

Meet Our Members: Joseph and Monica Pedano

Mother finds support from family, friends, TMA

It was summer when Joseph showed signs of sunburn, so Monica Pedano, his mother, wasn't immediately concerned. When the red color remained, she checked with his doctor and he confirmed that the Pennsylvania sun was likely the culprit.

Later, though, there were more alarming signs. Joseph, a strong 18-month old toddler who had been walking since he was nine months old, had difficulty bending down to retrieve something from the floor. "He would shake like an old man with the effort," Monica said. When he fell, she said, he looked like a turtle, with no control over his torso or legs. He simply didn't have the strength to get up.

By October, Joseph was finally diagnosed with juvenile myositis, partly from a blood test that showed his aldolase level was alarmingly high. Aldolase is a blood enzyme that's useful for measuring the presence and severity of myositis or muscular dystrophy. He was admitted to the hospital and remained there as doctors fought to get his muscle inflammation under control. Monica, a financial advisor, stayed by his side, often working from his hospital room.

Joseph's was to be a long struggle. His rash was difficult, his muscle weakness was serious, the side effects of high-dose steroids were unpleasant, and he developed calcinosis, hard and painful calcifications, in his legs.



Joseph has always enjoyed sports

For the next eight years, his life remained pretty much the same, his mother said. He was in and out of the hospital and doctors' offices, and on multiple drugs. Through it all, he remained as active as he could. "He did everything," Monica said. He was on sports teams in school, often in a leadership role, and he took setbacks in stride, working harder and harder to get back to his active life after a hospitalization or a spell of weakness. Try as they might, Joseph's doctors just couldn't find the right combination of drugs to break the cycle of flares and remission.

Finally, in 2008, Joseph was included in a study of rituximab, a drug that had been effective in treating children with juvenile myositis in a small, informal study. "He immediately was a million times better,"

Monica said. Joseph continues to be athletic and strong, as determined to succeed in competition as he was to recover from his disease.

From her long and stressful experience, Monica has some ideas for families just starting out on a juvenile myositis journey.

Find support

"I was lucky to have supportive family and friends," she said. Friends and relatives rallied around her, helping her family to have normal moments. "I couldn't give Joseph his cyclosporine shot," Monica said, "so we'd have 'taco night' every week at my sister's, (she's a physician) and she'd do it." Monica also found support at The Myositis Association, where she eventually became a member of its board of directors. Being active in TMA made a difference, she said, from finding out about trials to having access to medical advisors. "Everyone was always willing to answer my questions. If you remain isolated, you just don't know what's going on."

Joseph was about to have his second calcinosis surgery at the time of this interview, and Monica said TMA's medical advisors helped her find the best surgeon for this. "You just can't imagine how helpful this is," she said.

Find the best medical care

"I was very fortunate to have extremely good doctors," Monica said. She

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Monica and Joseph, cont'd from cover

advises parents to keep searching for someone who is knowledgeable about juvenile myositis and willing to keep up with new treatments.

Take care of yourself

"You've got to be healthy and find a way to manage stress; otherwise you just can't do it," she said. Not only that, but she found that her state of mind really influenced how Joseph and his brother handled the long struggle.

Remember that children see illness differently

"Joseph never even expressed the idea of 'why me?' or 'this isn't fair,'" Monica recalled. "I can't tell you why, but children just are different in how they approach their situation." In the long run, she believes his illness

strengthened Joseph emotionally. Joseph, now 12, handles adversity of all kinds very well, she said. "He never uses his illness as an excuse not to do something - if anything, he tries harder because of it."

Balance the needs of other children

Joseph has a brother two years younger, and the whole family took pains to make sure he had a life and interests of his own, apart from his brother. "Naturally, Joseph took up a great deal of our time," Monica said. "But we made up for it as best we could."

Learn everything you can

"This was another reason that I relied on TMA," Monica said. "The more you learn, the better able you are to make good decisions."



A family affair: Pedano Annual Golf Tournament

What to do when you have a child that you love, a passion for golf, and friends and relatives who want to help? Answer: You organize one of the most successful golf tournaments to ever benefit The Myositis Association and juvenile myositis research. In 2002, Monica Pedano and her family held their first of nine golf tournaments to raise funds and provide fun for everyone who cares so deeply for Joseph. The tournament is now entering its 10th year and has raised over a quarter million dollars. This year's tournament is scheduled for June 8. Monica's enthusiasm, boundless energy and determination caught the eye of TMA's board of directors, and in 2007 Monica was invited to add her perspective and passion to TMA's board. Her contributions to TMA have been substantial, and in 2011 the TMA board chose to honor Monica and her family by naming the next juvenile myositis research grant to be issued the "Pedano Family Juvenile Myositis Research Award."

Thank you to the entire Pedano family!



Arthritis, calcinosis, prednisone, new drugs

Dr. Brian Feldman, a pediatric rheumatologist, is Professor of Pediatrics, Medicine, Health Policy Management & Evaluation, at the University of Toronto and Division Head of Rheumatology at The Hospital for Sick Children in Toronto.

Dr. Feldman's main focus is clinical research in the field of childhood rheumatic disease. Recognizing the challenges involved in the study of rare disease, Dr. Feldman has worked to improve the tools available to assist in this research. He has worked at developing and refining outcome measurement tools for use in clinical trials and in outcome studies. He discussed treatment and outcomes in juvenile myositis in a recent live discussion offered by TMA. You can also read the transcript online at www.myositis.org.

I was diagnosed in 1989 with JPM at 12 years old with a biopsy. I am now 35. I was wondering what your research shows about aging and progression. I have a flare one or two times a year and my joints are getting worse. Do the symptoms always get worse with aging?

Dr. Brian Feldman: With age many patients with JDM get better and go into remission. We have much less information about JPM (than JDM) since it is much rarer. My suspicion, based on my patients, is that JPM is the same as polymyositis that develops at an older age - and therefore behaves just like polymyositis with aging.

My daughter was diagnosed at 6 and went into remission -- in terms of muscle strength -- after several months of treatment. Her rash has also improved. What are the chances that active juvenile myositis won't appear again?

Dr. Brian Feldman: It depends. If your daughter is in remission and no longer on any medications, then the chance of recurrence is low, and gets lower the longer she has been on remission. We found in our studies that a very small proportion of patients who were able to be off medications later had a recurrence.

Do you recommend that JM children get flu shots? My daughter's pediatrician is not sure, so I am waiting for her pediatric rheumatologist to get in touch with me, but the pediatrician mentioned that, since she is still immunosuppressed, it may not work, since the vaccine depends on a reaction.

Dr. Brian Feldman: I recommend that all my patients with JM get influenza vaccinations every year. This has been better studied in children with arthritis, and the vaccine in those children is safe and effective. For those children with JM that are still on medications, it is best to avoid influenza if at all possible, and the vaccine is a good way to do that.

My son has JM and complains about stomach pains. Is this common with JM?

Dr. Brian Feldman: There are many reasons for stomach pains to develop if you have JM, especially if you are on medications. JDM can have direct effects on the stomach and intestines when it is active - in this case the bowels are sometimes affected with constipation, diarrhea, or blood in the stools. Many of the medications that we use to treat JM can also cause stomach pains (in some patients, fortunately not in most). This is best checked out by your doctor to see why stomach pains are occurring and to see how best to treat them.

My teenage daughter's calcinosis is so severe that her doctor is recommending surgery. However, I have also read that this often does not help, and the calcifications just reappear. In your view, when is surgery warranted?

Dr. Brian Feldman: If the calcinosis is painful or troubling it can often be successfully removed surgically without recurrence. In the 'old days', when JDM was treated less completely, removing calcinosis while the disease was still active sometimes led to reappearance. Now, with more complete treatment of active JDM, my patients have had much more luck with surgical removal.

We waited for a long time to see a pediatric rheumatologist. The nearest one is more than four hours away and is extremely busy because of serving such a large area. I would like to involve my daughter's local pediatrician, who diagnosed her JM. How much, in your view, can be handled by a regular pediatrician and how does this work?

Dr. Brian Feldman: The key, in your situation, is for there to be a good working relationship between your pediatrician and your pediatric rheumatologist. If they are communicating well, and frequently, then much of your child's care can be done locally. Most pediatricians see very few children with myositis (or none at all) and feel inadequate when treating these children, unless they have the help of a dedicated specialist.

My son receives IVIG and it has made a tremendous difference in his recovery. His insurance covers it. Another child at the clinic who is severely ill cannot get this because her insurance does

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Ask the Doctor, cont'd from page 3

not pay. Is IVIG an approved therapy for JM?

Dr. Brian Feldman: The situation regarding IVIG varies in different parts of the world. In Canada, where I'm from, I am able to get IVIG for my patients without restriction. However, there have been no clinical trials in children establishing the benefit of IVIG. We just published an interesting study this year that I think proves IVIG's benefit, but it is not the type of clinical trial usually required by regulators, or by insurance companies, to grant coverage. Some insurance companies may not provide coverage because of the lack of formal clinical trials.

My pediatrician suggested my daughter be tested for Vitamin D deficiency, especially since she has covered herself from the sun for 2 years. She has a severe deficiency, so we are supplementing. I am going to ask the pediatric rheumatologist when I see him in March if it is okay, but I wonder if the deficiency could have something to do with her getting JM in the first place.

Dr. Brian Feldman: We don't have strong information about vitamin D levels and susceptibility to myositis. Many of my patients, though, become vitamin D deficient while on treatment - partly because they are wearing sunscreen all the time. We supplement routinely here to protect our patients' bones.

My son responds to prednisone in terms of it helping his muscle strength, but he can't sleep, is irritable, disfigured, not himself and has gained 40 pounds. Our whole family is suffering. Do you have any ideas for minimizing the side effects of prednisone?

Dr. Brian Feldman: Prednisone is one of the best medications for JM, but almost always is associated with

troubling side effects. For that reason, all of my patients are also started on methotrexate (and sometimes also IVIG) so that we don't need to use prednisone for a long period of time. In order to protect the bones from some of the prednisone side effects, I prescribe vitamin D and supplemental calcium. All of my patients are coached to follow a low sodium, low calorie "prednisone diet".

My 10-year old daughter is doing much better but she is just exhausted and weak. She is off medications but seems to be afraid to go back to her active life, which she needs to do in order to regain strength. What can I do to encourage her?

Dr. Brian Feldman: If your pediatric rheumatologist has determined that there is no longer any active myositis, and that there is no other medical reason for exhaustion and weakness, then a graded program of physical exercise (ideally prescribed by a physiotherapist experienced in myositis care) can work wonders to improve sleep, decrease fatigue, and improve strength. Some children who no longer need medications, still have muscle atrophy (thin muscles) with fat replacement when we check them with MRI. Those children, especially, need a good exercise program.

My 7-year-old daughter, diagnosed last summer, is taking high doses of prednisone orally. I read somewhere online that intravenous prednisone works faster and is better tolerated in children. I mentioned this to my daughter's rheumatologist and he said the thinking has changed on this. What do you think?

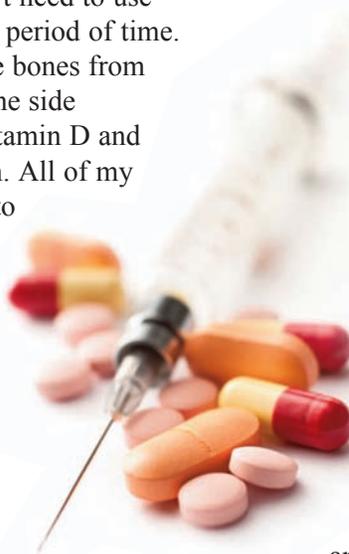
Dr. Brian Feldman: Some rheumatologists use intravenous forms of prednisone - especially early on in the

course of myositis - because some myositis patients don't absorb oral prednisone all that well from their stomach and intestines. With time, that improves, in my experience. For most patients, over time, it probably doesn't make much difference (we studied the differences after 3 years, and couldn't find any) - but in our study the most severe patients were all treated with intravenous forms of prednisone, so we didn't have any matches to compare them with. For that reason, for mildly, or moderately ill patients I start with oral prednisone, and only use intravenous forms if I suspect that there is a problem with medicine absorption from the stomach and intestines. For very severe patients, many rheumatologists will start with intravenous forms of prednisone.

What did the rituximab trial show about using this drug in children?

Dr. Brian Feldman: There weren't enough children in the rituximab study to be definitive (in my opinion). Almost all of the children in the study improved by the end of it, but the response was not as quick as anticipated. We are finding out with other diseases (like rheumatoid arthritis) that rituximab may take several months before it is effective. It may be that rituximab is effective in JM, but that it takes a long time to show an effect.

My daughter has been free of JM symptoms for 10 years and is now 25 and about to get married. I am unable to find out how JM might affect her children. Are there studies on this?



Dr. Brian Feldman: I am not aware of studies of pregnancies in women who are in remission from JM. In my opinion the JM will not have an effect on her children, and it is very unlikely - at this point - that having children will cause your daughter to have a flare of her JM.

My son wants to go back to playing basketball. He is no longer on medication but is weak. His coach is willing to work with him. I am afraid he will have a flare if he starts competing again. He is 15 and basketball is important to him. His doctor supports him in this. What do you think?

Dr. Brian Feldman: In my experience, exercise does not cause JM to flare, rather it is beneficial. There has been a limited amount of scientific study in children, and more in adults, that supports the safety of exercise, even while the myositis is active. I would say - go for it.

My daughter began to have symptoms of JM right after she had a bad flu with fever. The doctor said it was possible they were associated but no one really knows. Do you have any research on that?

Dr. Brian Feldman: Dr. Pachman's group in Chicago found that many children develop the symptoms of JM around the time of an infection. In Toronto, we confirmed those findings in another study. It is really hard to know - for sure - the role of infection in causing JM. Children get the flu and other infections all the time, so our findings may be the result of coincidences. However, I feel that susceptible children (that is, those with 'twitchy' immune systems) may react to infections, and perhaps other stimuli, by developing JM.



Send your child to camp

TMA offers camp scholarships to qualified children. It's easy, and those who have done it say it was great for their children. The guidelines are simple:

Camp Eligibility: Any camp that can handle the individual needs of your child is eligible for the program. If you would like a list of accessible camps, contact TMA.

Completing the Application: Once your child is accepted to camp, have the camp director or other administrator send the completed application to TMA's office. All applications must include the name of the camper's primary physician.

Payments: TMA will pay \$100 toward camp fees, either directly to the camp or to reimburse the parent.

Refunds: Parents must notify TMA in advance if their child will not be able to attend camp. TMA will contact the camp to request a refund.

Selection of Campers: In case there are more applications than available funds, TMA will base its awards on when the application was received.

Ready to send your child to camp? Download the Camp Scholarship Application from www.myositis.org.

Measuring inflammation with MRI

Dr. Warren Davis of the Rheumatology department at Royal London Hospital published an article in the December Issue of "Rheumatology" about using an imaging-based scoring system to measure active inflammation. Following is an edited version of the article's abstract:

Researchers set out to evaluate the consistency of a new scoring system to reliably measure the level of acute inflammatory change in JDM. This

system defines markers of inflammatory change in four muscle groups and the surrounding soft tissues. Forty-eight children (33 girls) underwent retrospective assessment of their MRI studies by two musculoskeletal pediatric radiologists to see if they could agree on the degree of disease activity.

Each observer performed the readings on two separate occasions. The agreement between the two observers and between the two readings was measured.

There was fair to moderate agreement between the two observers for all the examined disease activity markers in all muscle groups. There was good agreement between the two readings. There was no difference according to the side evaluated. Researchers concluded that the markers of inflammatory change in JDM can be observed reliably using MRI in a reliable fashion. The accuracy of the proposed scoring system is acceptable for the single reader, although there is more variability.

New warnings, precautions for acetaminophen

For years, parents have been advised to use acetaminophen (Tylenol) to control inflammation, pain and fever. In February, health authorities issued widespread warnings about its proper use.

Acetaminophen is an active ingredient in many over-the-counter and prescription medicines that help relieve pain and reduce fever. To find out if an over-the-counter medicine contains acetaminophen, look for "acetaminophen" on the Drug Facts label. If a prescription medicine contains acetaminophen, the label may not spell out the whole word or may have

Continued on back cover.

Watch the salt

Juvenile myositis patients are advised to follow a healthy diet very similar to that recommended for everyone. However, teenagers with juvenile myositis, especially those taking prednisone, are advised to be extra cautious about the amount of salt they consume. This isn't always easy: salt is everywhere. It's hidden in bread and sweet snacks as well as being the dominant taste in chips and other salty snacks. Here's a guide for those wanting to avoid "prednisone bloat."

General guidelines

Choose fresh instead of processed foods when you can.

Use the Nutrition Facts Label to check the sodium in packaged foods. Look for foods with 5% Daily Value (DV) or less. A sodium content of 20% DV or more is high.

Look for foods that say "low sodium" or "no salt added."

Take this list with you the next time you go food shopping:

Healthy Shopping List

Vegetables and fruits

Choose fresh or frozen fruits and vegetables when possible.

- Fresh fruits like apples, oranges, or bananas
- Fresh vegetables like spinach, carrots, or broccoli
- Frozen vegetables without added sauce
- Canned vegetables that are low in sodium or have no salt added
- Low sodium vegetable juice
- Frozen or dried fruit (unsweetened)
- Canned fruit (packed in water or 100% juice)

Dressings, Oils, and Condiments

- Choose condiments that are low in sodium or have no sodium at all.
- Unsalted butter or margarine
 - Vegetable oils (canola, olive, peanut, sesame)
 - Sodium-free salad dressing and mayonnaise
 - Vinegar

Breads, Cereals, and Grains

Compare labels to find products with less sodium. When you cook rice or pasta, don't add salt.

- Plain rice or pasta (Tip: If you buy a package with a seasoning packet, use only part of the packet to reduce the sodium content.)
- Unsweetened shredded wheat
- Unsalted popcorn

Seasonings

- Try these seasonings instead of salt to flavor food.
- Herbs, spices, or salt-free seasoning blends
 - Chopped vegetables, such as garlic, onion, and peppers
 - Lemons and limes
 - Ginger

Meats, fish, nuts, and beans

Choose fresh meats when possible. Some fresh meat has added salt, so always check the label.

- Fish or shellfish
- Chicken or turkey breast without skin
- Lean cuts of beef or pork
- Unsalted nuts and seeds
- Canned beans labeled "no salt added" or "low sodium"

Milk and Milk Products

Choose fat-free or low-fat milk and yogurt more often than cheese, which can be high in sodium. Milk and yogurt are also good sources of potassium, which can help lower blood pressure.

- Fat-free or low-fat (1%) milk
- Fat-free or low-fat yogurt
- Low sodium or reduced sodium cheese (like Natural Swiss Cheese)
- Soy-based drinks with added calcium (soymilk)

What is occupational therapy anyway?

All sorts of kids see occupational therapists. Many children with JM have trouble with everyday activities like dressing, tying shoes, feeding themselves, writing, drawing, or coloring in the lines.

Some kids with conditions like JM may need to use a wheelchair for a while. An occupational therapist can help kids in wheelchairs come up with a plan to go through the lunch line, get stuff out of their school locker, and make it to class on time.

An occupational therapist also offers aids and equipment like slings or splints to provide support to different parts of the body. They can help you find devices that make it easier to do things like opening a jar, putting your shoes on, or taking a bath or shower.

Often, the occupational therapist will work with a physical therapist,

who will test muscle strength and give you exercises to strengthen large muscles, such as the arms and legs, and keep joints flexible. A physical therapist can also help you get better coordination and movement.

Videos for you

Want to find out more about how an occupational therapist works? Watch this video:

http://kidshealth.org/kid/feel_better/people/occupational_therapist.html?tracking=K_RelatedArticle#cat20075

What's it like to have an MRI? This video, made for kids, will show you:

http://kidshealth.org/kid/closet/movies/video_mri.html#cat20580



Cool craft: Magnifying Waterscope

Ever wonder what's under the surface of the water? Watch through the private window of this waterscope and find out.



Materials

- Can opener
- Coffee or large juice can
- Duct tape
- Generic plastic wrap
- Wide rubber band

Instructions: Use a can opener to cut both ends off the can. If the can's cut edges seem sharp, cover them with duct tape. Stretch the plastic wrap over one end, then secure it with the rubber band and duct tape. Dip the covered end into the water and peer inside.

Tester's tips: To improve viewing, line the inside of the can with black craft foam. If your child is using the waterscope from a dock, have him lie flat on his stomach for safety.

Why it works: The pressure of the water against the plastic wrap bows it inward, creating a curved, magnifying lens.

Enjoy your summer

Although we have many weeks of cold, rain, wind and mud still left, it's time to start thinking about summer. TMA offers help for children to attend summer camp. Have your parents read page 5 of this newsletter, or go online to www.myositis.org to find an application.

Start with art

Swiss artist Paul Klee once said, "Drawing is like taking a line for a walk." Your child can put Klee's words into practice by using this continuous-drawing technique.

Place a piece of lightweight paper over a photo or picture and have your child trace the image without lifting his pencil. Or have him draw freehand, picking up the pencil only when he's done. Use markers or watercolors to finish the masterpiece -- and don't worry about staying inside the line!



New warnings, cont'd from page 5.

the abbreviation “APAP.” Giving your child more acetaminophen than directed on the label can cause liver damage and may lead to death.

To safely give acetaminophen:

- Make sure you know the active ingredients in all the medicines your child is using.
- Make sure your child doesn't get more than one medicine containing acetaminophen in a day.

Make sure you understand:

- how much acetaminophen you can give at one time

- how many hours you must wait before you can give acetaminophen again
- how many times you can give it each day
- when you should not give it
- when you should talk to your child's doctor

If the medicine is a liquid, use the measuring tool that comes with the medicine. If your child swallows a quantity of acetaminophen, get medical help right away, even if your child doesn't feel sick.

For immediate help, call Poison Control Center (1-800-222-1222) or 911.

Your child can develop liver damage after using too much acetaminophen. Symptoms may not

appear for days and early symptoms may seem like the flu: loss of appetite, nausea, and vomiting.

Ask your pharmacist for child safety caps on all your family's prescription medicines. Keep all medicines where they can't be seen or reached by children and pets. A locked box, cabinet, or closet is best.

For more information:

- talk to your doctor, nurse, or pharmacist
- visit FDA online at www.fda.gov/drugs
- call FDA at 1-888-INFO-FDA, or
- email questions to druginfo@fda.hhs.gov

