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Managing the risk of iatrogenic transmission of Creutzfeldt–Jakob disease in the UK

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SUMMARY

Background: With the emergence of bovine spongiform encephalopathy (BSE) and variant Creutzfeldt–Jakob disease (CJD) in the UK, there is concern about iatrogenic transmission, and the approach to managing this risk is unique.
Aim: To describe and review CJD incident management and the notification of individuals 'at increased risk' as a strategy for reducing iatrogenic transmission.
Methods: A description of iatrogenic CJD transmission, the CJD Incidents Panel's role, the number and nature of CJD incidents reported and the individuals considered 'at increased risk' by mid-2012.
Findings: Seventy-seven UK cases of CJD are likely to have resulted from iatrogenic transmission, among recipients of human-derived growth hormone (64 cases), dura mater grafts (eight cases), blood transfusions (four cases) and plasma products (one case). To limit transmission, the Panel reviewed 490 incidents and advised on look-backs, recalls of blood and plasma products, and quarantining and disposing of surgical instruments. Additionally, on Panel advice, around 6000 asymptomatic individuals have been informed they are at increased risk of CJD and have been asked to follow public health precautions.
Conclusion: The strategy to reduce iatrogenic transmission of CJD has been developed in a context of scientific uncertainty. The rarity of transmission events could indicate that incident-related exposures present negligible transmission risks, or – given the prolonged incubation and subclinical phenotypes of CJD – infections could be yet to occur or have been undetected. Scientific developments, including better estimates of infection prevalence, a screening test, or improvements in decontaminating surgical instruments, may change future risk management.

Introduction

In the late 1990s, with fears of a large epidemic of variant Creutzfeldt–Jakob disease (vCJD) in the UK following widespread exposure to bovine spongiform encephalopathy (BSE), the need for effective measures to reduce transmission of both vCJD and CJD through healthcare procedures (iatrogenic transmission) was recognized by the UK Health Departments.

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Whereas there have been many fewer vCJD cases than initially feared, and although the annual incidence of vCJD has declined since 2000, the continued incidence of sporadic and genetic CJD (Figure 1) and possible subclinical vCJD infections give serious cause for concern about iatrogenic transmission.[1–4]

All forms of CJD (variant, sporadic, and inherited prion disease) could be transmissible through the re-use of instruments that have had contact with potentially infectious tissue during specific procedures, as prions are resistant to conventional decontamination processes. In vCJD as tissue infectivity is more widespread, more procedures pose a risk of transmission.[5] Tissue and organ donations from infected individuals are another potential transmission route. Transmission of sporadic CJD has been reported among recipients of tissue transplants (dura mater, corneas) and of human-derived pituitary hormones (growth hormone and gonadotrophin).[6–8] Blood and plasma products are an additional transmission route in vCJD only.[9–14]

The UK strategy to reduce secondary transmission has focused on measures to safeguard donated blood and plasma products from vCJD. The strategy includes the introduction of universal leucodepletion and the importation of plasma to manufacture plasma products. To reduce the risk associated with surgery, guidance on infection control, instrument traceability and neurosurgical practice has been introduced.[15–17]

This paper aims to describe and review CJD incident management and the notification of individuals 'at increased risk' as a strategy for reducing iatrogenic transmission. This key aspect of the UK strategy is overseen by the Advisory Committee on Dangerous Pathogens Transmissible Spongiform Encephalopathy Risk Management Subgroup and, until recently, its associated committee, the CJD Incidents Panel.

Methods

Role of the CJD Incidents Panel

The CJD Incidents Panel (the Panel), which sat from 2000 to 2013, was composed of scientific and medical experts, ethicists, and lay people. The Panel was responsible for advising healthcare providers on managing incidents in which individuals may have been exposed to CJD risks through healthcare.[18] In March 2013 the Panel was dissolved, and responsibility for investigating, assessing, and managing CJD incidents (and where appropriate notifying patients) now rests with local hospitals and healthcare providers, health boards, and health protection teams. Public Health England provides assistance and advice as required.

The Panel also defined categories of individuals who can become 'at increased risk' following iatrogenic exposure. Individuals are 'at increased risk' if they are considered to have $\geq$1% risk of CJD in addition to the background risk in the UK population, following specific iatrogenic exposures, or if they are at risk of genetic forms of CJD. The 1% threshold level is used as a cut-off for implementing public health precautions and is not intended to be a precise measure of an individual patient's risk. The ten iatrogenic exposures which have led to individuals being considered 'at increased risk' of CJD are described in Table I.

Blood incidents

Blood and plasma are only considered transmission routes for vCJD. Blood incidents occur when an individual diagnosed with clinical vCJD previously donated or received blood components. This is identified by NHS Blood & Transplant (NHSBT) and the National CJD Research and Surveillance Unit, who run the Transfusion Medicine Epidemiological Review (TMER) study. Three patient groups are considered to have an increased vCJD risk following blood incidents and should be notified: recipients of blood from donors who later develop vCJD; blood donors to vCJD cases, and recipients of blood from donors to vCJD cases.[19–22] The latter two groups are considered 'at increased risk' as the donor to a vCJD case is considered to be a potential source of the recipient case's infection, and, if so, would be incubating the infection themselves and could have infected other blood recipients.

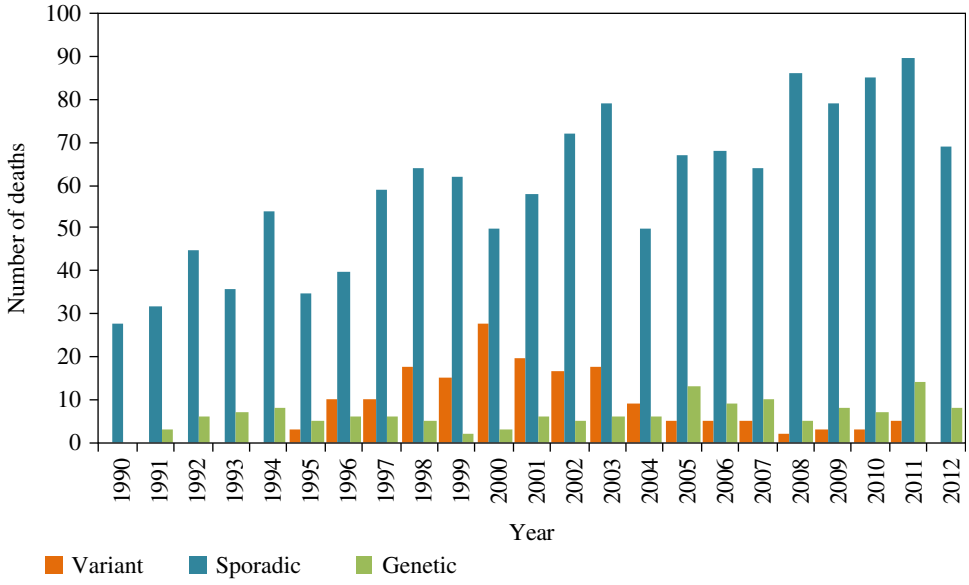

Figure 1. Deaths from Creutzfeldt–Jakob disease (CJD) in the UK 1990–2012 (confirmed and probable diagnoses). Data source: UK National CJD Research & Surveillance Unit. Data correct on 3 December 2012.