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Management of Frontotemporal Dementia in Mental Health and Multidisciplinary Settings

Mary Anne Wylie, RN, APN/PMH[1], **Adriana Shnall, MSW, RSW**[2,3], **Chiadi U. Onyike, MD, MHS**[1], and **Edward D. Huey, MD**[4,5]

[1]Division of Geriatric Psychiatry and Neuropsychiatry, Johns Hopkins University School of Medicine, Baltimore, USA

[2]Ross Memory Clinic, Baycrest Health Science Centre, Ontario, Canada

[3]University of Toronto, Faculty of Social Work, Toronto, Canada

[4]Taub Institute for Research on Alzheimer's Disease and the Aging Brain

[5]Departments of Psychiatry and Neurology, College of Physicians and Surgeons, Columbia University, New York, USA

Abstract

Diagnosis of frontotemporal dementia (FTD) in the mental health setting and issues pertaining to longitudinal care of this population in a specialty clinic are reviewed. FTD is often misdiagnosed as a psychiatric disorder, most commonly as a mood disorder. FTD has features that overlap with those of major depression, mania, obsessive-compulsive disorder and schizophrenia. We describe these features and how to differentiate FTD from these psychiatric disorders. This paper also describes practical issues in the management of FTD, specifically the issues that clinicians, patients and their families face in managing this disease. Areas of clinical care along the continuum are explored; FTD care involves collaborative management of symptoms and disability, and assisting patients and families in adapting to the disease.

INTRODUCTION

Frontotemporal dementia (FTD) features progressive disintegration of temperament and conduct; apathy, indifference, impulsive behaviors, compulsions, hyperactivity, disinhibition, overeating, and socially inappropriate behaviors are typical features. Thus FTD can mimic several psychiatric conditions, and it commonly presents in mental health settings. It is therefore important for psychiatrists and other mental health professionals to become familiar with this condition, the diagnostic process, and the types of care that patients and families will require. This article begins with a discussion of the diagnosis of FTD in the mental health setting, and the role that psychiatric care can play in the management of this condition. We describe comprehensive neuropsychiatric care for FTD and examine some frequently encountered clinical and psychosocial complications of FTD and their management.

Address correspondence to:, Edward D. Huey, MD, Taub Institute for Research on Alzheimer's Disease and the Aging Brain, College of Physicians and Surgeons, Columbia University, 710 West 168th Street, 3rd floor, New York, NY, edh2126@columbia.edu, Phone: +1-212-305-1134, Fax: +1-212-305-2426.

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Primary Care and Mental Health Issues in FTD Diagnosis

The Problem—There is a pressing need to improve the identification of FTD in mental health settings as these patients tend to be initially diagnosed with a psychiatric disorder, most commonly a mood disorder (Mendez et al., 2007; Woolley, et al., 2011). Early identification is important for several reasons. First, early treatment can mitigate suffering and minimize clinical and psychosocial complications. Second, it generally is frustrating for families to have their relative's diagnosis change after having been treated for an extended period (Chow, Pio & Rockwood, 2011). Third, when disease-modifying treatments become available, they will likely be most effective given early in the illness. If diagnosis of FTD is delayed, patients will not be able to receive treatment during a period of potential benefit. At present some patients with FTD never receive the proper diagnosis (Mendez et al., 2007).

There are several barriers to identifying FTD in the mental health setting. The first has to do with the substantial symptom overlap between certain psychiatric disorders and FTD, which can make it difficult to distinguish the conditions – especially early in the disease course (Table 1). Another barrier for the detection of FTD in mental health settings is the low prevalence of FTD compared to psychiatric disorders. It has been estimated that the point prevalence of FTD in the United States is 15 to 22 persons per 100,000 in the at-risk age category of 45–64 year-olds (Knopman & Roberts, 2011), whereas the point prevalence of Major Depressive Disorder (MDD) in adults is approximately 2.6% (Singleton el al., 2003), i.e., about 100 times more common than FTD. Based on the relative prevalence data, patients with, for example, social withdrawal, apathy, and anhedonia in the mental health setting are far more likely to have MDD than FTD. Another issue is that mental health settings often utilize non-physician mental health specialists for evaluation and treatment who usually have had little exposure to FTD and related disorders.

Symptom Overlap—The typical psychiatric misdiagnoses of FTD are MDD, bipolar affective disorder (BAD), obsessive-compulsive disorder (OCD), and schizophrenia (see Table 1). Symptom overlaps between FTD and psychiatric disorders may relate to shared neuroanatomic substrates, which can elucidate the neuroanatomic substrates of the symptoms of FTD and psychiatric disorders. For example, it has been suggested that MDD is comprised of two separate 'latent' factors, "cognitive/affective" (e.g., sadness, poor self-esteem, suicidal state) and "somatic" (e.g., decreased energy, appetite changes, poor concentration; Beck, Steer, & Brown, 1996; Steer et al., 1999). FTD symptoms overlap with the somatic, but not the cognitive/affective factors, suggesting these factors may be anatomically separable (Koenigs et al., 2008). A large literature indicates that the orbitofrontal and anterior cingulate cortices, basal ganglia, and thalamus are involved in the pathogenesis of OCD (Huey et al, 2008). These structures are also frequently damaged by FTD. Patients with FTD commonly have stereotyped behaviors that are similar to the compulsions of OCD, but without the obsessions that characterize OCD – suggesting a differential contribution of specific frontal cortical and subcortical structures to the OCD phenotype (Huey et al., 2008). Recent work suggests a larger "meta-structure" of psychiatric disorders with factors that transcend traditional psychiatric diagnoses. Two overarching categories of "internalizing" and "externalizing" symptoms have been identified with the "internalizing" category consisting of two sub-factors, "fear" and "despair" (Eaton et al., 2011; Krueger, 1999). In this structure, MDD, Dysthymia, and Generalized Anxiety Disorder are largely disorders of despair, Phobia and Panic Disorder of fear, and substance abuse and Antisocial Personality Disorder of externalization. Psychotic disorders such as schizophrenia were not considered in those studies, but one might hypothesize that the 'negative symptoms' of schizophrenia and some symptoms of FTD would constitute a fourth factor corresponding to social and emotional blunting and withdrawal. Further study of the behavioral, cognitive, and emotional effects of neurodegeneration in patients with

FTD has the potential to clarify the neuroanatomy of these core factors of psychiatric illness (Huey & Lieberman, 2012).

Red Flags—Certain "red flags" should alert mental health practitioners to the possibility of FTD; these include cognitive dysfunction, especially aphasia or executive dysfunction. Mood disorders are often associated with mild executive dysfunction on formal testing, but the degree of impairment is usually modest, when contrasted with that observed in dementia (Wright & Persad, 2007; see also Harciarek & Cosentino in this issue). A second red flag is a lack of distress. Emotional distress is a core component of mood and anxiety disorders (Eaton et al 2011) while behavioral-variant FTD (bvFTD) is usually characterized by a pathological lack of distress (i.e., anosognosia and anosodiaphoria). A third finding that should raise suspicion is if the illness is progressive and refractory to treatment. However, some psychiatric disorders, notably schizophrenia, can progress to a syndrome of dementia that includes features of frontal dysfunction with relative sparing of memory(Friedman et al., 2001; Harvey, 2012). Another red flag is an unusual psychiatric presentation, such as sustained manic-like states without grandiosity or euphoria, compulsions without obsessions, schizophrenia lacking hallucinations or complex delusions (although this has been complicated by the recent evidence that up to 38% of patients with C9ORF72 repeat expansions can present with psychosis; Snowden et al., 2012). New onset of a psychiatric disorder in middle age and older patients, while it certainly occurs, should prompt consideration of neurodegenerative disorder. So should any family history of dementia, parkinsonism, or motor neuron disease. Two studies found that abnormal gait, frontal release signs, functional decline, unusual calmness, and violation of personal boundaries distinguished FTD from psychiatric disorders (Panegyres, Graves & Frencham, 2007; Rankin et al., 2008). If FTD is suspected, referral to a center that specializes in the evaluation and treatment of dementia is usually warranted.

What is the Role of Psychiatry in FTD?

Psychiatry is poised to make important contributions to the investigation and treatment of FTD. Psychiatrists specialize in disorders of emotion, behavior, and cognition, all affected in FTD. Furthermore psychiatric medicines are commonly used to treat behavioral features of FTD. Strengths of psychiatry, such as the integration of pharmacologic and behavioral treatments, and involvement of the family in the treatment process, are essential for successful management of FTD. The study of FTD can benefit psychiatry by providing opportunities for examining the neuroanatomy and genetics of psychiatric symptoms (Huey & Lieberman, 2012). Psychiatry has been, historically, considered by some as the field that studies and treats mental disorders that are without an identified cause; this is a regressive view of psychiatry. If schizophrenia were found to be a prion or autoimmune disease, it would not cease to be a psychiatric disorder. Essentially all psychiatric disorders involve the frontal lobes of the human brain. Thus, the study of the frontal lobes, and the disorders that affect them, should be of particular interest to psychiatrists.

Neuropsychiatric care for FTD

One model for FTD management is the neuropsychiatric clinic, which involves multidisciplinary management and care from neuropsychiatry/geriatric psychiatry, neurology, neuropsychology, geriatric psychiatry nursing, palliative care, and social work. Here the integration of evaluation, pharmacologic treatment, behavioral care, and family engagement constitutes a comprehensive approach to patient care. This model of care offers assessment, treatment, case management, and family/carer support (e.g., education and counseling, psychotherapy and support groups), and access to a neuropsychiatry inpatient unit for behavioral exacerbations. These services are provided throughout the continuum of the disease process. While not yet established for FTD, multidisciplinary teams have been

shown to optimize health care delivery and survival for patients with related illnesses such as ALS (Miller et al 2009).

Initial Clinic Contacts and Managing Referrals

Most patients and families who come to an FTD Clinic seek diagnosis or understanding of confounding behaviors. Initial contacts include those seeking an evaluation, second opinion or specialist care, as well as information about the disease and/or access to resources such as research studies, community-based services, support groups, day programs, residential care and the like. Unfortunately, barriers to specialist care exist, including the afflicted individual's resistance, family disagreement about a course of action, dearth of specialists, lack of proximity, and type of insurance coverage. However even for those who do not attend a specialist clinic, print materials about FTD and information about support groups and advocacy organizations (particularly the Association for Frontotemporal Degeneration, AFTD).offered.

The initial encounter is at the diagnostic evaluation. Whether for the purposes of establishing a provider of care or a second opinion, it is important to establish the goals for the patient and the carer at the initial evaluation as there may not be agreement on the reason for the visit. If the patient is coming reluctantly, it is an opportunity to begin developing rapport. In the case of second opinion, review of the previous assessment may indicate that additional testing is needed. When discussing the diagnosis at the conclusion of the visit, it is important to ascertain whether goals for the encounter were addressed and the patient's and family member's reaction to the diagnosis.

Clinical Services

The care of the patient with FTD requires partnership with the patient, the family/carer and will usually include other health care providers and community agencies. Since there is no cure for FTD yet, care is focused on ameliorating symptoms, managing disability, promoting adaptation (the patient's and the family's), optimizing quality of life, and preparing for inevitable changes in the patient's life participation.

For interventions to be effective, the family and carers must learn about the disease they are confronting. The deterioration in social functioning, personality and behavior seen in FTD has deleterious effects on relationships (Bristow et al. 2008; Kaiser & Panegyres, 2007; Mourik et al., 2004). Assisting family members in understanding the patient's distressing and offensive behaviors as products of the disease facilitates empathy and adaptation. Opportunities for education occur throughout the illness and can be offered incrementally and through various sources and formats (e.g., reading materials, websites, support groups and referral to advocacy organizations).

Guiding the carer and providing support and encouragement is an integral focus of caring for the patient who has FTD. The carer should be seen as a partner who will support, promote and (eventually) manage the patient's participation in care. The carer is also a victim of the illness, in the sense of personal loss, being regularly confronted with asocial and offensive behaviors, raising dependent children, managing financial pressures, and maintaining employment. Devoting a portion of each visit to a private interview for the carer creates a safe space to discuss their needs. This time can be focused on supportive counseling, teaching of coping strategies, or problem solving specific targets. In some instances a carer experiencing a high level of psychological distress may benefit from formal psychotherapy, in which case a referral to a mental health professional should be made. Support groups comprised of FTD caregivers can be helpful to carers to discuss issues related to the illness and to feel less isolated and integrated into a community of carers. Time

spent with family members and the patient together can be diagnostically useful to assess social cognition and behavior as the clinician can observe how the patient interacts with family members.

The neuropsychiatry unit is reserved for cases where offensive behaviors cannot be safely managed in the home, because of their nature (for example, violent behavior) or their intensity – hospitalization has been arranged at Johns Hopkins for several cases featuring severe hyperphagia that became uncontrollable and caused violent confrontations or consumption of raw food and inedible materials. Admission to the ward provides opportunity to observe the behavior first hand for signs of environmental or socio-dynamic factors, and to investigate for causal or contributory physiologic derangements. The unit also facilitates rapid development and trial of behavioral interventions, as has been described (Lough & Hodges, 2002), and monitoring of pharmacologic trials (for insomnia, irritability/agitation or, rarely, psychosis). Admission has another benefit – affording a beleaguered family respite, enabling them to "recharge" for their reengagement in the patient's day-to-day care.

Financial Problems

Severe financial strain is not uncommon in the early stages of FTD (Hasse, 2005; Luscombe, Brodaty, & Freeth, 1998). Whereas the typical person with dementia is an elder, is no longer raising a family, and likely has retirement income, the typical FTD patient is younger, still working and, in many cases, still has dependent children at home (Bettie et al., 2002; Roach et al., 2008; Williams et al., 2001). Thus dementia in FTD can bring about an abrupt loss or reduction in family income, which in turn causes abrupt financial distress. Besides loss of income, in FTD financial trouble may arise from neglecting bills, impulsive spending (e.g., on the internet), compulsions (e.g., pornography), or poor judgment (e.g., scams, sweepstakes, gifts to strangers), or the costs associated with providing care for the affected (Hasse, 2005).

In the United States some patients encounter a serious dilemma before their diagnosis, wherein losing their job because of poor work performance causes the loss of health insurance that came with it. These dual losses can severely limit access medical care. Some patients are more fortunate, in that they receive the diagnosis before termination occurs – which affords them the protection and advantages of their employer's disability procedures and local regulations. In this situation, the clinic's role is to ensure timely documentation of disability and manage an early retirement.

All patients and families should be counseled about vigilance and practical safeguards such as controlling or changing access to accounts and credit and monitoring mail to remove solicitations if needed. Families should also be counseled to seek advice from an elder law attorney about securing assets and planning for their financial future. Patients and families should obtain long-term care and other types of insurance prior to genetic testing.

Driving

Persons with dementia inevitably lose the capacity to drive a vehicle. This does not mean that a diagnosis of dementia should lead immediately to orders to stop driving – although some patients will have this disability at diagnosis. In FTD visuospatial skills and memory are not impaired early (Hodges et al., 1999), so the patient may still be capable of operating a vehicle at the time of diagnosis. This is a nuanced question, requiring care and thought in reaching a clinical recommendation. Driving issues in FTD usually arise from poor judgment and conduct e.g., speeding, impulsive acts, and disregard for traffic rules. Thus cognitive tests may not be sufficient to identify patients who should no longer drive. It is

important, therefore, to inquire about driving capacity *and* driving behavior. Where there is concern about a patient's driving, it is best to obtain a driving assessment from the local jurisdiction or a formal driving assessment program. This assessment should include road testing, since many patients who can still drive safely may perform poorly on cognitive tests. When a patient passes a driving assessment and carers are not observing unsafe driving, they should probably be allowed to continue to drive. In this situation, it is best to agree the conditions for repeat assessment – typically time intervals, driving incidents, navigational disorientation or evidence of decline in praxis or visual perception.

Relying on a formal driving assessment to make decisions helps demonstrate fair process to the patient. The decision to recommend cessation of driving should never be made lightly. Terminating this privilege, if done prematurely, causes hardship by constraining the patients autonomy and life participation. It also causes avoidable conflict between patient and family, and it is always done at risk to the patient-physician relationship. Physicians and other providers should familiarize themselves with the local regulations pertaining to drivers with newly diagnosed cognitive or physical disability, since some jurisdictions have mandatory reporting requirements for specified diagnoses.

Continuity of Care between Visits

The interval between visits is determined by the nature of active problems, the level of stress the caregiver is expressing, the period between recent medication adjustments and the need to monitor effectiveness of medications. More frequent visits reduce the "stock piling of problems" that longer periods can foster. However, it is not unusual in FTD care to have frequent contact with carers between visits to manage worsened or newly developed problems. Sometimes phone contact suffices to solve a problem or make a prescribing decision. At other times an urgent office visit is scheduled for a clinical examination. Between-visit continuity is facilitated by a collaborative relationship with the primary care provider or a local neurologist or psychiatrist. In our experience, has become valuable in emergencies, particularly for those who live a long distance from the clinic – where we have been unfamiliar with their local medical system and travel to us would be time consuming and burdensome. In other cases an admission to the neuropsychiatry inpatient unit will be indicated.

Residential care

The decision to consider residential care has multiple antecedents reflecting characteristics of the patient, the carer, and the disease (Gaugler, et al., 2005; Yaffe, 2002). Reasons for placement include care needs exceeding the capacity of the carer to provide adequate and safe supervision as well as the severity of the dementia. In some situations the carer is ready or must transition from the role of primary caregiver because of milestone events in their life such as significant health care problems, the need to focus more attention on child rearing or because of workplace demands. It often takes a significant amount of time to prepare emotionally and practically for residential care, and so early conversations about this topic with the patient and carer are useful. Carers may express guilt about these decisions and it is usually helpful to discuss these. When the decision is made, the family requires active assistance in identifying and choosing the right residential care facility, since facilities vary widely in their design, services and medical sophistication (Leroi et al., 2007). Upon the patient's admission to the facility, the next step is to educate the family about the transition period (usually 4–6 weeks) and process. This will set expectations and instill the patience families as they adapt to separation, accept change, and develop relationships with the facility's medical staff. It is also very important to form relationships with the facility's medical staff, as it is possible to continue an active treatment role or provide 'as needed' consultation to the facility.

End of life care

Today it is inevitable, unfortunately, that our patients reach the end stages of FTD and can no longer be brought to the clinic. When that point is reached and crossed, the relationship between clinic staff and the carer often continues. This happens because the trust and expertise developed over years can be put to good use, helping a family navigate vexing challenges– cessation of eating, terminal symptoms, and comfort measures. Many families do not know what to expect in the final stages of dementia, and need the education, as well as support and guidance in making difficult decisions. It is at this time that referrals to palliative care and hospice are presented and then arranged. Upon the death of the patient, contact with the family serves to address results of post-mortem brain examination, if that has been done. We have learned from experience that this discussion of neuropathology reports inevitably leads to questions about the genetic implications of the illness for the family. Questions about what the final diagnosis means for the next generation usually arise. We have also faced situations where the neuropathology diagnosis differs from the clinical diagnosis, and have found that for some carers and families this is vexing. It is also the case that some carers and families request a final meeting with the clinic to ask and settle lingering worries about the care, such as "did I do enough?" and "did I make the right decisions?"

Community engagement and Advocacy

Educating the public about FTD has become a high priority for professionals involved with these patients, and for the patients, carers and families themselves. The increasing awareness and knowledge that these carers and families have about these conditions has, in turn, had positive effects on care and policy. In the United States, advocacy organizations such as the Association for Frontotemporal Degeneration (AFTD) are leaders in the effort to increase public awareness and education, advocate with public officials, promote research, and provide information and support to patients and their carers (Dickinson & Denny, 2011). AFTD offers a wide variety of resources. The Alzheimer’s Association also has resources for persons with FTD and their carers.

DISCUSSION

In this article, we have discussed the diagnosis, study, and management of FTD in the mental health and multidisciplinary care settings. These are important topics not only because of the importance of identifying patients with FTD in the mental health setting who are often misdiagnosed, but because of the contributions psychiatry and mental health can provide for patients with FTD and their families in the multidisciplinary care setting. We would like to increase education, involvement, and interest in FTD among psychiatrists and other mental health practitioners. The symptom overlap between FTD and psychiatric disorders can elucidate the neuroanatomic etiology of emotional, cognitive, and behavioral symptoms in FTD and psychiatric disorders. FTD patients can benefit from a multidisciplinary team that can address pharmacologic and behavioral treatments, family engagement, social work and placement needs, and neurological and palliative aspects of FTD. We discussed the management of several common issues in FTD including driving, finances, caregiver burden, residential care, and end-of-life issues.

References

Beattie AM, Daker-White G, Gilliard J, Means R. Younger people in dementia care: A review of service needs, service provision and models of good practice. Aging and Mental Health. 2002; 6:205–212. [PubMed: 12217088]

Beck, AT.; Steer, RA.; Brown, GK. Beck Depression Inventory - II. New York: Psychological Corporation; 1996.

Bristow M, Cook R, Erzinclioglu S, Hodges J. Stress, distress and mucosal immunity in carers of a partner with fronto-temporal dementia. Aging and Mental Health. 2008; 12(5):595–604. [PubMed: 18855175]

Chow TW, Pio FJ, Rockwood K. An international needs assessment of caregiversfor frontotemporal dementia. Canadian Journal of Neurological Sciences. 2011; 38(5):753–757. [PubMed: 21856580]

Dickinson S, Denny S. AFTD: Making Connections and Building Resources. Journal of Molecular Neuroscience. 2011; 45(3):704–705. [PubMed: 22428168]

Eaton NR, Krueger RF, Keyes KM, Skodol AE, Markon KE, Grant BF, Hasin DS. Borderline personality disorder co-morbidity: relationship to the internalizing-externalizing structure of common mental disorders. Psychological Medicine. 2011; 41:1041–50. [PubMed: 20836905]

Friedman JI, Harvey PD, Coleman T, Moriarty PJ, Bowie C, Parrella M. Six-year follow-up study of cognitive and functional status across the lifespan in schizophrenia: a comparison with Alzheimer's disease and normal aging. American Journal of Psychiatry. 2001; 58(9):1441–1448. [PubMed: 11532729]

Gaugler JE, Kane RI, Kane RA, Newcomer R. The longitudinal effects of earlybehavior problems in the dementia caregiver career. Psychology and Aging. 2005; 20:100–116. [PubMed: 15769217]

Harvey PD. Cognitive impairment in schizophrenia: profile, course, and neurobiological determinants. Handbook of Clinical Neurology. 2012; 106:433–445. [PubMed: 22608636]

Hasse, T. Early-onset dementia: the needs of younger people with dementia in Ireland. 2005. Retreived August 26, 2012, from: http://www.alzheimer.ie/eng/Resources/Research/Early-Onset-Dementia-The-Needs-of-Younger-People-with-Dementia-in-Ireland.

Hodges JR, Patterson K, Ward R, Garrard P, Bak T, Perry R. The differentiation of semantic dementia and frontal lobe dementia (temporal and frontal variants of frontotemporal dementia) from early Alzheimer's disease: A comparative neuropsychological study. Neuropsychology. 1999; 13:31–40. [PubMed: 10067773]

Huey ED, Lieberman JA. From known to unknown; old to new: Can lesion studies inform psychiatry about mental illness in the 21st century? Neuropsychiatry. 2012; 2(5):369–372.

Huey ED, Zahn R, Krueger F, Moll J, Kapogiannis D, Wassermann EM, Grafman J. A psychological and neuroanatomical model of obsessive-compulsive disorder. The Journal of Neuropsychiatry and Clinical Neurosciences. 2008; 20:390–408. [PubMed: 19196924]

Kaiser S, Panegyres PK. The psychosocial impact of young onset dementia on spouses. American Journal of Alzheimer’s disease and Other Dementias. 2007; 21:398–402.

Knopman DS, Roberts RO. Estimating the number of persons with frontotemporallobar degeneration in the US population. Journal of Molecular Neuroscience. 2011; 45:330–335. [PubMed: 21584654]

Koenigs M, Huey ED, Calamia M, Raymont V, Tranel D, Grafman J. Distinct regions of prefrontal cortex mediate resistance and vulnerability to depression. The Journal ofNeuroscience. 2008; 28:12341–12348.

Krueger RF. The structure of common mental disorders. Archives of General Psychiatry. 1999; 56:921–926. [PubMed: 10530634]

Leroi I, Samus QM, Rosenblatt A, Onyike C, Brandt J, Baker AS, Lyketsos C. A comparison of small and large assisted living facilities for the diagnosis and care of dementia: the Maryland Assisted Living Study. International Journal of Geriatric Psychiatry. 2007; 22(3):224–232. [PubMed: 17044133]

Lough S, Hodges JR. Measuring and modifying abnormal social cognition in frontal variant frontotemporal dementia. Journal of Psychosomatic Research. 2002; 53(2):639–646. [PubMed: 12169338]

Luscombe G, Brodaty H, Freeth S. Younger people with dementia: Diagnosticissues, effects on carers and use of services. International Journal of Geriatric Psychiatry. 1998; 13:323–330. [PubMed: 9658265]

Mendez MF, Shapira JS, McMurtray A, Licht E, Miller BL. Accuracy of the clinical evaluation for frontotemporal dementia. Archives of General Psychiatry. 2007; 64:830–835. [PubMed: 17606817]

Miller RG, Jackson CE, Kasarskis EJ, England JD, Forshew D, Johnston W, Woolley SC. Practice parameter update: the care of the patient with amyotrophic lateral sclerosis: multidisciplinary care, symptom management, and cognitive/behavioral impairment (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology. 2009; 73:1227–1233. [PubMed: 19822873]

Mourik JC, Rosso SM, Niermeijer MF, Duivenvoorden HJ, Van Swieten JC, Tibeen A. Frontotemporal dementia: behavioral symptoms and caregiver distress. Dementia and Geriatric Cognitive Disorders. 2004; 18:299–306. [PubMed: 15305107]

Panegyres PK, Graves A, Frencham KA. The clinical differentiation of fronto-temporal dementia from psychiatric disease. Journal of Neuropsychiatric Disease and Treatment. 2007; 3:637–465.

Rankin KP, Santos-Modesitt W, Kramer JH, Pavlic D, Beckman V, Miller BL. Spontaneous social behaviors discriminate behavioral dementias from psychiatric disorders andother dementias. Journal of Clinical Psychiatry. 2008; 69:60–73. [PubMed: 18312039]

Roach P, Keady J, Bee P, Hope K. Subjective experiences of younger peoplewith dementia and their families: implications for the UK research, policy and practice. Reviews in Clinical Gerontology. 2008; 18:165–174.

Singleton N, Bumpstead R, O'Brien M, Lee A, Meltzer H. Psychiatric morbidityamong adults living in private households. International Review of Psychiatry. 2003; 15:65–73. [PubMed: 12745312]

Snowden JS, Rollinson S, Thompson JC, Harris JM, Stopford CL, Richardson AM, Pickering-Brown SM. Distinct clinical and pathological characteristics of frontotemporal dementiaassociated with C9ORF72 mutations. Brain. 2012; 135:693–708. [PubMed: 22300873]

Steer RA, Ball RW, F Ranieri WF. Dimensions of the Beck Depression Inventory-II in clinically depressed outpatients. Journal of Clinical Psychology. 1999; 55:117–128. [PubMed: 10100838]

Williams T, Dearden AM, Cameron IH. From pillar to post – a study of younger people with dementia. Psychiatric Bulletin. 2001; 25:384–387.

Woolley JD, Khan BK, Murthy NK, Miller BL, Rankin KP. The diagnosticchallenge of psychiatric symptoms in neurodegenerative disease: rates of and risk factors forprior psychiatric diagnosis in patients with early neurodegenerative disease. Journal of ClinicalPsychiatry. 2011; 72:126–133.

Wright SL, Persad C. Distinguishing between depression and dementia in olderpersons: neuropsychological and neuropathological correlates. Journal of Geriatric Psychiatryand Neurology. 2007; 20:189–198.

Yaffe K, Fox P, Newcomer R, Sands L, Lindquist K, Dane K, ovinsky KE. Patient and caregiver characteristics and nursing home placement in patients with dementia. The Journal of the American Medical Association. 2002; 287:2090–2097.

Table 1

Syndrome	Commonly shared symptoms with FTLD	Rarely shared symptoms with FTLD
Major depressive disorder	Anhedonia, psychomotor agitation or retardation, decreased motivation and energy, decreased concentration and focus	Depressed mood, weight loss, insomnia, feelings of worthlessness, poor self-esteem, suicidal ideation
Mania	Logorrhea, flight of ideas, distractibility, increase in goal-directed activities, impulsive behavior	Elevated mood, decreased need for sleep
Obsessive-compulsive disorder	Compulsions (repetitive behaviors the patient feels compelled to perform)	Obsessions (unwanted, recurrent intrusive thoughts that cause anxiety)
Schizophrenia	Disorganized speech and behavior, affective flattening, alogia, avolition	Complex delusions, auditory hallucinations (more common in C9ORF72 mutation carriers)