

Update on PRN100: An innovative treatment for CJD

What is sporadic Creutzfeldt-Jakob disease (CJD)?

Sporadic Creutzfeldt-Jakob disease (CJD) is caused when healthy proteins which exist normally in the human body become spontaneously misshapen and build up in the brain.

These misshapen proteins, which are called prions, stick to other healthy proteins causing them to become misshapen too and the disease spreads through the brain.

Our immune system produces antibodies to fight infections which invade the body. However, as abnormal prions are made of one of the body's own proteins, our immune system does not make antibodies to fight them.

What is the PRN100 treatment?

Researchers at the Medical Research Council (MRC) Prion Unit at University College London (UCL) have developed a drug, called PRN100, for the potential treatment of Creutzfeldt-Jakob disease (CJD).

PRN100 is an artificially manufactured antibody which has been designed to bind tightly to normal proteins in the brain. The aim is to prevent abnormal prions from being able to attach themselves to healthy proteins, meaning that they cannot grow and cause devastation throughout the brain.

Have patients received the treatment?

In October 2018, a patient of University College London Hospitals (UCLH) became the first person in the world to receive the PRN100 treatment.

Since then, a further five patients of UCLH have also received the antibody. All six patients were at different stages of their disease when they began treatment. Three of the patients were able to consent to receiving the antibody for themselves. The other three patients did not have the capacity to consent, so, with the support of their families, UCLH sought the opinion of a judge in the Court of Protection. In each case, the judge confirmed that it was lawful and in the patient's best interests to receive the drug.

CJD is an invariably, and usually rapidly, fatal disease. Four of the six patients UCLH has treated with the drug have sadly now died as a result of their condition.

None of the six patients experienced side effects as a result of receiving the treatment.

Did the treatment have an impact?

We are currently looking at the clinical data and information we have gathered to determine if, or to what extent, the treatment has had an impact on the patients' conditions. We aim to report initial findings next year.

Will the treatment be available to more patients?

We are reaching the end of our supply of PRN100 and therefore will not be treating further patients with the drug at this time. We aim to report initial findings next year and will then consider next steps.

Where can I find out more information?

[October 2018 UCLH media release](#)

[December 2018 UCLH media release](#)

[January 2019 UCLH media release](#)

[October 2019 UCLH media release](#)

[UCLH Q&A](#)

[Cure CJD Campaign video](#)

You can also contact the National Prion Clinic (NPC) team on 020 7679 5142 or email uclh.prion.help@nhs.net