Disclosures

I have no conflicts of interest in relation to this presentation
Outline

- Palliative and Supportive Care
- Neuropsychiatric Symptoms
- Symptomatic Management
Cultural Adaptation is Required!
"Our most cruel failure in how we treat the sick and the aged is the failure to recognize that they have priorities beyond merely being safe and living longer"

Atul Gawande, Being Mortal
Overview

• Palliative or Supportive care is a medical specialty focused on caring for patients and caregivers affected by serious illness
  • Offers a comprehensive approach to health and well-being by focusing on the physical, emotional, social, and spiritual distress caused by serious illness
  • Focuses on improving quality of life for patients and family
    • Treat symptoms and distress (both physical and psychological)
  • Only one aspect is end-of-life and advance care planning
  • Supports informed decision making
  • Increasingly referred to as “Supportive Care” due to negative stigma and association of “palliative” with hospice and end of life
Overview

• Palliative care is applicable early in the disease and throughout the disease course
  • Starts when making the diagnosis of a neurodegenerative disease
    • ALS
    • Parkinson’s disease and related disorders
    • Alzheimer’s disease and related disorders
  • Offered simultaneously with all other appropriate medical treatment
Overview

• Myths:
  • “Why make the diagnosis when there is no treatment or cure?”
  • “There’s nothing I can do”

• Our care and treatment approach does not stop after someone has been diagnosed with a neurodegenerative disease without a cure – it may adapt or change
  • Start applying palliative care approaches to provide comfort and support to the patient and family

• Palliative care in neurodegenerative diseases has been shown to:
  • Improve quality of life
  • Reduce symptom burden
  • Reduce caregiver burnout and distress
  • Reduces complicated grief
  • Reduces healthcare costs

Katz, AAN Continuum – Movement Disorders 2022; Kluger et al., JAMA Neuro 2020
Palliative Care Approach

- **Serious illness communication** using evidence-based techniques
  - Communication is key!
    - Treat communication with care and preparation
    - Plan for what will be discussed and how
  - Communication varies by culture and must be adapted to what is most appropriate in your local culture and language
- Sharing guidance and facilitating advance care planning to be able to provide care that aligns with the patient and family goals
  - Most patients prefer this discussion earlier in the course of illness
- Emphasizes patient autonomy to decide preferences and goals of care
  - Determine how much the patient and family want to know
  - Family can speak on behalf of the patient when patient no longer able to understand their medical condition and communicate their preferences
  - “Has XX ever talked to you about what she would want if a situation like this ever arose...Have they said anything to you before?”
    - If not: “If XX was his normal self prior to these symptoms starting, what would he tell us was most important to him”
## Sharing the Diagnosis: SPIKES

<table>
<thead>
<tr>
<th>Term</th>
<th>Definition</th>
<th>Communication examples</th>
</tr>
</thead>
<tbody>
<tr>
<td>Setting</td>
<td>Establish an appropriate setting</td>
<td>Minimize disruptions, have easy access to tissues</td>
</tr>
<tr>
<td>Perception</td>
<td>Establish the patient’s and caregiver’s perception of their symptoms and health status</td>
<td>Ask questions such as:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>“Can you give me a sense of what you think might be causing your symptoms?”</td>
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<tr>
<td></td>
<td></td>
<td>“Have other doctors suggested a diagnosis, or have you read anything online that pointed you in one direction or another? It helps me to know where you’re coming from.”</td>
</tr>
<tr>
<td>Invitation</td>
<td>Obtain permission from the patient and caregiver to provide the difficult medical news</td>
<td>Ask questions such as:</td>
</tr>
<tr>
<td></td>
<td></td>
<td>“Would it be okay if I gave you my thoughts on where things are with your illness?”</td>
</tr>
<tr>
<td>Knowledge</td>
<td>Give a warning shot to allow the patient/caregiver to brace for bad news</td>
<td>“I have difficult news”</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Deliver the serious illness diagnosis in a bite-size chunk</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Example: “Your symptoms and examination are most consistent with Parkinson disease,” then pause to allow time for the patient and caregiver to process the distressing information</td>
</tr>
<tr>
<td>Emotions</td>
<td>Expect and validate the difficult emotions that will arise after hearing bad medical news with empathic communication</td>
<td>“That’s so difficult to hear. How are you doing with that information?”</td>
</tr>
<tr>
<td>Strategy</td>
<td>Develop a strategy for the future</td>
<td>“Let’s focus on helping you live as well as you can for as long as you can.”</td>
</tr>
</tbody>
</table>

Katz, AAN Continuum – Movement Disorders 2022;
Advance Care Planning Discussion: AVMAP

• When you would like to communicate that the disease is progressing and to identify goals of care for the future

• **A = Ask Ask Tell Ask**
  • **Ask Understanding**: Identify their understanding of prognosis and health:
    • “How do you think your health is now compared to 1 year go”
  • **Ask Permission** to share your impression
    • “Would it be ok if I shared my sense of where you are in the course of the disease”
    • If the patient/family do not want to know, then stop there and can ask about goals
  • **Tell Impression**: Share your impression in small pieces with a warning shot → main headline → then pause
    • “I’m worried that the disease has progressed, and time is now much shorter than we had hoped”
  • **Ask Impression**: Check in with their response to the bad news
    • “What are your thoughts” “How are you doing with this news”

Katz, AAN Continuum – Movement Disorders 2022; Childers et al., J Onc Prac 2017
Advance Care Planning Discussion: AVMAP

• **V = Validate their response and emotions**
  - Take time to listen and provide validation to their emotions and comfort
  - “I can’t imagine how difficult this must be to hear”

• **M = Map Out the Patient’s Goals of Care**
  - Identify 2 or 3 big picture values and goals
  - “When you look ahead, what your goals if your condition worsens? What worries you the most?”
  - “If time is short, what is most important to you? How do you want to spend your time?”
  - “Who do you want to make decisions if you are unable to?”

• **A = Align with the Patient’s Goals of Care**
  - “From what I’m hearing, spending meaningful time with family and being pain free are top priorities”

• **P = Plan Care Based on Goals and Values of patient**
  - Recognize this is an ongoing conversation
  - “Let’s make a treatment plan that focuses on these goals. Remember this is an ongoing conversation and we can change things as needed to help you live as well as you can”
Communication Pearls

• Be seated, make eye contact, and show you are listening with body language
• Can use light touch to provide support (shoulder, arm)
• Avoid technical language or medical jargon
• Less is more – spend more time listening than talking
• Demonstrate partnership and support
Communication Pearls

• Express support for the difficult decisions – “It sounds like this is what they would have wanted”

• Lines:
  • “I can only imagine” “I can’t imagine” “I can understand part of that feeling”
    • Avoid saying you understand what they are going through, unless you can share a specific example
  • ‘I wish” – ex: “I wish things were different”
    • Provides more alignment than saying “I’m sorry”
  • “Tell me more” when sharing hopes and fears
Caregiver Support, Grief, and Loss

- Caregivers experience 2 periods grief and loss when caring for someone with a neurodegenerative diseases
  - Grief at gradual loss of personhood of their loved one
    - There is a slow, progressive grief often experienced for years
  - Grief at Death
    - Some caregivers may feel relief and associated guilt
- Ask the caregiver: How are you doing? How can we best support you?
  - It takes a village to care for someone with a neurodegenerative diseases
    - Encourage to get as many hands on deck to help as possible
Prognosis

• **We cannot accurately predict the future**
  • Everyone patient is different with different diseases, genetics, environmental factors, and medical comorbidities that contribute to their presentation and stage at diagnosis
  • The best predictor of future progression in an individual patient is their past progression
    • Progression over last year is best predictor of degree of progression over the next year

• Staging – varies by person and disease
  • FAST scale for Dementia
  • Modified Hoehn and Yahr Stage for Parkinson’s Disease

• In the US, these criteria are used to predict <6 months prognosis in dementia:
  • Each of these:
    • Unable to ambulate, dress, or bath without assistance
    • Urinary and fecal incontinence (intermittent or constant)
    • No consistently meaningful verbal communication (<6 intelligible words)
  • At least one of these in the last 12 months:
    • Aspiration pneumonia, 10% weight loss, sepsis, pyelonephritis, stage 3-4 pressure ulcer, recurrent fever
# FAST Scale for Dementia

## Functional Assessment Staging Test

<table>
<thead>
<tr>
<th>Stage</th>
<th>Stage Name</th>
<th>Characteristic</th>
<th>Expected Untreated AD Duration (months)</th>
<th>Mental Age (years)</th>
<th>MMSE (score)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Normal Aging</td>
<td>No deficits whatsoever</td>
<td>--</td>
<td>Adult</td>
<td>29-30</td>
</tr>
<tr>
<td>2</td>
<td>Possible Mild Cognitive Impairment</td>
<td>Subjective functional deficit</td>
<td>--</td>
<td></td>
<td>28-29</td>
</tr>
<tr>
<td>3</td>
<td>Mild Cognitive Impairment</td>
<td>Objective functional deficit interferes with a person’s most complex tasks</td>
<td>84</td>
<td>12+</td>
<td>24-28</td>
</tr>
<tr>
<td>4</td>
<td>Mild Dementia</td>
<td>IADLs become affected, such as bill paying, cooking, cleaning, traveling</td>
<td>24</td>
<td>8-12</td>
<td>19-20</td>
</tr>
<tr>
<td>5</td>
<td>Moderate Dementia</td>
<td>Needs help selecting proper attire</td>
<td>18</td>
<td>5-7</td>
<td>15</td>
</tr>
<tr>
<td>6a</td>
<td>Moderately Severe Dementia</td>
<td>Needs help putting on clothes</td>
<td>4.8</td>
<td>5</td>
<td>9</td>
</tr>
<tr>
<td>6b</td>
<td>Moderately Severe Dementia</td>
<td>Needs help bathing</td>
<td>4.8</td>
<td>4</td>
<td>8</td>
</tr>
<tr>
<td>6c</td>
<td>Moderately Severe Dementia</td>
<td>Needs help toileting</td>
<td>4.8</td>
<td>4</td>
<td>5</td>
</tr>
<tr>
<td>6d</td>
<td>Moderately Severe Dementia</td>
<td>Urinary incontinence</td>
<td>3.6</td>
<td>3-4</td>
<td>3</td>
</tr>
<tr>
<td>6e</td>
<td>Moderately Severe Dementia</td>
<td>Fecal incontinence</td>
<td>9.6</td>
<td>2-3</td>
<td>1</td>
</tr>
<tr>
<td>7a</td>
<td>Severe Dementia</td>
<td>Speaks 5-6 words during day</td>
<td>12</td>
<td>1.25</td>
<td>0</td>
</tr>
<tr>
<td>7b</td>
<td>Severe Dementia</td>
<td>Speaks only 1 word clearly</td>
<td>18</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>7c</td>
<td>Severe Dementia</td>
<td>Can no longer walk</td>
<td>12</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>7d</td>
<td>Severe Dementia</td>
<td>Can no longer sit up</td>
<td>12</td>
<td>0.5-0.8</td>
<td>0</td>
</tr>
<tr>
<td>7e</td>
<td>Severe Dementia</td>
<td>Can no longer smile</td>
<td>18</td>
<td>0.2-0.4</td>
<td>0</td>
</tr>
<tr>
<td>7f</td>
<td>Severe Dementia</td>
<td>Can no longer hold up head</td>
<td>12+</td>
<td>0-0.2</td>
<td>0</td>
</tr>
</tbody>
</table>
Modified Hoehn and Yahr for Parkinson’s

<table>
<thead>
<tr>
<th>Stage</th>
<th>Modified Hoehn and Yahr Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Unilateral involvement only</td>
</tr>
<tr>
<td>1.5</td>
<td>Unilateral and axial involvement</td>
</tr>
<tr>
<td>2</td>
<td>Bilateral involvement without impairment of balance</td>
</tr>
<tr>
<td>2.5</td>
<td>Mild bilateral disease with recovery on pull test</td>
</tr>
<tr>
<td>3</td>
<td>Mild to moderate bilateral disease; some postural instability; physically independent</td>
</tr>
<tr>
<td>4</td>
<td>Severe disability; still able to walk or stand unassisted</td>
</tr>
<tr>
<td>5</td>
<td>Wheelchair bound or bedridden unless aided</td>
</tr>
</tbody>
</table>
Neuropsychiatric Symptom Management
Neuropsychiatric Symptoms (NPS) are Common

• 98% incidence of neuropsychiatric symptoms in dementia during disease course
• 75% of participants with dementia exhibit neuropsychiatric symptoms over a month
  • 55% of participants had 2 or more NPS
• Apathy - 36%
• Depression – 32%
• Agitation/Aggression – 30%
• Sleep – 27%
• Irritability – 27%
• Anxiety – 22%
• Delusions – 18%
• Aberrant motor behavior – 16%
• Hallucinations – 11%

Craig et al., AMGP 2005; Lyketsos et al., JAMA 2002; Kales et al., JAGS 2014
Non-Pharmacologic Intervention: First-line

• Evidence:
  • Reduces behavioral symptoms and problematic behaviors in patients
  • Reduces caregiver stress and burden
  • Reduces neuroleptic use and associated side effects without worsening of behaviors
  • Challenges in implementation
• Identify and address triggers as able in patient, caregiver, environment
• Includes:
  • Caregiver Education
  • Environmental changes
  • Behavioral changes

Brodaty and Arasaratnam, Am J Psyc 2012; Gitlin et al., JAMA 2010; Fossey et al., BMJ 2006; Livingston et al., Am J Psyc 2005
Approach to Non-Pharmacologic Interventions

- **DICE:**
  - **Describe** Behavior – context, environment, patient and caregiver perspective, degree of distress
  - **Investigate** Causes of the Behavior
    - Identify and address potential triggers for the behavioral change in the patient, caregiver, environment
    - Iatrogenic, pain, fear, boredom
  - **Create** a plan for intervention with specified follow-up time
  - **Evaluate** if intervention effective and safe

Gitlin and Lyketsos, JAGP 2014; Phan et al., Drugs in R&D 2019
Approach to Non-Pharmacologic Interventions

• Tips:
  • Avoid using logic and reason
  • Avoid criticizing, correcting and arguing
  • Establish a routine to engage in activities they enjoy with caregivers - exercise, arts and craft, household chores, scheduled check ins and visits from friends/family, looking at old photos
    • Set up an activity station for patient where you store items in a central area where the caregiver is still present
  • Communication strategy is key to minimize conflict – make sure to educate family

Gitlin and Lyketsos, JAGP 2014; Phan et al., Drugs in R&D 2019
General Approach to Medication Management

- Pharmacotherapy should be used judiciously in older adults
- Minimize unnecessary treatments, particularly psychotropic medications
  - Beers Criteria
  - Avoid typical neuroleptics and benzodiazepines if at all possible
    - When used, try to limit to one time uses
- Start low and go slow
  - Lowest possible dose and slowly uptitrate
  - 1 medication change at a time
- Use lowest necessary amount of time
  - Consider tapered withdrawal if no response in ~4 weeks
  - Consider trial of taper of antipsychotics after 4 months
- Treat factors that can support cognition
  - B12 deficiency, thyroid disease, renal disease, liver disease, vascular disease
  - Optimizing hearing and vision
Agitation and Aggression

- **Review and treat triggers**
  - Pain, infection, worsening of other medical illnesses, loneliness, boredom, environmental changes, fatigue
  - Remove psychotropic or contributing medications as safely able
- Cholinesterase Inhibitors – 1st line medication
- Memantine – 2nd line
- SSRIs – 3rd line
- Low-dose Atypical Antipsychotics – when dangerous to patient or others, or particularly bothersome
  - Quetiapine (preferred 1st line) or another atypical antipsychotic (2nd line)
    - Quetiapine: Start at 12.5mg nightly, increase as needed

Phan et al., Drugs in R&D 2019
Delusions or Hallucinations

• Remove psychotropic or contributing medications as safely able
• Use medications only when bothersome to patient or puts patient or others at risk of harm
  • Avoid treating hallucinations/delusions when not bothersome or dangerous (risk/benefit)
  • Antipsychotics associated with increased risk of overall mortality in older adults (1.5x)
    • Dose-dependent
• Quetiapine (preferred 1st line) or another atypical antipsychotic (2nd line)
  • Quetiapine: Start at 12.5mg nightly, increase as needed
  • Lowest known risk of mortality
• Avoid typical neuroleptics (such as Haldol) if at all possible – increased risk of mortality
• Avoid benzos if at all possible – increased risk of cognitive impairment and falls

Schneider et al., JAMA 2005; Phan et al., Drugs in R&D 2019
Depression and Anxiety

- SSRIs - 1st line
  - Can continue as long as tolerated
- Avoid Tricyclic Antidepressants if at all possible (anticholinergic effects)
Sleep Disorders

- Nonpharmacologic: Improved sleep hygiene
  - Reduce night-time fluids
  - Reduce caffeine
  - Increase daytime activity and exercise
  - Structured daily activities that minimize naps
  - Create optimal sleep environment – lighting, temperature, sounds, minimize risks
  - Remove psychotropic or contributing medications as safely able
- Medications:
  - Sleep disruption: Acetylcholinesterase inhibitor, melatonin (limited evidence), trazodone
    - Avoid benzodiazepines for this indication
  - REM Sleep Behavior Disorder: High-dose melatonin (1st line – up to 15mg if needed), benzodiazepine (2nd line)
Symptomatic Management
Pain

- Acetaminophen – can take around the clock
- NSAIDs – 2nd line
- End of life – Low dose opiates
Decreased Appetite (Anorexia/Cachexia)

- Part of the disease process and often more distressing to family than patients:
  - Loss of appetite is nearly universal among terminally ill patient
  - Cultural influences response
- Non-pharmacologic:
  - Provide appealing foods, favorite meals, and feeding assistance
  - Make sure the person is in a comfortable, upright position during feeding and ideally for 30 minutes afterwards for digestion
  - Adapt foods as needed for swallowing difficulty: soft, diced, grinded, pureed
  - Offer several smaller meals throughout the day
  - Clarify goals and expectations - “Their body does not need as much as they used to, and that’s ok”
- Can consider mirtazapine, but typically do not recommend medications as no evidence of improved outcomes
- **Do not recommend forcing foods or placing feeding tubes**
  - No evidence for improved outcomes (QOL or life expectancy)
Incontinence

- Non-pharmacologic:
  - Set a routine toileting schedule – keep record of when they go to see their natural routine, then schedule regular toileting intervals
  - Reduce liquids 2 hours before bedtime
  - Use a bedside urinal/commode
  - Use pads/briefs for occasional accidents
  - Use easy to remove pants
  - Avoid reacting negatively to accidents
End-of-Life

- Pain:
  - Acetaminophen – can take around the clock
  - NSAIDs – 2\textsuperscript{nd} line
  - Opiates – start low dose and increase as needed
- Agitation:
  - Atypical Antipsychotics – 1\textsuperscript{st} line
  - Benzodiazepines
- Dyspnea: Opiates for subjective improvement of breathlessness
- Continue acetylcholinesterase inhibitors, SSRIs, carbidopa/levodopa as long as possible to optimize quality of life
Resources

• Care Ecosystem: [memory.ucsf.edu/Care-Ecosystem](memory.ucsf.edu/Care-Ecosystem)
  • Amazing compilation of resources for dementia care and education:
    • [https://ucsf.app.box.com/v/CurriculumFINAL](https://ucsf.app.box.com/v/CurriculumFINAL)
    • Free Care Ecosystem self-paced online training course on the Canvas Network (you can also join using code R9B67G)

• Alzheimer’s Association: [https://www.alz.org/alzheimer_s_dementia](https://www.alz.org/alzheimer_s_dementia)

• AAN Continuum and Continuum Audio:
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I have no conflicts of interest in relation to this presentation

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  • memory.ucsf.edu/Care-Ecosystem