CREUTZFELDT-JAKOB DISEASE: Notes about the Disease

Creutzfeldt-Jakob disease (CJD) is an invariably fatal degenerative neurologic disorder that, in North Carolina and the remainder of the world, affects about one person per million each year. It is one of several so-called transmissible spongiform encephalopathies (TSEs) affecting humans and/or animals caused by abnormal self-replicating proteins called "prions." Prions behave in many respects like infectious agents with long incubation periods and were once actually called "slow viruses." Until the link between bovine spongiform encephalopathy (BSE, a TSE of cattle) and a new variant of CJD was noted in England, these diseases were largely considered curiosities of minor public health significance.

Scrapie is a TSE of sheep and goats recognized since the mid-1700s. BSE likely arose from feeding cattle rendered carcasses of sheep contaminated with the scrapie prion. A change in the rendering process in the United Kingdom around 1980 permitted the scrapie agent to survive. The UK BSE epidemic that began in 1986 was followed in 1996 by the first recognized cases of an aberrant form of CJD that was named "variant CJD" (vCJD). Previously, three forms of CJD were recognized: sporadic (accounting for 85-90% of cases), familial (5-10%), and iatrogenic (<5%). Even before the etiologies of BSE and vCJD were linked through molecular epidemiology, a number of rigorous changes in agricultural practices in the UK and elsewhere—including bans on importing British beef into the US—were instituted. The epidemic curve for BSE peaked in 1992 and has continued to decline.[1] In addition to eating beef contaminated with bovine neural tissue, human blood transfusion is now considered a possible vCJD transmission route.

There are important clinical differences between the classical forms of CJD recognized worldwide since the 1920s and vCJD: vCJD affects younger patients than classical CJD, features prominent psychiatric/behavioral symptoms with delayed neurologic signs in contrast to the dementia and early appearance of neurologic signs in other forms of CJD, and has a more protracted course.[2] Intensification of surveillance for CJD in NC, in order to increase the probability of detecting any cases of vCJD, has included making all forms of CJD reportable. The few cases of BSE and vCJD detected so far in the US most likely were imported rather than acquired here, but careful surveillance must be continued, as should current stringent preventive agricultural and human health measures.

In dealing with the news media and public about CJD, it is important to avoid confusion between vCJD and the near half-dozen cases of sporadic CJD that occur in NC each year. Confirmation by brain tissue examination of anyone with suspected CJD/vCJD who is less than 55 years of age is even more important.[3]

1. P. Brown, et al., "Bovine Spongiform Encephalopathy and Variant Creutzfeldt-Jakob Disease: Background, Evolution, and Current Concerns," *Emerging Infectious Diseases* 7, no. 1 (2001): 6-16, www.cdc.gov/ncidod/EID/vol7no1/brown.htm.
2. "CJD (Creutzfeldt-Jakob Disease, Classic)," *Centers for Disease Control and Prevention*, 13 April 2007, www.cdc.gov/ncidod/dvrd/cjd/.
3. "Questions and Answers Regarding Bovine Spongiform Encephalopathy (BSE) and Variant Creutzfeldt-Jakob Disease (vCJD)," *Centers for Disease Control and Prevention,* 4 January 2007, www.cdc.gov/ncidod/dvrd/vcjd/qa.htm#surveillance.