

Management of Advanced Neurodegenerative Diseases: Palliative and Supportive Care

American Academy of Neurology



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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!



Palliative and Supportive Care

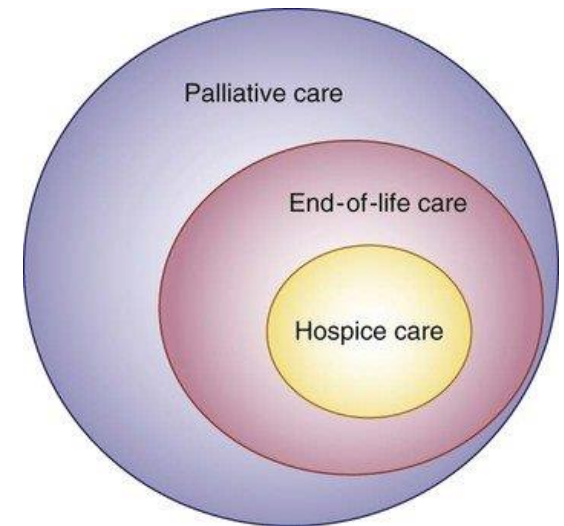
"Our most cruel failure in how we treat the sick and the aged is the failure to recognize that they have priorities beyond merely being safe and living longer"

Atul Gawande, Being Mortal



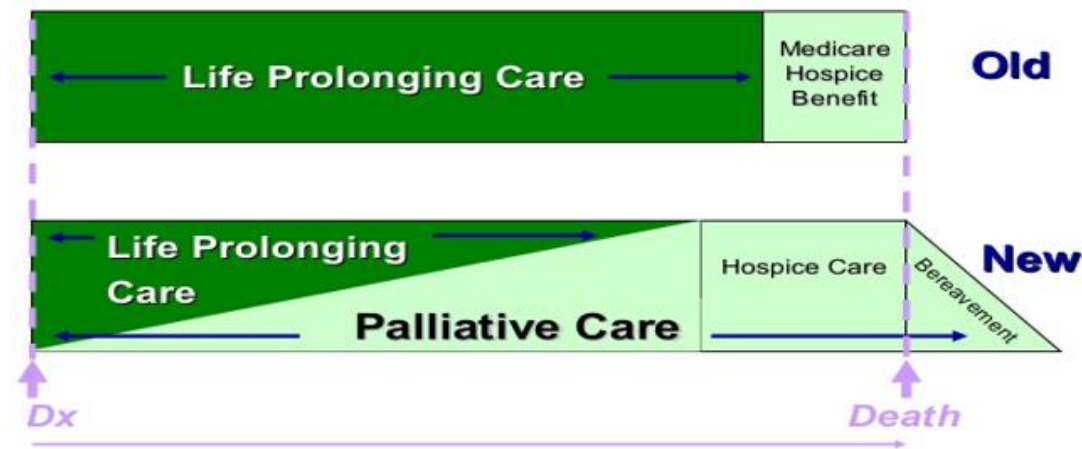
Overview

- 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

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

Term	Definition	Communication examples
Setting	Establish an appropriate setting	Minimize disruptions, have easy access to tissues
Perception	Establish the patient's and caregiver's perception of their symptoms and health status	<p>Ask questions such as:</p> <p>"Can you give me a sense of what you think might be causing your symptoms?"</p> <p>"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."</p>
Invitation	Obtain permission from the patient and caregiver to provide the difficult medical news	<p>Ask questions such as:</p> <p>"Would it be okay if I gave you my thoughts on where things are with your illness?"</p> <p>Assess how much information about the illness is desired. Ask questions such as:</p> <p>"Would it be okay for me to share my thoughts on what's causing your symptoms?"</p>
Knowledge	Give a warning shot to allow the patient/caregiver to brace for bad news	<p>"I have difficult news"</p> <p>Deliver the serious illness diagnosis in a bite-size chunk</p> <p>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</p>
Emotions	Expect and validate the difficult emotions that will arise after hearing bad medical news with empathic communication	"That's so difficult to hear. How are you doing with that information?"
Strategy	Develop a strategy for the future	"Let's focus on helping you live as well as you can for as long as you can."

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”

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

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

Modified Hoehn and Yahr for Parkinson's

Stage	Modified Hoehn and Yahr Scale
1	Unilateral involvement only
1.5	Unilateral and axial involvement
2	Bilateral involvement without impairment of balance
2.5	Mild bilateral disease with recovery on pull test
3	Mild to moderate bilateral disease; some postural instability; physically independent
4	Severe disability; still able to walk or stand unassisted
5	Wheelchair bound or bedridden unless aided

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%

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

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

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

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

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

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

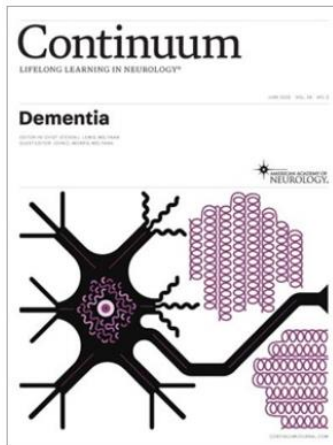
- Non-pharmacologic:
 - Set a routine toileting schedule – keep record of when they go to see their natural routine, then schedule regular toileting intervals
 - Reduce liquids 2 hours before bedtime
 - Use a bedside urinal/commode
 - Use pads/briefs for occasional accidents
 - Use easy to remove pants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

End-of-Life

- Pain:
 - Acetaminophen – can take around the clock
 - NSAIDs – 2nd line
 - Opiates – start low dose and increase as needed
- Agitation:
 - Atypical Antipsychotics – 1st 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
 - Amazing compilation of resources for dementia care and education:
 - <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
- AAN Continuum and Continuum Audio:



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I have no conflicts of interest in relation to this presentation

Thank You

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