

Comprehensive care in Huntington's disease A physician's perspective

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Abstract

Huntington's disease is a slowly progressive neurodegenerative disorder with wide-ranging effects on affected individuals and their families. Until a cure is found for the disease, patients and their families will continue to need care over years, even generations. The ideal care for HD is provided by a team, led by a physician, with input from rehabilitation therapists, nurses, psychologists, genetic counselors, social workers, and other health care providers. The goals of care are to maximize the quality of life at all points through the course of the disease, in part by anticipating problems that are likely to arise at the next stage of the illness. We describe below an approach to comprehensive care, and introduce the concept of the "Huntington disease molecule", in which the patient, in the center, is surrounded by a shell of immediate and extended family members, with bonds extended in multiple directions to provider who can give appropriate medical care, education, crisis management, research opportunities, address family issues, maximize function, and prepare for the future.

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1. Introduction

Research on the clinical management of people with HD and their families lags far behind basic research into disease mechanisms and potential pharmacotherapies, for many reasons. A comprehensive review of HD management is beyond the scope of this presentation; we focus on the concept of comprehensive care for this neurodegenerative disease. We present below a model for comprehensive care of HD, and make recommendations for future research.

The care-defining characteristics of Huntington's disease include the following:

1. It is a neurologic disorder, often classified as a movement disorder, suggesting a parallel or similarity to other movement disorders such as Parkinson's disease, multiple systems atrophies, and the ataxias. It can also be characterized as a dementing disorder, appropriately implying an analogy to Alzheimer's disease—although the differences between the clinical manifestations of two dementias are often emphasized more than their similarities.
2. It is a psychiatric disorder. There is no specific psychiatric symptom or complex of symptoms that characterizes HD; rather, a broad range of psychological effects are seen, with symptoms ranging from anxiety and depression to obsessive–compulsive behaviors, substance abuse, paranoid or suspicious thinking, or overt hallucination or schizophrenic-like manifestations.
3. It is a fatal neurodegenerative disorder. Symptom severity and functional disability increase over time, and the treating health team, patient, and family should all be prepared for the inevitable changes—including, at the end, death.
4. HD is a chronic disease. Progression is measured in years, or even decades, not in days, weeks, or months. In the ideal setting, the HD family will have a long-standing relationship with the health care team, perhaps even measured over generations.
5. HD is a dominantly inherited genetic condition. In many ways, the unit of care in genetic medicine is the family rather than the patient, as siblings and children of the affected person are often not just caregivers, but potential patients because of their at-risk status. Comprehensive care of HD requires management of these at-risk individuals as well as care for affected individuals. Involvement of at-risk family members

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in clinical research also provides a unique opportunity to understand the earliest stages of the disease.

6. HD is usually an adult-onset disease, with an average age of death in the mid-50s [1]. HD care is usually provided by health teams specializing in adult care. The subsets of patients with juvenile-onset HD have special, often poorly met needs, as practitioners with expertise in both pediatrics and HD are rare.
7. Thankfully, HD is not a multi-organ disease. Care can be coordinated throughout the course by a physician specializing in nervous and mental diseases, and medications can be used without special concern about renal or hepatic function. Cardiac, pulmonary, or endocrine complications are not expected. In the absence of other unrelated medical conditions, morbidity and death in HD are due to a relatively predictable set of complications of end-stage neurologic disease.

With these care-defining features in mind, we can review the three goals and the main objective of treatment in HD: the goals are to reduce the burden of symptoms, maximize function, and to eliminate unnecessary “surprises” as affected individuals pass expected disease milestones. Pursuit of these goals contributes to the primary objective, which is to improve the quality of life for people with HD and their families. Although we all look forward to the time when treatments slow, prolong, or reverse the course of the disease, the treatments of today are only of value insofar as they improve a patient’s quality of life. Pursuit of even this humble goal is challenging; it is both an art and a science to provide the education, support, and medical care that patients and families need throughout the changing course of a multi-year illness.

HD lends itself very well to multidisciplinary, team-based care. The ideal team would be led by a neurologist or psychiatrist specializing in HD, but would also include a general practitioner (particularly important in the later stages of the illness, when medical complications are likely to occur), and a dentist. A registered nurse or nurse practitioner can serve as clinic coordinator or case manager [3], and a research nurse can help to recruit for, coordinate, and perform clinical research. Allied health professionals important to an HD patient or family include: psychologist, neuropsychologist, physical therapist, occupational therapist, speech therapist, dietitian, social worker, and genetic counselor. We have found it helpful to have a chaplain and a lay group liaison attending our HD clinics as well.

Some HD specialty programs are based in residential care facilities. In such a setting, a non-neurologist might direct the clinical care, while additional therapies (music therapy, recreational therapy) might be added.

Many HD families do not have access to skilled HD care teams. Because of the dearth of evidence-based research publications supporting the use of one or another treatment, nonspecialists lack direction as they try to care for patients with this uncommon disease, and both patient and physician can become frustrated and negativistic. Past consensus meetings of HD clinical experts have ascertained the common practice among HD experts in behavioral and psychiatric management

and end-of-life care, and to identify areas for further research. Additional similar efforts seem warranted in the future, with improved dissemination of recommendations in places accessible to nonspecialists.

2. Care through the stages of HD

2.1. The stages of HD

The Shoulson–Fahn functional capacity scale was created for use in HD research, but because it measures functional skills, it is also very useful in the clinic [2]. This scale assigns points based on a patient’s ability to work, to manage money, to do household chores, to perform self-cares (“activities of daily living”), and whether the person is able to live at home or requires institutional care. Possible scores range from 0 to 13, and functional capacity is divided into five stages based on the score. In the clinic, the patient’s care needs can be predicted to a certain extent by the stage, with stage 3 representing the transition time between the early stages, when a patient can function independently, and stages 4 and 5, when the person needs full-time care. There are some limitations to the applicability of the scale: it cannot be used in “presymptomatic” individuals, individuals with severe psychiatric symptoms may function poorly and thus appear to be in a later stage of the disease than they really are, it does not apply to juvenile HD, and it has a floor effect. Patients may live for a number of years in stages 4 and 5, with clear progression during that time that is not measured by this scale [4]. Despite these minor limitations, the scale is very helpful to the clinician.

2.2. The role of the physician

In the narrowest view, the role of the physician is to identify patient symptoms for which there are medical treatments, and to write prescriptions for the appropriate medications. Most physicians assume a broader role, in which they also make appropriate referrals to allied health professionals such as therapists, social workers, and genetic counselors, when they recognize a symptom or a need that cannot be addressed by medication alone. In the HD specialty clinic, the physician often also serves as a team leader, an educator, an advocate and spokesperson, and a researcher.

The physician and the HD team have two additional duties that may be difficult to address adequately even in HD specialty centers. The first is to recognize and address issues that impact on the family or caregiver, and the second is to manage the disease in a proactive manner, so that both patient and family anticipate disease milestones and address them in a timely fashion, thus eliminating crisis and surprise from the disease course. Limitations of time, space, reimbursement, and privacy, may prevent the physician from seeing family members separately from the patient, from sharing with the patient concerns raised by the family, or ensuring that family members are appropriately attending to their own health. In addition, the patient, family, or physician may be struggling enough to manage the current situation that discussion and planning for the future never happen. Finally, the challenges of navigating the layers of patient or care-

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