Methods Since 1999, the Creutzfeldt-Jakob disease (CJD) Surveillance Committee collects data of all prion diseases by referring to registries to The Intractable Disease Treatment Research Program, reports of CJD as a notifiable disease and requests for prion protein gene or 14-3-3 protein analysis. In diagnosing prion diseases, all the referrals were assessed according to the case definition proposed by WHO. In the present study, patients with prion diseases over 65 years were analysed.

Results By August 2010, 1533 cases were confirmed and 996 (65%) were over 65 years of age. Among them, there were 798 (80%) sporadic CJD (sCJD), 35 (4%) dura mater graft-associated CJD (dCJD), 151 (15%) familial CJD (fCJD), 8 Gerstmann-Sträussler-Scheinker disease and three unclassified CJD. Proportions of sCJD and fCJD were higher in older patients than in younger patients. Percentage of definite cases which needs pathological confirmation were low (9% in sCJD, 46% in dCJD and 15% in fCJD) because only 12% underwent autopsy. The mean age at onset was 74 years. The latest follow-up survey revealed that 78% had died. The mean length of time from the onset to death was 15 months, which was 10 months shorter than in younger cases.

Conclusion In Japanese elderly, proportions of sCJD and fCJD were larger. Efforts to increase definite cases are needed to identify them correctly.

P2-540 EPIDEMIOLOGIC FEATURES OF PRION DISEASES IN JAPANESE ELDERLY: RESULTS FROM THE SURVEILLANCE

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Introduction Incidence and mortality of prion diseases has been increasing in Japan, especially in aged populations. The objective of this study was to investigate epidemiologic features of prion diseases in Japanese elderly.

Methods Since 1999, the Creutzfeldt-Jakob disease (CJD) Surveillance Committee collects data of all prion diseases by referring to registries to The Intractable Disease Treatment Research Program, reports of CJD as a notifiable disease and requests for prion protein gene or 14-3-3 protein analysis. In diagnosing prion diseases, all the referrals were assessed according to the case definition proposed by WHO. In the present study, patients with prion diseases over 65 years were analysed.

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