



## Frontotemporal Degeneration: An Urgent Need to Learn More Drives the CReATe Consortium

Frontotemporal degeneration (FTD) is a neurodegenerative disease that causes a combination of behavioral changes, speech and language problems, and movement disorders. It is caused by the degeneration of neurons (nerve cells) in two regions of the brain called the frontal lobes and the temporal lobes (see Figure). FTD is also sometimes referred to as frontotemporal dementia, frontotemporal lobar degeneration (FTLD), or Pick's disease.

FTD is included in the diseases covered by the CReATe consortium because of important overlaps between FTD and amyotrophic lateral sclerosis (ALS). "Some patients with ALS develop FTD, and some patients with FTD develop ALS," explains Rick Bedlack, MD, PhD, Director of the Duke University ALS Clinic, and Director of Advocacy and Outreach for the CReATe Consortium. Certain gene mutations that cause FTD also cause ALS, or both. The most common such mutation is in the C9orf72 gene. Mutations in the TARDP or FUS genes can also produce either or both ALS and FTD, in the same person or in different members of the same family. "This overlap suggests that the biology (and potentially the treatment) of these diseases may be linked," Dr. Bedlack says.

The symptoms of FTD fall into three categories:

- Behavioral changes. These may include personality change, loss of empathy, lack of inhibition, socially inappropriate behaviors, repetitive actions, and apathy.
- Speech and language changes. Symptoms may include difficulty finding the right word, use of inappropriate words, or slowed speech.
- Movement disorders. Symptoms may include tremor, rigidity, muscle spasms, and difficulty swallowing.

While they share some symptoms, FTD is not the same as Alzheimer's disease (AD), Dr. Bedlack emphasizes. Along with starting at an earlier age, "patients with FTD primarily have changes in their behavior and their personality," while those with AD have more trouble with memory or planning, while retaining socially appropriate behavior and language.

For family members with a loved one recently diagnosed with FTD, Dr. Bedlack offers this advice. "It is important to connect with health care professionals who can confirm the diagnosis and provide options for optimizing the safety and quality of life of the affected patient and family." The Association for Frontotemporal Degeneration (<http://www.theaftd.org>) is a great place to start to get information about the disease and to find professionals who can help.

In addition, there are important opportunities for getting involved in research to better understand FTD, and ultimately to develop new treatments. You can learn more about getting involved in research at <http://clinicaltrials.gov> (use the search term "frontotemporal degeneration").

The CReATe consortium is currently recruiting patients and family members for an observational trial to learn more about how FTD is related to ALS, and how the genes that cause



ALS-FTD relate to the clinical symptoms experienced by patients. You can learn more about this trial at <https://www.rarediseasesnetwork.org/CREATE/8001.htm>