

CLINICAL PRESENTATION, DIAGNOSIS, AND TREATMENT OF DEMENTIA

Interdisciplinary Colloquium: The Veracity of Memory (September 23, 2009)

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Slide 1.

The Veracity of Memory

The Human Dimension of Memory Loss



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Most individuals who suffer memory loss experience high levels of psychological distress, especially at the beginning of their illness when they are aware of the challenges that face them. Mercifully though, as the disease process advances, the individual who suffers from memory loss eventually loses insight into their downward cognitive decline. For their families however, the psychological pain is ongoing as they witness the slow multi year disintegration of their loved one's personhood.

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Let me tell you a story! I first met Mary some ten years ago at a mental health center in Lowell Massachusetts. Mary was a fifty year old, married white female, the mother of two teenage daughters. She was in good physical health. However, her family worried about Mary's memory because it was clear to them that she was experiencing difficulty remembering new information, and indeed on at least one occasion, she had become lost while driving in familiar local terrain. Mary had no history of

cardiovascular disease. She had never smoked or abused drugs and alcohol. She had never suffered a head injury. A CAT-SCAN of Mary's brain revealed mild atrophy of the cerebral hemispheres.

A Brief Story!

Mary's story!

Any likeness between Mary and anyone you know, or have known, is purely coincidental.

Mary held a Master's Degree in Nursing and had worked for a many years as an emergency room triage nurse at a well known Boston Hospital. By all accounts she had been quite adept at making clinical decisions in a fast paced and stressful environment. She had been both a valued and respected professional. However, she had been asked to resign her position some two years prior to my evaluation because she had evidenced an overall decline in her work performance. Her friends and colleagues had privately questioned the onset of dementia. I completed a full mental status evaluation of Mary (her Mini Mental Status Examination (MMSE) score being a 25), obtained a family history (completed a Genogram), referred Mary for neuropsychological testing, to further refine the assumed diagnosis of early onset dementia, and documented the reality that a maternal aunt had suffered from early onset dementia some twenty years earlier.

I next saw Mary two years later when she was being admitted to a nursing home in the greater Lowell area. By this time her MMSE score was 17. She had begun to wander, especially later in the day and at least once at night. She had left appliances switched on in the home, had allowed sinks and bathtubs to overflow, was experiencing delirium later in the day and at night, and was occasionally incontinent of urine and feces. However, she was in generally good physical health. Her speech was fluent and there was no clinical evidence of agnosia (not being able to identify common objects), apraxia (not being able to follow commands) or expressive or receptive aphasia (difficulty finding the right word or words when speaking, or not being able to understand the spoken word).

From that time onward, I would evaluate Mary and document her slow inexorable decline on a quarterly basis. Two years later, Mary's MMSE score was 10. She was oriented to place and person only. She was speaking in sentence fragments. She had become delusional believing that she was a nurse working on the unit where she was a patient. Although she could recognize her family, she could not name family members. She remained in good physical health.

Six years after first evaluating Mary, I could no longer conduct a MMSE because Mary could no longer communicate with, or engage in a meaningful fashion with others. She was oriented to person only. Her weight was declining. Her gait was unsteady (ataxia). She needed to be prompted to eat. She no longer “knew” her family. She was essentially mute, only responding very occasionally with “yes” and “no.”

Two years later, Mary was wheelchair bound. Her weight loss continued. She required total nursing care. She had to be fed. She had difficulty swallowing and had to be given thickened liquids to protect her against aspiration pneumonia. A year and a half later, Mary was confined to bed. She had by now adopted a fetal position as the neurons in her brain stem became impacted by the disease process. The family declined the insertion of a gastric tube to provide adequate nutrition, opting for a Do Not Resuscitate status and Care and Comfort only. Six months later, Mary died quietly in her sleep from pneumonia. She was 62 years of age.

Slide 3.

Memory Deficits---Cognitive Impairment

□ Four main diagnostic clusters of memory disorders:

- Delirium *
- Dementia *
- Amentia
- Amnestic Disorders



Diagnostic and Statistical Manual of Mental Disorders, Text Revision, (DSM-IV-TR) Fourth Edition (2005)

There are four diagnostic clusters of memory disorders. I will essentially confine my comments to delirium and dementia, the two diagnoses which are asterisked. However, let me speak briefly about amentia and amnestic disorders.

Amentia is the formal nomenclature for mental retardation or mental handicap. These unfortunate individuals never possessed a normal IQ because their cognitive abilities have been limited since birth. In the old days we referred to such unfortunate individuals as being morons, idiots and imbeciles. Over time, such medical terms took on a derogatory aura and consequently were dropped from the psychiatric lexicon.

Individuals who suffer from an amnestic disorder evidence a specific deficit of cognition, such as problems with short term memory without deficits in other areas of cognition. In other words, they do not suffer from dementia despite experiencing difficulties in highly circumscribed areas of their

cognitive processes, such as remembering new data. A good example is Korsakoff's Syndrome which is caused by long term alcohol abuse.

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Delirium

- ❑ The term delirium refers to the acute onset of cognitive impairment and changes in the level of consciousness---especially in children and in elders.
 - ❑ Delirium usually has an underlying physiological cause
 - ❑ Delirium usually waxes and wanes in intensity
 - ❑ Treat the cause and delirium usually resolves

Diagnostic and Statistical Manual of Mental Disorders, Text Revision, (DSM-IV-TR) Fourth Edition (2005), Pages 136 to 147.

Delirium reflects an acute change in mental status. The young and the elderly are impacted most frequently. Delirium develops quickly, usually in a matter of hours. It has a fluctuating course with the affected individual's level of consciousness ranging from acute disorientation to firm contact with reality. Individuals suffering from delirium often experience auditory and visual hallucinations, also delusions.

Delirium usually has an underlying physiological cause. Identify and treat the supposed underlying cause and the delirium usually resolves. In the elderly, delirium is often described as "sundowning," with changes of mental status occurring later in the day. The most common causes of delirium are infections, such as pneumonia and urinary tract infections, electrolyte imbalances, B12 and folate deficiency, thyroid pathologies, the use of narcotics, and the utilization of multiple medications which interact in problematic ways (polypharmacy), also drug and alcohol abuse, especially withdrawal. The take home message with respect to delirium is that if one can identify and treat the underlying cause, there are good prospects for its resolution.

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When we think in terms of dementia, we tend to think of Alzheimer's Disease (AD). AD is characterized by beta-amyloid plaques which form between the neurons, and tangles of twisted fibers of Tau within the neurons. Unfortunately, plaques and tangles can only be identified when the brain is examined after the death of the individual. The resultant accumulating damage to the neurons of the brain leads to a slow but inexorable cognitive decline which spans several or more years.

Vascular dementia is caused by abnormalities in the blood vessels of the brain. Such abnormalities include the restriction of blood flow to portions of the brain due to accumulations of athero-sclerotic

plaques, the total blockage of a blood vessel in the brain (an occlusive stroke) or the bursting of a blood vessel in the brain (a hemorrhagic stroke). All of these vascular accidents can cause damage to the brain's neurons. All strokes can cause cognitive deficits.

Dementia

Types of dementia.

- Alzheimer's Disease (Dementia) (Dr. Alois Alzheimer described the disease 1906)
- Vascular dementia
- Pick or Pick's Disease (Arnold Pick Described the Disease in 1892)
- Prion Disease (Stanley Prusner won the Nobel Prize in 1997 for his work on Prion Diseases)
 - Creutzfeldt-Jakob Disease in humans
- Brain trauma, drug and alcohol abuse and infections

DSM-IV-TR (2005), Pages 147 to 171



However, the specificity of the deficit depends upon which portion of the brain is damaged. As the number of vascular insults increase, neuronal damage advances. When that damage accrues in the cerebral hemispheres, dementia develops and advances. However, unlike AD where one sees a steady but inexorable cognitive decline, in vascular dementia we see the development of mild cognitive deficits which advance further with each successive stroke. As a result, we see a cognitive decline which follows a “stepped” trajectory, in which each stroke results in subsequent cognitive decline followed by a period of relative cognitive stability which is followed by additional cognitive deficits as strokes aggregate their damage to the brain's neurons.

Pick or Pick's Disease is known as a fronto-temporal dementia. It was first described by Arnold Pick in 1892. This disease often does not evidence itself, at least initially, by cognitive deficits, but via behavioral disturbance. The frontal lobes of the brain are the seat of our executive functioning, our higher human abilities, including judgment, anticipating consequences and editing our thoughts and restraining our actions such that we do not embarrass ourselves or offend others.

A number of years ago a well respected surgeon calved his initials into a patient's abdomen at the end of a surgical procedure. Obviously this act resulted in legal action on the part of the patient. During the preparation for a trial the surgeon was carefully examined by psychiatrists and neuro-psychologists. In addition, the surgeon submitted to a surgical procedure to obtain a biopsy of his frontal lobes. The trial concluded with a verdict of “not guilty by reason of insanity” because the biopsy of the surgeon's frontal lobes clearly showed the pathology associated with Pick's Disease.

Prion disease is known by various descriptors. In humans it is known as Creutzfeldt-Jacob Disease. In cows it is known as Mad Cow Disease. In sheep it is known as Scrapie. Collectively, all of these

diseases are known as variants of Spongiform Encephalopathy—because brain tissue when examined under a microscope resembles a sponge.

These diseases are caused by prions. Prions are self replicating proteins. Described simplistically, they have two shapes—a natural and a pathological form. In the natural form, prions serve a useful function in the brain. However, in their pathological form they self replicate, “eating away” brain neurons as they do so. In a short period of time, the victim suffers from dementia and usually dies within two to three years.

Stanley Prusner of the University of California at San Francisco (UCSF) earned the Nobel Prize in 1997 for his work on prions and the diseases they cause. His work demonstrated that prions cause pathology, that they were immune to the normal protocols we use to kill germs—autoclaving, bleach and boiling—and that they cause disease when ingested by humans and animals. The question not yet answered is whether we need to ingest a large single dose of prions once, or whether we need to ingest multiple small doses of prions over a period of time, for their pathological impact to be experienced.

Finally, brain trauma can cause dementia. We think of Dementia Pugilistic which is seen in individuals who have suffered frequent but relatively mild brain trauma due to Boxing. This disorder is also being seen in football players who suffer a number of brain insults during the course of their careers. We can also think in terms of Syphilis, where its causative agent, a spirochete, eats away brain tissue. Finally, brain infections by viruses and bacteria can end with cognitive deficits.

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Statistics on Alzheimer’s Disease (AD)

- ❑ AD affects an estimated 4.5 million Americans. The number of Americans with AD has more than doubled since 1980.
- ❑ AD is the most common cause of dementia among people age 65 and older.
- ❑ Increasing age is the greatest risk factor for Alzheimer’s. In most people with AD, symptoms first appear after age 65. One in 10 individuals over 65 and nearly half of those over 85 are affected.
- ❑ Rare, inherited forms of Alzheimer’s disease can strike individuals as early as their 30s and 40s.
- ❑ From the time of diagnosis, people with AD survive about half as long as those of similar age without dementia.

National Institutes of Mental Health (NIMH) (2009).




The statistics on AD are alarming. Individuals are more prone to developing AD with the passage of time. Although there are instances of early onset AD, think of Mary, most cases of AD are seen beyond the age of 65 years, becoming more frequent as an individual’s age advances.

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Dementia

- Initial Signs and Symptoms
 - Problems with short term memory
 - Difficulty assimilating new information
 - Becoming lost in familiar terrain
 - Difficulty balancing the checking account and paying the bills
 - Deterioration of language
 - Socially inappropriate behavior




I imagine that most of us are well ware of the signs and symptoms of dementia. Therefore, allow me to focus upon physical diagnostic criteria which help confirm the diagnosis. However, please remember that a firm diagnosis cannot be made until the brain of the individual is examined at autopsy when the pathologist looks for the characteristic plaques and tangles of AD.

A CAT SCAN of the brain of an individual suffering from AD will show atrophy or shrinkage of the cerebral hemispheres. One will also see corresponding enlargement of the brain's ventricles as the mass of the cerebral hemispheres declines. One may also see white matter changes if strokes have occurred. Obviously, with the passage of time, the individual suffering from dementia, irrespective of the cause, will present with a declining Mini-Mental Examination score. Generally speaking, what we learned last, we forget first. Therefore, grandma and grandpa can recall events of distant years ago while being unable to remember who visited them yesterday.

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Dementia

- Signs and Symptoms as Dementia Advances:
 - Failure of executive functioning
 - Changes in mood and emotions
 - Onset of psychotic pathology
 - Difficulty walking and swallowing
 - Total inability to care for self
 - Confinement to bed (in late stages)
 - Eventual death



So, for individuals who are suffering from dementia, long-term-memory tends to be good, at least until the later stages of the disease, whereas their short-term-memory is quite impaired. Two books provide vibrant and harrowing accounts of what it is like to look after an individual who is suffering from dementia — *The Long Goodbye* and *The 36 Hour Day*.

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“Treatments”

- ❑ Two classes of drugs are approved for the “treatment” of dementia:
 - ❑ The Cholinesterase Inhibitors
 - ❑ Aricept, Reminyl, Cognex and Exelon
 - ❑ Aspartate Receptor Antagonist
 - ❑ Namenda



Lexi-Drugs Comprehensive Guide to the Specialties (2009)
version 2/12/09

You will note that I have framed the word “treatments” in quotation marks because we cannot cure dementia. The reality is that the medications we currently prescribe to treat dementia serve only to slow the rate of cognitive decline, and that slowing is marginal at best. Giving a Cholinesterase Inhibitor or an Aspartate Receptor Antagonist may allow grandpa or grandma to live independently for an additional one to three months. These medications are also expensive, and as with all medications, they can cause significant side-effects. In fact I have had to discontinue these medications in many individuals because of problematic side-effects.

The current standard of care is to initiate and titrate upward a dosing of either Aricept or Reminyl (Cognex and Exelon are not used extensively anymore) to therapeutic levels before introducing Namenda, which is FDA approved for advanced dementia. However, I tend to discontinue both of these medications once an individual’s dementia has advanced to the advanced stage.

Slide 10.

Treating the Pathologies Associated with Dementia

- ❑ Many individuals who suffer from dementia also suffer from:
 - ❑ Depression
 - ❑ Anxiety
 - ❑ Lability of Mood
 - ❑ Psychotic Symptomatology

Individuals who suffer from dementia also may suffer from depression, anxiety, mood changeability (lability), and sometimes psychotic symptomatology. Various medications can be utilized to treat these conditions. The aim is to provide psychological comfort without causing sedation. If we can alleviate depression and anxiety, reduce mood lability and “take the edge off” psychotic thinking, we can reduce the psychological angst of the individual.

Slide 11.

Preventing Dementia

- ❑ The Nun Study
 - ❑ Chief investigator Dr. David Snowden, originally of the University of Kentucky
 - ❑ Ongoing longitudinal study of the Sister’s of Notre Dame
 - ❑ Multiple publications under the names of many authors



Simply Google “Nun Study”

The Nun Study is a longitudinal study of the Sisters of Notre Dame which is being led by Dr. David Snowden, formerly of the University of Kentucky and now at the University of Minnesota. The nuns who agree to participate in this study have to commit to annual evaluations of physical and mental health and to the donation of their brain for scientific study when they die.

The nuns are an ideal group of subjects because they all live similar lives. In other words, in the language of research, we have almost perfect control. In addition, a “perfect” record of the nun’s lives has been accumulated over the years. Many papers have been published by numerous authors based upon the findings of the Nun Study. These findings have added to our knowledge of dementia, its possible etiology and its possible prevention.

Notable findings include:

1. The nuns who wrote an excellent essay before they entered the Order, those who scored high on Idea Density, had a much lower incidence of dementia later in life than their peers who wrote essays low in Idea Density. One can hypothesize a number of explanations for this finding. For example, is dementia evidencing itself early in life or is education a protective factor against dementia?
2. Plaques and tangles were found in the brains of nuns who did not evidence any signs and symptoms of dementia during their life time
3. The more strokes incurred by an individual increases the risk of the development of AD. Plaques and tangles may not be associated with the development of AD but plaques and tangles in combination with strokes are associated with an increased risk for AD.
4. Tooth loss and poor dental hygiene, notably gum disease, is associated with an increased risk for dementia. This finding tie in with current thinking that the etiology of cardiovascular disease may be explained by bacteria causing atherosclerosis.

An overview of the findings of The Nun Study can be found in a Bantam Book titled: *Aging with Grace*.

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Can Dementia be Prevented?

- ❑ There are rare familial dementias that appear to be determined by genetic inheritance and as such are probably not preventable—Mary for example!
 - ❑ However, the Nun Study and a wealth of other studies strongly suggest there are life style choices implicated in the development of dementia.

Obviously there are some early onset dementias that have their etiology in genetics (think of Mary) and which at this point in time cannot be prevented. However, there is increasing evidence that there is much an individual can do to either delay or prevent the onset of dementia.

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What Can You do to Prevent or Delay the Onset of Dementia?

☐ Things you can do now!

- ☐ Reduce excess weight
- ☐ Eat a “Heart Healthy” diet
- ☐ Exercise regularly
- ☐ Treat high blood pressure and high cholesterol
- ☐ Maintain good dental health



Current scientific thinking strongly equates cardiovascular health with mental health. With the brain depending on a continuous and plentiful supply of oxygen and sugar (glucose) it stands to reason that any process which results in a decrease of blood flow to the brain could cause a deterioration of cognition. Therefore, a “heart healthy” lifestyle is also a “brain healthy” lifestyle. Everyone here knows what it is they have to do to promote cardiovascular health.

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So, let me talk about a few lesser known components of a “heart healthy” life style.

1. Alcohol in moderation is good for the cardiovascular system. However, the emphasis is on “moderation.” Two to three drinks a day for men and one to two drinks a day for women, are thought to be acceptable levels of drinking. But, remember, too much alcohol is not only bad for the cardiovascular system and the brain, it is also bad for the body in general.
2. Maintain your intellectual activities. For non-academics and older individuals, completing crosswords and playing Sudoku helps to keep the brain active.
3. Maintain your social connections as you age. Being engaged with life and involved with others is good for overall health. We all need something to get out of bed for each morning. Having a purpose in life, even after retirement is essential to physical health and psychological wellbeing.
4. Keep your stress hormone (cortisol) low by meditating or practicing yoga.

What Can You do to Prevent or Delay the Onset of Dementia?

▣ Things you can do now!

- ▣ Drink Alcohol in Moderation
- ▣ Reduce caffeine intake
- ▣ Do not smoke
- ▣ Do not abuse illicit drugs
- ▣ Keep your mind active---be involved
- ▣ Cultivate relationships and friendships
- ▣ Meditate



A mentor of mine once told me that as individuals' age they can follow one of two paths. The healthier group remains outwardly focused, engaged with others and socially involved. The less healthy group become inwardly focused and preoccupied with their own physical health and illness. As time advances they become disenfranchised from friends and society with a resultant further negative impact upon their physical and mental health.

Slide 15.

A Closing Personal Memory

A meaningful photograph!

Now to a degraded personal memory with respect to a photograph which was taken some forty or forty one years ago. Although this photograph captures a very important passage in my life, I cannot now

remember whether it represents the group after mid-course examinations or after final examinations. The group I am referring to is of me and my peer psychiatric nurses in England in the late 1960s.

Slide 16.

A Distant Memory!



Can you identify this speaker?

Slide 17: Any questions for any of the speakers?

* **Dr. IVOR PATTISON** earned his B.S. degree from the University of Massachusetts, Lowell, M.S. in Adult Psychiatric Mental Health Nursing from Boston College, and Ph.D. (Doctoral Fellowship) in Clinical Decision Making (Nursing) from Boston College. Ivor has practiced as a Psychiatric Nurse for the past 40 plus years. For the first half of his career he held a number of positions in psychiatric hospitals and units, both in the United Kingdom and in the USA. These positions ranged from working as Psychiatric Staff RN to serving as an Associate Vice President for Psychiatric Nursing Services. In his mid career, Ivor worked as a consultant to a variety of hospitals and nursing homes in Massachusetts, New Hampshire and Maine as they moved to establish, or improve, the quality of care on inpatient psychiatric and Alzheimer's Disease units. Since 1995 Ivor has taught within the Division of Nursing at Rivier College. He maintains a private mental health practice in Nashua, New Hampshire, where he provides care to adults and elders who suffer from depression, anxiety, mood disorders and dementia.