Chapter 1

Neuropsychiatric Symptoms Are a Core Feature of Neurocognitive Disorders

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Dementia as a Cognitive, Behavioral, and Functional Disorder

Dementia is described as a syndrome affecting cognition (memory and thinking), behaviors, and the ability to perform activities by the World Health Organization (WHO) [19]. WHO published a 2012 report outlining challenges due to dementia which impact the physical, psychiatric, and economic well-being of those with dementia, their families, and society. The report was part of a worldwide campaign to bring awareness and attention to dementia as well as hopes of reducing stigma and barriers to diagnosis and care for those living with dementia. The American Psychiatric Association (APA) last updated its *Diagnostic and Statistical Manual* to a fifth version (DSM-5) in 2013. APA now refers to the progressive neurodegenerative diseases that most people refer to as dementia with the phrase major neurocognitive disorders (MNCD). The central features of MNCD, as described in the

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DSM-5, are cognitive deficits and functional impairment in a person's ability to perform instrumental activities of daily living and activities of daily living. One challenge with the way DSM-5 conceptualizes neurocognitive disorders is the emphasis on cognition with some emphasis on functional impairment rather than the WHO triad of cognition, behavior, and function. Of the 53 pages in the DSM-5 devoted to neurocognitive disorders, less than one page addresses the definition and occurrence of behavioral disturbance. The WHO description of dementia is more consistent with the common view of dementia as well as how most living with dementia come to clinical attention.

Currently, there are no disease-modifying treatments for the leading types of dementia including Alzheimer's disease, Lewy body dementia, and frontotemporal dementia. There are no current treatments that reverse cognitive decline in any neurodegenerative disease. There are some pharmacological and behavioral interventions that appear to possibly slow the rate of functional decline in certain types of dementia. This evidence is not robust, meaning that there are a few suggestive studies but not a significant base of evidence [9, 13, 14].

Neuropsychiatric symptoms (NPS) are universal in dementia. The Cache County study followed a group of people with dementia longitudinally for 5 years and found that 97% of those with dementia experienced at least one NPS [16]. NPS, also referred to as behavioral and psychological symptoms of dementia (BPSD), are notable in two ways: they are the symptoms that cause caregivers and people living with dementia the most quality of life impairment, and they are the only symptoms of dementia currently modifiable on a reliable basis [5, 11]. Note that even the first person diagnosed with what is now known as Alzheimer's Disease, Frau A, came to the attention of Dr. Alzheimer in the early 1900s due to her severe NPS.

Frau A., the first person known to have Alzheimer's disease, came to medical attention due to persecutory delusions, hallucinations, screaming, and other NPS. [8]

Dementia Transitions to MNCD

Major neurocognitive disorder (MNCD) as a category in the DSM-5 describes degenerative dementia syndromes and other cognitive disorders. MNCD involves a complex combination of neuropsychiatric signs and symptoms. One reason the APA transitioned from dementia to major neurocognitive disorder involved differentiating between the degenerative diseases, particularly those that include most dementias, from other conditions that may be more static. Earlier detection of disease is another. Earlier and more accurate diagnoses will improve the chance of discovering disease-modifying treatments as well as identifying those who will benefit from treatment. Dementia is not a single entity. Dementia is a general phrase and represents several different syndromes. In both the medical literature, newspaper articles, and everyday speech, people frequently use the word dementia as if they were referring to one specific disease. This has likely led to some of the challenges in studying people who have a dementing illness. If the people being studied have different illnesses, yet are all placed in the same category, it can be difficult to differentiate what is helpful for some illnesses and not for others. Additionally, there is hope that changing the term from dementia to neurocognitive disorder will help address the stigma associated with the word dementia. In this chapter, the term dementia will be used when speaking about those specific major neurocognitive disorders associated with degenerative brain diseases; see Table 1.1 for those MNCD commonly thought of as diseases causing dementia. When a dementia is being referenced, the actual name of that disease will be used.

Neuropsychiatric Symptoms

As dementia advances, NPSs appear to become intrinsic to the neurodegenerative disease. It is quoted that anywhere from 80% to 90% of those with a degenerative dementia

Table 1.1 MNCD in DSM-5 that are considered degenerative dementias

Major neurocognitive disorders considered degenerative dementias
Alzheimer's disease
Vascular neurocognitive disorder
Neurocognitive disorder with Lewy bodies
Neurocognitive disorder due to Parkinson's disease
Frontotemporal neurocognitive disorder
Neurocognitive disorder due to Huntington's disease
Neurocognitive disorder due to prion disease

have behavioral and psychological symptoms of dementia (BPSD) also known as NPS over the course of their disease though one of the longest longitudinal studies referenced above found the actual percentage was nearly 100%. Behavior, by definition, is a normal and necessary part of life. This is one reason the authors of this text choose to describe BPSD as NPS. The NPS of dementia drive many if not most of the quality of life impairments, need for placement outside the home as well as caregiver distress [3]. Nearly all studies have shown the very high prevalence of NPS in those with dementia [20]. Some neuropsychiatric symptoms are best approached with behavioral and environmental interventions, while others are most safely approached using medications. There is a noteworthy evidence base supporting the use of specific medications in the hands of well-trained geriatric psychiatrists, behavioral neurologists, or others who specialize in that area [6, 10, 11, 12, 15]. Behavioral interventions, functional assessments, and strength-based care planning should be used with everyone. Although this text will not review in-depth specific medications used in treating neuropsychiatric symptoms, the reader may find the list outlined in Table 1.2 useful in knowing where the addition of medications may be helpful in decreasing suffering and where there may be little efficacy in using medications.

Table 1.2 Neuropsychiatric symptoms and responsiveness to medications

Neuropsychiatric symptoms (NPS)	
NPS not usually responsive to treatment with medications	**NPS usually responsive to treatment with medications**
Repetitive questions	Paranoia and delusions
Anxiety about memory loss or other deficits	Hallucinations
Psychomotor agitation *unless related to pain*	Pain
Sleep problems	Aggression
Calling out *unless related to pain*	Anxiety disorders – not the symptom of anxiety as a reaction to having cognitive impairment
Ambulating or wandering – otherwise generally known as walking	Major depressive episode – not grief, sadness, or apathy
Appetite problems	

Specific NPS include psychotic symptoms (paranoia, delusions especially persecutory delusions, hallucinations especially visual hallucinations, mood symptoms such as depression and elation, anxiety, agitation, anger outbursts, combativeness, resistiveness to cares such as bathing and feeding, disruptive motor or vocal activity, sleep disturbance (including insomnia, hypersomnia, rapid eye movement sleep behavior disorder, and circadian rhythm disturbance), apathy, wandering, disinhibition, hyperphagia, hoarding [1]). Other NPS include excessive motor activity such as pacing in the hall or repetitively picking something up and carrying it around, irritability, mood lability, and appetite disruption.

Dementia as a Leading Health Concern

According to the United Nations and its world population prospects report from 2017, there are an estimated 962 million people aged 60 and above [17]. Alzheimer's disease is by far the most common type of dementia. Age is the largest risk factor for developing Alzheimer's disease. Thus, we are facing a rapid increase in the number of people living with Alzheimer's disease. Newspaper headlines, medical journals, and magazines contain language such as the following: a "tsunami" or "tidal wave" of people with dementia is heading toward our shores. Even the most optimistic voices that predict a decline in the rate of new dementia cases continue to predict that the number of people developing dementia in any given year will remain roughly the same [18]. The Cognitive Dysfunction and Aging Study is a multicenter population-based study that sampled two English/Welsh groups in two different time periods. They report a 20% drop in incidence in dementia though there was no change in the actual number of new cases because:

> "Dementia continues to be a topic of major international interest with successive reports suggesting large increases throughout the world in the next decades. Following governmental concern and the 2013 G8 summit, the World Dementia Council has been established to facilitate greater attention both in research to support reduction of risk, better diagnosis and treatments, and support for those at risk of, and with, the dementia syndrome. [7, p. 2]"

Beyond the Medical Model to Person-Centered Care

We have come to define dementia clinically by a group of deficits and the loss of functioning. This corresponds to the medical model of diagnosis and allows us to name illnesses in a way that allows clear communication and research. Because we have a clear definition when people talk about Alzheimer's disease, we are likely to be talking about the same illness. This also applies to Lewy body dementia and many other types of dementia. Knowledge of the specific type of dementia a person has is essential to assist persons with dementia, their families, and caregivers, so they will know what to expect in the present and the future. This is most true in the early and middle stages of these diseases. Once someone has very advanced dementia, it becomes difficult or impossible to accurately differentiate the type of dementia.

Despite the utility of the medical model for diagnosis, the authors of this text believe that the medical model is not the best paradigm to use for care planning with those experiencing a MNCD. The authors here propose a new model based upon recognition and treatment of neuropsychiatric symptoms using functional assessments both to define assets as well as deficits. The functional strengths that a person has are what allow us to design the most respectful and comprehensive assessment and plan. Periodic reassessment of a person's ability to function enables people living with dementia to do as much for themselves as they can without unrealistically expecting them to do things they are unable to do. The team's goal is to use a strength-based functional framework for understanding behavior and assisting people by focusing on the highest quality of life possible. The team always includes the person with dementia, their family, as well as professional staff and personal caregivers.

By far the most common type of dementia is Alzheimer's disease. It is estimated that between 60% and 90% of people with major neurocognitive disorder have at least a component of Alzheimer's disease. Dementia due to Lewy body disease, Parkinson's disease dementia, and frontotemporal dementias are the next most common types of dementia [1]. One key to understanding neurocognitive disorders is that these are acquired over life rather than being the lack of development. Because these illnesses are defined by loss of functioning, we must attain that functioning first. Cognitive deficits are present in all serious mental illness. For example, schizophrenia, bipolar disorder, and major depressive disorder all involve impairments in executive functioning. The presence of significant cognitive impairment in many mental illnesses is often overlooked but is likely the main reason serious mental illness is so disabling. The presence of preexisting neurocognitive deficits due to serious mental illness complicates the ability to diagnose a neurocognitive disorder, even though people living with serious mental illness face an increase in the risk of developing dementia. Multiple studies of those with schizophrenia, bipolar disorder, and major depressive disorder show significant increased risk of developing dementia compared to the general population [2, 4]. Many primary care providers have difficulty differentiating NPS caused by dementia from the NPS caused by other psychiatric illnesses. Given that serious mental illness and dementia involve both a person's daily functioning and their neurological systems, it is not surprising that many of the same symptoms are found in those living with all these illnesses [1].

Dementia as a Life-Limiting Illness

A major component of person-centered care for those who have a life-limiting disease is maximizing functioning and maintaining or improving quality of life when possible. Palliative care is often confused with hospice. Palliation and the provision of palliative care emphasizing aggressive management of symptoms that cause or increase suffering are indicated whenever a life-limiting disease is diagnosed. When it is no longer possible to maintain or improve the quality of life due to the life-limiting nature of these progressive illnesses, it is the standard of care to consider a shift of the goal of care to comfort and relief of suffering. Medical professionals, as well as the public at large, have been reluctant to acknowledge the relentless and progressive nature of dementia. Some of these illnesses progress over as many as 20 years and therefore it is less obvious that they can be a cause of death. This attitude and the accompanying reluctance to diagnose people with dementia have been slowly changing. It is important to recognize that these illnesses are progressive until life ends. People who are dying, and their families, deserve the chance to process their grief and emotions related to dying. This is one way to decrease suffering. Too often family members only realize that someone living with dementia is near death when they are in their last weeks. As with all serious illness, it is advisable to complete advanced care planning and regularly reassess the goals of treatment and care. This allows for the provision of palliative care to minimize suffering whenever possible. This is reviewed more thoroughly in a later chapter.

Behavior Is Communication

All behavior is communication. This applies both to those who have dementia as well as all humans. Most communication, in fact, is nonverbal. Body language and tone of voice combined are thought to outweigh the content of words. Despite this fact, the human brain appears to be quite focused on verbal information. In clinical practice the tendency is to believe what people say with words even when their ability to use language is impaired and/or their actions repeatedly demonstrate something different. When working with those with dementia, this may be part of the laudatory desire to respect people as adults. Those with major neurocognitive disorder as well as other brain illnesses are often unable to use language with the level of specificity and accuracy that was used prior to the development of dementia. Further, due to memory and/or executive functioning deficits, people may mean what they say but be unable to carry through with actions. One of the advantages of language is that words enable rapid and succinct communication. However, because almost all forms of major neurocognitive disorder impair the ability of the brain to use and understand language, it is essential to include the metamessage that a person's behavior is communicating in addition to their words when planning and using communication. Note that the timing of the impairment of the verbal centers in the brain is different for different neurodegenerative illnesses. This is one of the many reasons why it is essential not to consider dementia or major neurocognitive disorder as a single entity. Because of these challenges with limiting communication merely to language, the authors of this text advocate analyzing what people are trying to communicate by combining information from verbal language, tone of voice, body language, and actions to best understand what people with MNCD want to communicate and plan for addressing this communication.

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