CJD and Prion Disease: Fact Sheet for Local Professionals

Introduction

CJD is one of a group of diseases called prion diseases, it affects 1 – 2 people per million of the population per year. The lifetime risk of CJD is around 1 in 5,000. CJD is a fatal neurodegenerative disease, and there no current treatments. Proactive symptom management and anticipatory care planning are extremely important. This fact sheet gives some information about CJD, care and infection control. The role of specialist services and possible local referrals are outlined here. Finally, information is provided on where local clinicians can get further information and support.

*"**We chose as a family to care for Mum at home**, our journey was as unique as our Mum and spanned 5 months. Within this time, we experienced positive and negative aspects of care, but focused on the **positive interactions** as these **resulted in positive interventions**.*

*Each positive interaction was based on high quality communication: the **professionals listened to us**, considered the cause, **discussed with colleagues** or other healthcare professionals, and **guided on the intervention**. This made the hardest of journeys, more positive for Mum and so for us."*

***Libby** (mum passed away with sCJD in 2023)*

There are four types of CJD

Sporadic CJD (sCJD) is the most common form, accounting for around 85 per cent of cases. The cause is unknown. It mainly affects the over 50s and has a rapid onset. The course of the disease is usually measured in months.

Inherited Prion Disease (eg familial or genetic CJD) occurs when there is a mutation in the prion protein gene, there are various different mutations which are known to be pathogenic. The symptoms experienced, age of onset and time course of disease can differ significantly depending on the type of mutation. Inherited prion disease is known as an autosomal dominant trait, meaning that children of an affected parent are at 50% risk of inheriting the mutation. Families are often aware of the condition as they may have seen it in their wider family.

Iatrogenic CJD (iCJD) occurs through contaminated medical procedures, for example, the use of now obsolete forms of human growth hormone, blood transfusion, or surgical instruments

Variant CJD (vCJD) is caused by exposure in the diet to the cattle prion disease, bovine spongiform encephalopathy, or BSE, and typically affects younger people. It has a relatively long time course – an average of 14 months from onset of symptoms to death. There is a combination of psychiatric, neurological, and physical symptoms.

Symptoms of CJD

Symptoms reflect brain dysfunction and certain features are common in all types: memory problems, confusion, clumsiness, unsteadiness, and jerky movements with progressive disability leading to loss of awareness, loss of mobility, loss of speech, incontinence, and the need for full nursing care. The precise symptoms and clinical time course of CJD vary according to the type of disease and to the individual. Unexpected symptoms may occur at unexpected timepoints. It is important that clinicians listen with openness and curiosity to what is being described by patients and their loved ones, and work to actively support them in what can be a rapidly changing situation.

Caring for a patient with CJD

Local teams already have the skills and experience necessary to care for patients with CJD. Anticipatory care planning and ensuring things are put in place promptly are important, as the disease can progress rapidly, and 'thinking outside of norms', as symptoms can occur and develop unexpectedly. Not all patients will follow a set path and, as multiple brain areas can be affected at once, there may be collections of symptoms which may not usually be seen. Communicate community referrals made to patients and their loved ones, informing them of the possibilities of other specialist services contacting them. For families wishing to care for their loved one at home, inform them of local services available to aid in the care management of the patient at appropriate time of condition progression.

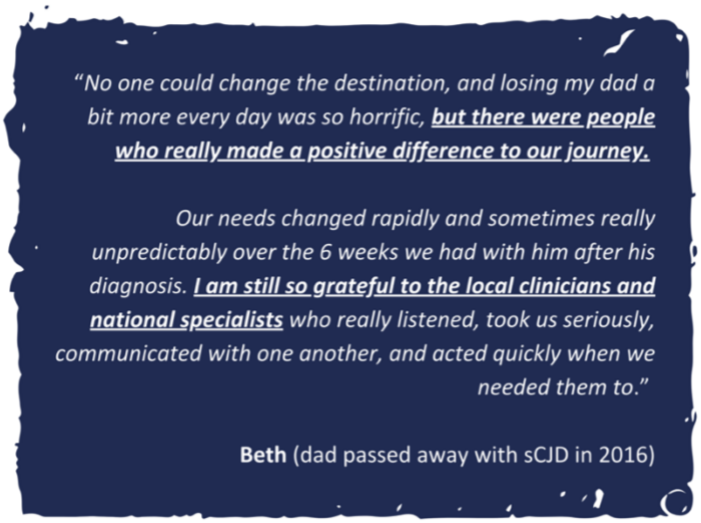
Infection control

There are no specific infection control precautions needed when caring for an individual with CJD, there is no risk associated with close physical contact and standard precautions should be taken for patient care. CJD is not infectious but is transmissible under certain circumstances such as brain surgery. The Department of Health and Social Care have published guidelines here. As a precaution, those with a diagnosis, or who are at risk, of CJD should not donate blood or organs.

CJD and Prion Disease: Fact Sheet for Local Professionals

Specialist Services

The Chief Medical Officer has requested that individuals suspected of having CJD should be referred to the National CJD Research & Surveillance Unit (NCJDRSU) in Edinburgh and the National Prion Clinic (NPC) in London who are able to contribute to the diagnosis with specialist tests.



If testing supports a diagnosis of CJD, or an individual is at risk of developing CJD, clinicians from the NCJDRSU and NPC are available to provide specialist advice, support and ongoing assessments alongside research with patient and family consent. The NCJDRSU and NPC will be in contact with local care teams. There are dedicated nursing services based in the NPC and in Edinburgh thatcan support patients, families, and local teams with specialist advice regarding symptom management, assessments (virtual and face-to-face), guidance on referrals/ referral completion, teaching sessions, resources and accessing additional funds via the CJD Care Fund The nursing services regularly meet to co-ordinate care and the support offered. The NPC also have a prion neuropsychologist who can offer psychological support to the patient and family.

Specialist services are aware that services differ throughout the UK and aim to support local teams to care for CJD patients by highlighting the likely progression of the disease and common symptoms, advising on which healthcare specialist maybe instrumental throughout the patient journey. The GP is ordinarily requested as the main contact for routine issues such as unexpected symptom management, and may also support with referrals to, for example, Palliative Care, District Nursing and other services.

Local Referrals

District Nursing – Referral should be made for continence and medication management. District nurses can also be helpful for assessments for Continuing Health Care (CHC) funding, as they often have local knowledge and connections with CHC, as well as experience with local processes.

Palliative Care – Referral to palliative care should be offered and encouraged as soon as possible. Early palliative care involvement can be helpful for discussions around the patient's wishes at end-of-life (EOL). Advanced care planning can ensure individuals wishes are expressed and documented, guiding families as the disease progresses and the individual is less able to communicate their wishes. Palliative care may be able to support with medication management of difficult symptoms, hospice at home, occasional overnight care, equipment, as well as possible peer and emotional support for carers, working alongside District Nursing to manage EOL care and supporting EOL admission to Hospice. While this is not always possible due to the rapidity of CJD in some cases, palliative care are experts in supporting families through EOL care and can usually offer bereavement support to families following loss, so referral is always recommended.

Occupational Therapy – Routine referral can facilitate access equipment such as grab rails, raised toilets, and adapted cutlery, hospital bed etc. . Referral to occupational therapy is most suited to those earlier in their disease and those families that wish to care for their loved ones at home.

Physiotherapy – Referral can be helpful for developing repositioning strategies for advance cases of sCJD and inherited prion disease where dystonia can make repositioning for personal care distressing for both patients and carers. Particularly for inherited prion disease, physiotherapy can be helpful for general wellbeing in the early stages of disease.

Speech and Language Therapy – Referral can be made to assist with communication issues in the earlier stages of the disease and swallowing issues in the later stages. CJD patients often need modified diet and advice on feeding strategies due to apraxia of swallow and associated risks.

CJD and Prion Disease: Fact Sheet for Local Professionals

Further information and support

The below specialist services and support organisations welcome contact from local clinicians, patients and families:

National CJD Research and Surveillance Unit

Western General Hospital, Crewe Road, Edinburgh EH4 2XU
Website - www.cjd.ed.ac.uk
0131 537 1980 telephone number for general enquiries and
loth.securecjd@nhslothian.scot.nhs.uk for Care Team enquiries/patient families

National Prion Clinic

National Prion Clinic, Institute of Prion Diseases, Courtauld Building, 33 Cleveland Street, London, W1W 7FF.
Website – www.nationalprionclinic.org
Helpline for National Prion Clinic - 020 7679 5142 / 020 7679 5036
uclh.prion.help@nhs.net

CJD Support Network

The CJD Support Network is a peer support charity offering information, support and connecting those with shared experiences.

Amin and general enquiries - admin@cjdsupport.net or +44 (0)7494 211476
Support - support@cjdsupport.net or 0800 774 7317
Website - www.cjdsupport.net
Post - PO Box 3936, Chester, CH1 9NG
Registered charity no. 1097173

Acknowledgements

The CJD Support Network gratefully acknowledges the assistance of Terri Awe, of the National CJD Research and Surveillance Unit, Rachel Williams, of the UK National Prion Clinic, and Libby Critchley, renal nurse specialist whose mum passed away with sCJD in 2023, in the preparation of this fact sheet.