Feeling Sad and Angry: What Can You Do About It?

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Huntington's Pathology

'Huntingtin pathological protein aggregation causes progressive cerebral deterioration starting in the striatum' E. van Duijn et al. The Journal of Neuropsychiatry and Clinical Neurosciences 2007; 19:441–448

Motor, Cognitive and Behavioral symptoms all reflect disruptions in connections between the frontal lobe and the striatum (which consists of the caudate and putamen).

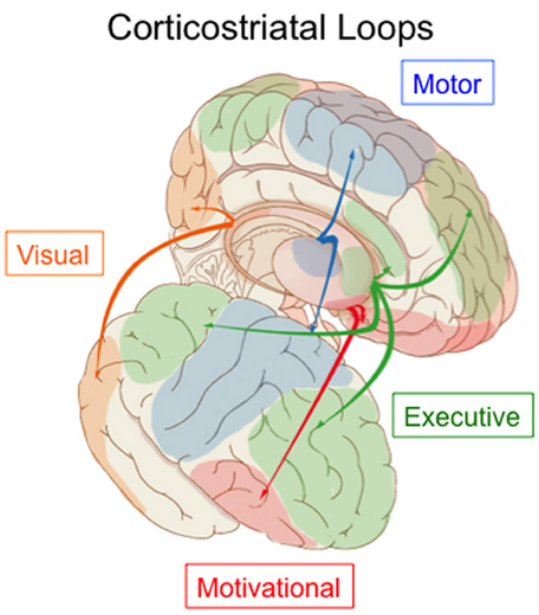
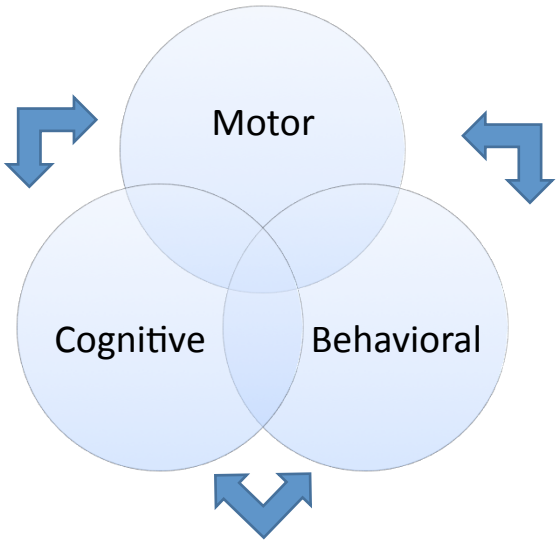


Image borrowed from CA Seger Frontiers in Systems Neurosci., 06 December 2013

Huntington's Disease



How Often?

- Over 2/3 of Huntington's Patients will have at least one symptom traditionally partitioned into the neuropsychiatric/ behavioral category.
  - Problems can develop as in the general population
  - Problems can emerge due to impaired coping from cognitive impairment
  - Some problems are symptomatic of Huntington's Disease itself

How Often-Registry (n=1993) van Duijn E, et al. J Neurol Neurosurg Psychiatry 2014;85:1411–1418.

- Apathy
  - 48%
  - Most dramatic correlation with severity of disease
- Depression
  - 42%
- Irritability and Aggression
  - 39%

How Often-Registry

- Obsessive Compulsive tendencies
  - 25 %
- Psychosis
  - 4%
  - Unlike other symptoms did not increase with disease severity
    - Associated more with family history of psychosis

Non-REGISTRY categories

- Mania- up to 10 % of HD Rosenblatt 2007
  - 5X historical risk, 2X contemporary
- Disinhibition- 35% of patients Paulsen et al 2001

Suicidality

- Twice as likely to have ideation (19% vs 10)
- 5 times as likely as general population to complete (5.7% vs. 1.2%)
  - AAM Hubers et al Journal of Affective Disorders 136 (2012) 550–557
- Early study indicating risk if family member had selection bias of family report
  - More likely to remember relatives who killed themselves and forget healthy relatives in composing family history
    - Di Maio et al, J Med Genet 1993; 30: 293-295

Suicidality

- Risk of suicide in HD correlates with depression, irritability, and benzodiazepine use
  - AAM Hubers Journal ofAffectiveDisorders151(2013)248–258
- Bipolar patients have 20 fold increased risk of general population
  - Pompili et al, Bipolar Disord. 2013 Aug;15(5):457-90
- 2/3 of suicides are due to major depression
  - 20 % lifetime prevalence in gen pop.
    - (White House Conference on Mental Health, 1999)

What Can You Do About It? Specific interventions

- Apathy
  - Maintain structured schedule
  - Stimulants can help if energy cause of refusal
- Depression
  - Beware dread
  - Psychotherapy and support
  - Medications if clinically depressed
    - 2 weeks of five key symptoms- any MD can diagnose

What Can You Do About It? Specific interventions

- Obsessive Tendencies
  - Redirection and Distraction
  - Response Prevention
  - Antidepressants if difficult to refocus
    - Antipsychotics if irrationally preoccupied
- Irritability / Aggression / Disinhibition
  - Consequences
    - Calmly implemented
    - Not debates or arguments
  - Medications under expert consultation
  - Watch out for anti-depressant induced irritability

What Can You Do About It?

- Don't ignore problems
- Don't jump to conclusions
- Identify and enlist trusted advisor(s)

References

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