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EXERCISING *with a* Muscle Disease

I don't exercise. If God had wanted me to bend over, he would have put diamonds on the floor.
~ Joan Rivers

Physical fitness is not only one of the most important keys to a healthy body, it is the basis of dynamic and creative intellectual activity.
~ John Fitzgerald Kennedy

If it weren't for the fact that the TV set and the refrigerator are so far apart, some of us wouldn't get any exercise at all. ~ Joey Adams

Lack of activity destroys the good condition of every human being, while movement and methodical physical exercise save it and preserve it. ~ Plato

I really don't think I need buns of steel. I'd be happy with buns of cinnamon. ~ Ellen DeGeneres

Exercise: Love it. Hate it. Gotta do it. On the following pages, Quest looks at the many sides of exercise for people with muscle diseases:

- What does the research tell us?
- Which types of exercises are OK and which aren't, and how can you tell the difference?
- What are other people doing — or not doing — and what are the results?
- And of course, the ultimate question: Is it worth it?

On the next page you'll see a box of warnings — please be sure to start there. Much of the conventional wisdom about exercising isn't true for people with muscle diseases, and doing it wrong may cause irreversible muscle damage.

As you read through the exercise articles in the magazine (see the table of contents), one thing is clear: Each body is unique, and its owner knows it best. Please filter all this information through your own wisdom and experience, and please share it with your physicians, therapists and trainers.

And if you want to share your exercise experiences with others, please visit Quest online (quest.mda.org) and post a message on the Feedback page. Because one thing is definitely true about exercise: It's more fun when it's shared!

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WARNING: Read Me First!

What to watch out for when exercising

Everybody agrees that exercise is a good thing — but only if undertaken safely. Here are some warning signs that say **STOP**.

When muscle fibers are undergoing damage, they usually — but not always — announce it pretty quickly, with cramping and pain. But sometimes muscle destruction doesn't make itself known until enough damaged muscle fibers have released a pigmented protein called myoglobin into the blood and eventually into the kidneys and urine. Dark, cola-colored urine hours after exercising indicates this kind of muscle destruction.

Signs of muscle damage or impending muscle damage are:

- cramping in muscles
  (probably related to insufficient energy supply for muscles)
- pain in muscles
- weakness of exercised muscles
- dark urine that looks like cola, following exercise
  (seek medical care immediately if this occurs)

Many muscle diseases affect the heart muscle, impairing its ability to pump blood fast enough to keep up with the demands of strenuous exercise. Others can cause an irregular heartbeat or one that's too fast or too slow, and strenuous exercise can exacerbate these abnormalities.

Signs of cardiac stress are:

- shortness of breath
- chest pain
- very elevated heart rate (150 to 160 beats per minute) with even moderate exercise
- weakness
- nausea
- sweating
- a gurgling sound in the chest with breathing

(seek medical care immediately if these symptoms occur)

It can't be stated enough: Always consult a physician before undertaking a new exercise program.

Exercise Q&A

Q: What is exercise? Why do it?

A: Exercise is muscle exertion (use), requiring the expenditure of energy. It can be very roughly divided into two major categories: aerobic and resistance (sometimes called "strengthening"). Aerobic exercise helps maintain overall fitness, especially cardiac and respiratory health. If muscles are intact and functioning, then resistance, or strengthening, exercise helps increase muscle mass and the force muscles can generate. However, some muscle diseases severely limit the ability of muscles to increase mass or force because they limit the regenerative capacity of muscle tissue. Additional benefits of exercise are maintenance of flexibility, preservation of bone density and mood elevation. Because exercise requires energy (calorie burning), it offsets the intake of dietary calories and may help keep weight in check.

A stationary bike offers excellent aerobic exercise. Elastic band stretching is a good resistance exercise.

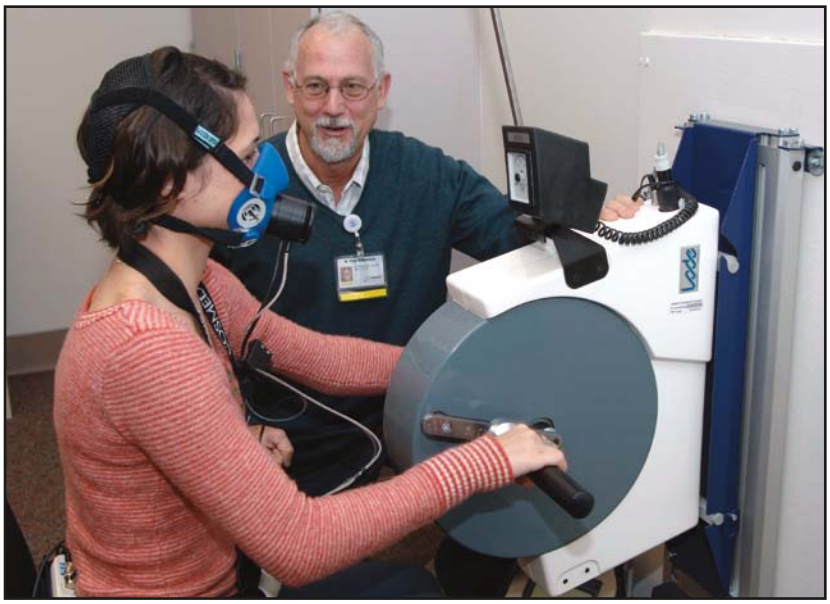
Q: What does or doesn't the medical literature tell us about exercise and neuromuscular disease?

A: Unfortunately, there isn't a lot known about exercise in neuromuscular disease, but some studies have been conducted that shed some light on the subject.

"We know so little," says Ted Abresch, director of research at the Research & Training Center for Neuromuscular Disease at the University of California at Davis. (The MDA Neuromuscular Disease Clinic at UC Davis is part of the MDA Clinical Research Network that supports studies in Duchenne muscular dystrophy.)

Abresch says some forms of exercise are beneficial in neuromuscular disease, but unfortunately, it's usually not prescribed unless patients specifically ask.

Studies of exercise, he says, have largely been "messy" and hard to interpret, because often there aren't adequate control groups (groups assigned not to exercise), and because people with different diseases are often studied together. At his center, he says, "We're looking at what people are actually doing, and we're looking at wearable technologies to see how much they're walking or sitting, so we can get a baseline and then see if an exercise prescription makes a difference." (A pedom-



Ted Abresch, who directs the Research & Training Center for Neuromuscular Disease at UC Davis, says studies of exercise in this area have been hard to interpret. Photo: Emi Manning, UC Davis

eter, which measures how many steps a person takes, is an example of a wearable technology. For more on this subject, see page 24.)

Abresch says the only thing he can say with certainty is that adults with slowly progressive muscular dystrophies (such as **limb-girdle, facioscapulohumeral** and **myotonic dystrophies**) showed improvements with moderate exercise. "The one program we did that seemed to work well was a walking program, where we put pedometers on people and asked them to walk 25 percent more over time than they did initially. They reduced weight, said they felt better and did better on quality-of-life testing."

Q: Can exercise hurt me?

A: If you have a neuromuscular disease (and even if you don't), some types of exercise can hurt you. It's always a good idea to consult with your physician before starting an exercise program and, ideally, to work with a physical therapist who understands your disorder. There are, however, some general precautions to take.

Many neuromuscular diseases affect the heart muscle, and that adds an extra requirement for caution. Cardiac abnormalities figure prominently in many muscular dystrophies, particularly **Duchenne, Becker, myotonic** and **Emery-Dreifuss** MDs, as well as in **Friedreich's ataxia**. If someone has a cardiomyopathy (heart-muscle degeneration), the pumping action of the heart is impaired, and the muscle won't respond to exercise the way a healthy heart would. It may thicken (hypertrophy) in an attempt to meet the increased workload of exercise, resulting in diminished blood-holding capacity. Or it may dilate, causing the heart to become floppy and relatively ineffective as a pump.

When cardiomyopathy or other types of heart disease render the heart's pumping action insufficient in supplying the body's cells with the

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Glossary of Exercise Terms

active exercise: exercise that a person does using his or her own strength (see passive exercise)

aerobic exercise: exercise that involves or improves oxygen utilization by requiring the heart and lungs to work harder. Aerobic exercises involve low to moderately intense activities performed for extended periods of time. Examples are walking, running, swimming and cycling.

anaerobic exercise: exercise that does not involve or improve oxygen utilization; weight lifting is an example

cardiomyopathy: degeneration of the heart muscle, such as occurs in some muscle diseases

conditioning: training to perform better for a long period of time; usually done through aerobic exercise, such as walking, swimming, etc.

coordination: the ability to integrate muscle movements to perform specific functions, such as walking, running or manipulation of small objects; eye-hand coordination is the ability to integrate what one sees with subsequent muscle action

concentric muscle contraction: a muscle contraction in which the muscle fibers shorten as they contract; flexing the arm at the elbow is an example

contraction: what muscles do when they're active; refers to protein filaments actin and myosin sliding over each other

contracture: permanent shortening of a muscle or tendon, resulting in a permanent "freezing" of a joint in a certain position; occurs when muscle weakness or spasticity prevents normal range of motion over a long period of time

eccentric muscle contraction: a muscle contraction in which the muscle fibers are pulled apart (lengthened) even though they're being activated to contract; for example, eccentric contractions occur when straightening the elbow gradually while holding a weight, so that the arm doesn't extend completely and suddenly

elliptical trainer: stationary exercise device that simulates walking or running without causing the joints to be subjected to much force

exercise: muscle exertion (use) involving expenditure of energy

fitness: the ability of the circulatory and respiratory systems to supply nutrients to skeletal muscles during sustained energy expenditure (exercise) and the ability of muscles to respond

forced vital capacity (FVC): the amount of air one is able to exhale after inhaling as completely as possible

interval training: repetitions of exercise interrupted by periods of rest or low activity

isometric muscle contraction: a muscle contraction in which the muscle fibers remain approximately the same length even though force is being exerted; an example is pressing the hands against a wall without extending the arms

maximal: full-out; exercising maximally means exercising as hard and as fast as one possibly can

myoglobinuria: the presence in the urine of myoglobin, which is released by damaged muscle fibers; myoglobin turns the urine brownish or cola-colored and can damage the kidneys; its presence indicates severe, acute muscle damage

oxygen consumption: the amount of oxygen taken in during exercise or at rest; can be determined directly by measuring oxygen intake and carbon dioxide exhalation through a metabolic mask or can be approximated indirectly by measuring heart rate

passive exercise: exercise that a person does without any exertion; for instance, having someone else move one's limbs in range-of-motion exercises (technically speaking, not really exercise, although it gives some of the benefits of exercise such as improving circulation, movement, comfort and flexibility)

physiatrist: a physician who specializes in maximizing functional abilities and quality of life for people with physical impairments; the specialty is called physical medicine and rehabilitation

physical therapist: health care professional who helps individuals develop, maintain and/or restore maximum movement and functional ability

range-of-motion exercise: exercise that involves putting a joint through its normal range of motion (as far as it can go in any direction); can be done actively or passively

resistance exercise: a form of exercise in which each effort is performed against a specific opposing force generated by resistance (for example, resistance to being lifted, pushed, squeezed, stretched or bent); sometimes used interchangeably with strength training

rhabdomyolysis: the rapid breakdown of skeletal muscle tissue

submaximal: exercise that's performed at less than maximum effort; sometimes defined as exercise that is not too strenuous to prevent talking, or exercise in which the heart rate does not go above 65-70 percent of maximum (maximum = 220 minus age)

tachycardia: abnormally rapid heart rate

trainer: a fitness professional who develops and implements an individualized approach to physical fitness, generally working one-on-one with a client; trainers generally do not have any medical background

treadmill: piece of indoor sporting equipment used to allow for the motions of running or walking while staying in one place

Recommended Exercises in Muscle Disease

Exercise purpose	Examples	Notes
improve endurance and conditioning	walking, running, swimming, cycling	• recommended for most people with neuromuscular diseases who choose to adopt an exercise regimen, with intensity and duration depending on ability and cardiac function • walking or running downhill not recommended • not to be done to exhaustion
build strength and muscle mass	weight lifting, working against resistance with equipment such as stretchable bands	• lifting light weights and working against light resistance are usually OK for most people with neuromuscular disease, unless in acute phase of an **inflammatory muscle disease**; stop at pain and before exhaustion
increase agility, coordination and balance	wheelchair sports, some video games, balance ball, horseback riding	• OK for most people with neuromuscular disease
maintain or increase flexibility	active or passive range-of-motion exercises; gentle stretching	• OK for most people with neuromuscular disease

Breathing Easier

In September 2008, Mary Gallo, 38, learned about a local pulmonary rehabilitation gym called Oxygym. She contacted her pulmonologist, who wrote a prescription for Oxygym's restrictive lung program.

"Oxygym is the first place I've ever attended that focuses on both breathing and physical therapy. I found a workout program tailored to my medical needs," says the Lindenhurst, N.Y., resident, who has congenital muscular dystrophy (CMD) with severe kyphoscoliosis and restrictive lung disease.

Gallo attends Oxygym for 90 minutes, three times a week. Each visit begins with a check-in, where her blood pressure and blood oxygen levels are checked.

Gallo works her arms by using a hand cycle, resistance band, and 3-pound weights.

She then spends 15 minutes on the treadmill, after which her blood pressure and blood oxygenation levels are checked again.

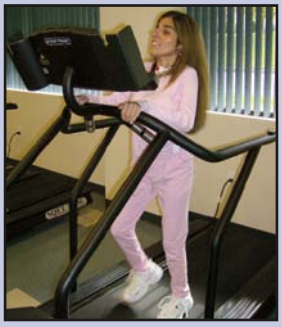
After the treadmill, she usually has a nutritional energy drink.

In addition to the leg press and leg curl machines, she does step-ups on a low aerobic-training step, while holding on to a bar. She hopes to advance to a stair-climbing machine.

The last stop is the breathing room, where she does group exercises to strengthen the muscles of the chest, back, neck and stomach.

In just two months at Oxygym, she's doubled the volume in her lungs, notices more muscle mass in her arms and finds walkways and ramps easier to tackle.

"Since I've been attending Oxygym, I feel physically and mentally better, and I'm happier when I'm working out."



Mary Gallo

Range-of-motion and light resistance exercise can be done in the water.

blood supply they need, the person is said to be in "heart failure."

The traditional view of exercise for people with heart failure has been that it should be undertaken with great caution, if at all. But at a meeting of the American Heart Association in November, 2008 investigators from Duke University and Thomas Jefferson University reported results of a long-term, large-scale study of heart failure patients assigned to an exercise or a usual-care group. The study found that exercise was safe for study participants with heart failure and lowered their risk of hospitalization or death.

The study included 2,331 patients from 82 study sites who were followed for an average of two-and-a-half years. Those in the exercise group started with three 30-minute sessions per week on a stationary bicycle or treadmill. After 18 sessions, they transitioned to 40 minutes five days a week. Those in the usual-care group continued their usual medical therapy and were encouraged to be active, but they were not given a specific exercise program.

It's imperative for anyone with a disease that affects the heart to consult a cardiologist before beginning an exercise program. Chances are, if your skeletal muscles are up to the task, some degree of exercise can be undertaken.

Then there's the critical issue of whether or not exercise can weaken and damage muscles already made fragile by a dystrophy. This is a particular concern in the dystrophies in which the muscle-fiber membrane is compromised. These are **Duchenne, Becker** and the **sarcoglycan-deficient forms of limb-girdle** MDs. In these diseases, a type of muscle contraction called *eccentric* can seriously damage already weakened muscle-fiber membranes and ulti-

mately destroy muscle tissue.

An *eccentric contraction* is what happens when a muscle is used in a braking action. When extending your arm slowly while holding a weight, your muscles act as a brake to prevent your arm from flopping down suddenly. Or when walking down a steep hill, your leg muscles use eccentric contractions to keep you from falling forward. This type of muscle activity is best avoided.

In **inflammatory muscle diseases** like **polymyositis** and **dermatomyositis**, active exercise during an acute flareup of the disease can exacerbate the inflammation and make things worse. Range-of-motion (ROM) exercise with someone assisting or doing all the work (passive ROM) is fine and may help keep joints flexible. Exercise should progress in duration and intensity as the disease comes under control with medication. When the disease is no longer active, recreational aerobic exercise, such as biking and swimming, and progressive resistance exercise, can be helpful in maintaining strength and cardiovascular health.

Weight machines can be adjusted to light resistance.

Recently, studies have shown that, in some muscle diseases where the fiber membrane is disrupted (such as **Duchenne** and **type 2D limb-girdle** muscular dystrophies), signals that normally tell blood vessels to open (dilate) during exercise don't occur. The signals aren't sent because a molecule called nitric oxide synthase, which starts the process, can't anchor itself to the membrane. The implications of this problem aren't yet clear. However, researchers believe this mechanism may explain some of the significant fatigue some patients feel with even mild exercise.

In **diseases of the motor neurons**, such as **spinal muscular atrophy (SMA)** and **amyotrophic lateral sclerosis (ALS)**, there's a theoretical concern that too much exercise can hasten the loss of these muscle-controlling nerve cells. These cells, the theory goes, not only are compromised by the disease, but they may be overburdened by trying to compensate for neighboring cells that have been lost.

Studies in mice with an **ALS**-like disease have shown that mice that ran regularly on a treadmill at a low speed survived longer than average, but mice that ran regularly on a treadmill at a high speed survived a shorter time than average. The researchers found the mice did best when exercise periods were interspersed with rest. However, they acknowledge that they don't know whether these findings

Exercise Brought Him Independence, Happiness

Eleven-year-old Jerry Huang is a happy camper these days, and his new exercise program is the reason.

Two years ago Jerry, who has congenital muscular dystrophy, had tendon surgery to relieve severe contractures in his legs. He also began a physical training regimen that wasn't much fun. "They had him doing things like throwing and catching balls," said his mother Tina. "It was very tiring for him, and after 10 minutes he was ready to quit."

Enter a new physical therapist at Jerry's grade school in Los Angeles. She started him on a schedule of swimming exercises and yoga, and got him both a stander and walker. He now exercises and stretches twice a week in PE class for an hour at a time.

His mom says the change in her son has been incredible. "When he's in the water, he can stand, he can walk, he can jump — all things he couldn't do before. It has been so good for him."

Jerry also doesn't have to wear his full-length braces during the day anymore, just at night. "Now I can get out of my wheelchair and stand, and I can walk about 30 feet. And my mom doesn't have to come with me to the bathroom anymore," he says.

The change has been as much emotional as physical. His mom says when Jerry was younger, he was quiet and withdrawn. Now, with his newfound physical independence, and with mentoring from an aide at school, he's become more outgoing and social, and he's an honor student.

Jerry Huang

Beth on the Go

Once a week, working mother Beth Bax of Altadena, Calif., works out at 24 Hour Fitness, where she's been a member since the early 1990s.

An environmental engineer, Bax, 39, received a diagnosis of Friedreich's ataxia in 1999.

Every other week she works with a personal trainer at the gym. "I originally signed up for sessions because I hadn't exercised for over two years, and I needed someone to show me what exercises to do and make me accountable for showing up," says Bax, whose neurologist supports her exercise routine.

She notes that for some people, a physical therapist might be a better alternative than a personal trainer.

"They're better accustomed to people with disabilities and knowing their limits," she says. "My trainer sometimes tries exercises with me that don't work out."

Bax's visits, which usually last 90 minutes, start with a 15-minute warm-up on the gym's stationary recumbent bike. Then it's over to arm, leg, and back exercises with the chest, leg and calf press machines and the lat pulldown. Depending on the machine, the weight is usually 40-50 pounds, though heavier for leg lifts. And then there's the 100 sit-ups.

It doesn't stop there. Bax rides her TerraTrike recumbent tricycle around her neighborhood for an hour twice a week. She even takes it along on family vacations so she can ride with her husband, Eric, and their two children, Natalie, 7, and Sarah, 4.

"I recommend a recumbent trike if you can sit in a seat and be stable," she says. "It's a great feeling to be going downhill super fast, and it's nice to do something on my own that I don't need help with.

"Exercise is proven to get you in a better mood, and I don't know anyone who can't use that in life."

Beth Bax

Water generally makes exercising easier and reduces stress on the joints.

apply to humans with **ALS** or other motor neuron diseases.

To add to the uncertainty, studies in mice with an **SMA**-like disease found regular running resulted in better motor function, a longer life span, and more surviving motor neurons compared to mice that didn't participate in the training. But again, extrapolation to humans is risky.

Perhaps the most dramatic instances of damage resulting from certain types of exercise are those that can occur in the **metabolic muscle diseases**, such as **Pompe disease** (acid maltase deficiency), **McArdle disease** (myophosphorylase deficiency), **carnitine palmityl transferase (CPT) deficiency**, and others in this category, including diseases involving the mitochondria, where much of a cell's energy is produced.

In these disorders, a biochemical defect is almost invariably associated with exercise intolerance, and normal exercise is impossible because of extreme fatigue, muscle pain and/or cramps.

If a patient pushes beyond the limits of fatigue or pain, acute muscle breakdown can occur, spilling muscle proteins like myoglobin into the bloodstream and potentially damaging the kidneys.

"Trainers like to say 'no gain without pain,' says Alfred Slonim, a pediatric endocrinologist at Columbia University, "but in **metabolic muscle disease** it's 'no gain with pain.'"

On the other hand, Slonim says, "When you turn a person into a couch potato or a wheelchair-dependent patient, their muscle status deteriorates from disuse."

He recommends light aerobic conditioning exercise, such as walking on a treadmill, or using a stationary bicycle, under the watchful eye of a physician or knowledgeable physical therapist. Light resistance training, using weights of no more than 5 or 10 pounds, is OK for some, he says, after they've done some aerobic exercise.

At the UC Davis center, respiratory and other functions are monitored during treadmill and other forms of exercise. Photo: Emi Manning, UC Davis

Q: Does proper exercise reduce pain?

A: "Proper" exercise sometimes can reduce certain kinds of pain. For instance, if pain is due to stiffness in the joints or prolonged sitting or lying in one position, gentle stretching or range-of-motion exercises can relieve it. However, severe muscle cramps or pain, either immediately or a day or two later, are an indication that the exercise may be doing some damage, and it should be either stopped or reduced in duration or intensity.

Q: Is there one kind of exercise that's good for anyone with a neuromuscular disease?

A: If there's any kind of exercise that comes close to being universally good for people with neuromuscular disease, it's probably moving in the water. The buoyancy of water makes exercise easier and reduces stress on the joints. However, actual swimming, especially for those with cardiac abnormalities, can be risky, and should be approved by a physician.

Not all exercises require fancy equipment.

Also, cold water can make **myotonia** (inability to relax muscles) much worse, especially in the condition known as **paramyotonia congenital (PMC)**. "If swimming, particularly in cold water, PMC patients are at increased risk of an attack [of myotonia], and in the water, this can be life-threatening," says MDA grantee Louis Ptacek, a neurologist and muscle physiologist at the University of California San Francisco. "No one should swim alone, but particularly not PMC patients."

Q: Will exercise cause me to lose weight?

A: Sometimes — or it may at least offset some of the weight gain that usually accompanies a sedentary lifestyle. Aerobic exercise is what's usually recommended for weight loss or maintenance, since it directly burns calories. However, recent research has suggested that resistance exercise, which builds muscle (at least in some people), also can be beneficial. Muscle is where most fuel is metabolized for energy, so if there is more muscle, theoretically, there is more fuel burned instead of stored as fat.

A program at UC Davis in which people with **slowly progressive muscular dystrophies** were given pedometers and asked to increase their walking time by 25 percent resulted in

Food Influences Performance, Says Multi-Athlete

Most athletes devote themselves to one sport. But Ryan Levinson of San Diego isn't most athletes.

Levinson, who has facioscapulohumeral muscular dystrophy (FSHD) and has weakness in his chest and leg muscles, excels personally and professionally at cycling, surfing, sailing, paddleboarding, kiteboarding, scuba diving, kayaking and the triathlon.

He builds up strength and skill in the pool, ocean, bay, beach, gym, and on the road. Aside from practicing his sports regularly, Levinson focuses on stretching all muscle groups, core strength and balance training, functional strength training with a combination of exercise physio balls, dumbbells, ropes hooked to cable weights, aerobics and swimming.

Levinson no longer runs because weakness in the muscles that give his knees and hips stability puts him at risk for injury, but his exercise program is designed to optimize his strength and aerobic fitness for his target events.

Working with a registered dietician, he found that a healthy diet has had a positive impact on his health and athletic performance.

His diet mainly consists of lean protein, vegetables, whole grains and raw nuts. He avoids processed foods and concentrated simple sugars, completely eliminating trans fat. Water is his beverage of choice during the day, and before going to bed and after waking up, he drinks a glass of whey protein, which is a mixture of whey concentrate, whey isolate and amino acids.

Levinson says he has found that good food helps his body compensate for some exercise stress. "Without good food, your athletic performance can actually decrease because your body's ability to recover will be impacted."

Ryan Levinson

Taking a Martial Arts Approach

For six years, teenager Dani Anderson of Newbury Park, Calif., trained regularly in the martial arts discipline of tae kwon do. Her dedication eventually earned her a coveted black belt rating.

An integral part of reaching that level of accomplishment was exercise, day in and day out.

Anderson, now 22, is even more a standout in her moves because she has Charcot-Marie-Tooth disease and uses a wheelchair for mobility.

"My tae kwon do school created a special curriculum for me," she explains. "We came upon ways for me to do kicks, punches, blocks, etc., from my wheelchair, equivalent to those the rest of the class was doing. When the rest of the class was doing push-ups, I would repetitively lift a weighted stick."

Anderson is no longer a full-time martial artist because she's attending school and working full time, but she still exercises regularly.

"I exercise at home by doing movements similar to those I did in tae kwon do. My physical therapist supports the exercise program completely, but she also encourages me to swim regularly," she says. "Now I swim at the gym. I do laps as well as walk in the pool. Along with tae kwon do movements, I expect these exercises to maintain the muscle mass I already have, and build strength."

Dani Anderson

When using free weights, be careful not to exceed 5-10 pounds.

weight loss, "feeling better," and scoring better on quality-of-life measures.

Q: How hard should a person with neuromuscular disease expect to work at aerobic exercise?

A: This, of course, depends on the disease in question and its severity, the age and condition of the person, and their cardiac and respiratory status.

But as a general rule, physicians recommend that patients with neuromuscular diseases do *submaximal* aerobic exercise, which means activity that maintains the heart rate at about 65 percent of its maximum.

The maximum heart rate can be calculated by subtracting one's age from 220. Multiply that number by 0.65 to get 65 percent. So, for example, the submaximal heart rate for a person who is 30 years old is: 220 - 30 = 190; 190 x 0.65 = approximately 123 beats per minute.

Another way of assessing aerobic exercise is to see whether you can talk while doing it. If you can't talk, says Ted Abresch of the UC Davis Center, you're doing something too fast or for too long.

Submaximal aerobic exercises include pool therapy, walking or bicycling on a flat surface.

Q: How hard should a person with neuromuscular disease expect to work at resistance exercise?

A: As above, it depends on disease type and severity and the individual patient's condition. In general, one should consider only light resistance exercise. Lifting more than 5 or 10 pounds or resisting more than 5 to 10 pounds (for instance, using a stretchable band or a moveable device you push against) is likely to cause muscle-fiber destruction. In most neuromuscular diseases, muscle fibers are difficult or impossible to regenerate.

Abresch says studies at UC Davis found high-resistance exercise wasn't any more beneficial than moderate-resistance exercise in patients with **slowly progressive muscular dystrophies**.

Q: How do rest and recovery fit into an exercise strategy?

A: At UC Davis, they use a "one-rep max" strategy, says director Ted Abresch. "That means they lift a weight, then we give them 15 minutes of rest, and then they get a weight that's a little heavier." Patients only go on to the next weight if the previous weight was easy to lift. They're told to stop when the weight

is no longer easily lifted.

Resting at intervals so that heart rate doesn't go above 65 percent of maximum is a general recommendation for aerobic exercise.

There's an exception to most rules, however. In **periodic paralysis**, it's during a rest following exercise that patients are most likely to experience an attack of paralysis.

In **hyperkalemic periodic paralysis**, attacks can come on within seconds or minutes or even up to an hour after ceasing vigorous exercise, says neurologist Louis Ptacek. For instance, sitting in a chair after doing housework can make someone too weak to get up from the chair 30 minutes later.

In **hypokalemic periodic paralysis**, the attacks are more likely to come on the morning following the exercise.

People can sometimes ward off an attack of paralysis by continuing their activity and slowing down very gradually or by "walking off" the weakness early in an attack.

Q: What about people who can't exercise in the usual sense? Can a daily power chair roll through the mall or the adrenaline rush of video games contribute to fitness?

A: It's not clear that this type of exercise can improve fitness. However, it may help restore energy and reduce fatigue, as well as reduce depression and, if done in a group, offset social isolation.

Passive range-of-motion exercise means another person manipulates the patient's joints.

Q: In the absence of clear answers, what should a person who wants to exercise do?

A: In most cases, people can do moderate-intensity aerobic training. Some can do moderate-intensity resistance training in muscles that can still work against gravity (for instance, muscles that can extend the leg from a seated position with the knee bent).

People always should check with their neuromuscular disease specialist and, if they have a disease that involves the heart, with a cardiologist, before beginning an exercise program.

People also need to know how to recognize the warning signs that tell them they've gone too far. See "What to watch out for," page 2. ❑

No Resistance to Resistance Training

Nick Johnson, Waltham, Mass., has been a big proponent of working out since he was a teenager, including throughout the 26 years since his diagnosis of Friedreich's ataxia (FA).

Now 45, Johnson, a mechanical engineer, still is working out, but has cut back on the types and frequency of his routines. "There's no question that FA has taken its toll on my muscles," he says. "But I still find value in exercising."

These days he mainly uses machines like Life Fitness that utilize resistance training.

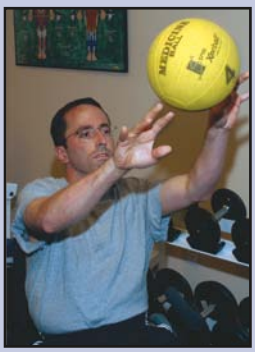
Johnson focuses on eight types of resistance training that exercise different muscle groups in his arms and legs. He goes for eight repetitions per type. Exercising once or twice a week, he relies on an attendant for help when at his health club, and his wife's assistance when exercising at home.

In addition to resistance training, Johnson spends 30 minutes on a handcycle, and occasionally tosses and catches a medicine ball.

"Exercise helps me in two ways," he says. "First, I feel it slows the rate of progression of my disease. Second, it helps me with my mind-to-body connection. It helps me maintain that, and it can help reduce the physical pain in my body."

Both his physician and physical therapist approve of his exercise regime, as long as he does it in moderation and with assistance.

"My advice to anyone with a disability contemplating exercise is 'just do it!' Be responsible, but try to keep pushing your comfort zone. Try not to get frustrated if your body isn't doing everything you wish it would."



Nick Johnson

Effect of Exercise on Different Muscle Diseases

Disease category	Nerve or muscle problem	Implications
Muscular Dystrophies Duchenne, Becker, congenital, distal, Emery-Dreifuss, facioscapulohumeral, limb-girdle, myotonic, oculopharyngeal	These are degenerative muscle diseases, meaning the normal ability of muscle to regenerate and repair damage is limited, resulting in a net loss of muscle tissue over time. Some (BMD, DMD and some LGMD forms) involve fragile membranes around skeletal and/or cardiac muscle fibers. Many muscular dystrophies (DMD, BMD, some LGMD, EDMD, myotonic MD) involve cardiac muscle deterioration or abnormal heart rhythms.	• Muscle fibers can be damaged by strenuous exercise, and the damaged sustained may be permanent. • In DMD, BMD and the sarcoglycan-deficient LGMDs, exercise involving eccentric (lengthening) contractions is particularly damaging. • If the heart's pumping ability or rhythm is affected by the disease, sudden, strenuous exercise could trigger an acute heart problem, respiratory problem or even death. A cardiologist should be consulted before undertaking an exercise program. Warning signs of an acute cardiac problem are chest pain, shortness of breath, nausea, sweating without strenuous exercise, a feeling of fullness or pressure in the chest, or a gurgling sound durinig breathing (a sign that fluid may be backing up into the lungs).
Motor Neuron Diseases amyotrophic lateral sclerosis, spinal muscular atrophy (all types), spinal-bulbar muscular atrophy	Nerve cells in the central nervous system (spinal cord and brain) that control muscle movement are lost, leaving muscles "orphaned" and undernourished.	• Theoretically, remaining nerve cells can become overburdened if too much is required of them with exercise.
Inflammatory Myopathies dermatomyositis, polymyositis	Muscle fibers are attacked by the immune system, causing inflammation and tissue destruction.	• Exercise should vary with disease activity. During severe disease activity, when strength is very poor, assisted or passive range-of-motion exercise can be undertaken. During periods of mild to moderate disease activity, when muscles can work against gravity, light aerobic exercise, isometric strengthening, and active range-of-motion exercise are recommended. When the disease is inactive (in remission), recreational aerobic and progressive resistance exercise are OK.
Diseases of the Neuromuscular Junction myasthenia gravis	Communication between nerve fibers and muscle fibers is compromised, in most cases by a mistaken attack on the neuromuscular junction by the immune system.	• Exercise can be undertaken when the disease is under good control. In periods of acute disease exacerbation, or when the disease is not well controlled, exercise may increase weakness. • Exhaustion, lasting joint or muscle pain, or shortness of breath means the exercise is too strenuous. • If on prednisone, mild exercise can help offset side effects, such as weight gain, bone loss, high blood pressure, high blood sugar, depression and anxiety.
Peripheral Nerve Diseases Charcot-Marie-Tooth disease, Dejerine-Sottas disease,	In CMT, and DSS, nerve fibers and/or the insulation around them are abnormal, compromising communication to muscle fibers.	• Exercise is good for the heart and lungs but will not overcome muscle wasting, which is the result of damage to nerve fibers and loss of input from the nervous system. • Low-impact exercise, such as swimming or biking, or yoga, is better than high-impact exercise
Friedreich's ataxia	In FA, an abnormal cellular distribution of iron leads to diminished cellular energy production, with damage to the heart and nerves	• Moderate exercise may help slow the loss of motor skills in FA and help offset weight gain. • Cardiac precautions apply. No exercise should be undertaken without consulting a cardiologist.

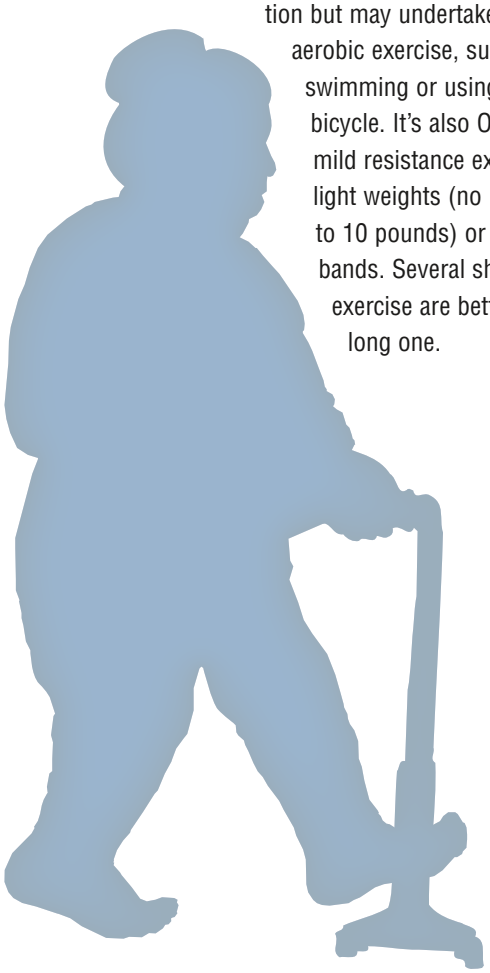
Metabolic Muscle Diseases acid maltase deficiency, carnitine deficiency, carnitine palmityl transferase deficiency, debrancher enzyme deficiency, lactate dehydrogenase deficiency, mitochondrial myopathy, myoadenylate deaminase deficiency, phosphofructokinase deficiency, phosphoglycerate kinase deficiency, phosphoglycerate mutase deficiency, phosphorylase deficiency	Processing of fuel (carbohydrates or fats) in muscle fibers is inadequate, leading to subnormal energy production and/or a toxic buildup of fuel.	• Muscle fibers may break down in an attempt to meet the energy requirements of exercise. • Do not engage in aerobic exercise that causes the heart rate to exceed 65 percent of maximum (220 minus age, multiplied by 0.65). • Light resistance exercise, such as lifting no more than 5 to 10 pounds, can help build muscle mass. • Warning signs to stop are pain, muscle tightness or cramps, exhaustion or cola-colored urine. Seek medical help if urine turns dark. • If the heart is involved, cardiac precautions apply.
Ion Channel Diseases paramyotonia congenita, periodic paralysis (hypokalemic & hyperkalemic)	Channels in muscle fibers that allow the passage of sodium or calcium are abnormal.	• People with periodic paralysis benefit from good fitness. However, vigorous exercise followed by rest can precipitate a bout of weakness or paralysis. (In hyperkalemic PP, attacks generally occur within an hour of exercising. In hypokalemic PP, they're more likely to occur the morning after exercise.) • Low-intensity exercise is best. Some people find they can ward off weakness by cooling down slowly. • In paramyotonia congenita, muscle cooling can increase myotonia (inability to relax muscles) and transition into weakness and paralysis. • If swimming, particularly in cold water, paramyotonia congenita patients are at increased risk of an attack and of drowning.
Centronuclear Myopathies myotubular myopathy, autosomal centronuclear myopathies	Muscle fibers show centrally placed nuclei, in contrast to their normal position, which is around the periphery (edge) of each fiber. Mutations in genes for the muscle proteins myotubularin, dynamin 2, MYF6, amphiphysin 2, and probably others result in weakness, the precise cause of which remains uncertain. Outcomes range from severe weakness affecting all muscles, including those involved in breathing, to weakness so mild it leads only to poor athletic performance.	Because these conditions are not progressive and don't have specific defects in the muscle fiber membrane that lead to its fragility, exercise is likely safe if the myopathy is not too severe. Check wih your doctor before beginning an exercise regimen. There are no data on exercise for children with severe centronuclear myopathies, such as the X-linked, myotubularin-related type.

What Kind of Exercise Can Be Done By …

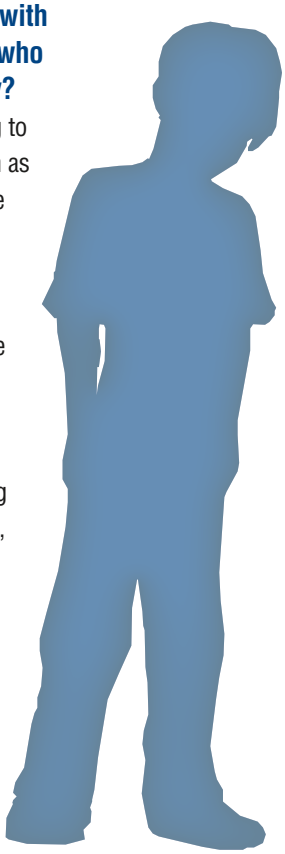
… a middle-aged, overweight person with myasthenia gravis who is able to walk?

Unfortunately, even when MG is under control, as it usually is these days thanks to effective medications, lack of endurance when exercising and excess weight gain as a side effect of medication and inactivity remain problematic.

People should not exercise to exhaustion but may undertake submaximal aerobic exercise, such as walking, swimming or using a stationary bicycle. It's also OK to try some mild resistance exercise, using light weights (no more than 5 to 10 pounds) or stretchable bands. Several short periods of exercise are better than one long one.



… an elementary-school child with Duchenne muscular dystrophy who uses a wheelchair occasionally?



The child with DMD who is beginning to need a wheelchair for distances, such as at the mall, is beginning to experience the effects of deteriorating muscle fibers. The fibers are degenerating because they lack dystrophin, which helps keep them intact, and so they're excessively fragile.

Doctors disagree about exercise in DMD, but most don't feel a child should become a couch potato. Doing no exercise at all leads to weight gain, depression and stiff joints, and may even contribute to muscle atrophy.

Children with DMD should avoid exercise that involves "eccentric" contractions. This type of exercise can severely damage muscle fibers that may not be able to regenerate. Eccentric muscle contractions occur when muscles are used as brakes, such as when walking down a steep hill or slowly lowering a weight. Weight lifting is an example of an activity that requires a lot of eccentric contractions.

After being cleared by a cardiologist, a child with DMD may undertake submaximal aerobic exercise, such as walking or swimming, while keeping an eye out for warning signs of overexertion. (See "What to watch out for," page 2.) Children with DMD always can do passive or active range-of-motion exercise, and physical therapists often recommend stretching of the heel cords to prevent stiffening of the ankle joints.

… a child with type 2 spinal muscular atrophy who uses a power chair full time?

The child with SMA is in a somewhat different situation from a child with DMD, even though they both may use wheelchairs.

In SMA, which is primarily a disease of the muscle-controlling nerve cells (motor neurons) in the spinal cord, there isn't as much concern about direct damage to muscles as there is in DMD. If the child is comfortable doing a particular exercise, the exercise is probably all right.

There's a theoretical concern that exercising too hard or too long with SMA might cause stressed motor neurons to wear out faster than they might otherwise, but that hasn't been proven.

In general, submaximal aerobic exercise, light resistance exercise and range-of-motion exercise are fine.

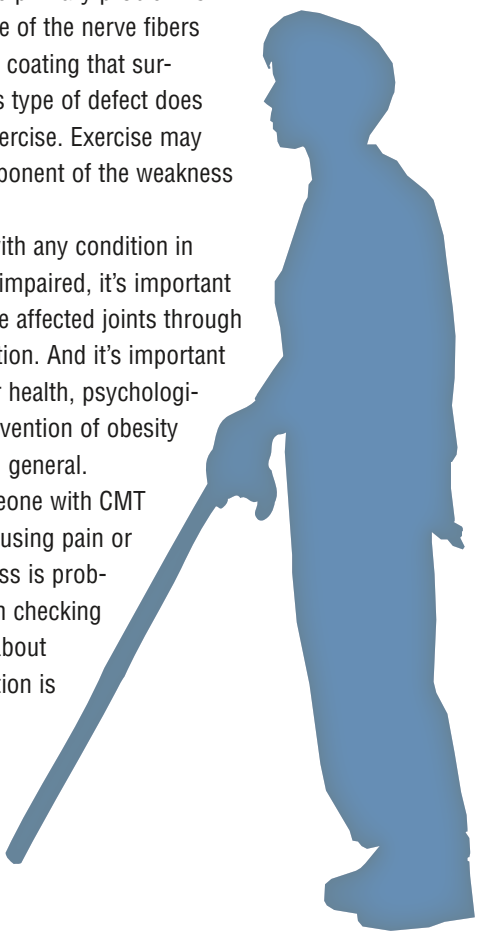
… a 25-year-old with Charcot-Marie-Tooth who can walk with the use of ankle-foot orthoses?

CMT is a disease of the peripheral nerves, which are made up of bundles of nerve fibers that connect the spinal cord with the muscles. It causes loss of motor function, particularly in the forearms and hands and in the lower legs and feet, and often some loss of sensation, especially in those areas.

Exercise may not change strength, dexterity or sensation in the affected areas, because the primary problem is faulty maintenance of the nerve fibers themselves or the coating that surrounds them. This type of defect does not respond to exercise. Exercise may help if some component of the weakness is due to disuse.

However, as with any condition in which mobility is impaired, it's important to keep putting the affected joints through their range of motion. And it's important for cardiovascular health, psychological health and prevention of obesity to keep moving in general.

Anything someone with CMT can do without causing pain or increased weakness is probably fine, although checking with a physician about any specific situation is always advisable.



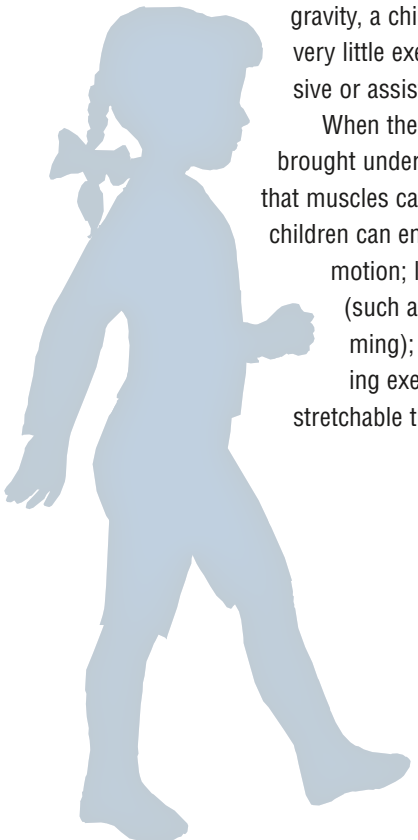
… a 10-year-old child with dermatomyositis whose disease has been well controlled by medication for several months, but who still tires easily with any exertion?

Children with DM (and their parents) face a complicated set of problems. In this disease, the person's immune system mistakenly sets up a severe inflammatory reaction that affects the blood vessels, muscle fibers, joints, skin and sometimes other organs. Chronic fever is not uncommon, and some patients feel lethargic much of the time.

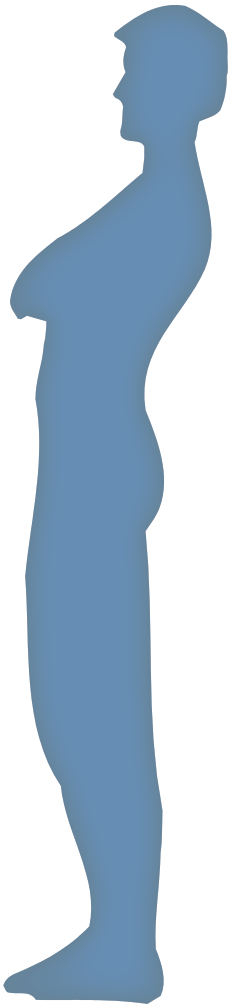
The goal should be to preserve as much normal growth and development as possible (which requires exercise), stave off excess weight gain from immobility and drug side effects, help joints stay flexible and not exacerbate the inflammatory process (which over-exercising can do).

During severe disease activity, when muscles are too weak to work against gravity, a child with DM should do very little exercise other than passive or assisted range of motion.

When the disease has been brought under control to the extent that muscles can work against gravity, children can engage in active range of motion; light aerobic exercise, (such as walking or swimming); and mild strengthening exercise, such as using a stretchable therapeutic band.



… a 20-year-old male college student with McArdle disease?



McArdle disease, which is caused by the lack of an enzyme that normally breaks down glycogen (animal starch) to generate energy in muscle cells, poses some unique challenges with respect to exercise.

In McArdle disease, unlike most other neuromuscular disorders, what one *can* do and what one *should* do are very different things. Moderate exercise, such as brisk walking, may be achievable but may cause severe muscle injury even if the pain or cramping it causes are tolerable. Weight lifting or picking up a heavy object of any kind can cause more injury than the initial pain might indicate.

In this condition, even mild cramping or burning or aching pain in the muscles can indicate muscle breakdown.

It's tempting for a young man who may look fit and strong to engage in strenuous workouts or sports, but if that young man has McArdle disease, he should never push past pain.

After exercising moderately for about 10 minutes, the typical McArdle patient is able to tolerate an aerobic activity much more easily. This "second wind" is believed to occur because fuel for energy production in the form of glucose (sugar) has arrived at the muscles through the bloodstream.

The "second wind" phenomenon has led to the idea of boosting energy by ingesting sugar right before exercising, such as a soda or a candy bar. Research confirms that this approach does help with exercise tolerance and injury prevention. However, it also can cause serious weight gain, so its use is limited.

Exercise generally should be limited to submaximal aerobic workouts, as long as they're comfortable, and perhaps light resistance exercise, lifting no more than 5 to 10 pounds and stopping when pain begins. ❑

Caregiver Exercises

Simple ways to resist injury

by Christina Medvescek

It just takes one little wrong move, like leaning over to tie a loved one's shoelace, for something to pop in the lower back and put you out of commission. For a caregiver, this kind of injury can be disastrous.

"Loads of people have had to give up caregiving due to injury," says Brenda Shaeffer, [formerly] a physical therapist at the MDA/ALS Center at Johns Hopkins University in Baltimore. "Then they have to find someone else to provide care, or in some cases find a nursing home."

Good body mechanics (like lifting with the legs, not the back) and assists (like a Hoyer lift or another person) significantly reduce the risk of injury and always should be used. But unless you're also taking care of your body, an injury may be just one wrong move away.

Muscles need flexibility and strength to resist injury — and stretching and strengthening exercises are the way to achieve those goals.

We're not talking about an hour-long trip to the gym, but exercises that fit neatly into odd moments of the day, like when you're watching TV or sitting at a traffic light.

And the beauty is, "whatever you can do will help," says physical therapist Jean Hill, [formerly] with the Kessenich Family MDA/ALS Center at the University of Miami. "A little here and there adds up."

Bad habits

Most adults have bad body habits, like poor posture or only moving in a few routine patterns.

The result of these habits is that muscles only stretch within a limited range of motion that works OK for everyday life, but doesn't stand up to stress.

"Injuries occur when you put your body into positions it's not used to," explains Lee Burkett, emeritus professor in the Department of Exercise and Wellness at Arizona State University in Tempe. "When your muscles are supple, you can put your body into more positions than you could before."

Beyond flexibility, leg and hip strength is vitally important for lifting, and strong abdominal and buttocks muscles stabilize the lower back region.

Opportune moments

There are dozens of opportunities during the day to fit in quick, gentle exercise.

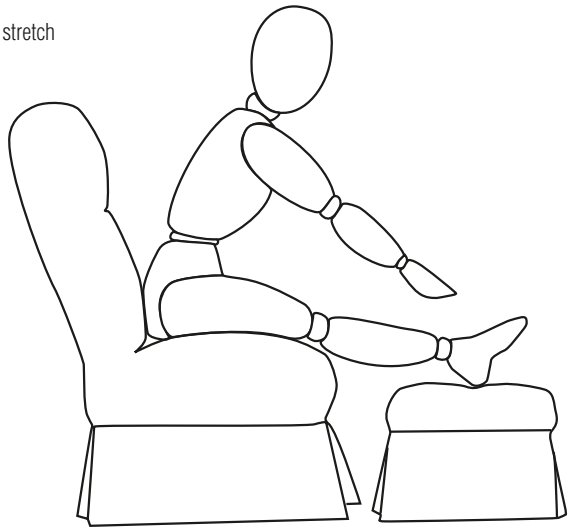
You can perform the exercises listed below basically anytime you have a spare 30 seconds. If you pair an activity with an exercise — for example, always doing partial squats while waiting for your toast to pop or hamstring stretches during TV commercials — you'll improve without even realizing it.

If you already have painful back, neck or shoulder muscles, check with your doctor before undertaking any of the exercises below.

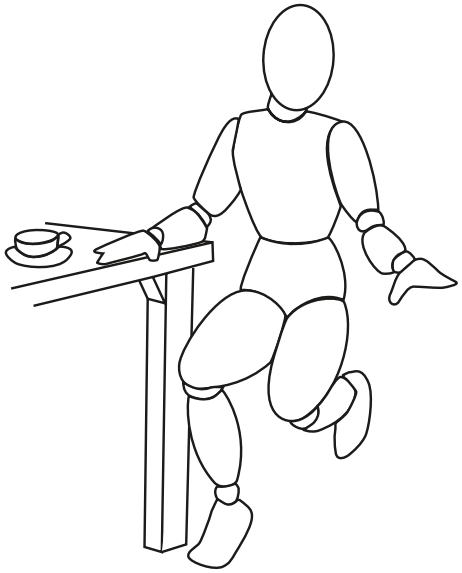
Stretch like a cat

Stretch to the edge of comfort but not beyond, or you may trigger a stretch reflex that actually contracts the muscle

Lower back stretch



Partial squat



and defeats your efforts. Pain isn't the point, nor is distance. Luxuriate in a stretch, the way a cat does.

Hold stretches about 15 to 30 seconds, repeating two or three times. Never bounce to get a greater stretch.

Lower back stretch: Tight hamstring muscles can cause back pain, says John Klune, physical therapist at the MDA/ALS Center at the University of Arizona Health Sciences Center in Tucson.

Stretch your hamstrings by sitting in a chair with your feet up on a footrest or coffee table. Keeping your knees straight, lean forward until you feel an easy, not painful, stretch.

Shoulder stretch: Scratch your back. First point your elbow in the air and scratch the top of your spine. Next, curl each arm around the front of your neck and scratch over your opposite shoulder. Finally, hold your elbow down by your side and twist your hand back up between your shoulder blades.

Chest stretch: Place your hands at shoulder height on either side of a doorjamb, then step into the doorway so your arms slightly wing out behind you. Feel the stretch through the middle of your chest.

Neck stretch: Lower your ear to your shoulder on either side.

Get strong

The rule of thumb for strengthening without weights is to tighten your muscles to their maximum, hold for a count of 10, relax for a count of 10 and do 10 repetitions. Be sure you're stable, either by taking a wide stance or by holding on to a secure object.

Legs and hips: Whenever possible, climb stairs instead of using an elevator. In addition, try partial squats. Hold on to a stable object like a countertop. Keep your back straight and slowly lower as if you're about to sit down. Go down as far as comfortable, then back up.

For greater effect, use only one leg at time. You also can work on leg strength each time you get up from a desk or table, by rising from the chair using only one leg. Hold on to the table for stability but don't pull yourself up. Alternate legs for an equal workout.

Lower back: Here's an exercise nobody will even know you're doing. Whenever you think of it, squeeze your buttocks or your abdominal muscles as tight as you can and hold. "You can't overdo it," says Klune.

A San Diego State University study of abdominal exercises found that they should be done frequently rather than for intense periods. Five minutes a day can make a difference if done regularly.

Shoulders and arms: Take a weight like a shoe or soup can and lift it straight up in front and to the side, from your waist all the way over your head.

Hold arms straight out to the sides at shoulder height, then bring one arm across in front of your chest, keeping the elbow straight. Go just to the point where your arm starts to bend, then back.

Force your shoulder blades together in the back, then relax, then push them together again. Do shoulder shrugs that bring your shoulders up toward your ears, as if you're saying, "I don't know."

Neck: Place your palm in the middle of your forehead and gently press, resisting the push by holding your head still. Repeat on each side and the back of your head.

These are just a few of the many exercises that will help keep your muscles supple and strong. For more information, Betsy Curless, [former] physical therapist at the MDA clinic at the Via Christi Medical Center in Wichita, Kan., recommends these books: ***Treat Your Own Back*** and ***Treat Your Own Neck***, both by Robin McKenzie (Orthopedic Physical Therapy Products). ❑

This article originally ran in the MDA/ALS Newsletter, Oct. 2003. Updates have been added for experts who were quoted.

"We can have positive influences on our bodies"

"It's frustrating I had to give it up"

"Any exercise will improve the psyche"

How Do You Feel About Exercise?

"There's no adrenalin rush"

"It's a huge pick-me-up"

"I'd rather be doing other things"

"I must exercise regularly to keep my strength"

As muscle weakness progresses, people give different answers

by Bethany Broadwell

In this age of obsession about body image, when someone asks me if I want a piece of strawberry pie for dessert, I'm apt to decline. "No thanks. I need to maintain my girlish figure."

My playful reply is designed to conceal the fact that I'm actually assessing my ability, as a 40-pound skinny mini with spinal muscular atrophy type 2 (SMA2), to eat the pie.

Do I have the strength to cut the berries and the crust? Will I have the oomph it takes to raise my fork to my mouth? Will I have the musculature to chew and swallow the dessert? Simply feeding myself and eating has become an exercise workout for me.

It is an awkward predicament to be in when everyone around me seems to be consumed with tight abs, low-carb diets and intense activities like spinning or step aerobics. I've never really had the strength or stamina for any significant exercise. Instead, I try to preserve as much of my strength as possible, call on assistance when necessary and realize that plenty of others with neuromuscular disease are dealing with similar challenges regarding loss.

People's degree of mobility and their past experiences significantly impact how they feel about exercise. Some choose not to work out. Others remain motivated and find ways to modify their routines to accommodate their weakness.

The following perspectives about exercise from people with neuromuscular disease may help you decide the extent of workout activity that fits your lifestyle.

Time to stop


Sue Charlip

Sue Charlip, 48, of St. Petersburg, Fla., received a diagnosis of myasthenia gravis (MG) in January 1997. She describes how the progression of the disease has gradually decreased her motivation for exercise, because continual

exertion causes her extreme physical weakness and fatigue.

"I was a big walker. I used to walk in the neighborhood, walk the mall," Charlip recalls. "I just walked and never gave it much thought until MG hit and walking became difficult."

She says she enjoyed the exercise because it was easy. It didn't take any special talent or equipment. Plus, walking didn't aggravate her asthma.

Repetitive motion exhausts her now, so Charlip uses a motorized scooter when she needs to cover long distances or avoid standing for long periods of time.

"I just try to accept my limitations," she concludes. "I was sick for two years, at least, before I got diagnosed; so in one way, I'm glad to know what is wrong with me and know how to take care of myself. It's just frustrating, depressing and sad that I had to give up the little bit of physical activity I was able to do and enjoyed."

Tina Baughman, 49, of Pacific Palisades, Calif., says living with adult-onset limb-girdle muscular dystrophy (LGMD) has completely taken away her desire to exercise. The reasons are multiple: "I can't stand up off the floor. I don't see any positive results. I would prefer to spend my time doing other things."


Tina Baughman

In her younger years, Baughman found it rewarding and fun pumping weights, developing her muscle strength, playing sports and working against her own personal best. Her experience is different now. "There's no adrenalin rush. It's like asking a competitive downhill snow skier to be happy spending time walking through the snow … for me, that's not going to happen."

With assistance, Baughman has successfully managed such adventure activities as tandem skydiving, High Sierra backpacking on horseback, indoor rock climbing and flying a single-engine aircraft. These opportunities help her hold onto the thrill of moving, and make it a little easier to listen when others share their excitement and interest about fitness.

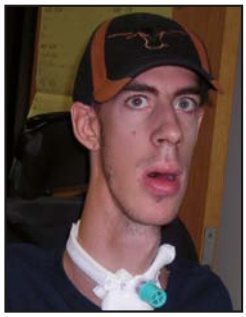
Even so, she admits, "There is a tinge of jealousy, longing and sense of loss when I hear about people doing things that are beyond my physical abilities."

Keeping going

Greg Halamicek, 59, of Lancaster, Calif., knows he needs to keep exercising to maintain his weight and body function, as his type 1 Charcot-Marie-Tooth disease progresses. He has modified his workouts and made swimming the primary focus of his routine, using an indoor pool near his home three or four times a week.

The limited extent to which he is able to exercise, Halamicek says, gives him a "huge pick-me-up" both emotionally and physically.

Joey Wells, 21, of Shadow Hills, Calif., became motivated to exercise two years ago after a hospital stay depleted his strength. Wells has myotubular myopathy and uses a power wheelchair. His workout routine includes arm cycling, stretching, stepping, kicking and weight training.


Joey Wells

Wells explains, "When I was younger, I had more strength to do what I needed. So, I could get away without exercising. Now I need to exercise regularly to keep what strength I have."

Nikki Young, 34, of Clarksville, Md., finds fitness and healthy living empowering as she struggles to deal with facioscapulohumeral muscular dystrophy. Through exercise, diet and physical therapy, she's lost more than 56 pounds and regained some independence. She advises, "I know that even with a neuromuscular disease, we can have positive influences on our bodies and keep them stronger."

Tyler Roope, 11, of Burbank, Calif., received a diagnosis of SMA2 at 18 months old. Although he doesn't always like to take time for stretching exercises, physical education is his favorite class in school.

Says Tyler, "I like baseball best. I want to manage the Cincinnati Reds someday. When we play, my friends hit for me and I run the bases in my chair. I like to play kickball at school in P.E. I pick someone to kick for me and I run the bases. When I am in the field, if the ball hits me, it's an out."

Of mind and muscles

For some who are very competitive or who deeply feel the psychological insult of neuromuscular disease, losing the ability to exercise can be hard to bear, says Carl Tishler, a clinical psychologist and professor at Ohio State University.

But, he says, there are other ways of "staying in shape" when moving the body is no longer feasible.

Exercising the mind can "strengthen the psyche and thus help the individual feel at ease," explains Tishler. As possibilities, he suggests learning a new language, solving crossword puzzles, playing computer games, following the stock market, reading or volunteering.

For those who have some degree of remaining mobility, Tishler urges: Make the most of the capability you do have. It might not help your muscles, but it might help your mind.

"Any exercise — no matter how brief or with little stress (weight) — will improve the psyche of the individual with MD." ❑

Bethany Broadwell is a freelance writer based in Traverse City, Mich. Her strength is wrestling with words.

Exercising Will Over Despair

by Michael P. Murphy

As a kid, I couldn't play — much less had any interest in — sports, so I wrongly assumed that exercise just wasn't necessary. I was a brainiac and a science fiction nerd, so I figured that exercise was just for the jocks, right? The only physical exercise I ever did on a regular basis were what I called Pumps.

To this day, we've never figured out why I had so many stomach ailments during my childhood. Once a month, almost like clockwork, I would be down for two or three days with nausea, vomiting, chills and sensitivity to light. Even in the midst of all this misery, I had a strange need for movement. I feared, I sensed, I was convinced that if I just lay still, my body would shut down. My mom, God bless her, would hold my ankles and pump my legs back and forth, 24 hours a day, sometimes in her sleep, until I felt better. I was only a kid, but I knew doing these Pumps saved my life, once a month.

This was my first inkling that some form of physical activity was important to good health. I've continued doing Pumps all my life, even though my stomach illnesses have long since subsided. I have my home health assistants pump each leg for five minutes (about 300 reps each) every morning before I go to work. Pumps help with circulation and also keep some limberness in my contracted limbs. These stretches eventually expanded to range-of-motion exercises with my arms and have now become such a normal part of my daily routine that if I should miss a day or two, I feel as if I've aged a hundred years.

In 2006, two things happened that further proved the need to keep physically fit. One: I turned 40, and my back, neck, and shoulders started telling me, "Hey, you don't have the stamina of a 20 year-old anymore. Let's get some stretches going here!"

Two: I was diagnosed with type 1 diabetes. I had suffered an ungodly summer: three months of weight loss (40 pounds), constant hunger, constant thirst, constant peeing, impotence and an out-of-control temper. Then, once a blood test revealed a blood sugar level of 540, I faced another few months of too much insulin, resulting in dangerously *low* blood sugars.

The diagnosis of diabetes knocked me on my butt, in every sense. I felt so lousy that all I wanted to do was stay in bed where I was most comfortable, where I could work, read, listen to music, watch movies and surf the Internet. The idea of just staying put was seductive, but I never let it stop my Pumps.

Once my metabolism settled down and I

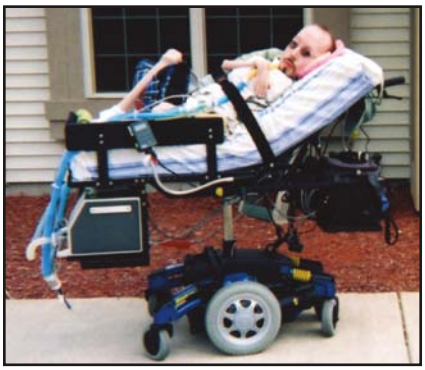
could feel some strength returning, I forced myself to get into my wheelchair and stay up a couple hours longer than I wanted to. Every day, I made myself keep up with my e-mail correspondence, my novel research, the screenplay I was writing — building the time a little bit each afternoon — because I feared turning into a decadent blob, like a skinny Jabba the Hutt. It wasn't easy. Sometimes, all that kept me from staying in bed was the fact that our new high-definition TV was two rooms away in the living room. But it worked! I got back to moving, exercising my will over my despair.

I knew then, and I know now, that I need to keep moving, mentally and physically. I'm fortunate that my current low-carb diet means that I only have to take one unit of insulin a day, but my daily exercise regimen helps with the quick metabolizing of that insulin. I don't know if pushing myself has been all that useful for me physically, but it has certainly helped with my morale. The body and brain must work together to keep the machine in good working order. Whichever side is lagging, the other side pushes to get up and go.

Having a disability means that many of us can never truly test the limits of our endurance. We don't have a whole lot of endurance to test! At that low point in my life two years ago, I was tempted to forget about the rest of the world, to be a spectator rather than a participant. By dragging my exhausted body *and* mind out of bed, I achieved a major victory over both middle age and this new disease.

Sometimes, just to keep moving IS the race. We might never score the winning touchdown, pass military training, or break the tape at the end of a marathon, but we have to savor every victory we can grab. And we win. Big time. We win through our determination to triumph over all those forces that attempt to slow us down. ❑

Michael P. Murphy, 42, has spinal muscular atrophy and lives with his family in Oconomowoc, Wis. He has written two science fiction novels, ***To Rule in Hell*** *and* ***Data Streets****, and a thriller,* ***Innocence Kills: A Paul Murdock Mystery****, available at authorhouse.com, Amazon.com and bookstores. He's hard at work on his latest novel,* ***Suka: A Paul Murdock Mystery****.*



Getting Creative with Adapted P.E.

Jacob Engers, a ninth-grader from Baltimore, switched from a 504 plan to an IEP once he qualified to receive adapted physical education (P.E.) services.

In most areas, schools require all students to complete a P.E. requirement. For this reason, adapted P.E., which can be provided by a physical education teacher or special education teacher, isn't considered a "related service," and occupational and physical therapy can't be used as substitutes.

Jacob, 14, has Duchenne muscular dystrophy and uses a power wheelchair full time. His mom, Cathy Engers, says Jacob did very well with a 504 Plan throughout elementary and middle school. The only difference between the 504 Plan and his new IEP is the adaptive P.E. program, she says.

Because the state allows some flexibility with the adapted P.E. curriculum, Cathy proposed that Jacob use the Nintendo Wii to fulfill his P.E. requirement. The school agreed and an adaptive program was designed by the head of the adapted P.E. program for Baltimore County Schools.

The Wii is a video game console that interacts with a wireless remote controller that users strap to their arm. The remote is activated by physical movements or by pressing buttons. The console typically connects to a television set.

To play a Wii sports game, the user straps on the remote and watches the TV screen for the virtual serve or pitch. The user then mimics the action of swinging a racket or a bat as if hitting the virtual ball. The console interprets the movement of the remote and the TV screen shows if the virtual ball has been hit, and, if so, where it went. The Wii can be used by people of all skill levels; as they improve, the skill level automatically increases.

Jacob still attends a general P.E. class. He participates in some activities with the rest of the class, such as when they're learning the rules of various games or completing written assignments. When he can't participate due to physical limitations, he goes to an office near the gym where he uses the Wii for the 90-minute period.

Jacob plays different sports on the Wii, including tennis and baseball. Oftentimes, another student comes in to play against him and keep him challenged. While using the Wii, Jacob is hooked up to a heart monitor, and his heart rate gets elevated, says his mom, showing "he's getting something out of it." ❑

Tracking Physical Activity and Energy Expenditure

Muscle-disease researchers say 'wearable technologies' may help in the quest to gain strength, lose weight

by Alyssa Quintero

Note: People with muscle diseases always should consult their physicians and MDA clinic team, particularly physical and occupational therapists, before beginning any exercise or weight loss program.

Low-tech devices such as step-counting pedometers can help people with slowly progressive muscle diseases track their steps and gradually increase physical activity levels.

When people exercise, they typically want to see results on the scale and in how they feel. Now there's another place to check for results.

"Wearable technologies," ranging from basic step-counting pedometers to high-tech "BodyBuggs," can give users concrete and immediate feedback about their physical activity — and in turn may help them achieve their exercise and weight-loss goals.

Some of these devices currently are being used by neuromuscular researchers to determine the effect of exercise or other therapies on people with muscle diseases.

What researchers want to know

At the NIDRR Rehabilitation Research & Training Center in Neuromuscular Diseases at the University of California at Davis, researchers are evaluating the role that wearable technologies play in monitoring and improving the health of people with muscle diseases.

Some researchers are evaluating the daily activities of people with muscle diseases by having them wear portable devices.

Because they aren't as active, people with neuromuscular diseases are more susceptible than the general population to "metabolic syndrome," says R. Ted Abresch, director of research at the facility. Metabolic syndrome is a weight-related condition strongly linked to the development of heart disease and type 2 diabetes.

Abresch says wearable monitoring devices provide researchers with insight into the kinds of physical activity being done by people with muscle diseases, when the activity is done, the duration of each activity and energy expenditure.

For example, UC Davis researchers gave a pedometer to 15 people with slowly progressive muscular dystrophies (such as limb-girdle and facioscapulohumeral muscular dystrophies). Participants tracked their steps for a week and then were instructed to increase their steps by 25 percent. Study participants were encouraged to look at the pedometer every day for six months and aim to increase their step counts.

Overall, the increased exercise had beneficial effects on quality-of-life measures, Abresch says. The participants felt better about themselves, and some experienced slight changes in weight.

"It's a simple, easy intervention that you can watch and do every day, so that's one that we think would be very effective," Abresch notes. "I would think that for most people who can walk, it would be helpful. But with our population, you always should ask your physician if it's OK or not."

In addition to simple pedometers, UC Davis researchers have conducted studies using the StepWatch Activity Monitor (SAM), a small pager-like device that's strapped onto the ankle and measures and records minute-by-minute step activity (or gait cycles). This device is not available to the general public.

SAM is programmed via computer to define the length of time for data collection and to make adjustments for different gait styles. It can be connected to a computer to upload data for further analysis, including frequency, duration and intensity of physical activity.

In a 2005 study, researchers investigated activity and heart rate patterns in ambulatory boys with Duchenne muscular dystrophy (DMD) for three days as they went about their normal daily routine. When compared to boys without DMD, the study participants had significantly more inactive minutes. When they were active, they took significantly fewer steps and spent fewer minutes at moderate and high step rates.

"The goal is to use the SAM to describe the type and level of activities that a person is doing in a normal day," Abresch explains. "You can tell how much time a person spends sitting, how much time a person is walking, how much time they're running and the length of time they're walking continuously."

Abresch hopes further research will determine if the SAM can be used both to modify behavior in people with muscle diseases and to gauge the effect of different therapies on activity levels.

For example, SAM is being used to measure outcomes in both the PTC124 clinical trial for boys with DMD and Insmed's Iplex trial in adults with myotonic muscular dystrophy. If the treatments improve strength, participants should walk more, which will be precisely recorded by SAM, explains Abresch.

He adds, "We like the step monitor and the pedometer because they're something that people understand. As long as people are ambulatory, using them makes sense."

That's an IDEEA

Accelerometers, which record how much energy (measured in calories) is used during physical activity, also are useful devices, says Abresch.

The MiniSun company, located in Fresno, Calif., has designed a computer-like device called the Intelligent Device for Energy Expenditure and Activity (IDEEA), which analyzes body motion, measures physical activity, monitors behavior patterns and estimates energy expenditure on a 24-hour basis.

Researcher Ming Sun (president of MiniSun) has worked with the UC Davis team to investigate new wearable technologies, and developed IDEEA as a way to determine energy consumption during all daily activities, not just prescribed intervals of exercise.

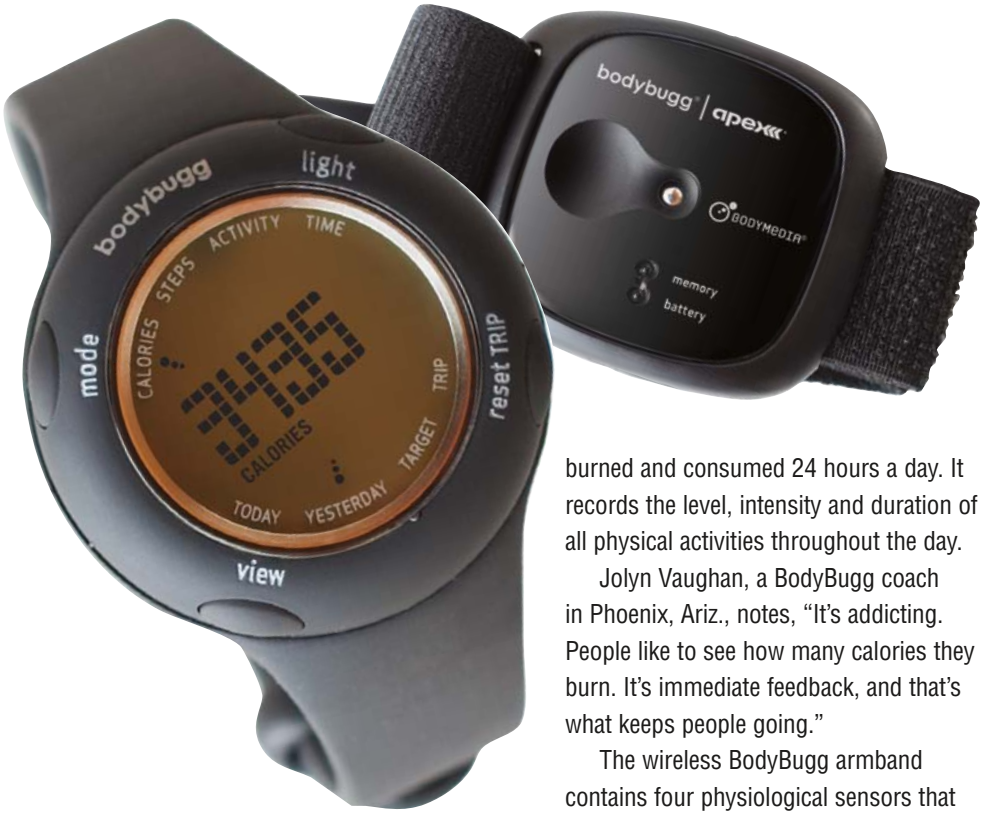
Now used extensively in research labs, IDEEA has five motion sensors, or accelerometers, that can identify more than 40 types of physical activity and provide detailed information on the duration, frequency and intensity of that activity.

The information can be uploaded to a computer and the MiniSun Web site will give a detailed analysis of physical activity and energy expenditure components in any given period of time.

The device takes into account a person's height, weight, age, gender and physical ability level when estimating energy consumption.

"The good thing is that it's very accurate and does a good job of measuring energy balance," Abresch says. "It's a very interesting device."

Sun's research team recently completed the new wireless IDEEA3 system that



monitors and records cardiac functions, including heart rate and electrocardiogram; electromyography (measuring the electrical activity of muscles); leg, trunk and head motions; and ground-impact force.

"Combined with these variables, IDEEA becomes ideal for simultaneous measurements of cardiac stress, energy expenditure, function and activity/exercise levels," Sun says.

Burning calories with the 'Bugg'

Although IDEEA's extremely high cost makes it appropriate mainly for research, there is an accelerometer option that's available for consumers — the BodyBugg.

Manufactured by Apex Fitness, located in Westlake Village, Calif., the BodyBugg is a small portable device that attaches to a user's arm via an armband. Used for calorie management, it's been featured on the NBC TV weight-loss show "The Biggest Loser."

The BodyBugg program helps users track and adjust the number of calories burned and consumed 24 hours a day. It records the level, intensity and duration of all physical activities throughout the day.

Jolyn Vaughan, a BodyBugg coach in Phoenix, Ariz., notes, "It's addicting. People like to see how many calories they burn. It's immediate feedback, and that's what keeps people going."

The wireless BodyBugg armband contains four physiological sensors that accumulate data about motion and physical exertion, body heat, core temperature and amount of sweat on the skin. The user then uploads the information to an online program (PC and Mac compatible).

Tailored to the user's age, gender, height, weight and physical ability, the BodyBugg program calculates, minute by minute, activity levels and calories burned.

"You don't necessarily have to go to a gym to do it," Vaughan says. "People get their physical activity in so many ways, like cleaning their house, or running around and playing with their children, and they're surprised at how many minutes of physical activity they get just by doing their normal routine."

(Unfortunately, the device isn't waterproof and can't be used during pool exercises, a preferred activity for many people with muscle disease.)

Because the key to weight loss is burning more calories than are taken in each day, the Web-based program tracks calories burned, calories consumed (based on the user's daily food logs) and the difference between the two figures. The online program also offers food options and a menu planning system.

When someone is in a wheelchair or has limited activity, "it can be easy to overeat," says Vaughan. BodyBugg users report that the device makes them more aware of food calorie values, allowing them to better monitor calories and keep weight under control.

The armband with digital display costs $348.95, which includes online food logging with customized meal plans, a six-month subscription to the Web-based program and a phone session with a BodyBugg coach. Consumers must fill out an extensive health history and food profile questionnaire before using the device.

After the initial subscription expires, users must renew in order to maintain access to the data collected by the BodyBugg and online program. Renewals range from $14.95 per month to $99 per year. (For more information, call (800) 656-2739, or visit www.bodybugg.com.)

And remember: Before using a BodyBugg, pedometer or any wearable technology, always check with your physician. ❑

Wear and Tear on the Chair

Care and feeding of the sports machine

by Bill Norman

Power wheelchair users are a growing presence in the world of sports. Players (and parents) need to be aware of the unique stresses that power chair sports exert on the chair's framework and machinery.

Common power chair sports include:

Power soccer. Two four-player teams square off against each other, typically on a basketball court. Players use their chairs (equipped with an add-on plastic or metal foot guard) to kick an oversized ball toward the goal. They also use their chairs to intersect or deflect an opponent's kick.

Power hockey. Similar to power soccer except players slap a puck (usually a plastic 2"- or 3"-ball) with lightweight hockey sticks. Sticks may be used by hand or taped to the wheelchair and the chair's power provides the "swing" energy.

Dog agility competition. Dog owners race alongside as their specially trained dogs navigate an outdoor obstacle course, striving for fastest time and accuracy.

Rural racing. Also sometimes called extreme racing, this sport entails competitors – usually equipped with protective clothing – striving for fastest time when negotiating multi-mile race courses that can include dirt, mud, water and other obstacles.

The sport of extreme racing has wheelchair pilots powering at high speed through the great outdoors.

All of these sports are high-speed and chairs get a real workout. In particular, wheelchair motors, controllers and batteries can be subjected to stresses their manufacturers may not have intended. The metal structure of the chair also can be bent, twisted and broken.

Heat is Enemy No. 1

Mark Smith, creator and operator of the Web site wheelchairjunkie.com, says the main issue in power soccer is "thermal foldback," where the constant acceleration demands and turning resistance run very high amperage through the controller, overheating the chair.

"Power chairs have been custom modified for individual users from time to time, but sacrifice everyday reliability, as there's long-term harm to constantly running power chair components at very high amperage and excessive heat," says Smith. "I've seen serious European players with custom-modified, unlimited electronics go through a controller and/or a set of motors every few games by eliminating thermal protection -- it makes for a very dangerous, expensive sport."

Adam Elix, program coordinator for Far West Wheelchair Sports in San Jose, Calif., (www.fwws.org) a nonprofit organization that promotes and organizes wheelchair sports, says power chairs can be modified to a certain extent for sports.

"We have a mechanic who can adjust the gear ranges — gear one for street cruising, gear four for athletes.

We've done a couple of things to deal with heat problems. One parent [of a soccer player] is a tech guy who hooked up a personal fan to blow on the battery compartment. We've also used ice packs on top of the batteries, and we've tried blowing [canned] compressed air on them. It comes out of the can very cold."

Component factors

Power chairs intended for rigorous sports use can benefit from utilizing specific types of equipment:

Belt drives rather than direct drives. A belt-drive chair, in conjunction with wheelchair motors that have a brake, can be abruptly switched into reverse gear from full-speed forward, says Dick Roberts, customer service representative for 21st Century Scientific, (www.wheelchairs.com) an Idaho firm that manufactures the Bounder brand of power wheelchair. He notes, "If you try that with a direct drive, you'll have parts flying everywhere."

6-pole motors rather than 4-pole. Motors have magnetic poles, just like simple magnets, but their poles are creating by winding magnetic wire in bundles. Motors with fewer poles spin faster (1,800 revolutions per minute, in the case of four poles) and have to work harder. Six-pole motors spin slower (1,200 rpms), but they have more torque (the twisting force that produces rotation) and can propel a chair at greater speeds. The 6-pole version also runs quieter.

John Cross, president of ASI Technologies, (www.asidrives.com) which designs and manufactures drive systems for wheelchairs, says another advantage of 6-pole motors is that "a lower mechanical gear ratio could be used with the higher pole motor."

However, apart from specialized applications like sports, Cross said, "The added complexity and cost of 6-pole vs. 4-pole is usually not justified in a wheelchair motor."

Bigger batteries are better. Wheelchair batteries usually come in three group sizes: 22, 24 and 27, with the larger numbers signifying larger external dimensions. Group 22 is standard for wheelchairs; group 24 is utilized to power heavier chairs; 27 is best for frequent chair use (as in sports) and traveling long distances over rough terrain. Larger batteries also often have the greatest power, reflected in their ampere ("amp") hour rating. Before attempting to move up in battery group size, chair users must ensure there's adequate space in the battery compartment and that their other electrical gear, such as the chair's controller, is compatible with a boost in power.

Mike Serhan, product manager and vice president of sales for Drive Medical Design and Engineering, (www.drivemedical.com) said gel cell batteries cost more and usually don't have the range of wet cell batteries, but they do provide more torque. Some wet cell batteries can leak or spill acid; gel cells are sealed (as are some wet cells), and that can be important in

Power soccer wheelchairs often have structural and mechanical modifications.

In power hockey, players sometimes tape their sticks to the wheelchair frame.

sports where impacts and steep-terrain can sometimes knock batteries (along with the wheelchair) onto their sides.

Shock absorbers. Most wheelchairs don't have them. Roberts said optional nitrogen-filled shock absorbers surrounded by coil springs greatly reduce the impacts felt by a wheelchair user when traveling over rough terrain.

Digital controls rather than analog. "The read-outs on your controller screen look the same with both types, but digital is more precise. You get more joystick sensitivity and ability to make a fine distinction between drive speeds," Roberts said.

TIG welds for frames. The welds that hold metal wheelchair structural members together can be subjected to front and sideways wrenching and twisting forces strong enough to break them apart during intense sports action. Tungsten insert gas (TIG) welds are stronger than those produced by common arc welding.

Different tires for different surfaces. Roberts said low-pressure wheelchair tires that are taller (14 inches vs. 12 inches) and wider (6 inches vs. 4 inches) offer a smoother ride and more flotation (less tendency for the tires to mire down on wet or muddy terrain). Serhan also suggested that flat-free wheelchair tires may be good for outback cruising, while pneumatic tires are preferable on the court.

Heading for the woods

John Mryczko is president of Extreme Chairing (extremechairing.com) and the Power Wheelchair Racing Association, based in the Chicago area. Both organizations organize outback wheelchair treks and races. Extreme Chairing, a 501(c)3 nonprofit group, aims to expand the popularity, corporate support and geographic range of events for a sport that is just getting off the ground.

"We're pretty much using everyday power chairs in the events right now, but competitors usually have their chairs' controllers reprogrammed so they can get more speed. When you first get up to 10 miles an hour, it seems really fast, but after a few miles it just seems normal. Sometimes I even wonder if I'm slowing down when I'm not," Mryczko said. He hopes they'll succeed in boosting the speed to 20-plus mph.

Some specialized all-terrain power chairs have four-wheel drive; others have literally replaced wheels with smaller versions of the metal treads used on tracked vehicles.

In that regard, the Wheelchair Site (www.thewheelchairsite.com), an independent consumers' guide to wheelchairs, scooters and accessories, says of all-terrain wheelchairs, "Sure, the most rugged models will travel over just about anything, but they drive like tanks!" The site suggests renting an all-terrain before buying, and notes that many resorts throughout North America offer all-terrain rentals to vacationing guests.

Shopping considerations

Abledata, which provides information about assistive technology products to the National Institute on Disability and Rehabilitation Research (part of the U.S. Department of Education), also has some guidance about power chair drive systems.

Power wheelchairs have either rear-wheel, front-wheel or mid-wheel drive. The drive type has a significant effect on maneuverability and handling.

Rear-wheel drive is better for outdoors travel, said Drive Medical's Serhan, while mid-wheel drive is better on a court (soccer or hockey) setting. "But mid-wheel drive is a little squirrely if you're going to be outdoors," he said.

The Abledata website, www.abledata.com, contains an exhaustive list of wheelchair manufacturers and distributors; a wheelchair-related organizations resources list; and information about approved wheelchair feature standards approved by the American National Standards Institute in cooperation with the Rehabilitation Engineering Society of North America. ❑

Are Power Chair Sports Really Exercise?

by Kathy Wechsler

Many kids have tried power soccer or power hockey at MDA summer camp and loved it.

Everyone agrees that these sports designed for power wheelchair users are a lot of fun — but are they also exercise? Absolutely, say the experts. And playing once a year may not be enough time to reap all the benefits power chair sports have to offer.

What's the game?

Power soccer and hockey are similar to the traditional versions of the sports. Players are males and females of all ages. Any child who can safely operate a power wheelchair should be able to play, says Greg Carter, who co-directs the MDA/ALS Center at the University of Washington Medical Center in Seattle.

Both sports are played on a regulation-size basketball court by two teams of four. Power soccer players use foot guard attachments to kick a 13-inch ball into a net, while power hockey players hit a small plastic ball with hockey sticks, which sometimes are mounted on their chairs.

A heart-y workout

While playing power soccer or hockey isn't a "locomotor" activity, such as walking, running or swimming, it still puts an aerobic stress on the heart, says J.P. Barfield, assistant professor of fitness and wellness at Tennessee Tech University in Cookeville.

"One of the ways that we quantify physical activity is by examining or documenting that increase in heart rate," says Barfield, who participated in studies in which power soccer players' heart rates were measured before and after a game. Heart rates increased during games to meet the players' energy demand.

Aerobic activity (which increases heart rate) may reduce the risk of heart disease, high blood pressure and obesity. It also can stimulate metabolism, burn calories, and improve mood and appetite.

Because the heart is stressed during

power chair sports, Barfield recommends that children with diseases that affect the heart (such as Duchenne and Becker muscular dystrophies and Friedreich's ataxia) first visit their doctors.

Keep it moving

Power sport players may be sitting in their chairs, but they're still getting exercise.

"You're balancing yourself in your chair, and as you're moving, you're shifting your body weight continuously to try to move where you're pointing your chair," Barfield says. "In muscular dystrophy, we see that kids are 'exercising' while playing in a motorized chair just because of the other movements that are occurring."

No matter their ability level, children try to get the most possible movement out of their bodies during play.

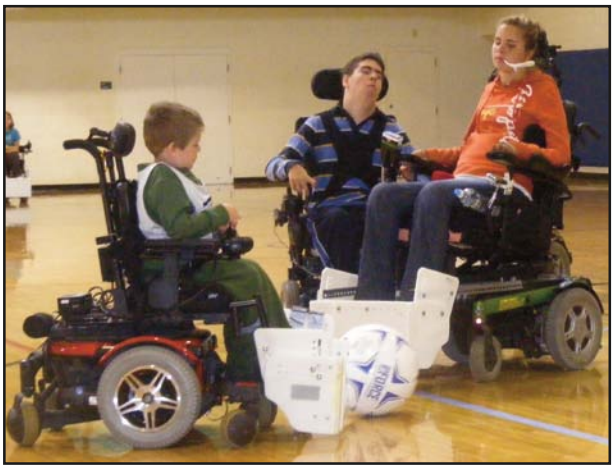
For example, 12-year-old Lauren C. Taylor of Lewisville, Texas, used to have her hockey stick taped to her wheelchair, because her congenital muscular dystrophy affects her arms. Eventually, she decided to try holding and swinging the stick instead of attaching it, because her teammates held and swung their hockey sticks. By challenging herself to keep up with her peers, Lauren gained some physical benefits.

"I use my arm strength to hit the ball, so my left arm has gotten a little stronger since I started," she says. "My left fingers are a little more flexible than my right, considering that's the arm I play with."

"Sometimes she'll move her legs like she's running," says her mother, Faith. "It's just overall — she's yelling, so she's exercising her lungs, and moving that stick exercises her arms and her shoulders." The sport also seems to help with Lauren's endurance.

Dena Miller of Haverhill, Mass., considers power soccer to be a great workout for her 6-year-old son, Joseph, who has type 2 spinal muscular atrophy (SMA2). When he started playing, he'd need a nap after practice, but now he stays awake and seems to have more stamina throughout the day.

"Everyone, even the adults in chairs, are tired once they're done — it's just great to see them be able to experience that release of exhaustion from a game well played," says Dena. "I played a lot of team sports and really didn't think he'd ever have that feeling of 'ahh,' totally tired from a great game."



Facing page: Lauren Taylor takes control of the ball and speeds down the court. Above:Power soccer allows Joseph Miller, left, to make friends of all ages.

Putting driving skills to the test

Power chair sports also can improve coordination because the body learns to process information faster, says Greg Carter.

Because players must zoom up and down the court without running into other players, practicing their coordination and driving skills is a must, says Dominic Russo, president of the United States Power Soccer Association (USPSA) in Carmel, Ind.

Each Saturday, Joseph's team spends three hours running drills to sharpen its wheelchair maneuvering skills and preparing both physically and mentally for power soccer plays. During drills, team members race in and out of cones, then stop suddenly and drive backwards through them. Dena has noticed these practices paying off.

"I thought he was a great driver before; now he can blow your mind," she says, noting Joseph excels at driving full speed backward. "He loves to show off his driving for people."

Power hockey adds an extra element of eye-hand coordination for players who hold their sticks.

Kickin' it with friends

Barfield says the chance to socialize is one of the greatest benefits of power chair sports.

"There aren't many activities that are designed for people with neuromuscular disorders, so the social opportunities that children have are limited," he says. "But power soccer is certainly one outlet for individuals with muscular dystrophy to be able to interact with all sorts of individuals."

Not only is this good for social skills but it also encourages independence, he notes.

Peer connections make children more likely to *want* to stay active so they can be independent and keep seeing their friends.

Like all athletics, power chair sports teach life skills such as self-confidence, discipline, teamwork, sportsmanship, independence and strategic options, says Russo, whose two children, Natalie, 20, and J.C., 18, both with SMA2, play power soccer for the Ball State University Cardinals.

Lauren Taylor admits she was a little intimidated when she began playing hockey at her local YMCA. She was the youngest player and the only girl.

"At first I was the shyest one there," says Lauren. "Then once I became friends with everyone, I became one of the most outgoing and outspoken."

Adds her mother, "I'd say confidence is the biggest thing that I see from her, because every time she makes a goal, or outplays somebody, she just lights up."

Faith says there's a friendship and camaraderie that comes with being on a team and cheering for one another. She thinks it's good for Lauren to learn to play along with others, and to experience winning and defeat.

Power sports help level the playing field, giving children with disabilities common interests with able-bodied kids.

"I can't tell you the number of times I've heard a player say they've always gone to soccer, basketball, softball games to cheer on their brother or sister and now their brother, sister, mom, dad are here to cheer them on," says Russo.

Players aren't the only ones getting benefits from power chair sports, says Russo. Parents enjoy being around other parents with similar situations to swap stories, advice and support.

"Power soccer has been so great for Joseph's dad and me, because a few years ago we didn't think this possible," Dena says. "I can finally say I'm a soccer mom, and that's priceless to me." ❑

Preventing Injury

Whenever there are power wheelchairs traveling at high speeds and maneuvering in close quarters, there's bound to be a fender-bender or two.

Lauren Taylor, who has congenital muscular dystrophy, has had her share. She's broken some footrests (her own and another player's) and a few hockey sticks.

Power soccer players have a foot guard that goes around the front of the chair but not so in power hockey, says Lauren's mom, Faith. "They're just out there, and when they crash into each other, it's very possible to get a leg hurt, or whatever. I haven't seen it happen too often, fortunately."

Power wheelchair sports must be carefully supervised, says Greg Carter, co-director of the MDA/ALS Center at the University of Washington Medical Center in Seattle.

"We've had a number of serious injuries over the years in power wheelchair sports," he says, describing a game in which a player ran over the ball with his power wheelchair and it flipped over on top of him, causing serious facial lacerations (but no head injuries, fortunately).

Kids need to be securely strapped into their wheelchairs and have plenty of head, neck, and spinal support, Carter emphasized. He suggests buying these supports from the wheelchair vendor.

Without wearing chest straps and seatbelts, there's significant risk of falling from the chair, particularly with the sudden stopping and starting in a game, he warns. Helmets, although not required, are probably a good idea, he adds.

Eugène Etsebeth

Am I Disabled or Aren't I Disabled?

by Eugène Etsebeth

While I lay on a plinth, a Swiss doctor gestured that I should lift up my left leg. He bear-hugged my leg and resisted its elevation with his full body weight. He grunted as he fought my movement.

He wasn't the only person prodding and inspecting me. A team of doctors, biokineticists and physiotherapists surrounded me. It was like the United Nations of medics. Besides the Swiss doctor, there was a lady from Israel, a young athletic Austrian and two Americans.

They were tallying up points of my strength on a clipboard. And things weren't going too well — it was turning out that I was too strong!

It wasn't suddenly apparent, but there was a growing unease and stillness in the room, and knowing glances were passed between the medical tribunal. I was about to be eliminated — not only from the "World Championships," but also from a man-made categorization that determines who is "disabled" and who is not.

A difficult irony

I had trained hard for the equivalent of the World Paralympic Championships (called the Open European Paralympic Championships), held under the auspices of the International Paralympic Committee (IPC). My training buildup had been intense. At night I would sweat on my stationary bike as I cycled more than two hours. On weekends and early mornings, I was on the cycle track practicing starts, sprints and keeping my top-end speed at over 26 miles per hour on the cliff-like embankment.

My proudest moment was at the airport just before the team flew to Holland, when I received my team kit with our national emblem, the Springbok, emblazoned on my tracksuit. I had achieved the ultimate — to represent South Africa at an international sporting event.

A day before my classification exam with all the examiners, I was starting to feel part of the disabled community. The opening ceremony had over 30 countries participating. We walked on to the famous Alkmaar cheese market with our country's flags waving in front of hundreds of onlookers. I was finally making peace with my body. I displayed my imperfections proudly — along with the other athletes. I felt a sense of belonging. My body was OK. I didn't need to conceal my imperfections.

Excommunicated

The Swiss doctor placed my leg down alongside the other. His perspiration matched my own. The medical team finished their examination by informing me that I do not fit into any particular locomotor category. I had too much muscle to fall into a category to take part in the championships. Luckily for me, in the spirit of the competition, I was allowed to compete even though I didn't fall into the classifications defined by the IPC, the international cycling body.

But this irony did not sit well with me. You see, muscle is actually my biggest problem. It is in a constant state of atrophy. I get weaker by the day. It is a slow weakening. I have facioscapulohumeral muscular dystrophy (FSHD), or in cycling jargon, a slow puncture.

This hereditary condition can vary by

degrees. Dale, my older brother, can walk only with the help of orthotics. My situation is that I can still walk, albeit with a slight drop foot. Push-ups are out of the question, along with sit-ups. I use trick movements to get myself out of bed in the morning. But to the untrained eye I look almost perfectly normal.

With a jacket, loose-fitting shirt and trousers, you don't see my winged scapulae, hunched shoulders or atrophied stomach and chest muscles. Casual onlookers also don't notice that I kick the lip of raised tiles, or feel unsteady on uneven surfaces.

Nevertheless, the damage had been done by the Paralympic organization. I had been excommunicated. My fears of where I fit in society had been amplified onto a world stage — am I or aren't I disabled? Which family do I belong to? It was like I was banished from the disabled family,yet I didn't neatly fit in with the able-bodied folk. I was alone in the middle somewhere — in limbo.

I can't lay the blame of this schizophrenic toggling purely on the medical examiners. With cock-crowing regularity I position myself as fully healthy. At work I don't trumpet my genetic flaws. I don't gush about my deterioration to new friends. I wear a cloak of normality, but underneath, the muscular truth lurks. One day in the near future, when my orthotic arrives to assist me to walk or a Segway is called upon for transport, I will finally land with my rightful family. For now, I am mentally dangling.

Finding my fit

Since being "out-classified" I have adopted a more philosophical approach to my body's deterioration. No man-made system can pigeonhole my physique or my life. I have climbed Mount Kilimanjaro, cycle-toured through Europe, gotten married and achieved success at work.

My latest mission — and my wife's — is to have a healthy child. We already have had one attempted in vitro fertilization cycle using preimplantation genetic diagnosis (PGD). This is a process whereby geneticists implant only embryos unaffected by my faulty gene. We are to return to England to re-attempt this process.

For me, this is the real world championships. The prize is the ability to give my child — or, hopefully, twins — a fresh start. To avoid all the nettles and barbs that can sting the soul of somebody who looks different. My soon-to-be role as a dad supersedes all concerns or worry about where I fit in. I will carve out my own sense of belonging.

Cycling still runs in my blood. On any given morning you'll find me readying my bike for a ride. I clip my shoes into my pedals and ride, oblivious to what my body looks like, oblivious to the deterioration of my muscles.

Rather, I revel in the clicking of my gears and the familiar tapping cadence of my pedal strokes. I cycle into the distance knowing full well that I am harnessing the power of my body and storing these sensations deep into my memory banks.

I know I will ride on, long after the pedaling stops. ❑

Etsebeth and his wife, Monique, live in Johannesburg, South Africa, where he is a senior business analyst for a software development company. His extensive travels include spending a year in the U.S. in the mid-1990s. Says Etsebeth, "I come from a family that has been hit hard by muscular dystrophy. We all love adventure and travel. We are a close-knit family, and that makes all the difference."