vCJD: Broad U.S. Response Required

In the United Kingdom, a deadly pathogen has jumped species—from sheep to cow to human—emerging about 5 years ago in two teenagers as a new variant form of Creutzfeldt-Jakob disease (vCJD). The cause: eating beef products contaminated with bovine spongiform encephalopathy (BSE) or "mad cow" disease. Rising panic pervades the United Kingdom and continental Europe as the number of people afflicted with vCJD approaches 100, and BSE peppers the continent. We see herds of cattle being slaughtered, ministries of health under fire, import restrictions on beef and human blood products within a "common market," meat sales collapsing, and a UK mandate that disposable instruments be used for tonsillectomies.

Public fear and scientific uncertainty surround this new human disease. Surely, there is no worse ending for a vigorous young person than a progressively moth-eaten brain, directing a many-month demise through stages of tremors, agitated depression, episodic psychosis, ataxia, motor and sensory loss, blindness, incontinence, dementia, and death. The culprit, discovered by Nobel laureate Stanley Prusiner, is a mysterious infectious protein called a prion, which self-replicates without DNA or RNA. Prions are radically different from the bacteria, viruses, parasites, and fungi that medicine understands, and they can neither be detected premortem nor destroyed by sterilization.

We must act now to combat this new human disease.

Hovering silently within its victims for years, vCJD hides in lymphoid tissue (tonsils, Peyers patches of the gut, lymph nodes, and spleen) and at some point within B lymphocytes, before its explosive assault on the brain. This long silence raises the specter of undetected transmission to others through contaminated surgical instruments or blood transfusion—a specter imputed from the involvement of lymphoid tissue and supported by several studies in animals. Susceptibility seems limited to people with a homozygous methionine at the 129 codon of their prion gene, but that is still some 40% of the population. We don't know how the infecting prion commands host prion proteins to misfold and replicate billions of times, how it spreads to the brain and incites amyloid deposition, or why it causes seemingly indiscriminate neuronal cell death. Without a blood test, scientists cannot predict whether human infection will be a small geographically contained event or a major global epidemic.

In the wake of this new human disease that so far has spared our continent, the U.S. Department of Agriculture and the Food and Drug Administration have instituted controls to minimize the risk of BSE entering our medicinals and our food chain. Blood centers are taking the necessary but crude step of excluding donors who have lived in places where BSE and/or vCJD have been identified. We must, however, do far more.

Urgently needed are simple and reliable blood tests for screening of humans and animals. This work is being conducted mostly in commercial laboratories, and success is estimated to be 2 years off. Meanwhile, surrogate markers could be life saving, as they would have been during the HIV epidemic before specific blood tests for the virus emerged. Methods to destroy infective prions and to sterilize potentially contaminated equipment are vital. The United States should expand its relatively small national surveillance center to better ascertain the presence of vCJD and BSE in suspected cases. Now is the time to establish well-designed banks of tissue from routine tonsillectomies, appendectomies, and lymph node dissections—as the United Kingdom and Switzerland are doing—to define and follow the prevalence of prions in the U.S. population.

Finally, resources must be mobilized strategically, substantially, and urgently by the National Institutes of Health (which invested less than $14 million in prion research in fiscal year 2000) and the Centers for Disease Control to seek the basic, clinical, and public health knowledge that is unlikely to come from commercial laboratories. Major investment is needed to recruit and train talent and to set up dedicated biosafety laboratories, transgenic facilities, and research instrumentation. Throughout, we in the United States must learn from our colleagues in Europe who are ahead of us in facing this agricultural and public health crisis, and we must communicate in an open and measured way to the public we serve.

If these efforts turn out to be unnecessary, we have advanced medical science. If, however, they turn out to be needed, we will have taken steps none too soon.

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