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FAMILY MEDICINE

Division of  
Palliative Care

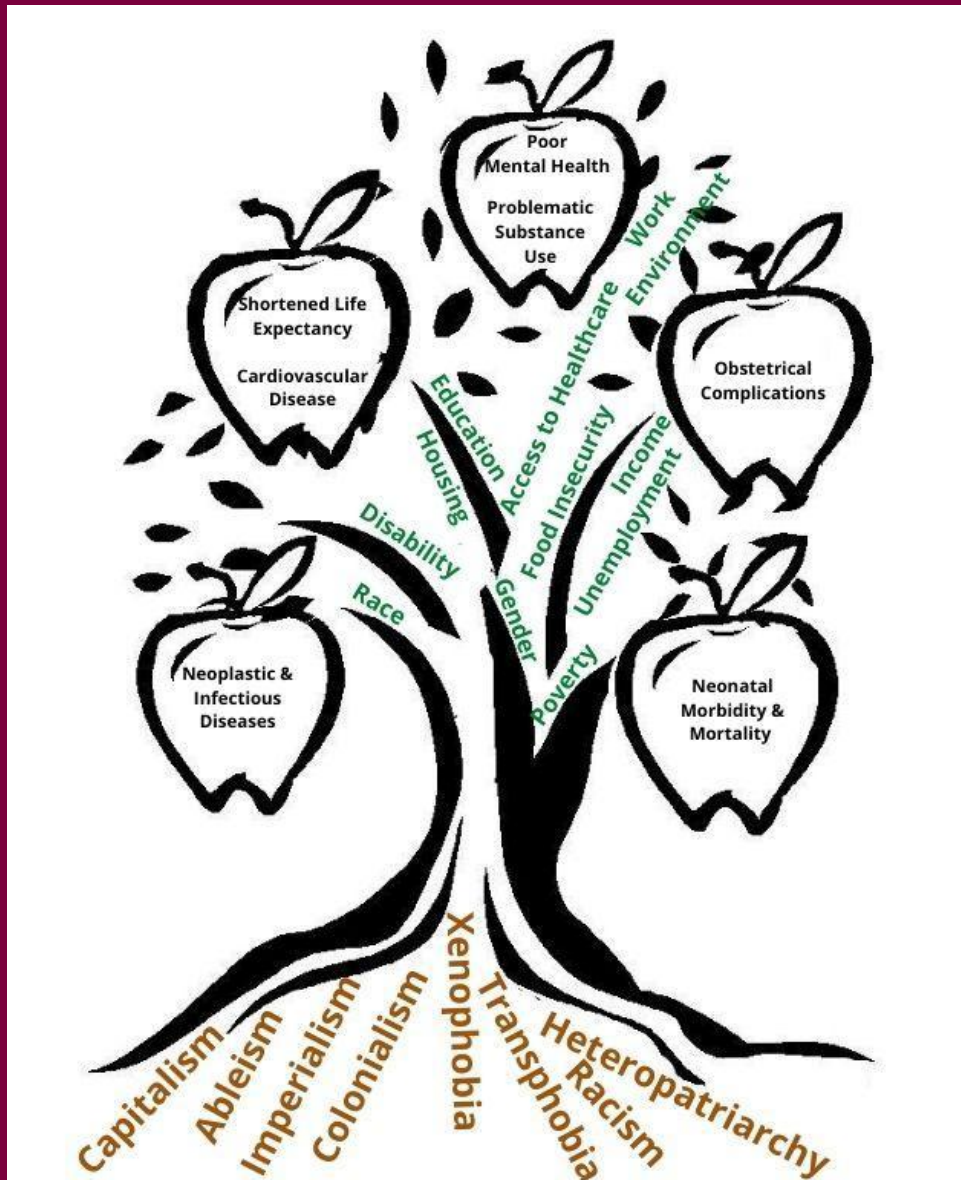
@McMasterFamMed

# Palliative Care in Sickle Cell Disease: A Missed Opportunity?

April 16, 2026

Alexandra Farag and Sarah Patterson





# Acknowledgements

Rai, 2017

Family Medicine

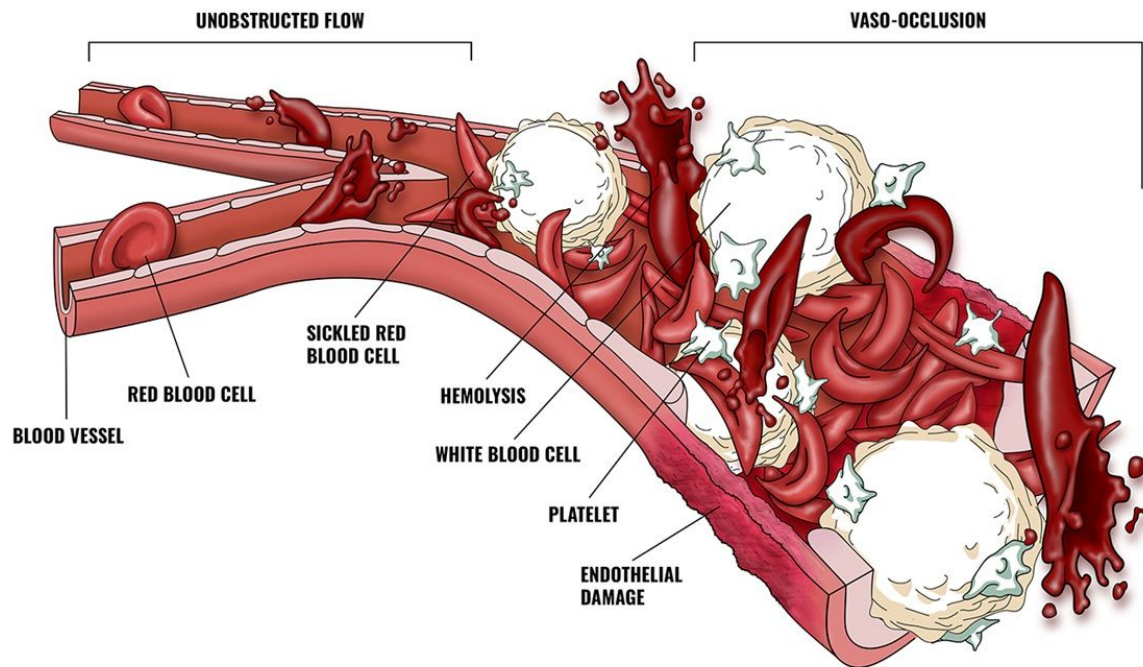
# Learning Objectives

By the end of this session, participants will be able to:

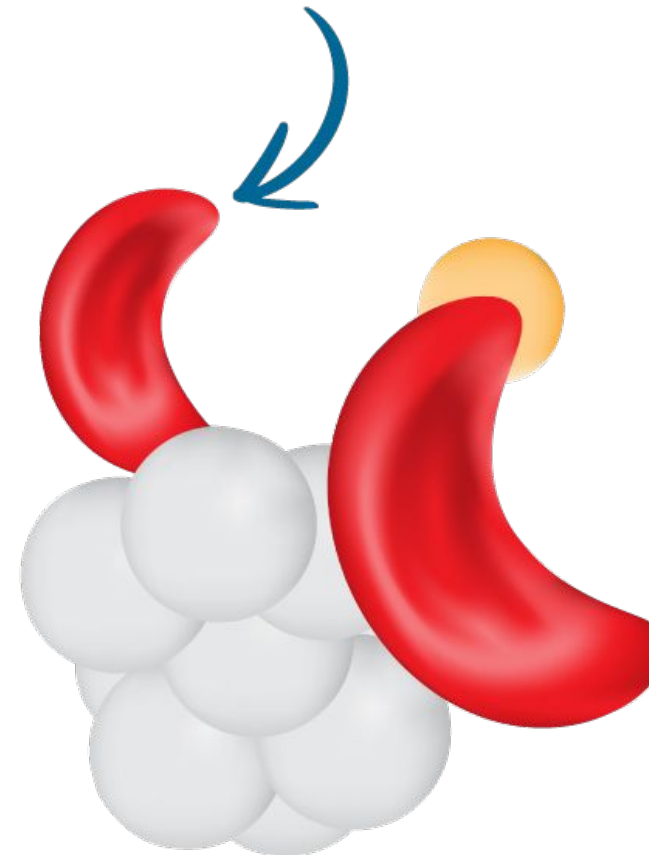
- Describe sickle cell disease and provide a brief overview of disease modifying treatment
- Understand how patients with sickle cell disease may benefit from a specialist palliative care
- Reflect on how you may be able to care for patients with sickle cell disease in your own practice

# Sickle Cell Disease

# Sickle Cell Disease



Sickle cell anemia



Kato, 2018

# Complications from SCD

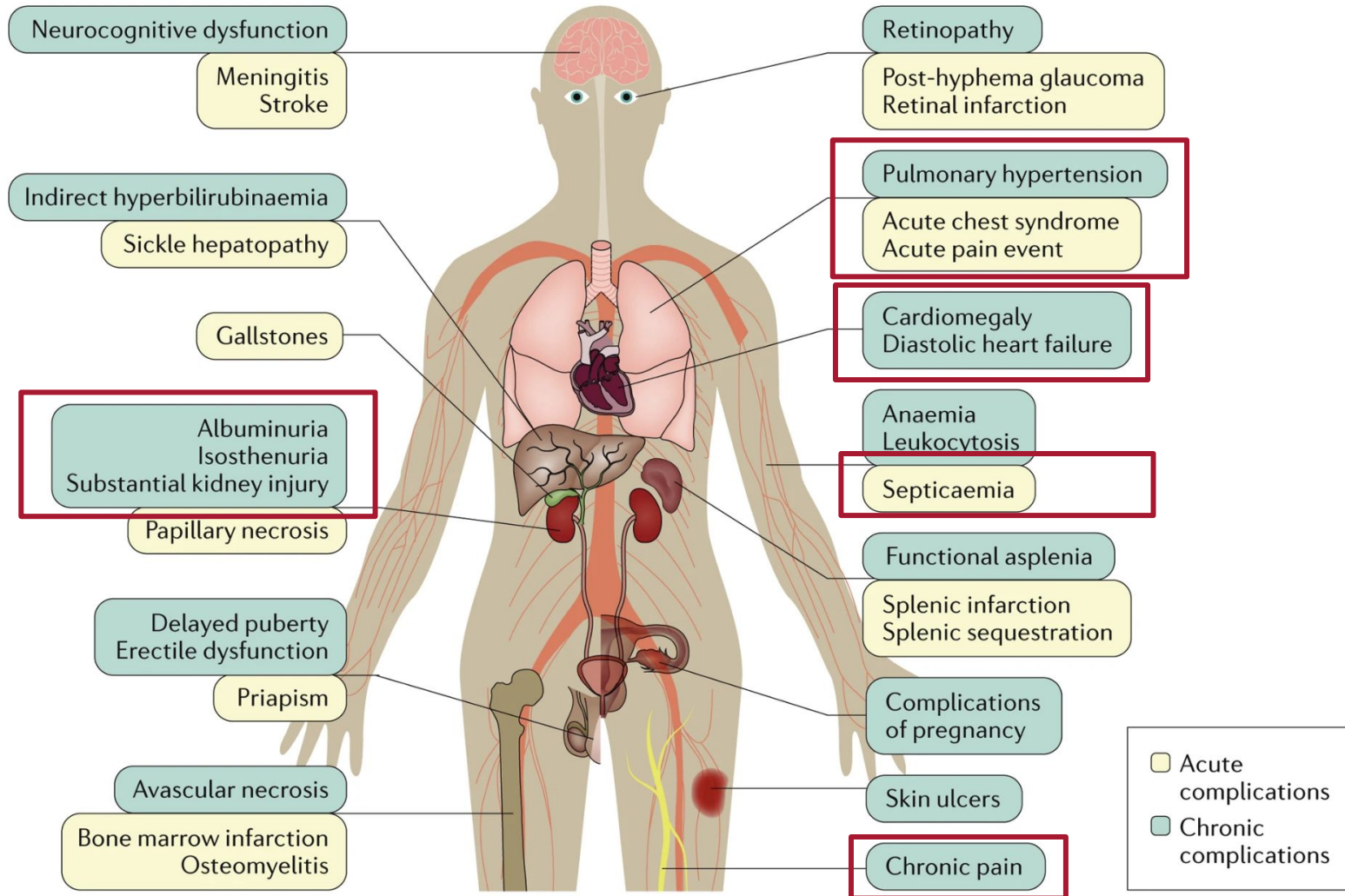
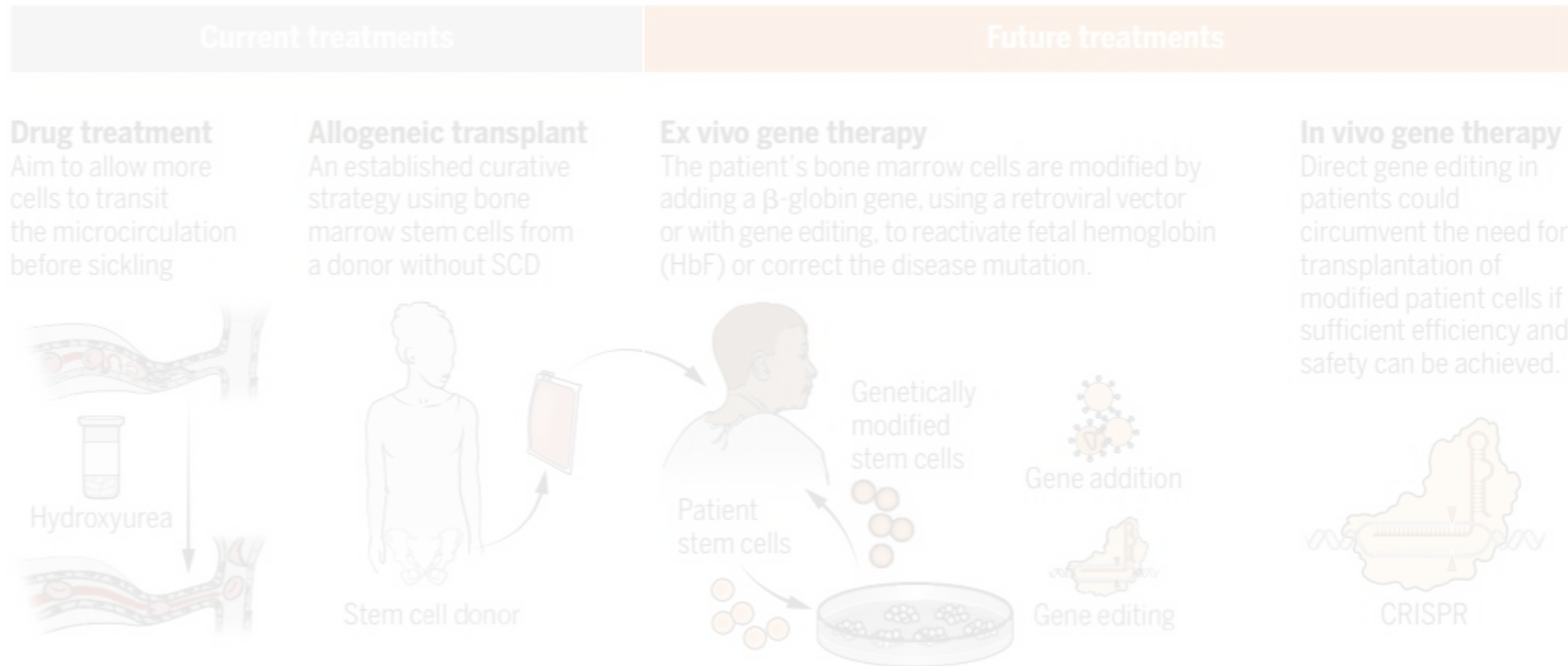


Image source: <https://www.nature.com/articles/nrdp201810>

# Current and future treatments for sickle cell anemia

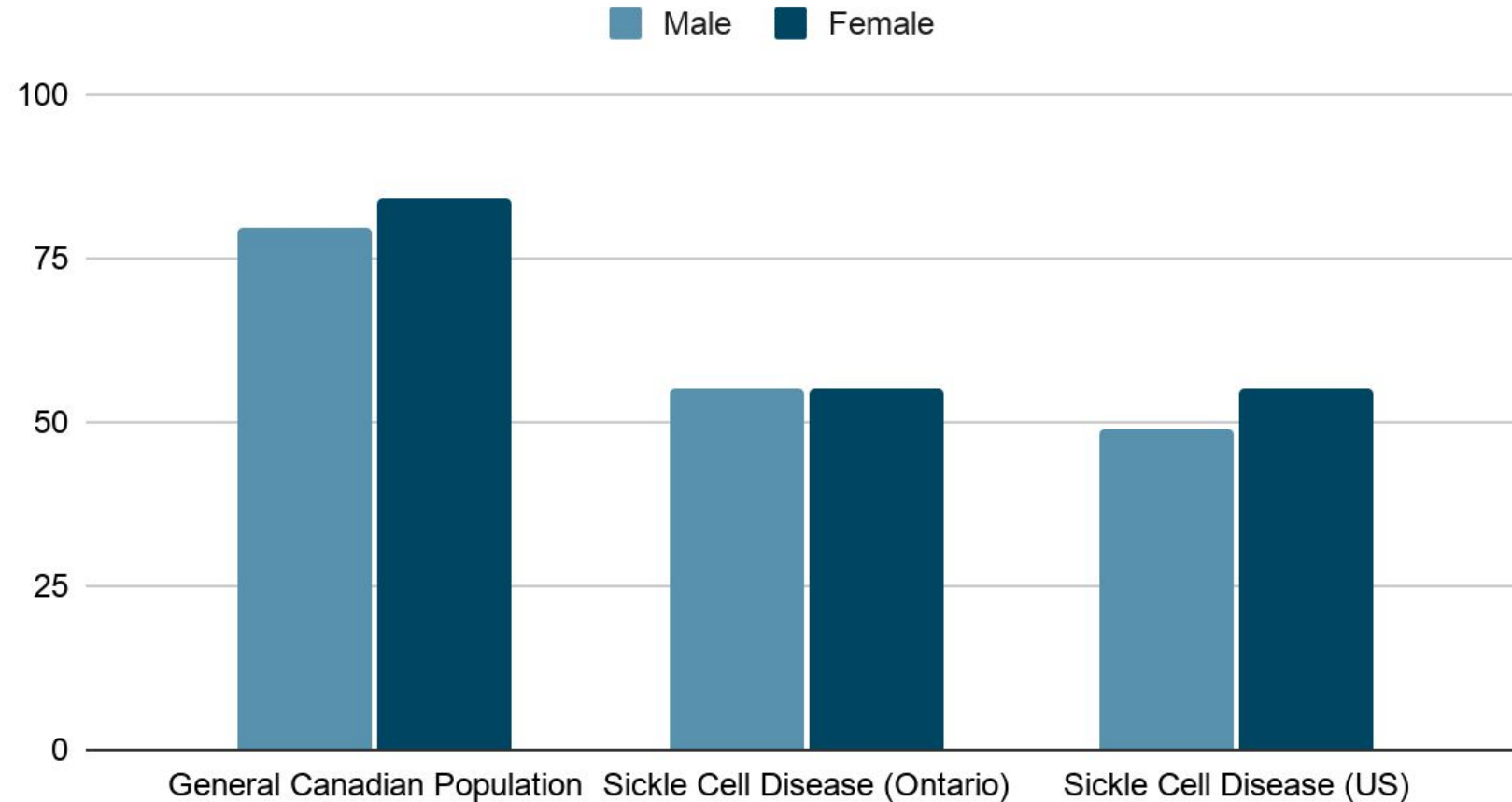
Numerous advances in the understanding of sickle cell disease (SCD) have allowed the development of curative therapies through allogeneic stem cell transplantation, with the promise of gene therapy-based treatments in the future.



# Why Sickle Cell Disease?

# Life- limiting illness

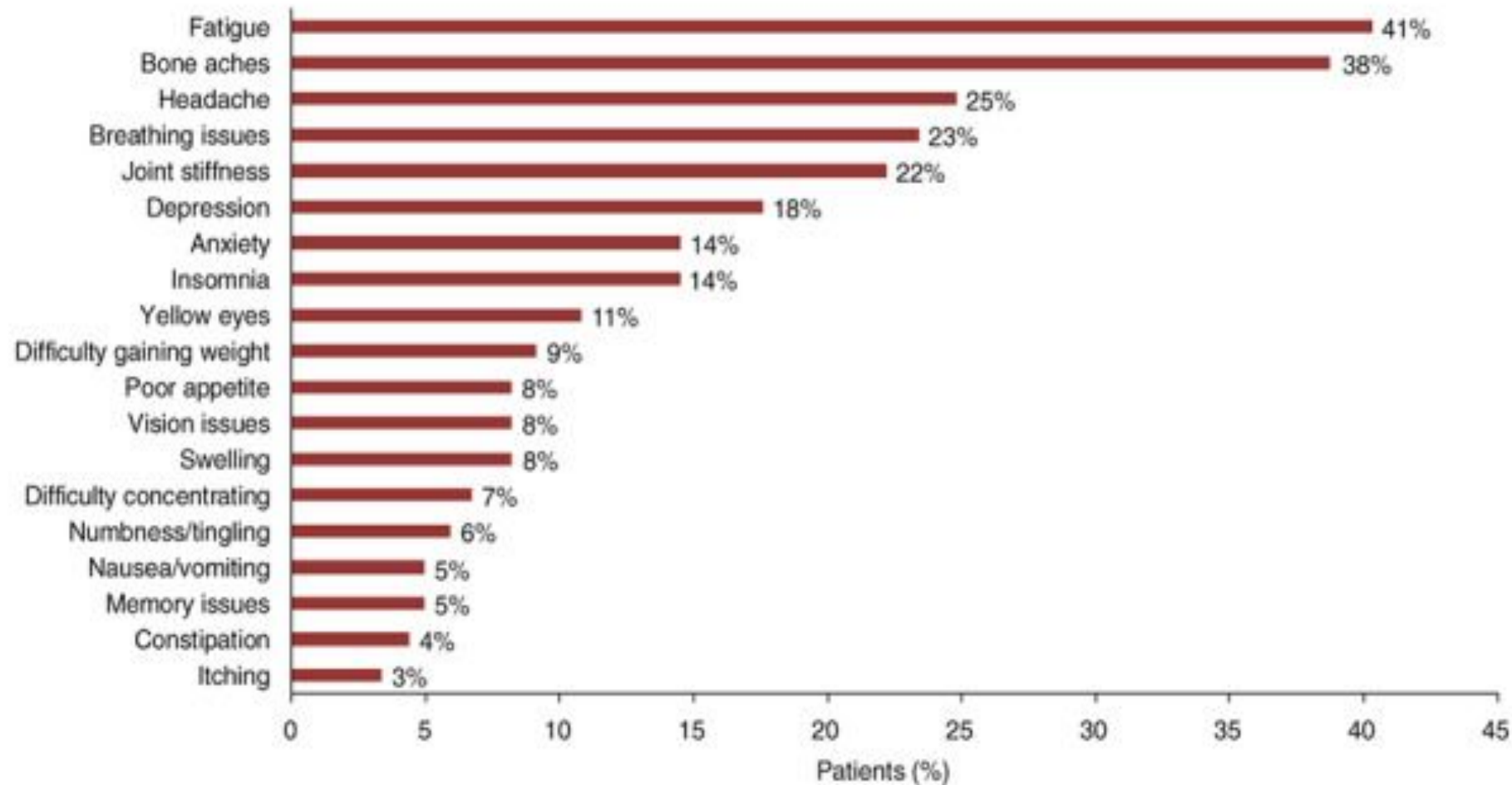
## Average Life Expectancy



People with SCD die in emergency departments and acute care settings (not at home or in palliative care settings)

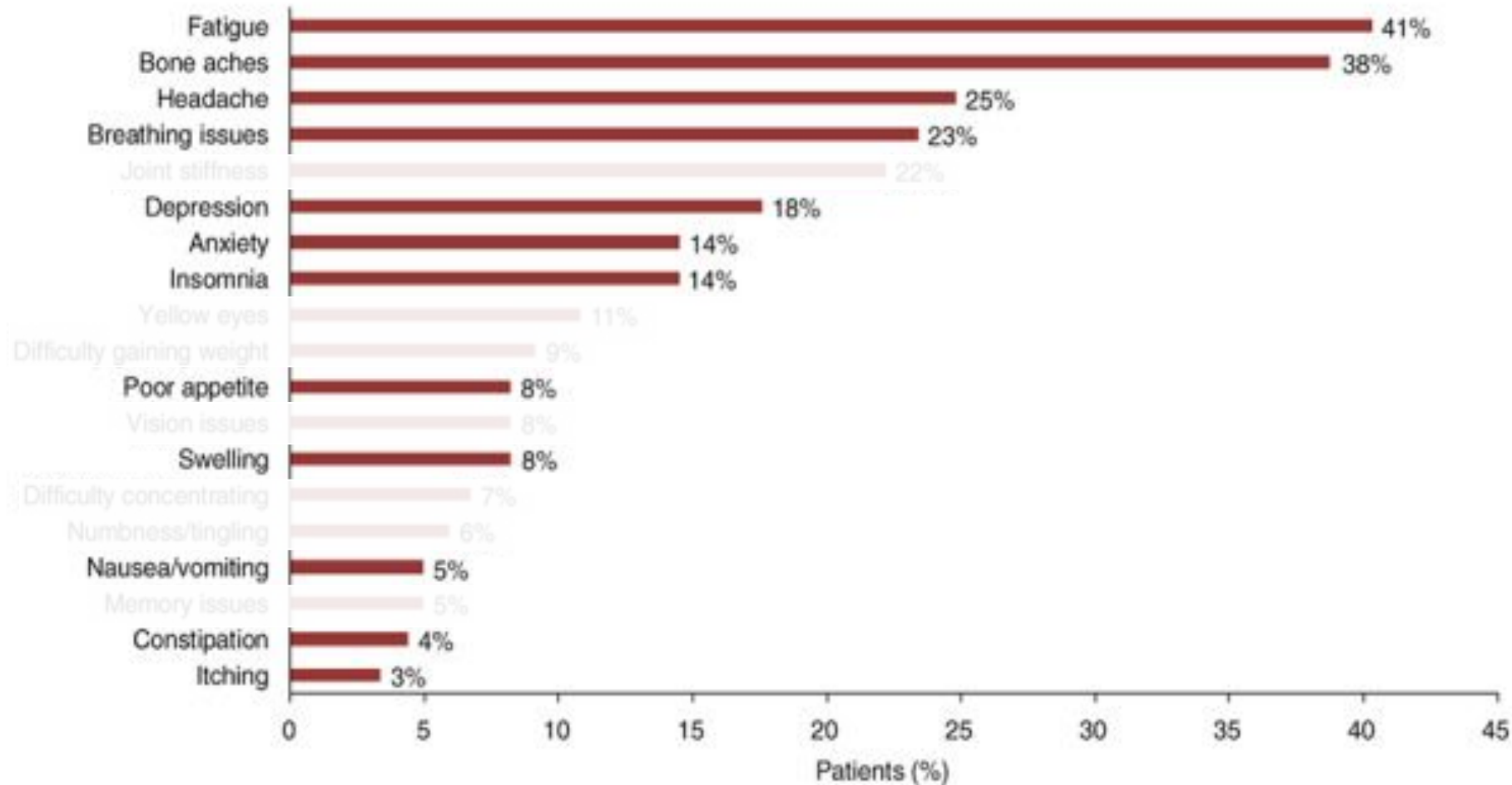
*Boshen, 2023, Pendergast, 2023, Lily 2026*

# Large and Complex Symptom Burden



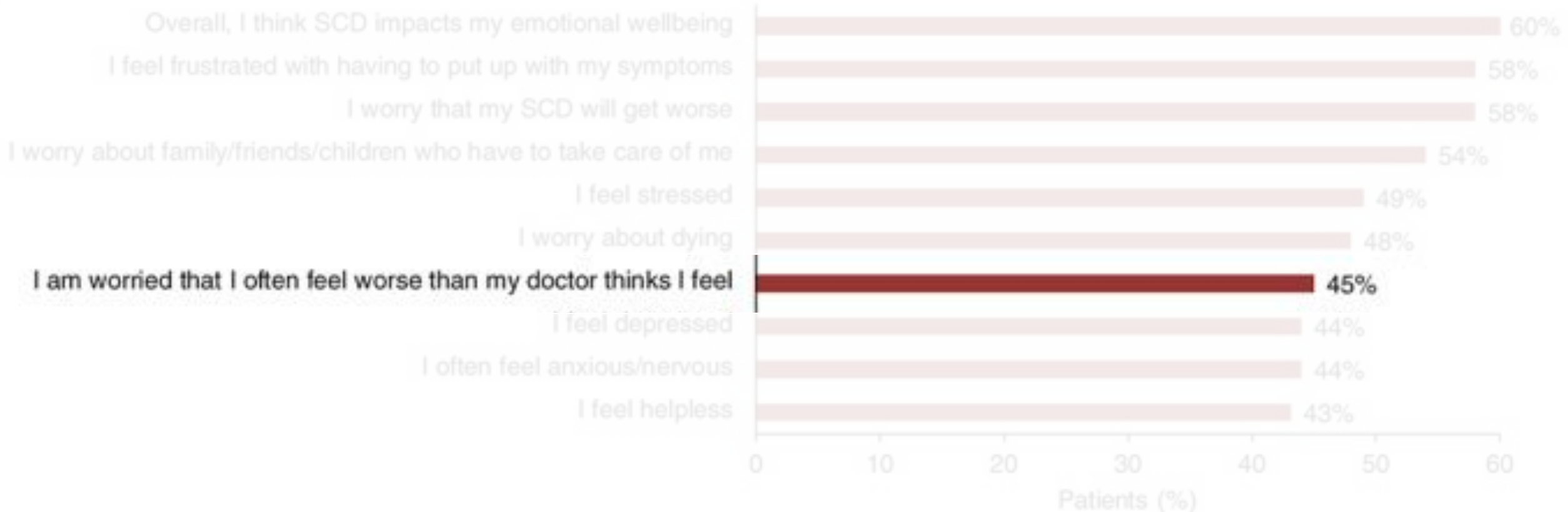
Osunkwo, 2021

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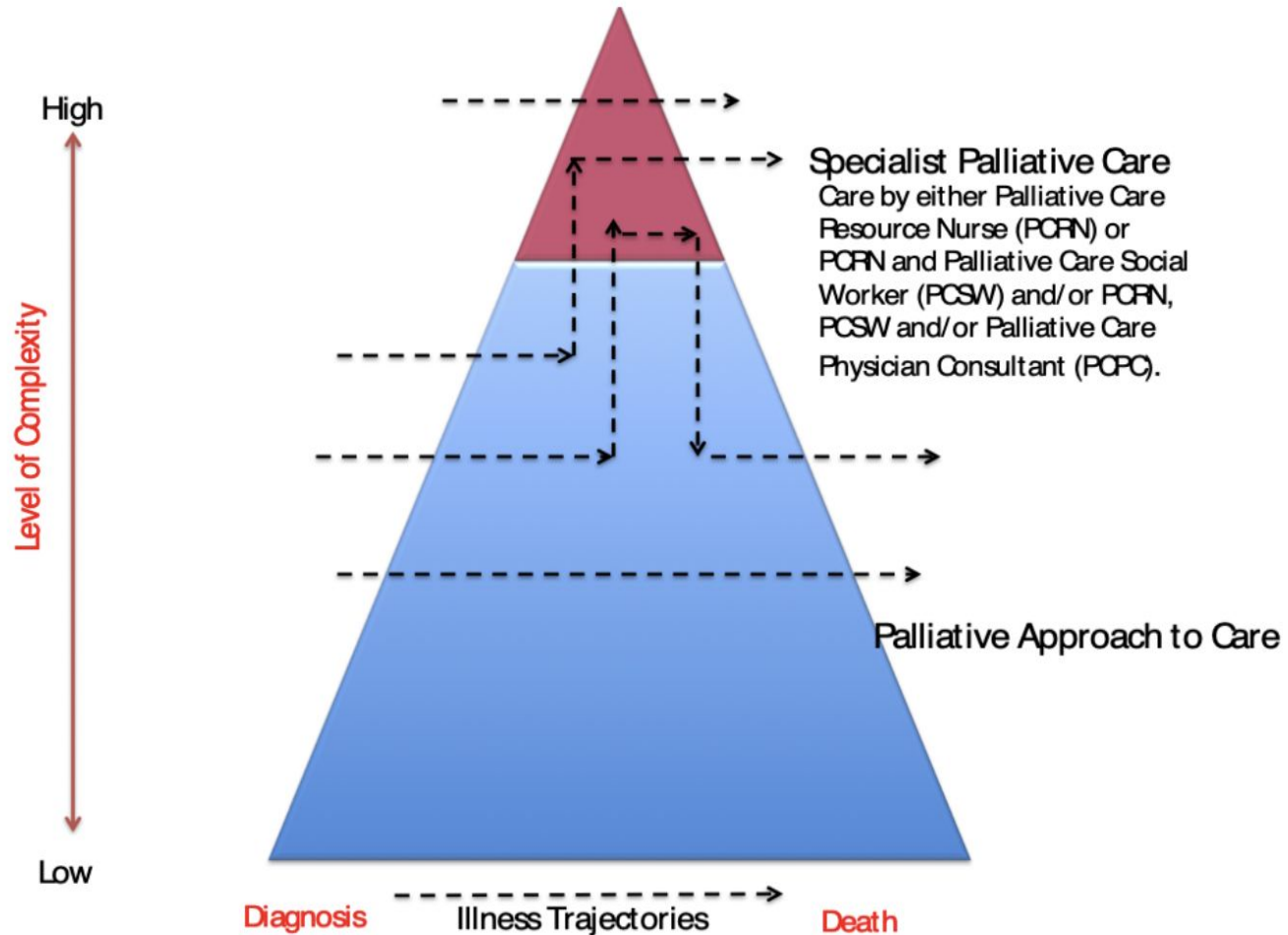


Osunkwo, 2021

# Impact on Quality-of- Life



# Access to Specialist Palliative Care



< 0.5% of patients with SCD receive a palliative care consultation during admission

Adapted from LEAP Core, Pallium Canada

*Nwogu-Onyemkpa, 2022*

What can we (as palliative care  
specialists) do?

Clarifying illness understanding

Supporting decision making

Symptom assessment and management

Anticipatory guidance and care planning

Supporting our colleagues

Rebuilding trust





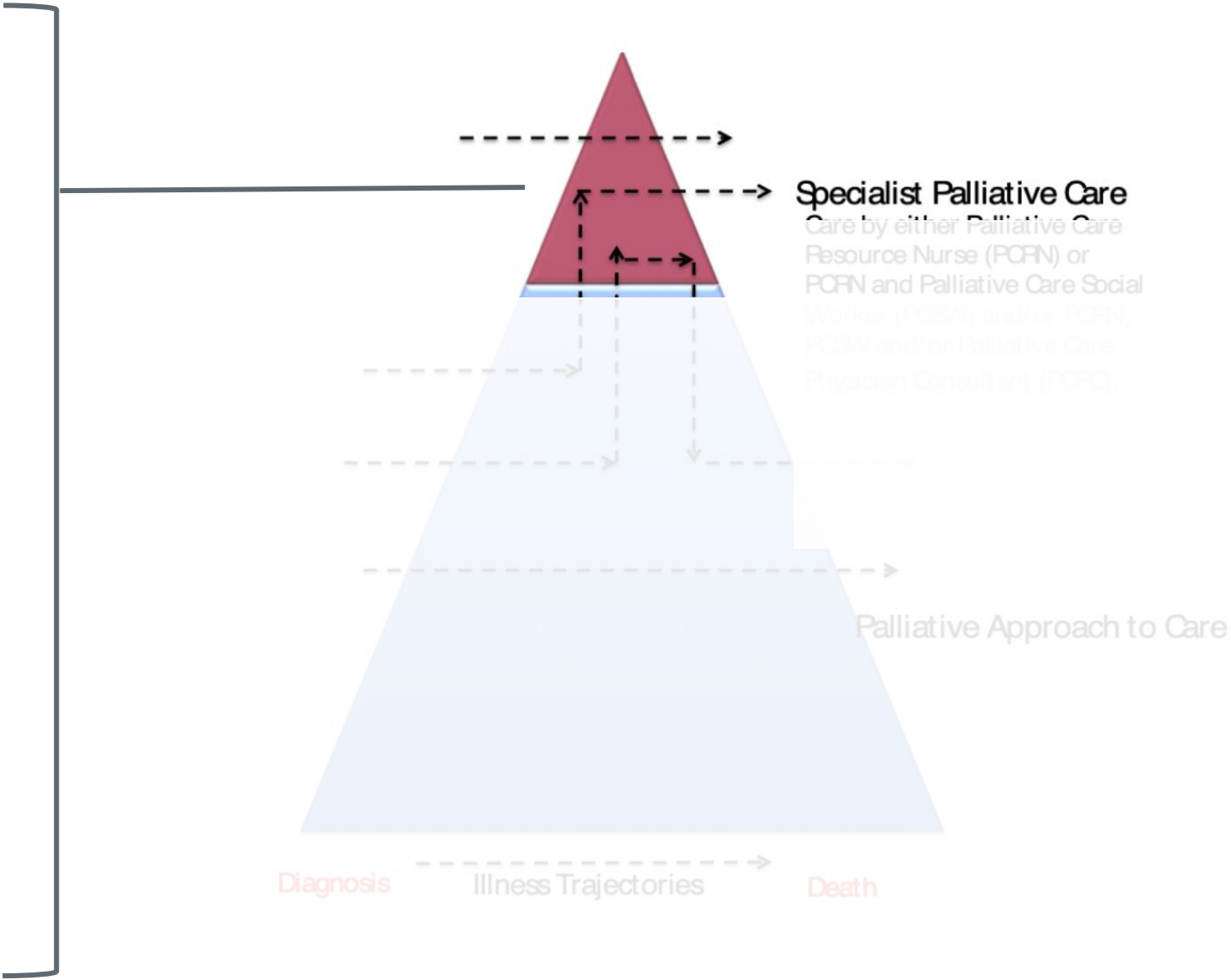
# Illness Trajectory

8% die “suddenly and unexpectedly”

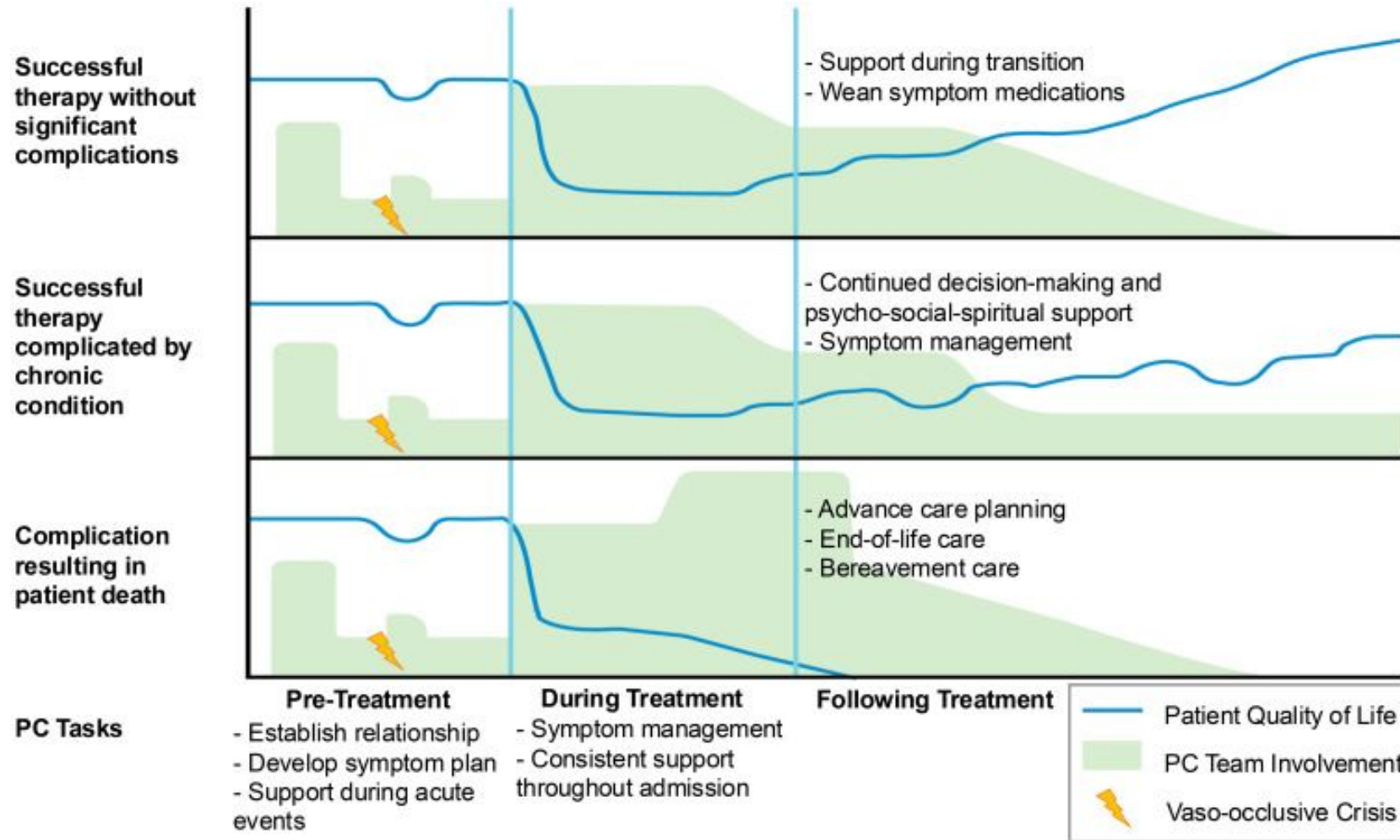


Repeat hospitalizations (VOC, line issues, clots, infections, volume overload)

- Clarifying illness understanding
- Supporting decision making**
- Symptom assessment and management
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# Supporting Decision Making



Collins et al, 2025

- Clarifying illness understanding
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# Symptom Assessment and Management

**Vaso-Occlusive Pain**

Recurrent episodes of hypoxic-ischemic reperfusion injury to bones and tissues due vaso-occlusion. This results in inflammation and tissue damage leading to repetitive nociceptive injury.

**Central Sensitization**

Increased responsiveness of nociceptors to normal painful and non-painful stimuli. Hypersensitivity to pain: both allodynia and hyperalgesia.

**Hyperalgesia**  
Increased pain perception incongruent to degree of noxious stimuli.

**Allodynia**  
Pain from a stimulus that does not normally cause pain such a light touch, pressure, heat, or cold.

**Opioid-Induced Hyperalgesia**  
Abnormally enhanced pain sensitivity to a familiar painful trigger that is associated with long-term opioid use. It causes generalized pain "all over".

**Neuropathic Pain**  
Pain caused by damage or disease to the somatosensory nervous system. Heightened response to both painful and non-painful stimuli.

**Cyclic Opioid Withdrawal**

Self-perpetuating cycle of pain exacerbation & opioid withdrawal triggered by abrupt discontinuation or tapering of opioids after prolonged use or high doses. Often under-recognized by providers and patients.

# Symptom Assessment and Management

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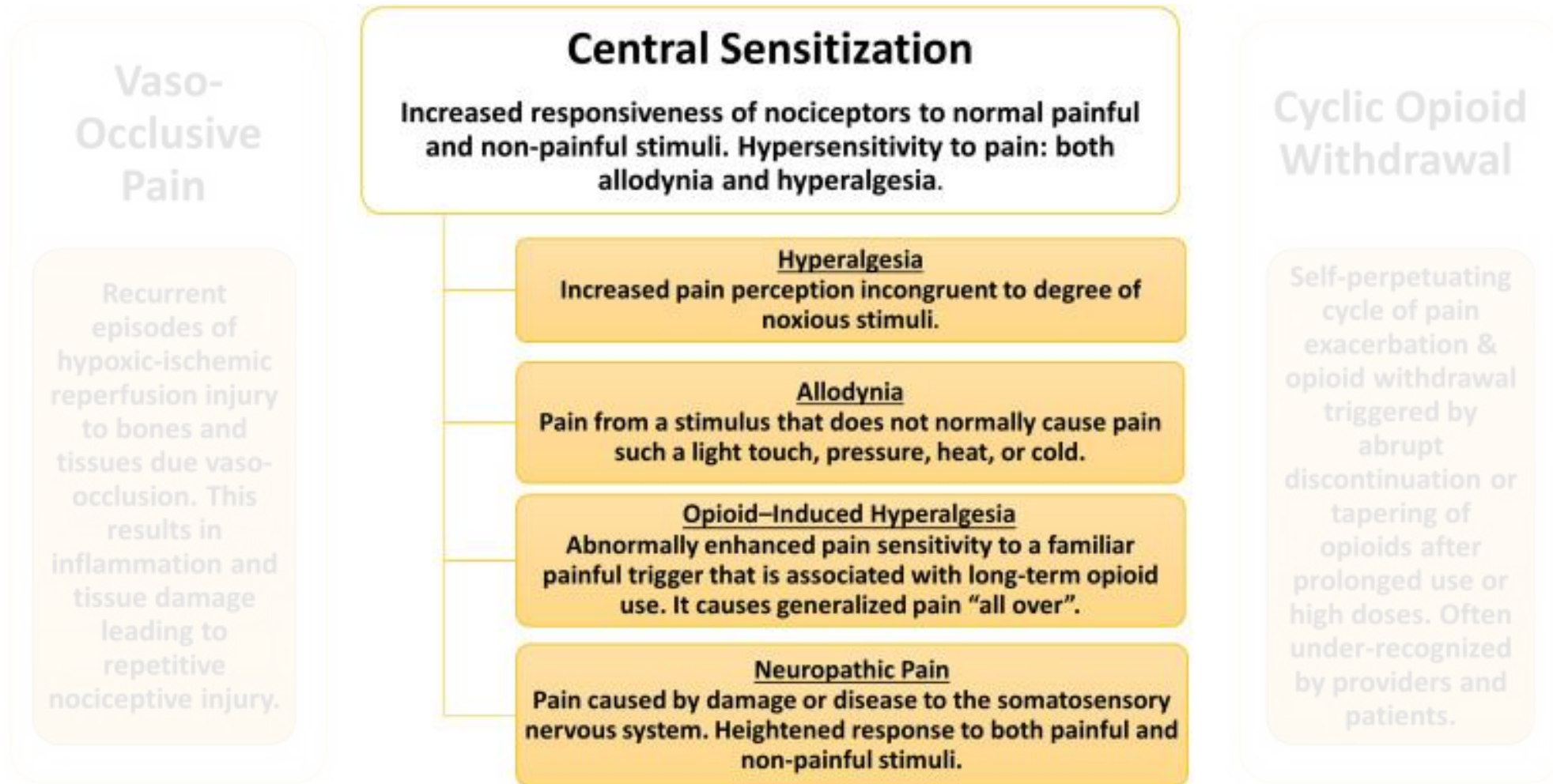
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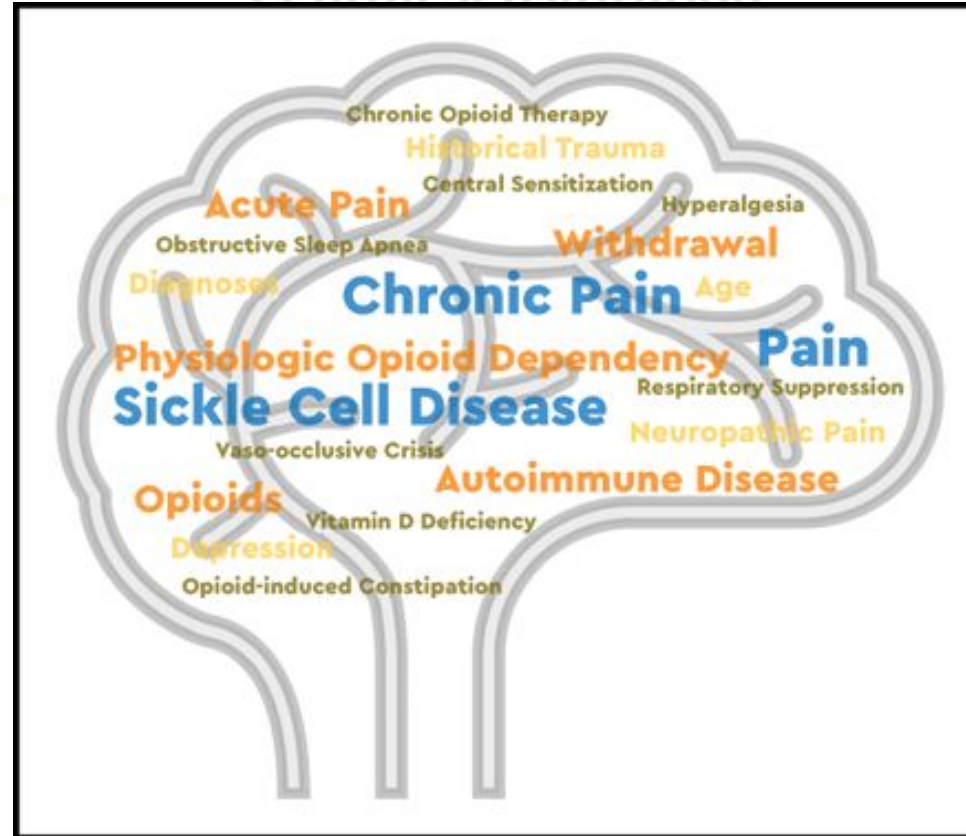
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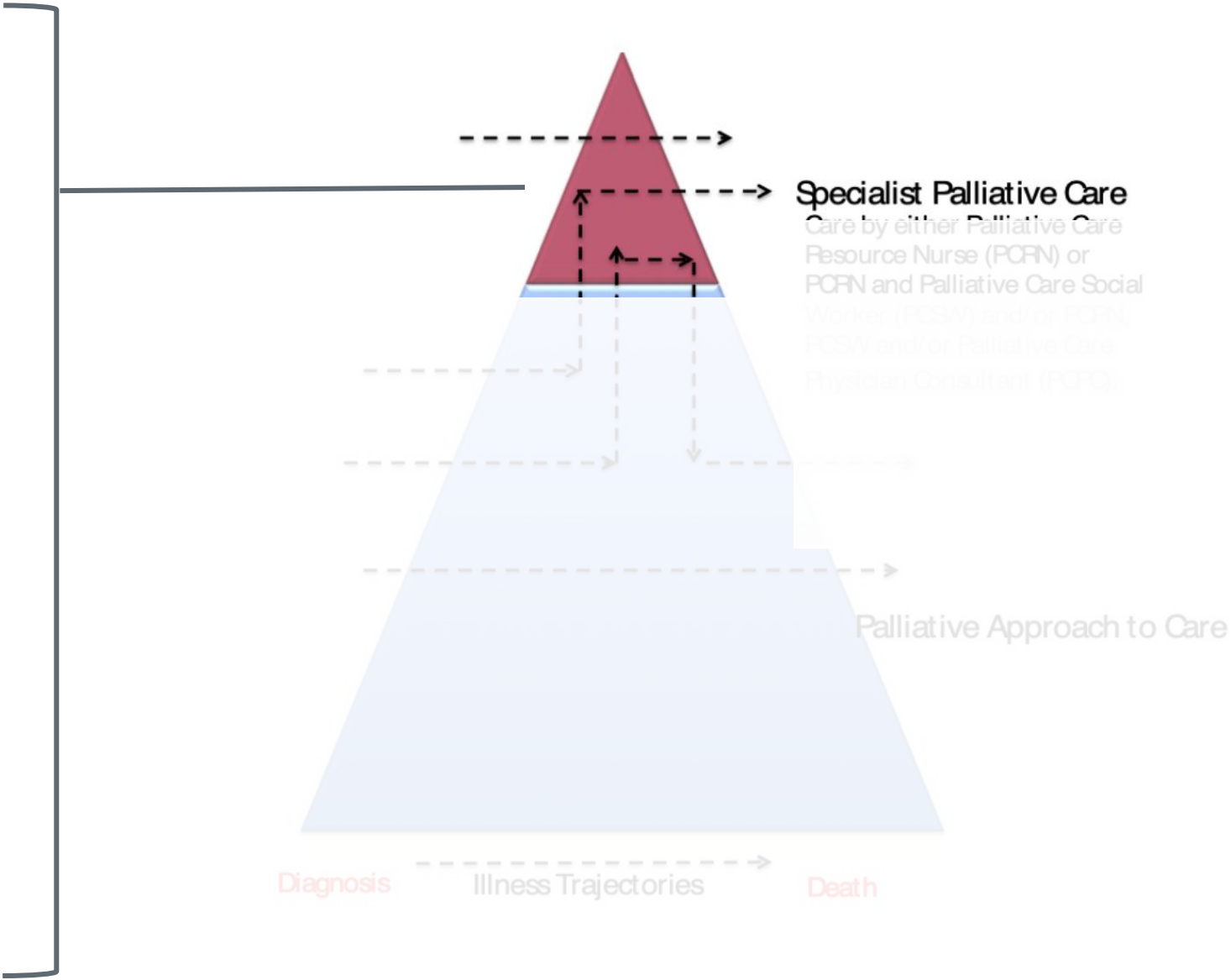


nervous system. Heightened response to both painful and non-painful stimuli.

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Self-perpetuating cycle of pain exacerbation & opioid withdrawal triggered by abrupt discontinuation or tapering of opioids after prolonged use or high doses. Often under-recognized by providers and patients.

- Clarifying illness understanding
- Supporting decision making
- Symptom assessment and management
- Anticipatory guidance and care planning**
- Supporting our colleagues
- Rebuilding trust

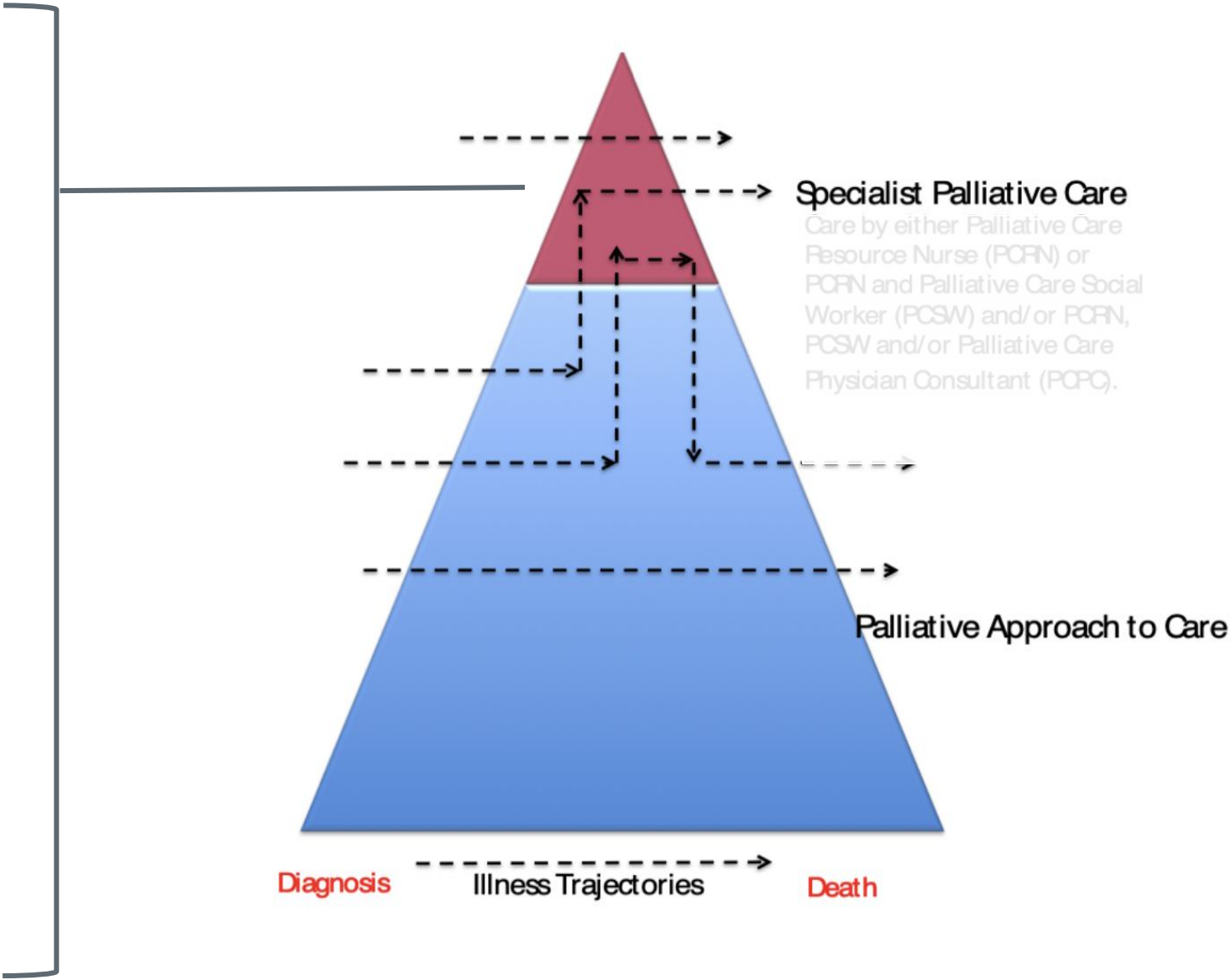


# Anticipatory Guidance and Care Planning

## INDIVIDUALIZED ER and INPATIENT CARE PLAN:

1. Patient has **VORTEX PORT** - Detailed instructions for care and accessing below. **MUST BE ACCESSED BY TRAINED NURSING STAFF, USING STERILE TECHNIQUE.**
2. **Analgesia (multidisciplinary plan which has been developed in consultation with hematology, medicine, and palliative medicine, with prior advice from substance use):**
  1. Continue **home doses of suboxone** as prescribed. (As of Jan 2026 dose is XX). No PRN suboxone. Routine prescription provided by the Hemoglobinopathy clinic.
  2. Start **Ketamine** 0.15mg/kg IV q4h scheduled (last weight 73.5kg, so 11 mg dose). Infusion contraindicated due to sedation.
    1. If dose ineffective, increase to 0.2mg/kg IV q4h scheduled (i.e. 14mg dose). May increase to 0.25 mg/kg IV q 4h scheduled (i.e. 18 mg dose) to a maximum of 0.3mg/kg IV q4h scheduled (i.e. 22 mg dose).
    2. Add Haldol 0.25mg subcut/po BID prn for dissociative symptoms, noted particularly at the 0.3mg/kg dose.
  3. **No opioids PRN** (central sleep apnea and history of over sedation), specifically no IV opioids. She has had multiple code blue events due to respiratory suppression previously.

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# Supporting Colleagues

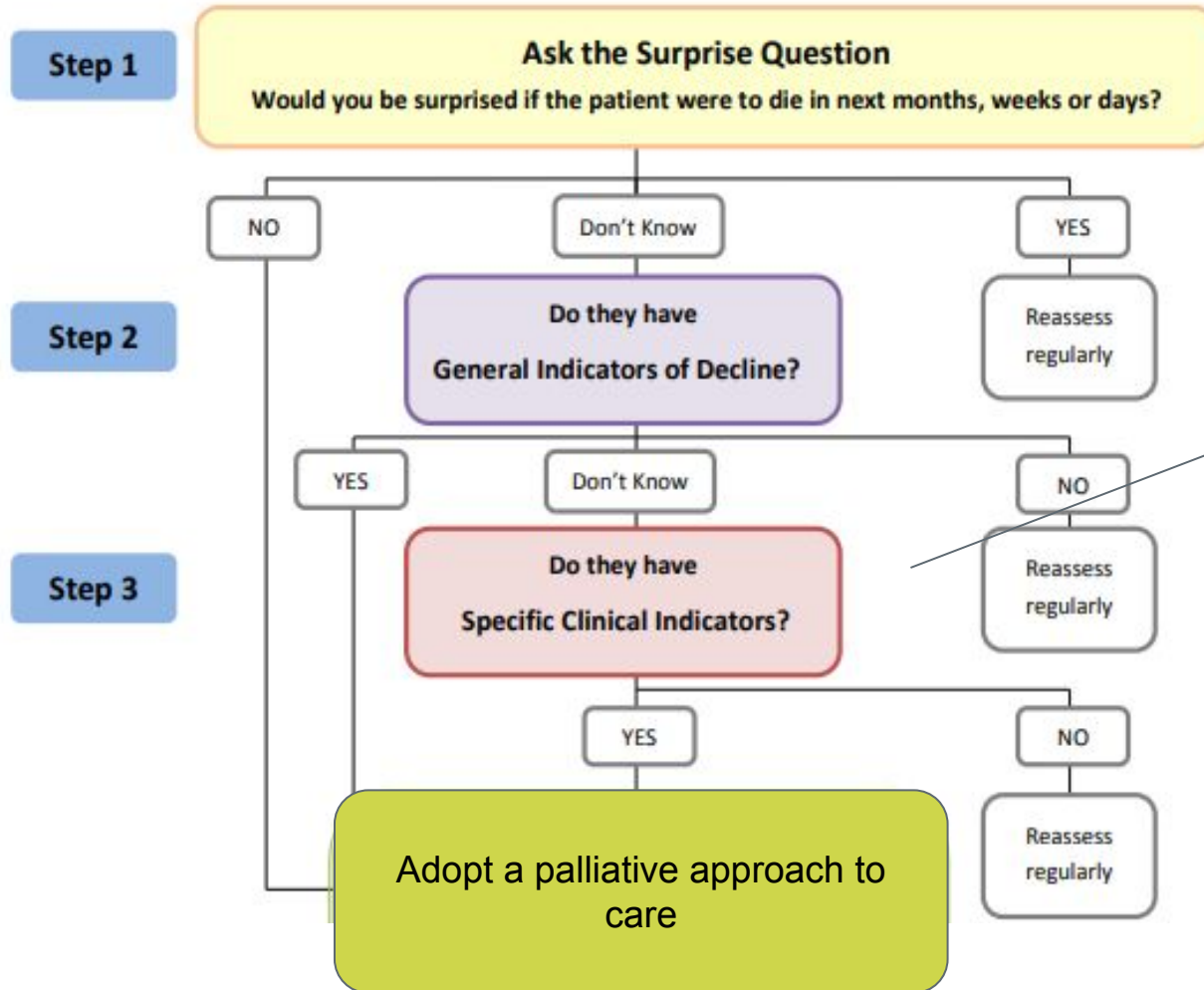


**When do we  
adopt a palliative  
approach to  
care?**

**How do we  
better assess  
symptoms?**

**How can we  
talk about  
illness?**

# Early Identification



## Sickle Cell Disease Specific Indicators:

- > 5x/year ER visits and hospitalizations
- Iron overload
- Pulmonary hypertension
- Reticulocyte count > 6-10% (depending on study)
- Low fetal hemoglobin (unclear cut off)

Adapted from GSF 2022 Maitra, 2017

# Symptom Assessment Tools

Edmonton Symptom Assessment System:  
(revised version) (ESAS-R)

Date: \_\_\_\_\_

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Please circle the number that best describes how you feel NOW:

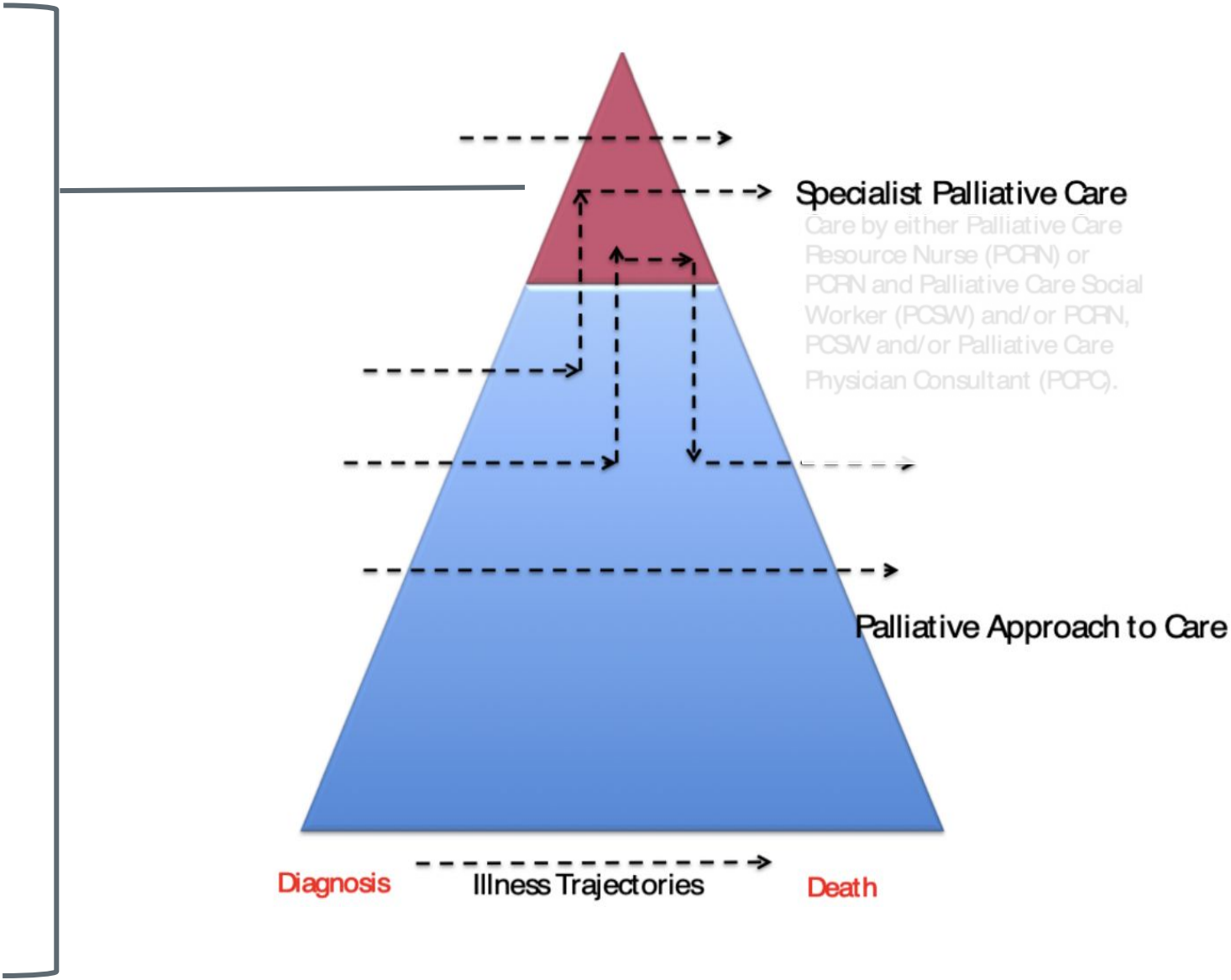
No Pain	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Pain
No Tiredness <i>(Tiredness = lack of energy)</i>	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Tiredness
No Drowsiness <i>(Drowsiness = feeling sleepy)</i>	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Drowsiness
No Nausea	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Nausea
No Lack of Appetite	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Lack of Appetite
No Shortness of Breath	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Shortness of Breath
No Depression <i>(Depression = feeling sad)</i>	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Depression
No Anxiety <i>(Anxiety = feeling nervous)</i>	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Anxiety
Best Wellbeing <i>(Wellbeing = how you feel overall)</i>	0	1	2	3	4	5	6	7	8	9	10	Worst Possible Wellbeing
No _____ Other Problem <i>(for example constipation)</i>	0	1	2	3	4	5	6	7	8	9	10	Worst Possible _____

Add cognitive changes, constipation, itching for the SCD population

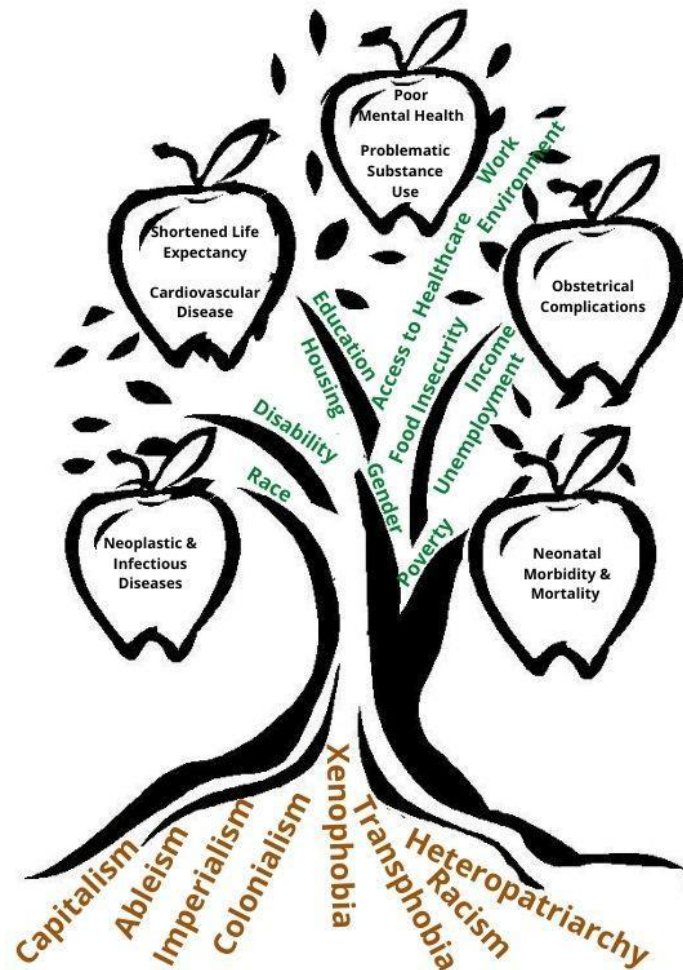
# Serious Illness Conversation Guides

Conversation flow	Patient-tested language
<b>1. Set up the conversation</b> <ul style="list-style-type: none"> <li>• Introduce purpose</li> <li>• Ask permission</li> </ul>	<b>Set Up</b> <p>"I'd like to talk about what is ahead with your illness and do some thinking in advance about what is important to you so that I can make sure we provide you with the care you want — <b>is this okay?</b>"</p>
<b>2. Assess illness understanding &amp; information preferences</b>	<b>Assess</b> <p>"What is your <b>understanding</b> now of where you are with your illness?"            "How much <b>information</b> about what is likely to be ahead with your illness would you like from me?"</p>
<b>3. Share prognosis</b> <ul style="list-style-type: none"> <li>• Frame with a "wish...worry", "hope...worry" statement</li> <li>• Allow silence, explore emotion</li> </ul>	<b>Share</b> <p>Prognosis: "I want to share with you <b>my understanding</b> of where things are with your illness..."            Uncertain: "It can be difficult to predict what will happen with your illness. I <b>hope</b> you will continue to live well for a long time but I'm <b>worried</b> that you could get sick quickly, and I think it is important to prepare for that possibility." Time: "I <b>wish</b> we were not in this situation, but I'm <b>worried</b> that time may be short as_ (express as a range e.g. weeks to months, months to a year)." OR Function: "I <b>hope</b> that this is not the case, but I'm <b>worried</b> that this may be as strong as you will feel"</p>
<b>4. Explore key topics</b> <ul style="list-style-type: none"> <li>• Goals</li> <li>• Fears &amp; worries</li> <li>• Sources of strength</li> <li>• Critical abilities</li> <li>• Trade-offs</li> <li>• Family</li> </ul>	<b>Explore</b> <p>"What are your most important <b>goals</b> if your health situation worsens?"            "What are your biggest <b>fears and worries</b> about the future with your health?"            "What gives you <b>strength</b> as you think about the future with your illness?"            "What <b>abilities</b> are so critical to your life that you can't imagine living without them?"            "If you become sicker, <b>how much are you willing to go through</b> for the possibility of gaining more time?" "How much does your <b>family</b> know about your priorities and wishes?"</p>
<b>5. Close the conversation</b> <ul style="list-style-type: none"> <li>• Summarize what you've heard</li> <li>• Make a recommendation; check in with patient</li> <li>• Affirm your commitment to the patient</li> </ul>	<b>Close</b> <p>"I've heard you say that _____ is really important to you. Keeping that in mind, and what we know about your illness, I <b>recommend</b> that we _____. This will help us make sure that your treatment plan reflect what's important to you"            "How does this plan seem to you?" "I will do everything I can to help you through this."</p>
<b>6. Document your conversation &amp; 7. Communicate with key clinicians</b>	

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# Developing Trust and Relationships



Commentary

Denied the right to comfort: Racial inequities in palliative care provision

Kavita Algu

*Temmy Latner Centre for Palliative Care, Mount Sinai Hospital, 60 Murray St, Toronto, M5T 3L9, Canada*

## Death Is a Social Justice Issue

### Perspectives on Equity-Informed Palliative Care

Reimer-Kirkham, Sheryl PhD, RN; Stajduhar, Kelli PhD, RN; Pauly, Bernie PhD, RN; Giesbrecht, Melissa PhD; Mollison, Ashley MA; McNeil, Ryan PhD; Wallace, Bruce PhD

[Author Information](#)

*Advances in Nursing Science* 39(4):p 293-307, October/December 2016. | DOI: 10.1097/ANS.0000000000000146

Commentary

## From Cultural Safety to Anti-Racism: Reflections on Addressing Inequities in Palliative Care

Seana Bulle<sup>1</sup>, Amit Arya<sup>2,3,4,5</sup> and Naheed Dosani<sup>6,7,8,9,\*</sup>

How do you care for people with SCD  
in your practice?



**FAMILY MEDICINE**

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Questions or Comments?

Email me!  
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Department of Family Medicine  
Michael G. DeGroote School of Medicine  
Faculty of Health Sciences

[fammedmcmaster.ca](http://fammedmcmaster.ca)  
[@McMasterFamMed](https://twitter.com/McMasterFamMed)

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