Managing Pain
in Marfan Syndrome

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Objectives

- Define pain
- Discuss the role of comorbid conditions that worsen pain
- Describe effective interventions for the treatment of chronic pain
Pain is made up of 2 parts:

- A sensory experience associated with physical manipulation
- An emotional response of distress and anxiety related to the sensory information

Frida Kahlo  *Without Hope*  1945
How academics think about pain?
How individuals think about pain?
Pain is ubiquitous in MFS

Adapted from Nelson Clin J Pain 2015

Pain is common
Speed Am J Med Genet 2017

Pain is undiagnosed

- Head 33%
- Shoulders 45%
- Back 78%
- Hips 46%
- Knees 43%
- Legs 37%
- Chest 28%
- Abdomen 18%
- Hands 25%
- Feet 49%
- Jaw 15%

41%
Chronic pain has consequences

- Pain
- Demoralization
- Apathy
- Fatigue
- Anhedonia
- Hopelessness
- Disability
- Social isolation
- Opioid/Benzo dependence
- Insomnia
- Risk Behavior
- Poor adherence
Pain in Marfan syndrome

89% pain
28% pain at onset
44% pain spreads

N = 245

Pain Disability

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Pain Catastrophizing

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Physical Health (SF-12)

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Mental Health (SF-12)

* p < 0.05, ** p < 0.01, *** p < 0.001
Why is it important to address pain?

- Relationships (spouse / partner, children, friends)
- Work
- Finances
- Energy
- Social activities
- Physical health
- Mental health
Where to begin?

I'm pretty tough. But some days my body just says: NOPE.
Diagnosis: Comorbidities

- Bereavement/grief
- Demoralization
- Stressful life events
- Major Depression
- Bipolar Disorder
- Anxiety Disorder
- Substance Use Disorder
- Insomnia Disorder
- Temperamental strengths/weaknesses
Why diagnose Depression?

- Major Depression
  - uncouples the reward system
  - increases reliance on avoidance coping
  - increases vulnerability to medication overuse
  - increases pain sensitivity

Walker AK Pharmacol Rev 2014
Symptoms of Major Depression

- Depressed or irritable mood [or feeling nothing]
- Decreased interest or pleasure in activities (anhedonia)
- Change in appetite or weight
- Sleeping more or less than usual
- Psychomotor agitation or retardation
- Fatigue or loss of energy
- Decreased concentration
- Feelings of guilt or worthlessness
- Recurrent thoughts of death or suicide
- Hallucinations and delusions possible
Why consider substance abuse? (alcohol, opioids)

- Increase liability to medication overuse
- Decrease social networks and support
- Decrease motivation to get well
- Diminish the baseline experience of being well

Sullivan Pain 2013
Pain and sleep frequently comorbid

BEHAVIORAL MODEL OF INSOMNIA

- Perpetuating Factors
- Precipitating Factors
- Predisposing Factors

THRESHOLD

Pre-Morbid  Acute  Early  Chronic

Spielman et al. 1987

Pain  50-80%  Insomnia

Finan et al 2015
## Treatment strategies

- Medications
- Psychotherapy
- Interventions/Stimulators
- Education
- Physical therapy
- Group therapy
- Exercise
- Family therapy

<table>
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<tr>
<th>N = 218</th>
<th>Current (%)</th>
<th>Past (%)</th>
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<tbody>
<tr>
<td>Over the counter meds</td>
<td>53.2</td>
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<td>Opioids</td>
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<td>Non-opioid medications</td>
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<td>Physical therapy</td>
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<td>Complementary/alternative</td>
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<td>Psychology pain interventions</td>
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<tr>
<td>Procedures</td>
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<td>Herbal supplements</td>
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<td>Occupational therapy</td>
<td>2.8</td>
<td>14.2</td>
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Speed et al 2017 (accepted)
Multimodal therapy

Pain education

Acetaminophen
Ibuprofen
Nerve medicines
SNRIs
TCAs
Anti-seizure medications
Opioids

Non-medication therapies

Antidepressants
Opioids
Acetaminophen
Anticonvulsants
NMDA antagonists

Opioids
Alpha\textsubscript{2} agonists
Local Anesthetics

Alpha\textsubscript{2} agonists
Anti-inflammatory drugs

Anti-inflammatory drugs
Topical Anesthetics
Toolbox of therapies

- Behavioral Approaches
- Relaxation
- Imagery
- Self hypnotic analgesia
- Distraction techniques
- Graded physical recovery exercises
- Assertiveness training
Cognitive Behavioral Therapy

Desire to feel a certain way to do a certain thing
More likely to do a certain thing
After a while it will make you feel a certain way

Adapted from Turk 2000
Relaxation Training

- Relaxation Therapies
  - Abdominal (Diaphragmatic) Breathing
  - Progressive Muscle Relaxation
  - Guided Imagery

- Yoga & Mindfulness Meditation
  - reduce inflammation

*practice during a time of relative low pain
Self efficacy

Belief in capability to perform a goal affects:

• the energy invested in the endeavor
• the persistence in the face of obstacles
• and the anxiety and demoralization in coping with stressors related to the endeavor
Final thoughts

• Individual pain experience is unique

• All individuals benefit from structure, hope, and advocacy

• Accept serendipity wherever you can find it

• Goal is functioning despite pain