Pain in Facioscapulohumeral Muscular Dystrophy (FSHD)

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Outline of webinar

- Katherine Mathews
  - Overview of literature on pain in FSHD
  - Brief discussion of medication as part of management
- Krista Kohl
  - Current concepts about pain
  - Introduction to some techniques that can be used to help manage pain
- Shelley Mockler
  - Physical therapy approaches to pain management

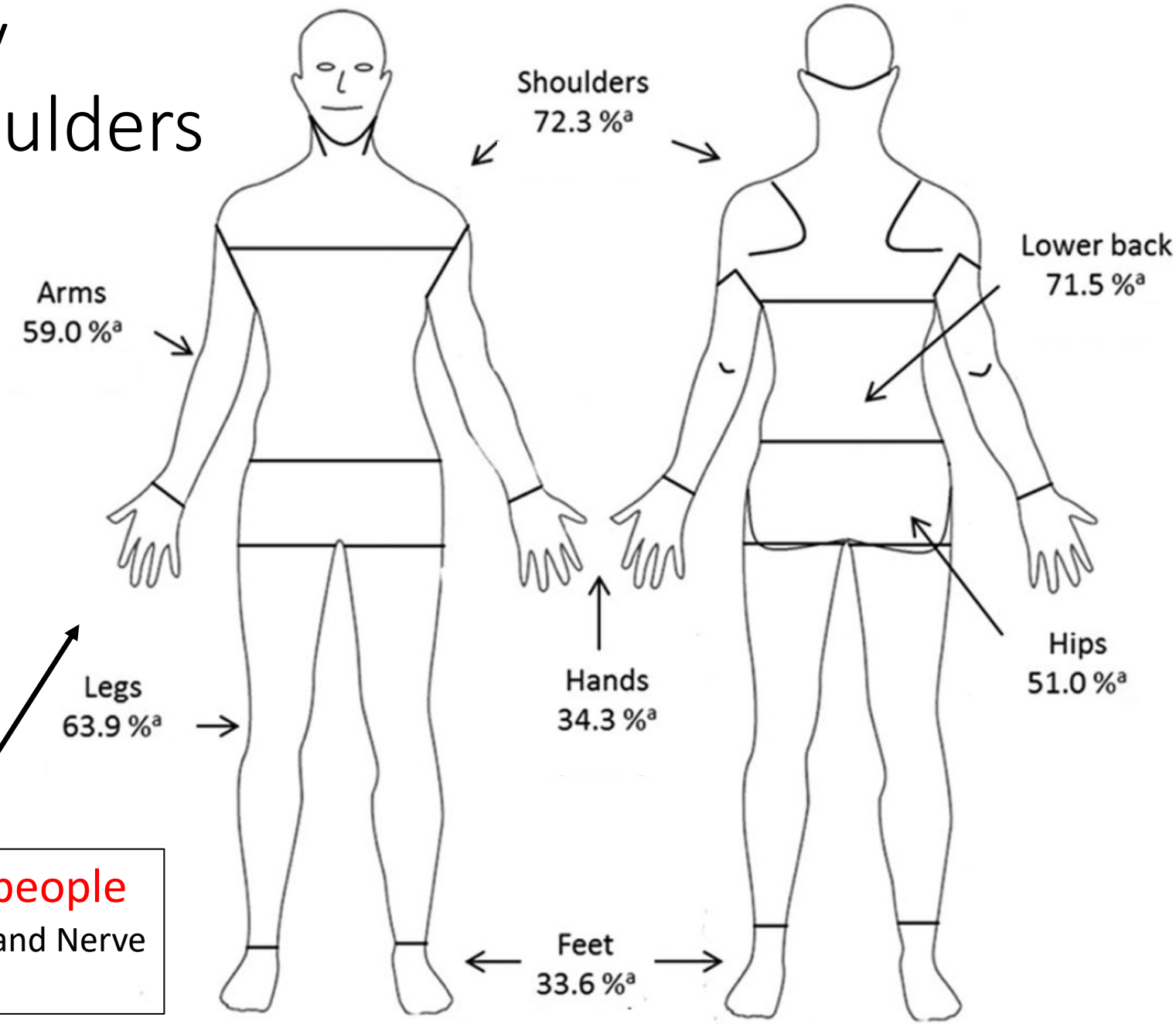
Pain is reported by 80-90% of adults with FSHD

- 82% of 127 people with FSHD (survey)
  - 19% with severe pain (>7 on 1-10 scale)
    - 2008, Jensen, et al
- 89% of 398 people with FSHD from the UK (Registry data)
  - 50% reported chronic pain (persistent pain experienced for at least 12 weeks within a year in the last 5 years)
    - 2018, Moris et al
- 87.7% 328 people with FSHD (survey)
    - 2019, Hamel, et al

- If you have pain, you are not alone.
- But there are approaches to management

Pain most commonly affects low back, shoulders

- Survey (127 people)
  - Low back (74%)
  - Legs (72%)
  - Shoulders (69%)
    - McDonald, et al 2018



Registry, 398 people

- Moris, Muscle and Nerve 2018

Pain has complex relationship with stage of disease

- No consistent relationship with D4Z4 fragment size (genetics in FSHD type 1)
- No consistent relationship with age (among adults) or duration of disease
  - In some studies, young and middle-aged adult patients were most likely to report pain
- No simple relationship with mobility status
  - Those with no limitations less likely to report pain in most series
  - No consistent difference seen between those walking and those using wheelchair

Pain affects quality of life

- Pain impacts
  - mobility
  - ability to do work
  - mood
  - sleep
  - enjoyment of life

    Jensen, et al 2008

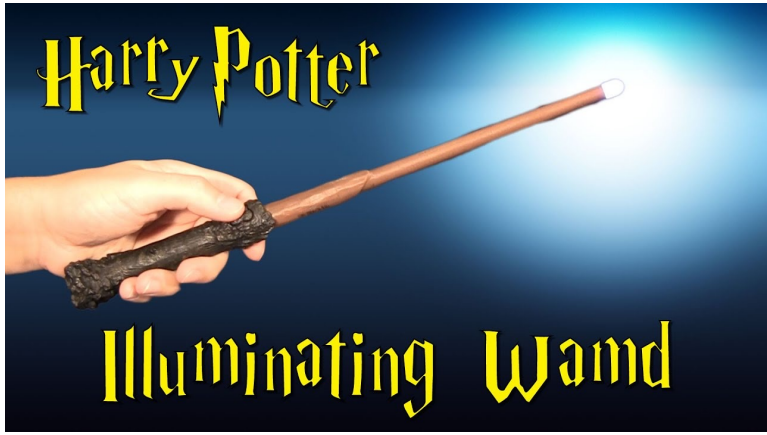
Pain can be a problem for patients with FSHD.
How do we approach it?

Initial evaluation of pain in FSHD

- Explore the details of pain (when, where, what kind of pain, what makes it better/worse)
  - The more information you can give, the better advice your healthcare team can give
- Based on history and exam:
  - Is specific testing needed (X-rays, etc)?
  - Is another type of specialist needed (ex. abdominal pain might need to see GI)?
- Determine significance of pain to you
  - Rare annoyance vs problem that needs management
  - How is it affecting life, mood, etc

Overall approach to pain management in FSHD

- Is there a clear modifiable cause (ex. it only hurts when I sit in this chair)?
- Treatments
  - Pharmacologic (medication)
  - Non-pharmacologic (physical and cognitive treatments)
    - Recall, pain occurs in the brain. If the brain is not involved (anesthesia) you don't feel pain
- Treatment is a process, there is no magic wand
  - Pain is complex and different for each person



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Medication classes that can be considered

- Over the counter pain medications (ex. ibuprofen, acetaminophen)
- Prescription non-steroidal medications (ex. celebrex, diclofenac)
- Anticonvulsants (ex. gabapentin, carbamazepine)
- Antidepressants (ex. amitriptyline)
- Muscle relaxants (ex. cyclobenzaprine)
- Creams or patches (ex. diclofenac, capsaicin, lidocaine)
- Opioids (ex. hydrocodone , meperidine, codeine)
- Injections (ex. joint injection)

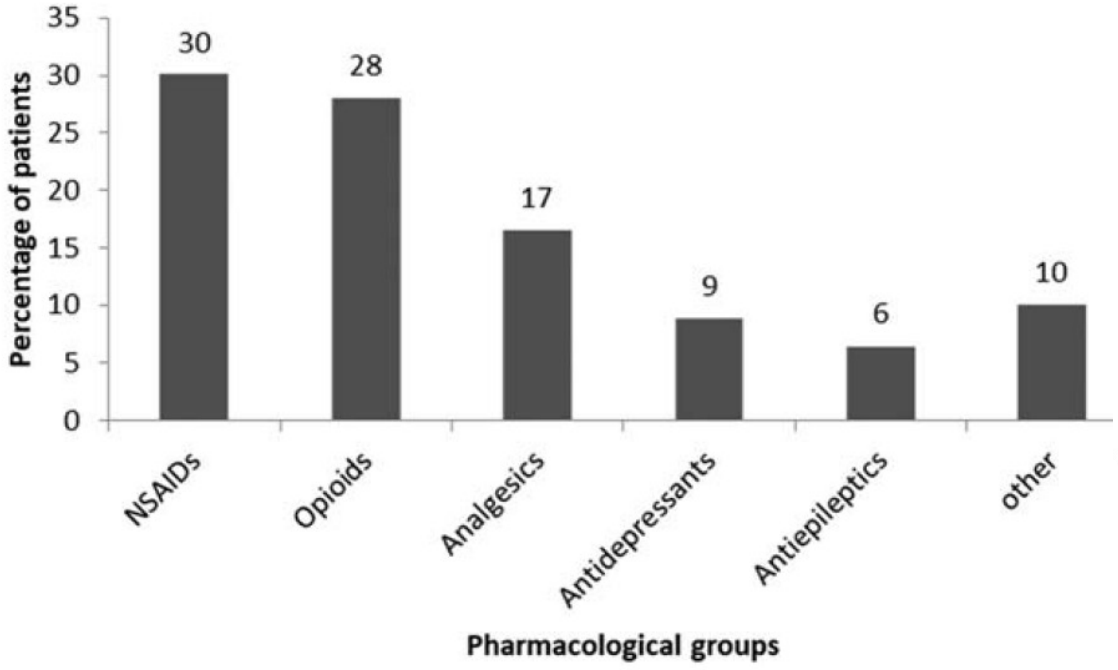
Medications in treating pain in FSHD

- Virtually all medicines have potential side effects
  - Use the lowest dose possible that achieves the desired goal for patient function and happiness
- Many of the medications are best managed by a pain specialist or pain clinic
  - Your neurologist or primary care doctor might start treatment, but then refer to a pain clinic if first line treatment(s) not effective
- Treatment of chronic pain is most successful when more than one approach is employed (medicine and non-pharmacologic approaches)

What do we know about how pain in FSHD is being managed?

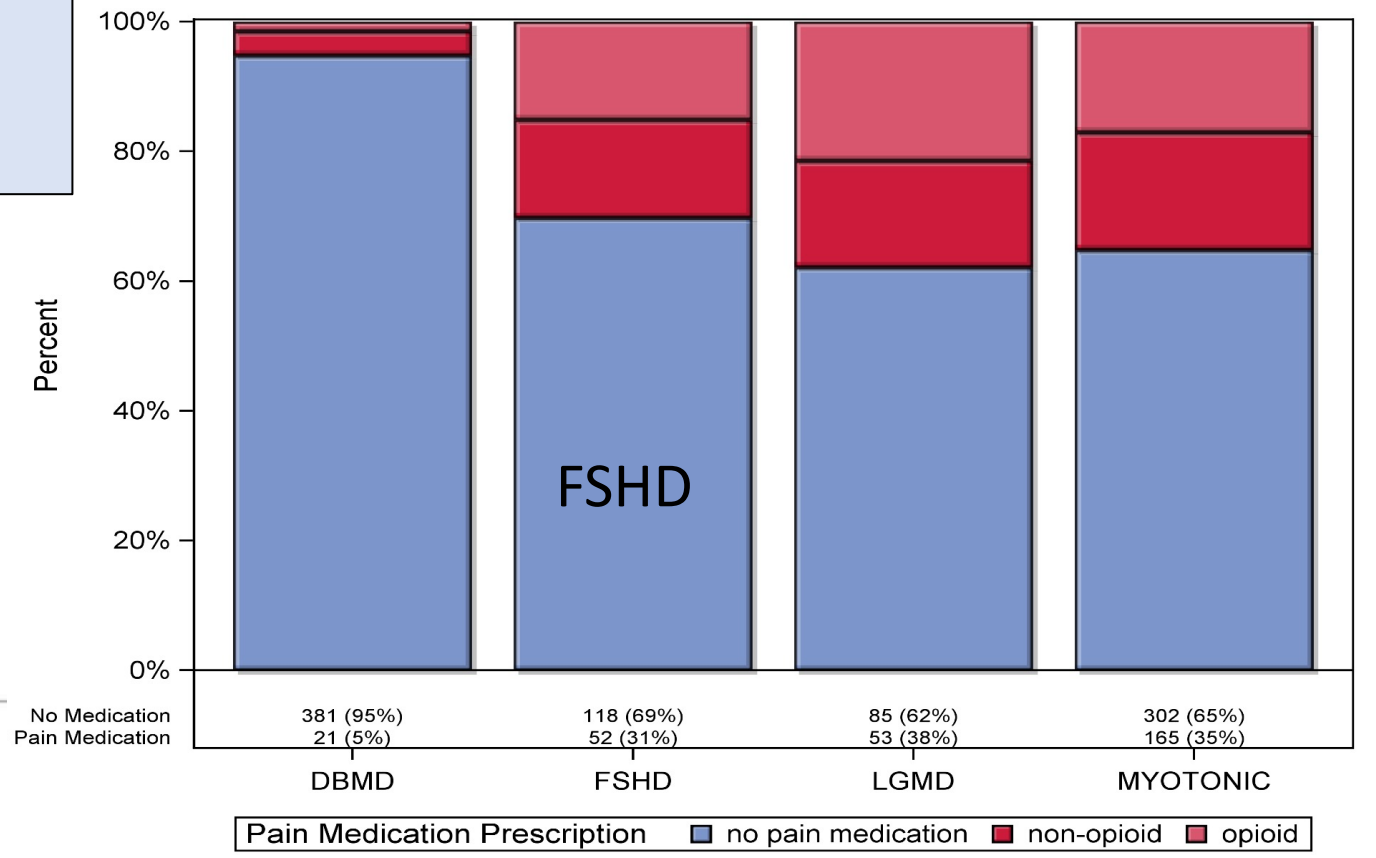
(Not a lot of information in the literature)

Pharmacologic treatment



- Non-steroidal anti-inflammatories and opioids were each used by ~30% of patients.
  (Patients lived in the UK.)

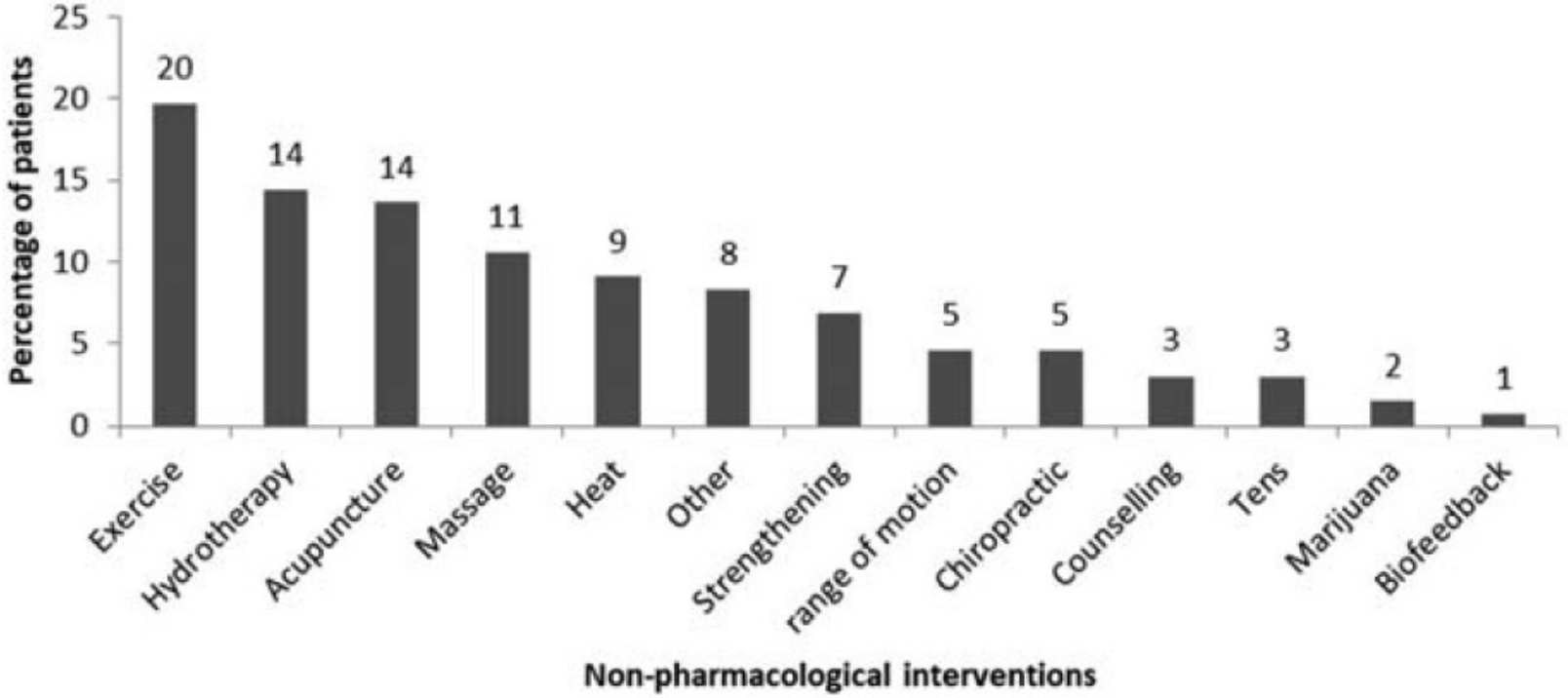
Moris, et al, 2018



Pain medications prescribed, by type of muscular dystrophy

- FSHD: 31% prescribed pain medication
- About half of these are opioids
  - Suhl, et al, MDSTARnet data presented as poster at MDA National Conference 2022

Non pharmacologic treatments



Moris, et al, 2018

<u>Top 5:</u>

- Exercise
- Aquatherapy
- Acupuncture
- Massage
- Heat

Continued use of pain treatments in FSHD + myotonic dystrophy

Treatment	Tried (%)	Continue to use (%)
Ibuprofen, aspirin	78	46
Acetaminophen	70	34
Heat	71	26
Strengthening exercises	64	29
Range of motion	44	29

Adapted from Jensen, Arch Phys Med Rehab, 2008

- These are most likely continued from a long list of possible treatments.
- Continued treatments
  - Readily available
  - Few or no side effects
  - Provide at least moderate pain relief

Summary

- Pain is a common problem that can affect quality of life for people living with FSHD
  - Severity and frequency similar to osteoarthritis and rheumatoid arthritis (Moris, et al. 2018)
- It is hard to predict who with FSHD will be affected by pain
- There is no single “best” management but lots of options
  - Balance of side effects and benefit
  - A certain amount exploring treatment options might be needed
- Treatment is not just medication!