

## Dr. Ann Reed answers questions about JM

*Dr. Reed is a professor of pediatric medicine, and the chair of pediatric rheumatology at Mayo Clinic Medical School in Rochester, Minnesota. Dr. Reed has served as a member of TMA's medical advisory board and presented at TMA's 2008 Annual Conference. This is a shortened, edited version of Dr. Reed's recent live discussion. For the full transcript, visit TMA's website, [www.myositis.org](http://www.myositis.org). Participation in TMA's live discussions is a benefit of TMA membership.*

**TMA member:** Is a child with juvenile myositis taking attention deficit disorder (ADD) medication at risk? Do the medications mix well with other myositis medications? Can the ADD medication trigger episodes?

**Ann Reed, MD:** It all depends on what the other drugs are. For example, when I look at methotrexate combined with those medications, no specific interactions come to my attention except that none of them should be used if you are pregnant or breast feeding. The most accurate way to assess this is to have your MD or pharmacist run a check of drug interactions. They can give you a good idea of the KNOWN interactions.

Children with myositis do take ADD medications while on steroids, methotrexate and IVIG. However, steroids themselves can cause emotional lability (instability) and that might counter the effects of the medication.

**TMA member:** Why is it that the muscle enzymes are never elevated in some JM patients, yet there is obvi-

ously muscle weakness and that weakness has been confirmed on an MRI?

**Ann Reed, MD:** Great question. We actually don't know why enzymes correspond or not at different times of the disease and in different people. We do know (CPK) levels have to do with muscle mass, so some people have increased levels because of their muscle mass, and others have low levels because they have small muscles. Different enzymes are located in different areas of the muscle cell.

When the muscle cell is damaged, enzymes are released into the blood, so if there is a lot of inflammation and little muscle cell perturbation, the enzymes might be normal. We also know that, after time, we see fewer of the enzymes released.

**TMA member:** What are your thoughts on every-other-day dosing of prednisone for "maintenance" of JDM?

**Ann Reed, MD:** Every-other-day steroids are not usually recommended in JDM, but are used when you are taking a patient off steroids. When steroids are used, they are thought to be needed on a daily basis. The goal is to continue to wean steroids while another agent is used for maintenance.

**TMA member:** Can you address the following regarding methotrexate: 1) Difference in absorption between oral vs. subcutaneous administration 2) Incidence of GI issues between oral vs. subcutaneous administration?

**Ann Reed, MD:** Methotrexate is thought to have better absorption when given subcutaneously in those children who have GI involvement.

Since we don't know how many children have GI involvement, some providers recommend subcutaneous methotrexate for everyone in the first few months. However, many will begin with oral methotrexate and if someone is not responding, will then switch to subcutaneous. Nausea is more common when methotrexate is taken orally.

**TMA member:** What is the maintenance dose of prednisone that you typically recommend for your patients?

**Ann Reed, MD:** There is usually no maintenance dose for children. Our goal is to control the disease and then wean the steroids. We prefer to use non-steroid medications to maintain disease remission. However, if steroids are the only medication that works, we try to use the minimum that is possible. Usually the dose is based on weight and response to treatment.

**TMA member:** Our daughter has been completely asymptomatic for 3-4 months with the exception of nail bed activity (which is only visible via magnification). Is it appropriate to leave prednisone at a maintenance dose and begin reducing other meds at this time?

**Ann Reed, MD:** The goal is usually to stop the prednisone and minimize the side effects. Also, sometimes in this situation, the IVIG might be weaned because of the cost and difficulty in getting an infusion.

**TMA member:** Our society emphasizes the importance of keeping our

Published by: TMA, Inc.

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immune system healthy. Immunosuppressive medications have become standard treatment for juvenile dermatomyositis. Have any studies been done that indicate possible long-term effects from turning off the immune system in children with myositis?

**Ann Reed, MD:** First, I know of no long-term studies that have been performed which look at the immune system 10 or 20 years later in JDM. Studies have asked patients how they are doing and most say they are doing well with minimal disability. A subgroup of children goes on to develop other autoimmune conditions as well. You need to think of the immune system as a balance of over-activity with potential areas that are not working as effectively. So the idea of immune suppression is to bring back the balance (or what we call homeostasis) in the system.

**TMA member:** Is it necessary to take hydroxychloroquine (Plaquenil) for treatment of JDM when you do not currently have skin involvement?

**Ann Reed, MD:** No, many children are not on Plaquenil. It is thought to help rash, but I do not use it in all my JDM patients, and sometimes it is not needed.

**TMA member:** How can you make fish oil supplementation palatable? My child does not swallow pills easily and has extremely sensitive and easily offended taste buds.

**Ann Reed, MD:** Fish oils do not taste good. Freezing the capsules actually can help. Sometimes other sources of Omega 3s are used which are more palatable. There are liquid forms of Omega 3s as well, and some can be sprinkled on food.

**TMA member:** What issues should we concern ourselves with in considering returning to school? What needs to be addressed in an Individual

Education Plan? How can we live a sane life and be vigilant about germs and viral contamination?

**Ann Reed, MD:** Returning to school is variable depending on the dysfunction a child has had. For some children, it's the emotional stress of returning to school caused by the change in their appearance with steroids. Others are anxious because they have been out a long time due to their illness. First, I would make sure there is a plan for re-entry set up. This means meeting with the teachers, making sure everything is accessible; making sure the child is comfortable with getting around school.

Stairs or gym class may be something they can't do at first. An IEP is great to help with the needs of your child at school.

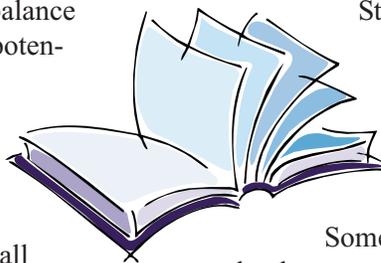
Sometimes it helps to have the doctor or a psychologist talk with the class about the illness and what the child has gone through.

Sometimes an older child does not want anyone to know. In that case, it's beneficial to prepare him or her with answers and ways to approach situations.

**TMA member:** Is it advisable for a JDM child to receive Enbrel injections for joint inflammation that may or may not be a result of the JDM?

**Ann Reed, MD:** It is not clear whether Enbrel or any of the TNF drugs are beneficial in JDM or may cause the disease to worsen. Early studies do not suggest these drugs are greatly beneficial in treating the disease. However, there are cases of children with ulcers and very hard to treat disease where Enbrel has been helpful. I am not sure I would choose this drug to treat arthritis with JDM unless everything else is in remission or quiet. I think we have other options we can use.

**TMA member:** After 7 years of constant treatment for JDM, my daughter



has not made it into remission. She is currently on CellCept, methotrexate and Plaquenil, and all her muscle markers are normal. We are weaning the methotrexate slowly over a year this time. Is it possible she could still get into remission after all this time has passed?

**Ann Reed, MD:** Yes, it is possible. Sometimes it takes the right combination of medications, time, and then whatever is stimulating the disease finally subsides.

**TMA member:** What long-term physical damage could realistically occur after treatment and serious bouts of the illness?

**Ann Reed, MD:** Damage from treatment is usually during the treatment itself. It depends on what the treatment is. Long-term problems from the disease range from mild difficulty walking, difficulty using muscles, tight muscles and scars on the skin to life-threatening disease in children with severe weakness and organ involvement (such as lung disease).

**TMA member:** How long does medication (excluding prednisone) continue after a child is asymptomatic?

**Ann Reed, MD:** We use our immune suppressive drugs for at least 2 years after the start of treatment. The goal would be to get a child asymptomatic and then minimize the medications. That usually means getting them off prednisone and other medications that were used to induce a remission, and leaving them on maintenance medication. Maintenance medications are usually, but not always, methotrexate, Imuran or cyclosporine. At times other medications are the maintenance ones, such as IVIG, Rituxan, Cytoxan and also prednisone.

**TMA member:** In your opinion, what research drugs for JDM are you most excited about? I thought the anti-TNF drugs were supposed to be very promising but based on a previous comment about Enbrel, it doesn't

appear you agree with this.

**Ann Reed, MD:** Enbrel is wonderful for some autoimmune conditions (arthritis) but not for others (myositis and lupus). There are new drugs that do show or suggest promise. There are medications that block interleukins that show promise. A medication being tested in adult myositis blocks interferon. Also Rituxan (an anti-B cell treatment) is under investigation in a large international trial. In reports before the study was done, positive results occurred in adults and children. Also new therapies against IL-6 and IL-17 are in development and testing, and all seem promising.

**TMA Member:** My son became quite overweight while on prednisone and is not losing it now that he has been off it for several months. I suspect part of it is he is still weak and doesn't get much exercise, and his prednisone appetite hasn't diminished. Any ideas for helping him without starving him?

**Ann Reed, MD:** This can be challenging, to say the least. Trying to get back to the pre-illness activity and dietary habits are key. Introducing a regular physical activity program not only helps muscle rebuild and normalize, but will get the child out of the routine they started when the disease was active and they could not exercise. Also making sure the entire household is changing their eating habits to those needed for the child (if you can) sometimes allows JDM children to achieve their goals.

**TMA Member:** Is there anything on the horizon that looks like it would be helpful for calcinosis? Do you know of any examples where surgery has been effective? When do you recommend surgery?

**Ann Reed, MD:** Surgery has been found to be helpful when the calcinosis is painful and in an area that it is causing problems for the child. No new medications have shown to be helpful but we do strongly feel calcinosis is related to ongoing inflammation and research is presently shedding light on that fact. Our hope is that the research will allow new therapeutic interventions to occur.

**TMA Member:** My daughter has started IVIG and sometimes she has chills afterwards and doesn't feel well. Is this a sign that she may be allergic or at risk?

**Ann Reed, MD:** You can have allergic reactions to IVIG that occur while a child is getting an infusion. However, more common is a response to the "protein" that is being infused. So it is a non-specific response which causes fever, chills, nausea etc. There are things that can be done to minimize this if it occurs.

**TMA Member:** Are there any diagnostic tools that can predict whether the disease will be a one-time flare, go on for years, or be a lifetime problem?

**Ann Reed, MD:** Not at this time, but we are actively working on that.

**TMA Member:** What are you seeing in the juvenile patients in the RIM study? Is the study still enrolling children?

**Ann Reed, MD:** The study is now closed to new patients. I do not know the number of patients that improved or did not improve. As someone who runs the study, I am not able to know this until all the subjects have completed the trial. However, in patients I treated prior to the onset of the study with Rituxan, the majority responded when previously they were resistant to treatment.



# STUDIES EXAMINE CALCINOSIS, QUALITY OF LIFE, COMMON SKIN PROBLEMS

## Researchers find clues to calcinosis

An international group of researchers, including several TMA medical advisors, cooperated in a study based at the Royal National Hospital for Rheumatic Diseases and the University of Bath in the U.K.

In this study, physicians treating JM patients wanted to see if the presence of certain autoantibodies had a connection with disease features and disease course.

Researchers examined autoantibodies in children recruited for the Juvenile Dermatomyositis National Registry and Repository for the United Kingdom and Ireland.

They collected clinical observations of children with juvenile myositis and their blood test results, and any blood tests that confirmed a certain protein (140-kd) were identified. DNA samples from 100 Caucasian children with myositis were compared with those from 864 randomly selected UK Caucasian control subjects.

Researchers found that 37 (23%) of 162 patients with JM had anti-p140 autoantibodies, which were detected exclusively in those patients and not in patients with JM-overlap syndrome (JM with other autoimmune diseases). None of the control subjects (those 864 children without JM) had any evidence of anti-p 140 autoantibodies.

Researchers also found that none of the antibody-positive patients were positive for other recognized autoantibodies. In children with anti-p140 autoantibodies, the presence of calcinosis was significant compared with

the rest of the children in the study.

The clinical features of patients with



anti-p140 autoantibodies were different from those of children with anti-p155/140 autoantibodies. The study is significant in that it establishes that anti-p140 autoantibodies represent a major autoantibody subset in JM. This may identify a further type within the juvenile myositis spectrum that includes an association with calcinosis.

## Measuring the quality of life with juvenile myositis

A study published in the April 2009, edition of "Arthritis & Rheumatism" reported on the health-related quality of life of patients with JM. The report, the collaborative effort of the Paediatric Rheumatology International Trials Organisation based in Genoa, Italy, was partially funded by TMA. Researchers set out to investigate the health-related quality of life and its changes over time, in patients with active JM.

The study assessed patients with

JM both at their first visit and after 6 months of follow-up. Healthy children 18 years and younger were also studied.

Some of the markers used to measure the quality of life were physician and parent observations, muscle strength, functional ability, disease activity, and laboratory tests. A total of 272 children with JM and 2,288 healthy children were enrolled from 37 countries. The JM children reported significantly lower quality of life than the healthy children, with physical well-being being the most impaired measurement.

Quality of life improved over time for children who responded to treatment, but remained unchanged or worsened in children who did not respond to treatment. Both physical and psychosocial summary scores decreased with increasing levels of disease activity, muscle strength, and parent's evaluation of the child's overall well-being.

The degree of disability and longer disease duration were the major determinants for poor physical well-being at the follow-up of the JM children, and physical well-being was most affected by the level of functional impairment in the children.

## Use of MRI in juvenile dermatomyositis

A study by the Department of Child Health at the University of Glasgow in Scotland examined the role of MRI in the assessment of the musculoskeletal system in children. Using MRI in children has important differences from its use in adults, because growth in children has significant impact on important structures, and there are radiological features that differ from those in adults. The study said that disease may alter structures

during a period of growth; and that the diseases themselves are at variance with adult forms of myositis, so many technical issues are different when a child is involved. These are important considerations in choosing the appropriate imaging. Researchers concluded that MRI is a powerful and valuable imaging technique in pediatric musculoskeletal pathologies, with considerable potential for a greater role in diagnosis, management, and therapeutic intervention for children.

### **Weakness in patients with polymyositis, dermatomyositis and juvenile dermatomyositis**

A study by a number of myositis experts coordinated at George Washington University School of Medicine and Health Sciences in Washington, DC, described the distribution and severity of muscle weakness using manual muscle testing in 172 patients with PM, DM and JDM.

The researchers also wanted to characterize individual muscle group weakness and determine how weakness was associated with functional status and myositis characteristics in a large number of patients with myositis.

Experts assessed strength in 13 different muscle groups and arrived at a total score, subscores based on functional and anatomical regions, and grades for individual muscle groups. Patient characteristics and secondary outcomes, such as clinical course, muscle enzymes, corticosteroid dosage and functional status were evaluated for association with strength.

The study identified a continuum of proximal muscle (muscle nearest the trunk) weakness, with PM the weakest, DM intermediate, and JDM strongest among the three myositis clinical groups. Hip flexors, hip extensors, hip abductors, neck flexors and shoulder abductors were the muscle groups with the greatest weakness

among all three clinical groups. Muscle groups were affected symmetrically.

### **Researchers review the effects and implications of skin manifestations in JM**

In “Dermatology Online Journal,” Drs. Elizabeth Dugan, Adam Huber, Frederick W. Miller and Lisa G. Rider, of the International Myositis Assessment and Clinical Studies (IMACS) Group, classified the various skin problems associated with JM. The report is accompanied by a comprehensive collection of photos, including photos of skin problems in children of color and those with long-term skin disease. The authors say that consideration of the manifestations of the disease in the skin is important for accurate and timely diagnosis, prediction of outcome and for the ongoing evaluation of response to treatment. This is particularly true when the cutaneous features come before skeletal muscle, lung or other systemic involvement.

In addition, the authors say, the skin may be the most active or difficult feature to manage in JM and may have a significant negative impact on patients' quality of life. They quote a study that evaluated the impact of active cutaneous (skin) symptoms of DM on quality of life in 71 adults with DM and found that the impact on quality of life was greater than for patients with psoriasis, atopic dermatitis or vitiligo. Pruritus (itching), an under-recognized symptom of DM, is a large component of the impact on quality of life, and one study found that pruritus occurred in 38 percent of children with JM.

In children, calcinosis, which is associated with long-term skin disease, can result in pain, ulcers, infection, scarring, and limited range of motion and physical dysfunction. Lipoatrophy (loss of fat tissue), most commonly seen in juvenile DM, can result in dramatic changes in appear-

ance, and may be associated with significant metabolic changes, including type II diabetes and insulin resistance.

There are significant negative psychological consequences to skin problems, particularly if located in cosmetically sensitive sites. Finally, the presence of persistent skin disease is recognized as being connected with ongoing systemic vascular disease, which may be associated with serious and potentially life-threatening systemic disease.

In this review, which can be found online, at <http://dermatology.cdlib.org/1502/reviews/photoessay/rideressay.html>, the authors consider recent advances in understanding the role of types of antibodies, with an emphasis on findings that are relevant to the understanding of skin disease. There's a photo-essay also online, that provides a comprehensive visual catalogue of JM skin disease, with photos of how the skin appears in people of color and in people with longstanding disease.



## Stories that heal

Kate Creskoff was in high school, the captain of her swim team and a hard-working athlete and student. When she began feeling exhausted, she chalked it up to training too hard and staying up too late. As time went by, though, her symptoms became too serious to ignore. Finally, in the midst of her senior year, she went to dozens of doctors, before finding out that she had Sjogrens Syndrome and other autoimmune diseases. The diseases kept attacking her for several years.

As Kate went through treatment, she was frightened and angry that her life had been derailed in such a dramatic way. She went from being a star swimmer to barely being able to follow the swim meet from the stands.

At one point, Kate realized that the effects of her medications were worse than the symptoms of her disease. She worked with her doctors to reduce her medications while she concentrated on setting goals for herself. She was accepted into Dartmouth University and graduated from there, meanwhile managing the ups and downs of flares and remissions with constant optimism. To this day, Kate believes that optimism was a huge factor in her recovery.

After graduating, Kate worked with the C. Everett Koop Institute and the S.T.A.R. (Steps to Adult Responsibility) program to collect the stories of teenagers with chronic disease. Kate's full story, along with those of other teens with chronic and, in some cases, near-fatal, diseases make fascinating reading. Find them at [http://www.starprogram.net/files/Nine\\_Stories.pdf](http://www.starprogram.net/files/Nine_Stories.pdf)

## Lead your own individual education plan (IEP) meeting

Who better than you to lead the discussion of those interested in your success in school? Many teenagers have done it and believe it to be a milestone in their lives. Whether you just need a few small adjustments, or require a significant change from business as usual at your school to receive the training you need for college or a career, now's the time to make it happen. There are dozens of resources available. Find them at [www.nichcy.org/InformationResources/Documents/NICHCY%20PUBS/st1.pdf](http://www.nichcy.org/InformationResources/Documents/NICHCY%20PUBS/st1.pdf)

## Learn how to communicate with health professionals

Maia Wroblewski is now in college, but she started several years ago to take over the management of her own health care. "It is completely normal for a youth with a special health care need to have frequent trips to their specialists," Maia writes: "Freedom to control our own appointments is a big step to independence. The next step is to be on your own at the actual appointment." Maia's tale of two appointments – one where not much was communicated and one where a great deal was accomplished – is a wonderful illustration of how all of us fail to make progress in our health care simply by not choosing the right words. In Maia's case, she used the word "okay" as an answer when the nurse asked her how her month had been. Maia was indeed "okay" during the visit, but she'd had ups and downs during the month and had some questions.

On the second visit, Maia told the nurse that she had questions for the

doctor about her medications. She also brought a complete list of medications so the doctor could review them.

The two visits, with their different conclusions, are published in full at <http://www.fvkasa.org/resources/files/health-communicating.html>.

### Suggestions from Maia:

- It is best to start weaning yourself off your parents' presence early. Try having your parents come into your appointment, but agree that they won't talk.
- Go over questions and your health status before the visit, and write anything down that you might forget. Remember: the doctor wants to talk to you, especially as you get older.
- Also, don't forget to be straightforward with your doctor. I know some things can be embarrassing or 'not cool' to talk about, but you have no reason to be embarrassed.
- Doctors also respond better if you use mature language. Instead of tummy, say stomach; instead of achy, try being specific about what type of ache you have.
- If you don't understand something have them write it down, or explain it slower.
- Finally, review the visit with your parents at the end. This will help you sort out what happened at the visit, and what each of you believe happened at the visit.

Find other tips for making the transition to adulthood at [www.fvkasa.org](http://www.fvkasa.org).





# JUST FOR ME

Fun facts and news for children affected by JM

## Ditch the Itch

It's summer, and everyone has an itch or two from time to time. Mosquitoes, poison ivy, flies, gnats and tons of other little bugs and biters set grownups and children to scratching once in a while.

If you have JM, though, you may have much more than a few little itches. The JM rash can be very itchy, and it often gets worse in the summer. If it gets very bad, you may have trouble sleeping or even playing. Instead of scratching all day, let your parents know that your rash is really bothering you, and your doctor will give you some medications to help. There are also a lot of things you can do to make things better. Here's how you can ditch the itch:

- Drink lots of water. The JM rash is made worse by dry skin. You can help prevent dry skin by drinking water and other drinks throughout the day even when you are not thirsty.
- Always use sunscreen. A rash that has gone away

can come back after too much sun. Light, cool clothing also provides you with some protection from the sun.

- If you're taking a new medicine and you begin itching, make sure to let your parents know. Some medicines cause skin to be extra sensitive to the sun and some cause itching on their own. Itching can be a sign that you're allergic to the drug and should stop taking it.

- Reduce the use of soap, which can make skin dry. You can rinse off after swimming with warm water. When you do use soap, use a mild one.

- Use lotions and moisturizers throughout the day. They are very soothing to the JM rash.

- If you see any change in your skin, let your parents know.



## Summer things that ITCH

- Mosquitoes
- Swimming in a pool
- Too much sun
- Poison Ivy
- Poison Sumac
- Certain medicines
- Some kinds of soap
- A rash
- Dry skin

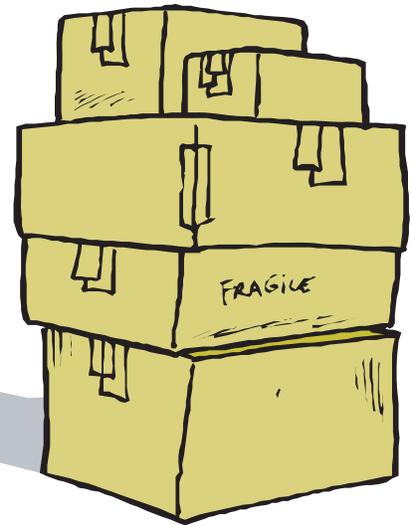
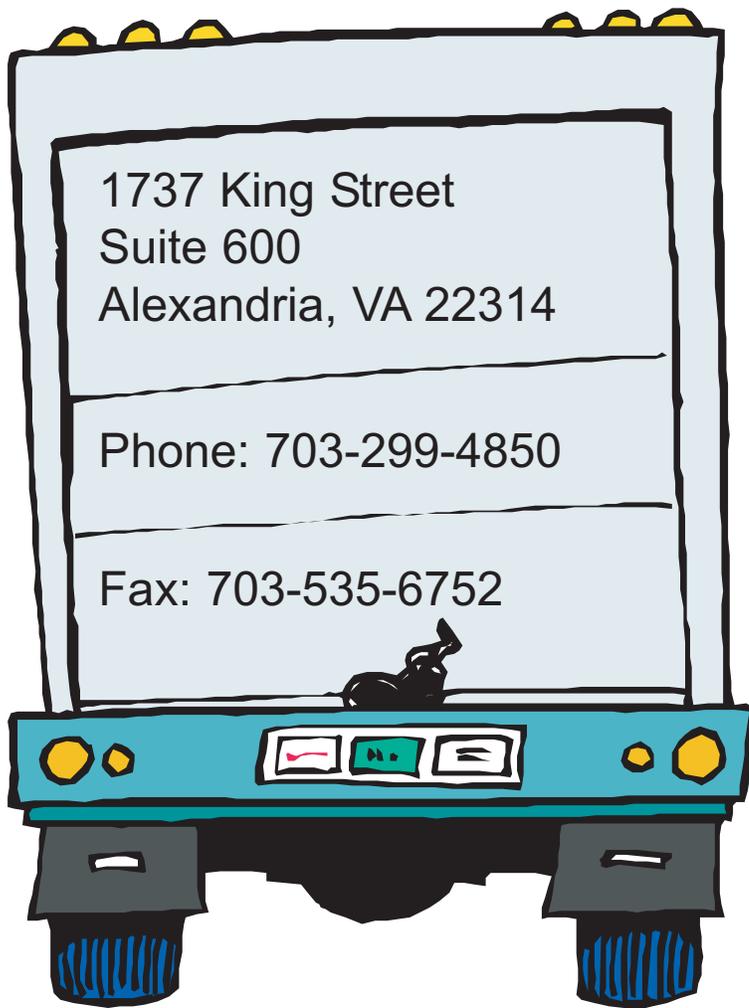
## Word jumble

Straighten out the letters in the following words related to itch!

1. traschc
2. tionlo
3. oispon vyi
4. llrgeya
5. shar
6. snik

1. Scratch 2. Lotion 3. Poison Ivy 4. Allergy 5. Rash 6. Skin

**TMA has moved!**  
**Update your records!**



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**JM COMPANION**