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Abilitylab[®]

**A PHYSIATRIC APPROACH TO PATIENTS WITH
FACIOSCAPULOHUMERAL MUSCULAR DYSTROPHY**

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WHAT IS A PHYSIATRIST AND WHAT DO WE DO?

- Physical Medicine and Rehabilitation physicians (PMR)
- Physiatrist complete 4 years of medical school and a 4 year residency program
- Specialty of medicine focusing on the physical and functional manifestations of a person affected by a physical or cognitive impairment

GOALS OF PHYSIATRIC MEDICINE

- Maximize an individuals function
- Maintain a persons independence
- Prevent secondary medical comorbidities and complications
- Prevent or limit physical deformities

WHY SHOULD A PHYSIATRIST BE INVOLVED IN THE CARE OF SOMEONE WITH FSHD AND WHEN SHOULD A PATIENT BE REFERRED?

- Usually referred through a primary care physician or a neurologist
- Referral should be made early in the diagnosis (pediatric physiatrist when onset before 18)
- Important to focus on maintaining function and endurance and preventing secondary medical and musculoskeletal complications

THE PHYSIATRIC EVALUATION: WHAT TO EXPECT?

- Complete medical and functional history
- Functional history:
 - Level of assistance needed for activities of daily living (ADL's) such as dressing, bathing, toileting, and feeding. Complex ADL's (CADL) such as cleaning, cooking, community activities, driving or working
 - Level of assistance needed for mobility-Use of assistive devices
 - Frequency of falls
 - Use of orthotics



THE PHYSIATRIC EVALUATION CONT.

- Review of systems:
 - Level of endurance
 - Pain
 - Nutrition/swallowing difficult
 - Speech/articulation
 - Breathing/shortness of breath with activities
 - Mood /adjustment to the disability
- Physical exam:
 - Special focus on musculoskeletal and neurologic systems:
 - UE and LE and trunk strength
 - ROM
 - Cranial nerves/Facial weakness
 - Speech
 - Gait[Pattern and stability]



THE PHYSIATRIC EXAM

- Upon completion of the exam recommendations may include:
 - Exercise prescription/recommendations
 - Referral to PT, OT, and or speech therapy
 - Referral to an Orthotist for bracing
 - Recommendation to use an assistive device for mobility (cane, walker)
 - Referral for a mobility device (wheelchair)
 - Recommendations for treatment of pain

All recommendations are **individualized** to the patients functional and medical status and **personal goals**

AN EXERCISE PRESCRIPTION FOR A PATIENT WITH FSHD

- Common questions asked:
 - Strength training VS. Aerobic training
 - Supervised (PT, OT personal trainer) or unsupervised
 - Will exercise make me stronger?
 - Will exercise make me worse or weaker?

LITERATURE REVIEW ON EXERCISE AND FSHD

- Paucity of literature looking at FSHD and exercise
- Cochrane database review (2013), reviewed only 2 studies pertaining to FSHD and strength training
 - Authors conclusion: “Moderate intensity strength training in patients with FSHD 1 (2 studies) appears to do no harm but there is insufficient evidence to conclude they offer benefit”

AEROBIC EXERCISE AND FSHD

- Anderson et al. (2015) investigated the effects of regular aerobic training (36 sessions, 30 min cycle ergometer) followed by ingesting a protein carbohydrate drink or placebo beverage
 - Results showed that all participants improved fitness, walking speed and self assessed health and that the post exercise protein drink did not add further benefit

AEROBIC EXERCISE AND FSHD CONT.

- A 12 week study by Olsen et al. (2015) looked at low intensity aerobic exercise (cycle ergometer at HR corresponding to work intensity of 65% VO₂ max (a measure of cardiac fitness pertaining to the amount of oxygen your body is capable of utilizing in 1 minute) at 35 minute weekly sessions and increased to 5 times week in 4 weeks.
 - After 12 weeks, participants showed improved maximum oxygen uptake and work load (exercise performance) with no signs of muscle damage (measured blood plasma CK which is a marker for muscle breakdown)

AEROBIC EXERCISE AND FSHD CONT.

- Bankole et al. (2016) evaluated the safety and efficiency of a 6 month home based exercise program(unsupervised) in 16 patients with FSHD
 - Patients were randomized to a control group with no HEP and a training group (cycle 3x a week for 35 minutes) for 24 weeks followed by the control group doing the HEP for 24 weeks
 - Found improvement in endurance, 6 minute walking test, and subjective improvement in fatigue with no detrimental effect on muscle tissue

- A recent study by Anderson et al. (2017) looked at high intensity training (HIT) and patients with FSHD
 - Number of participants: 13
 - Patients with FSHD: 1
 - HIT group (6 participants) underwent 8 weeks of supervised HIT (10 minute cycle ergometer, 3 times a week)
 - Control group underwent 8 weeks of usual care (not well defined)
 - Followed by all participants performed 8 weeks of unsupervised HIT
 - Supervised and unsupervised HIT participants improved fitness (VO2 max)
 - No training effect on muscle strength, 6 minute walk, 5 time sit to stand tests
 - Plasma CK (evidence of muscle breakdown) and pain scales were unaffected

CONCLUSION

- More studies need to be done looking at role of exercise in patients with FSHD
- No evidence to suggest strength training is beneficial, however it does not appear to be harmful
- Moderate aerobic exercise (and possibly HIT) may be beneficial in improving fitness and over all well being
- Any exercise program should be individualized due to the heterogeneity of FSHD (patient have different muscle involvement and rates of disease progression)
- When initiating an exercise plan, it should be initially be under the supervision of a professional.

PHYSICAL THERAPY AND FSHD

- Physical therapist will work on maximizing strength, gait, balance and ROM to maximize mobility
- May recommend an assistive device to aid in mobility/prevent falls
- Develop an **individualized goal oriented** home exercise program (HEP)-extremely important to follow-through to maintain gains
- Aqua therapy
 - No controlled studies looking at water therapy and FSHD
 - Theoretical Benefits
 - Buoyance of water acts to assist mobility
 - Many muscles can be worked simultaneously

OCCUPATIONAL THERAPY AND FSHD

- The most common initial finding in patients with FSHD is weakness of the scapular stabilizers, making it difficult to performing reaching and overhead activities
- OT's work on improving upper extremity mobility to aid with performing activities of daily living as well as community and work-related activities
- Also work on ROM to prevent contractures
- May recommend equipment to assist with ADL's (reachers, sock aids)
- May recommend bracing to improve function, prevent contractures
- Home Evaluation (PT and/or OT)
 - Evaluate home environment to maximize safety and independence
 - Need for/placement of grab bars
 - Look at kitchen/bathroom set-up to make items more accessible
 - Look at home obstacles (rugs, furniture) that may effect mobility

SPEECH THERAPY AND FSHD

- Due to facial weakness, patients may have difficulty with articulation, and more rarely swallowing
- Speech therapist aid in maximizing communication and articulation
- Will evaluate swallowing via a bedside swallow(in the clinic) or with a Videoflouroscopic swallow study(more sensitive, performed in radiology dept.)
 - If indicated will provide strategies to maximize swallow ability and safety
 - May recommend diet changes to prevent aspiration.

ORTHOTICS AND FSHD

- May be recommended by your M.D. or therapist
- Often referred to a professional Orthotist to customize braces
- Common orthotics in FSHD:
 - AFO: Ankle foot orthosis
 - Crosses the ankles and foot
 - Used for foot drop due to peroneal muscle weakness
 - Allows/aids in walking
 - Prevents patient from catching the toes with walking
 - ❖ Prevents falls
 - ❖ Normalizes gait pattern
 - ❖ Stabilizes knee to prevent hyperextension
 - ❖ Prevent contractures



ORTHOTICS AND FSHD

- KAFO- Knee ankle foot orthosis
 - Crosses the knee joint
 - Used when quadriceps (thigh muscle) is compromised/weak and more support is needed to prevent knee from collapsing
 - Stabilizes gait to prevent falls

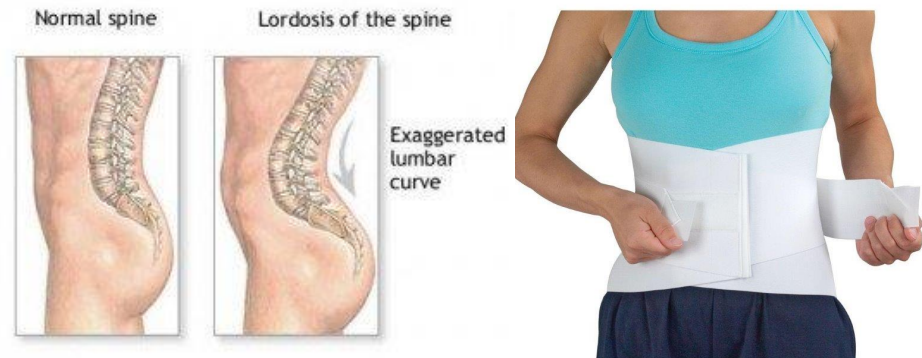
Due to weight of this orthotics often not tolerated especially if there is substantial hip girdle weakness



ORTHOTICS AND FSHD CONT.

- Abdominal binders
 - Due to weak core muscles often have hyperlordosis of the spine which can lead to pain
 - Used to aid weak core to stabilize back

- Scapular Bracing
 - Sometimes used to stabilize scapular muscles
 - Often not tolerated or effective



ASSISTIVE DEVICES AND FSHD

- Common assistive devices
 - Cane (straight, quad), walker
 - Equipment for ADL's (reachers, sock aids, raised toilet seat, etc.)



MOBILITY DEVICES AND FSHD

- 20% of patients will require a wheelchair for mobility after age 50
- Transport chair
 - Early in the disease, often used for longer community mobility, when patients are still walking but fatigue is an issue
- Power wheelchair
 - Due to upper extremity weakness, often difficult to use a manual wheelchair, power chairs are most commonly recommended
 - Referral should be made to specialized seating clinic
 - All wheelchairs should be customized to the individual
 - Support back, maintain postural alignment, minimize postural deformity

PAIN AND FSHD

- Limited literature on this topic
- Up to 80% of individuals suffer from acute or chronic pain
- Most common areas are the neck, shoulders, low back and legs
- Reasons for Pain
 - Muscle imbalance between opposing muscle groups, leads to a sustain stretch of some muscles and prolong contractures of others resulting in inflammation
 - Weak muscles around a joint can not protect the joint, resulting in improper joint alignment and pain
 - Weakness of the shoulder muscles can result in stretch injury to the brachial plexus causing pain and weakness down the arm
 - Weak abdominal muscles can not protect the low back resulting in low back pain

TREATMENT OF PAIN

- Bracing for stability
- Modalities (Heat/Ice)
- Pain medication
 - NSAID's, Acetaminophen, topical analgesic, nerve membrane stabilizers (lyrica, gabapentin)
- Rest if pain persists
 - Pain is the body informing us to stop doing an aggravating activity

CONCLUSION

- Integrated team approach to care
- Exercise can prevent fatigue and improve fitness
- Use of equipment/AD to maximize mobility and function
- The ultimate goal is to improve or maintain a persons **independence, safety** and

~~DISABILITY~~

A hand holding a red marker is shown crossing out the word "DISABILITY" written in black capital letters. The red marker is positioned over the word, with a red line striking through it from the top left to the bottom right.