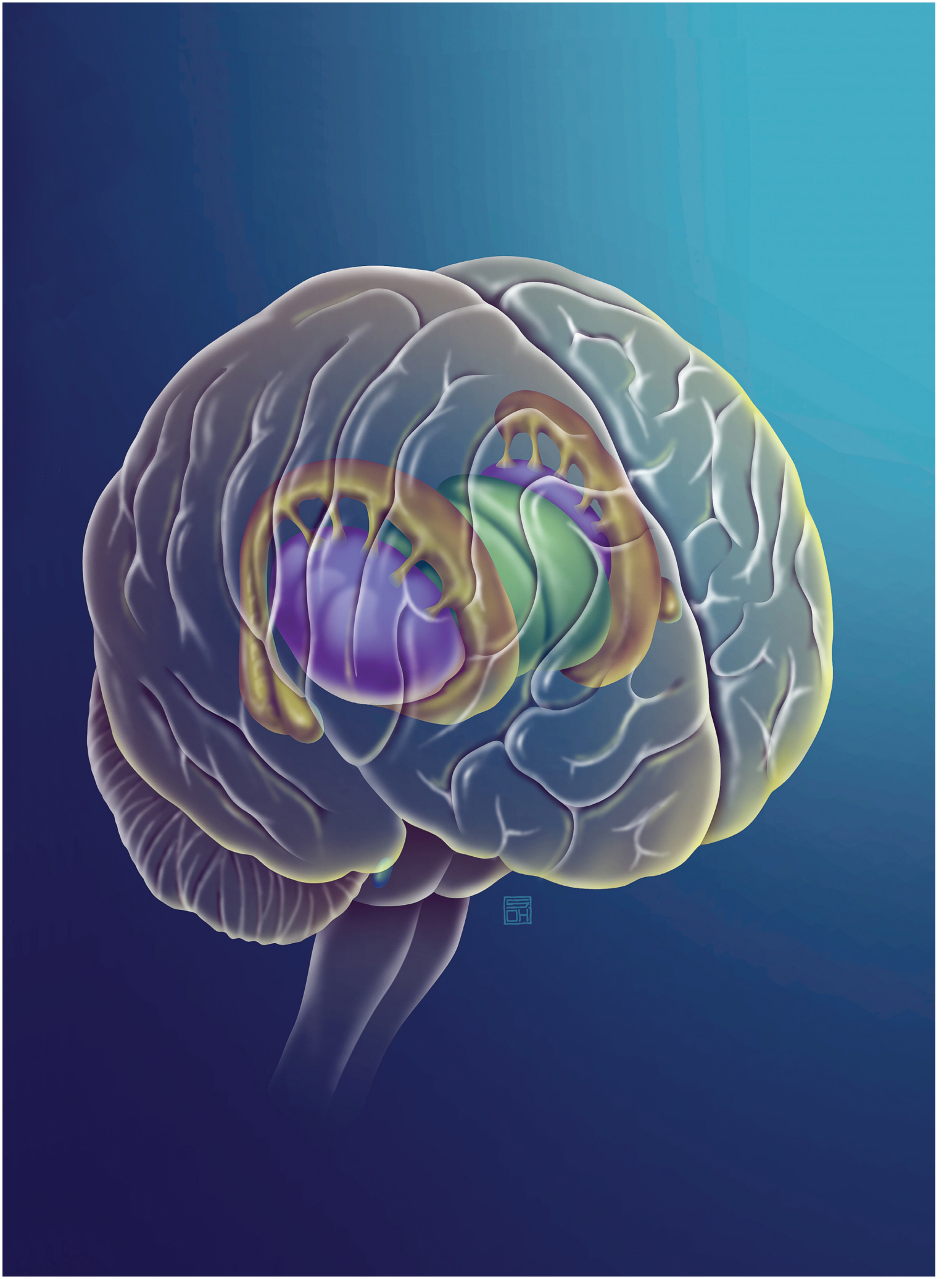


CE

1.0
ANCC
CONTACT HOUR

Adult-onset Huntington disease

An update

BY NANCY L. URRUTIA, EdD, MSN, RN, CNE

Abstract: Appropriate nursing-care strategies depend on the early recognition of Huntington disease (HD) to prioritize a plan of care. This article offers perspective on the clinical presentation, prognosis, diagnosis, and management of adult-onset HD.

Keywords: autosomal dominant disease, chorea, dystonia, Huntington disease, movement disorder, neurodegenerative disorder

BD, 42, WAS ADMITTED to the ICU from the ED for injuries sustained from a suicide attempt. Emergency medical technicians reported that he had jumped from a highway overpass into traffic. BD's mother was notified and informed the ED staff that BD had been diagnosed with Huntington disease (HD) at age 30. Recently, he had begun experiencing psychiatric symptoms of HD such as anxiety and hallucinations, but he had made no previous suicide attempts.

BD reported hearing voices that had encouraged him to jump from the overpass. Although he had been able to separate the voices from reality in the past, he had been unable to recognize the risk during this episode.

The fall fractured his pelvis and right femur. After a short stay in the ICU, BD was transferred to an ortho-

STEVE GH / MEDICAL IMAGES

pedic unit, underwent surgery to repair the fractures, and remained hospitalized until he was stable enough for discharge. Additionally, he received treatment for his psychiatric and HD signs and symptoms during his hospital stay, including pharmacologic and movement therapy.

Approximately 1 in every 10,000 people in the US has HD, which equates to about 30,000 patients.[1] It is the most common hereditary neurodegenerative disorder globally, with roughly 700,000 cases worldwide.[2] The global figure may underestimate its prevalence, as a recent screening of healthy individuals demonstrated that 1 in 400 may be at risk.[2] HD is most common among individuals of European descent, but it may affect those of Asian and African descent as well.[3] This article offers an overview of adult-onset HD, including risk factors, prognosis, diagnostic criteria, management, and nursing care.

Genetics

HD is an autosomal dominant inherited disease, meaning that only one copy of a defective huntingtin (HTT) gene is required to cause disease.[4] An inherited abnormal gene, even from just one parent, puts individuals at risk for the disease: An adult with HD has a 50% chance of transmitting it to each of his or her offspring.[5,6] Consequently, the rate of neurodegenerative diseases such as HD is estimated to double every 20 years, creating continued challenges for healthcare providers.[7]

HTT influences the production of the huntingtin protein. In patients with HD, the huntingtin protein is elongated and fragmented into toxic, binding pieces that disrupt cellular function.[4,8] The gene is characterized by the cytosine-adenine-guanine (CAG) trinucleotide repeat, in which the CAG sequence is repeated many times within an individual's DNA.[4,8] The more occurrences of the CAG trinucleotide repeat, the earlier the onset of HD.[4]

HD may occur when the number of CAG trinucleotide repeats is greater than or equal to 36. Typically, patients with more than 40 CAG trinucleotide repeats will experience HD onset, while individuals with 36 to 39 occurrences of the CAG trinucleotide repeat may or may not develop HD.[8] Those with 27 to 35 CAG trinucleotide repeats will not experience HD, but they may risk passing the disease along to their children.[4]

Pathophysiology

HD leads to progressive neuronal loss, specifically in the basal ganglia, which are interconnected with the cerebral cortex, thalamus, brainstem, and several other areas of the brain. These cells are responsible for processing messages communicated to the brainstem. Basal ganglia play an important role in the actions required to achieve goals, complete daily activities, and learn new actions.[9] When these cells degrade, movement and cognition are compromised.

HD may also cause a loss of brain tissue volume with marked gray and white matter atrophy. Atrophy in the frontal lobes accounts for many of the characteristic neurodegenerative signs and symptoms.[10]

Prognosis

HD is a progressive, insidious, and incurable disorder that leads to significant disability and shortens life expectancy.[11] On average, mortality occurs between 10 and 20 years after the onset of HD, but some patients may live up to 40 years.[12] Women typically survive longer than men.[11,12] Factors related to mortality may include aspiration pneumonia or infections due to immobility.[12]

Clinical manifestations

HD is a neurodegenerative neuropsychiatric disorder that is characterized by chorea, dementia, and psychiatric symptoms (see *Coming to terms with HD*).[10,13,14] Signs and symptoms typically begin between ages 30 and 50 and progress over the next 10 to 25 years, but the disease may emerge earlier or later.[10,15] Characteristics of adult-onset HD, such as physical and mental decline, are highly individualized.

- *Psychiatric symptoms*. Patients typically exhibit psychiatric impairment before motor and cognitive decline, with depression and anxiety being the most common manifestations.[14] Other common psychiatric symptoms include irritability and apathy, but obsessive-compulsive behavior and psychosis may also present in patients with HD.[12] Additionally, this patient population is at increased risk for suicide, with a 7% suicide rate and approximately a 20% rate of suicidal ideation.[12]
- *Motor signs and symptoms*. Early motor deficits may include subtle

Coming to terms with HD

Ataxia	Characterized by a reduction in muscle control or coordination in voluntary movements; often a sign of an underlying disorder.[19]
Bradykinesia	"Slowness of movement" characterized as difficulty actuating and following through with a movement.[52,53]
Chorea	Brief, abrupt, involuntary, nonstereotyped movements involving the face, trunk, and limbs.[12]
Dyskinesia	Described as fluid or dance-like; characterized as "involuntary, erratic, writhing movements of the face, arms, legs or trunk."[54]
Dystonia	Prolonged, sustained, abnormal postures.[12]

changes in handwriting and speech or a tendency to drop items. Patients may also experience changes in posture and gait, often progressing to bradykinesia, difficulty with eye-hand coordination, and restlessness.[10]

HD was previously known as Huntington chorea because chorea is a key feature and defining symptom at the time of diagnosis.[12] Chorea presents as brief, abrupt, involuntary, nonstereotyped movements involving the face, trunk, and limbs.[12] It may present as abnormal, involuntary movements in speech, gait, postural stability, and manual dexterity.[16] These include milkmaid grip, which is characterized by the involuntary squeezing and relaxing of a patient's grip; start hesitation when walking, which is characterized by stepping in place before moving; and ataxia, which is characterized by a reduction in muscle control or coordination in voluntary movements.[10,17-19]

Due to lack of coordination, chorea causes difficulty in activities of daily living. In later stages, chorea also affects the diaphragm, pharynx, and larynx, producing dysarthria, dysphagia, and involuntary vocalizations.[12] Dystonia (prolonged, sustained, abnormal postures) may be seen in the hands with such activities as walking.[12]

- *Eye movements.* Muscle dysfunction can lead to abnormal eye movements and disturbances in balance, placing patients at risk for falls.[1]
- *Cognitive impairment and dementia.* The patient may also experience cognitive impairment, with reduced recognition memory and self-awareness. Patients may not be able to communicate hunger, thirst, and the need to toilet. If not anticipated by a caregiver, a patient's basic needs may not be met, leading to malnutrition, dehydration, and other complications.

Cognitive impairments may manifest in various behaviors, including difficulty organizing, prioritizing, or focusing thoughts. Memory loss complicates the ability to retain information, so learning can be a challenge for these patients.[10] For those in the earlier stages of HD, these deficits can impair the ability to keep a job and maintain financial independence. Slow mental processing regarding the use of language and recall further complicates communication; for example, the loss of executive function, defined as difficulty with initiating and carrying out plans, and loss of verbal fluency.[10]

Irrational thought processes are also common, including poor impulse control and a lack of awareness of the impact of personal behaviors on others.[1,10]

Clinical progression

HD affects patients in a gradual decline, but they may fall into stages depending on their levels of motor and cognitive function. The transition of early-, middle-, and late-stage HD is characterized by progressive deterioration. Each stage features increasing signs and symptoms that patients may experience over the course of the disease.[12]

Overall, patients with early-stage HD can still function and maintain their independence despite the manifestation of mild symptoms. Middle-stage patients typically see a further decline in motor functions and cognition and may require caregiver assistance in daily life. Late-stage HD is characterized by increased morbidity and mortality. Patients require constant care as they experience further motor and cognitive decline. They may also have difficulty with speech and swallowing.[12]

Diagnosis

HD is typically preceded by a prodromal phase characterized by subtle cognitive and motor signs, which may be challenging to diagnose.[11] Because HD is a hereditary disorder, family history is especially important and may reveal premature mortality or mental illness related to HD in earlier generations. Predictive (presymptomatic) genetic testing is available for patients over age 18 with a confirmed case of HD in the family and administered with a corresponding neurologic exam.[20,21]

Currently, a definitive diagnosis for HD is achieved with gene mapping using a blood sample. Upon exam, an expansion of the CAG trinucleotide repeat in the HTT gene may indicate HD.[4,22]

Additional lab tests and imaging studies may be used to rule out other disorders.[10] These include thyroid profiles to identify hypo- or hyperthyroidism, which can negatively affect mood and metabolism. MRI or computed tomography of the brain may be used to rule out other pathology associated with cognitive changes, such as stroke or brain tumors. Other promising diagnostic approaches are under investigation (see *Additional research*).

The Unified Huntington's Disease Rating Scale is a comprehensive and reliable instrument to assess motor function, cognitive function, behavioral abnormalities, and functional capacity.[23]

Additional research[22,55]

Research continues to be a focus in understanding HD and finding alternative methods of testing. At this time, breath testing is not available, but a promising noninvasive method that tests for specific compounds in a patient's exhaled breath is underway. Potential biomarkers in metabolites, such as amino acids, lipids, and neurotransmitters, are also being researched. These are significantly altered in patients with HD, and testing would allow for early diagnosis and treatment.

Psychosocial impact

After receiving a definitive diagnosis, many patients with HD experience feelings of fear and inevitability, which can impact health management and decision-making.[20] Apathy, irritability, and depression may be persistent and difficult to treat despite interventions.[24] Frequent emotional outbursts and mood alterations can lead to family dysfunction and broken relationships. Strategies to cope with an HD diagnosis are highly individualized and depend on several factors, including the patient's level of social support, resources, and coping mechanisms.[20]

Although the HD diagnosis itself may impact a patient's psychological well-being, depression is also linked to brain injury and declining brain function as part of the disease process. Patients may exhibit withdrawal, insomnia, lethargy, and suicidal ideation as part of brain dysfunction.[1,25-27]

Weight gain or weight loss commonly accompanies these psychiatric and mood disorders: Patients may also abuse alcohol or drugs.[10]

Management

No cure or disease-modifying treatment is currently available for patients with HD. Therapy is focused on symptom management and supportive care to optimize quality of life.[28] Treatment cannot prevent physical, mental, and behavioral decline, but it may improve patient perceptions of well-being and empowerment as the disease progresses. Therapies available to manage the clinical manifestations of HD include pharmacologic therapy and nonpharmacologic interventions, including movement and psychotherapy.

Pharmacotherapy. Patients with HD require a multidisciplinary approach to medication management due to the disease's impact on movement, cognition, and mood. In addition, certain medications increase the risk of depression or suicidal ideation. Because patients are already at risk for self-injury during disease progression, caregivers must closely monitor their response to drug therapy. Pharmacologic therapy may include drugs to treat chorea, other movement disorders (including dystonia, bradykinesia and rigidity, tics, and myoclonus), psychosis and agitation, and depression (see *Pharmacologic therapy for chorea in HD*).

Movement therapy. Prescribed exercises can help patients manage the physical signs and symptoms of HD. For example, gait may be enhanced with structured rehabilitation.[29] As for healthy individuals, endurance training has been identified as having a positive impact on mitochondrial function in skeletal muscle.[30] Cellular and molecular activities contribute to the pathology of HD. Mitochondria are preeminent in the development of neurodegenerative disorders such as HD. Mutated HTT may have an impact on mitochondrial function.[31]

A 2018 study examined the value of dance therapy in improving gait, mobility, and balance in patients with neurologic disorders including HD. The study concluded that emerging evidence supports the use of dance as a feasible intervention for adults with neurologic disorders.[32]

Although no studies have concluded that therapy will prevent or delay motor complications in patients with movement disorders, movement therapy may be a valuable addition to the overall plan of care.[33] Prescribed movement therapy that focuses on quality of life and the physical, psychosocial, and spiritual problems caused by HD can help patients feel actively involved in their daily care. It may also help to delay long-term-care facility placement and maintain independent living arrangements.[34]

Psychotherapy. Neurodegenerative diseases affect the brain regions that regulate emotion. As HD progresses and motor signs become apparent, emotion recognition deficits become more pronounced.[35,36] This is characterized by difficulty understanding the emotions being conveyed in facial expressions or tone of voice.[37] Research to support the efficacy of psychotherapy delivered by trained therapists, such as cognitive and behavioral psychotherapies, is limited, but many caregivers recognize the importance of caring for their patients' emotional needs, regardless of HD stage.[38]

Pharmacologic therapy for chorea in HD[28]

Drug	Classification	Adverse reactions
Tetrabenazine Deutetrabenazine	Central monoamine-depleting agents; vesicular monoamine transporter 2 (VMAT2) inhibitors	Sedation, akathisia, parkinsonism, depression, suicidal thoughts and behaviors
Aripiprazole Risperidone Olanzapine	Atypical neuroleptics	Weight gain, hypotension, sedation, anticholinergic symptoms, hyperprolactinemia, extrapyramidal symptoms, cardiomyopathies, cataracts, sexual dysfunction
Haloperidol Fluphenazine	Typical neuroleptics	Extrapyramidal symptoms, tardive dyskinesia, hyperprolactinemia, neuroleptic malignant syndrome, QT prolongation, sudden death

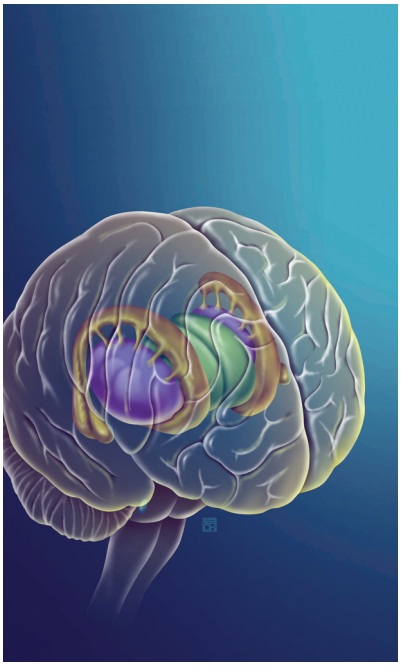
Nursing considerations during hospitalization

Patients with HD may be hospitalized for complications related to HD or for an unrelated acute care concern. As a plan of care is established, assessing patient needs is the nurse's primary role. Anticipating these needs through a team approach is an important strategy throughout a patient's stay.

Nutrition. Weight loss is common, particularly in late-stage HD. This may be due to metabolic dysfunction, eating and swallowing difficulties, loss of fine motor coordination, anorexia, or cognitive impairments. While the causes of weight loss are not entirely clear, the loss of metabolically active tissue may be a factor.[39]

Aspiration risk is an important consideration for patients taking oral nutrition and fluids, especially in later stages of the disease. Managing consistencies of food and liquids can be effective for safe swallowing in patients with HD.[40] Working closely with the family and the healthcare team, the nurse can help identify calorie-dense and safely swallowed meals and supplements for patients with HD in acute care settings. Nurses should consult speech-language pathologists to assess swallowing capabilities, and a consult with a dietitian may also be a resource.

Sleep hygiene. The acute care environment is not conducive to recuperative sleep, and disturbances affect many hospitalized patients. Sleep-wake disruption is one of the earliest reported symptoms of many neurodegenerative disorders.[41] Insomnia and circadian dysregulation are both associated with early-stage HD and occur throughout disease progression.[41] Even when patients are in familiar environments, sleep disturbances such as delayed sleep onset, frequent bedtime awakenings, and fatigue during the day are common.[32,42] These may become more pronounced when a patient with HD is admitted to the hospital.



As HD progresses and motor signs become apparent, emotion recognition deficits become more pronounced.

Traditional strategies for enhanced sleep in hospitalized patients are effective for patients with HD.[43] Proactive planning to reduce sleep interruptions, maintain routines, and limit noise and light help patients achieve more rest. Adjusting patient feeding and medication administration times may also be beneficial to maintain circadian rhythms and allow recuperative sleep.[44] Improved sleep quality can positively impact cognitive impairment in patients with HD, as well as easing anxiety, depression, and irritability.[42]

Elimination. Difficulty with elimination, which is often compromised by neurodegenerative disorders, may significantly reduce quality of life in patients with HD.[45] Bladder overactivity, urinary incontinence, and incomplete bladder emptying are common problems.[45] In addition, decreased anal sphincter tone and voluntary activation, lower sacral sensory thresholds, and decreased awareness of sensations are also common.[46] These changes, which reflect the degeneration of the central nervous system, typically worsen over time and may lead to fecal incontinence or constipation.

Supporting bladder and bowel function are nursing priorities for patients with HD. Depending on disease progression, patients may need help maintaining hygiene and recognizing occurrences of incontinence. Nursing interventions include prompt attention to elimination needs and maintaining skin integrity.

Safety. For patients with HD attempting to navigate environmental obstacles, ambulation may be complicated by poor balance and ataxia.[47] A decline in cognitive function and mobility lead to problems with gait, balance, and mobility.[48]

Nurses should initiate fall precautions per hospital policy. Interventions designed to improve mobility should address issues of balance and coordination, as well as cognitive impairment, specifically interventions that focus on training associated with movement and judgment.[48] Optimal functioning according to level of disease, safe use of assistive devices, and participation in self-care should be goals in nursing care.[10] Additionally, the nursing staff should focus on frequent rounding, alarms, low bed positioning, room placement near high-traffic areas, and bedside mats and padding to minimize patient injuries.

Family. Neurologic disorders are associated with chronic disability and increased need for patient care.[49] Family caregivers are deeply involved; provide substantial informal care, support, and advocacy at the bedside; and should be consulted

about treatment decisions, as desired by the patient.[50] Nurses should recognize family members in these important roles and seek their input.[51]

Collaboration between family and healthcare professionals can be a challenge, but it should be included in individualized patient care.[5] Connecting multidisciplinary teams, developing treatment guidelines, and involving families in quality improvements can offer healthcare delivery that meets individual needs.[50] Engaging both patients and their families in high-quality, compassionate care improves the lives of patients with HD.

Additionally, patients and families may require support from home care services, case management, counseling, or even legal interventions to manage the patient's needs as the disease progresses. Nurses should assess patients and their family dynamics for possible coping issues, neglect, or abuse.

Meeting challenges

BD's hospitalization was managed with the coordination of healthcare team services and family input. Once his orthopedic injuries began to mend, he was transitioned to a long-term-care setting near the family's home so BD's mother could continue to participate in his care. The acute care team's strategies to manage his HD symptoms through nutrition services, occupational and physical therapy, and medication therapy for his psychiatric symptoms helped to update his plan of care as his disease progression accelerated. Additionally, the family received ongoing case management and help from specialists, including physical therapists, to facilitate BD's return home.

Effective care for those affected by HD is achieved when realistic goals for improved quality of life and optimized functional status are set.[50] Acute care settings create challenges for patients with HD. The multidisciplinary team and the patient's family are valuable assets in managing HD.

Nursing strategies rely on early recognition of HD to prioritize the plan of care effectively. Nurses must be knowledgeable about HD and its progression to provide individualized and compassionate care. ■

REFERENCES

1. Mayo Clinic. Huntington's disease. 2017. www.mayoclinic.org/diseases-conditions/huntingtons-disease/symptoms-causes/syc-20356117.

2. Critchley BJ, Isalan M, Mielcarek M. Neurocardio mechanisms in Huntington's disease and other neurodegenerative disorders. *Front Physiol*. 2018;9:559.

3. Genetics Home Reference. Huntington disease: frequency. https://ghr.nlm.nih.gov/condition/huntington-disease#statistics.

4. Genetics Home Reference. Huntington disease: inheritance pattern. https://ghr.nlm.nih.gov/condition/huntington-disease#inheritance.

5. Røthing M, Malterud K, Frich JC. Family caregivers' views on coordination of care in Huntington's disease: a qualitative study. *Scand J Caring Sci*. 2015;29(4):803-809.

6. Huntington's Disease Society of America. Overview of Huntington's disease. 2019. https://hdsa.org/what-is-hd/overview-of-huntingtons-disease.

7. Dupont AC, Largeau B, Guilloteau D, Santiago Ribeiro MJ, Arlicot N. The place of PET to assess new therapeutic effectiveness in neurodegenerative diseases. *Contrast Media Mol Imaging*. 2018;2018:7043578.

8. Genetics Home Reference. Huntington disease: causes. https://ghr.nlm.nih.gov/condition/huntington-disease#genes.

9. Lanciego JL, Luquin N, Obeso JA. Functional neuroanatomy of the basal ganglia. *Cold Spring Harb Perspect Med*. 2012;2(12):a009621.

10. Bader MK, Littlejohns LR, Olson DM, eds. *AANN Core Curriculum for Neuroscience Nursing*. 6th ed. Chicago, IL: American Association of Neuroscience Nurses; 2016.

11. Sipilä JOT, Kauko T, Päivärinta M, Majamaa K. Comparison of mid-age-onset and late-onset Huntington's disease in Finnish patients. *J Neurol*. 2017;264(10):2095-2100.

12. Suchowersky O. Huntington disease: clinical features and diagnosis. UpToDate. 2018. www.uptodate.com.

13. Ahveninen LM, Stout JC, Georgiou-Karistianis N, Lorenzetti V, Glikmann-Johnston Y. Reduced amygdala volumes are related to motor and cognitive signs in Huntington's disease: the IMAGE-HD study. *Neuroimage Clin*. 2018;18:881-887.

14. Nogueira JM, Franco AM, Mendes S, Valadas A, Semedo C, Jesus G. Huntington's disease in a patient misdiagnosed as conversion disorder. *Case Rep Psychiatry*. 2018;2018:3915657.

15. Zarowitz BJ, O'Shea T, Nance M. Clinical, demographic, and pharmacologic features of nursing home residents with Huntington's disease. *J Am Med Dir Assoc*. 2014;15(6):423-428.

16. Mestre TA, Forjaz MJ, Mahlknecht P, et al. Rating scales for motor symptoms and signs in Huntington's disease: critique and recommendations. *Mov Disord Clin Pract*. 2018;5(2):111-117.

17. Scott BL. Huntington disease: chorea and other motor manifestations. Medscape. 2012. www.medscape.org/viewarticle/759293_2.

18. Ronthal M. Neurologic gait disorders of elderly people. UpToDate. 2018. www.uptodate.com.

19. Mayo Clinic. Ataxia. 2018. www.mayoclinic.org/diseases-conditions/ataxia/symptoms-causes/syc-20355652.

20. Hagen N. The lived experience of Huntington's disease: a phenomenological perspective on genes, the body and the lived experience of a genetic disease. *Health (London)*. 2018;22(1):72-86.

21. National Society of Genetic Counselors. Huntington's disease and predictive genetic testing. www.aboutgeneticcounselors.com/Genetic-Conditions/Huntingtons-Disease-and-Predictive-Genetic-Testing.

22. Graham SF, Pan X, Yilmaz A, et al. Targeted biochemical profiling of brain from Huntington's disease patients reveals novel metabolic pathways of interest. *Biochim Biophys Acta Mol Basis Dis*. 2018;1864(7):2430-2437.

23. Unified Huntington's Disease Rating Scale. 1999. www.neuropsicol.org/Protocol/Uhdrs.pdf.

24. Ruiz-Idiago JM, Floriach M, Mareca C, et al. Spanish validation of the problem behaviors assessment-short (PBA-s) for Huntington's disease. *J Neuropsychiatry Clin Neurosci*. 2017;29(1):31-38.

25. Brandt J. Behavioral changes in Huntington disease. *Cogn Behav Neurol*. 2018;31(1):26-35.

26. Regan L, Preston NJ, Eccles FJR, Simpson J. The views of adults with Huntington's disease on assisted dying: a qualitative exploration. *Palliat Med*. 2018;32(4):708-715.

27. Roman OC, Stovall J, Claassen DO. Perseveration and suicide in Huntington's disease. *J Huntingtons Dis*. 2018;7(2):185-187.

28. Suchowersky O. Huntington disease: management. UpToDate. 2018. www.uptodate.com.

29. Mirek E, Filip M, Chwała W, et al. The influence of motor ability rehabilitation on temporal-spatial parameters of gait in Huntington's disease patients on the basis of a three-dimensional motion analysis system: an experimental trial. *Neurol Neurochir Pol*. 2018;52(5):575-580.

30. Mueller SM, Gehrig SM, Petersen JA, et al. Effects of endurance training on skeletal muscle mitochondrial function in Huntington disease patients. *Orphanet J Rare Dis*. 2017;12(1):184.

31. Jodeiri Farshbaf M, Ghaedi K. Huntington's disease and mitochondria. *Neurotox Res*. 2017;32(3):518-529.

32. Patterson KK, Wong JS, Prout EC, Brooks D. Dance for the rehabilitation of balance and gait in adults with neurological conditions other than Parkinson's disease: a systematic review. *Heliyon*. 2018;4(3):e00584.

33. Fox SH, Katzenschlager R, Lim SY, et al. International Parkinson and Movement Disorder Society evidence-based medicine review: update on treatments for the motor symptoms of Parkinson's disease. *Mov Disord*. 2018;33(8):1248-1266.

34. Mestre TA, Shannon K. Huntington disease care: from the past to the present, to the future. *Parkinsonism Relat Disord*. 2017;44:114-118.

35. Dogan I, Saß C, Mirzazade S, et al. Neural correlates of impaired emotion processing in manifest Huntington's disease. *Soc Cogn Affect Neurosci*. 2014;9(5):671-680.

36. Löffler LA, Radke S, Morawetz C, Derntl B. Emotional dysfunctions in neurodegenerative diseases. *J Comp Neurol.* 2016;524(8):1727-1743.

37. Corcoran CM, Keilp JG, Kayser J, et al. Emotion recognition deficits as predictors of transition in individuals at clinical high risk for schizophrenia: a neurodevelopmental perspective. *Psychol Med.* 2015;45(14):2959-2973.

38. Zarotti N, Simpson J, Fletcher I, Squitieri F, Migliore S. Exploring emotion regulation and emotion recognition in people with presymptomatic Huntington's disease: the role of emotional awareness. *Neuropsychologia.* 2018;112:1-9.

39. Süssmuth SD, Müller VM, Geitner C, et al. Fat-free mass and its predictors in Huntington's disease. *J Neurol.* 2015;262(6):1533-1540.

40. Alves TC, Cola PC, Santos RR, Motonaga SM, Silva RG. Swallowing endoscopy findings in Huntington's disease: a case report. *Codas.* 2016; 28(4):486-488.

41. Abbott SM, Videnovic A. Chronic sleep disturbance and neural injury: links to neurodegenerative disease. *Nat Sci Sleep.* 2016;8:55-61.

42. Bellosta Diago E, Pérez Pérez J, Santos Lasaosa S, et al. Circadian rhythm and autonomic dysfunction in presymptomatic and early Huntington's disease. *Parkinsonism Relat Disord.* 2017;44:95-100.

43. Menear A, Elliott R, M Aitken L, Lal S, McKinley S. Repeated sleep-quality assessment and use of sleep-promoting interventions in ICU. *Nurs Crit Care.* 2017;22(6):348-354.

44. Wang HB, Loh DH, Whittaker DS, Cutler T, Howland D, Colwell CS. Time-restricted feeding improves circadian dysfunction as well as motor symptoms in the Q175 mouse model of Huntington's disease. *eNeuro.* 2018;5(1). pii: ENEURO.0431-17.2017.

45. Kolenc M, Moharić M, Kobal J, Podnar S. Bladder dysfunction in presymptomatic gene carriers and patients with Huntington's disease. *J Neurol.* 2014;261(12):2360-2369.

46. Kolenc M, Kobal J, Podnar S. No electrophysiological evidence for Onuf's nucleus degeneration causing bladder and bowel symptoms in Huntington's disease patients. *Neurourol Urodyn.* 2014;33(5):524-530.

47. Anwar N, Labuschagne I, Simpson K, Smith L, Georgiou-Karistianis N. Impairments in spatiotemporal gait adaptation during obstacle navigation in Huntington's disease. *Neurorehabil Neural Repair.* 2017;31(10-11):934-943.

48. Kloos AD, Kegelmeyer DA, Fritz NE, Daley AM, Young GS, Kostyk SK. Cognitive dysfunction contributes to mobility impairments in Huntington's disease. *J Huntingtons Dis.* 2017;6(4):363-370.

49. Gaskin J, Gomes J, Darshan S, Krewski D. Burden of neurological conditions in Canada. *Neurotoxicology.* 2017;61:2-10.

50. Edmondson MC, Goodman L. Contemporary health care for Huntington disease. *Handb Clin Neurol.* 2017;144:167-178.

51. McIntyre D, Fleming J, Foster M, Tweedy S. Experiences of adults with high-care needs and their family members with housing and support pathways in Australia. *Disabil Rehabil.* 2017;39(18):1829-1839.

52. Parkinson's Foundation. Bradykinesia (slowness of movement). 2019. https://parkinson.org/Understanding-Parkinsons/Symptoms/Movement-Symptoms/Bradykinesia-Slowness-of-Movement.

53. Robles IS. Bradykinesia. Huntington's Outreach Project for Education, at Stanford. 2010. https://hopes.stanford.edu/glossary/bradykinesia.

54. Parkinson's Foundation. Dyskinesia. 2019. https://parkinson.org/Understanding-Parkinsons/Symptoms/Movement-Symptoms/Dyskinesia.

55. Subramaniam NS, Bawden CS, Waldvogel H, Faull RML, Howarth GS, Snell RG. Emergence of breath testing as a new non-invasive diagnostic modality for neurodegenerative diseases. *Brain Res.* 2018;1691:75-86.

Nancy L. Urrutia is a nurse educator in the Cleveland, Ohio metropolitan area.

The author and planners have disclosed no potential conflicts of interest, financial or otherwise.

DOI-10.1097/01.NURSE.0000559914.46449.29