The following are clinical summaries of patients with pathologically confirmed dementia prepared by a dementia specialist neurologist unaware of the autopsy findings. These scenarios were derived from the patient’s entire medical record after personal identifiers, diagnoses, neuroimaging results, genetic analyses, and autopsy reports were removed.

We will ask the audience to review each case and complete a voting form twice – once after review of the scenario and once when both the clinical scenario and neuroimaging results have been presented. We will discuss features of the case that may be helpful after each vote and after neuropathology findings are presented.

Voting will be conducted through an audience response system. Like clinical practice, a diagnostic decision is necessary, even if there is some uncertainty. For each scenario, choose the most appropriate diagnosis and then indicate your degree of diagnostic confidence on a 3 level scale: VERY CONFIDENT, SOMEWHAT CONFIDENT, and UNCERTAIN. Votes will be tabulated and discussed.

FDG-PET scan results and neuropathology findings are not included to stimulate discussion. We hope you find this exercise interesting and informative.

Scenarios are listed in numerical order, except for scenario #2026, which will be discussed first.

Scenario #2026:

**HPI:** A 61-year-old gentleman with a 3.5-year history of progressive memory impairment. He is having occasional difficulty coming up with words during conversations and reports poor concentration while playing golf. It is difficult for him to follow a series of directions or commands. Three years into his illness he gave up doing the finances due to difficulties with calculations. He may get dressed too early in the day or leave too early for an appointment due to time confusion. He has also had recent problems operating power tools. He remains independent in personal care but may mismatch colors when dressing. No changes in sleep or appetite. Continues to drive without a history of getting lost.

**SH:** Retired billing specialist, high school education, married.

**FH:** Father with probable dementia, lived to age 75.

**Mental Status:** Oriented to season and was off by one month when given choices for month and year. Oriented to city but not to medical center name. Able to provide address and phone number with slight difficulty. Names simple objects but has difficulty with parts of objects. Repetition good. Spells world forward but not backward. Had difficulty with simple calculations. Attempted clock drawing but did not include numbers or positions for hands. Names only two professional golfers despite being an avid golfer himself. Registers 3 objects but recalls none at 3 minutes spontaneously, 2 with prompting. On formal neuropsychological testing, Verbal IQ 75, Performance IQ 65, MMSE 19.
**Neuro Exam:** Slight rest and postural tremor of upper extremities.

After 4 years he is no longer reading the newspaper other than looking at the headlines. Enjoys traveling and sporting events but needs to be coaxed to attend. On exam, good insight into his difficulties. Unable to name President. Makes errors with single digit calculations. Oriented to hospital but not city name. More difficulty naming items of clothing. Unable to count backwards from 20 as he reverses and begins to count forward half way through.

After 4 years 9 months, he continues to travel uneventfully. May reverse clothing when dressing himself. Able to prepare simple foods. Skips parts of the lawn when mowing and unable to figure out how to start a new lawn mower. No behavioral difficulties. On exam, oriented to year but not month. Needs hints to identify location as doctor’s office but does not name city. Impaired single digit calculations. Only able to follow single commands. Recalls 3 items briefly but not after a delay. Continued naming difficulties. General examination unremarkable.

After 5 years he developed anxiousness and agitation, which was not improved by buspirone. After 5 years 9 months he can still golf but with difficulty. No longer able to prepare simple foods but can assist with dusting or drying dishes. Needs more assistance with dressing and recently with bathing. Not incontinent. Has trouble opening the mail box or rolling down the car windows. Can not always find utensils when eating. Judgment now impaired. Will swallow everything put into a cup at once. On exam, does not speak unless spoken too. Unable to speak in full sentences. States name but not birthday. Unable to state full address or current location. Does not know the number or names of his children. Need choices to identify colors of objects presented to him. Unable to name most body parts or perform calculations. Can follow some one step commands. MMSE 9. No snout, glabellar, or grasp reflex elicited.

After 6.5 years, he needs significant assistance with bathing, shaving, and dressing. Not incontinent. Had brief hallucinations, possibly related to prior medication. Easily distracted when clearing dishes from the table. Has trouble opening a car door but still enjoys traveling with spouse. Taking alprazolam for anxiety and single episode of aggression. Speech consists of one to two word phrases. Walking slower. On exam, states name but not location or address. Unable to name most objects presented to him. Has difficulty understanding most components of general examination. Has moderate bradykinesia, right hand resting tremor and right hand postural tremor. Reflexes brisk and tone increased in upper extremities and neck. Arises and walks slowly with slowed turns.

Subsequent follow-up by phone. After 6 years 9 months, he resides in a nursing home and is very violent at night. Requiring haloperidol and lorazepam. Can feed self and remains continent. Otherwise needs all personal care provided.

After 7.5 years, he is wandering and striking out. Also displaying sexual behavior. Episodic crying spells. Remains continent. Receiving chlorpromazine.

After 8 years, still feeding self. Walking independently but wandering into other rooms and occasionally striking out. Does not participate meaningfully in activities. Occasionally speaks 1 to 2 words.

After 8 years 9 months, his behavior is calmer and he is receiving paroxetine. Follows simple commands by staff. Can participate in some group activities. Now incontinent and unable to feed self. Does not recognize staff but occasionally recognizes spouse.

After 9 years, he is having several falls and requires assistance for ambulation. Requiring complete care. Brightens when family visits. He died after 9 years of symptoms.

### Scenario #0037:

**HPI:** A 50 year-old right handed man with a longstanding history of bizarre behaviors, with progressive dementia with prominent behavioral disturbances, worse over the past eight years.

For many years, the patient’s wife had noted behavioral irregularities. Over twenty years ago, he managed his own business, but went bankrupt due to poor judgment. At that time he had been evaluated by a physician and
was simply told that he had a “chemical imbalance;” no further testing or evaluation was pursued at that time. Another example, also while in his 30’s, he had pawned all of the furniture in his home, despite not necessarily needing the money.

The first, more pervasive sign of functional decline began approximately eight years ago. At the time, he worked for an airline, where his initial role involved loading luggage. At that time, he required transfer to reservations due to back and knee arthritis. His wife reports that he was never as good at that position, and approximately four years ago he was suspended for inappropriate behavior. This was appealed and he was reinstated, but again fired for inappropriate behavior on the job. The second appeal was denied. He subsequently obtained another job in the reservations department of a different airline, where he remained until approximately two years ago when he was fired for being rude to a customer. Afterwards, he has remained effectively unemployed, although he has acquired several odd jobs, primarily in upholstering furniture for various companies. He remained able to drive himself to these sites independently without difficulty.

Additional examples of his behavioral changes include approximately 5 years ago, when he began speaking less and less. He also tended to repeat things over and over. Around that time he was started on galantamine, which improved his symptoms, enabling him to speak in longer sentences and to be more attentive during his odd jobs at upholstering. Despite this slight improvement, his wife had to take over financial and home management duties.

**Past medical and surgical history:** Obstructive sleep apnea and hypercholesterolemia. Remained physically active, regularly engaging in racquetball, basketball, and softball.

**SH:** Completed a high school level education, employed by airlines for many years loading luggage before transferring to reservations as noted above. Never smoked or drank alcohol on a significant basis. He lives at home with his spouse and children.

**FH:** Notable for dementia in the patient’s father, onset in later 50’s or early 60’s. Paternal grandfather died at age 82, markedly withdrawn before his death, although without clear diagnosis of dementia.

**Review of systems:** Positive for frequent awakening at night, general loss of interest, periods of anxiety and fearfulness, and trouble reading; gagging while eating, fluctuations in weight with a history of obesity, weight loss, and recent weight gain; cravings of sweets; chronic cough, shortness of breath with stairs; urinary hesitancy; back and knee pain. A Functional Activity Questionnaire also revealed an inability to manage business affairs, pay bills, work on hobbies, need for assistance with shopping, difficulty remembering appointments, and difficulty paying attention to TV or magazines.

**Neuro Exam:** The remainder of his comprehensive neurologic exam was normal. No snout, palmomental, grasp, or glabellar reflex.

**Mental Status:** Mini-Mental Status Exam score of 27/30. Not oriented to place. Could recall own phone number and address. Reported 3/3 items on immediate recall, 2/3 after 5 minutes, and 3/3 with a clue. Difficulty with copying a pentagon. Sentence was grammatically poor. Speech hesitant, with short sentences; often simply repeats “okay” to questions or commands. Naming intact. Able to mimic nonrepresentational hand movements, but had to correct 3 out of 5. Affect was distant, but cooperative, without anxiety or restlessness.

After eight years and eight months, there was significant decline in both cognitive function and ability to speak. He had quit working all together, and no longer spoke in complete sentences. He had word finding difficulties and would often simply repeat what was said to him. He was rarely oriented to time, date, day of the week, or season. His family felt that the preceding eight months represented the fastest decline that he had experienced in the last five years. His ability to play softball had declined, and he frequently became confused while running the plates, requiring a significant amount of verbal coaching. He exhibited irritability when dementia or Alzheimer’s was mentioned, and while he passed his drivers evaluation seven months prior (93rd percentile), he would become easily angered if anyone tried to stop him from driving. His behaviors became more repetitive. For example, he insisted on taking daily trips to the department store to buy a frame, return home, and frame one of his sports memorabilia pictures. He did this despite his wife’s repeated requests to stop. He saw a psychiatrist, who started him on memantine, which had no significant benefit. Review of systems revealed continued gagging while eating, and a five pound weight gain. On exam, Mini-Mental Status Exam score was 21/30. He was not oriented to place.
or time. He had deficits in recall, as well as significant difficulties copying a pentagon. He was unable to write a complete sentence. He exhibited hesitant speech with short responses and replied, often inappropriately, “yes” to most questions. The remainder of his neurologic exam was unchanged.

After nine years and five months, he had stopped driving. He spoke on average only once a week, in brief phrases, primarily stating “yes yes” or “what’s up?” He had developed many repetitive behaviors. For example, he would take his watch on and off repeatedly for hours, change his shoes as many as 50 times a day, or hide his clothes and then when he cannot find them, simply not wear that clothing item. He had become afraid to shower without assistance. He had become impulsive in eating, would occasionally swallow without chewing, and was unable to use utensils appropriately. He frequently coughed or choked when drinking or eating. He walked a lot, and had been sleeping better with the initiation of trazodone. He remained independent dressing and bathroom activities. On exam, he was dysarthric and exhibited frequent echolalia with prominent and persistent laughing. There was a positive glabellar reflex and a 2+ jaw jerk, but no grasp or snout reflex. Motor exam revealed increased tone at 2+ on the right persistently; no myoclonus or other abnormal movements. Reflexes were 2+ and symmetric throughout.

After ten years, he began to decline more rapidly. He could no longer read or write, and followed only very simple one-sentence instructions. He had not spoken in a month. He chewed his fingers and picked his nose. He was fearful of unknown situations. He developed tremors in both arms, and his movements seemed slower, although he continued to exercise daily, and walked around the block at least once a day. He often wandered off, and had become lost on two occasions. He needed help with dressing, bathing, and bathroom activities. His cough was weak, and he choked on saliva, pills, and thick or thin liquids, requiring a soft food diet. Swallowing study demonstrated that his epiglottis did not open with swallowing. He’d lost ten pounds within the last month. His family transitioned him to hospice care. On exam, his tongue exhibited fibrillations. Gag reflex required repeated attempts to illicit. No involuntary movements or grasp reflex seen. Jaw jerk was present. No snout or glabellar reflex. Fasciculation’s were noted on both forearms and upper arms. There was mild dorsal muscle wasting. There is a positive chadic reflex and positive cogwheeling rigidity in both upper arms. Gait, cranial nerves, and reflexes remained unchanged. EMG showed abnormal spontaneous activity with abnormal positive waves, fibrillation potentials, and fasciculations in muscles in all four limbs and the rectus abdominis muscles.

Scenario #0053:

HPI: A 34 year-old right handed man with a 1 year and 4 month history of progressive cognitive decline.

His wife first noted major personality changes and inappropriate behaviors just over a year ago, when he began working less. He at times said things in public that his wife felt were inappropriate, and had frequent arguments at work. On occasion, he was uncharacteristically aggressive towards his wife, and on several occasions, wandered into people’s homes. He appeared apathetic towards both his health and work activities, and he could no longer participate in extended conversations. Within approximately four months, he had quit his job, where he’d worked for the last fifteen years. He briefly found alternative jobs, but was let go due to persistent personality problems.

Approximately 7 months ago, he began smoking for the first time in his life. His habits became more restrictive. He began eating the same thing every day, tried to wear the same clothes every day, and refused to shave. He was inactive and often sat doing nothing, or simply watched television. He slept excessively, up to 14-15 hours per night. He was briefly admitted to a psychiatric ward, where the results of neuropsychiatric testing favored a primary neurological, rather than psychiatric, etiology.

SH: Three children, married. Recently started smoking as described above.

FH: Possible Alzheimer’s type dementia in grandfather, starting late in life. A cousin died at age 45, with an autopsy diagnosis of Creutzfeldt-Jakob disease.

Mental Status:  Mini Mental Status Exam score of 15/30. Bright, alert, and attentive, responded to questions quickly, although often perseverated. Not oriented to time, but quickly checked his cell phone in order to report the date correctly. Oriented to place. Correctly reported the current president. Recalled 3/3 objects immediately, but 0/3 at three minutes. Spelled “world” forward very slowly, unable to spell it backwards. Subtracted 7 from 100 once, but was unable to continue the task of subtracting serial sevens. Naming intact. Copied intersecting
pentagons correctly. Correctly read and performed the task of “close your eyes.” Correctly drew the numbers on the face of a clock, although placed the hands incorrectly to say “five minutes to three.”

**Neuro Exam:** The remainder of his comprehensive neurologic exam was normal.

After two years and one month, there was continued deterioration of both cognitive and behavioral aspects. He was no longer driving, and did appear to be upset about this. He continued to exhibit restrictive behaviors, and was bathing up to 20 times a day, often engaged in self-injurious behavior (e.g. hitting his head repeatedly on the back of the bed), and smoked excessively. His speech output became increasingly limited. He was independent in feeding, dressing, and bathroom activities, although required help with bathing and picking out clothes and had bladder and bowel incontinence on one occasion. He slept very poorly, although was in bed for 8 to 10 hours a night. Daily physical activity, such as walking, did not seem to improve his behavior significantly. He lost approximately ten pounds. On exam, he was orientation only to self, and perseverated on his name. He clapped his hands repeatedly, and paced the room repetitively. He was impulsive and walked away when spoken to. When asked to name his siblings, he named his children instead. He counted from 1 to 10, but could not count backwards. He echoed words and exhibited uninhibited utilization behavior. He mimicked motions of the examiner and would only stay in one place for a few minutes. Naming remained grossly intact.

After three years, he had been moved to a care center. His speech had progressively declined, and he rarely spoke in sentences; he continued to have frequent echolalia. His behavior was impersistent and he required constant direction. He paced and wandered. He remained continent, but sometimes inappropriate in his bathroom habits. Review of systems was notable for occasional choking and a 35 pound weight gain. On exam, he had difficulty with impersistence and was impulsive. He had decreased facial expressions and mildly nasal speech. 1+ jaw jerk and bilateral grasp reflex, with 3+ reflexes symmetrically, down going toes bilaterally, no clonus.

After three years and six months, his abilities continued to decline. He continued to wander continuously. He frequently drooled, and in the past four months, had become incontinent of urine. He began requiring a pureed diet. He rarely spoke, but occasionally would state his child’s name or mumble incoherently. Review of systems was notable for a 50 pound weight loss, representing significant fluctuation within the last year. On exam, he was mute. He was restless and unable to track objects. There was marked lack of facial expression. Reflexes were 3+ in the lower extremities; no glabellar, snout, or suck reflex, although there was bilateral grasp reflex.

After four years and one month, he had become completely mute, except occasional unintelligible mumbling. His movements seemed slower, and he often appeared lethargic. Exam showed an additional 20 pound weight loss. He was mute, and in contrast to previous encounters, sat quietly and no longer wandered. He did not make eye contact, and played with a toy telephone in a repetitive manner. Cranial nerves continued to show marked lack of facial expression. Tone and reflexes remained normal.

**Scenario #0145**

**HPI:** A 58-year-old female with a 4-year history of increasing inability to care for self. One year after her husband divorced her, the patient began to miss appointments and a year later she had difficulty with calculations and managing her money. She was fired from her job as an assembly line quality inspector for lapses in attention. Speech problems became more prominent a year later, with paraphasic errors and difficulty with expression. More recently she has lost interest in regular activities and has daily crying spells as well as occasional outbursts of anger. She has no prior psychiatric history. She is sleeping well and her weight is stable. She is independent in bathroom activities, but is unable to do more challenging household chores like cooking meals. She has been picking at sores and developed mild neurodermatitis. The family notes myoclonic jerking of her limbs several times a week.

**SH:** Retired secretary and quality inspector, high school education. No history of tobacco, alcohol, or drug use.

**FH:** Negative for dementia or other neurological disease.

**Mental Status:** MMSE 13/30. Oriented 3/5. Able to register three objects, but recalls none after a short delay. Unable to spell word backwards, repeat a sentence, follow a command, or copy a figure. Does not know her date of birth, the correct order of the months, and calls February “fairyberry.” Could not identify her shoulder or knee,
but successfully touched her ear when asked to. Unable to distinguish left from right and missed 5/6 nonrepresentational hand movements. The patient only speaks when spoken to and her answers are short. Normal affect and cooperative.

**Neuro Exam:** Easily distracted, myocolonic jerking of her stomach muscles.

After 6 months, she has declined considerably and needs help with basic tasks like dressing or remembering to wash her hair and brush her teeth. She has had two episodes of urinary incontinence and one of fecal incontinence. She is even more tearful than before and hit her daughter during one of her outbursts. She has more trouble expressing herself and is speaking very little. Sleep and appetite remain are not changed. Laboratory workup is negative, including CSF analysis. An EEG is unremarkable. She has not responded to a SSRI or TCA trial. MMSE 7/30.

After 1 year, the patient has been moved to an assisted living facility because she needs help with most ADLs and has had several falls climbing to the second floor of her daughter’s house. She has difficulty consolidating sleep and it is now the patient’s chief complaint. Caregivers report she has left the facility on a number of occasions and often wonders where she is and why. She goes long periods without talking and is unable to use the nurse call button when needed. She has largely adjusted to the new living situation, though, with fewer episodes of agitation and anger. MMSE 2/30.

After 2 years 1 month, there is continuous bowel and bladder incontinence. She is in a locked unit because of numerous attempts to elope and uses one to two word responses if she speaks, which is increasingly rare. She has regular outbursts of angry yelling of noises at herself while in front of a mirror. The patient has gained a large amount of weight and her access to food has to be restricted.

After 2 years 7 months, she is physically threatening to staff and transferred to a geropsychiatric unit. A variety of medication trials helped to find a combination of valproic acid, trazodone, and quetiapine that allowed the patient to go back to the locked unit. Falls have become more frequent and she now has no interest in eating or drinking and had to be admitted to a hospital because of severe dehydration. She only says two words, “there” and “babies.” The myocolonic jerks are much more common and she is now tremulous at baseline.

Seven years after the onset of symptoms, the patient died in at an outside hospital of an unknown cause.

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**Scenario #0175**

**HPI:** A 74-year-old male with a 6-year history of progressive anomia. The course has been insidious and at the time of his initial assessment, he continues to function at a high level, but now often has trouble remembering the correct names of familiar locations, friends, and even close family. He recalls past experiences in detail and manages his finances. More recently he developed paraphasias and confabulation, but refuses to be corrected. He has limited insight and minimizes his deficits. No changes in sleep or appetite. Continues to drive without getting lost. Had a trial of donepezil and memantine, which was discontinued because of anorexia.

**SH:** College educated and a retired corporate executive. Married for 53 years and no history of alcohol, tobacco, drug, or caffeine use.

**FH:** Father with probable late-onset dementia. Brother with dementia.

**Mental Status:** Alert and oriented x 4/4. Attention 5/5, short-term memory was 3/3 on immediate recall, but only 1/3 with cueing at delay. Able to recall the location of money hidden in the room – immediately and after a 30-minute delay. Long-term memory is preserved to personal events, but he is unable to remember the name of the President. He confabulates some details, saying he had gone to Cuba when he only went past it on a cruise. Language fluent, except for occasional word-finding difficulty. Naming impaired and difficulty producing representational items such as the okay sign. Visual-spatial function is intact. MMSE 26/30.

**Neuro Exam:** Normal except he has apraxia on heel-knee-shin. Unable to tandem. Bilateral grasp response and glabellar response.

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3ES.004 – page 33
After one month, he scores 30/30 on the MMSE. Routine laboratory workup for dementia is unremarkable. Patient’s wife reports he has nocturnal myoclonic jerks. A trial of clonazepam is initiated.

After two months, he has restarted memantine and is tolerating it well. Neuropsychological testing shows severely amnestic pattern of memory loss on tests of new learning, profound dysnomia/anomia, impaired semantic retrieval, and mild visuospatial impairment. Clonazepam is ineffective. Carbidopa-levodopa and galantamine started.

After eight months, he has occasional delusional ideation, such as being convinced he is on the correct road even after his wife shows him a map. Displaying obsessive-compulsive behaviors - repeatedly going to a room in a local hospital where he had been treated in the past and only stopping after they threatened to call the police. Carbidopa-levodopa was changed to pramipexole, which was also ineffective. MMSE 30/30.

After 1 year 4 months, he has started going on four to eight hour walks without explanation. He continues to walk across a golf course he has been told not to and crossed a freeway to pick up a coin he saw on the other side. Obsessed with visiting churches he has attended and going to old residences and demanding to be let in. More emotionally labile and no insight into his deficits. Galantamine is stopped and he becomes calmer without ill effect on his memory. MMSE 29/30. Repeat neuropsychological testing; 3MS-R 88/100, verbal intelligence 109, performance/perceptual intelligence 84, profound impairment in new learning, and visuospatial processing improved.

Tens years after the onset of symptoms, the patient died in a hospice of an unknown cause.

**Scenario #0219:**

**HPI:** A 69 year-old right handed man with a six-year history of gradual onset dementia.

Initially changes we noted by the patient’s wife six years ago, when the patient began doing things that were out of character for him. For example, on one occasion, she prepared a large amount of soup, which the patient then threw down the sink. This behavior was uncharacteristic of him, and when he was asked why he did it, he was unable to explain. In addition, he had become more irritable, rather than his previously calm demeanor.

Approximately five years ago he started having visual hallucinations. He experienced some abrupt falls, had become hyper somnolent, and exhibited rapid symptom fluctuations, changing over a matter of days. He began complaining of diplopia, and had trouble with swallowing both liquids and solids. He had been using a cane for the past year, and fallen 5 to 10 times over the past 5 to 6 months. During sleep, he acted out dreams and his wife noted hypnagogic jerks. This had become more frequent over the past years.

Four years ago, he stopped driving, per a doctor’s recommendation. Around that time, he also started going to an adult daycare. The patient denied symptoms of depression, although he did endorse occasional dull mood. He remained independent with his ADL’s.

**Past medical and surgical history:** Charcot-Marie-Tooth (CMT) diagnosed in 1972, with subsequent surgeries for bilateral foot drop.

**SH:** Married, lives with his wife. Bachelor’s degree in industrial technology with concentration on aviation. Retired 4 years ago because of age. Also used to act as a quality assurance specialist for the government. No significant alcohol, caffeine, or tobacco use.

**FH:** father with ALS, two younger brothers with CMT, mother with CMT and dementia.

**Review of systems:** remarkable for generalized fatigue, diplopia as described above, difficulty swallowing, poor peripheral circulation, GI disturbances including constipation and dysphagia; urinary frequency and urge incontinence, dizziness, generalized weakness, and memory loss for short and long-term events. Also positive for a mild tremor bilaterally, aches and pains, anxiety, depression and irritability, and frequent daytime naps.
**Mental Status:** Mini Mental Status Exam score of 27/30. Had significant difficulty learning word pairs, and scored 1.5/3 on immediate recall, 0/3 on the second try, and 3/3 on the third try. Also with difficulty copying intersecting pentagons. He otherwise scored full points. Correct recall of current president and vice president’s names, but had difficulty generating a list of prior presidents. Could recall some current events, although without specific details. Language notable for hypophonia, mild palilalia, and significant word finding difficulty. Naming and repetition were intact. Complex three-step comprehension was impaired. Reading and writing were preserved. Praxis was impaired to representational and nonrepresentational items; visuospatial function was impaired to clock drawing. Luria triangles and squares were extremely disorganized, with a tendency toward perseveration of parts of the prior figure and on squiggly lines. Calculation skills were significantly impaired, could not add 58 and 13, nor calculate the number of quarters in $6.75. Generated 11 animals in one minute. Mood and affect were normal, although there was mild abulia.

**Neuro Exam:** Steppage gait consistent with his longstanding bilateral foot drop. Posture mildly stooped. Could not perform tandem, Romberg positive. Retro pulsed on pull test. Small pupils bilaterally; notable delay in response during eye movement testing. Significant psychomotor retardation with hypomimia and paucity of movements, as well as micrographia, bradykinesia, hypokinesia, and hypometric movements. Increased tone with cogwheeling characteristics on facilitation maneuvers. Mild intermittent tremor in the upper extremities bilaterally. Power preserved except for 0/5 dorsiflexion at the ankle, secondary to his CMT. Sensory: Intact, except for graphesthesia impaired bilaterally. Coordination intact. Reflexes: 2+ throughout, except for 0 at the ankles; down going toes bilaterally. Positive bilateral grasp responses.

After seven years and eight months, there had been a notable decline in cognitive and physical abilities. His eating preferences became restrictive, preferring only sweets and junk food. His hallucinations continued; examples included seeing people that were not there or the entertainment center falling down. These hallucinations were bothersome, but not severely distressing. He required increasing assistance with his ADLs. On exam, Mini-Mental Status Exam score was 19/30. Not oriented to time or place. Scored 2/3 on registration, 2/3 on attention, 2/3 on comprehension, and 0 on copying a design. Voice was monotone and dysarthric. Facial expressions flat. Motor exam notable for a mild, infrequent resting tremor (right greater than left), slight rigidity (right greater than left), and mild bradykinesia and hypokinesia. Finger tapping and rapid alternating movements mildly impaired. Leg agility moderately impaired. Slow to rise from a chair with his arms crossed. Posture moderately stooped. Retro-pulsed on the pull test.

**Scenario #0258:**

**HPI:** An 80 year-old right handed man with a three month history of gait instability, falls, and memory loss. He had the gradual onset of gait difficulties over several months. He was unsteady, describing the sensation of “a seesaw in my head,” and began having frequent falls. He began requiring assistance to ambulate independently, and initially began using crutches, but soon progressed to a walker, and then a wheelchair at times. Prior to the onset of these symptoms, he had been in fairly good health and was taking only simvastatin, omeprazole, aspirin, and a multivitamin. Soon after the gait instability began, he began having difficulty with memory and speech. His wife reports his symptoms noticeably worsening from week to week. Due to difficulties with disorientation, he began having difficulty managing his usual affairs, and he was forced to stop his part-time job as a pharmacist. He remained able to perform basic activities of daily living (ADL’s).

**Past medical and surgical history:** hypertension, prostate cancer in remission, bladder cancer status post resection, hypercholesterolemia, and coronary artery disease status post CABG. Hip and knee replacements, shoulder surgery.

**SH:** married and living with his wife. Lifetime non-smoker and does not currently drink alcohol. Worked as a pharmacist, retired three month prior due to symptoms described above.

**FH:** Unremarkable for known neurologic disease.

**Review of systems:** positive for complaint of generalized weakness, dizziness, occasional diplopia, shortness of breath with exertion, swollen ankles on long trips, nocturia and incontinence over the past 2 weeks. Also complaint of back pain and a 7 pound weight loss in the last year.
Mental Status: Mini Mental Status Exam score of 16/30. Disoriented to time, except for month and year. Correct to type of facility and floor, but was otherwise disoriented to place. Able to state own address, but not telephone number or birth date. Registered 3/3 objects, but recalled 0/3 at five minutes. Unable to spell backwards and could not copy a design. Counted to 10, as well as backwards from 20. Repetition intact, but naming impaired. Unable to name his four children. Decreased speech output and fluency, with fragmented speech and a flattened affect; appeared withdrawn. Attention fluctuated, but remained alert throughout the encounter.

Neuro Exam: The patient arrived in a wheelchair and required significant assistance to stand. Stride length was slightly shortened, and gait was markedly unsteady. At times, he unpredictably lost control of his legs, but did not appear to be frankly ataxic. Gait was somewhat asymmetric and unstable, due in part to failure of attention. Notable for both an upward and downward gaze palsy, as well as an alternating dysconjugate gaze. No nystagmus. No fasciculations of the tongue, nor dysarthria. Normal tone. No tremor or startle myoclonus. Grossly normal coordination. Reflexes: 1+ and symmetric, with down going toes bilaterally, no clonus.

Shortly after initial evaluation, the patient was admitted to the hospital for approximately one week. Laboratory evaluation revealed CSF with a glucose of 82, total protein of 52, one RBC, no WBCs, and was negative for Borrelia burgdorferi, Cryptococcus, measles, and VDRL. Full CSF cytometry was negative for evidence of lymphoma, monoclonality or acute leukemia. CSF was sent for 14-3-3, which was pending on discharge. During hospitalization, he underwent a left frontal craniotomy for open biopsy. Upon discharge, he was transferred to a care facility for hospice care. On discharge exam, he was not oriented to person, place or time. He was able to follow only simple commands. There was poor repetition and fluency, with frequent paraphasic errors. Cranial nerves remained unchanged, with persistent upward gaze palsy. Strength and tone remained normal. Gait continued to be severely impaired, and the patient was no longer able to stand, despite two person assist.

Scenario #0387

HPI: A 58-year-old female with a 3-year history of progressive dementia with change in personality, disinhibition, compulsive behavior, and inattention. The patient lost her job one year prior for odd behavior such as repeatedly bringing her dog with her to work, despite being told not to. Since then her decline has been precipitous and she now lives in an assisted living facility. She has lost interest in hobbies and her social life. She speaks less and has trouble participating in conversation because she cannot focus for extended periods of time. The patient has very little insight into her decline. She cares for her activities of daily living, is continent, and has an appropriate level of hygiene.

SH: Retired secondary school administrator. Widowed five years prior.

FH: No known neurological disease.

Mental Status: MMSE 21/30. Patient is only oriented to place. She registers two objects, recalls one. Unable to spell world backwards or follow three step commands. Copied intersecting pentagons and named objects correctly. Too impersistent to mimic hand movements and had difficulty sitting still in her chair. Very little speech production and often would not answer direct questions. Monosyllabic replies and flat affect.

Neuro Exam: Walking is slowed, but it appears to be lack of initiative because she can speed up with encouragement. Arm swing and tone normal.

After 2 months, the patient has begun to wander all day long in and around her care facility. She walks more slowly and sometimes drools. She can become overly affectionate at times, but otherwise she does not engage or speak unless spoken to. Review of outside neuropsychological testing reveals significant deficits with word generation, judgment, and attention. A speech pathology evaluation shows no dysarthria or dysphagia. There is non-functional reading comprehension for long passages, but intact for shorter ones. Her responses to questions were often inappropriate and she has echolalia. MMSE 22/30.

After 4 months, she is overeating and has disinhibited affection for strangers. She rarely speaks, but giggles persistently throughout the day.
After 9 months, she is not attending to her hygiene and will not change her clothes unless told to do so. She wears adult diapers because of occasional urinary incontinence. She has undressed herself in public on a number of occasions. She is grabbing objects that are not hers and refusing to give them back. She persistently asks staff and residents at her care facility for money. When focused she can read, give cogent responses to questions and recall the name of a nursing student she only briefly met on a previous visit. She is impatient but easily calmed. Neurological exam is nonfocal. MMSE 22/30.

After 1 year 5 months, she has been moved to a locked dementia unit because of repeated elopement from her assisted living facility. She is now always incontinent of urine and often of stool. She has been physically threatening with staff when restrained from inappropriate or dangerous behavior such as stealing from other residents or making sexual comments. She takes all of her clothes out every day and piles them in a compulsive manner. She is hoarding candy. She will stuff her mouth so full of food she cannot chew and laugh hysterically while spitting it out. Her speech is limited to single word replies that usually demonstrate understanding of the question. She enjoys playing football during recreation time and takes long walks with staff. She is constantly touching various parts of her face and laughs and grunts inappropriately during the interview. There is evidence of mild self-mutilation on her face and arms.

Four years after the onset of symptoms, the patient died at an outside hospital of an unknown cause.

Scenario #0654:

HPI: A 61 year-old right handed woman with a four year history of progressive memory loss, aphasia, and personality changes.

Four years ago, the patient became more reclusive and sat in front of the TV for hours. She did not sleep well, and stopped cleaning her home. She became less talkative, showed less interest in family and social events. She began to exhibit uncharacteristic levels of affection towards others, and often wanted to shake other people’s hands at inappropriate times.

Two years ago, the patient lost her job as a first grade teacher due to an inability to interact appropriately with the children and other teachers. Upon evaluation by a psychologist at that time, she demonstrated an MMSE of 20/30. A trial of an anti-depressant was unbeneﬁcial. Repeat MMSE seven months later was 16/30.

At present, she is independent in bathroom activities, but requires reminders to shower. She is no longer able to cook, and can wash dishes only by hand, as she no longer can use a dishwasher or clothes washer appropriately. She continues to sew, and still remembers to feed the cats and dogs. She no longer walks the dog independently, but she and her husband walk one mile several times per week. She has not had any falls or difficulty walking. They occasionally go on excursions and she is appropriate in the grocery store and restaurant, although she is unable to shop for herself. She is sleeping well, and is emotionally stable.

Past medical and surgical history: Hypercholesterolemia.

SH: Married, teacher. Non-smoker, non-drinker. She lives with her husband and daughter.

FH: Her mother died in her 70’s with a progressive dementia syndrome. Her father had diabetes and Parkinson’s disease, but no diagnosed memory problems. She has five brothers, all living without memory complaints. She has eight children, all in good health.

Review of systems: Remarkable for weight gain, loss of interest, and anxiety.

Mental Status: Mini Mental Status Exam score of 10/30. Oriented only to state. Registered 3/3 objects, and recalled 2/3 after a delay. Naming was impaired; repetition was intact for a short phrase. Unable to write a sentence, and instead wrote “compleated sentence.” Traced, rather than reproduced a drawn figure. Exhibited limited verbal fluency, speaking only when spoken to, and in single words. Counted from one to five. Recalled her own birth date, but not her address.
Neuro Exam: Unremarkable, except for masked facies and dysarthria with both hypokinetic and spastic components. The was also an intermittent left hand tremor. 1+ symmetric reflexes, with absent Babinski bilaterally.

After 4 years, and two months, there was little change in day to day activities or behaviors, although, she was speaking less and less. Neuropsychological testing, using a severe impairment battery, was performed. The extracted MMSE score was 12/30. On exam, she was unable to write a sentence, and was completely disoriented. Language notable for perseveration and echolalia. Flat affect. 1+ tone in upper extremities, and unlike the previous visit, no tremor was noted. There was notable wasting of the first dorsal interosseus bilaterally; no fasciculations. Tone was normal in the lower extremities. Sensory and coordination grossly intact, reflexes 2+ in the upper extremities, 3+ at the knees and 2+ at the ankles without clonus. 3+ jaw jerk and bilateral grasp reflexes, but no glabellar or snout reflex. An EMG showed abnormal spontaneous activity in all four extremities with reduced motor unit recruitment and high amplitude motor units consistent with motor neuron disease. Speech pathology evaluation reported progressive expressive and receptive aphasia with mixed hypokinetic and spastic dysarthria. No evidence of swallowing difficulty.

After 4 years, and nine months, her speech and functional abilities continued to decline. She used only two or three words at a time, and spoke rarely. She had difficulty in understanding commands and could no longer use the picture book communication aid that had been provided by speech pathology. She continued to bathe and use the bathroom independently, although she required monitoring during eating, and often did not use utensils normally, often holding them upside down. On exam, MMSE score was 0/30; the patient is unable to follow commands or respond to questions. She was cooperative, placid, and happy, although repeatedly wanted to shake the examiners hand. Cranial nerves exhibited symmetric face, with mild weakness throughout and pooling of mucus in her mouth. No fibrillations, although there was mild atrophy of the tongue. Motor exam showed mildly increased tone in both biceps. 2+ increased tone at the knees. Bilateral wasting of the hands, with no tremor or fasciculations. Reflexes showed 2+ jaw jerk, 2+ at the biceps, 3+ at the knees, 2+ at the ankles, toes equivocal bilaterally, no clonus.

After five years and two months, there was continued deterioration of her speech. She would merely nod yes or no to questions occasionally. She began having difficulty swallowing. A modified barium swallow showed evidence of aspiration. She lost approximately 26 pounds, and experienced constipation. She seemed restless and often paced the house. Her temperament remained mellow. She participated in daily walks and car rides. On exam, MMSE score was 0/30. She paced around the room at times. Gait remained intact. Cranial nerves unchanged, with the exception of decrease in blink rate and flattened smile. Motor exam showed bilateral muscle wasting in the shoulders, arms, and hands. No fasciculations or tremor. Reflexes unchanged.

Scenario #0665

HPI: A 73-year-old female with a 3-year history of non-fluent aphasia. Her initial symptom was increasingly shorter sentences and eventually a frank slowing and paucity of speech. Subsequently she became incapable of expressing her thoughts in writing, but continues to manage most of her finances and sign her checks. In both speech and writing she has paraphasia. According to her daughters, who provide the majority of the history, the patient’s personality is largely preserved. She appears to understand written and spoken language and becomes very frustrated with her aphasia. She recently moved in with her daughter after her husband died and it became apparent she was not able to perform her IADLs.

SH: Worked as an RN and is retired for 10 years. Recently widowed and moved from Texas to Utah to be near family. No history of alcohol, tobacco, or drug use.

FH: A sister has vascular dementia. Her mother and a maternal aunt have Parkinson’s.

Mental Status: MMSE 17/30. Partially oriented, 2/5. Attention 0/5. Registration 3/3 and recall 2/3. Missed one stage of a three-stage command and could not read and obey “close your eyes.” Unable to copy intersecting pentagons or assess three word pairs. Finds three pieces of hidden money immediately and at half-an-hour. Knew the current President and Vice-President, but could only name two prior Presidents and was unfamiliar with current events. Severely dysfluent, difficulty producing words, but able to name objects and repeat syllables such
as pa/ta/ka without difficulty. Impaired praxia and visuospatial function. Unable to perform moderate difficulty calculations, and word list limited to two animal names per minute.

**Neuro Exam:** Significant for disordered smooth pursuit and saccadic tracking, mild oropharyngeal dysphagia, dysarthria, speech apraxia, and limitation of vertical gaze.

After 4 months, the patient refuses to perform the MMSE because of frustration with her aphasia. Routine screening laboratory studies for reversible causes of dementia reveal a B12 level of 200. Neuropsychological testing was technologically difficult because of the aphasia, but constructional praxis and higher executive function is markedly impaired. An ophthalmologist has diagnosed her with eyelid and oculomotor apraxia. A trial of memantine fails to help.

After 2 years 4 months, the patient is rarely speaking spontaneously and has problems with eye opening. She has begun to fall and broke her hip a year prior. She continues to fall at least weekly. Her dysarthria has worsened and she is frequently choking on liquids. She is very attentive and cooperative during the exam and appears to understand most commands, but her severe aphasia limits the remainder of the language exam. Vertical gaze is even further impaired, upgaze more than down. The right face is less mobile and has no nasolabial fold. The left face intermittently grimaces in a manner consistent with alien movement. Power is full throughout. The right upper extremity is held in a flexed posture at rest and demonstrates asymmetrically increased tone, writhing postural movements, downward drift, apraxia, and ataxia. She leans to the left with her walker and her right foot is externally rotated. There is no tremor. She is dependent on family for all of her ADLs at this point.

Five years after the onset of symptoms, the patient died of an unknown cause, but in the days prior to her death she had cataract surgery and a fall resulting in a cervical fracture.

**Scenario #2040:**

**HPI:** A 62 year old right handed woman with a 2 year history of changes in personality and memory. The most notable change has been a lack of participation in daily activities. She no longer does housework or participates in conversations, preferring to sit in a chair all day. She stopped driving after becoming lost on familiar routes 1 year into her illness. She is “very forgetful”. She has purchased items she does not need, and she is eating more. She may laugh inappropriately or become upset, and has made statements that people are “trying to dupe her”. Her behavior is more cooperative since haloperidol was initiated.

**SH:** Married, 8th grade education, homemaker.

**FH:** Unknown.

**Mental Status:** Alert and oriented to person, place, and time. Digit span 6 forward and 5 backward. Names current President and 4 of 5 previous Presidents. Single error with Serial 7 subtractions, but unable to calculate nickels in $1.35. Naming, repetition, and general speech intact. Draws a clock but not a cube. Recalled 3 of 5 objects at 5 minutes. Affect somewhat peculiar with occasional appropriate smiling but generally flat affect and poor awareness of cognitive changes. With formal neuropsychological testing, Verbal IQ 74, Performance IQ 85, Memory Quotient 108.

**Neuro Exam:** Unremarkable except slow, deliberate gait. No grasp, snout, root, palmomental, or glabellar reflexes.

After 2 years 3 months, a trial of nortriptyline has not affected her mood. She is speaking less and hums frequently. She complains of being tired and hungry all the time. She sleeps frequently during the day. Needs prompting to do household chores and spends most of her day watching television. On exam, markedly apathetic. Knows date and location. Names current but not prior Presidents. Registers 3 objects and recalls 1 after delay. Performed simple but not complex calculations. Excellent naming but occasional paraphasias. Poor comprehension with reading. No right/left confusion.

After 3.5 years, she is even less interested in activities. Incontinent of both urine and stool. Eating constantly and gaining weight. Will prepare food when it is available and will dress herself if clothing set out. Does not do other
household activities or personal grooming unless forced. Frequently hums and has prominent echolalia. Wanders frequently, walking to nearby homes of children without getting lost. On examination, frequently hums and repeats words. Recalls 3 objects at both 5 and 10 minutes. Names current President but refuses to name others. Names day and year but does not answer date or month. Can write name and address but has difficulty copying a cube or drawing a clock face. Names all objects presented to her and mimics hand movements well. 2+ snout reflex. No glabellar or palmomental reflex.

After 4 years, she remains incontinent. May scream suddenly for no apparent reason then start to laugh. Hums almost constantly while awake and wants to eat constantly. Frequently refuses to walk, but may wander outside in cold weather without a coat. On exam, humming almost constantly and picking at clothing in purposeless fashion. Occasionally repeated words of others but no paraphasic errors or neologisms. Paucity of spontaneous speech. Accurately followed all 1 step and some 2 step commands. Remembered 3 of 4 words at five minutes.

After 5 years 3 months, she is showing even less initiative and spends much of her time lying in bed. Frequently makes non-verbal sounds, i.e.: clapping her hands. Appetite uninhibited. On exam, initiated no speech but able to state name and type of car her husband owns. Frequently clapping. Followed simple 1 step commands variably. Grasp reflexes bilaterally. No snout or root.

After 6 years, she has no intelligible speech. Becomes agitated in new situations. Seems to recognize familiar faces. Becomes distracted, and may spend time simply staring at her hands. Now needs occasional encouragement to eat. Needs assistance for dressing and near total care for personal hygiene. On exam, had constant chewing motions. Vocalized with high pitched moans. Frequently attempted to leave the examination room. Frequently clapped. Unable to respond to any commands or cooperate with formal testing. Gait slightly slowed and stooped. Grasp reflexes present.

Patient died after 14 years of symptoms.

Scenario #2162:

HPI: An 80 y.o. woman with a 6 year history of progressive memory loss. She resides in a senior apartment, close to her daughter, but is becoming increasingly forgetful. She recently got lost after taking a bus downtown and required help from a stranger. She has been calling her daughter several times per day without realizing she has just phoned. She is able to do her own shopping and light meal preparation, though her daughter took the knobs off the stove to prevent her mother from leaving it on. She has significant visual and auditory impairment and bumps into objects when walking. She can no longer read or quilt.

SH: Unknown

FH: Unknown

Mental Status: Alert, oriented to year, season, and month but not day of week. Knows city, state, town, and clinic name, but not county. Registers 3 objects but recalls only one after several minutes. Unable to subtract 7 from 100 and only 3 letters correct when spelling world backwards. No difficulty with language. Mild impairment when copying a design.

Neuro Exam: Unremarkable except for visual (macular degeneration) and auditory impairment.

After 6 years and 9 months of symptoms, she has relocated to a nursing home where she socializes with others and remains in a good mood. She is generally incontinent of urine and occasionally of stool. On exam, oriented to own name and location, but not date or names of children. No knowledge of current events. Performs simple but not complex calculations, names most objects presented to her visually, and performs a simple two step command. 0 out of 3 recall after 5 minutes but 2 with clues. Speech fluent without word-finding difficulties. Possible extensor plantar response at right foot.

After 7 years and 3 months, she is able to feed herself without assistance but needs help with dressing and is often incontinent. Calm and without depression. On exam, oriented to city and “doctor’s office”, but unable to name children or name of current facility. Identifies body parts and simple objects. Fluent speech without word-
finding difficulties. Counts backwards from 20 without difficulty. 0 out of 3 recall after 5 minutes. Slight stoop when walking. Plantars both flexor.

After 7 years and 9 months, she is wandering from her facility. Paces in the hall. Assists others as possible, helping them get out of restraints. Plays piano but repertoire limited. Mood and sleep good. On exam, poorly oriented and unable to perform simple calculations. Does not learn 3 objects presented to her even after several trials. Mimics 50% of hand movements. Unable to name several simple items, and has obvious word finding delays, speaking in less than full sentences. Gait slightly slowed and stooped, but normal tone and no tremor or myoclonus. Right plantar response extensor.

At 8 years and 3 months, MMSE was 10. Boston Naming Test 5/60.

At 8 years and 9 months, she had experienced worsening gait in the setting of an infection, but this subsequently improved. Still feeding herself. On exam, oriented to city but inaccurate with personal information. Makes paraphasic errors when naming objects. Can point to named body parts. Makes several errors with subtractions but can repeat a lengthy phrase. Poor mimicking of hand movements. Mild facial and body bradykinesia. No extra movements. Possible right plantar extensor response.

After 9.5 years, she feeds herself with mild difficulty and performs practice lessons on the piano. Remains incontinent and occasionally wanders. Acts as though she is dusting furniture, but does not use a dust cloth if provided. On exam, does not speak unless spoken to, but is able to repeat phrases and use appropriate sentences. Oriented to city but inaccurate with personal information. Paraphasic errors with naming. Can count to 10 forward but not backwards. Accurately performs simple commands. Moderately stooped posture with significant imbalance and moderate bradykinesia. Increased tone in upper extremities. Right plantar response extensor.

After 10 years, she needs help with eating and speaks less frequently. Rarely plays piano. Frequently naps during the day. Occasionally pushes friend in wheelchair. Walks slowly but no falls. Nods head when name called. Remains incontinent. On exam, responses markedly delayed. Oriented to name but no longer to city. Follows 1 but not 2 step commands. Mixes up relationships of immediate family. Can distinguish two different colors. Does not visually track individuals or speak spontaneously. Has moderate kyphosis. Uses arms to stand, and crosses legs mildly when walking. Small steps do not improve with coaching. 4+ tone in neck with resistance to vertical movements. Eye movements absent to up gaze but full horizontally and with down gaze. Tone 3+ in right arm, 2+ left arm, and normal in legs. Reflexes brisk with upgoing toe on right.

Patient died after 10.5 years of symptoms.

**Scenario 2178:**

**HPI:** A 59 year old gentleman with a 1.5 year history of memory loss. He is having trouble remembering names, including those of his own children. He had difficulty remembering routes as a bus driver and was laid off. He has problems recalling conversations, and he uses an answering machine to record all phone messages. He will forget tasks unless written down. He is slower to come up with words during conversation. He remains independent in his self care. He navigates a boat and drives, but needs directional assistance in less familiar areas. No incontinence.

**SH:** College degree in industrial arts, prior service manager and bus driver, married.

**FH:** Mother with dementia developing at age 84. Possible memory loss in father in late 70’s.

**Mental Status:** Alert, oriented times 3, with slightly halting speech. Recalls 0 of 3 objects at three minutes, but 2 with significant prompting. Poor calculations and concrete interpretation of proverbs. Fund of general information poor. Knew little about WWII despite serving in this war. Normal constructions and right/left orientation. Naming correct 80% of the time. Occasional paraphasic error when reading, and poor recall of information read. Repetition and comprehension intact. No apraxia. With formal neuropsychological testing, Verbal IQ 86, Performance IQ 99, Memory Quotient 96, Boston Naming 14 of 60.
**Neuro Exam:** Unremarkable, though snout, glabellar, and trace suck reflexes present. Grasp and palmomental reflexes absent.

After 2 years, on exam, recalled 0 of 3 objects at 10 minutes. Digit span 5 forward and 4 backwards. Significant word finding problems during spontaneous speech and naming accurate only 50% of the time. Repetition and spatial orientation good. Able to recall place and date of birth.

After 2.5 years, he still drives but needs instructions if driving anywhere unfamiliar. He is having more difficulty reading to his children. Able to perform daily chores, but may take longer. Personality is more easygoing than in the past, though he interacts less often with friends. On exam, disoriented to hospital. Named President with significant coaxing. Abstraction poor. Could not discuss Pearl Harbor or other WWII events. Made word substitutions.

At 3 years, he is having more difficulty with word finding. Has difficulty following a television program or reading a story. Continues to drive, but wife accompanies him to give directions. No mood changes or crying spells. On exam, unable to name medical center or city, but knew state. Recalled birthdate but not anniversary. Named 5 of his 6 children with prompting. Right/left discrimination poor with evidence of perseveration. Unable to name any of 6 objects but could describe their use. With formal neuropsychological testing, Verbal IQ 77, Performance IQ 92, MQ 76.

After 3.5 years, he is no longer able to perform household repairs. Does not recognize family members by name. Able to make coffee, but having difficulty vacuuming the house or mowing the lawn. No longer reads, but plays bridge and watches sports on television. Needs prompts for showering. Does not handle money but still drives. Got lost once when walking, though able to give directions to driver who took him home.

After 3 years 9 months, he is more stubborn. Unable to recognize photographs of his mother or brother in family album. Performs personal hygiene with prompting. On exam, substitutes phone number for street address. Oriented to year but not month or date. Unable to name children and gives incorrect number of children. Able to copy simple figures and mimic hand movements. With formal neuropsychological testing, Verbal IQ 66, Performance IQ 81, Wechsler Memory Scale 67, and MMSE Score 9.

At 4 years 3 months, he frequently arises at night. Helping out less with housework. Sings in church at incorrect times and has hugged and kissed people inappropriately. On exam, oriented to name but not age or place. Unable to name most objects or body parts. Able to write a sentence spontaneously but content and grammar inappropriate. Grabbed medical staff inappropriately.

After 5 years, he is still able to prepare his own breakfast but may do so much earlier than usual. Dresses himself and walks the dog. Will lick the dishes at a restaurant, so social outings curtailed. Also continues to exhibit inappropriate affection toward strangers. No incontinence. On exam, unable to state name or age. Had difficulty cooperating with much of the examination and tried to kiss the examiner.

After 5.5 years, he has a home health aide and behavior is deteriorating. Urinates in public and attempts fondling. Becomes physically abusive when frustrated. Demonstrating day/night confusion. On exam, disoriented with no ability to follow commands. Behavior inappropriate with frequent laughter. Does not seem to recognize wife. Started on haloperidol.

The patient did not return to clinic and died after 8.5 years of symptoms.

**Scenario #2184**

**HPI:** A 61 y.o. right handed woman with a nearly four-year history of gradual memory decline. Symptoms in the first year included becoming easily frustrated and tearful. She had difficulty using equipment at work, forgot to take her medications, and lost track of the day of the week. She put dishes away incorrectly at home. In the second year she had trouble caring for grandchildren, feeding them the wrong foods at the wrong times. Was making mistakes with her checking account. She quit working and stopped driving. An antidepressant helped her mood, but not her memory loss. After three years of symptoms, she had stopped cooking. In the fourth year she had difficulty dressing, putting clothes on inside-out or backwards. She still dusts, vacuums, and washes dishes,
but needs prompting for laundry. She does not get lost when walking within the grocery store or within one block of home. She feeds and bathes herself. She watches television but is unable to recall details of recent programs. She has had no changes in speech or continence. Sleep and appetite are good.

**SH:** High school graduate, lives with husband, former courthouse employee.

**FH:** Mother had memory loss beginning at age 75 with significant impairment at the time of her death at age 90. Father lived to age 75 with normal memory. No siblings affected.

**Mental Status:** Alert, oriented to person. Able to state address and phone number. Disoriented to year and state. Fluent speech with mildly impaired naming but intact repetition. Difficulty with two step commands. Left/right orientation intact. Digit span of four forward. Recalled 0 of 3 objects at 3 minutes and 1 with choices. Unable to discuss current events but could name her grandchildren correctly. Unable to mimic hand movements. Poor clock drawing. No obvious neglect. With formal testing, MMSE score of 12, verbal IQ 72, performance IQ 59, and Wechsler Memory Scale score 57. Named 19 out of 60 objects presented visually.

**Neuro Exam:** Unremarkable except glabellar, snout, and grasp reflexes; palmomental and suck reflexes absent.

After 4.5 years of symptoms, she needs supervision in all dressing. She can perform limited dusting and vacuuming, but folds towels incorrectly and cannot perform other household activities. Her energy is low and she has occasional crying spells. Insight is limited. She does not prepare meals, and occasionally needs assistance to find the bathroom in her home. She had a single recent episode of possible hallucinations. Does not always recognize daughters. On examination, she is disoriented to day, year, and unable to describe place. Difficulty following simple commands. Unable to name or point to most body parts. Cannot mimic any hand movements or provide address. Grasp, snout, and glabellar reflexes persist, with new finding of palmomental reflexes.

After 5 years of symptoms, she has hired assistance at home. She spends more time in the bathroom, but does not know what to do when she gets there. Plays with toilet water and/or excrement. Not incontinent. Occasional crying spells, but sleeps well and appears interested in activities around her. On exam, can point to some body parts but very poor naming and inability to follow most commands. Orientated to person only with fluent spontaneous speech, though content irrelevant. Needs to be led to the examination room due to visual spatial difficulties, but general exam otherwise unremarkable.

At her last visit, 5.5 years after symptom onset, she has relocated to a nursing home. She needs more help with eating and using the bathroom, but is not incontinent. No behavior disturbances. Bumps into things frequently. On exam, speaks in full sentences that are tangential. Protrudes tongue to command but can not close eyes to command. Points to some body parts but unable to name any objects presented visually. Able to state name and previous address, but not names of family members. Hesitant gait but examination otherwise unremarkable.

Patient died 8.5 years after symptom onset.

**Scenario #2233:**

**HPI:** A 65 y.o. right handed gentleman with a 9 month history of changes in personality and cognition. Initial symptoms included irritability and difficulty filling out invoices at work. He avoids making decisions at work. He is helping in the building of his new home without apparent difficulty.

**SH:** 12th grade education, partner in a plumbing and heating contracting firm, married.

**FH:** Negative for memory loss.

**Mental Status:** Alert, oriented to date and location. Normal affect. Knows current but not prior Presidents. Poorly informed about World War II details. Makes paraphasic errors when reading. Has difficulty drawing a clock and unable to copy a cube. Requires multiple attempts to register 3 objects and then recalls 2 at five minutes. Performs simple but not complex calculations. Makes paraphasic errors with repetition and names half of objects presented to him.
**Neuro Exam:** Mild peripheral neuropathy.

After 1 year he is retired and less irritable. He is driving without difficulty and still able to help in the construction of his home (doing plumbing/handiwork). Received a new hearing aide, with significant improvement in communication skills. On exam, naming for simple objects now normal, but other deficits (poor recall of prior Presidents and historical events, difficulty repeating phrases) remain. Formal neuropsychological testing revealed Verbal IQ 74, Performance IQ 89, Wechsler Memory Scale score 84, MMSE score 24. Named 39 out of 60 objects presented visually. Some evidence of mild depression.

After 1.5 years he can still perform plumbing and other construction tasks, but is more forgetful and needs to make multiple trips to get the correct supplies. Still driving without difficulty. On exam, oriented to name, place, and date. Knows address but not phone number. Difficulty naming parts of objects. Able to follow 2-3 step commands. Even simple calculations poor. Could not copy cube. Thinking concrete and judgment vague. Unable to discuss current events. Registered 2 of 3 objects and recalled none at five minutes. General exam normal.

After 4 years, he is driving limited distances without difficulty. Does numerous chores around the house. Mood good. On exam, oriented to month but one day off on date. Has difficulty with historical and geographic knowledge questions. Markedly impaired confrontational naming. Unable to recall phone number or address, but repetition intact and able to mimic 4 of 5 non-representational hand movements. Recalls 1 of 3 items without clues after delay. No improvement with clues. General exam unremarkable.

Subsequent follow-up by phone, and after 4.5 years, he is able to prepare his own lunch and order food when eating out. Memory fluctuates in relationship to chemotherapy he is receiving for systemic malignancy. Becoming frustrated about loss of skills.

At 5 years he needs some minor verbal cues. No behavior difficulties. At 5.5 years he had an episode of getting lost, but was able to recognize this and notify the police. Dressing, shaving, and showering on his own. Cognition still fluctuating with chemotherapy.

At 6 years, he is still independent with eating, bathing, and dressing. He attends a day center. Helps wife with household tasks. May get dressed in middle of night, but never wanders. He died shortly after the last phone contact, after 6 years of symptoms.

**Scenario #2236:**

**HPI:** 70 y.o. gentleman with a 5 year history of progressive difficulties with visual tasks. The first few years of his illness were characterized by marked difficulties driving, such that his wife traveled with him during his trips as a salesman. She took over financial management during the fourth and fifth years of his illness. He no longer cooks unattended and is unable to perform many simple tasks around the home. He can become angry due to his visual difficulties and is slightly more emotional but does not have inappropriate crying or laughter. There is no history of hallucinations or delusions. He has not gotten lost in his home. No incontinence. He performs well on activities of daily living other than reversing articles of clothing when dressing.

**SH:** Retired salesman, lives with wife.

**FH:** Older brother with a rapidly progressive dementia (became incapacitated within 2-3 years of onset) attributed to Alzheimer’s disease; father had significant personality changes and violent behavior before his death, but in a more slowly progressive fashion. No other family members with neurologic illness.

**Mental Status:** Oriented to date and location. Speech loquacious and somewhat tangential. Normal mood and affect. Naming intact for simple but not complex objects. Paraphasias with repetition of lengthy phrases. Registers three items but recalls none at five minutes spontaneously and two with prompting. Mild errors with calculations and identification of presidents. Concrete descriptions of similarities but able to provide some abstract answers for proverbs. Followed a three step command and had no apraxia. Intact writing but marked errors when copying a complex figure. Unable to draw a clock face, due to multiple distortions of the clock and
corresponding numbers. Formal neuropsychometric testing revealed verbal IQ 93, performance IQ 62, and MMSE score 22. Naming low at 17 but verbal fluency normal at 14. Wechsler Memory Scale MQ of 84 with 38% recall for Part 4 and 75% for Part 6.

**Neuro Exam:** Motor impersistence on visual tasks. Slight left pronator drift with equivocal left plantar reflex. Diminished dexterity with rapid alternating movements on the left. Mild simian posturing during gait examination. Intact graphesthesia.

After 5.5 years of symptoms, he remains independent in most activities of daily living. He maintains an active social life and takes initiative with certain household chores (taking out garbage). He is not able to cook without supervision or perform complex repair jobs. He has difficulty distinguishing bills of different denominations and spoke with a stranger in a grocery store, not realizing it wasn’t his wife. He is unable to tell time on analog watches and has difficulty dialing the telephone. Stopped driving. On exam, he demonstrates an excellent vocabulary and is orientated to place but one year off on date. Intact naming, repetition, and writing to dictation. Recalls recent world events and three of three items at five minutes, with a small clue for one item. Poor performance on calculations and clock drawing. Makes several errors with nonrepresentational hand movements. Difficulty with similarities and proverbs. Motor impersistence when following targets and less attentive to stimuli in left visual field, though no field cut noted. Slightly stooped posture with gait. Suggestion of a rooting reflex but no other frontal release signs elicited.

After 6 years of symptoms, the patient is no longer able to read the newspaper and will wear his clothing inside out. Still golfing. Has trouble recognizing objects handed to him, and is no longer cooking at all. On exam, fully oriented with fluent speech. Registers four objects and recalls two after two minutes. Significant difficulties with calculations, proverb interpretation, and constructional tasks. Normal praxis. Vague about recent world events. With formal testing, MMSE 20, performance IQ 61, verbal IQ 76, memory quotient 70 with no recall after a delay. General exam unremarkable.

After 6.5 years, he has become more emotional but remains socially active, including continuing his volunteer activities. Still dressing self but with difficulty. Able to make coffee. On exam, disoriented to month, requires multiple attempts to register three objects and recalls none spontaneously after five minutes.

After 7 years, he is attending adult day care. He is having trouble choosing the correct utensil, and dressing skills are variable. Can prepare coffee but not assemble a sandwich correctly. Disoriented to day, particularly after napping. Speech is fluent with occasional paraphasic errors and significant word finding difficulty. Unable to count backward from 20 past 17. Vague about historical and political events. Disoriented to day, date, and month. Has extinction to the left with double simultaneous stimulation 50% of the time, otherwise unremarkable exam.

After 8 years of symptoms, patient moved to a nursing home, and after 8.5 years he had trouble recognizing his wife. Remained continent but needed assistance with bathing and dressing.

He died 8 years 9 months after symptom onset.