THE MYOSITIS ASSOCIATION

OUTLOOK EXTRA

WINTER/SPRING 2006 Treatment Issue

Chronic disease + inactivity = pain

Remember the old prescription for pain? "Stay in bed until you feel better." Now, doctors are recommending just the opposite. Almost every chronic condition benefits from activity, they say. Staying in motion despite chronic disease helps prevent depression, fatigue and de-conditioning. Maintaining flexibility and energy has the added benefit of reducing pain, according to Dr. Edward Laskowski, a physical medicine and rehabilitation specialist at the Sports Medicine Center at Mayo Clinic, Rochester, Minnesota. He writes about the relationship between activity and pain in the Mayo Clinic publication *Housecall*.

Laskowski says exercise prompts your body to release endorphins, the chemicals that block pain signals from reaching your brain and lessen feelings of anxiety and depression. Both anxiety and depression can make chronic pain worse. Also, by building strength in the muscles that are not affected by myositis, you help spare muscles that hurt, and take some of the strain from your bones and cartilage. If you stay flexible, it means your joints are able to move through their full range of motion and are less likely to be plagued with aches and pains.

In this edition of TMA's "Treatment Extra" we provide many suggestions for exercising at your own level, with a program of aquatic therapy from the Annual Conference, suggestions for resistance and reasons to keep exercising at some level no matter what your circumstances.

Progress and promise in dermatomyositis

The past few years have advanced our understanding of dermatomyositis (DM), said Dr. Chet Oddis, allowing researchers to design meaningful trials and physicians to effectively treat the disease. Oddis, a member of TMA's Board of Directors, said one thing is certain: DM is not polymyositis (PM) with a rash. Disease targets and prospects for treatment distinguish these complex diseases. In PM, the target is the muscle fiber; in DM, it's the blood vessel.

Patients often report a DM skin rash that's as troubling or worse than the muscle weakness associated with their disease; and often it seems less responsive to treatment. Patches of redness accompanied by itching and pain appear on the face and on the knees and knuckles of DM patients. The rash is accompanied at times by extremely dry skin and sometimes also covers the scalp. Patients describe itching that nearly drives them crazy. Another, less common symptom is an inflammation in the fatty tissue in the upper arm, with swelling, redness and pain. DM is a systemic disease, sometimes affecting the lungs, the joints and the gastrointestinal system as well as the skin.

The different classifications of myositis, Oddis said, can predict both the disease symptoms and the prospects for treatment. One, called "Anti-Mi-2," includes the V-sign and shawl-sign rashes, so called because they distribute themselves across the arms and upper back like a shawl. These signs signal to the physician the probability of a good response to treatment, with a nearly 100 percent five-year survival rate.

Most cases of DM respond to conventional treatment, usually prednisone with one or more immunosuppressive agents. Intravenous immunoglobulin (IVIg) is also used in DM, particularly when there is no response or a bad reaction to prednisone.

Newer treatments borrowed from cancer therapy target the tumor necrosis factor and are called anti-TNF agents—etanercept (Enbrel) and infliximab (Remicade) are two anti-TNF agents that have been tested in clinical trials on small numbers of patients, with larger studies in progress. A multi-center etanercept trial is now underway.

Another medicine addresses itself against the surface marker of the B lymphocytes and is called a monoclonal anti-B cell agent. Rituximab (Rituxan) has been tested in a small pilot and showed promise. Now, there's a larger, multi-center study investigating the efficacy of rituximab in both adult and pediatric DM. Oddis said the trial (a double-blind, placebo-controlled trial of refractory PM and DM in adults and DM in children), still underway, has shown excellent results in 13 of 14 patients, with no serious complications. Go to www.myositis.org to find either of these trials near you.

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Dear Reader,

By now most of you have read the Winter *OutLook,* reporting on many of the 2005 Annual Conference presentations. This special *OutLook Extra* issue provides details on the sessions that focused on treating dermatomyositis, inclusion-body myositis, polymyositis and juvenile myositis.

You'll note that much of this publication is devoted to exercise; in particular, water exercise, and we hope this will inspire readers to talk to their physicians about this and other helpful forms of exercise. Including these articles along with Conference reports on new drug studies and conventional therapies is meant to emphasize the importance of exercise in the array of healing options. For those patients who do not respond to drug therapy, exercise is particularly important.

At TMA's Medical Advisory Board meeting in January, it was noted how encouraging it is to have two major multi-center drug trials now underway for myositis, with the support of two different NIH institutes. To find a center near you to enroll in either the etanercept or rituximab trials that Dr. Oddis mentions in his report on dermatomyositis, visit TMA's web site at www.myositis.org.

Families at the Annual Conference found Dr. Rennebohm's explanation of the disease process in juvenile myositis very enlightening, and we've summarized it in "Treating the JDM Mistake." We also report the latest on treating dysphagia and provide a glossary for drugs commonly used in treating myositis. In the next regular issue of the *OutLook,* you'll meet two new TMA Board members and learn more about the 2006 Annual Conference.

Until then, mark your calendars for August 31-September 3, the dates for the 2006 Annual Conference in Orlando, Florida.

Sincerely,

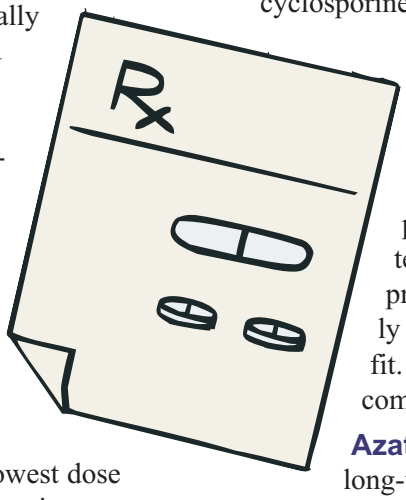
Bob Goldberg
Executive Director

A glossary of myositis drugs

Corticosteroids

Prednisone and Solumedrol are commonly used for long-term immunosuppression with fairly fast results in polymyositis, juvenile myositis and dermatomyositis patients.

Originally hailed as miracle drugs, corticosteroids featured in newsreels from the '40s and '50s show patients on crutches literally throwing them down and running across a field. Now we know there are possible side effects, particularly in high doses. Some side effects are brittle bones, cataracts, stomach upset, weight gain, and changes in blood sugar. Because of these troubling side effects, doctors generally prescribe the lowest dose possible, and for as short a time as possible. Some physicians prescribe them every other day, rather than every day, and those working with patients who experience insomnia suggest patients take them in the morning. Some patients are unable to take corticosteroids because of extreme side effects; and some have a form of their disease that does not respond to steroid treatment.



Physicians are likely to increase the dose in the event of a flare. Increasingly, doctors are prescribing combinations of drugs in order to reduce the amount of corticosteroids. There is a wide range in the size of the steroid dose that physicians prescribe, depending on the weight of the patient and the severity of the disease. Never discontinue or reduce your dose without checking with your physician.

Disease-modifying anti-rheumatic drugs

"We've stolen a lot of our drugs from other specialties," Lawrence Phillips, MD, told participants at the Annual Conference. None of the drugs used in myositis treatment were originally developed for the inflammatory myopathies. Hydroxychloroquine (Plaquenil) was developed to combat malaria; methotrexate and chlorambucil were borrowed from oncologists who used it for treating cancer; and cyclosporine was developed by transplant specialists as an anti-rejection drug.

Cyclosporine A is an alternative to prednisone for long-term immunosuppression and relatively rapid onset of benefit. Side effects are not common.

Azathioprine provides long-term immunosuppression with few side effects. It may be useful to reduce needed doses of corticosteroids, but there may be a long interval before improvement.

Methotrexate generally works a bit faster than other medicines, and can be useful when corticosteroids or Cyclosporine A are ineffective or cause side effects. With routine monitoring, serious side effects are uncommon.

Cyclophosphamide is used in cases with life threatening features and in B-cell mediated disorders that respond to few other treatments. It can have serious side effects.

Biologic response modifiers

These types of drugs were borrowed from rheumatoid arthritis treatment, and work well in some people. They work by inhibiting cytokines, key players in some myositis inflammation. Some used by myositis patients are **etanercept** (Enbrel) and **infliximab** (Remicade). These drugs are very expensive and many insurance companies do not pay for them. They provide a more targeted treatment and are being studied in several clinical centers. For more on biologics under study, see page 1.

Plasma exchange (PE) and **human immune globulin (IVIg)** are used for rapid onset, short-term benefit when patients have life-threatening signs such as respiratory insufficiency, dysphagia, or severe weakness. PE and IVIg are expensive but are occasionally used for chronic therapy when other treatments have failed.

Novel Agents

Rituximab (Rituxan) may be useful in neuropathies with associated serum IgM autoantibodies. Over the short term rituximab appears to have few side effects. (See more on rituximab, page 1.)

Mycophenolate mofetil may be used for long term immunosuppression as the primary agent or to spare toxicity due to other medications.

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In this issue:

Aquatic exercise: Perfect for all abilities

Carrie Baldwin is a certified aquatic therapy and rehabilitation instructor who works with chronically ill patients.

Go to a pool these days and you're likely to see a number of people stretching, bending, walking and even marching through the water, along with those swimming laps. They're exercising, taking advantage of the qualities of the water to protect their joints, cushion their backs and increase resistance.

"That's because of the basic nature of water," said Carrie Baldwin, speaking at the Annual Conference in Cleveland. "Water is hundreds of times more supportive than air. It's the force that acts in opposition to gravity. That's why we can float." At the same time, the density of the water provides a gentle resistance, so the free movement of limbs or trunk through the water requires a great deal more strength than the same movement through the air.

One reason that water exercise is ideal for those with weak muscles is the capacity of the water to help bear their weight, Baldwin said. The deeper the water, the less your own body weighs you down. If you're in the deep end, with water lapping at your chin, you're bearing the equivalent of only 10 percent of your body weight; if you stand with the water about waist high, that changes to 50 percent. The effects of this sudden weight loss are obvious for myositis patients: they can move more freely, and the support of the water allows them to exercise without the fear of falling down.

Besides buoyancy, the water provides a type of consistent pressure—called hydrostatic pressure—against your skin. This pressure is exerted by water molecules on any submerged surface and increases with the depth of the water, so there is greater pressure at your feet than at your waist if you're standing in chest-deep water. The pressure acts a bit like a support stocking, gently squeezing to return blood from the legs. This helps your circulation and decreases swelling. An added bonus for those who dread the post-workout muscle soreness is that water exercise drastically cuts down on this common side effect to vigorous exercise.

Although the pool makes exercise easier in many ways, you are actually working harder when you work against the resistance of water: twelve times harder, Baldwin said. Water provides twelve times the resistance of air, in all directions and against every movement, which is good news for those hoping to gain strength. "Water provides a very balanced workout," Baldwin said. "It works the muscles evenly." An added bonus is your posture during exercise: while the resistance slows you down, you are more likely to learn the correct way to do the movement. She advises those building strength to increase resistance as they are able, by increasing speed, changing direction and adding equipment.

While you're having fun, strengthening your muscles and improving your balance you're also helping your heart, Baldwin said. As the hydrostatic pressure of the water improves the flow of blood to your muscles, it also improves the blood flow to your heart. With each beat, your heart is pumping out a greater volume of blood so it does not need to pump as fast to get blood to exercising muscles.

Researchers think this is one of the reasons people experience a heart rate on average that's 10% lower when they exercise in the water. You're helping your lungs, too, once you slip into the deep end: The work of breathing increases by about 60% with immersion to the neck because the pressure on the muscles helps you in your breathing. This is a wonderful benefit of water exercise, except in the case of someone with extreme pulmonary weakness, Baldwin said. The increased blood flow also promotes kidney (renal) health.

Some considerations, before you head for the pool:

- What's the right temperature? Opinions vary, Baldwin said. It is very difficult to come up with an "ideal" water temperature because there are so many different populations, ages, and body mass types. Therapeutic pools are usually kept between 84-94 degrees Fahrenheit (F). Community-based lap swimming pools are usually 80-82 degrees F. Warm water relaxes the muscles, decreases chances of swelling and is generally comforting.
- Air temperature should ideally be a few degrees warmer than pool temperature, humidity around 50%.
- If your illness requires aquatic physical therapy (aquatic exercise taught individually by a licensed physical therapist), look for a therapist with advanced training in aquatic therapy techniques from the Aquatic Therapy and Rehabilitation Institute (866-462-2874 or www.atri.org). This level requires a program designed for you and your specific needs, requires a referral from your physician, and is

SWIM AROUND PAIN

University of Alabama geriatrician Andrew Duxbury, MD, offers a tip for those who want to exercise and are on pain medication. "Pain medication should be taken about half an hour prior to exercise so it is peaking in the system during exercise," he said.

usually paid for by insurance if you have a diagnosis considered appropriate.

- If you're seeking a regular aquatic exercise class, look at local health clubs, YMCAs, public pools, rehab centers, sometimes even hotels.
- There are all kinds of specialty classes: arthritis classes, water aerobics, toning, Ai Chi (aquatic Tai Chi), water Pilates, and many other specialties.
- Ask about instructor training and experience in dealing with special populations.
- Check out the pool before your first lesson. Determine depth, temperature (air and water), accessibility (ladders, ramps, lift chair, railings), floor (will pool shoes be necessary for better traction?), locker rooms, lifeguard, water quality and acoustics.

- Water exercise may not be for you if you have active illness, fever or infection, bowel or bladder incontinence, open wounds, skin rashes, uncontrolled seizures, severe cardiovascular disease, unstable angina, or excessively high or low blood pressure. If your own fear of water is a factor, Baldwin recommends starting with an aquatic physical therapist offering one-on-one assistance.
- Once you've become comfortable with water exercise, you may want to increase the intensity of your workout. Baldwin recommends speeding up, working harder. Or move away from the wall and perform exercises freestanding to increase challenge to balance; or add equipment. Baldwin demonstrated wonderful water "props": barbells, "noodles," balls and floats.
- Don't forget: Never swim alone. Never.
- Always consult your physician before beginning any exercise program, Baldwin cautions. Remember to listen to your body and slow down if you need to. Plan ahead to save enough energy to get out of the pool, change, and go home. Don't work to fatigue—it is very easy to overdo it in the water. If it hurts, slow down or stop.
- BREATHE.

Why water?

Buoyancy and resistance provide opportunities for strengthening, says TMA medical advisor

By MICHAEL HARRIS-LOVE, DSc, MPT, CSCS

Dr. Harris-Love is a member of TMA's Medical Advisory Board and assistant professor for health care sciences at George Washington University in Washington, DC.

Various forms of aquatic rehabilitation have been employed for more than a thousand years, but we seldom read about this method of therapeutic exercise for the treatment of myositis.

A question that immediately comes to mind is, "What took us so long?" Aquatic exercise is perhaps most closely associated with conditions marked by significant joint pain, such as arthritis. In recent years, the scope of diagnoses deemed appropriate for aquatic exercise has expanded to include post-polio syndrome, multiple sclerosis, and other complex movement disorders.

Many physical therapists have come to recognize that a key characteristic of water—buoyancy—is ideal for strengthening people who have significant muscle weakness. Carrie Baldwin, a physical therapist with an aquatic therapy practice in Northern Michigan, notes an interesting paradox in her presentation above: movement in the water may be easier, or more difficult, than land-based activity. The interaction of water buoyancy and resistance allows for slow movements to occur with little effort, while rapid movements are part of a more challenging workout.

Physical therapists manipulate these qualities of water for the purposes of rehabilitation and create exercise programs appropriate for people with various levels of ability. It should be noted that additional work needs to be done to better understand the effectiveness of aquatic exercise for individuals with myositis.

Also, it is important for a plan of care involving aquatic rehabilitation to be linked with land-based goals and functional outcomes. However, the research findings concerning aquatic exercise for people with neurological and rheumatic conditions suggest that this type of physical activity may help to improve strength and cardiovascular fitness. Through the advocacy of dedicated clinicians like Ms. Baldwin, and the focused efforts of investigators and clinical trial participants, aquatic exercise may begin to play a more prominent role in the rehabilitation of people with myositis.

For more on resistance, see page 12.

For fun, strength and relaxation, just add water

Carrie Baldwin recommends some of the following aquatic exercises. Please note that they are for general purposes only and are not intended to diagnose or treat any specific illness or replace exercises recommended by your physician, therapist or health-care provider. As always, consult with your physician before beginning any exercise regimen.

Warm up

Do these exercises in waist- to chest-depth water, using opposite arms and legs. To work on balance, you may also do them backwards!

- **Regular water walk.** Opposite arm moves with opposite leg. Heel strikes the floor first, then ball of foot, then toes when moving forward, and the reverse when moving backward. Try various swimming arm patterns, like crawl stroke, breaststroke, and straight-arm pull.
- **March.** Opposite arm moves with opposite leg, landing on flat foot. Can vary arm patterns here, too.
- **Sidestep.** Arms move upwards towards the surface as legs step apart sideways, and then pull back to midline together.
- **Lunge.** Take a large step forward, bending at the hips and knees. Do not let your knee bend forward beyond your foot. Move opposite arm with opposite leg.
- **Crossover.** Step one foot over and across your other leg, then bring the leg from behind, out and to the side. Alternate stepping in front and then in back of your opposite leg while moving sideways, as in the "basket weave step."

Stretch

Once you've warmed up, try some gentle stretches against the pool wall.

- **Hamstrings.** Stand with your back against the pool wall and lift your leg toward the water's surface, keeping your knee straight and letting the water support your leg. Try to keep your hips back against the pool wall, and pull your toes up towards your body. You should feel this stretch along the back of your leg.
- **Heel cord.** Stand facing the pool wall, then step backwards with one leg, pressing your heel down against the floor. Lunge forward into the water until you feel a stretch up the back of your calf on the back leg.

Leg strengthening

Only work within your comfortable range of motion.

- **Squats.** Stand facing the pool wall and bend at the hips and knees like you are going to sit down in a chair. Try not to let your knees bend forward beyond your toes.
- **Toe raises.** Push up your toes and tighten the muscles on the back of your calves.
- **Hamstring curl.** Lift your heel up behind you like you are trying to touch your buttocks, tightening the back of your thigh. Keep your knee pointed toward the pool floor.
- **Knee lift.** Lift your knee like you are marching and use a noodle (a flexible bouyant device shaped like a giant macaroni) under your foot for increased resistance.
- **Straight leg kick.** Keeping your leg straight, kick up towards the water's surface and then pull back down. Use a noodle for increased resistance.
- **Side leg kick.** Keeping your leg straight, kick sideways and then pull back in. Use a noodle for increased resistance.
- **Circles.** Keeping your leg straight, circle your leg from your hip in both directions. Use a noodle for increased resistance.

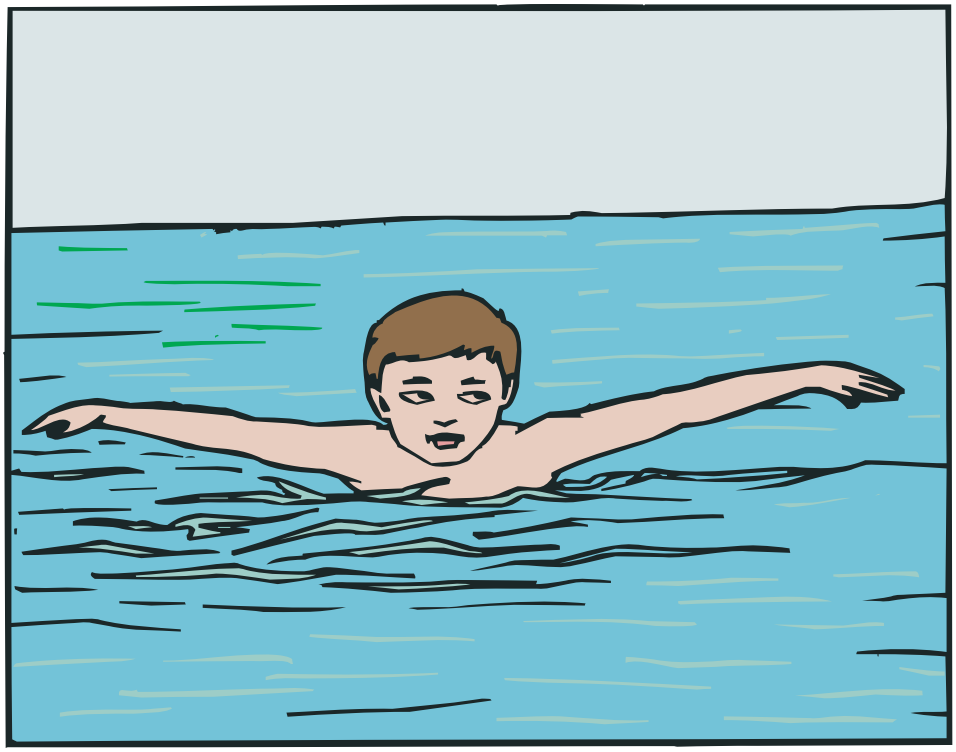
Abdominal strengthening

In a seated position against the pool wall, perform a pelvic tilt and flatten your lower back against the pool wall. Hold a noodle on the water's surface. Push the noodle just below the water's surface, maintaining your low back against the pool wall. You should feel your abdominal muscles tighten to hold your trunk steady.

Shoulder strengthening

Only work within your comfortable range of motion, and try to keep your shoulders at or below the water's surface.

- **Shoulder flexion and extension.** Keeping your arms straight, lift them up towards the water's surface in front of you and then return to your sides.
- **Shoulder abduction/adduction.** Keeping your arms straight, lift them up towards the surface to the side and return back down to your sides.
- **Horizontal abduction/adduction.** With your arms straight in front of you just under the water's surface, push them outward until they are straight out from your sides, then return to the front.
- **Circles.** Circle your straight arms in both directions. Movement is from your shoulder, and arms can be in front or to the side.
- **Chest press.** With your arms straight out in front of you, bend your elbows, pulling them back towards your sides. Squeeze your shoulder blade back towards your spine in a rowing-type motion.
- **Internal/external rotation.** Press your arms against your sides, with your elbows bent at 90 degrees. Keeping your elbows at your side, rotate your forearms outwards and then inward.



Children look forward to their turn in the pool

Water therapy for children

Most children enjoy swimming and splashing around. For children with myositis, water exercises can be especially valuable, both physically and emotionally.

Physically, JM children experience muscle weakness, often accompanied by muscle pain and joint stiffness. Studies have shown that water therapy is effective in increasing joint flexibility, reducing pain, and decreasing the difficulty children may have performing daily tasks. Daily tasks for children—playing, running, sports, and more—are important to their feelings of normalcy. Water therapy may also help lower or prevent the undesirable and discouraging weight gain that is common with prednisone use.

Making it work

Children often dread the thought of doing exercises and rehabilitation day after day, leading to a lack of enthusiasm and negative overall feelings. Parents may struggle to convince children to stick to it. Therefore, participating in an aquatic therapy program—one that may not feel quite like a traditional exercise regimen—can in effect keep your child interested enough to stay loyal to the program for the long term. The social interaction that comes with swimming benefits children as well.

"Aquatic therapy provides an opportunity to improve strength, movement patterns, communication, and functional skills in a warm and relaxing environment," says Jeanne Baldwin, DPT, Physical and Aquatic Therapist at Providence Neurodevelopmental Center for Children. Therapists work with families to develop a program that is appropriate for each child and then families can carry out these programs on their own.

Though JM children should avoid the sun, there are ways around this obstacle. Families have found local YMCA and community pools available for their children to use; some have been allowed to swim in local hotel pools. Other families swim in the evenings to avoid the harsher rays of the sun during the daytime hours.

WHAT DOES THAT MEAN?

For those of you who have lived with JM for a while now, the terms used in conjunction with therapies, medicines, and other aspects of JM are familiar. They may not be, however, for families who are new to JM. Here is a guide to the terminology that you may hear over the course of your child's JM treatment:

Calcinosis: hard, often painful lumps of calcium that form under the skin's surface, especially in juvenile dermatomyositis.

Pulse: dose of medicine given intravenously (through a needle) over a short period of time, in many cases to "jump start" the treatment.

Refractory: resistant or unresponsive to treatment.

Remission: period of time when a patient shows no symptoms of disease and has been off all medicines for a period of six months or longer. The definition of remission varies some from doctor to doctor, and some specialists consider patients on low doses (or maintenance doses) of medicine to be in remission.

Taper: process of slowly lowering the dosage of medicine to reach a maintenance dose or to stop taking the medicine completely.

Ulcerations: breaks in the skin or mucous membranes resulting in the loss of tissue and disintegration of endothelial tissue. Ulcerations of the skin and gastrointestinal tract are possible in JM though not common.

Vasculitis: swelling of blood vessels under the skin that causes a visible rash.

Visit the Juvenile Myositis pages of TMA's web site at www.myositis.org for more information and materials specific to JM children and families.

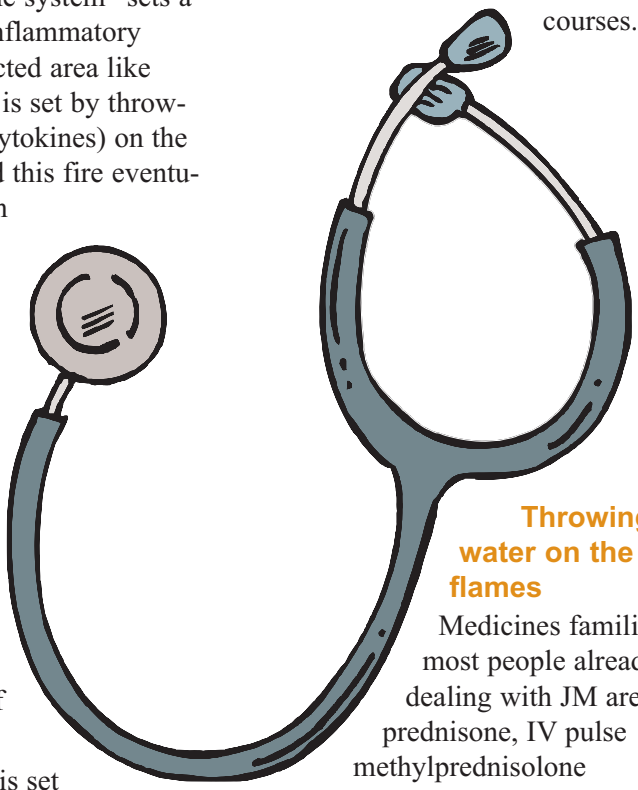
Treating the JDM mistake

Robert Rennebohm describes what happens with children's immune systems in juvenile dermatomyositis (JDM) as *the JDM Mistake.* "It is inappropriately and unnecessarily attacking the endothelial cells in the small vessels that feed the child's muscle and skin," said Dr. Rennebohm, Associate Professor of Pediatrics at The Ohio State University College of Medicine and Chief of the Division of Pediatric Rheumatology at Columbus Children's Hospital.

With an appropriate immune response, our immune system "sets a fire," or creates an inflammatory response, in the infected area like your throat. The fire is set by throwing more wood (or cytokines) on the fire in the throat, and this fire eventually kills the virus. In return, the throat becomes red, inflamed and sore, but this is accepted as a part of the normal healing process. The immune system then stops throwing wood on the fire, and the throat returns to normal in an acceptable and appropriate period of time.

In JDM, this fire is set in the small blood vessels of the muscle and skin. "The immune system gathers as much fuel as it wants," Rennebohm said, "and fuels the fire at whatever level of intensity it wants." Though the immune system may think it is acting appropriately, it is mistaken.

"We do not know how to reform the fire-setter," Rennebohm said. "Fortunately, though, the immune system is capable of self-correction." When the system actually corrects itself varies from patient to patient and typically happens only gradually. Until this self-correction happens, doctors "throw water" on the fire, firewood and fire-setter in the form of medicines. The challenge is to find the appropriate amount of water, or medicine, to use depending on the child's individual course. A mild course, for example, does not need excessive amounts of medicine; and continuing treatments during flares in a polycyclic course will unnecessarily add to the side effects. (See *The Different Courses of JM* for an explanation of these courses.)



Throwing water on the flames

Medicines familiar to most people already dealing with JM are oral prednisone, IV pulse methylprednisolone (SoluMedrol), methotrexate, and intravenous immune globulin (IVIG). "It is always a guess as to how much we can taper the prednisone," Rennebohm said. Luckily, it is an educated guess, using measures of muscle strength to determine the best tapering schedule.

If a child is not tolerating the prednisone tapering, steroid-sparing medicines are introduced; otherwise, too much prednisone is needed. Methotrexate is increasingly introduced sooner rather than later in therapy, except in mild cases. Other medicines used, depending on the disease severity and symptoms, are etanercept (Enbrel), cyclosporine A, IV pulse cyclophosphamide, and rituximab (Rituxan). Cyclosporine is often used for those children experiencing a chronic continuous course with a lot of skin disease; cyclophosphamide for ulcerations.

"What we need to develop," said Rennebohm, "is a new therapy that either totally disables or totally reforms the fire-setter. This would convert every child's disease into a very brief, harmless monocyclic course." Research continues into the different subsets of JDM, to find out the most important mistake the child's immune system is making to better understand how to stop, or even prevent, that mistake.

Medicines used to fight JM

Lisa Rider, MD, Deputy Chief, National Institute of Environmental Health Sciences at the National Institutes of Health, agreed that finding the best treatment for each individual child is a process rather than a fixed prescription. As has been the case for years, daily oral corticosteroids like prednisone remain the mainstay for treating JM, she said. Typically, prednisone is given at 1-2 mg/kg/day ("mg/kg/day" refers to number of milligrams per kilogram of your child's body weight given daily). There are a number of complements to this standard treatment, Rider said: sunscreen and avoiding the sun as much as possible, topical corticosteroids, calcium and vitamin D, and rehabilitation. All of these therapies make up the typical first line of attack against JM.

For especially frustrating itches, moisturizing creams or antihistamines may be introduced, along with topical medicines like tacrolimus (Protopic) or pimecrolimus (Elidel).

Antihistamines are often prescribed to treat allergies, and these medicines may combat more difficult skin symptoms in JDM. Dapsone, an oral medicine used to treat other skin conditions, is a possibility for those children with severe skin disease.

Skin disease that is unresponsive to some of the first-line treatments may require methotrexate, intravenous immune globulin (IVIG), cyclosporine, or mycophenolate mofetil. Some research supports the use of these medicines; however, this research is limited and larger studies are needed. Studies of methotrexate, for example, have shown improvement in disease activity and remission in some children. The initial effects of methotrexate may take four to six weeks, with peak effects taking up to several months, Rider noted. It's important to weigh benefits and risks, as methotrexate is a strong medicine with possible side effects of mouth sores, increased sensitivity to the sun, nausea, and increased chance of infection. Doctors report that injectable methotrexate is better absorbed by the body than oral methotrexate (pills).

Testing IVIG, researchers found that 1-2 g/kg/day (grams per kilogram of body weight per day) for two days a month over a period of three to nine months led to approximately 30% of the subjects normalizing, 65% showing improvement in strength and rash, and several resolving or improving their calcinosis. Side effects were minimal and mostly infusion-related, though the high expense of these infusions can be prohibitive as well.

Cyclosporine is not used as often as some of the other medicines introduced in JM, but for refractory JM, or treatment-resistant cases, cyclosporine has been successful in improving children's strength and allowing them to decrease their corticosteroids. Most of the side effects (like hair growth, kidney problems, and high blood pressure) are reversible.

Becoming more common is the use of these once second-line agents, especially methotrexate, at disease onset instead of waiting. Rider cited two studies suggesting the benefits of introducing medicines along with prednisone:

- Oral prednisone and methotrexate versus prednisone alone: less corticosteroid exposure, therefore fewer side effects.
- Intravenous pulse corticosteroids and methotrexate versus oral prednisone alone: more rapid improvement in strength, function and rash, and less calcinosis.

When should these second-line agents be introduced? In the past, these medicines were commonly used for children whose disease doesn't respond to corticosteroids, when children become dependent on corticosteroids, when there is an unacceptable level of steroid toxicity, or when children do not reach remission after ongoing disease activity. More and more, they are part of the initial treatment plan as doctors aggressively fight JM while trying to avoid some of the corticosteroid-related side effects.

THE DIFFERENT COURSES OF JM

"When your child is first diagnosed," Rennebohm said, "it helps to know the spectrum of the disease but also to know the most likely course your child will follow." Early on in the disease course, this is difficult to determine.

Three factors—intensity, duration and fluctuation—explain why doctors see three main clinical courses of JM:

- monocyclic (single cycle)
- polycyclic (many cycles)
- chronic continuous

Within these categories are mild, moderate and severe versions of the disease. In terms of physical activity, children who have no strength to get up have a severe version, children who can lift up for only short periods of time have a moderate version, and children who can maintain a lift for about two minutes have a mild version.

"By definition," he said, "all patients who follow a monocyclic course go into remission and are off all medications within two years after the onset of the JDM." The JDM mistake is not made again. These children's immune systems are capable of correcting the JDM mistake within a year or two.

In a polycyclic course, children experience two or more cycles with full remissions between these flares of the disease. Like those children following a monocyclic course, their immune systems are able to correct themselves but make the same mistake again later.

As the name suggests, chronic continuous courses continue for more than two years without periods of remission, though there can be periods where the mistake is made to a lesser degree. The child's immune system is not as capable of correcting the mistake.

Under the current aggressive therapy for JDM, about 40% of patients follow a monocyclic course; 10-20% polycyclic; and 40-50% chronic continuous. Within these categories, 40% will experience a widely fluctuating course, 40% constant, 10% predominantly cutaneous (skin involvement), and 10% ulcerative (ulcerations of the skin or gastrointestinal tract). The ulcerative course is the least common but most severe and is the most difficult to deal with as both the patient and doctor.

Treatment, tools and techniques for swallowing

Something formerly accomplished without a thought becomes an ordeal for myositis patients with dysphagia, a condition defined as trouble with any phase of the swallowing process.

Presentations at The Myositis Association Annual Conferences allowed members to listen to and question the experts about dysphagia. Violet Cox, MS, CCC-SLP, Clinical Coordinator of Speech and Audiology at Cleveland Clinic Health System, asked patients at this year's Conference about their first clue that this basic function wasn't going smoothly: "I can't eat in a hurry anymore," one TMA member replied.

A review of the swallowing process

Understanding what's involved in a "normal" swallow helps us understand more about what happens in myositis patients who have difficulty swallowing. Swallowing is a complex process, involving three stages and dozens of muscles and nerves. Cox explains:

Stage One: In the first stage, the tongue manipulates the food in the mouth for chewing, and saliva mixes with the food to soften and moisten it. In healthy people, the saliva starts even before the first bite is taken, in anticipation of the meal. Chewing makes the food the right size for swallowing. This stage is voluntary and usually remains normal even in people with muscle weakness.

Stage Two: The tongue pushes the chewed food to the back of the mouth, triggering the swallowing reflex. This is where the process becomes automatic. Food goes from the mouth to the esophagus through the pharynx, and the airway closes to prevent food or liquid from entering the lungs.

Stage Three: Food enters the esophagus and passes through the canal to the stomach.

It's a complicated system, said Todd Levine, MD, Director of Samaritan ALS Clinic and Director of the Department of Neurophysiology, Good Samaritan Hospital, Phoenix, AZ. The trachea, which passes air to your lungs, and the esophagus, which passes food to your stomach, are very close together. Because the swallowing process is so complex and involves both voluntary and automatic actions, complicating it with a muscle disorder quite often leads to the system breaking down in some way.

Disrupting the normal swallow

A normal swallow takes between one and three seconds, said Michelle Reuthers, a speech therapist. When your muscles become weak, this timing slows down. Your dysphagia symptoms will depend on where in the process you have trouble. Don't be surprised if your symptoms vary from those of someone else. Your treatment will also depend on your own unique set of circumstances.

Do you have dysphagia?

Signs and symptoms of dysphagia are:

- Pain when swallowing
- Inability to start the swallowing process
- Persistent coughing or choking while swallowing
- Low-grade fevers
- Sensation of food stuck in the throat or chest
- Frequent heartburn

Parents of children with swallowing problems notice them being distracted during meals, tensing their bodies, taking more than 30 minutes to finish, leaking or spitting foods or liquids, and coughing or gagging during meals. Poor weight gain or growth may also be evident in children.

Many adults first realize they're having problems when it takes longer than usual to eat, Levine said. If you need five or ten swallows for each bite of food, that's too many.

Signs of dysphagia are often more obvious when you're eating out, Reuthers said, as this is a time when you're not paying as much attention to your food. It's especially important to concentrate on eating when you have any swallowing problem.

Diagnosing swallowing disorders in myositis

There are a number of diagnostic tools available, but Levine finds one tool in particular is the most effective when he diagnoses dysphagia. He has his patients drink water, then say their names. If their voice sounds wet, like they're talking under water, there is a problem. In this situation, the water is trapped in the top of the esophagus at the trachea, where the vocal chords are located.

Doctors and speech-language pathologists are trained to test for and treat swallowing problems. The most common test is the modified barium swallow, or videofluoroscopy. Reuthers encourages myositis patients, or those thought to have a swallowing problem, to have a modified barium swallow. This test looks at the entire swallowing process and shows the therapist exactly where the procedure runs into trouble. Knowing if the problem happens before, during or after the swallow tells doctors what will be of greatest benefit. Remember that swallowing problems don't happen all the time, so if you notice certain foods or textures continuously causing problems, bring them with you for the test. Otherwise, Reuthers said, the test may mistakenly show a perfect swallow.

Help for dysphagia

Treating dysphagia begins with changing what you eat and how you

eat it. Though it may not seem logical, Reuthers said, thinner foods and liquids are not easier to swallow, as they are difficult to control in the mouth. Thicker consistencies are easier to control during the swallowing process. Even with thicker consistencies, different people will require different levels of consistency. Thickeners are available in different consistencies, called nectar, honey and pudding. Nectar is similar to tomato juice in consistency, honey like a soft gel, and pudding is thick, Cox said. These additives are generally tasteless, although it's difficult not to taste them when they're added to water. Thickening liquids slows them down in the swallow, giving you a better chance to control and complete the swallow correctly.

Health professionals and myositis patients with experience with dysphagia share helpful hints:

- Tuck your chin down—this shortens the distance for the food or liquid to travel and protects the airway.
- Turn your head to the weak side to shut it off, using the strong side to swallow.
- Find what types of food stimulate a swallow for you. Cold, tart beverages may work for some, while spicy foods work well for other people.
- Spray your mouth to moisten it before taking a drink of water. Keep your mouth from becoming dry throughout the day by using the spray bottle or sucking on mints.
- Make your own "nosey cup" by cutting a piece out of the top of a Styrofoam cup for your nose to fit while tipping the cup.
- Eat less food more often, especially when eating a meal takes more time for you than for other people. Try eating six smaller meals each day.
- Drink small amounts of liquid often enough to keep the hypopharynx (the "pocket" at the base of your tongue near the epiglottis) clear.
- Take smaller portions of food and liquid at a time, and make sure your mouth is clear before taking any more.
- Focus on eating—avoid distractions and don't eat when you're overly tired.
- Swallow when you feel the urge to clear your throat. Swallowing is a smoother action than clearing your throat and won't irritate your vocal chords.
- Avoid foods that cause the most difficulty. Try to find which foods cause problems and which stimulate your swallow.

If these strategies don't help, there are surgical procedures to consider. For esophageal dilation, a dilator is inserted into your esophagus to stretch the opening when it is too narrow. Crycopharyngeal myotomy is a bit more invasive in that a small cut is made in your muscle to allow food to pass through more easily.

When you're losing weight because of swallowing problems, Levine encourages you to consider a feeding tube. Also called a G-tube or PEG tube, this is often seen as a last resort or an extreme measure. Levine doesn't consider this to be the case. The PEG tube is less invasive than many people think, agreed Dr. Lawrence Phillips, II, Professor of Neurology at the University of Virginia School of Medicine. It secures itself by means of a balloon, he said, and is easily removed by deflating the balloon. The tube allows the food to bypass the swallowing phase of eating altogether, as it is inserted directly into the stomach. This is then your main source of nutrition, but you can still eat small amounts to enjoy the sensory pleasures of eating. Levine especially recommends feeding tubes for his patients who have recurring pneumonia (see *Aspiration Pneumonia,* page 12), who are losing weight, and for whom eating isn't enjoyable anymore.

Dr. Phillips explained that, in some cases, throat surgery is a good solution for myositis patients with swallowing difficulty. "It releases the pressure," Phillips said. "The upper third of your swallowing mechanism is largely skeletal muscle, and that's where most myositis patients have the problem." The lower two-thirds are smooth muscle. In contrast, he said, dogs have skeletal muscle all through their swallowing mechanism and that's why they and other animals have more voluntary control over their swallows.

Trends in research

Swallowing troubles aren't limited to those with myositis, so researchers studying these problems have a larger pool of patients to observe. Some studies highlight the use of botulinum toxin A in treating dysphagia. This is still an experimental treatment and Levine points out that a larger study in myositis patients would be of help in understanding long-term effects.

One study looked specifically at two inclusion-body myositis patients, injecting botulinum toxin A (known as Botox) into the upper esophageal sphincter. This Canadian study reported that this treatment successfully alleviated esophageal sphincter pressure and dysphagia in both patients. Other studies shared similar results. A team of researchers in New York stated: "Injection of Botox A in the office should be considered when there is failure of the cricopharyngeus muscle to relax after the swallow, significant pooling in the cricopharyngeal region, and a risk for penetration and aspiration." An Italian study noted that patients with mild impairment shown by videofluoroscopy and a higher pressure of pharyngeal contractions best predicted response to Botox treatments. If Botox treatment is unsuccessful, surgery remains an option.

For resources on living with dysphagia, go to www.myositis.org.

In or out of the water, resistance is the key to building strength

Resistance training—strength-building exercises like weightlifting—has been a consistent focus of those who exercise. Resistance training builds muscle and prevents bone loss, injury and weight gain. The Mayo Clinic reports that for people who must avoid resistance training because an injury or chronic health condition makes weightlifting impossible, there are several effective alternatives. One is aquatic exercise (see page 4). For those who cannot use water for resistance, there are still others:

Isometric exercise uses your own body for resistance. For example, stand with your back to a wall and press the back of your arm into it. Hold for three to five seconds and repeat 10 times on each side. If you have hypertension, check with your doctor before trying isometric exercise. Some doctors believe it may raise blood pressure.

Pilates is a system of exercises derived from dance and ballet moves. Exercises are performed on the floor or with special machines, and are designed to strengthen the muscles of your lower back, abdomen and buttocks.

Resistance bands are latex bands that offer resistance by stretching. The more you stretch them, the more resistance they offer. If used incorrectly, they can irritate injuries, so learn the proper form before trying exercises and be sure to buy the right bands for your fitness level. People who are allergic to latex can look for latex-free versions.

Dysphagia puts patients at risk for aspiration pneumonia

If you have trouble swallowing, aspiration pneumonia is a major concern. Aspiration means that foods or liquid are going into your lungs instead of following the correct path down the esophagus. These small particles can lead to aspiration pneumonia, or swelling in the lungs and bronchial tubes.

There are two basic types of aspiration: silent (covert) and overt. In silent aspiration, you don't know you've taken food or liquid into your lungs. After some time, you may end up with a fever, but this is the first sign of a problem. On an x-ray, this shows up in the right side of the lung, said Violet Cox, MS, CCC-SLP, Clinical Coordinator of Speech and Audiology at Cleveland Clinic Health System. With overt aspiration, the food entering the lungs causes you to cough and/or choke immediately.

Some symptoms of aspiration pneumonia are coughing, wheezing, fever, and shortness of breath. Antibiotics are typically used to treat aspiration pneumonia, and for those with swallowing problems, doctors may assess swallowing function and discuss alternative feeding methods, if needed.

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