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Medical Management of Advanced HD

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

“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

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
Function & Duration of Life

Time (years)

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

Predictors of Extended Care Placement
- Important conclusion: motor variables alone predicted nursing home placement

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)
- Whee lock, 2003
  - 52 years old
  - Disease duration = 8.6 years
  - 63% women

Medical Issues in Advanced HD
- Behavioral disturbance
- Movements — Including falls
- Dementia
- Delusions & other forms of psychosis
- Swallowing & Nutrition — Weight loss/gain
- Mood & anxiety

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, . . .
- Weight in the patients who no longer walk
- How to keep patients entertained and out of trouble
**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

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**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

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**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 mirtazapine
- Behavioral dyscontrol
  - Anything that can get into the patient and sedates

**Small Things Can Make a Difference**

- The "Tara" Tewksbury hospital
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!