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Health Tips

FSHD is highly variable, even among affected family members. Each person is possesses a unique combination of genetic and environmental factors that influence his or her body and health. This information is provided for general informational and educational purposes only. The FSHD Society does not provide medical advice or recommendations. Licensed physicians and other medical professionals who are familiar with an individual's specific health situation should be consulted for diagnosis and treatment of FSHD and any other medical conditions.

Keep Healthy Habits

Managing FSHD requires, first and foremost, managing one's overall health. Healthy habits and practices will help with managing FSHD, while poor ones such as smoking, poor diet, not getting enough sleep and high stress will make it harder.

Speak Up and Be Persistent

FSHD can affect things in ways that are not obvious or direct. For example, a doctor who lacks experience in muscular dystrophy may not be able to find the cause of a specific pain and basic tests such as an x-ray will not offer any indications as to the pain source. A neurologist, physiatrist, orthopedist or physical therapist with experience in muscular dystrophy may have an educated hunch about the cause of pain and may be able to provide the appropriate relief measures.

Medications Will Vary from Patient to Patient

The side effects of some medications and other medical treatments may be more problematic for some FSHD patients than for people without FSHD. If a patient has FSHD and has been prescribed specific medications by a physician, the patient should monitor any side effects closely and notify their doctor accordingly.

Flu Shots, Surgery and Inactivity

FSHD patients may be concerned about the effect of injections on their muscles, and others worry about exposure to chemicals in vaccines, but FSHD medical experts agree that any potential risk is greatly outweighed by the benefits. "Please encourage them to all get the flu shot," said Kathryn Wagner, MD PhD, of the Kennedy Krieger Institute. Older patients and anyone with respiratory issues should especially get immunized against flu and also pneumonia.



Beware of Blood Clots During Long Periods of Immobility (e.g. Flights)

FSHD patients who can't walk may be at risk of developing blood clots on long airplane flights or other situations where one is required to be immobile for an extended duration. On the advice of their doctors, some people take an anti-blood clotting prescription medication before flights to reduce the risk of clotting.

Dealing with Pain

A majority of FSHD patients report pain, which often can be chronic. Pain may be related, at least in part, to overuse of muscles and to fatigue. Not many studies have been done about pain in FSHD. Whether and how to treat pain is a decision for each patient in consultation with his or her doctor.

For a discussion of treating pain, please see our Physical Therapy and FSHD <u>brochure</u> written by Katy Eichinger, PhD, and Shree Pandya, PT, MS.

Cardiac Involvement and FSHD

FSHD per se is not thought to cause cardiac complications, and patients in general have normal longevity. This predisposes them to the usual age-related cardiac complications, and management of these problems is the same as in non-dystrophic patients.

Pulmonary and Respiratory Health and FSHD

Pulmonary complications may occur with FSHD, particularly impaired ability to expel carbon dioxide. This can lead to a build-up of CO2 in the blood, or hypercarbia. Evaluation of the symptoms and signs of respiratory insufficiency should be sought during routine clinic visits in patients with moderate to severe FSHD. Patients are encouraged to regularly monitor respiratory functions, as one might experience insufficiency over a long period of time without presenting signs.

Fatigue is often part of FSHD because the muscles have to work harder than normal muscles, but fatigue may also result from respiratory impairment, including sleep apnea and lower than normal forced vital capacity. If a patient feels fatigue or exhibits signs of sleep apnea (such as breathing in a labored manner or momentarily not breathing during sleep), this should be discussed with a doctor. This may require persistence because some doctors, even experienced neurologists, don't associate FSHD with respiratory problems. **Blood carbon dioxide should be monitored**.

If a patient is experiencing respiratory insufficiency, it can be initially managed with nighttime noninvasive pressure support, typically a BiPAP machine. In very severe cases, patients may require the use of a ventilator. For FSHD patients with respiratory insufficiency, care should be taken not to suppress respiratory drive with narcotics. In trauma, emergency room, surgery and anesthesiology settings, it is important to notify the doctors about FSHD and any respiratory problems the patient might have or be at risk for.



Physical Therapy and FSHD

A physical therapist (PT) who is experienced with FSHD patients can be invaluable. For example, physical therapists can:

- Administer some exercises directly (e.g., stretching)
- Teach patients how to perform other exercises

FSHD patients ask their doctors to prescribe physical therapy when:

- It feels like their FSHD is progressing more rapidly than usual
- They want periodically to monitor how they perform their exercises
- Rehabilitating specific conditions or injuries such as strained or pulled muscles or injuries from falls.
- They want to maintain their current level of function. This type of maintenance PT is now covered by Medicare, but some practitioners may not be aware of this and "fire" a patient for not showing improvement.

Exercise and FSHD

As with all exercise, it's important for FSHD patients to consult their doctors first. As a rule of thumb, moderate aerobic exercise is considered to be beneficial. Intense exercise, such as heavy weight training, is thought to put FSHD muscles at risk. Patients have reported beneficial results with muscle activation technique, Pilates, and gentle yoga. The AlterG "antigravity" treadmill provides support to enable individuals with leg weakness to get a good walking workout.

Many FSHD patients swim regularly as it has a low impact on the joints and bones, with a low risk of injury. The buoyancy of the water provides resistance, which affords the opportunity for gentle exercise and stretching. It also enables some people to do things they are unable to do on land - for example, someone who can't stand or walk on land may be able to stand and walk in water.

Watsu, a form of massage done in the water, combines the benefits of ordinary massage and being in the water. Both massage and Watsu should be done only by certified professionals. www.Watsu.com.

Standing Frame

For some people with FSHD who can no longer walk, standing with the aid of a standing frame can be beneficial. A standing frame is a piece of equipment that can straighten and elevate a person into a standing position, and support him in that position. Using a standing frame can help maintain range of motion and reduce the possibility of contractures, relieve pressure on the posterior, increase circulation, reduce stress, realign the internal organs and provide weight bearing to preserve bones.



Surgical Treatments in FSHD

A number of surgical methods have been developed to address some of the debilitating loss of functional muscle such as <u>scapular winging</u>. Not every patient is a suitable candidate for surgery. A surgeon must have a thorough understanding of FSHD, experience with the procedure, and must carefully assess each individual to determine the risk and potential benefit, including whether a patient will be able to go through physical rehabilitation after surgery.

Consult an occupational therapist (OT)

An occupational therapist (OT) is a healthcare professional trained in rehabilitation who helps people learn how to do the activities of daily life. OTs can help FSHD patients improve function, increase comfort and reduce stress, fatigue and risk of injury. They can also provide home visits to observe the actual conditions of daily living and recommend ways of improving safety, efficiency, and comfort. Consider the viability of coordinating a joint home visit by an OT and a physical therapist as this can be especially effective. Some OTs are also experts on workplace ergonomics. Many employers are willing to pay for an ergonomics consultation. If an employee has a disability as defined by the Americans with Disabilities Act, the employer may be legally required to do so. Simple and inexpensive modifications in the workplace can often improve the ability of someone with FSHD to perform efficiently, comfortably and safely.

Use voice recognition software

Computer use can strain and fatigue the hands, wrists and arms of people with FSHD; this can be alleviated via voice recognition software.

Assistive technology

As FSHD progresses, patients may need to use assistive technology and equipment including wheelchairs, scooters, canes, walkers, lifters, reachers, ramps, electric beds and commode/shower chairs. OTs are experts in assistive technology and equipment – consult them for more guidance.

Driving

Some people's FSHD progresses to the point where they begin to feel uncomfortable or unsafe driving. At this point it is critical to have their driving evaluated by an adaptive driving expert. Some hospitals have adaptive driving programs which are usually staffed by an OT who is also a certified driving instructor. Evaluations are expensive and insurance usually doesn't cover them. But if driving is necessary to maintain employment, the rehabilitation agencies of some states may pay part or all of the cost of an evaluation, adaptive driving equipment and training. An increasing variety of adaptive driving technology is available, from simple mechanical hand controls to sophisticated electronic controls.