

**FAST FACTS AND CONCEPTS #201  
PALLIATIVE CARE FOR PATIENTS WITH HUNTINGTON'S DISEASE**

**Sean Marks MD, Serena Hung MD, and Drew A Rosielle MD**

**Background** Huntington's disease (HD) is an incurable neurodegenerative disorder inherited in an autosomal dominant fashion. It is characterized by progressive movement disorders, psychiatric manifestations, behavioral abnormalities, and cognitive impairment. This *Fast Fact* will focus on supportive and terminal care for patients with HD and their families.

**Natural History and Prognosis**

- Symptom onset is usually between 33 and 44 years; subtle cognitive and motor changes may precede diagnosis by many years. Mean duration of illness from onset to death is 15-20 years with average age of death of 60 years; there are no proven therapies which slow the progression of HD.
- Patients show signs of progressive dementia and become unable to walk, talk, take in nutrition, and care for themselves. Life threatening complications may result from aspiration, chronic infections, poor nutrition, falls, or cardiovascular disease.
- ~1/3 of all patients with HD are institutionalized in long-term care facilities.

**Impact on Families** HD often begins during a time when family life is most complex and therefore most disruptive to the family structure (e.g. child-rearing, career development). Children can be particularly affected: distress is aggravated by concerns about their own genetic susceptibility, and as many as 40% of children of HD patients describe HD as splitting their family apart. Careful assessments of familial coping and psychosocial needs are an integral part of ongoing care for the HD patient.

**Common Symptoms and Supportive Care** Patients are best served by an interdisciplinary team familiar with caring for patients with HD.

- **Motor Manifestations.** Abnormal involuntary movements include: chorea, dystonia, rigidity, bradykinesia, tremor, and myoclonus; other motor manifestations include gait and balance problems leading to frequent falls, slurred speech and swallowing difficulties
  - First-line strategies are non-pharmacologic and include gait/balance training, speech therapy, and orthotics and leg weights to assist with upright posture.
  - Chorea is the most frequently targeted symptom for pharmacologic therapy. Tetrabenazine (a dopamine depleting agent) has been shown to reduce chorea in a well-designed placebo controlled trial; it is undergoing approval in the US. Haloperidol and other antipsychotics are also used for chorea, although trials evaluating their effectiveness have shown mixed results.
- **Psychiatric manifestations** are present in over half of HD patients.
  - Depression is a significant psychiatric problem and rates of suicide are higher in HD patients than the general population. Case reports support using tricyclic as well as newer antidepressants.
  - Agitation is also common, and a small number of patients develop psychosis. Atypical neuroleptics are commonly used. Emotional lability (including episodes of extreme anger) can respond to propranolol.
  - Low doses of scheduled benzodiazepines before meals or propranolol are used to control motor manifestations and anxiety related to eating.
  - Establishing strict daily and hourly routines can help lessen anxiety, short-term memory deficits, intrusive thoughts, and fear of abandonment.
- **Cognitive Deficits:** gradual loss of memory and executive function are common. Consequently, increasing impairments in initiating movements and conversation occur. Yes/no questions may be preferable over open-ended questions when cognitive impairments become severe.

**Advance Care Planning** Advanced care planning should be performed as early as possible, prior to cognitive impairment. Of particular importance is establishing a health care power of attorney, as well as documenting guidance to families for likely decisions they will face (such as tube feeding and mechanical ventilation). Some states require clear evidence that a patient would want tube feeding withheld or

withdrawn at the end of life and patients should be instructed to document this if consistent with their wishes. See also *Fast Facts* #12, 65, 162, and 178 for further discussion of advance care planning.

**Terminal Care** There are no evidence-based criteria for determining a 6 month prognosis in HD; web-based reference 13, however, provides some guidance regarding hospice eligibility. A retrospective, multi-centered study of over 100 HD patients, suggested that the hospice length of stay is longer than non-HD hospice patients and only a significant minority of HD patients are able to die at home. Labored breathing, excessive secretions, and restlessness are common terminal symptoms – see *Fast Facts* #1, 60, 109, 158, and 176.

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**Version History:** Originally published April 2008. Version re-copy-edited in June 2009; then again July 2015 – reference #15 added and incorporated into text, web resources updated.

**Fast Facts and Concepts** are edited by Sean Marks MD (Medical College of Wisconsin) and associate editor Drew A Rosielle MD (University of Minnesota Medical School), with the generous support of a volunteer peer-review editorial board, and are made available online by the [Palliative Care Network of Wisconsin](#) (PCNOW); the authors of each individual *Fast Fact* are solely responsible for that *Fast Fact's* content. The full set of *Fast Facts* are available at [Palliative Care Network of Wisconsin](#) with contact information, and how to reference *Fast Facts*.

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