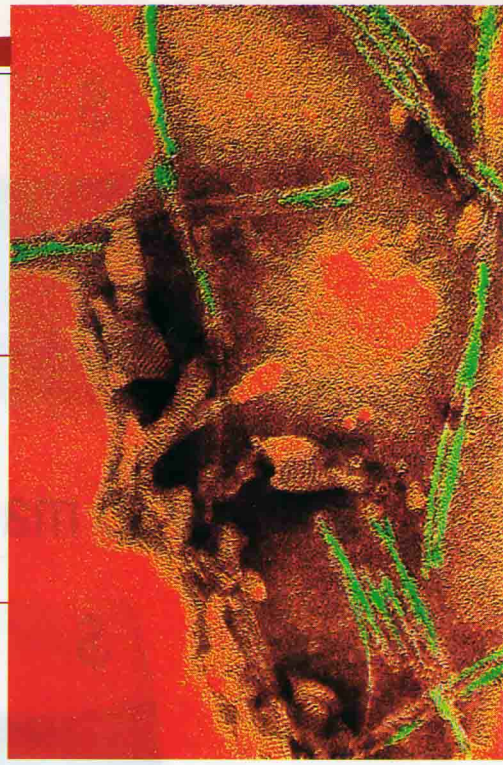
Ever since the British quelled a 1967 outbreak of FMD by slaughtering 447,000 animals, Western European health officials had thought the disease was licked. But the virus has long been endemic in Africa, the Middle East, Asia and South America. In an increasingly global market, where people and animals can move freely across borders, a disease like foot-and-mouth can spread quickly from one continent to another. At the end of last week there were no confirmed cases of the disease in mainland Europe. If it does spread, last week's alarm will seem pretty tame.

The USDA is downplaying the new risk, but it banned the importation of pork products from Britain as soon as it heard of the outbreak. And independent scientists are far less sanguine. To infect the U.S. meat supply, "all someone would have to do is go to a farm in England and then go to a farm here," says Dr. Lewis Thomas, West Virginia's state veterinarian. "We have hundreds of millions of dollars' worth of livestock, and they would have to be slaughtered. It would be beyond the realm of comprehension."

With SCOTT JOHNSON *in Paris and* JULIE SCELFO *in New York*

BIRTH OF A PLAGUE

A type of dementia called Creutzfeldt-Jakob disease was first described 80 years ago. But a new variant of the disease was identified in England in 1995, after bovine spongiform encephalopathy (BSE) spread to humans.



PRION FIBRILS in a mad-cow-infected brain. The green strands are believed to be aggregations of the infectious prion proteins.

1 A cow in England contracts BSE, or mad cow. The disease may have arisen spontaneously, or it may have spread from sheep.

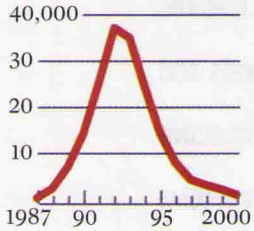
ANIMAL CASES

Herds from Portugal to France and Switzerland have been affected.

Top European totals
1987-2000

U.K.	180,401
Ireland	587
Portugal	509
Switzerland	366
France	241

United Kingdom

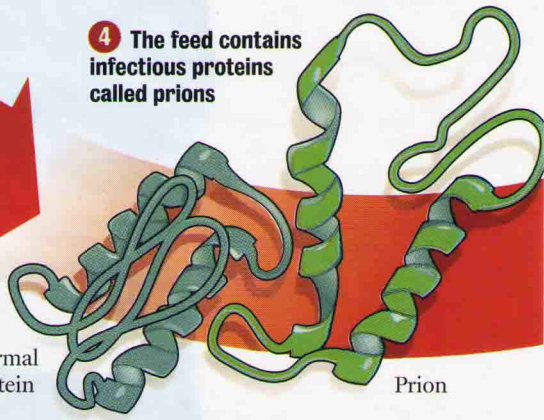


2 Remains of the infected cow are ground up into meat-and-bone-meal feed and given to other cows

Cows are herbivores and do not normally feed on other cows

3 Contaminated feed is distributed within England and shipped to other countries

4 The feed contains infectious proteins called prions



PRION DISEASES IN ANIMALS

Scrapie in sheep has been known for centuries, but it doesn't seem to infect humans. Variants in other species have emerged more recently.

Scrapie

1730s: sheep

Transmissible mink encephalopathy

1965: minks

Chronic wasting disease

1980: elk

1980: deer

Bovine spongiform encephalopathy

1985: cattle

1990: cats

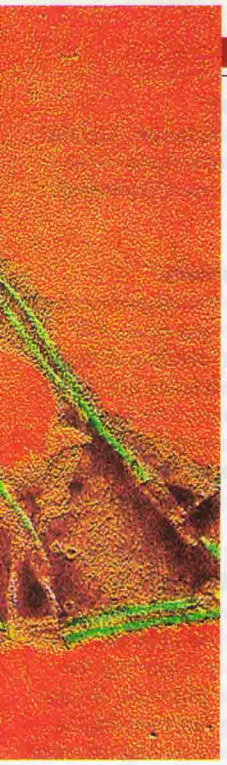
1992: zoo monkey

sumption is falling," says Alisa Harrison of the National Cattlemen's Beef Association. "Actually, beef demand is on the increase."

But it's a bit early to conclude that America is prion-proof. Spongiform encephalopathies have turned up in American sheep, deer, elk, mink and people in recent decades. And though BSE has not been seen in U.S. cattle, some experts suspect we're just not looking hard enough to find it. Over the past decade, animal-health officials have examined brain tissue from 12,000 "downer" (nonambulatory) cattle without finding any BSE. Some 2,300 downers tested negative last year—proving, says Gary Weber of the Beef Association, that the U.S. infection rate is fewer than one cow in a million. Other experts say he's wrongly assuming that only downer cattle can have BSE, when infected cows may look healthy for five years. Germany appeared BSE-free when it tested only downers, says Marcus Doherr, an epidemiologist who helped design Switzerland's testing program. "Now, with very intensive screening, they have found over 30 cases within two months."

If BSE did show up in this country, could we keep the disease from spreading? Agriculture officials say the ban on bovine cannibalism would prevent the kind of explosion Britain experienced early on. Perhaps, but this country's feed rules are neither as strict nor as well enforced as you might think. Since cows and sheep are prone to TSEs, the government bars the use of cow and sheep byproducts in their feed. But since pigs and poultry don't exhibit TSEs outside the laboratory, they can eat anything. That's not a problem in itself, unless pigs are more susceptible than we realize. The trouble is, it's nearly impossi-

CLOCKWISE FROM TOP: SPL-PHOTO RESEARCHERS, HARTMUT SCHWARZBACH—STILL PICTURES, BRUCE COLEMAN, TIM DAVIS—PHOTO RESEARCHERS, L. M. STONE—BRUCE COLEMAN, PHOTO RESEARCHERS (2), BRUCE COLEMAN (2), JOHN COLWELL—GRANT HEILMAN



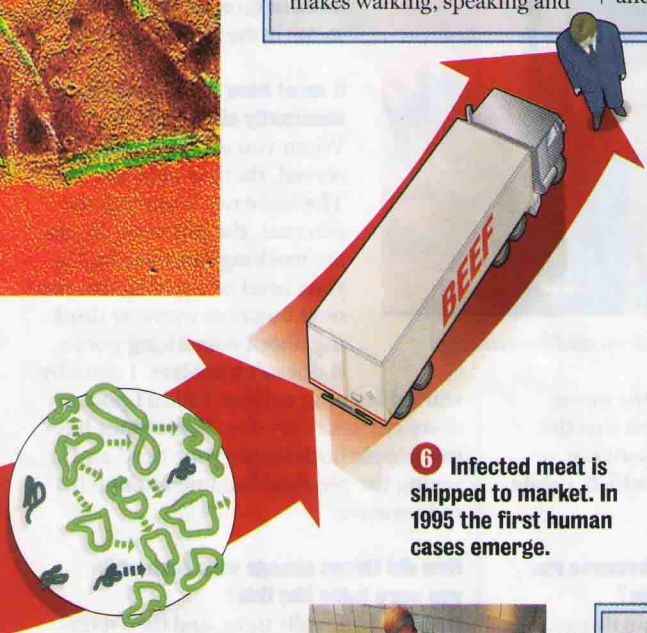
PRION DISEASES IN HUMANS

Creutzfeldt-Jakob: Begins subtly with depression and memory problems, but in 4 to 6 months progresses to dementia, uncontrollable jerking of muscles and finally death.

Gerstmann-Sträussler-Scheinker syndrome: Damage to motor portions of the brain makes walking, speaking and even swallowing difficult. Dementia comes much later. Death within 2 to 6 years.

Fatal insomnia: Progressive insomnia leads to panic attacks, phobias, hallucinations and finally dementia. Patients die within 18 months.

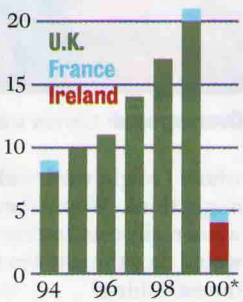
Kuru and variant Creutzfeldt-Jakob: Anxiety yields to trembling, loss of coordination and death after 16 months.



6 **Infected meat is shipped to market. In 1995 the first human cases emerge.**

HUMAN CASES

Most victims so far are British. But France and Ireland have had a few.



*FIGURES INCLUDE STILL-LIVING PERSONS WITH PROBABLE VCJD OR DECEASED PERSONS WHOSE DIAGNOSES HAVE NOT YET BEEN CONFIRMED.

5 **These prions cause normal proteins in the brain to refold into abnormal shapes. Infected cows are slaughtered for meat.**

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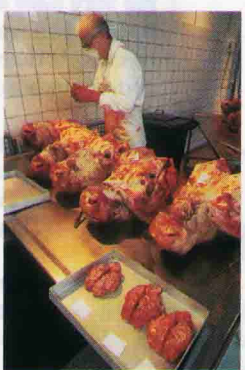
SOURCES: WORLD ORGANIZATION FOR ANIMAL HEALTH; CENTERS FOR DISEASE CONTROL; FOOD AND AGRICULTURE ORGANIZATION. GRAPHIC BY TONIA COWAN—NEWSWEEK



Examination of cattle imported into Germany from Britain

BAD FEED FOR SALE

Official British figures show that more than 80 countries imported cattle feed that was probably infected. A partial list of deliveries, 1980-1996:

Imports, in metric tons	
Indonesia	600,000
Thailand	185,000
Taiwan	45,000
Philippines	20,000

ble to keep the different products apart—and a feed kernel the size of a peppercorn can transmit BSE. From 1988 until 1996, Britain used the current U.S. standard. There was so much cross-contamination—in rendering plants, in feedlots and in barnyards—that an estimated 60,000 cattle were infected as a result. Would that system work any better here? The record isn't encouraging. Just six weeks ago a Purina outlet in Gonzales, Texas, called a feedlot in Floresville to explain that an employee had inadvertently spiked a recent shipment of cattle feed with offal intended only for other animals. More than 1,200 cattle were already munching the stuff when the call came.

There is no evidence that the feed was tainted. Even so, Purina purchased the affected cattle and pledged to keep them out of the food chain (presumably by destroying them). Purina has also announced that it will stop using sheep or cattle in any of its products. And the American Feed Industry Association is now pushing its members to create separate "production lines" within their factories to prevent such commingling. But these reforms are voluntary, and the feed companies' past record doesn't inspire confidence. In a study published last fall, the General Accounting Office found that 20 percent of the 1,700 U.S. companies handling both restricted and unrestricted offal "did not have a system ... to prevent commingling and cross contamination."

The United States lags in other areas as well. Studies (and common sense) suggest that brain-destroying prions are more likely to show up in meat torn from a cow's spinal column than, say, a rump roast. Though Britain now bars the sale of such high-risk tissues, U.S. law still permits it (unless the tissue comes from a high-risk country). An American hot dog, for example, can contain up to 20 percent "mechanically separated meat," which the government describes as "a paste-like and batter-like meat product produced by forcing bones with attached edible meat under high pressure through a sieve ..." And because dietary supplements remain largely unregulated in this country, their manufacturers can peddle the most potentially dangerous tissues as tonics. Atrium Inc.'s Brain 360 promises 360mg of "raw brain concentrate (bovine)." Atrium's Pituitary Whole provides 40mg of raw pituitary, also from cows. And PhytoPharmica's Adrenal-Cortex Fractions include raw bovine lung, heart, kidney, spleen and brain, all in one caplet.

Grotesque oddities or public-health threats? It's impossible to say. We tend to assume the best until confronted with evidence to the contrary. But if the story of BSE teaches us anything, it's that paranoia pays. Who would have worried about eating a cheap British burger in 1985? Who would have deprived a stunted child of a chance to grow tall in the 1960s or '70s? Wendy Nofi reached nearly five feet with the help of human growth hormone. She married, had three kids and lived happily in Bethpage, N.Y., until 1995, when she started losing her balance. "She always felt like she was on a boat, kind of rocking," her husband, Michael, recalls. You know the rest of the story. Her vision blurred. She stopped walking and swallowing and lost all bowel and bladder control. By the time she died in 1998, she had spent two years on a feeding tube. "There were absolutely no guidelines for screening the pituitaries," her husband now marvels. "Really, they didn't screen anything." Of course not. No one had gotten sick.

With ANNE UNDERWOOD *in New York,* ADAM ROGERS *in Washington,* ANDREW MURR *in Los Angeles,* KAREN SPRINGEN *in Chicago,* WILLIAM UNDERHILL *and* MICHELLE CHAN *in London and* SCOTT JOHNSON *in Paris*