Alzheimer's Disease

(Dementia, Chronic Brain Syndrome, Organic Brain Syndrome, Aphrenia, Athymia)

Description of the Disability

Dementia is a generic term to describe a general deterioration of the brain's ability to think and function. In particular, dementia involves problems with the ability to speak, to remember, to understand where one is, and to perform other cognitive tasks. There are often changes in emotional behavior and personality, especially an increase in paranoia, in aggression, and in general confusion. Dementia is most common in the elderly, especially in those over age 85, but some forms can occur at much earlier ages.

There are several types of dementia, but none of them are considered a normal part of aging. Some types of dementia are reversible, and are simply the brain's reaction to some form of stress on the body. The causes of **Reversible Dementia** include:

- Metabolic problems – including renal failure, liver failure, electrolyte imbalance, hypoglycemia, hepatic disease, and pancreatic disorders.
- Vision or hearing problems – leading to confusion and disorientation
- Malnutrition
- Endocrine Disorders – including hypothyroidism, hyperthyroidism, and adrenal problems
- Infections – in the elderly, the stress of fighting infection may increase confusion and disorientation.
- Internal pressure on the brain – including Subdural Hematoma (blood clot on the surface of the brain), Normal Pressure Hydrocephalus (gradual accumulation of spinal fluid pressure), and benign brain tumors
- Depression
- Side effects of medication taken for other reasons.

Other types of dementia are permanent and progressive, sometimes called **Cerebral Degenerative Diseases**. The most common cause of irreversible dementia is Alzheimer's Disease. Other causes include Atherosclerosis (also called Vascular Dementia, or VaD) (reversible if caught early), Multi-Infarct Dementia (see Stroke entry), Parkinson's disease (see entry), Huntington's Chorea, Pick's Disease, prion diseases (such as Creutzfeld-Jakob's Disease), AIDS Dementia Complex (see HIV/AIDS entry), and Multiple Sclerosis (see entry).

The first symptom of dementia is usually forgetfulness. As the disorder increases, the person will begin to experience reduced problem solving skills and problems with language (forgetting words, not understanding directions, etc.). As these difficulties grow worse, personality changes may occur. For example, the person may become stubborn or very nervous and anxious. When a

person with significant dementia feels frustrated, confused, ignored, or rushed, they may start to experience "**Agitation**" – which includes irritability, repetitive questions, repetitive phone calls, demands for attention and reassurance, pacing, rummaging, stubbornness, shouting, cursing, and, on occasion, physical aggression. Agitation is an expression of emotional distress, and it can often be diminished through reassurance or distraction with pleasant activities.

The person with moderate to advanced dementia may also begin to have **delusions** about things in their environment. Some particular types of delusions are especially common among people with dementia, including that they are in danger from criminals, that things of theirs have been stolen, that a spouse is unfaithful, that unwelcome guests are living in their house, that a relative is really an imposter, and that there are strangers in their yard. Delusions are, by definition, firmly held beliefs, so it can be difficult for caretakers to convince the person that these ideas are not true.

Individuals with moderate to advanced dementia may also become sensitive to changes in temperature and light or upset by a rearrangement of their environment. They may become frustrated with their own inability to do things that used to be easy for them. As they become more forgetful, they may have trouble recognizing family members or they may forget where they are. At times they may become confused and wander away, loosing track of where they are and how to get home. Routine and familiar surroundings become very important.

Although dementia is not part of the normal aging process, it is normal to see some reduction in memory with age. Researchers call this Age-Associated Memory Impairment or Benign Senescent Forgetfulness, and it is not considered a disease or disorder. Dementia is a much more significant reduction in memory and other cognitive functions, interfering with activities of daily life.

"**Sundowning**" is a sleep disorder specific to dementia. In some individuals, the internal clock controlling our sleep/wake cycle deteriorates. Although these individuals may be clear-headed during the day, in the evening they find themselves wakeful, restless, and confused. They become convinced that it is actually daytime and get themselves out of bed, get dressed, and get ready to begin the day.

Depression is a common coexisting disorder, and one that compounds the severity of dementia. Fortunately, depression is treatable. In some cases, treatment of depression leads to a significant improvement in symptoms even for individuals with irreversible and progressing dementia. Because of this, it is important to watch individuals with dementia for signs of clinical depression.

Alzheimer's Disease

Alzheimer's Disease is the most common cause of irreversible dementia in individuals over age 65. It was originally thought of as a "pre-senile" dementia because the first case, described by Dr. Alois Alzheimer in 1906, was a woman who died in her mid 50s after years of progressive dementia. When Dr. Alzheimer performed an autopsy, he found abnormal clumps and tangles in the nerve cells of her brain. Today these clumps (called "**Neuritc Plaques**") and tangles (called

"**Neurofibrillary Tangles**") are the definitive symptom of Alzheimer's disease. But we now know that most people who acquire the disease are older than 65.

Researchers do not know exactly what causes the plaques and tangles in the neurons, but they are somehow associated with the death of the cells. The neuritic plaques are made up of a protein called amyloid-beta. The neurofibrillary tangles seem to be damaged parts of the cell's internal protein skeleton, the microtubules. No one knows if either of these are side effects, causes, or consequences of the cell degeneration from Alzheimer's Disease. There is also a significant decrease in the amounts of neurotransmitters acetylcholine, serotonin, and norepinephrine present in the brain, and in the levels of glucose in the brain. Again, no one knows if these decreases are side effects, causes, or consequences of the damage. All that is known for certain is that these structural and chemical changes accompany the degeneration of nerve cells in specific parts of the brain as the disease progresses.

The degeneration starts with neurons in the Entorhinal Cortex and the Hippocampus, which are important for memory formation. From there, the damage spreads to other parts of the brain, including the Cerebral Cortex, which handles language and reasoning. In each affected area, the nerve cells gradually loose their connections to each other, and many die. This essentially disconnects parts of the brain from each other.

There are several theories about root causes of Alzheimer's Disease, including:

- Chemical Theories – At one time, researchers reported finding increased levels of aluminum in the brains of people with Alzheimer's Disease, but follow-up studies have not supported this claim. There is still interest in whether aluminum, zinc, calcium, or some other chemical might be involved.
- The Genetic Theory – There seems to be a genetic predisposition for Alzheimer's in some families, but in most cases the case for a genetic link is much weaker. One subtype of the disease, Early Onset Alzheimer's Disease (see below), is clearly genetic.
- The Autoimmune Theory – Some researchers think the damage looks like the result of an autoimmune disease, but the evidence is not conclusive.
- The Slow Virus Theory – Some similar types of dementia are caused by a "slow" virus that incubates for years before causing symptoms. Some researchers think there may be a virus behind Alzheimer's, but so far there is not much evidence for it.

A definitive diagnosis of Alzheimer's Disease is difficult while the individual is still alive. As noted above, the diagnostic symptom is plaques and tangles in the nerve tissue of the brain, which can only be examined after death. There is currently no blood or tissue test to prove the presence of Alzheimer's Disease. Instead, physicians have to use "Diagnosis by Exclusion" – documenting that the cognitive symptoms match, mapping brain damage (if any) through brain scans, ruling out everything else, and presuming the diagnosis from that. Because of the wide variety of other causes for dementia, ruling out everything else can be a challenge. As a consequence, some people consider Alzheimer's Disease to be vastly over-diagnosed.

There are two types of Alzheimer's Disease, Early Onset and Late Onset. In Early Onset Alzheimer's, recognizable symptoms show up in people in their 30s, 40s, and 50s. It is a rare form of the disease and appears to be due to an inherited dominant gene and occurs in only a handful of families. In contrast, most cases of the disease are Late Onset Alzheimer's, in which symptoms typically show up after age 65. For individuals who are 65 years old, only one to two percent have the disease. By age 85, the frequency has climbed to twenty percent. By age 90, the frequency approaches fifty percent.

Although Alzheimer's Disease is progressive, the rate of deterioration varies from individual to individual. For a particular person the rate will usually keep a steady pace, so an individual who experiences only a very slow deterioration will not likely have it speed up later on. Some individuals will only live 4 to 8 years after diagnosis, but many individuals will live 20 years or more. In the most advanced stages of the disease, individuals are unable to care for themselves and need almost total support in activities of daily living. Alzheimer's Disease is not directly fatally, but the degeneration of functional abilities contributes significantly to eventual death. The direct cause of death may be a coexisting condition, such as heart disease, or a result of their dependence and generalized weakness, such as pneumonia, a urinary infection due to a catheter, or complications from a fall. In particular, difficulty swallowing can cause people to inhale some of what they eat/drink and lead to pneumonia.

It is important to note that during the early stages of the disease, many individuals continue to live successfully by themselves. Symptoms, although progressive in the long term, will flare up and recede on a day to day basis. A person with mild to moderate Alzheimer's will have good days and bad days. With planning, support, and environmental modifications (see treatment, below), the person can continue to function successfully for many years. In fact, with supports, most individuals can live at home into very advanced stages of the disease. However, during advanced stages, any family member acting as a caregiver may themselves need supports and counseling.

Incidence Statistics

- An estimated 1.8 million Americans have severe dementia
- 1 to 5 million Americans have mild to moderate dementia
- An estimated 4 million Americans have some stage of Alzheimer's Disease (AD)
- Early Onset Alzheimer's is documented in approximately 120 families world wide.
- Age and family history are the dominant known risk factors for AD. Significant head injury and low education level are also risk factors.
- It is estimated that up to 25% of all individuals with AD live alone.
- At age 65, 1-2% of the population have AD. At age 80, 20% have it. At age 90, nearly 50% have the disease.
- Vascular Dementia (VaD) (from Atherosclerosis, or hardening of the arteries) is the second most common cause of Dementia in the elderly, after Alzheimer's.

Common Treatments, Medications, and Side Effects

So far, Alzheimer's Disease cannot be cured or sent into remission through medical treatment. However, the FDA has approved three drugs that seem to slow down the progression of the disease and temporarily improve some of the symptoms: Tacrine (Cognex), Donepezil (Aricept), and Rivastigmine (Exxelon) (See Drug entry on Dementia Drugs for side effects). Similar drugs are under development.

Some research suggests that three of the NSAIDs - ibuprofen (Advil, Motrin, Nuprin), naproxen sodium (Aleve) and indomethacin (Indocin) - reduce the risk of Alzheimer's by 30 percent to 60 percent. Other NSAIDs such as aspirin do not have the same effect, and no one knows why. The evidence is still uncertain and the use of NSAIDs has not been officially approved as preventative treatment for individuals at risk for Alzheimer's. Researchers have also found that, for post-menopausal women, hormone replacement therapy has a similar preventive effect. As with the NSAIDs, this therapy is not approved as sufficient justification by itself to receive hormone replacement therapy.

Intriguingly, some research shows that higher education may also help prevent or reduce the severity of Alzheimers Disease, but know one knows exactly why.

Other medications are sometimes used to reduce specific symptoms of dementia, such as memory problems, behavior problems or sleeping problems. These include antipsychotics (especially conventional antipsychotics), SSRIs, vasodilators, CNS stimulants, and some antiseizure drugs (especially carbamazepine) (see Drug entries for side effects).

Environmental modification is often a more successful solution, but it may be a short-term solution if the condition is progressive. The following changes seem to help, when appropriate to the person's functional level:

- Post notes about tasks (how to use the microwave oven, how to use the washing machine) in prominent places
- Post notes on cupboards and drawers describing their contents
- Post important numbers by the telephone and in a notebook
- Prepare a list of the activities for that day
- Follow a regular schedule
- Have someone go with them when they go out
- Let the person stay in a familiar environment or have familiar objects placed prominently in the environment
- Limited changes in environment
- Provide a quite environment with few distractions

- Provide a well-lit environment
- Maintain a comfortable temperature
- Provide a safe environment, with sharp objects or other sources of injury removed
- Install night lights

By keeping in mind the following, family members and service providers can improve interaction with a person with moderate to advanced Alzheimer's:

- When speaking, stand where they can see you. Use their name frequently. Touch them lightly to get their attention if they seem to drift.
- Identify yourself by name.
- Speak slowly and in simple sentences. Present one idea at a time. Don't rush and don't rush their replies. Ask simple questions if they seem to become confused.
- Use gestures and cues in addition to language. For example, if it is time to take them somewhere, gesture towards the door while speaking, or perhaps get their hat and coat out.
- Do not show your anger, it will only agitate the person more.
- Try to address underlying fears rather than fixate on specific complaints of the person. For example, if they insist you did not visit them yesterday, rather than debate the issue, calm their insecurity by telling them you will not forget about them.
- Long-term memories are often unaffected by the dementia. Asking them questions about their childhood or looking through a photo album can be a good way to help the person relax or stop fixating on something else.

Possible Functional Issues

- Difficulty problem solving
- Short-term memory problems
- Misplacing items
- Difficulty understanding and following complex instructions
- Difficulty speaking (recall of words, understanding others, etc.)
- Getting lost, even in familiar places
- Confusion about where they are, why, and what is going on around them
- Agitation and disruptive behavior when frustrated or upset
- Perseveration, repetition
- Passivity

- Personality changes
- Difficulty recognizing acquaintances (in advanced stages)
- Hallucinations/delusions (in advanced stages)
- Aggressive behavior (in advanced stages)
- Difficulty with awareness of immediate needs, such as thirst, need to use the restroom, feeling cold or warm (in advanced stages)
- Tendency to wander off and get lost (in advanced stages)

Initial Interview Considerations

Initial Questions

- What functional issues is the person having?
- How recently have they had these particular problems? (Gets at the pace of degeneration)
- At what particular times of the day do they experience more problems? In the evening, for instance?
- How significant are the functional issues when they are at home?
- How well are they able to live by themselves?
- What are their long-term plans for living arrangements, etc.?
- What hobbies do they have? How have those been affected by the disability?
- Do they ever have times when they get easily upset or when other people complain that they are being especially difficult or stubborn (more than is typical for them)? Is there any pattern to when these periods happen? Particular settings or times of day?

Initial Observations

- Does the person seem to keep track of the conversation, or do they loose it occasionally?

Interview Accommodations (if any)

- Hold the interview in a quiet place with few distractions
- Make sure the room is not too cold or too warm. Make sure it is well lit. Ask them if they are comfortable.
- Always introduce yourself, even in follow-up meetings.
- Speak directly to the person. Use their name often and, if appropriate, touch them lightly on the shoulder to get their attention if they get distracted.

Possible Accommodations and Assistive Technology

- Instructions to co-workers on helping the person if he or she is having a difficult day (be patient, explain where they are and why, let them do things their own way if it doesn't hurt anything, etc.). Perhaps provide a contact person and phone number in case an emergency arises.
- A co-worker mentor to help them if they become confused about job tasks.
- A flexible schedule with frequent breaks
- Regular visits by a job coach or advocate to assess any changes in support needs over time.
- Periodic review of employment by a support team to assess any changes in support needs.
- A well-lit, climate-controlled environment that is familiar to the person.
- Signs or notes posted in the environment as cues
- A notebook for notes to themselves
- Alternative transportation to reduce the chances they will get lost
- For individuals at risk of wandering off, the Alzheimer's Association provides identification products, support services, and a national registry through its Safe Return program (see resources at the end of this entry for contact information).

Career Planning Issues

- Individuals with mild Alzheimer's disease can still have many functional, satisfying years ahead of them. New ways are emerging to slow down, if not stop, the progression of the disease. Although the specter of advanced dementia may seem to loom ahead, that may be 2 decades or more away. It is important to stay focused on current abilities and symptoms.
- Try to plan ahead for the next potential phase of the disease and develop a graduated system of supports for the person to use as needed. Discuss with the individual and their physician what the expected pace of the disease will be. See if you can get an estimate of how current functional abilities may slowly decline and what additional abilities are most likely to become affected. Think about the supports that might be transitioned in if and when that happens.
- The individual's work history and social skills will probably not be affected.
- Coordination and dexterity will probably not be affected.
- Reading skills will remain unaffected through mild and moderate stages.
- Consider whether home-based employment might be a useful option. For some individuals with dementia, the home setting is comforting and calming. Also consider

whether the individual will need supervision or human contact in a home setting. Safety concerns should also be addressed.

- An environment with frequently changing features, tasks, or schedules may be challenging for the individual as the disease progresses.

Emerging Issues

- Treatment
- Supports for activities of daily life
- Causes
- Risk factors

Additional Information Resources

- ADEAR (The National Institute on Aging, Alzheimer's Disease Education And Research Center): www.alzheimers.org
- The Alzheimer's Association: www.alz.org
- Safe Return (A registry and support service of the Alzheimer's Association, for those at risk of wandering off): www.alz.org/caregiver/programs/safereturn.htm (800-272-3900)