Frontotemporal lobar degeneration (FTLD) is now the third most common cause of dementia, following Alzheimer’s disease (AD) and dementia with Lewy bodies. FTLD has unique clinical features different from those of AD in which loss of memory is often the initial presentation. Another characteristic is that FTLD often accompany amyotrophic lateral sclerosis (ALS). In this field, a number of Japanese works have made significant contributions, including description of ALS with dementia, well-known in Japan as Yuasa-Mitsuyama type. The pathological features of FTLD are also different from AD, and the recent discovery of trans-activation DNA binding protein with a molecular weight of 43kd (TDP-43) has provided further evidence for linkage of FTLD and ALS. As a result, FTLD is now of pressing interest.

The aim of the session is to take a fresh look at FTLD, and to bridge clinical, pathological and molecular information. In the session, a historical review including Japanese data will be shown first carried out, then unique clinical features including language problems, writing errors and acquired Savant syndrome will be introduced. Subsequently, the pathological aspects of FTLD/ALS with a focus on TDP-43 will be presented. The session will be consisted of the following 5 topics.