Dementia is not a specific disease. It is a descriptive term for a collection of symptoms that can be caused by a number of disorders that affect the brain. Dementia can be defined as having diminished mental skills to the extent that it impacts normal activities and relationships. A person with dementia may lose their ability to solve problems and maintain emotional control. Dementia is not a normal part of the aging process. Many people live into their 90s and even 100s without any symptoms of dementia. Some forms of dementia
manifest with memory loss, others with behavioral, language or understanding losses. ¹

**Why is “Alzheimers’ Disease” and “Dementia” used interchangeably?**

“Alzheimers Disease” is often *inaccurately* used interchangeably with the term “dementia”. There are many causes of dementia. Alzheimers Disease is a major cause of dementia in the elderly, however, it is only one of the many causes of dementia. People with Alzheimers Disease have dementia. However, not all people with dementia have Alzheimers Disease.

**Location of Brain Abnormalities Impacts Specific Symptoms:**

The memory center is in the temporal lobe. A person’s ability to plan and understand (executive functioning) are controlled in the frontal region of the brain. Therefore, disturbances in that region will have an impact on those type activities and thought processes. ² Since the temporal lobe of the brain controls a person’s memory, disturbances in the temporal lobe will impact memory. ³

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² Lecture Notes: “Other Dementias” Genie Pritchett, MD. October 12, 2011 Alzheimer’s Association Education Symposium
Disturbances of the brain:
Disturbances to the brain are slow and cause distinct patterns of atrophy (reduction in the size of the brain) and others occur rapidly and may only involve a small area of the brain (such as a stroke). Some disturbances to the brain are “primary”—meaning they are the primary cause of the disease and not the result of another disease. In other instances the damage is “secondary”—meaning the dementia occurs as a consequence of another disease or an injury.
DIAGNOSING DEMENTIA

Patient History—when symptoms developed; overall medical condition; assessment of emotional state;

Physical Examination—to help rule out treatable causes of dementia; If a person is taking meds that may be causing or contributing to these symptoms the doctor may suggest discontinuation or replacement to see if the symptoms go away.

Mini-Mental State Examination—(MMSE) A test used to assess cognitive skills in people with suspected dementia. The test examines orientation, memory, and attention, as well as the ability to name objects, follow verbal and written commands, write a sentence spontaneously, and copy a complex shape.

Neurological Evaluations—A physician will assess balance, sensory function, reflexes, etc.

Imaging Studies—abnormal brain structure (neuroanatomy) or functioning can now be observed

Genetic biomarkers—genes that are associated with a disease

Protein biomarkers—proteins that are different than in a normal brain

TYPES OF DEMENTIAS

Fronto-temporal Dementia (FTD) FTD has three variants:

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1. Progressive Non-fluent aphasia (PNFA) primary manifestations are slow, stuttering, effortful speech with lots of mistakes, impaired repetition problems with verbs more than nouns, difficulties with complex speech and frequent misuse of grammar, eventually suffering from limited speech or mutism and impaired comprehension.

2. Semantic dementia (SD) fluent but empty speech, using many “canned” phrases (it’s that thing) inability to name things (cat vs. Chihuahua) loss of object identity (loss of the concept of dog and all domains of dogness-eventually can’t recognize a picture of a dog or the smell of a dog). Preserved calculation and visual-spacial skills.

3. Behavioral variant (BV) change in personality, decline in social conduct, inappropriate remarks, risk taking, impulsiveness, loss of insight, apathy, compulsive use of instruments (combs, pencils), poor control and eventual limitations in memory. Commonly misdiagnosed as depression.

**Biology of FTD:** FTD is the second most common form of dementia for those 65 years and younger. Average age of presentation is 60. The disease lasts for only 2-8 years on average. 40% of the familial cases are associated with a known gene. There is common protein pathology with other diseases such as Parkinsonism, Amyotrophic lateral sclerosis (ALS), progressive supranuclear palsy (PSP) and motor neuron disease (MND).

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6 Lecture Notes: “Other Dementias” Genie Pritchett, MD. October 12, 2011 Alzheimer’s Association Education Symposium
**Genetics:** There is a mutation of an arm of Chromosome 17 that causes a disruption of the gene progranulin. As a result of the mutation there is an aggregation of two proteins, tau and ubiquitin which will eventually destroy the neuron.

**What can we do about FD?** There is no cure but we can treat symptoms such as depression and agitation. Antidepressants, can be used, with the most common class being SSRI’s (selective serotonin reuptake inhibitors). Although antipsychotics are sometimes used, they cause many severe side effects and should be only used as a last resort under the guidance of a skilled physician. Non-pharmacologic interventions are effective. Addressing the needs of the caregivers, providing support groups and counseling are helpful. Learning behavioral management techniques can be empowering for the families that are struggling with this disease. And remembering that the bizarre features of the disease are part of the pathology and are not directed at the caregiver.

**Lewy Body Dementias:** There is a spectrum of diseases all of which have the Lewy Body as its primary pathology. The location of where the Lewy Body is found and the co-existence of other conditions determine whether a person has dementia with Lewy Bodies (DLB) or Parkinsons Disease Dementia (PDD). Similar genetic and environmental factors are present in both diseases. Exposure to high level of pesticides and high soil manganese content as well as the exposure to synthetic heroin and certain antipsychotics have all been associated with the disease. Both dopamine and choline deficits are identified with DLB and PDD. When the protein alpha-synuclein is aggregated a Lewy Body is created causing disruption of the neuron.⁷

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Key symptoms of Lewy Body Dementias: Both cognitive and motor symptoms are present. Executive function impairment is seen with notable visual spacial deficits. Hallucinations is a hallmark feature often accompanied with delusions and agitation. Another key feature is the dramatic fluctuations within the course of a day ranging from fatigue to extreme agitation. Memory is often preserved. Falls, incontinence, dizziness and hypotension are common because the autonomic system is impacted. Other key features are brady-kinesia, rigidity and postural instability as well as REM sleep disorder.

Vascular dementia:
Many different names are used to describe the process of neuron damage such as multi-infarct dementia, stroke related dementia, vascular dementia. Major risk factors that can contribute to vascular compromise are hypertension, smoking, diabetes, coronary artery disease, and atrial fibrillation.

Another way in which brains can be damaged and cause dementia is low blood oxygen to the brain (hypoxemia). Genetic vascular diseases occur, although rarely. Vascular lesions can occur throughout the brain and will therefore cause different symptoms depending on the place of injury.

If the injury occurs in the cortex one is likely to experience memory language thinking or behavior problems. If the damage occurs below the cortex emotions and movement are likely to be impacted.

Common symptoms of vascular dementia are memory impairment, language disorders, motor weakness, gait abnormalities and emotional lability. A stepwise deterioration is characteristic of this
type of dementia. Vascular dementia accounts for up to 20% of all dementias.  

CADASIL – Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarts and Leukoencephalopathy. This is a genetic small artery disease causing ischemic events resulting in mood disorder and apathy. Most individuals who have this disease have suffered with migraine headaches. The disease is seen in younger individuals with an average age of 45. There are many ethical issues surrounding genetic testing.

Progressive Dementias:

Some dementias get worse over time, gradually interfering with more and more cognitive abilities. These are called progressive dementia.

Some examples of progressive dementias are:

- Syphilis
- Lyme disease
- HIV
- Certain vitamin deficiencies
- Exposure to heavy metal
- Autoimmune Diseases such as thyroiditis and lupus
- Tumors that secrete certain substances that can cause brain inflammation

Creutzfeldt-Jacob Disease (CJD) or Mad Cows Disease. This is a grave disease with a very short course – usually between 5-12 months. It affects individuals between the ages of 50-70.

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It impacts cortical brain (cognition) and cerebellar brain (motor) as well as the extrapyramidal region causing behavioral and movement disorders. There are characteristic findings on the EEG and MRI that can help make the diagnosis.

**Limbic Encephalitis** is an inflammation of the brain caused by antibodies that have reacted to a tumor, typically small cell lung cancer. It can cause memory loss, degeneration in the cerebellum, movement disorders such as dystonia, rigidity and can cause profound sleep disturbances.

**Hashimoto’s encephalopathy** is caused by elevated thyroid antibodies and is often associated with other autoimmune diseases such as diabetes and lupus. Patients can present with tremors, gait abnormalities, confusion and behavioral problems.

**Alcohol-related dementia.** This is a significant and under-diagnosed cause of dementia in our culture where we support “letting granny have her drink”. Prolonged consumption of alcohol will impact judgment, decision-making, insight and can have motor symptoms that last beyond the time of intoxication. By the time the symptoms of dementia have occurred, most will not recover from the cognitive losses. Treatment often includes replacing thiamine.

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9Lecture Notes: “Other Dementias”  Genie Pritchett, MD. October 12, 2011 Alzheimer's Association Education Symposium
DIABETES AND DEMENTIA

Both too little blood sugar and too much blood sugar can cause immediate problems. Over time too much blood sugar in the brain will cause small vessel disease in the brain causing dementia that manifests like vascular dementia. When correcting low sodium (which occurs with very high blood sugar) a locked in syndrome can occur.

TREATMENT OF DEMENTIA

Aricept (donepezil)
Cognex (tacrine)
Namenda (memantine)
Exelon Patch (rivastigmine)
Reminyl (galantamine)  

PREVENTING DEMENTIA

The most important intervention for preventing dementia is correcting underlying diseases or risk factors such as high blood pressure, diabetes, alcoholism. Equally important is to detect early sign and symptoms of cognitive impairment and address potential causes. If it is a neurodegenerative process, once other causes have been ruled out, begin education and support.

PLANNING SUPPORT

Support for those persons caring for the person with dementia is essential and will ultimately affect the quality of life of both persons. Having the courage to address end of life issues, to establish each individual's values and goals is a choice that everyone can make. Using resources such as Alzheimer's Association, Frontotemporal Dementia Association, Parkinson's Association are wise choices to assist families.
1. Exposure to heavy metal can cause progressive dementia. a. true b. false

2. One of the key symptoms of Lewy Body Dementia is that both cognitive and motor functioning is affected. a. true b. false

3. A primary treatment for alcohol-induced dementia is: a. aspirin
   b. a bolus of alcohol c. thiamine d. a, b, & c

4. The Mini-mental state examination tests the following:
   a. drawing a complex shape
   b. drinking a glass of water without spills
   c. walking without stumbling
   d. speaking without stuttering

5. Injury to the temporal region in the brain will usually impact vision. a. true b. false

6. Aspirin is often used in the treatment of memory issues. a. true b. false

I have received, read, and understand the Continuing Education Module, DEMENTIA.

Signed: ________________________________

Name (printed): ____________________________Title: _______

Date: ____________________