

Help your child take control

It's a frightening thought: Just when your child is facing huge physical changes and emotional challenges, you'll be letting go of some of the control involved in managing his or her disease. Like handing over the car keys, handing over the management of juvenile myositis may have a lot of apprehension mixed in with the parental pride in your child's maturity.

Most likely, you've talked to your child about assuming more and more responsibility, so it's not happening all at once. "Think of it as you would think about teaching your child any other adult responsibility," says Ann Robertson, an adult who was diagnosed with JM at 16. "You've been teaching your children how to cook, do laundry and clean up after themselves. Becoming responsible for their medications, appointments, diet and exercise is not that different from, say, balancing a checkbook."

In fact, Robertson says, the more you can allow your teenager to control his environment, the better. "It's frustrating when you have so little to say about how your disease is affecting you," she said. "It helps to have the feeling that you can be in charge of something in your life." If your child is sensible and responsible -- which often happens when children are exposed to major challenges at a young age -- you may want to acknowledge it by showing trust in his or her ability to manage school work, laundry, and finances without much parental involvement.

Of course, young adults begin the process of assuming the management of their health long before they can drive to the clinic or know when their medication isn't working the way it should. It's a process that should begin as soon as they're diagnosed.

One difficulty parents mention is the lack of age-appropriate health information for teens. There are materials directed towards young children and others directed towards adults. This situation is rapidly changing: and teenagers, like most young adults, are increasingly finding health information on the internet, where links, visual illustrations, and case studies allow them to delve as deeply as they are able. Most of these materials are written in understandable language, so you may be surprised to find your teenager becoming an expert on JM research; or drug interactions; or signs of a flare.

Finding information is the easy part. Grappling with peer pressure is harder. Robertson points out that drugs prescribed for juvenile myositis are extremely potent. If your teenager is taking methotrexate, for instance, experimenting with alcohol has potentially severe implications.

The Double Whammy

Susan Sawyer, Director of the Center for Adolescent Health in Melbourne, Australia, notes that physicians haven't dealt much with the issues of sexually-transmitted diseases and drug and alcohol abuse in chronically ill teenagers because they may have

assumed that somehow these teens would behave differently from their peers. Just the opposite is true, Sawyer writes in "Adolescents with Chronic Disease: the Double Whammy." For whatever reasons, research indicates that chronically ill teens take the same sorts of risks as other adolescents. This is a "double whammy" for many reasons.

"When you have JM, you are even more susceptible to the effects of alcohol," says Cynthia J. Mears, an Assistant Professor of Pediatrics at Northwestern University's Feinberg School of Medicine. In "Approaching Adolescence," a chapter in "Myositis and You," Mears writes: "Your liver, which breaks down the alcohol, might not function normally. This is because of the myositis-related inflammation or some of the treatments for the myositis itself. In particular, prednisone, methotrexate, azathioprine and other JM medicines can impair your liver function and your ability to break down the alcohol." Mears warns that an impaired liver makes young bodies much less able to handle alcohol and recreational drugs, so that even very small amounts can have a big impact.

Another issue particularly hard for parents is grappling with the information that your teenager is more likely to catch -- and have more serious consequences from -- sexually transmitted diseases, than are teenagers without JM. Teens who are taking medica-

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Take Control, continued from cover.

tions for JM have suppressed immune systems and are at higher risk for Human Immunodeficiency Virus (HIV).

They're also likely to have more serious complications and reactions from common sexually-transmitted diseases like gonorrhea, both because of their existing illness and because the medications used to treat the new illness are designed to strengthen the immune system, an approach that counteracts the drugs that are stepping down the immune system to lessen the JM inflammation.

Protecting your daughter from pregnancy is also more complicated because of the interaction between hormonal methods of birth control and myositis medications, Mears writes. This is true both of birth control and the "morning after" pill. Make sure your daughter consults her physician before taking these medications.

On the other hand, there are serious implications for pregnancy while your daughter is on certain medications. Some of the common JM medications can be harmful to an unborn baby, causing deformities or death.

Dr. Mears give us some statistics:

- 12-15 million sexually-transmitted infections occur every year in the United States. Adolescents have the highest infection rate of any age.
- Almost one-fourth of the population over 12 years of age is infected with the herpes simplex virus type 2.
- 40 to 80 thousand people are infected each year with HIV; about half are people younger than 25 years old.
- The most common indication that a teenager has a sexually-transmitted infection is no symptom at all. This is true even in teenagers who have HIV.

Dr. Sawyer notes several ways that chronically-ill adolescents are influenced, both by their disease and by their parents' response to their disease.

- Adolescents with obvious signs of illness (like skin symptoms, scars or disabilities) may feel less attractive. This can affect their ability to make friends and form intimate relationships.
- The very understandable desire to belong to a peer group and feel 'normal' can cause medication neglect because of embarrassment taking medication publicly.
- Some experimenting that might be considered normal in healthy adolescents may have dire consequences.
- Your child's self-discipline in school may be subtly affected by lowered expectations from parents or school.
- There may be a lack of role models for successfully getting through adolescence with a chronic disease.
- Over protecting your child may delay independence.

There's good news along with all the worries. A study earlier this year examined teens with rheumatoid arthritis, and what they needed (according to their own judgment) to assume the responsibility for their care. The study, published in the January issue of *Arthritis Care & Research*, looked at web-based programs for managing chronic disease in 36 Canadian students.

The teens, from 12 to 20 years old, were very clear about what they needed in the way of support. They reported that they were gradually learning to take control of their health care from their parents and health care providers.

The teens expressed a need for more information on managing their disease. They also reported their own worries about peer pressure and the

social and psychological issues involved in being a teenager with physical limitations. Many of them asked for ways to get more social support and acquire more skill in social communication. They saw the internet as a way to fill all these needs.

Obviously, writes Dr. Sawyer in "Double Whammy," reaching the age of 18 does not automatically insure that teenagers can assume responsibility for their own care, nor does it mean that a 17 year old might not be ready. She says teens show readiness for this transition if they:

- Can name and explain their condition
- Can list their medications, treatments or other management practices (like diet and exercise)
- Can explain why each medication or management practice is necessary
- Remember to take their medication most of the time
- Can answer questions asked by doctors or other health professionals
- Can ask questions of their doctors or other health professionals
- Can arrange (and cancel) appointments
- Can consult with doctors or other health professionals without a parent
- Have their own insurance information
- Remember to obtain more medication before it runs out
- Know how to have prescriptions filled at a pharmacy
- Have a desire for their health care to be independent of their parents
- Can choose their health over other desires

"Transitions" in this issue of the Companion has information directed at young adults.



Ask the doctor

Dr. Lisa Rider, a pediatric rheumatologist with a long-time interest in juvenile myositis, is a TMA medical advisor and deputy chief of the National Institute of Environmental Health Sciences, at the National Institutes of Health, and has been closely associated with TMA since its beginning. She led a Live Discussion on TMA's website early this year. The following questions and answers are taken from her discussion. For the full transcript, check the "Community" section of TMA's website at www.myositis.org.

Question: How can lab results be normal when disease symptoms are present?

Lisa Rider, MD: First, some of the muscle enzyme tests are not that sensitive in detecting active disease in children with JM. Chances are greater of picking up an abnormality in the enzymes if a whole panel of them is followed, rather than just 1 or 2 tests. This would include CK (creatinine kinase), aldolase, LDH (lactate dehydrogenase) and transaminases. We have found LDH to be the best marker of active disease among these.

Other reasons why the tests may be normal, even when the disease is active: there are likely inhibitors of the enzymes (especially CK) in the blood of JM patients that would cause the levels to be falsely low. Also, if the illness has been going on a while, there can be muscle atrophy (thinning), which causes the enzyme levels to be low- so they will not elevate when the disease is active. Finally, enzymes can be normal and the disease still active because the activity can be outside the muscles- in the skin, joints, intestines, etc- and this activity often does not cause the muscle enzymes to be elevated.

Question: Have you ever had patients

with JM whose liver function tests stay elevated even after they are in remission?

Lisa Rider, MD: Unless the liver function tests are elevated for another reason (an underlying liver problem), I would say no. We like to define remission as absolutely no sign of active disease- so if the tests are still abnormal and this seems to be related to the myositis, I would think of the illness as still being active, not in remission.

Question: What is your definition of remission?

Lisa Rider, MD: We have worked in a collaborative international group of myositis experts, called IMACS Group (International Myositis Assessment and Clinical Studies Group). The IMACS Group considers remission to be no signs or symptoms of active disease, stable muscle strength and function, normal enzymes and other blood tests, no active skin rashes or other activity in other organ systems, and the patient being off medication for a period of 6 months or longer. A more achievable goal is a complete clinical response, with the patient still on some medication.

Question: Can you have continued gastro-intestinal vasculitis involvement, even though labs continue to be stable, with the exception of LDH, which continues to stay high but stable? My 11 year-old daughter has struggled with GI pain before diagnosis and ever since. Can GI vasculitis affect absorption of nutrients?

Lisa Rider, MD: Some children with JDM develop skin ulcerations- a true opening in the skin tissues (dermis and epidermis) that we think is related to the same vasculopathy that occurs in the muscles. It seems the capillaries in the tissue are attacked by the immune system, and this leads to their not being able to supply the tissues with nutrients, which then

leads to the open holes, or ulcerations, which we think is from lack of nutrients or dying tissue.

Divots, sometimes in the inner canthus of the eyes, sometimes other places, are smaller versions of these ulcerations in the skin; they may also not run quite as deep- they may be erosions. These lesions do indicate the illness is active. Although not a lot is yet known about the prognosis of children with these ulcerations, we do know that they are associated with some pro-inflammatory cytokines, including TNF-a, and some anecdotal reports suggest these may indicate a more difficult course of illness. Some children only develop the skin ulcerations, others develop skin and gastrointestinal tract ulcerations, some only gastrointestinal tract ulcerations.

Question: Typically how long do kids receive intravenous immune globulin (IVIG) if they are doing well and the Prednisone is being reduced? My daughter's labs have been excellent and she is going to 10 mgs from 60. Her doctor said she does not know how long she will have her remain on the IVIG.

Lisa Rider, MD: IVIG can be continued a while, or stopped after only a few months. It varies with each patient. Fortunately, many JDM patients, unlike adult DM patients, seem to continue to respond to IVIG over a long period of time. IVIG is expensive, often in short supply, and does require an intravenous treatment, so these are some of the reasons it is often discontinued after three or six months.

Question: What are the most common misunderstandings about JDM that you have encountered among patients, their families, and doctors who do not have a lot of experience treating this condition?

See **Ask the doctor**, on back.

Orthopedists to parents: Choose the right backpack

Many children and teenagers carry backpacks during the school year for schoolbooks and other supplies.

“When used correctly, backpacks are the most efficient way to carry a load and distribute the weight among some of the body’s strongest

muscles,” says Eric

Wall, director of orthopedic surgery at Cincinnati Children’s Hospital Medical Center.

In May 2002, the U.S.

Consumer Product Safety

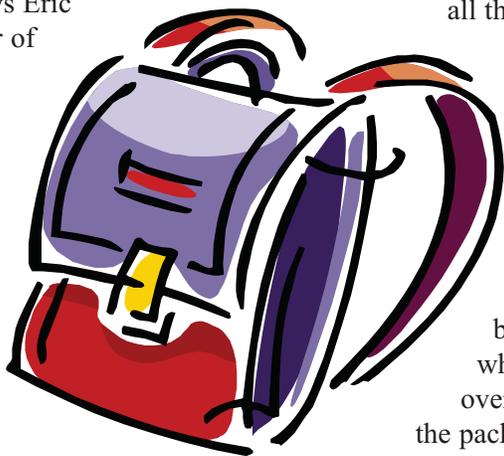
Commission reported

backpack related injuries sent almost 6,000 students each year to emergency rooms. The list swells to more than 13,260 reported injuries to children between the ages of five and eight, when those treated in doctors’ offices and clinics are included.

The American Academy of Orthopedics confirmed that backpack injury is a significant problem for children. Cincinnati Children’s Hospital Medical Center recommends these simple guidelines to prevent any unnecessary injuries to your child throughout the school year:

- When choosing the right backpack, look for one that is lightweight, has two wide and padded shoulder straps, a cushioned back, and waist straps. A rolling backpack is another good option if your child has to carry a very heavy load, but school terrain is much more challenging to negotiate with wheels than your average airport.

- Always encourage your child to use both shoulder straps and make sure the straps are tight.



- Limit your child’s backpack to no more than 15-20 percent of his or her body weight.

- Organize your child’s supplies and books so that the heaviest items are closest to the center of his or her back and all compartments are used.

- Persuade children to stop at their lockers often so they’re not carrying all their books throughout the day.

- When wearing or lifting a heavy backpack, remind your child to bend using both knees.

- Do not leave backpacks on the floor where others can trip over it, and do not swing the pack around where it may hit other people.

- Back and shoulder discomfort is common with heavy backpack use. If your child complains of persistent back pain, consult with your child’s pediatrician.



Progress in treating calcinosis

Calcinosis (the formation of hard lumps under the skin) continues to be a problem for children with juvenile myositis.

If your child is forming new deposits, his or her physician will check to see if the myositis is still active and if the calcinosis itself, which can be associated with a lot of inflammatory changes, is in an inflammatory stage. Pediatric rheumatologists often try a variety of drugs to directly decrease the deposits of additional calcinosis. These include probenecid, and also calcium channel blockers, bisphosphonate therapies (also used to treat osteoporosis) and phosphate binders.

“None of these agents has been tried in a randomized controlled trial,” said Dr. Lisa Rider. “The experience with them is very mixed- some successes, some failures, and we just don’t know enough to know whether any of these really work. It is best to discuss these options with your child’s myositis doctor and see if they think it is appropriate to try any of these types of medications for the calcinosis.”

A recent trial using intravenous infliximab in children with JM has added another possible treatment to those mentioned above. The small trial, by Drs Riley, McCann, Maillard, Woo, Murray and Pilkington, studied this drug, also called by its brand name, Remicade, in five juvenile dermatomyositis patients whose disease course proved refractory to multiple drug treatments. All of them had calcinosis, and improvements were seen in all five patients. The findings were published in *Rheumatology*.

The researchers classified “improvement” according to a previously agreed-upon system of measurements from observation, range of movement and, in some cases, diminished calcinosis and skin symptoms. There were no major side effects. “Calcinosis can be a debilitating complication of JDM, resulting in joint contractions and calcinotic infections,” said the authors. “Unlike adult DM, increasing calcinosis in JDM is considered to be a sign of active disease.”

The authors repeated Dr. Rider’s statement that there is evidence that prolonged active disease is related to calcinosis, and that its incidence can be reduced by intervening in the disease earlier.

Once established, though, calcinosis is difficult to treat, with only isolated case reports detailing any success. For many reasons, the authors wanted to try one of the anti-TNF therapies in the young patients

who hadn't responded well to other drugs. They chose infliximab because it ensured compliance and allowed close monitoring of clinical changes and side effects.

The goal was both to control inflammation and to wean the JM children from corticosteroid therapy. All five patients had the typical JDM rash, muscle weakness and sharply raised creatine kinase (CK) levels. Four of the five patients had gotten confirmation of their disease by biopsy. All still had evidence of active disease, including progressive calcinosis. They'd all received multiple treatments, including physical therapy and drug treatment with at least two drugs.

The researchers used the combination of clinical exam and X-ray to trace the progress of the calcinosis. They found that plain X-ray films will pick up calcinosis that has not yet been discovered in the clinic, so this combination became their guide as to the current extent of calcinosis. They also used laboratory measures – creatine kinase and lactate dehydrogenase – to document muscle disease activity.

During the course of the study, they increased both the amount of the infliximab given and its frequency, after discovering that this protocol was more beneficial to the young patients.

In their summary, the researchers say the therapy produced major clinical improvement in all five cases they studied. The children who were most improved by this treatment were those with muscle weakness, calcinosis and joint contractures. The treatment was less effective on the skin disease.

“Calcinosis was still present in these cases,” the researchers report, “but notably, it was softer, painless and, in four cases, was less extensive.” In all five cases, the infliximab treatment allowed for the reduction of corticosteroids, including completely eliminating them in three cases.

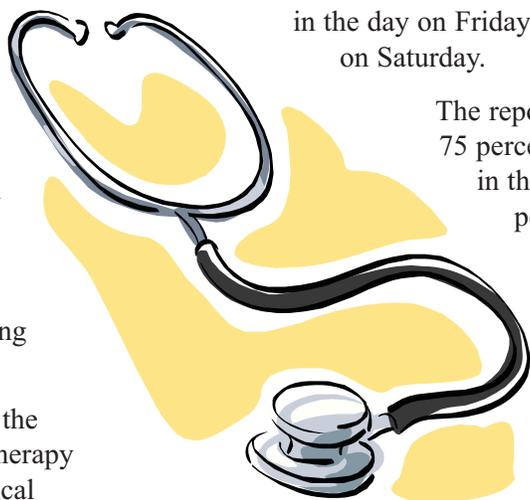
All of the five cases studied were having physical therapy, and showed a better response to the muscle building exercises after the infliximab.



Shortage of pediatric rheumatologists felt by JM families

According to a report by the Health Resources and Services Administration, an agency of the Department of Health and Human Services, the severe shortage of pediatric rheumatologists continues to plague this country.

This is not news to Lois Graves who has a five-hour round trip to appointments for her son, Tony. Graves can only manage the appointments if she schedules them for late in the day on Friday and returns on Saturday.



The report calls for a 75 percent increase in the number of pediatric rheumatologists. It also polled pediatric rheumatologists for suggestions as to how to best attract new

people to the field and help families in areas where there are no specialists.

The report found there are fewer

than 200 certified pediatric rheumatologists in this country, and that there are 13 states that do not have any pediatric rheumatologist within its boundaries. On average, children travel 57 miles to reach the nearest pediatric rheumatologist, but in some cases, the trip is much longer.

Legislation named the Arthritis Prevention Control and Cure Act (S. 626, H.R. 1283) has been introduced to authorize loan repayment programs for pediatric rheumatology, generate an increase in the number and size of institutional training grants to institutions supporting pediatric rheumatology, and expand public-private partnerships to encourage pediatric rheumatology education and training. The bill also recommends that some innovative “distance” treatment techniques be adopted, using closed circuit television and other technological tools to treat young patients.

To find the pediatric rheumatologist nearest you, go to the website of the American College of Rheumatology (www.rheumatology.com) and click on your state. There's a box at the bottom that enables you to limit your search to rheumatologists who specialize in children.

If you are unable to find a pediatric rheumatologist near your home, you may find it works to see the closest one several times a year. It's possible that you can arrange for coordination between your family doctor or regular pediatrician and the far-away specialist, so the two work together for the good of your child.

In some seriously underserved areas, adult rheumatologists have made an effort to consult with pediatric colleagues from other states on cases involving children.



Be your own boss

In Canada, teenagers with rheumatoid arthritis attend sessions called “Be Your Own Boss.” In Los Angeles, an 18 year-old diabetes patient gets reminders to take his medicine, make medical appointments and eat right delivered by cell phone text messages he arranges in advance. And all over the world, young people are educating themselves by looking up information about their health on the internet.

These teenagers, age 12-18, have decided they’re ready to take charge of their own health care, including managing their juvenile myositis. “It kind of happens naturally once you leave home,” says Ann Robertson, an adult who had JM as a child, “but it’s much easier if you start gradually to take responsibility for your medication and doctor’s appointments while you’re still at home.”

That doesn’t mean, she said, that you always have to go to the doctor alone. One young college student said she meets her mother at the doctor’s office and they have lunch or shop afterwards. “It’s helpful to have someone else with you to help you remember what the doctor told you,” she said. “And this allows us to visit.”

As you approach adulthood, you’ll face some difficult issues.

Sports

Exercise can improve muscle strength and physical function. Remember not to exercise to the point of muscle soreness. Experts say muscle soreness is a sign you’ve exercised too long or too hard. Talk to your coach and your

doctor to find the best path for you to remain active in the sports you love.

Birth control

Birth control pills and creams contain estrogen, which may have a role in flaring autoimmune diseases, so it’s a good idea for your rheumatologist to coordinate with your gynecologist. If you go instead to one of the public health clinics in all 50 states that offer confidential birth control information, make sure you note your disease and every medication you take.

Alcohol

If you are taking methotrexate, do not drink any alcohol. Discuss the reactions of different drugs and alcohol with your physician, or read further about this in chapter 32 of “Myositis and You,” available from the TMA website, www.myositis.org. Any JM patient is likely to have a more severe reaction to alcohol and drug use because of the treatment and the disease itself. An impaired liver from your disease and its treatment mean that even small amounts of toxins can affect your body more severely.

Sexually-transmitted diseases

JM patients are more likely to get these diseases, and to have more severe cases, since their natural immunity is suppressed by the medications they take for their disease. Once they contract a disease, the treatment is likely to interfere with the JM treatment. If you are sexually active, learn everything you can about disease prevention. “Myositis and You” has more information in chapter 32.

Pregnancy

Methotrexate is harmful to an unborn

fetus and can cause abnormalities in the fetus, so it should be discontinued prior to pregnancy. Talk to your doctor to see if you still need medication or if your illness could remain in remission without medications. If you do need medicine, prednisone or azathioprine would be safer to use during the pregnancy, and they should be used in the lowest doses possible. The book “Myositis and You” discusses this issue further in chapter 33.

Transition to an adult specialist

If your JM is still active when you become an adult, or if you relapse, you will fall under the care of adult specialists (rheumatologists, neurologists or dermatologists). TMA Medical Advisor Dr. Lisa Rider said in her February discussion that adult specialists tend to be a little bit less aggressive in treating the disease than most pediatric rheumatologists. If your adult specialist is not comfortable with the treatment of JM, she/he should consult with pediatric rheumatologists with special interest in JDM. You can ask your adult specialist to consult with your pediatric rheumatologist or find a pediatric myositis specialist by contacting TMA.

You can find additional information through the following resources:

www.teenshealth.com - Teens Health

www.myositis.org - TMA website

www.medlineplus.com

www.pubmed.gov

www.medlineplus.com

“Myositis and You,” available through TMA website, www.myositis.org.



Is there a bully in your school?

Sometimes kids with JM are afraid that someone will notice they look different and treat them differently. Or you may notice a child in your school is sometimes picked on by another child. How can you tell if it is bullying? Kids Against Bullying has some answers.

What bullying is

Bullying can be lots of things. They are hitting, pushing, name calling, teasing, threatening, sending mean e-mails, taking or ruining another person's things, leaving someone out and lots of other nasty stuff.

How can you tell if something is bullying?

Just remember this:

It's bullying if...

Someone is hurting another person on purpose

and

The kid who is doing it has more power.

What You Can Do

Now that you know how a bully thinks, you are in control. There's a lot you can do to protect yourself from bullying.

What a Bully Looks for?

Differences!

How You Can Protect Yourself?

Respect differences!

Everyone has differences. Just look around you. When you value and respect differences, you let the bully know that being different doesn't bother you a bit. In fact, you like it! If a bully makes fun of your red hair or wheelchair or how you talk, just smile

and agree.

"Yes, I do have red hair."

"Yes, I do use a wheelchair."

"Yes, I do stutter."

After responding, just go away from the bully.

Kids who bully want you to be upset. They're counting on it. So DON'T give them what they want. Even if you feel scared or upset, try not to show it. Crying, showing fear, or acting mad encourages the bully to pick on you again.

Kids Against Bullying has a great website, with games, puzzles and plenty of information on dealing with bullies, at www.pacerkidsagainstbullying.org.



Get ready for fall

It's still warm in many parts of the country, but it's easy to tell fall is coming! Look for grasshoppers, chrysanthemums, apples and colorful leaves.

About fall or autumn

■ Autumn is the time that comes between summer and winter. We usually call this season fall because it is a time of falling leaves. The first day of autumn (fall) is celebrated on the fall equinox - a time when both night and day are equal. That means that daylight will be around for about 12 hours and night will also last 12 hours.

■ In the northern half of the world, autumn begins about September 23. In 2008, the Autumn equinox falls on September 22. In the southern half of the world, autumn begins and ends at the same time spring begins and ends in the north.

■ After September 22, nights begin to get longer each day. The darkness will last longer than the light until the Winter Solstice in December.



Ask the doctor, from page 3.

Lisa Rider, MD: Some of the most common things people don't know about JM are:

1. That the disease can really still be active, even when the CK and a number of the other enzymes are normal, and this does require treatment.
2. That corticosteroids are best taken in the morning, not evening, to decrease the side effects.
3. That when a patient is having new calcinosis form, there needs to be an aggressive search for active disease and inflammation- immunosuppressive agents can often be helpful when calcinosis is first depositing- often the illness is active somewhere.
4. The need for adjunctive approaches- physical therapy (exercise in a graded way) and sun protection.
5. As the disease is ongoing, there is a mixture of activity (which can be helped by the immunosuppressive medications) and damage (longstanding changes, like scarring or muscle thinning (atrophy)). The damage can improve in children but will not reverse with immunosuppressive medications, but if there is a lot of damage, it can make it challenging to assess how much activity is present. Even in patients with damage, the activity needs to be treated, or the damage will get worse over time.



THE MYOSITIS ASSOCIATION

1233 20th Street, NW, Suite 402
Washington, DC 20036