

**FTD/PPA Caregiver and Professional Education Conference
 August 10, 2009**

Question and Answer Session

Answered by: G. Peter Gliebus, MD, Emily Langendorf, Darby Morhardt, MSW, LCSW, Mary O'Hara, MSW, LSW, Emily Rogalski, PhD, Sandra Weintraub, PhD, and Christina Wieneke.

Evaluation and Treatment of FTD/PPA

These questions deal with the process of obtaining an evaluation, the necessity of follow-up visits, and medications that are sometimes used for either disease.

At what point should a patient be referred to a specialist?

It is important to contact a specialist in FTD/PPA to evaluate the cognitive and behavioral changes very early in the course of illness. One of the problems frequently encountered by patients and their families is that they have had 2 or more evaluations prior to getting a final diagnosis. Many of the symptoms in FTD/PPA can be caused by other diseases or conditions, some treatable. A specialist is trained to differentiate between FTD/PPA and other conditions.

How do we find a FTD clinic in our area or find a good doctor in our area?

The Association for Frontotemporal Dementias is a good resource for finding a doctor in your area: <http://www.ftd-picks.org/support-resources/us-regions>

Are there any medical centers that are known for care of patients with a diagnosis of FTD with motor neuron disease (MND)?

FTD refers to the cognitive and behavioral changes while motor neuron disease refers to accompanying symptoms of MND/ALS (amyotrophic lateral sclerosis). There are clinics that specialize in ALS but they typically focus on the motor symptoms. Most of the centers that work with FTD can also help or refer patients with ALS to their colleagues at these centers. Northwestern has a FTD specialty and we also work with a clinic known for their expertise in ALS. If our center is not convenient for you, please refer to the AFTD website for a list of referral centers across the country. (www.ftd-picks.org)

Are there any tips on how to make the process of obtaining a diagnosis easier?

Getting a diagnosis of FTD/PPA can be frustrating. It is important to find someone who is familiar with these types of disorders so they can exclude other potential causes of the symptoms. Don't hesitate to ask if the center you are consulting has many patients with PPA or FTD. Be prepared for your appointment by writing down questions beforehand. The visit can be overwhelming and you are likely to forget questions if you don't write them down. Finally, educate yourself on the disease. You're more likely to understand what is happening and what you are being told if you are familiar with the terms.

“Your allies” includes “your doctor” – as a facilitator of a caregivers group, I hear, often, that a diagnosis is difficult to obtain and doctors are not forthcoming with information. How do we

educate the medical community to refer/what are suggestions when a caregiver cannot obtain this information?

Unfortunately, this is very common. People are often misdiagnosed for years before they find a center to provide a more accurate diagnosis. If a family member is feeling that they are not getting their questions answered, or that the physician is not referring them to the appropriate specialist, we would suggest they get a second opinion. They can find a center that specializes in FTD care, resources and research by contacting the AFTD. The AFTD maintains information about diagnostic centers that specialize in FTD by region. The web address is:

<http://www.ftd-picks.org/support-resources/us-regions> or call: 866 507-7222

What kind of doctor(s) should we be seeing? What kind of doctor should be coordinating care? Neurologist? Psychiatrist? Neuropsychologist? Other?

A team of neurologists, neuropsychologists and social workers is most helpful with each professional adding their particular expertise in dealing with medical, behavioral and psychosocial issues. A psychiatrist may also be required for consultation with respect to extreme behavioral issues.

The doctor who diagnosed my loved one is a psychiatrist with a specialty in cognitive disorders. We like him very much and he is a part of a university-based neurology practice. Is there any reason why my loved one should see a neurologist instead of a psychiatrist?

Many different disciplines can be involved in caring for individuals with FTD. In modern times, neuropsychiatrists and behavioral neurologists overlap in their areas of interest. Although your loved one may be well managed by the psychiatrist for the behavioral management issues, if you have never had a neurological examination by someone who specializes in FTD, it might be worthwhile to obtain a one time consultation.

With the advanced technology of brain imaging, along with information from data concerning the appearance of the hippocampus, amyloid and tau presence in addition to the APOE testing, why can't a more positive diagnosis of Alzheimer's be offered before death and brain donation?

All of the advances you mention are all very exciting and promising but not at the point where they can be applied to diagnosis at the individual level. There would be too many false positive diagnoses and false negative diagnoses with knowledge at the current level. However, this is the hope for the future.

How common is it for physicians to be unable to distinguish between FTD and Alzheimer's disease based on symptoms in patients with moderately advanced dementia, even after neurological examination, neuropsychological testing and brain MRI?

It is very, very common. Even in established centers using strict clinical criteria, the final diagnosis relies on the brain autopsy and in the case of FTD the correspondence between the clinical diagnosis and what is found at autopsy is not 1:1. This is an area of research that is receiving a lot of attention in order to improve our ability to predict. In the absence of true biomarkers (that is, blood tests or other biological tests that predict the brain pathology with 100% accuracy) we can only use the best estimates, which rely on the clinical examination.

How can a team be assembled when a professional is limited by confidentiality? How can non-medical professionals alert doctors of the need for a differential diagnosis?

If a non-medical professional notices a problem, they should first speak with the patient and caregiver and urge them to obtain an evaluation from a doctor who specializes in FTD/PPA. Additionally, you can educate other professionals about the importance of the differential diagnosis without disclosing a person's name or identifying information.

How often should neuropsychological testing be done?

Depending on the stage of illness, testing done annually gives us markers for the rate of progression. In these diseases, changes usually are apparent at 12-month intervals – any sooner and they may not be observed.

Should we be having regular neuroimaging? What kind?

Depending on the disease and its progression the necessity for repeat imaging is determined by your physician on an individual basis. Repeat imaging is usually done for research purposes.

Can functional MRIs help to show if PPA patients comprehend things?

Functional MRIs are used experimentally for research at this point and they are most powerful to examine differences between groups of individuals.

What medications are used for FTD?

So far there is no FDA approved treatment for FTD. The behavioral and psychiatric symptoms could be managed by the appropriate medications evaluating each patient individually.

Can you explain medications for behavior management? My brother has developed a dangerous obsession with alcohol and I've been told that Ritalin could be effective in curbing his obsession.

There is no evidence that Ritalin is helpful in treating any symptoms of FTD. Antidepressant medications could be an option for treatment obsessive symptoms of FTD. It is important to obtain consultation with a geriatric psychiatrist or other medical professional who is experienced in FTD.

Is the Exelon patch strictly for Alzheimer's Disease?

It could also be used for Diffuse Lewy Body disease, vascular dementia, and if one suspects atypical Alzheimer's disease presenting as FTD or PPA. The above-mentioned indications for use in other than Alzheimer's disease conditions are off-label.

My father with FTD was recently put on Risperdal, but I've heard it shouldn't be used for patients with dementia. Is that true?

There is evidence that antipsychotic medications (and Risperdal is one of them) increase mortality in patients with dementia, so it should be used only if behavioral and psychiatric symptoms are very severe and cannot be controlled by other interventions.