JM COMPANION

FALL/WINTER 2005

For patients, families and friends affected by juvenile mycsitis

Tips from JM families

Dr. Lisa Rider's question-andanswer period at the Conference was truly an interactive session, with parents adding their own suggestions learned over time. Here is a sampling of what families shared:

Fosamax: Fosamax is often initiated to prevent or slow bone loss, as prednisone can negatively affect bone density. However, anyone taking Fosamax must stand upright for 30 minutes after taking the medicine. That's hard enough for adults, so how do you convince your children to do this? Jennifer plans for her daughter to take a shower after her daily dose of Fosamax, keeping her upright as directed.

Schools: We have discussed dealing with schools in the past, and this remains an important topic, especially to families of newly-diagnosed children. Families who have worked with their schools encouraged others to be the "enforcers," not assuming that the schools are doing as they've agreed to do. This doesn't mean you need to be aggressive – just check up on things to be sure your son or daughter is receiving what was promised.

Contractures: One mom noticed that her daughter walked on her toes, not using her heels at all. Dr. Rider suggested heel cord stretches for these contractures, to strengthen and stretch her heel and allow her to

The JDM mistake

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

In his talk, *How Does Juvenile Myositis Behave Over Time?*, Rennebohm introduced common concepts and ideas to help everyone grasp the medical terminology and processes that accompany JM.

What s happening to my child?

The primary sites of abnormality, Rennebohm said, are the small blood vessels in the muscles, skin and gastrointestinal tract. In JM, the child's immune system injures the endothelial cells of the blood vessels, narrowing the channel. Therefore, not enough oxygen or nutrients reach the muscles and skin that are fed by these vessels. The muscles and skin then suffer. This is, as Rennebohm described it, the JDM (juvenile dermatomyositis) mistake – the child's immune system is responding incorrectly.

Using familiar analogies, Rennebohm further explained what is happening. In an appropriate immune attack, our immune system "sets a fire," or creates an inflammatory response, in the infected area, for example the throat. The fire is set by throwing more wood (or cytokines)

on the fire in the



throat, and this fire eventually kills the virus. In return, the throat becomes red, inflamed and sore, but this is accepted as a part of the normal healing process. The immune system then stops throwing wood on the fire, and the throat returns to normal in an acceptable and appropriate period of time.

In JDM, this fire is set in the small blood vessels of the muscle and skin. "The immune system gathers as much fuel as it wants," Rennebohm said, "and fuels the fire at whatever level of intensity it wants." Though the immune system may think it is acting appropriately, it is mistaken. Fortunately, Rennebohm continued, the immune system is capable of correcting itself at some point, but that point varies from patient to patient and is typically only gradual.

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THE MYOSITIS ASSOCIATION

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

P: 202-887-0088 P: 800-821-7356 F: 202-466-8940 Email: tma@myositis.org Web: www.myositis.org

Executive Director: Bob Goldberg Editors: Theresa Reynolds Curry, Kathryn Spooner

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Kathryn Spooner and sons Rhys (left) and Caden (right).

Dear Families:

This issue of the *Companion* reviews information gathered at this year's Annual Conference in Cleveland, and there will be more in upcoming issues of the *Companion*, the *OutLook Extra Treatment Issue*, and the *Research Reporter*. Medical professionals discussed the various courses that juvenile myositis may take in different children, how art therapy helps children

and families deal with chronic illness, and what you can do as a parent to be an integral part of your child's health care team.

As always, we found the many stories and recommendations passed on by parents and children living with myositis daily to be just as valuable as the professional presentations. These bits and pieces, shared so openly, were beneficial to every family there. We can't stress enough the value of continuing to share your thoughts, shortcuts, suggestions, and ideas with us and with each other.

Thanks to everyone who made the JM track of this Conference a positive experience. I enjoyed meeting each one of you and your families over the weekend.

Warmest regards, Kathryn

Spotlight on JM: TMA-funded research highlighted at ACR conference

At this year's annual meeting of the American College of Rheumatology (ACR), TMA Medical Advisor Ann M. Reed, MD, and her team of researchers at the Mayo Clinic presented their findings on the role of dendritic cells in juvenile dermatomyositis (JDM). Dendritic cells play a central role in cell-mediated immunity, and one subset of these cells is often found in inflamed tissues. This subset is thought to influence the immune response.

In this study, funded in part by The Myositis Association, researchers compared samples of muscle tissue from children with JDM to tissue from children with other disorders. Some immature dendritic cells stay in the muscle tissue, and the team sought to determine if the dendritic cells are activated inside or outside the muscle tissue. They discovered that these cells are activated within the muscle tissue and may then move out of the tissue and into the bloodstream. These findings suggest that chronic muscle inflammation in JDM likely results from these mature dendritic cells, possibly working together with maternally derived cells.

These insights into JDM may lead to advances in the understanding of other autoimmune diseases as well.

Mistake, continued from cover

Factors influencing how JDM behaves

Rennebohm introduced three factors determining how a child's JDM behaves over time – intensity, duration and fluctuation. How intensely your child's immune system makes the JDM mistake varies from mild to moderate to severe, depending on how much firewood the system continues to throw on the blaze. "With severe intensity," he said, "the system is intent on setting a huge blaze."

A second factor is duration: how long will the JDM mistake continue? The mistake can occur for a short or prolonged period of time, and for these durations, the intensity can be mild, moderate or severe. Finally, the child's immune system can make the mistake continuously without fluctuation or with periods of varying (or fluctuating) severity.

Considering these three factors, and the degrees to which the JDM mistake is made within these three factors, there are a number of different paths each child's JM can take. The worst case, Rennebohm said, is when the mistake is made with severe intensity, prolonged duration, and constantly severe (no fluctuation); the best case: mild severity, short duration, and constant but diminishing (again, no fluctuation). And then there are all of the "in-betweens."

The courses of JM

With these factors of intensity, duration and fluctuation influencing how your child will respond to JM, how do doctors determine what treatments to use for each child? "When your child is first diagnosed," Rennebohm said, "it helps to know the spectrum of the disease but also to know the most likely course your child will follow." Early on in the disease course, it's difficult to determine which course each patient will follow. These three factors explain why doctors see three main clinical courses of JM – monocyclic (single cycle), polycyclic (many cycles) and chronic continuous. Within these categories are mild, moderate and severe versions of the disease. In general terms of physical activity, children who have no strength to get up have a severe version, children who can lift up for only short periods of time have a moderate version, and children who can maintain a lift for about two minutes have a mild version.

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

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

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

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

Progress in understanding JDM

It's obvious that many questions remain about why children develop juvenile myositis, why they follow certain courses, and why they respond to treatment differently. International groups bring doctors and researchers together to develop standard definitions of improvement. The Paediatric Rheumatology International Trials Organisation (PRINTO) and International Myositis Assessment and Clinical Studies Group (IMACS) are two groups where physicians collaborate to move forward toward a clearer understanding of the different aspects of JM. Using uniform clinical assessment tools developed by these groups (i.e. CMAS measure of muscle strength), doctors are able to work together and more effectively treat children with JM.

The upcoming *OutLook Extra Treatment Issue (Winter)* will highlight Dr. Rennebohm's discussion of the available treatments for juvenile myositis, using the same fire-setter analogy to better explain his thoughts on treating JM. The *Extra* is available to all members of The Myositis Association, both online and in print.

Tips, continued from cover

walk from heel to toe. At first, passive range of motion should be used, having someone gently manipulate and stretch the heel cord by flexing the foot. (Always discuss these and other exercises with a physical therapist or your child's doctor to ensure they are done correctly.)

Everything's right in art therapy

Most people like to draw, or at least doodle, and art therapy expands this natural enjoyment of art to promote healing, said Barbara Greenwood, MAAT, ATR, art therapist at Rainbow Babies and Children's Hospital near Cleveland. And even more important – no one is ever right or wrong in art therapy.

Greenwood began her session for the juvenile myositis track of the Annual Conference by asking every-

one how they felt about art: I like it, but I'm terrible at it; I can't draw a straight line; I love art but can only doodle. "I believe that everyone can do art," she said. Art can become an important part of your lives as you and your



family deal with chronic illness.

Scribble drawings

Greenwood introduced scribble drawings to the Conference attendees, encouraging everyone to take part in this therapeutic activity. So don't just read this – do it! You'll need a big piece of paper and some chalk.

Set the paper on a table and stand up. In the air in front of you, imagine that you're painting a fence. Brush up and down in the air. Once you feel ready, take a piece of light-colored chalk and move it along the paper, scribbling at random. Don't worry about a pattern or picture, and don't lift the chalk from the paper. Just move your chalk around the paper. When you feel you're done, stop.

Look at your paper. What do you see? If you need to, turn your paper around to find something in your "scribble." Choose one part of your drawing – whatever you "see" – and

pull it out using a darker piece of chalk. The rest of the lines are no longer important to your picture. Give what you pulled out an environment. For example, if you brought out a pair

> of sunglasses, draw a face behind them. Have fun with it – remember, nothing is wrong in art.

How does art therapy work?

This sounds fun, and a little messy, but does it really help your child? There are many different ways art can help children and parents alike, Greenwood said: "It builds self-esteem, and it encour-

ages independence." Self-esteem is so important – and often equally elusive – when children are dealing with the normal issues of growing up along with the added challenges of chronic illness. Creating a work of art gives them a sense of achievement, an alternate method to communicate their thoughts and feelings, and a sense of control over their environment when they may feel they've lost some of the control they previously had.

Unfortunately, art therapy is not available in every hospital or institution. Help your children work on their own. Buy a sketchbook and encourage drawing. Greenwood urged everyone to actively participate, making this a family activity. This can be a successful stress reliever for every member of the family. "Do it on your own time," she said. "Don't rush." You can set up space to get away whenever you need a break.

One family uses their side porch as their getaway, giving them a place out of the way where their art and supplies are readily

available any time anyone needs some art therapy.

Always remember to relax and have fun. Everyone has his or her own level of creativity, Greenwood said, and you may just discover you're more creative than you thought!

MORE THAN JUST A PRETTY PICTURE

Art therapy helps in different ways, depending on children's personalities and needs. Greenwood highlighted some common benefits of this unique therapy.

Creating art:

- becomes an outlet for excess energy
- allows time to pass more quickly
- helps children develop new skills
- reinforces qualities like patience, persistence and acceptance
- relaxes the artist, and
- helps people understand themselves better.

Be a part of your child s health care team

Anyone who has been on a sports team, in a band or on a committee knows the importance of every team member. Your health care team is no different.

Your child with juvenile myositis (JM) may have several doctors: pediatric rheumatologist, general pediatrician, dermatologist, and other specialists. At doctors' visits, your child may see nurses, nurse practitioners, rehabilitation specialists, and social workers. For some, psychologists, naturopathic doctors and dieticians play important roles on the team, too. These are just the members of your medical team – you also have home and community teams available to you. Parents, brothers, sisters, and extended family make up your home team; friends, teachers, school personnel, organizations like The Myositis Association, and other community resources are all a part of your community team.

Why take the team approach? "Each team member has his or her own expertise and strengths," said Jill Karnes, MS, MSW, LISW, social worker at Columbus Children's Hospital. Since JM brings its own complexities, different team members work on those parts they know best so your child receives the most complete care possible. Working together produces greater benefits. Your home and community teams will also help alleviate stress that inevitably comes from a child's chronic illness.

A parent s role

"Take an active role in managing your child's disease," Karnes said. Parents are the team members who are involved with everyone else, so you must work closely with all of them. Become as educated as you can about JM, its treatments, possible side effects, and other aspects of the disease, and discuss your child's treatment options and possible barriers with the doctors. Be honest: if your child is refusing to take medicines, let the doctors know. If you've introduced any complementary therapies, including dietary supplements, discuss these so that your doctor can inform you of any possible interactions with your child's prescriptions.

"Ask questions, give feedback, and be assertive, but not overly aggressive," Karnes said. Track your child's progress and take your records and notes to your child's appointments. These will help everyone get a better grasp on the whole picture, not just their part of it. Karnes highlighted four important roles of parents:

Be a role model for your child. "If you take an active role in disease management," she said, "they will, too, as they get older."

Gradually transition the active partnership from you to your child. As your child gets older, he or she can take on more responsibility. This involves such things as talking to the doctors, keeping up with the medication schedules, and sitting in on meetings with school personnel.

Be supportive. Fulfilling this role is more difficult than it sounds, but your child will recognize your signs of support and respond to them.

Set goals for you and your child in partnership with your teams. Setting goals is a key to success in any situation. For children with JM, goals can be long- or short-term, and they may change over time, Karnes said. No matter what age your child is, he or she needs to take charge to some degree. These goals will help. It's important that these goals are reasonable and not dream-like, she said.

She suggested a simple worksheet for each goal, with the following information:

- 1. Something I want to do this week
- 2. How, when and where I can do this

- 3. Possible barriers I might face
- 4. Plans to overcome these barriers
- 5. How confident I am that I can accomplish this goal (1-10)
- 6. Follow-up plan

Benefits of the team approach

Working with others will help everyone better understand your child's JM and how to best manage it. The team approach leads to more confidence in the situation, both in you and in your child, Karnes said, and overall satisfaction with your child's treatment in general.

KEEPING TABS ON THINGS Karnes suggested three main categories to keep you organized so you can more effectively track your child's progress:

Health Record Notebook: Ask for copies of all of your child's health records, and keep them in one notebook. Organize them by date or by doctor, and be sure to include any notes you have from the appointments. Take this to all appointments.

Medication Chart: List all of your child's medicines, including doses and when your child takes the medicine. Keep track of any side effects of the medicines as well as any benefits you notice over time. Some parents have told us that they also "rate" their child's energy/strength level each day, using a 5- or 10-point scale to compare from one day to the next. Take this chart to each appointment.

Goal Chart: Make a plan of action related to your child's goal. Once she has determined what she wants to accomplish, write down how, when and where she will do this, what possible roadblocks may arise, ways to get past these blocks, and a follow-up plan.

Ask the Doctor with Lisa Rider

Working Together

There are many questions that crop up when your child is first diagnosed with juvenile myositis. The answers may not be as easy to find as parents would like, but more research is going into learning the right answers. At the Annual Conference, Lisa Rider, MD, discussed the importance of collaborating in research. Dr. Rider's comments on treatment will be included in this Winter's OutLook Extra Treatment Issue.

"Collaborative research is advancing knowledge of juvenile myositis," Dr. Rider said. She highlighted four specific groups working toward this goal:

International Myositis Assessment and Clinical Studies Group (IMACS) has developed standards for clinical trial designs and outcome measures, making it easier to compare results from clinical studies completed at different research centers. Without these standards, comparisons would be all but impossible. IMACS is also working on classification and diagnostic criteria for juvenile myositis.

Childhood Arthritis and Rheumatology Research Alliance (CARRA) works with pediatric rheumatologists to provide alliances to conduct clinical studies and to actively work toward developing a scientific agenda in the area of childhood rheumatic diseases like myositis.

The JDM Research Registry, established by TMA Medical Advisor Lauren Pachman, MD, has enrolled more than 300 newly diagnosed children to study the incidence, demographics, environmental factors, and long-term outcomes of juvenile myositis.

Paediatric Rheumatology International Trials Organisation (PRINTO), formed by two European physicians, has also developed clinical trial outcome measures, planning a clinical trial to study the effects of methotrexate versus cyclosporine.

The Myositis Association supports these groups by participating in their annual conferences, providing research funding, and publicizing announcements of clinical trials and other important information. Through these groups, scientists from around the world work together to create definitions that will hopefully help lead to answers every researcher – and every parent – is pursuing.

Research notes

The calcinosis challenge

Calcinosis is an irritating and painful complication of JM, and prevention remains the best treatment. Aggressively treating the JM itself helps prevent calcinosis, but no standard treatments have been found to effectively remove the calcinosis once the deposits have formed.

A group of Australian physicians reported dramatic success using alendronate need popular drug name that people will recognize. Is this fosomax? to treat a 6-year-old boy's calcinosis. which had been unresponsive to diltiazem and probenecid. The group noticed improvement in one month; by 12 months, the calcinosis had virtually disappeared. As with many studies of JM, further studies with larger groups are necessary to validate these findings.

[Ambler GR, Chaitow J, Rogers M, McDonald DW, Ouvrier RA. Rapid improvement of calcinosis in juvenile dermatomyositis with alendronate therapy. The Journal of Rheumatology. 2005 September; 32(9):1837-9.]

Watching your child's eyes

Some doctors recommend routine formal eye exams for children with juvenile myositis. A Canadian Children's Hospital decided to record the frequency and range of eye involvement in their JM patients to determine the need for formal evaluations.

More than half of the children had abnormal eye exams, most commonly problems with eyelids. Some children also had corticosteroid-induced cataracts; other abnormalities were rare. These findings suggested that formal evaluations are not needed as both of the more commonly-found abnormalities are as easily detected by non-ophthalmologists. Ask your child's doctor to let you know of any concerns about cataracts or other possible eye problems.

[Akikusa JD, Tennankore DK, Levin AV, Feldman BM. Eye findings in patients with juvenile dermatomyositis. The Journal of Rheumatology. 2005 October;32(10):1986-91.]

Methotrexate as a first-line treatment

Side effects of prednisone are all too familiar to most children with JM. A group of Canadian researchers tested methotrexate as a part of the initial treatment with corticosteroids rather than its traditional role as a secondline medicine. They treated their patients with high-dose but aggressively tapered corticosteroids along with methotrexate at diagnosis. A control group consisted of children already on high-dose corticosteroids only.

Their results support the use of methotrexate and corticosteroids as a combined first-line therapy, showing it is as effective as the traditional high-dose corticosteroids alone in treating JM. The combined therapy also allows for a lower overall dose of prednisone.

[Ramanan AV, Campbell-Webster N, Ota S, Parker S, Tran D, Tyrrell PN, et al. The effectiveness of treating juvenile dermatomyositis with methotrexate and aggressively tapered corticosteroids. Arthritis and Rheumatism. 2005 November;52(11):3570-8.]





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Feel good about drawing

Did you know that every time you color, draw or paint, you may just be helping yourself? Art is not only fun to do, it can relax you, make you feel

good about yourself, and give your family time to work together.

Many hospitals use art therapy to give their patients a way to express themselves and deal with frustrations, pain and sadness. Even if you don't have an art therapy program near

you, you can start one for yourself in your own home.

Find an out-of-the-way place and

Your myositis teams

You may not think of your doctors, nurses, parents, schoolteachers, brothers, and sisters as a team, but that's exactly what they are. They

make up different teams to help you, and you're the most important player on all of these teams.

Who's on your teams? Medical team = your doctors, nurses,

social workers, therapists, dieticians, and anyone else from the hospitals and doctors' offices who treat you. *Community team* = your friends,

set up a table with paper, pencils, chalks, paints, crayons, or whatever you like to use to create art. Then start drawing! Don't worry about being perfect. Art isn't about being exact - it's about expressing yourself

> in your own unique way. There are no "wrongs" in the world of art.

Send your drawings to TMA, and we'll use them in publications. on the web site or on note cards. Include vour name.

address, age, and diagnosis. TMA will only use your first name, age and diagnosis on any

materials. Thanks for sharing your creations with us!

schoolteachers, principals, and organizations like The Myositis Association.

Home team = your mother, father, brothers, sisters, and other family members.

These three teams have members that help you in many different ways, depending on what each one of them knows best. Doctors and nurses decide what medicines will help you the most. Friends and teachers make sure you are doing well in school and give you the time and support you need. Most importantly, your family is there for you for just about anything – giving you medicine, helping with your exercis-

Activity: Scribble drawing

Put a piece of paper on the table in front of you. With a piece of chalk in your hand, pretend you're painting a fence in the air in front of you. Once

> you're relaxed, put the chalk on the paper and scribble without lifting the chalk from the paper. Stop when you feel like you're done. Look at your scribble and find a picture in your lines. Using a darker piece of chalk, trace the lines of the picture you've found. Now forget about the other scribbles and draw

things around your picture that fit with it. Have your parents, brothers and sisters join you - then have fun looking at what everyone in the family created.

es, and listening to what you have to say.

How can you help? Be a big part of your teams: talk to your doctors and nurses about what's hurting or feeling better; tell your teachers if you're having trouble with anything in school; and be sure your parents know how you're feeling, too. Depending on how old you are, you can take charge of different parts of your treatment – for example, know when you're supposed to take your medicines each day and tell your parents when it's time.

Working together with the other members of your team helps everyone!



CRAFTS FOR KIDS

Handmade Cards

These make great thank you cards for all your holiday gifts!

What you'll need: Construction paper Sponge Scissors Paint Paintbrush

What to do: Fold the construction paper in half to make a card.

Draw a design on the sponge; cut it out. This is now your stamp.

Brush paint onto the stamp.

Firmly press your painted stamp on the paper then remove.

Allow this to dry, then add your greeting to the inside.



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

Placemats for Any Occasion

What you'll need: Fabric (100% cotton or 50/50 cotton-polyester) Fabric markers Scissors

What to do: Cut fabric into a rectangle for the placemat.

Use fabric markers to color your picture or design.

Once you're finished, have your mom or dad "heat set" the design either by ironing on the reverse side for four minutes or putting in the dryer on the hottest setting for 30 minutes. (This way, you can wash the placemats later.)

Extras: Use zigzag scissors to add an interesting border. Consider designs that can be used at different times of the year, like snowflakes, colorful leaves or spring flowers, or for different occasions, like birthdays and holidays. Or personalize your placemats for each member of the family.

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