

3<sup>rd</sup> Annual Congress on

# INFECTIOUS DISEASES

August 21-23, 2017 San Francisco, USA



## Michael D Geschwind

University of California, USA

### The spectrum of human prion diseases

**Statement of the Problem:** Diagnosis of human prion diseases can be difficult as they can present similar to many other conditions, and many other conditions can clinically mimic prion disease. Correct diagnosis of prion disease is important in order to prevent accidental transmission of the prions, to prevent further unnecessary diagnostic testing and to provide a realistic prognosis to the patient and family.

**Methodology & Theoretical Orientation:** Our center has evaluated more than 2500 cases of rapidly progressive dementia (RPD), including more than 600 cases of prion disease through our clinical research program. Most patients undergo a comprehensive evaluation including clinical history, cognitive testing, CSF analysis, research brain MRI protocol and other testing. These data are analyzed to identify measures that might improve diagnostic accuracy of prion disease compared to other non-prion RPDs.

**Findings:** The clinical presentation, including presenting symptoms, duration of disease, and laboratory findings are quite varied in prion disease. Brain MRI with diffusion sequences showed high diagnostic accuracy for human prion disease. Unfortunately, radiologists in the USA often miss the radiological diagnosis of prion disease, despite the MRIs showing classic features. A relatively new CSF test called RT-QuIC shows high specificity, although not as good sensitivity, for prion disease diagnosis.

**Conclusion & Significance:** Our ability to diagnosis prion disease has improved over the past few years to the point at which brain biopsies are rarely needed. Improved diagnosis will be important for future treatment trials and prevention of accidental transmission of these potentially infectious diseases.

### Biography

Michael D Geschwind is a Professor of Neurology at the UCSF Memory and Aging Center who specializes in the assessment, treatment and management of rapidly progressive dementias, including prion diseases such as Jakob-Creutzfeldt disease (JCD) and autoimmune encephalopathies, and other cognitive/movement disorder syndromes. He helped to establish a program for the assessment of rapidly progressive dementias at UCSF Medical Center, the first of its kind in the country. He helped to run the first US treatment trial for sporadic disease, at UCSF. He has also helped to establish and co-direct a clinic for patients with autoimmune encephalopathy. He Co-directs the Huntington's Disease Society of America Center of Excellence (HDSA COE) and Ataxia Clinic at the UCSF Memory and Aging Center. His research interests include rapidly progressive dementias, cognitive dysfunction in movement disorders, such as Huntington's disease, spinocerebellar ataxia, corticobasal degeneration (CBD), progressive supranuclear palsy (PSP) and other Parkinsonian dementias.

Michael.Geschwind@ucsf.edu

### Notes: