

Week 16

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

KW27 : Huntington's Disease

DSM-IV Dementia due to Huntington's Disease

- Presence of Dementia, judged to be due to HD
- HD Symptoms
 - insidious development
 - movement disorder
 - early : increased fidgeting
 - late: chorea
 - cognitive / emotional changes
 - early: personality changes, irritability, anxiety, depression
 - late: dementia
- Note: the DSM-IV diagnosis is not terribly detailed.

HD Prevalence

- Prevalence:
 - very rare - 5 to 10 out of 100,000 adults worldwide
 - more common in white Europeans
 - 100% genetically-based

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

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

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

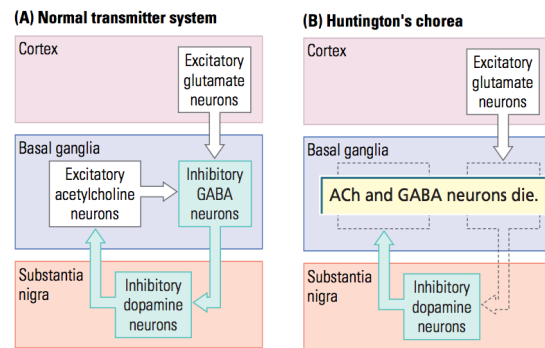
- Not completely understood
- Basal ganglia atrophy (primary cause)
- Cortex shrinks (side effect)
- Balance of neurotransmitters in Basal Ganglia disrupted:
 - Glutamate
 - GABA
 - Dopamine
 - Acetylcholine
- Loss of GABA inhibition allows Dopamine overactivity?

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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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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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