Brain Bank for Human Prion Diseases

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Neuropathologic approach, autopsy, is important to confirm the diagnosis as well as to understand the pathomechanism of human prion diseases. However, autopsies of patients who died from prion disease are always difficult due to lack of proper facilities and/or fear of disease transmission. (In Japan, the rate of autopsy is 14.7% [< 20 cases per year)].)

In order to collect brain resources for prion disease, we have taken the following actions for the last 10 years at our brain bank:

- Actively accept patients with prion diseases in our hospital to develop trust relationships between patients' families and hospital staff.
- Be on call around the clock to participate in the transport of the body in cases of autopsy requests from other hospitals. Carry out all autopsies on call.
- If an autopsy room is available for prion autopsy at other hospitals but no pathologist is available, a member (MT) goes to the hospital.
- Carry out all processes free-of-charge, including transportation of the body and pathologic diagnosis. The patient's family incurs no cost.
- Carryout general autopsy if permission is obtained from the patient's family.
- Store half of the brain and part of general organs at -80° C to use for biochemical and molecular analysis. In some instances, the neural retina is obtained
- Fix the left cerebral and cerebellar hemispheres and the brainstem in 20% buffered neutral formalin for neuropathologic diagnosis and research.

Since 2007, we have carried out more than 50 autopsies for prion diseases. Those cases include sporadic CJD, familial CJD, dura matter grafted CJD and Gerstmann-Sträussler-Scheinker diseases. We have also published the results of human prion diseases (1-5). We believe that

our system of brain banking may contribute to future research of prion diseases.

References:

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