Progression of FTD: A Caregiving Perspective

Introduction.

One of the more distressing feelings for the family that accompanies a diagnosis of FTD and caring for a loved one with the disease is not knowing what to expect in the future as the disease progresses. In many cases the family has already endured a prolonged and frustrating journey just getting to the point of receiving a diagnosis of FTD, a disease they probably never even heard of, only to discover that there are no treatments available for it. There are many uncertainties and questions about what the future holds, not only for the person just diagnosed with FTD but the entire family, for which many physicians are not adequately equipped or experienced to provide insight, counseling or advice.

In the following paragraphs the information about the progression of FTD is based primarily on the personal observations and experiences of some who been through the journey with a loved one suffering from FTD and cared for them over an extended period of time. While there may be more objective clinical criteria for defining stages of FTD and dementia in general, what is presented here is from a caregiving perspective. Although the information is subjectively divided into disease stages for the purpose of establishing a frame of reference, the intent is to provide answers to some questions about what to possibly expect as the FTD advances and the caregiving needs become more demanding for which the defining or characterization of the stages themselves is not that important. Also, while there are those in their sixties and older who are diagnosed with FTD, the disease is generally regarded as an early onset form of dementia that primarily appears between the approximate ages of 40 to 60. Consequently, much of the focus and descriptions here relate to those with FTD in this age range.

Frontotemporal dementia is comprised a several variants of the disease that with onset initially affect either behavior or language abilities. Consequently, each case of FTD can vary to a great extent from one another in the specific behaviors and symptoms that arise. The information here is not in any way intended to be an authoritative compilation of symptoms and behaviors that will develop. It is only intended to provide some insights gained from others who have witnessed and had to deal with some of the devastating and progressive effects of FTD with the prospect that the knowledge may in some manner better prepare or at least give some understanding for what future challenges and issues may surface in a journey with someone suffering with FTD.

Also, with each case of FTD having its own unique characteristics an accurate prediction or description of the actual course of the disorder and likely life expectancy for any specific case is not possible. By describing what others have experienced, though, it may provide some markers by which others may at least find partially useful in assessing the progression of FTD with their loved one.

Rate of Disease Progression.

There is no typical pattern for progression of FTD. With each case of FTD the rate with which the disease progresses and the type of symptoms and the order with which they appear can vary.

The mental, physical and emotional decline brought on by FTD may occur gradually in a somewhat continuous process over time. Alternatively, these declines may occur in more of a step-wise fashion characterized by comparatively short periods, perhaps days to weeks, of rapid decline followed by longer periods of relative stability. With the step-wise decline pattern the periods of relative stability may become shorter in the later stages of the disorder.

Life Expectancy.

The life expectancy for someone diagnosed with FTD reported in the scientific literature varies. Generally articles state from about 3 to 10 years after diagnosis on average but some FTD patients can live more than 20 years. Ultimately, not enough is known or understood about FTD to give any reasonably accurate prediction of life expectancy. In some reported cases FTD patients have declined rapidly and lived for only two years or less after symptoms were recognized. With others the disease has progressed much more slowly and patients have lived for 10 years or more following the appearance of the first symptoms. With all the difficulties recognizing and diagnosing FTD, in general, there is an inherent high degree of uncertainty with just as to when onset really occurs and therefore life expectancy.

The Beginning Stage: Onset of Early Symptoms.

FTD patients are generally categorized into one of two main groups depending on the initial primary symptoms in the beginning stages, those relating to either behavior or language problems. As the FTD progresses to later stages, though, regardless of whether the initial symptoms manifested were behavior or language based, it is not uncommon for the caregiving issues and challenges to become increasingly similar in nature.

At the onset of behavior variant FTD and usually before a diagnosis is even made those with the disorder will typically exhibit behaviors ranging from what may simply seem as uncharacteristic for them to what is unquestionably socially inappropriate for anyone. Many uncharacteristic behaviors, as out place as they may be, may draw no more attention than a momentary quizzical pause or be regarded as just an anomaly without additional thought. By themselves they are likely not to raise any major concerns with those close to the person and likely be even less noticeable as unusual with others. The uncharacteristic behaviors might fall into a broadly defined range of “normal” in a general population but normally would be out of place for the specific individual. Much more noticeable are the socially inappropriate behaviors that may be exhibited. As one example when reading or hearing about other cases where a diagnosis of behavior variant FTD is eventually made, it is not uncommon for sexually inappropriate behaviors being exhibited prior to the diagnosis. While these behaviors bring negative attention to the individual, make others around them feel uncomfortable, or even draw reprimands with the more egregious trespasses, most of the time the individual may appear to function and behave normally like their usual self. As unsettling and disturbing as the occasional aberrant behaviors can be, at first there may be not an immediate sense that anything is seriously wrong.

For those with a language variant of FTD their ability to communicate and or understand communications are the early symptoms manifested. In comparison to those with the behavior variant of FTD, it is typical for much thought or notice, if any, to be given by others to these early communication lapses of those with a language variant of the disorder. It is very normal for people to occasionally forget a word that they want to use or misunderstand what is said. Where persons with the behavioral variant of FTD often do not recognize anything is odd or inappropriate about their behavior, persons with a language variant of FTD are more likely to be aware that they are having greater difficulty finding the right words to use or understanding what is being said to them. Other than the language problems there may not be any other indications to suggest that something is wrong. Often a person with this variant of FTD may be referred to a speech therapist or language specialist early on for their communications impairment.

Unfortunately, it is not uncommon for someone in the onset stage of FTD to be initially misdiagnosed with depression or some other psychiatric issue, regardless of whether they have the beginnings of a behavioral or language variant of FTD. It may be particularly difficult for the person with the early onset symptoms of FTD to be told they are depressed when they strongly believe that that is not their problem.

The Second Stage: Early Symptoms Progress and New Ones Develop

It may have taken a while but a diagnosis of FTD has been made based mostly on symptoms and perhaps some brain scans. The onset stage of FTD will be followed by a period where the early symptoms are likely to become much more apparent with the uncharacteristic or inappropriate behaviors becoming much more frequent. The person with FTD may also have a growing obliviousness and/or lack of concern for what others may think about them as they appear to become unaware of their growing deficits. In general any awareness of any effects from their actions is likely become greatly diminished to nonexistent in this stage. To the immediate family members it will become quite obvious that there is definitely something wrong with their loved one but to most others they still may not recognize that there is anything really amiss with them. At some point during this stage, probably earlier rather than later, it is no longer safe for the person to be driving.

There is a broad spectrum of specific uncharacteristic behaviors that can be manifested by persons with a behavioral variant of FTD. Whatever behaviors are being manifested, though, it is likely they will become increasingly much more the norm than the exception. What had been an occasional display of poor judgment in making routine everyday decisions may become so frequent that you will likely feel very uncomfortable leaving them alone at home or allowing them to go out by themselves. In some cases the person may become more aggressive or assertive making it more physically difficult for the caregiver to manage them.

During this stage there are specific behaviors that may develop that can be categorized as one or more of several general common characteristics or traits that are frequently associated with FTD: perseveration, obsessive compulsiveness, hyperorality, loss of empathy, apathy, disregard for normal social conventions, and a lack of interest in personal hygiene.

Perseveration is the persistent repetition of a verbal response or action regardless of whether or not it fits the situation. Sometimes the repetitive behavior can be totally harmless such as continually saying a simple meaningless word or phrase in any conversation. Other times it may be something that sometimes can cause a problem but not always such as laughing in response to any situation. Then there are more offensive repetitive behaviors such as always addressing others with a profanity or using threatening gestures as a normal response. Obsessive compulsive behaviors are those that are done without really thinking about it and may or may not be a problem. A fairly common obsessive compulsive behavior for someone with FTD seems to be going through the belongings of another person. Some examples of this would be thumbing through the papers on someone else's desk while meeting with them or simply walking into someone's home without any invitation because they want to see what is in there.

Hyperorality is the insertion of inappropriate items in the mouth. With loss of empathy the person may show no signs of emotion or caring when someone else is hurting. They may even exhibit emotions exactly opposite of what would be appropriate in specific situations. One display of apathy that seems to be fairly common is no longer having any interest in the life of their grade school children. Disregarding normal social conventions and losing interest in personal hygiene are two of the more embarrassing general behaviors that can develop.

Diet and weight control can also be a challenge. Eating may become an obsessive compulsive behavior rather than a means to satisfy ones hunger and nourish the body. Substantial weight gain is a common occurrence in this stage when eating is not adequately monitored.

For those with a language variant of FTD during the first several years following onset the dysfunctions are more typically related to communication skills: speaking, writing, reading or language comprehension. There can be an increased frequency of difficulties comprehending the meaning of words being spoken to them, finding the right words to use to express themselves, or being grammatically correct when speaking. Over time their language skills may erode to the point of not being able to communicate very easily if at all. It seems, though, that it is not atypical for the person to retain at least one mode of communication during this time. While with some language variant cases of FTD there may be little deterioration in judgment and behavior accompanying their deterioration in language skills, it is not uncommon for uncharacteristic or socially inappropriate behaviors to start after several years or more following the initial onset of the language and communication problems. It is during this stage where behavioral dysfunctions similar to those associated with behavioral variants of FTD can become readily apparent in those with a language variant of the disease. If behavior anomalies do arise, they are likely to develop over a period of time and not just suddenly appear.

From a caregiving perspective this stage following the onset of symptoms may be one of the more frustrating and some ways one of the most tiring ones. Early on in this stage it is common for the person with FTD to still function reasonably well by them self. They may be capable of taking care of all the basic activities of daily living (eating, bathing, walking, toileting, dressing and continence) reasonably well by them self despite their inappropriate behaviors and poor judgment. It is likely, though, they will have increasing difficulty managing the instrumental activities of daily living such as shopping, cooking and taking care of their finances. This can often lead to conflicts because the person is insistent on doing these instrumental activities of

daily life but they cannot be trusted or depended on to do them correctly by them self anymore. The caregiving challenge will be balancing between allowing the person to do the activities that they can still manage and to somehow restrict or assist them with those activities that they are no longer able to manage by them self. It is also possible that during this stage the person reaches a point where they are no longer able to recognize that there is anything wrong with them.

Issues with safety in general will become a major concern. Besides problems with the person possibly hurting them self, their poor judgment may place them in potentially unsafe situations. Some examples of this are they may indiscriminately invite any stranger who comes to door into their home or they may decide to take an evening walk by them self in a dark secluded area.

Sleep patterns may also become disrupted with the person not adhering to any consistent schedule.

A fairly common story to read about in FTD cases is for the patient to indiscriminately to spend or give away their money and valuables at some time during this stage. They may accept any solicitation made to them to purchase a product or service regardless of the affordability or need for it. A related behavior may be to give away money or other valuables to family, friends or strangers.

As mentioned earlier preventing them from driving is likely to be a particularly challenging situation when it is no longer safe for them to do so. Other behaviors that can be challenging may relate to eating. As an example you may have just finished eating a meal and they are insistent on eating again. You may find that the person is becoming less cooperative when trying to get them to do something. As an example you may be going to visit friends or family and the person simply refuses to get into the car unless they are allowed to drive even if they do not know where you are going or how to get there anymore.

Family and caregivers will likely experience additional frustrations and stresses with the person suffering from FTD while still able to perform the activities of daily life because of their poor judgment. As an example the person may still want and for the most part be able to cook but they may no longer always distinguish between plastic and metal containers to use in an oven, in a microwave, or on the stove. Consequently, they may use a plastic container on a stove burner with which cook. Ultimately, you will likely reach a point where you feel you need to monitor the person’s every action even though much of the time it may not be necessary to do so.

One common frustration that the family is likely to experience will come from the lack of understanding, regret or dismay expressed by the person for any negative outcomes or consequences that result from their poor judgment. While you know their FTD is the cause of this, it will still be difficult for you to not feel they are doing it on purpose and are capable of controlling them self more than they are.

One interesting frequent observation is that those with FTD seem to often enjoy or relate to music. They may like to listen to it, sing songs, or in some rare cases even show a new found talent for it. For those who have a language variant of FTD with a severely limited vocabulary

and not able to verbally express themselves anymore they may still be able to sing when shown the words to a song as if they had no language problems.

There is one final development to be mentioned here that is likely to occur during this stage, incontinence. For many this is a caregiving challenge that will at first seem overwhelming. There are, however, strategies and products that can make dealing with this matter manageable. The incontinence will likely begin as an occasional accident that escalates quickly over a period of perhaps weeks. Whether the person is actually incontinent, i.e., unable to control their functions, or just unaware of when and where it is appropriate to relieve them self, really does not matter from a caregiving perspective, the challenge is the same.

Based just on the differences in the length of survival after onset symptoms the duration of this stage can vary by several years or more. While some with FTD may not live more than 2 years after onset, for others this stage can last 2 to 4 years or longer.

The Advanced Stage: Need for 24/7 Care

At some point in the previous stage it was likely recognized that it was becoming much more difficult for the person with FTD to manage all the basic activities of daily living by them self. If it has not happened yet, in the advanced stage of FTD the person is most likely no longer able to manage some if not all the basic activities of daily living without assistance. This is one way to mark or characterize the advanced stage of the disease. Besides having issues with incontinence, there may have developed problems with them feeding them self, bathing, brushing their teeth, walking, getting up out of bed, or dressing and undressing. There may be a marked decline in the person's quality of life because of these developments.

By this stage of the disease the symptoms and therefore the caregiving challenges become much more consistent between patients regardless of what variant of FTD with which they were initially diagnosed. The person is likely to show little if any interest, emotion or response to any situation or stimuli. They may show little or no any indication of being in pain if they are hurt. One common development is that the person may sleep much of the time if allowed to do so. In the advanced stage it is likely that the person will not perform any activity either basic or instrumental without someone else first initiating and then assisting them with it. It may seem that there is nothing that interests them. There is only apathy towards everything.

It is not uncommon for motor neuron deficits such as Parkinson's like symptoms to now be evident if they hadn't appeared earlier. Much less often amyotrophic lateral sclerosis, also known as Lou Gehrig's disease, which affects voluntary muscle movement, may be associated with FTD. The Parkinson's like motor neuron deficits include bradykinesia, constipation, muscle rigidity, impaired posture or balance, loss of automatic movements, and loss of facial expressions. Bradykinesia is where movements are slower than normal. One sign of this may be walking with short, shuffling steps rather than with a normal gait. Constipation can become a major problem resulting from the slowing of the colon muscles used for elimination. Muscle rigidity may also cause body limbs to become stiff with a decrease in the range of motion. In more severe cases muscle contractures can cause limbs to remain in a contracted position.

Problems with posture or balance are serious issues because it makes the person much more prone to falling and injuring themselves, in particular they are much more at risk of falling and fracturing their hip. The lack of eye blinking, sneezing or swinging of the arms when walking are some examples of loss of automatic movements.

If up to now the progression of the FTD appeared to be in more of step-wise pattern, it might be noticed in the advanced stage that the periods of relative stability are becoming shorter in duration. Since by now the person may have lost their ability to carry out many of the basic and instrumental activities of daily life, it can be more difficult to recognize when they lose any more abilities unless it was a major one remaining.

For those patients with the behavior variant of FTD if they haven't shown any problems with language skills, they may start to appear now but not necessarily. For those with a language variant of FTD they are likely, but not necessarily, to become totally mute. With some patients with a language variant of FTD you may now be totally unable to communicate with them in any effective manner. Besides being mute they are unable to understand any form of communication and are unable to write. Others may still retain an ability to partially communicate either orally or in writing though probably in short phrases rather than in full grammatically correct sentences.

As with the earlier stages, the length of this advanced stage is variable. It is quite likely, though, to be shorter duration than the previous stage.

The End Stage: Nearing the End of the Journey

For the person with FTD the progression of the disease ultimately reduces their physical and mental capabilities to the point where vital functions begin to be impacted. One indication the FTD has progressed to the end stage of life can be the development of eating difficulties in the person. In the transition period from the advanced to the end stage a gradual decline in their ability to properly chew foods before swallowing might be observed. It may also have become necessary to cut their food into smaller pieces to make it easier for them to consume it. In the end stage it may now be that the person can only eat soft foods that require little or new chewing to swallow without choking. Feeding them may take much longer as they are slow to consume their food. One challenge may be distinguishing between eating slowly and being reluctant to eat at all. Drinking any fluid with a viscosity similar to water may become very difficult for the person without choking on it. Even though their intake of calories may be about the same as before, there still could be a loss of weight occurring.

If neuromuscular weakness or degeneration symptoms had been evident earlier, ambulation, the ability to walk, may now either be greatly impaired or completely lost. The person may also lose their ability to stand or sit upright without being propped up. The first signs of this may come when the person is sitting in a chair and for no apparent reason falls out of it and on to the floor. As a caregiver this may occur a few times before it is realized that they are now having difficulty just sitting upright in a chair.

Another possible manifestation that may arise is spasticity in arms or legs where they might become rigidly folded with a significant loss in their range of motion. The muscles in the limbs remain in a contracted state so it extremely difficult if not impossible to completely straighten them without causing pain or injury. As brain cells continue to die the normal controls for muscle contraction and relaxation can become impaired leading to the continuous increased muscle tension. For practical caregiving purposes such muscle contractures in an arm or leg it can make it much more difficult any time you need change, dress and undress the person.

Although starting out each case of FTD may have different caregiving challenges, in this final stage it is most likely, regardless of what the symptoms and behaviors were in the earlier stages of the disease, that there will be much more of a commonality in the caregiving needs and challenges that exist. Questions are likely to arise that require difficult decisions regarding the ongoing care that should be provided:

Do they still have a reasonable quality of life?
Is it time to start hospice care?
What are the implications of starting hospice care?
How much longer will they live?
Is the end near?
If they can no longer swallow any solid food or liquids without a high likelihood of choking is it time to stop feeding them and let nature take its course?

These questions and others will weigh on you as you cope with this end stage. There are no set answers, only difficult choices. Each family must determine what is best for their loved one and when it is time to compassionately and humanely let go of them.