

IVIG: a guide for parents

Many of TMA's juvenile myositis patients receive IVIG, an abbreviation for intravenous immunoglobulin. Writing in "Myositis and You," Drs. Rider, Pilkington, Lang and Malleson tell us that this treatment (called a "second-line" treatment because it's usually a supplement rather than the primary treatment) has been useful for treating long-term JM to allow the dosage of prednisone to be lowered. It's also used in situations where a child is extremely ill or when there are life-threatening complications, such as trouble swallowing.

IVIG "turns down" parts of the process causing inflammation in children, and usually does it within a few days. Its advantage over other JM treatments is that it is able to stop the immune response that causes JM without affecting the entire immune system, so your child can heal quickly from a cold, or an infection of any kind.

How is IVIG made?

IVIG comes from human plasma. Human plasma is the liquid part of the blood that does not contain red blood cells. There are 5 types of Immunoglobulins, Vaughan said: IgG, IgA, IgM, IgE and IgD. Current IVIG preparations are highly concentrated, containing 90-98 percent IgG; however, there are small amounts of IgA, IgM and IgE in IVIG products. Preparations are screened thoroughly for blood diseases like HIV and hepatitis B and C before pooling.

Once purified, the solution is delivered intravenously. It's been use-

ful to treat three categories of illnesses: immunodeficiencies, autoimmune neuromuscular disorders, and certain rheumatologic conditions. IVIG takes advantage of the factors at work in a large number of healthy immune systems. Doctors often call this "passive immunity." Although its benefits have been well documented, no one understands precisely how it works.

IVIG side effects

IVIG is generally well tolerated in JM patients, write the authors of "Myositis and You." A great many precautions, including screening the donors and refining the manufacturing process, are taken to make sure IVIG doesn't carry any trace of virus, bacteria or any other infections from the multiple donors.

Although it's considered to be a very safe treatment, there are sometimes side effects, the most common of which are low-grade fever, chills, headache, nausea, changes in blood pressure, flushing, an increase in the rash, fast heart rate, back pain, muscle aches, shortness of breath and chest tightness.

What causes adverse reactions to IVIG?

A group of Canadian researchers noticed that most prospective trials for adults taking IVIG found no difference in efficacy and safety between different IVIG products.

However, they heard repeated reports about children who experienced side effects that seemed to depend on the type of IVIG pre-

scribed. They'd be nauseated, for example, when infused with one brand of IVIG, and do fine with another.

Drs. Manlhiot, Tyrrell, Liang, Atkinson, Lau and Feldman studied whether differences in the immunoglobulin A content could be a risk factor for adverse events. All of the children reviewed had been seen in the juvenile myositis clinic at the Hospital for Sick Children in Toronto, Canada, from 1986 to 2005. They found 135 children had been treated for JM. The children who were prescribed IVIG either were not responding adequately to prednisone therapy or could not be weaned off long-term prednisone without a return of active disease.

During the time period studied, they found that 38 patients with juvenile dermatomyositis received 1,056 infusions at the Hospital for Sick Children. There were 92 adverse events (in about 9 percent of the infusions) affecting 25 patients (66 percent), a frequency higher than that usually reported in adult patients, where only 1-5 percent of infusions result in adverse events.

Children who reported problems with one infusion often went on to have many more infusions without any problems, so most of the adverse events were at the first infusion. Often, once they had a bad experience, they were treated before later infusions with antihistamines, corticosteroids (prednisone), or both. The

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course of infusion for the children studied was typically about 2.5 years.

The study determined that adverse events were reported more often (nearly twice as often) with products that contained high immunoglobulin A.

Specifically, the adverse events were fever (8 percent with high immunoglobulin A vs 1 percent with the low immunoglobulin A), lethargy or malaise (2 percent vs .1 percent), and nausea or vomiting (5 percent vs 1 percent).

For the infusions, healthcare workers used products that were readily available at the time needed. However, they tended to prefer Iivegam, when available, because physicians had learned from personal experience that it seemed to cause fewer adverse reactions. They did find that adverse events varied significantly between IVIG preparations. Gammagard was associated with the lowest frequency of adverse events, followed by Iivegam, and finally by Gamimune. The reactions most commonly reported were fever, headaches, nausea or vomiting, and lethargy. There were no life-threatening reactions to IVIG.

Based on the ingredients of each product, the researchers were able to classify Iivegam and Gammagard as having low IgA levels, whereas Gamimune N was classified as having high IgA levels.

This study, the only one of its kind, seems to confirm informal reports that products with high IgA concentration are associated with more adverse reactions than products with low IgA content. The most common complaints were nausea and lethargy. The researchers found that headaches seemed not to be related to the IgA content, suggesting they may be reactions to the intravenous pro-

cess or to other unstudied aspects of IVIG.

Be informed

Time is often short with healthcare providers, but you're sure to have questions about your child's treatment. Here are some questions you should ask well in advance of your child's first infusion:

- Which IVIG therapy have you chosen for my child and why?
- What will he or she be able to do while the infusion is going on?
- Who should I contact if I have concerns about the infusion? Is there an after-hours contact in case of an emergency?
- Can your office or the IVIG company help me understand insurance coverage for IVIG?

Work with your child to overcome fear

Carolyn Kidwell likes the way the child life specialists used play therapy to help Savannah, her four-year old grandchild, overcome the fear of the IV starts. Cathy Harrison brought medical supplies to the room and let Savannah practice all the steps involved in an IVIG infusion on a specially-made puppet.

Savannah learned everything from preparation to inserting the needle and tube under close supervision. Harrison sent more medical supplies home with Savannah to practice on her dolls.

"For weeks the dolls and stuffed toys at Savannah's house were almost always wrapped in brightly colored Coban bandages!" Carolyn said. Harrison also was helpful in suggesting reading material for children. Savannah's family reads "Franklin Goes to the Hospital" often, and it helped Savannah remember what to expect.

Another book addresses a fear that children don't always express. "Why is Keiko Sick?" deals with that ques-

tion in a non-judgmental way, explaining that disease is not a child's fault, sent because he or she was bad." "Savannah was helped with her acceptance and attitude about her illness by that book," Carolyn said.

Another tip, from the experienced JM grandmother: "The attitude of the parents is picked up by the child and I can't stress that enough to those I have talked to. Even if you are falling apart on the inside don't let the child see it. You have to remain calm and matter-of-fact about what needs to be done."

Savannah's family makes a special occasion of infusion day. Savannah, who is still young enough and hungry enough from the prednisone to like hospital food, is allowed to order whatever food she wants from the hospital menu. Her grandmother and she consider it their special time and bring movies and games to play at the hospital.

Savannah's attitude has improved to the point where her nine-year-old sister was shocked last month when she heard Savannah singing "Tomorrow is IVIG day." Her sister asked, "Why in the world are you happy?" Savannah just said, "It's not so bad."

"I'm impressed by her attitude," Carolyn said. "I don't know if I could handle all that she has so well."

Another way that Savannah's family turned a stressful situation into fun was by having a party when Savannah had her methotrexate injection. "Savannah loves to dance, so after her injection, which was given at bedtime, she was allowed to stay up 30 minutes later and dance to her favorite music," Carolyn said. "The whole family turned out for that, to make it as special as possible."

Savannah, now a shot veteran, doesn't need the party anymore and handles her shots in a matter-of-fact way.

Other families report their own ingenious ways to put some fun into infusion day and to introduce their children to the idea of being resilient and happy despite difficulties. One mother has her son, 12, take photos of the hospital and the people he deals with every time he has an infusion. He's learned the favorite candy and sports teams of his favorite nurses and brings candy some weeks. Other weeks, he may wear a temporary tattoo of a rival team just before a big game. Often, a different family member will come for a visit, and they have special dinners, picnics and movies on infusion days.

Older children are more likely to worry that something will happen during the infusion, especially if they've seen it happen before, either to themselves or someone else. It's important for parents not to magnify the hardships their children may encounter: if they feel sick, they'll soon feel well; if the nurse has a hard time finding a vein, they'll drink more water next time; if the numbing cream fails to numb the infusion site sufficiently, the pain will soon fade. Children rely on their parents to provide realistic scenarios to counter their own fantasies, which often come from bits and pieces of misinformation and the fear inherent in being powerless and not knowing the overall picture.



Caregivers need care

Andrew White, a Colorado professor and counselor, presented a session on caregivers and stress at the 2009 Annual Conference in Charlotte,

North Carolina. See the Outlook, page 11, for more on this session. His remarks below are aimed specifically at parents and other relatives who find themselves in the role of caregivers for their chronically ill children.



It is very important for parents who have children with chronic illnesses to be relieved of their duties on a regular basis. Burnout is very common among parents who are taking care of their children in these difficult circumstances on a daily basis. Because they are providing in-depth care, often on an hourly basis, parents can easily become physically exhausted, emotionally drained and psychologically imprisoned by stress. Parents MUST make a consistent effort to have other family members and close friends relieve them so they can get away and get some time to themselves, and if married, then have a couple's night out several times per month. It would be good for parents to get into the habit of having alone time once a week if at all possible.

Parents who have chronically ill children need to get connected with other parents in the same or similar situations. They need to be involved in a support group together or even a social group getting together twice a month. In regards to a support group, design the group to have open discussions on whatever topic comes up for the week and have a set agenda where parents take turns conducting research on their child's illness and bringing it back to the meetings to share and educate everyone else. A collective

and collaborative sharing of research and engaging in discussions is not only essential but is very powerful. The collective social get-togethers among such parents serves as a positive outlet for them - it is their time to unwind, laugh and have fun together as a group.

Finally, it is important for parents not to become enablers for their children to the point where they are smothering and doing everything for their children. Certainly, children having chronic illnesses are indeed limited in what they can do. However, it is to the child's benefit (and to the parents' benefit too) to be allowed to do all the things that are within their capabilities. For example, if the child can bathe him or herself and provide other personal care needs, then allow the child to continue to do these things instead of parents taking over. There is a profound difference between nurturing and assisting and smothering and controlling. Having said all of that, I want to express that parents sometimes can go to the extreme on the other end of the spectrum too. Parents can push their children too much or want to do too many things with them as they realize that time is short. There has to be an evaluation done and parents should be able to come to a balance of some kind.

Andrew White, who teaches counseling and has a family counseling practice in Colorado, has polymyositis. Find his full presentation online at www.myositis.org.

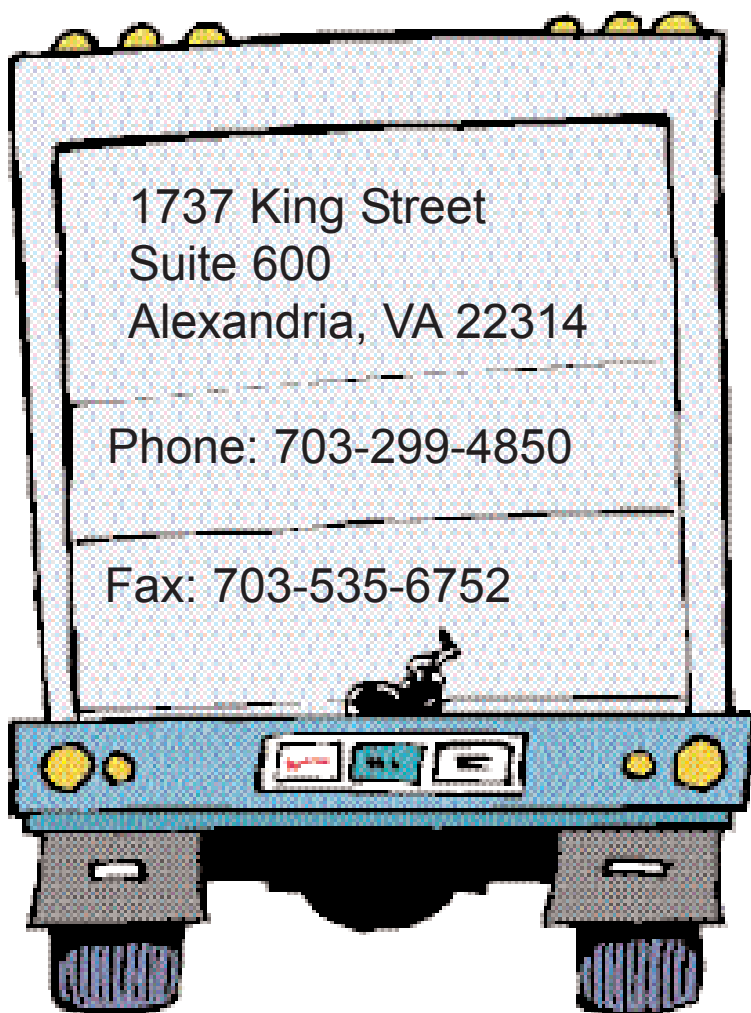


This issue of JM Companion is shortened to make room for the TMA 2009 Conference summary. Read the Outlook for sessions of interest to JM patients. Sessions are also on the TMA website at www.myositis.org.

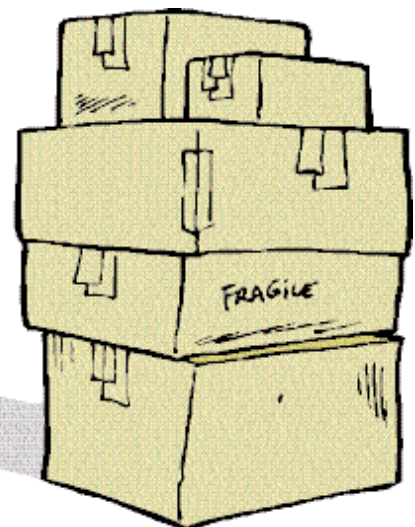


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