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Medical Management of Advanced HD

Samuel Frank, MD
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Overview

- What is advanced HD?
- Progression of disease
- Issues in management
- Ideas for management
- Dignity in advanced stages



Presenter Disclosures

Samuel Frank, MD

The following personal financial relationships with commercial interests relevant to this presentation existed during the past 12 months:

No relationships to disclose or list



“Technically,” Advanced HD is

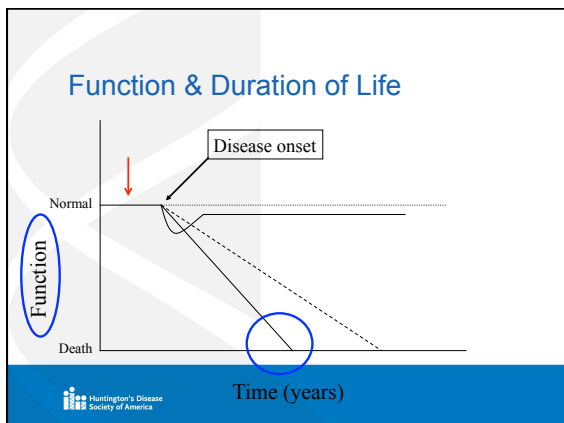
- Total Functional Capacity (TFC) score of 0-2
 - 13 point scale used for staging
- “Functional” definition
 - Patients who are no longer able to independently care for themselves due to physical, cognitive or behavioral reasons



Rate of Progression

- Early in disease, loss of TFC is 0.5-1 point per year until total is 5-6.
 - Average TFC of a patient admitted to an extended care facility is typically 4-6.
- Later in HD – not well studied
- Patients may spend 5-10 years or longer under near total to total care





Predictors of Extended Care Placement

- Nursing home residents had worse motor function:
 - Chorea, slow movements, walking troubles, and imbalance
- More likely to have behavioral disease:
 - Obsessions, compulsions, delusions, auditory hallucinations
 - Aggression, disruptive and irritable behaviors

Huntington's Disease Society of America
Wheelock, 2003

Predictors of Extended Care Placement

- Important conclusion: motor variables alone predicted nursing home placement

Huntington's Disease Society of America
Wheelock, 2003

Average Characteristics of Patients in Extended Care

- Nance, 1996
 - 45 years old
 - Disease duration = 10.0 years
 - 47% women
 - Previously employed
 - High school graduate
 - Unmarried (44% never, 33% divorced)
- Wheelock, 2003
 - 52 years old
 - Disease duration = 8.6 years
 - 63% women

Huntington's Disease Society of America
Nance, 1996

Medical Issues in Advanced HD

- Behavioral disturbance
- Movements
 - Including falls
- Dementia
- Delusions & other forms of psychosis
- Swallowing & Nutrition
 - Weight loss/gain
- Mood & anxiety

Huntington's Disease Society of America

Everyday Issues in Advanced HD

- Medication use (and when to stop)
- How to assess pain and anxiety in the non-verbal patient
- Preventing purposeful physical violence
- Falls, falls, falls, falls, falls, . . .
- Weight in the patients who no longer walk
- How to keep patients entertained and out of trouble

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Everyday Issues in Advanced HD

- Posture
- Communication with patients
 - Communication devices
 - Non-verbal behavior
- Screaming
- Eating/feeding with swallowing issues and time
- End of life management
 - Hyperthermia and other signs of imminent death
 - Pain
 - Brain donation



Nance & Sanders, 1996

- We believe that the most important challenges to management of HD in the long-term care facility are:
 - Management of severe behavioral disturbances in one third of patients
 - Dietary management, with the goal of increasing weight and prolonging the time that eating is pleasurable to the patient
 - Adjustment by patient, family, and staff to the patient's gradually decreasing ability to walk, talk, eat, sit, or even lie down safely



Small Interventions Can Make Large Differences

- Prepare for advancing disease
 - CPR, feeding tubes, hospitalization, etc.
- Revisit use of medications
- Positioning
- Routine/predictability
- Adjunctive therapies such as speech & language, music, recreational, and art therapies may improve quality of life



Dignity in Advanced Disease

- Permitting 'old self' to be present as much as possible
- Institutionalization vs. at home care
- Freedom vs. safety
- Eating
- Toileting
- Smoking



The 'new' Tewksbury hospital



Medications Can Be Used in Advanced HD for:

- Chorea
 - Neuroleptics, tetrabenazine, amantadine, clonazepam
- Mood
 - Anti-depressants, mood stabilizing agents
- Weight loss
 - Megace, cannabinoids, olanzapine, low dose mirtazipine
- Behavioral dyscontrol
 - Anything that can get into the patient and sedates



Small Things Can Make a Difference



Nance, 2007

An Important “Small” Thing - Advocacy

- Incentive payments for nursing homes
- Specific programming
 - Education through HDSA as a start
 - Ongoing daily activities and therapies focused on HD
- HR 678 - HD Parity Act
- An intervention that everyone can help with

Thank you!