FSHD, Activity, and Exercise

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Background

Symptoms

- Facial and scapular muscle weakness (early stage)
- As condition progresses, weakness develops in the pelvic girdle (hips), tibalis anterior (lower legs), and abdominals
- Asymmetrical weakness
- Fatigue



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What research is showing





Both aerobic exercise and cognitive-behavioral therapy reduce chronic fatigue in FSHD

- 57 patients randomly assigned to: AET, CBT, or UC
- 16 weeks of intervention with a 12 week follow-up.
- AET=30 min cycling 3x/week (2 at home, 1 with a PT).
 - Goal for 50-65% HRR. Min of 40 sessions.
- CBT=50 min sessions conducted by a cognitive-behavioral therapist, based on 6 possible modules dealing with known fatigue-perpetuating factors.
 - Minimum of 3 sessions.
- Results:
 - After intervention, AET and CBT had less fatigue relative to the UC group and benefits lasted through follow-up.
 - CBT had increase in registered and experienced physical activity, sleep quality, and social participation. AET only had increase in registered physical activity.
 - The registered physical activity in the AET and CBT were present at follow-up, as well as the increased social participation in the CBT group.

(Voet N et al., 2014)

Safety and efficacy of a 6-month home-based exercise program in patients with facioscapulohumeral muscular dystrophy

- 16 patients assigned to TG or CG.
 - Additional CGT group where the original CG participated in the intervention for weeks 24-48.
- TG=cycling 3x weekly for 35 minutes for 24 weeks
 - 2 sessions at 60% MAP followed by sets of near-maximal revolutions. 1 session of interval training.
- Results:
 - TG: Significant improvements in VO2 peak (same for CGT at T48). No change in CG.
 - Significant increase in muscle endurance and MVC for TG. (CGT: MVC unchanged but muscle endurance increased). No changes for the CG.
 - 6 MWT increased for TG, similar for CGT. No changes for CG.
 - Muscle fiber cross-sectional area and citrate synthase activity increased in TG. Dystrophic pathophysiologic patterns were not exacerbated.
 - No changes in QOL for any of the 3 groups. Fatigue decreased significantly in TG and CTG group.

High–intensity interval training in facioscapulohumeral muscular dystrophy type 1

- A randomized controlled clinical trial
- Sample size of 13 patients, 1 person dropped out due to abdominal surgery unrelated to intervention or FSHD1.
- Trial consisted of 2 parts:
 - Part 1-Supervised high intensity training (HIT) with FSHD patients and control healthy participants performing HIT for 8 weeks.
 - Part 2- Unsupervised HIT for 8 weeks with FSHD patients only.
 - Upright stationary bike used for HIT training.
- Results:
 - HIT found to be safe.
 - Low fluctuations in plasma-CK levels in patients.
 - Activity levels were unchanged and patients preferred HIT over other forms of training.
 - No complaints of muscle pain or injury, change in pain scores, drop outs or worsening of self assessed health.
 - Strength and functional assessments were unchanged by HIT.
 - 4 hours of HIT in 8 weeks was found to be sufficient enough to improve fitness (VO2max and workload) significantly in supervised and unsupervised patients.

Strength training and albuterol in facioscapulohumeral muscular dystrophy

- Randomized, double-blind, placebo controlled design study
- 4 groups (training plus albuterol, training and placebo, non-training and albuterol, and non-training and placebo)
- Sample size : 65 participants
- Results:
 - Training group demonstrated a significant increase in dynamic strength in elbow flexors compared to placebo group.
 - Training plus albuterol group demonstrated an increase that was significant in maximal voluntary isometric strength vs. placebo treated group.
 - 11 out 12 of the non-trained muscles in the albuterol group showed a positive effect on the MVIC compared to placebo group.
 - Ankle dorsiflexion strength decreased in all groups.
 - Muscle volume decreased in the placebo group and increased in albuterol treated group.
 - Study conclusion, strength training and albuterol appear to be safe interventions with limited positive effect on muscle strength and volume.

FSHD and What We Know



- Exercise is beneficial and safe
- Still determining the different types and expansion of muscles tested for safe recommendation of exercise performance. Further research needs to be conducted on the more impacted muscle groups.
- Future research needs: Effects of specific modes of exercise including aquatic exercise, yoga/pilates, and strength training.

Exercise Safety

- Not overdoing it
- Start slow with exercise and activities
- HR zone (target heart rate intensity)
 Use of heart rate reserve
- Proper tolerance of exercise including:
 - Pain free
 - If you feel fatigue or muscle burn during exercise, stop and take a rest





General Types of exercise/activities

Stretching

 Stretching tight muscles groups

• Bike

• Recumbent, pedal, upright bike

Yoga/Piliates

• Sitting, standing , quadruped







General Types of exercise/activities: continued

• Pool

- Buoyancy of water helps you to move easier and decreases the workload of the weaker muscles.
- Resistance training, core strength, aerobic exercise, stretching, balance

Walking

• In a mall, neighborhood, treadmill, Alter-G









Maintaining Function

- Compensations are okay
- Maximize safety with mobility
- Energy conservation
 - Performing activities at the time of the day when you have the most energy
 - Taking rest breaks when needed
 - Use of power mobility aids to conserve energy (scooters, power wheelchair)

Types of Assistive Equipment

- Foot orthotics (SMO, AFO, Ritche brace, Dictus bands, Turbo-Med)
- Seat assist
- Abdominal binder
- Wedge pillow
- Walking aides
 - Canes, loftstrand crutches, hiking sticks, walkers

Equipment









Equipment







Equipment









Multidisciplinary Approach

• Clinical Care

- Multidisciplinary Clinic
 - × Make contact with neurologist, PT, OT, seating clinic, nutrition, social worker, cardiology, pulmonologist, genetic counselor
- KKI Resource Center
- Adaptations to home (community rehab)
 - × Ramps, stair lift, modifications for bathroom (raised toilet seat, anti-slip mats, handrails), vehicle adaptations

• How to be your own BEST ADVOCATE !!!!!!



Resources

- Voet N, Bleijenberg G, Hendricks J, Groot I, Padberg G, Engelen B *et al.* Both aerobic exercise and cognitive-behavioral therapy reduce chronic fatigue in FSHD. American Academy of Neurology 2014; 83:1-9.
- Bankole LC, Millet G, Temesi J, Bachasson D, Ravelojaona M, Wuyam B *et al.* Safety and exercise of a 6-month home-based exercise program in patients with facioscapulohumeral muscular dystrophy. Medicine 2016;95:31.
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