

## Two perspectives on myositis treatment

"The goal of therapy in inflammatory myopathies," says Dr. Marinos Dalakas, "is to improve muscle strength and activities of daily living." Dalakas, Chair of TMA's Medical Advisory Board and Chief of Neuromuscular Diseases section of NINDS, NIH, notes that when muscle strength improves, CK levels drop; however, the reverse is not always the case because immunosuppressants can lower CK levels without having a positive effect on muscle strength. This is sometimes misinterpreted as an improvement, he says, so doctors will erroneously follow the CK levels instead of the muscle weakness.

### Corticosteroids

To begin treatment, Dalakas prefers high-dose prednisone at 60-80 mg/day for 3 to 4 weeks followed by a tapering schedule over a 10-week period. His goal is to reach 60-80 mg/day on an alternate-day schedule at the end of the 14 weeks. If prednisone is effective and doesn't produce excessive side effects, he reduces the prednisone by 5-10 mg every 3 to 4 weeks until reaching the lowest dose possible to maintain stability. If, however, there are no signs of improvement by the end of the 14-week taper, he considers this patient unresponsive and tapers quickly to begin immunosuppressive therapy.

Dr. Chester Oddis suggests a divided daily dose at 60 mg/day for PM and DM patients. Once CK

*See Perspectives, page 3*

## Myositis 101: a beginner's guide

What is myositis? What causes it?  
How do I know that's what I have?  
What should I use to treat it?

When you're diagnosed with any form of myositis, the questions seem endless. At the Annual Conference, Dr. Fred Miller, Senior Investigator and Chief, Environmental Autoimmunity Group, NIEHS, at the National Institutes of Health, explained myositis in straightforward terms and basic concepts for those just learning about their conditions.

### What is myositis?

The general definition of myositis seems basic enough - "myo" means muscle and "itis" means inflammation or swelling. But there's much more to it when referring to the chronic form of myositis. Myositis is the general term used in dermatomyositis, polymyositis and inclusion-body myositis; but in medical terms, these conditions are more specifically referred to as idiopathic inflammatory myopathies (IIMs), or diseases of the muscles with inflammation but no known cause. Breaking it down into the specific disease types, polymyositis (PM) means affecting many muscles; dermatomyositis (DM), involving the skin; inclusion-body myositis (IBM), with inclusions - a kind of cellular hole showing up in the muscles; and juvenile myositis (JM), occurring in childhood, typically at 18 years of age or younger.

Even the more homogeneous groups of PM, DM, IBM and JM have a number of factors that cause different people to show symptoms in

various ways, and it's important to know more about each case to plan the most effective course of treatment. The dilemma lies in the fact that different patients are affected in almost unique ways, says Miller. Some symptoms develop slowly, others quickly; people experience different levels of muscle weakness and pain; and about 30 percent have joint involvement, he says. Many symptoms are shared by other diseases, often leading doctors to other possible diagnoses first.

### How do you treat myositis?

First, make sure the diagnosis is correct. There aren't clear-cut lines between the different forms of myositis as well as other diseases: "One size doesn't fit all for myositis," Miller says. This is still the art of medicine, not yet the science. Some variables to consider: how much is disease activity or active inflammation versus disease damage or scarring? What are the risks and benefits of particular drugs used to block the immune system? In forming a treatment plan, he recommends a holistic approach, taking into account your own expectations along with possible adverse effects from treatment. "Your job is to become more active in your treatment," he says.

The primary therapy remains oral or intravenous (IV) corticosteroids - prednisone, methylprednisolone, solumedrol. Along with this first-line treatment, Miller stresses the importance of beginning physical and occupational therapy, calcium and Vitamin D supple-

*See Myositis 101, page 21*

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Dear TMA member:

This is the second Annual Special Treatment Issue of the *OutLook Extra*. It is devoted entirely to providing updates on the latest treatment and research news as well as reports from TMA's Annual Conference, held this year in Las Vegas.

As you read through this issue, you will find a range of information - some for those who have recently been diagnosed (Myositis 101: a beginner's guide) to more technical reports for those better versed in the disease. Likewise, you will find information from a wide range of researchers - those who have studied myositis for decades as well as those who are relatively new to the field and of another generation. The focus of the researchers also varies from those nearly exclusively studying only one form of myositis to those who are looking at the "big picture" of what environmental factors may be contributing to or causing the full range of inflammatory myopathies.

You will also find questions and comments from people like you - those who are living with myositis and have learned firsthand the perplexing and frustrating aspects of this terrible disease.

TMA is trying, through its publications, online services, support groups and conferences, to provide a forum for all those involved with myositis to share their experiences and information. We believe that through collaboration and information sharing, everyone will benefit - patients, family, physicians and researchers.

The Special Treatment Issue is another step along this path.

I hope you find it helpful and informative.

Sincerely,

A handwritten signature in black ink, appearing to read "Bob Goldberg". The signature is fluid and cursive.

Bob Goldberg  
Executive Director

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## Perspectives on treatment

continued from cover

levels have normalized, usually within one to two months, he changes to a once daily dose and tapers by 20 to 25 percent every 3 to 4 weeks to reach 5-10 mg. The patient then maintains this dose until he or she has been on corticosteroid therapy for one full year. This, he says, is for an ideal patient, so there are obviously other factors to keep in mind - side effects, lack of response and repeated relapses when tapering. In some cases, other agents are added.

### Adding other agents

Which immunosuppressant your doctor chooses, says Dalakas, often depends on your doctor's past experience and personal preference. More common immunosuppressive therapies are methotrexate, azathioprine, cyclophosphamide, cyclosporine, and mycophenolate mofetil. There are no standard dosages for treating myositis, but Drs. Dalakas and Oddis share the more commonly prescribed regimens. Your own doctors will consider your medical histories and symptoms to determine the right dose for you.

■ **Methotrexate:** 7.5 mg per week orally for three weeks, given at 2.5 mg three times at 12-hour intervals, increasing the dose by 2.5 mg per week to a 25-mg dose. Methotrexate works faster than azathioprine (below), so it is often considered as the first addition to first-line therapies. This medicine can be given orally, intravenously or subcutaneously.

■ **Azathioprine:** 3 mg/kg orally, though 1.5-2 mg/kg is more common. Azathioprine is well-tolerated but takes up to six months to show whether it's effective.

■ **Cyclophosphamide:** 0.5-1 mg/m<sup>2</sup> monthly for those with interstitial lung disease.

■ **Cyclosporine:** 150 mg twice daily or 3-3.5 mg/kg daily, but no more than 5 mg/kg per day.

■ **Mycophenolate mofetil:** up to 2 g daily. This is a promising, well-tolerated treatment, though it may take up to three months to notice any clinical benefit.

### Treatments to consider

For difficult-to-treat cases, both doctors introduce intravenous immunoglobulin at 2 g/kg. Dalakas proposes this over two to five divided daily doses every five to eight weeks. For DM, Dalakas typically sees a noticeable improvement 15 to 20 days after the first treatment. Those with DM may need repeated therapy every 6 to 12 weeks to maintain these results, he says. In his opinion, IVIg is best reserved as a third-line treatment for those who are steroid-resistant, though he says it is often a second-line treatment for children. His own experience shows that IVIg is effective in about half of his PM patients, but this treatment is not effective in IBM except for those with swallowing problems. Oddis presents several studies on the use of IVIg in combination with other therapies, showing positive results for many refractory or relapsing patients.

Treatments typically aren't as straightforward as this, though, as there are a number of symptoms to take into account.

Hydroxychloroquine (Plaquenil) at 200-400 mg per day, for example, fights the DM rash more aggressively, says Oddis. Also, topical tacrolimus (Protopic) has been shown effective in studies to combat the skin rash. Lung problems call for stronger therapies, and Oddis describes one option: oral corticosteroids at 60-80 mg daily (divided) plus pulse solumedrol at 1 gram per day for three consecutive days, with the addition of an immunosuppressive early in treatment.

### Additional treatments

Adjuncts to medicinal therapy are key, says Dalakas. Physical therapy should be an integral part of the treatment plan from the start to address

muscle weakness, loss of joint motion, and fatigue, adds Oddis. For those with less strength, passive range-of-motion should begin the therapy, changing to assisted range-of-motion then a more aggressive program as strength increases.

Nutrition is also an important consideration - he recommends a low-carbohydrate, low-salt and high-protein diet; antacids for any gastrointestinal problems brought on by the medicines; calcium at 1 g per day; Vitamin D at 50,000 units per week; and supplements to protect bone loss. (See extensive osteoporosis section, beginning on page 10). For IBMers, he adds Co-Q10, Vitamin E and creatine to his treatment approach.

[Dr. Dalakas' article, "Therapeutic Approaches in Patients with Inflammatory Myopathies," and Dr. Oddis' article, "Idiopathic Inflammatory Myopathy: Management and Prognosis," appeared in the December 2003 issue of TMA's electronic full-member publication, Update, and are included. Dr. Miller's introduction to myositis was a 2004 Annual Conference presentation.]



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# Prednisone: saves lives, creates havoc

It's the drug everyone loves to hate, the pharmaceutical paradox of modern times. Prednisone, in its many forms, saves lives while it creates a multitude of problems. Once in the bloodstream, prednisone hitchhikes itself to certain proteins and enters nearly every cell in the body indiscriminately. Once there, receptors draw it to the nucleus where it binds to genes, causing a variety of effects. In the muscle cell, it works very well to decrease inflammation, ease pain, and restore normal function. In the bone, it decreases the ability of cells to incorporate calcium, causing osteoporosis; in fat cells, it causes the swollen ("Cushingoid") face and the characteristic "buffalo hump" across the back of the neck. The fat cells also increase as the brain cells are stimulated by prednisone to think your body requires more and more food. In your immune system, cells are slow to respond, increasing the chance of infection of all types. That's because prednisone blocks the speed that blood travels to the affected cells.

Meanwhile, the brain cells are also changed in a way that causes depression, anxiety, or other mood swings. To combat all these effects, physicians tinker with the type of prednisone, as well as the timing and strength of the dose, but it's always a balancing act: with less side effects, it's often less effective; the more effective dose often has more side effects. Prednisone can be inhaled, injected, given orally or intravenously. It can be administered once daily, twice daily, three times daily, as well as every other day.

Most inclusion-body myositis patients do not benefit from prednisone, but because inflammation is also part of this disease, there may be some initial and continued response.

This is especially true if the patient has another autoimmune disease. Because of the very slow progress of IBM, it's difficult to assess the benefits of prednisone therapy. Many clinicians, including Dr. Chester Oddis and Dr. Richard Barohn, will try prednisone for a short time if the patient requests, discontinuing it fairly quickly if there's no progress.

Prednisone's side effects are as inconsistent as they are troubling. In many

cases, they disappear when the drug is tapered and discontinued. One of the most troubling side effects, corticosteroid-induced osteoporosis, is discussed separately in a section devoted exclusively to osteoporosis, beginning on page 10.

Other side effects are caused by the very properties that make it effective. Since it suppresses the immune system, it also increases susceptibility to infections, especially with long-term use. In turn, infections may flare the underlying disease, as they provide extra stimulation for the suppressed immune system. The moon-face or "Cushingoid syndrome" disappears rather quickly, and most people are able to lose the extra weight caused by increased hunger and fat accumulation. "Lock the fridge" was the advice to those wanting to know how to avoid the weight gain almost always experienced by prednisone users. Drs. Oddis and Barohn and Dr. Marinos Dalakas give their prednisone patients a low-salt, low-fat, low-sugar diet, and report good results if people are able to ignore the prednisone-induced hunger and follow the regimen.

Children taking regular prednisone can experience slower growth and may be more vulnerable to childhood diseases. Long-term prednisone users

are at greater risk for vision problems: glaucoma and cataracts as well as distortion in their vision, especially when tired. At first, prednisone can cause elevated blood pressure, but the body often adjusts and the blood pressure returns to a normal range. Tapering prednisone after the body has made this adjustment may cause the blood pressure to drop below normal. Since the drug is always tapered very slowly, the body usually adjusts once again.

Women of childbearing age are very concerned about the effects of prednisone on pregnancy and birth. It does not cross the placenta to the baby during pregnancy, although Dr. Oddis - who has written extensively about myositis and pregnancy - cautioned that some other steroids do cross over to the baby and can cause harm. In studies, Dr. Oddis and others found that women who had their myositis under control before pregnancy generally did better than those who bore and delivered a child during a flare. All physicians involved - family practitioner, neurologist, rheumatologist and obstetrician - should consult and treat the pregnancy as one with some risks.

## The birth of a medical miracle in mid-century

In an age where it seems each new day brings another miracle drug, it's hard for us to imagine the scene in 1949. The place was the Mayo Clinic; the drug was "substance X." With extreme secrecy, patients with severe rheumatoid arthritis were tested in trials. The reason for the secrecy was that preliminary results were so amazing that investigators couldn't believe their eyes. One spring day, they shared the results of the "substance X" trials with the staff and prepared themselves for the onslaught of publicity and all the hype and hope it was to create.

**Prednisone's side effects are as inconsistent as they are troubling.**



Wire services around the world told stories and showed pictures of people - previously barely able to walk - throwing down their crutches, leaping up from their wheelchairs and running upstairs.

"Substance X" was cortisone, a drug that had taken nearly a decade to identify. The late Dr. Phillip Hench devoted his life to finding a treatment for rheumatoid arthritis, a disease that was poorly understood and seemed, even to the most optimistic, to be a heartbreaking diagnosis, often disabling and life-threatening.

Like other researchers of his day, Dr. Hench looked for the rare cases where the disease under investigation appeared to improve or remit spontaneously. He and his colleagues noted that this happened in pregnant women and patients with jaundice. The researchers theorized that those patients were producing a substance related to their condition that reversed the disease. Dr. Hench thought it was probably a hormone.

By the late 30s, a Mayo Clinic colleague, Edward Kendall, had isolated six hormones from the adrenal glands of cows. He identified the extracts as compounds A, B, C, D, E, and F. Compound E seemed chemically similar to a compound excreted by patients with jaundice, so Dr. Hench thought that might be his "substance X." But it was so difficult to obtain that it took him

years to accumulate enough "compound E" to treat one patient. With

exciting response in that one patient, Hench

had more resources at his command. He gave the drug to several

more Mayo Clinic patients in the days leading up to that historic spring staff meeting. He and Kendall traveled to Stockholm in 1950 to accept the Nobel Prize for Medicine. The American scientists were joined by Swiss biochemist Tadeus Reichman, whose work with cortisone paralleled their own.

The next breakthrough was the development of synthetic cortisone, or prednisone, and it was used throughout the world to fight inflammation. As use spread, so did awareness of the side effects, and in the last two decades, physicians have used it in combination with other drugs. What scientists are hoping for is another discovery on the level of Dr. Hench's - a substance that will suppress targeted reactions of the immune system without disabling it.

#### Protection from steroid-induced osteoporosis

Corticosteroids cause bone loss and are the most common cause of drug-related osteoporosis. For many people who depend on these drugs for survival, the loss of bone and consequent risk of injury is the most frightening side effect of the treatment. Osteoporosis is the major cause of bone fractures in postmenopausal and elderly women, and prednisone treatment adds to the risk. Your bones don't remain static as they may appear, but change constantly, with old bone broken down and replaced by strong new bone. Bone mass declines in early middle age, and women lose more bone mass after their estrogen level drops. Bones with less mass are more likely to break or fracture, even in a minor fall.

Prednisone damages bone by increasing the bone loss and also reducing new bone formation, calcium absorption and estrogen levels. For those taking prednisone, the amount of bone loss depends on the size and duration of the dose, and how strong the bones are when beginning the medication. A daily dose of

prednisone higher than 7.5 milligrams over several years results in bone loss and an increased risk of fractures, but short-term prednisone treatment is less harmful.

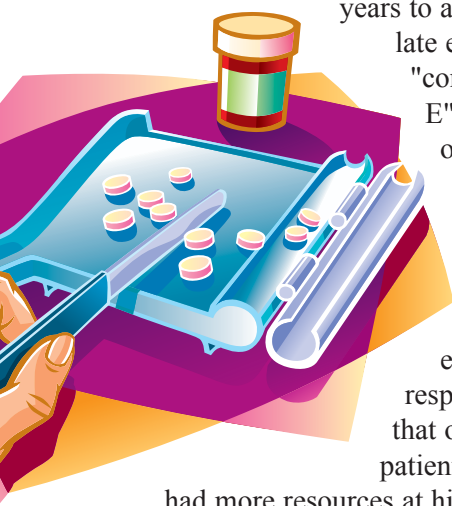
The most important step a myositis patient on prednisone therapy can take is to discuss osteoporosis prevention with his physician and get regular bone density tests. If you're under the age of 35, you can prepare for bone loss in the future and counteract some of the effects of prednisone by increasing your calcium intake. Although most people associate calcium with dairy products, it's important to know that there are other very good sources for calcium for people who don't enjoy or don't use milk or milk products. Green leafy vegetables, shellfish, sardines, oysters, Brazil nuts, tofu, almonds and calcium-fortified breads and cereals are also sources of calcium. Most adults need 1,000 milligrams of calcium each day. As you age, you may need more than 1,500 milligrams each day. Dr. Oddis and other physicians recommend taking calcium in two or more doses. Doctors are now prescribing a number of drugs and combinations of drugs in the hope of reducing the risk of osteoporosis in myositis patients. For new research on preventing and treating osteoporosis, see page 10.

#### Do you have prednisone-induced osteoporosis?

If you are taking prednisone, or have taken it for periods of time to treat myositis, you can find out if you have lost or are losing bone mass by taking a bone density test. Most doctors believe that anyone who is taking steroids should be tested regularly. Follow-up tests are then made to keep track of your progress.

What should you expect from the test? It's not painful or time-consuming. It's done with a bone densitometer using a special X-ray. The scan can measure as little as one or two

*Continued, next page*



percent loss in bone. CAT scans and ultrasound machines may be used in the future to measure bone density.

Once your bone density is determined, your physician may want to find out if there are factors other than your prednisone use that may contribute to osteoporosis. He may order lab tests, such as a serum calcium test, a phosphorus test, a protein test, a thyroid hormone test, an alkaline phosphatase test and liver and kidney function tests. He may also prescribe calcium. Some studies claim that calcium carbonate supplements contain the highest amount of useful calcium, and the Arthritis Foundation recommends calcium in this form. This form can be harder for your body to absorb, so it's recommended that the calcium be taken with meals. Calcium citrate and calcium gluconate are easier for some people to absorb. Another measure you can take - for overall health as well as to combat bone loss - is to stop smoking. Calcium absorption is reduced in smokers; women who smoke generally experience menopause earlier; and smoking reduces the benefits of estrogen on calcium absorption.

Another habit that affects bone loss is alcohol consumption. This effect is seen in heavy drinkers, who lose more bone mass and lose it faster than those who drink moderately or not at all. Dr. Oddis also recommended weight-bearing exercise to protect bones. "Weight-bearing" means any physical activity that causes your bones to bear weight or increases the force of gravity against them. These are exercises like dancing, running, walking, tennis, and cross-country skiing. It is not known how long you need to exercise, but aim for 30 minutes most days. Myositis patients, as always, are advised to check with their doctors about exercise. TMA will publish a full discussion of exercise in the March issue of *The Outlook*.

Preventive measures help reduce

the risk of osteoporosis, and the same kinds of measures are recommended for those who have already suffered bone loss because of prednisone therapy - exercise, supplements, calcium through food. Biphosphonates are compounds that slow the loss of bone from osteoporosis, and restore bone density and strength.

In a person with myositis, certain lifestyle changes can also help prevent further injury. With bone loss coupled with loss of muscle strength, falls are always a threat. It's important to do everything possible to prevent falls. Our members suggest keeping living areas well-lit and carrying flashlights when traveling, installing nightlights, freeing your house of obstacles, installing sturdy handrails and railings (and using them), and designing kitchens that are efficient and safe.

#### How TMA medical advisors prescribe prednisone

Prednisone is still the first treatment for dermatomyositis and polymyositis. Despite its side effects, it's a lifesaver, making death from myositis a rare exception. "We all struggle with this," said Dr. Richard Barohn, "but we've certainly gotten better at working with the drugs and preventing side effects."

Oral prednisone is usually given in a single daily dose of 0.5 to 1.5 mg/kg until the strength is normal or the maximum improvement has been obtained. Then, prednisone is tapered slowly over a 12-month period. Often after 1 or 2 months on therapy, he switches the patient to every other day treatment. It's been the experience of many of his patients that this process takes a couple of years.

For more on how different doctors prescribe prednisone, see "Myositis treatment," cover.

We've learned that early diagnosis and treatment are critical for predicting a good outcome. A study of 113 patients demonstrated that one-third

of patients with a short delay in diagnosis responded completely to prednisone, compared with much less satisfying improvement by all patients with a long delay. If you see good results in blood tests but still have muscle symptoms, be patient.

Sometimes clinical muscle strength improvement lags behind improving lab tests, and sometimes you feel stronger before the tests confirm an improvement.

#### Side effects: sometimes more than you bargained for

Some patients respond very well to prednisone and have very minor side effects. It's more common, according to our members, to respond well in improvement of strength, but to experience one or more of these side effects: weight gain, cataracts, diabetes, stomach symptoms, osteoporosis, and insomnia. When Dr. Barohn prescribes prednisone for his patients, he addresses the side effects from the beginning. "Each one of our patients on prednisone is given a calorie-restricted diet," he said. It's been his experience, and also that of Dr. Marinos Dalakas, that most patients who adhere to a low-salt, low-fat diet of between 1000-2000 calories, with complex carbohydrates and no junk food or sugar-loaded desserts and drinks, do avoid the weight gain, bloating, diabetes and "moon-face" associated with prednisone use. "It's a difficult diet, but it does work," Dr. Barohn said. "It's very similar to the diabetes diet."

He also prescribes supplemental calcium and Vitamin D for patients with normal bone density; and the bone density test is repeated every year. Those who test below normal are prescribed alendronate sodium as well as the supplements. Most patients don't experience stomach upsets. "If that's the case, I leave well enough alone," Barohn said. Otherwise, he prescribes Zantac.

## Gardens and yards: good for the bones

Researchers have linked regular yard work to the prevention of osteoporosis, finding that women aged 50 and older who gardened at least once a week showed higher bone density readings than those who performed other types of exercise - including jogging, swimming, walking and aerobics. By knowing which exercises provide the greatest benefit, women can design a workout regimen that will ensure strong bones as they age. Such preventive measures may reduce the number of people who develop osteoporosis.

To gain a comprehensive look at the effects of exercise on older women, researchers looked at the National Health and Nutrition Examination Survey, a dataset collected by the National Center for Health Statistics, which contains information on more than 40,000 women.

Younger subjects were eliminated, leaving a pool of 3,310 women aged 50 and older, and researchers studied how often these women performed different activities, including yard work, calisthenics, bicycling, dancing, aerobics, swimming, jogging, walking and weight training. Each activity was related to bone mass, finding that bicycling, aerobics, dancing, yard work and weight training were linked to higher levels of mineral density.

After adjusting for other factors, the results showed only two activities to be significant for maintaining healthy bone mass - yard work and weight training. Researchers certainly hadn't expected yard work to be significant, since it's not considered an athletic activity. There's a lot of weight-bearing motion going on in the garden - digging holes, pulling weeds, pushing a mower. An additional benefit of gardening is the fact that it's performed outdoors, where exposure to sunlight boosts vitamin D production, which aids the body in calcium absorption. While weight-

bearing activity and vitamin D work directly to strengthen women's bones, yard work provides indirect benefits as well. Of all the activities, yard work proved the most popular, with nearly half of the subjects - 1,384 women - claiming to garden at least once a week. Such popularity makes it a highly effective preventive measure.

In the end, the best thing about yard work is that so many people are willing to do it. They don't dread it as exercise, and they'll probably continue to do it as long as they're able. And as long as people stick with an exercise, they can harvest the benefits well into old age. In addition to exercise, women must take other precautions, and they must take them as early in life as possible: at least 1,000 milligrams of calcium a day, a healthy body weight, and no smoking or excessive alcohol consumption.



## Stand up straight and call the "bone phone"

Often the first sign of osteoporosis is a fracture or a fall. Now, research shows that good posture can help people with osteoporosis reduce their risk of broken bones, says University of Alabama registered dietitian Beth Kitchin, R.D., of the Osteoporosis Treatment and Prevention Clinic. "For people with osteoporosis even movement as simple as bending over to unload a dishwasher or wash your face can lead to a vertebral fracture. Learn to use your hips and knees when bending, rather than hunching or rounding your shoulders forward, to reduce the risk."

The University has become a regional leader in osteoporosis management, and has also established a hotline known as the "bone phone." The UAB osteoporosis hotline answers questions about osteoporosis. Call toll-free, 1-888-934-BONE.



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# Juvenile myositis and prednisone: battling side effects

*Editor's note: Prednisone is the mainstay of therapy, especially for those with juvenile myositis. It typically works quickly and effectively, but along with the positive impact, it brings a number of undesirable effects. Richard Gay, who had JM as a child, explains his personal experience with the side effects of prednisone therapy. Fortunately, doctors have discovered new ways to deal with adverse effects of prednisone - including introducing steroid-sparing medicines earlier in treatment - since Rich was treated for JM 40 years ago.*

## Getting more than you bargained for

The doctors at the recent TMA Conference called juvenile myositis (JM) a "devastating disease." The realization that juvenile dermatomyositis (JDM) will be with me for a long time does not come right away when diagnosed. Even when the doctor makes an early and correct diagnosis, I'm not aware of the serious nature of the illness. The long-term effects of JDM are not on the front burner. The first thing on my mind is to recover my strength, be able to swallow food again, and make the itching go away.

Thus, the knight on the white horse is prednisone, the most widely used medication for JM, and deservedly so. The value of prednisone can be most readily appreciated by observing that in 1964, when I was 13 years old and first diagnosed, prednisone was the primary treatment - in fact, the only treatment - for JDM. Now forty years later, with amazing progress in medical treatments, prednisone is still the primary medication. It works, and we have to give credit where it is due.

## Immediate effects - the bad with the good

But prednisone has a dark side that comes in the form of side effects. In

1964, I knew none of the short- or long-term side effects of prednisone. I had the immediate reaction of the "moon face," but it was not a major concern to me. In today's world of make-overs, fad diets, and personal trainers, Madison Avenue is teaching us that appearance is the most important thing in our lives.

Prednisone makes a rapid and dramatic change in appearance. Fortunately, it didn't bother me and my doctor was able to warn me that it was coming.

In addition, prednisone also causes an increase in acne. As a young teenager, my acne was difficult to control, and prednisone made it worse. I went to a dermatologist many times for treatments. I had pimples across my back, and my dad would clean my back when I took a bath just to try to control the acne. As it turned out, time was the only effective remedy for the acne.

Prednisone also makes us susceptible to infections and common illnesses. I remember having many colds during high school. It seemed that I was always taking Contact - the favorite cold remedy of the time - while in school. The colds were controllable but just didn't seem to go away. Now I know why: prednisone makes us susceptible to every opportunistic bug we encounter.

## What to watch for

The long-term effects are more subtle but can be much more serious. Sometimes, you get more than you asked for and there is no easy way to say, "No, take it back." The first side effect I had to deal with was stomach ulcers. I took 30 mg/day of prednisone for about five years. Within a few months, I noticed trouble with my stomach, and my doctor recommend-

ed a liquid antacid called Maalox. Maalox is a mixture of magnesium hydroxide and aluminum hydroxide, both chemical bases that neutralize the excess stomach acid produced by the prednisone. I took so much Maalox that the first calcium deposit I got under my skin came on the left thumb I used to open the lid of

the Maalox bottle. I still have that calcium deposit, and it's pretty big. Doctors today recommend calcium carbonate tablets (Tums) to neutralize stomach acid and provide supplemental calcium. However, antacids don't stop the damage to the stomach caused by prednisone, and some permanent sensitivity remains today, even though I have been off prednisone for 23 years. The stomach acid issue goes beyond antacids: it creates an awareness of the acid contents of foods, which most kids don't consider. I had a rapid learning experience to decide what foods to eat and what foods to avoid.

A second side effect, which is just as serious, is the loss of calcium from our bones. After about a year on prednisone, I began to experience short spasms in my back. At that time, there were no medications to retain calcium in our bones, such as Fosamax and Actonel today. In fact, I took dilute sodium fluoride solution nominally for this purpose, but it had no benefit. The back

**Now forty years later, with amazing progress in medical treatments, prednisone is still the primary medication.**





spasms were the result of the leaching of calcium from my back by the prednisone. There was nothing to stop this from happening. The solution was to wear a back brace. Today, many lightweight back braces made of durable plastic are available. In 1965, the brace was made of metal that curved around the front of my body to the back, with straps to hold a pad across my back against the metal frame. It was uncomfortable and hot; I wore out a number of pads. There was a serious psychological effect because there was no expectation that I would ever be out of the back brace.

I also noticed that I was not gaining in height or weight, and I asked my doctor if I would grow any more. He told me no. I know now that prednisone suppresses our growth. I did not have the weight gain that many people ascribe to prednisone, though my appetite was always good.

I had the life-giving benefits of suppressing the autoimmune response of JDM while experiencing the detrimental side effects of the medication itself. There are other serious effects that I did not experience, such as cataracts and mood swings. In fact, I have never had to take anti-depressants in the 40 years I have had this illness.

*By Richard Gay (JM), Co-chair Los Angeles Keep In Touch support group. Edited for space - to receive a copy of Rich's article in its entirety, email [tma@myositis.org](mailto:tma@myositis.org)*



### Another patient, different side effects

Joanne has dealt with JM since childhood, but she doesn't remember much about the medicine's side

effects. With some help from her mother, here's what she does recall:

As far as I can remember, I was on prednisone, methotrexate, cyclosporin and an aluminum-based liquid medicine for calcifications, which messed around with my stomach. I had years of stomach cramping, diarrhea, and irritable bowel syndrome, even years after I stopped taking it, and it didn't help with the calcium, as I had and still do have a lot.

My parents remember my ravenous appetite and how they had to monitor my diet so I didn't put on too much weight, which they said was hard because I was always hungry. They also watched my sugar intake due to steroid-based diabetes.

Mum said I went through sleeplessness but didn't know of anything to help with that. I still have problems sleeping and now take a natural tablet called Valeria.

For my hair loss, Mum said we basically used sorbalene because they advised against anything perfumed - nothing of help for the hair loss.

One thing that we found for ulcers on the skin was EDP powder. It comes in a little bottle and is like an antiseptic powder that helps dry out the ulceration.

Mum said I often had this creepy crawly feeling in my skin, which she thinks may have been my muscles breaking down. Luckily, I can't actually remember this.

As far as long-term side effects, I have diabetes, hypertension, osteoporosis, and cataracts. I have depression, which has a link to steroids and to diabetes and, as we know, is just a plain old common complaint nowadays anyway. There are other medical problems, but as I've been told, there has been so little research done into the long-term side effects that they just can't confirm if they are related to the steroids or not.



### Strategies to help along the journey

By Richard Gay (JM)

Given the serious nature of the side effects of prednisone, what is a successful strategy to deal with both the JDM and the side effects?

■ Recognize that time is our ally in this endeavor. Surprisingly, my doctor expected time to be a major factor in beating the disease. He expected it to "burn out" within a couple of years. It did not, but it did become inactive after 14 years.

■ Identify ways to lower the prednisone dose while not allowing the illness to get worse. Today we see many combinations of medications used to supplement the prednisone precisely with the goal of reducing the amount of prednisone used. In 1970, I was in a trial study using methotrexate with prednisone, effectively lowering my dose from 30 mg/day to 15 mg/day. Within a few months, my back strengthened and I have not worn a back brace since then. The long-term decrease in calcium in my back is only slowly recovering.

■ Use your muscles as much as possible through exercising and stretching. This encourages the healthy part of our body to maintain and build the remaining muscles that aren't damaged in the initial illness.

■ Maintain a healthy emotional and spiritual life. The center of who we are is our own spirit; if we allow it to atrophy, our physical bodies will follow. There is a feeling of isolation with JDM. I was always active in various organizations at school and church, and JDM did not interfere so the isolation was minimized.

It's a risky balance choosing between the illness and the side effects of the medication. Medical science is making significant progress every day, and there are some very promising medicines for JDM and adult dermatomyositis.



# Osteoporosis - new approaches for crippling disease

Prevention and treatment of osteoporosis - a serious side effect of prednisone therapy and a costly public health problem - continues to be a high priority for research, and was the subject of several presentations at the American College of Rheumatology 2004 Annual Scientific Meeting. Osteoporosis weakens bones, leaving them subject to fracture in one out of every two women, primarily in those over the age of 50. In the U.S. alone, some eight million women and two million men run the risk of fracture leading to chronic pain, long-term disability and even death from this silent disease. The studies presented here are very new: please discuss them with your doctor.

## Vitamin D linked with improved muscle strength and function

This research studied patients with knee osteoarthritis, not osteoporosis, but we include it here because it is the first look at vitamin levels in relation to pain and disability. The 221 patients had an average age of 67 years and were measured for changes in pain, physical function, muscle strength and blood levels of vitamin D two or more times across a 15- and 30-month period. At the beginning, the 48 percent of patients with low levels of vitamin D (at or below the amount needed to satisfy the body's requirements) experienced more pain and disability than those with levels above the minimum. Researchers also found that those with low levels of vitamin D were weaker.

Vitamin D, which comes primarily from exposure to sunlight, promotes the absorption of calcium and phosphorus in bone mineralization, growth and repair. Vitamin D also comes from dietary sources like oily fish, liver, fortified breakfast cereals and dairy products. The elderly are less efficient at producing vitamin D from sunlight and absorbing it from food. To address their higher risk for D

deficiency, the elderly population is often directed to take a vitamin D supplements, of 400-600 International Units per day. (Exposure to sun should be limited to five to 15 minutes on the face, hands or arms, or arms and legs. Dermatomyositis patients should always check with their physicians before exposing themselves to sunlight.)

## New oral medication appears to reduce fractures

Strontium ranelate, a new oral medication, may reduce spinal, non-spinal, hip and other fractures in older women with osteoporosis, a large ongoing study suggests. Post-menopausal women were randomly assigned strontium ranelate or a placebo, along with calcium and vitamin D supplements for three years and testing was broken down into two multi-national, double-blind controlled studies. One focused on the possible reduction of fractures of the spine in nearly 1,650 women, average age 69; the other studied non-spinal fractures in more than 5,000 women, average age 76. All women studied had low bone density.

In both studies, participants experienced a significant reduction in fracture risk. Over the three-year period, 36 percent fewer women 74 years of age or older suffered hip fractures. Concurrently, spinal and non-spinal fractures were reduced by 32 percent and 31 percent, respectively, in the subgroup of elderly women ages 80 and older. Strontium ranelate appeared to both increase bone formation and decrease bone density loss in the majority of patients, demonstrating a good bone and general safety response.

"Strontium ranelate is the first compound to simultaneously decrease bone resorption and stimulate bone formation," said Jean Yves Reginster, MD, Dept. of Public Health, Epidemiology and Health Economics,

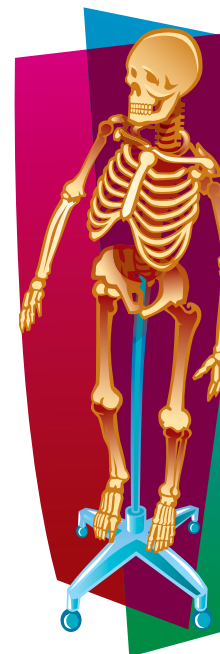
University of Liège, Belgium and World Health Organization Collaborating Center for Rheumatic Diseases, who was an investigator in the study. "Given this and its outstanding safety profile, strontium ranelate could prove to be a first-line treatment option for women with low bone mineral density with or without prevalent fractures as well as for elderly women with increased risk factors of hip fractures."

## Osteoporosis cocktail stimulates new bone growth

The combination of teriparatide (Forteo®), an injectable parathyroid hormone medication, when administered with the selective estrogen receptor modulator, raloxifene (Evista®), improves bone density formation, a new study suggests. Previous research had shown that another antiresorptive agent, alendronate (Fosamax®), appeared to diminish the gain in bone density seen with teriparatide alone.

To determine if the benefits of teriparatide can be enhanced with the addition of raloxifene, a selective estrogen receptor modulator (SERM) which slows bone loss and slightly increases normal bone growth, researchers conducted a six-month randomized, double-blind study comparing the use of teriparatide against the combination therapy. The 137 postmenopausal women participating in the trial, none of whom had prior osteoporosis treatment, also received calcium and vitamin D supplements throughout the course of the study.

Groups on the single and double agents showed similar significant increases in bone formation in months



one, three and six. However, bone resorption was reduced (as measured by markers of bone turnover found through urine and blood tests) in the group taking both medications by month three, an effect which persisted to month six. Those taking both teriparatide and raloxifene had higher bone density at the spine and hip (significantly higher for the hip site) than those on teriparatide alone. "The results are encouraging since we are looking for agents or combination of agents given together or sequentially that will further reduce the rate of fracture in high risk patients," said Chad Deal, MD, Cleveland Clinic Foundation, Cleveland, Ohio, and an investigator in the study. "The most important question is effect on fracture reduction and, although no studies are underway at this time to assess fracture reduction with this combination, the bone density and marker data are a promising start."

#### Antibody prevents bone loss

A novel treatment administered semi-annually to postmenopausal women with low bone density appears to rapidly inhibit the bone resorption process, resulting in improvements in bone mineral density at 12 months. Bone is living tissue that is in constant regeneration. This means tissues that form bone are constantly being created and resorbed by the body. During adolescence and early adulthood, bone growth resulting in peak bone density is due to the significantly greater new bone formation as compared to bone resorption. With aging and the loss of estrogen after menopause, the balance between bone resorption and new bone formation shifts. More bone is lost than can be replaced, leaving bones thinner and structurally weaker. This results in osteoporosis and fragility fractures.

Researchers recently studied a new treatment antibody, AMG 162 that binds to a receptor protein on the surfaces of osteoclasts - the cells that function in the absorption and

removal of bone tissue. AMG 162, which is still in clinical trials, may prevent bone loss resulting in osteoporosis and the bone erosions that lead to rheumatoid arthritis.

To assess AMG 162's effectiveness when administered every six months, researchers conducted a year-long test of different doses of the antibody use in 411 women, average age 63 years, who are participating in an on-going, randomized study. Eight of the nine treatment groups received double-blind, subcutaneous injections of AMG 162 (six, 14 or 30 milligrams every three months, or 14, 60, 100 or 210 milligrams every six months) or a placebo. The last group received open-label 70-milligrams of oral alendronate once weekly. Urine and blood tests as well as X-rays were used to evaluate results.

An anti-resorptive response, as measured by bone turnover markers (urine and blood tests), was almost immediately evident in all patients taking the antibody, and continued to improve through month four. Depending on AMG 162 dose levels, increases in bone mineral density also were observed as early as month one. The most common adverse effect, indigestion, occurred in only a small portion of all groups. Overall, AMG 162 administered once every six months was well tolerated and caused a rapid, dose-dependent increase in bone formation and bone density. "If ongoing clinical trials demonstrate fracture risk reduction, this therapy should lead to a dramatic improvement in patient compliance due to the ease of administration compared to presently available osteoporosis treatment," said S.B. Cohen, MD, Radiant Research, Dallas, Texas, and an investigator in the study.

#### Protecting young bones

Juvenile myositis is hard on children's bones, said Mona Calvo of the Center for Food Safety and Applied Nutrition at the Food and Drug Administration. Parents at TMA's Juvenile Myositis

conference were aware that the prednisone prescribed for JM, especially in large doses, contributes to the loss of children's bones. There are other contributors, though, Dr. Calvo explained. The active myositis itself affects bone density, as does the reduced activity endured by JM patients, the decreased sun exposure because of the rash, and possibly poor protein and calorie intake combined with poor calcium and vitamin D intake that can be a side effect of illness and poor appetite. She gave the parents some ideas for preventing bone loss and encouraging good bone formation in children to reduce the chances of future fracture risk. Her advice for the families was to increase the amount of calcium, vitamin D and protein intake while also making sure children were exercising as much as possible under the circumstances.

Children with myositis need more than the 800 to 1300 mgs of calcium and the 200 international units of vitamin D required by healthy children, Dr. Calvo explained, because the prednisone increases the amount of calcium the body loses and interferes with the way vitamin D functions. Dr. Calvo encouraged parents to look closely at the "Nutrition Facts" panel on the label of the foods they buy for their families; to encourage children to drink milk and vitamin D-rich beverages; and to avoid the typical US teen's dietary habits, which have changed over the years to include huge amounts of carbonated soft drinks beginning at age 11. Because of the higher than usual needs of JM children, Dr. Calvo encouraged parents to search out calcium and vitamin D fortified foods. Some foods presently fortified with Vitamin D are milk, cereal and fruit juices.



*Find Quick tips for healthy bones on page 30, and read how gardening can help prevent osteoporosis on page 7.*

# Dysphagia: what you should know

Dysphagia, or trouble swallowing, is a serious side effect of myositis and is often ignored, says Dr. Todd Levine. This topic is of special interest to him, both because he is a neurologist treating people with swallowing problems and because he has personal experience using a temporary feeding tube following an injury.

Swallowing is one of the most complicated things we do neurologically, he says. Half the brain stem is used to get your food from your lip to the back of your throat. It's a complicated system, as the trachea, which passes air to your lungs, and the esophagus, which passes food to your stomach, are very close together. There are intricate connections between the brain, nerves and muscles. Once something else enters the equation, like muscle disease, the system breaks down in some way. Michelle Reuther, a speech therapist, notes that a normal swallow occurs in one second. A number of muscles are involved in this one second of swallowing - the posterior wall and front muscles come together, your soft palate moves up, the back of your tongue stretches and pulls, your epiglottis closes to cover your airway, and your Adam's apple comes up. When your muscles become weak, she says, the timing slows down.

## Signs of dysphagia

The first complaint for many is frequent choking, but Reuther says you may only feel like you're choking. The food is not blocking the airway but instead is stuck in a "pocket" (hypopharynx) at the base of your tongue near the epiglottis. If the hypopharynx fills up before you're ready to swallow, some of the build-up spills over the epiglottis into your airway, leading to aspiration. Aspiration pneumonia, which is brought on by small particles of food in the lungs, is a risk for those with

swallowing difficulties.

How do you know if you have dysphagia? You may realize it's taking much longer to eat meals than it should - requiring up to 5 to 10 swallows to take in a single bite, says Levine. For him, the easiest diagnostic tool is to have his patients drink water then say their name. If their voice sounds wet, there's a problem. In this situation, the water is sitting in the top of the esophagus at the trachea, where the vocal chords are located. His patients end up sounding like they're talking under water.

Other signs include:

- Pain when swallowing
- No interest in eating
- Frequent choking
- Low-grade fevers
- Coughing overnight

Signs of dysphagia often happen when you're eating out, a time when you may not be paying as much attention to your food as you should be, says Reuther. It's important to keep from being distracted while you're eating. If you're already having trouble swallowing, she says, you must concentrate in order to get the food down.

## Diagnosing the problem

Different doctors can diagnose swallowing disorders, but speech therapists are the most effective. "And they can give you techniques to help you overcome the problem," says Levine. Ear, nose and throat doctors are able to look down your throat with a scope, and speech therapists can look into your esophagus while you talk or eat to see which muscles are moving and which are not. Doctors are also interested in how you form a bolus, or the mass of chewed food - if you don't form a proper bolus, you'll end up with a long string of food or liquid to try to

swallow, he says.

Reuther encourages everyone to have a modified barium swallow. "With dysphagia," she says, "everyone is different." The barium test shows the therapist exactly when the problem occurs - before, during or after the swallow. Knowing this determines what exercises, techniques or procedures will benefit each individual. However, swallowing problems don't happen all the time for everyone. If you notice certain foods continuously causing problems, bring them with you for the test. Otherwise, the test may show a perfect swallow, she says, even in the presence of a problem.

## Treating dysphagia

There are many ways to treat dysphagia, and their success can be measured. Dr. Levine keeps track of everyone's weight in his clinic to make sure they are receiving adequate nutrition. His experience indicates that people with chronic disease do much better if they maintain a reasonable weight. If you have trouble swallowing, you need to take in more calories in an easier format, he says.

Though it doesn't seem logical, thinner foods and liquids are not the answer to dysphagia, as water and other thin liquids are more difficult to control in the swallowing process. Thicker foods are actually easier to control and are therefore easier to swallow. Reuther recommends alternating liquids and solids when eating to prevent any buildup in the hypopharynx. There are also thickeners that produce consistencies of nectar, honey and pudding. The additives are tasteless, although users may detect them in water. Thickening these thinner liquids slows them down, providing a better chance to swallow correctly.

Reuther also encourages patients to be aware when they're eating; distraction leads to more problems. She recommends that patients keep their

chin down to help close off the airway and bring the muscles closer together. She gives her clients three exercises to try - her "holds":

- **Tongue hold:** Stick your tongue between your teeth then swallow. This stretches all of your muscles.
- **Breath hold:** Take a deep breath, hold it and swallow. This is difficult, so you have to concentrate to be able to do this.
- **Adam's apple hold:** Swallow; then halfway through the swallow, when you feel your Adam's apple going up, leave it there for a second. Then finish your swallow. This is a difficult one to learn.

When techniques and exercises don't work, different procedures are available. In esophageal dilation, doctors insert a dilator into your esophagus to stretch it when the opening is too narrow. For cryopharyngeal myotomy, a small incision is cut in your muscle to allow food to pass through more easily.

If you aren't maintaining your weight, Levine encourages you to consider a feeding tube. A feeding tube, also called a G-tube or PEG tube, allows food to bypass the swallowing phase of eating by inserting a tube directly into your stomach. This tube then provides your main source of nutrition. "People often equate feeding tubes with ventilators as extraordinary measures," he says. "They're two vastly different things." Feeding tubes have many advantages: it's a 30-minute procedure, it's not visible to others, it's not permanent, and, maybe most importantly to you, you can still eat with a feeding tube. Eating is a sensory pleasure, says Levine, so with a feeding tube, you can enjoy a meal with friends and family without feeling pressured to eat more quickly. He recommends a feeding tube for anyone who has recurring pneumonia, who is losing weight, or for whom eating just isn't pleasurable anymore. Caregivers get

worn down, too, trying to help someone who has difficulty swallowing, adds Reuther, and a feeding tube may alleviate some of their workload and their worry.

#### Resources:

#### The Dysphagia Cookbook

The Dysphagia Cookbook, with a foreword by Dr. Todd Levine, is available at TMA's online Marketplace or by calling 1-800-821-7356.

Written especially for those with swallowing problems, this cookbook offers tips for preparing and serving food, information on food consistency, tools and foods to have on hand, and suggestions for eating out. Recipes include vegetable dishes, fish and meats, breakfast foods, salads, sauces, desserts, fruits, and drinks.



#### Dealing with dysphagia: more helpful hints

Michelle Reuther had a number of suggestions to take home with you. Here are her recommendations, along with general tips from previous TMA publications:

- Have a spray bottle with water next to your bed at night, then spray to moisten your mouth before taking a drink of water. Throughout the day, suck on a Lifesaver or something similar to keep your mouth from becoming too dry.
- Cut a piece out of a small Styrofoam cup for your nose to fit in while taking pills. This allows you to drink water without tipping your head back. You can buy these "nosey cups," but they're just as easy to make yourself, she says.
- Eat less food more often. Eating six smaller meals a day is better than three larger ones, especially when eating takes more time. Take in small amounts of liquid over time to keep the hypopharynx clear.
- Take smaller bites of food and smaller sips of liquid, and be sure your mouth is clear before taking another bite.
- Don't eat when you're overly tired. Focus on eating, and avoid distractions like television.
- Don't clear your throat when you feel the urge. Instead, swallow. This is a smoother action that doesn't irritate the vocal chords and has the same effect.
- Suck on lozenges, candies or mints when you're coughing a lot. Also take in plenty of water and avoid caffeine.
- Avoid foods that cause the problem. It's not always easy to determine which foods are the culprits, but if you can pinpoint any type - spicy or sweet, thick or thin, hot or cold - stay away from these. On the other hand, some foods may stimulate a swallow. Try mixing flavors or textures to see what works for you. Also, avoid foods that are most difficult to control in your mouth.
- Turn your head to the left or right. Decide what position of your head gives you the least problem when swallowing. Sit upright while eating and 1 to 2 hours afterwards to let gravity help.
- Maintain good oral hygiene. Research shows that this prevents aspiration pneumonia in individuals with swallowing disorders.

# Overlap syndrome: when myositis isn't your only obstacle

Myositis affects such a small percentage of the population that doctors don't often see patient after patient with the disease. When you have another autoimmune disease along with myositis, this causes even more confusion. It's often hard to sort out the symptoms of each disease, says Dr. Fred Miller: you typically need to meet the criteria for all of the diseases for them to be considered overlap.

Some more common overlapping illnesses are rheumatoid arthritis, lupus, scleroderma and Sjogren's. One research group from Hungary says myositis (mainly polymyositis)/rheumatoid arthritis overlap affects about 3 to 5% of myositis patients. In terms of the general population, a Romanian group states: "The connective tissue diseases comprise a group of syndromes of unknown etiology affecting as many as 1 person in 40, often with a predilection for the female sex." Rheumatoid arthritis affects about 1% of the US population, mainly with inflammation of the joints; lupus is less common, afflicting 0.1% of the population with joint, kidney, blood, and skin problems. Even more rare is scleroderma (0.01%), a connective tissue disease characterized by thickening and hardening of fibrous tissue. As with other autoimmune conditions, joint stiffness, fatigue, GI and lung involvement, Sjogren's syndrome (dry mucous membranes), and muscle aches are common complaints with scleroderma.

## Treating overlaps

There are differences between overlap and non-overlap myositis patients. In epidemiological studies, Miller notes that myositis is less severe when there is an overlapping condition than when a patient has myositis alone. In either case, though, doctors are treating the symptoms of the diseases because

there are no known causes for most of them. In reality, doctors treat each person's individual problems rather than the disease itself.

The treatments used are basically the same, as many of the treatments used for myositis are applied to the different autoimmune diseases. There are similar risk factors for the different autoimmune diseases as well - for instance, HLA alleles are found to be

associated with myositis, other autoimmune diseases, and overlap

**As with myositis in general, the first step is making sure the diagnosis is correct.**

syndromes.

As with myositis in general, the first step is making sure the diagnosis is correct. Is it really myositis, or is another condition mimicking myositis? Once the diagnosis is accurate, determine if the disease is active or if it's in the chronic, damaged phase. In the latter, physical therapy and rehabilitation are started to help with existing muscle function. Finally, it's essential to keep your own wishes and needs in mind in forming an acceptable, effective treatment plan.

Unfortunately, there are more unanswered questions than answered questions in treating myositis and overlap syndromes. There are some guides, including certain factors that can predict in many cases how you'll respond to therapy, but these factors apply to larger groups and not always to individuals. The ultimate goal, says Miller, is prevention.

Which treatment works best? Miller admits that all myositis patients are guinea pigs when they step into their doctor's offices. That said, doctors aren't randomly choosing medicines but making educated guesses using information from trials and other sources. In the end, they're

trying to fix what's bothering you, not treating each particular overlapping condition you may have.

More research will lead to more answers, and that's where you can help: contribute toward the funding of research projects, write your representative in Congress asking for support for autoimmune disease legislation, educate others about myositis, and participate in clinical trials when appropriate. There are currently many basic research projects, with promising trials on the horizon.

## Questions from you

**Is it worth trying alternatives in the absence of effective conventional medicines?** If conventional therapies aren't working for you, alternative treatments are available. However, be sure the risks are low. Alternatives aren't regulated like traditional medicines, so doctors don't know all of the possible side effects, says Miller. There aren't good controls over these methods.

**How long should you stay on a particular medicine to see if it's working? And how do you know the medicine is actually working?** A three-month trial period is adequate for most medicines, says Miller, but different medicines will have different time periods for showing effectiveness. To assess whether it's working, besides simply feeling better, doctors need to come up with better measures of disease improvement and other factors. Currently, a group of more than 100 doctors, called IMACS (International Myositis Assessment and Clinical Studies Group), is working together to standardize definitions in order to better compare results from research trials and clinical observations. The group is focusing on definitions for remission, flare, improvement, deterioration, and other terms. This standardization will lead to clearer answers in the future.

**How do you know you're in remission?** Again, says Miller, IMACS is working on definitions to help with questions like this. For now, remission means that a person is off all medicines and has had no evidence of disease activity for a period of six months. This is different from a complete clinical response, where you have no evidence of disease activity but are still taking medication to control the disease. Keep in mind that damage may still be there from past disease activity, he says, so even in remission, you may not feel quite "normal" even with normal test results.

**Why do previously effective drugs become ineffective?** Doctors don't always know why the medicines worked in the first place, says Miller. In cancer studies, it has been documented that patients can develop a resistance to certain medicines, so this may be the case with myositis patients as well, though there have been no studies on this.

**When you're being treated with a number of medicines for overlap, how do you know what's helping?**

This depends on a number of factors, he says. It could be related to the sequence in which you started each of the medicines, but it remains difficult to determine which medicine is actually working for you. Dr. Miller keeps his patients in a clinically controlled response for three to six months before tapering, then begins decreasing the most dangerous medicine first. He prefers to taper slowly and steadily to avoid flares.



**! Acupuncture helps in osteoarthritis of the knee**

Acupuncture, as a complementary therapy to drug treatment for osteoarthritis of the knee, is more effective than drug treatment alone, find researchers from Spain.

Osteoarthritis of the knee is com-

mon, affecting almost a tenth of the population aged over 55. The role of acupuncture in osteoarthritis remains controversial and few studies comparing acupuncture and drug treatment have been conducted. A total of 88 patients with osteoarthritis of the knee were randomly divided into two groups, one receiving acupuncture plus diclofenac (an anti-inflammatory drug) and the other dummy (placebo) acupuncture plus diclofenac. Treatment lasted 12 weeks and levels of pain, stiffness, and physical function were monitored using recognized scales. The acupuncture group had a greater reduction in pain and stiffness, improved physical functioning and quality of life than the placebo group. Although the 12-week monitoring period may be insufficient to evaluate the effects of treatment in the medium term, authors say acupuncture as a complementary therapy to drug treatment for osteoarthritis of the knee is more effective than drug treatment alone.



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# Confused about anti-inflammatories? Learn the facts

Drugs like Celebrex and others called "Cox-2s" are generally tolerated very well by many patients, which is fortunate because these drugs are often very helpful for people with pain and inflammation. Most side effects are minor and easily reversible by discontinuing the drug. The risk of serious side effects is small, but a little bit of understanding of the possible side-effects of these drugs can make them safer to use. If any of the following guidelines are not clear, or if you think it does not apply to you, discuss the issue with your physician.

Stop the drug and call your physician immediately if you have any severe abdominal pain or a black, tarry stool or any blood in your stool, which are signs of internal bleeding. Although the manufacturers say that these drugs can be taken on an empty stomach, to help reduce irritation of the stomach and prevent an ulcer, take non-steroidal anti-inflammatories (NSAIDs) at the end of a full meal, or with antacid. Limit alcohol intake since alcohol can irritate the stomach.

Celecoxib and valdecoxib should not be prescribed for patients with allergy to sulfa-containing drugs (sulfonamides). Such drugs include the antibiotic trimethoprim-sulfamethoxazole (Bactrim, Septra). If you are thinking of taking celecoxib or valdecoxib, make sure that you do not have a history of allergy to such drugs. If you aren't sure if a drug you had a bad reaction to fits in this category, ask your physician.

## Why Cox-2 agents became so popular

Cox-2 agents, like Celebrex, Vioxx and Bextra, became immensely popular in a very short time, not because they were found to be more effective than traditional NSAIDs like aspirin, ibuprofen and naproxen, but because of their perceived reduction in side effects. A study presented at the American College of Rheumatology

annual meeting said this attribute contributed substantially to a dramatic increase in prescriptions and influenced the way physicians prescribe rheumatology medication.

Traditional or non-selective NSAIDs are effective inhibitors of the enzyme cyclooxygenase (Cox) 1 and 2 and so are able to reduce inflammation and musculoskeletal pain. Since Cox-1 enzymes help maintain the body's internal stability and Cox-2 enzymes signal pain and inflammation, the agents that attack both sets of enzymes increase the risk for gastrointestinal complications like bleeding or ulcers in some patients. In 1999, the first selective Cox-2 inhibitor, celecoxib (Celebrex®), appeared on the market, followed by rofecoxib (Vioxx®), and valdecoxib (Bextra®). These Cox-2 agents selectively inhibit cyclooxygenase 2, the enzyme responsible for inflammation and pain, but they have no impact on the beneficial action of Cox-1, thereby reducing the incidence of GI side effects. This was seen to be a tremendous benefit to people suffering from chronic inflammatory diseases, including myositis.

Using source data on prescription sales from independent, chain, super-market, mass merchandisers and deep discount pharmacies, researchers evaluated the influence of the introduction of these Cox-2 inhibitors on the number of overall NSAID prescriptions written between 1998 and 2003. Results indicate that NSAID therapy prescriptions increased 67.7 percent during the five-year period primarily because of the availability of Cox-2 inhibitors. During the first year following their introduction, these Cox-2 agents accounted for more than two-thirds of the increase seen in NSAID prescriptions.

As might be expected, the new generation of inflammation fighters had a huge impact on the anti-inflammatory market, and they now repre-

sent roughly 44 percent of the total anti-inflammatory drugs other than steroids prescribed by doctors, said Bethany Fedutes, a Clinical Specialist in Drug Information at the University of Pittsburgh and an investigator in the study. Of course, traditional NSAIDs are available over-the-counter in lesser strengths than the prescription forms of these drugs.

## Savings on ulcer prevention never materialized

Because of the general expectation of reduced stomach symptoms in patients taking Cox-2 therapy, researchers were surprised to find that physicians have not decreased the number of prescriptions written for ulcer prevention medications such as proton pump inhibitors in patients with rheumatoid arthritis (RA) and osteoarthritis. Patients taking conventional NSAIDs for chronic inflammation often required additional medications to eliminate the potential stomach distress caused by such a regimen. Because Cox-2 therapies are thought to be less likely to cause ulcers, this was an unexpected finding, especially since those promoting the new class of drugs maintained that the additional expense of Cox-2 NSAID would be justified because of the assumption they would decrease the number of prescriptions for medications to prevent traditional NSAIDs' side effects.

Researchers conducted a three-year, semi-annual evaluation on 10,392 rheumatoid arthritis and osteoarthritis patients and found that those taking the drugs that were assumed to be without stomach complications were just as likely to be prescribed ulcer prevention therapy as those taking the cheaper NSAIDs.

## Your own heart and stomach should help you choose the right drug

The recent announcement that the drug Vioxx was being pulled from



pharmacy shelves because of heart and stroke risks left a lot of pain patients stunned, confused and worried. Some are even wondering if they should abandon medications that are in the same family as Vioxx, but doctors from the University of Michigan Health System say there are plenty of options to relieve pain and arthritis symptoms, and that patients should talk with their own doctors about what medication or combination of medications is right for them. In light of the effectiveness of traditional NSAIDs like aspirin and ibuprofen, patients may have more choices than they think.

The choice may come down to letting your heart or your gut decide - literally, say the doctors. Patients should be treated with medications based in part on their individual risks for heart disease and stomach problems, says Mark Fendrick, MD, a U-M professor of internal medicine who has studied the use of the family of medicines that includes Vioxx. In 2002, he co-created a pain-medication guideline for doctors that was published in *Pharmacy and Therapeutics* and adopted by the Michigan Quality Improvement Council. It took into account early evidence that Vioxx and its cousins might raise the risk of a heart attack.

"Every patient is different, but there are clear options even in the wake of the Vioxx situation," says Fendrick. "Patients should work with their clinicians to determine the best combination for them, and to persist until they get relief. No one should have to live with pain and other symptoms that interfere with their daily life." Fendrick's guide for treating patients' pain can be boiled down to a four-box grid that takes into account risk for heart disease and risk for gastrointestinal problems caused by NSAIDs. (See back cover.)

It's based on research showing that Vioxx and similar prescription drugs, traditional NSAIDs, are avail-

able both by prescription and over-the-counter. But either way, they're much less expensive than Cox-2 inhibitors. The grid also takes into account the fact that many people take another NSAID - aspirin - every day to reduce their risk of a heart attack. Taking aspirin with any other NSAID, including Cox-2 inhibitors, creates a combined effect that markedly increases the risk of gastrointestinal complication including ulcers and bleeding.

"Most patients and many clinicians are unaware of the fact that adding aspirin to a Cox-2 inhibitor takes away a great deal, if not all, of the gastrointestinal safety benefit," says Fendrick. "In fact, a recent national study showed that over 50 percent of Cox-2 users also take aspirin, and are therefore putting themselves at risk for ulcers and gastrointestinal bleeding." Add to that the added heart risk that Cox-2 inhibitors appear to carry, and it turns out that people who have suffered heart attacks, chest pains or strokes, or have a high risk for them, probably shouldn't take Cox-2 drugs at all. In fact, a warning to that effect was placed on Vioxx years ago. Many patients who take both kinds of NSAIDs benefit by adding a stomach-protecting drug called an acid blocker, available by prescription (under the names Nexium and Prevacid) or over the counter (sold as Prilosec).

"So, the bottom line is, patients should talk with their clinicians about their pain, their heart risk, and their risk factors for gastrointestinal complication from NSAIDs. Don't assume that what works for one person will work for you, or that risks or side effects are the same for everyone," says Fendrick. "And no matter what, be frank with your doctor about pain that you're feeling, because in the end you should be able to get relief."

## ! Painkillers, antioxidants found in tart cherries

The chemicals that give tart cherries their red color may relieve pain better than aspirin and may provide antioxidant protection comparable to commercially available supplements like vitamin E, according to Michigan State University researchers. The new findings "suggest that the consumption of cherries may have the potential to reduce cardiovascular or chronic diseases in humans (such as arthritis and gout)," write the scientists.

The research was published in the January 28, 2004 web edition of the peer-reviewed *Journal of Natural Products*, published by the American Chemical Society, the world's largest scientific society.

While cautioning that studies have not yet been conducted with human subjects, lead author Muralee G. Nair, PhD, says their laboratory assay results suggest that a person eating about 20 tart cherries could realize antioxidant or anti-inflammatory benefits. That number of cherries contain 12-25 milligrams of the active compounds, called anthocyanins, according to the authors.

In the study, anthocyanins were found to prevent oxidative damage, caused by oxygen or free radicals, about as well as compounds in commercial antioxidants. They also inhibited enzymes called cyclooxygenase-1 and -2, the targets of anti-inflammatory drugs, at doses more than ten times lower than aspirin. "It is as good as ibuprofen and some of the nonsteroidal anti-inflammatory drugs," says Nair.

"Daily consumption of cherries has the potential to reduce pain related to inflammation, arthritis and gout," adds Nair. While reiterating the need for human studies, he says a market may one day exist for putting the anthocyanins in pill form: "Then people can pop a pill instead of eating a whole bowl full of sour cherries. That's pretty hard to do."



## Experience helps when diagnosing IBM

*Anthony Amato, MD, is Chief of the Neuromuscular Division and Director of the Clinical Neurophysiology Laboratory at Brigham and Women's Hospital, Boston. He is also Associate Professor of Neurology at Harvard Medical School.*

By now, Dr. Anthony Amato trusts his clinical instincts when examining an IBM patient: "Here's a man over 50 who's weak, who has noticeable atrophy of his thigh muscles and says he's been getting slowly weaker for years but just thought he was out of shape. Now, you know and I know what he has. What is it?" The audience reply is unanimous: "IBM."

That's not only the first choice, Amato said, it's likely to be the second and third choices as well. Addressing the crowd at the Annual Conference, he asked another question: "How many of you were diagnosed with another disease - PM or ALS or some mixed connective tissue disease - before the correct diagnosis?" Seeing almost half of the IBM patients raise their hands, Amato nodded. The clinician's experience is the best tool for diagnosis, he said: an MRI will only diagnose signal changes, an EMG might miss it altogether. Even with some response to medicines, said Dr. Amato, he still questions the diagnosis. Steroids, for instance, may slow the progress of the disease, so there's a modest benefit. However, this improvement doesn't dissuade him from thinking the disease may be a dystrophy or IBM. Without a major response to treatment, he "scratches his head," considering different scenarios to reach the right diagnosis.

Polymyositis is not so easy to pin down, Amato said: "Most often it's a collection of syndromes that do not always have a lot of features in common." It's a hodge-podge, and if you don't respond to treatment, chances are you really have IBM. If it's estab-

lished that you truly have polymyositis, and standard prednisone therapy doesn't help you, it's time to really work with your physician, said Dr. Chester Oddis. "This is where it's essential to find someone who will communicate with you, who will try one treatment and then another and another until something is effective," he said. "All of us here get the very worst cases; and we also consult with other physicians from all over the world." Oddis said that treating the lung symptoms aggressively is extremely important. "If you have the JO-1 profile and lung disease, please pursue treatment," he said. There's no magic bullet, just an arsenal of choices that a skillful clinician can offer and monitor. "DM is something you can hang your hat on," he added. "With PM, that's often not the case." Dr. Fred Miller acknowledged that it's frustrating for both the physician and the patient when no exact diagnosis is possible. "We hope this will change," he said, "as we learn more."

Amato, who has published some well-received commentary on the confusion surrounding polymyositis, believes that the disease is very different from the classification accepted by many physicians. "It is not just dermatomyositis without the rash," he said. Neither is it one single disease with symptoms and syndromes alike in every patient. "What we are really seeing is that it is a collection of diseases that varies with every patient," he said. "With PM, I advise people to be proactive, to understand as much as possible about their own particular disease, and to keep working with the team of doctors they trust."

Even when patients diagnosed with assumed PM are stabilized using a combination of treatments like prednisone, methotrexate and cyclosporine, Amato still recommends retesting for IBM. He'd still wonder about the diagnosis of PM, he

said. A muscle biopsy doesn't always show inclusions, so the pattern of weakness holds clues to the proper diagnosis. If you need three drugs to control your disease but you still experience weakness, this raises questions.

### Questions from patients

For the Question-and-Answer session at the Annual Conference, Dr. Amato was joined by Frederick Miller, MD, PhD, the Senior Investigator and Chief of the Environmental Autoimmunity Group, NIEHS, at the National Institutes of Health. Dr. Miller serves on The Myositis Association's Board of Directors and as Vice Chair of the Medical Advisory Board.

### Treatment goals

**Q** With current treatments, are we treating the disease or the symptoms?

**A** "The reality is we don't know the cause of these diseases," said Dr. Miller, "so we are not directly treating those diseases with immunosuppressants." Since the immune system seems to be involved at some point in the disease process, doctors are currently blocking this system to prevent symptoms and further muscle damage. The fact that doctors can't say people are cured, he said, points to the fact that doctors are treating the immune response and not the disease itself.

**Q** I'm willing to take anything that might work for IBM. How does that fit into medical ethics?

**A** "We as clinicians take an oath to first do no harm," said Dr. Amato. Doctors must continually weigh risks and benefits of treatments, and the decision is individual to each patient. For some patients, having a greater risk for certain side effects or other conditions doesn't mat-



ter; for others, this decision is not as straightforward.

#### Alzheimer's treatment for IBM?

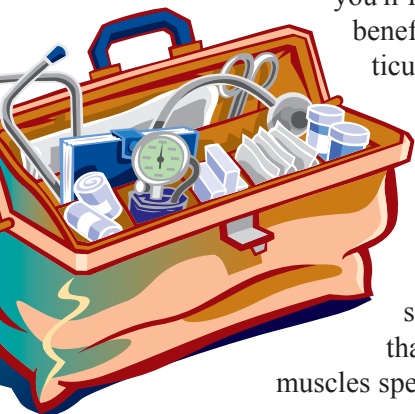
Q Medicines used to treat myositis at this point are off-label. Is there any anecdotal evidence that Aracept, a medicine used for Alzheimer's Disease, will be effective in treating myositis?

A "It's a misunderstanding," said Dr. Amato, "to think of IBM as Alzheimer's of the muscles." Researchers use Alzheimer's as a model to show that IBM could be a degenerative disease where inflammation is present, he said. In Alzheimer's, however, the problem is extracellular; in IBM, intracellular. Do the amyloids have something to do with the disease (IBM), or are they just innocent bystanders? That remains to be answered, he said. In the meantime, he sees no real reason to consider Aracept as a possibility in treating myositis, as Aracept targets the neurotransmitters of the brain.

#### Intravenous immunoglobulin

Q My husband takes intravenous immunoglobulin (IVIg) for his IBM, and the doctors feel his disease is stabilized because of this. They're afraid to take him off of this treatment. Is there any proof that IVIg is effective in treating IBM?

A In large double-blind trials testing IVIg for IBM, the treatment didn't show significant benefit in muscle strength, for example. Chances are, said Dr. Amato, you'll find some benefit in a particular muscle group, but it's hard to pinpoint unless you're studying that group of muscles specifically.



It's also difficult to determine if IVIg does slow the progression of IBM, even in the absence of dramatic improvement, as you'd need a relatively large number of subjects and a longer period of time for this type of study. In deciding whether or not to continue with IVIg, you should weigh the risks versus benefits, considering time and cost as well, he said.

#### Enbrel and Imuran

Q I've heard Enbrel loses its effectiveness over time, so my doctor is considering adding Imuran. Should I be concerned about long-term use of Imuran and possible liver damage?

A Enbrel has been shown to be effective in treating rheumatoid arthritis but not proven for myositis, said Dr. Amato. People do use it to treat myositis; some with benefit. There is no evidence of long-term liver damage. If damage were to occur, he said, you would see it early on in the treatment so that you could stop taking the medicine.

There is a higher risk of malignancy with certain medicines, but even with the risk being greater than the normal population, it is still a tiny risk, he said.

#### Stem cell research

Q Can you tell us more about stem cell research as it relates to myositis?

A There are some promising leads, said Dr. Miller, but clearer answers are still 5 to 10 years off. Researchers are limited by the Bush administration's decisions on stem cell research, and it's certainly a topic of much debate.

#### Oxandrolone for IBM

Q How effective is oxandrolone for IBM?

A Dr. Rutkove did one study in Boston looking at oxandrolone, but it didn't prove to be effective in treating IBM, said Dr. Amato.

#### Approval for new treatments

Q Other countries seem to be less rigid in their guidelines for approving and legalizing medicines. Is this the case?

A This is in fact the case, said Dr. Miller, but the Food and Drug Administration (FDA) seeks safety above all else. There is an increasing trend toward completing Phase I and II trials in other countries. These countries still follow a set of guidelines, but they are less rigid than those in the U.S. The FDA accepts some information from these trials, possibly helping to speed up the process for approving medicines.

It can take from several to 10 years to gain FDA approval for medicines. The first phase tests safety and tolerability; the second, efficacy. Further phases are needed to study the treatment in larger groups and with different conditions before approval is given.

#### Myostatin

Q I've heard myostatin mentioned in the news recently. Is there any hope in this for those with myositis?

A Myostatin, a protein that occurs naturally in your body, acts to slow muscle growth. There have been recent reports of animals and even one child without the ability to produce myostatin, resulting in unusually large muscles, said Dr. Amato. There are animal models to study the effectiveness of medicines aimed at inhibiting myostatin. This seems to improve muscle strength and to show some benefit under the microscope, he said.

Studies are currently underway to look at myostatin and certain muscular dystrophies. This is promising for those with myositis, he said. He is optimistic further studies are on the horizon, focusing on different disease groups including myositis.



# Smart bombs vs. shotguns: new medicines attack the disease, not the whole body

Dr. Todd Levine's fascination with some rare nerve diseases led him to the discovery that rituximab - a chemical specifically designed to deplete B cells - would also work for patients suffering from dermatomyositis.

Speaking at the Annual Conference, Levine said that a couple of myositis patients who crossed his path while he was using rituximab (marketed as Rituxan) for patients with IgM neuropathies had such a good response that he suggested it for a young woman whose dermatomyositis wasn't responding to conventional myositis therapy. The Arizona neurologist was pleased with the quick, dramatic results. After her roommate in the rehabilitation center requested the drug and had similar positive results, Levine designed a small trial of his patients and those referred by TMA. Improvement in the eight patients was impressive and the one side effect reported (due to a nursing error) was quickly resolved. Since one of the patients was a 12-year-old, Levine expects rituximab to be a welcome change from conventional therapy for juvenile as well as adult myositis patients.

A couple of slides Levine showed the conference audience demonstrated why B cells are considered the culprit in DM. B cells cluster around the portions of slides taken of muscle with obvious damage. B cells are what researchers call transient cells: they appear in the cellular transformation from stem cells to plasma cells. The need for a DM treatment that targets these cells is obvious. Levine said: conventional treatment with prednisone has the side effects of diabetes, bone thinning, weight gain and mood swings, sometimes so severe that patients can't continue taking it. (See Prednisone, page 4). That's because prednisone and virtually every other myositis drug targets the

entire immune system. The new targeted therapies - those that zero in on the disease process - are like smart bombs, Levine said. They attack the very specific cause of the disease, rather than using the shotgun approach of prednisone, methotrexate and others. Although Levine found improvement in all but one of the patients using rituximab, he can't explain exactly why. "We don't know how it works," Levine told his audience. "That's more common with medicines than you might think." One patient who had never had any treatment at all for her myositis did especially well and didn't require any other medication, which causes researchers to speculate that the "smart bombs" might eventually be desirable as the first choice for DM rather than the last resort.

In conducting his small private study, Levine worked with myositis patients from all over the country. A couple were not taking prednisone, and some were taking prednisone and other drugs when they enrolled in the study. Levine worked with their personal physicians, who gradually tapered the other medication as the rituximab took effect. Each patient improved quickly and sustained the improvement for several months. They relapsed at different time periods following the last treatment, and Levine found this could be reversed with sporadic continued treatment. Several continued to augment their rituximab treatment with small maintenance doses of prednisone.

Levine pointed out that the involvement of B cells is central to the disease process of dermatomyositis, which targets the blood vessels around the muscles rather than the muscles themselves; in polymyositis, the T cells appear to be more involved as they attack the actual muscles. But new rituximab studies,

led by TMA Medical Advisory Board member Dr. Chester Oddis, will begin at several centers, and they will include polymyositis as well as dermatomyositis patients, children as well as adults. The studies will begin in fall, 2005, and TMA will publish trial enrollment information as soon as it is available.

## A patient's story

*Lori Fisher is a long-time member of The Myositis Association and recently participated in a clinical trial studying rituximab (Rituxan). She shares her experience with us:*

At the time I heard of Dr. Levine's rituximab (Rituxan) study in relcalcitrant and long-term DM patients, my neurologist had just told me to take the big family vacation to DisneyWorld because there was nothing more he could offer me. I'd recently been hospitalized for 14 days with a pulmonary embolism following plasmapheresis, and the neurologist felt I would have to "wait for modern medicine to catch up with my condition." Fortunately, through TMA, I'd learned of the Rituxan study. When I visited my rheumatologist, Dr. Kathleen Price, I was doubly fortunate because her PA had worked with an oncologist and was familiar with Rituxan.

My first treatment with Rituxan was a resounding success although it didn't seem that way at first when two months afterwards, I had a DM rash, the beginning of my usual springtime flare. With the addition of 10 mgs of prednisone a day, I immediately felt stronger than I had in decades.

Thanks to the Rituxan/decadron (a corticosteroid medicine) infusions followed by maintenance methotrexate and prednisone, I was soon going hiking several times a week on the rugged paths of Bull Run Mountain in

Thoroughfare, Virginia. One fine day, I hiked a 6-mile loop trail at Manassas Battlefield Park. Thanks to Dr. Levine's research, the summer of 2003 is one for the memory books.

By December of that year, I needed additional treatment, and we repeated the Rituxan. I learned to respect the role B lymphocytes play in fighting infection. I think I used the strength from those infusions to fight off several different kinds of infections. (See JM and prednisone, page 6.)

I've recently completed my third course of Rituxan. Amy, Dr. Price's PA, has told me that everyone in the field is very excited about Rituxan, especially when it's combined with daily prednisone. Even though I don't take well to prednisone, I've agreed to try 5 mgs a day, to see if it helps boost me into another remission like the one in the summer of 2002. I don't know if I'll ever go back to the top of Bull Run Mountain, but if I close my eyes, I can remember what it was like to stand there on top of the big white rocks overlooking the Piedmont at 1,300 feet elevation. A priceless memory, thanks to Dr. Todd Levine's research!

- Lori Fisher

### Why a muscle biopsy?

When prednisone was the only tool available, it didn't matter so much whether the disease being treated was PM, DM or a collection of syndromes, Dr. Levine said. "Since it was just about all we had and it treated everything, it wasn't so important." Not only is this procedure, which gives the clinician a picture of the affected muscle, important for diagnosis, it's also very important as a beginning marker in the treatment process. Levine expects muscle biopsies to become routine in myositis as treatment becomes more and more targeted to very specific diseases.



### Myositis 101,

continued from cover

ments, and, for those with DM, sun precautions, topical corticosteroids and hydroxychloroquine (Plaquenil) for the skin involvement. If you have other factors like lung involvement, intravenous corticosteroids are used to control the disease more rapidly. Dr. Chester Oddis, a TMA medical advisor and professor at the University of Pittsburgh School of Medicine, suggests IV pulse methylprednisolone with the presence of lung problems but notes that immunosuppressants are often required as well. IV methods are also preferred by many when treating children to diminish long-term adverse effects.

The most common second-line therapies are methotrexate and azathioprine (Imuran). Intravenous immunoglobulin (IVIg) and cyclosporine also fall into this category. Often, combinations of medicines are used for more aggressive treatment. There is no standard for what combination works most effectively. "The reality is we don't know which patients will respond best," says Miller. Tacrolimus (Prograf), mycophenolate mofetil (CellCept), cyclophosphamide, and chlorambucil are all considered third-line agents.

Experimental therapies show some promise, including biologic agents like infliximab (Remicade). A current NIH study showed initial positive results, with some patients clearly responding to Remicade and others definitely not responding. The question of how many would fall into each category remains. In a couple of years, results from this trial will explain this disparity more clearly.

### Response to treatment

Doctors can't predict how you'll respond to any particular therapy, says Miller, but there are some general rules. As with all rules, he points out, there are exceptions. Doctors look at poor prognostic factors - for

example, older patients; those with cardiac, pulmonary or gastrointestinal involvement; and patients with a longer delay between disease onset and the start of treatment - when formulating a treatment plan.

With more research, Dr. Miller is optimistic that answers to the yet-unanswered questions are on the horizon. He encourages everyone to help raise funds for research, write Congress to increase funding for autoimmune research, and participate in clinical trials as they apply to you.



### Prednisone,

continued from page 6

Rather than adding another supplement, he advises patients to eat bananas for potassium. He recommends periodic eye exams to screen for cataracts.

"Sometimes adjusting the dose of prednisone will improve insomnia," he said. "Other times, the patient makes a natural adjustment after a few days." He has reports from patients who have been helped by melatonin, available over the counter.

*This material is compiled from lectures given by Dr. Chester Oddis, interviews with TMA medical advisors, and articles previously published by TMA.*



### Questions about nutrition? Use the hotline

Increasingly, research supports the notion that nutrition may be a key player in preventing illness and promoting healing. For instance, tomato sauce is high in lycopene, which may help prevent or slow the progress of prostate cancer. Many other fruits and vegetables may also have preventive powers. The Nutrition Information Service maintains the EatRight Nutrition Hotline to answer questions about good nutrition and health. Call the hotline at 1-800-231-3438.



# Patients find relief from dry skin, itching, rash

Many myositis patients, young and old, must deal with extremely dry, itchy skin, especially the cracked and scaly hands known as mechanic's hands. With the helpful advice of others, you can soothe this uncomfortable condition especially common in dermatomyositis (DM) and polymyositis (PM).

"Even when both skin disease and muscle disease is present, topical cream- and ointment-based medications can be applied to the skin rash in order to minimize the doses of internal medications that are required to control the entire disease," says Dr. Richard Sontheimer, John S. Strauss Endowed Chair in Dermatology, Professor and Head, Department of Dermatology, University of Iowa College of Medicine, and TMA Medical Advisory Board member.

## Ointments, oils, creams, and lotions

Moisturizers come in different forms - ointments, oils, creams, and lotions. Ointments most effectively trap the moisture in your skin, says Dr. Sontheimer, but they can also leave a greasy feeling. Use Aquaphor, Vaseline Petroleum Jelly, or another ointment in a small amount on the affected area, and rub it in well to avoid excessive greasiness. Oils, including baby and mineral oils, are still effective without being as greasy as ointments. Creams tend to be more popular since they disappear when rubbed in. Dr. Sontheimer cites Original Eucerin Cream, Nivea, and Neutrogena Hand Cream/ Norwegian Formula as popular brands that are frequently recommended by dermatologists. Lotions often contain alcohol, which may cause the skin to dry even more if used too often.

## Patient-to-patient recommendations

What do patients themselves recommend to each other when ordinary lotions don't do the trick? Dale Sjogren, a PM sufferer, stumbled upon Badger Balm at his local hardware

store. He coats it on his rough spots and covers them with a Band-Aid overnight. "It generally heals a crack within 12 hours," he says.

Beth, another PM patient and Bulletin Board regular, uses No-Crack hand cream she found at [www.restorationhardware.com](http://www.restorationhardware.com). "My fingertips still split and crack at times," she says. "But not near as much when I am using this."

Two more frequent posters on the web site's Bulletin Board, Dorris Norris and Lea Jaeger, both praise MSM Cream. Dorris spotted this remedy at her health food store; Lea discovered a mail order web site, Kala Health Inc., willing to send it to her in Israel. "I have IBM and also have fingers that split, bleed and get very sore," says Dorris. "[MSM Cream] heals the places quickly."

Other Bulletin Board users volunteered these helpful products: L'Oreal's Ombrelle 60, a light, non-greasy, and fragrance-free sunscreen that protects against both UVA and UVB rays; Aquaphor, a fragrance- and preservative-free lotion that comes in original or healing ointments; and Dermoplast, a pain relieving spray that numbs and soothes the affected area. You can find most of these products at your local pharmacies, including CVS ([www.cvs.com](http://www.cvs.com)) and Walgreens ([www.walgreens.com](http://www.walgreens.com)); Cheri (DM) came upon L'Oreal's Ombrelle 60 online at [www.feelbest.com](http://www.feelbest.com).

## Scalp solutions

"Because the hair interferes with the application of creams and ointments, the scalp requires a corticosteroid chemical dissolved in other vehicles such as solutions, gels, sprays, or foams," says Dr. Sontheimer. "A potent corticosteroid solution massaged into the still-moist scalp after shampooing can provide the greatest benefit from this form of therapy." In answer to a fellow Bulletin Board user, Jane, a DM patient, recommends

Luxiq (betamethasone valerate), a topical corticosteroid, for scalp itch. This medicine is available as a prescription foam for scalp rashes. "It actually works like a mousse and gives your hair fullness while helping your scalp," she says. She also offers fluocinolone acetonide topical solution (also known as Derma-Smoothe/FS) and desonide ointment, both prescription medications.

## Mummy treatment

Sometimes the substances alone are not enough. For Andrea Clausen, nothing compared to the wet-pack, or mummy treatment prescribed by the Mayo Clinic to help her skin more effectively absorb her medicated creams.

Andrea applies wet dressings of soft, white, 100 percent cotton material with lukewarm water (avoid hot or cold), squeezing out the excess water so the material was damp but not dripping. She then applied the prescribed cream to her skin, covering it with the wet dressings. She treated her stubborn rash for thirty minutes twice a day, noticing dramatic results within two weeks.

When finished with the treatment, she reapplied the creams or ointments. If mechanic's hands bother you, try Dertpak Gloves to cover the lotion, ointment or cream you choose. Andrea treats her hands when they're irritated, spreading cream over her hands and wearing the gloves to bed until her rash improves.

## Prevention through sun protection

Protection from the sun's harmful rays guards against drying out your skin even more. Use sunscreen with Sun Protection Factor (SPF) 15 or higher, and apply as much as you would lotion on dry skin. If you don't use the proper amount of sunscreen, the SPF actually decreases. Be aware that certain medications, including methotrexate and tetracycline, increase your

sensitivity to the sun, so read the labels carefully. Using the Bulletin Board to solicit peer advice, Cheri sought information on sunscreens with mexoryl to prevent a sun-induced flare, as recommended by her dermatologist. Liesl Dutro offered Anthelios "L" SPF 60 Sun Block as an option, though Chris's response mentioned his recollection of its strong scent and greasy feeling.

Sun protection is also available in certain cosmetics, including Dermablend and Covermark. These products blend physical sunscreen with cover-up. Dermablend Corrective Cosmetics promise natural-looking coverage that won't clog your pores, are fragrance-free, and resist smudges. Even more important to those with skin problems, however, is the SPF 30. Covermark also offers sun protection, with SPF 20 "Face Magic" and SPF 16 "Leg Magic" that won't rub off or smudge. Covermark's cosmetic camouflage is 100 percent waterproof and highly pigmented.

The increasing emphasis on sun safety has spurred companies to create more stylish, comfortable sunprotective clothes - clothes with the protection woven directly into the fabric. You'll find swimwear, business and polo shirts, track pants, hats, driving gloves, canopies, umbrellas, and much more. It's important to understand the labeling, which the Federal Trade Commission monitors carefully. The Ultraviolet Protection Factor, or UPF, measures how much of the sun's radiation is absorbed by the fabric. So a UPF of 20 means that 1/20th of the radiation passes through the clothing; or in other words, the fabric reduces your exposure by 20 times. A UPF value of 15-24 is considered good protection against UV rays; 25-39, very good; and 40-50, excellent. Some UV protection may be lost if the clothing is overly worn or washed, wet, or stretched out. Enter "sun protective clothing" in your Internet search engine to find a list of companies stocking these products.

There are times you may not consider sunscreen to be essential, like while driving or going for a short walk. Windows and car glass do not block the UVA rays, prompting some companies to offer UV light blocking film. Fluorescent lights also emit some UV rays, and you can place acrylic diffusion shields over the bulbs for protection. For these protective films and shields, Dr. Sontheimer suggests the following companies: Southwall Technologies Inc., North Solar Screen, and Llumar. The film coatings absorb or reflect from 96 to 99 percent of the sun's harmful UV rays. North Solar Screen shades roll up and down so that you can use them according to your different needs for day and night, and they offer bulb jackets to provide UV protection from fluorescent lights.

#### Direct treatments for JM rash

Though prednisone and other medicines help the rash that comes with JM, sometimes it's necessary to put ointments directly on the rash to make it heal. Tacrolimus (Protopic) and pimecrolimus (Elidel) are two of the topical creams used to more aggressively fight the rash.

Julia Becker, now in remission from her JM, participated in a research study looking at the effectiveness of Protopic on the JM rash. She applied 0.1% topical tacrolimus ointment twice a day only on the parts of her skin with a rash. "We didn't really see any effect at first, but within a few weeks, Julia's rash started getting noticeably better," says Ralph Becker, Julia's father. "Over the following months, she continued to show a gradual improvement in the rash and after about nine months of use, her rash was gone and we were able to discontinue using Protopic." The researchers concluded that Protopic may be useful in helping the JM rash, but more studies are needed.

"We had a strong family history of eczema, so it seemed likely that our kids would get it," says Shari, a JM

mom. "The most helpful topical cream we have found is Elidel." Her son Ricky, another JM patient, has also found success with nightly applications of Elidel cream.

*This information first appeared in the 2003 Outlook Extra Treatment Issue.*



#### Dermatologist gives tips for winter skin care

In the winter, it feels good to wrap yourself up in heavy sweaters and turn up the heat in your house. But have you given any thought to what's happening to your skin as it gets colder? That dry itchy skin you get in the winter can be a direct result of trying to keep warm. However, winter skin can be prevented with a few simple changes to your skin care routine.

"The hot air inside your home or office can do more damage to your skin during the winter than the cold air outside," said dermatologist Stephen Webster, MD, Clinical Professor of Dermatology at the University of Minnesota Medical School. "When the relative humidity inside drops below 60 percent, your skin begins to lose moisture, causing that dry, itchy, flaky skin that irritates so many during the winter months."

The skin is made up of several layers of cells. The epidermis, the top layer of the skin, along with the oil glands, produce lipids, and these lipids keep the skin from losing moisture and make it soft and supple. But your skin is constantly losing moisture into the air and every time you wash your skin, you strip away these lipids, letting more moisture evaporate and drying the skin. However, in humid conditions, the skin can replenish itself by soaking up moisture from the air. So, when the humidity drops, as it does in many places in the winter, your skin loses another opportunity to moisturize itself. Couple that with the low

*See Skin tips, next page*

## Skin tips,

continued from page 23

humidity of indoor heating, and hotter showers and baths, and your skin can become dry and irritated.

It's these cold weather activities, such as taking hotter showers and turning up the heat that can cause dry skin during the winter. To correct some of the misconceptions that can lead to dry winter skin, Dr. Webster provides the following facts:

**Myth:** The hotter and longer the shower, the warmer I'll feel.

**Fact:** Sure, you'll feel warm, says Dr. Webster, but only for a few minutes. As soon as you step out of the water, your skin begins to lose moisture because hot water removes natural oil from the skin, making it dry and itchy. Bathe or shower in lukewarm - not hot - water, and limit your showers to 5 to 10 minutes.

**Myth:** Switching your brand of soap can "confuse" skin, leading to irritation.

**Fact:** All soaps have the potential to cause contact dermatitis, especially if you are allergic to certain ingredients in the soap, says Dr. Webster. It is perfectly fine to use the same brand of soap throughout the year. However, if you find your skin becoming drier in the winter months, look for a milder soap that is fragrance-free. In fact, many soaps today contain moisturizing ingredients, like oils and vitamins, which can be beneficial for your skin all year round.

**Myth:** Completely dry your skin before applying lotions and creams.

**Fact:** Dr. Webster recommends applying moisturizers to skin within three minutes of stepping out of the shower or bath. Putting on a cream, ointment or lotion helps trap the water in the upper layers of the skin and decreases dryness and itching. Find a cream or lotion with a lighter texture to avoid greasiness.

Since severely dry skin is less effective at providing a barrier against

infection and can split and bleed, creating a greater chance for an infection, Dr. Webster gives these tips for healthier winter skin:

■ Remember, snow can reflect more than 80 percent of the sun's damaging rays, so be sure to always wear a broad-spectrum sunscreen with an SPF of 15 or higher. Reapply it every two hours for maximum benefit.

■ Don't forget to protect your lips. Look for a lip balm with an SPF of 15 to help prevent chapped lips.

■ Consider purchasing a humidifier to keep the humidity in your home higher during the winter.

■ Dab petroleum jelly on problem areas to seal in moisture and heal very dry skin.

■ After washing your hands, immediately put on hand cream to seal in moisture.

"The winter months don't have to be torture for your skin," says Dr. Webster. "Remember to place a greater emphasis on moisturizing and visit your dermatologist if your skin becomes infected or if you don't see any improvement in your skin."



## Short comments from the conference

During the medical panel and the question-and-answer period, Drs. Oddis, Miller, Amato and Levine answered questions from the audience.

### On Campath

With B cells eliminated in DM patients, they improve: why not try this approach to the T cells in IBM and possibly PM? "It's a dangerous thing to wipe out all of your T cells," says Dr. Levine. That's what's happened to AIDS patients. Campath, a treatment now being studied at NIH, affects the T cells and may have some promise, Dr. Oddis says. To find out more about the study, go to [www.myositis.org](http://www.myositis.org) (resources) or call NIH at 301-496-4000.

### On cancer and transplant drugs

Why use the "old" drugs taken from cancer and transplant treatment, when these disciplines have moved on to new drugs? "We're looking for some kind of history," says Levine. "Even the old drugs don't have a history with myositis." He added that it was easy to wipe out the immune system. The hard part is retaining the helpful immunity that everyone needs to survive.

### On stem cell therapy

It's political and poorly understood, say Levine, Amato and Miller; and all believe it shows promise in myositis, noting that everyone is born with the same amount of muscle fiber they will die with. Miller says working with the tools available to scientists under current Bush administration policy is like having a hand tied behind your back.

### On the placebo effect

Scientists must have blind studies with placebos before truly knowing the potential of a new drug. Here's why: Levine quoted a headache study where 40 percent of those getting the placebo reported improvement. To get a better sense of the effect, researchers released the 40 percent from the trial and tried again. The percentage of those receiving the placebo and reporting improvement remained at 40 percent, indicating that the placebo effect is more powerful than the researchers had anticipated.

### On cancer and dermatomyositis

Cancer is present in 20 to 30 percent of DM patients over 50 within five years, says Levine. If the cancer is treated and resolved, the DM often goes away. If more than 20 years have passed between the time of the discovery of DM and the discovery of cancer, they are not related, he says. Levine recommends a full body scan for anyone with DM: a procedure that will screen for the cancers that most commonly affect DM patients.





# Anti-TNF agents: here for the long term?

Etanercept (Enbrel), adalimumab (Humira), and infliximab (Remicade) are self-injectable medications known as biologic response modifiers that work to suppress the tumor necrosis factor (TNF) proteins associated with inflammation. Suppressing tumor necrosis factor has been an effective treatment in the day-to-day lives of many patients with rheumatoid arthritis, and a number of myositis patients have found etanercept helpful.

All three TNF agents are known to be very effective in the treatment of rheumatoid arthritis (RA) but since they are fairly new drugs there is limited information about their long-term effectiveness and safety. A seven-year study of treatment of patients with early and long-standing rheumatoid arthritis with etanercept found continued effectiveness and no increase in toxicity over time, findings that may be useful in predicting their effect in myositis patients taking them over the course of several years. Similarly, a four-year study of adalimumab plus methotrexate in the treatment of long-standing moderate to severe rheumatoid arthritis reported sustained improvement in arthritis with no new safety concerns. Both treatments allowed patients to take lower doses of corticosteroid medications and methotrexate.

Etanercept was approved in 1998 for RA and is prescribed to reduce the signs and symptoms of RA. It inhibits the progression of structural damage, and improves physical function in patients with

moderate to severe disease; and it can be used alone or in combination with

methotrexate. Neither etanercept nor any other drug has been approved for treatment of myositis, but there are studies and informal reports of its use in myositis. Adalimumab was approved in 2002 and is prescribed for reducing signs and symptoms of RA and inhibiting the progression of structural damage in adult patients with moderately to severely active RA who have had an inadequate response to one or more disease-modifying anti-rheumatic drugs (DMARDs); it can be used alone or in combination with methotrexate or other DMARDs.

"The efficacy and safety of adalimumab seen in six-month clinical trials continues in long-term follow-up for up to four years," said Michael Schiff, MD, Director, Clinical Research, Denver Arthritis Clinic, and lead investigator in the study.

Schiff was encouraged by the continued effectiveness and safety of etanercept: "We report the longest experience with etanercept and show that it continues to be effective with long-term therapy in patients with active rheumatoid arthritis and not only allows for a reduction in background corticosteroid doses but is well tolerated and serious side effects are rare," said Michael Weinblatt, MD, Professor of Medicine, Brigham and Women's Hospital and Harvard Medical School, and lead investigator in the study. Dr. Weinblatt presented his study at the annual meeting of the American College of Rheumatology.

Proper management can reduce post-surgical infections in patients taking etanercept

One potentially serious side effect of TNF can be prevented and was dis-

cussed in a recent study. The same TNF protein that causes symptoms in autoimmune disease also plays a key role in suppressing infections with certain bacteria in the body.

Researchers found that continued use of TNF inhibitors before surgery could increase the risk of many types of infections, such as septic arthritis, osteomyelitis or deep wound infection, following surgery.

To assess this post-operative risk, researchers evaluated the outcome of 91 rheumatoid arthritis patients, average age 59.5 years, who underwent

bone or joint surgery between January 1, 1999 and March 15, 2004. Patients who developed deep bone or soft tissue infections within 30 days after surgery

were identified and their medications were reviewed.

Of 35 patients receiving treatment with a TNF inhibitor at the time of surgery, seven developed a post-operative infection. In contrast, only three of 56 patients not receiving a TNF inhibitor at the time of surgery developed an infection. TNF inhibitor use was associated with a four-fold increase in risk for infection.

TNF inhibitors such as etanercept, infliximab and adalimumab can be discontinued and restarted without impairing the health of patients. This study concludes that, since each drug has a distinct half-life, patients should ask their physician for pre-surgery guidelines.

TMA members using infliximab (Remicade) report careful monitoring by their doctors, who discontinue treatment at any sign of infection, then resume it when the infection is past.

Since they are fairly new drugs, there is limited information about their long-term effectiveness and safety.





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## Recent Changes in Medicare May Affect Where You Receive Your IVIG Treatments

Effective January, 2005 Medicare has changed the way it pays for IVIG. If you have been receiving IVIG, your treatment will still be covered. However, you may need to receive your treatment in a different location.

IgG America, a national pharmacy specializing in IVIG, can provide your treatment at home if you have secondary insurance.

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## Medicines and their side effects

Side effects are an unwanted but often common part of taking certain medicines. Some of these effects are better known – like weight gain with steroid medicines – while others are less familiar. In some cases, these possible side effects are enough to make you think twice before starting a treatment. Below is a list of potential unwanted effects of medicines more commonly used in treating the different types of myositis. Not all effects are listed, and not all that are listed require immediate medical attention.

Be sure to talk to your doctor about what you should expect when taking medicines, and keep a journal of how you feel each day when you start a new treatment. This will help you explain to your doctor how the medicine makes you feel – physically and emotionally.

Generic name	Brand name(s) *	Possible side effects
<b>Alemtuzumab (IV)</b>	Campath	Black stools; chills; cough; diarrhea; dizziness; fever; headache; hives; nausea/vomiting; shortness of breath; sore throat; stomach aches; back pain; tingling; indigestion; decreased appetite.
<b>Azathioprine (Oral, IV)</b>	Imuran	Reduced immunity; loss of appetite; nausea/vomiting; skin rash; cough; fever/chills; back pain; painful urination; unusual fatigue; liver problems; anemia; delayed onset cancers possible.
<b>Corticosteroids (Oral, IV, Topical)</b> [Corticosteroids are a type of medicine; this is not the generic name. There are a number of common names for corticosteroids, listed here.]	Prednisone, Prednisolone, Methylprednisolone, Dexamethasone, Betamethasone; <i>Topical</i> : Elocon, Luxiq	Lower resistance to infection (also harder to treat infections); increased appetite; weight gain; restlessness; rounded, “moon”-like face; stunted growth (in children); acne; nausea; indigestion; moodiness, irritability; decreased appetite; thinning bones; abnormality of fat distribution; diabetes; menstrual irregularities; hypertension; cataracts; insomnia; increased thirst; continual stomach pain; vision changes; headache; irregular heartbeat; increased hair growth. Your body needs time to adjust when stopping this treatment. Tapering, or slowly lowering the dose you take, is essential. Report any side effects to your doctor as you are tapering or when you are completely off this medicine.
<b>Cyclophosphamide (Oral, IV)</b>	Cytoxan	Darkening of skin, fingernails; loss of appetite; nausea; vomiting; diarrhea; abdominal pain; facial redness; headache; temporary hair loss; cough, hoarseness; fever, chills; pain in low back or side; dizziness; shortness of breath; pulmonary fibrosis; blood in urine (even after treatment stops - check with doctor); secondary malignancies; sterility. Some effects of cyclophosphamide may not occur for months or years after using this medicine.
<b>Cyclosporine (Oral, IV)</b>	Neoral, Sandimmune	High blood pressure; kidney problems; increased hair growth; trembling hands; tender or bleeding gums; fever; chills. Doctors may monitor kidney function while on this medicine.

<b>Etanercept (IV)</b>	Enbrel	Abdominal pain, nausea, vomiting (more in children); throat pain; runny/stuffy nose; diarrhea; loss of energy; chills; cough; fever; skin rash; sneezing; chest congestion; dizziness; fast heartbeat; frequent urination; headache; joint or muscle pain or stiffness
<b>Hydroxychloroquine (Oral)</b>	Plaquenil	Diarrhea; headache; itching; decreased appetite; nausea/vomiting; stomach pain; dizziness; restlessness; change in vision; ringing ears; weakness; drowsiness/headache/excitability signs of possible overdose
<b>Infliximab (IV)</b>	Remicade	Abdominal pain; cough; dizziness; headache; congestion; nausea; shortness of breath; unusual fatigue; back pain; chest pain; fever/chills; facial flushing; hives; itching
<b>Interferon beta-1a (IV)</b>	Avonex	Heartburn; indigestion; trouble sleeping; chills; diarrhea; fever; flu-like symptoms; unusual fatigue
<b>Intravenous immunoglobulin (IVIg)</b>	(Also called immune globulin intravenous or IGIV)	Fever/chills; backache; headache; joint pain; nausea/vomiting; leg cramps; rapid heartbeat; skin flushing; troubled breathing; feeling lightheaded; changes in blood pressure; feelings of discomfort
<b>Methotrexate (Oral, IV) [also called amethopterin]</b>	Methotrexate, Rheumatrex	Kidney, stomach or liver problems; temporary hair loss; acne; loss of appetite; nausea/vomiting; diarrhea; stomach pain; malignancies; lung problems/inflammation; delayed onset cancers
<b>Mycophenolate (Oral, IV)</b>	CellCept	Constipation; diarrhea; headache; indigestion; nausea/vomiting; cough; fever/chills; back pain; shortness of breath; mouth sores; white patches in mouth
<b>Pimecrolimus (Topical)</b>	Elidel	Burning or itching in hairy areas of skin; skin blemishes; earache; stomach pain; chills; flu-like symptoms; congestion; fever; loss of appetite; muscle aches; headache; warmth on skin; hives; swelling
<b>Rituximab (IV)</b>	Rituxan	Back pain; increased cough; loss of strength; muscle aches; night sweats; pain; rash; sore throat; stomach pain; anxiety; dry mouth; fatigue; headache; increased hunger and thirst; fever/chills; nausea
<b>Tacrolimus (Oral, IV)</b>	Prograf	Kidney problems; reduced ability to fight infections; high potassium in blood; low magnesium in blood; high cholesterol; high blood pressure; abdominal pain; anxiety; chills; diarrhea; fever; flu-like symptoms; decreased appetite; nausea; tingling; difficulty sleeping; delayed onset cancers
<b>Tacrolimus (Topical)</b>	Protopic	Itching skin (in children); headache; fever; bothersome cough; loss of appetite; hives; chills; flushing; swelling; increased skin sensitivity

\* There are a number of brand names for certain medicines. The most common are included here.

## Stem cells from human blood help repair mouse muscles

Regeneration of damaged hearts using blood stem cells now appears to be clinically promising, say Texas researchers who show that in mice, human stem cells use different methods to morph into two kinds of cells needed to restore heart function - cardiac muscle cells that contract the heart as well as the endothelial cells that line blood vessels found throughout the organ. Using a sophisticated way of examining the "humanness" of mouse heart cells, researchers report in the December 21 issue of the journal *Circulation* (which was published online December 13) that two months after mice with ailing hearts were treated with human stem cells, about two percent of cells in their heart showed evidence of a human genetic marker.

Furthermore, researchers described, for the first time, how these human master cells use different ways to become two distinct kinds of cells needed in the heart. Human stem cells primarily "fuse" onto mouse cardiac cells to produce new muscle (myocyte) cells that have both human and mouse DNA. But to form new blood vessel cells, they "differentiate" or mature by themselves, presumably to patch damaged mouse blood vessels with human cells.

These findings should help resolve debate within the field as to whether stem cell transfer actually creates new types of cells that last within a heart, says the study's lead author Edward T. H. Yeh, MD, professor and chair of The University of Texas M. D. Anderson's Department of Cardiology.

"We have shown that these stem cells create both types of tissue needed to repair areas of damage, that they use two different ways to develop them, and that these cells can persist for up to a year, which is a long time in the life of a mouse," he says. "Most of all, this study is important

because it begins to explain why stem cells can help a heart heal," he adds. "Clinical trials that use bone marrow stem cells in people with heart damage have shown promise, but no one knows how it works. This starts to provide an explanation."

Yeh and his research team, which includes investigators from the Texas Heart Institute, have been looking for a relatively simple way to help restore the functioning of hearts damaged by chemotherapy, which can occur in up to 10 percent of cancer patients treated with such drugs, he says.

"Deriving stem cells from bone marrow is a complicated matter. It would be much easier for patients if the stem cells were taken from blood. It would be as simple as a blood donation. Cell fusion has been important for muscle growth, Yeh said. Myocytes fuse to form a muscle and the muscle regrows fused. "This paper shows that fusion is a predominant event in muscle cell generation, and it may work by allowing a cell to enter the cell cycle and divide and produce new progeny, but all of this is new and needs to be studied further," Yeh says.



## Complementary and alternative medicine use widespread in pediatric pain management programs

The use of alternative medicine has increased dramatically in the last decade, and complementary and alternative medicines are now used by 50 to 70 percent of children with chronic conditions, yet there was no data on complementary and alternative medicine in university-affiliated pediatric pain management programs.

In a study by Yuan-Chi Lin, MD, MPH, visiting associate professor, Department of Anesthesia, Children's Hospital Boston, designed to assess the availability of complementary and alternative medicine in the United

States and Canada, 52 pediatric pain management programs were identified and 43 (83 percent) responded to a telephone survey directed to a physician or nurse from the pain service program. The survey revealed that complementary and alternative medicines are used in more than 70 percent of university-affiliated pain management programs. Services offered included biofeedback (at 63 percent of the centers); imagery (60 percent); relaxation therapy (60 percent); massage (49 percent); hypnosis (44 percent); meditation (44 percent); acupuncture (40 percent); art therapy (28 percent); music therapy (23 percent); self-help groups (19 percent); therapeutic touch (14 percent); herbal remedies (9 percent); chiropractic (7 percent); yoga (5 percent); and Tai-Chi (2 percent).

Of these 43 institutions, 30 (70 percent) have pediatric pain clinics for chronic pain. According to Lin, who is the director of Pediatric Pain Management Service at Lucile Packard Children's Hospital at Stanford University and associate professor of Anesthesia and Pediatrics at Stanford University School of Medicine, currently on sabbatical at Harvard, research comparing the clinical outcomes and cost effectiveness of complementary and alternative medicine to conventional therapies is of pressing importance.



## Quick tips for healthy bones:

- Increase calcium by choosing milk, cereal and fruit juice fortified with vitamin D, leafy greens, cheese and other calcium-rich foods.
- Get plenty of protein.
- Exercise as much as possible.
- Avoid the "teen-age" diet of high-fat, high-sugar junk food and high-sugar soft drinks.





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## 2004 Nobel Prize Winner to participate in TMA IBM Conference

Dr. Aaron Ciechanover, winner of the 2004 Nobel chemistry prize, will be participating along with nearly 30 physicians in TMA's conference on sporadic inclusion-body myositis scheduled for January 26-28, 2005 in Marina Del Rey, California. Dr. Ciechanover of the Israel Institute of Technology was one of three researchers to share the Nobel prize for research in ubiquitin and proteasome inhibition in neurodegenerative disorders. There is growing evidence that proteasome inhibition plays a role in IBM, and Dr. Ciechanover will present on this topic as well as coordinate the final Conference session, "What

are the most promising aspects of pathogenesis leading to treatment of IBM?"

The Myositis Association is sponsoring this first-ever international scientific conference, "Frontiers of Research Potentially Relevant to Treatment." It is by invitation only and includes the world's foremost s-IBM physicians and researchers. TMA Medical Advisory Board members Drs. Valerie Askanas and Marinos Dalakas are organizing this event for invited experts to present and exchange ideas on s-IBM and to discuss promising research directions for s-IBM such as the role that aging

plays, unique aspects of the disease and its symptoms, and the roles of inflammation, beta-amyloids, cholesterol, oxidative stress and protein folding. This exciting, historic exchange of ideas in an atmosphere of collaboration and trust is expected to both advance the global understanding of IBM and conclude with a consensus on the most fruitful avenues for future research.

*The Muscular Dystrophy Association is providing funding to cover the costs of publishing and distributing the s-IBM Conference report.*



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