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# CLINICAL SAVVY

## Huntington's Disease: Helping the Patient Retain Function

By Jeanne Kimball France

I was once called to help subdue a patient who, frustrated that no one could understand him, was becoming quite disruptive. He had Huntington's disease (HD), an inherited illness characterized by rapid, jerky, purposeless movements (chorea), speech problems, and mental deterioration. Prior experience with HD had taught me how to help this man put his fear, anger, loss, and loneliness into words. Our conversation calmed him. After we talked for a while, he smiled, sighed, and kissed my hand.

Patients with symptomatic HD have unique physical, psychosocial, environmental, and nutritional requirements. But there are strategies for meeting these needs. Outlined below are some ways you can help patients with HD—and others with psychomotor disorders—retain function and hope through improved communication, mobility, personal care, safety, and nutrition.

### **Q** How common is HD and what's its typical course?

**A** In the United States, about 25,000 people have the disease and another 150,000 have a 50-50 chance of developing it. Initial symptoms usually appear between the ages of 35 and 40, but may occur as early as adolescence. After that, it follows a progressively debilitating 10- to 25-year course.

As Huntington's advances, it affects cognitive functions, such as the abilities to plan, reason, think creatively, and make judgments. It becomes more dif-

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Although there's still no cure, there's plenty you can do to help the patient with Huntington's disease communicate and remain mobile, strong, and independent for as long as possible.

icult and takes more time for the person to solve problems, store language, concentrate, and conceptualize. All of this contributes to his inability to communicate. This can be extremely frustrating for the patient, as well as for the family or health care provider.

### **Q** Is there any treatment for HD?

**A** Currently, there's no cure. Chorea may be slowed by medications, such as haloperidol (Haldol) and chlorpromazine (Thorazine). But now that the Huntington's gene has been isolated, there's hope of developing drugs that will limit the progression of the disease or prevent symptoms altogether.

### **Q** What can I do to help the patient overcome communication problems?

**A** Allow him plenty of time to respond—even minutes. In Huntington's, a person receives the message, but has difficulty interpreting it and forming a response. He may still be an-

swering the first question while you're asking a second or third. Try asking questions that require one-word responses. Use short, simple questions or statements. Break complex subjects into smaller parts. Speak slowly and clearly.

If you're having difficulty understanding the patient, ask that he say one word at a time. Repeat each word so he knows you understand him. (This reduces his frustration.) You might also use visual aids, such as a "point board," and ask him either to point to the correct response or to look to the right for yes and to the left for no. If available, a speech therapist can help evaluate the problem and develop strategies for improving or maintaining communication.

### **Q** How can I teach a patient with progressive mental deterioration something as complex as new ways to speak and move?

**A** Try to involve the family as well as the patient in the education. Sometimes they can help you identify how the patient learns. For example, does the patient learn best by repetition or by association? This information will help you personalize education.

Frequently, adaptive skills practiced while the patient is in the early stages of HD can be recalled later in the disease process. For example, one patient, who'd been taught to use the call bell, didn't use it until the day she got caught behind her bed.

### **Q** Why do people with HD have problems walking? What can I do to help the patient become more mobile?

**A** As HD progresses, degeneration of the neuromuscular system causes the

posture and coordination to deteriorate. The patient develops a wide-based, staggering gait. Moreover, as chorea worsens, he spends more energy in constant movement and tires easily. Advise the patient to take naps and pace activities. Energy conservation generally improves endurance, reduces agitation, and lessens choreic movements.

To maintain leg strength and coordination, the patient might try riding a stationary bicycle. (Some HD patients find that easier than other exercises.) To exercise the patient's upper body, suggest that he practice throwing and catching a lightweight ball.

Supportive devices, such as handrails, can stabilize the patient's balance. You might suggest he steady himself by pushing a wheelchair. Then, if he gets tired, he can sit down. (Before allowing the patient to use a wheelchair, teach him how to brake and maneuver it. Oversee his practice until he masters the skill.)

**Q** *How can I help a patient with advanced HD perform activities of daily living?*

**A** Help him break each activity into simple steps. For example, a transfer from bed to chair becomes: stand up, pivot, sit down.

Independence to dependence is the course of HD, but along the way people have ups and downs. Encourage the patient to do as much of his own care as possible. Assist as necessary, helping him straighten his clothes or fasten buttons and laces, after he's completed as much as he can. To make dressing easier for the patient, clothing should be a size too large. A jogging suit with a wide neck, loose fit, and no zippers and buttons is ideal (plus, it's easy to launder). Sturdy shoes with Velcro closures or elastic laces make it easier for the patient to slip shoes on and off. Avoid slippers and sandals as they don't provide good support.

When the patient's balance deteriorates, encourage him to sit on a sturdy chair while washing, brushing his teeth, dressing, or combing his hair. Seated, he has more support and conserves energy. Be sure that the patient has plenty of extra space to maneuver safely and that his environment is free of clutter.

Keep in mind that weather will affect

his physical condition. Generally, he'll be more stable and will function better in cool weather. During hot, humid months, endurance and capacity wane.

**Q** *What other health problems are associated with HD?*

**A** As HD progresses, the patient may lose bladder and bowel control. A toilet schedule, such as first thing in the morning and then every two hours thereafter until bedtime, may prevent the need for incontinence aids.

Since the patient may have difficulty wiping properly, look for signs of urinary tract infections, such as restlessness, visible pain during urination, and frequent trips to the toilet. Also monitor bowel movements. Huntington's patients may be unable to swallow sufficient fluids or roughage and often don't realize or can't communicate that they're constipated. An unexplained rise in agitation may be your first clue. Intervene as appropriate with supplemental dietary fiber and bowel training.

Many HD patients are depressed. They might have watched a parent or other relative suffer with the disease. Then, when they develop symptoms, they may feel hopeless or irritable, have trouble sleeping, or experience a rise or fall in appetite. Some respond by isolating themselves. Antidepressants, support groups, therapy, and acceptance by family and friends can help relieve these symptoms.

Brain-tissue changes also affect HD patients' ability to control impulses. They may abuse alcohol and other substances, become hypersexual, or engage in risky sex. (Safety awareness drops off.) In the later stages of HD, a patient who smokes will find it difficult to hold a cigarette without dropping it. Supervise any smoking.

**Q** *What safety precautions should be taken in the hospital?*

**A** As chorea worsens and the patient is in bed for longer periods, it's necessary to reevaluate the bed area for safety. Be sure that the mattress fits the frame snugly and that there's no room between the frame and the lower edge of the side rail. (Otherwise, the patient may catch his head in the gap and strangle, or

break a limb.) The bed's side rails and foot board may be padded to protect the patient from bruising himself.

To enhance his quality of life, the patient is encouraged to be out of bed as much as possible. Try to provide a sturdy chair with safety and comfort features, such as safety belts, padding (on the seat, sides, and leg rest), and the ability to tilt for easier positioning.

**Q** *Does the Huntington's patient need a special diet?*

**A** Yes, one that's high in calories and fiber. Choreic movements raise the caloric requirements in the HD patient, who may require 4,000 to 5,000 kcal or more a day to maintain his weight. Patients whose movements aren't yet excessive may be able to maintain weight on a regular diet.

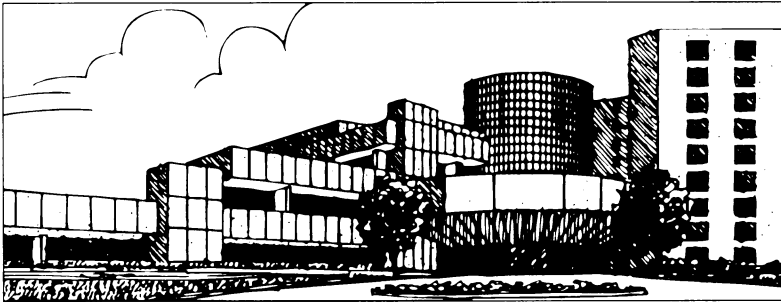
Monitor weight weekly. Try to maintain HD patients at a higher-than-normal weight. Those who lose even one to two pounds in a week seem to experience more chorea, agitation, and swallowing difficulties. And since HD patients may be prone to constipation, check that the diet is high in natural fiber.

As it becomes harder for the HD patient to move food from the front to the back of his mouth, you can start him on a soft or soft-moist diet (normal foods, but with creamy sauces or gravies to ease food through the mouth and down the esophagus). Eventually, he may need pureed foods; again, try blending normal foods, which offer more variety in flavor and contrast. Thin liquids can be difficult to swallow and may cause choking. Liquids such as water, fruit juice, coffee, and milk can be thickened by adding instant food thickener, breakfast powder, yogurt, smooth ice cream, pudding, or pureed fruits (which also add calories).

**Q** *What other feeding problems should I watch out for?*

**A** Choreia can make it difficult for the HD patient to hold his head still. Seat him in a chair with armrests so that he can steady one elbow and support his head in that hand, while leaving the other hand free to pick up the food. Normal utensils and dishes may be difficult for the patient to handle.

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Try using adaptive feeding equipment: Covered "sippy" cups allow the patient to drink without spilling the liquid. High-rimmed plates let him scoop up food with a spoon or fork. Nonskid mats keep the plate in place.

Since HD patients are prone to dysphagia, always have the patient sit upright for meals to prevent choking and aspiration. If you're feeding him, allow him sufficient time to chew and swallow each bite before placing the next one in his mouth. Offer him a straw for drinking liquids and encourage sips of no more than 5 to 10 mL, or less than a mouthful, at a time.

If the patient speaks or makes noises during meals, listen carefully. If his voice sounds muffled or like he's talking underwater, ask him to cough. This will clear any fluids from his larynx.

If possible, speak with the patient and family about advanced preferences for feeding long before gastrostomy tubes are needed. If the patient has an opportunity to consider the various options at an earlier time, it can make the eventual transition less emotional for all.

**Q** *How can I get more information on Huntington's disease? What groups conduct research on HD?*

**A** Write the Huntington's Disease Society of America (HDSA), 140 W. 22nd Street, 6th Floor, New York, NY 10011, or call the HDSA National Helpline at 1-800-345-HDSA. Huntington's disease research is supported by the Hereditary Disease Foundation (213-458-4183) and the Foundation for Cure and Care of HD, 81990 Overseas Highway 5-202, Islamorada, FL 33036.

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