

# National Human Genome Research Institute



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# CONQUERING PAIN AND FATIGUE IN ECD

Experience with the ECD cohort and application  
from the medical literature

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OCD/NHGRI /NIH

Erdheim-Chester Disease Patient and Family Gathering

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# The Challenge of Pain Rests in the Brain:



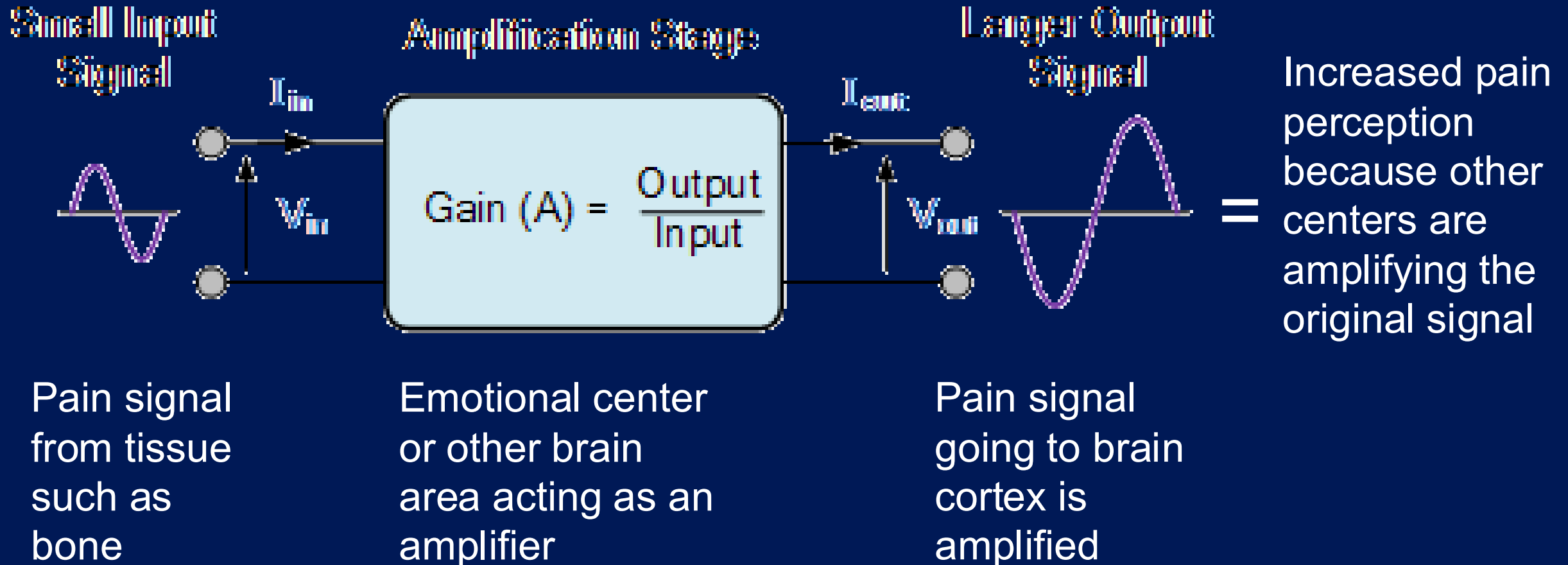
- Multiple studies confirm the commonness of pain and fatigue in ECD, other histiocytosis, and other serious illnesses (cancer, neurological diseases, autoimmune diseases).
- Pain signals can arise from multiple body structures, sometimes simultaneously
- Common areas include bone, especially long bones, but also muscle, tendon, and other joint tissues.
- Pain is much more than a noxious signal from the tissues. It is an experience interpreted by the brain
- ***Your pain is real; you are not making it up***

# Conquering Pain and Fatigue in ECD

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- Pain is interpreted by multiple brain areas, and some are not directly involved in the signal from the affected tissue
- Other brain areas such as emotional centers or centers that create a fear response can be triggered with the experience of pain
- When other brain areas are activated, pain may be perceived as being worse, and suffering may occur
- These other areas in the brain that get activated by the repeated experience of pain act like amplifiers in an electrical circuit

# Non-tissue Amplifiers of Pain

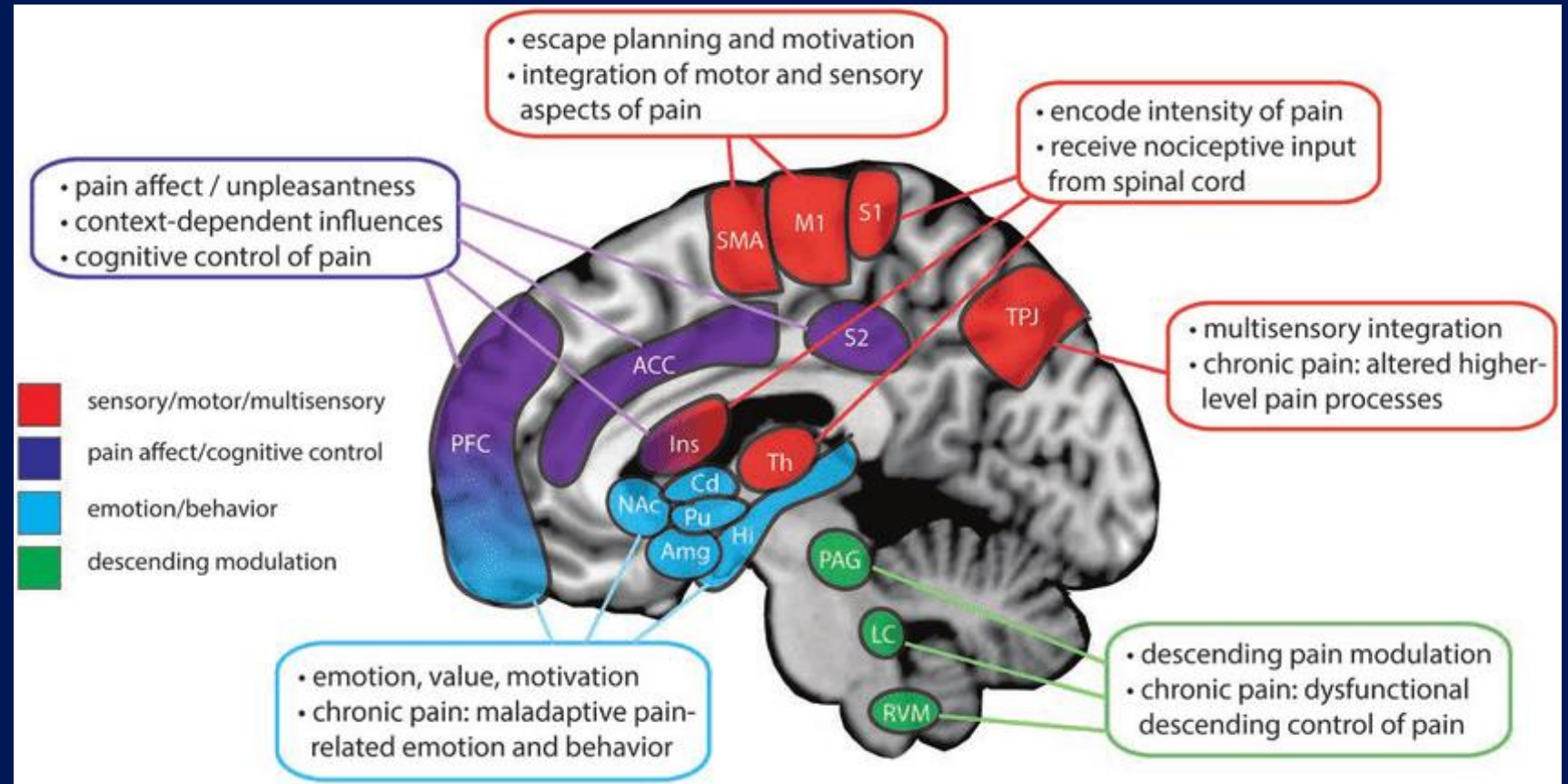


# Other Brain Areas Can Change Pain Perception

Some brain areas act like amplifiers increasing the perception of pain

Other areas can function as suppressors decreasing the pain signal or decreasing the meaning of the signal in the highest brain centers

YOU can modulate the signal and its meaning



Katherine T. Martucci, Sean C. Mackey; Neuroimaging of Pain: Human Evidence and Clinical Relevance of Central Nervous System Processes and Modulation. *Anesthesiology* 2018; 128:1241-1254  
doi: <https://doi.org/10.1097/ALN.0000000000002137>

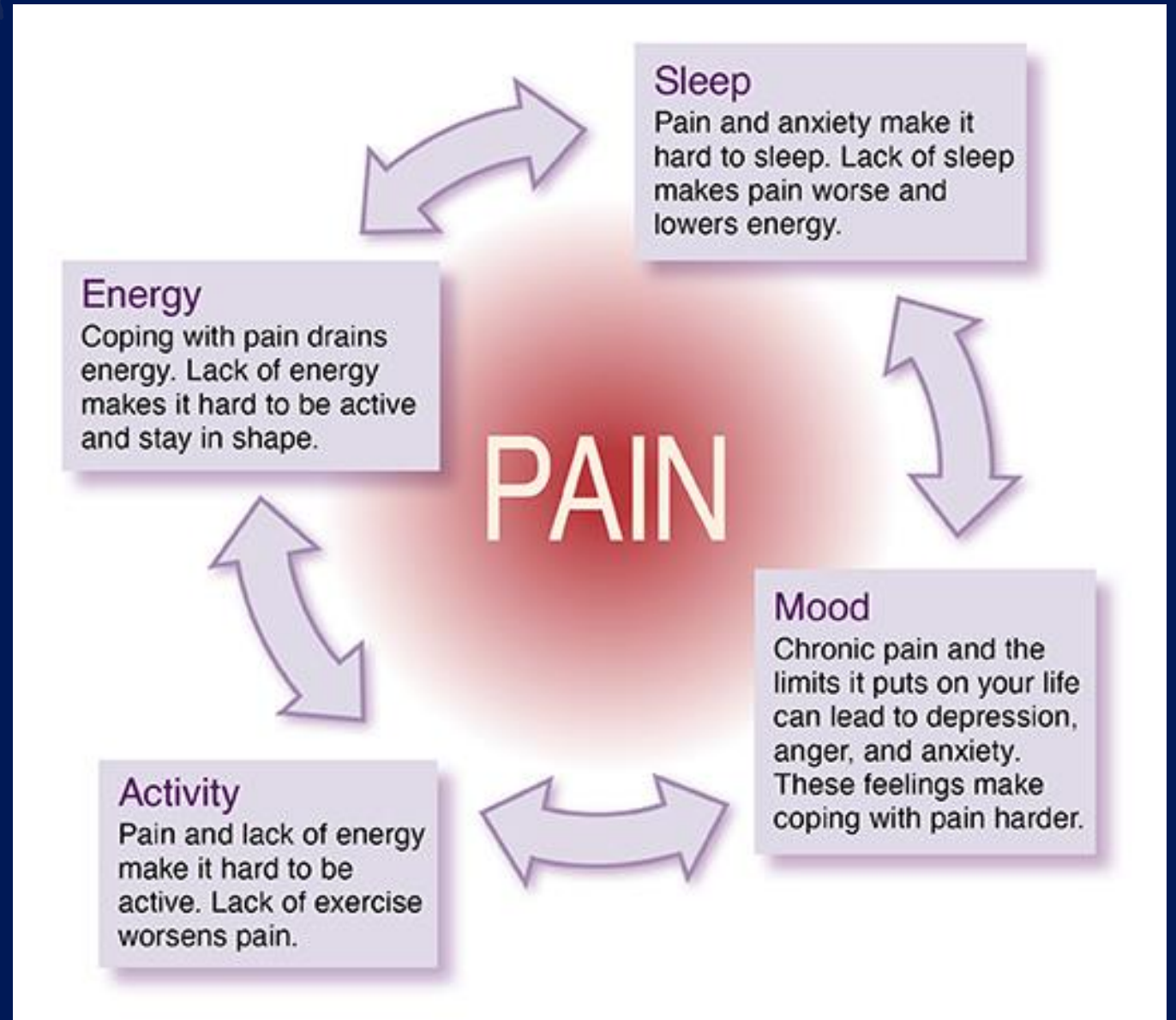
# Conquering Pain in ECD: Chronic Pain

Acute pain can become chronic

Chronic pain can create a vicious cycle

Other factors, like sleep deprivation worsen pain and cause fatigue

The chronic pain cycle can be broken !!



# Conquering Pain in ECD

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*The first step is to tell your provider that you have uncontrolled pain*

*Don't say everything is OK when it's not*



**Don't try to tough-out the pain because that will only reinforce the pain cycle**

**Be willing to take medication to reduce your pain while you work on other strategies to control pain**



<https://tenor.com/view/thumbs-up-gif-18124999>

# Conquering Pain in ECD

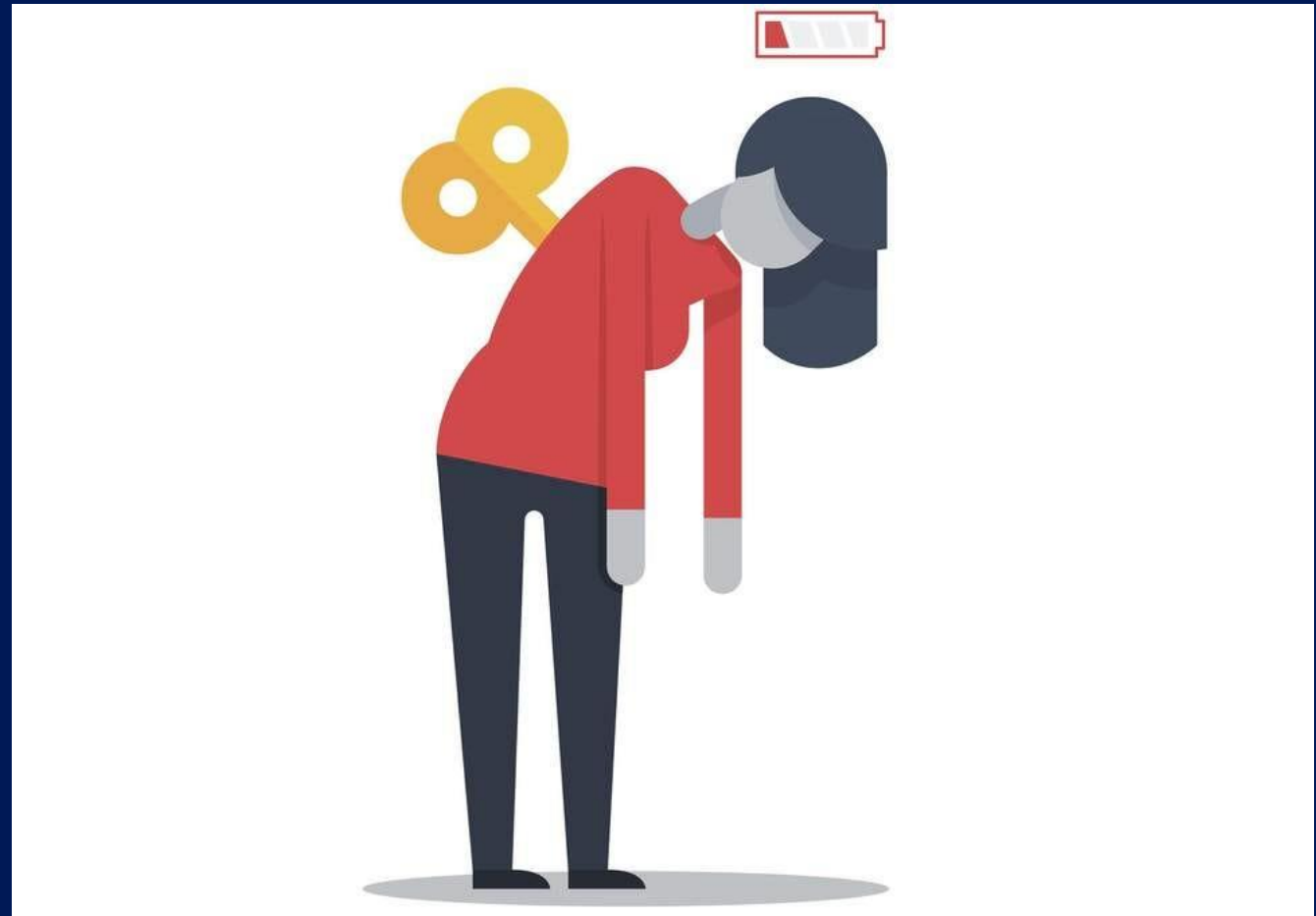
- IT IS OK TO TAKE NARCOTICS UNDER SUPERVISION
- When used appropriately, narcotics are effective
- Narcotics can help control acute pain while you work on other pain reduction strategies
- Help reduce nocturnal pain by reducing anxiety and inducing sleepiness



# REDUCING FATIGUE IN ECD

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- Fatigue is another vexing problem in ECD that has multiple causes
- Important causes include: poorly managed pain, sleep deprivation, medication side effects, untreated depression and anxiety, poor diet, inactivity, and disease activity affecting energy production or organ function
- Your fatigue is real, it is not “in your head” or made up. Discuss this with your provider



<https://medium.com/@kularti/dealing-with-fatigue-and-persistent-chronic-pain-after-stroke-7ccca03ab09f>

# STRATEGIES TO REDUCE FATIGUE IN ECD

- Medication-induced fatigue may lessen as your body acclimates to the drug, if not, then other treatments can help
- ***Fixing sleep deprivation and treating poorly-controlled pain are two of the most effective treatments for fatigue in ECD.*** This may require medications
- Don't be afraid to take medications to help control pain and to restore your sleep cycle
- Paradoxically, REGULAR exercise reduces fatigue over the long-term, but at first may make you feel more tired because you are not used to be so active.

# STRATEGIES TO REDUCE FATIGUE IN ECD

- Stimulants useful for severe fatigue to help the person get moving again while other strategies are being enlisted
- Treatment for cancer-related fatigue has been effective and safe
- Have your provider determine if you are a candidate
- Examples of stimulants include: Methylphenidate, dextroamphetamine, and modafinil. Coffee works too
- Dose adjustment may be necessary

# STRATEGIES TO REDUCE FATIGUE IN ECD

## RESTORING THE SLEEP CYCLE:

- Important long-term treatment that takes months --or even a year
- Requires coordinated effort between the patient, provider and caretakers. Everyone must try and all must be patient
- Can be frustrating to find a pattern and many people give up too soon
- Discipline to the plan is key and difficult to maintain
- Medications short-term are OK to help establish the cycle

# STRATEGIES TO REDUCE FATIGUE IN ECD

## EXERCISE AND PLANNED REST BREAKS:

- Get clearance from your doctor and you can work with a PT or trainer for initial guidance
- Walking is great, but a stationary bike or arm bike is also good for mobility-impaired
- Exercise also reduces pain when done regularly
- Learn to manage your daily energy levels

# STRATEGIES TO REDUCE PAIN AND FATIGUE IN ECD

- GRADED EXERCISE IS THE BEST INTERVENTION TO REDUCE PAIN AND FATIGUE. BETTER THAN MEDICATIONS!
- TIGHT JOINT STRUCTURES HURT WHEN YOU BEGIN EXERCISING AFTER PROLONGED INACTIVITY. LIKE WHEN YOU STRETCH TIGHT HAMSTRINGS
- TAKE MEDICATION BEFORE EXERCISE TO MAKE THE SESSION TOLERABLE
- MEDITATION IS ANOTHER EXCELLENT INTERVENTION TO REDUCE PAIN AND FATIGUE
- MEDITATION TAKES PRACTICE AND LEARNING TO BREATHE INCREASES EFFECTIVENESS

# PEACE FOSTERS HOPE: BE STILL AND BREATHE



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Paul C. Hendrie, MD PhD

## Patients, Caregivers and Families living with ECD

University of Washington School of Medicine  
Fred Hutchinson Cancer Center.

[Dr. Paul Hendrie, MD, PhD](#)

# SELECT REFERENCES:

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# Personal Challenges in ECD

## Henderson's 14 Needs as Applied to Maslow's Hierarchy of Needs

<https://nurseslabs.com/nursing-theories/>

