

ACKNOWLEDGMENT

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Letter to the Editor:

Dear Dr. Blizzard and Editors of GGH:

I've been reading GGH for years, and have found it so useful. This month's timely release of the intersex review really "hit the mark". I work in a state birth-defects surveillance department. The non-physicians have expressed tremendous interest in the management of ambiguous genitalia, either as an isolated finding, or related to exstrophy. This review will serve as the focal point for our next monthly teaching session to be supplemented by your review (*GGH* Vol. 19, No. 1).

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Dear Dr. Linn: Thank you!

Dear Other Readers:

Please let us know your positive and negative comments – and your agreements or disagreements regarding the abstracts and their comments and the lead articles. Your input is absolutely necessary for us to maintain, upgrade, and disseminate your agreements and disagreements. We encourage you to respond quickly after your thoughts and criticisms come to mind.

Robert M. Blizzard, MD

Abstracts from the Literature

Factors Determining the Pattern of the Variant Creutzfeldt-Jakob Disease (vCJD) Epidemic in Great Britain

Editorial Preface: Growth hormone (GH) extracted from human pituitaries obtained at autopsy was first given to children in 1958. Twenty-seven years later (1985), the first cases of Creutzfeldt Jakob Disease (CJD) resulting from such injections were observed in individuals who had received GH injections 8 to 10 years prior to that time. The fact that no cases of CJD were reported reflects the long latent period between exposure and the onset of symptomatic disease.

The exact number of the pituitary injections that may have been contaminated with the CJD prion is unknown. GH from only one of three laboratories in the U.S. extracting pituitaries has been associated with CJD. All three of the laboratories extracting GH used different procedural techniques. In retrospect, the GH extraction procedure of two of the three laboratories eliminated the active prion from the final product. From 1985 until April 2003 only 26 cases of CJD were recorded among several thousand (7,700) recipients in the U.S. who had received native human growth hormone. All U.S. patients with CJD received GH prior to 1977; afterward a new purification step was added to the GH extraction procedure.

The early symptoms of CJD consist of degenerative neurological function. Death unfortunately follows within a period of 6 to 36 months. The number of catastrophes to date in the United States have been relatively small, particularly in light of the number anticipated in 1985 when the first two deaths were reported within a month of each other. Postulation, with reasonable justification, was that the incubation period and susceptibility to the disease were influenced by the dose of contaminated material, possibly the age of the recipient, and possibly by an individual's genetic susceptibility. The latter was suspected on the basis of a few studies using scrapie disease in sheep as a prototype since CJD, occurring primarily in humans, is similar to scrapie disease in sheep. These diseases produce degenerative neurological alterations; although the histology of the pathological findings in the central nervous system are different. They are known as spongiform cerebral encephalopathies.

Abstract: In 1985 and 1986 a similar but different spongiform encephalopathy manifested itself in England when humans were first diagnosed with "mad cow