Australian National Creutzfeldt-Jakob Disease Registry

https://www.neurodegenerationresearch.eu/survey/australian-national-creutzfeldt-jakob-disease-registry/

Title of the register

Australian National Creutzfeldt-Jakob Disease Registry

Name of Principal Investigator - Title

Prof

Name of Principal Investigator - First name

Steven

Name of Principal Investigator - Last name

Collins

Address of institution -Institution

The Florey Institute

Address of institution - Street address

30 Royal Pde

Address of institution - City

Parkville

Address of institution - Postcode

3052

Country

Australia

Website

http://ancjdr.path.unimelb.edu.au

Contact email

Q1a. Please indicate below if your cohort includes or expects to include, incidence of the following conditions?

Prion disease

Q2. In a single sentence, what is the stated aim of your register?

The detection and surveillance of Cruetzfeldt-Jakob diseaseand associated prion diseases in Australia.

Q2b. What distinguishes this register from other disease registers?

The Registry is unusal as it is tasked with the public health surveillance of prion diseases for local and the Commonwealth Health Departments. The Registry provides specialist advice and diagnostic services, similar to other prion disease units oversease. As a specialist reference centre, the ANCJDR manages the clinical case evaluation and disease classification of individuals prior to cases inclusion in incidence and epidemiology reports. Classification uses WHO surveillance clinical criteria.. The Registry is also unusual as it involves clinically suspect notifications under noifiable diseases, while patients are usually alive and the illness isnt always clearly characerised due to the complexity of diagnosis and the lack of a single specific ante-mortem diagnostic test. Cases are routinely evaluated after death by the ANCJDR, relying either on pathological autopsy findings or ANCJDR evaluation of clinical details and test findings. Medico-demographic information is accessed from medical records and surviving family mmebrs assisting teh ANCJDR.

Q3a. i) Number of publications that involve use of your register to date

>15

Q3a. ii) Please give up to three examples of studies to date (PI, Institution, Title of Study)

Collins S. Creutzfeldt-Jakob disease surveillance in Australia: update to December 2013. Commun Dis Intell Q Rep. 2014 Dec 31;38(4):E348-355.| Collins S. Enhanced geographically restricted surveillance simulates /Masters CL. Creutzfeldt-Jakob disease cluster in an Australian rural city. Annals of Neurology. 52(1):115-118, 2002 Jul. sporadic Creutzfeldt-Jakob disease cluster. Brain. 2009 Feb; 132(Pt 2):493-501. |

Q3b. If data on research outputs are already available please paste the publication list/other data or provide a link to where these data are publicly available?

CDI publications - http://www.health.gov.au/internet/main/publishing.nsf/content/cda-cjdanrep.htm and an extensive list of collaborative peer reviewed publications.

Q3c. If no research has been done as yet, please explain in a few sentences what information (i.e. research findings) you expect will be gained from the register

n/a

Q4a. Study criteria: what is the age range of participants? Age in years: from

1

Q4a. Study criteria: what is the age range of participants? Age in years: to

until death

Q4b. Study criteria: what are the inclusion criteria?

The clinical suspicion of prion disease

Q4c. Study criteria: what are the exclusion criteria?

Insufficient cliinical evidence to support a CJD diagnosis, the diagnosis of an alternative illness and exclude CJD, or alternatively pathology proven not CJD after brain autopsy.

Q5. What is the size of the register (i.e. how many patients have been enrolled)?

1001-5000 clinical cases

Q6a. Please describe what measures are used to characterise participants

Clinical review of the illness raising concern for prion diseases, accepted as statistical case hen case criteria meets WHO surveillance criteria for CJD and associated prion diseases.

Q6b. Are there defined primary and secondary endpoints (e.g. defined health parameters)?

No

If YES, please describe
Q7a. i) Is the register of fixed duration?

Yes

Q7a. ii) Please enter the data collection start date

01/01/1970

Q7a. iii) Please enter the data collection end date

Nil

Q7b. Could you provide some information about the data collection for this register?

Data collection ongoing|Data analysis ongoing

Q8. Funding of the register - How is the register funded?

Funded by the Commonwealth Health Dept.

Q8. Funding of the register - Is this funding expected to continue

Yes

Q8. Funding of the register - If so, for how long (months)?

Current contract to mid 2017, funding may continue.

Q9. Could you provide information about data sweeping? - How many data sweeps have taken place?

0

Q9. Could you provide information about data sweeping? - When was the most recent data sweep?

n/a

Q9. Could you provide information about data sweeping? - When is the next data sweep?
Q9. Could you provide information about data sweeping? - How many more data sweeps are planned on current funding? e.g 0,1,2…..

0

Q9. Could you provide information about data sweeping? -How many more data sweeps are planned in total (with funding and with funding yet to be secured) e.g. 0,1,2…

0

Q10. Is the clinical (phenotypic) information that is held in the register from patients and other participants such as family members:

Only collected through the study

Q11. Is there a limit on the number of studies that can be based on this set of patients?

No

If YES, please give details
Q12a. Please give information on the format and availability of data stored in a database (1)

Data summarised in database

% Available
Q12a. Please give information on the format and availability of data stored in a database (2)

No

% Available

Q12a. Please give information on the format and availability of data stored in a database (3)

No

% Available
Q12a. Please give information on the format and availability of data stored in a database (4)

No

% Available
Q12a. Please give information on the format and availability of data stored in a database (5)

No

% Available
Please specify language used

English

Q12b. Please give information on how data is held as individual records (1)

Data is held as individual records

% Available
Q12b. Please give information on how data is held as individual records (2)

No

% Available
Q12b. Please give information on how data is held as individual records (3)

Data held on computer based records

% Available
Q12b. Please give information on how data is held as individual records (4)

Data held on cards

% Available
Please specify language used

English

Q13a. Is data available to other groups?

No

Q13b. If data is available to other groups what is the access policy/mechanisms for access?

Apply to PI or co-ordinator at resource

Q14. What data sharing policy is specified as a condition of use?

Data made publicly available after a specified time point

Q15a. Are tissues/samples/DNA available to other groups?

Yes

Q15b. i) If yes, please describe below:

Post-mortem donors: brain

Q15b. ii) In what form are tissues/samples/DNA supplied?

Primary Samples: Stabilised samples (frozen or fixed)

Q15b. iii) Is the access policy/mechanism for obtaining samples the same as that for obtaining data (Q13b above)?

No

Q16a. Is information on biological characteristics available to other groups?

If available for a subset please specify number of patients and % of total cohort

Number of patients
% of total cohort
Q16b. If yes, is the access policy/mechanism for obtaining samples the same as that for obtaining data (Q13b above)?

Yes

Types:
Disease Registers

Member States:
Australia

Diseases:
Prion disease

Years:
2016

Database Categories:
N/A

Database Tags:
N/A