MEDICAL PRACTICE

For Debate . . .

Bovine spongiform encephalopathy

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Press announcements released last year about an outbreak of a brain disease, spongiform encephalopathy, in the cattle of south west Britain were received with alarming indifference by the medical profession as well as by the general public. Fears that transmission of the disease to man might occur through the sale of animal products were immediately allayed by reassurances largely from the veterinary profession, but no contribution was made from the food industry, and the basis for this confidence was not adequately explained. It has generally been accepted that the slaughter of animals showing characteristic signs of infection—such as behaviour changes—as well as the usual processes of sterilisation and pasteurisation, are enough to remove any risk to the consumer. Unfortunately, this is a view that is naive, uninformed, and potentially disastrous.

We are dealing with a condition thought to be related to Creutzfeldt-Jakob disease, scrapie, and kuru—all forms of degenerative encephalopathy—which are transmissible by artificial inoculation and, in the case of kuru, by dietary means. Kuru is notorious for its probable transmission by cannibalism among the Fore people of New Guinea. It is said that the sharing out of the various parts of the carcass led to a more common occurrence of the disease in women and children, the brains being given to them in preference to the adult men. The epidemiology of this disease has provoked much controversy, but few doubt that kuru can be transmitted through dietary means, even if the importance of this is not clear. Before the abolition of cannibalism in New Guinea kuru had become the commonest cause of death among the Fore people.

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Fatal course of disease

Creutzfeldt-Jakob disease has a much wider distribution than kuru, cases having been reported world wide. Spongiform degeneration occurs in the infected brain in the absence of an inflammatory response,[1] no antibodies appearing in the serum or in the cerebrospinal fluid. The incubation period is variable, but may be several years. Once symptoms begin the disease runs a progressive and invariably fatal course. The transmissible agent of Creutzfeldt-Jakob disease can apparently withstand high temperatures and sterilisation with formaldehyde. Transmission to man has occurred by mistake in a variety of circumstances. One case was known to be the result of corneal grafting,[2] another was due to transmission by deep cortical electrodes which, despite sterilisation with formaldehyde vapour, passed on the Creutzfeldt-Jakob disease agent from one patient to another.[3] A further example is the few cases of Creutzfeldt-Jakob disease thought to be related to the use of human growth hormone derived from pituitary glands removed at necropsy. In this instance the number of cases is small, but its possible implications, both medical and legal, led to the withdrawal of human growth hormone from the market at a time when no satisfactory alternative was available.[4]

Scrapie is a disease of sheep known at least since the eighteenth century. Several reports of possible transmission to man (resulting in Creutzfeldt-Jakob disease) are found in the literature, but all are based largely on circumstantial or anecdotal evidence.[5-7] The incidence of Creutzfeldt-Jakob disease is 30 times higher than expected among Libyan Jews in Israel, and one possible explanation is their consumption of sheep's eyeballs, as well as other delicacies containing brain extracts.

Bovine spongiform encephalopathy was first recognised and recorded as a clinical entity in November 1987, although media reports at the time stated that cases had been known as early as December 1986. The disease is not mentioned in the HMSO publication *Animal Health 1986*, and the 1987 edition is not yet available. Unpublished information from the press branch of the Ministry of Agriculture, however, reports 246 cases of the disease since records began in November 1987. These cases were distributed among 223 farms in England and Wales, and these figures were last

updated in February 1988. New cases are still being diagnosed.

Bovine spongiform encephalopathy has been observed in both pedigree and commercial herds and is most common (according to reported cases) in the Friesian and Holstein cattle. Apart from one single case report of a bull with the disease, all other animals have been female and usually, but not invariably, milking cattle. The life span of a milking cow is considerably longer than that of animals bred for meat.

Bovine spongiform encephalopathy is diagnosed only on necropsy evidence. Farmers who own and rear cattle are not obliged to report suspected cases, but if the animal becomes unmanageable they can ask for it to be culled. It is the farmers who own the cattle who are responsible for bringing cases to attention, and they have a financial incentive to defer the diagnosis. The official guidelines are that if a farmer suspects, for instance, a milking cow to be afflicted, he has two alternatives. Either he can carry on milking the animal until it becomes unmanageable (at which point it can be culled), or he can send it straight to the abattoir for use in the meat industry. Undoubtedly many infected cattle have been used to make meat products, and the reported numbers only represent those animals with well established clinically manifest disease.

Use of brain derived products

Most of the epidemiological work concerning the dietary basis for the transmission of slow viral encephalopathies has assumed that the brains are found only in certain local delicacies.[7] This prompted us to investigate the use of brain derived products in the British food industry.

At first sight it seems as if the law is fairly tight. Brain is classified as a "prohibited offal," and cannot therefore be included in uncooked meat products,[8] including sausages. No regulation, however, prohibits the inclusion of brain in cooked products (such as meat pies), although this offal cannot then be counted as meat in the ingredients list. (The incentive from the manufacturer's point of view is therefore restricted to the added bulk gained rather than a higher meat content.) Coming from an industry not exactly renowned for its genuine concern over public health, this is little comfort to those of us aware that the Creutzfeldt-Jakob disease agent can survive high temperatures. Moreover, the maximum penalty for breaking the law is only £1000. The chances of being caught are also small, prosecutions resulting usually from sporadic complaints on behalf of individual consumers. As well as the possible contamination of supermarket products two other factors cause concern.

Firstly, it has always been possible to buy raw brain over the counter in butchers' shops as an ingredient for stews and casseroles, quite popular among elderly people. Secondly, other tissues such as spinal cord, which might conceivably contain the transmissible agent, are commonly included in such items as pork chops. This is not illegal because the food is "meat" rather than a "meat product." It seems as if the law protects us only against the inclusion of brains in processed foods. It is legal therefore for a large scale caterer to buy brains from a butcher to use as an ingredient in, for instance, canteen food.

In summary, we are faced with the fact that spongiform encephalopathy, whether or not we are at risk from it ourselves, is now established in the cattle of this country. This is a disease for which there is no serological marker, and the incubation period is probably long. There is no way of telling which cattle are infected until features develop, and if transmission has already occurred to man it might be years before affected individuals succumb. It is possible, but unproved, that many asymptomatic cattle are nevertheless as infective as those symptomatic animals which are immediately destroyed for public health reasons. So should not the use of brains in British foods be either abolished outright or more clearly defined? Then in the absence of more compelling evidence those of us who wish to exclude it from our diets at least have that choice.

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MATERIA NON MEDICA

A slice of the sweet life

Carnival had come to Tolfa that year. The news came in a telephone call from my uncle in his villa in Via San Giovanni decollato. I was to meet him there the following morning at 9 o'clock sharp. He was making one condition, however. I was not to ask for money or favours.

My uncle had spent most of the second world war languishing in a Roman jail. He had been found in possession of a British passport. But now he was a citizen of Rome and, what's more, he was mayor of Tolfa. I did not dare to argue with an uncle who also happened to be a mayor. So, as dawn broke, from my apartment by the Piazza San Pietro near the Vatican I rode north to Tolfa.

Carnival was already in full swing, the pazzarielli cavorting. The pazzarielli are an eighteenth century Neapolitan invention, a kind of singing commercial by a troupe of four entertainers. They tour the cities and large villages of southern Italy, advertising in sound and movement. As I arrived, they were singing the praises of white wine, strong and fresh from the slopes of the great Vesuvius and sweet as a virgin's milk. The children screamed with glee and anticipation. The pazzarielli were their heroes.

I walked away in search of a cool drink, pushing my Vespa. I met a girl who told me her name was Maria. We talked, or tried to. You see, I had learnt my Italian the previous summer, walking the narrow streets of Rocca di Papa and Frascati trying to write an article on wine for the *Times of Malta*. It seemed Maria spoke the dialect of Tarquinia. Communication was soon made worse by her mother appearing as if from nowhere. She obviously was acting as chaperone. She told me her name was Lucia. I think she tried to tell me also how she was born in Vinci, home of the great Leonardo, that her brother was the village parroco, and how life in Tolfa mimicked a kind of nineteenth century dolce vita. To cap it all, she boasted that she was a personal friend of the mayor. I tried not to blush.

I attempted to tell her of my humble origins in a Maltese hamlet, but also how I came to be born under the shadow of the Basilica of San Giovanni, no less. I was trying to write a "Penguin" on Italian wine while studying to become a medico. I had an urgent appointment in Tolfa and already I was very late. Would she and her beautiful daughter excuse me?

Lucia, however, was in no mood for listening. Her face was beginning to light up. Would I care to be introduced to the mayor in case I needed money or I really preferred to become not a medico, but a giornalista, perhaps?

My finals seemed a million years away.—JOSEPH BRIFFA, senior clinical medical officer, Essex.