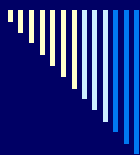


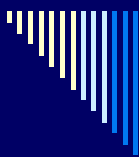
Bovine Spongiform Encephalitis

Epidemiology
Pathogenesis
Public Health



Iowa State University Extension Service

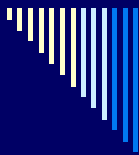
Nolan R. Hartwig, DVM
Extension Veterinarian
Professor, Department of Veterinary
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Transmissible Spongiform Encephalopathies

□ TSEs

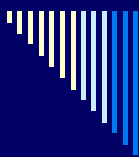
- Caused by abnormally folded prion proteins
- Prions accumulate in central nervous system
- Long incubation period
- Progressive dementia
- Invariably fatal
- Affect several species



Species Affected by TSEs

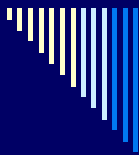
□ Sheep—scrapie

- First recognized in 18th century
- Progressive encephalitis
- Definite genetic predilection in sheep
- Much more widespread in U.K. than U.S.
- May have been original source of BSE infection in British cattle
- Eradication program in progress in USA
- Transmissible experimentally to other ruminants, primates, cats, variety of rodents



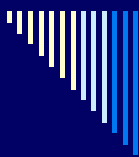
Transmissible Mink Encephalopathy

- Transmissible mink encephalopathy
- Long relationship between feeding sheep offal to mink and TME
- Rare disease
- Does not occur in wild mink population



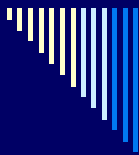
Chronic Wasting Disease of Deer and Elk

- Colorado, Wyoming, Nebraska, Wisconsin, Illinois (few cases), Canada
- First recognized in captive deer and elk
- Occurs in white tail and mule deer, elk
- Has spread fairly rapidly in wild population
- Method of spread unknown
 - Licking of birth fluids, placenta, dead animals, rutting season may all be involved
 - Direct injection of CWD agent into brain of cattle causes disease in small number—suggests caution is in order
- No evidence of human disease



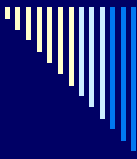
Human TSEs

- Kuru
 - “Fore” tribe of New Guinea
 - Spread related to ritualistic cannibalism or handling of dead
 - Rapidly disappearing since these practices stopped
- Gerstmann-Straussler Syndrome--Rare
- Creutzfeldt-Jakob Syndrome (CJD)
 - “natural,” sporadic dementia of older people
 - Worldwide
 - Prevalence of about 1/1,000,000



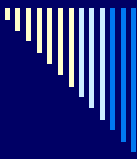
Human TSEs, Con't

- Variant Creutzfeld-Jakob Disease (vCJD)
 - First recognized in United Kingdom in 1996
 - Often strikes people much younger than CJD
 - Course of disease slower than CJD
 - Nature of causative prion undistinguishable from BSE
 - Prion different from sporadic CJD
 - 153 cases world-wide, 143 in U.K. 12/01/03
 - Incidence of new cases is declining in UK
 - No case where did not have history of living in a country where BSE occurred.



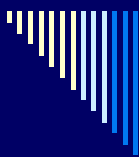
vCJD (con't)

- One case of vCJD in U.S. (Florida)
 - Victim native of U.K., spent many years there
- Epidemiology modeling at one point predicted there could be several thousand cases in U.K.
 - Number of actual cases better than “best case” projections
 - Some people may have very long incubation period
- May be human genetic susceptibility differences (codon 129 of PrP gene)



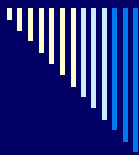
Bovine Spongiform Encephalopathy

- First appeared in U.K. in mid-80s
- Approximately 1000 cases/week in U.K. at height of epidemic
 - Approximate total of 180,000 cattle affected
 - 35,000 herds
- Found in about 19 countries
- Number of cases rapidly declining
- Usually only 1 or 2 cases per herd
 - Indicates perhaps major differences in susceptibility
 - Major losses for British cattle industry



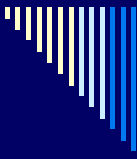
Other Species

- Can be transmitted to some laboratory animal species
- BSE found in domestic cats, transmissible to several other felines
 - Via raw food
 - Zoological parks have had some cases
- No TSEs in poultry or avian species
- No TSEs in swine
- No TSEs in dogs, horses
- Transmitted experimentally to mice, mink, sheep, pigs, non-human primates



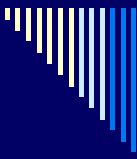
Transmission and Pathogenesis of BSE

- Prions are proteins
 - Contain no DNA
 - Still some controversies about prions
- Disease producing prions have abnormal architecture (folding)
 - Prevents enzymatic breakdown
 - Serve as template to modify “new” prions
- Found primarily in nervous tissue
 - Brain, spinal cord, dorsal root ganglia, distal ileum, trigeminal ganglion
 - “Exact degree of infectivity in lymphoid tissues has been impossible to determine due to lack of a sensitive infectivity test.” C.I. Lasmezas, Rev. Sci. tech. Off. Int. Epiz., 2003, 22, 23-36.
 - Apparently fewer BSE prions in peripheral nerve tissues than scrapie



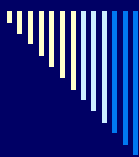
Diagnosis of TSEs

- Immuno-histochemistry (IHC)
 - Considered gold standard
 - Laborious, takes 3-4 days
 - Requires trained pathologist
- Histopathology
 - Indicative, highly accurate
 - Time consuming, requires trained pathologist
- Screening tests:
 - “Quick” tests under development
 - Western blot, others
 - Under development
 - Used for screening for Scrapie in sheep



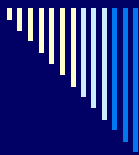
Pathogenesis

- Highest concentration in brain stem
- Absorbed from gut, transferred to CNS via nerve tissue
- Prions not found in:
 - Milk
 - Blood
 - Muscle
 - Connective tissues
 - Glandular tissues other than possible lymph nodes associated with lower gut



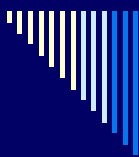
BSE Epidemiology

- Primary transmission by ingestion
- Possibly first developed in cattle population by consuming meat and bone meal from scrapie infected sheep
- May have developed spontaneously in cattle, perhaps many years ago
 - This is a very important “unknown”
- Rarely found in cattle less than 36 months of age



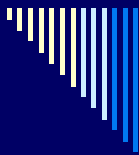
BSE Epidemiology

- British rendering processes were much lower temperature than traditionally used in USA
- Prions are extremely heat resistance
- May 20 case in Canada and Dec. 9 case in U.S.A. may be due to feeding contaminated meat and bone meal
 - Unknown at this time



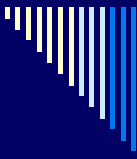
Preventive Measures in U.S.A.

- Banned import of cattle, beef, organ meats from affected countries when diagnosed
- Monitoring of U.S. cattle population
 - 20,000 per year for 2 years
 - Most samples from downer animals presented for slaughter at federally inspected plants
 - Sampling frequency designed to detect 1/1,000,000 with 95% confidence
- 1997 ban on feeding meat and bone meal to ruminants



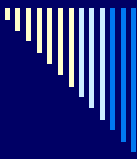
BSE and Food Safety

- Prions found in brain, spinal cord, associated nerves
 - Not in muscle
 - Cannot say with 100% assurance that there could be no nervous tissue in meat
 - BSE prion not found in sciatic, other peripheral nerves
 - Brain, spinal cord now removed in cattle older than 30 months



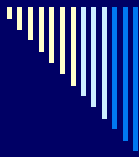
BSE and Food Safety

- Ban on harvest of any downer animals
- Risk for vCJD in U.S.
 - Not zero
 - So small that is impossible to estimate the odds ratio
- This predicated on control of BSE in U.S. cattle population
 - Harvard study, epidemiologic projections, experience in U.K. indicates it will be controlled at very low levels in U.S. cattle population, probably eradicated



BSE Concerns

- Difficult disease to understand
- Need research on translocation of prions in nervous system, other tissues
- Still need to know more about prions
 - Some disagreement by research scientists about some aspects
 - Stanley Prusiner received Nobel Prize for identification of prions as causative agent
- Need accurate, quick screening test



Concerns

- Scientific understanding of epidemiology and pathogenesis
 - Consumer understanding
 - How the science is presented to the public
 - Sensationalism
 - People with “another” agenda
 - Politics of international trade
-