Huntington's Disease on the inpatient psych unit

Last updated October 2023.

Introduction. Huntington's Disease (HD) is a genetic neurodegenerative disorder. This disease occurs due to an abnormality in the HTT gene located on chromosome 4, specifically, the repetition of a particular sequence of DNA, known as CAG repeats, more times than normal. This genetic mutation leads to the symptoms associated with Huntington's Disease. If a patient has a parent with the condition, their risk of inheriting the disease is 50%. There's no cure, but supportive care helps manage symptoms.

Symptoms:

• **Neuromuscular:** Patients exhibit chorea, characterized by unpredictable movements of the face, limbs, and trunk, as well as rigidity and dystonia.
• **Cognitive:** Includes declines in memory, attention, and executive functions.
• **Psychiatric:** Symptoms like depression, irritability, and anxiety can precede neuromuscular symptoms by years, with psychosis developing in advanced stages.

Disease Onset and Progression:

• The age when symptoms first appear and their severity correlate with the number of CAG repeats.
• A repeat count above 40 guarantees disease manifestation.
• Most patients develop symptoms between 30 and 50 years old, but juvenile-onset HD can emerge before age 20.
• Life expectancy is shortened, with the average life expectancy 15-20 years after onset of symptoms.

Diagnosis: If a patient displays a mix of movement, cognitive, and psychiatric symptoms, especially with a family history of HD, obtain genetic testing. Prior to testing, ensure your patient receives genetic counseling, given the profound implications a diagnosis can have for them and their family.

Management:

- Multidisciplinary approach is essential, e.g. with neurologists, physical therapists, others.
- Medication Management:
    - VMAT-2 inhibitors: Tetrabenazine, deutetrabenazine and valbenazine can mitigate choreic movements. Watch for side effects, including depression and suicidal thoughts.
    - Antidepressants: SSRIs or SNRIs help with depression and obsessive-compulsive symptoms.
    - Antipsychotics: Useful for controlling both chorea and psychosis but monitor for worsening tremors and muscle stiffness**.**
    - Mood Stabilizers: For mood swings.
    - Benzodiazepines— for anxiety but use cautiously as they can worsen cognition.
- Psychotherapy: Cognitive-behavioral therapy (CBT) helps in managing anxiety, obsessive-compulsive symptoms, and depression.
- Physical therapy: to enhance mobility and reduce fall risk.
- Nutritional Guidance: to assess the patient's dietary needs and make recommendations, e.g. for pureed food to prevent choking.
- Speech Therapy: Helpful for patients who struggle with speech due to dysarthria.
- Capacity Evaluation: Regularly assess the patient's ability to understand and make decisions about their care; maintain a clear line of communication with their designated decision-makers.
- Advanced Care Planning: Initiate early, before the patient's decision-making capacity becomes compromised.
- Family Support: Family therapy or support groups on the unit can be invaluable for relatives feeling stressed or overwhelmed.
- Post-discharge Planning: Ensure patients and families are aware of community resources, such as the Huntington's Disease Society of America at https://hdsa.org.