CHALLENGES IN LATE-STAGE HD:

PALLIATIVE CARE AND THE CALIFORNIA END OF LIFE OPTIONS ACT

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HUNTINGTON DISEASE STAGES

- Stage 1
  - Slightly lower performance at work; independent at home.
- Stage 2
  - Can still work (lower level), still mostly independent at home.
- Stage 3
  - Difficult to work, starts to needs help with financial, home activities
- Stage 4
  - Unable to work. Needs major assistance with care and 24 hour caregiving
- Stage 5
  - Bedbound. Full-time nursing care required.

USUAL COURSE OF HD: ERODED FUNCTIONAL ABILITIES

- Loss of capacity to work
- Loss of capacity to parent
- Impaired ability to perform activities of independent living such as driving and shopping
- Impaired ability to perform personal activities of daily living
- Eventual need for 24 hour caregiving to address issues of mobility, safety, nutrition, and emotional well-being
- Death after 15 – 20 years

CHALLENGES IN HD

- Long timeframe from diagnosis to death
- Loss of functionality is traumatic to patients and their families
- Unawareness or denial of symptoms or diagnosis, mood disorders, cognitive impairment, poor judgment may prevent discussion or completion of Advanced Directives

HD PEER WORKGROUP PRIORITIES FOR HD CARE (2001)

- Autonomy
- Dignity
- Meaningful social interaction
- Communication
- Comfort
- Safety and order
- Spirituality
- Enjoyment, entertainment and well-being
- Nutrition

Huntington's Disease Workgroup (2001). Promoting excellence in end of life care. A National Program of the Robert wood Johnson Foundation.

PALLIATIVE CARE

"Palliative care is an approach that improves the quality of life of patients and their families facing the problem associated with life-threatening illness, through the prevention and relief of suffering by means of early identification and impeccable assessment and treatment of pain and other problems, physical, psychosocial and spiritual."

World Health Organization, http://www.who.int/cancer/palliative/definition/en/

WHAT IS PALLIATIVE CARE ?

- specialized care for people with serious illnesses
- focused on providing patients with relief from the symptoms, pain, and stress of a serious illness - whatever the diagnosis
- goal is to improve quality of life for both the patient and family
- provided by a team of specialists who work with a patient's doctors to provide an extra layer of support
- appropriate at any age, and at any stage, of a serious illness
- can be provided together with curative treatment, or as the sole focus of care.

Courtesy of Dr. Nathan Fairman, UCD Palliative Care Physician
Center to Advance Palliative Care 2011.

PALLIATIVE CARE

- provides relief from pain and other distressing symptoms;
- affirms life and regards dying as a normal process;
- intends neither to hasten or postpone death;
- integrates the psychological and spiritual aspects of patient care;
- offers a support system to help patients live as actively as possible until death;
- offers a support system to help the family cope during the patients illness and in their own bereavement;
- uses a team approach to address the needs of patients and their families, including bereavement counselling, if indicated;
- will enhance quality of life, and may also positively influence the course of illness;
- is applicable early in the course of illness, in conjunction with other therapies that are intended to prolong life, such as chemotherapy or radiation therapy, and includes those investigations needed to better understand and manage distressing clinical complications.

World Health Organization, http://www.who.int/cancer/palliative/definition/en/

PALLIATIVE CARE FOR CHILDREN: JHD

- Palliative care for children is the active total care of the child's body, mind and spirit, and also involves giving support to the family.
- It begins when illness is diagnosed, and continues regardless of whether or not a child receives treatment directed at the disease.
- Health providers must evaluate and alleviate a child's physical, psychological, and social distress.
- Effective palliative care requires a broad multidisciplinary approach that includes the family and makes use of available community resources; it can be successfully implemented even if resources are limited.
- It can be provided in tertiary care facilities, in community health centers and even in children's homes.

World Health Organization, http://www.who.int/cancer/palliative/definition/en/

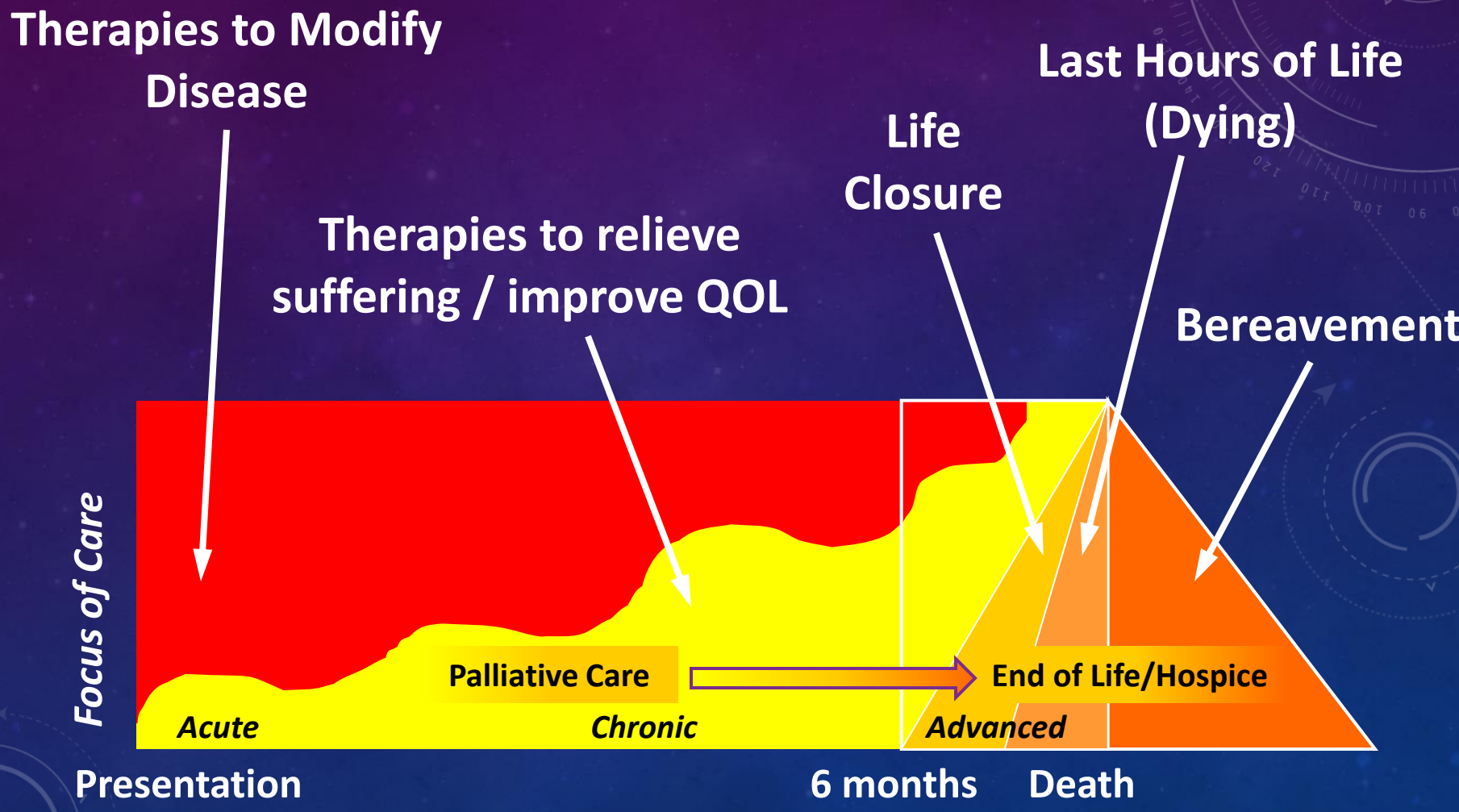
HOSPICE CARE

Provides ENHANCED Palliative Care

- Last phase of life-threatening illness
  - Safe, comfortable dying
  - Self-determined life closure
- Bereavement period
  - Effective grieving

Courtesy of Dr. Nathan Fairman, UCD Palliative Care Physician
Ferris F. JPSM 2002; 24:106.

Palliative Care



Courtesy of Dr. Nathan Fairman, UCD Palliative Care Physician

LATE-STAGE HD CARE ISSUES: LIFESTYLE, MOBILITY AND ENVIRONMENT

- Padded furniture and fixtures to prevent injury and bruising
- When walking is no longer safe, transition to wheelchair or IPU Ultimate Walker
- Custom wheelchairs needed to accommodate postures and movements of late-stage HD
- Seatbelts, seat vests, pummel seats
- Broda chair: cushioned tilt-in-space wheelchair
- Padded bumpers on beds

Klagen J, Duckett A, Sandler S and Moscowitz, C. "HD: A Caring Approach to the End of Life." Case Management Journals 9;2008:75 - 81

LATE-STAGE HD CARE ISSUES: MEDICATIONS

- Movement disorders include chorea, dystonia, rigidity and slowness
- "The chorea is not as disabling as the underlying emotional problems of stress, anxiety or depression that may cause the movements to worsen."
  - Chorea can be managed with low-dose neuroleptics, benzodiazepines and dopamine depleters (tetrabenazine).

Klagen J, Duckett A, Sandler S and Moscowitz, C. "HD: A Caring Approach to the End of Life." Case Management Journals 9;2008:75 - 81

LATE-STAGE HD CARE ISSUES: MEDICATIONS

- "Although medicating for chorea may has cosmetic value, medicating the underlying emotional problems proves most helpful."
  - Medications commonly used: atypical antipsychotics, antidepressants and benzodiazepines

Klagen J, Duckett A, Sandler S and Moscowitz, C. "HD: A Caring Approach to the End of Life." Case Management Journals 9;2008:75 - 81

OTHER SYMPTOMS IN LATE-STAGE HD

- Urinary difficulties (retention, catheters)
- Constipation, feeding difficulties
- Weight loss
- Skin breakdown
- Pain

PAIN IN LATE-STAGE HD

- Use of padded bed railings, comfort mattress (air or other), reclining chair
- Frequent turning, re-positioning, stretching, range of motion exercises, touch and massage
- Simple analgesics to start
- Step up to stronger pain meds including opioids as needed
    - Monitor for worsening constipation and respiratory compromise

ADVANCED DIRECTIVES

- Definition: a document expressing a person's wishes about critical care when he is unable to decide for himself.
- Types of decisions to make:
  - Decision to hospitalize
  - Decision to resuscitate
  - Decision about feeding tubes
  - Decision about organ donation for those living, and brain donation for research
- All patients should have significant input into what their wishes for end of life would be

WHEN TO CONSIDER HOSPICE REFERRAL

- Generally, for patients in late stage HD – Stages 4 - 5
- Most patients will be bed-bound or at least partly bed-bound
- Rapid or significant weight loss
- Recurrent serious infection – pneumonia, urinary tract
- Recurrent or significant skin breakdown
- Additional serious medical illness: heart disease, cancer, other

MEDICARE HOSPICE BENEFITS

- If you have Medicare Part A (Hospital Insurance) AND meet these conditions, you can get hospice care:
  - Your hospice doctor and your regular doctor (if you have one) certify that you're terminally ill (with a life expectancy of 6 months or less).
  - You accept palliative care (for comfort) instead of care to cure your illness.
  - You sign a statement choosing hospice care instead of other Medicare-covered treatments for your terminal illness and related conditions.

https://www.medicare.gov/Pubs/pdf/02154.pdf

HOSPICE CARE

- You can get a one-time only hospice consultation with a hospice medical director or hospice doctor to discuss your care options and management of your pain and symptoms. You can get this one-time consultation even if you decide not to get hospice care.
- Medicare will cover the hospice care you get for your terminal illness and related conditions, but the care you get must be from a Medicare-approved hospice provider.

https://www.medicare.gov/Pubs/pdf/02154.pdf

WHAT SERVICES DOES HOSPICE PROVIDE?

Hospice care is usually given in your home. Depending on your terminal illness and related conditions, the plan of care your hospice team creates can include any or all of these services:

- Doctor services and nursing care
- Medical equipment (like wheelchairs or walkers) and medical supplies (like bandages and catheters)
- Prescription drugs
- Hospice aide and homemaker services
- Physical and occupational therapy, speech-language pathology services
- Social worker services
- Dietary counseling
- Grief and loss counseling for you and your family
- Short-term inpatient care (for pain and symptom management)
- Short-term respite care
- Any other Medicare-covered services needed to manage your terminal illness and related conditions, as recommended by your hospice team

https://www.medicare.gov/Pubs/pdf/02154.pdf

EUTHANASIA AND PHYSICIAN-ASSISTED SUICIDE IN THE NETHERLANDS

- Termination of Life on Request and Assisted Suicide Act, 2002
- Two main requirements for physicians:
  - Know that the request was voluntary and carefully considered
  - The patient's suffering was unbearable and without prospect of improvement, and there is no reasonable alternative

Intl Psychogeriatrics 25;2013:339-340.

EUTHANASIA AND PHYSICIAN-ASSISTED SUICIDE IN THE NETHERLANDS

- This option may be requested in the Advanced Directive, initially intended for those with dementia
  - The insidious progression of dementia allows patients to adapt to the disease and may lead to a change of opinion, called “response shift.”
  - Surveys showed >50% of physicians would be reluctant to perform euthanasia based on an Advanced Directive
  - The California End of Life Options Act specifically excludes the option of requesting an end-of-life medication based on an advanced directive. The patient must be able to request the prescription verbally and in writing at the time that they wish to take this action.

Intl Psychogeriatrics 25;2013:339-340.

EUTHANASIA AND PHYSICIAN-ASSISTED SUICIDE IN THE NETHERLANDS

- The second condition is suffering.
  - The court has stated that "the prospect of unbearable suffering, progressive deterioration and when knowing that dying with dignity is possible now, but most likely not in the future can qualify as suffering."
- Between 2007-2011, 30 HD patients requested this option.
  - Estimated prevalence is 1700 cases, with 50-60 deaths/year and 5 – 10 requesting a prescription for an end-of-life medeication

Intl Psychogeriatrics 25;2013:339-340.
J Neurol 261;2014:2184-2191

OREGON AND WASHINGTON STATE DEATH WITH DIGNITY ACTS

- "Attitudes of Patients with ALS and Their Caregivers Toward Assisted Suicide."

  Ganzini et al, NEJM 339;1998:967-973

- N = 100 patients and n = 91 care partners
- "Under some circumstances I would consider taking a prescription for a medicine whose sole purpose was to end my life."
  - 56% of patients indicated yes
  - 56 of the 91 care partners would support this decision

UNDERSTANDING CALIFORNIA'S END OF LIFE OPTION ACT*

What is the End of Life Option Act?

- This is a new California law that will allow a terminally ill patient to request a drug from his or her physician that will end the patient's life.
- Physicians who help their patients with this process will be providing a new, legal form of end of life care and will not be subject to legal liability or professional sanction for doing so.
- This end of life option is voluntary for both patients and their physicians.

*This summary is based on information provided by the UCSF/UC Hastings Consortium on Law, Science, & Health Policy and does not constitute legal advice.

CALIFORNIA'S END OF LIFE OPTION ACT
(continued)

Who can use this option? The patient must:

- Be 18 or older and a resident of California.
- Have a terminal disease that cannot be cured or reversed and is expected to result in death within 6 months.
- Have the capacity to make medical decisions.
- Not have impaired judgment due to a mental disorder.
- Have the ability, physically and mentally, to take the drug at the time they want to take it.

Patients cannot request aid-in-dying in advance directives or other documents.

CALIFORNIA'S END OF LIFE OPTION ACT
(continued)

How does a patient obtain aid-in-dying?

- If the above conditions are met, the patient must make two verbal requests at least 15 days apart to his or her physician, plus a special request in writing.
- The physician and patient must discuss this decision privately to make sure the decision is completely voluntary.
- A second physician must confirm the patient's diagnosis, prognosis, and ability to make medical decisions.
- The patient and physician must discuss several specific issues related to the aid-in-dying drug and alternatives, including comfort care, hospice care, palliative care, and pain control.

CALIFORNIA'S END OF LIFE OPTION ACT
(continued)

Do patients have to take the drug if they've requested it and received a prescription for it?

- Patients who receive the drug can take it whenever they want – or they can choose not to take it at all.
- Taking the drug is the patient's choice alone.
- The patient must take the drug himself or herself. Others can help prepare the drug and sit with the patient, but the patient must be the one to physically take it.

Do doctors have to give patients aid-in-dying if they ask for it?

- No. A physician's participation is voluntary – and so is the participation of facilities like hospitals and nursing homes.

CALIFORNIA'S END OF LIFE OPTION ACT
(continued)

Does the law protect patients from being pressured to end their lives?

- Yes. There are several provisions to assure that a patient's choice is completely voluntary, backed by criminal sanctions.
- The law also prevents insurers from denying life or health insurance or annuities based on requesting aid-in-dying.
- Wills and contracts cannot require patients to receive aid-in-dying or prevent them from doing so.

For more information about California's End of Life Option Act, see http://www.ucconsortium.org/wp-content/uploads/2015/12/FACT-SHEET-End-of-Life-Option-Act-Updated-01.15.16.pdf

BEREAVEMENT

- “Bereavement is the period of grief and mourning after a death. When you grieve, it's part of the normal process of reacting to a loss. You may experience grief as a mental, physical, social or emotional reaction. Mental reactions can include anger, guilt, anxiety, sadness and despair. Physical reactions can include sleeping problems, changes in appetite, physical problems or illness.”
- “How long bereavement lasts can depend on how close you were to the person who died, if the person's death was expected and other factors. Friends, family and faith may be sources of support. Grief counseling or grief therapy is also helpful to some people.”

https://www.nlm.nih.gov/medlineplus/bereavement.html

NEXT STEPS

- HDSA Center of Excellence at UC Davis will conduct an anonymous survey of attitudes about End of Life Care Act in summer-fall 2016
- Respondents will be 18 and older, and will either have HD or be at risk of HD, or be a care partner
- Graduate student Megan Hereen, Lisa Mooney LCSW, Dr. Josh Dayananthan and others

Thank you