



Dementia

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Case Presentation 1

Case Presentation:

- 69 yo man living with daughter and her family.
- Very set routine.
 - Up at 6 AM
 - Washes up and dresses
 - Assists the family with getting ready for the day
 - Walks down the block to get the paper
 - Returns, reads the paper and eats breakfast.
 - Etc...

Sudden change...

- One day, the news stand was closed.
- Man was lost for hours.
- When found he seemed unaware of what happened, failed mental status testing.





Case Presentation #2

Case Presentation 2

- Previously healthy 72 year old woman
- Owns a dress shop in a small town
- Began having difficulty with managing the budget
- Husband became concerned
- Woman was seen by PCP
- Testing was performed and indicated cognitive decline.

Diagnosis?





Definitions



Encephalopathy

- Latin for:
 - Encephalo: the brain
 - Pathy: something is wrong
- It implies disturbed cognition and impaired alertness.
- Many types of encephalopathy.
 - Congenital or Acquired
 - Temporary or Permanent



Delirium

- Latin for:
 - to leave the furrow or track
- Reduced awareness or interaction with the environment.
- Four key features:
 - Acute, fluctuating course (hours to days)
 - Decreased attention
 - Disorganized thinking
 - Memory disturbance
 - Often restless and incoherent

Definition of Dementia

- Gradual deterioration of:
 - Memory
 - Mood
 - Concentration
 - Reasoning and Planning
 - Language
- Affects a person's ability to perform activities of daily living
- Lasting more than 6 months
- Without alteration of consciousness

Multiple types of Dementia


- Alzheimer's Disease
- Frontotemporal Dementia
- Vascular Dementia
- Dementia with Lewy Bodies
- Chronic traumatic encephalopathy
- Parkinson's Disease Dementia
- Creutzfeldt Jacob Disease
- Normal Pressure Hydrocephalus
- Medical causes of dementia

Medical causes of apparent dementia

- AIDS
- High fever
- Dehydration
- Hydrocephalus
- Systemic lupus erythematosus
- Lyme disease
- Long-term drug or alcohol abuse
- Depression
- Vitamin deficiencies
- Poor nutrition
- Hypothyroidism
- Hypercalcemia
- Multiple sclerosis
- Brain tumor(s)
- Side effects of prescription medications.

The background is a blue-toned abstract composition. On the left, a portion of a globe is visible, with a dark silhouette of a person standing on its surface. The globe is surrounded by glowing, wavy lines and a bright light source that creates a lens flare effect across the upper right portion of the image. The overall aesthetic is futuristic and professional.

Case presentation 3



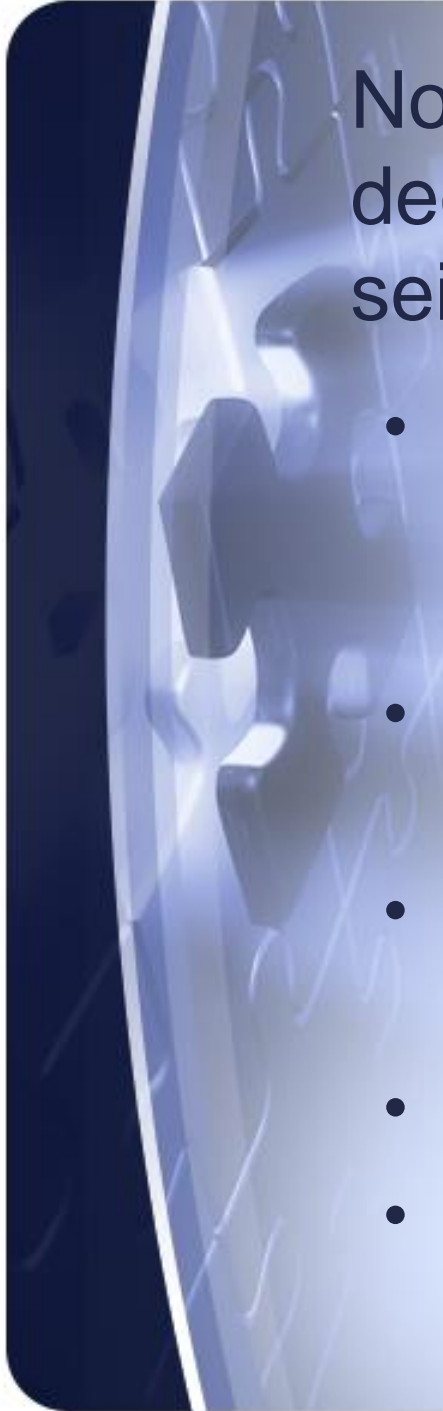
52 yr old woman with moderate cognitive impairment due to post-natal infection. Presented for follow up of seizure disorder.

- Averaging one recognizable seizure per month, but....
 - Visual hallucinations are increasing.
 - Episodes of becoming confused
 - Speech noted to be slurred
 - Does not recognize objects (example: threw away the dishes as she didn't know what they were for.)
 - Not exactly sure of the onset.

Past medical history and Medications:

- Intractable complex partial seizures
- Moderate mental retardation
- History of visual hallucinations
- Lamictal
- Tegretol
- Dilantin
- Primidone





Now 66 yr old woman with ongoing decline. Onset of rare myoclonic seizures.

- Speech is dysarthric, and she only speaks one or two words in response to questions.
- She did not know the name of her long term direct care staff member
- She did not know the name of her sister (guardian).
- No longer able to walk
- Exam otherwise unremarkable

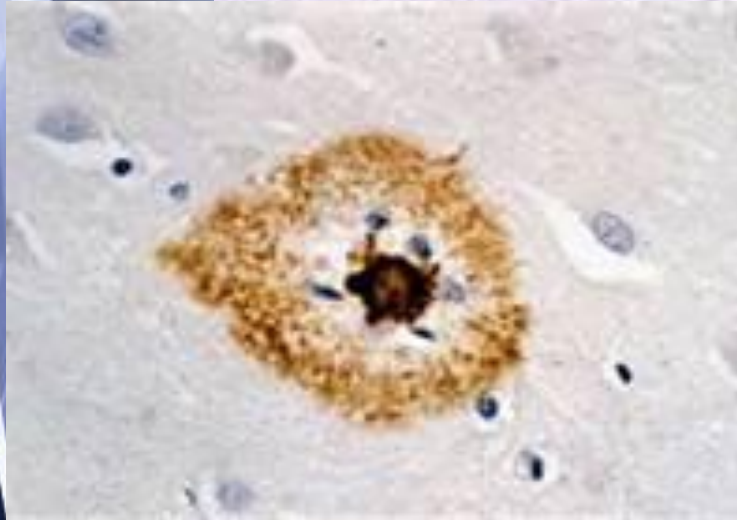
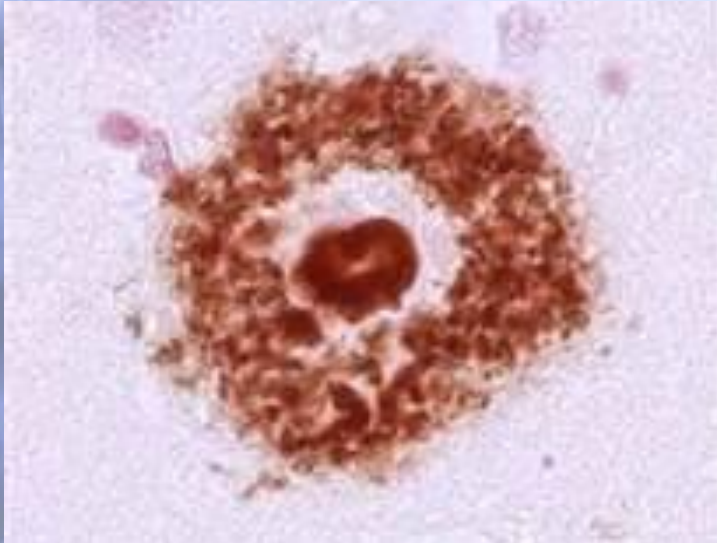


Alzheimer's Disease


Alzheimer's Disease - history

- Initially referred to as “presenile dementia”.
- 1901 – Alois Alzheimer, a German psychiatrist.
 - Mrs Auguste D – 51 yr old woman with short term memory loss
 - Died Apr. 1906.
 - Brain reviewed on autopsy with neurofibrillary tangles and amyloid plaques identified
 - Presented and Published, 1907

Amyloid plaques



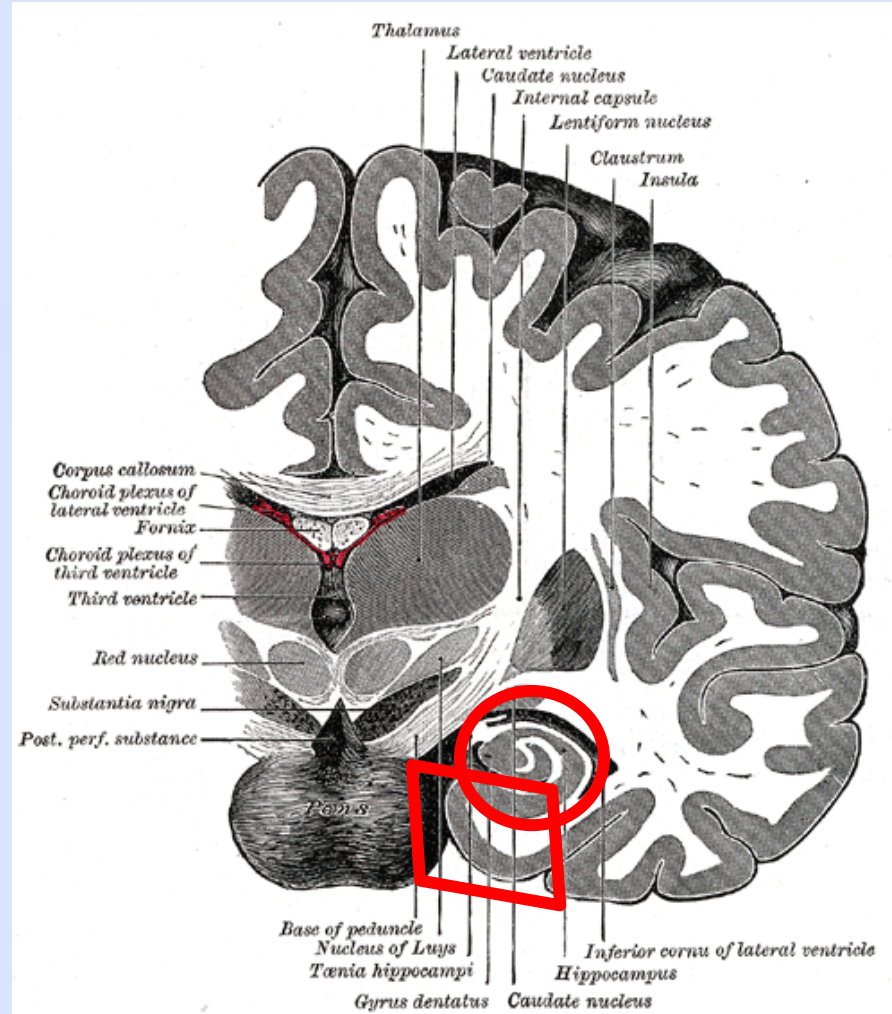
- Beta-Amyloid
 - normal protein byproduct.
 - Some are considered “toxic”
- Composed of:
 - beta-amyloid center
 - Surrounded by reactive glia
 - Degenerative neurites
- Function
 - uncertain
- Found in 76% of cognitively normal



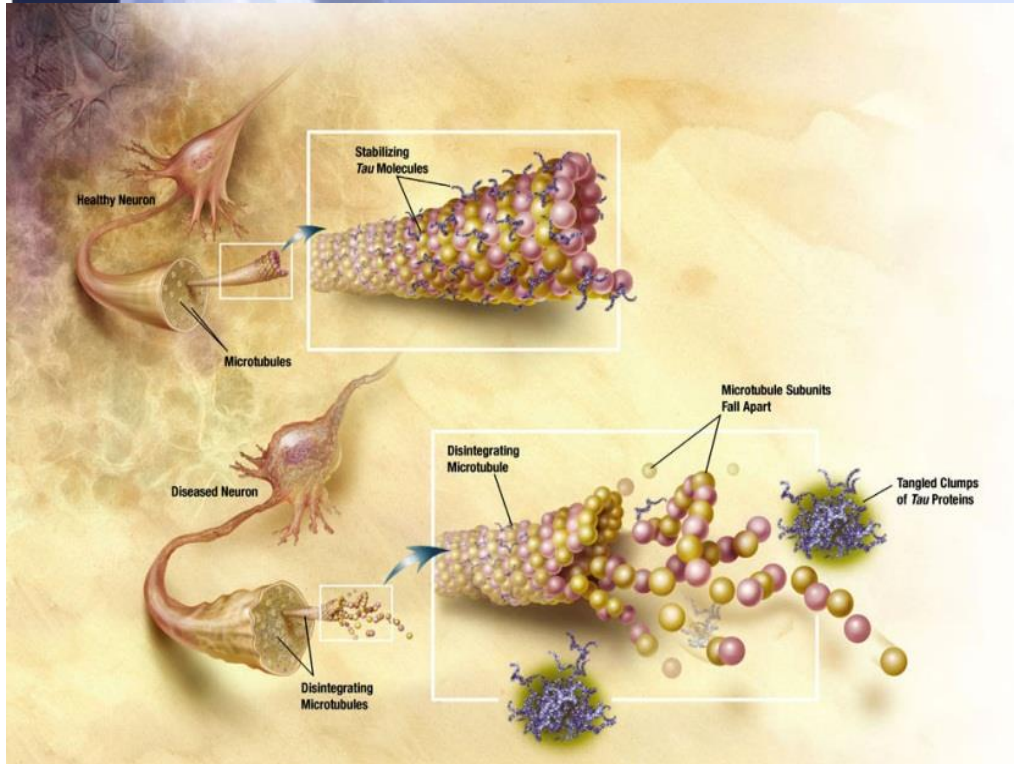
Pathophysiology of amyloid plaques

- Different types of Amyloid Plaques
 - Sporadic Alzheimer's dementia
 - Accumulation of toxic amyloid plaques
 - Familial Alzheimer's dementia
 - Decreased ability to clear amyloid plaques
 - Adversely affects the neurons ability to function

- Accumulation begins in the transentorhinal cortex, then spreads to the Hippocampi.
- Affects on tau proteins.



Pathology



- Associated with Chromosome 17.
- Tau proteins stabilize the microtubules
- Important for nutrient transport
- Phosphorylation of Tau protein is affected
- Tau protein undergoes a chemical change
- Microtubules breakdown
- Synapses breakdown
- Neurofibrillary tangles develop



Questions?

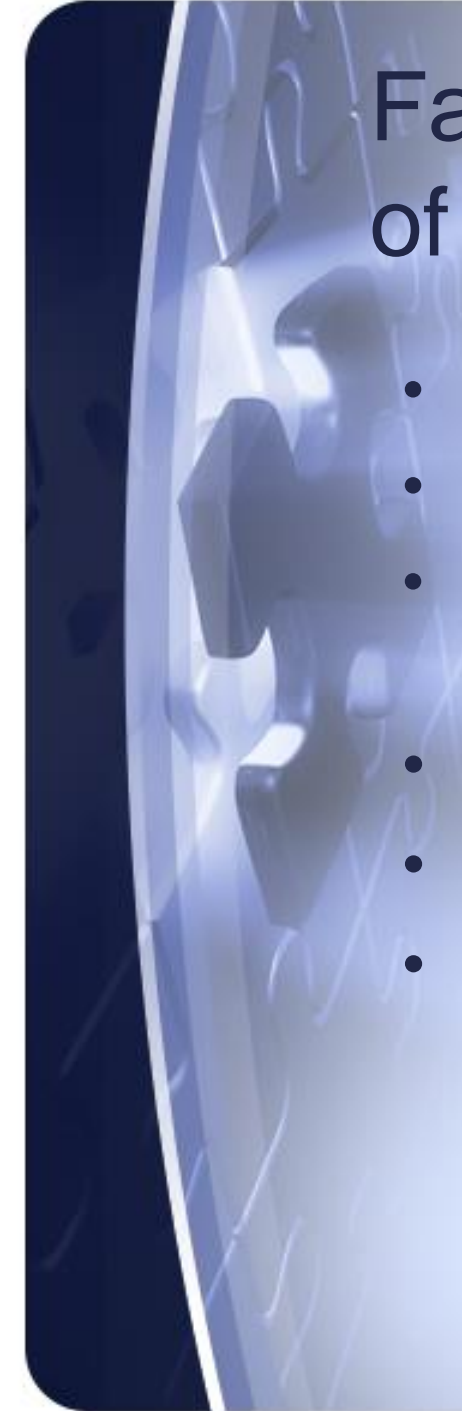
- What is the result of microtubules breaking down?
 - The cells don't get enough nutritional support and die.
- Do amyloid plaques cause damage?
 - The toxic ones can, and if they aren't clearing normally they can.
- Do neurofibrillary tangles cause damage?
 - No, they are a byproduct of damage.

Epidemiology

- 50 – 80% of all dementias
- Risk factors:
 - Age at 65 years is 2%, doubling every 5 years.
 - Apolipoprotein E (ApoE); 3 possible alleles
 - $\epsilon 2$ – rare
 - $\epsilon 3$ - most common
 - $\epsilon 4$ – associated with increased risk of AD
 - Homozygous $\epsilon 4$ confers >90% risk.

Genetics vs environment

- 25% clearly familial
- Remainder unknown
 - Combination of environment and genetics
 - Sporadic
- Length of survival
 - Variable
 - Median 5.9 years



Factors leading to increased rate of decline

- Early age of onset.
- Use of neuroleptic medications.
- APOE ϵ 4
 - Studies support and refute this as a factor.
- Male gender
- Medical comorbidities
- Level of functional abilities.

Symptoms

- Memory loss – especially of:
 - Recent events
 - Names
 - Placement of objects
 - New information
- Confusion of time and place
- Difficulty completing ADLs
- Poor judgment when making decisions

Symptoms

- Language difficulties:
 - Finding appropriate words
 - Completing sentences
 - Following directions
 - Following conversations
- Difficulty with complex mental assignments
 - Balancing a checkbook
 - Other tasks involving numbers

Changes in mood or personality

- Depression in ~50%
- Paranoid delusions including fears of:
 - personal harm,
 - Theft of property
 - Marital infidelity
 - “phantom boarder”
- Hallucinations
 - Primarily visual (20%)
- Agitation (70%)
 - Physical aggression
 - Verbal aggression
 - Motor restlessness w/ pacing

Cognitive impairment

- Executive function difficulties such as:
 - Understanding complex or multistep instructions
 - Solving problems
 - Reasoning
- Verbal blocking:
 - Losing your train of thought in the middle of a sentence
- Apathy and bradyphrenia – common
- Spatial or geographic disorientation:
 - Lost in the mall or your own home.
- Misidentification errors:
 - Failure to recognize people you know
- Fluctuations – a defining feature.

Diagnosis

- Laboratory studies – to exclude other causes
- MRI showing entorhinal and/or temporal lobe atrophy
- Spinal fluid
 - Increased $A\beta_{1-42}$
 - Decreased tau protein
 - Neither are specific, but together increase the odds.

Treatment

- Cholinesterase Inhibitors
 - Donepezil (Aricept)
 - Galantamine (Razadyne)
 - Rivastigmine (Exelon)
 - Tacrine (Cognex)
- NMDA receptor antagonist
 - Memantine (Namenda)
- Neuroprotective approaches
 - Selingiline
 - Vitamin E



Why cholinesterase inhibitors?


- Many of the cortical neurons involved early in Alzheimer's disease secrete acetylcholine as their neurotransmitter.
- Well-documented decrease in cholinergic activity.

Proposed treatments

- Non-steroidal Anti-Inflammatory drugs
 - Chronic use appears to reduce the risk
 - Once AD is established, not helpful
- Estrogen replacement therapy
 - Women's Health Initiative Memory Study – failed to find a positive impact.

The background is a deep blue gradient. On the left side, there is a stylized globe with white and light blue segments. Several thin, wavy white lines, resembling data or signal paths, curve across the globe and extend into the background. A bright, horizontal light streak or lens flare effect is visible in the upper right quadrant, creating a sense of depth and focus.

Case presentation 4



CC: 34 yr old female with Trisomy 21 presents with cognitive and behavioral changes:

- Decreased interest in things she previously enjoyed for 1 year.
- Previously “happy-go-lucky”.
- Enjoyed talking with family, now doesn’t want them to come into her room. Tells them to “go away”.
- Memory is unchanged.
- Speech is unchanged.
- Recent thyroid tests were normal.
- Recent diagnosis of right hip arthritis.
- Medications:
 - Zoloft,
 - Motrin
 - Vitamin C

Case 4

- 49 yr old previously independent in ADL.
- “2 years ago” began to require assistance with almost all ADLs.
- Medications:
 - Namenda
 - Exelon Patch
- Speech is now dysarthric, and she rarely speaks.
- She was able to tell me the name of her dog.
- She has developed myoclonic seizures.



Alzheimer's Disease and Down Syndrome

- People with Down Syndrome are at increased risk to develop Alzheimer's disease but:
 - The reason is unknown
 - Alzheimer's disease is diagnosed in:
 - 10 - 25% of people 40 – 49 years old
 - 20 – 50% of people 50 – 59 years old
 - 60 – 75% of people 60 years and older
 - Presenting symptoms differ from the general population.



Presenting symptoms

- Reduced interest in:
 - Social activities
 - Conversing
 - Expressing thoughts
- General decreased interest in participating
- Decreased ability to attend to environment
- Increased sadness, irritability, and anxiety




Presenting symptoms

- Restlessness or sleep disturbances
- Seizures beginning in adulthood
- Changes in gait or coordination
- Increased noisiness or excitability
- Memory loss is less noticeable

Pathophysiology

- Beta-Amyloid precursor protein is coded for on the proximal part of the long arm of chromosome 21.
- The triplicate chromosome 21 therefore, may produce excessive beta-amyloid precursor protein → increased potential for misfolding of proteins or perhaps decreased ability to clear the amyloid plaques → cellular destruction.



Alzheimer's Disease should be a diagnosis of exclusion for people with Down Syndrome

- Consider:
 - Depression
 - Hypothyroidism
 - Alantoaxial instability
 - Sleep apnea
 - Osteoarthritis
 - Sensory loss
 - Celiac disease



Treatment


- Same medications may be used, but they have not been studied for effectiveness

Questions:

- Does the presentation for Alzheimer's Disease differ in people with Down Syndrome compared with people without Down syndrome?
 - Yes, people with Down Syndrome are more likely to have a change in personality and behavior rather than memory problems early on.
- Is Alzheimer's disease “a given” in people with Down Syndrome?
 - No, even into their 60's and older ~25% will remain free of Alzheimer's Disease

The image features a blue-toned abstract graphic. On the left, a globe-like structure is composed of various 3D shapes, including a prominent dark blue cube and several white, translucent, rounded rectangular blocks. A bright, glowing light beam originates from the upper right and shines across the scene, creating a strong lens flare effect. The background is a gradient of blue, with a curved, metallic-looking band at the bottom. The overall aesthetic is modern and technological.

Case Presentation 5



61 year old woman with cognitive impairment due to prenatal injury

Presents with:

- New onset of hand tremor
- Intermittent confusion
- Past medical history:
 - Moderate cognitive impairment
 - Behavioral difficulties
 - Osteoporosis
 - Tardive dyskinesia
 - Hypothyroidism
 - Hypertension
 - Hypertriglyceridemia
 - History of seizure disorder, resolved
 - Thoracic kyphosis
 - Chronic renal failure



Review of daily activities

- No problem waking up in the AM
- Independent with toileting
- Requires verbal cues with showering
- Eats independently
- Requires verbal prompts to take dishes to the sink.
- Brushes her teeth independently
- Previously unloaded the dishwasher and put the dishes away, now requires verbal prompts
- Used to put her own clothes away in correct drawers, no longer does this.



Medications

- Zoloft
- Zyprexa
- Tegretol
- Atenolol
- Colace
- Oxybutinin
- Benztropine
- Prilosec
- Actonel
- Levothyroxine
- multivitamin
- Calcium supplement

Exam:

- General: Alert, cooperative, constant mouth movements consistent with tardive dyskinesia
- Speech was soft, but spoke in short phrases
- Decreased blinking
- Mild cogwheeling in the left wrist and elbow
- Gait: stride was decreased, and she needed to take more than one step to turn 180 degrees
- Reflexes: increased on the right
- I did not observe any tremoring

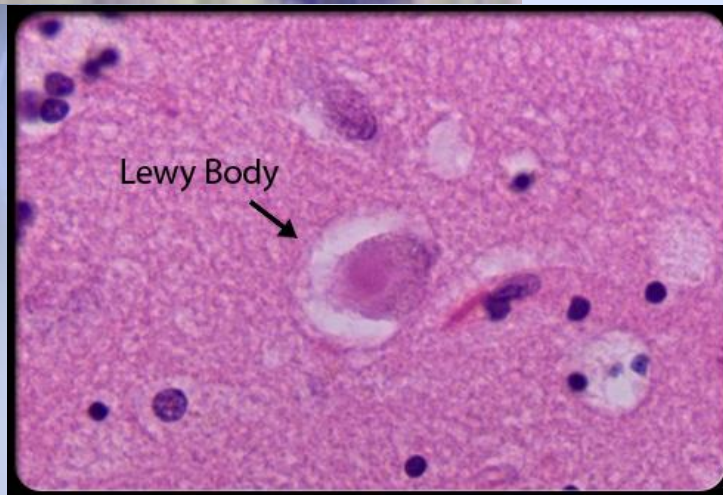
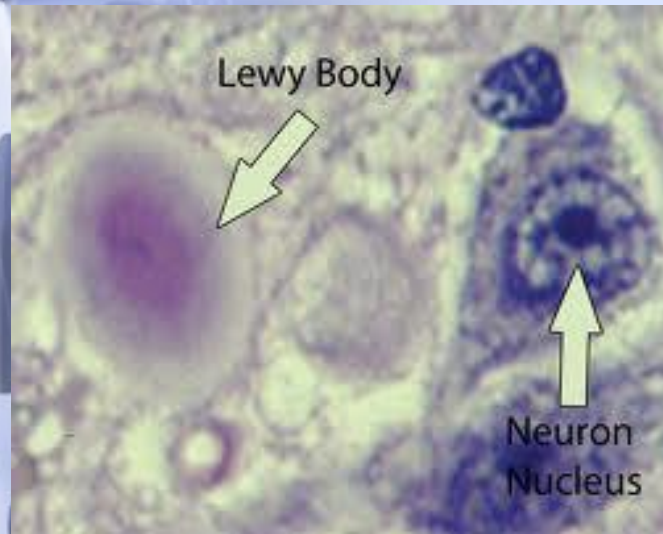
What is the diagnosis?



The image features a blue-toned abstract graphic. On the left side, there are several wavy, vertical lines that resemble stylized letters or symbols. In the center, there is a dark, three-dimensional, blocky figure that looks like a stylized letter 'H' or a similar character. The background is a gradient of blue, with a bright, glowing horizontal band across the middle. The overall effect is futuristic and digital.

Dementia with Lewy Bodies

Dementia with Lewy Bodies



- First described in 1961 by Okazaki and colleagues.
- Second most common form of dementia
- Frequency 15 - 20%



Clinical Features

- 5 domains affecting function:
 - Cognitive impairment
 - Neuropsychiatric features
 - Motor dysfunction
 - Sleep disorders
 - Autonomic dysfunction



Cognitive Impairment

- Gradual deterioration in memory.
- Fluctuating attention (a defining characteristic).
 - Prolonged runs of drowsiness
 - Prolonged times of staring into space
- Confusion

Neuropsychiatric features

- Visual hallucinations (often a first sign):
 - Vivid and well-formed
 - May be insects, animals, or people
 - Recognition of incorrect perception variable
- Visual illusions:
 - Objects are perceived incorrectly
- Delusions:
 - Usually paranoid quality
 - Belongings stolen
 - People are invading the home.
 - Capgras syndrome
- Depression – very common
- Anxiety - common

Motor Dysfunction

- Development of Parkinsonism (a defining characteristic)
 - Shuffling gait
 - Increased muscle tone with slowed movements
 - Imbalance
 - tremors
- Myoclonus – less common



Sleep disorders

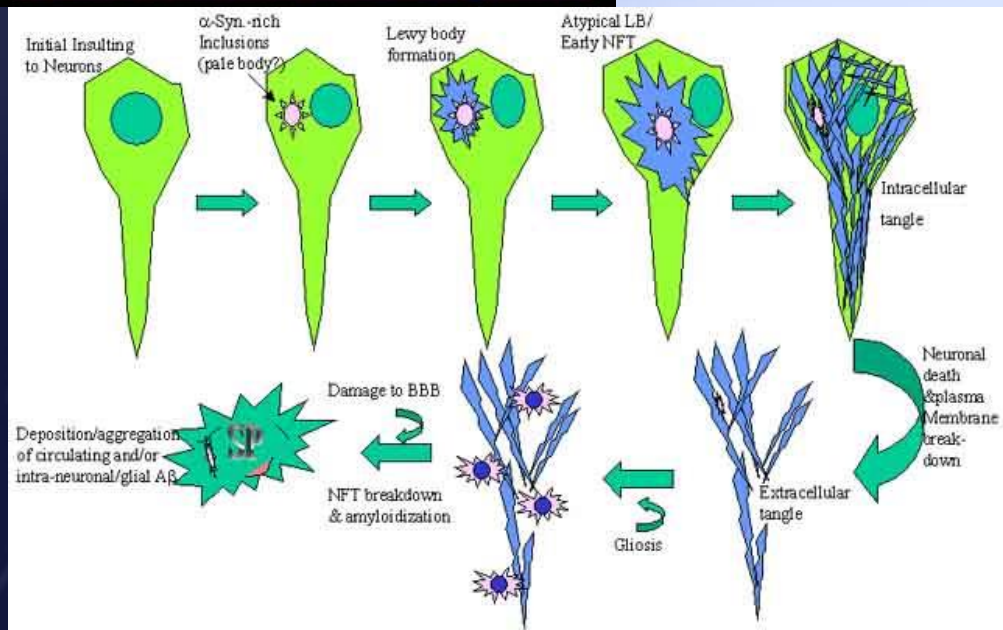
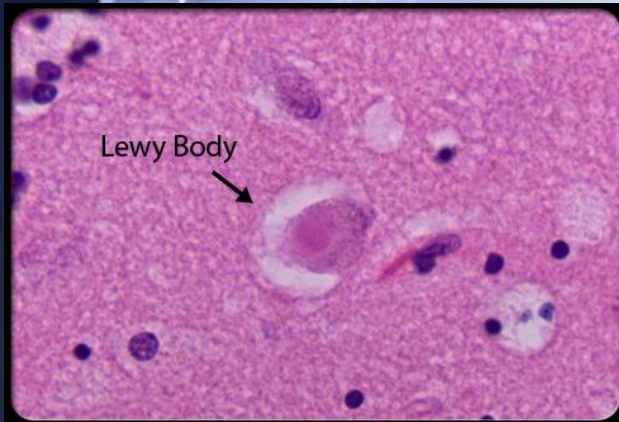
- REM Behavior Disorder
 - Patients act out their dreams
 - May begin years or decades before cognitive or motor symptoms develop.
- Daytime somnolence
- Other disorders:
 - Obstructive sleep apnea
 - Central sleep apnea
 - Restless leg syndrome
 - Periodic limb movements



Autonomic dysfunction

- Orthostatic hypotension
- Impotence
- Urinary incontinence
- Constipation

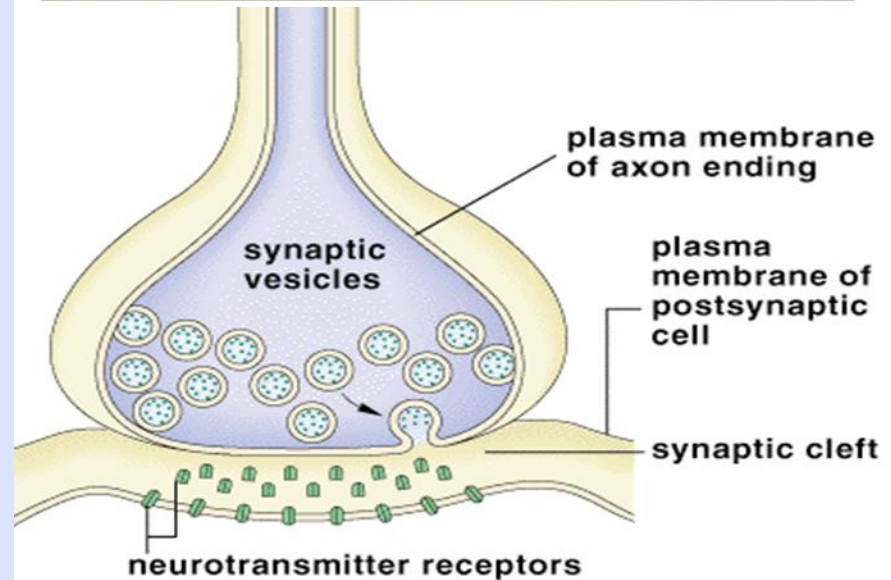
What are Lewy Bodies?



- Accumulations of misfolded proteins
- Composed of:
 - A-synuclein
 - Ubiquitin
- Leading to formation of neurofibrillary tangles

Pathology

- The problem is with α -synuclein
- Alpha-synuclein is important in the function of synaptic vesicles.





Pathology of Dementia with Lewy Bodies

- Forty percent of people have pathologic findings consistent with AD
- Significant debate exists whether Dementia with Lewy Bodies and Parkinson's disease with dementia.
 - Are they the same process with different presentations?
 - Are they separate disease states?



Risk Factors:

- Male gender
- Family member with diagnosis of Lewy Body Dementia
- 60 years of age or older

Longevity

- Average 8 years

Diagnostic testing

- Blood tests – none
- Urine tests – none
- Spinal fluid tests – none
- EEG – non-specific
- Neuroimaging – less hippocampal atrophy compared with AD or vascular dementia
- Functional neuroimaging – not helpful
- Polysomnography
 - loss of normal REM atonia

Treatments

- Cholinesterase inhibitors are helpful
- Carbidopa/levodopa and dopamine agonists
- Clonazepam may help REM Behavior disorder

Treatments:

- Neuroleptics should be avoided due to sudden onset severe irreversible parkinsonism.
- Serotonin Reuptake inhibitors
 - Fluoxetine may worsen insomnia
- Conservative Treatments
 - Modify Communication
 - Modify the environment
 - Encourage physical activity

Conservative treatments





Communication

- Look at the person when you speak to them.
- Touch their shoulder to maintain attention
- Speak slowly and use gestures or point
- Don't rush them
- Don't constantly correct them
- Validate their concerns
- Reassure them
- Break down tasks into individual steps
- Focus on success
- Structure and routine helps them to feel secure



Environmental modifications

- Reduce Clutter
- Reduce extraneous noises
- As behavior is worse at night
 - Establish bedtime rituals
 - Keep environment quiet at night
 - Use night lights
 - Limit caffeine late in the day
 - Discourage daytime naps

A person is walking on a path that is part of a larger, blue-tinted image. The path is made of light-colored, textured blocks, and the person is walking away from the viewer. The background is a gradient of blue, with some faint, abstract shapes. The overall image has a soft, ethereal quality.

Encourage physical activity

- Routine exercise that is not excessive
 - Improves symptoms of depression
 - Helps retain motor skills, flexibility, and balance
 - Decreases risk for falls
 - Helps with sleeping at night
 - Provides distraction for taking daytime naps



Questions:

- What are the three main clinical findings suggesting a dementia is Dementia with Lewy Bodies?
 - Fluctuating course
 - Early onset of visual hallucinations
 - Parkinsonism
- Which class of medications should be avoided?
 - Neuroleptics as it can lead to severe irreversible Parkinsonism



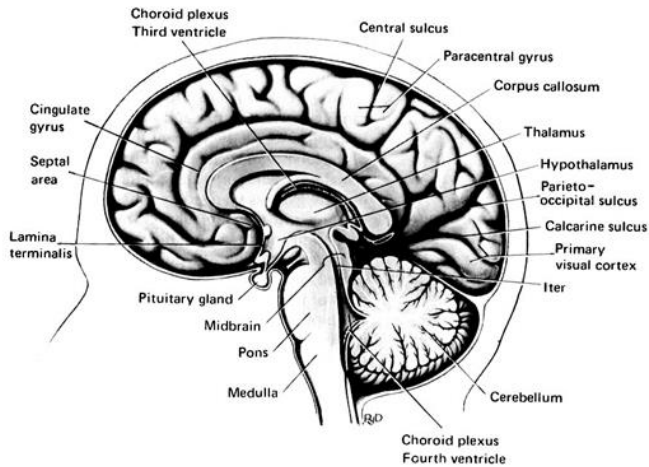
End Stage Parkinson's Disease



Parkinson's disease

- Very common ~ 1% of people > 60 years
- May be seen rarely in people < 40 years
- Men slightly more common than women
- Known genetic cause in ~10%
- Believed to be due to combination of genetic and environmental factors

Pathophysiology



- Loss of dopaminergic neurons in the substantia nigra
- Development of Lewy bodies
- Destruction of the neurons



Clinical presentation

- “Pill rolling tremor” at rest
- Decreased dexterity
- Decreased arm swing when walking
- Decrease in volume of speech
- Decreased facial expression
- Decreased blink response
- Sleep disturbances
- Autonomic symptoms
 - Constipation
 - Sexual dysfunction
 - Sweating abnormalities



prognosis

- Neurodegenerative disorder
- Cognitive decline
- Sleep disturbances become more prominent
- Visual hallucinations develop
- Longevity is extremely variable
 - Age
 - Onset of rigidity vs tremor

Treatment


- Replacement of dopamine with levodopa/carbidopa
- Monoamine oxidase inhibitors B
- Dopamine agonists
- Symptomatic treatment of
 - Sleep disorders
 - Dementia
 - Behavior disturbances

Questions:

- What are the 3 primary clinical characteristics that define Parkinson's disease?
 - Pill rolling tremor at rest
 - Rigidity or increased muscle tone, but not spasticity
 - Bradykinesia - they move slowly
- What is the primary distinction between Dementia with Lewy Bodies and Parkinson's Disease Dementia?
 - The dementia precedes the movement disorder in Dementia with Lewy Bodies
 - The movement disorder precedes the dementia in Parkinson's Disease Dementia.



Case Presentation 6



70 yr old woman presents with her son, who complained of decline in memory over the past 10 months

- Increased suspicion of family members
- Begun hoarding things
- Can't remember ingredients while cooking
- Has lost interest in activities previously enjoyed.
- Needs reminders to take her medication.

Past medical history

- Hypertension
- Diabetes
- Coronary artery disease
- Osteoarthritis
- Osteoporosis



An abstract digital background with a blue color palette. On the left, a stylized globe is visible, composed of white and blue segments. A bright blue beam of light originates from the top right and illuminates the globe. The background is filled with faint, glowing blue lines and patterns, suggesting data or neural connections. The overall aesthetic is futuristic and technological.

Vascular Dementia

Vascular Dementia

- One of the most common forms of dementia
- Frequency, dependent on cultural background.
- The range is 4% to 30%
- Incidence – male:female ratio 1:1
- This is really a cluster of syndromes
- Ultimately preventable.

Epidemiology

- Japan – 50% of all dementias.
- Western European – 25% of all dementias
- Latin American – 15%
- Australia – 13%

- Mortality Rate
 - Higher compared to Alzheimer's Disease due to concomitant problems with vascular disease.

History

- 1899 – Presenile Dementia and dementia related to atherosclerosis were two different entities.
- 1969 – Mayer-Gross determined that ~50% of dementia patients had hypertension as the underlying cause.
- 1974 – Hachinski et al labeled it as multi-infarct dementia
- 1985 – Loeb used the term vascular dementia.

Pathophysiology

- Vascular disease produces either a focal or diffuse effects on the brain.
- Common areas of the brain prone to causing cognitive decline include:
 - The deep white matter
 - The basal ganglia



Risk Factors:

- Hypertension – found in 50%
- Diabetes
- Tobacco use:
 - ≥ 2 packs per day for 40 years confers $>100\%$ risk
- Older age
- Lower educational level
- Family history of dementia
- Left sided strokes
- Large or multiple strokes

Types of Vascular Dementia

- Multi-infarct Dementia:
 - Multiple small infarcts have a combined effect to cause dementia
- Single infarctions can cause dementia:
 - Anterior Cerebral Arteries – affecting the frontal lobes
 - Distal Middle Cerebral Artery – affecting the parietal lobes
 - Small perforating vessels – affecting the thalamus
 - Infarction of the cingulate gyrus.

Types of Vascular Dementia

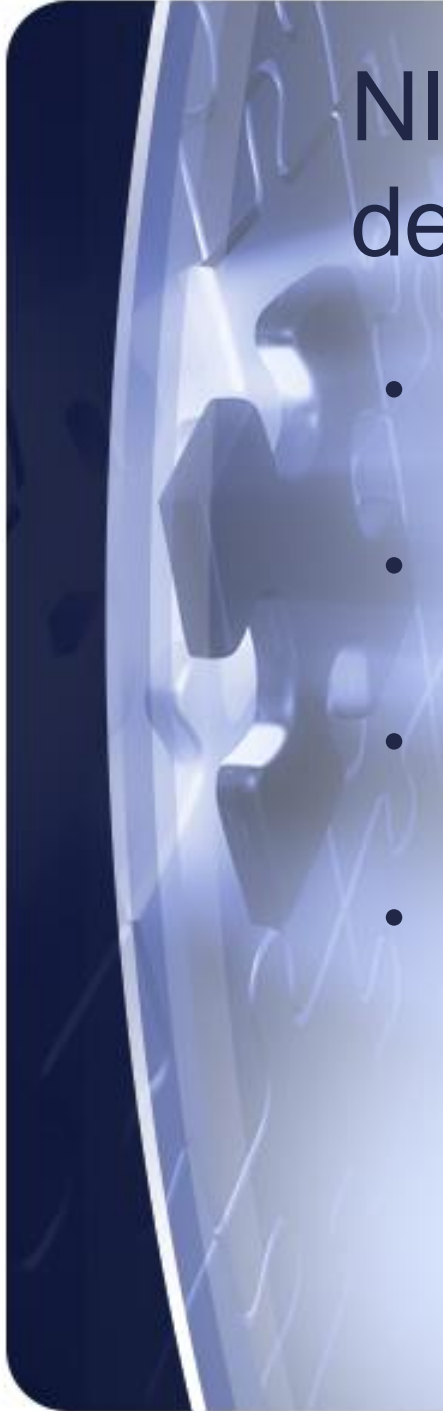
- Small Vessel Disease leading to:
 - Lacunar Infarctions:
 - Small vessel occlusions producing multiple small cavitory lesions.
 - The multitude affects the neural circuits and efficacy of the brain.
 - Binswanger's Disease (aka subcortical leukoencephalopathy):
 - Fibrinhylinolysis affects the small cortical vessels
 - Fibrinoid necrosis affects the larger vessels
 - Leading to diffuse impairment from poor circulation

Types of Vascular Dementia

- Cerebral Amyloid Angiopathy:
 - Amyloid builds up in the walls of the small cortical vessels
 - Amyloid causes stenosis and sometimes aneurysms
 - Causes a diffuse damage to the subcortical white matter → dementia
 - Patients often present with cerebral hemorrhages prior to 40 years of age.
- Others which are Very Rare.

Definitions of Vascular Dementia

- **DSM IV Criteria:**
 - Clinical evidence of stroke based on signs from neurologic exam or imaging.
 - No requirement for the dementia to have a temporal link to the stroke
- **California Based Consortium of Dementia**
 - Cognitive decline
 - Evidence of 2 strokes in different vascular territories either clinically or on imaging.



NINDS-AIREN criteria for vascular dementia

- Decline in cognitive function in 2 or more domains from a previous level.
- Evidence of cerebrovascular disease on examination.
- Evidence of cerebrovascular disease on imaging
- A relationship between the dementia and cerebrovascular disease.
 - Onset of dementia either abruptly or within 3 months of a recognized stroke
 - Stepwise progression of cognitive deficits.

Hachininski Ischemic Scale

- Abrupt onset
- Stepwise deterioration
- Somatic complaints with neurologic symptoms and signs.
- Emotional incontinence
- History of hypertension and/or stroke



3 basic criteria in common

- Presence of a stroke temporally related within 3 months.
- Presence of bilateral gray matter infarcts in the frontal, temporal or parietal cortices, basal ganglia or thalamus
- Symptoms or evidence on physical exam, consistent with prior strokes.



Important distinction

- No matter how dramatic the MRI findings, the clinical assessment of function and cognition is what determines dementia.
- Twenty percent of older adults have had at least one lacunar (clinically silent) infarction, but this increases the risk for developing dementia.



Clinical Manifestations

- Executive dysfunction is a more common presenting symptom than memory loss.
- Depression is more common and more severe in people with vascular dementia than in those with Alzheimer's Disease.
- Many people with vascular dementia will develop psychosis, delusions, hallucinations, or paranoia, and they can become violent.



Work up

- Imaging study of the brain
- Laboratory studies:
 - Including Serum Protein Electrophoresis
- Carotid Doppler Studies
- Echocardiography
- Holter Monitoring
- Cerebral Angiography



Treatment

- Aspirin or other antiplatelet agent
- Treatment of hypertension
- Treatment of hyperlipidemia
- Treatment of diabetes
- Smoking Cessation
- Cholinesterase inhibitors have not been shown to be beneficial.
- Anti-depressants
- Anti-psychotics

Summary

- The presence of clinical strokes temporally related to dementia onset or worsening or two infarcts on imaging are the two cardinal features necessary to consider the diagnosis of vascular dementia.

Questions


- What are the 3 biggest risk factors for vascular dementia that can and should be treated?
 - Hypertension, diabetes, tobacco use, or hyperlipidemia
- What is the more likely presenting feature of vascular dementia?
 - Loss of executive function rather than memory loss.

A blue-toned 3D graphic. On the left, a large, dark blue number '4' is prominent. To its left, there are several wavy, metallic-looking lines. A bright, glowing light beam originates from the top right and shines across the scene. The background is a gradient of blue, with a curved, metallic-looking surface at the bottom. The overall aesthetic is futuristic and digital.

BREAKTIME!!!!

The image features a blue-toned abstract graphic. On the left, a stylized globe is visible with a dark silhouette of a person standing on it. A bright beam of light emanates from the person, extending across the upper right portion of the frame. The background consists of horizontal blue bands and wavy lines, creating a sense of depth and movement. The overall aesthetic is modern and professional.

Case Presentation 7



62 yr old man, successful graphic designer began having a significant personality change:

- Trouble finding names for people and objects
- Trouble filling out order forms
- Began to make frequent spelling mistakes
- He was caught stealing a shiny necklace from a client's store
- Soon afterward he began to comb the beach for seashells, spending hours there.
- At home he played solitaire compulsively for up to 6 hours per day.
- What does this man have?

The background is a deep blue gradient. On the left side, there is a stylized globe with white and light blue segments. Several thin, wavy white lines, resembling data or neural connections, are scattered across the scene. A bright, horizontal light streak or lens flare effect runs across the upper right portion of the image. The overall aesthetic is futuristic and scientific.

Frontotemporal Dementias

Frontotemporal Dementia



- Dementia syndromes associated with circumscribed regions of atrophy.
- First one described was Pick's Disease
 - Arnold Pick, MD, 1892
 - Patient with severe atrophy localized to the frontal lobes.
- Frequency – 1 – 7%

4 primary recognized syndromes

- Frontotemporal dementia
- Non-fluent progressive aphasia
- Semantic dementia
- Logopenic Progressive Aphasia (aka Prosopagnosia)

- Fluent progressive aphasias?
 - Several known
 - Not included in the frontal temporal dementias because:
 - Other areas of the brain involved
 - Motor deficits associated.

Pathophysiology

- This group of disorders is strongly tied to:
 - Genetic mutations on Chromosome 17
 - Occasionally mutations on Chromosome 9
- Finding of Pick Bodies on autopsy
 - Collection of Tau Protein fibrils
 - Involves Microtubule Associated Protein Tau (MAPT)
 - Considered 'Tauopathies'

Tau Protein

- Six isoforms
 - 3 are “3 repeat (3R) forms” because they have 3 repeated sequences that have microtubule binding sites.
 - 3 are “4 repeat (4R) forms” because they have 4 repeated sequences that have microtubule binding sites.
- These binding sites become impaired → microtubular degeneration → cell death
- Why they are focal is unknown

Tauopathies

- Alzheimers disease
- Amyotrophic lateral sclerosis
- Argyrophilic grain disease
- Autosomal-recessive juvenile parkinsonism
- Corticobasal degeneration
- dementia pugilistica
- Diffuse neurofibrillary tangles with calcification
- Down's syndrome
- FTD and parkinsonsim linked to chromosome 17
- Gerstmann-Straussler-Scheinker disease
- Hallervourden-Spatz disease
- Myotonic dystrophy
- Niemann-Pick type C
- Picks disease
- Postencephalitic parkinson's disease
- Progressive supranuclear palsy
- Subacute sclerosing panencephalitis
- Tangle-only dementia

Epidemiology and Mortality

- Male:female are 2:1
- Age of onset is often < 65 years old
- 40 – 50% have a first degree relative with FTD
- Mortality
 - Variable depending on which type
 - People may go up to 12 years with progressive aphasia, and no signs of dementia
 - Others may have profound dementia in a few years



Core criteria for the frontotemporal dementias

- Insidious onset
- Gradual progression
- Focal atrophy on imaging studies:
 - Frontal Temporal Dementia – both frontal lobes
 - Progressive nonfluent aphasia – the dominant hemisphere (usually left)
 - Semantic Dementia – left anterior temporal lobe
 - Logopenic Progressive aphasia – right anterior temporal lobe

Frontal Lobe Dementia

(Formerly Pick's Disease)

- Early findings:
 - Emotional blunting
 - Very disinhibited
 - Loss of insight of how their behavior effects others.
 - Or may become apathetic
- Altered speech output
 - ↓ speech output
 - Echolalia
 - Perseveration
 - Mutism
- ↓ in personal hygiene
- Hyperorality
- Sexually inappropriate
- Utilization behavior
- Sociopathic personality changes
- Dietary changes
- Perseverative and stereotyped behavior

Progressive Nonfluent Aphasia

- Nonfluent spontaneous speech which precedes cognitive decline by at least 2 years
- Preservation of social skills
- Late behavioral changes similar to FTD.
- Word finding difficulties
- Abnormal speech patterns
- Stuttering
- Oral apraxia
- Anomia
- Impaired repetition
 - ***later***
- ↓ comprehension
- Alexia
- Agraphia
- Impaired spelling

Semantic dementia

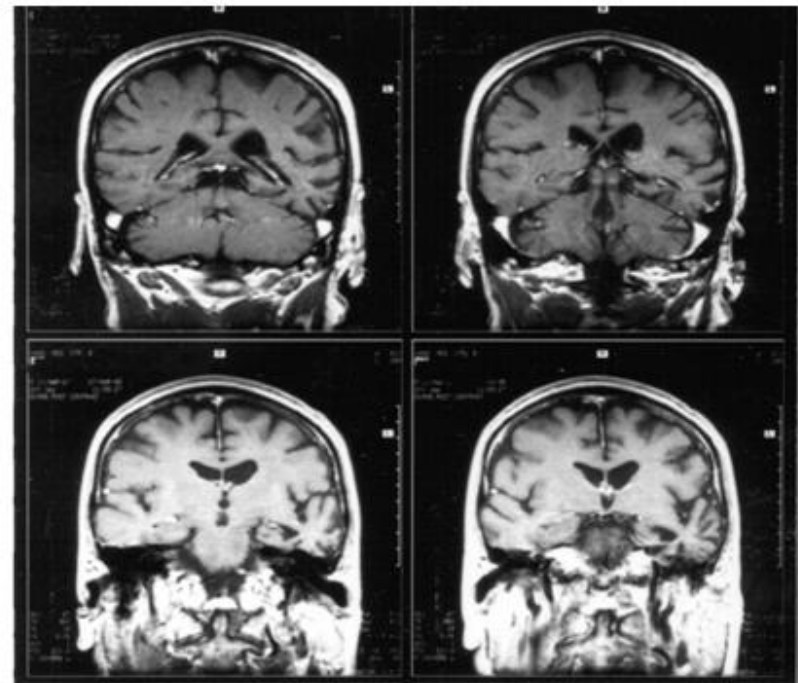
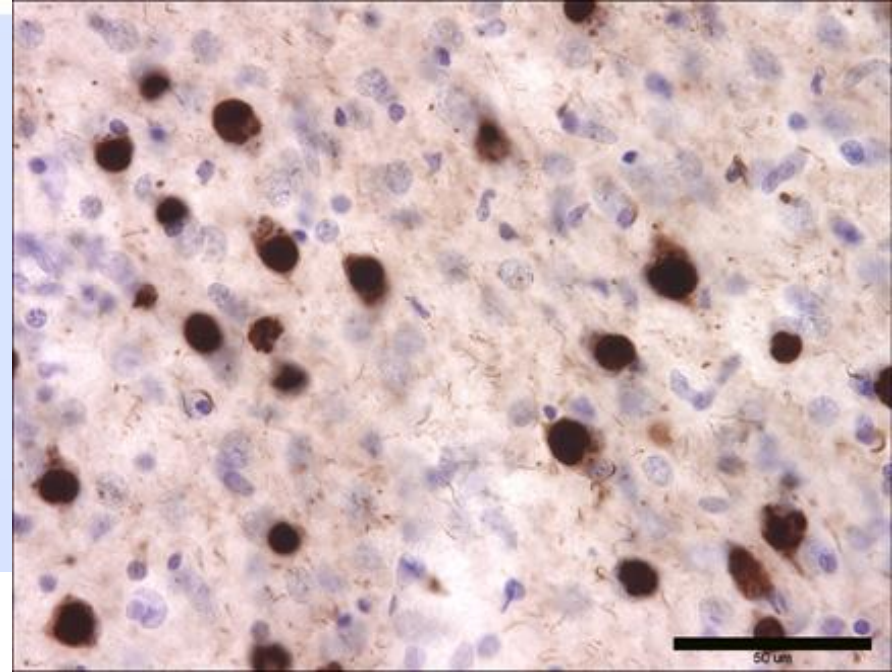
- Progressive fluent aphasia
- Press of speech
- Loss of word meaning
 - Impaired naming
 - Impaired comprehension
 - Semantic paraphasias
- Empty spontaneous speech
- Idiosyncratic word usage
- Surface Dyslexia
- Dysgraphia

Logopenic progressive aphasia

- Impaired Naming
- Impaired single word retrieval
- Impaired repetition
- Speech sound errors
- Spared motor speech
- Spared comprehension
- As the disorder progresses:
- Perceptual disorder:
 - Prosopagnosia (impaired recognition of familiar faces)
 - Associative agnosia (impaired recognition of object identity)

Work up

- EEG
 - Focal slowing over one or both frontal and/or temporal lobes
- MRI of the brain
 - Focal atrophy
- Blood tests
 - None specific



A person wearing a white protective suit and a mask is walking through a tunnel. The tunnel has blue lighting and a grid-like pattern on the walls. The person is walking away from the camera.

Therapy

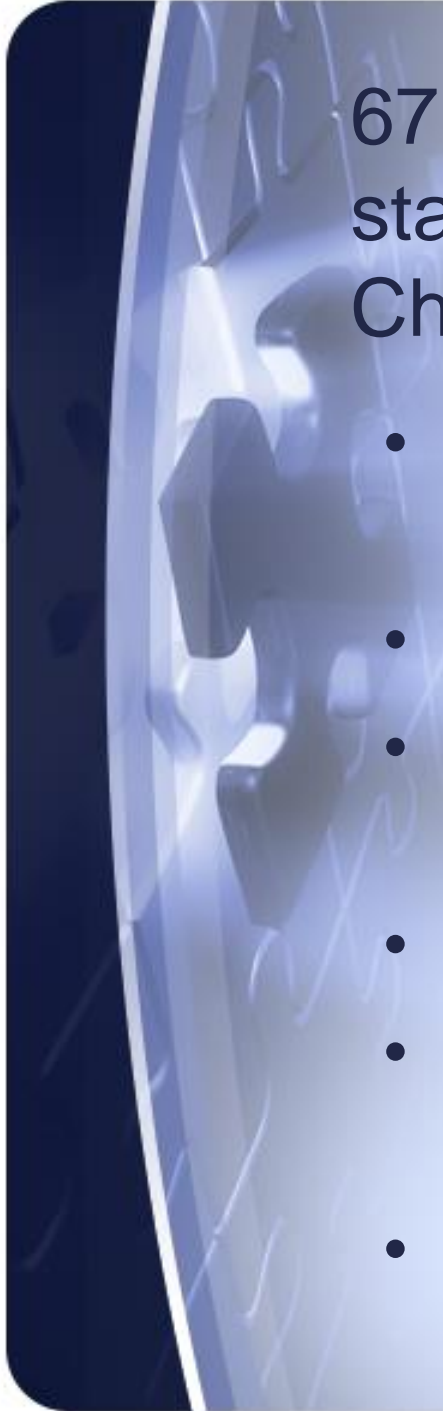
- No specific therapies
- Cholinesterase inhibitors may sometimes help, but are not proven.
- Vitamin E has not been helpful.
- SSRIs may be helpful for treatment of challenging behaviors.
- Dopamine may help with motor impairments

Questions

- The frontal temporal dementias begin with memory loss in all cases?
 - No, usually the presenting symptom is development of speech aphasia
- What is pathologically the biggest difference in the Fronto-Temporal Dementias and other forms of dementia?
 - Focal atrophy on imaging.

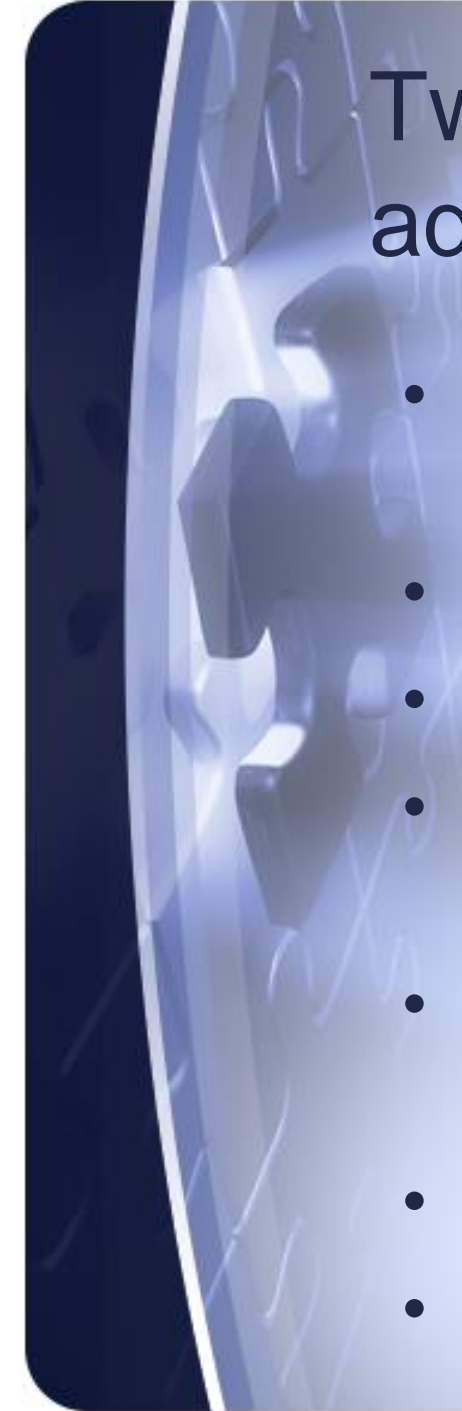


Case Presentation 8



67 yr old woman, who was in her usual state of health during the recent Christmas holiday.

- In January, she noted difficulty managing her checkbook.
- Inability to plan activities
- She couldn't seem to complete household chores
- Had difficulty with ADLs
- Noted an onset of occasional right arm, hand, and leg jerking.
- Intermittent visual blurring



Two months later she was admitted to the local hospital:

- She was having visual and auditory hallucinations
- She could no longer walk
- She was blind
- She was having occasional episodes of rigidity, nystagmus, and seizure activity
- She vacillated between periods of lucidity and profound dementia.
- She died 1 month later.
- What does she have?



Creutzfeld-Jakob Disease

Creutzfeldt-Jakob Disease

- Three main forms:
 - Spontaneous (~80%)
 - Genetic (~ 15%)
 - CJD Variant (~5%)
 - Acquired from bovine spongiform encephalopathy
 - What is bovine spongiform encephalopathy?



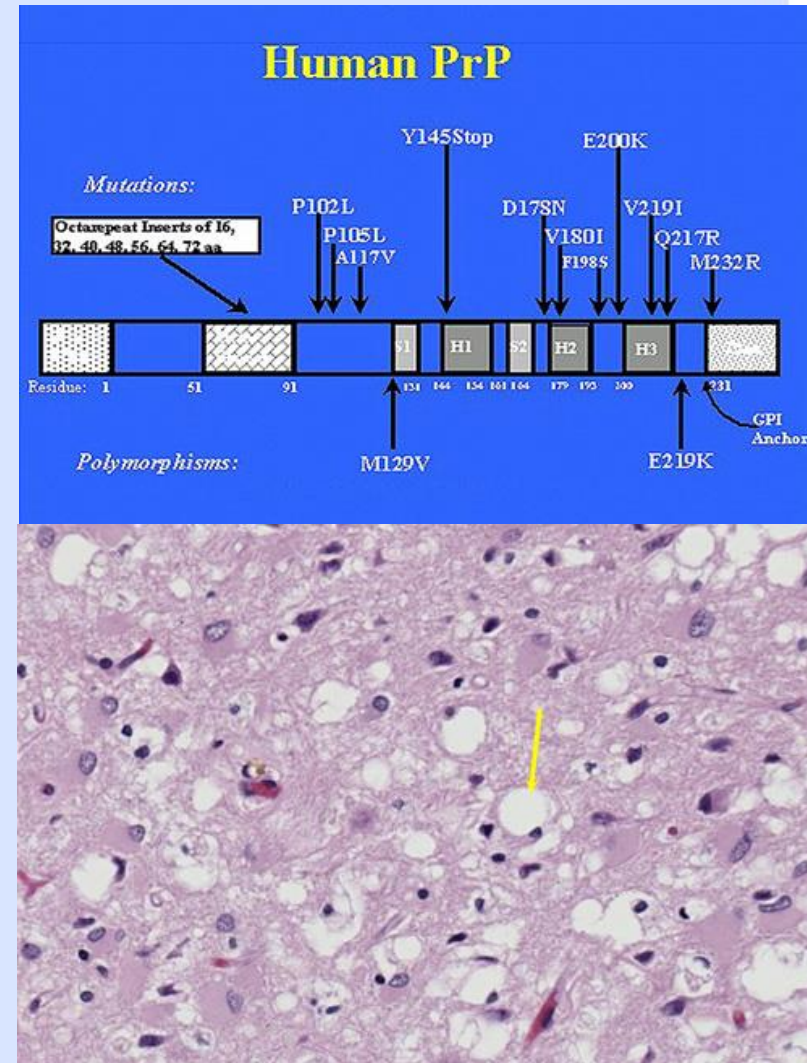
History



- Described in 1920 by:
 - Hans Gerhard Creutzfeldt, MD and Alfons Maria Jakob, MD
- Incidence
 - 1.5/million
- Risk factors
 - Age > 50 years
 - Variable for vCJD

Pathology

- Chromosome 20
- Prion-related protein is naturally occurring
- Change in conformation → amplification → cell destruction.
- May be inherited or transmitted through infected tissue or by ingestion.



Other prion diseases

- Bovine spongiform encephalopathy
- Gerstmann-Straussler-Scheinker
- Kuru
- Familial Fatal Insomnia
- Feline spongiform encephalopathy
- Scrappie

Clinical findings

World Health Organization Criteria for the Diagnosis of Sporadic CJD

- I Rapidly progressive dementia

- II A - Myoclonus
 B - Visual or cerebellar signs
 C - Pyramidal or extrapyramidal features
 D - Akinetic mutism

- III A – EEG show periodic wave complexes
 B – Positive 14-3-3 analysis of CSF

Possible CJD: I and 2 of II with a maximal duration of 2 years

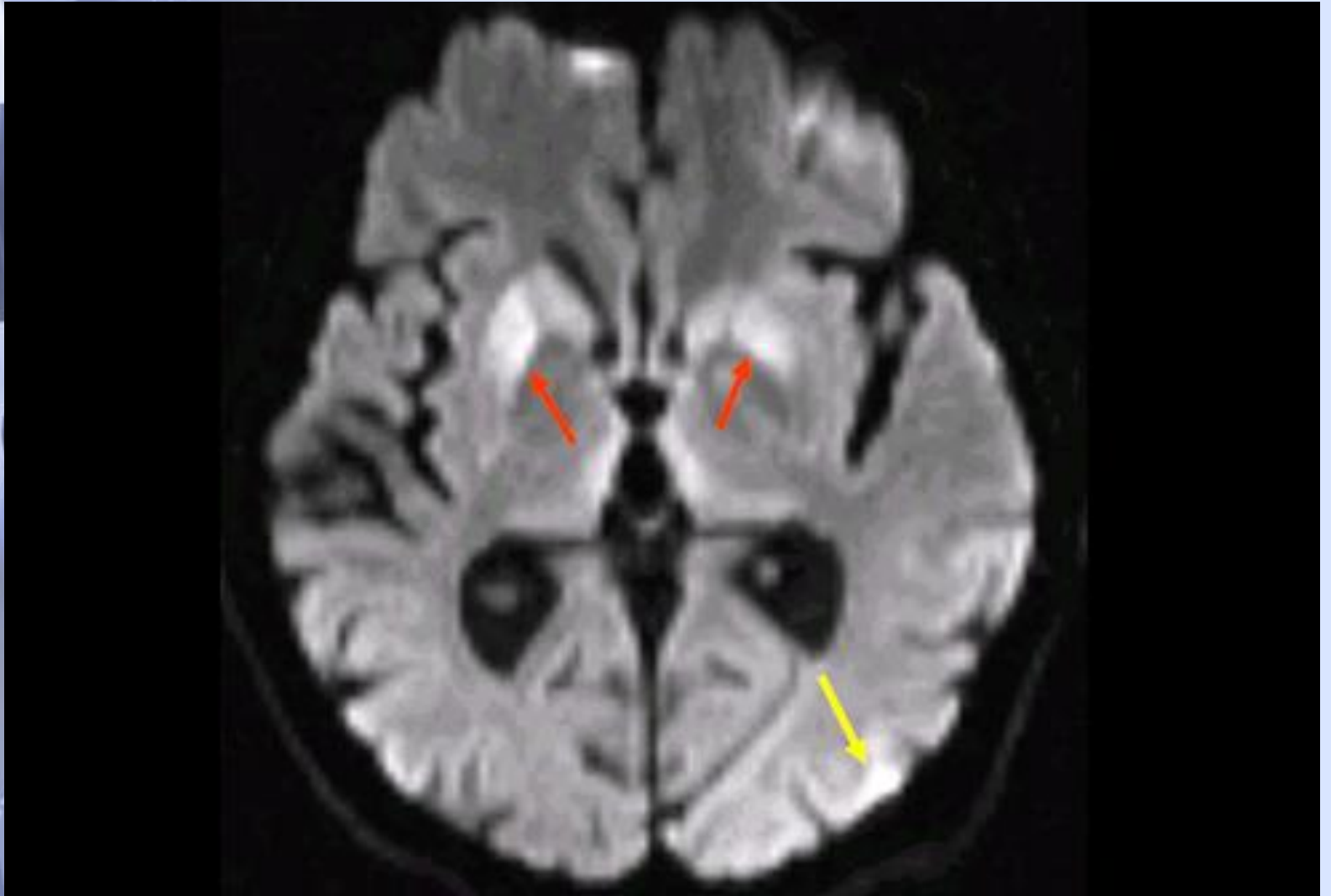
Probable CJD: I and 2 of II and III A and/or III B

Definite CJD: Neuropathologically confirmed case of CJD

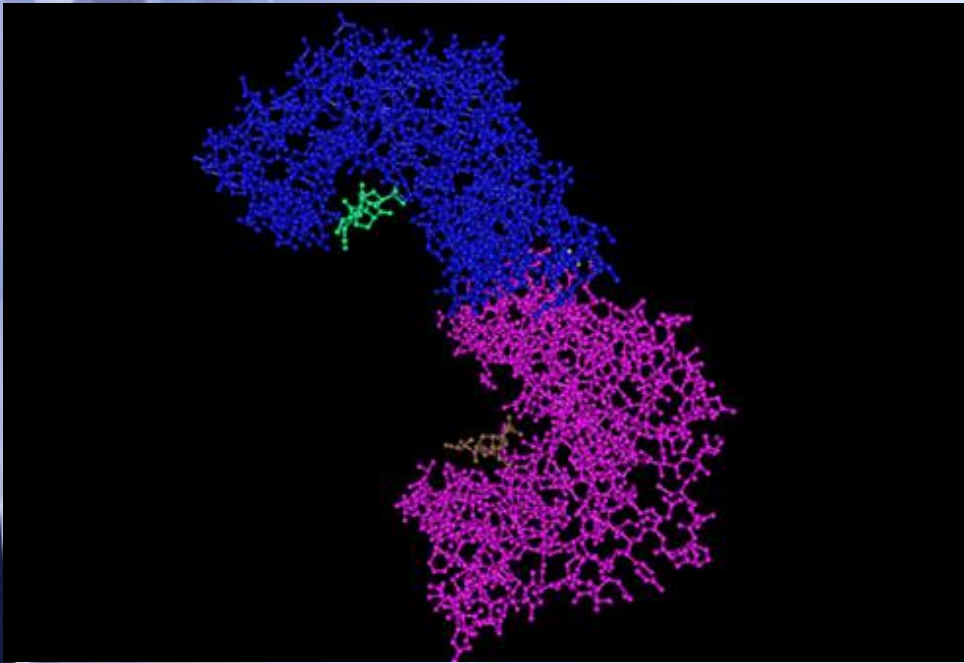
EEG – periodic pattern seen in 2/3



Brain MRI



Diagnostic studies



- CSF
 - 14-3-3 protein
 - >35 ng/ml
 - Neuron specific enolase
 - > 1200 pg/ml
- Individually not sensitive or specific
- Together increase likelihood

Treatment and Mortality


- No known treatments
- Duration:
 - 8 months for classic CJD
 - 15 months for variant CJD

Questions:

- Creutzfeldt-Jakob Disease is categorized as what type of disorder?
 - Prior disease or spongiform encephalopathy
- What are the three main diagnostic criteria?
 - Rapidly progressive dementia
 - Myoclonus
 - Periodic pattern on EEG.

The background is a deep blue gradient. On the left side, there is a stylized globe with white and light blue segments. Several thin, wavy white lines, resembling data or signal paths, curve across the globe and extend into the background. A bright, horizontal light streak or lens flare effect is visible in the upper right quadrant, creating a sense of depth and movement.

Case presentation 9



65 yr old man with hypertension presents with:

- Difficulty walking for 1 year.
 - Hesitancy in initiating steps
 - Difficulty turning corners
 - Would freeze at times while walking
- Forgetfulness for 6 months
 - Difficulty with word finding
 - Difficulty with money handling
 - Forgetting recent events
 - Names of distant relatives
- Urinary urgency for 5 months

Exam:

- Mental status exam showed mild impairment
- Slow reaction time
- Muscle strength and tone were normal.
- Gait: wide-based, steps were slow, shuffling, as if his feet were glued to the floor.



An abstract graphic with a blue and white color scheme. It features several wavy, vertical lines on the left side, resembling stylized waves or data points. In the center, there is a dark, three-dimensional, blocky shape that looks like a stylized letter 'H' or a similar geometric form. The background is a gradient of blue, with a bright, glowing area on the right side that fades into a darker blue towards the bottom. The overall composition is modern and technical.

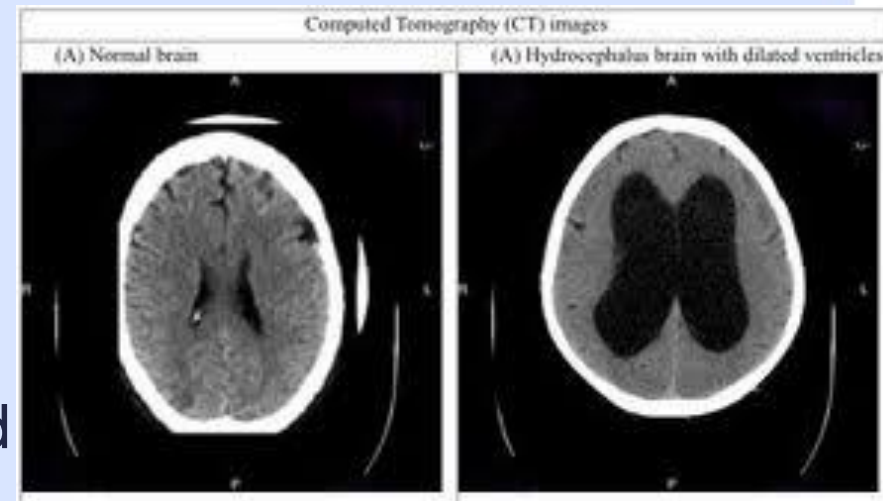
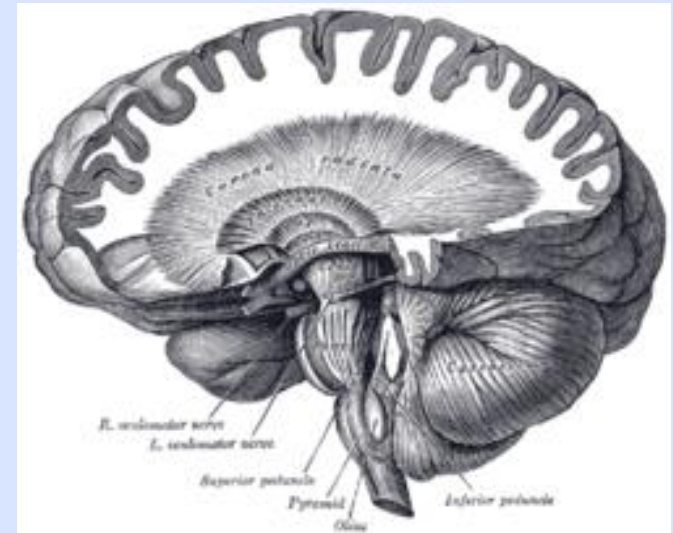
Normal Pressure Hydrocephalus

Normal Pressure Hydrocephalus

- Characterized by findings of:
 - Dementia
 - Urinary Incontinence
 - Gait abnormalities
- First described by:
 - Dr. Hakim in 1965
- Important as it is potentially reversible

Pathophysiology

- Enlargement of the ventricles impinge on the corona radiata
- Especially posteriorly affecting lumbo-sacral nerve fibers.
- Dementia possibly results from compression of the periventricular limbic system.
- Intermittent increased pressure in the ventricles



Epidemiology

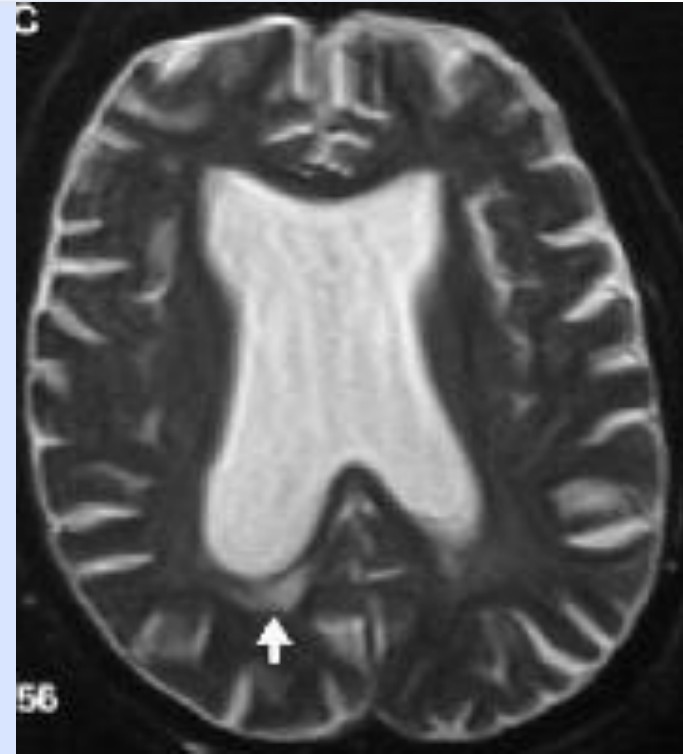
- Estimated at 21.9/100,000
- Possibly as high as 14% of all dementias
- Race – no association recognized
- Gender – no association recognized
- Age – increases risk

Clinical presentation

- Initial symptom
 - Gait apraxia
 - Bradykinetic
 - Magnetic or shuffling
- Urinary symptoms
 - Begin initially as increased frequency
 - Becoming urgency
 - Develop incontinence
- Dementia
 - Bradyphrenia
 - Decreased attention
 - Inertia
 - Increasing forgetfulness
 - Aphasia develops late
- Pyramidal tract
 - findings are common

Causes

- Idiopathic in ~ 50%
- Head injury
- Meningitis
- CNS tumor
- Subarachnoid hemorrhage
- Previously compensated congenital hydrocephalus





Work up

- Lumbar Puncture with removal of a large volume of spinal fluid
- Imaging study – ventricular enlargement with subependymal flow
- Nuclear medicine study of CSF flow

Treatment

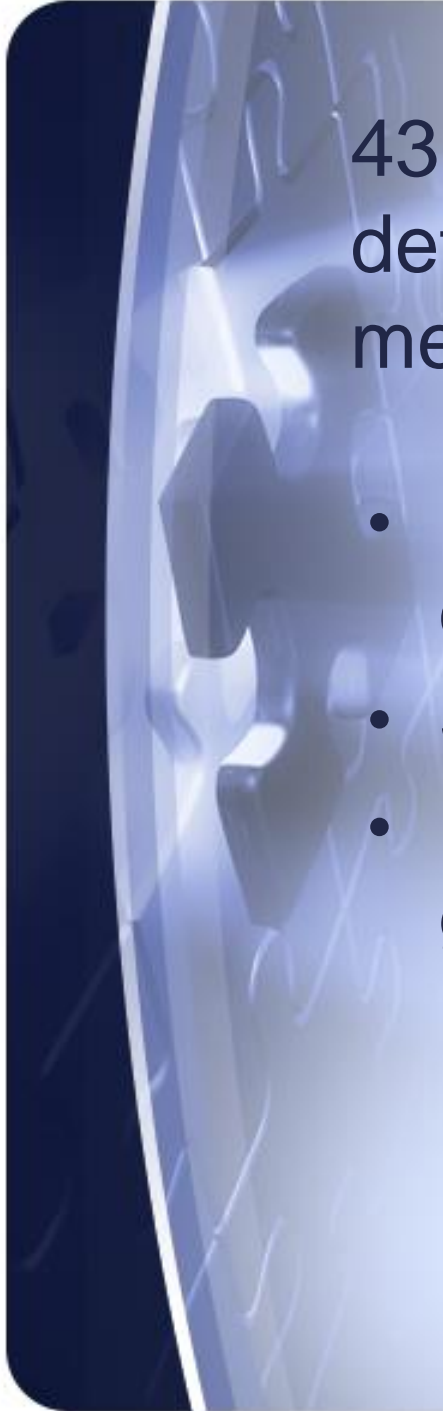
- Ventriculoperitoneal shunt placement
 - This is helpful early in the course
- Once dementia begins, it is unlikely to change the course

Questions:

- What is the classic triad for Normal Pressure Hydrocephalus?
 - Gait apraxia
 - Urinary incontinence
 - Dementia
- Is treatment always effective?
 - If intervention is begun early, if dementia has begun, it is unlikely to be beneficial.



Case Presentation 10



43 year old man who was referred to determine ongoing need for anti-seizure medication.

- Hit on the head 4 yrs prior and was “in a coma for 3 to 5 months”.
- Started on anti-seizure medications.
- Never had a documented seizure in his group home.



Past medical history

- Coronary artery disease s/p bypass surgery
- Depression
- Sleep apnea
- TBI
- Polysubstance abuse
- Mental retardation
- Self injurious behaviors (head banging)

Examination:

Alert and able to follow simple commands

Speech - dysarthric.

- Medications:

- Aspirin

- Niaspan

- Flonase

- Zocor

- Prevacid

- Vistaril

- Plavix

- Nitro-Dur

- Vitamin B

- Colace

- Depakote

- Dilantin

- Calcium

- BuSpar

- Klonopin

- Monopril

- Celexa

- Abilify

- K-Dur

- Fish oil

- Folate



Return Visit:

49 year old man referred back to the clinic due to cognitive decline.

- He has not had any seizures
- He is mildly ataxic
- Some days are good, and he will talk
- Other days are bad, and he will attack people, and tear up the house.
- He refuses to eat most days, and has lost 65 to 75 pounds
- He refuses to shower, brush his teeth, change clothes, and refuses personal hygiene
- He sleeps only intermittently and only for a few hours
- Type 2 Diabetes



Exam:

- General: agitated disheveled foul smelling man
- Speech was slurred, and limited to one or two words, and he perseverated most of the time.
- He moved all limbs
- Gait: mildly ataxic.

Medications at follow up

- Vitamin D
- Multivitamin
- Plavix
- Simvastatin
- Metoprolol
- Colace
- Magnesium
- Risperdal
- Zolpidem
- Isosorbide
- Nitroglycerin patch
- Lorazepam
- K-Dur
- Metformin
- Flonase
- Prilosec
- Gax-X
- Neurontin
- Sucralfate
- Klor-Con
- Aspirin
- Doxazosin
- Fish oil
- Depakote
- Buspirone
- Ciprofloxacin

- 
- Vitamin D
 - Multivitamin
 - Plavix
 - Simvastatin
 - Metoprolol
 - Colace
 - Magnesium
 - Risperdal
 - Zolpidem
 - Isosorbide
 - Nitroglycerin patch
 - Lorazepam
 - Metformin
 - Flonase
 - Prilosec
 - Gax-X
 - Neurontin
 - Sucralfate
 - Klor-Con
 - Aspirin
 - Doxazosin
 - Fish oil
 - Depakote
 - Buspirone
 - Ciprofloxacin

diagnosis



Imaging Studies

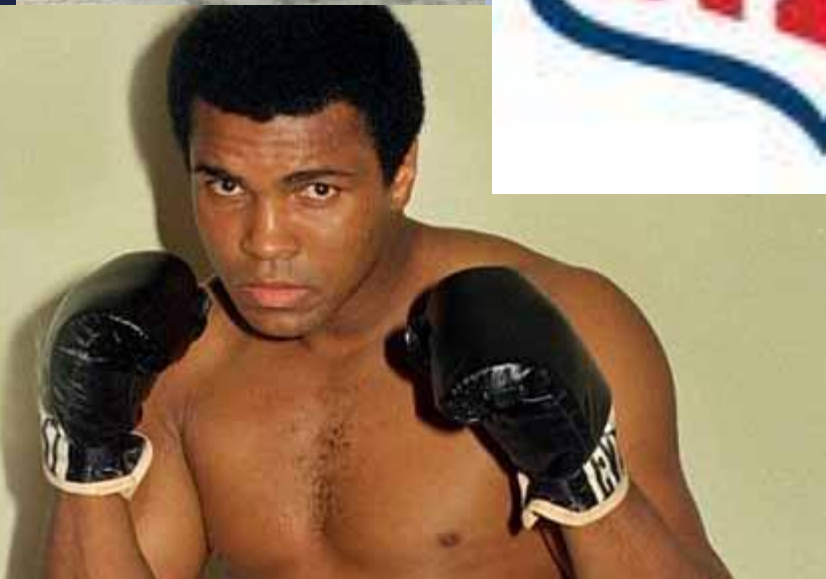
- CT of the brain in 10/2000:
 - 3 cm hemorrhagic contusion with edema lateral left temporal lobe
 - Small contusion , right gyrus rectus
 - Small anterolateral left frontotemporal subdural hematoma with mild mass effect
 - Subdural blood along the anterior interhemispheric fissure
 - No fractures

Imaging studies

- CT of the brain 2009
 - Advanced Cerebral Volume loss for age.
 - Evidence of prior traumatic brain injury with mild left frontal polar and left inferior temporal encephalomalacia.

An abstract digital background featuring a glowing blue sphere on the left side, with a dark, stylized figure or object positioned in front of it. The background is filled with horizontal blue lines and a gradient of light to dark blue. The overall aesthetic is futuristic and high-tech.

Chronic Traumatic Encephalopathy



History

- Originally described by Harrison Martland, MD in 1928
 - Progressive neurologic deterioration in boxers “punch drunk”.
 - 1937 called “Dementia Pugilistica” by Dr. Millspaugh.
- Other activities noted to cause syndrome
 - 1949 - Progressive traumatic encephalopathy
 - Now Chronic Traumatic Encephalopathy

Mechanisms of cerebral injury

- Direct concussion:
 - Front to back injuries are not as severe as lateral (side-to-side) injuries.
 - A fluid wave in the lateral ventricles produces a shearing force on the septum pellucidum, believed to cause a septum cavum pellucidum.
 - Damage to the blood-brain barrier releases local neurotoxins, and may contribute to the formation of neurofibrillary tangles

Mechanisms of injury

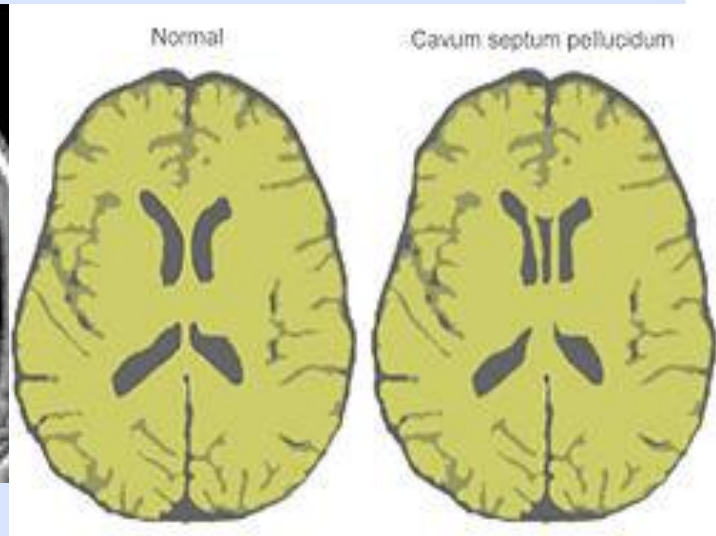
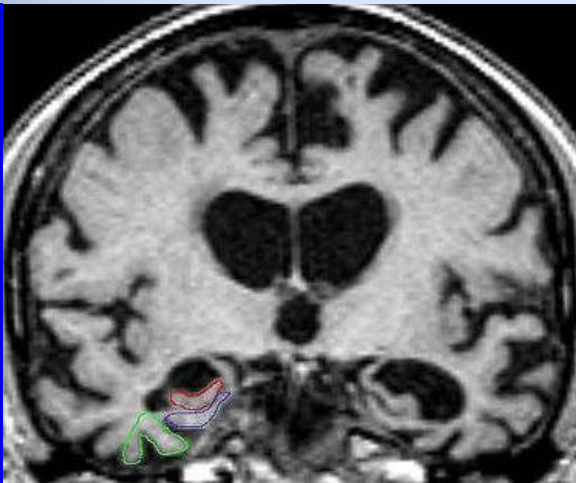
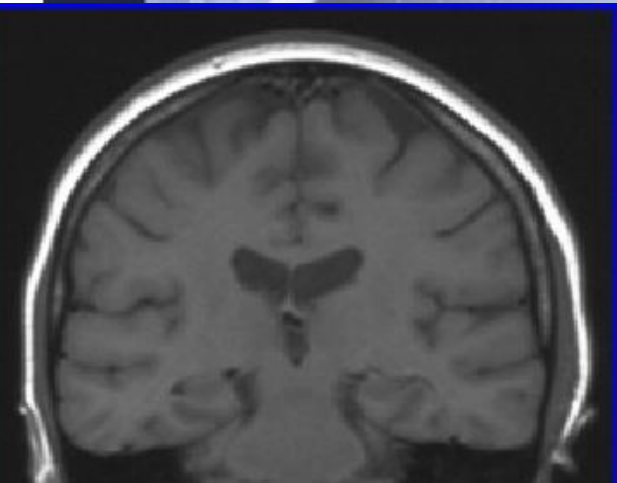
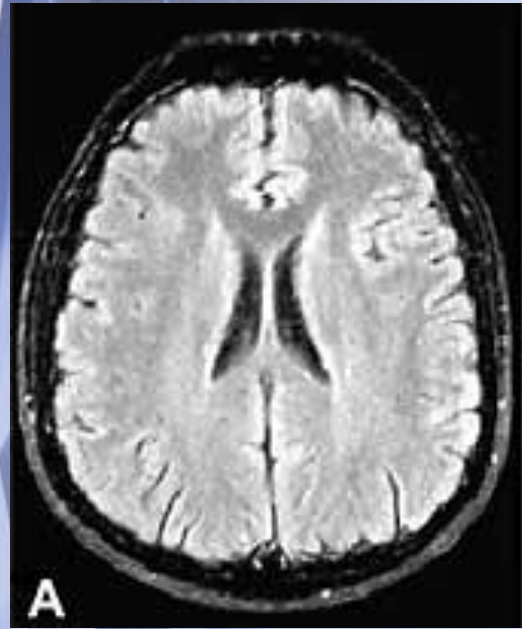


- White matter changes
 - Stretching the axons (shearing injury)
 - damages the microtubules
 - Damages mitochondria
 - Leads to uncontrolled influx of calcium
 - Leads to cellular destruction



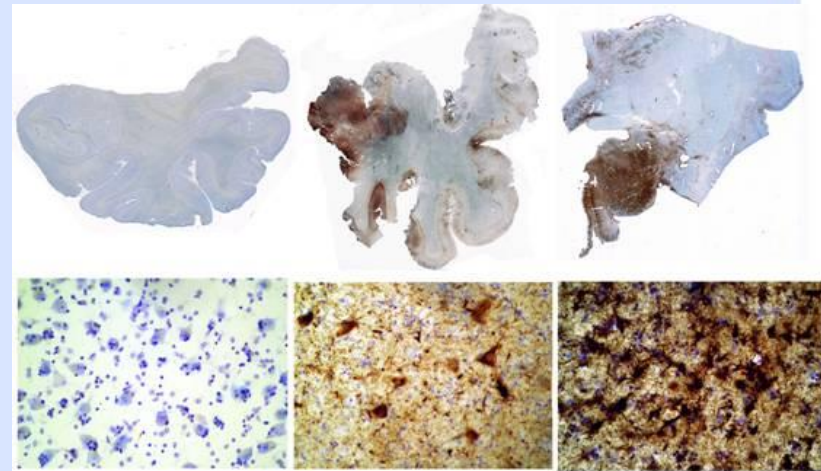
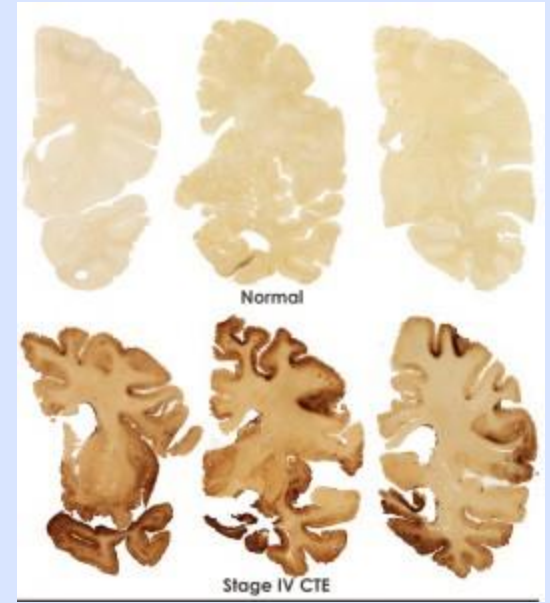
pathophysiology

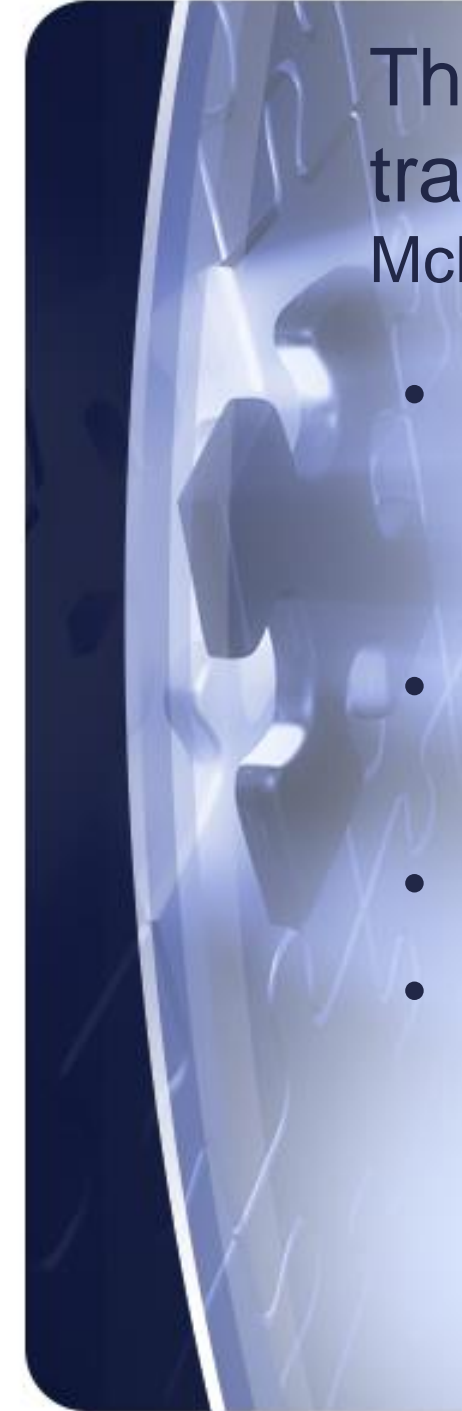
- Atrophy of the cerebral hemispheres
- Atrophy of the medial temporal lobes
- Atrophy of the mammillary bodies and diencephalon
- Enlargement of the ventricles
- Development of cavum septum pellucidum



pathophysiology

- Extensive neurofibrillary tangles
- Astrocytic tangles
- Extensive degeneration of the axons
- Relative absence of beta-amyloid deposits





The spectrum of disease in chronic traumatic encephalopathy

McKee A et al. Brain 2013;136;43-64.

- 85 deceased subjects
 - 58 football players
 - 1 male with history of SIB (head-banging)
- Interviews of family members re: cognitive status prior to death
- Pathologic evaluation
- Classified four levels of impairment

Clinical manifestations

Stage 1

Several were asymptomatic

Decreased attention span

Decreased concentration

Short-term memory difficulties

Aggressive tendencies

depression

Stage 2

- Depression
- Mood swings
- Headaches
- Short-term memory loss
- Explosivity
- Loss of attention and concentration
- ↑ impulsivity
- ↑ suicidality
- Language difficulties

Clinical manifestations – stage 3

- Memory loss
 - Executive dysfunction
 - Explosivity
 - Difficulty with attention and concentration
 - Depression
 - Mood swings
 - Visuospatial difficulties
 - Aggression
 - Impulsivity
 - Apathy
 - Headaches
 - suicidality
- * 75% in this category are considered cognitively impaired.

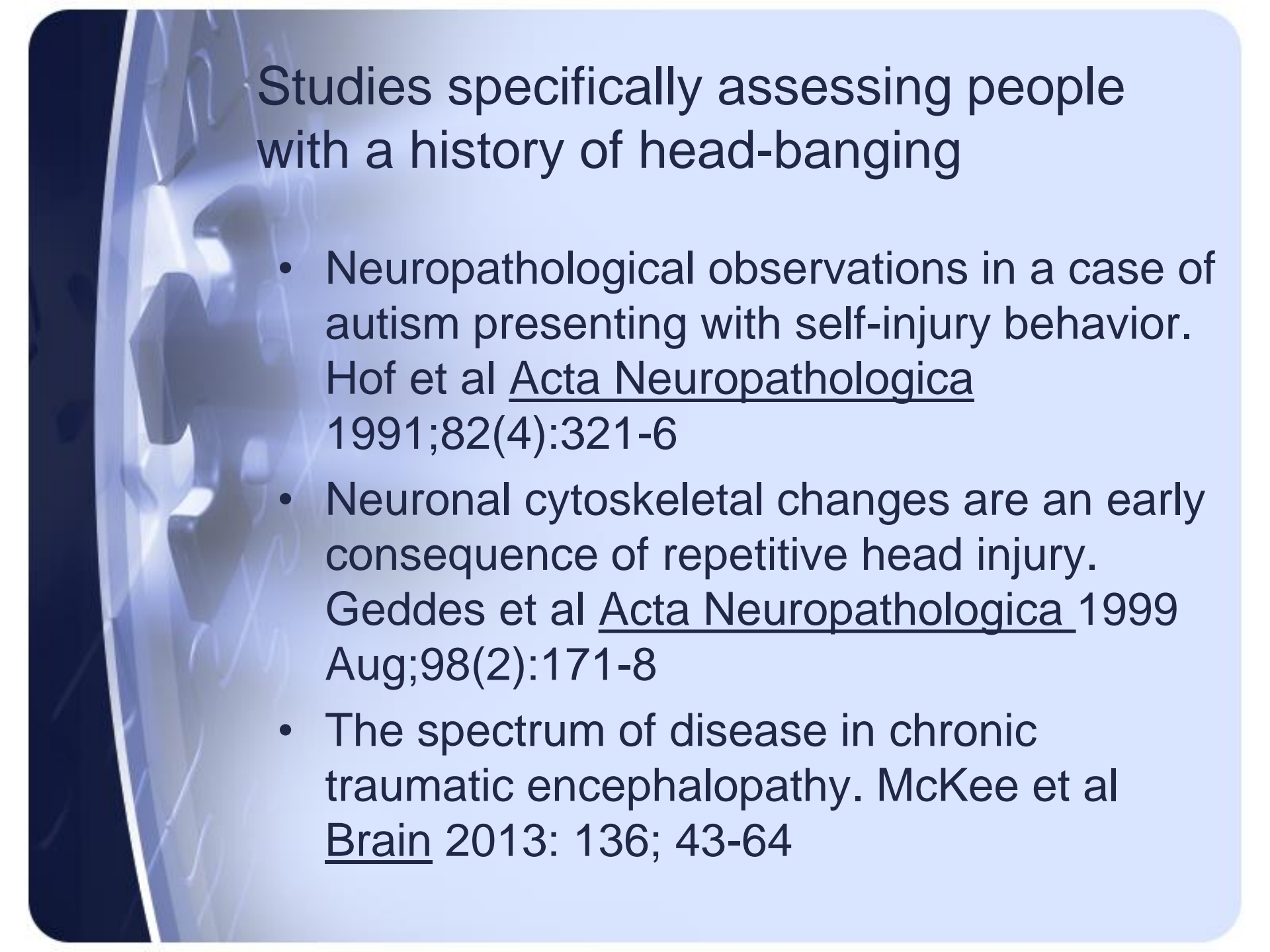
Clinical Manifestations – Stage 4

- Profound loss of executive function
- Profound loss of Memory
- Profound loss of attention and concentration
- Explosivity
- Aggressive tendencies
- Paranoia
- Depression
- Gait difficulties
- Visuospatial difficulties
- Dysarthria
- Parkinsonism
- Suicidality in 31%



Treatment

- Symptomatic



Studies specifically assessing people with a history of head-banging

- Neuropathological observations in a case of autism presenting with self-injury behavior. Hof et al Acta Neuropathologica 1991;82(4):321-6
- Neuronal cytoskeletal changes are an early consequence of repetitive head injury. Geddes et al Acta Neuropathologica 1999 Aug;98(2):171-8
- The spectrum of disease in chronic traumatic encephalopathy. McKee et al Brain 2013: 136; 43-64




Questions:

- Does the severity of the impact matter with regard to the development of dementia?
 - A single severe impact can cause severe brain injury, but so to can repetitive mild impacts over time.
- What's worse, side-to-side impact, or front-to-back impact?
 - Side-to-side impacts are more damaging.



Case Presentation 11



22 yr old woman with history of profound cognitive impairment, referred for concerns of decline

- Sent with direct care staff member who has worked with the patient one month
- She has been with the agency for < 1 year.
- Lived with mom, who is now deceased, prompting placement.



Past medical history:

- Hypothyroidism
- Seizure disorder – none observed to date
- Profound cognitive impairment
- Etiology- unknown
- Exam:
 - Non-verbal, unable to follow commands
 - Disinterested in surroundings or people
 - Has cataracts both eyes
 - Unremarkable motor, sensory, cerebellum, reflexes, and gait.

Diagnosis?



The image features a blue-toned abstract graphic. On the left, a globe-like structure is composed of several interlocking, semi-transparent blue spheres. A bright, glowing light beam originates from the center of the globe and extends horizontally across the upper right portion of the image. The background is a gradient of dark blue, with a curved, metallic-looking band at the bottom. The overall aesthetic is futuristic and scientific.

Testing for Dementia

Why is this important?

- The life expectancy of a person with Down Syndrome:
 - in the 1930s was 11 years of age.
 - In 1999, increased to > 60 years.
- For people with cognitive impairment from any cause:
 - In 1931, was 22 years of age.
 - In 1976, increased to 59 years.
 - In 1993, increased to 66.1 years.

How do we test for dementia?



Mini Mental Status Exam

Mini-Mental Status Examination

The Mini-Mental Status Examination offers a quick and simple way to quantify cognitive function and screen for cognitive loss. It tests the individual's orientation, attention, calculation, recall, language and motor skills.

Each section of the test involves a related series of questions or commands. The individual receives one point for each correct answer.

To give the examination, seat the individual in a quiet, well-lit room. Ask him/her to listen carefully and to answer each question as accurately as he/she can.

Don't time the test but score it right away. To score, add the number of correct responses. The individual can receive a maximum score of 30 points.

A score below 20 usually indicates cognitive impairment.

The Mini-Mental Status Examination

Name: _____ DOB: _____

Years of School: _____ Date of Exam: _____

Orientation to Time	Correct	Incorrect	
What is today's date?	<input type="checkbox"/>	<input type="checkbox"/>	
What is the month?	<input type="checkbox"/>	<input type="checkbox"/>	
What is the year?	<input type="checkbox"/>	<input type="checkbox"/>	
What is the day of the week today?	<input type="checkbox"/>	<input type="checkbox"/>	
What season is it?	<input type="checkbox"/>	<input type="checkbox"/>	
			Total: _____

Orientation to Place	Correct	Incorrect	
Whose home is this?	<input type="checkbox"/>	<input type="checkbox"/>	
What room is this?	<input type="checkbox"/>	<input type="checkbox"/>	
What city are we in?	<input type="checkbox"/>	<input type="checkbox"/>	
What county are we in?	<input type="checkbox"/>	<input type="checkbox"/>	
What state are we in?	<input type="checkbox"/>	<input type="checkbox"/>	
			Total: _____

Immediate Recall

Ask if you may test his/her memory. Then say "ball", "flag", "tree" clearly and slowly, about 1 second for each. After you have said all 3 words, ask him/her to repeat them – the first repetition determines the score (0-3):

Ball	<input type="checkbox"/>	<input type="checkbox"/>	
Flag	<input type="checkbox"/>	<input type="checkbox"/>	
Tree	<input type="checkbox"/>	<input type="checkbox"/>	
			Total: _____

Maximum
score

Score

5

—

Orientation

What is the (year) (season) (date) (day) (month)?

5

—

Where are we: (state) (county) (town or city) (hospital) (floor)?

Registration

Name three common objects (e.g., "apple," "table," "penny"); Take one second to say each. Then ask the patient to repeat all three after you have said them. Give one point for each correct answer. Then repeat them until he or she learns all three. Count trials and record.

Trials: _____

Attention and calculation

Spell "world" backwards. The score is the number of letters in correct order.

(D__L__R__O__W__)

5

—

Recall

Ask for the three objects repeated above. Give one point for each correct answer.

(Note: recall cannot be tested if all three objects were not remembered during registration.)

3

—

Language

Name a "pencil" and "watch."

Repeat the following: "No ifs, ands or buts."

Follow a three-stage command:

"Take a paper in your right hand, fold it in half and put it on the floor."

Close your eyes.

Write a sentence.

Copy the following design.

2

—

1

—

3

—

1

—

1

—

1

—

Total
score: _____



Mini mental status exam


- Most common test
- Assesses the following domains:
 - Orientation
 - Attention
 - Immediate and short term recall
 - Language
 - Ability to follow simple verbal and written commands.

Mini Mental Status Exam

- Takes about 10 minutes.
- Useful in objectively tracking progression.
- Can not differentiate the different types of dementia.

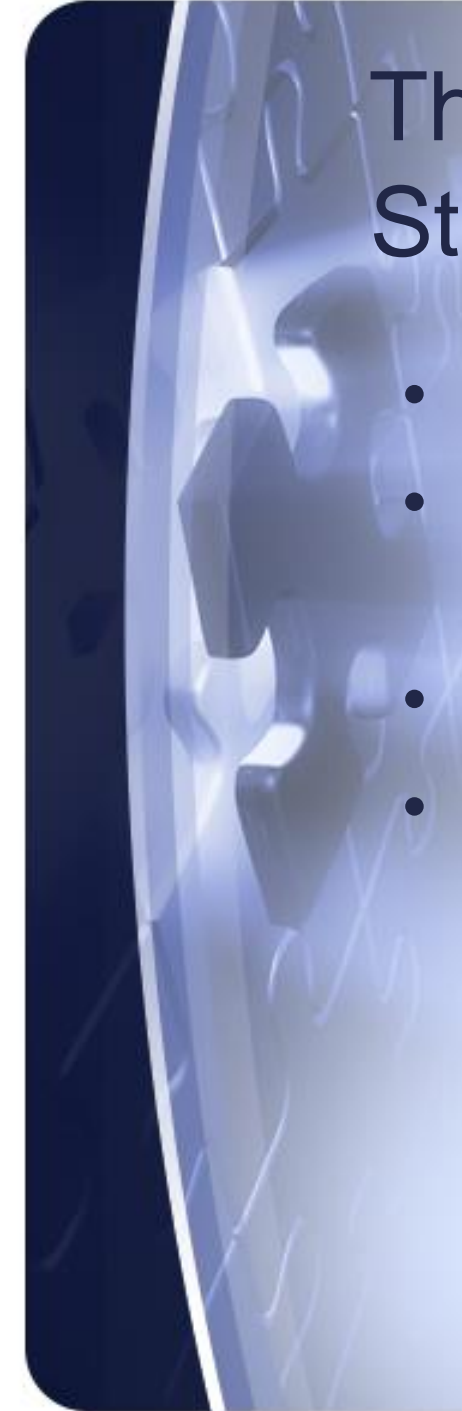
Dementia Rating Scale

- Assesses cognition in 5 areas:
 - Attention
 - Perseveration and initiation
 - Construction
 - Memory
 - Conceptualization
- Simple to administer
- Typically takes 20 to 40 minutes to administer.
- Sensitive and specific



The Neurobehavioral Cognitive Status Examination

- Assesses cognition in 5 areas:
 - Language
 - Constructional ability
 - Memory
 - Calculation skills
 - Reasoning and judgement
- Three general factors:
 - Level of consciousness
 - Attention
 - Orientation



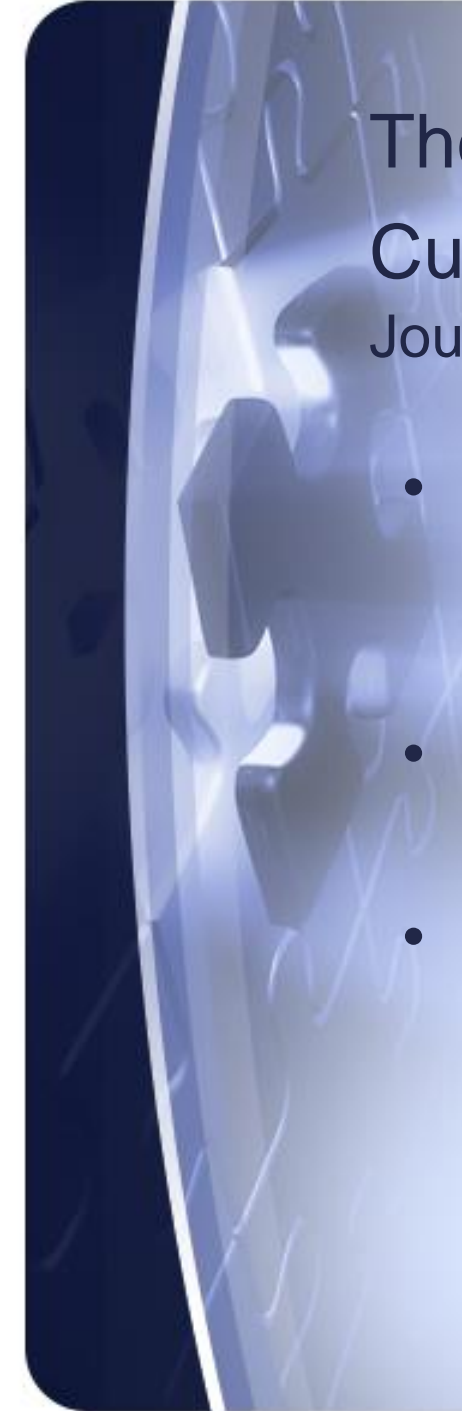
The Neurobehavioral Cognitive Status Examination

- Highly sensitive
- Can be administered 5 minutes or less in “normal” individuals.
- Not specific, with 70% false positives
- Determined that it should not be used as the sole measure of dementia.



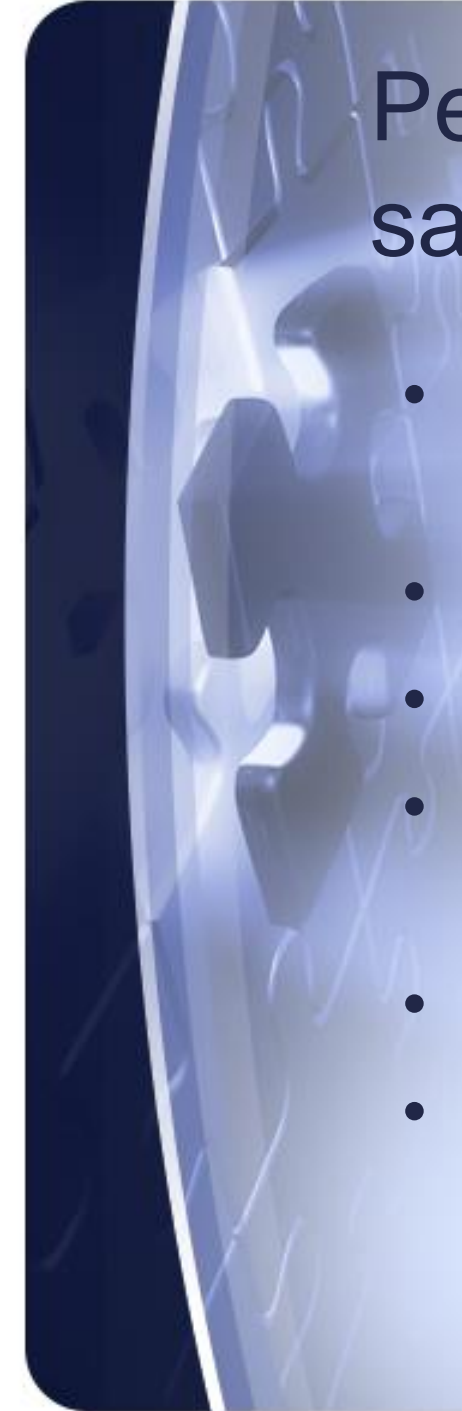
The problems with these tests.

- At baseline people with DD have:
 - Memory impairments
 - Impairments in communication
 - Poor executive functions
- They cannot perform the standard tests at baseline.
- People who are DD generally do not present with dementia in the same way as the general population.



The Elderly Mentally Retarded Person: Current Perspectives and Future Directions. Journal of Applied Gerontology,(1995)14 (3): 275-288

- They noted a lack of literature discussing elderly people with developmental disabilities.
- They noted, lack of consistency with cohorts.
- They note that people with developmental disabilities appear to age earlier than a non-developmentally disabled cohort.



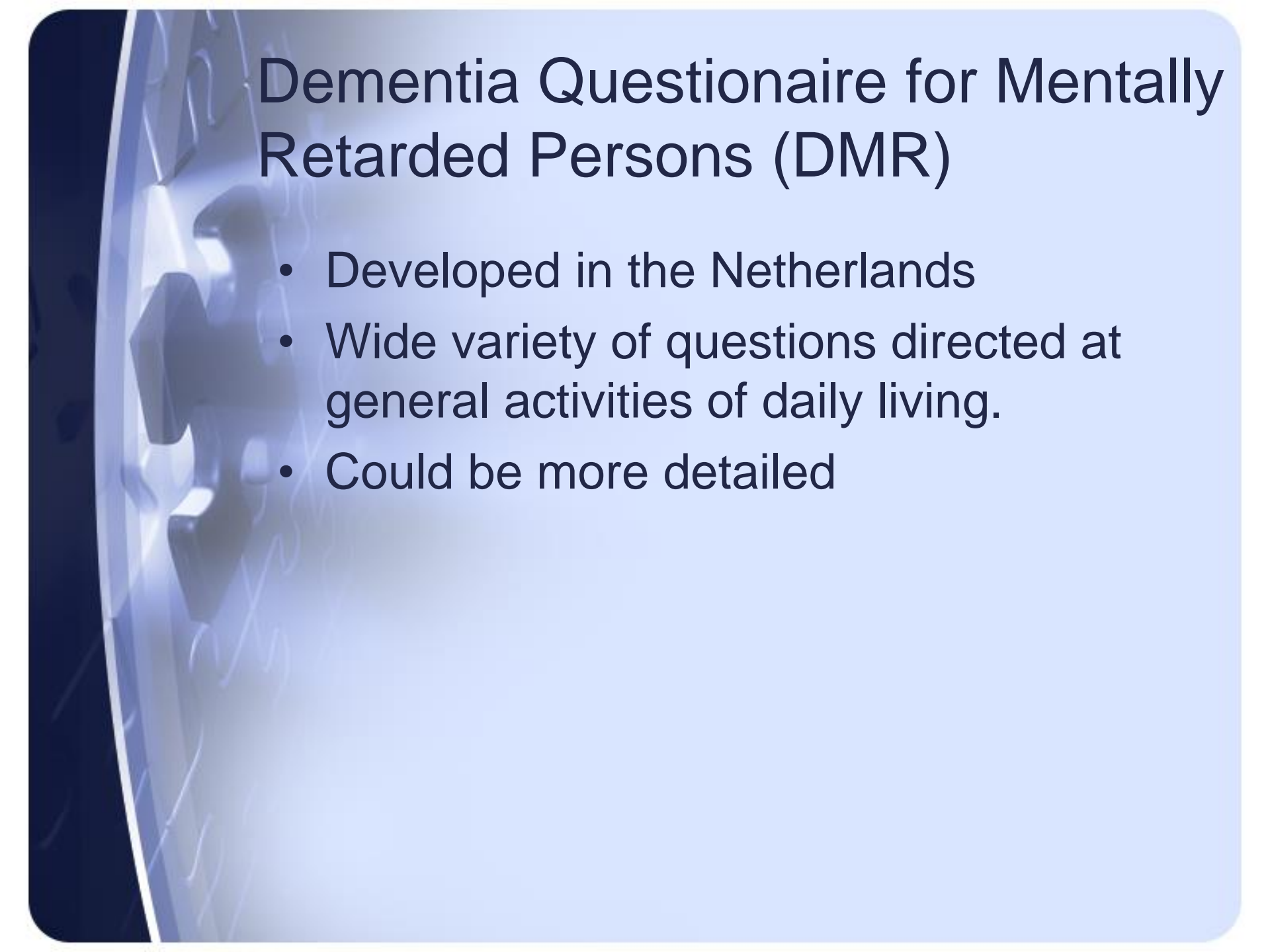
People are susceptible to the same changes as anyone else

- Increasing impairment of vision and hearing.
- Decreased muscle mass and flexibility.
- Increased incidence of arthritis.
- Increased risk of hypertension and heart disease.
- Diabetes
- They are at ↑ risk of osteoporosis



What is needed?

- To diagnose dementia in a developmentally disabled person you need to determine the following:
 - A change in the individual's baseline functioning.
 - Knowledge of the individual's baseline personality.
 - A reliable reporter.




Dementia Questionnaire for Mentally Retarded Persons (DMR)

- Developed in the Netherlands
- Wide variety of questions directed at general activities of daily living.
- Could be more detailed



E-Chat Health Assessment Tool

- Developed by DDS
- More detailed, but developed for nursing care assessments

- 
- The Working Group for the Establishment of Criteria for Diagnosis of Dementia in Individuals with Intellectual Disabilities
 - Recommended that all individuals with intellectual disability be tested as a baseline by 25 years of age.
 - How is the paperwork going to follow the person?



Case Presentation 1



Case Presentation:

- 69 yo man living with daughter and her family.
- Very set routine.
 - Up at 6 AM
 - Washes up and dresses
 - Assists the family with getting ready for the day
 - Walks down the block to get the paper
 - Returns, reads the paper and eats breakfast.
 - Etc...

Sudden change...

- One day, the news stand was closed.
- Man was lost for hours.
- When found he seemed unaware of what happened, failed mental status testing.
- What is the diagnosis?



Alzheimer's Disease

- When people have a very structured routine, and minimal demands, they can become severely demented before it is recognized.



Causes for acute alteration

- Abrupt acceleration in degenerative illness can be due to superimposed delirium due to:
 - Urinary Tract Infections
 - Pneumonia
 - Medication errors
 - Trauma



Case Presentation #2




Case Presentation 2

- Previously healthy 72 year old woman
- Owns a dress shop in a small town
- Began having difficulty with managing the budget
- Eldest son became involved, and recognized a problem
- Woman was seen by PCP
- Referred to Neurologist, testing indicated cognitive decline.

Diagnosis?





Patient was diagnosed with dementia – probable Alzheimer's Dementia

- She continued to deteriorate, and had to be placed in an assistive living apartment
- She was seen by a second physician.
- She was diagnosed with Pernicious Anemia (Vitamin B12 deficiency)
- She began to receive B12 injections
- She is now back to living with her husband and running a smaller scale store.



Summary

- Dementia should be a diagnosis of exclusion, other medical problems should be assessed for.
- Multiple types of dementia exist
- Treatment is often with cholinesterase inhibitors or symptomatic treatments.
- Differentiating the types of dementia a patient may have is not always clear.
- New treatments are continuing to emerge.

A person in a wheelchair is shown on a ramp, moving upwards. The image is semi-transparent and serves as a background for the text.

Summary:

- People with developmental disabilities are living longer than they ever have.
- We don't have a good way to accurately measure dementia in the elderly developmentally disabled person.
- Research is needed to help clarify types of dementia and appropriate treatments.

Questions

- What is the most common form of dementia?
 - Alzheimer's Disease
- Which Dementia is the most rapidly progressive?
 - Creutzfeld-Jakob Disease with a longevity of 8 to 15 months.
- How are people with developmental disabilities different from the non-disabled population with regard to presenting signs of dementia?
 - They are more likely to have personality changes rather than memory impairment.