Introduction

The transmissible spongiform encephalopathies (TSEs) are a group of neurodegenerative diseases caused by novel infectious agents, called prions. Stanley Pruisner first reported them in 1982 and described them as “proteinaceous infectious particles”. The infectious agent is the prion protein (PrP), a glycoprotein, which is remarkably stable and resistant to breakdown within cells due to its tertiary structure. Once prions enter a brain cell, they accumulate to form amyloid-like deposits. This leads to degeneration and death of the cells, causing voids to occur within the brain (like a Swiss cheese or sponge), hence the term spongiform encephalopathy. They are also remarkably resistant to conventional methods of disinfection and sterilisation.

The human TSEs are:

- Creutzfeld-Jacob disease (CJD)
- Gerstmann-Straussler-Scheinker syndrome (GSS)
- Fatal familial insomnia (FFI)
- Kuru.

The CJD group is further divided into classical sporadic, familial, iatrogenic and (new) variant (nvCJD or vCJD) subtypes. vCJD is thought to be linked to bovine spongiform encephalopathy (BSE) in cattle and was first described in 1995. It is suggested that vCJD may be transmitted by ingesting foods from BSE-infected cattle. Person to person transmission of TSEs does not occur.2,3

All human TSEs are very rare. The global incidence of CJD in all its forms is of the order of one per million people per year. Two other TSEs, (GSS and FFI) are exceptionally rare, with an annual incidence of 1:10-100 million and appear to have an hereditary basis for transmission. Finally, Kuru has been known about since the 1950s, and was first described in the Fore indian tribe of Papua, New Guinea. It is associated with cannibalistic funeral rites where the entire body is eaten (including the brains) of infected individuals. Over 2,000 cases had been reported before 1958, when ritualistic cannibalism ceased. Most cases were seen in women, because, in the hierarchy of the Fore society, they were left to eat the offal and the brains of the victims (see Table 1).

Transmission of prion diseases in the health care setting

Iatrogenic transmission of CJD has been reported via neurosurgical and ophthalmic operations and by the administration of growth hormone derived from infected human pituitary glands. There is the theoretical possibility that vCJD may also be transmitted by these routes, although there are no reported cases to date. Iatrogenic transmission of CJD has been associated with:

- Dura mater grafts (~110 cases worldwide)
- Human cadaver pituitary-derived hormone (~130 cases worldwide)
- Contaminated medical equipment (~seven cases worldwide, two probable, five possible)

The majority of dura mater graft cases all relate to one particular commercial product (“Lyodura”). This product was withdrawn from the Irish market in 1987. It is possible that the product may have been used in a small number of neurosurgical procedures up until 1993.4

There have been three cases of CJD associated with corneal transplantation (one definite, one probable, one possible). A detailed risk assessment is currently underway in the United Kingdom to determine if receipt of a corneal graft should be considered a risk factor for development of CJD.5,6

One iatrogenic case of CJD was reported in Ireland in 2001, which was linked to prescription of human pituitary-derived growth hormone.

There is some evidence that other surgical procedures could potentially be associated with iatrogenic vCJD transmission. Unlike CJD the vCJD prion has been identified not only in brain and neural tissue, but also in peri-oral tonsillar tissue and spleen. The incubation period for vCJD is unknown and

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