COGNITIVE NEUROLOGY AND ALZHEIMER'S DISEASE CENTER

of the Northwestern University Feinberg School of Medicine

FTD/PPA Caregiver and Professional Education Conference Question and Answer Session

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Risk Factors/Diagnostic Relationships/Genetics

These questions ask about potential risk factors and related diseases to FTD/PPA. They also explore the risk to family members for developing FTD/PPA.

Are there environmental risk factors for FTD/PPA?

The risk factors for PPA/FTD are largely unknown. The e4 allele of the apolipoprotein E is a risk factor for Alzheimer's disease but not a risk factor for PPA/FTD. Research suggests that learning disabilities, especially dyslexia, may be more frequent in PPA patients and their first-degree relatives. However, the exact nature of the risk is unknown.

Does it hurt for the person with FTD/PPA to drink alcohol?

Excessive alcohol use is not good either for patients or for healthy adults. There is some evidence that several glasses (no more than 1-2 per day) of red wine per week might be helpful for our brain and health overall.

How does lifestyle (diet, exercise, alcohol, drugs) impact onset of PPA/FTD?

We know that there are lifestyle factors that can influence Alzheimer's disease in older individuals but at present we have almost no information on these factors in FTD and PPA.

Any further exploration of the idea that vasectomies are connected to PPA?

There is no further information on this issue.

Are there any gender differences in FTD/PPA?

The epidemiology and risk factors of FTD/PPA are largely unknown. Early reports indicated PPA is more common in men than women. It is not clear whether this demographic profile remains true today.

There appears to be a dominance of FTD/PPA with the male population (vs. female). Any research to explain?

There are more women than men with Alzheimer's disease. We do not yet understand what makes one gender more vulnerable than another to different forms of neurodegenerative disease.

Have you found any correlations with weight loss and PPA?

There has been no research to date investigating the connection between weight loss and PPA. Weight gain or loss may occur in bvFTD if the individual has had a change in the type of food he/she likes.

Are Parkinson's symptoms relevant to this disease? In our case, Parkinson's is worsening. Perhaps it's not FTD?

There are forms of FTD that have Parkinsonism (symptoms of Parkinson's disease but not the entire spectrum) associated with them. Some familial forms of FTD caused by tau genetic mutations on

chromosome 17 are associated with Parkinonism. In addition, there are other motor disorders, corticobasal degeneration (CBD), that cause FTD and also produce motor symptoms. FTD is a clinical diagnosis. CBD and any of the other forms of frontotemporal lobar degeneration that cause the FTD can only be diagnosed by brain autopsy.

Explain digestive issues. My husband is having a terrible problem digesting, eating, and swallowing. He is losing weight and has intestinal pain. The neurologist says it is from FTD and suggests a feeding tube soon. What does this have to do with FTD? (GI tests negative other than irritation in stomach).

One of the causes of PPA/FTD is corticobasal degeneration that also produces motor symptoms as the illness progresses. There are other forms of degeneration that also affect motor systems. This can lead to problems with swallowing but we have not heard of intestinal issues otherwise.

Can you speak about a connection between FTD and ALS? My husband has FTD and his sister recently died after a brief, but very devastating experience with ALS (first sign of ALS in February and death in June). Mayo Clinic suggested a connection.

They seem to be related at the level of the pathology in the brain that produces both disorders and the proteins that may be involved in the pathology. We do no have a great deal of information at present.

Is there evidence that head trauma, e.g. multiple concussions, can lead to PPA? My husband had 4 "football" and one car accident concussions.

We think that head trauma is one of the risk factors for several different types of neurodegenerative diseases of the brain but there is no direct link from head injuries to PPA per se.

What is the relationship between FTD and Bipolar Disorder? My husband was diagnosed with FTD at 52 years old. His daughter was diagnosed bipolar in her early 50s – many symptoms are the same.

It appears that in family members of individuals with FTD, there may be a higher frequency of psychiatric disorders than in the general population. This may have to do with inherited predispositions to develop FTD or some other chronic psychiatric disorder in different family members. However, a diagnosis of bipolar disorder can also often be erroneously made in someone with psychiatric symptoms of FTD.

Have you noticed a relationship with FTD, sleeping disorders, and Asperger's syndrome?

There has been no research to date investigating the connection between sleeping disorders, Asperger's syndrome, and FTD/PPA.

Could there be a connection between sleep disorders such as sleep apnea and PPA brain cell deterioration?

We do not know of any connection between sleep disorders and PPA. However, sleep disorders can affect brain functioning independently from PPA. Some individuals with sleep disorders have difficulty concentrating when they are awake. This could contribute to cognitive symptoms but does not cause the symptoms of PPA.

Is PPA any way related to cancer? My husband had 2 types of cancer (renal and carcinoid), now last year he was diagnosed with PPA.

The link is not an obvious one since cancer is fairly frequent and PPA infrequent. However, there has been no research to date investigating the connection between cancer and PPA.

Is there any relationship between FTD/PPA and illnesses such as diabetes and hypoglycemia?

There has been no research to date investigating the connection between diabetes or hypoglycemia and PPA.

Is FTD/PPA hereditary?

Cases of FTD/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% chance of developing it. If the disease is inherited, it is most likely that there is an instance of FTD/PPA in every generation of the family (meaning that the affected person would have an affected parent). In familial cases, family members of an affected individual have an increased risk of developing the disease, although this increase is not well defined. A case of FTD/PPA is defined as familial when there is a family history (in first degree relatives, such as children , parents or siblings) of FTD/PPA, a related dementia such as Alzheimer's or related neurodegenerative condition such as ALS. 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. Source and more information: http://www.ftd-picks.org/frontotemporal-dementias/genetics

What are the genetic risks if there are two FTD-type dementias in a family, e.g. to same-sex child of one of a pair of siblings?

We do not know the answer to this and many other questions because we don't have enough information on a sufficiently large number of individuals with FTD. However, the only way to know if there is a genetic mutation in a family is to have the affected members genetically tested for known mutations. There are no tests for unknown mutations.

Dr. Mesulam mentioned that a new gene related to PPA has been found. Can you elaborate on this, please?

In 2007, we published a report (in the journal Archives of Neurology) of two families with a mutation in the progranulin gene and several family members affected with PPA. However, this is very rare.