

FROM CARE TO CURE

NEWS FROM THE MGH - FRONTOTEMPORAL DISORDERS UNIT

FALL 2014

Welcome to the FTD Unit: A Letter from Dr. Brad Dickerson



It's hard to believe that it's been seven years since we started the MGH FTD Unit. In this first issue of our newsletter, I'd like to extend a warm welcome from myself and our growing team. The mission of the MGH FTD

Unit is "to improve understanding of FTD, PPA and related disorders with the goals of finding a cure and providing better comprehensive care of patients and families." To achieve this mission, we aim to:

- Provide interdisciplinary clinical evaluation and care management services to maximize and maintain quality of life;
- Perform clinical and translational research to develop better tools for early diagnosis, prediction, monitoring, and treatment;
- Interface with investigators performing basic science research on these conditions, aiming to use this knowledge to find a cure;
- Provide targeted education to health care professionals, family caregivers, and community leaders across political, judiciary, and social service systems for

increased understanding of the many facets of managing an FTD diagnosis

The momentum in the fight against FTD is accelerating. We have launched the first-ever multi-center clinical drug trial of a medicine to try to treat the behavioral variant of FTD. The National Institutes of Health has just agreed to fund several major new FTD-related projects, going well beyond their previous portfolio. A number of pharmaceutical companies are building programs to try to treat FTD and related conditions. The 9th International Conference is approaching soon.

We cherish our partnerships with patients and families: our best hope for a future without FTD is to invest in research, which can't be done without you. While we work toward better therapies or hopefully cures, we need to proactively join together to develop better care resources and maximize quality of life for patients and families. Our team, and the larger community, is here to accompany you through the journey, and we are working tirelessly to improve what we can offer.

I'd like to express special appreciation to the Tom Rickles/Krupp Foundation PPA Research Fund for their generous support.

NEW PATIENT INQUIRIES:

(617) 724-6387

mghftdunit@partners.org

RESEARCH INQUIRIES:

Christina Caso,
*Clinical Research
Coordinator*

(617) 724-7092

mghftdunit@partners.org

Upcoming FTD Conferences:

The 9th International Conference on Frontotemporal Dementias (ICFTD) will take place in Vancouver, from October 23 – 25, 2014.

The ICFTD is held every two years and is the only regularly scheduled international conference devoted to frontotemporal dementia (FTD). This is the largest meeting bringing together both clinicians and scientists from all over the world.

In addition, from its beginning ICFTD has provided a day long informative session for caregivers. This has allowed both caregivers and family members to attend the conference, as well.

If you are interested in registering for this conference, or learning more information,

please visit: www.ftdvancouver2014.com

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The SIRS: A New Tool to Predict Social Symptoms in Brain Disease

Genevieve Wanucha, FTD Unit Staff Science Writer



THE SOCIAL BRAIN IN FTD

For people developing behavioral variant frontotemporal degeneration (bvFTD), and those around them, social life is difficult. These patients may become too trusting of strangers, drink from another person's water glass at the dinner table, or lose interest in family and friends. They can get into big trouble. For 20 years,

scientists and clinicians have documented how this disease degrades the human ability to communicate and empathize. Many experts call bvFTD a unique disorder of the social brain. Some patients with other forms of FTD, including Primary Progressive Aphasia (PPA), also may develop changes in social behavior.

Traditionally viewed as random and unpredictable, the hallmark socio-emotional deficits in early bvFTD have received less dedicated study than cognitive and memory problems. Families often leave the clinic without answers about the causes of a loved one's odd behavior or a roadmap for what's to come.

However, dozens of brain-imaging studies show that the behaviors in bvFTD are far from random. Specific socio-emotional changes, such as diminished empathy or emotional connectedness to loved ones, relate to the brain areas most damaged in the disease. Now, emerging from a big step forward

Drawing conclusions from a second-hand description of social behavior has notorious pitfalls. In the past, most of the instruments used to measure these abilities are brief questionnaires filled out by family members. Answers may be vague, biased, or may omit crucial details or context. A patient's wife might say 'yes' to a question asking if her husband communicates less with his family. But has he lost the desire or ability to engage in conversation? Or does she mean that he is withdrawing because can't follow along in conversations? The SIRS interview takes up to an hour or more as the clinician probes for examples of behaviors to get at the true nature and severity of the deficits. The final profile of ratings in each of the 6 domains of the scale ultimately pinpoints and quantifies the big problem areas for an individual in social contexts.

"The SIRS is a tool that we hope will be useful for measuring different kinds of social symptoms in FTD, allowing doctors to inform families, for example, that a patient is more likely to have progressive symptoms of a socially extraverted type rather than a more emotionally detached one," said social neuroscientist Kevin Bickart, who worked on this research during his PhD dissertation under the mentorship of Dr. Brad Dickerson, a behavioral neurologist and Director of the MGH FTD Unit, and Dr. Lisa Feldman Barrett, a social neuroscientist and director of the Affective Neuroscience Laboratory at Northeastern University.

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in this line of neuroscience research, investigators in the MGH Frontotemporal Disorders Unit have designed a new clinical tool called the Social Impairment Rating Scale (SIRS) that measures the types and severity of social symptoms in people with FTD.

MEASURING A SOCIAL LIFE

The SIRS is a structured interview in which a trained clinician asks a close family member of a person with FTD a series of questions about the patient's everyday social behavior. The questions fall into different social domains, including the lack of attention to social cues; being overly trusting of or willing to approach strangers; inappropriateness in social situations; and socio-emotional detachment from family members or friends. The clinician rates the symptoms described in the interview on a 5-point scale indicating their severity.

PATHWAYS OF THE SOCIAL BRAIN

The work that led to the SIRS actually started out as an investigation into the healthy social brain (some of the work done by the MGH FTD Unit aims to better understand healthy human brain function). Several years ago, Dickerson, Barrett, and Bickart had become fascinated with the amygdala, an almond-shaped structure deep in the brain known to process emotional information in social contexts. In fact, they had recently found that the amygdala is bigger in people who maintain larger real-world groups of friends. But the amygdala doesn't do its social work alone. Functional fMRI (imaging that reveals patterns of activity in the living brain) shows that the amygdala works as part of networks of communicating brain areas broadly implicated in emotional and social experience, including distinct regions within the frontal, temporal, and insular lobes.

In a 2012 brain imaging study of healthy people published in the *Journal of Neuroscience*, the team discovered that the amygdala brings more to the party than anyone knew, working as a common node in three distinct brain networks. Making correlations between the psychological traits of the study participants and the strength of connectivity in these three networks, the group hypothesized that each one handles a different aspect of social life: an affiliation network that promotes pro-social bonding behavior; an aversion network involved not only in physical pain but also in avoiding social threats, such as untrustworthy people; and a perception network that decodes and interprets social signals from others.

These findings made intuitive sense to Dickerson, who as the leader of the MGH FTD Unit sees time and again how FTD patients develop problems in affiliation, aversion, and social perception. He suggested to his team that these patients, and their brain scans, might help them better pin down specific brain network-behavior relationships.

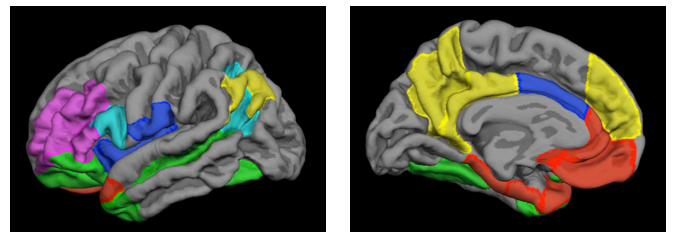
To do so, the team needed a way to measure socio-emotional function—just like a neurologist measures memory deficits. “But there was no instrument that had previously been developed to quantify the severity of impairment in a broad array of social domains,” said Kevin Bickart. “So we had to develop our own scale in order to get at very specific social impairments.” Since Bickart was in a combined MD/PhD program, he had been seeing patients with Dr. Dickerson in the FTD Unit, seeing how memory, language, and other symptoms were measured. Together with the group’s hypotheses about the specific functions of the three brain networks described above, he worked with Drs. Dickerson and Feldman Barrett to develop the SIRS, a detailed framework for measuring impairments in social behavior, modeled after a scale widely used in Alzheimer’s disease research to measure memory and other cognitive impairments.

A BRIGHTER FUTURE

Bickart, Dickerson, and team piloted the SIRS interview with 20 family members of patients with FTD. When the team matched up these detailed profiles of social impairments with the patients’ brain scans, a surprisingly clear pattern emerged. The patients with the worst atrophy (shrinkage) in the affiliation network were the most emotionally disconnected from family and friends, whereas patients with the greatest atrophy in the aversion network showed the greatest lack of social apprehension, giving away personal information to people they didn’t know, for example, or inviting strangers into their homes. Patients with the greatest damage in the perception network showed the most severe lack of awareness of social cues, such as others’ gestures, eye contact, personal

distance, changes in facial expression, or even head nods or waves. Similar observations had previously been made in patients with brain damage due to strokes or head injuries, but have not been previously incorporated into this kind of a framework.

In essence, the team found that the extent of brain damage in these three social brain networks in the study’s patients predicted the severity of distinct social impairments measured using the SIRS. The findings, now published in the *Journal of Neurology, Neurosurgery, and Psychiatry*, not only validate a new clinical tool, but also support MGH FTD Unit team’s hypotheses about the healthy social brain.



Most interesting about the neuroimaging results, says Bickart, is the potential for better diagnosis and monitoring of FTD spectrum disorders. “The study shows that MRI is useful for identifying specific regions that researchers can focus in on to look for differences in brain structure and function that relate to distinct social deficits. It can simplify how we look for these types of abnormalities.”

The MGH FTD Unit team sees potential benefits to formally recognizing different types of social symptoms in neurodegenerative disease. With the SIRS, it may be possible to predict, from early damage to a person’s aversion network, that she could soon be in danger of falling victim to a financial scam or getting into a stranger’s car. Families could prepare for, and possibly lessen the impact of, the worst incidents that FTD can bring. In addition, the SIRS may be valuable in monitoring symptoms in the context of studies aiming to treat social impairments.

The science behind the SIRS reveals something that the MGH FTD Unit has always known. FTD is not, especially in the beginning stages, a diffuse problem in the brain that unleashes an incomprehensible array of symptoms. Some of the symptoms affecting patients’ interactions with loved ones arise from damage to specific circuits of the social brain, and this knowledge may help to demystify some of the most painful symptoms of the illness.

RESEARCH OPPORTUNITY:

Study of Families at Risk for FTD or Related Disorders

OUR RESEARCH. YOUR STORY.

Research is happening at MGH that is focused on learning about the cognitive abilities, personality, and other traits of people who have more than one blood relative affected with FTD, Alzheimer's disease, and related disorders.

YOU CAN CONTRIBUTE TO THE CAUSE

Diagnosis of FTD is often challenging and current treatment options are limited. We hope that the scientific research we are conducting at MGH will change that for future generations.

We are seeking people who had more than one blood relative with a diagnosis of FTD (including behavioral variant FTD, also known as Pick's disease or Primary Progressive Aphasia), Amyotrophic Lateral Sclerosis (ALS), Corticobasal Degeneration (CBD), Parkinson's, Progressive Supranuclear Palsy (PSP), Alzheimer's disease, or a related disorder, to participate in our research.

By learning about you and your family, we hope the knowledge gained from this work will help researchers and doctors to learn if there are certain cognitive or behavioral traits that can help predict the development of these disorders.

YOU CHOOSE WHAT YOU LEARN

You do not have to learn anything about your test results by participating in research. Our clinical research time respects that decision to participate in research is an important one. We will honor the gift of your time and dedication to our research efforts by maintaining a responsive partnership. To individualize this experience, we will keep an open dialogue and provide feedback for your questions and concerns.

For more information or to find out if you are eligible for this study, please contact:

Christina Caso, *Clinical Research Coordinator*
(617) 724-7092
cdcaso@mgh.harvard.edu

CAREGIVER CORNER:

Katie Brandt, Co-Facilitator, Boston-area FTD Support Group



I am someone who likes to travel. It might be because as a former FTD caregiver and a current Alzheimer's caregiver, I feel like I actually need to leave the house to let my mind take a vacation. When I know I am headed out the door on a trip, I like to plan ahead. I want to know how I will arrive at my destination. Plane, train or automobile? I want to gather any necessary documents or supplies. Passport? Pocket-sized translator for quick reference? Whenever possible, I want to enlist a friend to travel with me. It helps to have someone you can borrow a pair of underwear from when your baggage gets lost, or to navigate the unmarked roads when the GPS fails.

Caregiving has been a journey for me. This trip has proven difficult to plan, with an ever-shifting itinerary and many unmarked roads. My passport into this country of care was simply that I loved someone with a Frontotemporal Disorder. What I thought would be a solo journey turned out to be a group trip worthy of a caravan. The caring clinicians of the FTD Unit offered themselves as travel guides. They provided supplies, navigation, and connections to a powerful network of caregivers who had traveled the road before. FTD Unit staff employ a holistic approach to caring for patients that involves support for the caregiver too. They refer families to caregiver resources through The Association for Frontotemporal Degeneration and the Alzheimer's Association. The Boston-area FTD Support Group is a community where caregivers share stories, ask questions, and learn practical tips and resources for the road ahead.

Every journey is better with a friend. If you are caregiving alone, reach out and make a connection. You will find that our bags are packed to travel with you.

www.theaftd.org

www.alz.org

FTD Support Group email: ftd.boston@gmail.com

**Thank you for considering a donation to the
Frontotemporal Disorders Unit at Massachusetts General Hospital.**

For your reference, our non-profit tax ID number is 04-3230035.

Checks should be made payable to MGH Dr. Dickerson Lab.

In the memo line, please put **Dementia Research**.

They can then, be mailed to:

MGH Development Office

100 Cambridge Street, Suite 13108 • Boston, MA 02114

Donations may also be made online by following these procedures:

1. Go to this MGH website: <https://secure.massgeneral.org>
2. Click on "Donate Now"
3. Select amount, and click on Designation and select "Other" Then, enter "Dickerson Dementia Research Fund" Then, enter details below and submit (In comment line, write: Direct the donation to Dr. Dickerson's Alzheimer's and Dementia Research Fund)

We can't thank you enough for contributing to this important cause.

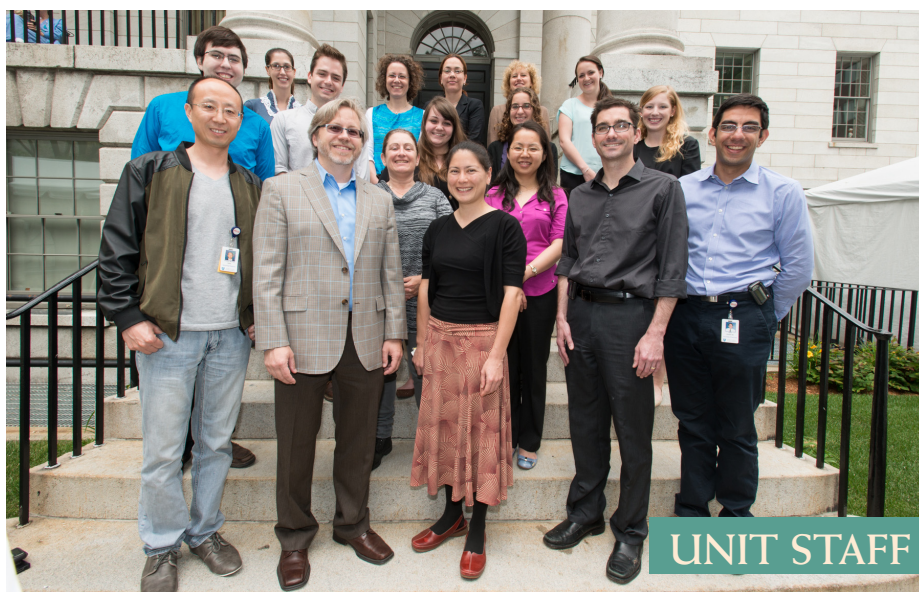
PATIENT PERSPECTIVE: *Dennis White*



I'm a 63 year old male with a diagnosis of Primary Progressive Aphasia (PPA). I'll start by talking about the Kübler-Ross five stages of grief: Denial, Anger, Bargaining, Depression, and most importantly, Acceptance. I thought I was experiencing these emotions out of order. I definitely felt denial at first (not me), followed by anger (why me), and then I seemed to skip bargaining and go right to acceptance. I had it all wrong. I followed these emotions exactly in order. I'm now feeling depressed after bargaining with all my research on Wikipedia.

Joyce Shapiro Gordon, my speech therapist, mentioned that she had a friend that was at the Berklee School of Music, and qualified as a music

therapist. They were interested in putting together a group of people with PPA as a speech/music therapy exercise. Some were worse off than me and some were better. It was amazing to listen and talk to people who have what I have. Joyce and Megan Quimby, another Speech Language Pathologist at MGH, asked how we all dealt with phone conversations. All I could think of is that I try to avoid phone conversations, like the plague. But one of the participants said when she answers the phone she starts by saying: "I have a speech disorder." Why should I not move from feeling depressed to acceptance of what I have?



INTRODUCTION TO THE UNIT STAFF:

Bradford Dickerson, MD: Behavioral Neurologist, Director of FTD Unit

Daisy Hochberg, MS, CCC-SLP: Senior Speech Language Pathologist

Diane Lucente, MS, CGC: Senior Genetic Counselor

Scott McGinnis, MD: Neurologist, Director of Clinical Trials, FTD Unit

Mark Eldaief, MD: Neurologist

David Perez, MD: Neurologist

Elena Ratti, MD: Neurologist

Sara Mitchell, MD: Neurology Fellow

Chenjie Xia, MD: Neurology Fellow

Jaya Padmanabhan, MD: Psychiatry Fellow

Jessica Collins, PhD: Research Fellow

Megan Quimby, MS: Speech Language Pathologist Fellow

Mike Brickhouse: Research Assistant

Mike Stepanovic: Research Assistant

Christina Caso: Research Assistant

Katie Kelly: Research Assistant

Sara Makaretz: Research Assistant

Claire Cordella: PhD student Harvard-MIT

Barbara H. Maxam, LSW: MGH Social Worker

Liz Lynch: Administrative Assistant to Dr. Brad Dickerson

Rose Gallagher: Clinical Patient Service Coordinator

Katie Brandt: FTD Unit Consultant, Patient-Family Liaison

Genevieve Wanucha: FTD Unit Staff Science Writer

From Care to Cure



An exciting and informative event specifically designed for caregivers and family members of those with Frontotemporal Degeneration (FTD).

This event will focus on providing appropriate resources for you and your loved one.

You will hear from professionals in the fields of:

- FTD Research
- Life Care Management
- Behavioral Management
- Elder Law
- Family Caregiving

**An FTD Caregiver Education Event sponsored by:
Massachusetts General Hospital, Frontotemporal Disorders Unit**



This event will be located at The Westin Waltham Boston Hotel, 70 3rd Avenue, Waltham, MA 02451

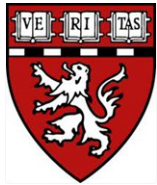
Please contact Liz Lynch for more information: 617-643-2847

Date: November 13, 2014

Time: 9:00 AM—3:00 PM

*Registration/Resource Fair to begin at 9 am





Massachusetts General Hospital
Frontotemporal Disorders Unit
Tel: 617-643-2847

From Care to Cure: An FTD Education Event

An exciting and informative event specifically designed for caregivers and family members of those with Frontotemporal Degeneration (FTD). This event will focus on providing appropriate resources for you and your loved one.

Date: November 13th, 2014
Time: 9:00 AM—3:00 PM
***Registration/Resource Fair to begin at 9 am**

This event will be located at
The Westin Waltham-Boston Hotel
70 3rd Avenue, Waltham, MA 02451

10 am-12 pm: Research Update from the MGH FTD Unit

1:30 pm-3:00 pm: Professional Workshops

- **Lindsay Brennan: Executive Director of Life Care Directions of New England, LLC**
Transitioning Through Care Options: Determining the best supports for your family
- **Paul Raia, Ph.D: VP, Professional Clinical Services Alzheimer's Assoc., MA/NH Chapter**
Behavioral Sleuthing: Examining the Clues that Cause Challenging Behaviors and Interventions.
- **Pamela Greenfield, Esquire: Senior Associate, Oalican Law Group, LLC**
Practical Planning: Legal Issues to Consider with an FTD Diagnosis
- **Patient Q & A with MGH FTD Unit Clinicians**

Please register by emailing the information below to EALYNCH1@partners.org or please mail the form below to
Attn: Liz Lynch, MGH FTD Unit, 149 13th Street, Ste 2691, Charlestown, MA 02129

Massachusetts General Hospital: FTD Unit

MGH FTD Unit
149 13th Street, Suite 2691
Charlestown, MA 02129

Phone: 617-643-2847
Fax: 617-726-5760
E-mail: EALYNCH1@partners.org

Name _____

Address _____

Phone _____

Please put the corresponding number next to your choice of workshops:

i.e. first choice - 1, second choice - 2, etc

LIGHT BREAKFAST AND LUNCH TO BE SERVED

Workshop #1 - 1:30-2:15 pm

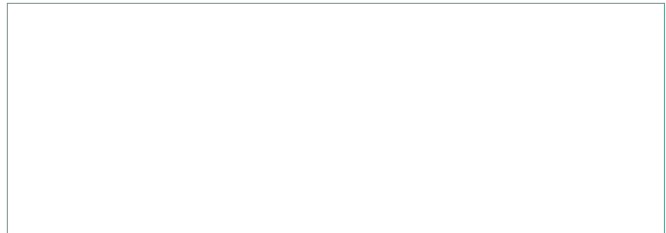
- Paul Raia, Ph.D
- Lindsay Brennan
- Pamela Greenfield, Esq
- Patient Q & A

Workshop #2 - 2:15- 3:00 pm

- Paul Raia, Ph.D
- Lindsay Brennan
- Pamela Greenfield, Esq
- Patient Q & A

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www.FTD-Boston.org

OUR MISSION:

Our Frontotemporal Disorders (FTD) Unit is a comprehensive clinical-research unit focused on Frontotemporal Degeneration and related disorders,

INCLUDING:

Behavioral Variant FTD
Primary Progressive Aphasia (PPA)
Semantic Dementia (SD)
Corticobasal Degeneration Syndrome (CBD)
Progressive Supranuclear Palsy (PSP)
Posterior Cortical Atrophy (PCA)
Atypical forms of Alzheimer's
FTD-ALS (amyotrophic lateral sclerosis)

Our team is committed to sophisticated, compassionate, multidisciplinary care of patients with neurodegenerative disorders.

WE PROVIDE SERVICES IN THE AREAS OF:

- Neurology
- Neuropsychology
- Speech Pathology
- Neuro-Ophthalmology
- Psychiatry
- Genetic Counseling & Testing Services
- Physical & Occupational Therapy
- Social Work
- Neuropathology

