

Week 16

- KW27 - Dementia (Huntington's)
- Case Report: TAL 492 : Dr. Gilmer and Mr. Hyde

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KW27 : Huntington's Disease

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DSM-5 Cognitive Disorders Compared

	Delirium	Mild Neurocognitive Disorder	Major Neurocognitive Disorder
Timeframe	acute (hours to days)	insidious (months)	insidious (months)
Domains affected	Attention Consciousness Orientation (and others)	One or more	Two or more*
NP Deficits	(untestable)	1-2 SD below normal	2+ SD below normal
Activities of Daily Living affected?	Yes	Little	Yes

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DSM-5 : NCD due to Huntington's Disease

- A - NCD criteria are met
- B - insidious onset, gradual progression
- C - Clinically established Huntington's disease (or risk based on family history or genetic testing)
- D - no other etiology can explain the symptoms

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HD Motor Symptoms

- Motor Activity : + or -
 - PD : *hypokinetic*
 - HD : *hyperkintetic*
- Early Symptoms
 - initial reduction in activity / interest
 - within a year “fidgeting” (whole or part limb), irregular patternless movements
 - difficulty sustaining muscle contractions (fluctuating hand grip, can't keep tongue protruded)
- Later Symptoms
 - movements increase, “chorea” (dance-like) involving much of the body
 - walking, talking, eating impacted

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HD Prevalence

- Prevalence:
 - 100% genetically-based
 - very rare, but depends on ancestry
 - White Europeans: 5 to 10 per 100,000
 - Asians: less than 1 per 100,000

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HD Cognitive Symptoms

- Nearly all HD patients have cognitive/emotional symptoms
- Early
 - initial reduction in interest
- Emotional
 - anxiety, irritability, depression, mania, psychosis
- “Subcortical Dementia” (like PD)
 - cognitive slowing
 - executive functioning
 - memory deficits
 - retrieval rather than learning
 - recognition better than recall
- “Cortical” features are rare:
 - Apraxia, agnosia

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HD Course

- Insidious onset
 - First symptoms around age 30-50
- Progression is variable - typically 12-20 years from start until death
- Cause of death
 - 33% pneumonia
 - 25% heart disease
 - 7% suicide

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HD Causes - Genetics

- Genetics
 - Autosomal Dominant gene on chromosome 4
 - 100% genetically-based

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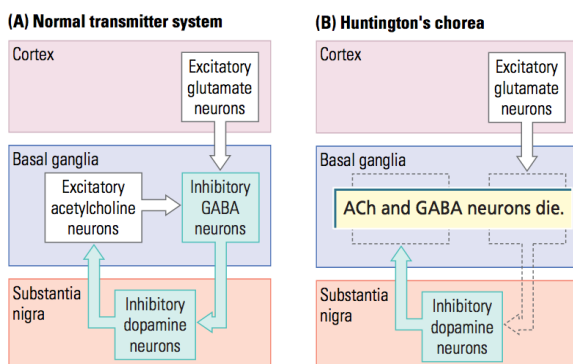
HD Causes - Biology

- Not completely understood
- Basal ganglia atrophy (primary cause)
- Cortex shrinks (side effect)
- Balance of neurotransmitters in Basal Ganglia disrupted:
 - Glutamate
 - GABA
 - Dopamine
 - Acetylcholine
- Loss of GABA inhibition allows Dopamine overactivity?

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HD Causes - Neuroanatomical



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HD Treatment

- No Cure. No single NT to target
- Chorea:
 - Tetrabenazine -
 - inhibits Vesicular Monoamine Transporter (VMAT2) which increases breakdown of Dopamine
 - treats chorea primarily (not dementia)
- Psychiatric symptoms:
 - various drugs
- Other
 - physical therapy (walking, eating)
- Surgery
 - Feeding tube

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HD Prevention

- Entirely genetically based
- No known prevention

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HD Diagnosis & Ethical Issues

- 50% chance for offspring
- Symptoms after child-bearing years
- Genetic testing is available

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TAL 492 : Dr. Gilmer and Mr. Hyde



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Story

- Cane Creek Clinic, North Carolina
 - Dr Vince Gilmer
 - “beloved family doctor”
 - no criminal history
 - at age 41, murdered his father who had “Alzheimer’s Disease”
 - Dr. Benjamin Gilmer (no relation)
 - took over job after Vince went to prison
- Small town, patients confide in both doctors
- Benjamin wants to understand Vince’s crime

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Vince’s History 1

- As child, reportedly had ADHD (mother)
- Wife Karen (also MD)
- Age 40, needed to take medical board exams
 - Extreme anxiety
- Crashed his truck
 - at hospital was disoriented for 24 hours
 - Amnesia: “I’m Bobby Brown”, didn’t recognize his wife
 - several friends worried the crash was intentional
 - to get out of taking boards
 - to get a new truck or as suicide attempt?
- Soon after, Vince left his wife, abruptly
- Began drinking heavily (though reportedly fine at work)

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Vince’s History 2

- Vince picked up his father to transfer him to nursing home
- Father
 - 60 years old
 - taking antipsychotic medication
 - wheelchair bound, could not walk
- Vince “voices telling him to kill his father”
 - Note: father reportedly sexually abused Vince & his sister
- Vince strangles father & mutilates the body
- Then acts normally, begins lying about what happened.
- Body is found, detectives begin investigating
 - suspicious of Vince’s odd behavior
 - lack of emotion / curiosity about the crime
 - threatening detectives

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Vince's History 3

- Detectives felt murder was pre-meditated
 - Vince had obtained rope, gloves and pruning shears
 - Vince was father's guardian, but had not been paying his bills (\$270k debt)
 - Vince had Alaskan vacation planned
 - had changed to one-way ticket
- Vince fled, was caught
 - confessed to the killing
- His defense: "Serotonin"
 - had stopped taking Lexapro days before the crime
 - irritability, hearing voices, compulsions
 - "my brain wasn't working right"

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Vince's History 4

- Many people felt he was "hamming it up"
 - odd movements of head & arm
 - emotional outbursts
 - observers videotaped him
 - odd movements only happened when officers were nearby
- Clinical Psychologist
 - fidgeted, stooped
 - symptoms not consistent with "known anxiety or psychotic" diagnoses
 - "exaggerating his symptoms"
 - Diagnosis
 - Atypical Depression
 - Malingering

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The Trial

- Vince's behavior erratic
- Dismissed his lawyers
- Presentation was illogical
- Calls former girlfriend as witness:
 - Q: "Have I hurt or killed anyone"
 - A: "Your dad"
 - Q: "Well, besides what happened with my father..."
- Vince convicted
 - jurors convinced he was faking
 - life in prison

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Theories

- ASPD / Sociopathy / Psychopathy
- PTSD secondary to sexual abuse as child
- Traumatic Brain Injury due to truck crash
- SSRI withdrawal

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MSE

- Benjamin Gilmer - still trying to understand the crime
- Prison visit, 6 years later
 - Vince requesting specific SSRI dosage/forms
 - only 50, looks much older
 - missing teeth (due to aggression, violence from guards)
 - normal at times
 - odd mannerisms
 - losing track of conversation
 - emotionally labile
 - Reportedly: getting worse
 - Vince begs for help
- Psychiatrist Dr. Buie comes to visit
 - "Could this be Huntington's?"

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Diagnosis & Treatments

- Positive for Huntington's Disease
- Tx: Lexapro (SSRI)

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