

Documenting your family's journey

The link at the end of this article is where the entire blog story of Josie's disease can be found. It is a moving and graphic account of Josie's illness that contains illustrations and descriptions of severe disease. While it can be a rich resource of information, it can also be disturbing to readers. Please be aware that Josie's disease is not typical of the great majority of cases of JM. Caution is advised before allowing children to view the blog pictures without a parent present. TMA thanks Jay Adams and his family for sharing their difficult story.

"What's going on with Josie?" That's what Jay Adams asked shortly after his family's July 4 vacation at Detroit Lakes in Minnesota more than four years ago. It was the first sentence of the first chapter in a story of tremendous pain, courage and bewilderment for 2 ½ year-old Josie and her family. Josie's father decided to tell the story online day by day, even as one day's ordeal was followed shortly by one even more frightening.

Adams, who works in the online scheduling industry, said he had maintained a family website before Josie got the rash that started her long bout with juvenile myositis (JM). "I wasn't really doing a good job with it," he said. "I didn't post regularly or anything like that."

Josie's introduction to JM was extremely dramatic, with severe swelling and rashes everywhere. There were trips to the emergency room, multiple referrals and a great deal of anxiety before Josie's disease

was even diagnosed. "Naturally, everyone we knew wanted to know what was happening," Adams said. Some nights, after dealing with the hospital, his own work, and the other children in the family, he found himself talking to 10 or 15 concerned people. "We appreciated their concern, of course," he said, "but it was exhausting." He found himself sending out an email every day. "This dovetailed nicely into a web log," he said. "People knew they could come to the blog and find out the latest news about Josie. It was very simple to put together in writing, and it was easier to do it that way than talking, when I sometimes couldn't remember who knew what. I could make it very clear and detailed, and everybody could get the whole story." Adams backtracked a little to start at the beginning, but his posts after the first couple of weeks were entered day by day, with no knowledge of the outcome or even what would happen in the next few hours. "We were never prepared, but always hopeful," he said.

There were many reasons why Adams chose this way to record Josie's daily struggles besides the friends and family members who logged on every night to check on Josie's condition. "We're the type of people who approach things from a practical point of view," he said. "Rather than throwing up our hands, our instinct is to do whatever we can to move forward, to understand the difficulties and try to fix them." He found that taking the role of the

detached observer every evening, recording Josie's progress or lack of progress, helped him to step back and gain some perspective.

He also decided that he would not spare the graphic details, either in words or photos. "This is definitely real life," he said. "It was horrible for Josie and horrible to watch." He knew that glossing over some of the shocking photos or gruesome descriptions would be, in essence, trivializing Josie's ordeal. As Josie grew old enough to understand, she agreed with this approach. "She wanted a true record, as we did," Adams said. "I remember a time in the hospital when she was covered with blood she'd thrown up. She wanted me to take a photo." The hospital staff protested, but there were a number of reasons Adams did what Josie wanted.

"When you're in the midst of a life emergency, you don't remember things so well," he said. "Recording the horrible as well as the good helps you remember." During Josie's long illness, Adams at times forgot which medicines she'd taken, who she'd seen, what reactions she'd had, what seemed to characterize her worst moments. "All I had to do was go back to the blog and I knew exactly what happened, and when it happened."

Another advantage of showing the raw details was providing a guide for families just starting out with a juvenile myositis diagnosis. "Someone could read about Josie's troubles and

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know that she and we had survived them, and that there was hope for improvement," he said. That said, he always cautions new families that Josie's case was extremely difficult, and that most JM patients can expect a much easier path than his daughter's.

There's another, less tangible reason that Josie's real story is important. "She's as feisty a little girl as you can imagine," Adams said. "If she ever has doubts about her courage, she can read what she went through and realize she can get through just about anything." Josie and her dad will look at the photos as well as the physical sites of Josie's calcinosis and make a game of predicting when one of the surface deposits will break open. "By joking about it, we deal with it," he says.

Josie was in the hospital for many months, and the Adams family juggled their time so the other children, now 13, 10 and 9, could have somewhat of a normal life. One parent took the children to sports and school events, while the other parent was with Josie at the hospital. "It was remarkable, how the other children coped," Adams said. "They never showed any resentment, only concern."

Adams settled into a routine. "I'd be at the hospital and then I'd spend a couple of hours writing about it." He found it calming, as though writing about it helped him work through his worries and prepare for the next day.

The family slowly built a following for their blog, and began hearing from well-wishers all over the country. Many of TMA's adult members followed Josie's progress and cheered her small gains in health. "We never heard anything at all negative," Adams said, "and it helped us to know that the world is such a place that even strangers care about a small, brave girl." Jay found out that the medical staff was reading the blog.

"Some of the rheumatology students were directed to pictures of Josie so they'd know what the DM rash looks like," he said.

Adams fields a lot of questions from other parents: "I am always glad to answer them, although I realize her case is much more extreme," he said. How does Josie, now an active and happy 7-year old, feel about her illness? "Her attitude is really one of 'this is my life;' she doesn't know any different," he said. "Certainly, she's been molded by her own experiences. She seems to be thriving, although it's hard to tell if her personality has anything to do with the struggle she went through, or if she would have been especially feisty anyway."

Adams has some advice for others wanting to keep a web log for their children:

- Be conversational. He likes to picture himself having coffee with a friend or close relative while he writes. "If you get bogged down in formal writing, it won't be the real story," he said. He uses a lot of side notes, parentheses and non-sequiturs. "That's how we remember the details."

- Use photos. "That's been tremendously important in telling Josie's story," Adams said. From rashes to calcinosis, he captured the progress of her illness in a way that has been helpful to both his memory of her progress and that of her health care team. One of his photos is used in training.

- Know your own level of comfort with strangers. "As you can probably tell already, we're not an incredibly private family as we've posted Josie's story for all to see," Adams said. "However, you can restrict access very easily with a simple password, so only those with the password can log on to see your reports."

Find Josie's story at <http://www.deebers.com/josie/>

CURRENT JM REPORTS

JM patients who didn't take steroids

In the study, *Favorable Outcome of Juvenile Dermatomyositis Treated without Systemic Corticosteroids*, Drs. Levy, Bingham, Kahn, Eichenfield and Imundo, from the Department of Pediatrics at Columbia University Medical Center in New York, describe the course of patients with juvenile dermatomyositis (JDM) treated effectively without corticosteroids such as prednisone.

Researchers in this retrospective study found eight patients who had never been treated with corticosteroids out of 38 with JDM treated at a children's hospital.

Patients in the “no steroid” group were followed for a period of two to nine years. Treatment was primarily with intravenous immunoglobulin (IVIG) (75 percent) and methotrexate (50 percent), with favorable response in all. There were no serious treatment complications, although headaches were reported by three patients receiving IVIG. Two patients had a myositis flare after discontinuing all medications for more than one year, but complete absence of symptoms was observed after either one or two further doses of IVIG. Two patients had calcinosis (at one and nine years of disease); however, no patient had joint contractures, muscle atrophy, lipodystrophy, or functional limitations.

The authors concluded that systemic steroids can be avoided in some patients with JDM. Alternative agents such as methotrexate and IVIG may be prescribed to effectively treat JDM and prevent complications.

Muscle and skin symptoms: what a long lapse tells us

Drs. Scott Lieberman and David Sherry report on the case of a child with years of delay between a diagnosis of polymyositis and the develop-

ment of the typical juvenile dermatomyositis rash, a report that might be of interest to those with a child diagnosed with polymyositis.

The authors, in the Rheumatology and Pediatric Divisions of The Children's Hospital of Philadelphia and at the University of Pennsylvania, reported in the September 2009 edition of the *Journal of Clinical Rheumatology*.

They note that the annual incidence of juvenile myositis of all kinds is from 1.7 to 4.1 cases per million children, but that juvenile polymyositis (JPM) accounts for only 3 to 12 percent of juvenile cases. Allowing for a few other unlabeled cases of childhood myopathies, juvenile dermatomyositis accounts for between 80 and 92 percent of all cases of myositis in children, so polymyositis in people less than 18 years of age is very rare. It's not uncommon, the authors say, for children with JDM to come to them with muscle weakness but no rash, but the skin signs usually appear within a few weeks or a few months. Often, the fingernails are the first JM skin symptoms. In one study they reference, the longest time from muscle weakness to the beginning of the rash was 20 months. In that study, the median lapse (for those with muscle weakness but no rash) was about three months.

An 11-year-old black boy with no significant past medical history had weakness in his upper and lower extremities and had fallen down twice at school the week before he was first seen by doctors. His first complaint was lower back pain from the second fall, but when the doctors questioned him, he remembered difficulty climbing stairs and performing daily activities.

He had no other neurologic symptoms, or any reason to explain his weakness. He had no family history of rheumatic illness, no heart or circulatory symptoms, no rash and no fin-

gernail abnormalities. An exam showed he had significantly decreased strength. He was unable to sit up from a lying position but was able to stand from a seated position. He had two of the myositis-specific antibodies, and an EMG showed he had pronounced muscle disease from an inflammatory process. A muscle biopsy was done and showed muscle fiber degeneration.

Because of his reported weakness and subsequent laboratory, imaging and blood tests, and the absence of rash, he was diagnosed with JPM and treated with pulse intravenous corticosteroids and IVIG, then oral steroids, with the addition of cyclosporine A when his clinical and laboratory response was disappointing.

Over the next three years, his symptoms waxed and waned, and muscle enzymes remained elevated with brief intervals in or near the normal ranges. No tests over the years showed all muscle enzymes within the normal range at the same time. The treatment regimen included (at varying times) methotrexate, etanercept and adalimumab in addition to IVIG, corticosteroids and cyclosporine A.

With each new treatment his weakness and muscle enzymes improved, allowing for further tapering of corticosteroids; however, each time his medication was reduced, his muscle enzymes increased, with or without development of weakness. Various medications were reduced when they weren't effective, or when they caused a reaction.

Forty months after his diagnosis, the young patient came for a routine follow-up with the typical JDM heliotrope discoloration over his eyelids. He also had Gottron's papules. The doctors checked carefully to make sure the skin signs weren't caused by infection or other factors. The rash was noted in late summer,

suggesting possible excess sun exposure. He has since been treated with hydroxychloroquine, low-dose cyclosporin, and three rounds of rituximab, the last round being almost two years ago.

Currently, his strength is back to normal, his nail fold capillaries remain normal, and only a mild rash remains on his fingers. His medication regimen includes prednisone, hydroxychloroquine, IVIG, and IV methylprednisolone monthly. His most recent laboratory tests were mostly indicative of mild disease, with some of the tests being normal.

“To our knowledge,” the authors write, “this is the longest reported lag between diagnosis of an inflammatory myopathy and subsequent development of typical JDM rash. The JPM-like pathology in this child who subsequently developed classic JDM adds to the continuing debate over whether JPM is really a distinct entity or, rather, just a subset of JDM.”

Do vaccines trigger JM?

Drs. Orbach and Tanay at Wolfson Medical Center in Israel published a study in *Lupus*, noting that we give vaccines credit for being among the greatest medical discoveries, eradicating some diseases and improving survival and quality of life.

However, vaccines are among the environmental factors implicated as triggers for the development of inflammatory myopathies. The sporadic reports on vaccine-induced inflammatory myopathies include hepatitis B virus, tetanus, influenza, smallpox, polio, diphtheria, diphtheria-pertussis-tetanus, combination of diphtheria with scarlet fever and diphtheria-pertussis-tetanus with polio vaccines.

However, a significant increase in the incidence of dermatomyositis or polymyositis after any massive vaccination campaign has not been reported in the literature. In this study of

patients with inflammatory myopathies, no recent immunization was recorded in any of the patients. Prospective multicenter studies are needed to identify potential environmental factors, including vaccines, as potential triggers for inflammatory myopathies.

Advances in understanding

Several recent reviews of research over the past five years shed light on how the mystery of JM is slowly unraveling. TMA presents research on its website under “Research” at www.myositis.org. Read the full reports by clicking on “Juvenile Myositis” under the “Published Research” category.

Dr. Adam Huber, a TMA medical advisory board member, took a look at the newest body of JM research and summarized it in his report, *Advances in Pathogenesis, Evaluation and Treatment*. Published articles called “reviews” are of tremendous help to physicians treating rare diseases, and their patients because they compile the results of recent patient trials. Among some of the questions Dr. Huber asks and answers:

How is JDM related to infection?

Compelling evidence of the role of infections in JM has been lacking. There have been a variety of studies of a potential link between specific infections and JM, including coxsackievirus, hemolytic streptococcus, hepatitis B virus, influenza, parainfluenza, *Borrelia* spp., *Toxoplasma gondii* and parvovirus. However, these studies have not been consistent and, in some cases, have not been able to be replicated.

Dr. Huber says it is reasonable to conclude that, if JM is caused by an infectious agent, either the specific organism has not been found or JM is not associated with a single infection, but with many. Dr. Lauren Pachman and others reported that 63 percent of

children diagnosed with JDM had a history of an infection in the preceding three months (the type of infection was not identified) compared to a control group of 42 percent. A more recent study led by Dr. Pachman again documented that infections are common in the three months before the symptoms of the disease are noticed. In this study, using interviews within six months of diagnosis, researchers found that 57 percent of children with JM had respiratory infections, 30 percent had previous gastrointestinal infections, and 63 percent of the children with infectious symptoms had taken antibiotics. This last study did not include any data that would determine if these children with JM differed from normal, healthy children, and did not include testing for any specific infectious agents.

JM TREATMENT ROUND-UP

Dr. Huber reviewed treatments from recent reports.

Anti-TNF Therapy

The association of JDM with tumor necrosis factor (TNF) has led to consideration of anti-TNF medication for JDM. There has not been a published clinical trial to date, although there have been results in abstract form for a small number of patients with JM treated with etanercept (Enbrel). Dr. Huber reviewed them.

Ten children with chronic disease that did not improve with intravenous pulse corticosteroids and other immunosuppressant medications received Enbrel. There was a statistically significant improvement in disease activity, but the change appeared quite modest, and there was no change in muscle enzyme levels or percent of B cells. Another anecdotal report on Enbrel from a large center that sees many JM children has not been favorable, either, Huber notes.

Another study reported on the open-label treatment of five JDM patients with infliximab, another anti-

TNF medication. All patients had not responded to standard therapy, including intravenous corticosteroids, methotrexate, and at least one other disease modifying anti-rheumatic drug. They all had progressive calcinosis. The authors reported that all patients had improvements in muscle strength and function, disease activity and contractures, and all were able to discontinue other medications. However, Huber notes, this is a small trial, with no control group. He recommends more studies of anti-TNF medication in JM patients, with infliximab being the most promising, although there might be other agents, as yet unstudied, that might hold promise.

Rituximab

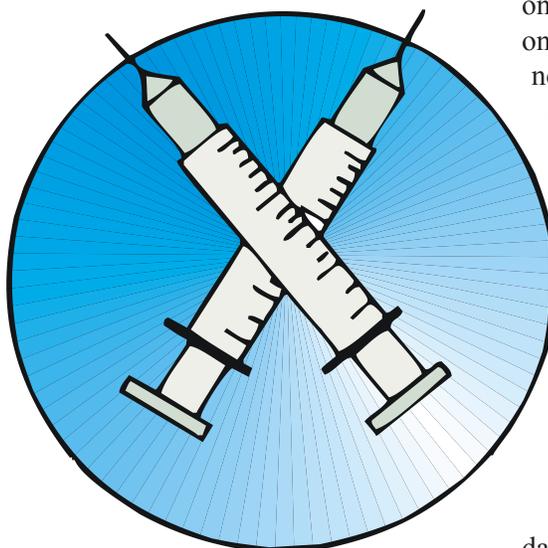
Rituximab is a monoclonal antibody, which depletes B cells. It is thought that B-cell depletion is important for a response in patients with JM. The use of rituximab in children was first reported by TMA Medical Advisory Board Member Dr. Todd Levine, who treated five patients, two of whom were children with refractory JDM. These patients experienced B-cell depletion, but otherwise tolerated the treatment well. The JDM patients were reported to experience 40 percent and 45 percent improvements in muscle strength, but no other details were available.

Another report included four JM patients with disease lasting from five weeks to 27 months, with markedly active disease despite treatment with corticosteroids, methotrexate, and other immunosuppressant agents. Two patients were retreated one year later, and all had depletion of their B cells (one after a second dose). Three patients had excellent responses, with the rash healing, strength becoming normal, and weaning or discontinuation of other medications. Responses lasted for 12 months or more; two patients with flares responded well when treated again. One patient had

progression of her disease with no evidence of response, despite appropriate B-cell depletion.

Rituximab for skin symptoms

One study reported the use of rituximab for chronic skin features of JDM. Two of the three patients had JM and had been treated with oral corticosteroids, methotrexate, hydroxychloroquine, and cyclosporin, with apparent response in muscle disease, but not much improvement in skin disease. Both patients were treated with rituximab for four weeks, and



experienced dramatic improvements in their skin disease. They were also able to discontinue or markedly wean all other immunosuppressant medications.

In this small number of reports available, rituximab has been well tolerated in JDM, with no serious adverse effects (including infections) reported. There is an ongoing trial being conducted, with participating centers in North America and Europe, to determine the efficacy of rituximab in both adults and children with refractory DM. Note: recruitment for the “Rituximab in Myositis” trial is closed. Contact TMA at TMA@myositis.org if you want to consult with a physician experienced with using rituximab in children.

Methotrexate

Although there has never been a clinical trial to support its use, methotrexate has become commonly used as a steroid-sparing medication in JDM. In one report, thirty-five patients with JDM seen over a ten-year period were all treated with high-dose oral or intravenous corticosteroids, followed by methotrexate within six weeks if there was no improvement in muscle enzyme levels. At follow-up, 26 patients were in remission (no rash, weakness or elevation of enzyme levels), although whether they were still on medications was not stated and only five had developed mild calcinosis. Those who developed calcinosis had longer disease duration before treatment, longer time to normalization of muscle enzyme levels, and a longer time to remission. Huber notes that this study did not compare this regimen with another, making it difficult to determine if these outcomes would have been achieved without methotrexate.

In another report, the authors used data from one of the larger JM clinics in the world, and compared JDM patients before and after a change in clinical practice. Prior to the change, most patients were treated with oral corticosteroids with reduction by 10 percent every four weeks, depending on response. After the change, most patients were treated with oral corticosteroids, but methotrexate was started at the beginning, and corticosteroids were reduced every two weeks depending on the child’s response. This resulted in a markedly shorter period of exposure to corticosteroids. Not surprisingly, the patients with the more rapid corticosteroid wean had a better growth velocity and less weight gain, and there were significant trends towards fewer cataracts and fractures.

Otherwise, the authors did not detect any other differences in outcome, including muscle strength,

Do your own research!

Once you're making your own appointments with doctors and other health professionals, it's a good time to start learning about your own disease by going to expert sources of information. Learning to read reports of studies involving real-life juvenile myositis patients and those with similar diseases will be helpful to you throughout the course of your illness. Many reports relevant to chronic disease, inflammation, exercise, bone loss and medication that appeal to a wider audience are explained by health communicators on sites like "Web MD" or "Medline." Most articles specifically about JM do not reach these popular outlets because the disease is so rare.

If you go directly to articles appearing in professional journals, you'll find them very technical, but there is usually an "abstract" online that gives the most important points, including the overall outcome of trials. Usually the "conclusion" part of a journal article (which is often in the abstract) will say whether the medication or other treatment appeared to be effective and safe, and may call for more studies. Not all articles are about the effects of drug treatment. Some talk about exercise, or the pathology of the disease, or possible side effects of medication.

Find the Journals

Go to www.pubmed.com. You will find several ways to search for information about juvenile myositis. Most likely, you will be searching for information about your condition in general, or information about a certain drug

you are taking. There will be a search box at the top of the pubmed page, so type in "juvenile myositis," or "juvenile myositis, methotrexate" to find articles in peer-reviewed journals about the topic that interests you. "Peer-reviewed" means that the article is not an isolated physician's opinion or observations, but that a team of specialists in his or her field have reviewed the research and approved it. You can sign up to have any articles in future journals that correspond to your search terms delivered to your email.

Go to TMA's website

TMA has a wealth of knowledge about juvenile myositis, and it's on the website at www.myositis.org. You'll also want to check out the information on dermatomyositis, which has many of the same characteristics and medications as JM. TMA also hosts an online message board for JM patients and their families to talk, and interact periodically in live discussions with myositis experts. You'll find that asking direct questions of the experts and of other people with JM is a good way to find the answers you need. TMA also has information about coping skills, disability rights, mobility aids and joining a medical trial. If you don't find the information you're seeking, email TMA@myositis.org and TMA staff will find the answers for you.

Get the book

A number of international JM experts compiled a book, "Myositis and You," full of information about medication, career planning, alternate treatments, individual educational plans, emotion-

al issues, dealing with family matters and other challenges that patients and families face. You can purchase it on TMA's website, www.myositis.org.

What to do with what you know

As you read about your disease, make note of any questions you'd like to ask your family doctor, physical therapist or specialist. Talk to your parents and other family members. Don't make changes in medications on your own, but discuss ideas you have about what you've read. Ask about changes in diet, medications and exercise you would like to make based on your research.

Share your knowledge

Some young people with JM maintain a blog, an online history of their life with their illness. It can range from a simple way to keep track of doctor's visits and medications, to the full story of everything you've experienced and learned. Blogs can be extremely helpful to those just beginning their course of treatment for JM. You can learn a lot from the blogs of people with myositis and similar illnesses, especially how they handle the illness day to day while dealing with school, work, family and friends. Remember that each person's experience with JM is very different, and that families experiencing the most difficulties are the families most likely to keep a blog. If you would like to offer support to someone just diagnosed with JM, email TMA@myositis.org.





JUST FOR ME

Fun facts and news for children affected by JM

What does an EMG feel like?

The EMG test has two parts. You know how sometimes in the winter you walk across a wool rug, then touch someone and feel a little spark? That's the first part, called a "nerve conduction study." It measures how fast your nerves are working with your muscles and your brain.

The second part is the actual EMG, the electromyogram. The doctor puts tiny wires into your muscles and listens to the muscle's electrical activity. You will feel a tiny pinch when these wires go into your muscles. You may be able to listen to the sound of your muscles, too. Children at Cincinnati Children's Hospital said the noise of their muscles sounded like rain on the roof or a motorcycle.

What does a muscle biopsy feel like?

The muscle biopsy is a test that takes a small piece of your muscle or skin so the doctor can see what is happening. The doctor will give you something to help you fall asleep, so you won't feel the test. Take along a favorite toy or book so you will have

it when you wake up. Remember that you will wake up and feel confused from the sleep medicine, and looking at something you like will make you feel better. Your parents will come into your room when you wake up. There will be a small bandage where the test was done, and it will feel as though you got a bruise on the playground. Most likely, the doctor will

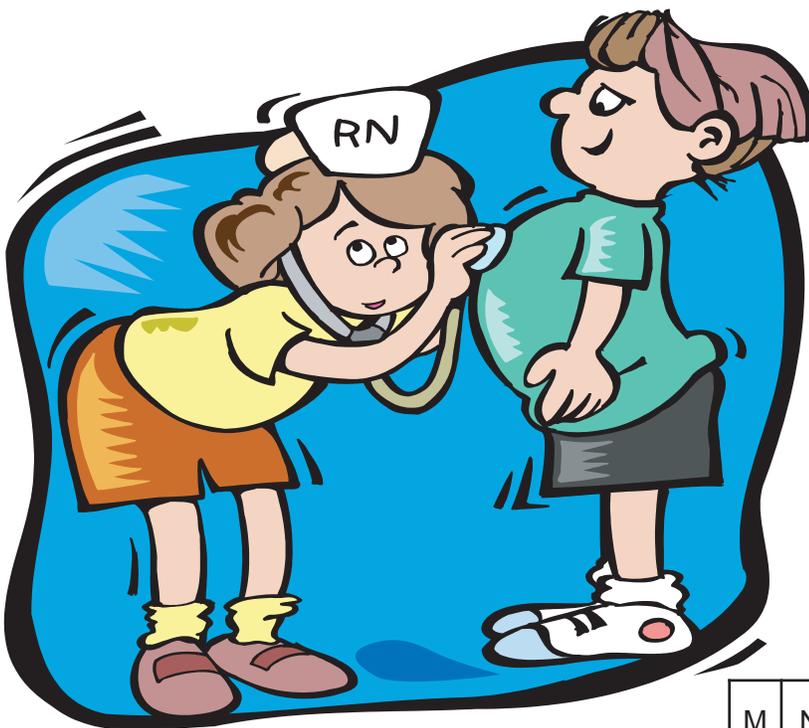
exactly where he wants the picture taken. You may be covered with a special guard so that the x-ray only goes into the exact place the doctor wants to see. X-rays are fast, don't hurt, and leave no marks or scars.



Can you find them all?

Word Puzzle!

1. X-RAY
2. NURSE
3. TEST
4. DOCTOR
5. SHOT
6. NEEDLES
7. HOSPITAL
8. BANDAGE
9. MUSCLE
10. MEDICINE



tell you to rest quietly for a day or two, and you should be able to do whatever you feel like after that.

What does an x-ray feel like?

X-rays are a way of taking a picture of the inside of your body. You won't see or feel anything different, but you will be asked to sit still. This is important, because the doctor knows

M	N	D	O	C	T	O	R
U	E	A	U	S	H	X	B
S	E	D	E	V	O	R	A
C	D	T	I	D	S	A	N
L	L	A	N	C	P	Y	D
E	E	Z	B	X	I	P	A
Q	S	H	O	T	T	N	G
N	U	R	S	E	A	O	E
R	E	B	H	P	L	Z	V



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TREATMENT

Continued from page 5

physical function, persistence of rash, calcinosis, disease flare or need for other medications. This study is limited by the use of a non-randomized control group, Dr. Huber notes, but if these results are supported by further follow-up, this is an attractive step in reducing prednisone-related damage in children with JM.

Other medications

Although the majority of patients with JDM do well with standard treatment (corticosteroid therapy with or without methotrexate), there remains a smaller number of patients who have either very severe disease, especially those with skin or bowel vasculitis, or disease that simply fails to respond. For these patients, Dr. Huber notes, additional medications are needed. *See Dr. Huber's comments on cyclophosphamide and tacrolimus*

in the full research article at TMA's website, www.myositis.org.

Dr. Huber concluded his review by saying, "This indeed is an exciting time to be involved in the care of these children. Studies on pathogenesis are providing important insights into the sequence of events that lead to JDM. These studies may, in the future, allow determination of patients at highest risk for poor outcomes, and point the direction to safer and more effective treatment. Finally, new medications such as rituximab, and new ways of using older medications, such as methotrexate, promise to lead to improvements in treatment for children with JDM and associated reductions in morbidity. There remains much work to be done, but it is clear that research is leading to a much better understanding of this complex and fascinating disease."



JM COMPANION