



Public Health
England

Protecting and improving the nation's health

PHE update

Estimating national Wilson's disease
prevalence rates:

Exploring the utility of clinical and routinely
collected data

**BASL Wilson's Disease Special Interest Group Meeting
14th June 2018**

PHE NCARDRS

The National Congenital Anomaly and Rare Disease Registration Service
(NCARDRS)

Project overview

- Collaboration with PHE NCARDRS, WDSIG, BASL and BSG
- Proof of concept
- Aims
 - Provide a national estimate of the prevalence of Wilson's Disease.
 - Identify people in England diagnosed with Wilson's Disease for inclusion on the NCARDRS database (CARA)
 - Assess the utility of data already collected routinely (i.e Hospital Episode Statistics) to identify cases of Wilson's Disease
 - Identify systems by which we can establish effective, prospective reporting of rare disease, including Wilson's Disease
 - Support the work of the Wilson's Disease Special Interest Group (WDSIG)

Progress update

- National prescribing data
- HES data
- ONS mortality data
- Transplant data

National prescribing data

Primary care data

Trial of prescriptions data matching

- 48 patient details sent for prescriptions matching
- 7 months of data available for matching (April – October 2015)
- 36 cases matched

Of these:

9 were prescribed Wilzin and/or Trientine (Wilson specific drugs)

18 were prescribed zinc sulfate, penicillamine or/and distamine

9 were not prescribed any drugs to indicate the presence of Wilson's disease

HES data

The number of patients with episodes coded to E83.0 for 2011/12-2015/6 by hospital trust. Number of patients: 863

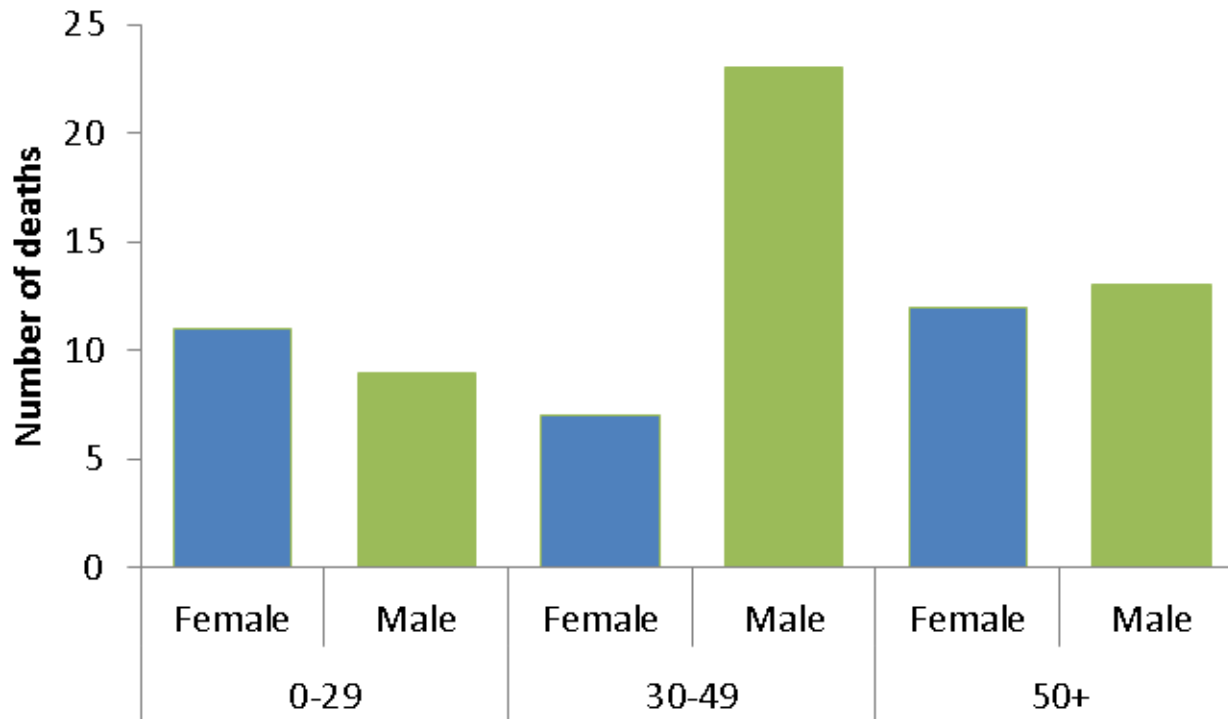
Number of trusts	Number of patients coded to ICD10 E830
117	<10
23	10-19
4	20-25
1	80
1	174

ONS mortality data

- Death in years 2001-2015
- E83.0 is cited in any category on the death certificate
- Refined using text as written on death certificate
(Wilson's Disease or Hepatolenticular Degeneration)

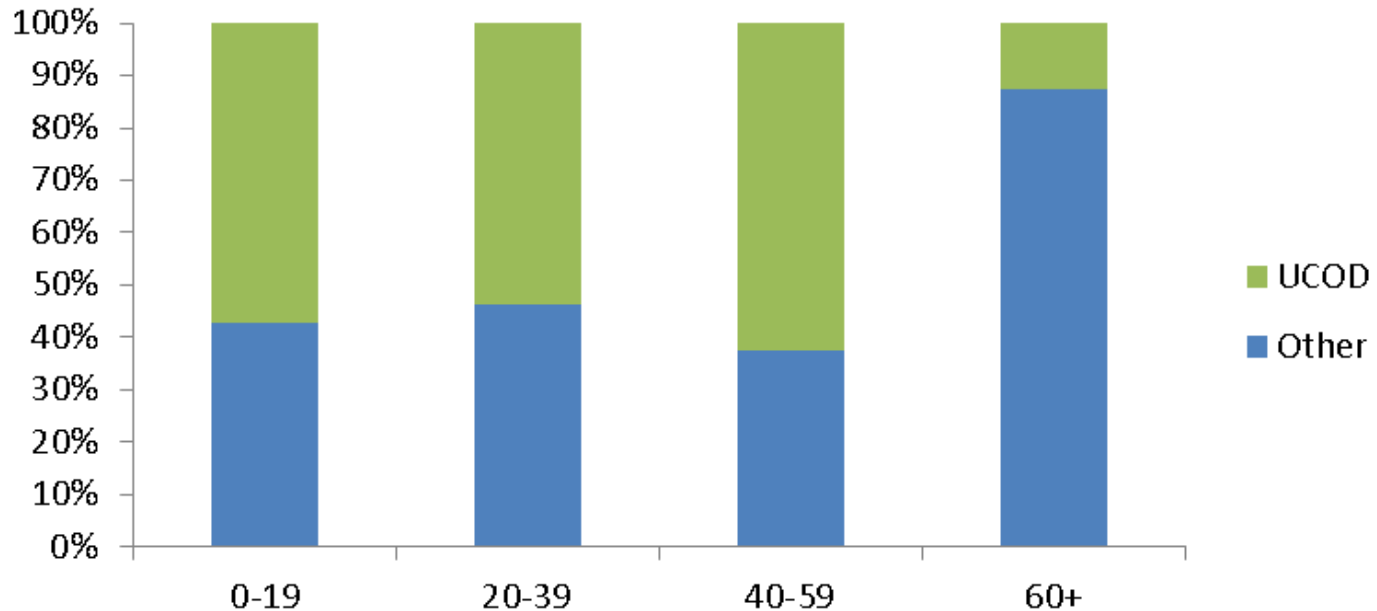
ONS mortality data (provisional)

Number of deaths certificates with mention of Wilson's Disease by sex & age (2001-2015)



ONS mortality data (provisional)

Placement of Wilson's Disease on death certificates by age (2001-2015)



Transplant data

NHS Blood and Transplant data

The number of liver transplants for patients resident in England, with Wilson's disease reported at either transplant or registration, 01/01/2007 – 31/12/2017

	No. of transplants	Notes
No of liver transplants performed 2007 - 2017	56	
Indication of Wilsons at both registration & transplant	46	
Indication of Wilsons at registration only	3	
Indication of Wilsons at transplant only	7	
Retransplants		2 received their first liver transplant before 2007

Next steps

- Continue case confirmation exercise
- Identified cases to be sent for prescriptions matching
- Interrogate ONS identifiable mortality data for cases containing E83.0 and references to Wilson's disease anywhere on the death certificate.
- Confirm vital status of confirmed cases & check deceased patient records against ONS mortality death certificate records for these patients.
- Detailed analysis will be carried out by NCARDRS staff and medical statisticians at Newcastle University.
- Work with the SAS Trace Elements subcommittee to support prospective data collection to NCARDRS.

- We welcome further support and suggestions about our outputs from this group.