



## An Overview of Anesthetic Concerns for Persons Diagnosed with FSHD

by Raymond A. Huml, MS, DVM, RAC

### Introduction

I became part of the FSHD family when my daughter was diagnosed with FSHD at Duke University's Muscular Dystrophy Association Clinic in 2003. While I have come to understand that, as yet, we cannot change FSHD genetics or cure FSHD, I believe that some potentially life-threatening situations for persons with FSHD can be minimized or eliminated. After a recent trip with my daughter by ambulance to an emergency room (ER) for a suspected seizure, I have come to the conclusion that every person with FSHD and their caregivers should be prepared to give details about FSHD and key potential anesthetic complications associated with it, while in the ER, the intensive care unit (ICU), or prior to elective surgery.

This article will examine several key anesthetic concerns for persons with FSHD and is intended to encourage those with FSHD (and their caregivers) to be proactive in providing information about FSHD to those delivering medical care. *It is not intended to serve as medical advice.*

### Anesthetic concerns

Even within the high-quality medical care system in America, not all medical staff—including nurses, residents, and interns—are familiar with muscular dystrophies and neuromuscular diseases.

- Regarding my recent trip to the ER, it would have been helpful to have a laminated one page description, with websites, available to give to the medical personnel. The FSH Society can also act as a resource in providing information and [fact sheets](#).

The basic anesthetic concern for all persons with FSHD is that the respiratory system, due to a weak diaphragm muscle or chronic response to elevated carbon dioxide levels, can be additionally compromised with oral, intravenous, or inhaled drugs (or even the inappropriate use of oxygen). For example, muscle relaxants, like valium, can suppress respiratory function by compromising the diaphragm. Opioids (for pain), while not directly affecting the lungs, can have central (i.e., brain) effects that can compromise the respiratory system.

Respiratory insufficiency can occur in all persons with FSHD. The symptoms and signs of respiratory insufficiency should be evaluated during routine clinic visits; this is especially important in people with moderate to severe FSHD. The first pulmonary visit can be used as to document baseline function. Subsequent visits can be used to determine if pulmonary function is changing over time.

- *After meeting with a pediatric neurologist with my daughter as a follow-up to the ER visit, I discussed my concerns regarding anesthesia, and we agreed it would be prudent to get a baseline pulmonary function exam.*

Regular monitoring of respiratory function is important because insufficiency can occur gradually over a long period of time without clinical signs. Symptomatic respiratory insufficiency can initially be managed with night time non-invasive pressure support like a Bilevel Positive Airway Pressure (BiPAP) machine which helps get more air into and out of the lungs. In severe cases, FSHD patients may require the use of invasive mechanical ventilation (although some practitioners dispute this).

In standard practice, trauma, ER, ICU, surgery and anesthesiology settings, care should be taken not to suppress respiratory drive with narcotics. Oxygen supplementation can be detrimental to patients with hypercarbic (high CO<sub>2</sub>) respiratory failure and lead to worsening (i.e., higher) carbon dioxide (CO<sub>2</sub>) levels. Oxygen should generally not be administered unless BiPAP or similar ventilatory support is also being used.

- *During the 15-minute ambulance ride to the ER with my daughter, I found it impossible to convince the EMS staff that oxygen supplementation was exacerbating the symptoms—especially since there were no obvious signs of respiratory insufficiency—like increased or labored breathing or blue lips—and normal pulse oximetry readings.*

Your physician and pulmonologist can help you periodically monitor CO<sub>2</sub> levels in the office or evaluate pulmonary function in the hospital, or even by a nocturnal oxymetry study. The timing for the periodic monitoring of pulmonary function will be determined by the results of the baseline visit, any new clinical signs, you, your pulmonologist and your neurologist may discover.

### **Elective surgery, surgery for dentistry, or surgery that allows some planning time**

Anyone having surgery should be aware of the general risks associated with the surgical procedure and/or complications that may arise during and/or after it. While certain risk factors are increased in people with FSHD who undergo anesthesia, each individual is different and reactions to anesthetics may be different.

As with all medical procedures, you should talk to your physician about the specifics of your case and what you should anticipate and hopefully have baseline data available for physician review—like pulmonary function data. The following is a brief description of complications that may occur among people with FSHD who have surgery.

### **Complications from anesthesia**

While this list is not exhaustive, complications in patients with FSHD who have had anesthesia during surgery can include the following:

- respiratory distress
- rhabdomyolysis (muscle degeneration)
- cardiac complications
- myotonia (decreased ability to relax the muscles)
- generalized muscle spasms (particularly the jaw muscle)
- hyper and hypothermia (abnormally high and low body temperature).

Some of these complications appear to be related to the symptoms, severity and stage of FSHD, while other complications, such as respiratory or heart problems, appear to be related to secondary conditions. For example, while rhabdomyolysis appears to be a complication related to NMD symptoms, breathing problems are most often associated with a respiratory problem that is secondary to the muscle weakness caused by the disease process.

There are no definitive ways to completely alleviate all risks. However, there are several precautions, like pre-operative testing, that physicians can take to lessen the likelihood of these complications.

### **Pre-operative testing**

One way to prevent or alleviate complications from surgery in individuals with FSHD is through pre-operative testing. These tests may or may not be done based upon the information already contained in your medical records (i.e., symptoms and severity, whether or not you are known to have heart or respiratory problems, etc.). If your physician does not have sufficient information, she/he may run a variety of tests (see below for examples of preoperative tests) to determine:

- what types of anesthetics to use (i.e., local vs. regional vs. general);
- what drug combination to use for anesthesia;
- what types of pre- and post-operative measures should be taken to lessen the likelihood of complications.

For patients with FSHD who have not had these tests performed earlier, these pre-operative tests are recommended:

- baseline pulmonary function testing;
- baseline neurologic testing (clinical or diagnostic like EEG);
- baseline cardiac testing such as an electrocardiogram (ECG).

### **Identification of a potential problem to medical persons**

Without a caregiver present or the ability to speak coherently, medical personnel may not know that you have FSHD. FSHD patients should take care with their advanced medical directives and health care proxies to be sure that the “do not resuscitate clause” does not put them at risk. Here are some ideas to consider:

- Carry a MedicAlert card in your wallet (such as our [downloadable version](#));
- Use of a bracelet, necklace, etc., from MedicAlert (or a similar service) to notify medical personnel about potential problems associated with FSHD;
- Carry medical information (in PDF format) on a USB flash memory drive.

Note that the use of a bracelet or necklace in adolescents can be problematic because, at this age, it identifies the person afflicted with FSHD to a nontargeted (e.g., non-medical) audience. It may be possible, in lieu of a bracelet, to carry such identification card in the wallet or purse.

## Conclusions

- Not all medical personnel are well-informed about FSHD or may not know that you have FSHD unless it is communicated to them either verbally or via a MedicAlert-type device.
- Persons with FSHD manifest a wide variety of clinical symptoms of the disease.
- There are some complications of anesthesia that are more prevalent amongst people diagnosed with moderate or severe FSHD.
- You and your caregiver should be an informed participant in any surgical procedure. It is prudent to learn more about the respiratory implications of anesthetics before they become a problem.
- Be aware of the potential complications that may be associated with the use of anesthesia with FSHD and discuss the risks and benefits with your physician before choosing a particular procedure.
- Review estate planning documents to ensure that they are consistent with the medical intervention you wish to receive if you are unable to communicate.

See also [Anesthesiologists](#). **References:**

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