Erratum: Creutzfeldt-Jakob disease surveillance in Australia: update to 31 December 2020

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Errata

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Details of the number of ‘incomplete’ suspect case notifications, in the second paragraph of the report’s Abstract as originally published, were incorrect. The corrected Abstract is published below.

Abstract

Nationwide surveillance of Creutzfeldt-Jakob disease and other human prion diseases is performed by the Australian National Creutzfeldt-Jakob Disease Registry (ANCJDR). National surveillance encompasses the period since 1 January 1970, with prospective surveillance occurring from 1 October 1993. Over this prospective surveillance period, considerable developments have occurred in pre-mortem diagnostics; in the delineation of new disease subtypes; and in a heightened awareness of prion diseases in healthcare settings. Surveillance practices of the ANCJDR have evolved and adapted accordingly. This report summarises the activities of the ANCJDR during 2020.

Since the ANCJDR began offering diagnostic cerebrospinal fluid (CSF) 14-3-3 protein testing in Australia in September 1997, the annual number of referrals has steadily increased. In 2020, 510 domestic CSF specimens were referred for 14-3-3 protein testing and 85 persons with suspected human prion disease were formally added to the national register. As of 31 December 2020, just over half (44 cases) of the 85 suspect case notifications remain classified as ‘incomplete’; 12 cases were excluded through either detailed clinical follow-up (8 cases) or neuropathological examination (4 cases); 18 cases were classified as ‘definite’ and eleven as ‘probable’ prion disease. For 2020, sixty percent of all suspected human-prion-disease-related deaths in Australia underwent neuropathological examination. No cases of variant or iatrogenic CJD were identified.

The SARS-CoV-2 pandemic did not affect prion disease surveillance outcomes in Australia.