AFTD’s March 5 EL-PFDD Meeting: Glossary of FTD Terms

As you observe and participate in the March 5 Externally Led Patient Focused Drug Development meeting on FTD, you may encounter some specific terms that it could benefit you to understand in advance. These and a range of other terms are also defined on AFTD’s website.

**ALS/FTD** — A clinical syndrome where both amyotrophic lateral sclerosis (ALS) and FTD occur in the same person. Symptoms include muscle weakness and atrophy, muscle contractions and twitches, and difficulty speaking or swallowing in addition to changes in behavior, personality or language. Also referred to as FTD with motor neuron disease (FTD-MND).

**Amyotrophic lateral sclerosis (ALS)** — Also known as Lou Gehrig’s disease, ALS is a neurodegenerative disease that results from the death of nerve cells in the brain and spinal cord responsible for controlling voluntary muscle movement. Up to 30% of people diagnosed with FTD also have symptoms of ALS; this form of FTD is called FTD-ALS or FTD-MND.

**Behavioral variant FTD (bvFTD)** — Form of FTD characterized by changes in personality, apathy, disinhibition, and a decline in judgment, self-control, and empathy. bvFTD is the most common form of FTD.

**Biomarker** — Physiological characteristic that can be objectively measured as an indicator of underlying biological or pathological process. Blood pressure — an indicator of heart disease — is an example of a biomarker. The identification of FTD biomarkers to guide diagnosis and drug development is a critical research priority; such a biomarker could be a protein or other factor measured in the blood or cerebrospinal fluid or features detected by brain imaging.

**Clinical diagnosis** — A diagnosis based on signs, symptoms, and medical history, in conjunction with laboratory tests or imaging procedures, but without confirmatory pathology. A clinical diagnosis of FTD can only be confirmed post-mortem by examining brain tissue for characteristic pathological features.
Clinical trial — Type of research study designed to evaluate the safety and effectiveness of a new medication or other type of treatment. Every clinical trial follows a protocol that includes clear criteria for participation, the dosage and duration of treatment, the methods that will be used to measure the effects of the treatment, and the statistical procedures that will be used to analyze the data.

Corticobasal degeneration (CBD) — The pattern of brain degeneration associated with the clinical symptoms of corticobasal syndrome (CBS). The affected brain regions include the frontal and temporal lobes as well as the brain stem, cerebellum, and substantia nigra. These regions play important roles in initiating, controlling, and coordinating movement.

Corticobasal syndrome (CBS) — The clinical diagnosis given to the symptoms associated with CBD. One of the movement-predominant FTD disorders, CBS is characterized by difficulty controlling limb movement and uncontrollable muscle contractions. People with CBS may also exhibit behavioral and language symptoms common to other forms of FTD.

Disease-modifying treatment — Medication or other form of treatment aimed at slowing or reversing the underlying disease process. Because no disease-modifying treatments are currently available for any of the FTD disorders, identifying such treatments is a high priority for FTD researchers.

Familial — Indicates that a disease has been documented in multiple family members. While a familial pattern of occurrence often occurs because of a gene mutation passed down from generation to generation, it can also be caused by common exposure to an environmental factor.

Frontotemporal spectrum disorder — Comprehensive term that places FTD without motor symptoms, FTD-ALS, and ALS on a disease continuum that accounts for the clinical, pathological, and genetic overlap between FTD and ALS.

Genetic testing — The analysis of an individual’s DNA to identify gene mutations associated with a disease.

Hereditary — Describes a disease genetically inherited by a child from an affected parent.

Magnetic resonance imaging (MRI) — Form of brain imaging that uses radio waves and magnets to visualize brain structure or function. While MRI scans cannot provide a definitive diagnosis of FTD, they can, in conjunction with other evidence from a neurological exam, medical history, laboratory or neuropsychological tests, and procedures such as lumbar puncture, support the diagnosis. MRI is also an important research tool.
Neuroimaging — A collective term for procedures that enable visualization of the structure or function of the brain and spinal cord, including changes associated with FTD and other brain disorders. Examples include CT, PET and MRI scanning.

Nonpharmacologic therapies — Treatments that are not medications. Transcranial direct current stimulation (tDCS) is an example of a nonpharmacologic treatment currently being tested in FTD.

Parkinsonism — Motor symptoms commonly observed in Parkinson’s disease, such as tremors, rigidity, abnormally slow movement, and difficulty maintaining balance. These symptoms (but not other symptoms of Parkinson’s disease) are also observed in two movement-dominant FTD disorders, corticobasal syndrome and progressive supranuclear palsy. As a result, these diseases are sometimes referred to as atypical parkinsonism.

Pathology — The medical specialty that focuses on the study of organs and tissues to determine the specific changes characteristic of diseases.

Primary progressive aphasia — The umbrella term for three FTD syndromes characterized by the progressive loss of the ability to speak, read, write, or understand spoken language.

Progressive supranuclear palsy (PSP) — A movement-predominant FTD disorder characterized by a progressive inability to move the eyes, especially vertically, as well as difficulties with balance, coordination, and movement of the muscles controlling the mouth and throat. People with PSP can also exhibit symptoms common to individuals with Parkinson’s disease, such as rigidity and abnormally slow movement. As a result, PSP is sometimes referred to as an example of atypical parkinsonism.

Sporadic — Refers to FTD cases in which the affected individual has no known family history of FTD disorders.

Symptomatic treatment — Medication or other form of treatment aimed at relieving symptoms, without affecting the process causing the disease. For example, medications are sometimes prescribed for people with FTD to decrease agitation and other behavioral symptoms, while speech-language therapy can preserve language skills in people with PPA.

Transcranial direct current stimulation (tDCS) — Nonpharmacologic, noninvasive form of treatment for brain disorders that applies a weak electrical current to the scalp to stimulate an increase or decrease in brain activity.