

Module 1 – Introduction to Memory Loss

Purpose: To introduce patients and families to the basic concepts of dementia and Alzheimer's disease, and to help them to understand the diagnostic process

Introduction: What is the difference between dementia and Alzheimer's disease?

Illnesses causing memory loss and neurologic degeneration are common, affecting more than four million Americans. Although Alzheimer's disease is the most common disease causing memory loss, many neurologic conditions have similar symptoms and are managed in approximately the same way.

When a person begins to develop personality changes, become forgetful – especially for recent events, and have difficulty performing tasks they were once able to do these are often symptoms of a condition called “dementia.” Dementia means there is something, usually a disease, affecting the “thinking and doing” parts of the brain. There are numerous illnesses and injuries that cause dementia.

- The condition of dementia is a lot like the condition of an infection. When a person has an infection, no matter where it is in the body, they will generally have pain, tenderness, heat, redness, swelling, and perhaps some drainage. There are numerous “germs” that cause infections. So the word “infection” is simply the body's pathological response to the germ. Successful treatment of the infection involves identifying the germ and treating it with the correct antibiotic for the causative germ.
- Dementia is the body's response to an illness in the brain. Typically there are four groups of symptoms we see with dementia:
 - Personality changes – There may be a blunting of the personality, apathy, decreased inhibitions, and increased self-absorption (selfishness with lack of ability to consider the needs of others). These changes are not something the person can control.
 - Changes in “cognition” - This means the person has difficulty remembering recent events, may distort past memories, difficulty with

learning new things, make judgments, slowly lose written and verbal language skills, and with understanding the concept of time.

- A slow progressive decline in day to day functioning – This is mostly due to a decreased ability to plan, initiate, and carry the activity through to a goal. The person knows what they want to do, and may be able to complete the task intermittently, HOWEVER when thinking about the task the person is unable to put the steps into the sequence to get the job done. And, the more the person thinks about the activity, the more difficult it becomes for them. So asking the person to “think about it,” or “try harder” only makes the problem worse and causes anger or frustration. Caregivers often think the person is being “lazy” or “willful,” however this is really just another symptom of the dementia. “Module 2: The Seven Stages of Dementia” outlines the usual loss of abilities with disease progression.
- A decreased ability to tolerate stress – The person becomes fatigued more easily and may anger easily. Situations and groups may irritate or overwhelm the person. This becomes more evident and problematic as the disease progresses. “Module 7 : Day to Day with Dementia” discusses what triggers stress-related behaviors and how to prevent them.
- There are numerous diseases that can cause dementia. By far the most common is Alzheimer’s disease, so while every person with Alzheimer’s disease develops dementia, all people with dementia do not have Alzheimer’s disease. Some of the more common causes include the following:
 - Alzheimer’s disease
 - Dementia from vascular disease, often due to multiple strokes
 - Frontal lobe syndromes including frontotemporal disease, multiple sclerosis, and ALS
 - Lewy Body disease
 - Parkinson’s disease and Parkinson’s Plus diseases
 - Normal pressure hydrocephalus
 - Alcoholism and substance abuse

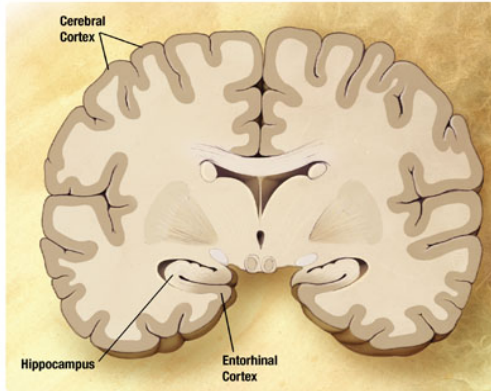
- AIDS
- Creutzfeldt-Jacob disease
- Severe liver disease
- Meningitis and encephalitis
- Brain injury
- “Prizefighters dementia” - Dementia pugilistica
- The symptoms of dementia vary depending on the places in the brain where the disease occurs, the person’s premorbid personality, the person’s culture, the environment where they live, and what is happening around them

The average family spends about \$200,000 on care for a loved one after they have been diagnosed, yet the costs of the illness far exceed the money spent. Families selflessly devote their love, time, energy, and creativity to care for their loved ones while watching the slow agonizing progression of the disease

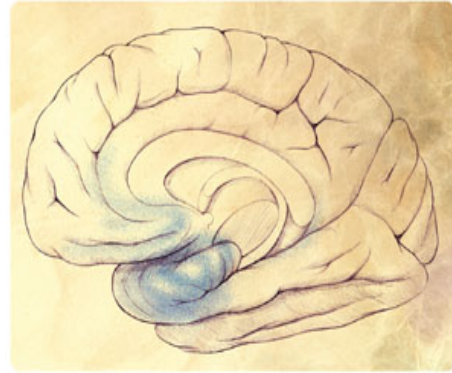
Brain Deterioration due to Alzheimer's disease

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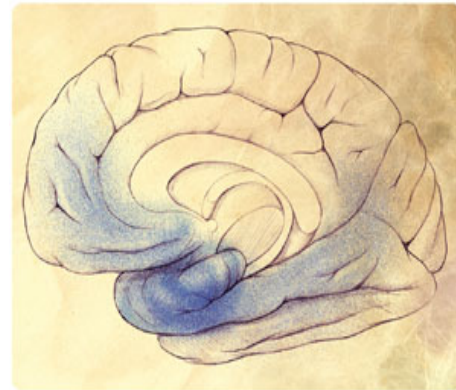
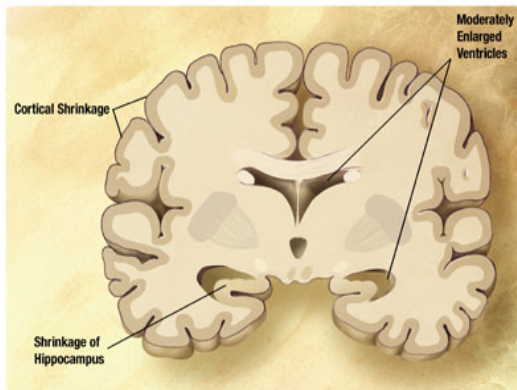
Cross-section of brain



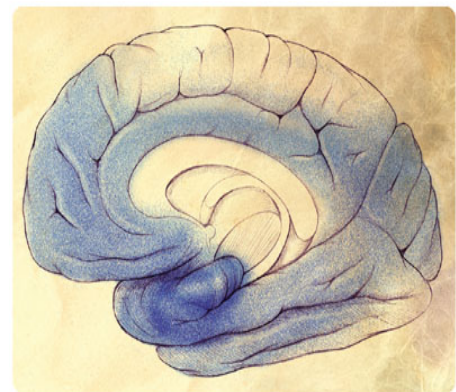
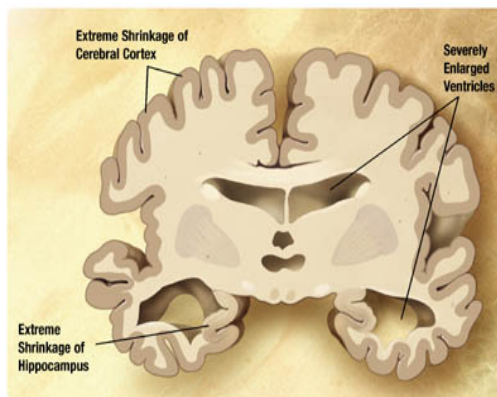
Side view of brain:
Shading indicates affected areas.



Very Mild (No or very mild symptoms years 1-3)



Mild to Moderate changes (Symptomatic years 3-6)



Advanced changes (Symptomatic years 7-death)

When behaviors equal symptoms

Dementing illnesses -- including Alzheimer's disease, Parkinson's disease, and strokes -- have a variety of symptoms that can baffle and overwhelm family members. Some of the most challenging and frightening problems rarely occur early in the disease but may "pop up" when it is least expected as the disease progresses. Examples might include the following:

- Demanding to leave during an activity or event they had been looking forward to
- Waking up in the middle of the night to get dressed and start the day
- Not recognizing familiar settings, home, or family late in the afternoon
- Accusing family members of stealing items the patient has hidden -- or blaming "outsiders" for taking things
- Threatening family members, especially children with physical violence
- Becoming irritated or belligerent late in the day
- Refusing to bathe, go to the doctor, or out socially for no apparent reason
- Walking away from home or getting lost
- Telling stories you know aren't true
- Thinking there are extra people or children in your home
- Seeming selfish

These behaviors may seem mean-spirited and purposeful, but they are simply symptoms of the person's disease. These behaviors are normal and expected in mid-disease. It is important to realize that the person can not control these sudden behavior changes, but you can help to prevent or minimize them. While not everything works all the time, we can eliminate much of the "acting out," behavior by making simple changes in our behavior and the environment. The changes help the patient to succeed. Unexpected changes in behavior are often triggered by things you can control -- once you know how.

Diagnosing a dementing illness: Why bother?

If four million people have a dementing illness and about half of those illnesses are caused by Alzheimer's disease, as many as two million people have non-Alzheimer

dementias. There are dozens of non-Alzheimer type (atypical) dementias, yet diagnosis is often overlooked after an initial evaluation for whether the disease is reversible. People with different types of dementia have different care needs. If we provide care for everyone with dementia the same way we care for people with Alzheimer's disease, it results in poor fit.

Families of people with atypical dementias are often mainstreamed into Alzheimer's support groups. They report frustration with having their questions answered or not receiving understanding support from professionals or other support group members. It is therefore important that health providers understand and assess for those behaviors that may indicate atypical presentations of dementia. The following information describes what is done to diagnose Alzheimer's disease and other dementias. If the person with dementia doesn't "fit" the picture of Alzheimer's disease described below it is reasonable to suggest the family seek diagnosis from a behavioral neurologist.

When to seek a diagnosis

Often aged people with memory loss come to service providers having had no formal diagnosis. Families may have been unsure about whether to seek a diagnosis, feel that memory loss is normal for older adults, that the memory loss is not severe enough to warrant an evaluation, or are generally fearful of what may be found. The following list offers guidelines for families when encouraging to seek a diagnosis for dementing illness.

- Memory impairment severe enough to impair function
- Episodes of confusion
- Problems with finding or substituting words
- Change in abilities in day to day functions
- Personality change
- Changes in motor skills

Diagnosis of dementing processes

The three critical questions that are posed during the diagnostic process:

1. Is memory loss present?

The answer to the first question comes from a combination of information from the family, primary care provider (family doctor), or health professional. The information should include the following:

A. History from the patient and their family

- Changes in memory and function from previous abilities
- Changes in behavior patterns
- History of mental health problems/substance use/OTC and prescribed medications
- Other symptoms such as becoming unstable on their feet, spells, or tremor
- Medical history and medications
- Social history such as degree and/or changes in social interaction, and supportive family
- Prior employment
- How the person functions on a usual day
- Onset and duration of the symptoms
- Waxing and waning of symptoms (good days and bad days)

B. A mental status screen

- Mini-Mental Status Examination
- Clock Drawing
- Figure copying

C. Observation of behavior and interaction with others

- Language Function
- Ability to cope with noise or groups
- Motor Functions
- Evidence of paranoia or psychosis
- Ability to navigate the environment and see
- The person's insight into their memory

D. Barriers to diagnosing memory loss

- Some patients can pass mental status tests despite their memory limitations and declining function
- Patients who are either very bright at baseline or who have strong personality characteristic may be able to "bluff" their way through the evaluation
- The physician or diagnostician has a strong relationship with patient and has some difficulty seeing decline
- The providers are concerned about the family motives in pursuing a diagnosis, especially in light of a history of family conflict
- Some conditions take years to diagnose. ALS with frontal lobe presentation, diffuse Lewy body disease, and other frontal lobe dementias frequently are misdiagnosed until several years after the family reports changes.

Once the provider is certain that cognitive loss is present, issues of cause must be addressed. The usual medical evaluation answers the second question using the following test results.

2. What could be causing it?

- A. The goal is to rule out all other causes of progressive dementia
- History & thorough physical examination
 - Imaging - should be negative
 - Done to rule out tumor, strokes, trauma, hydrocephalus
 - May see areas of localized atrophy (shrinkage)
 - Can be CT scan, MRI, PET
 - Neuropsychological testing – a complex battery (minimum 2 hours) that tests each type of mental functioning
 - Reconfirm memory loss
 - Defines areas of weakness and therefore areas of brain affected
 - Begins the care plan
 - Laboratory evaluation
 - CBC - anemia and blood dyscrasias
 - TSH - hypothyroid
 - Electrolytes - imbalances, renal disease, dyhydration
 - Blood sugar - diabetes
 - B12 & Folate - B12 deficiency
 - Urinalysis - infection, signs of renal disease
 - HIV, STD testing
- B. Other tests as indicated (rarely done unless there is a high degree of suspicion for the diseases listed below)
- Lumbar puncture – To rule out tumor, infection, NPH
 - EEG – Looking for seizures or infectious dementias
 - Chest X-ray – Looking for tumor
 - Cardiogram, Holter monitor – Looking for irregular heart beat (arrhythmia)
 - EMG – ALS The EMG is a test for nerve conduction in arms and legs
 - Overnight oxymetry and sleep studies- Looking for sleep apnea, restless legs, or REM sleep disorders.
 - Arterial biopsy – Looking for an immune disease called arteritis

All of the diagnostic data are compiled and interpreted to answer the last question....

3. Are the symptoms and course of the disease consistent with what we know as Alzheimer's disease?

A. If the symptoms are consistent with Alzheimer's Disease, the following may be noted:

- The history and neuropsychological tests will reflect losses in cognition and
- Imaging studies and laboratory tests will be negative.
- The symptoms and behavioral presentation will be consistent with AD
 - Slow onset, insidious progression

- Global losses in cognition, planning, language, memory, and visual-spatial perception
- Changes in short-term auditory (what the person has heard) and/or visual memory
- Subtle intensifying of negative personality characteristics
- Decreased ability to inhibit
- Increased self-absorption
- Decreased tolerance for noise, crowds, change
- Increased symptoms with fatigue
- Uneven symptoms presentation
- Episodes of depression
- Gradual loss of functional abilities because of problems with planning and sequencing

B. You might suspect other diseases if the following symptoms are present (especially early in the disease process). This is a partial list of some of the more common atypical dementia syndromes.

Presenting Symptoms	Possible Syndrome
Early onset of language problems, ataxia, incontinence	Normal pressure hydrocephalus
Early loss of language	Progressive aphasia, Frontal lobe degeneration, Corticobasal ganglionic degeneration (CBGD)
Severe disinhibition as evidenced by making poor decisions couples with hyperactivity, or socially inappropriate behavior	Frontal dementia syndromes including Pick's disease, frontal lobe degeneration, ALS
Abulia (failure to initiate activities) Apathy	Frontal dementia syndromes including Pick's disease, frontal lobe degeneration, ALS
Changes in motor function, tremor, spasticity, weakness, incoordination.	ALS, CBGD, Diffuse Lewy Body disease, Progressive supranuclear palsy
Psychosis	Bitemporal dementia, Diffuse Lewy body disease
Loss of vision	Visual variants of Alzheimer's disease, Progressive asimultanagnosia, Agnosia, CBGD
"Spells"	Seizure disorder, stroke disorders, amyloid angiopathy
Muscle wasting	ALS

C. Reasons for pursuing a diagnosis of a non-Alzheimer-type dementia

Once the diagnosis of a dementing illness is established, responsibility for planning and providing daily care is assumed by the family and non-physician health professionals. Enormous strides have been made in both the understanding of pathophysiology of chronic dementing illnesses and the development of research-based techniques for providing care to victims of Alzheimer's disease and related disorders (ADRD) and their families. Yet, while neuroscientists have identified multiple variants of dementing illnesses, care improvements still focus primarily on typical presentations of Alzheimer's disease, assuming people with dementia have similar symptoms and therefore, similar care needs. Thus, care programming for people with dementia has become essentially a "one size fits all" strategy.

This approach falls short with patients presenting with atypical presentations, rare, or multifactoral dementias -- perhaps as many as 20% of the patient population with irreversible dementias. Identifying unusual presentations, whether linked to Alzheimer's disease or not, can only help practitioners and caregivers to understand observed behaviors, distinguish symptoms, and plan care accordingly. For example, people with Lewy body disease have special medication needs that, if not understood, can worsen their symptoms dramatically. Or, expecting a person with a frontotemporal dementia to respond to environmental modifications in the same way as a person with Alzheimer's Disease, will result in frustration and disillusionment in caregivers.

Care of the person with an atypical dementias may be modified to accommodate subtle differences in disease presentation or an entirely new strategy may have to be developed - especially early in the disease. Caregiving professionals need to understand both the functions of the areas of the brain affected and how these areas influence symptom presentation in order to make decisions about care. Only then can the professional be truly helpful to the family.

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NOTES:

Key Points to Understanding the diagnosis of cognitive decline

1. The word “dementia” means that a person has changes in their memory, thinking, personality, and ability to do things because they have a disease affecting their brain. There are many diseases that cause dementia; Alzheimer’s disease is the most common in older adults.
2. Memory loss and behavioral change severe enough to change the way a person functions is never a normal part of aging and should be evaluated by a physician
3. Early diagnosis of memory loss is important because there are medications and programs of care that MAY slow the illness and because the person and family can receive.
4. Diagnosis of a dementing illness asks three questions:
 - a. Is memory loss present?
 - b. What could be causing it?
 - c. Are the behaviors and symptoms consistent with those we know of as Alzheimer’s disease?
5. Some dementias take several years to develop. In the early stages they look like Alzheimer’s disease but may change into a new diagnosis over 1-2 years.