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

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

- Apathy
  - 48%
  - Most dramatic correlation with severity of disease

- Depression
  - 42%

- Irritability and Aggression
  - 39%

- 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 \textsuperscript{Rosenblatt 2007}
  • 5X historical risk, 2X contemporary

• Disinhibition- 35% of patients \textsuperscript{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)

• 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 of Affective Disorders 151(2013)248–258

- Bipolar patients have 20 fold increased risk of general population

- 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

- van Duijn E, et al. J Neurol Neurosurg Psychiatry 2014;85:1411–1418