



# CDB SEMINAR

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Tuesday, September 18, 2012

16:00~17:00 C1F CDB Auditorium

### How do misfolded proteins and altered RNA metabolism cause ALS?

#### Summary

Accumulations of aggregated proteins are a key feature of the pathology of all of the major neurodegenerative diseases. Amyotrophic lateral sclerosis (ALS) was brought into this fold quite recently with the discovery of TDP-43 (TAR DNA binding protein, 43 kDa) inclusions in nearly all ALS cases. In part this discovery was fueled by the recognition of the clinical overlap between ALS and frontotemporal lobar degeneration, where ubiquitinated TDP-43 inclusions were first identified. Later the identification of TDP-43 mutations in rare familial forms of ALS confirmed that altered TDP-43 function can be a primary cause of the disease. However, the simple concept that TDP-43 is an aggregation-prone protein that forms toxic inclusions capable of promoting neurodegeneration has not been upheld by initial investigations. Our research is focused on using cell culture and animal model systems to understand the relationship between aggregation of RNA binding proteins, altered RNA metabolism, and neurodegeneration in ALS.

#### Host:

##### Hideki Enomoto

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