

MEDICAL EQUIPMENT, ACCESS SOLUTIONS AND RESPIRATORY THERAPY

By Howard L. Chabner

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I have **facioscapulohumeral muscular dystrophy (FSHD)**, began using an electric wheelchair in 1990 and have used one full-time since 1996. Over the years fellow FSHD patients and friends with other mobility disabilities have asked for recommendations about medical equipment. Needless to say, each of us is different, but as our condition progresses, many of us confront similar decisions. This memo is intended for people with mobility disabilities, especially those with muscular dystrophy. It updates and supersedes my memo of January 2013.

This memo covers a wide range of topics, many of which, hopefully, will not be relevant for a significant percentage of readers. For some readers, it will be sobering and perhaps difficult to read about some of these subjects, but I chose to include the full range because over the years friends and acquaintances have asked about each of these things. I hope the disadvantages of being introduced to solutions you may never need and that may be unpleasant to contemplate are outweighed by the risks of not learning about something you may need to know until your need is imminent and your choices limited. Managing a progressive and unpredictable condition requires fighting the progression as much as one can while at the same time anticipating and planning for new needs on the horizon without making them a self-fulfilling prophecy. Choosing the right equipment can greatly improve one's quality of life and can liberate one from some of the limitations of having a disability.

Many thanks to **Alan Brown** for reviewing this memo (and the previous version) and making valuable, insightful comments.

Other memoranda I wrote that may be of interest are:

“Managing FSHD - One Patient’s Perspective”

“Tips for Dealing with Medical Insurance Companies” about making the case for coverage of medical equipment

“Condom Catheter - Men’s Liberty by Bioderm”

“Disability Insurance and the Decision to Stop Working”

My wife **Michele DeSha** and I have written articles about wheelchair accessible travel in France, Italy, Israel and Spain. They are published on **WheelchairTraveling.com** and **Global Access News**. Search our names and wheelchair accessible travel, or contact me for a list of links.

www.wheelchairtraveling.com

<http://www.globalaccessnews.com/travelarchives.htm>

The sections of this memo are:

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This memo is not medical advice. It is imperative that you consult your own medical professionals (primary care doctor, neurologist, physiatrist, occupational therapist, physical therapist, respiratory therapist, etc.) to advise you about medical equipment you may need.

1. Some General Observations

Document your equipment, parts, dealer, repairs and insurance company dealings with the same organizational skills, resourcefulness and rigor as you use in your job or professional career or serious hobby. Write down model numbers, contacts, phone numbers, prices, distributors, repair details and the like. It will take time initially, but you will save time and frustration in the long run.

Many repairs to medical equipment don't require specialized knowledge or even parts. If your spouse or friend is mechanically inclined, or if you have a good handyman or auto mechanic, they can do some repairs and maintenance. This will avoid having to schedule repairs with an equipment dealer and will save a lot of time. Some bike shops, for example, are happy to replace a wheelchair tire if you provide the tire.

Give feedback, positive and negative, but always specific and constructive, to manufacturers, dealers and technicians. Often they will listen and be glad that you've taken the time to appreciate their work and help them improve their product.

As change is a constant in life and in the world, so it is with medical equipment. Products are constantly changing. Technology advances, but this doesn't necessarily mean that every change is an improvement for every user. Engineering and design involve trade-offs, and a change that may be positive for many users may not be positive for everyone, especially considering the wide range and unique combination of abilities and limitations people have. Even for one individual, some changes in an item of equipment may be improvements while others make it less useful.

Engineers and designers like to design, marketers and salesmen like to market and sell, and there is an economic imperative to increase sales. There is constant pressure to control costs and reduce inventory. Industry consolidation may or may not be a good thing. A small, innovative company may be acquired by a competitor, a conglomerate or a private equity firm, which may have different priorities and values than those of the founders. A visionary founder may retire, and his vision may be diluted or even erased by successors or acquirers who don't share his vision or passion. Some components of a product are likely to be made by a third party, which itself may well be subject to the aforementioned factors, which means that the manufacturer probably doesn't have control over every component.

If there's a product that really works for you and you can afford to, it's a good idea to buy a second one as a backup and buy extra parts if they are available. For example, I have a great trackball but I'm concerned that one day it may no longer be made, so when I replace it every few years I buy an extra one just in case.

When you have a significant disability, it's essential to get a product that is reliable and easy to repair, and where the product, the manufacturer, the dealer and replacement parts are likely to be around for many years. When one relies heavily on equipment for day-to-day living, reliability is priceless. Be wary about buying an unusual, out of the mainstream product, especially one that's new to the market. Innovation and high-technology are great, but consider carefully the pros and cons of being a guinea pig. The medical equipment industry has matured over the decades, technology has advanced and products have improved, but for some types of equipment (for example, lowered floor wheelchair accessible minivans), the total number sold each year is far less than their non-disability analog, which means it takes a lot longer for the manufacturer to acquire engineering learning and real-world experience, incorporate customer feedback, and work out the kinks.

For example, several years ago a specialty company offered a wheelchair accessible **Dodge PT Cruiser**. I've heard of many problems with this vehicle, which is not surprising considering it combines wheelchair access, which is difficult to achieve, with an unusual vehicle of which relatively few standard ones are produced, and only a tiny number of accessible conversions, and it's a small vehicle to boot. Similarly, there is a power wheelchair manufacturer that has a highly unusual design and sells direct to consumers; I've known people who have had bad experiences with them, and there are many poor reviews online. Recently there was an article in the newspapers about a woman in the UK whose electric wheelchair (the article doesn't say which brand) was badly damaged by an airline, and because there are only

500 such wheelchairs in the world, it's been difficult for her to have her chair repaired or replaced.

The iBOT stairclimbing wheelchair is a cautionary tale about overreliance on technological solutions, and about the importance of considering whether an item has the potential for truly broad appeal or is likely to be a niche product. Its one truly revolutionary feature - the ability to climb stairs - came at a steep cost in maneuverability, size, stability, positioning support, and simplicity (plus wear and tear on residential stairways, and the stairclimbing feature itself had limitations, such as not being able to climb wedge-shaped stairs). These costs were unacknowledged by the designer and manufacturer. Most of the other features its designer and manufacturer showcased as unique were already available on certain other advanced power wheelchairs. I saw it demonstrated at a trade show: the young man who demonstrated it was able-bodied and had to hold onto a railing, balance his torso and shift his weight as the machine climbed each stair. Most of its fans were paraplegic veterans with strong upper bodies who otherwise would have chosen a manual wheelchair, and for whom a power chair was, at least arguably, not medically necessary. They prided themselves on their fitness, and were strong and agile enough to execute the tricky maneuver required as the chair traversed each stair. Although it was expensive, the iBOT wasn't hugely more costly - perhaps 20% more - than other advanced power wheelchairs which had been on the market a long time and which incorporated continuous improvements based on many years of real-world experience. Not many were sold, and it was produced for less than 10 years.

In 2016 there were news reports that the inventor of the iBOT, Dean Kamen, has partnered with Toyota to bring a new, improved version to market. It's a noble endeavor and I wish them well.

In short, a good product combines appropriate innovation, technology and engineering with real-world experience, a deep understanding of the needs, desires, abilities, limitations and other conditions of the users, and a consideration of the trade-offs that are inevitable in any design.

2. Equipment Decisions are Interrelated

Each item of medical equipment is used together with other items and in your own particular environment. In considering any item, it's critical to think about how well it can be used with your other equipment, how it will affect the other equipment, and how all of this will fit into your environment. For example, the decision about what type of mobility device to get has implications for what type of vehicle you will need, and vice versa. For various reasons I chose to get an accessible minivan without a kneel feature; this was feasible only because my wheelchair has a tilt feature, which enables me to compensate for the steeper minivan ramp. The conditions in your home may constrain and influence your choice of mobility device, and the decision to get a particular type of device may necessitate home modifications.

The vendors for one type of equipment aren't the same as for others. Understandably, each vendor is focused on selling the type of equipment he specializes in; even with the best

intentions, he may not be fully aware of how well or poorly that equipment fits in with your other equipment and your environment. You, the consumer, need to be proactive, creative and wide ranging in considering these interrelationships. Measure three times and cut once. It's also important to think ahead - a solution that just barely meets your needs today may become obsolete for you in a year or two. Although no solution is permanent or definitive, failing to plan ahead can be costly in safety, stress, emotions, time, money and logistics.

3. Insurance; Funding Sources; Taxes

It may take several rounds of fighting with your health insurance company to get a major item covered. Don't give up. But it may be better to pick your battles and pay for small items yourself if the insurance company refuses or if the hassle of the paperwork (including hassle for your doctor) outweighs the dollars involved.

Many states offer funding for durable medical equipment and items such as wheelchair accessible vehicle conversions through state vocational rehab agencies. Eligibility and funding levels are based on income, and often on whether the item is necessary to enable someone to work or to continue working. Some states also have loan programs to finance medical equipment and related items at below market interest rates.

Achieving a Better Life Experience (ABLE) accounts are tax-advantaged savings accounts designed to enable people with disabilities and their families to save for and pay for disability-related expenses. See IRS Publication 907 - Tax Highlights for Persons with Disabilities.

Many car manufacturers offer rebates (up to \$1,000 as of 2016) for adaptive equipment, whether a complete vehicle conversion or an external scooter/wheelchair carrier.

For medical equipment that isn't covered by insurance, the acquisition cost may be deductible as a medical expense for federal (and some states') income taxes if you itemize deductions. Insurance companies, Medicare and Medicaid may differ as to what they consider medically necessary equipment. Regardless of the insurer, the definition for insurance purposes is often narrower than for tax purposes. For example, I'm not aware of any insurer that pays for the conversion cost of an accessible vehicle. But for 2016 (and prior years) the IRS says that "You can include in medical expenses the difference between the cost of a regular car and a car specially designed to hold a wheelchair." (I'm not aware that this is expected to change for 2017 or future years.)

For 2016 medical expenses are only deductible for federal income tax purposes to the extent they exceed 10% of adjusted gross income. For those 65 and older, the threshold is lower. See IRS Publication 502 - Medical and Dental Expenses. If you or your spouse has a **Health Savings Account**, it can be used to reimburse the cost of medical equipment. See IRS Publication 969 - Health Savings Accounts and Other Tax-Favored Health Plans.

4. Occupational Therapy

An occupational therapist (OT) is a health professional who works with patients to help maintain the highest possible level of independence, safety and quality of life. Occupational therapists are concerned with helping patients in their “occupations” - the day-to-day tasks that “occupy” their time at home, at work and elsewhere. OTs are knowledgeable about medical equipment, assistive technology and other products that can aid people in their daily living activities.

Those whose mobility limitations have progressed to the point where daily living activities are becoming more difficult can benefit by periodic consultation with an occupational therapist for assessing and adjusting daily living activities to increase function and reduce stress, fatigue and risk of injury. Importantly, this often includes equipment evaluations and recommendations. If you are considering getting a mobility device such as a walker, wheelchair or scooter, an OT can be invaluable in helping you choose equipment and ensuring that your seating and positioning are optimum. Occupational therapy is typically covered by insurance; you must get a doctor’s order.

Home visits are essential - there is no substitute for having an OT observe the actual conditions of daily living in your particular environment and recommend ways to improve safety, efficiency and comfort. Home visits also benefit caregivers by teaching them how to reduce stress, burden and risk. Ideally, an OT and a physical therapist should do the home visit together.

OTs can also help you maintain the ability to work. There are occupational therapists who specialize in workplace accessibility and ergonomics. Many employers are willing to pay for an OT consultation at the workplace, and where the employee has a disability per the Americans with Disabilities Act, the employer is legally required to do so if appropriate. The ADA requires employers to provide qualified individuals with disabilities a reasonable accommodation in the workplace, including physical changes such as accessible restrooms, modifications to workstations, and voice recognition software. (A “qualified individual” is someone who is qualified to perform the job, by virtue of his or her education, training and experience, with or without a reasonable accommodation.) See “**A Guide for People with Disabilities Seeking Employment**” at <http://www.ada.gov/workta.pdf>. The US Department of Justice has a comprehensive website about disability rights at www.ada.gov.

5. Sources of Information; Disability Trade Shows; Abilities Expo

A good way to learn about medical equipment, assistive technologies, and services for disabled people is by attending disability trade shows. There you can talk to vendors, see and try medical equipment and other products, get literature and exchange information with other disabled consumers. If your condition has reached the stage where you realize it’s time to begin considering equipment such as scooters, wheelchairs and accessible vehicles, but you’ve put off thinking about these things concretely, attending a trade show is a great place to start. As in so many other aspects of coping with muscular dystrophy and other progressive medical conditions,

it's important to think ahead in order to make well-informed, deliberate decisions, maximize your choices and plan your finances, while at the same time not obsessing about things, thinking so far ahead as to make the need for equipment a self-fulfilling prophecy or getting equipment before it's truly necessary.

The largest and best-known show is the **Abilities Expo**, held annually in several cities throughout the US. There are also smaller local shows.

www.abilitiesexpo.com

Quest magazine, published by the **Muscular Dystrophy Association**, is a good source of information, which can be found in its feature articles and advertisements.

<https://www.mda.org/quest>

6. Wheelchairs and Scooters

The Decision. Deciding to begin using a mobility device is one of the most difficult decisions for people with muscular dystrophy and other mobility-limiting progressive medical conditions. The decision is always difficult and painful, and almost everyone fights hard against beginning to use a device. There is no one right moment. The decision is best made with the help of your spouse, partner and other close family members, and your doctor, physical therapist and occupational therapist.

I began using a power wheelchair in 1990 and have been unable to walk at all since 1996. Initially I began using a wheelchair all the time outdoors and most of the time indoors. I never used a scooter, resisting any mobility device (with the result that my activities became increasingly limited) until an occupational therapist convinced me to try a manual wheelchair. After it sat unused in my basement for months, I began using it, and it soon became clear that my arms weren't strong enough to push myself more than short distances on flat surfaces, so I needed a power wheelchair. Using a manual chair was an essential first step, however - it got my feet wet in a practical sense, liberated me from the fear that using any mobility device would be a form of surrender, made me realize I would still be able to stand and walk a bit, freed me and my wife from constant worry about falling, liberated her to focus more energy on herself, and enabled me to notice and enjoy things about the world again that had been escaping me because standing, walking and simply not falling were taking so much energy and concentration.

Choosing a mobility device is a huge decision. Before deciding, it's imperative to test drive the equipment for several days - not hours - around your home, your workplace, your community, and the other environments and conditions of your everyday life. Wheelchairs can be individually customized along many variables such as features, configuration and dimensions, and small changes in any variable can make a big difference. Your vendor should work with your occupational therapist to ensure you are making a choice that is right for you. A power wheelchair is a large financial investment. Insurance should pay most of the cost, but if you are

unhappy with your decision, you're unlikely to get another chance for many years unless you pay for it yourself or your physical needs change significantly.

Scooters. A scooter can be good for part-time use for someone who is finding it increasingly difficult to walk and needs a device to enable them to go longer distances, conserve energy, reduce the risk of falling and navigate crowds. However, in my opinion, as one's progression continues and he or she needs to use a mobility device for increasing amounts of time, and if their arms are too weak to push a manual wheelchair for long distances and over different slopes and types of surfaces, they should seriously consider a power wheelchair.

A good wheelchair is much more maneuverable than a scooter. The difference is apparent everywhere: at home, at work, in stores, restaurants, theaters and public transportation. They can come much closer to desks and tables. Power wheelchairs can be equipped with seat functions that scooters cannot, including an elevating footrest, reclining back and tilt-in-space (where the seat can tilt backwards, so the front edge is higher than the rear and the seat is no longer parallel to the ground). Like wheelchairs, scooters can be equipped with a seat elevator, but it generally can't go as high. And some scooters have reclinable backs, but they are not electronically operated and cannot recline as far as a power wheelchair.

As one spends more time seated in a mobility device, it becomes increasingly important to have an appropriate, well-designed, high-quality, supportive, comfortable seat cushion and back; here, too, wheelchairs have a great advantage. Also, for people who can't stand, it's much easier to transfer in and out of a wheelchair than a scooter. Finally, although this may be counterintuitive, in my experience a good power wheelchair is more conducive to closer physical contact and interaction with other people. Unlike a scooter, there is no mast and handlebars to get in the way.

Permobil. My first power wheelchair was a **Permobil**, which lasted 12 years. In 2002 I replaced it with another Permobil. I got my third Permobil in September 2012. The Permobil is made in Sweden and was originally designed by **Per Udden**, a medical doctor and visionary who was decades ahead of his time. I believe it is absolutely the best wheelchair on the market. It is more expensive than others, but besides being better in absolute terms, it's a better value for the money considering its comfort, design, maneuverability, features, look and feel, quality, reliability and durability. My Permobil invites social interactions - people feel comfortable getting close to it, and it's a conversation starter. Over the years I've tried other power wheelchairs as demos, loaners and backups, seen others at disability expos, and my enthusiasm for Permobil has only grown.

www.permobilusa.com

Seat Functions. Electrically operated seat functions available on Permobil include elevating, articulating footrests; reclining back; tilt-in-space; and a seat elevator that can raise the seat eight inches or more. I highly recommend these four features. Permobil also can be equipped with a standing feature, where the user's knees and chest are held in place by brackets and he or she is elevated into a standing position. But if the standing feature is chosen, there may be trade-offs or limitations with respect to the other features. I have a stationary standing

frame in my home (see Section 13, below), so it's more important for me to have the four other features without any trade-offs.

Paradoxically, it was my first Permobil that enabled me to continue walking for as long as possible. It had become extremely difficult to get up from a seated position. With my Permobil I was able to do this with much less difficulty by raising it to its highest position, raising one armrest, pivoting to the open side and leaning on the wheelchair to stand. Because I had to overcome gravity much less with this maneuver than when I had been getting out of an ordinary chair at normal height, I got out of the wheelchair and walked (indoors) fairly often.

What's more, these seat functions enable me to remain in my chair while having my teeth cleaned and for certain eye examinations, my annual physical and even electrocardiograms and other diagnostic procedures.

Drive Wheels. Permobil is available in front-wheel drive and mid-wheel drive versions. I've always had the front-wheel-drive model and have found it to be marvelously maneuverable, but there are pros and cons of each version, and it's a good idea to try both before making a decision. If you are intending to drive an automobile from your wheelchair, you'll most likely need front-wheel-drive.

Footrests. Two types of footrests are available: a single, large, continuous footrest; and two separate, independent footrests. I prefer the former. Try both versions before deciding.

Joystick. Permobil offers several joysticks, all manufactured by **PG Drives Technology**, which also makes joysticks for other wheelchair brands. I am thrilled with the shape, size and feel of the joystick assembly on my old (2002 - 2012) chair, **Pilot Plus**, which is a curved, slightly triangular shape, but it isn't available on the new chairs. I tried the new standard joystick, **R-Net/ICS**, and found the assembly to be too long, too large, at an uncomfortable angle and not ergonomic. R-Net/ICS has a display screen with graphics, and high tech features including sophisticated diagnostics, advanced programmability and multiple memory modes (similar to a car seat, where the system remembers several favorite seating positions), which I found unnecessary for my needs. The rehab technology specialist I worked with mentioned that other clients with muscular dystrophy have similar opinions about this joystick. It's important to keep in mind that wheelchairs are designed for users with many types of disabilities, and features that are essential and worth major trade-offs for some people are unimportant (or even detrimental) for others.

Instead, I ordered the **LED R-Net** joystick, which has the same advanced driving capabilities but no display screen, less advanced diagnostics and programmability, and no memory modes. The LED R-net joystick assembly is smaller and more ergonomic than the R-Net/ICS, and similar in shape to my old joystick. (It's good, but I still prefer my old Pilot Plus, which is a bit more compact.) Yet another option is the **VR-2**, which has a more triangular assembly and is the most similar in shape to the Pilot Plus, but it has less advanced driving capabilities and, a deal breaker for me, cannot support four seat functions. However, this may have changed since I got mine. Permobil has a lot of options; it's important to ask what's available and to try more than one.

Another choice for the joystick is how it swings away. My joystick can be swung away on one pivot point, enabling the chair to come very close to a table or desk. I'm accustomed to driving the last several inches with the joystick at an angle when bringing myself close to a table or desk, and it's easy to compensate for the angle while steering. An option I tried but didn't like is where the joystick swings away by two hinge points, an arrangement that enables it to remain facing forward (perpendicular to the user) while swung away from the armrest. This enables the user to drive while the joystick assembly is swung away without having to compensate for the angle. But with it swung away in this manner, it was quite far from my arm, difficult to reach and awkward to drive. Also, there is a lot more metal than with the single pivot point design, making the whole mechanism less compact. This feature might be good for someone with a cognitive impairment, but for many people with muscular dystrophy the simpler, single pivot point option may well be better.

Seat Function Controls. A standard feature of Permobil is that the seat functions can be operated in two ways: by the joystick (you scroll through a menu), and by individual switches in a small box. I almost never operate the seat functions by the joystick, but it's a good feature for someone who has the use of only one hand, and it's also useful if the individual switches are broken. And if a seat function doesn't work in response to its individual switch, one can troubleshoot the problem by seeing whether or not the function can still be operated through the joystick. If it can, the problem is the switch, not the seat function motor or wires.

You can choose to have the seat function switch box on the same side as the joystick or the opposite side. Mine is on the opposite side because it's more comfortable physically (the switch box can be farther forward on its own than if it were behind the joystick assembly) and because, as described below, it's important for me to be able to adjust the seat functions while driving.

Another choice is whether the seat function switches can operate only with the joystick power on, or with the joystick power either on or off. For me it's important to be able to operate the seat functions with the joystick power on or off. Often when going uphill or downhill I adjust the tilt, backrest and footrest while driving. I adjust these angles as I proceed, fine-tuning or adjusting them if I encounter an obstacle, change of slope or cross slope. Other times it's important to be able to operate the seat functions with the joystick power off, to ensure that the chair won't move - for example, at a restaurant or in a restroom, or when transferring to bed. In these situations I can adjust the footrest and backrest while ensuring that the chair won't move if I (or someone who is helping me, or a waiter, or a customer at the next table) accidentally touch the joystick.

Tires. The drive wheels can have either pneumatic tube tires or flat-frees (solid tires or tube tires with solid inserts). The small non-drive wheels are always flat-frees. On my first two Permobils I had pneumatic tube tires because the ride is more comfortable, but there was the occasional flat tire. On my third Permobil (delivered in September 2012) I ordered solid tires, but found the ride was not as comfortable and the chair not quite as maneuverable as with pneumatic tires, so I replaced them with pneumatics. (Maneuverability and a comfortable ride

are among the reasons why automobiles use pneumatic tires, not solid ones. So it's logical that the same applies to wheelchairs.)

Stan's Tire Sealant is a liquid that solidifies when exposed to air. You remove the metal valve stem, remove some air, apply the product inside the tube, reflate, and reattach the valve stem; as the tire rolls, the product solidifies and seals when it touches the leaky hole. It's fast and easy to apply, and comes with a small wrench for the valve stem. I tried it and the sealant held perfectly for almost a year. The only potential problem is that the product may solidify around the valve, making it difficult or impossible to add air to the tire, but this hasn't been a problem. One bike mechanic recommended applying this product as a prophylactic, not just if there is a leak. Stan's Tire Sealant is not well known, especially among wheelchair technicians. A bike shop is the best place to find it. There's a similar product called **Slime**, which I haven't tried but which bike mechanics say is messy and not nearly as effective.

www.stanstiresealant.com

Batteries and Charger. Batteries will be included with your wheelchair or scooter. I strongly recommend sealed gel cell batteries, which are maintenance-free, non-spillable, safer, and better environmentally than wet batteries. They are also acceptable to airlines, which prohibit or strongly discourage wet batteries. (Air travel is difficult enough for passengers in power wheelchairs; wet batteries compound the problems for everyone.) Sealed gel cells have become standard, but it's a good idea to make sure you get them.

Different insurance companies have different policies about how frequently they will pay for new batteries. It's a good idea to learn about your insurance company's rules.

A battery charger will also be included. **Lester Electrical** and **MK Battery** are two of the leading charger manufacturers. Lester is an employee-owned company that makes its products in Nebraska, so it's the way to go if you want to buy American.

www.lesterelectrical.com
www.mkbattery.com

Lights and Other Accessories. Headlights and tail lights are available as an option on Permobil and other power wheelchairs. They are essential for safety, and also useful in many indoor situations (for example, going into a dark theater, and entering a room in your house where the lights are off). Another useful option is a carrying case, which, among other things, can be used to carry your wheelchair tools at all times. These options typically are not covered by insurance, and lights aren't cheap, but they are a worthwhile investment.

Insurance Coverage of Wheelchairs and Scooters. Whether you have private insurance or Medicare, be prepared to fight for the wheelchair and features you need. Thorough preparation is essential - enlist the help of your doctor, occupational therapist, physical therapist and wheelchair dealer in preparing a comprehensive letter of medical necessity, which must be based on a recent physical exam. Ask for an opportunity to review and comment on the letter before it is submitted to the insurer. Rarely will everything be covered, but ultimately most of it will.

The insurer's initial response may be to cover some items and deny others; be prepared to appeal.

Medicare will cover only whatever type of mobility device is necessary for the patient to move around in his or her home, but not outside the home. So, at least in theory, if you are able to move around in your home in a manual wheelchair and only require a scooter or power wheelchair outside the home, Medicare will only cover a manual wheelchair. (Of course, a doctor who understands the patient's needs will not fall into that trap.) This is an unfairly and unrealistically narrow definition of medical necessity. Is it reasonable to expect that people remain in their homes?

Under the same theory, lights are not covered (by Medicare and by many private insurers) because they are deemed not medically necessary for one to be able to move around in one's home. (This is illogical. Either your wheelchair will have lights or it will not. Should you be forced to forgo an essential safety feature because it is only essential outside your home? Or should you be expected to stay at home at night because your wheelchair doesn't have lights and you don't want to run an increased risk of being hit by a car when crossing the street? This may seem trivial, but it isn't: a good light package on a power wheelchair is expensive and can represent a nontrivial investment.) Also, Medicare categorically will not cover a seat elevator, claiming it is never medically necessary. These distinctions and policies are illogical, medically incorrect and morally unjustified, but they are unlikely to change anytime soon, so you must be aware of them in order to plan and budget accordingly.

Private insurance may be more flexible, but many of the guidelines of private insurers are based on Medicare.

See my memo "**Tips for Dealing with Medical Insurance Companies**" about making the case for coverage of medical equipment.

7. Accessible Vehicles; Adaptive Driving

Related to the decision about getting a mobility device is how to transport it and, more broadly, what implications it has for driving and transportation.

External carriers are available that attach to the rear of a car and can carry scooters and some wheelchairs. This is a good solution for people who can get out of their wheelchair or scooter, stow it and walk to the driver or passenger door.

For those who are unable to do this, or who want to remain in their wheelchair, or whose wheelchair is too large for an external carrier, the two solutions are lowered floor accessible minivans and modified full-size vans. Lowered floor minivans began to be produced around 1988 and are now the most popular option. They are available with a ramp on the passenger side or at the rear; a side ramp is the most popular by far, except for taxis, which typically have a ramp in the rear. **Chrysler/Dodge, Toyota and Honda** minivans can be converted. The leading converters of minivans are **BraunAbility** and **Vantage Mobility (VMI)**. Years ago Braun

acquired **Independent Mobility Systems (IMS)**, which at the time was one of the largest converters. The industry has become mature, and quality, reliability, finishes and options have improved. There is a robust market for used accessible minivans.

Full-size vans have lifts (not ramps) at the side or rear.

There are specialty companies that convert other vehicles for wheelchair access, such as **Chrysler PT Cruisers** (this particular conversion is no longer made), but production quantities are small, experience is limited, replacement parts can be scarce, and these conversions can be problematic.

There are several seating choices for accessible minivans. The driver or front passenger seat can be removed and the disabled person can remain in his wheelchair as the driver or front passenger. There are also power transfer seats, available on the driver or passenger side, which slide along a track on the floor and can be swiveled, so the disabled person can position his wheelchair in the middle of the minivan, transfer to the transfer seat, and slide it along the track into the driver or front passenger position. A transfer seat is a good choice for those who are able to transfer without much difficulty because it permits an able-bodied person also to use the seat, unlike when a front seat has been removed to make room for a wheelchair and the able-bodied would-be passenger is confronted with a big empty space. When a transfer seat is not installed, a standard feature of accessible minivans is that the front passenger seat is removable, which provides several options and offers flexibility for future changes.

If you are tall or your wheelchair is high, it's especially important to consider the interior height of the minivan and the doorway height. Typically the interior height of a standard conversion is around 10 inches higher than a standard, non-converted minivan, but this can vary depending on the make of the minivan. Conversion companies now offer an option with extra interior height, which is accomplished by lowering the floor even more than usual (and compensating by raising the entire vehicle so it has sufficient ground clearance), not by raising the roof.

Lockdowns are used to secure the wheelchair to the vehicle floor. **Sure-Lok** and **Q'Straint** are the leading makers of securement systems. Besides manually operated, spring-loaded retractable lockdowns, they offer electric docking systems that are mounted to the floor of the vehicle. Electric docking systems require a bracket to be attached to the bottom of the wheelchair, which reduces the wheelchair's ground clearance.

For those who want to continue to drive, it's important to consider hand controls. Hand controls include simple, manual controls and sophisticated, expensive electronic controls. This is a complex decision involving safety, cost, independence and many other things. Some hospitals offer **adaptive driving programs**, where an occupational therapist who is also a certified driving instructor will evaluate a disabled person's driving ability and offer potential solutions including different types of hand controls.

My first lowered floor accessible minivan had a driver's side transfer seat on a track. I transferred to the seat, left my wheelchair in the middle, and was able to continue driving from

the driver's seat for many years without needing hand controls. My wife was also able to drive from this seat. (I no longer drive. I reached the point where I would have required complex, expensive electronic hand controls, which at that time were fairly new to the market. With my wife's support, I decided to stop driving.)

In 2015 we bought a new Toyota Sienna lowered floor minivan converted by VMI. So far we are happy with it. When we began shopping we thought we preferred a folding ramp, as our previous minivans had, rather than in-floor, because of slightly higher ground clearance and because we thought there would be less that could go wrong, and despite the fact that folding ramps are noisier. Moreover, and importantly, because we live and park in a city with many high curbs, we were concerned that an in-floor ramp would hit high curbs, and thought we wouldn't be able to park on streets with high curbs. However, it appears that the industry is moving toward in-floor ramps only and eliminating folding ramps. So we ended up getting an in-floor ramp.

We bought a minivan with a manually operated ramp and without a kneeling feature. (This version is called "caregiver" or "companion" because the person in a wheelchair can't operate it independently). Getting a manually operated ramp is counterintuitive, but when my wife tried deploying the ramp it was quite easy. And with a manual ramp, one has more control to physically lift the ramp to meet a high curb, instead of pressing a button and keeping one's fingers crossed that it won't bang into the curb. Because I'm tall and my wheelchair is high, we got the extended height version. With the extra doorway height, it's easy for me to tilt my wheelchair when entering and exiting, so the steeper angle of the ramp compared to a kneeled minivan isn't an obstacle. We also figured that much less can go wrong with a manual version - the ramp isn't electric, there is no kneeling mechanism and the passenger rear door isn't electric.

Accessible vehicle companies bring demo models to disability trade shows. You can't test drive them, but you can go inside, check out the dimensions, and try the ramps and seats. Shows are a good place to begin learning about accessible vehicles.

Many car manufacturers offer rebates (up to \$1,000 as of 2016) for adaptive equipment, whether a complete conversion or an external scooter/wheelchair carrier.

For 2016 and prior years, the cost of the conversion is deductible as a medical expense for federal (and some state) income tax purposes. (I'm not aware that this will change for 2017 or future years.) See **IRS Publication 502 - Medical and Dental Expenses**.

www.braunability.com
www.vantagemobility.com
www.sure-lok.com
www.qstraint.com

Accessible Vehicle Rental. Unfortunately, regular car rental companies don't offer accessible vehicles, although they do offer standard vehicles with manual hand controls, enabling some drivers who use manual wheelchairs to rent from them. There are specialized companies that rent accessible minivans. **Wheelchair Getaways**, the largest rental provider, is a nationwide

association of small, locally owned businesses. Prices are much more expensive than renting an ordinary vehicle, and these companies don't have physical locations or parking lots, so you must arrange for delivery and drop-off, usually for a fee. Because there are so many accessible vehicle configurations, it's essential to confirm that the rental company has the seat configuration and interior height you need.

www.wheelchairgetaways.com

8. Shower/Commode Chairs

A shower/commode chair with wheels is a terrific solution for someone who can no longer stand, is experiencing increasing difficulty transferring from a wheelchair to the toilet, and is finding it more difficult to shower or bathe. It saves time and reduces the number of transfers, conserving energy and reducing strain.

For many years I've used a shower/commode chair made by **Nuprodx**, a company based in Sonoma, California, and cannot say enough good things about it. The Nuprodx chair is superbly designed, sturdy, comfortable, durable and easy to use. I use it at home and when traveling; it's easy to disassemble and fits in the overhead compartment of most airplanes. It's expensive, but the Nuprodx chair is very well made and has lasted many years. (I've replaced the seat cushion a couple of times). Most of the components are made in California, so an added benefit is that you are buying an American product. I've become friends with the founder/owner/designer, **Bruce Hammer**, a champion motorcycle racer who became a quadriplegic as a result of a motorcycle accident.

For those who have a roll-in shower, a shower/commode chair is ideal for showering and toileting. If you don't have a roll-in shower, it's still useful for toileting. In addition, for those who don't have a roll-in shower, **Nuprodx** makes transfer and tub chairs that enable the user to sit on the chair and slide it over the bathtub, in effect using the tub like a shower.

For those who are able to push a manual wheelchair, the Nuprodx shower/commode chair is available with large wheels and hand rims. For those who can't and need assistance, or whose space is limited, it's available with small wheels and push handles; this is the version I have. The small wheels have brakes, operated by levers.

I have the **Multi-Chair 4000**. I don't use the footrests because it's more stable to put my feet directly on the floor. Also, I have an older version in which the armrests are attached to the seat frame, not the back. I prefer that configuration because of its shape and stability.

The seat cushion/seat frame is available in two shapes - square and hourglass. Mine is square, which is better for people with muscular dystrophy. The hourglass version is designed for those with spinal cord injury.

The standard seat cushion/seat frame size is 17" x 17". I was able to order a customized version 20" wide by 17" deep. The extra width provides more stability. The seat cushion may be available in a choice of thicknesses; mine is 2 inches thick.

Nuprodx offers various options of seat size, seat shape, armrest style and other elements.

www.nuprodx.com

9. **Bidet - Biffy**

The **Biffy** is a bidet that attaches to a standard toilet. It's made by **American Biffy Company**. Many wheelchair users have found it helpful. However, if you use a shower/commode chair, the Biffy is unlikely to fit on the toilet.

www.biffy.com

10. **Electric Bed**

As it becomes more difficult to transfer into and out of bed, an electric bed can help. This is a home version of a hospital bed, in which the entire bed can be raised, the upper part can recline, the foot part can be raised, and these adjustments can be combined. Raising the bed to transfer out of it and elevating your wheelchair when transferring to bed can ensure that you are always transferring downward. Also, raising the head, the foot, or both can help you reposition, reducing stiffness, strain and fatigue. It's also beneficial to raise the head if you have a cold.

For many years I've been using an electric bed manufactured by **Volker**, a small German company. It's well designed, easy to operate, comfortable, not institutional looking, and it has been trouble-free. Until 2012, **Hertz Supply** of Allentown Pennsylvania (now **ProCare Medical**) was the US distributor. Over the years I've had a few questions and Hertz was always helpful. In 2012 Volker was acquired by **Hill-Rom**, a large US hospital bed company, and ProCare no longer carries Volker. ProCare now offers other brands of electric beds, which are on its website and look interesting.

I've had to replace the hand control unit a couple of times over the years, and the unit made by Hill-Rom, while similar to the original Volker one, isn't quite as good. I don't know whether Hill-Rom has made other changes to the Volker bed; if it has, they may not be improvements.

Some electric bed manufacturers also sell mattresses, or you can buy a mattress separately. If you buy a separate mattress, make sure it is flexible enough to be used with an electric bed. I have a comfortable, durable mattress made in San Francisco by **McRoskey Mattress Company**. One disadvantage of many electric beds, including Volker, is that they can't be used with a box spring, so it's especially important to get a comfortable mattress.

In considering an electric bed, it's important to find out exactly how low the bed can go and its total height in the lowest position, including the mattress. This is key when considering whether the bed can be lowered to a compatible transfer height relative to your wheelchair, scooter and shower/commode chair. (The highest position of the bed is usually less relevant, because it will almost certainly be possible to raise the bed higher than your wheelchair and other devices.)

www.volker.com

www.hill-rom.com

<https://www.procare-medical.com/>

www.mcrosskey.com

11. Lifts

As it becomes more difficult to transfer between wheelchair and bed, the time may come to consider getting a lift. Broadly speaking there are two types of lifts - freestanding, portable ones with wheels that roll on the floor (sometimes referred to as "Hoyer lifts," after a leading manufacturer) and ceiling lifts (also known as overhead lifts).

Freestanding lifts are simpler, less expensive, and can be moved throughout one's home, but they occupy a great deal of floor space, have a large mast, require clearance under the bed for their legs, and can be cumbersome to operate. They are available with electric motors or with manual operation using levers or cranks.

Overhead lifts have motors, enable the floor to be kept clear and are easier to use and more versatile, but are much more expensive and, generally, must be fastened to the ceiling joists. Some manufacturers also offer versions where the horizontal overhead rails don't need to be fastened to the ceiling but instead are supported by vertical bars that can be assembled and disassembled fairly easily. Ceiling lifts are available with a single track, which can be straight, curved or a combination, going from point A to point B. They are also available in an X/Y axis arrangement, with two parallel rails fastened to the ceiling, a perpendicular rail that slides along the fixed rails, and a motor that slides along the movable rail, thereby enabling the lift and the person being transferred to be moved anywhere within the footprint of the rails.

With both freestanding lifts and ceiling lifts, the person being transferred is typically in a mesh sling that is attached to the lift by straps. A variety of shapes and styles of slings are available. Initially it feels strange to be in a sling, and for most people it takes some time to get used to being in one.

In my bedroom I have a ceiling lift made by **Handi-Move International**, a Belgian company whose product name and US distributor are **SureHands**. This device has a unique design. Instead of a sling, the person being transferred is held in a contraption called a **Body Support**, consisting of a metal tubular frame with curved, hard plastic supports that fit around the sides of the person's rib cage, and metal leg brackets ("thigh supports") that support the person's thighs and are suspended from the tubular frame. It's an odd looking device, and it took

a few times for me to get comfortable with it, but I find it much more comfortable than a mesh sling, and one's body remains in a more natural and better supported position, more like being seated than with a mesh sling. As you are lifted, the frame tightens around your chest enough to secure your body in the Body Support, but not so much as to be uncomfortable. (SureHands also offers traditional mesh slings as an alternative to the Body Support.)

SureHands makes a well-designed, sophisticated product. Don't be put off by how strange the Body Support looks. The ceiling lift is available in a version that must be operated by a caregiver, and a version that can be operated by the person being transferred if he or she has sufficient strength and dexterity. SureHands also makes pool lifts and tub lifts.

The SureHands thigh supports come in several sizes, and it's important to get the right size. This is obviously a highly individual decision. When I first tried a demo model, I was given the shortest thigh supports, which were painful and didn't position me securely. Next I tried a medium size, which was better, and finally the longest, which are comfortable and work well for me. They're around 10 inches long at the longest point, so my weight is spread across a larger area of my thighs. They are contoured, meaning curved - an arc of a circle when viewed in section. The non-contoured style is angular when viewed in section. It's easier for my caregivers to put my legs in, and remove them from, the contoured ones, and it's much more comfortable for me.

SureHands makes an electrically operated freestanding lift that can accommodate the Body Support. A couple of years ago I bought a used one in Chicago, where I travel to visit family, but found that the size and style of thigh support from my ceiling lift was no longer manufactured. I contacted Handi-Move in Belgium and they were willing to fabricate a pair for me at a fair price. The long, contoured, gray, non-slip-proof thigh supports I have are #1056. Note that these are not the designated "muscular dystrophy" type, nor are they "slip-proof" (the slip-proof style has fabric inserts). They are not slippery, however, and my legs have never slipped out of them.

Besides being easier and safer for the person being transferred, lifts are much safer, healthier and easier to use for caregivers than lifting with brute force, which is why many hospitals are including ceiling lifts in their accessible patient rooms. Some hotels in Las Vegas even have them.

As with so many types of medical equipment, when considering getting a lift, it's imperative to try different types, measure your available space, and carefully consider the details and dimensions of your environment.

Medicare and most private insurers don't cover lifts. Not only that, but my ceiling lift was subject to California sales tax because the medical equipment sales tax exemptions are narrowly and specifically defined, and anything not on the list is taxable.

In addition to SureHands, leading manufacturers of lifts include **Guldmann**, a Danish company; **Liko**, a Swedish company now owned by the American company **Hill-Rom**; and **Prism Medical**, a Canadian company.

www.surehands.com
www.guldmann.net
www.liko.com
www.prismmedical.ca

12. Overbed Table for Reading

When it became difficult for me to hold a book, I bought an overbed table made by **Volker**. It's now carried by **Hill-Rom** and is item number **RT-985**. I use it only for reading, not eating, and it's in my living room. It's height adjustable, the reading surface can be tilted, the reading surface has a raised edge at the bottom to hold a book, the table moves easily on four small wheels, and the undercarriage is U-shaped, making it easy to roll my wheelchair under the table without the footrest hitting the table legs. The surface has enough space for more than one book.

<http://www.hill-rom.com/international/Products/Products-by-Category/furniture/overbed-table-rt-985/>

13. Standing Frame

For those who can no longer walk, standing in a standing frame is a beneficial form of passive exercise. A standing frame is a stationary metal frame that enables the user to stand in a safe position while the frame bears some of their weight. The user pulls their wheelchair up to the device and aligns their knees in the knee supports, a helper puts a sling around the user's posterior, the sling is attached to metal brackets, and the helper operates a lever that controls a piston and elevates the brackets. As the brackets are raised, so is the user. Standing this way enables weight bearing to strengthen bones, helps maintain range of motion and prevent contractures, relieves pressure on the posterior, increases circulation, reduces stress, improves alignment and functioning of the internal organs, and is psychologically (besides literally!) uplifting. A physical therapist should check to ensure the frame's settings are correct, your posture is good and you don't overdo it. A drawback of standing frames is that they occupy a lot of floor space.

Ever since I lost the ability to walk, I've used the **Grandstand**, manufactured by **Prime Engineering**, which is based in Fresno, California. The Grandstand is excellent - it is comfortable, safe and durable. Back in those days, my health insurance covered it, although I did have to fight the insurance company a bit.

www.primeengineering.com

14. Exercise Equipment

Over the years I've used exercise equipment manufactured by **Biodex**, an American company, in both inpatient and outpatient physical therapy. After experiencing the benefits of this equipment I decided to get a **Biodex Upper Body Cycle** to use at home. Biodex equipment is institutional quality, durable and well designed for wheelchair access. The Upper Body Cycle comes with a removable chair; with the chair removed, a manual wheelchair user can roll over the machine's horizontal base and use the equipment. The top of the base is around five inches above the floor, which is too high for my Permobil, so I had someone build a simple wooden ramped platform that I can roll on in order to clear it. The arm cranks are height adjustable and can be reached from a wheelchair.

The Biodex website doesn't appear to have the Upper Body Cycle currently. Biodex may have reduced its range of exercise equipment.

www.biodex.com

15. Ramps

Prairie View Industries of Fairbury, Nebraska manufactures reasonably priced, high-quality portable lightweight, non-skid aluminum ramps with safety edges on the sides. PVI makes folding portable ramps, solid ramps and large modular ramps with handrails. Over the years I've used PVI ramps and have recommended them to many stores and restaurants where it isn't feasible to renovate the entrance and install a permanent ramp. They are well made and durable. PVI ramps are the best I've seen.

Some manufacturers offer two-piece ramps. Each separate ramp is narrow, and the left wheelchair tires roll on one ramp and the right tires on the other. The concept of this design, especially for long ramps, is that it's lighter weight than a one-piece ramp, which is wider than the two pieces combined. This sounds good in theory, but I've tried two-piece ramps and strongly recommend against them. For most wheelchairs, whether power or manual, the front and rear wheels are not completely aligned with each other, so each ramp must be wide enough to accommodate the offset between the front and rear wheels, and also the two ramps must be placed an almost perfectly uniform distance apart. In practice, this just doesn't work. The industry may be phasing out this style.

www.pviramps.com

16. Voice Recognition Software; Microphones; Trackball; Center for Accessible Technology

Dragon Naturally Speaking voice recognition software by **Nuance** is an excellent computer solution for those whose hands and wrists get fatigued easily. I eliminate a large percentage of keystrokes by dictating, saving my hands for the mouse and some editing. (You can command the mouse by voice, but it's cumbersome.)

For a long time Dragon supported only the Windows operating system, but a Mac version became available several years ago. I use a PC with Windows and don't know anyone who uses Dragon on a Mac, so I don't know how good the Mac version is. Also, Apple also makes its own voice recognition software.

Dragon software is inexpensive but requires a computer with a fast processor and a lot of memory.

Dragon has some flaws, but each version gets better. Antivirus software, such as Norton, can interfere with it and degrade your voice files, especially over time as upgrades to the antivirus software are automatically downloaded to your computer. To avoid this, go into the antivirus software settings and set them so they don't run a virus scan on the Dragon files. Voice file degradation can also happen over time as updates are added to your operating system. Also, Dragon is improved if some of the language and speech features that are built into Windows are disabled. However, I've experienced these problems much less on the current version of Dragon, and these fixes may no longer be necessary.

If you don't want to use a headset microphone or a Bluetooth, another option is a desktop microphone. I have a superb one, the **SpeechWare 9-in-1 TableMike**. Made in Belgium, it has way more features than I use (the 3-in-1 and 6-in-1 TableMike versions are simpler), but has an extra-long boom so the base can be placed out of the way, far from my keyboard and trackball. To use a desktop microphone effectively you need to make sure there is no background noise in the room. I bought mine online from **Speech Recognition Solutions**, a small business owned by a medical doctor. Its website has an extraordinary amount of useful, detailed information.

A key tip for using Dragon is to make sure that the microphone is always in the same position relative to your mouth.

Instead of a mouse I use a trackball made by **Clearly Superior Technologies**. The base is very large and the ball is almost as big as a billiard ball, making it easier to use and less fatiguing.

I learned about many of these computer-related products from **The Center for Accessible Technology**, a nonprofit in Berkeley, California that provides information and training about computer access for disabled people.

www.nuance.com
<http://www.speechrecsolutions.com/>
<http://clearlysuperiortech.com/>
<http://www.cforat.org/>

17. Respiratory Impairment; Ventilatory Support; Cough Assist; Stacked Breathing Exercises; Anesthesia and Emergencies

Fatigue is often part of FSHD because our muscles have to work harder and they tire easily, so people with FSHD often accept fatigue as a constant condition and assume that nothing can be done except getting more rest and reducing their activities. But fatigue may also be caused by interrupted sleep due to respiratory problems. Doctors have become increasingly aware that some FSHD patients have respiratory impairment, including sleep apnea and daytime respiratory insufficiency. Whether the cause is due to FSHD, other factors or a combination has not been conclusively proven. Sleep apnea, for example, is common in middle-aged men, especially those with extra weight in their midsection. But it's logical to expect that weak chest, diaphragm and abdominal muscles have a negative effect on breathing, so as these muscles weaken in people with FSHD, breathing can be affected.

If you feel fatigued, especially if you feel that fatigue is increasing faster than the progression of your muscle weakness, and if you feel less alert during the day even though you seem to be getting enough hours in bed, mention these symptoms to your doctor. Also, tell your doctor if your spouse or partner notices unusually loud snoring, breathing cessation, or abrupt awakening and shortness of breath while you are sleeping. Your doctor could order **pulmonary function tests (PFTs)**, including **forced vital capacity** and **nocturnal oxymetry** tests. These are easy, non-invasive tests that don't require a hospital stay. Forced vital capacity can be measured in the doctor's office, and a nocturnal oxymetry test merely requires attaching a small, simple machine to your index finger using an alligator clip finger probe before going to bed one night. These tests measure the amount of oxygen in your blood. (You can buy an inexpensive probe oximeter, but it doesn't record the reading.) A full **sleep study**, requiring an overnight stay at a hospital or clinic, is a much more comprehensive test and, typically, would only be performed if your doctor concludes that the other tests haven't yielded enough information. In some cases a **blood gas study** may be performed in order to determine whether unhealthy levels of carbon dioxide are building up in your blood.

Because some doctors, even experienced neurologists, still don't associate FSHD with respiratory problems, your doctor may be reluctant to order pulmonary function tests; be persistent if you feel these symptoms.

If indicated by the tests, using a **BiPAP** at night or another mechanical ventilation device such as **CPAP** can increase oxygen and improve sleep. The pulmonary function tests should be repeated periodically to ensure that the machine's settings are appropriate. Your doctor should consult a respiratory therapist as early as possible and the RT should remain involved in monitoring your progress on an ongoing basis. A good RT is worth her weight in gold. Note that these machines do not provide oxygen therapy; there is no oxygen tank involved. They use room air to provide mechanical assistance in exhaling, thereby ensuring that carbon dioxide is expelled and doesn't build up in the bloodstream.

I've been using a **BiPAP** at night for many years. From the moment I began using it the difference in my energy level was dramatic. Over the years the machines have gotten smaller, and the interface has become less cumbersome and more comfortable. My current machine is a **Respironics AVAPS System One**. Respironics is owned by **Philips Healthcare**. The machine is smaller than a shoebox. It has an internal automatic voltage converter, so I can use it when traveling to Europe, where the voltage is 220/240, without having to make any adjustments.

These machines have filters to filter out dust, which must be replaced periodically. They also have a small, removable computer card that records a multitude of data, including hours that the machine has been on, your breathing rate and lung volume. A respiratory therapist can download the data, interpret it and decide whether any adjustments are needed.

A humidifier can be attached to BiPAP and CPAP machines. Your respiratory therapist and doctor can help you evaluate this option. These machines can make your nostrils dry. I find it very helpful to use an over-the-counter nasal saline solution when needed.

There are a wide variety of interfaces, ranging from full face masks to small interfaces with “nasal pillows.” I use the **Swift FX** by **ResMed** with nasal pillows; it’s the smallest and most comfortable interface I’ve tried. If you try a BiPAP or CPAP and find it uncomfortable, your respiratory therapist should work with you and help you try different interfaces until you find one that’s right for you. Many interfaces have silicone or plastic components that touch your cheeks and can irritate your skin; fabric covers are available to mitigate this. The interface and tubing should be replaced periodically because they stretch out over time and the seal becomes less tight.

Those with respiratory weakness have difficulty coughing and their cough is weak, making it more difficult to recover from a simple chest cold and increasing the risk of pneumonia. There is an electromechanical machine called a **Cough Assist** by **Philips Respironics** that helps clear secretions. Your doctor and respiratory therapist may recommend having this machine in your home so it will be available to you in case of a chest cold.

Stacked breathing exercises can be helpful for people with respiratory weakness. (But they are not a substitute for mechanical ventilation.) They involve a facemask attached to a football-shaped plastic bladder. The device is called a **bag valve mask (BVM)**. A helper holds the facemask around the mouth of the person doing the exercise. The person inhales rapidly several times in succession without exhaling while the helper squeezes the bladder to push air into the person’s lungs. This exercises the chest and diaphragm muscles and increases oxygen intake. These exercises don’t require much time and a family member can help you do them.

Respiratory therapy can be complex, and there are many therapies and types of equipment available. If you have respiratory weakness, a knowledgeable, patient, determined respiratory therapist and doctor can make a huge difference in your quality of life.

Anesthesia. People with FSHD who have respiratory impairment may also need to be especially careful when undergoing anesthesia. If you have respiratory problems, it’s imperative to discuss this issue with your doctor, and ask him or her to discuss it with the anesthesiologist if you are undergoing surgery.

In Case of Emergency. **Note: I am not a doctor or other health professional, and the following is not medical advice. It is my recollection of what doctors have told me. It’s included here for general information purposes; it’s imperative to discuss these issues with your own doctor.**

As the carbon dioxide level goes up, the oxygen level goes down. Weak diaphragm, trunk and chest muscles can cause carbon dioxide to go up. Pulse oximeter finger probes measure oxygen but not carbon dioxide. Giving oxygen when carbon dioxide is up can blunt respiratory drive, making carbon dioxide go even higher. If a patient with FSHD who has respiratory weakness is in the emergency room with respiratory problems, blood gas should be checked, not just oxygen via pulse oximeter. If carbon dioxide is up, oxygen should only be given with extreme caution.

For people with normal respiratory function, when their body senses increased carbon dioxide, that is a trigger to increase breathing - it triggers their respiratory drive. Another trigger is decreased oxygen level. But people with respiratory problems, for example emphysema, function with higher than normal levels of carbon dioxide; their body gets used to this, so they can't detect increased carbon dioxide levels the way someone with normal respiratory function can. So for them, decreased oxygen level is their only trigger to increase their breathing, not increased carbon dioxide. But if they are given oxygen, they have no trigger to increase breathing, and their carbon dioxide level can remain dangerously high.

According to my doctor, emergency room doctors and nurses would know about the mechanism described above, but they might not necessarily know that people with FSHD or other types of muscular dystrophy may have respiratory weakness similar to someone with emphysema. So it's important to make sure they are aware of this.