Creutzfeldt-Jakob Disease: Hypothesis for High Incidence in Libyan Jews in Israel

We would like to suggest a possible link between two recently reported findings concerning Creutzfeldt-Jakob disease (CJD).

Kahana *et al.* (*1*) investigated the incidence of CJD among Jews of different origin now living in Israel. They found a remarkably high incidence among Libyan Jews (31.3 per million) in contrast to other groups. Iraqi Jews had an incidence of 1.9 per million; Jews from western and central Europe, 1.0 per million; and native-born Israelis, also 1.0 million.

This finding may be related to the dietary habit of eating sheep's eyeballs, which are a gastronomic delicacy among Bedouin and Moroccan Arabs and also Libyans. A disease of sheep, scrapie, has clinical and histopathological features similar to those of CJD, and the two diseases have been included in the group of subacute spongiform virus encephalopathies (*2*). If the CJD agent is found in the cornea, retina, or optic nerve, the ingestion of eyeballs of sheep harboring the scrapie agent might possibly lead to the development of CJD in susceptible individuals and thus account for the high incidence of the disease in Libyan Jews. Many investigators have wondered whether the virus of scrapie might be identical to the virus of CJD. However, there is no laboratory study that can yet be used to determine this possibility.

This suggestion was prompted by a report of the apparent human transmission of CJD by corneal transplantation (*3*). A 55-year-old male died of pneumonia after a 2-month history of progressive failure of memory, with ataxia, involuntary movements, and myoclonus. The diagnosis of CJD was not made until after death. The recipient of this patient's cornea was a 55-year-old female who developed symptoms 18 months later. She became lethargic and ataxic, with increasing neurological deterioration manifested by dysphagia, mutism, and myoclonus. She died 8 months after the onset of symptoms, and the autopsy showed the characteristic changes of CJD, as did autopsy of the donor.

Kahana *et al.* (*1*) note the rarity of CJD and consider it unlikely that its occurrence in both the donor and the recipient of the graft was merely coincidental. They suggested that proof of a cause-and-effect relationship required other studies. There is as yet no proof that the CJD virus is present in corneal cells. In the surgical procedure of removing the eyeball from a donor and storing it on saline-moistened gauze in a sterile container until the cornea is taken for transplantation, the cornea is likely to become contaminated with virus present in the optic nerve. In an attempt to provide such proof, the corneas of a moribund capuchin monkey with experimental CJD were transplanted to two healthy capuchin monkeys in this laboratory. This procedure was performed on 2 April 1974, and the grafts have taken well. Signs of disease in the recipient cannot be expected before many months, for 31 months elapsed between the intracerebral inoculation of CJD material into the donor monkey and the appearance of clinical signs. In capuchin monkeys, incubation periods of CJD have ranged from 11 to 39 months.

L. HERZBERG, B. N. HERZBERG
C. J. GIBBS, JR., W. SULLIVAN
H. AMYX, D. C. GAJDUSEK
Laboratory of Central Nervous System Studies, National Institute of Neurological Diseases and Stroke, Bethesda, Maryland 20014

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The exotic food habits of people in other cultures never cease to amaze "enlightened" Westerners. This may explain why strange food so often feeds our speculation about disease. In calling attention to the unusually high incidence of Creutzfeldt-Jakob disease (CJD) among Libyan Jews as compared to several other national groups in Israel, we, like Herzberg *et al.*, considered food habits as a possible explanation.

We knew that brain and spinal cord, mainly from sheep, was a delicacy among Libyan Jews. Inquiries even revealed that a favorite method of preparation is light grilling, which could conceivably leave an infectious agent viable. However, ingestion of grilled nervous tissue is not limited to Libyan Jewish immigrants in Israel. It is eaten by all North African groups and quite a few European groups as well. Therefore, we were reluctant to suggest that this food habit could account for the excess of CJD observed only among Libyan Jews, so we deleted reference to it in our final manuscript.

A word of caution is in order before moving from speculation that CJD might be transmitted by corneal transplantation to speculation that ingestion of tissues can transmit this disease. I recently discussed evidence on modes of natural transmission of slow viruses and emphasized that a distinction should be drawn between transmission by *inoculation* (or transplantation) and by *ingestion* (*1*). Although good evidence exists that slow virus diseases may be transmitted by inoculation, the evidence that these diseases may be transmitted by eating infected tissue is tenuous in man. Even in animals, for which there is better evidence for occasional oral transmission (*2*), transmission is far more reliable by intracerebral, intraperitoneal, or intramuscular inoculation.

The suggestion of Herzberg *et al.* is nonetheless appreciated, and questions about eyeball consumption will be incorporated in a planned epidemiological study of CJD among groups of different national origin living in Israel. Quantitative considerations alone, however, make brain a more likely source of the putative CJD agent than eyeballs.

MILTON ALTER
Epidemiology and Genetics Unit, Department of Neurology, University of Minnesota, Minneapolis 55455

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L. Herzberg, B. N. Herzberg, C. J. Gibbs Jr., W. Sullivan, H. Amyx, D. C. Gajdusek and Milton Alter

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