



CREUTZFELDT-JAKOB DISEASE
FOUNDATION, INC.

Supporting Families Affected by Prion Disease



CAREGIVING IDEAS

Families who receive a prion disease diagnosis are often shocked, confused and overwhelmed. They must rapidly accept the news that their loved one – who probably appeared quite healthy just weeks ago – is suffering from a terminal disease that has no treatment. And they must quickly determine who will care for the patient, and where. Here are some thoughts to consider.

DEVELOP A CARE PLAN

Families who receive a prion disease diagnosis should contact hospice as early as possible. While hospice care may not be immediately needed in the early stages, the patient's needs are likely to evolve rapidly. Choosing a hospice provider and setting up care in advance will free the family to focus more on the patient later. Hospice providers bring knowledge and resources that are invaluable to a family that is confronting rapidly evolving care needs.

When receiving a diagnosis of prion disease, a family will need to quickly consider several issues. Consulting hospice, the medical care provider, or hospital social worker at this time can be very valuable. Issues include:

- Is it safe for the patient to be alone at any time? If the patient lives alone, who will monitor them on a daily basis, to determine when this assessment changes? (This can change rapidly as the disease progresses)
- If the patient is about to be released from the hospital, is the best place for them going to be at their home, at another home, or at a hospice or other care facility?
- Should the patient be moved to another community or another

state where more family or friends are available to caregive? Is the patient still able to comfortably and safely travel?

- Which family members or friends are available to help? Can a schedule be arranged for them to care for the patient, or serve as companions, in shifts?
- Can private health aides be hired if needed?

EXPECT FREQUENT CHANGES

A patient can experience many or just a few of the common symptoms associated with prion disease. One consistent factor, though, is frequent change. For a caregiver, it is important to stay attuned to the shifting needs of the patient. This becomes challenging as the patient loses the ability to communicate and to understand. It would be helpful for caregivers (family members and professionals) to communicate with each other regularly, or possibly keep a notebook, with any changes in the patient's symptoms, ideas on what seems to soothe them, and concerns.

CREATE A SOOTHING ENVIRONMENT

Heightened sensitivity is common for prion disease patients. Reactions occur more commonly to unexpected touch, loud or sudden noises and music. This may produce immediate signs of distress and agitation.

Caregivers should limit stimuli to make the patient comfortable, and stay attuned to factors that distress them. If reflections seem to cause visual disturbance, the family can

cover reflective surfaces such as mirrors, windows, or glass tables or shiny appliances. A flickering television can be disturbing, or could even induce hallucinations. Soft lighting is ideal, as is quiet music if the patient tolerates it well. It may be necessary to avoid loud sounds and large groups of people if they appear disruptive to the patient.

OFFER SUPPORT FOR ACTIVITIES OF DAILY LIVING (ADLs)

Over time the patient is likely to need increasing help with eating, bathing, dressing, toileting, and transferring. Hospice and health aides can provide invaluable assistance in planning and problem solving. They have knowledge and access to countless approaches, tools, and resources that can aid in patient comfort and care. Often they may suggest a caregiving aid (such as a shower chair or hospital bed) before the family realizes the patient needs it.

Handling Incontinence

Incontinence can make the patient agitated, restless, and distressed. It can give them a feeling of losing dignity and independence, and in later stages, the movement required to change the patient can be jarring. Working with the hospice or a medical care provider, the family can assess options for addressing incontinence and learn how to minimize movement when changing bedding and clothing.

Nourishment

As prion disease progresses, the patient may have difficulty with appetite, chewing, focusing on a meal, and ultimately with swallowing.



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At early stages, ergonomic utensils may be helpful to enable self-feeding. As dysphagia progresses, pureed foods and thickener in fluids may become necessary.

The caregiver should balance nourishment with enticing foods. If the patient prefers less healthy foods such as milkshakes, or begins focusing on a very limited set of preferred foods, it is fine to move in that direction as long as the patient is obtaining adequate nourishment and is not encountering digestive difficulties. The patient's desire for foods may fade. As long as they are hydrated, caregiver should follow the patient's lead where possible, and should not force foods. Mouth care is important to keep the patient comfortable. Options include a washcloth, and a child's toothbrush, and swabs.

Handling psychiatric symptoms

The patient may suffer many or few psychiatric symptoms. While the patient's temperament and personality may change radically, it is important for the family to remember that it is often the disease talking, and not the patient. Remaining calm and patient, and not arguing when the patient expresses delusions and hallucinations, is ideal. As long as the patient is not a hazard to themselves or others, reassuring them and working through episodes with patience is best. If symptoms worsen, the caregiver should communicate frequently with the physician to ensure medications and dosages are as effective as possible in managing symptoms.

Handling Myoclonus (Tremors or Seizures)

If the patient develops myoclonus, caregivers can help by minimizing touching and movement in caring for

the patient. At times, gentle massage may help relax the muscles, and the family may ask the physician about medications to help reduce spasticity and tremors. While myoclonus spasms are not painful for the patient, they are certainly disruptive, so reducing their effects can enhance the patient's comfort.

COMMUNICATE WITH YOUR PHYSICIAN

As the disease progresses, you will want to contact the physician, hospice, or healthcare provider about any concerns, such as:

- New symptoms that are not easily managed, particularly hallucinations, and extremely fearful or violent behavior
- Suspected infections such as UTI or pneumonia
- Options for reducing patient's discomfort and severe distress
- How to maintain nourishment and hydration when swallowing becomes difficult

Be bold in reaching out to the healthcare professional if you are not hearing a response. With prion diseases, the patient's condition may change more quickly than the doctor is accustomed to.

MANAGE VISITORS

After prion disease diagnosis, your family may feel compelled to invite large groups to your home to visit the patient or say goodbye to them. Before inviting a large group, assess whether the patient might adjust well to the group. If visits make the patient restless or agitated, it is best to ask visitors to come in pairs or small groups, keep the visit short,

and have low expectations for the patient's level of involvement in the discussion.

Although the patient may be confused or even unable to communicate, you can try to include them in conversations, and speak to them kindly about your shared history, your love for them, pride in their accomplishments, and happy memories together.

TAKE CARE OF THE CAREGIVER

Relieving your exhaustion and stress is crucial for both you and the patient. You may have little time to care for yourself while caring for the patient, but self-care is vital. Exercising, getting adequate sleep, and eating healthfully will benefit you and enable you to continue the grueling tasks of caring for your loved one. You may also need to ask for help from professional caregivers or family and friends.

If friends offer to help, you may want to ask them to provide respite care, set shifts for sitting with the patient, or provide a break or support for you in some way. Friends could organize meal trains or offer to do errands and help with maintaining the home. As the caregiver, you can also benefit from stress reduction through therapy, meditation, massage, and exercise.

For more information on Caregiving for a prion disease patient, contact the CJD Foundation Helpline to ask questions or request a Caregiving Guide: 800-659-1991