

Notification and support for people exposed to the risk of Creutzfeldt-Jakob disease (CJD) (or other prion diseases) through medical treatment (iatrogenically) (Review)

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[Intervention Review]

Notification and support for people exposed to the risk of Creutzfeldt-Jakob disease (CJD) (or other prion diseases) through medical treatment (iatrogenically)

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ABSTRACT

Background

Creutzfeldt-Jakob disease (CJD) and variant CJD (vCJD) are rare and always-fatal diseases transmissible via certain medical procedures. If a person is exposed to the disease risk through medical treatment, they may need to be notified of this to prevent them passing the risk to others in healthcare settings and to enable additional infection control measures to be put in place for certain procedures. As CJD is incurable, and unable to be screened for or effectively treated, communicating this risk information after an exposure incident may have significant implications for the person at risk, their families/ carers and healthcare professionals. The best ways to notify people of their exposure to the risk of CJD or vCJD, and to support them subsequently, are currently unknown.

Objectives

To evaluate the effects of interventions to notify and support consumers (patients and their family members or carers) in situations where exposure to the risk of CJD or vCJD has occurred as a result of medical treatment (iatrogenically), on consumer, healthcare provider and healthcare system outcomes.

Search strategy

We searched the Cochrane Consumers and Communication Review Group Specialised Register (10 February, 2009), the Cochrane Central Register of Controlled Trials (CENTRAL, *The Cochrane Library*, Issue 1 2009), MEDLINE (OVID SP), EMBASE (OVID SP), PsycINFO (OVID SP), CINAHL (EBSCO Host), Current Contents (OVID SP) and Dissertation Abstracts (Proquest) from start date to February 2009. We searched MEDLINE In-process and Other Non-indexed Citations (OVID SP) and Sociological Abstracts (CSA) in November 2009. We searched reference lists, websites, and contacted consumer groups and experts for details of relevant research.

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Selection criteria

Randomised and quasi-randomised controlled studies, controlled before-and-after studies and interrupted time series analyses assessing the effects of any intervention to communicate with (notify or support) people exposed to the risk of CJD or vCJD through medical treatment were included. We sought outcomes relevant to consumers, health providers and health services, including both benefits and harms.

Data collection and analysis

Cochrane review

Two authors independently assessed studies for inclusion against selection criteria, and would have applied standard Cochrane review methodology were any studies identified.

Thematic synthesis

We also conducted a thematic synthesis by systematically identifying and screening those studies that met the same population, intervention and outcome criteria as the Cochrane review, but that were identified from the broader literature providing evidence on policy implementation and consumer experiences. We systematically extracted and synthesised the data from these studies to produce a thematic synthesis, presented in appendices to this Cochrane review, which assembles evidence on the views, experiences and acceptability of notification and support strategies for people at risk.

Main results

Results of the Cochrane review

No studies meeting the study design criteria were identified for inclusion in this Cochrane review.

Results of thematic synthesis

In total, 49 studies and pieces of literature meeting the same population, intervention and outcome criteria as the Cochrane review, but identified from the broader literature providing evidence on policy implementation and consumer experiences, were included and formed the basis of a thematic synthesis, and which is presented in appendices to this Cochrane review. The thematic synthesis indicates that ideally communication may be considered as a longitudinal multicomponent programme, ensuring that notification and support are coordinated; that communication is tailored and responsive to need; and that activities to support individual risk communication, such as widespread education and monitoring of access to health care for those at risk, are in place. The thematic synthesis also indicates that poor communication practices may have negative impacts or cause harm, such as discrimination in accessing health care.

Authors' conclusions

There is insufficient rigorous evidence to determine the effects of interventions to notify people at CJD or vCJD risk and to support them subsequently, or to identify the best approach to communication in these situations. The thematic synthesis can be used to inform policy and practice decisions for communicating with people at risk in the absence of rigorous evaluative studies.

PLAIN LANGUAGE SUMMARY

Notification and support for people exposed to the risk of Creutzfeldt-Jakob disease (CJD) (or other prion diseases) through medical treatment

CJD and variant CJD (vCJD) are rare and fatal diseases with very long incubation periods. While CJD can occur spontaneously or genetically, this review focuses on those forms of CJD and vCJD in which the disease risk has been acquired through one of a small number of specific medical procedures (iatrogenically), such as brain surgery or the use of human pituitary hormone products.

Routine healthcare sterilisation techniques may not completely destroy the infectious agent responsible for causing CJD or vCJD. If a person is exposed to the disease risk through medical treatment, they may need to be notified of this to prevent them passing the risk to others in healthcare settings, and additional infection control measures may be needed for some procedures. As there is no screening test or effective treatment for CJD or vCJD, communicating this risk information after a healthcare incident where exposure to risk occurred may have a significant impact on the person at risk, their family members or carers, and their healthcare professionals. With no way to test for CJD or vCJD, those exposed to risk can be told only that they are at an increased risk of developing the disease. The

seriousness of the disease and the uncertainty of the risk can make notification a highly distressing event, and may cause significant anxiety and psychological harm. Notification of risk and living with at-risk status may also lead to ongoing problems over time, such as delays or discrimination in accessing routine or other health care.

For these reasons, communication about CJD and vCJD risk must be considered carefully. The risk information must be conveyed in a way that causes the least harm to individuals and their families. However, the best ways to notify people of their exposure to the risk of CJD or vCJD, and to support them after notification, are not known. These questions were the subject of this review.

That CJD and vCJD are very rare diseases creates challenges for researchers aiming to conduct rigorous quantitative studies in this area. Although we searched widely to identify all relevant research evaluating the effects of interventions to communicate with (notify and support) people at risk, we did not identify any studies that met the criteria for inclusion in this Cochrane review.

However, systematic searches did identify a number of pieces of relevant research and literature that provided evidence about policy implementation and consumer experiences in situations of iatrogenic exposure to risk. This research formed the basis of a thematic synthesis. The synthesis identified several activities that aim to improve the experiences of people at risk of CJD and vCJD. It indicates that communication may be best considered as a longitudinal multicomponent programme occurring over time, ensuring that notification is coordinated and considers impact; that support is in place and is offered over time; that communication is flexible, tailored and responsive to need; and that supporting activities, such as widespread education of the healthcare workforce, the public and the media, and monitoring of access to health care for those at risk, are in place. The thematic synthesis also indicates that poor communication practices may have negative impacts or cause harm, such as discrimination in accessing health care. In the absence of rigorous evaluative studies the results of this thematic synthesis can be used to inform policy and practice decisions for communicating with people at risk.

BACKGROUND

This review evaluates the effects of interventions to notify and support consumers exposed to the risk of Creutzfeldt-Jakob disease (CJD) through medical treatment. We planned this review knowing that it may not identify any relevant studies for inclusion. However, it is an important aim that interventions for notification and support should be identified, described, categorised, evaluated and this data assembled for summary; an aim that is facilitated by the preparation and updating of Cochrane reviews. It cannot be assumed, for instance, that all interventions for these purposes are equally beneficial in terms of their effects, or conversely, that they are without harms.

The aim of this Background section is to describe in depth the population of people 'at risk' of CJD which form the focus of this review. Becoming a person 'at risk' arises from several complex and overlapping treatment circumstances, yet the population shares sufficiently similar experiences to be considered collectively. We also aim to describe the ethical issues implicated in interventions targeting this at-risk population, and to outline the complexities that must be taken into account in providing and evaluating interventions to notify and support people at CJD risk.

Creutzfeldt-Jakob disease and other human prion diseases

CJD is a rare and always-fatal disease that cannot be screened for, nor treated effectively (NINDS 2008). CJD is the most common of the Transmissible Spongiform Encephalopathies (TSEs), or prion diseases, affecting humans, and yet only occurs in one person per million or fewer in the general population (HPA 2008; NINDS 2008; NPC 2008a). CJD is unusual in that it may be both heritable and transmissible, as well as occurring sporadically (DHA 2007; NPC 2008a; NPC 2008b; Will 2003). The cause of prion diseases is not understood clearly, but is thought to be associated with changes to the prion protein. This protein exists normally within the brain, but can also occur as an abnormally-folded form. The abnormal form of the prion protein is thought to be able to cause other normally-occurring prion proteins to similarly change shape. This leads to further abnormal forms of the prion protein, which aggregate (form clumps). It is thought that this protein accumulation and deposition within the brain leads to progressive and irreversible neuronal damage that characterises the prion diseases (Knight 2006; NINDS 2008; NPC 2008a; Pedro-Cuesta 2006; Watts 2006; Ward 2008).

CJD is extremely rare, and a high degree of uncertainty remains about its aetiology, diagnosis and clinical course (CJD IP 2005; CJD SU 2002; NINDS 2008). CJD has an incubation period of variable duration before symptoms emerge, but for some acquired forms this may be as long as several decades after a person's exposure to the infectious prion agent (Brown 2006a; NPC

2008a; Will 2003). Once the symptoms of CJD appear, however, the disease progresses rapidly and around 70 to 90% of people die within one year of symptoms first emerging (CJD Foundation 2008; NINDS 2008; NPC 2008a). CJD causes brain damage, leading to a range of symptoms which may include profound cognitive changes and dementia, behavioural changes, movement abnormalities and other neurological symptoms such as seizures and visual problems (Appleby 2007; Barnett 2005; CJD SGN 2008; CJD SN 2007). These symptoms progress rapidly to become severe and disabling (Du Val 1997; Pollock 2007; Turner 2003b). At present there is no test to diagnose CJD accurately before the onset of clinical symptoms, and a definitive diagnosis of CJD can be confirmed only by neuropathologic examination (Knight 2006; NINDS 2008; vCJD CGAG 2007). Doctors typically make a clinical diagnosis by excluding other neurological or psychiatric diseases with symptoms similar to CJD (Barnett 2005; vCJD CGAG 2007). However, CJD causes a range of symptoms of varying intensity, and not all patients show all symptoms, or show them in the same sequence. This can cause significant problems such as long delays in reaching a diagnosis. In turn, this can delay the provision of appropriate medical, health and social services to consumers (patients and their families and carers) (Barnett 2005; Cegelka 2002; Douglas 1999; vCJD CGAG 2007). There may also be delays for health professionals in accessing the services and support they need to be involved effectively in the patient's care; for example, the need for accurate, up-to-date information on CJD; or access to experts and specialists for professional support (Barnett 2005; vCJD CGAG 2007). Such problems suggest that communication with people affected by CJD, their families, carers and health professionals may often be a complex process occurring over time, rather than a one-off event.

How and why do people develop prion diseases?

Many uncertainties surround prion diseases, and research into diagnosis, prevention, treatment and cure are high priorities, despite the rarity of the diseases (NINDS 2008; Sutton 2006; vCJD CGAG 2007). The prion diseases include a number of different conditions of different aetiology, but can be divided into several major types (NINDS 2008; NPC 2008a; Will 2003), as follows:

1. *Sporadic CJD* accounts for the majority (approximately 80 to 85%) of human TSE cases, although it remains a rare disease in the general population with an incidence of approximately one case per million per year (HPA 2008; NICE 2006; NINDS 2008; NPC 2008a; Pedro-Cuesta 2006). It is not yet clear how or why people spontaneously develop sporadic CJD (Knight 2006).

2. *Genetic (inherited) TSEs* are rare, and include familial (genetic) CJD, Fatal Familial Insomnia (FFI) and Gerstmann Strüssler Scheinker disease (GSS) (NICE 2006; NINDS 2008; NPC 2008a). A number of specific genetic mutations in the prion protein gene are now known to be associated with the inherited prion diseases (Knight 2006; NPC 2008a; Pocchiari 2004), and differ-

ent mutations influence the length of disease and determine which symptoms are most dominant (NINDS 2008). Genetic screening and predictive prenatal testing are now available for familial CJD (CJD Foundation 2008; Pedro-Cuesta 2006).

3. *Acquired TSEs* are caused by exposure to the infectious prion protein. These include kuru as well as iatrogenic (medically-acquired) CJD and variant CJD (vCJD, previously called new-variant CJD, nvCJD) (NPC 2008a; Pedro-Cuesta 2006), the latter two of which are relevant to this review.

a) Iatrogenic CJD: Exposure to prions can occur accidentally through medical care. To date, CJD has been acquired through the following medical procedures:

- the use of contaminated batches of intramuscular human pituitary hormones (hPH), including human growth hormones (hGH) and human pituitary gonadotrophins (hPG). People exposed in this way form the largest group to date of those exposed to the risk of CJD through medical treatments (NPC 2008b). In these cases, some people were treated with batches of hormones that were produced using organs (pituitary glands) of deceased people who themselves had CJD, or were incubating CJD (were not yet showing symptoms) but which was not recognised at the time of donation (Allars 1994; NINDS 2008; NPC 2008b; Will 2003);

- the transplantation of corneas and dura mater grafts (used in neurosurgery). These cases occurred because people inadvertently received grafts from people who themselves had CJD or were incubating CJD at the time of donation (NINDS 2008; NPC 2008b; Will 2003); and

- exposure to contaminated neurosurgical equipment. As the infectious prion agent may not be completely destroyed by conventional sterilisation procedures, transmission can occur, for example, via neurosurgical instruments that have been used previously on a person who has CJD that is not recognised at the time of surgery; or who was incubating CJD at the time of surgery and then later goes on to develop CJD (Hart 2004; NICE 2006; NINDS 2008; NPC 2008b; Pollock 2007).

Substantial uncertainty surrounds the transmissibility of CJD through non-neurosurgical procedures or other routes. At present there is little consensus as to how much of the infectious prion agent is required for person-to-person transmission of CJD to occur. It is also unclear whether there are further routes through which CJD infection might occur (Blajchman 2004; Duncan 2005; Farrugia 2005; Hart 2004; Larke 1998; Wilson 2000). For example, there is speculation whether surgery (other than neurosurgery) might also represent a route of transmission from person to person (Ward 2008). The long latency and low prevalence of CJD in the general population has made it difficult to trace people with CJD and their contact with healthcare systems; or to demonstrate with certainty that CJD is not transmitted through routes such as blood, where it still remains a 'theoretical' risk of transfusion (Du Val 1997).

b) Variant CJD (vCJD, previously called new-variant CJD, nvCJD):

The variant form of CJD is acquired through the diet, by consuming meat contaminated with bovine spongiform encephalopathy (BSE) (NICE 2006; NPC 2008a; NPC 2008b). This variant form differs in several ways from other forms of CJD; for example, people with vCJD have infectious prion proteins distributed more widely throughout their body in tissues such as the spleen and lymphoid tissues (NINDS 2008; vCJD CGAG 2007).

While vCJD is initially acquired through the diet, it can also be spread in a secondary way through medical procedures. It has recently been confirmed, for example, that vCJD can be transmitted from person to person through blood, and four cases of probable transmission via transfusion have been documented to date (TMER 2010). This means that a person might acquire a risk for vCJD by eating BSE-contaminated meat, and that this risk could then be passed on to others through blood donation (Brown 2006a; Hewitt 2006a; Hewitt 2006b; Llewelyn 2004; Sutton 2006). Other medical routes of vCJD transmission, such as surgery, cannot yet be ruled out as possible risks to public health (NICE 2006). It is also not known how many people might be asymptomatic but incubating the disease and so at risk of passing on vCJD through healthcare routes (Brown 2006a; vCJD CGAG 2007).

Any measures to reduce the risk of transmission of CJD or vCJD in healthcare settings must therefore take into account the unusual features of the prion diseases. This includes their extremely long incubation periods; the lack of any tests to determine definitively who is at risk, or incubating the disease; and the current lack of adequate decontamination techniques for the prions in healthcare settings (Will 2003).

Medically-acquired (iatrogenic) CJD and vCJD

Internationally, medically-acquired CJD makes up only a very small proportion (less than 1%) of deaths due to CJD (Ladogana 2005; NINDS 2008; NPC 2008b; Ward 2008). Deaths associated with the transmission of CJD via human pituitary hormones and dura mater grafts are declining (Brown 2006a). However, as there is no way to screen for CJD or vCJD, issues of infection control and transmissibility through healthcare routes (such as surgery, blood or tissue donations, or other possible routes) are of ongoing public health concern (Brown 2006a; NICE 2006; Pocchiari 2004; Ponte 2006; vCJD CGAG 2007; Ward 2008). There is concern that people have, and will continue to be exposed to, CJD and vCJD through a range of healthcare procedures. It is these people who form the focus of this review: those at risk of CJD or vCJD acquired through medical procedures (iatrogenically), together with their families, carers and healthcare professionals.

The clinical course of medically-acquired CJD varies, and is affected by several factors (Pocchiari 2004), including the dose of the infectious prion to which an individual is exposed through treatment. Genetic factors also play a role: the presence of particular prion protein gene polymorphisms (variations) affect an in-

dividual's susceptibility to, or the incubation period of, iatrogenic CJD (NICE 2006; NPC 2008b; Pocchiari 2004; Will 2003) and may even influence the symptoms that people experience (Knight 2006).

The route of CJD transmission, described as central or peripheral, also seems to play a role in the disease's clinical course (Will 2003). People with CJD acquired via intracerebral (central) procedures, such as neurosurgery, tend to have a relatively short incubation period (approximately 18 months to 4 years, but this may be as long as 15 years). In contrast, those exposed to CJD via peripheral treatment (that outside the central nervous system), such as via intramuscular injection of contaminated hGH, typically experience longer latent periods of approximately 4 to 25 years before symptoms develop (Furtner 2008; Llewelyn 2004; NPC 2008a; NPC 2008b; Will 2003). Although there is considerable individual variation, peripherally-acquired CJD is, in general, characterised by a longer incubation period, prolonged illness and early symptoms of coordination and balance problems. In contrast, centrally-acquired CJD tends to have a shorter latency and rapid progression, with memory problems and other cognitive symptoms predominating (NPC 2008a; NPC 2008b).

The symptoms of vCJD are different again from other forms. For example, people with vCJD tend to experience more prolonged total disease duration than those with sporadic CJD, and initial symptoms are often predominantly psychiatric, such as depression and anxiety (Appleby 2007; CJD Foundation 2008; NPC 2008b). People with vCJD also tend to be younger, with most under 30 years of age (Knight 2006; NPC 2008b). It is not yet clear how long the vCJD incubation period is when people are exposed via diet, although some estimates suggest it may be as long as 10 to 30 years after exposure (CJD IP 2005). Among recipients who appear to have developed vCJD as a result of blood transfusion, the incubation period seems shorter (approximately 6.5 to 8 years), although once again it may vary between individuals (Hewitt 2006a; Hewitt 2006b; Llewelyn 2004). That the incubation period of vCJD appears to be a matter of several years - whether acquired from diet or secondarily via blood products - is significant: protection of the public health must take account of people potentially incubating the disease as a result of either exposure and passing this on to others via blood products in healthcare settings.

Internationally, there is wide variation in the number and geographic distribution of people at risk of CJD and vCJD. For example, compared with other countries, France has a high proportion of iatrogenic CJD cases, which are decreasing over time; whereas the UK shows the highest proportion of vCJD cases which is also declining over time (Pedro-Cuesta 2006). Different emerging patterns of CJD and vCJD will dictate who needs to be informed of their exposure to risk, and notification policies will need to be aligned with the latest evidence on CJD and vCJD. The recent confirmation of vCJD transmission through blood, for example, has led to large numbers of individuals being notified of their elevated risk for vCJD above background levels (Dolan 2006;

Hewitt 2004; Hewitt 2006a; Hewitt 2006b; Llewelyn 2004). This includes people with haemophilia, a group with significant experience with iatrogenic risks associated with blood transfusion. Other people notified will include those with no personal prior experience of iatrogenic risks or infections (Hewitt 2006a; Hewitt 2006b). Notification and support processes for iatrogenic CJD and vCJD risk communication must be able to accommodate the experiences and needs of people at both ends of this spectrum, yet the best ways of doing this are not known.

Ethics and notification: public health and individual issues

Regardless of the route of medical exposure, CJD and vCJD are serious diseases and there is considerable debate about the ethical implications of notifying people about exposure to the risk of a fatal disease that cannot be screened for and about which so much medical uncertainty exists (Barnett 2005; Blajchman 2004; Hewitt 2004; Hewitt 2006a; Shalowitz 2009; Steinberg 2001). Guidance in these situations might be informed by the evidence on communicating bad news to patients and their families or carers, and on consumers' expressed needs and preferences in similar situations. Recent studies on communicating bad news to oncology patients and their family members, for example, suggest that communication must take account of the personal impact of the information, and that it must be a planned encounter involving two-way communication between doctors and their patients, as well as family members or carers (Eggle 2006; Parker 2001). Such encounters must provide information that is comprehensive and understandable, provide support for decision making, and be responsive to the needs of patients and their family members or carers (Eggle 2006). There is less evidence on the best ways to communicate prognostic information for cancer patients, or on the needs and preferences of oncology patients and their families in these situations over time (Hagerty 2005; Parker 2001).

Possible guidance in situations where people have been exposed iatrogenically to the risk of CJD or vCJD could also be informed by similar experiences with other medically-acquired diseases. Worldwide, several incidents have occurred in which people were infected with serious diseases such as Human Immunodeficiency Virus (HIV) or Hepatitis C (HCV) through medical procedures (Bowker 2004; HPA 2008; Myers 2003; Wilson 2007; Worthington 2002). In the last two decades, large numbers of people have been potentially exposed to these diseases through blood transfusion. This led to large recalls of potentially contaminated blood, and notification of the individuals involved, so that people could choose whether to be screened, and then to access medical and other support services if, or when, needed (Cagle 2007; Heddle 1997; King 1998; Langley 2001; Wilson 2007).

The high degree of uncertainty associated with medically-acquired risk of CJD and vCJD may, in many ways, distinguish these situations from others involving communication of bad news to patients and families or carers. No effective screening or treatment

options yet exist for people at risk of CJD or vCJD; and there is uncertainty about the meaning of the acquired risk for possible future disease. Such uncertainties may make the case for non-disclosure of possible CJD or vCJD exposure stronger than for a disease that has been diagnosed, such as in the case of communicating a diagnosis of cancer; or for medically-acquired diseases such as HIV or HCV, for which screening and increasingly effective treatment options exist (Howe 2001; Steinberg 2001).

However, notifying people around iatrogenic incidents is in line with recent moves to support services to disclose openly to patients and their families the facts around adverse incidents that occur when receiving medical care (Iedema 2008; Levinson 2009; Mazor 2004). There is also a need to protect the public health and to prevent people at risk of CJD or vCJD from donating blood or undergoing medical procedures without appropriate infection control measures in place where required (DHA 2007). The balance between precautionary public health priorities, the individual's rights, and the potential harms of notification of CJD or vCJD risk exposure therefore remains contentious (Bird 2004; DHS 2006; Hewitt 2006b; NINDS 2008; Ponte 2006; Wilson 2004b).

What are the options for notifying and supporting people following exposure to the risk of CJD or vCJD through medical treatment?

The consequences of being notified of iatrogenic exposure to the risk of a serious disease such as CJD or vCJD may be profound for consumers. It may also have implications for the healthcare professionals involved and for the health systems in which such incidents occur (DHS 2006; Dolan 2006; Steinberg 2001; Turner 2003; vCJD CGAG 2007). Notification of exposure to the risk of a life-threatening disease, whether the person goes on to develop the disease or not, may cause significant distress, anxiety and lasting psychological harm (Duncan 2005; Larke 1998; vCJD CGAG 2007). People notified of exposure to CJD or vCJD risk may also face additional difficulties, such as delays or discrimination when accessing appropriate medical care, including restricted access to invasive procedures such as surgery, or other care such as dentistry (CJD SGN 2008; DHS 2006; Howe 2001; Steinberg 2001; vCJD CGAG 2007).

Ensuring the best possible clinical care and support for people exposed to the risk of CJD or vCJD through medical treatment is therefore essential (vCJD CGAG 2007). 'Lookback' programmes identify people who have been exposed to a potential source of infection (e.g. recipients of blood potentially contaminated with HIV), notify them of their exposure, and offer the opportunity for testing if this is available. Evaluations of lookback programmes used to notify people of potential exposure to HIV, HCV or CJD have suggested that people are generally positive about receiving such notification. A high proportion of people notified via lookback activities, for example, undergo testing for HIV or HCV

following notification (Heddle 1997; King 1997); and notification may enable people to access counselling, therapeutic interventions and additional support services (Heddle 1997; Larke 1998; Younossi 1998). Research also suggests that recipients, in general, would rather be informed than not (Blajchman 2004; Hewitt 2004; Larke 1998), even though the notification may itself be distressing (King 1995; King 1998). Importantly, however, some people explicitly wish not to be notified of exposure to a disease risk in such situations (Blajchman 2004; Caulfield 1997; Hewitt 2004), and general approaches to notification remain controversial, at least in part because of these people who may be inadvertently notified against their wishes (Bowker 2004; Caulfield 1997; King 1997; Reesink 2003; Ricketts 1997).

The impact of notification

Relatively little is known about the effects of notification of exposure to CJD or vCJD risk on psychological, emotional and social outcomes for consumers. Research indicates that several factors, such as the individual's personality, may influence the impact of notification (Langley 2001; Sibbald 1998). This suggests that the effects of notification may differ markedly between individuals (Hewitt 2004; Larke 1998); and even those who are most certain they wish to be notified of exposure to a disease risk may suffer harm as a result, and require ongoing support (Larke 1998). However, the range of impacts on people notified of exposure to CJD or vCJD risk is not well documented. There is little information to identify: those who might be adversely affected, or most at risk of harms arising from notification - other than those inadvertently notified by the media or some other accidental route. Issues include: what types of harms might arise; and how to prevent or minimise potential harms associated with notification. Given that a range of responses to notification of the risk of iatrogenic CJD/vCJD exposure may exist, there may not be a single best way to notify all people in these situations (Farrugia 2005; Sibbald 1998).

Notification of at-risk status

Iatrogenic exposure to CJD or vCJD risk raises questions about how and by whom notification should be carried out (Larke 1998). There are many different ways in which people might be notified of their exposure to risk. This includes:

- conveying the information using a range of different formats (e.g. consultation with healthcare provider, letter, telephone call);
- providing the information through different people or organisations (e.g. treating physician, specialist, health service);
- presenting the risk information on CJD/ vCJD in different ways (e.g. as an absolute or relative risk, a natural frequency, or as a lifetime probability); and

- using a range of delivery features (such as varying the timing of the notification, or different components of the notification strategy, providing training to those involved in communicating with people at risk, and establishing notification processes ahead of time) (Callum 1999; DHS 2006; Farrugia 2005; Freedman 1997; Hewitt 2006a; Hewitt 2006b; King 1998; vCJD CGAG 2007).

The notification strategy may also incorporate more than one component and/or format. There are potentially, therefore, many different approaches to notification. The evidence on what people need or would prefer in these situations, however, has yet to be systematically assembled and evaluated.

Support and follow-up post-notification

Once people have been notified of their at-risk status for CJD or vCJD, there are different ways in which they might be followed-up and supported over time. These include strategies such as accessing specialist telephone help lines, follow-up appointments and referral to specialists, counselling, and provision of detailed information on CJD and vCJD (DHS 2006; Freedman 1997; Hewitt 2006a; Hewitt 2006b; King 1998; Reesink 2003; vCJD CGAG 2007). Again, approaches might incorporate more than one component. The necessity of living with the uncertainties of risk and prognosis once notified, possibly over many years, may significantly affect people's ongoing psychological, emotional and physical wellbeing (Meek 1998; Steinberg 2001). An international forum of experts recently recommended that extensive follow-up and referral to specialists be provided routinely to people notified of exposure to CJD (Reesink 2003). However, the needs of people at risk immediately following the notification, and over time, are not well documented. As a result, the best ways of meeting their needs are unknown.

The long latency of CJD and vCJD suggests that the responses and needs of people at risk may change over the person's lifetime. It is also likely that, in time, some of the uncertainties of CJD and vCJD risk, and the meaning of this risk for future disease, will become clearer. A recent report highlighted, for example, that as the uncertainties around CJD/ vCJD start to be resolved, or if a sensitive and specific test becomes available, the people involved will need to be informed and supported appropriately at each stage (vCJD CGAG 2007). As knowledge about CJD and vCJD grows or changes over time, there will therefore need to be strategies in place to appropriately convey this information to patients and their families, as well as to healthcare professionals.

Relationship to other relevant reviews

The scope of this review covers a wide range of interventions, such as information provision, education, notification and disclosure, counselling and supportive care. These interventions are likely to

overlap with the scope of existing Cochrane and non-Cochrane systematic reviews. However, the focus of this review on medically-acquired CJD/ vCJD risk is narrow and we have not identified any existing reviews which focus specifically on these situations or populations.

Several Cochrane reviews and protocols are relevant to the scope of this review. One particularly relevant review is 'Methods of communicating a primary diagnosis of breast cancer to patients' (Lockhart 2007), which found no studies for inclusion. Other Cochrane and non-Cochrane reviews, such as those dealing with communication in serious diseases, and breaking bad news, have been used to inform our choice of outcomes (such as Davies 2003; Leliopoulou 2001; NICE 2004; Walsh 1998). Literature on rare diseases and communication about risk, diagnosis and treatment options is also relevant to the current review. However, we are not aware of any systematic reviews, Cochrane or non-Cochrane, which deal with these issues in a way that has clear implications for the current review and we therefore rely on primary research undertaken in the area to inform our discussions.

Why this review is important

Little is known about the best ways to notify people of potential exposure to the risk of CJD or vCJD through medical treatment, what people need or want from the notification process, or how best to support them during and after the notification.

In this review we focus solely on people at risk of CJD or vCJD following potential iatrogenic exposure. The rationale for this is the high degree of uncertainty around the diagnosis, transmission, treatment and clinical course of CJD and vCJD, which sets these people apart in important ways from those exposed to other serious, medically-acquired diseases. As the body of knowledge on CJD and vCJD grows and informs practice and policy, this accumulated knowledge will need to be communicated accurately and sensitively to consumers. In the interim there are individuals, families and groups facing the uncertainty of CJD and vCJD risk, and the best ways of notifying and supporting these people in the face of what is known about the disease has yet to be systematically assembled and evaluated. Effective ways to inform and support people at risk over time are needed in relation to infection control measures and to promote people's capacity to manage their health care, as well as to promote healthcare responses that are appropriate to the person's individual risk level.

Poor practices in the past mean that these issues remain important. For instance, although the widespread therapeutic use of cadaver-derived hPH ceased over two decades ago, the long latency of CJD means that cases may continue to emerge amongst this population (Brown 2006a; Furtner 2008; Pedro-Cuesta 2006). In addition, those cases arising spontaneously, those associated with heritable disease, and cases of vCJD will continue to emerge and the people affected will invariably have contact with healthcare systems (Knight 2006; Will 2003). Even if effective screening and treat-

ment options become available, it may not be possible to identify all people with emerging CJD or vCJD as they engage with healthcare systems. This means that other people will continue to be exposed to these risks iatrogenically. Effective ways to notify people of potential risk in these circumstances, to support them and to best organise their care should they develop CJD or vCJD will therefore remain a high priority into the foreseeable future.

We planned this review with awareness that it may be an 'empty Cochrane systematic review', that is, a review without included studies to form the basis of analysis, synthesis and conclusions (Green 2007; Lang 2007) - and this proved to be the case. We believe that, nevertheless, the review has:

- identified interest in the processes of effective communication with people at risk of CJD or vCJD, acquired iatrogenically;
- highlighted research gaps;
- contributed to the systematic identification of the range of interventions and outcomes that could form the basis of rigorous studies in this area; and
- provided a status report of research that will be regularly updated (Lang 2007).

Furthermore, this review was commissioned by the Public Health Division of the Health Department of Victoria, Australia, which concurrently commissioned us to conduct a separate review (thematic synthesis) on consumers' views, experiences and the acceptability of notification and support strategies in relation to being at risk of CJD or vCJD as a result of medical procedures. We have provided further information about the methods and findings of this thematic synthesis (see Appendix 12 to Appendix 15) and have used it to inform the Discussion of this Cochrane review.

As more is known about the aetiology and treatment of CJD and vCJD, we will revise the scope of this review to fit the changing evidence base. As for policy and practice, it will be important to keep this review regularly updated to reflect new evidence (vCJD CGAG 2007). Although the importance of strategies for notifying and supporting people in relation to iatrogenic exposure to CJD and vCJD risk is well recognised, the best ways of doing so are not known. These strategies are the focus of this review.

OBJECTIVES

To evaluate the effects of interventions to notify and support consumers (patients and their carers or families) in situations where exposure to the risk of CJD or vCJD has occurred as a result of medical treatment (iatrogenically), on consumer, healthcare provider and healthcare system outcomes.

METHODS

Criteria for considering studies for this review

Types of studies

Current advice is that Cochrane reviews summarise the effects of interventions which have been the subject of rigorous, controlled evaluative studies (Higgins 2008). This is because these studies are less prone to bias than other types of studies, with the result that Cochrane reviews are most commonly reviews of randomised controlled trials (RCTs) alone. However, when reviewing the evidence on the effects of complex interventions, and particularly in situations in which there are likely to be few RCTs or none at all, it may be appropriate to include a limited range of other study designs in addition to RCTs in order to answer questions of effectiveness (Ryan 2009).

In this review we decided to include a wider range of study designs, and so considered (individual and cluster) RCTs, quasi-RCTs, controlled before-and-after (CBA) studies and interrupted time series (ITS) analyses eligible for inclusion. Although non-randomised studies may be more prone to bias, RCTs may not always be possible, practical or ethical to conduct.

Given the importance of this topic and the authors' view that there was a need to summarise and synthesise the available research on this topic, we also conducted a separate thematic synthesis alongside this Cochrane review. For this thematic synthesis, we systematically identified and screened those studies that met the same population, intervention and outcome criteria as the Cochrane review, but that were identified from the broader literature providing evidence on policy implementation and consumer experiences. These studies formed the basis of the thematic synthesis (presented as [Appendices](#) to this Cochrane review) and informed the [Discussion](#) in the absence of research meeting the study design criteria for the Cochrane review.

Types of participants

We included people of any age, gender or ethnicity at risk of CJD or vCJD acquired via medical treatment (whether or not they were also genetically predisposed to CJD or other prion diseases); their families and/or carers, and/or the healthcare professional(s) involved in their care.

The risk of CJD or vCJD could be associated with any medical or surgical treatment, and included those people at secondary risk of vCJD through blood transfusion.

We excluded:

- People for whom the risk of CJD or vCJD was not acquired through medical treatment, including:
 - People exposed to the risk of vCJD through diet (BSE-contaminated meat consumption).
 - People and families for whom the risk for CJD or other prion disease was solely genetic in origin. The rationale for this exclusion was that families in which familial CJD risk exists may

choose to undergo genetic screening to identify this risk. This process involves a range of different communication issues to those where an individual's risk of CJD is acquired iatrogenically.

- People with probable or confirmed CJD or vCJD, and their families and carers. The rationale for this exclusion was that people dealing with actual disease, rather than the possibility of exposure to the risk of disease, have a distinct range of needs and experiences, and face a different range of issues and decisions, to those of people at risk.

Types of interventions

We included any intervention aiming to notify or support people exposed to the risk of CJD or vCJD through medical treatment. In this review, we use the term 'notify or support' broadly to mean all interventions to communicate with, notify, educate, inform, seek the participation of and support and follow-up people in these situations.

For people at risk and their families/ carers, examples include interventions to:

- Notify or inform people of their exposure to the risk of CJD or vCJD - e.g. letter, telephone call, consultation with GP or other healthcare professional.
- Inform or educate people about the disease risk to which they may have been exposed - e.g. information about the disease or risk of disease and the current state of knowledge and uncertainty about prion diseases; the risk of transmission and/or precautionary measures that must be taken to protect the public health (e.g. when receiving medical or other health treatment; when considering blood, tissue or organ donation).
- Support people following notification - e.g. supportive counselling or family-based interventions.
- Improve the continuity of care or provide follow-up for people notified of the risk of CJD or vCJD, or the healthcare professionals involved in the notification process and/or the provision of care, including interventions in time periods immediately following notification, as well as at later time points.

Interventions could be delivered in any setting (e.g. hospital, home, community); via any format or medium (e.g. letter, telephone, face-to-face consultation); and by any provider (e.g. clinician, community health worker, peers or family).

The intervention could have one or several components. It could be delivered solely to the patient (person at risk), and/or to their families or carers; and could be delivered to individuals or to groups or families.

Interventions could also be delivered to healthcare professionals involved in notifying and/or supporting consumers, but only where the intervention aimed specifically to equip the professional with the skills and resources to communicate more effectively. In such cases the study must have reported at least one consumer-oriented outcome to have been eligible for inclusion.

We included the following comparisons:

- Interventions to notify or support people versus no intervention.
- Interventions to notify or support people versus standard or usual care.
- One form of intervention to notify or support people versus another - including simple versus complex interventions.

We excluded:

- Interventions aiming to notify people or support them in relation to genetic risk of CJD or other prion disease.
- Interventions aiming to communicate with or support patients or their families and carers in relation to a probable or confirmed diagnosis of CJD/ vCJD.

Types of outcome measures

A range of outcomes relevant to consumers, to healthcare providers and to healthcare systems may be influenced by or the result of interventions to notify and support consumers following iatrogenic exposure to the risk of CJD or vCJD. There were no exclusions based on the outcomes reported by studies that were otherwise eligible for inclusion in this review, other than where the intervention targeted healthcare providers (where at least one consumer-oriented outcome must have been reported).

We sought data on the outcomes listed below (for a more detailed list of outcomes sought see [Appendix 1](#)). Given the long latency of CJD and related diseases, we sought information on outcomes in both the short term (e.g. immediately following intervention delivery) and at all longer-term time points for which data were available.

Primary outcomes

Consumer-oriented outcomes, including:

- Knowledge and understanding, e.g. understanding of how the risk was acquired, perceived risk.
- Health status and wellbeing, e.g. physical or psychological health outcomes.
- Communication, e.g. satisfaction with communication.
- Evaluation of care, e.g. satisfaction with care or interventions received.
- Support, e.g. whether people used practical or psychosocial support (such as information, counselling, services), and the effects of support (such as perceived or actual support, social function, isolation).

Healthcare provider-oriented outcomes, including:

- Knowledge and understanding, e.g. levels of knowledge about CJD/ vCJD and prognosis, infection control.
- Consultation processes, e.g. level of patient-centred care.
- Support, e.g. support and/or training received in notifying patients.

We also sought data on potential harms and adverse events associated with the interventions, for the patient, family members and/ or carers, and for the healthcare providers involved in delivering interventions or care and management. This included consideration of emotional and psychological harms, such as anxiety, psychological distress and fear. We also specifically considered harms relating to changes in access to medical and health treatments; and perceived and actual discrimination. We also identified any complaints regarding services or personnel.

Secondary outcomes

Consumer-oriented outcomes, including:

- Patient involvement in care, e.g. involvement in decision making.
- Skills acquisition, e.g. self-care skills.
- Health behavior, e.g. attitudes towards a disease or health care.
- Treatment outcomes, e.g. clinical outcomes.

Healthcare provider-oriented outcomes, including:

- Health service use, e.g. provider behaviour such as appropriate referral.
- Health status and wellbeing, e.g. psychological health outcomes for the provider.

Health service delivery-oriented outcomes, including:

- Service delivery level, e.g. use of care plans or teams, communication between care teams.
- Societal or governmental, e.g. healthcare policy/ legislation/ procedures and revisions of these.

Search methods for identification of studies

Electronic searches

We searched the following electronic databases:

- Cochrane Consumers and Communication Review Group Specialised Register;
- Cochrane Central Register of Controlled Trials (CENTRAL) (*The Cochrane Library*, issue 1 2009);
- MEDLINE (OVID SP), including MEDLINE In-Process and Other Non-indexed Citations;
- EMBASE (OVID SP);
- PsycINFO (OVID SP);
- CINAHL (EBSCO Host);
- Current Contents (OVID SP) (Social & Behavioural Sciences, Clinical Medicine);
- Dissertations and Theses (Proquest); and
- Sociological Abstracts (CSA).

Searches of the Specialised Register, CENTRAL, OVID (MEDLINE, EMBASE, PsycINFO, CINAHL, Current Contents) and

Dissertation Abstracts were run on February 10 to 12, 2009. Searches in MEDLINE In-Process and Other Non-indexed Citations and Sociological Abstract and were run on November 12 and 24 2009 respectively.

We searched all databases from their start date to the present. The search strategy for MEDLINE was used as a basis, appropriately tailored, for searching of other databases. Search strategies can be found in full in Appendices ([Appendix 3](#) to [Appendix 10](#)).

There were no language or date restrictions. Nor were there search filters or restrictions applied based on study design.

Searching other resources

We searched grey literature, including relevant government, health agency and consumer websites. Full details of websites searched are presented in [Appendix 11](#).

We contacted experts in the field and authors of key studies for advice on other relevant studies, both published and unpublished. We did not handsearch journals but searched reference lists of relevant studies and reviews; and cross-checked relevant papers and authors using ISI Web of Science and PubMed (related articles function) to locate additional relevant materials.

Data collection and analysis

Selection of studies

Two authors working independently screened the titles and abstracts of studies identified from searches for inclusion. We retrieved in full text any papers identified as potentially relevant by at least one author.

Two review authors then independently screened full text articles for inclusion or exclusion, with discrepancies resolved by discussion to reach consensus, and by consulting a third author where necessary. All papers excluded from the review at this stage were listed as excluded studies, with reasons provided in the [Characteristics of excluded studies](#) table.

We also provided citation details and any available information about ongoing studies (see [Characteristics of ongoing studies](#)) or those awaiting assessment (see [Characteristics of studies awaiting classification](#)).

As no studies were identified for inclusion in this review, details of studies and the reasons for their exclusion were entered into RevMan by a single review author. Data entry into RevMan of a selection of approximately 20% of these excluded studies was checked for accuracy by a second review author.

Data extraction and management

No study met the inclusion criteria. Should any eligible studies become available for inclusion in future updates of this review we will use the methods described in [Additional Table 1](#) to extract data (see [Additional Table 2](#)), assess the risk of bias of included studies and analyse data.

Table 1. Methods for future review updates

<p>Data extraction and management</p>	<p>Two review authors will independently extract data from all included studies, with any discrepancies to be resolved by discussion to reach consensus, or through consultation with a third author wherever necessary. A data extraction form will be developed and piloted, using the Cochrane Consumers and Communication Review Group Data Extraction Template (available at: http://www.latrobe.edu.au/chcp/cochrane/resources.html) as a basis. Further details of the data to be extracted from included studies is given in Additional Table 2. All extracted data will be entered into RevMan by one review author, and checked for accuracy against the data extraction sheets by a second review author working independently.</p>
<p>Assessment of risk of bias in included studies</p>	<p>Two review authors will independently assess the risk of bias of each included study, with disagreements resolved by discussion and consensus, and by consulting a third review author if necessary. Studies of different designs will be dealt with separately throughout this review in both the risk of bias assessment and analysis. For RCTs (and quasi-RCTs), we will assess and report on the following elements that contribute to bias, according to the guidelines outlined in Higgins 2008:</p>

Table 1. Methods for future review updates (Continued)

	<ul style="list-style-type: none"> ● sequence generation; ● allocation concealment; ● blinding of participants, personnel and outcome assessors; ● incomplete outcome data; and ● selective outcome reporting. <p>We will describe the study and assign a judgement relating to the risk of bias for each item, following the guidance in Higgins 2008. We will rate each item as 'yes' (indicating a low risk of bias), 'no' (a high risk of bias), and 'unclear' (risk of bias is unclear). For each study we will summarise the risk of bias for each outcome.</p> <p>We will also assess a range of other possible sources of bias and indicators of study quality, in accordance with the guidelines of the Cochrane Consumers and Communication Review Group (Ryan 2007), including:</p> <ul style="list-style-type: none"> ● baseline comparability of groups; ● validation of outcome assessment tools; ● reliability of outcome measures; ● other possible sources of bias (e.g. contamination or co-intervention). <p>If studies other than RCTs and quasi-RCTs (CBA and ITS studies) are included in the review, we will also assess the risk of bias and quality of these studies. We will adapt the risk of bias criteria for these study designs, based on guidance in the Cochrane Handbook for Systematic Reviews of Interventions (Higgins 2008) and that of the Cochrane Consumers and Communication Review Group (Ryan 2007), which reflects the guidance from EPOC; and will describe the study and assign a judgement relating to the risk of bias for each item.</p> <p>We will incorporate the results of the risk of bias and quality assessment into the review through systematic narrative description and commentary about each of the assessed items, for each type of included study. This will lead to an overall assessment of the risk of bias across the included studies and a judgement about the possible effects of bias on the effect sizes of the included studies.</p> <p>We will attempt to contact authors of included studies for additional information about the included studies, or for clarification of the study methods, particularly to obtain details relevant to risk of bias assessments, as required.</p>
<p>Measurement of intervention effects</p>	<p>Where quantitative data are available, we will extract means and variances, numbers of participants on which results are based, and results of any tests of statistical significance. Where possible we will use RevMan to report quantitative data.</p> <p>For individual RCTs, quasi-RCTs and CBA studies we will calculate relative risks (RR) and 95% confidence intervals (CI) for dichotomous data. For continuous data where outcome scales are similar, we will calculate the mean difference (MD) and 95% CI. Where outcome scales are variable we will calculate the standardised mean difference (SMD) and 95% CI. For ITS studies, we will calculate the mean difference in outcomes before and after intervention delivery.</p>

Table 1. Methods for future review updates (Continued)

Unit of analysis issues	If cluster-randomised controlled trials are included, we will check for unit of analysis errors. If required and sufficient data are available we will recalculate the results using the appropriate unit of analysis (Higgins 2008).
Dealing with missing data	Where data are missing, we will attempt to contact authors of included studies to obtain complete data. Where possible, we will conduct analysis on an intention-to-treat (ITT) basis; otherwise data will be analysed as reported. We will report on the levels of loss to follow-up and assess this as a source of potential bias.
Assessment of heterogeneity	<p>We anticipate that a substantial degree of heterogeneity will exist due to differences in the interventions and outcome measures, study designs and the methodological quality of included studies. Although the scope of this review is narrow in terms of the disease risks considered eligible, we also expect that there may be variation in the populations included, such as whether people at risk have been exposed to a confirmed or theoretical risk of CJD or vCJD, or the age of the people involved. There will also be variation between people based on risk level. For example, there are people at a higher risk level as a result of exposure through human pituitary growth hormone treatment, or haemophiliacs exposed through blood transfusion, when compared with neurosurgical routes of exposure which are considered to confer a lower risk.</p> <p>We will therefore consider systematically any differences in populations, interventions and outcomes in the synthesis of data to determine whether statistical pooling of results is appropriate and likely to yield meaningful results.</p> <p>Where studies are considered similar enough clinically to consider pooling data, we will assess the degree of heterogeneity by visual inspection of forest plots and by examining the I^2 statistic (Higgins 2008).</p>
Assessment of reporting biases	We will examine funnel plots for asymmetry to check for possible publication bias (Higgins 2008).
Data synthesis	<p>We will conduct a narrative synthesis of results. We will present the major outcomes and results, organised by intervention categories according to the major types and/or aims of the identified interventions, such as: interventions to notify people of risk; to inform or educate; to support people following notification or diagnosis; to improve continuity of care or follow-up; and to improve self-care or self-management.</p> <p>Depending on the assembled research, we may explore the possibility of organising the data by population (e.g. people 'at risk' of CJD or vCJD; people 'at theoretical risk' of CJD; healthcare providers involved in notifying and/or supporting people at risk); or by setting (e.g. home, hospital). The decision on which way to categorise and organise the data will also be informed by the thematic synthesis.</p> <p>Within the data categories we will group results by study type. Within each data category, the main comparisons will be:</p>

Table 1. Methods for future review updates (Continued)

	<ul style="list-style-type: none"> ● Intervention versus no intervention. ● Intervention versus standard or usual care. ● One form of intervention versus another. <p>Where studies compare more than one intervention, we will compare each separately to no intervention/ control; and with one another.</p> <p>In the case of limited data being included in this review, we will also provide a description of which notification and support strategies have been tested, what their key components were, their content, format and other relevant features. This description will also be informed by the thematic synthesis. We will explore possible reasons for variability in findings in the systematic narrative analysis. Exploration of potential effect modifiers may include investigating characteristics of the interventions; variations in the populations assessed (e.g. CJD risk, vCJD risk, theoretical CJD risk, the age of the individuals involved); the different risk levels associated with the route of exposure (e.g. neurosurgical routes, blood transfusion, hormone treatment); the timing of notification relative to the medical treatment through which exposure to risk occurred; or the influence of different settings upon the effects of the interventions examined. Exploration of potential effect modifiers in the narrative analysis will be performed with the aim of informing the development of best practice recommendations based on the available evidence.</p> <p>Where studies are sufficiently similar in terms of populations, inclusion criteria, interventions and/or outcomes, we will consider pooling the data statistically. We will use a fixed-effects model unless substantial heterogeneity is detected. If substantial heterogeneity is detected, either through visual inspection or by an elevated I^2 statistic, then a random-effects model will be used as a more conservative method of analysis. We will conduct separate meta-analysis, where appropriate, to pool the results of studies within major intervention types (e.g. interventions to notify, educate, support) and within each included study type (RCT, quasi RCT, CBA, ITS). We will analyse and present the findings for each major intervention type organised by the outcomes: consumer-, provider- and system-oriented outcomes respectively. Adverse effects and harms will be analysed and presented separately.</p>
<p>Subgroup analysis and investigation of heterogeneity</p>	<p>We do not anticipate identifying enough studies to justify statistical subgroup analyses to explore underlying causes of variability in the findings. Although we have not planned subgroup analysis, this review and the accompanying thematic synthesis may help to identify potential subgroup analyses for investigation in future versions of the review; for example, considering different sub-populations of people at risk (e.g. CJD risk, vCJD risk, theoretical CJD risk; the age of those involved); or considering different risk levels associated with routes of exposure (e.g. neurosurgical routes, blood transfusion, hormone treatment).</p>
<p>Sensitivity analysis</p>	<p>If adequate data are available, we will conduct sensitivity analyses to investigate the effects of methodological quality (risk of bias). We will investigate study quality as a possible source of heterogeneity by comparing the results of studies of lower methodological quality with those of higher</p>

Table 1. Methods for future review updates (Continued)

	relative methodological quality.
Consumer involvement	For each included study, we will collect information about the involvement and role of consumers in the development and evaluation of the interventions.

Table 2. Planned data extraction

Planned data for extraction
<p>STUDY DETAILS - including:</p> <ul style="list-style-type: none"> • Authors and year of publication. • Details of the study - including aims, recruitment, selection criteria, ethics approval, funding sources and consumer involvement. • Country and setting. • Time span of the trial. • Details of analysis performed. • Consumer involvement and participation - including their role in developing and evaluating the intervention.
<p>PARTICIPANT DETAILS - including:</p> <ul style="list-style-type: none"> • Pre-trial calculation of sample size. • Participant characteristics - such as age, gender, ethnicity, diagnosis and details of comorbidities. • Participant numbers - recruited, randomised and analysed.
<p>INTERVENTION DETAILS - including:</p> <ul style="list-style-type: none"> • Intervention aims. • Development of the intervention. • Characteristics of the intervention - such as the medium/format, content, intensity and other delivery features (e.g. individual versus group, setting where delivered, frequency and timing, provider and training/support of the provider), and the different components of the intervention. <ul style="list-style-type: none"> • Characteristics of control (usual care) or alternative interventions will also be extracted, as well as co-interventions where applicable. • Quality and integrity of the intervention.
<p>OUTCOME DETAILS - including:</p> <ul style="list-style-type: none"> • Primary outcomes and outcome measures. • Secondary outcomes and outcome measures. • Adverse events. • Method and timing of outcome measurement - for each reported outcome. • Validity of outcome measures - for each reported outcome. • Reliability of outcome measures - for each reported outcome.
<p>RISK OF BIAS DETAILS - including rating (yes, no, unclear) and information on each of the following:</p> <ul style="list-style-type: none"> • Sequence generation. • Allocation concealment. • Blinding of participants, personnel and outcome assessors. • Incomplete outcome data. • Selective outcome reporting.

Table 2. Planned data extraction (Continued)

Other study quality indicators: <ul style="list-style-type: none">• Baseline comparability of groups.• Validity and reliability of outcome assessment tools.• Other possible sources of bias (such as contamination or co-intervention).
OTHER INFORMATION - including <ul style="list-style-type: none">• Details of author contact and results

Relation to the thematic synthesis

As outlined above, we concurrently conducted a broad literature review (thematic synthesis) unrestricted by study design (Ryan 2008). The central question of this second review is: what are people's needs, experiences and preferences for notification and support post-notification in situations where exposure to the risk of CJD (or vCJD) has occurred as a result of medical treatment? This thematic synthesis included studies of diverse methods that identify consumers' and carers' views and experiences of being notified of their at-risk status for CJD or vCJD, and their needs with respect to notification and support. Drawing from the research and other literature reviewed, we identified key themes and used them to organise and analyse the data. We also used these themes to develop a model for a comprehensive, multi-component framework for improving notification, communication with, and support for people in relation to CJD or vCJD risk acquired medically. The model has informed the identification of the range of possible interventions and outcomes outlined in this Cochrane review. We present further details on the methods used for this thematic synthesis in [Appendix 12](#).

Consumer involvement

This review and thematic synthesis have been reviewed by consumer referees, as per standard editorial processes of the Cochrane Consumers and Communication Review Group, to ensure the applicability and acceptability of the review to consumers and to ensure that consumers' views are represented appropriately.

RESULTS

Description of studies

See: [Characteristics of excluded studies](#); [Characteristics of studies awaiting classification](#).

Cochrane review

We screened over 300 studies in full text to assess eligibility for inclusion in this review. Several studies or reports of incidents, lookback exercises or tracking of CJD/ vCJD cases were published in more than one paper.

No studies met the inclusion criteria for this Cochrane review. We excluded most studies on the basis of the population considered (i.e. they did not deal with people at medically-acquired risk for CJD or vCJD); or because they failed to either describe or assess any aspect of the impact of notification, communication or support for those at risk. We also excluded several studies because they reflected the broader literature on policy implementation and consumer experiences, rather than providing evidence about the effectiveness of interventions by meeting study design criteria for inclusion in the Cochrane review.

Thematic synthesis

In total, 49 pieces of research and other literature (reports, consensus statements, complaints data and other sources) were identified as relevant and provided evidence from the broader literature about policy implementation and consumer experiences. We included these studies that otherwise described or assessed the impact of notification, communication and support for people at medically-acquired risk of CJD or vCJD in the thematic synthesis. We present the results of the thematic synthesis in [Appendix 13](#) and [Appendix 14](#), and outline the patient-centred communication framework developed from the thematic synthesis in [Appendix 15](#). We present a summary and overview of the results and their implications in the [Discussion](#).

Risk of bias in included studies

No studies were eligible for inclusion in the Cochrane systematic review; the risk of bias was not assessed.

Effects of interventions

No studies were eligible for inclusion in the Cochrane systematic review.

DISCUSSION

Summary of main results

Summary of the Cochrane review

No studies were included in this Cochrane review. There is currently no evidence from rigorously- conducted studies to inform decisions about the effects of interventions to notify people, or to support them post-notification, in the context of medically-acquired risk for CJD or vCJD.

Summary of the thematic synthesis

In the absence of rigorous studies on the effectiveness of interventions to communicate with people at risk, and given the importance of this topic and the need to inform decisions with the evidence available, we present here a summary of the thematic synthesis which includes 49 pieces of research and other literature that provides evidence about policy implementation and consumer experiences.

To our knowledge there is no other comprehensive review that systematically assembles systematically the research and literature on people's documented views, experiences and preferences, and on how their experiences of notification and support post-notification might be improved in the context of CJD or vCJD risk.

The results of this thematic synthesis has enabled us to outline a comprehensive approach to communicating with people at risk. This is described as a 'patient-centred communication framework', which is outlined below and presented with supporting evidence in [Appendix 15](#). Although the proposed communication framework is comprehensive, it is not intended as a model ready for implementation in its current form. Instead it represents a summary of the available evidence on communication with those at risk and so is intended as an input to decision making in these situations. The communication framework therefore requires tailoring to individuals, to specific incidents and to the requirements of local contexts, as well as requiring adjustment in response to practical constraints such as availability of resources and personnel.

Although this thematic synthesis focussed on people with medically-acquired CJD or vCJD risk, several subgroups were identified in the included literature. This included people at risk of CJD via confirmed routes (e.g. those people treated with contaminated hPH); vCJD through confirmed routes (e.g. blood transfusion); theoretical risk (e.g. CJD transmission through blood); as well as

possible candidates for future vCJD screening, subject to screening test availability. These subgroups may have unique communication needs and preferences, however, data from all subgroups were mapped to concepts in the data and contributed collectively to the communication framework. Identifying the unique needs and experiences of people based on different assessed degrees of risk of developing CJD or vCJD remains an issue to be addressed by future research.

The assembled research and literature on people's views, experiences and preferences for notification of their at-risk status and support post-notification identifies several features of good communication. It also clearly indicates that poor communication practices may have negative impacts or cause harm. Both of these findings may inform future communication strategies.

Although protection of the public health may mean that people at risk may not have a choice about whether to be notified of their at-risk status, there may be choices in how risk is communicated and support is provided. Notification in at least some circumstances may cause unavoidable psychological harms due to the burdensome nature of the information conveyed. Alongside this, however, may be harms that are avoidable if the notification and subsequent support are well-planned, developed with consideration of people's expressed needs and preferences, and with recognition of the impact of this information in people's lives.

A patient-centred communication framework

Our thematic analysis led to development of a patient-centred communication framework with 10 major components (denoted as F1 to F10; see also [Appendix 15](#)):

- F1: Ethical principles underpinning the proposed framework
- F2: Research findings associated with consumers' experiences - towards an assessment of harms and preferences
- F3: Programmatic approach to notification: a proposed model
- F4: Longitudinal model of support: a proposed model
- F5: Education of healthcare workers
- F6: Consumer group role
- F7: Evaluation, improvement and research priorities
- F8: National and jurisdictional issues
- F9: Public communication
- F10: Media issues

The framework identifies major sets of activities aiming to improve the experiences of people at risk of CJD and vCJD by addressing people's expressed needs for notification, follow-up and support. Embedded within the framework are two patient-centred models of communication:

- a model for notifying people of their at-risk status (F3);
- a model for supporting people post-notification (F4).

Taken together, these models form a continuum of communication with people at risk over time. Other major components of the

communication framework specify key activities occurring at the community and policy level (F5, F7 to F10). These activities do not directly interface with people at risk yet may form an enabling network of activities that support patient-centred communication with at-risk individuals (depicted by the notification and support models).

The framework therefore specifies a complex set of interrelated communication activities involving communication within and between multiple levels of the health system. We outline major features of the framework below.

The need for a purposeful, coordinated and programmatic approach to notification and support for people at CJD or vCJD risk; see particularly framework components F1, F2 and F8.

This aspect of the framework specifies that an alternative to ad hoc or poorly planned communication approaches is possible and desirable. It also makes explicit that harms may occur if notification is poorly planned or conducted.

In essence this means that incidents involving exposure to CJD or vCJD risk in healthcare settings require standardised processes to be in place, and these must specify what needs to happen, including how people are to be notified (F3).

Practically this is a more complex task, requiring coordinated communication processes that incorporate standardised content, formats and delivery to be established. It also means that support structures must be in place before individuals are notified.

Such planned, coordinated communication may promote high-quality, sensitive notification of at-risk status, and ensure that appropriate support is available. It also supports health professionals involved in notification by identifying the resources and training they need to effectively communicate risk and to provide support (F3).

The need for a longitudinal model of communication which considers impact; see particularly framework components F1 and F4.

A second major aspect of the framework is that communication with people at risk should take into account the potential impact of the notification (risk information) in people's lives. There should be acknowledgement that it is burdensome and introduces long-term uncertainty. This means that communication with those at-risk might be most useful if approached as a longitudinal program, rather than as a one-off event. For instance, planning how and when new research information should be communicated to people could be part of the program.

The need for communication that is flexible, tailored and responsive to needs; see in particularly framework components F2, F3 and F4.

A further major feature of the framework is that the communication strategies it proposes can be tailored to individuals and responsive to expressed needs. These needs include that for:

- accurate, up-to-date information on CJD or vCJD;
- access to comprehensive, informed support services; and
- monitoring of access to health care to prevent

discrimination.

Component F2 of the framework deals particularly with people's documented preferences and needs.

The framework's notification and support models incorporate multiple components to address these and other needs identified in the research, or to an individual's choice where possible (F3, F4). Since people may often live with knowledge of their at-risk status for many years, and their needs are not uniform or static, the framework incorporates processes that are responsive and can be tailored, including options to:

- receive emerging information (F4);
- consult with healthcare professionals (F3, F4);
- receive support from knowledgeable individuals (F4); and
- engage with consumer support groups (F6).

Importantly, such communication processes incorporate communication in many directions, including to and from consumers, between consumers and between different levels of health systems.

The need for infrastructure to support effective communication; see in particularly framework components F5, F8, F9 and F10.

Finally, while the notification and support models are individually complex, these may function best when embedded in a wider context of supportive infrastructure and activities. These broader activities may not directly involve people at risk, but may promote effective communication with individuals.

These wider activities include the need for education of the healthcare workforce, the public and the media (F5, F9, F10) and the need for national guidance on communication with people at risk, including review and monitoring of access to health care (F8). These are indicated by reports of poor understanding of CJD, vCJD and risk, and by the documented discrimination that people at risk report when accessing care (F1).

Improving community and healthcare workforce understanding of risk, infectivity and infection control guidelines in health settings may:

- diminish discrimination following disclosure of at-risk status;
- promote responses that are appropriate to an individual's level of risk;
- support sensitive communication in recognition of the harms associated with poor communication; and/ or
- encourage people at risk to disclose their status and share in public health protection.

Improving community understanding of CJD and vCJD may also help to destigmatise at-risk status. Standardised communication approaches outlined in regional or national protocols (F3, F8) might help to achieve this and might present mechanisms for monitoring access to health care to ensure that discrimination based on at-risk status does not occur.

Overall completeness and applicability of evidence

There are many gaps indicated by the research assembled in this Cochrane review and accompanying thematic synthesis. These include the lack of reliable research to inform choices about effective notification and support interventions identified by the empty Cochrane review; as well as gaps identified in the accompanying thematic synthesis.

Research gaps and priorities identified by the Cochrane review

There is a lack of reliable, valid research with which to inform decisions about the effectiveness of interventions with which to notify, support and follow-up people at risk. There is a need for rigorous evaluation of interventions to improve communication with at-risk individuals, in order to identify the best communication approaches over time. However, it may be challenging to design and conduct such research to rigorously address all of the communication issues related to notification and support strategies.

Research using rigorous methodology (such as an RCT) might be both theoretically possible and appropriate to assess the effectiveness of some strategies for communicating with people at risk. For example, it may be possible to conduct rigorous evaluations of the effects of support and follow-up interventions. This might include assessing alternate forms of peer or personal support, either in comparison to standard care or in head-to-head comparisons. It may be more challenging to evaluate rigorously the effectiveness of interventions to notify people of their at-risk status. Randomised or quasi-randomised controlled trials may not be ethical or practical in situations where people need to be notified of exposure to an iatrogenic risk without delay, in order to protect the public health. Similarly, it may be difficult to evaluate such notification interventions using other rigorous forms of evaluation (e.g. CBA or ITS studies that might fit Cochrane review inclusion criteria) that rely on pre-intervention assessment of outcomes: such approaches are unlikely to be possible from a practical standpoint, and may also carry ethical implications.

Many of the barriers to research in this area are practical: for example, the need to protect the public health by notifying people of their risk in a timely way once the incident or exposure is recognised may preclude an RCT or quasi-RCT. Establishing the research design for such studies ahead of time, together with ethics

approval, might mean that such study designs could be used to evaluate alternate forms of notification (e.g. notification via letter compared with notification via telephone call). However, a further significant practical barrier to any future research in this area, whether evaluating the effects of notification or support interventions, is the rarity of the disease risk. Even large incidents and notification exercises do not typically involve hundreds of people and the rarity of medically-acquired risk for CJD and vCJD would present significant challenges to researchers, in terms of recruiting adequate numbers of people to rigorous quantitative studies. Similarly, the rarity of the disease risk for CJD and vCJD suggests that the necessary resources and planning needed are unlikely to be devoted to the study of optimal communication strategies in the near future. Furthermore, the lack of clear consensus on whether it is more ethically sound to inform people (or not) of their risk of exposure to the risk of CJD and vCJD, and that what is known about these diseases and their associated risks changes over time, may also represent barriers to research in this area.

If rigorous studies are conducted in future it will be imperative that they seek and assess a range of benefits of interventions, as well as explicitly seeking information on harms. Such information may not be easily captured in standard ways and we would encourage anyone undertaking such research to consider collecting in-depth qualitative data on people's experiences (positive and negative) of notification and support post-notification, alongside quantitative data.

Research gaps and priorities identified by the thematic synthesis

The difficulties in designing rigorous studies to inform decisions about all aspects communication with those at risk, but particularly in relation to notification strategies, underscore the importance of the thematic synthesis that accompanies this Cochrane review.

The literature, systematically assembled and assessed in this thematic synthesis, indicates several clear gaps in the evidence (see also framework component F7). These include specific issues, such as the appropriate risk communication formats for accurately conveying CJD or vCJD risk and associated uncertainty; as well as larger gaps such as the ongoing social and emotional impacts of notification. There is a need for qualitative and other research to fill the gaps indicated.

Many of the components of the communication framework, which have been explicitly constructed from the available research literature evidence, could be rigorously evaluated in trials or other experimental research. However, we believe that a more pressing need may be to fill gaps in the assembled research identified both by the Cochrane review and by the framework, considered together.

This might include the following areas of research to inform future communication strategies, by identifying:

- The ongoing social and emotional impacts of notification on people at risk and their families.
- How people may be supported to deal effectively with knowledge of their at-risk status; and what ongoing needs or preferences people have regarding support and follow-up.
- How notification practices might be improved, for example, to minimise avoidable psychological and other harms; or to identify how those experiencing unavoidable harms could be best cared for.
- Risk communication formats which most accurately convey the risk of CJD or vCJD, as well as the uncertainty associated with the risk.

Research into the social, emotional and psychological impacts of notification of CJD or vCJD risk on patients and their families is a priority as the pool of people notified of risk grows. Such research may inform the development of future notification and support strategies.

There is a further issue on which this thematic synthesis was unable to provide guidance, and which might be usefully considered in future research: this concerns differential levels of risk. People at risk of CJD or vCJD, acquired medically, may be at different degrees of risk of developing the diseases in the future based on how their exposure happened. For example, people who received a transfusion with blood products known to have come from a person with vCJD are considered to have a relatively higher risk of developing vCJD than people exposed through other routes, although there may also be uncertainty associated with degrees of risk. The different degrees of risk carry with them different impacts and will have implications for the needs and experiences of the individuals involved. However, this thematic synthesis was unable to determine separately the different needs or experiences of people based on different levels of individual risk as this was not reported in the included research and literature. This issue needs further attention in future research.

One further point is that even if rigorous studies of intervention effectiveness are performed in the future, these will not necessarily supersede the information assembled and analysed in the thematic synthesis. There is, for example, important information available from the thematic synthesis that would not be identified easily in other research. In particular, we were able to capture the experiences of individuals concerning harms (psychological, emotional) and discrimination in relation to healthcare access by including documented experiences collected by consumer support groups and others. This type of information is not commonly reported in trials or other rigorous research, yet reveals that there are major potential harms associated with poor notification and support strategies - practices that health services should aim to remediate urgently.

Implications of the thematic synthesis for the Cochrane review and other future research

Our thematic synthesis assembles a range of research and other literature to develop a patient-centred communication framework for people at risk of CJD or vCJD. It highlights the need for further research in this area, which is underscored by the empty Cochrane review. This body of work assembled to date may inform future rigorous research in the area, including future primary studies (trials and other study types) as well as future updates of this Cochrane review, and systematic reviews in related areas.

One major implication for future research lies with the interventions to be assessed. The thematic synthesis describes two complex, multi-component models for communication (notification and support). Future studies in this area will need to consider and report a range of features of the specific communication interventions to be employed, such as the intervention aim, content and components; staging and timing; training and support needs; and information and other resource requirements for successful and appropriate delivery. Such intervention details were sought in this Cochrane review, based on key features identified by the thematic synthesis.

The thematic synthesis and framework also clearly indicate that individual communication on CJD and vCJD risk may best happen with attention to the wider context. Future studies will therefore need to consider carefully the range of contextual factors that may affect successful implementation of these kinds of complex interventions in real-world settings. These factors include higher-level coordination of communication processes and stages, involving several individuals and organisations (e.g. health departments, health professionals, support groups); community-level and health professional education with different purposes (e.g. destigmatising CJD and risk within the community; promoting accurate knowledge of infection control among health professionals); and processes and mechanisms for assessing and revising infection control guidelines and other materials to align them with the latest knowledge about CJD and vCJD.

The thematic synthesis may also inform the choice of outcomes to be assessed in future research, just as it has informed the range and types of outcomes sought in this Cochrane review. These include consumer, provider and health system outcomes that will be important for understanding the effects of complex communication interventions in people with CJD or vCJD risk. As indicated above, an essential consideration for future research will be an in-depth assessment of harms and adverse effects associated with communicating CJD and vCJD risk. Such harms may be associated with the distressing nature of the risk information. Just as harmful, and perhaps more alarming, may be the discrimination arising as people - who have acquired the risk for CJD or vCJD through health care - attempt to access the health care they require and have a right to receive. Future research must make every effort to identify and capture such adverse event data.

Practical and ethical implications associated with research on peo-

ple at risk means that future research and updates of this Cochrane review may need to take a broader approach. The thematic synthesis identified several communication interventions, particularly those aiming to support and follow up people at risk, that might be assessed further using a randomised trial or other rigorous design, although the rarity of the disease risk may present several practical challenges for trialists (see above). It seems neither possible nor feasible to conduct studies on the effectiveness of notification strategies, for example using RCT, CBA or ITS designs. Realistically, the most rigorous types of studies possible for investigating the effects of notification interventions might be certain types of non-randomised studies, e.g. retrospective cohort studies or other studies with an historical control, or case control studies. Quali-

tative research, too, will be invaluable.

Future updates of this Cochrane review will seek and include these additional types of research, should they become available. Methods for integrating such research within Cochrane reviews are in development (Higgins 2008) and may provide important information about interventions such as their suitability, acceptability or people's preferences, and adverse effects, as well as of effects of interventions. All of this information will be key for informing future policy and practice decisions about how to communicate most effectively with people at risk, and with the minimum associated harms. Proposed recommendations for future research, based on this Cochrane review and the thematic synthesis are given in Additional Table 3, using the EPICOT+ structure (Brown 2006b).

Table 3. Implications and suggestions for future research

Using the EPICOT+ format to develop future research directions		
Element to consider in future research	Implications and suggestions for future research arising from Cochrane review	Implications and suggestions for future research arising from thematic synthesis
E - Evidence	No rigorous studies to inform decisions about effectiveness exist.	A range of research and other literature exists (none methodologically rigorous for assessing intervention effectiveness); but with gaps identified (e.g. ongoing social and emotional impacts of risk information).
	Research is warranted clinically and ethically to inform decisions about communication with people at medically-acquired risk for CJD or vCJD.	
P - Population	People at risk for CJD or vCJD, where the risk has been acquired medically. Note that the issue of differential degree of risk of CJD and vCJD also needs to be investigated in future research.	
I - Intervention	Intervention choice for future studies might be informed by the thematic synthesis and framework. Interventions have multiple specific and identifiable aims - e.g. to notify people of risk, to support them over time, to improve follow-up and continuity of care. In all cases, consideration of range of features of the interventions themselves will be required - e.g. aim, contents and components, staging, timing, training and support needed. Consideration of a range of contextual factors (indicated by the thematic synthesis) in which the interventions are implemented will also be required.	Interventions are indicated for specific communication aims by the assembled research and literature. These can inform the choice of interventions to be assessed in rigorous studies. Gaps in the assembled literature on communication are also indicated - e.g. research to inform future communication strategies, by identifying: <ul style="list-style-type: none"> • Ongoing social and emotional impacts of notification. • Ways to support people to deal with knowledge of at-risk status. • Ongoing needs or preferences for support and follow-up. • How notification practices might be improved.

Table 3. Implications and suggestions for future research (Continued)

		<ul style="list-style-type: none"> ● Risk communication formats which most accurately convey the risk of CJD or vCJD and uncertainty.
C - Comparison	<p>Head-to-head comparisons of alternate communication intervention types may be most appropriate for certain interventions (i.e. support and follow-up) - to identify the most effective strategy.</p> <p>For notification interventions the most appropriate control may be retrospective (i.e. inability to obtain pre-intervention data before the notification event).</p>	Comparative analysis of data arising from different sources to identify the best ways of communicating with those at risk.
O - Outcome	<p>In future studies it will be imperative that both benefits and harms are comprehensively assessed and reported.</p> <p>Outcomes should include those relevant to consumers, providers and health systems; the choice may be informed by the thematic synthesis with consideration of the Cochrane Consumers and Communication Review Group taxonomy of outcomes (reflected in the planned outcomes for this Cochrane review).</p>	
T - Time stamp	October 2010	
Other aspects of research		
appropriate study types	<p>For notification interventions: retrospective cohort, case control studies may be the most rigorous possible design in this population.</p> <p>For support and follow-up interventions: RCT, quasi RCT, CBA or ITS may be appropriate and possible; selection of design will depend on aims and features of study and intervention, as well a practical considerations.</p> <p>Qualitative research is also indicated.</p> <p>Observational data may be used to provide information on adverse events, harms or complaints.</p>	<p>Qualitative research to fill gaps is needed. Research not constrained by design but approaches might include surveys, interviews or others to elicit preferences, needs and views, and including explicit assessment of harms and complaints data.</p>
disease burden or relevance	<p>Disease risk is rare: prevalence of 1/1,000,000 in general population (all forms CJD/vCJD). Medically-acquired CJD or vCJD may represent ~1% of this total.</p> <p>However:</p> <ul style="list-style-type: none"> ● Over time the number of people classified as being at-risk is growing. ● Exposure through medical routes continues, without ways to identify people through screening or when incubating the disease. ● CJD and vCJD are both 100% fatal; there is no effective treatment and even symptomatic treatment is not very effective. 	

Table based on the EPICOT+ structure proposed by [Brown 2006b](#).

Agreements and disagreements with other studies or reviews

As described in the [Background](#) to this review, notifying people of medically-acquired CJD or vCJD risk may share some similarities with other serious medically-acquired diseases (e.g. HIV, HCV), but there are also significant points of departure in the experiences of such diseases. Communication for CJD or vCJD risk may instead share more common ground with the experience of communication for rare diseases. A recent survey by the European Association for Rare Diseases, for example, highlighted several striking similarities between the experiences of those with a rare disease and those at-risk for CJD or vCJD ([EURODIS 2009](#)). This included poor notification practices, with 35% of respondents indicating that they had been informed of their diagnosis in an unacceptable manner or situation. The survey also documented a widespread loss of confidence in the health system as a result of delays in receiving care or needing to seek care from multiple doctors; and - most disturbingly - almost 20% of respondents indicated that they had been rejected by a healthcare professional because of the complexity of their disease or symptoms. A recent qualitative study in which people with rare diseases were interviewed about their experiences reiterated many of these concerns. These included people's desire for humane and sensitive disclosure of their diagnosis, for relevant, actionable information, and to be listened to and supported by health professionals ([Huyard 2009](#)).

Such findings suggest that many of the experiences of those at risk of CJD or vCJD are not isolated occurrences. Rather, the documented views and experiences of those living with a rare disease or with the risk for a rare disease indicate that coordinated, patient-centred models of communication and care are needed to improve people's experiences of health care, and health outcomes, and that the burden of living with and managing the disease or risk should not be left solely to the individuals affected ([Anonymous 2009](#); [EURODIS 2009](#)).

Adopting patient-centred models of communication and care may be the means of achieving better outcomes for people living with a rare disease or with risk of a rare disease such as CJD or vCJD. Given the documented negative consequences of poor or ad hoc communication approaches, and the growing recognition of the role of effective communication in improving health and other outcomes, there are compelling reasons and a clear ethical imperative to focus on improving communication with people with medically-acquired risk for CJD or vCJD, and with other rare diseases.

AUTHORS' CONCLUSIONS

Implications for practice

Implications of the Cochrane review

There is a lack of methodologically rigorous evidence of effectiveness to inform decisions about interventions to notify and support people at medically-acquired risk of CJD or vCJD. Research is needed to address this gap and to inform future practice and policy decisions for communicating with people at risk.

Implications of the thematic synthesis

Despite the absence of methodologically rigorous evidence of effectiveness to inform decisions about interventions to notify and support people at medically-acquired risk of CJD or vCJD, there are compelling ethical reasons to promote and implement effective communication strategies with these people.

Protection of the public health means that people may need to be notified of their at-risk status, but adopting an evidence-based approach to communication means that there may be better or worse ways to impart this information and to communicate subsequently with those at risk.

Considering the experiences, needs and preferences of consumers placed at uncertain risk of acquiring a rare and fatal disease through medical treatment clearly indicates both the negative consequences of poor communication and the need for patient-centred and responsive communication strategies that ensure that individuals are communicated with in a sensitive, appropriate manner.

Communication with people at risk may best happen via a planned and coordinated longitudinal programme of notification and support, acknowledging the impact of the information in people's lives and that harms may occur if notification is poorly planned or conducted. Ideally, individual communication would be flexible, capable of being tailored, and responsive to need. Such communication may be best embedded within a wider supportive infrastructure to enable effective risk communication with individuals by considering community, health system and health professional infrastructure, and aiming to diminish stigma and discrimination for those at risk.

The above findings identify key aspects of communication with those at risk, based on the evidence available. These might be used as an input to decision making by health services about how communication (notification and support) happens when these incidents occur, appropriately tailored to local populations, settings and need.

Implications for research

Implications of the Cochrane review

There is a lack of reliable research with which to inform decisions about interventions on notification, support and follow-up for people at risk. There is a need for rigorous evaluation of interventions to improve notification, support and follow-up, to identify the best approaches over time and to inform future decisions about communicating with those at risk. Intervention choice for future studies, as well as outcomes for assessment, might be informed by the thematic synthesis and framework.

We present suggestions for future research, based on this Cochrane review and the thematic synthesis and using the EPICOT+ structure (Brown 2006b) in Additional Table 3.

Implications of the thematic synthesis

Many of the components of the communication framework, which we have explicitly constructed from the available research literature evidence, could be rigorously evaluated in trials or other experimental research. However, we believe that a more pressing need is to fill gaps in the assembled research identified both by the Cochrane review and by the framework, considered together. There is a need for qualitative and other research to fill gaps indicated by the thematic synthesis and framework.

This might include the following areas of research to inform future communication strategies, by identifying:

- the ongoing social and emotional impacts of notification for people at risk and their families;
- people's preferences and needs for support and follow-up post-notification; and

- the optimal risk communication formats for accurately presenting and communicating the risk of CJD or vCJD, as well as uncertainty.

Research into the effects of differential levels of risk for CJD or vCJD is needed. Research into the social, emotional and psychological impact of notification of CJD or vCJD risk on patients and their families is a priority as the pool of people notified of risk grows, and may inform the development of future notification and support strategies.

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* Indicates the major publication for the study

CHARACTERISTICS OF STUDIES

Characteristics of excluded studies *[ordered by study ID]*

Study	Reason for exclusion
Aakko 2004	Risk communication; not communication with people at medically-acquired CJD/ vCJD risk.
Alaszewski 2003	Risk communication; not communication with people at medically-acquired CJD/ vCJD risk.
Allars 1994	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Anonymous 1996a	Description of award made for accurate reporting of BSE risk (journalism); not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1996b	BSE/ epidemic and reporting by newspapers; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1996c	Description of surveillance; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1996d	Description of BSE and food safety; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1996f	Description of hospital use of graft tissue; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1996g	Discussion of BSE risks and transmissibility; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1998	Description of medically acquired risk and screening for; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1999a	Comment on BSE and reporting of cases; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 1999b	Description of the issues associated with knowing at -risk status; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2000a	Description of issues for STD risk transmission; not communication with people at medically-acquired CJD/ vCJD risk. Note: exclusion not based on full translation (French).
Anonymous 2000b	Description of risk transmission; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001a	Description of technical aspects of infection control in medical settings/ transmissibility; mentions need to inform patients about risk of possible exposure, but no description or evaluation of the impact of notification. Not communication with people at medically-acquired CJD/ vCJD risk.

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Anonymous 2001b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Anonymous 2001c	Description of restriction on donations to prevent onwards transmission of BSE; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001d	Surveillance data only; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001e	Not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001f	Not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001g	Infection control but may be relevant re tracking of instruments for those at risk/ quarantining instruments used on suspected/at -risk individuals; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001h	Infection control issues; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001i	Transmission risks for vCJD; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2001j	Sterilisation techniques to reduce the risk of medical CJD transmission; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2002a	Technical discussion of decontamination; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2002b	Discussion of infection via bacterial, viral agents more generally; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2003a	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Anonymous 2003b	Description of BSE and meat consumption risk; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2004a	Refers to but does not describe notification and process to be used; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2004b	Not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2004c	Notification report, clinical exposure to risk; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2004d	Details of a case of vCJD contracted from blood donation; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2004e	Description of BSE and public health protection; not communication with people at medically-acquired CJD/ vCJD risk.

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Anonymous 2005b	Editorial on emerging pandemics; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2005c	May be an additional paper for others relating to NBS/ DoH UK notification exercises for blood products. Excluded based on study design.
Anonymous 2005d	May be an additional paper for others relating to NBS/ DoH UK notification exercises for blood products. Excluded based on study design.
Anonymous 2005e	Report of court approval to allow pentosan polysulphate for treating CJD patients; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2006a	Description of possible transmission via blood transfusion but no process evaluation and no description for notification; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2006b	Technical paper on BSE and tissue infectivity levels; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2006c	Description of haemophilia therapies/ risks of vCJD based on use of blood. Transcript of interview with authors including Ironside, Dolan, Pipe; may be additional reference for these papers (but excluded based on study design criteria).
Anonymous 2006d	Discussion of availability of a test for screening for BSE/ vCJD in humans; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2006e	Technical paper on BSE risk, infectivity, vCJD projections for numbers of infections; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2007a	Describes decontamination technique for prion removal; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2007b	Primary research into therapy; not communication with people at medically-acquired CJD/ vCJD risk.
Anonymous 2008a	Individual's account of family member with sporadic CJD; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Anonymous 2008b	Individual's account of family member with CJD as a result of Human Growth Hormone treatment in teenage years; not communication with people at CJD/ vCJD risk.
Appleby 1997	Description of BSE/vCJD risk and public risk perception; not communication with people at medically-acquired CJD/ vCJD risk.
Armengaud 2003	Description of role of doctors during epidemic; not communication with people at medically-acquired CJD/ vCJD risk. Note: excluded based on English abstract only.
Barash 2008	Technical analysis of surgery as risk factor for CJD; not communication with people at medically-acquired CJD/ vCJD risk.

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Barnett 2005	Management of health and social care provided for patients with CJD within the UK; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Baron 1991	Neurological/ scientific/ technical report on BSE and transmission risks; not communication with people at medically-acquired CJD/ vCJD risk.
Batthey 1999	Science communication; not about CJD specifically; not communication with people at medically-acquired CJD/ vCJD risk.
Bellaby 2003	Risk perception of acquiring vCJD from food; not communication with people at medically-acquired CJD/ vCJD risk.
Bennett 2005	Technical, transmission rates via surgery; not communication with people at medically-acquired CJD/ vCJD risk.
Blajchman 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Brooke 2004	Description of risk epidemiology or assessments for iatrogenic CJD; not communication with people at medically-acquired CJD/ vCJD risk.
Brown 2001a	Technical description of screening test requirements; not communication with people at medically-acquired CJD/ vCJD risk.
Brown 2001b	Technical description of infectivity and transmissibility, screening; not communication with people at medically-acquired CJD/ vCJD risk.
Brown 2005	Description of CJD pathogenesis, and experimental transmission and infectivity studies. Not communication with people at medically-acquired CJD/ vCJD risk.
Butler 1996a	Description of BSE risk and management, political enquiry. Not communication with people at medically-acquired CJD/ vCJD risk.
Butler 1996b	Epidemiology and surveillance reporting; not communication with people at medically-acquired CJD/ vCJD risk.
Butler 1996c	Discussion of decision-making and policy re BSE; not communication with people at medically-acquired CJD/ vCJD risk.
Butler 2001	Clinical description of patients with diagnosed CJD and issues for psychiatric management. Not communication with people at medically-acquired CJD/ vCJD risk.
Callum 1999	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Calman 2002	Overview on risk communication; not specifically communication with people at medically-acquired CJD/ vCJD risk.
Carruthers 2001	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.

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Caulfield 1997	Discussion of ethics of notification; not communication with people at medically-acquired CJD/ vCJD risk.
CDHFS 1995	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CDHFS 1996	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Cegelka 2002	Qualitative study to identify the thoughts, feelings, problems and experiences of caregivers whose family member has died from CJD; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Check 2002	Technical report on surveillance/ infection control (screening of tissue samples); not communication with people at medically-acquired CJD/ vCJD risk.
CJD IP (Framework) 2005	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD IP 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD IP 2005a	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD IP 2005b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD IP 2006	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD IP, HPA, CDO 2006	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD SGN	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD SGN 2006a	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD SGN 2006b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD SGN 2007a	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD SGN 2007b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
CJD SGN 2008	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Clark 1997	Description of graft susceptibility of recipients; not communication with people at medically-acquired CJD/ vCJD risk.
Clarkson 2000	Description of hospital disinfection guidelines for CJD and vCJD, risk minimization guidelines; not communication with people at medically-acquired CJD/ vCJD risk.
Coulthart 2001	Review on BSE/vCJD transmissibility and infectivity, epidemiology; not communication with people at medically-acquired CJD/ vCJD risk.

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Creuzot-Garcher 2004	Risk assessment, screening for at-risk people; not communication with people at medically-acquired CJD/ vCJD risk.
Culver 2000	Theoretical discussion of disclosing risk, especially ethical aspects but no notification / evaluation of notification; not communication with people at medically-acquired CJD/ vCJD risk.
Darlington 1996	Description of BSE risk and implications; not communication with people at medically-acquired CJD/ vCJD risk.
Day 1999	Description of screening/ diagnostic testing issues; not communication with people at medically-acquired CJD/ vCJD risk.
Dealler 1996	Discussion of risk of transmission through diet, BSE risk and transmissibility; not communication with people at medically-acquired CJD/ vCJD risk.
DHAC 2001	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
DHS 2006	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Dickson 1998	Description of compensation for hPH recipients; not communication with people at medically-acquired CJD/ vCJD risk.
Dolan 2006	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Douglas 1999	Qualitative study to identify the needs of families and carers for information and practical support, from the perspectives of families and healthcare professionals who had been involved in their care; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Du Val 1997	Description of the ethics of notification; not communication with people at medically-acquired CJD/ vCJD risk.
Duncan 2005	Description of the ethics of notification and benefits of particular decision rules for personal autonomy versus public health protection; not communication with people at medically-acquired CJD/ vCJD risk.
Edwards 2003	General discussion on risk communication, not CJD specifically; not communication with people at medically-acquired CJD/ vCJD risk.
Evatt 1998	Description of epidemiology of transmission; considers theoretical possibility of blood transfusion risk; not communication with people at medically-acquired CJD/ vCJD risk.
Farrell 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Farrugia 2002	Complements Farrugia 2005 ; vCJD transmission via blood and blood products. Excluded based on study design.
Farrugia 2005	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.

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FDA 2006a	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
FDA 2006b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Fernandez 1998	Description of clinical and other features of CJD, transmissibility, infection control guidance and risk management; not communication with people at medically-acquired CJD/ vCJD risk.
Fishman 1998	Technical report on infection control for CJD; not communication with people at medically-acquired CJD/ vCJD risk.
Fitzpatrick 1996	Letter stating no link between BSE and CJD; not communication with people at medically-acquired CJD/ vCJD risk.
Forbes 2004	Policy analysis; technical link between BSE and VCJD; not communication with people at medically-acquired CJD/ vCJD risk.
Freedman 1997	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Frewer 2003	Survey of risks and trust associated with food; not communication with people at medically-acquired CJD/ vCJD risk.
Galloway 1996	Description of patient death from CJD, not at-risknot communication with people at medically-acquired CJD/ vCJD risk.
Giovannini 2007	Review of book on BSE crisis and risk perception/ public policymaking; not communication with people at medically-acquired CJD/ vCJD risk.
Girardi 1998	Analysis of media coverage of BSE; not communication with people at medically-acquired CJD/ vCJD risk.
Goodnough 2004	Description of blood service notifications, may be additional to other papers within the thematic synthesis. Excluded based on study design.
Grant 1992	Opinion letter, outlining the desirability of notification; not communication with people at medically-acquired CJD/ vCJD risk.
Green 2001	Description of BSE and food and blood safety; historical review of BSE worldwide and policy responses. Not communication with people at medically-acquired CJD/ vCJD risk.
Green 2005	Discussion of food safety, BSE and food safety as well as public risk information; not communication with people at medically-acquired CJD/ vCJD risk.
Gunasekera 1996	Discussion of BSE risk perceptions and behaviour (reports of); not communication with people at medically-acquired CJD/ vCJD risk.
Hamilton 2004	Infection control in people at risk; not communication with people at medically-acquired CJD/ vCJD risk.

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Hammersmith 2004	Case report of clinical care for a confirmed CJD patient (deceased); not communication with people at medically-acquired CJD/ vCJD risk.
Hammersmith 2005	Discussion of issues of informed consent for eye surgery (corneal transplant); not communication with people at medically-acquired CJD/ vCJD risk.
Hampshire 2003	Description of diagnosis and care of people with CJD; not communication with people at medically-acquired CJD/ vCJD risk.
Harakeh 2003	Survey of students' knowledge of BSE risk, modification of risk behaviours (meat consumption); not communication with people at medically-acquired CJD/ vCJD risk.
Harrington 1998	Outline of risk communication and public health risk perceptions, communication and policy; BSE/ CJD as an example. Not communication with people at medically-acquired CJD/ vCJD risk.
Harrison 1996	Description of risk communication generally; not communication with people at medically-acquired CJD/ vCJD risk.
Hart 2004	Discussion of informed consent for transfusion and vCJD risk; not communication with people at medically-acquired CJD/ vCJD risk.
Hartley 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Harvey 1996	Discussion paper on risk communication; not communication with people at medically-acquired CJD/ vCJD risk.
Healy 1993	Description of chronology of CJD transmission via pituitary hormones; not communication with people at medically-acquired CJD/ vCJD risk.
Healy 2001	Description of disease and need for screening tests and investment to find treatment/ cure; not communication with people at medically-acquired CJD/ vCJD risk.
Hewitt 2006a	Analysis of transmission through transfusion; epidemiological study of transmission and blood transfusion; not communication with people at medically-acquired CJD/ vCJD risk.
Hewitt 2006b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Higgins 2004	Technical description of case study of instrument decontamination and associated issues for re-use of instruments. Not communication with people at medically-acquired CJD/ vCJD risk.
Hinchliffe 2001	Policy analysis focusing on animal feed policy in the context of BSE; not communication with people at medically-acquired CJD/ vCJD risk.
Howe 2001	Discussion of ethical implications of notification and discussion of individual variation in response to disclosure. Theoretical; not communication with people at medically-acquired CJD/ vCJD risk.
HPA 2005	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.

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HPA 2007	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
HPA/HPS 2006	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Hunt 1996	Ethical argument over freedom of information re BSE; ethics and transmission of BSE via food; no consideration of notification/ impact of information. Not medically-acquired CJD risk.
Ironside 1996	Description of clinical features, epidemiology of human prion diseases; not communication with people at medically-acquired CJD/ vCJD risk.
Ironside 2006	Discussion of epidemiology of vCJD; includes some discussion of issues for haemophiliacs and need to notify. Not communication with people at medically-acquired CJD/ vCJD risk.
Jacob 2000	Description of policy making and science in the context of BSE; not communication with people at medically-acquired CJD/ vCJD risk.
Jacobson 2004	Editorial on BSE and food chain in US, BSE/ vCJD food safety risk; not communication with people at medically-acquired CJD/ vCJD risk.
Jansen 2003	Epidemiology/ surveillance numbers; not communication with people at medically-acquired CJD/ vCJD risk.
Jefferson 2003	Report on disposable equipment; not communication with people at medically-acquired CJD/ vCJD risk.
Jeffries 2006	Overview of history of BSE/vCJD in UK and discussion of current policy on surgical instruments, decontamination and infection control; not communication with people at medically-acquired CJD/ vCJD risk.
Jelinek 2008	Malaria; not communication with people at medically-acquired CJD/ vCJD risk.
Johnson 1998	Review of information on CJD/ TSEs; not communication with people at medically-acquired CJD/ vCJD risk.
Johnson 2007	Description of aetiology and technical information on current state of knowledge on TSEs; not communication with people at medically-acquired CJD/ vCJD risk.
Johnston 2001	Technical discussion on infection control and aetiology; not communication with people at medically-acquired CJD/ vCJD risk.
Kapoor 2001	Description of disposable instruments and tonsillectomy; not communication with people at medically-acquired CJD/ vCJD risk.
Kemmann 1998	Discussion of informed consent re ART and theoretical CJD risk, refers to a disclosure program regarding albumin product used on embryo culture: theoretical risk to embryo. No information about the disclosure program; not communication with people at medically-acquired CJD/ vCJD risk.

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Kewell 2008	Textual analysis and language used, policy and BSE/vCJD link; not communication with people at medically-acquired CJD/ vCJD risk.
Khashia 2006	Primary research into therapy; not communication with people at medically-acquired CJD/ vCJD risk.
King 1998	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
King 2001	Description of anaesthetic equipment and infection control, not specifically CJD; not communication with people at medically-acquired CJD/ vCJD risk.
King 2008	Background, news report on French trial (GH cases) on how people were exposed to CJD medically; not communication with people at medically-acquired CJD/ vCJD risk.
Kirby 2007	Discussion of policy development process, reference to disclosure policy as an example, no description of processes; not communication with people at medically-acquired CJD/ vCJD risk.
Kirkpatrick 2002	Technical paper on infection control, single use instruments; not communication with people at medically-acquired CJD/ vCJD risk.
Kirkup 2003	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Kitchin 1972	Description of technical genotyping study, frequency of genetic variation associated with Kuru; not communication with people at medically-acquired CJD/ vCJD risk.
Klein 2000a	Editorial on policies around BSE/ vCJD risk, Phillips Report; not communication with people at medically-acquired CJD/ vCJD risk.
Kmietowicz 2004	Description of notification; already have detailed policy papers for (Hewitt, Blachjman papers) included in thematic synthesis. Not a primary report of notification/ communication.
Krebs 2001	Description of BSE/ food risk and safety, role of FDA and development of relevant policy; not communication with people at medically-acquired CJD/ vCJD risk.
Kurth 2007	Discussion of public health risks; not communication with people at medically-acquired CJD/ vCJD risk.
Kuster 1999	Description of speech and language resources for dementias; not communication with people at medically-acquired CJD/ vCJD risk.
Landals 2004	Description of veterinary issues; not communication with people at medically-acquired CJD/ vCJD risk.
Lanska 1998	Discussion of risk and policy around beef, notification, BSE and vCJD policy analysis and risk management; not communication with people at medically-acquired CJD/ vCJD risk.
Larke 1998	Editorial, includes case studies on beef consumption; not communication with people at medically-acquired CJD/ vCJD risk.

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Lee 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Lemyre 2009	News report on setting up surveillance for BSE and risk communication; not communication with people at medically-acquired CJD/ vCJD risk.
Lewis 1994	Description of BSE/vCJD surveillance in animals; not communication with people at medically-acquired CJD/ vCJD risk.
Liberski 2005	Description of neurodegeneration, specifically Alzheimer's disease; not communication with people at medically-acquired CJD/ vCJD risk.
Llewelyn 2004	Description of recipients of blood transfusions donated by individuals who later developed vCJD and elevated risk of vCJD and the need to notify; no description of the notification or other communication or impact of communications on those at risk.
Lovasik 2004	Technical overview of CJD and TSEs, including patient management; not communication with people at medically-acquired CJD/ vCJD risk.
Lowe 2006	Description of project on blood substitutes (to decrease blood-borne transmission risk); not communication with people at medically-acquired CJD/ vCJD risk.
Lukasiewicz 2001	Comparison of different ways of presenting risk to GPS (including risk of nvCJD); not communication with people at medically-acquired CJD/ vCJD risk.
Lurie 2004	Description of aetiology and BSE/ vCJD link in US; not communication with people at medically-acquired CJD/ vCJD risk.
Lyons 1992	Comment on disease surveillance and case finding; not communication with people at medically-acquired CJD/ vCJD risk.
Lysons 2004	Disease surveillance measures; not CJD specifically; not communication with people at medically-acquired CJD/ vCJD risk.
MacEvelly 2001	Food safety and public risk communication; not communication with people at medically-acquired CJD/ vCJD risk.
Mannion 2000	Technical report on hospital-acquired infections; not CJD specifically; not communication with people at medically-acquired CJD/ vCJD risk.
Marmot 1996	Editorial on risk communication principles; not communication with people at medically-acquired CJD/ vCJD risk.
Martin 2006	Analysis of risk of transmission of vCJD via blood donation; not communication with people at medically-acquired CJD/ vCJD risk. Note: decided on English abstract only (fulltext in French).
Martinez 2003	Patient case descriptions; not communication with people at medically-acquired CJD/ vCJD risk. Note: decided on English abstract only (fulltext in Spanish).

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Masters 2001	Description of surveillance, risk containment and infection control; not communication with people at medically-acquired CJD/ vCJD risk.
McCarthy 1996	Surveillance in USA on CJD; not communication with people at medically-acquired CJD/ vCJD risk.
McConnell 1997	Brief report on status of number of cases of vCJD and the BSE/vCJD link; not communication with people at medically-acquired CJD/ vCJD risk.
McCrea 2003	Discussion of risk communication principles using BSE/vCJD as an example; not communication with people at medically-acquired CJD/ vCJD risk.
McCrea 2005	Risk communication principles re risk communication and food; not communication with people at medically-acquired CJD/ vCJD risk.
McCullough 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
McDade 2000	Description of new pathogens; not communication with people at medically-acquired CJD/ vCJD risk.
McDonald 2002	Description of BSE/vCJD link, surveillance, infection control; not communication with people at medically-acquired CJD/ vCJD risk.
McIntosh 2002	Description of TSEs; not communication with people at medically-acquired CJD/ vCJD risk.
McMahon 2001	Description of BSE, features, transmissibility re vCJD in medical settings (issues for nurses); not communication with people at medically-acquired CJD/ vCJD risk.
McMahon 2004	Description of link between BSE and vCJD; not communication with people at medically-acquired CJD/ vCJD risk.
McPherson 1996	Description of BSE/vCJD surveillance and use of pre-operative questionnaire re risk of CJD; not communication with people at medically-acquired CJD/ vCJD risk.
Mead 2005	Description of use of screening questionnaires to detect those at risk; US FDA policy on blood risk and lookback; not communication with people at medically-acquired CJD/ vCJD risk.
Menache 1996	Description of transfusion transmissibility, aetiology, background rates; not communication with people at medically-acquired CJD/ vCJD risk.
Milner 2000	Description of CJD incidence/ case report; letter to editor from sibling of CJD patient. Not communication with people at medically-acquired CJD/ vCJD risk.
Mocsny 1998	Description of CJD, management, infection control; not communication with people at medically-acquired CJD/ vCJD risk.
Molesworth 2006	Description of surveillance/ deaths from CJD; not communication with people at medically-acquired CJD/ vCJD risk.

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Morel 1998	Description of haemovigilance and tools for ensuring transfusion safety (identifying risk factors and monitoring risks to reduce transmission risks); not communication with people at medically-acquired CJD/ vCJD risk.
Morita 1998	Description of disease surveillance, infectious diseases; not communication with people at medically-acquired CJD/ vCJD risk.
Morris 2007	Description of counselling and support for patients with CJD and their families; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Myles 2002	Estimation of the costs (both direct and indirect) borne by families caring for people with vCJD; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Najjar 2005	Description of informed consent for eye surgery; case report of CJD and corneal transplantation. Not communication with people at medically-acquired CJD/ vCJD risk.
Neatherlin 1988	Description of epidemiology, aetiology, management of patients with CJD and their families; not communication with people at medically-acquired CJD/ vCJD risk.
Newcombe 1996	Technical paper on risk associated with specific neurosurgical procedures; mentions tracing dura graft recipients. Not communication with people at medically-acquired CJD/ vCJD risk.
Normile 2004	Meat testing standards for BSE, surveillance in animals; not communication with people at medically-acquired CJD/ vCJD risk.
NSW DoH 2004	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
O'Brien 2001	Description of agencies and roles where cases of CJD occur; communicable diseases policy and surveillance report. Not communication with people at medically-acquired CJD/ vCJD risk.
Olsen 2005	Technical analysis of risk of transmission of CJD risk through surgery; not communication with people at medically-acquired CJD/ vCJD risk.
Operskalshi 1995	Description of incidence/ surveillance re blood transfusion risk and CJD; not communication with people at medically-acquired CJD/ vCJD risk.
Painter 2000	Technical report of TSEs, risk and risk containment; not communication with people at medically-acquired CJD/ vCJD risk.
Pauls 1996	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Pfister 2001	Focus on risk communication, with BSE as an example; psychology of risk communication to general populations about BSE. Not communication with people at medically-acquired CJD/ vCJD risk.
Pipe 2006	Overview of emerging pathogens and blood supply; not communication with people at medically-acquired CJD/ vCJD risk.

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Pitrelli 2007	Discussion of public health threats, review of policy and communication review. Not communication with people at medically-acquired CJD/ vCJD risk.
Podger 2001	Description of release of BSE report, food standards. Not communication with people at medically-acquired CJD/ vCJD risk.
Populus 2007	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Ramasamy 2003	Technical discussion and modelling of the risk of transmission following surgery; not communication with people at medically-acquired CJD/ vCJD risk.
Ramsey 2004	Description of management of blood component recalls; not specifically vCJD/ CJD. Not communication with people at medically-acquired CJD/ vCJD risk.
Ratzan 1998a	Discussion of risk communication; not communication with people at medically-acquired CJD/ vCJD risk.
Ratzan 1998b	Discussion of risk communication re BSE/vCJD link; not communication with people at medically-acquired CJD/ vCJD risk.
Reesink 2003	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Reingold 1996	Description of CJD surveillance; not communication with people at medically-acquired CJD/ vCJD risk.
Rentz 2008	Technical report on patients with CJD, nursing issues etc; not communication with people at medically-acquired CJD/ vCJD risk.
Ricketts 1997	Discussion of epidemiology and theoretical nature of risk of transmission for CJD through blood; not communication with people at medically-acquired CJD/ vCJD risk.
Riesner 2001	Technical paper on science and theory of prion proteins; not communication with people at medically-acquired CJD/ vCJD risk.
Roberts 1999	Description of case study re hospital infection control for patient with vCJD and decontamination required; not communication with people at medically-acquired CJD/ vCJD risk.
Robotin 2002	Evaluation of ANZCJD Registry (surveillance and adequacy of services); not communication with people at medically-acquired CJD/ vCJD risk.
Rohwer 2004	Description of assessing the BSE/ vCJD link and transmissibility through blood transfusion; not communication with people at medically-acquired CJD/ vCJD risk.
Roos 2001	Discussion of Infectivity/ transmissibility of BSE; not communication with people at medically-acquired CJD/ vCJD risk.
Ross 2002	Technical report including guidelines for surgery; patients notified of possible exposure to CJD risk but no description/ evaluation. No communication with people at medically-acquired CJD/ vCJD risk.

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Ruef 2002	Infection control and prevention of transmission in medical settings; not communication with people at medically-acquired CJD/ vCJD risk.
Salamon 1994	Discussion of blood transfusion risks, surveillance. Not communication with people at medically-acquired CJD/ vCJD risk. Note: decided based on English abstract only (fulltext in French).
Sanders 2000	Surveillance data and discussion of blood transmission of prion diseases. Not communication with people at medically-acquired CJD/ vCJD risk.
Sandler 2006	Ethics and risk communication in the context of vCJD and blood transmission; opinion piece on accuracy of contemporary testing methods. Not communication with people at medically-acquired CJD/ vCJD risk.
Schneider 2007	Huntington's disease (and similar genetic risk diseases). Not communication with people at medically-acquired CJD/ vCJD risk.
Sehulster 1998	Description of decontamination/ risk prevention for transmission; also patient characteristics and disease latency. Not communication with people at medically-acquired CJD/ vCJD risk.
Seitz 2005	Technical description of TSEs including risk (transfusion, transmission, sterilisation, screening), and course of disease. Not communication with people at medically-acquired CJD/ vCJD risk.
Senate 1997	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Shalowitz 2009	Discussion of the ethics of notification and knowledge of the exposure to risk/ risk information. Not description or impact of communication with people at medically-acquired CJD/ vCJD risk.
Shamrock Marketing 2002	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Sheff 2005	Description of BSE and vCJD risk/ link; supportive nursing for CJD patients, and also some generic information about what to tell patients about the risk from food. Not communication with people at medically-acquired CJD/ vCJD risk.
Shickle 2000	Public risk communication, including BSE as a risk and portrayal in the media; not communication with people at medically-acquired CJD/ vCJD risk.
Sibbald 1998	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Sicard 2000	Description of blood transfusion safety; not communication with people at medically-acquired CJD/ vCJD risk. Note: decided based on English abstract only (fulltext in French).
Sickmuller 1997	Description of link between BSE contamination and vCJD risk, regulation of animal-derived pharmaceutical products. Not communication with people at medically-acquired CJD/ vCJD risk.
Siegel 2007	Guidelines for infection control, general discussion of transmission of infectious agents in healthcare settings (background, mode, exposure risk). Section on CJD mentions notification, but not the process of communication; not communication with people at medically-acquired CJD/ vCJD risk.

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Siegenthaler 2004	Technical, risk control of new agents such as prions in transfusion medicine; not communication with people at medically-acquired CJD/ vCJD risk. Note: decided on English abstract only (fulltext in French).
Simpson 1996	Perceptions of BSE risk (doctors surveyed about BSE risk); not communication with people at medically-acquired CJD/ vCJD risk.
Skegg 1997	Description of TSEs and BSE generally; not communication with people at medically-acquired CJD/ vCJD risk.
Smith 1999	Description of food safety and consumers' perspectives on BSE risk; not communication with people at medically-acquired CJD/ vCJD risk.
Smith 2003	Describes several cases of CJD and/or risk exposure associated with dental procedures, infection control needs as illustrative of the current guidelines to manage known cases. Not communication with people at medically-acquired CJD/ vCJD risk.
Smith 2003a	Editorial outlining papers in issue, mentions one on CJD and clinical uncertainty. Not communication with people at medically-acquired CJD/ vCJD risk.
Smith-Bathgate 2005	Description of nursing care for patients with CJD, diagnosis issues, nursing treatment and surveillance issues. Not communication with people at medically-acquired CJD/ vCJD risk.
Sofer 2004	Discussion of biopharmaceutical processing to remove prion (BSE) risk; not communication with people at medically-acquired CJD/ vCJD risk.
Soldan 2005	Description of the need for and process of notification for blood donors/ recipients re vCJD risk exposure. Does not describe the processes or impacts of communication with people at medically-acquired CJD/ vCJD risk.
Spencer 2002	Technical report on sterilisation/ decontamination techniques and practices; not communication with people at medically-acquired CJD/ vCJD risk.
Srinivasan 2005	Informed consent for eye surgery; letter discussing a case report. Not communication with people at medically-acquired CJD/ vCJD risk.
Stainsby 2004	Description of monitoring of adverse events associated with blood transfusion; measures to reduce risk of blood transmission of vCJD. Not communication with people at medically-acquired CJD/ vCJD risk.
Steelman 1994	Technical discussion of infection control and sterilisation processes/ decontamination; not communication with people at medically-acquired CJD/ vCJD risk.
Steinberg 2001	Discussion of ethics of notification, decision not to inform of risk; not communication with people at medically-acquired CJD/ vCJD risk.
Steinberg 2002	Discussion of ethics of notification; not communication with people at medically-acquired CJD/ vCJD risk.

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Stephenson 2007	Technical description of decontamination techniques, measures to prevent spread of CJD risk in health-care settings (infection control and sterilisation techniques); not communication with people at medically-acquired CJD/ vCJD risk.
Stratton 1997a	CJD cases, epidemiology and tracking in Canada including notification of authorities where diagnosis is confirmed; not communication with people at medically-acquired CJD/ vCJD risk.
Stratton 1997b	Description of epidemiological trends in CJD mortality; not communication with people at medically-acquired CJD/ vCJD risk.
Strength 1992	Technical description of dementias, including CJD, and clinical care; not communication with people at medically-acquired CJD/ vCJD risk.
Stricof 2006	Description of lookback at surgical records regarding two confirmed CJD diagnoses; not communication with people at medically-acquired CJD/ vCJD risk.
Sullivan 2005	Technical chapter on blood supply, donation, transfusion and international trends in blood banking policies; not communication with people at medically-acquired CJD/ vCJD risk.
Sulser 2006	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Tammelleo 1996	Description of legal case re medical malpractice (dura matter labeled for investigational use only, subsequent court case arising as patient contracted and died from acquired CJD). Not communication with people at medically-acquired CJD/ vCJD risk.
Tamura 2007	Counselling, information and support for people with genetic CJD risk; not medically-acquired risk for CJD/ vCJD.
Tien 2007	Not communication with people at medically-acquired CJD/ vCJD risk.
Tordoff 2002	Discussion of protein contamination of specific medical equipment; not communication with people at medically-acquired CJD/ vCJD risk.
Tubiana 2000	Description of precautionary principle; may be relevant to medically at-risk populations but not communication with people at risk.
Tubiana 2001	Description of precautionary principle; may be relevant to medically at-risk populations but not communication with people at risk; appears to be same review as Tubiana 2000 .
Tullo 2003	Description of practicalities associated with infection control for people at risk undergoing ophthalmic surgery; not communication with people at risk.
Tullo 2006	Technical case report of possible iatrogenic risk transmission via ocular transplant; relevant population. Does not describe the processes of notification, communication or support.
Turner 2003	Screening blood for vCJD, technical report; not communication with people at medically-acquired CJD/ vCJD risk.

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Turner 2004	Discussion of issues for patients and delivery of care, service requirements and delivery. Not communication with people at medically-acquired CJD/ vCJD risk.
Turner 2009	Overview; discusses implications of notification of positive screening test results and notification as a result of blood products. Covers incidents already described in detail in other included papers (secondary report, not much detail). No focus on communication with people at medically-acquired CJD/ vCJD risk.
van den Burg 1998	General discussion of blood donation policies and donor selection; not specifically CJD/ vCJD risk; not communication with people at medically-acquired CJD/ vCJD risk.
Vaughan 1996	Discussion of ethics and transmissibility (risk to blood supply); not communication with people at medically-acquired CJD/ vCJD risk.
vCJD CGAG 2007	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Verbeke 1999	BSE risk and public communication of risk re food safety; not communication with people at medically-acquired CJD/ vCJD risk.
Verity 2000	Follow-up surveillance in children re suspected vCJD risk (similar data to Verity 2006; different time frames sampled); not communication with people at medically-acquired CJD/ vCJD risk.
Verity 2006	Surveillance in children re suspected vCJD risk; not communication with people at medically-acquired CJD/ vCJD risk.
Wallis 2004	Technical discussion of post-transfusion mortality generally; not communication with people at medically-acquired CJD/ vCJD risk.
Ward 2008	Assessment of blood transmission of risk of vCJD (from confirmed cases); not communication with people at medically-acquired vCJD risk.
Weller 2003	Discussion of palliative care issues for patients with CJD; not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
WHO 2006a	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
WHO 2006b	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.
Wilks 2000	BSE/ vCJD, food risks and risk communication for public and policy making; not communication with people at medically-acquired CJD/ vCJD risk.
Willesee 2008	Description of a case of a 58-year-old woman who developed CJD (sporadic); not communication for people at CJD/ vCJD risk, nor specifically medically-acquired risks.
Wilson 2001	Analysis of policy decisions; not communication with people at medically-acquired CJD/ vCJD risk.
Wilson 2004a	Analysis of media coverage on BSE; not communication with people at medically-acquired CJD/ vCJD risk.

(Continued)

Wilson 2004b	Public communication for BSE risk; not communication with people at medically-acquired CJD/vCJD risk.
Wilson 2006	Discussion of risk of vCJD in different genotype group; not communication with people at medically-acquired vCJD risk.
Wroe 2006	Case report of patient with vCJD (who may have been exposed to vCJD risk through blood); not person with risk for vCJD.
Yamauchi 2002	Deals with care of patients with CJD, not people with risk for CJD.
Yuasa 2002	Deals with care of patients with CJD, not people with risk for CJD.
Zekauskas 1990	Did not meet study design criteria but met all other criteria for inclusion in thematic synthesis.

Characteristics of studies awaiting assessment *[ordered by study ID]*

Anonymous 1996e

Methods	
Participants	
Interventions	
Outcomes	
Notes	French; requires translation.

Anonymous 2005a

Methods	
Participants	
Interventions	
Outcomes	
Notes	German; requires translation.

Anonymous 2005f

Methods	
Participants	
Interventions	
Outcomes	
Notes	German; requires translation.

Bastin 1997

Methods	
Participants	
Interventions	
Outcomes	
Notes	Italian; requires translation.

Boixiere 2004a

Methods	
Participants	
Interventions	
Outcomes	
Notes	French; requires translation.

Boixiere 2004b

Methods	
Participants	
Interventions	
Outcomes	
Notes	French; requires translation.

Chambaud 2003

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Chapuis 2006

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Dubouis 2001

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Nau 2002

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Nau 2004

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Nau 2005

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Proust 2001

Methods	
Participants	
Interventions	
Outcomes	
Notes	French, requires translation.

Quimper 2006

Methods	
Participants	
Interventions	
Outcomes	
Notes	Full text not obtainable.

Rigal 2006

Methods	
Participants	
Interventions	
Outcomes	
Notes	German, requires translation.

Tull 2000

Methods	
Participants	
Interventions	
Outcomes	
Notes	Swedish; requires translation.

Van Roon 1999

Methods	
Participants	
Interventions	
Outcomes	
Notes	Dutch; requires translation.

DATA AND ANALYSES

This review has no analyses.

APPENDICES

Appendix I. Outcomes sought

We planned to collect information on a range of outcomes relevant to consumers (patients and their families/carers), health professionals and health systems.

We sought data on the following outcomes:

Primary outcomes

Consumer-oriented outcomes, including:

- Knowledge and understanding - e.g. Understanding of how the risk was acquired; understanding of the risk status of the patient and the risk levels (infectivity) associated with different tissues and medical procedures; understanding of the disease, its progression and outcome; knowledge of availability of health and support services; knowledge of treating professionals, eligibility for additional services, such as counselling; perceived risk, accuracy of, or change in accuracy of perceived risk; patient's or family members' wishes regarding whether to be informed or not;
- Health status and wellbeing - e.g. quality of life; physical or psychological health outcomes for the patient, carer and/or family, including emotional outcomes such as fear, anxiety or distress;
- Communication - e.g. use of communication aides; satisfaction with communication;
- Evaluation of care - e.g. perceptions/ ratings of care or interventions received; satisfaction with care or interventions or processes or decisions; and
- Support - e.g. whether people accepted practical or psychosocial support (such as information, counselling), and the effects of support (such as perceived or actual support, social function, isolation, burden).

We also sought data on potential harms and adverse events associated with the interventions, for the patient, family members and/or carers, and for the healthcare providers involved in delivering interventions or care and management. This was to include consideration of emotional and psychological harms, such as anxiety, psychological distress and fear. We also would have specifically considered harms relating to changes in access to medical and health treatments; and perceived and actual discrimination. We also sought to identify any complaints regarding services or personnel.

Healthcare provider-oriented outcomes, including:

- Knowledge and understanding - e.g. attitudes and behaviours; levels of knowledge (such as, about CJD and prognosis, infection control and transmissibility, specialist and other care requirements, patient's or carer's wishes);
- Consultation processes - e.g. level of patient-centred care; choices offered in the provision of care;
- Support - e.g. support and/or training received in notifying patients.

Secondary outcomes

Consumer-oriented outcomes, including:

- Patient involvement in care - e.g. involvement in decision-making; decisional conflict; preferences (e.g. for information, treatments, services or support); patient-held information;
- Skills acquisition - e.g. communication skills; self-care skills; caregiving skills;
- Health behavior - e.g. attitudes towards a disease or health care; adherence to recommended care or acceptance of health care; risk-taking behaviours; use of services including screening/ diagnostic services; and
- Treatment outcomes - e.g. physiological/ clinical outcomes, follow-up, adverse events including access to medical treatment and health services.

Healthcare provider-oriented outcomes, including:

- Health service use - e.g. provider behaviour such as active and appropriate referral; co-ordination of care and follow-up; and
- Health status and wellbeing - e.g. physical or psychological health outcomes for the provider.

Health service delivery-oriented outcomes, including:

- Service delivery level - e.g. service use (such as use of specialist care, hospital admissions); coordination and continuity of care; use of care plans and/or teams; communication between sectors and/or care teams or care manager; and
- Societal or governmental - e.g. healthcare policy/legislation/procedures and revisions of these; healthcare monitoring, health care planning, procedural changes; health professional training; quality improvement strategies.

Appendix 2. CENTRAL search strategy

#1	MeSH descriptor Disclosure explode all trees
#2	MeSH descriptor Referral and Consultation explode all trees
#3	consultation:kw
#4	MeSH descriptor Counseling explode all trees
#5	MeSH descriptor Population Surveillance explode all trees
#6	“infection control”:kw
#7	disclos* or notif* or warn* or recontact* or contact* or surveillance or “bad news” or communicat* or inform* or counsel* or consult* or referral* or educat* or ((social* or psycho*) near/4 support*)
#8	(#1 OR #2 OR #3 OR #4 OR #5 OR #6 OR #7)
#9	(*cjd or creutzfel* or jakob*):ti,ab,kw
#10	MeSH descriptor Prion Diseases explode all trees
#11	((spong* or transmissible or subacute) next encephalopath*) or prion or spongiform* or “brain spongiosis” or ((tse or tses or bse) and (brain* or encephalopath*)) or heidenhain*
#12	(#9 OR #10 OR #11)
#13	(#8 AND #12)

Appendix 3. MEDLINE search strategy

1. communication/
2. exp disclosure/
3. information dissemination/
4. duty to recontact/
5. contact tracing/
6. exp population surveillance/
7. disease notification/
8. exp “referral and consultation”/
9. exp counseling/
10. social support/
11. patient education as topic/

12. health education/
13. (disclos* or notif* or recontact* or contacted or contacting or to contact or surveillance or bad news or communicat* or informed or informing or inform or ((provid* or provision or give* or giving or suppl* or convey* or present*) adj5 information) or counsel* or consult* or referral* or educat* or ((social* or psycho*) adj4 support*)).tw.
14. or/1-13
15. (cjd or vcjd or nvcd or creutzfel* or Jakob? or ((spong* or transmissible or subacute) adj encephalopath*) or prion disease* or prion disorder*).tw.
16. exp prion diseases/
17. or/15-16
18. iatrogenic disease/ or iatrogenic*.tw.
19. exp cross infection/
20. exp tissue donors/
21. exp blood/
22. exp blood transfusion/
23. (blood or plasma).tw.
24. dura mater*.mp.
25. exp equipment contamination/
26. exp postoperative complications/
27. exp surgical procedures, operative/
28. (donor* or recipient*).tw. or blood born*.mp.
29. (transplant* or graft* or operat* or surgical* or surgery or neurosurgical* or neurosurgery).tw.
30. exp disease transmission/
31. (transmission or transplantation).fs.
32. disease outbreaks/
33. risk factors/
34. or/18-33
35. (prion* or spongiform* or brain spongiosis or ((tse or tses or bse) and (brain* or encephalopath*)) or heidenhain*).tw. or exp prions/
36. 34 and 35
37. 17 or 36
38. 14 and 37
39. (animals not (humans and animals)).sh.
40. 38 not 39

Appendix 4. EMBASE search strategy

1. interpersonal communication/
2. mass communication/
3. information dissemination/
4. parental notification/
5. patient information/
6. duty to recontact/
7. contact examination/
8. disease surveillance/
9. infection control/
10. patient referral/ or consultation/
11. exp counseling/
12. social support/
13. patient education/
14. health education/
15. (disclos* or notif* or recontact* or contacted or contacting or to contact or surveillance or bad news or communicat* or informed or informing or inform or ((provid* or provision or give* or giving or suppl* or convey* or present*) adj5 information) or counsel* or consult* or referral* or educat* or ((social* or psycho*) adj4 support*)).tw.

16. or/1-15
17. (cjd or vcjd or nvcjd or creutzfel* or jakob?).tw.
18. (((spong* or transmissible or subacute) adj encephalopath*) or prion* or spongiform* or brain spongiosis or ((tse or tses or bse) and (brain* or encephalopath*)) or heidenhain*).tw.
19. exp prion disease/ or exp prion/
20. brain spongiosis/
21. or/17-20
22. 21 and 16
23. (nonhuman not (human and nonhuman)).sh.
24. 22 not 23

Appendix 5. PsycINFO search strategy

1. exp communication/
2. information/
3. information dissemination/
4. exp counseling/
5. at risk populations/
6. (disclos* or notif* or warn* or recontact* or contacted or contacting or to contact or surveillance or bad news or communicat* or informed or informing or inform or ((provid* or provision or give* or giving or suppl* or convey* or present*) adj5 information) or counsel* or consult* or referral* or educat* or ((social* or psycho*) adj4 support*)).ti,ab,hw,id.
7. or/1-6
8. (cjd or vcjd or nvcjd or creutzfel* or jakob?).ti,ab,hw,id.
9. (((spong* or transmissible or subacute) adj encephalopath*) or prion* or spongiform* or brain spongiosis or heidenhain* or ((tse or tses or bse) and (brain or encephalopath*))).ti,ab,hw,id.
10. or/8-9
11. 10 and 7
12. limit 11 to human

Appendix 6. CINAHL search strategy

S6	s5	Limiters - Exclude MEDLINE records Search modes - Boolean/Phrase
S5	s3 and s4	Search modes - Boolean/Phrase
S4	cjd or vcjd or nvcjd or creutzfel* or jakob* or (spong* N2 encephalopath*) or transmissible encephalopath* or subacute encephalopath* or spongiform* or brain spongiosis or prion* or ((tse or tses or bse) and (brain* or encephalopath*)) or heidenhain*	Search modes - Boolean/Phrase
S3	s1 or s2	Search modes - Boolean/Phrase
S2	disclos* or notif* or warn* or recontact* or contact* or surveillance or bad news or communicat* or inform* or counsel* or consult* or referral* or educat* or ((social* or psycho*) and support*)	Search modes - Boolean/Phrase

(Continued)

S1	MH communication+	Search modes - Boolean/Phrase
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Appendix 7. Current Contents search strategy

1. (disclos* or notif* or warn* or recontact* or contact* or surveillance or bad news or communicat* or inform* or counsel* or consult* or referral* or educat* or ((social* or psycho*) adj4 support*)).mp.
2. (cjd or vcjd or nvcjd or creutzfel* or jakob?).mp.
3. (((spong* or transmissible or subacute) adj encephalopath*) or prion* or spongiform* or brain spongiosis or heidenhain* or ((tse or tses or bse) and (brain or encephalopath*))).mp.
4. or/2-3
5. 4 and 1
6. (beha or clin).sb.
7. 5 and 6

Appendix 8. ProQuest Dissertations & Theses search strategy

(disclos* or notif* or warn* or recontact* or contact* or surveillance or bad news or communicat* or inform* or counsel* or consult* or referral* or educat* or ((social* or psycho*) W/4 support*)) AND (cjd or vcjd or nvcjd or creutzfel* or jakob or ((spong* or transmissible or subacute) W/1 encephalopath*) or prion* or spongiform* or brain spongiosis or ((tse or tses or bse) and (brain* or encephalopath*)) or heidenhain*)

Appendix 9. Sociological Abstracts search strategy

KW=(disclos* or notif* or warn* or recontact* or contact* or surveillance or bad news or communicat* or inform* or counsel* or consult* or referral* or educat* or ((social* or psycho*) within 4 support*)) and KW=(cjd or vcjd or nvcjd or creutzfel* or jakob or ((spong* or transmissible or subacute) within 1 encephalopath*) or prion* or spongiform* or brain spongiosis or heidenhain* or ((tse or tses or bse) and (brain or encephalopath*)))

Appendix 10. MEDLINE In-Process and Other Non-Indexed Citations search strategy

1. (disclos* or notif* or warn* or recontact* or contact* or surveillance or bad news or communicat* or inform* or counsel* or consult* or referral* or educat* or ((social* or psycho*) adj4 support*)).mp.
2. (cjd or vcjd or nvcjd or creutzfel* or jakob?).mp.
3. (((spong* or transmissible or subacute) adj encephalopath*) or prion* or spongiform* or brain spongiosis or heidenhain* or ((tse or tses or bse) and (brain or encephalopath*))).mp.
4. or/2-3
5. 4 and 1

Appendix I I. Details of websites searched

We searched websites in the period October to December 2007, with supplementary searches and follow-up of additional materials into early 2008.

Website searched	Contained within website
http://ancjdr.path.unimelb.edu.au/	CSF 14-3-3 Protein Test vCJD Testing Diagnostic Test Fees http://ancjdr.path.unimelb.edu.au/notify/ links page Fact sheets Prion Diseases - overview Sporadic Creutzfeldt-Jakob Disease Inherited Prion Disease Acquired Prion Disease: Iatrogenic Creutzfeldt-Jakob Disease http://www.hpa.org.uk/infections/topics_az/cjd/menu.htm Acquired Prion Disease: Variant Creutzfeldt-Jakob Disease Diagnosing Prion Diseases Tonsil biopsy in Variant Creutzfeldt-Jakob Disease Managing Symptoms New research findings (6 March 2003): Antibodies as potential future treatments for CJD and other prion diseases http://www.mrc.ac.uk/index.htm Infection control Part 1: The experience of dementia Part 2: Denial and challenging behaviour Part 3: Seeing, Perceiving and Believing: Elaborating Perceptual Problems in Dementia Part 4: Speech, Language and Communication Part 5: Disorders of memory Part 6: Working with People, Making Sense of Dementia Part 7: Swallowing problems - How to Help Forthcoming events Clinical trials MRC Prion-1 Study Consumer Workshop on Clinical Trials for CJD Contacts CMO referral form Download CMO referral form Prion genetics consent forms Professional guidelines Department of Health publications Guidelines for social workers At risk questionnaire Download the questionnaire
National Prion Clinic	National Prion Clinic at the National Hospital for Neurology and Neurosurgery University College London Hospitals: National Hospital for

(Continued)

	<p>Neurology & Neurosurgery (NHNN) - National Prion clinic Research into Prion diseases Research into Prion diseases Other sources of help www.doh.gov.uk/cjd/cjd`stat.htm www.open.gov.uk/doh/coin.htm www.cafamily.org.uk http://www.cafamily.org.uk/Direct/c79.html http://www.cjd.ed.ac.uk/ http://www.cjdsupport.net/ http://www.hbsef.org/ http://www.nationalprionclinic.org/ www.gig.org.uk www-micro.msb.le.ac.uk www.candid.ion.ucl.ac.uk</p>
National Creutzfeldt-Jakob Disease Surveillance Unit	<p>CJD Disease Surveillance http://www.cjd.ed.ac.uk/figures.htm http://www.cjd.ed.ac.uk/PROTOCOL.htm http://www.cjd.ed.ac.uk/referralsystem.htm http://www.cjd.ed.ac.uk/cmo.htm http://www.cjd.ed.ac.uk/reportingform.htm http://www.hpa.org.uk/infections/topics`az/cjd/inheritedCJD.pdf http://www.cjd.ed.ac.uk/report15.htm http://www.cjd.ed.ac.uk/guidance.htm http://www.cjd.ed.ac.uk/TREAT.htm Information on Variant CJD http://www.cjd.ed.ac.uk/lancet.htm http://www.cjd.ed.ac.uk/letter.htm http://www.cjd.ed.ac.uk/seac.htm http://www.cjd.ed.ac.uk/geo.html http://www.cjd.ed.ac.uk/cjdq56.pdf http://www.cjd.ed.ac.uk/vcjdworld.htm Care and support http://www.cjd.ed.ac.uk/carerep.html http://www.cjd.ed.ac.uk/care.html http://www.cjdsupport.net/ http://www.hbsef.org/ http://www.cjd.ed.ac.uk/advice.htm http://www.doh.gov.uk/scg/homecarecharges/ Practical information about CJD http://www.cjd.ed.ac.uk/CJDtype/cjdtype.htm http://www.cjd.ed.ac.uk/clinfeat.htm http://www.cjd.ed.ac.uk/investigations.htm http://www.cjd.ed.ac.uk/path.htm http://www.cjd.ed.ac.uk/glos.htm CJD Research http://www.cjd.ed.ac.uk/image.htm http://www.cjd.ed.ac.uk/unitrefs.htm Links The European and Allied Countries Collaborative Study</p>

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	<p>Group of CJD (EUROCJD/NEUROCJD). Transfusion Medicine Epidemiology Review (TMER). NEUROPRION Scottish TSE Network Transfusion Guidelines Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection. Department of Health CJD Policy Unit CJD Incidents Panel vCJD Trust The Association of British Neurologists National Prion Clinic. PRION-1 trial MRC New Therapies Scrutiny Group for Prion Disease (NTSG) Food Standards Agency The European Food Safety Authority (EFSA) BIOHAZ Panel and TSE Assessments. TSE Reagent Resource Centre. CJD Resource Centre CJD Alliance American CJD Foundation Centers for Disease Control and Prevention, Atlanta, Georgia National Prion Disease Pathology Surveillance Center USA infection control and TSEs FDA British Medical Journal. World Health Organisation Blood Recall/Withdrawal - Creutzfeldt-Jakob disease (CJD). Position statement on BSE from the Institute of Food Science & Technology.</p>
Human BSE Foundation	
www.prion.ucl.ac.uk	
www.ctu.mrc.ac.uk	PRION-1: randomised trial of quiacrine in human prion disease.
www.clinicaltrials.gov	CJD trials, communicating risk Showing Health Information Value in a Community
www.controlled-trials.com	
www.pubmedcentral.nih.gov/articlerender.fcgi?artid=59587&rendertype=abstract	

(Continued)

www.cjdsupport.net	what is CJD? CJD and Prion Disease Audio who we are get involved Fundraising common questions news and events At risk through a blood transfusion how we can help you In memoriam carers' views links Fact Sheets Variant CJD Sporadic CJD Iatrogenic CJD Familial CJD Swallowing problems Aggressive behaviour in CJD The autopsy in patients with suspected CJD vCJD and blood transfusions CJD guidelines for social workers
Official Mad Cow Disease Home Page	
Department of Health CJD Guidance	Department of Health publications Guidelines for social workers
http://www.vcjdtrust.co.uk	
The Brain & Spine Foundation	
www.alzheimers.org.uk	Information sheet 400 Information sheet 410 Information sheet 427 Information sheet 442 Information sheet 412 Information sheet 440 Information sheet 405 Services List Zoe Appleyard to launch International CJD Day
http://www.patients-association.com/	
http://www.mrc.ac.uk/index.htm	Medical Research Council - New Therapies Scrutiny Group for Prion MRC: Medical Research Council - News Memory and behavioural problems

(Continued)

	<p>MRC: Medical Research Council - News Prions block cell recycling</p> <p>MRC: Medical Research Council - News Alzheimer's prevention role ...</p> <p>MRC: Medical Research Council - Location list MRC Prion Unit</p> <p>Medical Research Council - Prion disease could be combated with ...</p> <p>MRC: Medical Research Council - News vCJD case highlights blood ...</p> <p>MRC: Medical Research Council - News Major academic/industry ...</p> <p>Medical Research Council - Document library</p> <p>MRC: Medical Research Council - People Behind Discovery Giovanna ...</p>
<p>http://www.dh.gov.uk/en/Policyandguidance/Healthandsocialcaretopics/CJD/CJDgeneralinformation/index.htm</p>	
<p>The European and Allied Countries Collaborative Study Group of CJD (EUROCCJD)</p>	<p>Objectives of the current project</p> <p>Australia</p> <p>Austria</p> <p>Canada</p> <p>France</p> <p>Germany</p> <p>Italy</p> <p>Netherlands</p> <p>Slovakia</p> <p>Spain</p> <p>Switzerland</p> <p>UK</p> <p>Advisers</p> <p>Results</p> <p>Definition of terms</p> <p>Reference list-</p> <p>Other links</p>
<p>NEUROPRION</p>	<p>http://www.neuroprion.com/en/home.html</p> <p>http://www.neuroprion.com/en/np`act`report2006.html</p> <p>http://www.neuroprion.com/en/ev`events.html</p> <p>special Prion2007 Newsletter issue</p> <p>abstract book</p> <p>http://www.neuroprion.com/en/ne`news.html</p> <p>Prion2007 orals - Synchronised video and slides ... Coming out soon!</p> <p>The Prion2007 online poster session is now opened</p> <p>Prion2007 - More than just a meeting for the scientists!</p> <p>New faculty positions available</p> <p>New Post-doc positions available</p> <p>Confronting CJD and Other Prion Disorders</p>

(Continued)

	<p>A new breakthrough in prion diseases The foundation Alliance BioSecure launches its first call focussing on prion diseases The December Issue of the NeuroPrion Newsletter is out! A special Newsletter issue, focussing on the Prion2006 conference in Torino is now available! The abstract book of the Prion2006 in electronic format WHO TSE REFERENCE MATERIALS www.who.int/bloodproducts/tse/en www.who.int/bloodproducts/tse/en -Prion2006 posters now accessible for Prion2006 participants, NeuroPrion members and associates! -Seven Prion2006 participants rewarded for their stunning posters... -Come and see the photos of the Prion2006 conference -Inovative technologies to communicate science to the public -NeuroPrion strives for a closer cooperation with patients organisations. --Alzheimer's may seed itself like mad cow disease -Prion2006 conference - Early bird registration deadline extended to the 31st of July 2006 -BSE could incubate in people 50 years or more before symptoms show -UK regulator considers action against BSE-type disease in sheep -New Mad Cow Disease Found in Texas, Alabama Cases -74th Annual General Session of the International Committee of the World Organisation for Animal Health (OIE) 21 - 26 May 2006 -1st mad cow case reported in West Austria -OIE Finally Recognises New Zealand's BSE-Free Status -Genetic finding raises fear of wider vCJD spread -The April Issue of the NeuroPrion Newsletter is out! -Estonia dismisses suspected case of mad cow disease -Transplanted Nerve Tissue Can Cause Human 'Mad Cow' Disease -A new blood-linked vCJD case found! -Post-Doc position at the Institute of Virology of the Technical University of Munich -Post Doc position available to study small ruminant diseases -PhD positions available in "Molecular and Cellular Life Sciences" -The Prion2005 abstract book is now available for download http://www.neuroprion.com/en/jb`jobs.html http://www.neuroprion.com/en/media.html http://www.neuroprion.com/en/doc`download.html</p>
World Health Organisation	http://www.who.int/topics/creutzfeldtjakob`syndrome/en/FACT SHEETS

(Continued)

	<p>Bovine spongiform encephalopathy (BSE) Variant Creutzfeldt-Jakob disease</p> <p>RELATED LINKS</p> <p>Disease outbreaks: Creutzfeldt-Jakob disease Variant Creutzfeldt-Jakob disease Impact of BSE Variant Creutzfeldt-Jakob disease Surveillance and control Information dissemination Information resources WHO fact sheet on BSE WHO fact sheet on Variant Creutzfeldt-Jakob disease Prion diseases Blood products: Transmissible spongiform encephalopathies (TSE) WHO Reference Reagents for In Vitro Assays of CJD Specimens. ECBS 2003. WHO/BS/03.1965 Rev.1 Catalogue of WHO International Reference Materials (for CJD specimens see Miscellaneous) Distribution of WHO International Reference Materials</p> <p>Related documents</p> <p>-WHO Guidelines on Tissue Infectivity Distribution in TSEs 2006 Report on International Reference Materials for Diagnosis and Study of Transmissible Spongiform Encephalopathies (TSEs). Working group fourth meeting, Geneva, Switzerland (April 2002) -Report on International Reference Materials for Diagnosis and Study of Transmissible Spongiform Encephalopathies (TSEs). Working group third meeting, Geneva, Switzerland (March 2001) -Report on International Reference Materials for Diagnosis and Study of Transmissible Spongiform Encephalopathies (TSEs). Working group second meeting, Geneva, Switzerland (May 2000) -Report on International Reference Materials for Diagnosis and Study of Transmissible Spongiform Encephalopathies (TSEs). Working group first meeting, Geneva, Switzerland (Sep 1999) -Report on WHO Consultation on Diagnostic Procedures for Transmissible Spongiform Encephalopathies (TSEs): Need for Reference Reagents and Reference Panels. Geneva, Switzerland (March 1999)</p> <p>Related links</p> <p>WHO Communicable Disease Surveillance and Response (CSR) Health Protection Agency; variant CJD and blood products UK CJD Surveillance Unit Transfusion Medicine Epidemiology Review</p>
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(Continued)

CJD Resource Centre	<p>http://www.nibsc.ac.uk/cjd/whoarewe.html http://www.nibsc.ac.uk/cjd/listofavailablesamples.html http://www.nibsc.ac.uk/cjd/requisitionforms.html http://www.nibsc.ac.uk/cjd/charges.html http://www.nibsc.ac.uk/cjd/FAQ.html http://www.nibsc.ac.uk/cjd/World.html http://www.nibsc.ac.uk/cjd/bookmarks.html http://www.nibsc.ac.uk/cjd/References.html</p>
Scottish TSE Network	<p>http://www.stn.ed.ac.uk/ http://www.stn.ed.ac.uk/public/ http://www.stn.ed.ac.uk/public/bse.html http://www.stn.ed.ac.uk/public/vcjd.html http://www.stn.ed.ac.uk/public/scrapie.html http://www.stn.ed.ac.uk/public/cwd.html http://www.stn.ed.ac.uk/public/links.html</p> <ul style="list-style-type: none">• Roslin Institute, Neuropathogenesis Unit http://www.ri.bbsrc.ac.uk/• Institute for Animal Health www.iah.bbsrc.ac.uk/• Scottish Blood Transfusion Service: www.scotblood.co.uk• National CJD Surveillance Unit: www.cjd.ed.ac.uk• Veterinary Laboratory Agency: www.defra.gov.uk/corporate/vla/• The University of Edinburgh: www.ed.ac.uk• The Roslin Institute: http://www.ri.bbsrc.ac.uk/ <p>Information on TSE</p> <ul style="list-style-type: none">• The European and Allied Countries Collaborative Study Group of CJD (EUROCJD).• Transfusion Medicine Epidemiology Review (TMER).• Transfusion Guidelines• Transmissible Spongiform Encephalopathy Agents: Safe Working and the Prevention of Infection.• Department of Health CJD Policy Unit• CJD Incidents Panel• vCJD Trust• The Association of British Neurologists• National Prion Clinic at St. Mary's Hospital in London and the MRC Prion Unit, Institute of Neurology, Queen Square, London.• PRION-1 trial• Food Standards Agency• TSE Reagent Resource Centre.• CJD Resource Centre• American CJD Foundation• USA - Centers for Disease Control and Prevention• USA - National Prion Disease Pathology Surveillance Center

(Continued)

	<ul style="list-style-type: none">● USA - CDC's website on infection control and TSEs● USA - Food and Drug Administration (FDA)● British Medical Journal.● World Health Organisation● Research Report on Transmissible Spongiform Encephalopathies● Blood Recall/Withdrawal - Creutzfeldt-Jakob disease (CJD).● The Spongiform Encephalopathy Research Campaign Funders <ul style="list-style-type: none">● the Biotechnology and Biological Sciences Research Council (BBSRC)● Department for Environment Food and Rural Affairs (DEFRA)● Medical Research Council● The Wellcome Trust
American CJD Foundation	About Us Mission Staff History Activities Conference Volunteering Contact Us About CJD Pamphlets Glossary Symptoms Fact Sheet Health Concerns Suggestions Advocacy History Current Year Writing a Letter Links Family Support Suggestions Support Groups From Our Desk Foundation Store Donations Memory Quilt Memorial Quilt Information Medical Information Medical Pamphlet Medical Education Project From Dr. Gambetti

(Continued)

	Clinical Trials End of Life Issues Handling Loss From Dr. Gambetti Legal Issues Autopsy Information Guestbook Helpline Newsletter Funeral Directors Guide Privacy Policy External Resources
British Medical Journal.	78 total articles
Blood Recall/Withdrawal - Creutzfeldt-Jakob disease (CJD).	

Appendix 12. Methods for thematic synthesis

Objectives

To evaluate systematically the documented communication needs, preferences and experiences of people at medically-acquired CJD or vCJD risk, and to identify the best ways to notify people of risk and to support them post-notification.

Methods

Criteria for considering studies for this review

Types of studies

Given the importance of this topic and the need to evaluate the available evidence, we did not exclude studies based on their design. We sought and included all relevant research studies and literature, including quantitative and qualitative research, anecdotal reports, process evaluations, consensus documents, surveys, policy documents, adverse event data and individuals' documented experiences. CJD and vCJD are rare diseases and even large notification exercises do not typically involve hundreds of people. Furthermore:

- there is not yet clear consensus on whether it is more ethically sound to inform people (or not) of their risk of exposure to the risk of CJD and vCJD.
- what is known about CJD and vCJD, and their associated risks changes over time.

We therefore expected that few, if any, controlled studies would exist to assess the range of interventions that form the focus of this review.

Types of participants

We included people of any age, gender or ethnicity at risk of CJD or vCJD acquired via any medical treatment (whether or not they were also genetically predisposed to CJD or other prion diseases); their families and/or carers, and/or the healthcare professional(s) involved in their care; see [Types of participants](#) for full details.

Types of interventions

One of the aims of this review was to identify the best ways to notify people of CJD or vCJD risk acquired through medical treatment, and to support them post-notification. We took an inclusive approach, using the term 'notify or support' broadly to mean all interventions to communicate with and notify, educate, inform, seek the participation of and support people in these situations; see [Types of interventions](#) for full details.

In this evidence synthesis we also included all studies of people's experiences of the processes of communication (notification, information, support), as well as their expressed preferences and views of such communications.

Types of outcome measures

A range of outcomes relevant to consumers, to healthcare providers and to healthcare systems may be affected by interventions to notify and support consumers following iatrogenic exposure to the risk of CJD or vCJD. There were no exclusions based on the outcomes reported by studies that were otherwise eligible for inclusion in this review, other than where the intervention targeted healthcare professionals (where at least one consumer-oriented outcome must have been reported).

A detailed list of outcomes sought for consumers, healthcare providers and health services is given in [Appendix 1](#).

Search methods for identification of studies

See [Search methods for identification of studies](#) and Appendices 2 to 11 for full details of electronic searches and other search strategies.

Data collection and analysis

Selection of studies

To identify eligible studies for this thematic synthesis, we screened citations retrieved from systematic database searches undertaken for the Cochrane systematic review.

Two authors working independently screened the titles and abstracts of studies identified from database searches against the selection criteria. We retrieved in full text any papers identified as potentially relevant by at least one author.

Two review authors then independently screened full text articles for inclusion or exclusion, with discrepancies resolved by discussion to reach consensus, and by consulting a third author where necessary. All papers excluded from the review at this stage were listed as excluded studies, with reasons provided in the [Characteristics of excluded studies](#) table. Those studies that were identified as relevant and which provided evidence about policy implementation and consumer experiences were included in the thematic synthesis.

One author (RR) screened and assessed documents identified via website searches and from contact with experts and consumer groups.

Data extraction and management

One author (RR) extracted and summarised data from included studies including the following (where available): population and setting features; study design and/or research characteristics; intervention features; outcomes assessed; and results.

We could not assess the methodological quality of the studies included in the thematic synthesis due to the diversity of included literature (study designs and types). Instead, we categorised studies based on their research design or documentation type, then grouped them as empirical and non-empirical studies. The process of data synthesis took into account whether the source was empirical or non-empirical, although this did not mean that we placed no weight on non-empirical studies, only that uncertainty might be greater with these data ([Ryan 2010](#)).

Data synthesis

This thematic synthesis includes studies of diverse methods that identify consumers' views and experiences of being notified of their at-risk status for CJD or vCJD, and their needs with respect to notification and support. See [Appendix 13](#) for studies included in the synthesis, classified by research or documentation type.

Drawing on the evidence reviewed, and on consensus views, we identified key themes and used them to organise and analyse the data ([Barnett-Page 2009](#); [Thomas 2008](#)). We also used these themes to develop a model for a comprehensive, multi-component framework for improving notification, communication with, and support for people in relation to CJD or vCJD risk acquired medically ([Ryan](#)

2010); see [Appendix 15](#). The model was also used to inform the range of possible interventions and outcomes considered by the Cochrane systematic review.

Appendix 13. Studies included in thematic synthesis - by research type

EMPIRICAL STUDY TYPES	
Survey - Evaluation of past notification strategies - with or without evaluation of preferences for future notification strategies	Carruthers 2001 [S] CJD IP 2005a [*,**] CJD IP 2005b [*,**] Freedman 1997 [S] King 1998 [S]
Evaluation/ description of past notification strategy (non-survey)	Hewitt 2006b [**]
Description of past notification strategy - selected qualitative responses to/ choice of notification strategy - with or without evaluation of preferences for future notification strategies	Lee 2004 [*] Pauls 1996 [S] Sibbald 1998 [**]
Survey - past information and/or support	CDHFS 1995 [*] CDHFS 1996 [*]
Interview	CJD SGN 2008 [*] Kirkup 2003 [*]
Group interview/ market research	Blajchman 2004 []
Issues/ complaints data	CJD SGN 2006a [*] CJD SGN 2007a [*] CJD SGN 2007b [*]
Focus group	DHAC 2001# [*]
Consensus conference/ expert meeting/ consultation with range of stakeholders/ workshop	CJD IP 2004 [*,**] DHS 2006 [*] HPA 2005 [] McCullough 2004 [] Populus 2007 [] Reesink 2003 [] Shamrock Marketing 2002 [*,**] vCJD CGAG 2007 [**] WHO 2006a [**] WHO 2006b [*,**]
Ministerial inquiry/ submissions	Allars 1994 [*] Senate 1997 [*]

(Continued)

Legal cases	Farrell 2004 [*]
Unsolicited letters - letter to the editor on past 'lookback' exercises	Callum 1999 [§]
Individual inquiry about risk	CJD IP 2006 [*,**]
NON-EMPIRICAL STUDY TYPES	
Departmental/ organisational policy papers or guidelines	CJD SGN [*] CJD IP (Framework) 2005 [*,**] CJD IP, HPA, CDO 2006 [*,**] Dolan 2006 [**] Farrugia 2005 [**] FDA 2006a [**] FDA 2006b [**,] HPA 2007 [**] HPA/HPS 2006 [*,**] NSW DoH 2004 [*,**]
Description of issues for people at risk	Anonymous 2001b [*] Anonymous 2003a [*] CJD SGN 2006b [*] Hartley 2004 [**] Sulser 2006 [**] Zekauskas 1990 [*]

Note: this study also included survey evaluating CJD Support Group Network Incorporated (CJD SGNI) role/services.

* CJD [iatrogenic source, confirmed CJD risk transmission route]

** vCJD [iatrogenic source, confirmed vCJD risk transmission route]

§ tCJD [iatrogenic sources, theoretical CJD risk transmission route]

|| vCJDfs [iatrogenic, future screening (currently 'theoretical')]

Appendix 14. Description of included studies and results of thematic synthesis

Results: Description of studies

After screening a large body of research literature and other documents, we included 49 pieces of research and documented views and experiences in this thematic synthesis.

None were rigorous, controlled quantitative studies or in-depth qualitative research; instead we included a range of studies using diverse methods. This included surveys, process evaluations, consensus documents, complaints data, individuals' accounts, policy documents and others. Approximately two thirds of the identified research and documentation was empirical research of some kind (that is, studies incorporating primary research or directly accessing the views, experiences or expertise of individuals). Empirical studies ranged from formal evaluations of notification or support strategies in people at risk or their families, to consensus conferences and workshops reporting on the views of experts or consumers or both.

Tables containing extracted, summarised data for all included studies are available on the Cochrane Consumers and Communication Review Group website, at: <http://www.latrobe.edu.au/chcp/assets/downloads/CJDDDataTables.pdf>

Although this thematic synthesis focussed on people with medically-acquired CJD or vCJD risk, several subgroups were identified in the included literature. They included:

- people at risk of CJD via confirmed routes (e.g. those people treated with contaminated hPH; or undergoing certain neurosurgical or ophthalmic procedures);
- people at risk of vCJD through confirmed routes (e.g. blood transfusion);
- people at theoretical risk (e.g. CJD transmission through blood); and
- possible candidates for future vCJD screening, subject to screening test availability.

These subgroups may have unique communication needs and preferences, however, data from all subgroups were mapped to concepts in the data and contributed collectively to the communication framework.

Our analysis and synthesis of the evidence identified ten major components of a patient-centred communication framework for communication with people at risk, comprising notification, follow-up and support. The main features and supporting evidence for each component are given in [Appendix 15](#). An overview of the major components of the communication framework, and their interconnections, is also given in the [Discussion](#) of this Cochrane review.

Appendix 15. Patient-centred communication framework

Framework component	Key activities and features
<p><i>F1: Ethical principles underpinning the proposed framework</i> Considers the ethical principles that operate in situations of risk communication of an uncertain risk for a rare and fatal disease.</p> <p>Included studies contributing evidence to this component: [Allars 1994; Anonymous 2001b; Blajchman 2004; Callum 1999; CJD IP 2005a; CJD IP (Framework) 2005; CJD IP 2006; CJD IP, HPA, CDO 2006; CJD SGN 2006a; CJD SGN 2006b; CJD SGN 2007a; CJD SGN 2007b; CJD SGN 2008; DHS 2006; Farrell 2004; Farrugia 2005; FDA 2006a; FDA 2006b; Freedman 1997; HPA 2007; HPA/HPS 2006; King 1998; Lee 2004; McCullough 2004; NSW DoH 2004; Senate 1997; Pauls 1996; Populus 2007; Reesink 2003; Shamrock Marketing 2002; Sibbald 1998; Sulser 2006; WHO 2006b; Zekauskas 1990]</p>	<p>Ethical principles operating include consideration of the need to:</p> <ul style="list-style-type: none"> • Notify people of their at-risk status - due to public health measures needed; principles of open disclosure around adverse events; and people's expressed preference to know about their medically-acquired at-risk status. • Notify people of their risk in a manner appropriate to their risk level. • Recognise explicitly the burdensome nature of risk information and the uncertainty associated with it, and the probable necessity of an ongoing and supportive relationship with people at-risk and their carers. • Protect confidentiality of people at risk, and to ensure that discrimination in accessing health care does not occur based on at-risk status.
<p><i>F2: Research findings associated with consumers' experiences - towards an assessment of harms and preferences</i> Considers the evidence on harms arising from poor communication or inadequate notification approaches, and preferences for different strategies for communicating with consumers in a notification exercise.</p> <p>Included studies contributing evidence to this component: [Allars 1994; Anonymous 2003a; Callum 1999; CJD IP (Framework) 2005; CJD IP 2005a; CJD IP 2005b; CJD IP 2006; DHS 2006; Dolan 2006; Farrell 2004; FDA 2006a; FDA 2006b; Freedman 1997; Hewitt 2006b; HPA 2005; HPA 2007; HPA/HPS 2006; King 1998; Lee 2004; NSW DoH 2004; Senate 1997; Populus 2007; Reesink 2003; Sibbald 1998; vCJD CGAG 2007]</p>	<ul style="list-style-type: none"> • Considers the experiences of people notified of their at-risk status. • Serious harms can occur through poor, inadequate or ad-hoc inadvertent notification of risk, including psychological harms, family breakdown, stress. <p>Identifies the features of notification that are <u>least</u> preferred (acceptable), including:</p> <ul style="list-style-type: none"> • If the person delivering the notification deviates from the planned timing and process for notification. • If the person delivering the notification is not known to the person at risk. • If the person delivering the notification is not knowledgeable about CJD and risk, or the person at risk has to seek information elsewhere. • If there is inadequate support or follow up post-notification. The expectation should be that additional consultation or support will be needed routinely post-

(Continued)

	<p>notification.</p> <p>Identifies those notification features that are <u>most</u> preferred and includes the following:</p> <ul style="list-style-type: none">• People feel better supported if the notification comes from a service with which they have previous experience.• Evidence on preferences is mixed for communication medium and process. Telephone call may be least preferred as a method of contact. Other research indicates that people may prefer either a consultation with their doctor (although there is also some evidence that notification via appointment or telephone call may be disadvantaging); or notification via letter plus back-up information and support.• Since evidence is mixed on people's preferences for notification, decisions about how future notifications should happen should include consultation with consumers from at-risk groups. <p>Several different notification models exist. Individually these models specify different components of the notification approach, but collectively include consideration of the following major features:</p> <ul style="list-style-type: none">• <i>Communication medium</i>: e.g. notification via letter, consultation, telephone call, combined approaches.• <i>Content of the communication</i>: e.g. reason for communication; wording of messages and language used; key messages; how to convey the risk information and associated uncertainties; supplementary information; information on support services.• <i>Timing</i>: e.g. length of appointments; timing of letters; staging of notification to people at risk, treating doctors and experts.• <i>Personnel</i>: e.g. identification of person with existing relationship with the person at risk to conduct the notification; training and support for those involved in notification/ support; designation of a coordinator of information for the person at risk. <p>For people who may <u>not yet have to be notified</u> of risk (e.g. where risk is theoretical):</p> <ul style="list-style-type: none">• People should not be notified against their will unless public health protection measures are needed.• The potential harms of notification of theoretical risk need to be acknowledged.• Where possible, people should be given individual choice about the way they are notified: a model that incorporates choice may help to minimise adverse outcomes.
<p>F3: Programmatic approach to notification: a proposed model Details the components of a comprehensive, planned and programmatic model for notification of people of at-risk for CJD or vCJD.</p> <p>Included studies contributing evidence to this component: [Allars 1994; Blajchman 2004; Carruthers 2001; CJD IP</p>	<p>Proposes a programmatic, deliberate approach to notification of at-risk status, accounting for the following issues:</p> <ul style="list-style-type: none">• The need for a national guideline for notification and public risk communication in relation to CJD or vCJD risk. This should include standardised advice and collaborative

(Continued)

(Framework) 2005; CJD IP 2004; CJD IP 2005a; CJD IP 2005b; CJD IP 2006; CJD IP, HPA, CDO 2006; DHAC 2001; DHS 2006; Farrugia 2005; FDA 2006a; FDA 2006b; Hewitt 2006b; HPA 2005; HPA 2007; HPA/HPS 2006; King 1998; Kirkup 2003; Lee 2004; McCullough 2004; NSW DoH 2004; Senate 1997; Pauls 1996; Populus 2007; Reesink 2003; Shamrock Marketing 2002; vCJD CGAG 2007; WHO 2006a; WHO 2006b]

communication processes so that when iatrogenic incidents occur people know what needs to happen; and there are clear processes in place that determine how the risk should be communicated, the content of the information conveyed, who should notify and how the notification should be performed.

- The need for notification to be planned (coordinated), deliberate, and consider impact, and to ensure that people at-risk receive the best possible information, care and support.
- The need for a supportive network of services to be in place prior to notification

Proposes a multi-component model for notification of at-risk status, accounting for the following elements:

- Communication medium for first stage of notification: people should be notified via letter from the health service or via consultation with a known clinician. A standardised approach should be used in either case (i.e. standardised wording for letter; script for consultation).
- Communication content for first stage of notification: must be clear and concise, focus on key messages, and be written in understandable language. It must also be up-to-date and should be personalised and evidence-based wherever possible.
- The initial notification should include a rationale for the notification; and should acknowledge explicitly the degree of uncertainty associated with at-risk status.
- Backup information must be provided to supplement the notification letter or consultation. This should include general information on CJD/vCJD; referral to other information sources; and referral and contact details for support services (consumer support groups, helplines, counselling services).
- People should have access to further advice and support from their GP, as needed.

The need for communication and support for the GP of person at risk (and other health professionals), including the following elements:

- The person's GP should be informed if their patient is to be notified of at-risk status. The person's GP should be notified in advance, and provided with comprehensive information.
- If the GP is involved in conducting the notification they should receive training and support to fulfil the role of the initial notification as well as a follow-up support role.
- GPs and other health professionals of the person at risk should have access to public health experts knowledgeable about CJD/ vCJD; and these experts supporting GPs in notification should also have advanced warning of the notification exercise.
- GP should record in the person's notes they are at risk for public health purposes; ensure confidentiality; and encourage the person at risk to share the responsibility for protecting the public health.

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<p>F4: Longitudinal model of support: a proposed model Considers that an ongoing relationship with people at risk is needed, articulated through a planned, supportive and co-ordinated health service model, for reasons of providing support over time and in response to changing need; and providing updated information to assist with health management and infection control.</p> <p>Included studies contributing evidence to this component: [Allars 1994; Blajchman 2004; CDHFS 1995; CDHFS 1996; CJD IP (Framework) 2005; CJD IP 2005a; CJD IP 2005b; CJD IP 2006; CJD IP; HPA, CDO 2006; CJD SGN 2006a; CJD SGN 2006b; CJD SGN 2007a; CJD SGN 2007b; CJD SGN 2008; DHAC 2001; DHS 2006; Farrugia 2005; FDA 2006a; FDA 2006b; Freedman 1997; Hewitt 2006b; HPA 2005; HPA 2007; HPA/HPS 2006; King 1998; NSW DoH 2004; Senate 1997; Reesink 2003; Shamrock Marketing 2002; Sibbald 1998; Sulser 2006; vCJD CGAG 2007; Zekauskas 1990]</p>	<p>Argues for an ongoing (longitudinal) model of communication with people at risk which includes the need for the following elements:</p> <ul style="list-style-type: none">• Appropriate and accessible support services in place prior to notifying people of their at-risk status. People need to be informed about support services available to them, and these must be accessible and appropriate, as well as responsive to changing need.• An ongoing relationship with people at risk and their families, in order to provide new or additional information and support, subject to informed consent for contact.• Responsive follow-up and support tailored to individual need and response to at-risk status, and to be adaptive over time.• Information to allow people at risk and their families to make informed decisions about health care. This information should be of high quality and reflect the current state of knowledge about CJD/ vCJD. It must also be understandable, accurate and accessible.• Professionals involved in counselling to be appropriately trained and knowledgeable about CJD and VCJD, as well as about the emotional impact for people at risk.• Ensure that people at risk are not discriminated against and that they receive the same access to health care and treatment as others in the community.• Health professional awareness of the latest CJD/ vCJD infection control guidelines and required infection control measures; for an appropriate level of response to risk by healthcare professionals; and for clear processes where people at risk are identified through screening questionnaires or self-disclosure of risk status in medical settings.
<p>F5: Education of healthcare workers Considers education for health professionals on CJD and vCJD, and the impacts of notification and infection control.</p> <p>Included studies contributing evidence to this component: [CJD IP 2005b; CJD IP; HPA, CDO 2006; CJD SGN 2006a; CJD SGN 2006b; CJD SGN 2007a; CJD SGN 2007b; CJD SGN 2008; DHAC 2001; Farrugia 2005; FDA 2006a; Hewitt 2006b; HPA 2005; NSW DoH 2004; Sibbald 1998; vCJD CGAG 2007]</p>	<p>Considers the supporting role for health professional education in effective communication with people at-risk, including the need for:</p> <ul style="list-style-type: none">• Education about CJD and vCJD, risk and infection control guidelines.• Training in communicating with people at risk, and awareness of the potential negative impact of poor notification or communication strategies.• Access to comprehensive, up-to-date information about CJD or vCJD.• Access to expert advice and support from specialists.
<p>F6: Consumer group Considers the key role played by consumer groups in supporting people at risk and their families.</p> <p>Included studies contributing evidence to this component: [DHAC 2001; Senate 1997]</p>	<p>Outlines the role and functions of consumer support groups, including the following:</p> <ul style="list-style-type: none">• Because people at risk may seek information from consumer support groups following notification, such groups must be informed, in advance, when incidents occur in order to respond effectively to contacts from people at risk or their families.

(Continued)

	<ul style="list-style-type: none"> • The need for consumer groups to raise awareness with at-risk people, of the group's activities and functions. • The need for consumer groups to provide relevant, up-to-date information, and to attempt to respond to the identified needs of people at risk and their families.
<p>F7: Evaluation, improvement and research priorities Documents the substantial research gaps identified by the review, and proposes a research agenda, focusing particularly on the ongoing impact of being notified of CJD or vCJD risk status.</p> <p>Included studies contributing evidence to this component: [Allars 1994; Callum 1999; Carruthers 2001; CDHFS 1995; CDHFS 1996; CJD IP 2005b; CJD IP 2006; DHS 2006; Freedman 1997; Hewitt 2006b; HPA 2005; King 1998; Lee 2004; McCullough 2004; Reesink 2003; Sulser 2006; vCJD CGAG 2007]</p>	<p>Considers the research gaps that exist on communicating with people at risk and proposes research to inform future communication strategies.</p> <p>This includes the following major issues:</p> <ul style="list-style-type: none"> • The ongoing social and emotional impacts of notification for people at risk and their families. • What assists people to deal effectively with knowledge of their at-risk status? What ongoing needs do people have regarding support and follow-up? <ul style="list-style-type: none"> • How can notification be improved - e.g. to minimise avoidable psychological and other harms? How could people experiencing unavoidable harms be best cared for? • Which risk communication formats are best to accurately convey the risk of CJD or vCJD, as well as uncertainty?
<p>F8: National and jurisdictional issues Considers national issues such as the need for standardised communication strategies around CJD or vCJD and risk.</p> <p>Included studies contributing evidence to this component: [CJD IP, HPA, CDO 2006; HPA 2007; HPA/HPS 2006; DHS 2006; Farrugia 2005; Lee 2004; Senate 1997; Reesink 2003; vCJD CGAG 2007; Zekauskas 1990]</p>	<p>Considers the need for standardised approaches to communication, including the need for:</p> <ul style="list-style-type: none"> • Standardised and consistent approaches to notification, so that when iatrogenic incidents occur the process is clear, consistent and people know what needs to happen. • Standardised and consistent approaches to public risk communication around iatrogenic incidents. • Updating individuals and the public as new evidence emerges or public health protection measures change. • Periodic review to ensure that people at risk of CJD or vCJD are not discriminated against based on their at-risk status.
<p>F9: Public communication Considers strategies to improve public understanding of CJD and vCJD and features of communicating with the public about specific incidents.</p> <p>Included studies contributing evidence to this component: [CJD IP (Framework) 2005; DHS 2006; HPA 2005; Lee 2004; Pauls 1996; Shamrock Marketing 2002; WHO 2006a; WHO 2006b]</p>	<p>Outlines the supporting role for public education and communication about CJD and vCJD, including the need for:</p> <ul style="list-style-type: none"> • Community education about CJD and vCJD, areas of uncertainty, healthcare-associated risk and measures taken to minimise risk in healthcare settings. • Standardised approaches to public communication around specific incidents, which includes provision of general and incident-specific information; and referral to further information and support (e.g. helplines), including information about how to contact consumer support groups.
<p>F10: Media issues Proposes a proactive approach to communication with the media, given the potential for significant harm if people inadvertently learn of their risk through this route rather than from health professionals or services.</p>	<p>Outlines a proactive approach to communicating with the media, which includes the following:</p> <ul style="list-style-type: none"> • The need to notify people of at-risk status directly and ahead of releasing information to the media, to avoid potential significant psychological harm that may occur:

(Continued)

Included studies contributing evidence to this component:
[Allars 1994; Blajchman 2004; CJD SGN 2006a; CJD SGN 2006b; CJD SGN 2007a; CJD SGN 2007b; FDA 2006a; HPA 2005; Lee 2004; Kirkup 2003; Senate 1997; Shamrock Marketing 2002]

- If people learn about their at-risk status through a third party, such as the media.
- If new cases or probable deaths due to CJD or vCJD arise among people in similar situations (e.g. exposed to risk in a similar way or cohort) and people are informed through a third party, such as the media.
 - The need to routinely brief the media to keep them informed of developments in the area and the issues around notification, including the ethical and practical issues related to communicating with people at risk.
 - The need to avoid the perception in the media that iatrogenically-acquired CJD or vCJD risk are the result of negligence or failure to implement proper procedures.

Appendix 16. Limitations and commentary on the thematic synthesis

This thematic synthesis has assembled and synthesised all available evidence on people's experiences of notification and how best to notify and support people at risk. We have included a large number of individual pieces of diverse types of research and literature relevant to this area; an approach which has both strengths and limitations.

State of the evidence

It is important to acknowledge that this thematic synthesis represents only a snapshot in time of the available evidence on CJD and vCJD and associated uncertainties. As knowledge about CJD and vCJD changes, or areas of uncertainty are resolved, some of the communication issues identified in this review may become more or less significant. It is therefore important that the research evidence on communication is maintained and aligned with the most recent knowledge about these diseases.

Searching for and locating the evidence

We searched exhaustively to locate all relevant research. This included comprehensive medical database searches as well as website searches and contact with experts for further information about relevant research. We searched for both published and unpublished research.

Eligible research was highly diverse, however, and much was unpublished, fragmentary or difficult to locate. For example, research in this area was often scattered across health agency and government websites and so was difficult to trace and identify systematically. It is therefore possible that we missed key documents. There was also a large volume of research and literature on related topics, including infection control issues and guidelines; pathophysiology of the prion diseases; and information on the ethical implications of notification of risk. We sorted systematically through this literature; however it is possible that we missed key pieces of information embedded within the literature on these related topics.

This area is a sensitive one and communication of risk and issues of patient confidentiality must be dealt with accordingly. In some cases this has contributed to the difficulty in locating relevant research. For example, we know of at least one incident occurring in Australia recently for which we have been unable to locate any documentation on the notification process, and so it has not been included in this review. There may also be other similar examples involving other incidents of which we are not aware.

A number of papers in languages other than English have also been identified and are awaiting translation and assessment (see [Characteristics of studies awaiting classification](#)) and it is possible that some of these studies may in fact be eligible for inclusion in this synthesis. We also know of one qualitative study on people with vCJD, which seems likely to be relevant to this thematic synthesis, but for which we have not been able to obtain data or further information from the authors.

Review methods

A single author undertook much of the work in completing this thematic synthesis and this may introduce bias or errors. However, study selection was done by two authors, as was thematic data analysis, increasing our confidence in this step of the analysis that was crucial for synthesis and interpretation of the data. Having input from more than one researcher at key steps in the analysis of data represents a strength of this thematic synthesis.

This thematic synthesis represents a systematic review of studies of diverse methods, and so sits between quantitative and qualitative research methods. We were unable to identify any guidance in this area, in particular to guide our decisions around including more than formal research, such as including documented complaints as a source of data.

The rationale for our decision to take an inclusive approach to eligible research and literature was based on the choice between virtually no data (quantitative and qualitative) being available and a larger pool of data from multiple sources. Given the need to inform decision making with available evidence, we chose the latter approach.

We also considered as a valid source of data that collected periodically by consumer groups (Baggott 2005; Ryan 2010), for which there are no agreed criteria to appraise methodological quality. We decided to itemise included studies or reports as empirical or not, to make the distinction between formal research and processes whereby documented views were gathered and synthesised (e.g. consensus approaches).

Our approach in this thematic synthesis therefore reflects a deliberate shift away from focusing on methodological quality as the standard for inclusion in systematic reviews to an approach mediated by the context and criteria of high public and professional interest, little available guidance, and best available evidence. While there are methodological shortcomings in the included research, and limitations of the review itself, we have adopted transparent methods of study selection, data extraction and synthesis designed to minimise bias in the review's conclusions. The various components of this framework, which have been explicitly constructed, could be rigorously evaluated in trials or other research, although we believe that a more pressing need is to fill gaps in the assembled research identified by the framework.

Implicitly we have recognised within this thematic synthesis that different sorts of research are best suited to different purposes. The purpose of this synthesis and framework developed from the evidence is not to estimate quantitative effect sizes of outcomes but rather to propose a framework and specific communication models, indicated where possible by data from diverse sources, within which notification of risk and support might best occur. These models of communication might be used by health services and decision makers to develop protocols for notification and support for people at CJD or vCJD risk appropriately translated to local or regional settings.

The identified research and literature

Although we included a large number of studies and documents in this synthesis, not all was empirical (that is, based on primary research data, or accessing the views, experiences or expertise of individuals). Approximately one third of included studies were classified as non-empirical, and the results of these studies should be interpreted with particular caution as they may be limited in applicability to real populations and settings.

One strength of this thematic synthesis is the diversity of included research and literature, including complaints data, government and health agency documents and policies, formal evaluations and surveys of notification and/or support exercises. Including these varied types of research has provided a rich source of data. However, we did not identify any rigorous quantitative or in-depth qualitative research to include in this synthesis. This is a limitation of the existing research and represents a major gap in the evidence.

The quality of studies included in this review was variable: because a wide range of research was identified and included, which employed a range of disparate study designs, we did not attempt to evaluate formally and systematically the quality of included studies, but took the research at face value. This is a limitation of the synthesis.

We included numerous individual pieces of research and literature in this thematic synthesis, however the assembled evidence is quite fragmentary and in some areas sparse. This limits conclusions based on the evidence. For example, we identified relatively few papers that addressed the questions of how best to support people at risk of CJD/ vCJD in an ongoing way after notification has happened. Similarly, there was little information available on the impacts of notification; and none on risk communication formats for CJD risk. Even in those areas of this synthesis where more data were available, for example, related to the notification process itself, the evidence was generally mixed and did not give definitive answers. In part, these difficulties reflect those inherent in conducting research in this area. However, it also highlights the need for further high quality research on several important questions arising from this thematic synthesis and Cochrane review.

Gaps in the evidence

Although we included a large number of pieces of research and literature in this review the assembled evidence is sparse in some areas, which limits the conclusions that can be drawn. The research gaps documented by this synthesis are numerous and highlight a need for further high-quality research. This includes qualitative research to better understand the impacts of notification, as well as research on the effectiveness of the full range of communication interventions available, which is currently lacking.

See the [Discussion](#) section of the Cochrane review for a detailed discussion of research gaps and implications, and Additional [Table 3](#) for recommendations for future research arising from the Cochrane review and thematic synthesis.

This synthesis cannot produce evidence where none exists, and so is limited in scope and applicability by the research that has preceded it. These gaps are explicitly highlighted as priorities for future research, and we hope that this will encourage policy makers and researchers to prioritise this important research, particularly as people will continue to be exposed to the risk of CJD and vCJD through health care, and the best ways to notify and support these people is therefore an ongoing concern.

HISTORY

Protocol first published: Issue 1, 2009

Review first published: Issue 3, 2011

CONTRIBUTIONS OF AUTHORS

RR: developed the review scope and protocol; searched for papers and contacted experts; extracted data, assessed papers, performed analysis for the thematic synthesis and development of the framework; and wrote the review and thematic synthesis.

SH: initiated the review and developed the scope and protocol; assisted with analysis for the thematic synthesis and development of the framework; contributed to writing the review and thematic synthesis.

DL: assisted with locating and screening relevant papers; contributed to protocol drafts; assisted with data checking and contributed to the review.

KA: assisted with methodological advice; assisted with screening papers; and contributed to protocol and review drafts.

MT: assisted with screening papers and contributed to protocol and review drafts.

CM: assisted with screening papers and contributed to protocol and review drafts.

DECLARATIONS OF INTEREST

None known.

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INDEX TERMS

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*Disease Notification; *Iatrogenic Disease [epidemiology]; Creutzfeldt-Jakob Syndrome [epidemiology; *transmission]; Prion Diseases [epidemiology; transmission]

MeSH check words

Humans