

COGNITIVE NEUROLOGY AND ALZHEIMER'S DISEASE CENTER

*of the Northwestern University Feinberg School of Medicine presents the*

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**FRONTOTEMPORAL DEGENERATION  
AND  
PRIMARY PROGRESSIVE APHASIA  
CAREGIVER EDUCATION AND SUPPORT CONFERENCE  
MONDAY, MARCH 21, 2011**

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ROBERT H. LURIE MEDICAL RESEARCH CENTER • HUGHES AUDITORIUM  
303 E. SUPERIOR STREET • CHICAGO, IL 60611  
[WWW.BRAIN.NORTHWESTERN.EDU](http://WWW.BRAIN.NORTHWESTERN.EDU)



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# FRONTOTEMPORAL DEGENERATION (FTD) AND PRIMARY PROGRESSIVE APHASIA (PPA) CAREGIVER EDUCATION AND SUPPORT CONFERENCE

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**MARCH 21<sup>ST</sup>, 2011**

## **AGENDA**

- |                |   |
|----------------|---|
| 7:30-8:30 AM   | <b>Registration and Vendor Fair</b>   |
| 8:30-9:00 AM   | <b>Welcome</b><br>Marsel Mesulam, MD, CNADC Director<br>Darby Morhardt, MSW, LCSW, CNADC Education Director<br>Christina Wieneke, BA, CNADC Research Project Manager                          |
| 9:00-9:15 AM   | <b>Opening Remarks</b><br>Sharon Denny, MA, Association for Frontotemporal Degeneration (AFTD)<br>Ellayne Ganzfried, MS, CCC-SLP, National Aphasia Association (NAA)                          |
| 9:15-9:55 AM   | <b>Keynote Address: “Perspectives on FTD and PPA”</b><br>Bradley F. Boeve, MD, Professor of Neurology, Mayo Clinic, Rochester   |
| 9:55-10:15 AM  | <b>“Assessing and Treating Mood and Behavioral Symptoms in FTD and PPA”</b><br>Deborah Reed, MD, Assistant Professor of Psychiatry, Northwestern University                                   |
| 10:15-10:30 AM | <b>Break and Vendor Fair</b>  |
| 10:30 AM-noon  | <b>Question-and-Answer Session</b><br>Dr. Boeve, Dr. Reed, Eileen Duhig, Nancy Gapinski, and Lorene Schlie  |
| noon-1:00 PM   | <b>Lunch and Vendor Fair</b>  |
| 1:15-2:15 PM   | <b>Breakout Session 1</b>   |
| 2:30-3:30 PM   | <b>Breakout Session 2</b>   |
| 3:45-4:15 PM   | <b>Closing Plenary: “Affirm Yourself for Caregiver Challenges”</b><br>Janet Edmunson, MEd, Author of <i>Finding Meaning with Charles: Caregiving with Love Through a Degenerative Disease</i> |



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## THANK YOU!

The Cognitive Neurology and Alzheimer's Disease Center of the Northwestern University Feinberg School of Medicine thanks the Association for Frontotemporal Degeneration and the National Aphasia Association for their generous support of this special event.



The Association for  
Frontotemporal Degeneration  
*Opening the gateway to help and a cure*

**aphasia**  
NATIONAL  
**APHASIA**  
ASSOCIATION

# THANK YOU!

2011 Planning Committee Members:

CNADC Faculty and Staff

Joseph Boyle  
Darby Morhardt  
Mary O'Hara  
Kristen Oshyn  
Jaimie Robinson  
Emily Rogalski  
Christina Wieneke  
Kristine Zachrich

*and all volunteers who have made this day a success!*

The CNADC appreciates your dedication and commitment to making this day possible.



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# PROGRAM HIGHLIGHTS

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# PROGRAM HIGHLIGHTS

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**Keynote Speaker**  
**Bradley F. Boeve, MD**

**Professor of Neurology**  
**Mayo Clinic College of Medicine**  
**Mayo Clinic Rochester**

Dr. Bradley F. Boeve received his Doctor of Medicine degree from the University of Florida in 1991. He underwent training in Internal Medicine (internship), Neurology (residency), Behavioral Neurology (fellowship), and Sleep Medicine (fellowship) in the Mayo Graduate School of Medicine at Mayo Clinic in Rochester, Minnesota. He joined the Mayo Clinic staff in 1997 and now serves as Chair of the Division of Behavioral Neurology of the Mayo Foundation, Head of the Behavioral Neurology Section at Mayo Clinic Rochester, Director of the Behavioral Neurology Fellowship Program at Mayo Clinic Rochester, Co-Director of the Clinical Core of the Mayo Alzheimer's Disease Research Center, and is Professor of Neurology in the Mayo Clinic College of Medicine. He is a Diplomate of the American Board of Psychiatry and Neurology, American Board of Sleep Medicine, and United Council on Neurologic Subspecialties (Behavioral Neurology and Neuropsychiatry), and is a member of the American Medical Association, American Academy of Neurology, American Neurological Association, American Academy of Sleep Medicine, Society for Cognitive and Behavioral Neurology, and Movement Disorders Society. He also serves on the Medical/Scientific Advisory Councils of the Association from Frontotemporal Dementias and Lewy Body Dementia Association, and state chapter of the Alzheimer's Association.

Dr. Boeve's clinical and research interests include normal aging, mild cognitive impairment, Alzheimer's disease as well as the non-Alzheimer degenerative dementias (e.g., dementia with Lewy bodies, frontotemporal dementia, primary progressive aphasia, corticobasal degeneration, posterior cortical atrophy), the neurologically-based sleep disorders (e.g., REM sleep behavior disorder, narcolepsy, restless legs syndrome), prion disorders, neurogenetics, and the autoimmune/inflammatory encephalopathies. He has received grant support from the Alzheimer's Association, National Institute on Aging, and National Institute of Neurological Disorders and Stroke. He has authored 26 book chapters and has contributed to over 200 papers in peer-reviewed journals.

## Deborah A. Reed, MD

Dr. Deborah Reed is Assistant Professor of Clinical Psychiatry and Behavioral Science, a college mentor, and the former Division Director of Geriatric Psychiatry at Northwestern University Feinberg School of Medicine. She is also the Consulting Psychiatrist at the Cognitive Neurology and Alzheimer's Disease Center (CNADC).

Dr. Reed received her bachelor's degree in Biology from the University of Pennsylvania, her Doctor of Medicine degree from the Medical College of Pennsylvania, and completed her residency in Adult Psychiatry at Northwestern Memorial Hospital. She has participated in clinical drug trials for patients with FTD and PPA at Northwestern. Her interests include the pharmacologic management of cognitive and behavioral syndromes in patients with dementia, attention deficit disorder and other neurologic conditions.

## Janet M. Edmunson, M.Ed

While working full-time, Janet took care of her husband, Charles, during the five years he fought a movement disorder with dementia. Janet wrote about her experience in her book, *Finding Meaning with Charles: Caregiving with Love through a Degenerative Disease*.

Janet has over 30 years' experience in the health promotion field. She retired in May 2007 as Director of the Prevention & Wellness for a staff of 20 at Blue Cross Blue Shield of Massachusetts. Since retirement, she has been a national inspirational speaker having spoken to over 200 groups in the last couple of years.

Janet is Chair of the Board of Directors for the *Foundation for PSP | CBD and Related Brain Diseases*. She is also on the Board of Trustees for the Employee Ownership Foundation. She is a former President of the Association for Worksite Health Promotion. Janet has a Master's degree from Georgia State University.

# PROGRAM HIGHLIGHTS

## Afternoon Breakout Sessions for Caregivers

### **Newly Diagnosed FTD**

A new FTD diagnosis brings on feelings of devastating loss and uncertainty. Families may feel overwhelmed trying to manage the personality and behavioral changes as they search for guidance about what to expect, how to plan for the future, and what living with FTD will be like. This group will focus on the experience of a new diagnosis, the first steps to take, and where to find ongoing guidance, resources, and support. It will explore both immediate concerns and longer-term issues about how this illness will affect a loved one and a family.

### **Newly Diagnosed PPA**

Families may be uncertain about what to do next when a person is diagnosed with PPA. How should the frustrations and struggles resulting from changing communication abilities be addressed? There are important considerations for the present and the future that families should discuss early on. This group will focus on both the first steps and where to find ongoing guidance, resources, and support for the person with PPA and the family.

### **Legal Planning**

Once a person is diagnosed with a dementia, family and friends should help him or her to make legal plans early on so that, if possible, the person is able to participate. Lack of insight can result in the person's being unable or resistant to making these plans. In some cases, guardianship is necessary. Making legal plans for health care and long-term care includes discussing finances and property and naming another person to make decisions on behalf of the person. This group will discuss what families should know about legal planning and how to begin this process.

### **Managing Behaviors**

The range of possible new behaviors of persons with bv (behavioral variant) FTD may be difficult for families to manage. This group will discuss environmental modifications, psychosocial approaches, and pharmacological interventions for inappropriate, impulsive, aggressive, compulsive, paranoid, and other challenging behaviors. It will also explore how these behaviors affect the family and how the family can cope.

### **Communication Challenges in PPA**

The primary goal of speech and language therapy for PPA is to improve a person's ability to communicate by helping him or her to use remaining language abilities, compensate for language problems, and learn other methods of communication. This group will discuss alternative and augmentative forms of communication, the use of low- and high-tech communication devices, and other methods of supporting language abilities.

# PROGRAM HIGHLIGHTS

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## Afternoon Breakout Sessions for Caregivers

### **Families with Children and Teens**

Since PPA and FTD have a young average age of onset, a parent may become affected when children are still living at home. The stress brought on by the diagnosis is profound; families face uncertainty about not only the disease, but also its effect on children. This group will explore the experiences of children and teens who have parents with FTD or PPA.

### **Planning for Future and Palliative Care**

Planning for future care can feel complicated and overwhelming for caregivers. There is a great deal to consider in exploring what care options work best for the person diagnosed and his or her family. This group will provide a comprehensive review of care options throughout the course of the disease. It will also review palliative approaches to care, a comfort-based approach that focuses on maximizing quality of life.

### **Starting a Support Group in Your Area**

Support groups offer a safe environment in which family caregivers share their experience with others who truly understand. There is a need for more support groups specific to FTD and PPA. This group will discuss how -- with the help from the AFTD, the NAA, and the Alzheimer's Association -- to begin, sustain, and maintain a group specifically for caregivers of people with FTD or PPA.

### **Genetics and FTD/PPA**

FTD and PPA can be described as inherited, familial, or sporadic. In inherited cases, each child or sibling of the person with the disease has a 50 percent chance of developing it. In a familial case, family members of an affected individual have an increased risk of developing the disease, although this increase is not well defined. The majority of FTD/PPA cases are sporadic, meaning the disorder develops by chance and the children or siblings of the affected individual do not have an increased risk for developing the disorder. This group will discuss the genetics of FTD and PPA, including what we know and what we still hope to uncover through future research.

# PROGRAM HIGHLIGHTS

## Afternoon Breakout Sessions for Caregivers

### Group Facilitators

**Nicole Batsch** has over 15 years of experience working for not-for-profits developing programs in senior centers, Alzheimer's care, applied research, and family caregiving in Florida, Chicago, and Arizona. She works for the Alzheimer's Association Greater Illinois Chapter as the Director of Early Stage & Support Services developing programs that directly help the diagnosed individual in the early stages of Alzheimer's disease experience an increased sense of security, self-esteem and hope. She also supervises the over 100 support groups in the Chapter's 68 county service area.

**Bradley Boeve, MD** is Chair of the Division of Behavioral Neurology of the Mayo Foundation, Head of the Behavioral Neurology Section at Mayo Clinic Rochester, Director of the Behavioral Neurology Fellowship Program at Mayo Clinic Rochester, Co-Director of the Clinical Core of the Mayo Alzheimer's Disease Research Center, and Professor of Neurology in the Mayo Clinic College of Medicine. He has authored 26 book chapters and has contributed to over 200 papers in peer-reviewed journals.

**Henrietta Boudros MA, CCC-SLP** is a Chicago native with over ten years of clinical experience working with adults who have cognitive communication disorders. She has worked closely with the team at CNADC to assist in establishing appropriate treatment plans for individuals diagnosed with PPA/FTD.

**Joan Brzezinski** is a full-time worker, mother of a now 27-year old daughter, Theresa, and caregiver to her father for some 20 years and then to her husband Tom. Tom was diagnosed with FTD at the age of 53 and passed away at the age of 59 in July 2009. The Brzezinskis are a family of caregivers through and through, and Joan and Theresa continue to stay involved to try to make a difference.

**Theresa Brzezinski** was a 19-year old college student when her father was diagnosed with FTD at age 53. Theresa received a degree in Public Relations at Bradley University; however, after graduation and one year of employment, Theresa stopped working full-time to assist her mother in her father's care. Theresa's father passed away at the age of 59, in 2009. Theresa and her mother, Joan continue to help many families by talking about their experience and what they learned along the way.

**Derin Cobia, PhD** is a postdoctoral fellow in the Department of Psychiatry and Behavioral Sciences at the Northwestern University Feinberg School of Medicine. He earned his degree in clinical psychology, specializing in neuropsychology. His research interests are on the use of neuroimaging techniques to understand how changes in brain structure relate to language functioning in PPA.

## Caregiver Group Facilitators, continued

**Joseph Cooper, MD** completed his undergraduate degree in Neuroscience at Johns Hopkins University. He then completed medical school and residency in Psychiatry at the University of Chicago. In July 2010, he joined the Cognitive Neurology and Alzheimer's Disease Center at Northwestern University as a fellow of Neuropsychiatry and Behavioral Neurology.

**Sharon S. Denny, MA** is the Program Director at the Association for Frontotemporal Degeneration where she directs patient and caregiver support and education initiatives. Since coming to AFTD in 2008, her focus has been on the development and delivery of high-quality services for people with FTD, their families and the professionals working with them. Ms. Denny has a Masters degree in Clinical Psychology and more than twenty-five years experience with nonprofit organizations that serve people with medical and psychiatric disabilities.

**Amy and Eileen Duhig** are daughters of Marty, who was diagnosed with FTD in March 2005. They are both employed full-time and go home to provide their mother respite on a weekly basis.

**Janna Dutton** has been practicing in the area of elder law for more than 30 years. As a result, Janna has the knowledge and depth of experience to skillfully navigate through a diverse assortment of elder law matters, including trusts and estate planning, Medicaid and Medicare planning and applications and probate and estate administration. She is certified by the National Elder Law Foundation as an elder law attorney, and has held this certification since 1995.

**Janet Edmunson, M.Ed** is Chair of the Board of Directors for the *Foundation for PSP | CBD and Related Brain Diseases*. She was a caregiver to her husband, Charles, during the five years he fought a movement disorder with dementia. Janet wrote about her experience in her book, *Finding Meaning with Charles: Caregiving with Love through a Degenerative Disease*.

**Ellayne Ganzfried, MS, CCC-SLP** is a speech-language pathologist and the Executive Director of the National Aphasia Association. She is Past President of the NYS Speech Language Hearing Association (NYSSLHA), Long Island Speech Language Hearing Association (LISHA) and the Council of State Association Presidents for Speech Language Pathology and Audiology (CSAP) and remains active in these associations. Ellayne is a Fellow of the American Speech Language Hearing Association (ASHA). Ellayne has written articles and presented regionally and nationally on a variety of topics including aphasia, rehabilitation and leadership skills.

**Nancy Gapinski** is a wife, married 4 1/2 years to Chris, who was diagnosed with Primary Progressive Aphasia in August 2008. They have 9 nephews, 3 nieces and 1 great-niece. Nancy is employed full-time for a private company in the Risk Management Department.

**Tamar Gefen** is a clinical psychology doctoral student at Northwestern University Feinberg School of Medicine, specializing in clinical neuropsychology under the mentorship of Dr. Sandra Weintraub. She is conducting research at the CNADC on Primary Progressive Aphasia, Alzheimer's disease, and on the underlying mechanisms of aging.

## Caregiver Group Facilitators, continued

**Josh Hauser, MD** is Assistant Professor of Medicine and Palliative Care at Northwestern's Feinberg School of Medicine, Director of Education at the Buehler Center on Aging, Health and Society and Director of The Education on Palliative and End of Life Care (EPEC) Project. He practices in the palliative care program at Northwestern Memorial Hospital. As Director of the EPEC Project, he oversees all of its conferences and curricula, including the development of the EPEC for Veterans Curriculum, the creation of EPEC-distance learning and the creation of a curriculum on communication skills for patients and caregivers, entitled *Navigating the System*.

**Julianne Hill** is an award-winning journalist, filmmaker and documentarian whose work has focused on brain diseases, including essays aired on *This American Life* and run in *Health and Real Simple*. Hill also writes and produces videos for caregivers via the Alzheimer's Foundation of America. She lives with her bright and beautiful 16-year old son, Nicolas, founder of Cognitive Neurology & Alzheimer's Disease Center's "Tri-4-Daddy" fundraiser, and they deeply empathize with young families struggling with frontal lobe dementia.

**Diana Kerwin, MD** is a board certified internist and geriatrician specializing in cognition and dementia. She has an interest in risk factors, biomarkers and the effect of obesity and body weight on cognition and brain function. Dr. Kerwin is involved in clinical trials investigating new therapies for FTD, and the identification of biomarkers and neuroimaging studies for the diagnosis of dementias such as Alzheimer's disease.

**Jennifer Medina, PhD** is currently completing a post-doctoral fellowship in clinical neuropsychology at the University of Illinois at Chicago. She completed her PhD at Northwestern University Feinberg School of Medicine where her clinical and research training were focused on working with individuals with neurodegenerative disorders. Her research specifically examined mood disorders experienced by individuals diagnosed with Primary Progressive Aphasia.

**Darby Morhardt, MSW, LCSW** is Research Associate Professor, Director of Education and Social Worker for the CNADC at Northwestern. She has 25 years clinical experience with persons with dementia and their families. Her research interests focus on the evaluation of quality of life enrichment programs for patients and families.

**Mary O'Hara, AM, LCSW** is a social worker and the Assistant Director of Education at Northwestern's Cognitive Neurology and Alzheimer's Disease Center (CNADC). In addition to helping facilitate the CNADC's Quality of Life Enrichment Programs, Mary also provides education and support to diagnosed persons and their families in the Memory Health Clinic.

**Kristen Oshyn** is a social work intern at the Cognitive Neurology and Alzheimer's Disease Center. She is currently a second-year MSW candidate in the Older Adults program of study at the University of Chicago's School of Social Service Administration. Kristen plans to work with older adults and their families when she graduates.

**Julia Rao, MS** joined the CNADC in September 2008 as a graduate student in Northwestern's Clinical Neuropsychology program and works in the lab of Dr. Sandra Weintraub. She is a graduate of psychology from the University of Wisconsin-Madison and is now involved with projects researching Primary Progressive Aphasia as well as SuperAging.

*Caregiver Group Facilitators, continued*

**Deborah Reed, MD** is Assistant Professor of Clinical Psychiatry and Behavioral Science, a college mentor, and the former Division Director of Geriatric Psychiatry at Northwestern University Feinberg School of Medicine. She is also the Consulting Psychiatrist at the Cognitive Neurology and Alzheimer's Disease Center (CNADC). Her interests include the pharmacologic management of cognitive and behavioral syndromes in patients with dementia, attention deficit disorder and other neurologic conditions.

**Mary Beth Riedner** and her husband Steve have been married for 38 years. Steve was diagnosed with Primary Progressive Aphasia in the fall of 2006. They have three grown children and two adorable grand-daughters. Mary Beth retired in 2008 from Roosevelt University where she served as University Librarian for nine years.

**Jaimie Robinson, MSW, LCSW** is the Resource Navigator and Young Onset Coordinator at the Neurobehavior and Memory Clinic of the Cognitive Neurology and Alzheimer's Disease Center. In addition to providing patients and families with symptom specific ideas, interventions and resources, she provides education and support for patients and families living with a young onset form of dementia.

**Emily Rogalski, PhD** is an Assistant Research Professor at the CNADC. Dr. Rogalski is a neuroscientist whose research has focused for more than seven years on the language-based dementia PPA, including identifying risk factors, genetic features and structural neuroimaging markers of the syndrome.

**Lorene Schlie** is a mother and grandmother, married 43 years to Ron, who was diagnosed with FTD March 2006. She is employed full time and is Ron's advocate and caregiver. In March 2008, she started an FTD support group with eight people that has grown to fifteen plus caregivers and now includes other dementias.

**HyungSub Shim, MD** is a behavioral neurology fellow at the Cognitive Neurology and Alzheimer's Disease Center and Department of Neurology of Northwestern University Feinberg School of Medicine. He earned his medical degree at the Medical College of Wisconsin, and did his neurology residency at the University of Iowa.

**Sandra Weintraub, PhD** is Professor of Psychiatry, Neurology and Psychology at Northwestern University Feinberg School of Medicine and the Director of Neuropsychology in the CNADC. Her research focuses on the neuropsychology of primary progressive aphasia and frontotemporal dementia.

**Christina Wieneke** joined the CNADC in late 2006. She is the research project manager for the Language in PPA research study and a facilitator of the PPA Education and Support Group. She received her bachelor's degree in Behavioral Neuroscience from Purdue University in 2006.

**Kristine Zachrich** has been with the CNADC since August 2009 and is a Research Study Programs Coordinator. She received her bachelor's degree in Communication Disorders from Bowling Green State University in 2008.



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# ABOUT THE CNADC

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# WHO WE ARE

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## Cognitive Neurology and Alzheimer's Disease Center Northwestern University Feinberg School of Medicine

### **Mission:**

The Cognitive Neurology and Alzheimer's Disease Center (CNADC) is a multidisciplinary organization dedicated to the following pursuits:

1. Conducting research to discover how the brain coordinates cognitive functions such as memory, language, attention, and emotion.
2. Discovering causes and treatments for diseases that disrupt these functions, such as Alzheimer's disease, frontotemporal degeneration and primary progressive aphasia.
3. Transferring the benefits of this research to patients and their families.
4. Training researchers and clinicians who want to work in this field.

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*320 East Superior Street, Searle 11th Floor, Chicago, IL 60611*  
*Phone: 312-908-9339, Fax: 312-908-8789, CNADC-Admin@northwestern.edu*  
*<http://www.brain.northwestern.edu>*

## *Dedicated to*

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### **Patient Care**

Neurobehavior and Memory Clinic  
 Northwestern Medical Faculty Foundation  
 676 North St. Clair Street, Suite 945  
 Chicago, IL 60611  
 312-695-9627

### **Research and Education**

Alzheimer's Disease Center  
 320 E. Superior Street, Searle 11  
 Chicago, IL 60611  
 312-926-1851  
[www.brain.northwestern.edu](http://www.brain.northwestern.edu)

## *Making a Difference Every Day*

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### **Research directions**

- Research programs for people with and without dementia
- Clinically testing new drug therapies for Alzheimer's disease (AD) and Frontotemporal Degeneration (FTD)
- Studying the effect of sleep deficits on memory and cognitive function
- Measuring the impact of stress on AD's and dementia's progression
- Causes and treatments of Primary Progressive Aphasia (PPA), FTD, and other early onset dementia
- Identifying brain factors associated with "SuperAging"
- Treatment and prevention of dementia diseases
- Nature of cognitive and behavioral changes in dementia

### **Programs that inform and support**

- Annual Alzheimer Day
- The Buddy Program™
- The Memory Ensemble™
- Early-Stage Memory Loss Education and Support Group
- FTD/PPA Caregiver Conference
- FTD and PPA Caregiver Support Group
- Spousal/Partner Support Group
- Support Group for Adult Children
- Younger Onset AD Education and Support Group
- PPA Patient Support Group
- Mindfulness Practice for Stress and Depression

### **Community outreach and partnerships**

- Alzheimer's Association - Greater Illinois Chapter
- Association for Frontotemporal Degeneration
- National Aphasia Association
- Coalition of Limited English-Speaking Elderly
- Erie Family Health Center
- Francis J. Atlas Regional Senior Center of the Chicago Department of Senior Services
- South Side Dementia Consortium

# NEUROBEHAVIOR AND MEMORY HEALTH CLINIC

## NORTHWESTERN MEDICAL FACULTY FOUNDATION

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### Care for Patients and Families:

The Neurobehavior & Memory Clinic is designed to meet the needs of persons experiencing memory loss or other symptoms of dementia, and their families.

### Services Include:

- Evaluation and follow-up care by behavioral neurologists who specialize in the diagnosis and treatment of dementia syndromes
- Evaluation of memory and other thinking abilities with the use of specialized tests given by a clinical neuropsychologist
- Management of medication for memory disorders
- The opportunity to participate in clinical research and clinical drug trials
- Psychiatric evaluation and treatment for mood and behavior disorders associated with neurological disease
- Education and counseling for patients and families
- Symptom specific interventions and strategies
- Information and referral to other supportive services

### A Dedicated Clinical Team

#### Behavioral Neurologists

M.-Marsel Mesulam, MD, Director  
 Darren R. Gitelman, MD  
 Jay Gottfried, MD, PhD  
 Jaime Grutzendler, MD  
 Joseph Cooper, MD  
 HyungSub Shim, MD

#### Geriatrician

Diana Kerwin, MD

#### Neuropsychiatrist

Deborah Reed, MD

#### Resource Navigator & Young Onset Coordinator

Jaimie Robinson, MSW, LCSW

#### Neuropsychologists

Beth Borosh, PhD  
 Nancy Johnson, PhD, ABPP-CN  
 Sandra Weintraub, PhD, ABPP-CN

#### Social Workers

Darby Morhardt, MSW, LCSW  
 Mary O'Hara, AM, LCSW

#### Clinic Manager

Cori Malone

#### Patient Access Representative

Anthony Nowaske

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676 North St. Clair Street, Suite 945, Chicago, IL 60611  
 Phone: 312-695-9627, Fax: 312-695-6072

# CURRENT FTD/PPA RESEARCH AT NORTHWESTERN

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**Language in Primary Progressive Aphasia:** The purpose of the study is to gain a better understanding of the progression and characterization of primary progressive aphasia (PPA). To do this, we look at many different aspects of the disease: neurological, defined by a clinical exam and MRI; neuropsychological, defined by a large battery of tests examining memory, attention, naming, and others; and linguistic, measured through picture, word, and sentence comprehension.

The study lasts three days total, about 7 hours each day, including breaks. The individual diagnosed with PPA and their study partner are compensated for travel, meals, and accommodations. Participants also receive a daily stipend for their time.

To participate, one must have a diagnosis of PPA, be a native English speaker, have the ability to read large print, have adequate hearing to follow conversation, have no significant medical illness that would interfere with future participation, safe for an MRI, and may not be claustrophobic. For more information, contact Christina Wieneke: 312-908-9681 or [c-wieneke@northwestern.edu](mailto:c-wieneke@northwestern.edu).

**Memory Disorders Research Core:** The purpose of the research core is to better understand various dementia syndromes, including Alzheimer's disease, frontotemporal degeneration, primary progressive aphasia, and other related disorders. This project supports many different research studies on aging and dementia as well as helping to understand the needs of diagnosed individuals and families to provide improved counseling, education, and referrals to community services.

Tasks include an interview with the diagnosed individual and family members, a series of paper and pencil tests to evaluate memory and thinking skills, and a meeting with a social worker. Each research visit is approximately 1.5 hours in length. There is no cost for participation. For more information, contact Mallory Swift: 312-926-1851 or [memoryresearch@northwestern.edu](mailto:memoryresearch@northwestern.edu).

**The Utility of Namenda® in the Treatment of Frontotemporal Dementia:** Researchers are evaluating the use of memantine (Namenda®) in those with a diagnosis of frontotemporal dementia (FTD). Memantine is currently an FDA approved treatment for Alzheimer's disease. Evidence from studies of persons with Alzheimer's disease suggests memantine may reduce damage to brain cells and therefore may also be effective in people with FTD. This study is designed to evaluate the safety and tolerability of memantine in FTD and to evaluate whether memantine will slow the rate of decline in thinking and in problem behaviors in FTD.

Participation in the 26-week study will include 6 office visits two of which will include a blood draw and ECG. Study participants will be randomized to either a placebo group or a treatment group. At each office visit, there will be testing of memory and thinking skills, and at certain visits there will be a physical and neurological exam. For more information, contact Kristine Lipowski at 312-503-2486, or email at [k-lipowski@northwestern.edu](mailto:k-lipowski@northwestern.edu).

All eligible participants must:

- Have a diagnosis of FTD
- Have a reliable caregiver to assist with medication and study visits
- Be between the age of 40 to 80
- Not currently taking a prescription of memantine, donepezil (Aricept®), rivastigmine (Exelon®), or galantamine (Razadyne®)

# THE IMPORTANCE OF BRAIN DONATION

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## **Brain donation is one of the most important contributions to research.**

As researchers work to better understand disorders that affect mental function with aging, brain donations are essential to their progress.

By studying the anatomy, pathology, and chemistry of the brains of people with memory problems or cognitive disorders, we are able to expand our knowledge of diseases such as Alzheimer's and Frontotemporal Lobar Degeneration and take steps toward prevention and treatment.

While major advances have already been made possible through the generosity of brain donation, there is still much more to be learned and a need for continued support.

## **Brain donation provides a valuable service to families.**

A comprehensive brain autopsy is performed on each person who is in our Alzheimer's Disease Core Center study and makes a brain donation to our Center. The family of the donor receives a full report detailing the neuropathologist's findings. At present, neurodegenerative diseases can only be diagnosed with 100% certainty through a brain autopsy, so families are provided with a definitive diagnosis.

Such information is useful if other family members develop a problem with memory or thinking in the future or if there is a known family history.

Making this generous donation provides the family with a way to potentially help others, which can create a sense of hope and power over the illness that affected their loved one.

## **Make the decision to be a brain donor with your family.**

The decision to become a brain donor requires careful thought and planning. As you and your family consider making this important contribution, please keep in mind that the bereavement period is not the optimal time to begin planning for a brain autopsy. It is best to make arrangements as far in advance as possible, even though death may be years away. There are several things that you and your family can do to prepare in advance.

Begin talking about brain donation with your family now. Early discussion can reduce stress at the time of death.

Brain donation is a private matter. Northwestern's Alzheimer's Disease Center respects the decisions of each individual and his or her family. Our Autopsy Coordinator is available to assist you and your family during the decision-making process. Our staff can be reached Monday through Friday, from 8am to 5pm.

(312) 926-1851 or [memoryresearch@northwestern.edu](mailto:memoryresearch@northwestern.edu)

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# DISEASE INFORMATION

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## FREQUENTLY ASKED QUESTIONS ABOUT PRIMARY PROGRESSIVE APHASIA

### **What is Primary Progressive Aphasia (PPA)?**

- ◆ Progressive difficulties in word-finding, word usage, word comprehension or spelling leading to a diagnosis of PPA. Memory for recent events, finding personal objects, recognizing faces and the essential features of personality remain mostly intact.
- ◆ Most people with PPA are in their 50s and 60s at onset but it can affect adults at any age.

### **What causes PPA?**

- ◆ PPA arises when parts of the brain that control language malfunction. The underlying diseases are called “degenerative” because they involve nerve cell death but cannot be attributed to clear-cut causes such as head trauma, stroke or cancer.

### **Why have I never heard of PPA?**

- ◆ PPA is a relatively rare form of dementia that affects many fewer people than the most common type of dementia, Alzheimer’s Disease.
- ◆ Because PPA affects individuals at a younger age than most with dementia, doctors may not recognize the features and misdiagnose the individual with another condition. Increasing awareness about PPA in the medical community will facilitate more efficient accurate diagnoses.

### **How does PPA progress?**

- ◆ The progression is variable and unfolds over many years. Word finding and word comprehension become more difficult over time. Sometimes additional problems can arise in personality (irritability, apathy or inappropriate behaviors), problem solving, memory or dexterity.

### **How is PPA different from Alzheimer’s Disease (AD)?**

- ◆ In scientific language, the term AD denotes a specific pattern of abnormalities in the brain. These can only be detected by microscopic examination after death. In the vast majority of cases, AD starts by affecting the parts of the brain that control short-term memory so that the symptoms consist of forgetfulness instead of word-finding difficulty.
- ◆ In about 30% of people who receive a diagnosis of PPA, the examination of the brain after death shows changes of AD. In such cases, PPA can be considered an atypical (unusual) form of AD.
- ◆ In the majority of people with PPA, the brain shows changes of Frontotemporal Lobar Degeneration (FTLD), also known as Frontotemporal Degeneration (FTD). FTLD and AD are two completely different diseases of nerve cells even though they both may cause PPA.
- ◆ Some types of PPA are more likely to be caused by AD, others by FTLD. Researchers are working to identify reasons for this.

### **How is PPA different than aphasia caused by a stroke or brain injury?**

- ◆ Aphasia due to stroke or brain injury often suddenly develops. In contrast, people with PPA develop language difficulties slowly over time.
- ◆ People with aphasia from stroke or brain injury can often improve with speech therapy, while people with PPA continue to worsen.

## **FREQUENTLY ASKED QUESTIONS ABOUT PRIMARY PROGRESSIVE APHASIA, CONTINUED**

### **Is there treatment for PPA?**

- ◆ There is no medical treatment that has been proven to work for PPA.
- ◆ Because of the 30% probability of AD, some physicians will prescribe AD drugs such as Exelon, Razadyne, Aricept or Namenda. None have been shown to improve PPA.
- ◆ Speech therapy may offer benefits in the early stages by teaching more effective communication strategies and ways to compensate for language difficulties.
- ◆ Quality of life enrichment and support programs offer individuals and families ways of coping with a diagnosis of PPA.

### **Is PPA hereditary?**

- ◆ PPA can be caused by hereditary forms of FTLD. However, it is extremely rare for more than one member of the same family to have PPA.

### **What are some features of PPA?**

- ◆ Slowed or halting speech
- ◆ Word-finding hesitations
- ◆ Sentences with abnormal word order
- ◆ Substitution of words (e.g., “table” instead of “chair”) or words that are incomprehensible
- ◆ Difficulty understanding conversation topics despite normal hearing
- ◆ Inability to think of names, even though the person is recognized
- ◆ New impairments in spelling

### **Does the diagnosis of PPA mean the end of an active life?**

- ◆ People with PPA usually have to make major adjustments at work since almost all professions are heavily dependent on verbal communication.
- ◆ Many people with PPA remain independent for many years, participate fully in social and civic activities, travel widely and take up novel hobbies ranging from gardening to square dancing, painting, carpentry, etc.

### **Where can I go for more information?**

- ◆ Northwestern University Cognitive Neurology and Alzheimer’s Disease Center: [www.brain.northwestern.edu](http://www.brain.northwestern.edu)
- ◆ IMPPACT, the International PPA Connection: [www.ppaconnection.org](http://www.ppaconnection.org)
- ◆ The Association for Frontotemporal Degeneration: [www.ftd-picks.org](http://www.ftd-picks.org)
- ◆ The National Aphasia Association: [www.aphasia.org](http://www.aphasia.org)

## FREQUENTLY ASKED QUESTIONS ABOUT FRONTOTEMPORAL DEGENERATION

### What is Frontotemporal Degeneration (FTD)?

- ♦ FTD is a condition characterized by gradual changes in behavior, personality, reasoning, and problem solving.
- ♦ Most people with FTD are in their 50s and 60s at onset but it can affect adults at any age.

### What causes FTD?

- ♦ FTD arises when parts of the brain that control behavior and personality malfunction. The underlying diseases are called “degenerative” because they involve nerve cell death but cannot be attributed to clear-cut causes such as head trauma, stroke, or cancer.
- ♦ Specifically in FTD, cells in the frontal lobe (the area of the brain which controls behavior, judgment, personality and emotion) begin to die.

### Why have I never heard of FTD?

- ♦ FTD is a rare form of dementia that affects fewer people than the most common type of dementia, Alzheimer’s disease. However, frontotemporal degeneration is one of the most common dementia diagnoses of people under the age of 65.
- ♦ Because FTD affects individuals at a younger age, doctors may not recognize the features and often misdiagnose the individual with mental illness or a psychiatric disorder. Increasing awareness about FTD in the medical community will facilitate more efficient accurate diagnoses.

### How does FTD progress?

- ♦ The progression of FTD is variable and can unfold over many years.
- ♦ While behavior and personality changes are typically the first symptoms, memory and language abilities can change as the disease progresses and spreads to other areas of the brain which control these functions.
- ♦ Eventually, as the illness progresses, a person becomes dependent upon others for all aspects of their care.

### How is FTD different from Alzheimer’s Disease (AD)?

- ♦ In scientific language, the term Alzheimer’s Disease (AD) denotes a specific pattern of abnormalities in the brain. These can only be detected by microscopic examination after death. In the vast majority of cases, AD starts by affecting the parts of the brain that control short-term memory so that the symptoms consist of forgetfulness instead of behavior changes.
- ♦ The abnormalities found in the brains of individuals with FTD are more complex since there are at least three different disease proteins implicated in FTD pathology: tau, TDP-43 and FUS. When tau is responsible for the disease process the FTD pathology the term FTLT-TAU is used to describe the pathology. Tau abnormalities can cause Pick’s disease, corticobasal degeneration and progressive supranuclear palsy. The terms FTLT-TDP or FTLT-FUS are used to describe the pathology in FTD when abnormal TDP-43 or FUS proteins accumulate in nerve cells. Therefore people may have the same clinical diagnosis of FTD but different pathologic diagnoses (e.g., FTLT-TDP or FTLT-TAU or FTLT-FUS). Pathologists are still working to understand these pathologies; therefore, terminology may change as we learn more about this complex condition.

## **FREQUENTLY ASKED QUESTIONS ABOUT FRONTOTEMPORAL DEGENERATION, CONTINUED**

- ◆ In the early stages of FTD, there is usually no true memory loss of the type that is seen in Alzheimer's dementia. Instead, there are changes in personality, ability to concentrate, social skills, motivation and reasoning.

### **How is FTD different than dementia caused by a stroke or brain injury?**

- ◆ Dementia due to stroke or brain injury develops suddenly. People with FTD develop changes slowly over time.
- ◆ People with stroke or brain injury can improve with therapy. People with FTD continue to worsen.
- ◆ According to the Association for Frontotemporal Degeneration (AFTD), there has been no evidence to date investigating the connection between traumatic brain injury and FTD. Frontal dysfunction is common in traumatic brain injury and can result in symptoms similar to FTD.

### **Is there treatment for FTD?**

- ◆ There is no approved medical treatment that has been proven to work for FTD.
- ◆ Some physicians will prescribe AD drugs such as Exelon, Razadyne, Aricept or Namenda. It is not known if these medications are of any benefit.
- ◆ Quality of life enrichment and support programs offer individuals and families ways of coping with a diagnosis of FTD.
- ◆ Medications may be helpful with certain features of the disorder, e.g. some types of antidepressants may reduce the behavior problems associated with FTD.
- ◆ The Northwestern CNADC actively conducts research and clinical trials to improve the diagnosis and treatment of FTD. Your participation helps us reach those goals. Please go to the CNADC website ([www.brain.northwestern.edu](http://www.brain.northwestern.edu)) to learn more about current research opportunities.

### **Is FTD hereditary?**

- ◆ The majority (50-70%) of cases FTD are sporadic, meaning the disorder develops in that person by chance rather than being inherited and the risk to family members is the same as that of any individual in the general population.
- ◆ According to AFTD, in a very small percentage (5-10%) of FTD patients, a family history suggests a hereditary condition with an autosomal dominant pattern of inheritance. This means there is a clear pattern of FTD-type diagnoses being passed from parent to child, with virtually every patient having an affected parent and each child of an affected person having a 50% chance to inherit the disorder.
- ◆ In about 20-40% of FTD cases the disorder appears to be "familial", meaning that it is not directly inherited but members of the family are at elevated, though undetermined, risk for developing the disorder.

### **What are some features of FTD?**

- ◆ Impairments in social skills and inappropriate or bizarre social behavior
- ◆ Lack of awareness (insight) about the changes
- ◆ Change in activity level; apathy, withdrawal, loss of interest, lack of motivation
- ◆ Decreased judgment in financial decisions, impulsive spending

## ***FREQUENTLY ASKED QUESTIONS ABOUT FRONTOTEMPORAL DEGENERATION, CONTINUED***

- ◆ Changes in personal habits; lack of concern over personal appearance
- ◆ Alterations in personality and mood
- ◆ Changes in one's customary emotional responsiveness
- ◆ A lack of sympathy or compassion in someone who was typically responsive to others

### **Does the diagnosis of FTD mean the end of an active life?**

- ◆ People with FTD usually have to make adjustments or leave their jobs, since all professionals are heavily dependent on interpersonal communication, appropriate behaviors and sound judgment.
- ◆ Many people with FTD remain active for many years. While they may need additional supervision, they can continue to participate fully in social and civic activities.

### **Where can I go for more information?**

- ◆ Northwestern University Cognitive Neurology and Alzheimer's Disease Center (CNADC): [www.brain.northwestern.edu](http://www.brain.northwestern.edu)
- ◆ The Association for Frontotemporal Degeneration: [www.ftd-picks.org](http://www.ftd-picks.org)

# Clinical Recommendations for Families and Individuals Diagnosed with Primary Progressive Aphasia and Frontotemporal Dementia

Jennifer Medina, BS, Sara Banks, BSc, and Sandra Weintraub, PhD

PPA is a dementia that causes progressive breakdown of language, and as such the needs of individuals with this diagnosis are unique. FTD is a related disorder that causes a progressive breakdown in behavior. The two disorders often overlap, with PPA patients eventually showing behavioral change and FTD patients showing language symptoms. Here are some suggestions to help FTD and PPA patients and their families improve quality of life:

- 1. Avoiding Depression.** Depression is quite common in PPA, where the patient is usually unaware of their language problem. The family and physician should watch for signs of depression, which include tearfulness, changes in sleeping or eating patterns, irritability, and withdrawal. In some cases, a physician might choose to introduce an antidepressant to boost the patient so they may become more engaged and hopefully feel better.
- 2. Improving Communication.** Language problems can be extremely frustrating, so we recommend introducing alternate modes of communication as early as possible in the disease to help minimize frustration. Many families choose to work with a speech-language pathologist to tailor-make alternative communication strategies for their situation. This may include a communication notebook (pages devoted to family, emergency information, and everyday information such as groceries and prescription details) to help bolster independence and allow the patient to get their point across. We also recommend that all language-impaired patients carry a wallet card with a brief explanation of their condition and pertinent emergency information so they can communicate their situation quickly to anyone they interact with.
- 3. Avoiding Confrontation.** Especially in FTD, where patients may not appreciate that there is anything wrong with them, confrontational situations may emerge. In FTD, poor judgment is common, and inappropriate behaviors such as telling offensive jokes, approaching strangers, sexual disinhibition, and indiscriminate spending are common--leading to difficulty with other family members. If a confrontation emerges, remember not to argue or try to reason with the patient. Try to identify exactly what is causing the situation, and understand triggers or warning signs. Often, changes in volume, tone of voice, or body language can indicate that the patient is upset. Pick your battles, and only intervene in really disruptive cases. Try to keep a sense of humor. Keeping decision-making to a minimum decreases confusion and frustration. Validate feelings and make them feel safe. Finally, certain medications may be introduced to minimize some aberrant behaviors.
- 4. Maximizing Activity.** When an individual has language or behavioral problems, keeping them active can be a challenge, yet it is important to maximize brain health and improve mood. Adult day health and leisure programs provide socialization and structured daily activity. A hired companion who comes to the home may be able to provide some stimulation, help with language practice, or help to get the patient out of the house for some exercise. Nonverbal activities such as listening to music, art activities, spending time with a pet, or completing nonverbal puzzles may all be soothing.
- 5. Caregiver Health.** Finally, caregivers are extremely precious and need to be cared for. Dementia caregiving can be very stressful, and it is important to stay healthy and fit. Make sure you take the time for yourself, recruiting friends, family, and professionals to provide respite. Identify activities that are relaxing and fun for you, and make time for them. Your mood is important too--many seek psychotherapy to help them cope. Support groups are available through Northwestern, the Alzheimer's Association, and the community, and many find such groups useful.

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# THE FTD/PPA MEDICAL CARE TEAM

Assembling a care team is important when someone in the family has a diagnosis of FTD or PPA. Together, your team can help you navigate the various questions, concerns and uncertainties that can affect the well-being and quality of life for the person and the family. Below is a list of possible Medical Care Team members. Keep in mind not all of the professionals listed below may be available in your area, covered by your health insurance, or familiar with FTD or PPA.

**Primary Care Team:** This team can consist of a medical doctor, nurse practitioner, and physician's assistant who will provide ongoing care and treatment for a variety of common medical conditions. When your primary care team suspects an illness outside the scope of their practice, they will refer you to a specialist. This team will still be in charge of your overall care even if you see a specialist for a particular condition.

**Neurologist:** This clinician is trained to identify and treat illnesses related to the nervous system and is typically the first to diagnosis FTD and PPA. A behavioral neurologist is specially trained in neurological disorders that affect cognition. Behavioral neurologists are most often found in university health care settings.

**Neuropsychologist:** This clinician evaluates a person's cognitive abilities using specialized paper and pencil tests which pinpoint the exact areas of cognition that are affected and to what degree. These tests help the neurologist either make a diagnosis or understand more specifically what may be causing the symptoms.

**Psychiatrist:** This clinician specializes in evaluating behaviors and moods of individuals. A psychiatrist may prescribe medications to modify challenging behaviors and moods specific to the diagnosis that are otherwise unmanageable. A special type of psychiatrist, called a neuropsychiatrist or geriatric psychiatrist, is specially trained in treating psychiatric problems in neurological disorders.

**Speech and Language Pathologist:** This clinician sees individuals with language changes. They evaluate different aspects of language in detail and can make recommendations and offer strategies to improve communication. Family members should be included in parts of the treatment sessions so they can receive education on how to facilitate communication.

**Social Worker:** This clinician can help you and your family by providing local resources, helpful information about your particular diagnosis; in addition to, counseling and support. Some local resources include home care agencies, long-term care options, adult day centers, support groups, specialized programs, meaningful activities & financial/disability resources.

**Occupational Therapist:** This clinician works with individual patients and their family members to improve or maintain the patient's daily functioning and reduce the burden on the family caregiver by developing ways to modify or adapt activities of everyday life. This can be particularly helpful for patients with movement or motor changes.

**Palliative Care Team:** Health care practitioners on this team promote comfort and dignity for anyone experiencing a terminal illness and can be implemented as early as the first day the diagnosis is made. This care focuses on comfort and symptom relief. Hospice care, a Medicare benefit, is a form of palliative care and is implemented when a person is expected to live for six months or less.

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**“PERSPECTIVES ON  
FTD AND PPA”**

**BRADLEY BOEVE, MD**

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## Perspectives on Frontotemporal Dementia and Primary Progressive Aphasia

COGNITIVE NEUROLOGY AND ALZHEIMER'S DISEASE CENTER

FRONTOTEMPORAL DEMENTIA AND PRIMARY PROGRESSIVE APHASIA

CAREGIVER EDUCATION *and* SUPPORT CONFERENCE

Cosponsored by the Association for Frontotemporal Dementias and the National Aphasia Association



**Bradley F. Boeve, M.D.**

**Division of Behavioral Neurology  
Department of Neurology  
Mayo Clinic  
Rochester, Minnesota**

**Alzheimer's Disease Research Center  
Mayo Foundation**

## Disclosures

### Financial/Other

- Investigator for clinical trials sponsored by Cephalon, Inc., and Allon Pharmaceuticals
- Royalties from the publication of a book entitled Behavioral Neurology Of Dementia (Cambridge Medicine, 2009)
- Honoraria from the American Academy of Neurology
- Research support from the NIA, NINDS, and the Alzheimer's Association

### Off-label and/or Investigational Use

- I will discuss use of some medications not FDA-approved for the indications to be reviewed

## Perspectives on FTD and PPA Outline

- Symptoms
- Brain-Behavior Correlations
- Genetics
- Pathophysiology and Future Perspectives
- Resources/Websites

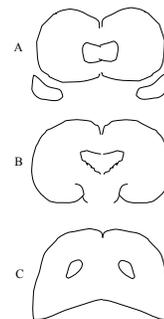
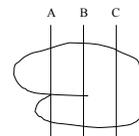
## Symptoms

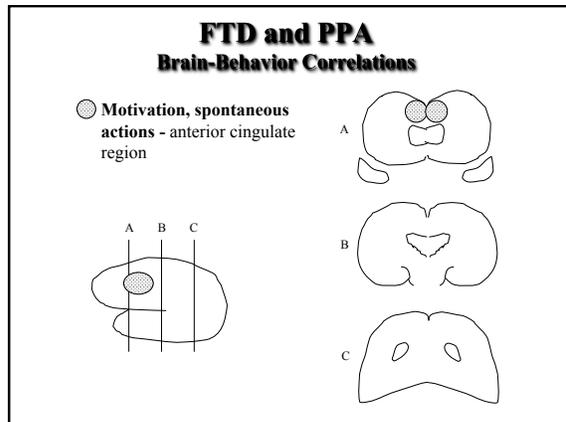
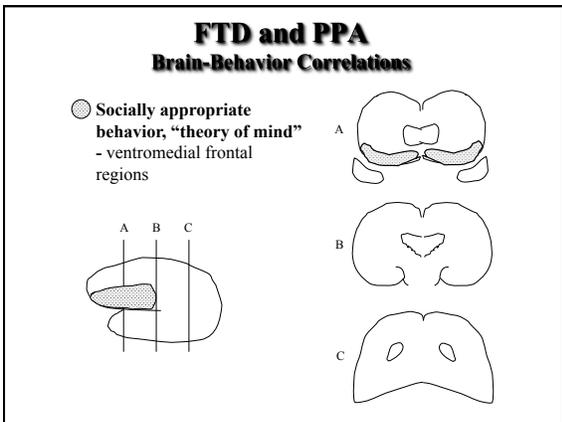
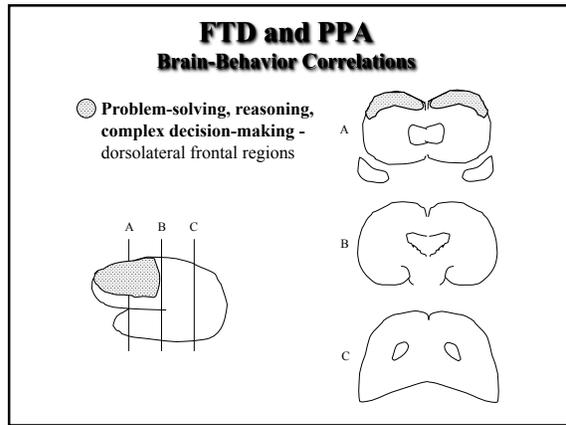
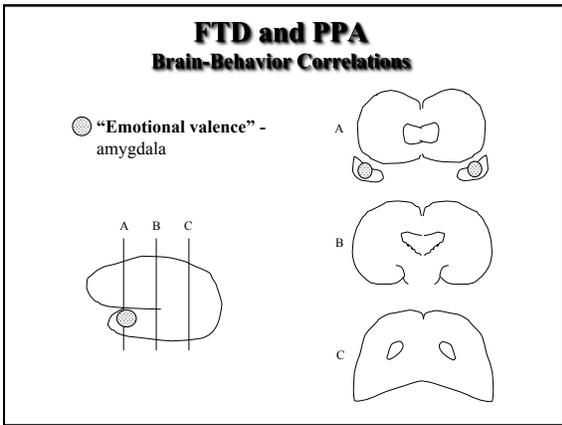
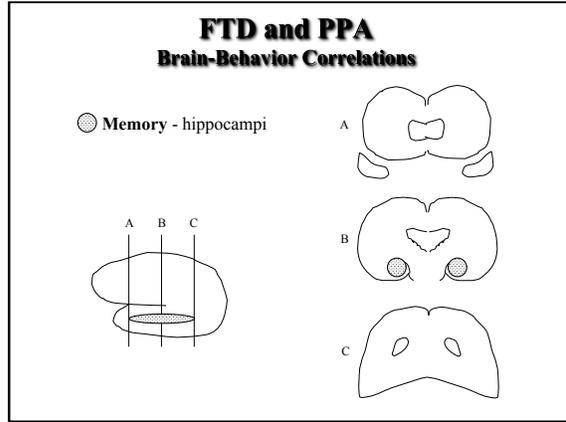
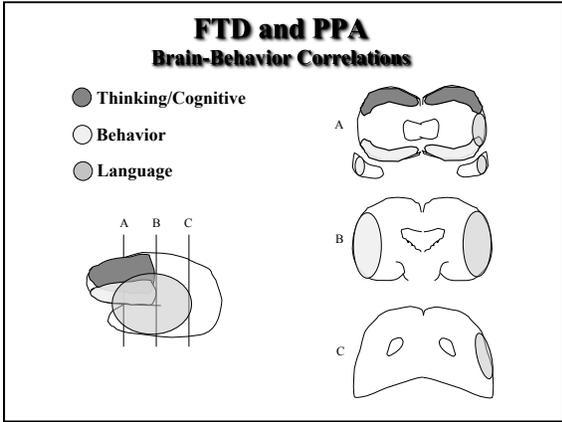
## Patient Examples

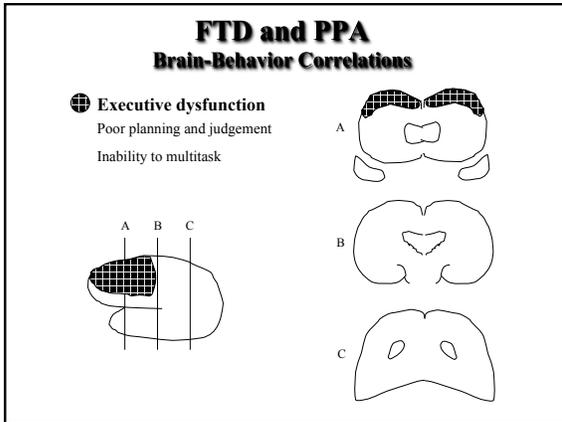
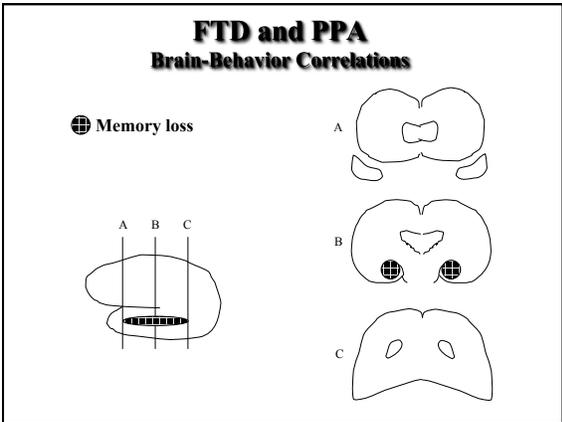
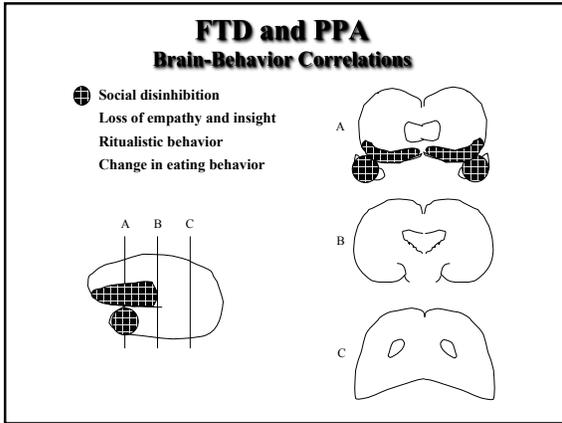
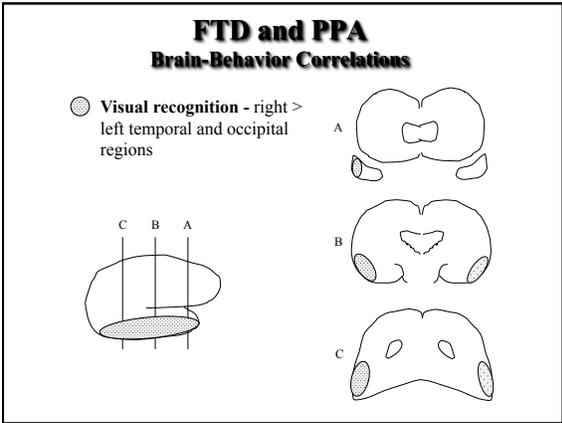
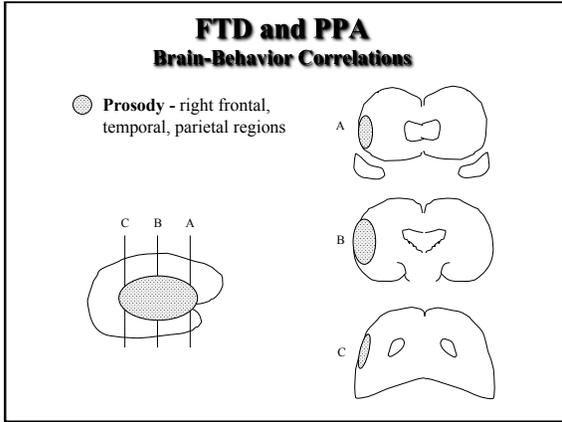
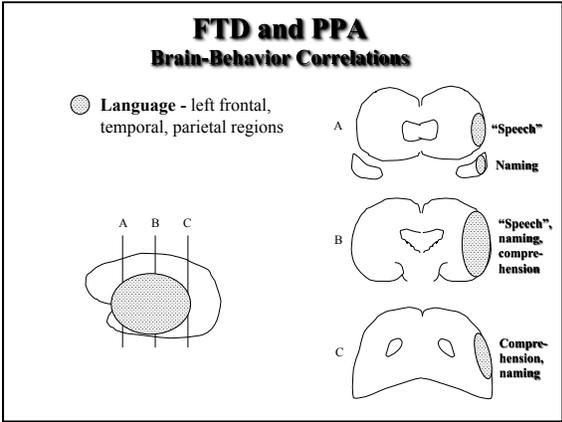
## Brain-Behavior Correlations

## FTD and PPA Brain-Behavior Correlations

*Templates*







### FTD and PPA Brain-Behavior Correlations

**⊕ Loss of motivation**  
Tendency to sit, not initiative conversations or actions

A B C

### FTD and PPA Brain-Behavior Correlations

**⊕ Poor verbal expression**  
**● Poor naming of objects and people**  
**● Poor comprehension and repetition**

A B C

### FTD and PPA Brain-Behavior Correlations

**⊕ Aprosodia**  
Monotone voice  
Unable to interpret inflections in voice another person's voice

C B A

### FTD and PPA Brain-Behavior Correlations

**⊕ Visual agnosia**  
Unable to recognize objects and people

C B A

### FTD and PPA Brain-Behavior Correlations

**FTD**

**Core features**

- Behavioral disinhibition
  - Socially inappropriate behavior
  - Loss of manners or decorum
  - Impulsive, rash or careless actions
- Apathy or inertia
- Loss of sympathy or empathy
- Perseverative, stereotyped or compulsive/ritualistic behavior
- Hyperorality and dietary changes
  - Altered food preferences
  - Binge eating
  - Oral exploration or consumption of inedible objects

Rakovsky et al, International bvFTD Criteria Consortium

### FTD and PPA Brain-Behavior Correlations

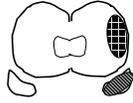
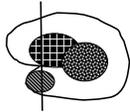
**PPA**

**Core features**

- Most prominent clinical feature is difficulty with language
- These deficits are the principal cause of impaired daily living activities
- Aphasia should be the most prominent deficit at symptom onset and for the initial phases of the disease

Mesulam MM. Ann Neurol 2001  
Gorno-Tempini et al, Neurology 2011

## FTD and PPA Brain-Behavior Correlations



PPA

### Core features

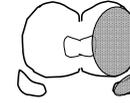
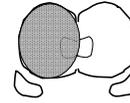
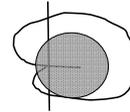
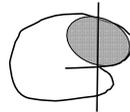
- Most prominent clinical feature is difficulty with language
- These deficits are the principal cause of impaired daily living activities
- Aphasia should be the most prominent deficit at symptom onset and for the initial phases of the disease

### Variants

- Nonfluent/agrammatic
- Semantic
- Logopenic

Mesulam MM. Ann Neurol 2001  
Gorno-Tempini et al, Neurology 2011

## FTD and PPA Brain-Behavior Correlations

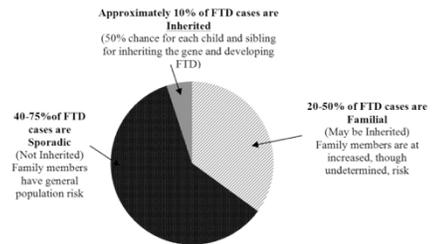


FTD

PPA

## Genetics

## FTD and PPA Genetics



## FTD and PPA Genetics

- ? Inherited form – if you have 1 or more 1<sup>st</sup> degree relatives with dementia and/or parkinsonism and/or ALS
- Genetic considerations:
  - Microtubule associated protein **tau** (clinical testing available)
  - **Progranulin** (clinical testing available)
  - Valosin-containing protein (**VCP**) (clinical testing not yet available)
  - **CHMP2B** (clinical testing not yet available)
  - **TDP-43**
  - **FUS**
  - Chromosome 9 gene not yet found

## Pathophysiology and Implications for Future Therapies

## FTD and PPA Underlying Disorders

### FTLD - the pathologies

- Pick's disease
- Corticobasal degeneration
- Progressive supranuclear palsy
- Multisystem tauopathy
- Frontotemporal dementia with Parkinsonism associated with mutations in microtubule associated protein tau on chromosome 17 (FTDP-17*MAPT*)
- Frontotemporal lobar degeneration (FTLD) with motor neuron disease (MND)
- Frontotemporal lobar degeneration (FTLD) with ubiquitin/TDP-43-positive inclusions
- Frontotemporal dementia with Parkinsonism associated with mutations in progranulin on chromosome 17 (FTDP-17*PGRN*)
- Dementia lacking distinctive histopathology (DLDH)

Alzheimer's Disease

## FTD and PPA

### Pathophysiology and Implications for Future Therapies

#### Tauopathies

- Pick's disease
- Corticobasal degeneration
- Progressive supranuclear palsy
- Argyrophilic grain disease
- Frontotemporal dementia with Parkinsonism associated with mutations in microtubule associated protein tau on chromosome 17 (FTDP-17*MAPT*)

**tau**

#### TDP-43opathies

- Frontotemporal lobar degeneration (FTLD) with motor neuron disease (MND)
- Frontotemporal lobar degeneration (FTLD) with ubiquitin/TDP-43-positive inclusions
- Frontotemporal dementia with Parkinsonism associated with mutations in progranulin on chromosome 17 (FTDP-17*PGRN*)

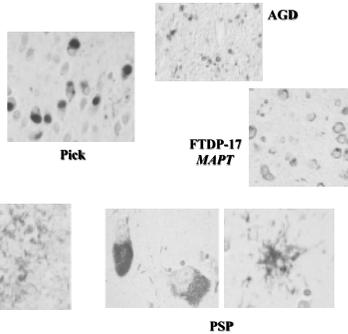
**TDP-43**

## FTD and PPA

### Pathophysiology and Implications for Future Therapies

#### Tauopathies

- Pick's disease
- CBD
- PSP
- AGD
- FTDP-17-*MAPT*

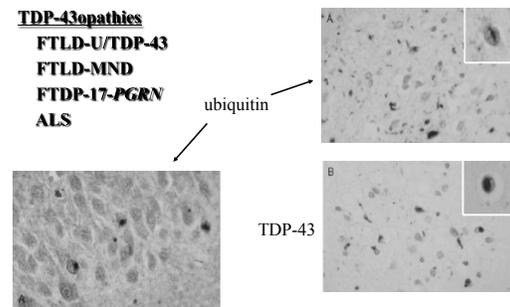


## FTD and PPA

### Pathophysiology and Implications for Future Therapies

#### TDP-43opathies

- FTLD-U/TDP-43
- FTLD-MND
- FTDP-17-*PGRN*
- ALS



## FTD and PPA

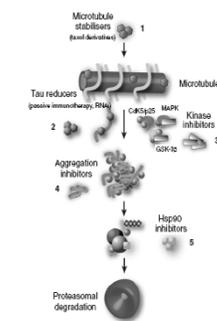
### Pathophysiology and Implications for Future Therapies

Tauopathies

TDP-43opathies

## FTD and PPA

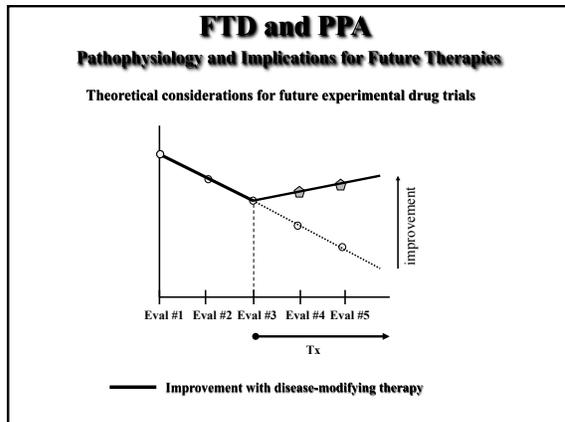
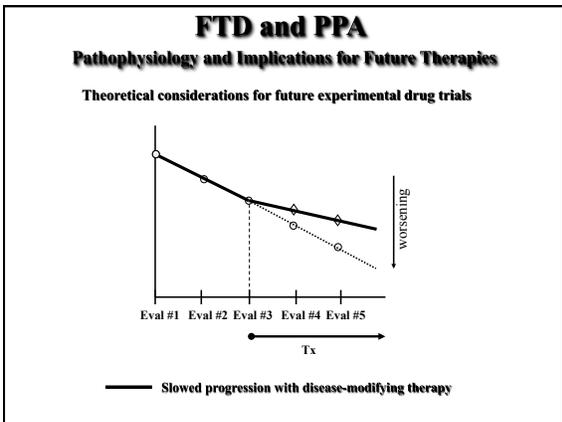
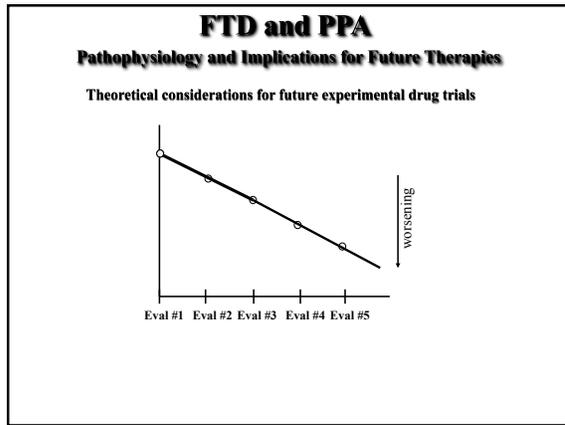
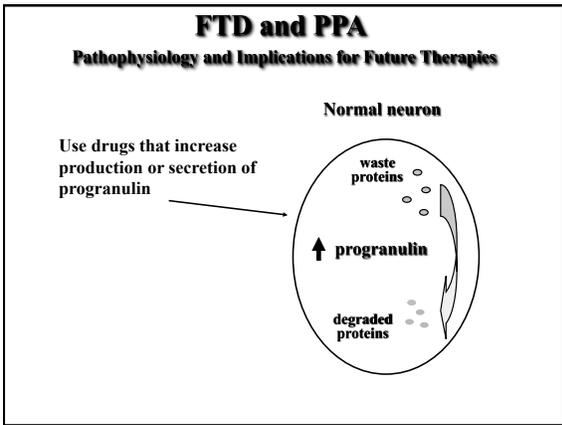
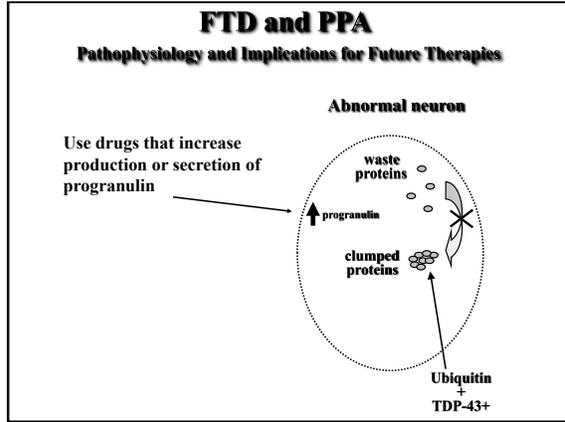
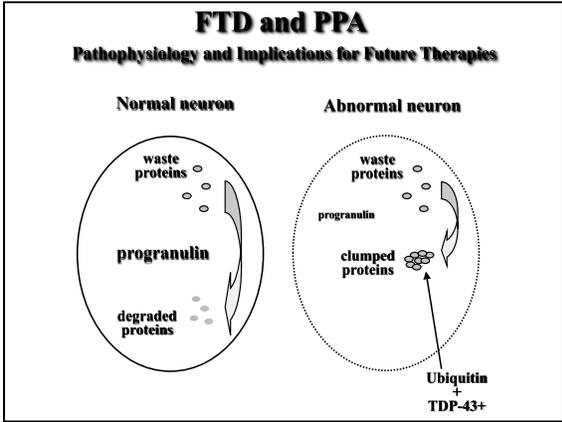
### Pathophysiology and Implications for Future Therapies



#### ? Tauopathy therapies

- Microtubule stabilizers
- Tau reducers (vaccine)
- Tau kinase inhibitors
- tau aggregation inhibitors
- Hsp90 inhibitors

Dickey & Petrucelli, Expert Opin Ther Targets 2006



**FTD and PPA**  
**Pathophysiology and Implications for Future Therapies**

Without knowledge about the natural history of patients with FTD and PPA, we will not know where to start when promising medications are identified/developed

**This is one of many reasons involvement in research studies are so critical**

**FTD and PPA**  
**Pathophysiology and Implications for Future Therapies**

*If you participate in research - thank you!!*

*If you are interested in participating in research – please contact any of the staff at this conference*

**FTD and PPA**  
**Resources - Websites**



**Cognitive Neurology and Alzheimer's Disease Center**

<http://www.brain.northwestern.edu/index.html>



The Association for Frontotemporal Degeneration  
Opening the gateway to help and a cure

<http://www.theaftd.org>

**FTD and PPA**  
**Resources - Websites**



The Association for Frontotemporal Degeneration  
Opening the gateway to help and a cure

**FTD/PPA Genetics**  
<http://www.theaftd.org/frontotemporal-dementias/genetics>



**The Frontal Lobes**  
**Scientific American Frontiers**  
**Make Up Your Mind**  
<http://www.pbs.org/saf/1302/>



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# AFTERNOON SESSION HANDOUTS

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# NEWLY DIAGNOSED CHECKLIST

## FOR INDIVIDUALS WITH FRONTOTEMPORAL DEGENERATION (FTD)

- *Tell Someone.* Share this life-changing event with others who can support you.
- Educate yourself about the diagnosis. Become knowledgeable, but don't obsess over it.
- Acknowledge that your life will be very different and that the person cannot control their behavior.

### Legal and Financial

- Meet with an Elder Law Attorney to discuss:
  - ◊ Durable Power of Attorney for Health Care and Property
  - ◊ Legal and financial planning
  - ◊ Social Security Disability
  - ◊ Veteran's benefits, if applicable
- If possible, begin discussions about future care wishes and options.
- Find out about financial supports available through caregiver tax credits or prescription drug coverage.
- Inquire about Short Term and Long Term Disability Benefits through diagnosed individual's employer.

### Support

- Join a support group or seek out individual counseling.
- Find out about local programs and services for the person and your family through the local offices on aging and disability.
- Take time for yourself and ask others to help you commit to this promise.
- Inquire about Family Medical Leave Act (FMLA) benefits for family members who may take time from work to care for the diagnosed individual.

### Safety

- Consider a Medical Alert Safe Return bracelet for you and the person with FTD.
- Be watchful of day-to-day activities (checkbook, paying bills, eating, going places alone, driving, hygiene) and regularly evaluate the safety of leaving the person alone.
- Be aware of their decision making or ideas so that you can intervene if necessary.
- Carry with you a card that states "This person has FTD."

### Quality of Life

- Take lots of pictures with family and friends.
- Establish structure and routine in their day.
- Adapt to their behaviors and needs; don't argue or try to change their behavior - distract instead.
- Help them continue to enjoy their favorite activities as long as they can.
- Help them maintain their dignity and focus on their strengths.
- Exercise together.

### Medical Care

- Find a doctor who understands FTD and see yourself as a partner with this doctor and medical team.
- Begin a list of questions that you can ask the doctor at the next visit.
- Keep a journal/log to track changes in behavior so the neurologist can better understand the changes you are seeing.
- Ask your doctor about getting involved in research.
- Ask your doctor's office who you can call there when you need help urgently.

# NEWLY DIAGNOSED CHECKLIST FOR PRIMARY PROGRESSIVE APHASIA (PPA)

## SUPPORT

- Tell someone. Share this life-changing event with others who can support you. Consider *how* and *when* to tell family and friends.
- Educate yourself about the diagnosis. Become knowledgeable, but don't obsess over it. Realize each person is affected differently.
- Join a support group that best fits you and your family's needs (online forum, telephone, age-appropriate, face-to-face group).
- Find out about local agencies and services for the person and your family.
- **Take time for yourself** and ask others to help you commit to this promise.
- Join a research study.
- Make a list of ways that friends can help if they ask, "Is there anything I can do?"

## MEDICAL CARE

- Find a neurologist who understands PPA.
  - ◊ See yourself as a partner with the doctor and medical team.
  - ◊ Make a list of questions that you can ask the doctor at the next visit.
  - ◊ Take notes when at the appointments.
  - ◊ Keep a journal of/document changes in behavior and communication between doctor visits.
- Find a speech therapist and start to learn other ways to communicate in addition to speech (gestures, pictures, communication devices), etc.
- Find a psychiatrist, if appropriate.
- Find a social worker that can assist in being an advocate for you.

## COMMUNICATION

- Continue to include the person in conversations but prepare for the conversations to take more time than before.
- Speak clearly and slowly to the person.
- Give simple instructions and use simple sentences. Don't bombard the person with too much information at once.
- Minimize noise and number of people in a conversation at a time.
- Speak face-to-face. Phone conversations may be difficult.
- Wait for the person to finish verbalizing their thought. Don't interrupt unless you have permission. Decide on a strategy that indicates, "I'm not finished talking yet."
- Agree on cues that the person can use in social settings to indicate that they need your help.
- Don't pretend to understand when you don't.

# ***NEWLY DIAGNOSED CHECKLIST***

## ***FOR PRIMARY PROGRESSIVE APHASIA (PPA), CONTINUED***

### **LEGAL AND FINANCIAL**

- Meet with an elder law attorney to discuss:
  - ◊ Durable power of attorney for health care and property
  - ◊ Legal and financial planning
  - ◊ Social Security Disability (PPA is now on the Compassionate Allowances list)
  - ◊ Veteran's benefits, if applicable
- Begin to discuss future care wishes and options, as a family.
- Evaluate options regarding continuing employment to retain health insurance and maintain adequate income. Check with the person's employer about short-term and long-term disability benefits, as well as family-medical leave for yourself.

### **QUALITY OF LIFE**

- Adapt to the person's behaviors and needs. Anticipate changes and frustrations. Use tools to help with day-to-day activities (e.g., ready-made grocery list, prepared card with favorite restaurant order, calendars, etc.).
- Help them adapt their favorite activities so that they can enjoy them for as long as possible.
- Exercise and keep active. Find volunteer opportunities for the person.
- Be on the lookout for signs of depression.
- Help them maintain their independence and dignity. Focus on strengths.
- Keep a daily routine to provide structure.

### **SAFETY**

- Be watchful of day-to-day activities (e.g., checkbook, paying bills, going places alone, driving). Regularly evaluate the abilities of the person and their decision-making skills.
- Don't argue or try to change behaviors unless they pose a danger to the person or others.
- Have the person carry an "I have PPA" card, as well as a card with emergency contact and medical information.

### **COPING**

- Be prepared to take on increased responsibilities that the other person used to handle.
- Seek individual counseling to deal with personal grief, depression or anger.
- Don't be afraid to ask others for help. Don't be embarrassed to accept help from others.
- Keep up with your own doctor visits.

### **RESOURCES/WEBSITES**

- [www.brain.northwestern.edu](http://www.brain.northwestern.edu) - Northwestern University Cognitive Neurology and Alzheimer's Disease Center
- [www.aphasia.org](http://www.aphasia.org) - National Aphasia Association
- [www.ftd-picks.org](http://www.ftd-picks.org) - Association for Frontotemporal Degeneration
- [www.ppaconnection.org](http://www.ppaconnection.org) - IMPPACT, The International PPA Connection
- [www.caregiver.org](http://www.caregiver.org) - Family Caregiver Alliance

# LEGAL PLANNING

Advanced Directives outline a person's future wishes and instruct others how to carry out their wishes after they can no longer make decisions. In cases of dementia, it is important to get these documents in order as soon as possible because the disease progression will affect the person's ability to participate in decision-making. The documents *MUST* be prepared while the person is legally able to execute and understand them, so it is encouraged to make these arrangements early on.

Speaking with an elder law attorney to begin legal planning and making arrangements for future care can reduce anxiety about the future for you and your loved one. An elder law attorney is an expert in the field of legal planning and is knowledgeable in the areas of disability and guardianship.

## TERMS TO KNOW

### **Power of Attorney for Health Care**

This document allows the diagnosed person (agent) to designate another person (principle) to make health care decisions on their behalf in the event the doctor determines they are unable to do so. The power of attorney for health care speaks for the diagnosed person and is legally authorized to act, including withholding or withdrawing life support and making other health care decisions.

### **Power of Attorney for Property**

This document allows the diagnosed person (agent) to designate another (principle) to act for them in financial matters and property transactions in the event a doctor determines they are unable to do so. The power of attorney for property is legally authorized to speak on the diagnosed person's behalf.

### **Living Will**

A Living Will records the diagnosed person's wishes for medical treatment near the end of life. It expresses preferences to forgo life support, in the event it would only prolong the dying process. It does *not* authorize another person to make health care decisions, but communicates preferences to a health care provider.

### **Will**

A Will indicates how the diagnosed person's assets and estate will be distributed. It can also include funeral arrangements.

### **Living Trust**

A Living Trust allows for properties of the diagnosed person to be owned in trust while the person is living. It appoints a designated trustee to control the assets and requires that the trustee manage the property according to the terms of the trust.

# LEGAL PLANNING, CONTINUED

## GUARDIANSHIP

When all other options have been exhausted, in some cases, a person with dementia may need guardianship. Guardianship essentially takes away a person’s rights, so it is a difficult process, but in some cases necessary to protect the person and family. A guardian is a surrogate decision-maker who is appointed by the court to make either personal and/or financial decisions in the best interest of the person. It is very important to speak with a lawyer to determine if guardianship is the right option for you and your family.

## PLANNING FOR LONG TERM CARE COSTS AN OVERVIEW OF MEDICARE VS. MEDICAID

MEDICARE	MEDICAID
Health insurance for seniors age 65+ or persons receiving Social Security Disability Insurance (SSDI).  One is eligible for Medicare 2 years after SSDI benefits begin.	Needs-based health care program for the poor.
Federally controlled, uniform across the country.	Controlled by state, different eligibility criteria and regulations in each state.
Does <i>not</i> cover long-term care costs. Pays for first 20 days of nursing home care after a 3-day inpatient hospital stay if the person has “skilled needs.” Days 21-100, a co-pay is required.	Pays long-term care costs to participating home health and skilled nursing home providers.
Pays for primary hospital care and related medically necessary services and some medications.	Pays for all medications and health care costs if providers accept Medicaid as payment.
Must have contributed to Medicare system to be eligible.	Must meet income and asset limits to be eligible. Guidelines protect well spouses from financial impoverishment.

\*\*If your loved one is eligible for VA Benefits or has a Long-Term Care Insurance Policy, inquire about these additional benefits early on.\*\*

## LEGAL PLANNING, CONTINUED

### WHO CAN HELP

#### 1. National Academy of Elder Law Attorneys

The National Academy of Elder Law Attorneys, Inc. is a professional association of attorneys who specialize in legal services for seniors and people with special needs. To find an attorney in your area, contact: 703-942-5711 or visit [www.naela.org](http://www.naela.org).

#### 2. Financial Planning Association

A financial planner can help you plan for the cost of long term care. To find a financial planner in your area, visit: <http://www.fpanet.org>.

#### 3. Social Security Disability

The Social Security disability program provides financial assistance to people with disabilities who have paid into social security. Both FTD and PPA are on the Social Security list of Compassionate Allowances, which means that individuals with these diagnoses are automatically approved for disability benefits. For more information visit: <http://www.ssa.gov/disability/>, 1-800-772-1213.

#### 4. Medicare

Medicare provides health insurance to those over 65 or persons with Social Security Disability (eligible after a 2 year wait). For information, visit: <http://www.cms.gov/home/medicare.asp>, 800-MEDICARE (800-633-4227)

#### 5. Medicaid

Medicaid is a state run health insurance program for low-income individuals. For information on eligibility requirements by state, visit: <http://www.cms.gov/home/medicaid.asp>. Local phone numbers vary by state.

### TIPS FOR PLANNING

#### Start Discussions Early

It can be especially difficult to make these plans for people with bvFTD who lack insight into their condition. They may resist or even undo arrangements that have been made. In some cases, it is helpful to have the doctor or someone other than family begin this discussion.

#### Review Plans Over Time

Personal situations can change over time. Review plans regularly and update documents as needed.

#### Organize Your Documents

Gather copies of your documents and put them together in a file in a safe place. Make sure someone close to you knows where you keep these papers. Give copies to your lawyer, doctor and family members/friends involved in care.

## LEGAL PLANNING, CONTINUED

### IMPORTANT DOCUMENTS CHECKLIST

DOCUMENT	DATE COMPLETED	LOCATION
Create Power of Attorney for Health Care		
Create Power of Attorney for Property		
Create Will		
Create Living Will		
Do Not Resuscitate (DNR), if applicable		

### LEGAL PLANNING CHECKLIST

	DATE COMPLETED	NOTES
Meet with an Elder Law Attorney		
Apply for Social Security Disability		
Investigate options to pay for long-term care		
Discuss intentions for future care with family		
Discuss intentions for future care with doctors		
Speak with your family and physician about options for research and brain donation		

# MANAGING BEHAVIORS

## CHANGES IN BEHAVIOR

Significant changes in behavior and personality are the main symptoms of behavioral variant frontotemporal degeneration (bvFTD). This means that a generally active, involved person could become apathetic and disinterested. The opposite may also occur. A usually quiet individual may become more outgoing, boisterous and disinhibited. Personality changes can also involve increased agitation, irritability, anger and even verbal or physical outbursts toward others (usually the caregiver). Not all people with bvFTD will adopt one symptom or another. Symptoms don't occur in "stages" but rather existing symptoms will worsen and new symptoms may appear in an unpredictable manner. Remind yourself that these are not the behaviors of the person you love - **these behaviors are the result of an illness.**

<b>BEHAVIOR</b>	<b>SUGGESTED INTERVENTIONS</b>
<p style="text-align: center;"><b><u>APATHY/LACK OF MOTIVATION</u></b></p> <p>Lack of interest, drive and/or inability to initiate activity. Often confused with depression.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Unable to take the steps to go on a bike ride on their own, but if guided to a stationary bike, they will engage in riding.</li> <li>• Unable to follow the steps to make a bowl of cereal. However, if the objects involved are laid out for them and they are cued appropriately, they can execute the numerous steps involved.</li> </ul>	<ul style="list-style-type: none"> <li>• Don't rely on the person to initiate activities on their own.</li> <li>• While they might be having trouble starting an activity, they may be able to participate if others do the planning or divide the task into small successive steps and provide assistance when needed.</li> <li>• Limit and offer specific choices; e.g. "Do you want to walk to the park or to Jim's house?" instead of a more open-ended "What do you want to do today?"</li> <li>• If they resist, do not force the activity.</li> </ul>
<p style="text-align: center;"><b><u>PERSEVERATION</u></b></p> <p>Repeating the same activity over and over when it no longer makes sense to do so.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Repeatedly doing the laundry even if there is only one item to wash.</li> <li>• Continuously talking about the same topic over and over.</li> </ul>	<ul style="list-style-type: none"> <li>• Distract by getting their attention focused on something else. Do not feel you need to explain why you are doing this.</li> <li>• If the activity is not dangerous or costly, let them do it.</li> </ul>
<p style="text-align: center;"><b><u>DISINHIBITION</u></b></p> <p>Acting impulsively without considering the social effects of inappropriate behavior or lacking insight that the behavior can offend others or cause harm.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Making offensive comments to others or to strangers.</li> <li>• Speaking about personal issues with strangers.</li> <li>• Approaching other people's children as if they were acquainted, or hugging and kissing children.</li> <li>• Shoplifting.</li> </ul>	<ul style="list-style-type: none"> <li>• Let friends and neighbors know about the diagnosis so they understand the behavior is not intentional</li> <li>• Go to places where the person is known well.</li> <li>• Distract by getting their immediate attention onto another activity.</li> <li>• It's okay to be firm by ending the conversation with, "Thank you, we have to go now," even though it may seem abrupt.</li> <li>• Use "The person I am with has FTD" card.</li> </ul>

# MANAGING BEHAVIORS

## CHANGES IN BEHAVIOR, CONTINUED

BEHAVIOR	SUGGESTED INTERVENTIONS
<p style="text-align: center;"><b><u>LACK OF EMPATHY/EMOTIONAL CHANGES</u></b></p> <p>Showing no emotions (seeming flat and disinterested) or showing exaggerated, jocular or improper emotions. A lack of sympathy or compassion to others' distress.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Seems to withdraw in familiar company.</li> <li>• Displaying emotions that are inappropriate, e.g., laughing at a funeral.</li> <li>• Seems to "not care" about another's distress, e.g., acting indifferent to spouse's diagnosis of cancer.</li> </ul>	<ul style="list-style-type: none"> <li>• Although it is very difficult, <b>do not take this personally.</b></li> <li>• Do not try to "get through" to the person by explaining why the behavior is not appropriate.</li> <li>• Let others know about the diagnosis so they are not offended.</li> <li>• Find emotional support and companionship from other friends, family or a support group.</li> <li>• Seek professional counseling.</li> </ul>
<p style="text-align: center;"><b><u>UTILIZATION BEHAVIOR</u></b></p> <p>Difficulty resisting impulses to operate or manipulate objects that are within reach; "automatic" behavior, the kind of action we all have experienced when an elevator door opens and you automatically exit despite the fact that it is the wrong floor.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Seems to be drawn to objects or actions in the immediate environment (e.g., picks up objects that are part of others' activities, seems to imitate others' behaviors) even when those objects or actions are not purposeful or appropriate for the moment.</li> <li>• Picking up the phone when walking by it even if it is not ringing or there is no intention of making a call.</li> </ul>	<ul style="list-style-type: none"> <li>• Determine if the behavior is putting the person or others at risk. If so, distract with other objects that get the person's attention immediately, such as calling them on a cell phone to interrupt an activity - the person is likely to answer it because it is an automatic behavior.</li> <li>• Note that calling their name may not work to get their immediate attention.</li> <li>• If the behavior is not dangerous or costly, let them do it.</li> </ul>
<p style="text-align: center;"><b><u>HYPERORALITY</u></b></p> <p>Compulsive eating; craving carbohydrates and sweets; no ability to regulate intake or "feel full."</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Taking food from someone else's plate at a dinner table.</li> <li>• Gorging on food to the point of vomiting.</li> <li>• Eating anything in sight with no consideration to how much has been eaten.</li> <li>• Eating uncooked meat from the fridge.</li> <li>• Eating only a certain type or brand of food.</li> </ul>	<ul style="list-style-type: none"> <li>• Provide supervision while eating, setting out portions.</li> <li>• If necessary, lock up foods and keep raw foods out of sight.</li> <li>• Use distraction.</li> </ul>

# MANAGING BEHAVIORS

## CHANGES IN BEHAVIOR, CONTINUED

BEHAVIOR	SUGGESTED INTERVENTIONS
<p style="text-align: center;"><b>RITUALISTIC/COMPULSIVE BEHAVIORS</b></p> <p>Acts that are completed over and over again, without purpose and unrelated to the circumstances in which they occur.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Person needs to continuously walk on the same route for 2 hours every day at 2pm.</li> <li>• Continuous whistling, drumming fingers in certain patterns.</li> <li>• Rigidity and inflexibility, and insistence on having his/her own way, increasing difficulty adapting to new or changing circumstances.</li> </ul>	<ul style="list-style-type: none"> <li>• If it is safe, accept the behavior and arrange for necessary supervision.</li> <li>• If unsafe (e.g., scratching at a sore until it bleeds), consult with a physician to consider medications that can minimize compulsive behaviors.</li> </ul>
<p style="text-align: center;"><b>AGGRESSION</b></p> <p>Because many individuals with FTD are not aware of their illness, they may become frustrated at limitations and constraints that they do not understand and consider to be unfair and punitive. As a result, the person may occasionally strike out at the caregiver or resist assistance. These behaviors can occur suddenly, with no apparent reason, or can result from a frustrating situation.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• Shouting or name-calling.</li> <li>• Physical abuse (hitting, pushing).</li> </ul>	<ul style="list-style-type: none"> <li>• Stay out of the person’s way if they are combative. In extreme cases, call police but explain the person with FTD’s condition.</li> <li>• Never point out the problem to the person, try to reason about their behavior, or argue about the “logical” solution.</li> </ul>
<p style="text-align: center;"><b>REASONING</b></p> <p>Not able to categorize information or think in the abstract; very literal interpretations. Lack flexibility in thinking and unable to pursue an alternative solution if the first one doesn’t work. May increase safety risk since they have difficulty recognizing consequences of behavior.</p> <p><i>Examples:</i></p> <ul style="list-style-type: none"> <li>• The person cannot understand explanations about their own illness and is resistant to continued attempts to make things clear.</li> <li>• May behave as if the caregiver is “bossy,” unreasonable or trying to control them</li> <li>• Cannot reason logically about the solutions to simple problems (e.g., how to respond in the event of a fire).</li> </ul>	<ul style="list-style-type: none"> <li>• <b>Do not argue. No amount of reasoning will make the person able to grasp the ideas.</b> Instead, distract.</li> <li>• Tell them firmly what is going to happen and repeat the information from time to time, without explanation. E.g., “We are going to see a lawyer to make sure that we have the proper documents to sell the house.” If asked for an explanation, say, “We will both have time to talk.”</li> <li>• Obtain power of attorney (in some cases guardianship is necessary) so that decision-making is not left to the person with significant reasoning deficits.</li> </ul>

**General Communication Tips**

- Always avoid confrontation. Do not argue, lecture or try to point out the truth.
- Avoid retaliation and do not shame the person. Do your best not to take out your frustration and anger on them. If you must, leave the room to calm yourself down.

# MANAGING BEHAVIORS

## CHANGES IN BEHAVIOR, CONTINUED

- Try not to take the person's behavior personally. There is no intent to hurt others, but only the inability to have normal reactions and feelings.
- When it is helpful for the person, keep decision making to a minimum by offering two choices. Don't put the person in a situation that stresses their failing reasoning capacity.
- Approach the person with a calm, reassuring tone. Smile. Individuals with bvFTD are better at understanding positive emotional expressions than negative ones. So if you are frowning or looking sad or angry, the person may not understand. A smile will elicit a more positive response.
- Note that the caregiver's emotions and facial expressions can elicit the same expression in the person (imitative behavior) even though he/she may not be feeling that way.

### Meaningful Activities

- Provide materials that are readily available and not dangerous. Jigsaw puzzles, drawing materials, coins to be sorted, and laundry to be folded.
- The person should be provided with physical activities within their capacity. They may require support, such as a "trainer," an individual who can be hired to take the person out for a walk daily or do other types of stimulating activities. Using such a label may be more acceptable to the person than a "companion" or a "caretaker."

When the individual with behavioral changes shows new symptoms, don't assume that it is the disease. Because people with bvFTD find it increasingly difficult to articulate such things as pain or discomfort, they may manifest such things as agitation or irritability. It could be the disease or it could be something else that can be addressed with a visit to the primary care doctor. With all new behaviors that you observe, go through the following checklist to determine what is causing the change and find the most appropriate intervention:

- ✓ Could this be a separate **medical problem** that is causing the change in behavior? For example, the person may have a toothache but be unable to articulate the precise problem. Another example is an imbalance of thyroid function, an infection, or other chemical imbalance in the body that temporarily makes the bvFTD symptoms look worse.
- ✓ Identify triggers of certain behaviors; is the **environment** triggering the behavior? Although many behaviors are erratic and have no explanation or precedent, some may be reactions to certain types of situations. For example, the person becomes agitated when there are more than three people talking. If so, what in the environment can be changed? In this example, the solution might be to reduce the number of people the person interacts with at one time. Invite one adult child to dinner, instead of the whole family. Try to identify if there are triggers and what they might be.
- ✓ Is this behavior **safe for them?** Is this **safe for me and others?** Some behaviors are very annoying but are not injurious to the person or others. On the other hand, if the person does not recognize that an 18-month child cannot be left on the living room floor with the front door open and a flight of stairs not far away, precautions need to be taken to make sure that the person is not put in a situation where they cannot exercise appropriate judgment. Even though the person may be able to play with the 18-month old in an appropriate way, they are unable to be left alone with the child in this instance.

### When to consider medications:

Trying the above strategies is always the first step in responding to changing behaviors; however, sometimes medications can also help. Some serotonin reuptake inhibitors are often prescribed for **carbohydrate craving, disinhibition and impulsivity**. Persons who experience uncontrollable **aggression or delusions** are sometimes prescribed low doses of antipsychotic medications. It is important to consult with a specialist in this area, such as a psychiatrist with expertise in dementia and pharmacology.

# COMMUNICATION CHALLENGES IN PPA

Loss of language is the main presenting symptom of PPA. This means that an active reader, writer and conversationalist could lose his/her grammar, vocabulary, and ability to find the correct word. The opposite may also occur; a usually quiet individual may become more vocal but others may not understand his/her speech. Every individual diagnosed with PPA is unique and no one will experience the disease progression identically. Symptoms don't occur in "stages" but rather existing symptoms will worsen and new symptoms may appear in an unpredictable manner. However the symptoms present, every individual with PPA has a unique set of communication strengths and preferences. Identifying and supporting these strengths and preferences can help them communicate most successfully and effectively.

## Accepting PPA and Adapting to New Forms of Communication

When symptoms first present, families are forced to let go of previous patterns of communication (the individual with PPA may have been the primary communicator in family/relationship) and with the use of new strategies and tools, begin to communicate differently. **Communication Strategies** are alternative ways of communicating that the person and family can learn to help them compensate for the loss. **Communication Tools** refer to high tech or low tech devices that assist the person in communicating with others.

### On a Daily Basis

- Remember that the goal is communication, not perfection.
- Do not speak for the individual. Establish a cue to use that implies they would like your help.
- Speak slowly, allow enough time for the individual to respond to questions.
- Help the person to avoid stress. It makes the communication challenges more difficult.
- Continue to participate as much as possible in social activities (gatherings with friends/family).
- If a response is incomplete or not clear, ask for clarification; repeat back what you heard. Do not pretend to understand if you do not.
- Be aware of signs of depression. Because persons with PPA are often aware of the changes, they may experience depression, anxiety, frustration, and loneliness.
- The person will have good days and bad days. Do your best to provide additional support on the more difficult days.

### Educate Yourself

- A Speech and Language Pathologist (SLP) can help teach new ways of compensating for losses and help identify the most appropriate strategies/tools at each stage of the diagnosis.
- Become familiar with communication tools for all modes of communication. Using multiple modes of communication (speaking, writing, drawing, gesturing) can make communication more effective.
- It is best to implement and practice use of strategies/tools in various communication situations **before** they are absolutely needed so that diagnosed individual and family members feel comfortable using them. For example, create the communication notebook or become familiar with communication technology before it is needed.

### The Role of the Supportive Communication Partner

- Try not to interrupt or supply words unless your help is requested.
- Ask a question that can be answered with YES/NO: "Do you want eggs?" or "Do you want to go for a walk?" Ask a question, which requires a choice between two items/possibilities: "Do you want eggs or cereal?" or "Do you want to go for a walk or take a nap?" Try to avoid open-ended questions, like: "What do you feel like eating?" or "What do you want to do today?"
- Have realistic expectations based on the communication strengths and areas of difficulty of the individual diagnosed with PPA.
- Manage the environment (background noise, number of people present).
- Manage your feelings, language, thoughts, and expectations.
- Continue to reevaluate the person's changing needs as time goes on.

## COMMUNICATION CHALLENGES IN PPA, CONTINUED

METHOD	STRATEGIES FOR EFFECTIVE COMMUNICATION
<b><u>LISTENING</u></b>	<ul style="list-style-type: none"> <li>• Listen for key pieces of information: who, what, where, when, why?</li> <li>• Be aware of distracting background noise, and reduce it if possible</li> </ul>
<b><u>SPEAKING</u></b>	<ul style="list-style-type: none"> <li>• If speaking is difficult, or someone has difficulty finding the words, use the following word-finding strategies:               <ul style="list-style-type: none"> <li>• Synonym Substitution: another word to convey the same meaning</li> <li>• Antonym Substitution: state the opposite of the target word</li> <li>• Association: think of/name words associated with the target word (e.g., bread goes with butter)</li> </ul> </li> <li>• Describe the object of thing you are trying to think of:               <ul style="list-style-type: none"> <li>• Purpose or function</li> <li>• General category (fruit, clothing, sport)</li> <li>• Physical description (size, shape, color)</li> <li>• Location</li> </ul> </li> </ul>
<b><u>WRITING</u></b>	<ul style="list-style-type: none"> <li>• Can be used when reading and writing are relatively well preserved</li> <li>• Individual with PPA, or communication partner, can do the writing</li> <li>• Communication partner writes a question with 3 or 4 choices, (1 of which is “other”), individual with PPA chooses appropriate response. Example: Where were you born? East Coast, West Coast, Midwest, Other</li> </ul>
<b><u>GESTURES</u></b>	<ul style="list-style-type: none"> <li>• Gestures can be developed for core daily functions</li> <li>• Practice and have gestures in place, before you actually need them</li> </ul>
<b><u>TOUCHING</u></b>	<ul style="list-style-type: none"> <li>• Firm handshake, pat on the back, holding hands, giving a hug, sitting close</li> </ul>

### Alternative/Augmentative Communication Tools

- Low Tech Devices
  - Communication Notebooks: Commercially available are ready-made books for general communication needs with some room to personalize (example is Alimed)
  - Personalized Communication Book includes a collection of pictures of family, friends, activities, and commonly used phrases. Pictures are arranged by category in different sections of the book and words and descriptive phrases are attached to each picture
  - Communication Board (dry erase)
  - Index cards with pre-written statements (“I would like to order a chicken salad”)
- High Tech Devices
  - Dynavox
  - Communication Applications for smart phones
  - Proloquo2go (for iPhone/iPad)

# FAMILIES WITH CHILDREN AND TEENS

## WHAT TO TELL THE CHILDREN?

When a family member is diagnosed with Frontotemporal Degeneration (FTD) or Primary Progressive Aphasia (PPA), parents often worry about what to tell the children. Children are affected by everything that happens in the family. Given the number of changes brought on by FTD or PPA (moods, behavior, communication changes, etc.), children soon realize that family life is different. If someone does not communicate with them about what is happening, they imagine their own reasons for changes. They may attempt to make sense of the situation by listening into adult conversations, looking at written materials brought into the home or by questioning other children. Children can feel isolated from adults, confused and overwhelmed. Parents can help by telling the children the truth and addressing their children's concerns regarding cause, contagion, care, connection and communication.

**Cause:** Tell children the name of the disease or condition. Explain how and/or where in the brain it began. Give clear and concrete explanations geared to their age and level of understanding. Make sure children understand that the illness is a medical condition that they did not bring on with their thoughts or actions. Give explanations gently and over time if needed.

**Contagion:** Children may be concerned that they or other family members will get FTD or PPA, too. Older children may have questions regarding the illness as a hereditary condition. If heredity is a truth for the illness, remember that each one of us needs information in order to cope. Answer questions to the best of your ability and in a manner that is developmentally appropriate. If the form of FTD/PPA is not hereditary, reassure the children that they or other family members cannot get the illness.

**Care:** Reassure children that they will continue to be cared for and loved even though more attention will be given to the person with the illness. Children deal better with changes in the family when they know they are still important. Also, encourage children to ask specific questions about the impact of the illness on their daily routine.

**Connection:** Children need to maintain a connection with the person with FTD and PPA. This can be difficult as the individual begins to have more personality changes (with bvFTD) and language decline (with PPA). Be aware of what safety issues need to be addressed. Although the person with FTD or PPA cannot be in charge of the child's care; with proper supervision, he or she may be able to assist or engage in playful activities with the child; for example board or video games, going out for a walk, or preparing lunch.

**Communication:** Children need to be reassured that it is okay to ask questions as they arise. It is important for them to know that it is safe for them to express their thoughts and feelings. If parents are unable to address their children's questions, thoughts or feelings due to their own experience of the family member's illness, be sure to designate one or two trusted adults to whom the child can turn. These adults may be family members, clergy persons, bereavement counselors, school personnel or anyone with whom the family and child have a trusting relationship.

Informational resource: *How to Help Children Through a Parent's Serious Illness* by Kathleen McCue

*Adapted and used with permission by Elise Gaul, MS, LPC, CT  
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# **FAMILIES WITH CHILDREN AND TEENS**

## **COPING WITH FTD AND PPA:**

### **HOW ADULT FAMILY MEMBERS CAN HELP CHILDREN, CONTINUED**

*The following are some suggestions for helping children when an important person in their lives has FTD or PPA. All families are different and all children are unique—so some suggestions may not apply to all cases. Speaking to a children’s mental health professional can help you clarify how to best take care of the children in your family.*

- ❖ Learn as much as you can about the illness
- ❖ Use correct medical terms. Don’t over explain, but be honest. Fear and fantasies are often worse than reality.
- ❖ If the person with FTD or PPA lives in another setting, offer children choices of phone calls, letters, drawings, and/or visits with the person with FTD or PPA. Prior to a visit, describe what the child can expect to experience at a hospital, hospice, or nursing home-- including how the person who is ill will look and act.
- ❖ Reassure children that noting they did or didn’t do caused the illness. Find time to give them love and attention.
- ❖ Take care of yourself and find support to overcome personal fears and anxieties. Children model behavior and coping skills from the adults they live with.
- ❖ Encourage communication. Don’t assume lack of questions mean lack of interest. Children are more likely to express themselves in art, play or actions than in words. Find time to observe them during these activities. Ask teachers & others for their observations. Share your own true feelings to help them understand their own.
- ❖ Headaches, stomach aches and behavioral problems may be caused by repressed feelings. Provide healthy outlets for energy release and expression with creative and physical activities.
- ❖ Feelings of abandonment, helplessness, despair, anxiety, apathy, anger, guilt and fear are common in a family with serious illness. Children often act them out aggressively when there are no healthy outlets. Anxiety may lead to hyperactivity and behavioral problems. Try to maintain as normal a routine as possible. Children need structure to feel secure during stressful times.
- ❖ Children need to be involved in appropriate ways. If they try to assume caretaker roles, remember they need to grow up normally without being burdened with adult responsibilities.
- ❖ Coping with illness, financial and personal needs may overwhelm the parent who is not ill. Children need increased support from grandparents, neighbors and friends. They need to grow up knowing there is someone to count on or they may become too independent and distrustful.
- ❖ There is a significant correlation between duration of illness and a child’s behavioral difficulties. Illness lasting more than a year requires more intervention because the family focus is on something other than the child. Learn what services are available for your situation and needs through hospitals, churches, schools, community agencies and professional counseling. Inform pediatrician about family problems.

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# PLANNING FOR FUTURE AND PALLIATIVE CARE



The Association for  
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*Opening the gateway to help and a cure*

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## Hospice - A Valuable Service to Consider

If a family member has a terminal illness, it is never too early to learn about hospice and investigate hospice agencies as part of planning for health care. Hospice is a philosophy of care that holds as fundamental a patient's right to be treated with respect and dignity during his or her final period of life. It refers to a holistic approach that encompasses a range of compassionate services for both the patient and family. Hospice is also a specific federally funded program within Medicare in the United States.

The goal of hospice is to maintain the patient's quality of life and to relieve pain and suffering as much as possible. It focuses on the emotional, spiritual, psychological, and physical needs of the patient. Hospice recognizes the important role family and friends play as caregivers, and will provide practical and other supports for them in that capacity.

The approach prioritizes the wishes and choices of the patient concerning how and where they want to live out their final days. Living wills, advanced medical directives, and do not resuscitate orders (DNR) may express some of these choices. Hospice will also provide the medical equipment needed to make it possible for the patient to remain at home, if they so choose.

### What Hospice Offers

#### **Palliative Medicine**

Hospice provides palliative medical care. Palliative medicine is a specialty that focuses on the treatment and management of the symptoms of a terminal illness. The aims of palliative medicine are to provide comfort, relieve pain, and improve a patient's quality of life. It will deliver any treatment or medication necessary to ensure those aims at any stage of a terminal illness, not just the late stages.

In contrast, medical care aimed at curing, stopping, or preventing a terminal disease is called curative care. In order to participate in public health insurance programs such as Medicare or Medicaid hospice agencies can only provide palliative medical care, not curative care. Hospice in other parts of the world, and some private pay hospice plans, may not make this distinction and will provide palliative care along with medications or treatments intended to cure or stop the progression of a terminal illness.

Hospice will usually offer 24 hour access by phone to doctors or nurses to answer questions and offer guidance. If in-patient care is needed to manage pain and symptoms related to a terminal illness a hospice agency can arrange a short-term hospital stay.

#### **Non-medical Healthcare**

In addition to palliative medical care, hospice offers a range of non-medical health services. Listed below are some of services provided by hospice and a brief description of how they may benefit a person with FTD. This list is not all inclusive, and there may be differences between hospice agencies in terms of the services offered.

# ***PLANNING FOR FUTURE AND PALLIATIVE CARE, CONTINUED***

For more information on the professionals trained to deliver these services see AFTD's website section on Other Health Professionals.

## **Physical Therapy**

Physical therapy can help manage pain and discomfort, increase mobility, and provide other health benefits associated with routine physical activity. Physical therapy can also alleviate specific muscle control problems and help maintain and extend an individual's physical independence.

## **Massage Therapy**

Massage provides some of the same benefits as above and can improve flexibility and mobility. Massage can also have a calming and restive effect.

## **Speech-Language Therapy**

This can help with communication problems either directly by addressing specific speech problems or by providing alternative communication strategies. A speech therapist that specializes in swallowing can also address problems with eating and drinking to help avoid choking, or breathing food or liquid into the lungs.

## **Art/Music Therapy**

FTD can rob people of their ability to be self aware and communicate their feelings and experiences. Art/music therapy provides a way for patients to express themselves and can help maintain emotional stability and some cognitive abilities.

## **Routine Personal Care and Hygiene**

A common symptom of FTD is a decrease in self-care. Help with routine activities such as bathing and brushing teeth can prevent related health problems, especially infections.

## **Nutrition/Diet Planning**

Another frequent FTD symptom is idiosyncratic eating habits. Meeting a body's basic dietary requirements helps maintain general health.

## **Supportive Counseling**

Hospice offers social, emotional and spiritual services for the terminally ill patient and his or her family through a variety of counselors and therapists both religious and secular. Some hospice programs include special services that address the needs of children in the family.

## **Other Benefits**

The hospice approach is recognized and valued for its holistic approach to supporting all aspects of the patient's and families experience at the end of a loved ones' life. Below is a list of some additional benefits available through hospice.

## **Respite Care**

Caring for a loved one with FTD is emotionally, spiritually, and physically draining. If the primary caregiver becomes sick, or needs a break hospice can arrange a short term stay in a nursing or assisted living facility for the patient.

# ***PLANNING FOR FUTURE AND PALLIATIVE CARE, CONTINUED***

## **Durable medical equipment and supplies**

Equipment, such as a hospital bed, needed to allow a patient to remain in his or her own home or current living situation can be provided through Hospice. Supplies such as bandages or bed sheets are also provided.

## **An extra pair of hands**

Many hospices have dedicated and trained volunteers to help with household chores, like shopping or cleaning.

## **Case Management**

FTD can raise a host of problems in addition to the health issues. Hospice agencies often provide a case manager to help families' access additional community resources, and make plans around providing care for a loved one.

## **Bereavement supports**

Grief counselors are available through hospice after the patient's death, in some programs this benefit extends for up to 13 months.

## **Services Not Available through Hospice**

### **Emergency Medical Care**

In addition to curative care hospice will not provide emergency medical care not included or contrary to the wishes of the patient as stated in an advance medical directive or similar document. Any emergency medical services provided by hospice must be pre-arranged and addressed in the hospice care plan. This includes ambulance transport to an emergency room, and any emergency medical treatment or in-patient hospital care related to the terminal illness.

### **Room and Board**

Even though hospice care is often provided to people in nursing homes or other residential facilities; room-and-board, and any other cost associated with a residential facility will not be covered by hospice, with the exception of an in-patient hospice program.

## **How to Arrange Hospice**

Hospice services are all provided and coordinated by an interdisciplinary team within a single agency led by a medical director. To comply with Medicare rules, services must be provided by a certified agency and that agency must be used for all hospice care. Services can be provided in the home, at a residential facility like a nursing home, or at an in-patient hospice facility.

## **The Hospice Team**

### **Hospice Medical Director and/or Primary Care Physician**

The Medical Director, usually a MD, oversees and directs the medical care of hospice patients. It is possible to arrange to have your primary care physician be included as part of the hospice team to work with the Medical Director to direct and guide services and to ensure the continuity of medical care. The Medical Director is also responsible for determining whether a patient is medically eligible for hospice and for re-certifying the need for continuing hospice care.

# ***PLANNING FOR FUTURE AND PALLIATIVE CARE, CONTINUED***

## **Primary Care Provider**

The primary care provider is usually a nurse practitioner who monitors the patient and coordinates care with the Medical Director. The nurse will establish a routine of scheduled visits and provide all the skilled nursing care required to manage symptoms and control pain. A hospice nurse will be available 24 hours a day to respond to situations which need immediate attention.

## **Social Worker**

Hospice care is coordinated by a social worker, including arranging respite care and other short-term in-patient stays. Social workers will also help families access other services and agencies in the community as needed.

## **Periods of Care**

Because of the uncertainty and the time-limited nature of end of life care, hospice is delivered in periods of care lasting 90 days initially, then 60 days thereafter. At the start of each period the hospice medical director must recertify the patient, based on continuing need. There is no limit on the number of times a person may be recertified for an additional 60 days. A patient or legal guardian may choose to stop hospice at any time, and has the right to switch providers once during each certified period of hospice care.

## **How to Pay for Hospice**

Hospice is covered by almost all health insurance plans including federal Medicare and state Medicaid programs. There are also hospice agencies, usually not-for-profit organizations that provide care regardless of the patient's ability to pay and have funds for patients who may not be eligible or able to afford any health insurance.

## **Medicare and Medicaid**

Hospice benefits are included in Medicare part A (the hospital plan) and through Medicaid in 47 US states and the District of Columbia. To use the Medicare Hospice Benefit people must sign a statement choosing hospice instead of other Medicare-covered benefits to treat a terminal illness. Medicare coverage will still be available for any health care services not related to a terminal illness, so if a hospice patient falls and breaks a wrist, they will still be covered by Medicare. Medicaid (state funded) hospice benefits may have a similar requirement which may vary in the details from state to state.

## **Military benefits**

The Veterans Health Administration (VHA) includes hospice benefits in the basic eligibility package available to enrolled participants. VA medical centers will either provide hospice care directly in their facilities or purchase it from a community hospice agency. The US military health plan TRICARE includes hospice for active duty service members and retirees of the seven uniformed services, their family members, and survivors. TRICARE also provides hospice benefits for National Guard and Reserve members and their families after the member turns 60. Hospice benefits through TRICARE are only available to patients residing in the fifty United States, Washington D.C. and U.S. territories.

## **Private health insurance**

Private health insurance companies offer policies which include hospice benefits that resemble government plans but may vary in the details. Always check with your insurance provider and the hospice agency on what benefits and services are available and how they will be paid for. Insurance plans will likely cover the majority of costs, including medications, but there may be co-pays and other expenses for which you are responsible.

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# ***PLANNING FOR FUTURE AND PALLIATIVE CARE, CONTINUED***

## **Considerations in FTD**

### **When to Consider Hospice**

Hospice is an underutilized healthcare program. There are many reasons for this but chief among them is that hospice is too often considered only when death is imminent. If it is discussed at all it is usually offered as a last resort or when a doctor or nursing home staff feels that further medical treatment is in vain. You do not need to wait for someone else (ie: the doctor) to suggest hospice as an option. If a loved one has a terminal illness, hospice is an option to look into at any time, not just in those very last days or weeks of a person's life.

Choosing hospice is too often thought of as an admission of defeat; a sign that the caregiver is abandoning hope and giving in to death. Instead, the mission of the hospice movement is to affirm life, preserve dignity, and provide comfort during the final period of life. The benefits are greater when the patient and family can work with the hospice team over time. Most hospice agencies will provide a free initial consultation to review payment options and assess whether a patient and their family is ready for hospice.

This can be an excellent opportunity to make long term care plans, and help a family become aware of all the options available. Some agencies offer transitional or partial hospice plans, often called "bridge" programs, for patients who do not fully qualify for hospice care, or when families are not ready to stop medical treatments aimed at a cure. An initial consultation also provides the opportunity to assess whether the agency is capable of caring for your loved one, and the unique challenges presented by a FTD patient.

### **Six month life expectancy requirement**

Most hospice programs in the US are designed to meet Medicare rules. One eligibility requirement of Medicare is that the person's physician and the hospice medical director must certify that the patient has six months to live given the usual course of the disease. However, the program recognizes that doctors cannot make this claim with absolute certainty, and hospice services can stop and start as needed, depending on the patient's condition.

The natural course and end stages of neurodegenerative diseases such as FTD are especially difficult to predict and may not be well understood by physicians and hospice programs. It is helpful to express an interest in hospice as a health care option early with the physician and prospective hospice providers to educate, build collaborative relationships and advocate for services as needed. Attentive documentation of small changes in functioning can be made before and during hospice services to support continued eligibility.

### **Deciding to stop curative treatment**

In the U.S. in order to receive hospice care through public health benefits the recipient must agree to use their primary insurance plan exclusively for palliative care through a certified hospice agency and forego coverage of curative care by that insurance plan. Public health plans in the US will not pay for both forms of care for the same illness at the same time. This is a deeply personal choice and is the fundamental decision caregivers and patients must make before choosing hospice care.

# ***PLANNING FOR FUTURE AND PALLIATIVE CARE, CONTINUED***

## **Hospice and Dementia**

Because hospice began primarily serving cancer patients its traditions and standards do not always apply to dementia patients. Dementia differs from cancer and other terminal illnesses because it affects the mind first then the body, but waiting until the mind has degenerated to the extent that it can no longer control the body before accessing hospice is not necessary, and diminishes the potential benefits of using hospice care.

To qualify for hospice Medicare initially required dementia patients to be rated on a scale developed to track the progression of Alzheimer's patients. This scale, known as FAST, required patients to be, bed-ridden, unable to talk, and without any basic motor skills such as the ability to sit up or turn their head. Such a strict requirement likely added to the general belief that hospice could provide little help for dementia patients and may be among the reasons why doctors and nursing home staff rarely suggest hospice care for FTD patients. There is now less strict adherence to FAST ratings and doctors are able to consider other factors such as the clinical history of the patient, the overall rate of decline, and other health concerns which could be exacerbated by dementia and contribute to the prognostic picture.

Even with a broader view of eligibility, several common barriers persist to considering hospice for people with degenerative neurological disorders such as FTD. They include:

- Recognition and acceptance of FTD as a terminal illness by the patient and family, or the physician.
- Challenges with predicting the long- term course of the disease and with a making a prognosis.
- Difficulty in making decisions around such things as treatment of acute infection, placement of feeding tubes, and hospitalization.

These barriers can be addressed through discussion with involved family and friends and consultation with physicians and hospice providers.

# STARTING A SUPPORT GROUP IN YOUR AREA

Support Groups offer a safe environment for family caregivers to share their experience with others who truly understand. Currently few support groups are available specific to Frontotemporal Degeneration (FTD) and Primary Progressive Aphasia (PPA). There is a need for more groups of this kind. In this session we will discuss how to start a support group in your area with the support of AFTD and the Alzheimer's Association. Attendees will learn about how to begin, sustain, and maintain a group that specifically offers support to those caring for someone with FTD or PPA.

## **Planning Steps**

### **1. Assess need.**

A support group often starts small and grows over time. You can gauge the level of potential interest or need for a group in your area based on 1) the existence and location of other FTD groups, and 2) an awareness of FTD caregivers through a local medical center, Alzheimer Chapter, care facilities. AFTD can provide a count of registered caregivers in an area as another measure of potential interest.

### **2. Assess the facilitator's readiness.**

Most support group facilitators have a personal or professional connection to the disease. Family caregivers that would like to facilitate a group should consider their own needs before taking on this additional responsibility. The general recommendation is to wait at least one year after the passing of the person with the diagnosis before the family member can dedicate themselves to helping others. However, as few supports exist, current family members are often eager to start a group. Finding a co-leader who is another family member or local professional can help ease the burden of organizing and leading a group.

Qualities of an effective facilitator include:

- Compassionate
- Non-judgmental
- Good listener
- Able to separate own situation from the needs of the group
- Not the expert or problem-solver
- Knowledgeable about the disease, current treatments, research trends and coping skills
- Can manage group personalities, differences of opinion and unique challenges as they arise
- Organized and prepared for each meeting and conducts reminders prior to meetings

### **3. Determine appropriate location, meeting format, and time of day.**

Locations appropriate for hosting support groups are often community-based such as libraries, community centers, churches or synagogues. Consideration may be given to adult day programs that could assist with supervision for the person diagnosed while the caregivers are meeting nearby. Long term care facilities are possibilities, but keep in mind that not everyone is comfortable visiting these settings until they are ready which may limit group attendance. However, sometimes a facility has the resources to provide the group a meal or refreshments during the meeting.

In any setting, be mindful that the group's purpose is education, support and coping around the diagnosis and not marketing of the host site's religious affiliation or product. Conversations with a

## **STARTING A SUPPORT GROUP IN YOUR AREA, CONTINUED**

potential host site should involve expectations of each party involved. You want your members to feel comfortable and welcome regardless of their race, sex, sexual orientation or religion.

While choosing a location, consider the meeting space. Is it private and does it provide an environment for confidentiality? Does it have a door that closes? Is it close to the bathroom? Is it easy to find? Is there comfortable seating? Is it handicap accessible? Is adequate parking available?

When scheduling your meetings consider how often you will meet - monthly, twice a month or a different time frame. If meeting in the daytime, your primary audience will be retirees. However, if you decide to meet in the evening, family members that work are more likely to attend.

### **4. Make a list of places to conduct outreach to recruit possible members.**

Create a flyer for your group and take it to local referral sources such as the diagnostic center, neurologists, hospitals, pharmacies, home health agencies, assisted living, nursing homes, a senior service agency and/or the local senior center. Think about the places you would see a notice about a group like this.

The local paper often has a health calendar. This would be a good location to advertise the support group. Church bulletins are also a good place for a paragraph about the group.

### **5. Learn about facilitation techniques for managing group dynamics.**

If you've never led any type of group before, you may want to participate in training to practice group dynamic skills. Every group is unique and each person within a group brings their own knowledge, history, values, coping methods and personality. There may also be differences in each member's relationship and time spent with the person diagnosed. Some caregivers are responsible for the person's care 24-hours-a-day at home and some are long-distance caregivers for a person who lives in a care facility. As the group leader, your job will be to help people find common ground on which to share their emotions, strategies and strengths.

You will want to learn techniques for engaging a group participant that is withdrawn or quiet, as well as members that try to dominate the conversation. Also remember, caregivers have a range of emotions such as anger about their situation or sadness about the loss of the future they had planned. The facilitator's role is to provide a safe, respectful environment where everyone feels accepted and heard.

You can learn more about training and support for group facilitators by AFTD and the Alzheimer's Association below.

### **Support Groups for Diagnosed Persons**

Groups are beneficial for people with a diagnosis of PPA by providing a safe place to discuss their feelings, coping strategies and useful information. Diagnosed persons report a feeling of isolation and loneliness and appreciate meeting others with similar challenges. These types of groups are most helpful in the earliest stages of the disease process after the diagnosis has occurred. A facilitator must possess a special set of skills and training as compared to groups for the care partners and would most likely be a professional with expertise in engaging people with a specific diagnosis. For example, a PPA group would benefit from having a facilitator with training in speech therapy techniques. Inquire with your local diagnostic centers if this is a type of group you would like to see in your community.

## ***STARTING A SUPPORT GROUP IN YOUR AREA, CONTINUED***

### **FTD/PPA Support Group Training & Support**

#### **Association for Frontotemporal Degeneration (AFTD)**

The Association for Frontotemporal Degeneration (AFTD) is a non-profit organization whose mission is dedicated to helping people who live and work with the frontotemporal disorders. A central aspect of AFTD's mission is to provide information, education and support to people diagnosed with frontotemporal disorders and their families. Services include a website ([www.ftd-picks.org](http://www.ftd-picks.org)) and HelpLine, by email or toll-free telephone, which reach thousands of people per year.

AFTD sponsors telephone support groups and provides assistance, resources and continuing education to facilitators of independently operated local groups. AFTD helps new groups to assess need in their area, plan for success and notify potential participants of a new resource. Once groups are established, AFTD posts information on its website and informs people contacting the HelpLine. The Association provides group leaders with information on developments in the field, facilitates resource sharing, problem-solving and provides continuing education with experts in the fields of frontotemporal disorders and caregiving. Individuals interested in starting a support group should contact AFTD's Program Director at 267-514-7221 or [info@ftd-picks.org](mailto:info@ftd-picks.org).

#### **Alzheimer's Association**

Although FTD and PPA are different from Alzheimer's disease, similarities exist with how a family copes with the diagnosis and the community resources that are available. Through a network of 70 Chapters throughout the U.S. the Alzheimer's Association provides information and referral, education, support and advocacy. Resources through the 24-hour Helpline and website include non-Alzheimer's related dementias.

Until more specialized FTD and/or PPA support groups become more widely available, care partners are welcome to attend already existing Alzheimer's support groups in their community. Individuals interested in starting an FTD and/or PPA care partner support group can contact the Alzheimer's Association for support and training. Support group facilitators become volunteers of the Association and complete pre-requisite education, a background check screen and participate in a leader training program. Once the group begins, the Chapter can assist leaders with continuing education, mentoring and providing resources to use in groups. Many Chapters also provide a website listing of the group and information about the group on the 24-hour Helpline. Contact your local Chapter by calling 1-800-272-3900 or visiting [www.alz.org](http://www.alz.org).

#### **National Aphasia Association (NAA)**

NAA is a consumer-focused, not-for-profit organization that was founded in 1987 as the first national organization dedicated to advocating for persons with aphasia and their families. Although PPA is different from acquired aphasia, there are similarities in the need for increased awareness, education and support. The NAA is able to provide information and guidance for those persons with PPA and/or their families. There is great need for support groups specifically for PPA and those interested in starting a group should contact the NAA for resources. Additionally, the NAA currently lists more than 400 aphasia community groups across the country and many may be appropriate for persons with PPA and/or their families. You can reach the NAA by calling our hotline (800) 922-4622 or visiting our website [www.aphasia.org](http://www.aphasia.org).

# STARTING A SUPPORT GROUP IN YOUR AREA

## CONSIDERATIONS FOR FRONTOTEMPORAL DISORDERS

### SUPPORT GROUPS

The need for support groups for caregivers of people with frontotemporal disorders is significant. A lack of awareness among the public and health professionals contributes to the burden that patients and families face. People may experience frustration getting a diagnosis, and finding knowledgeable professionals; they may feel socially isolated when others don't understand. The effective support group will allow people to share their frustrations while learning about the disorders and developing coping and care management skills.

Some particular considerations for leading these support groups include:

#### **1. FTD research and language are evolving rapidly.**

Advances in understanding these diseases contribute to a steadily changing landscape of terminology, nomenclature and information. Doctors may not use the same terms for diagnosis which can be confusing for patients and families. Acronyms are many and arcane. Group leaders need to be sensitive to how difficult this can be for families. They can clarify where possible, direct people to other resources and re-focus on support and skill-building.

#### **2. There is a wide variety of clinical presentations and presenting needs.**

The frontotemporal disorders are identified clinically by the symptoms that develop first and are most prominent. People may experience early language changes (Primary Progressive Aphasia), behavior and personality changes (behavioral variant Frontotemporal degeneration) or difficulty with movement (Cortical Basal Syndrome or Progressive Supranuclear Palsy).

The challenges associated with these early symptoms can be quite different. Support group facilitators, especially in a caregiver led group, may be most familiar with one clinical presentation. It is important to appreciate the range of possible symptoms and their associated caregiving challenges.

#### **3. The pathology and genetics are complex and frustrating to many.**

Scientists are just beginning to understand what happens in brain cells that lead to Frontotemporal lobar degeneration (FTLD). Many support group participants struggle to understand what caused their loved one's disease and if other family members may be at risk. These are complex questions that remain difficult for medical and genetic experts to answer. Facilitators should be prepared to offer basic information, direct people to other resources and re-focus on what caregivers can control.

#### **4. Progression is unpredictable.**

Over time additional symptoms emerge as other parts of the brain become affected. All patients will experience increased dependency later in the disease progression, but the specific symptoms and rate of decline cannot be predicted. The continual changes and uncertainty about the future can contribute to anxiety. The group can help caregivers to focus on the present, identify retained strengths and create a positive atmosphere for both patient and caregiver.

# ***STARTING A SUPPORT GROUP IN YOUR AREA***

## ***CONSIDERATIONS FOR FRONTOTEMPORAL DISORDERS***

### ***SUPPORT GROUPS, CONTINUED***

#### **5. Concerns differ based on an individual's relationships to the disease.**

A single FTD caregiver group may address the needs of people with different perspectives on the disease. A spouse, adult child, parent with children at home or friend of a patient will have unique emotional and practical caregiving concerns. Solid group facilitation skills can ensure that everyone has a chance to contribute and benefit from the experiences of others. While currently limited, leaders can become aware of additional resources tailored to these different perspectives that can supplement the local group.

#### **6. The group can be a hub for local resources.**

Most caregivers know how difficult it is to find local resources that have experience working with someone with one of the frontotemporal disorders. Local Alzheimer's Association Chapters and Offices of Aging can be helpful; AFTD does not yet have good command of local resources. A support group that compiles resources with knowledge and experience serving people with FTD provides added value to current and future members.

#### **7. Members can be passionate about advocacy and awareness.**

The lack of awareness about frontotemporal disorders and limited services often propel support group members to want to become involved in related activities. Support groups can be a catalyst for making a difference, but not all members will be similarly motivated. The facilitator must ensure that the group remains focused on caregiver support and education, but can help interested individuals to volunteer with AFTD, the Alzheimer's Association or other organizations or projects promoting their cause.

#### **8. The group can't be all things to all people.**

Within relatively rare diseases like frontotemporal degeneration people are hungry to find understanding and resources. Sometimes the needs may be overwhelming and adequate resources may not exist. Focus on what you do well and recognize that the support group cannot be all things to all people.

# GENETICS AND FTD/PPA

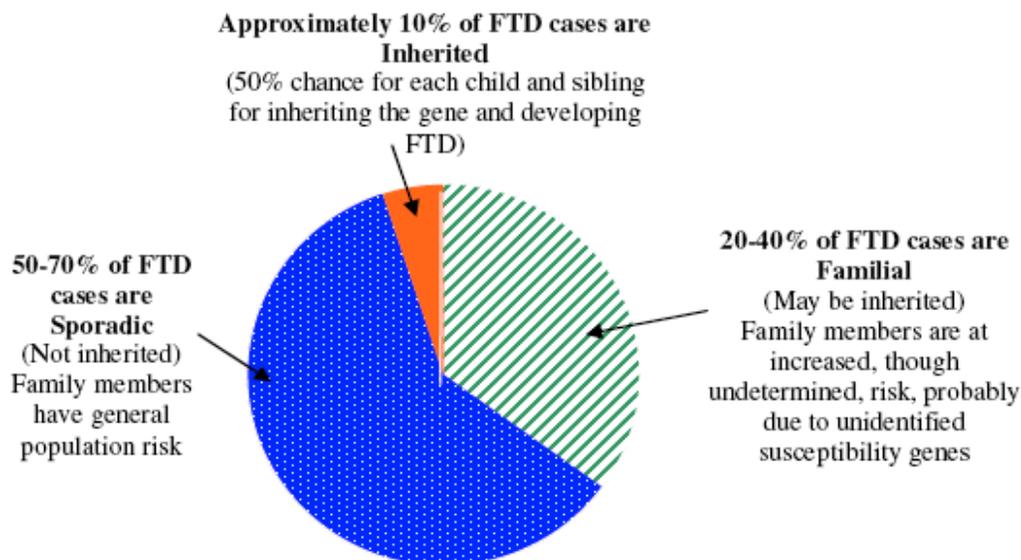
## The Genetics of FTD: Should you worry?

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### Summary

In 2011 there is still more that we DON'T understand about the genetics of FTD than we DO understand. Researchers and families are working together around the world and each year the picture comes into focus a bit more.

Our current understanding of the genetics of FTD is summarized in the following graph:



- Much of FTD (50-70%) is **sporadic**. This means the disorder is not likely to be inherited, and the risk to family members is near that of the general population.
- For a small minority (about 10%) of families, FTD is being **inherited**, and each first-degree relative of the patient (siblings and offspring) has a 50% chance of having the same disease causing gene mutation and thus developing the disorder. This type of inheritance pattern is called autosomal dominant. Estimates of the percent of autosomal dominant cases vary by country and by the method employed by the research study.
- A large group of families (20-40%) lie in a mid-range, where family history reveals that there is a heritable component to the disorder, but does not raise the risk to family members as high as in the "inherited" group. This inheritance pattern is termed **familial and is much more complex than that of the autosomal dominant type of inheritance.**

## ***GENETICS AND FTD/PPA, CONTINUED***

Given that advances are continually being made, it is important to stay abreast of developments in this rapidly changing field. One of the best ways of doing this is to stay connected with AFTD.

This article contains the following sections:

- **Introduction**
- **The Importance of Your Family Medical History**
- **Genes and Inheritance: Understanding the Basics**
- **The Genetics of FTD**
- **Genetic Counseling and Genetic Testing**
- **Discussing These Issues with Your Family**
- **References**
- **For More Information**



### **Introduction**

One of the most distressing aspects of a diagnosis of FTD is the fear that the disorder may be inherited, and that the patient's children or siblings might be at increased risk for the same condition. Although the genetics of FTD are not completely understood, research programs throughout the world are increasing our knowledge each year, and, thanks to the generous participation of patients and families in genetic research studies, the picture is beginning to come into focus.

Research continues to show that the proportion of FTD cases with a strong genetic component is small, although the extent to which genetic susceptibility plays a role in the majority of cases is not as clearly understood (see graph above). This review provides a summary of our current understanding of the genetics of FTD, some basic information on genetics that will help you better understand this summary, and advice on important steps you can take to best clarify the inheritance risks in your own family.

When asking questions about your own family's situation, it is important to keep a few general principles in mind:

- The answer to: "Is FTD being inherited in my family?" can best be found by working with a trained health professional (a genetic counselor or a specialist in neurogenetics) and by gathering accurate information from family members. Sometimes it is difficult to determine an answer to this question. Family members may have died of other causes before the age at which they would have developed FTD or contact with family members may have been lost. Regardless, getting as much information as possible is helpful.
- Accurate genetic risk can only be determined if there is an accurate diagnosis. Some FTDs cannot be accurately diagnosed until autopsy; this is an important issue to discuss with your clinician. Genetic counseling is essential to help identify and accurately interpret risk, provide

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## **GENETICS AND FTD/PPA, CONTINUED**

education, and explore the benefits and limitations of genetic testing for you and your family.

- Whereas we once thought of disorders as either being genetic (inherited) or not, modern science is revealing that many, if not most, medical conditions fall in a middle, gray area. The genes one has inherited provide a general background risk (low, average, or high) for a condition, and specific factors in an individual's life - behaviors, environment, even chance—determine whether the person actually develops that disorder. Although no specific environmental risk factors have been linked to FTD, in all likelihood, overall risk for most people will be a combination of heredity and environment.
- For many families, the answer will lie (frustratingly!) in this middle, gray area, where it appears that the risk to family members is higher than that of the general population, but no specific risk number can be quoted, and the precise cause of this elevated risk is not yet known. In such cases it is a good idea to stay in touch with your medical team (and AFTD), as new information on your family and research advances may enable them to re-evaluate and give your family a more specific risk in the future. There is no question that genetic testing will improve and prove to be a valuable diagnostic tool in the future for more families and individuals with FTD.

One of the difficulties of researching the genetics of FTD is that it is so rare. Scientists need cases and cases are hard to find. Research programs are critically important to this effort; it is only through the generous participation of patients and their families that we will advance our understanding of the role genetics plays in FTD. If your family would like to be involved in genetic research, ask your doctor or the AFTD how to get involved in a study.



### **The Importance of Your Family Medical History**

When a physician or healthcare provider is evaluating a patient for a diagnosis of FTD or similar neurodegenerative condition, information regarding the family history can help to determine a diagnosis. Thus, a detailed family history is a valuable diagnostic tool. It is worth the time and effort to contact relatives and obtain the most accurate details of family structure and medical information. It is important to document the information in a meaningful way that is accessible and easy to read. (For more information on the importance of knowing your family's health history, and tools to help you gather and organize the information, see the U.S. Surgeon General's Family History Initiative at [www.hhs.gov/familyhistory](http://www.hhs.gov/familyhistory).)

Another important point to consider in gathering information about your family history is the variable way a genetic disorder can be expressed. In other words, a mutation in the same gene can cause FTD in one person, a Parkinson's disease-like condition in another relative, and ALS (Lou Gehrig's disease) in another relative. In one of the rare inherited forms of FTD, a muscle disease or a bone disease can be caused by the genetic mutation. It is therefore extremely important to gather as much medical information as you can about your relatives, even if you may think that some of the information is not relevant.

## GENETICS AND FTD/PPA, CONTINUED

Geneticists and genetic counselors create a **pedigree**, which is a graphic description of family structure and health history, to record information collected from patients and families. Determining the quantity and quality of information to collect can be difficult. Ideally, one should research at least three generations of relatives, which includes:

- First-degree relatives: children, siblings, and parents
- Second-degree relatives: half siblings, aunts, uncles, nieces, nephews, grandparents, grandchildren
- Third-degree relatives: cousins

The type of medical information to obtain on relatives can include:

Medical Information to Gather on Relatives:	
Vital Status (Living or deceased)	
Age (Date of birth)	
Age at death	
Cause of death	
Autopsy (if one was performed)	
Pregnancy, miscarriages and stillbirths	
Infertility	
Individuals with previous genetics evaluation	
Environmental exposures	
	Radiation, alcohol or drug abuse, tobacco
Health history	
	Birth defects
	Mental retardation
	Deafness, blindness
	Chronic childhood illness
	Cancer
	Neurological conditions (e.g. epilepsy, migraines, strokes, multiple sclerosis, Parkinson's disease, ALS (Lou Gehrig's disease))
	Mental illness (e.g. bipolar disorder, schizophrenia, OCD)
	Dementia (Alzheimer disease, senility)
	Ages of diagnosis for all of the above

When going back and obtaining medical information from previous generations, it is important to note that many of the medical terms we use today, such as FTD and Corticobasal Degeneration, were not used. Therefore, many individuals with neurodegenerative conditions would have been told that they had “dementia” or “senility”. In such cases, it can be useful to try and gather more descriptive information. For example, it is useful to ask about early disease symptoms (e.g. if the individual had problems speaking as a first symptom, or if he/she had a personality or behavior change). It is also important to try to determine an estimate for the age of onset of symptoms.

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## ***GENETICS AND FTD/PPA, CONTINUED***

Another way to determine diagnosis in deceased relatives is to inquire about autopsy. If an individual had an autopsy, the autopsy records (as well as other records, MRI reports and brain biopsies) can be requested and these records can be most informative. Many discoveries have resulted when brain tissue originally obtained from an autopsy that was performed years or decades previously was re-examined with modern techniques. Therefore, if a relative underwent an autopsy years ago, it is extremely helpful to work with the physicians and staff at the facility where the autopsy had been performed, and request that the tissue and slides be sent to a neuropathologist with expertise in FTD. Confirmation of diagnoses with medical records from previous evaluations and laboratory studies is also crucial. If anyone had genetic testing, a copy of the test result is essential.

The family medical history, or pedigree, can be a powerful diagnostic tool to a clinician evaluating a patient. The pedigree can be utilized as a diagnostic tool in the following ways. It helps to:

- establish pattern of inheritance;
- identify individuals in the family at risk for the condition;
- determine strategies for genetic testing; and
- screen for medical risks (such as cancer and heart disease).

Family history information needs to be respected and treated appropriately by healthcare providers and individual family members. Contacting relatives and asking about personal information is not an easy task. Navigating through the complex interpersonal relationships and personalities in a family can be emotionally difficult and stressful. This is a give-and-take process. When calling a relative, it is important to state your intentions or reason for collecting the information. Offer to re-contact family members with information that you learn about your loved one's diagnosis and how it may affect them. Respect an individual's right for privacy – some individuals do not want to contribute any information and this point of view must be respected. If you go through the effort of obtaining a family history, be sure to document the information clearly and secure it in a location that is accessible to other family members and future generations.



### **Genes and Inheritance: Understanding the Basics**

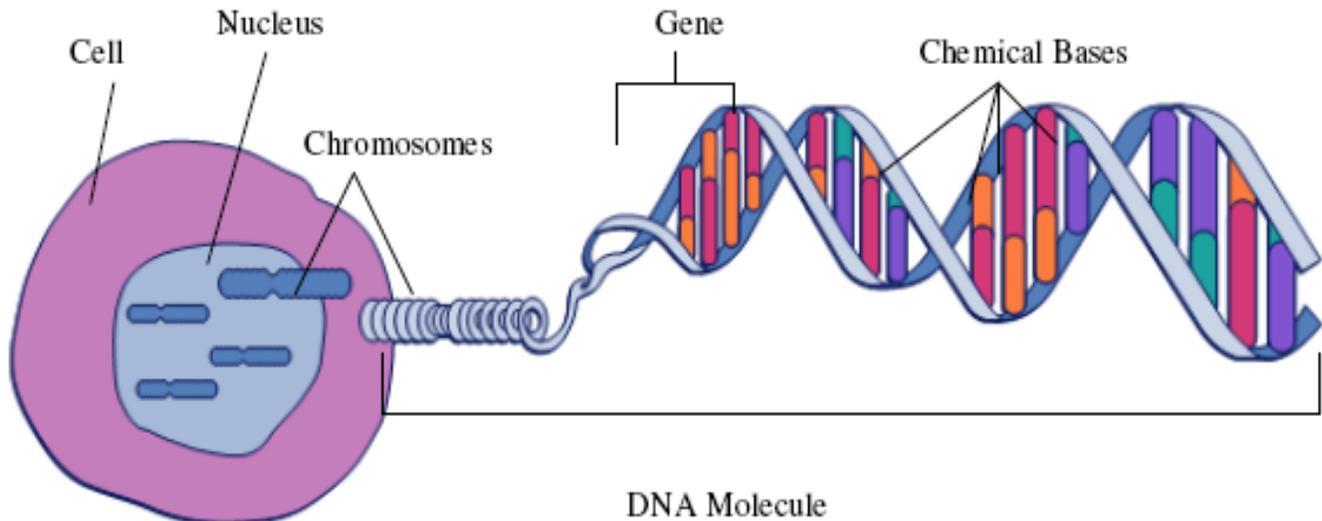
**DNA** (deoxyribonucleic acid) is a chemical that is the most basic unit of genetic information.

**Chromosomes** are highly organized structures containing DNA in long strands (see illustration).

Most cells in our body contain a complete set of 46 chromosomes, or 23 pairs. The chromosomes are numbered 1-22 (largest to smallest) and the twenty-third pair are the sex chromosomes, which determine our gender (two "X" chromosomes = female; one "X" and one "Y" chromosome = male). We inherit our chromosomes at the time of conception: one set of 23 from our mother and one set of 23 from our father. As we grow from a single cell into a complex human being, our chromosomes are copied into each new cell.

## GENETICS AND FTD/PPA, CONTINUED

**Genes** are specific subunits or groups of DNA along the chromosomes. Just as our chromosomes come in pairs, so do our genes. Most genes code for proteins (or chemicals) that have a specific function in the body.



The following analogy may be helpful: One can think of a gene as a long word. Every letter in the word is a piece of DNA. Just like words, genes must be correctly “spelled”, or have the correct DNA code in order to function properly. There are two types of “misspellings” that can occur in our DNA.

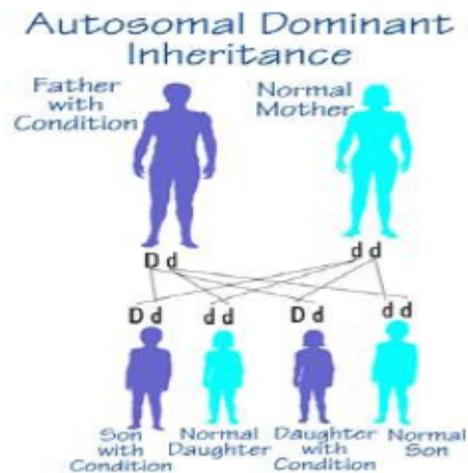
- One type includes words with multiple spellings but with the same meaning or a misspelling that is silent and allows the word to still be read correctly. For example, the word “theater” is sometimes spelled as “theatre”. Despite this alteration you still understand the word and its meaning. This type of alteration in the DNA code is called a **polymorphism**.
- The second type of misspelling involves changes to the word that alter the meaning or make the word unreadable. An example is if the word “good” were changed to “gxod”. One would not be able to make sense out of “gxod”. This type of misspelling or change in the DNA code is called a **mutation**. **Mutations** alter the function of the gene and are often associated with disease.

Inherited conditions can be passed on (inherited) in families in different ways.

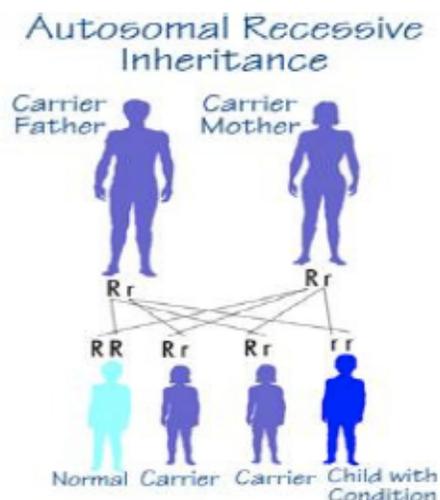
**Autosomal dominant** conditions affect males and females equally, and only one gene of the pair needs to be abnormal for the individual to have the condition. Every child of an individual with an autosomal dominant condition has a 50% chance of inheriting the mutation and having the disorder (Huntington’s disease and achondroplasia (a common form of dwarfism) are examples of autosomal dominant conditions.) When examining a family history for an autosomal dominant condition, often times one will identify multiple individuals in each generation with the condition. It is important to understand that **if an individual did not inherit the abnormal gene then he/she cannot pass it on.**

## GENETICS AND FTD/PPA, CONTINUED

**Features of Autosomal Dominant Inheritance:** The condition appears in multiple individuals in multiple generations, and each affected individual has an affected parent. Any offspring of an affected parent has a 50% risk of inheriting the condition. Non-affected individuals do not transmit the condition to offspring. Males and females are equally likely to have the condition



**Autosomal recessive** conditions affect males and females equally, but **both** copies of the disease gene need to be abnormal for the individual to have the condition. Autosomal recessive conditions can be passed on when each parent is a “carrier” for the condition, and their offspring have a 25% risk of inheriting the condition. “Carriers” have one abnormal copy of the gene but do not have clinical symptoms and **are not at increased risk to develop the condition**. A family history of a recessive condition can reveal multiple individuals in a single generation (brothers and sisters) with the condition; in the case of small families, however, there may be only one affected individual. Autosomal recessive conditions also appear more frequently among individuals with the same ethnic background or among individuals who marry within the same family. (Sickle cell anemia and cystic fibrosis are examples of autosomal recessive conditions.)



**Features of Autosomal Recessive Inheritance:** The condition appears in multiple members of a sibship, not in the parents or offspring. If the family is small there may only be one affected individual. The recurrence risk for each sibling is 25%. Males and females are equally likely to have the condition. Parents may be consanguineous (related).

Other types of inheritance include conditions that are linked to the sex chromosomes (X-linked) or those that are only passed on through maternal transmission. Neither of these patterns has been identified in the inheritance of FTDs.

Based on genetic research, we now appreciate that when one examines the family tree not all “genetic conditions” show a clear pattern of inheritance. This is because not all are caused by a single gene. Rather, many conditions, especially neurodegenerative conditions, are caused by

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## GENETICS AND FTD/PPA, CONTINUED

changes in *multiple* genes that create a *susceptibility*, or increased risk, for the condition. When individuals with this increased risk encounter an additional environmental influence (e.g. head trauma or infection), the medical condition then appears. Conditions that are caused by both genetic and environmental influences are called multifactorial.

Often, multifactorial conditions are seen in multiple family members but without a specific pattern to the inheritance. Thus, another term used for these conditions is “**familial**”.



### Genetics of FTD

FTD can be **sporadic**, **familial**, or **hereditary**.

In the majority of cases, FTD is **sporadic**, meaning it is a disorder that develops by chance rather than being inherited. When FTD is diagnosed in a patient with no family history of FTD or dementia, it is an isolated (sporadic) case, which appears to pose minimal (if any) elevated risk to family members.

Approximately 20% to 40% of patients have a positive family history for FTD or a related neurodegenerative condition or dementia (e.g. Alzheimer disease, Parkinson’s disease, or ALS). This may indicate that there is a predisposition for this type of neurological disease in the family, and that members of the family may be at increased risk to develop one of these disorders. The term “**familial**” is used to describe this unspecified, but likely increased, risk in relatives. In such a family, a meeting with a genetic counselor, who will take a detailed family history and discuss the possibility of such a risk, may be informative.

Worldwide, only about 10% of FTD patients have a family history that suggests a **hereditary** condition with an autosomal dominant pattern of inheritance. This means there is a clear pattern of FTD-type diagnoses being passed from parent to child, with virtually every patient having an affected parent and each child of an affected person having a 50% chance to inherit the disorder. In some countries, research studies indicate that hereditary FTD is more common.

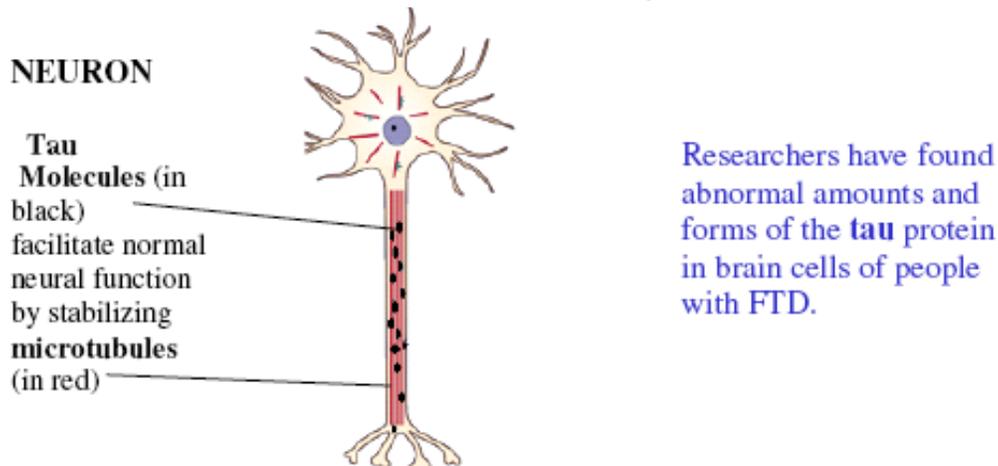
In cases of familial and inherited FTD, the age of onset can be younger than those with sporadic presentations (thirties, and forties for familial FTD), and it is not uncommon for the condition to progress more rapidly. A similar profile has been observed in a subset of Alzheimer’s patients, and dubbed “familial Alzheimer’s”.

For several years researchers have been collecting blood and tissue samples from individuals whose symptoms and family history suggest that their disease has a significant genetic component (either autosomal dominant or familial), to try to determine the specific genetic cause for this group of diseases. This has enabled scientists to identify the fact that, in families with a hereditary pattern of FTD, the disorder is caused by a mutation in one of two specific genes.

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## GENETICS AND FTD/PPA, CONTINUED

These genes are called microtubule associated protein tau (*MAPT*) and progranulin (*PGRN*). **The *MAPT* gene** is located on chromosome 17, and codes for the protein called **tau**. Abnormal amounts of the tau protein have been described in neuropathology (brain tissue) investigations among individuals with FTD, Alzheimer's disease and other neurodegenerative conditions.



The families that were originally linked to the *MAPT* gene were given a more specific diagnosis called frontotemporal dementia with parkinsonism (FTDP-17). This condition was clinically described by the presence of dementia and/or parkinsonism, frontal and/or temporal atrophy of the brain, as well as two or more similarly affected family members consistent with an autosomal dominant pattern of inheritance. Scientists estimate that approximately 20-25% of the hereditary and familial FTD families have a mutation in the *MAPT* gene.

More than 40 mutations in the *MAPT* gene have been found. Many of these families have “unique” mutations that have only been described once. However, there are a few mutations that have been seen in multiple families around the world. In such cases, where the same mutation is seen in different families, specific phenotypic (clinical) features have been compared. These types of studies are called genotype/ phenotype correlations. Such information can be useful for counseling families about the associated clinical findings and prognosis when a known mutation is identified. For example, some mutations are associated with a more rapid progression of the condition.

Many polymorphisms (genetic changes) have been described in the *MAPT* gene that are not directly linked to disease, although there are some specific patterns of polymorphisms, called haplotypes, that have been implicated by association with FTD and related diseases such as CBD and PSP. Researchers are currently trying to determine how specific haplotypes may modify risk for FTD. Studies of individuals with sporadic FTD have not yielded mutations in the *MAPT* gene.

Although inheriting a mutation in *MAPT* means that individual will very likely develop FTD, the exact symptoms and onset cannot be predicted. Even people in the same family with the same mutation may have different ages of onset and different symptoms. Some individuals may develop parkinsonism or motor neuron disease where as others do not. This variability is not understood but may have to do with the influence of other genes.

## GENETICS AND FTD/PPA, CONTINUED

In July, 2006 two different research groups published the finding that mutations in the *PGRN* gene are responsible for causing FTD in another approximately 20 -25% of hereditary and familial patients. More than 60 mutations in the *PGRN* gene have been found. Together **the *MAPT* gene and the *PGRN* gene appear to be the cause of FTD in less than 50% of families with hereditary and familial FTD.**

**The *PGRN* gene** (also located on chromosome 17) contains instructions that tell cells how to make the protein progranulin. Progranulin is the precursor of a factor that stimulates cell growth and wound repair, and too much progranulin has previously been linked to some cancers. Although the precise role of progranulin in brain cells is unknown, FTD patients with *PGRN* mutations do not produce enough functional progranulin. How this relates to the premature degeneration of nerve cells in the frontotemporal region of the brain in FTD is not yet known, but in October, 2006 AFTD awarded a research grant to a group of scientists to develop a mouse model to investigate this question.

Although more studies need to be conducted on patients with a *PGRN* mutation, early results indicate that they have a higher chance of developing progressive aphasia (one of the language disorders known to occur in some FTD patients) and are less likely to have motor neuron disease than are FTD patients without a *PGRN* mutation. Like *MAPT*, *PGRN* mutations show variability in symptom expression. Some family members may present with aphasia while others have behavior disorders. Additionally, the age of onset is even more variable than *MAPT*. In some families, mutation carriers in their 70s do not show symptoms.

Michael Hutton, PhD., who led one of the groups of scientists who made this discovery (and is on the AFTD Medical Advisory Council) adds the following thoughts: “Although only around 5% of FTD patients appear to have mutations in *PGRN*, a much larger proportion have the same neuropathological changes in their brains. As a result, by understanding how *PGRN* mutations cause FTD we are likely to have a much clearer idea of the disease mechanism in this larger group of cases.”

The brains of *PGRN* mutation carriers, as well as some other non-tau FTD cases, have deposits of a protein called **TDP-43**. This substance has also been found in the brains of individuals with other neurodegenerative diseases such as ALS. This substance may lead to important information about common pathways for neurodegeneration. More recently, a certain polymorphism within the *PGRN* gene has been identified as a possible susceptibility factor for familial and sporadic FTD. More studies are necessary in order to duplicate and clarify this finding. *For more information on the *PGRN* research finding see Understanding the Science on [www.theaftd.org](http://www.theaftd.org).*

Several other genes have been associated with various FTD subtypes. These genes are very rare. Research continues to investigate links between some FTDs and ALS (Lou Gehrig’s disease) and some FTDs and Alzheimer’s disease. All of this data provides tantalizing clues, but none of it is yet ready to be applied to patient care.

In the future, genetic research will:

- Identify genotype/phenotype correlations for *MAPT* and *PGRN* mutations that will be useful in providing genetic counseling to families;
- Investigate the role of progranulin and TDP-43 in neuronal functioning and whether substances related to progranulin might serve as the basis for future drugs to treat FTD.

## GENETICS AND FTD/PPA, CONTINUED

- Since the majority of families with hereditary FTD do NOT have a mutation in any known disease-causing gene, the search continues to find additional FTD-associated genes (such as *MAPT* and *PGRN*) which will help researchers further understand the cause and biology of the FTDs.



### Genetic Counseling and Genetic Testing

Genetic counseling is a communication process between an individual (or the family) and a healthcare provider with special training in genetics. Some of the activities in a typical genetic counseling session are:

- Construction of a family history and analysis of that history for patterns of inheritance and genetic risk;
- Education about natural history, genetics and inheritance of a condition;
- Discussion of recurrence risk for inherited and multifactorial conditions;
- Discussion of benefits and limitations of genetic testing; and
- Psychosocial support for individuals and families coping with a diagnosis.

A genetic counselor can also be an important ally in advocating health care needs for patients and families.

Genetic testing can be extremely complex. Not all tests are 100% diagnostic, and often there are ethical and social concerns that influence one's decision to have or not to have genetic testing. Genetic testing for FTD has been offered on a research basis mostly to individuals with a suspected familial or hereditary FTD. There is an important distinction between *research* genetic testing and *clinical* or *commercial* genetic testing. Research testing is not a diagnostic test: There is no guarantee of a result that will be of benefit to the patient. The goal of research testing is to advance scientific knowledge about the condition. This research is usually conducted in academic laboratories. A study coordinator or physician should review the details of the study and obtain informed consent from the patient and/or family members for participation. You should be provided with an informed consent form that has been approved by an Institutional Review Board (IRB) at the university or hospital where the research is being performed.

In contrast to research testing, clinical or commercial genetic testing involves a diagnostic test: The patient will receive an official result from the test. The testing is performed in a laboratory that has been approved for this type of diagnostic testing, to ensure the integrity of the result. Such laboratories are located in hospitals, universities, biotechnology companies and commercial laboratories. Often, clinical genetic testing is only offered after the patient and family have received genetic counseling to explore the benefits and limitations of testing.

## GENETICS AND FTD/PPA, CONTINUED

Clinical genetic testing for *MAPT* and *PGRN* is available for diagnosis of individuals with or at-risk for hereditary forms of FTD. However, these tests will not be useful to all individuals. For example, genetic testing to identify disease-associated mutations in the *MAPT* or *PGRN* genes will usually only be recommended to individuals with a dominant family history of FTD because of the improbability of finding a mutation without a family history. As more causative genes for FTD are identified, research testing will move to the clinical arena, further expanding the role of genetic testing for diagnosis of FTD.

Many individuals who are presently asymptomatic but believe that they are at increased risk for FTD based on family history have expressed an interest in genetic testing so that they can learn if they “inherited the gene.” Presymptomatic or predictive genetic testing is not a new concept in coping with neurodegenerative conditions. Such testing has been offered to individuals at risk for Huntington’s disease, familial Alzheimer’s disease and other genetic conditions. Predictive testing is *only possible* when a mutation in a known causative gene has been identified in a family member affected with FTD.

For the vast majority of these conditions, FTD included, there is no treatment. Therefore, an individual who has the presymptomatic genetic testing may learn that he/she is destined to have a progressive, debilitating disease, and that current medical knowledge cannot prevent this. Obviously, such information can be devastating to the individual. For this reason, genetic counseling *before* the test is performed is critically important. Many clinical centers and laboratories require that the individual go through a formal protocol of pre- and post-test genetic counseling along with psychiatric evaluation.

During genetic counseling, the individual will have an opportunity to discuss his/her motivation for testing, explore the possible outcomes and anticipated reactions, identify coping strategies for each, develop a support system, and discuss the risks associated with receiving a diagnosis (such as adverse psychological outcome, insurance or employment discrimination). The genetic counselor will also work to document the specific genetic abnormality or mutation in an affected family member, to confirm the diagnosis before testing other at-risk individuals in the family. Genetic testing for untreatable conditions is not recommended for at-risk children because they are not able to provide informed consent and giving a child a “label” may do great harm. Most experts will not pursue genetic testing in anyone younger than 18 years of age.

Genetic counselors have Master’s level training in human or medical genetics or genetic counseling and are certified by the American Board of Genetic Counseling. To locate a genetic counselor who specializes in FTD, contact your local university-based FTD or dementia program. If this is not feasible, consult with your clinician or contact the National Society of Genetic Counselors via the internet at <http://www.nsgc.org> and go to the Resource Link to identify someone near you.



# GENETICS AND FTD/PPA, CONTINUED

## Discussing These Issues with Your Family

Genetic issues present a difficult dichotomy within a family. Your genes are what make you unique: no one else in the world has the same exact combination of genes that you do. Yet, you share 50% of your genes with each of your first-degree relatives (parents, siblings, and children). So any genetic issue is both extremely personal and common to all blood relatives.

For this reason, discussing genetic risk can be extremely difficult, even within the most supportive families. When the disorder in question is an adult-onset, progressive, neurological disease like FTD, the issues can be even more complicated. The patient's siblings are often raising their own families, and at the prime of their careers. The patient's children may be too young to understand genetic issues, may be considering marriage and a family of their own, or may already be raising their own children. None of these individuals will want to hear that they might be at risk for the disorder that is afflicting their loved one.

What can a caregiver do regarding communicating potential genetic risk to family members? To what extent is this your responsibility? While there is no one answer for every circumstance, here are some general guidelines and tools to help.

- Meet with a genetic counselor or have a serious discussion with your neurologist about the patient's family history and the likelihood that FTD might be inherited in the family. This is especially important if you have a high level of concern based on family lore about older relatives. This professional can help separate fact from "fiction" and, if there is valid reason for concern, they will devise a plan to get more information.
- If the patient's children are under age 18, there is no rush to have the discussion. Because FTD does not present a risk this early in life, no clinician would perform a genetic test on a minor; the policy is to test only adults who can understand the complicated issues involved and thus give truly informed consent. As with all complex, emotional issues, it is best to follow a child's lead, letting them know that you welcome their questions and making a judgment call as to how much information they are ready to know at any given time.
- Broach the subject with adult members of the immediate family. Chances are that if there is some family history, these people have been wondering about heritability, but have not wanted to broach the subject themselves. Perhaps they are worried about appearing to have selfish concerns about the future, when you are dealing with the daily demands of your loved one. Opening the door to the topic will give them permission to pick up on the discussion, if they want to.
- There are different ways to introduce the topic. You could share copies of a letter from the neurologist that outlines the possibility that FTD is being inherited in the family. Use the exercise of researching the family history to introduce the topic with relatives who may have more information about ancestors. Share this booklet. Remember, it is OK that you do not have the answers to all of the questions; often it can be comforting just to know that others in the family have the same concerns.
- People within the family may react very differently to the subject. Some will want to talk about it; others may be scared and get angry. This is normal, and you should remember that they are not reacting to you, but to the threat of FTD. The best you can do is to let them know that there is a support network, and professionals who can help them find answers if and when they want to.

## GENETICS AND FTD/PPA, CONTINUED

- Different members of the family will choose different paths to address this risk. Some will ignore it; some will want to discuss it with family; still others will pursue professional advice in private. It is important to know that a geneticist or genetic counselor will hold each individual's consultation in the strictest confidence. Information is not shared with other family members without an individual's consent.

You may not know which course each of your relatives takes. All that is important is for you to know that you opened the door to let each of them address it in his or her own way.



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Farmer, J. "The Role of Genetics: A Piece in the FTD Puzzle" (2003). What If It's Not Alzheimer's? (L. Radin, G. Radin, eds). Amherst, New York. Prometheus Books.

Farmer J, Grossman M. (2005) Frontotemporal Dementia: An Overview. *Alzheimer's Care Quarterly*. 6:225-232.

Goldman JS, Farmer JM, Van Deerlin VM, Wilhelmsen KC, Miller BL, Grossman M. (2004) Frontotemporal Dementia: Genetics and Genetic Counseling Dilemmas. *The Neurologist*. 10: 227-234.

Alzheimer's Disease and Frontotemporal Dementia Mutation Database  
<http://www.molgen.ua.ac.be/ADMutations/default.cfm?MT=0&ML=0&Page=Home>

FTD Overview  
<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=600274>

FTDP-17 – mutations in *MAPT*  
<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=157140>

FTDP-17 – mutations in *PGRN*  
<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=138945>

FTD – mutations in *VCP*  
<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=167320>

FTD – mutations in *CHMP2B*  
<http://www.ncbi.nlm.nih.gov/entrez/dispomim.cgi?id=600795>



## ***GENETICS AND FTD/PPA, CONTINUED***

### **For More Information**

- What If It's Not Alzheimer's?, A Caregiver's Guide to Dementia, Lisa Radin and Gary Radin, editors, Prometheus Books, 2008. The first caregiver manual devoted exclusively to FTD, this valuable book is a wealth of information for patients and caregivers living with FTD. To order a copy, see [www.prometheusbooks.com](http://www.prometheusbooks.com)
- AFTD: [www.theaftd.org](http://www.theaftd.org)
- National Society of Genetic Counselors: [www.nsgc.org](http://www.nsgc.org)
- U.S. Surgeon General's Family History Initiative: [www.hhs.gov/familyhistory](http://www.hhs.gov/familyhistory)

*This article has been updated and expanded from the chapter "The Role of Genetics: A Piece in the FTD Puzzle" in What If It's Not Alzheimer's, edited by Lisa Radin and Gary Radin. (Prometheus Books, 2003)*

*Content updated April, 2009.*

# **RULES OF PPA**

## **BY THE EARLY-STAGE PPA SUPPORT GROUP**

1. It's okay to just listen.
2. Don't be afraid to ask for help.
3. Difficulties with speaking, writing and reading do not begin at the same time. Some challenges are easier to deal with than others.
4. There are good days and bad days, don't be afraid of the bad days.
5. It's not the worst thing that is going to happen. It won't get better but there are people out there with other problems.
6. Speech therapy is good practice.
7. Write out notes for what you're going to say and practice it.
8. Tell other people (e.g., friends and family) about your condition.
9. Carry an "I have PPA card" - it's good for emergencies and to quickly explain your difficulty with language to people. Also keep a phone card with emergency numbers.
10. See a good neurologist who is knowledgeable about PPA.



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**“AFFIRM YOURSELF  
FOR CAREGIVER  
CHALLENGES”**

**JANET EDMUNSON, MEd**

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# AFFIRM YOURSELF FOR CAREGIVER CHALLENGES

## Charles' symptoms

Autopsy Confirmed  
Corticobasal Degeneration (CBD)  
(an FTD disease)



### ***Cognitive Issues & Behavior Changes***

- Word find problems
- Slowed thinking
- Couldn't do math
- Yes/no reversal
- Apathy
- Perseveration
- Impulsivity
- Poor organization
- Lack of judgment
- Obsessions (due to meds?)
- Inappropriate behaviors
- Depression
- Sleep issues

### ***Movement & Other Issues***

- Eye movements
- Didn't know where he was in space (visual-spatial issues)
- Fine motor movements (cortical sensory loss)
- Rigidity (one sided)
- Loss of balance
- Speech
- Swallowing
- Couldn't do movements he had to think about (apraxia)
- Feet didn't feel a part of his body (alien limb phenomenon)
- Difficulties with bladder & bowel function
- Involuntary muscle jerks

 **JANET M. EDMUNSON** author, speaker

[www.janetedmunson.com](http://www.janetedmunson.com)  
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## Affirm Yourself for Caregiver Challenges Worksheet

### **Affirmation #1: *I hold on to my passions***

List your passion(s): \_\_\_\_\_

\_\_\_\_\_

### **Affirmation #2: *I remember the difficult personality changes are NOT my loved one***

What might work in handling changes in personalities? \_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_

### **Affirmation #5: *I look for the gifts***

What gifts have you experienced? \_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_

### **Affirmation #6: *I let the love flow, even when all else is lost***

What are ways you can let the love flow? \_\_\_\_\_

\_\_\_\_\_

\_\_\_\_\_



*www.janetedmunson.com*

*Free weekly caregiver affirmation:*

*www.getresponse.com/archive/caregiveraffirmations*

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I hold on to  
my passions,  
because they  
are  
the essence of  
who I am

I will remember  
that the  
difficult  
personality  
changes are not  
my loved one  
- they are the  
disease

I have a  
reservoir  
of strength  
within me

I give myself  
grace when I  
occasionally  
blow it

I look for the  
gifts that only  
this type  
of challenge can  
afford

I let the  
love flow,  
even when all  
else is lost



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# 2011 CAREGIVER RESOURCE LIST

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**Cognitive Neurology and Alzheimer's Disease Center (CNADC)  
Frontotemporal Degeneration (FTD) & Primary Progressive Aphasia (PPA)  
Caregiver Information and Resources**

**CNADC**

**Cognitive Neurology & Alzheimer's Disease Center**

[www.brain.northwestern.edu](http://www.brain.northwestern.edu)

CNADC-Admin@northwestern.edu

**Clinical Services**

Northwestern Memorial Faculty Foundation

Neurobehavior and Memory Health Clinic

676 N. St. Clair Street, Suite 945

Chicago, IL 60611

For appointments: 312-695-9627

**Education**

FTD and PPA Caregiver Education and Support Conference

Contact: Mary O'Hara, 312-503-0604

**Caregiver Support Group**

Northwestern FTD/PPA Family Support Group

Contact: Darby Morhardt, 312-908-9432

**PPA Patient Support Group**

Contact: Christina Wieneke, 312-908-9681

**National Organizations**

**The Association for Frontotemporal Degeneration (AFTD)**

[www.ftd-picks.org](http://www.ftd-picks.org)

866-507-7222 (Toll Free Helpline)

**National Aphasia Association (NAA)**

[www.aphasia.org](http://www.aphasia.org)

800-922-4622

**National Institute of Neurological Disorders and Stroke (NINDS)**

[www.ninds.nih.gov](http://www.ninds.nih.gov)

800-352-9424

**National Organization for Rare Disorders (NORD)**

[www.rarediseases.org](http://www.rarediseases.org)

800-999-6673 (voicemail only)

203-744-0100

## Legal and Financial Resources

### **Social Security (SSA)**

www.ssa.gov or 800-772-1213

*If the person is working and needs to file for disability, it is best to speak to their employer as well as the local security office. As of October 2008, FTD (including PPA) is one of 50 “Compassionate Allowance” diseases that the SSA has marked for expedited approval.*

### **Social Security Disability**

<http://www.ssa.gov/disability>

### **Medicare**

www.medicare.gov or 800-MEDICARE

*Medicare is a health insurance program for people over the age of 65 or under 65 with certain disability. Part A covers inpatient care without monthly premiums. Part B covers outpatient care with a monthly premium.*

### **Medicaid**

<http://www.cms.gov/home/medicaid.asp>

*Medicaid is a state run health insurance program for low-income individuals.*

### **The National Academy of Elder Law Attorneys**

www.naela.org or 703-942-5711

*An elder law attorney can assist you with legal and financial planning.*

### **Financial Planning Association**

www.fpanet.org

*A financial planner can help you plan for the cost of long term care.*

## Care Services and Resources

### **Eldercare Locator**

www.eldercare.gov or 800-677-1116

*The Eldercare Locator contains information about local agencies that offer day programs, support groups, respite providers, in-home care, case management and care giving services.*

### **National Association of Professional Geriatric Care Managers**

www.caremanager.org or 520-881-8008

*Geriatric Care Managers help families care for older relatives. They act as a guide and an advocate and provide ongoing assessments, referrals and care to an older adult.*

### **Care Services and Resources, continued**

#### **National Private Duty Home Care**

[www.privatedutyhomecare.org](http://www.privatedutyhomecare.org) or 317-663-3637

*The National Private Duty Association (NPDA) represents more than 1,200 care agencies throughout the US. Members of the NPDA meet specific standards for private duty homecare, staff education, ethics and best practices within the home care industry.*

#### **ARCH National Respite Network and Resource Center**

[www.archrespite.org](http://www.archrespite.org)

*Families can search the database for local respite agencies/services.*

#### **Palliative Dementia Care Resources**

[www.pdcronline.org](http://www.pdcronline.org)

*Connects family caregivers with information and resources for palliative dementia care.*

### **Support and Support Groups**

#### **FTD Support Forum**

[www.ftdsupportforum.com](http://www.ftdsupportforum.com)

*An excellent online support forum set up by caregivers of people with various types of FTD, including PPA. While most posters are caregivers, there are special selections dedicated to individuals with PPA and FTD.*

#### **PPA Support Group Online**

<http://health.groups.yahoo.com/group/PPA-support>

#### **Pick's Disease Support Group Online**

[www.pdsg.org.uk](http://www.pdsg.org.uk)

#### **FTD Caregiver Support Center**

[www.ftdsupport.com](http://www.ftdsupport.com)

*Includes a listing of support groups by country.*

#### **Support Groups, by State**

[www.ftd-picks.org/support-resources/us-regions](http://www.ftd-picks.org/support-resources/us-regions)

#### **Share the Care**

[www.sharethecare.org](http://www.sharethecare.org)

*A unique site developed by caregivers that discusses how to organize a group to care for someone who is seriously ill.*

### Special Topics

#### **The International PPA Connection, IMPACT**

[www.ppaconnection.org](http://www.ppaconnection.org)

#### **Camp Building Bridges**

<http://www.freewebs.com/campbuildingbridges08/> or 417-933-2030

*A summer camp for teens 12-16 that have a family member diagnosed with dementia.*

#### **Alimed**

[www.alimed.com](http://www.alimed.com) or 1-800-225-2610

*Source of the Daily Communicator® and other communication materials.*

#### **Dementia and Driving**

[www.thehartford.com/alzheimers](http://www.thehartford.com/alzheimers)

#### **The Genetic Alliance**

<http://www.geneticalliance.org/>

*A resource for information and support about genetic conditions.*

### Alzheimer's Resources

*Although geared toward Alzheimer's Disease, many of the resources listed below are helpful for FTD and PPA as well.*

#### **Alzheimer's Association**

[www.alz.org](http://www.alz.org) or 800-272-3900

#### **Alzheimer's Disease Education and Referral Center (ADEAR)**

[www.nia.nih.gov/Alzheimers](http://www.nia.nih.gov/Alzheimers) or 800-438-4380

### Books

What if It's Not Alzheimer's? A Caregiver's Guide to Dementia

Lisa Radin and Gary Radin, editors. Prometheus Books, 2003.

*The first caregiver manual devoted exclusively to FTD.*

Losing Lou-Ann

Erb, Clinton A. Brandon, VT: Holistic Education Press, 1996.

*An inspiring account of a spouse caring for his wife with Pick's disease.*

Finding Meaning with Charles: Caregiving with Love Through a Degenerative Disease

Edmunson, Janet. JME Insights: 2006.

**Books, continued**Pick's Disease and Pick Complex

Kertesz, Andrew and Munoz, David G. Chichester, U.K.: Wiley-Liss, 1998.

*A comprehensive reference that clarifies Pick's diagnosis compared to other forms of dementia.*

The Banana Lady and Other Stories of Curious Behavior and Speech

Kertesz, Andrew. Trafford Publishing: 2006.

*Nineteen lives are chronicled as told by caregivers, followed by tips for caregiving, a useful glossary of terms, plus FTD references.*

An Evolution of Love: Life and Love with Frontotemporal Dementia

Marie Sykes and Michelle Stafford. Georgetown Publishing, LLC: 2007.

*A memoir describing the life of Bob Sykes, Jr., a man who passed away from FTD at age 50.*

When a Family Member Has Dementia: Steps to Becoming a Resilient Caregiver

McCurry, Susan M. Praeger Publishers: 2006.

The Executive Brain: Frontal Lobes and the Civilized Mind

Goldberg, Elkhonon. Oxford University Press: 2001.

The Dysphagia Cookbook: Great Tasting and Nutritious Recipes for People with Swallowing Difficulties

Achilles, Elayne. Cumberland House Publishing: 2004.

*A recipe book for individuals with dysphagia, a condition of swallowing difficulty that sometimes occurs in dementia.*

A Slender Thread

Davis, Katharine. NAL Trade: 2010.

**Cut out the cards below to keep in your wallet for emergency situations.**



<p><b>The person I am with has Frontotemporal Degeneration.</b></p> <p>This is caused by a condition in the brain that impairs behavior and judgment.</p> <p>Sometimes they may say things or act in a way that seems strange or inappropriate. They are unaware that they are acting in this fashion.</p> <p><b>How you can help:</b> Please be patient. Do not laugh if they speak or act inappropriately. Speak simply and directly.</p> <p>Northwestern CNADC: <a href="http://www.brain.northwestern.edu">www.brain.northwestern.edu</a></p>	<p style="text-align: center;"><u>In Case of Emergency</u></p> <p>Name: _____</p> <p>Address: _____</p> <p>Phone: _____</p> <p style="text-align: center;">Please Contact:</p> <p>Name: _____</p> <p>Phone: _____</p> <p>Relationship: _____</p>
<p><b>I have Primary Progressive Aphasia.</b></p> <p>This is caused by a condition in my brain that makes it difficult for me to say the words I mean to say.</p> <p>Sometimes I may also have difficulty understanding what others are saying to me.</p> <p>I am not under the influence of alcohol or drugs. There is nothing wrong with my hearing, memory or thinking abilities.</p> <p><b>How you can help:</b> Give me time to communicate. Speak simply and directly to me. Do not shout; it does not help. Ask yes/no questions.</p> <p>Northwestern CNADC: <a href="http://www.brain.northwestern.edu">www.brain.northwestern.edu</a></p>	<p style="text-align: center;"><u>In Case of Emergency</u></p> <p>Name: _____</p> <p>Address: _____</p> <p>Phone: _____</p> <p style="text-align: center;">Please Contact:</p> <p>Name: _____</p> <p>Phone: _____</p> <p>Relationship: _____</p>





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- Assistance with ADLs
- All Care Givers are Bonded and Insured

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