

December 2012: Summary of "Sick Protocols for Mito Patients"

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Dr. Korson, a founding member of MitoAction, has years of experience with Mito patients and unfortunately sees plenty of "sick kids." His discussion today is to help Mito patients and parents of affected children to think about what to do if they (or their child) gets sick. Over the years Dr Korson believes he has learned much from his patients and their families about how they navigate through the health care system - sometimes with ease, other times with more difficulty. And he has seen how advocates can be a great help to these families to clear the way for them.

The focus of today's discussion will be on **infections and viruses**, not on Mito symptoms or other illnesses. Infections, like the common cold, are temporary, but since Mito patients may need hospital support during illness, advocacy is as important as prevention. How Mito patients respond to an infection is variable and individual (just as Mito symptoms). Since all children and adults with Mito respond differently, some may do fine at home with support while others very quickly get into situations where they are better supported in the hospital. Adult patients and caregivers should know how they respond to infections and need to be ready to respond at the first signs.

When a person gets an infection like the common cold, the body responds aggressively to fight off the infection, thereby using lots of energy. Anyone with an infection feels tired and "low" on energy. This is multiplied many fold for the person with mitochondrial disease. For the Mito patient, the symptoms of a runny nose, cough or sore throat don't necessarily get worse, but the fatigue can be profound and severe. In addition, recovery and very low energy is typically prolonged - often days to weeks.

Autonomic nervous system symptoms can also be more severe during bouts of infection. The autonomic system controls those parts of the body that we do not think about - for example, breathing, sweating, heart rate, & digesting food. Often times these autonomic symptoms are exacerbated when the body is stressed by illness or environment. For example, if the child or adult Mito patient is overheated, the gut may slow down. During illness, symptoms often "flare" or get worse due to the high energy demand of an infection. Viral infections typically slow gastric emptying for all patients, but this effect can have much more of an impact on the Mito patient.

Hormones also naturally respond to the stress of an infection; for example, the pancreas' control of blood sugar is altered during stress. Adrenal gland function also responds to stress. In some patients their adrenal glands function normally until they are ill; at this time, patients have what is commonly called "sick adrenal syndrome." This syndrome prolongs the recovery time from even mild infections.

Many Mito patients are familiar with the "Mito crash" as a response to even a minor illness. The majority of Mito patients don't get more infections, but rather they take much longer to recover and can experience significant complications along the way to recovery. However, some Mito patients do experience a compromised immunity as part of their symptom presentation, making prevention of infection and active care during illness even more important.

What signs & symptoms are urgent and require immediate care?

This depends very much on the individual patient or child. Look for signs or patterns that led to complications in the past. For example, if in the past the child vomited and then became dehydrated, do not wait for dehydration to occur this time. Intervene early. Sometimes if a child is vomiting and can't keep anything down, their gut is simultaneously slowing down and it becomes increasingly difficult to keep them hydrated. Often there is no easy way to increase fluids without IV support in the hospital. If infections trigger seizures, then at the first sign of infection, take note and follow your neurologist's orders or plan. Anything which impedes breathing can be serious - during an illness the muscles are weak due to energy loss and those that control the diaphragm & walls of the chest may be weakened.

When to call the doctor

Parents and/or their families often don't know which doctor to call during an illness like a cold or virus or flu: the specialist or the PCP? Dr. Korson highly recommends keeping the PCP or pediatrician involved. If parents find that their pediatrician or internist seems reluctant to get involved because the case is "complex", then try to find another primary care physician. There are pediatricians and primary care physicians who do not mind having a "complex" case and who appreciate always being "kept in the loop" and involved. This is important because you do not need to drive miles to get to the medical center where your specialist is every time a child has a cold. These are symptoms a PCP can handle especially if he/she has been kept up to date with the child's condition and appropriate protocols by the family and the specialist.

When the early symptoms or patterns of an infection (which you have seen before), be proactive and call your primary care physician or pediatrician right away. Have a plan – when will you go to have your child evaluated? Should you go directly to the ER? Approach illness as a team – call your primary at the first sign of illness and have a plan before the illness, fever, etc. gets worse. Go to the hospital when you feel you cannot manage at home - do not wait. If attempts to hydrate or nourish are failing, then the magnitude of the problem will only get worse. Do not wait until the patient is dehydrated.

Some infections evolve slowly. Caregivers commonly lose several nights sleep in a row and are exhausted. Before you, the caregiver, collapse also, get help! Consider talking to your doctor about admitting your child or family member for a day or two because there is no one at home to continue the needed care. This gives the parent/spouse a day or so to catch up. Parents need to care for themselves so that they can care for their child. Hospitalizing a child is not a sign of failure on the parent's part - it is a necessary part of good medicine and good care.

Another key variable in this discussion is communication and cooperation between the PCP and the Mito specialist. For example, if there is a predictable pattern that your child has followed with past infections, then the Mito specialist and PCP might develop/share a plan about what to do next time. Orders can be written, for example, to initiate an IV for fluids right off the bat at the first sign of a fever.

In general, when Dr. Korson is called about a patient who is "ill", he suggests that the parents go to the PCP first and have the child assessed. PCPs assess common illnesses all the time in their office. Then parents can ask the PCP/pediatrician to call the specialist while they are all there together in the office. Together a plan can be developed - a combined plan that includes the PCP, the specialist, and the parents/caregiver and child/patient. Everyone wins with this approach and the PCP is not out of the loop nor is he/she afraid of the complex disease. Based on experience, this is the ideal process to manage non-emergency illnesses.

Symptom Guide MitoAction offers a comprehensive online Mito symptom guide for the management of all kinds of symptoms, written by Dr. Korson and Margaret Klehm RN. Because this guide is based on specific signs and symptoms presented by patients, it is an easy guide for physicians not trained in the care of Mito patients. PCPs can also use this guide to look for the causes of symptoms. MitoAction also offers postcards with information about the online clinicians guide which are great reminders for patients to share with their doctors (send postcard requests to info@mitoaction.org).

Parents are reminded, however, if they are going to bring written information to their PCP or pediatrician, to only take a few pages - those related to the specific symptoms your child is exhibiting at this time. Otherwise they can be overwhelmed by pages and pages of information. The online guide is configured so that pages can be "printer-friendly" and printed one section at a time.

What kinds of illnesses can be safely managed at home?

The answer to this really depends on how the child/patient deals with or fights infections. You need to consider what you can do to intervene first. Assume that there is a pattern that you have seen before and that the onset is fairly gradual. Nourishment and hydration at home can often keep other symptoms in check. If they do not, then you may need to see the physician or go to the ER. Once vomiting and dehydration begin, it is difficult to manage at home. Further, if the GI system shuts down, you cannot do anything at home, unless you have a central line and can use all IV fluids.

If knowing limits of safe care at home is difficult, discuss the situation with your physician after 1 or 2 episodes of illness, and develop a plan in advance for what to do the next time these symptoms appear.

Steps to take if admitted to hospital

If the infection causes a metabolic decline or "Mito crash", a cascade of complications can occur and the patient is likely to need hospitalization for support. At this point an emergency protocol would be useful and helpful. Put together a short (2 pages) document which highlights your concerns, what has worked and what has not worked in the past. This can help advocate for you - the doctors won't have to recreate the wheel each time the child is admitted. Templates of emergency protocols can be found at <https://mitoaction.com/diagnosis-care/mito-sick-protocol/>.

Also, it is very helpful to ask your pediatrician or primary care physician call the emergency room or admitting floor in advance of your arrival. This prevents confusion in the ER when you arrive, and helps the admitting team to focus on the symptoms and care of the current infection rather than becoming sidetracked by the complexities inherent to mitochondrial disease. When you speak to the admitting physician, it is also helpful to bring short documentation which tells what the past pattern of infections and responses have been. Help to quickly educate the team: tell them what you expect to happen, what has worked in the past, and what should be avoided. Sometimes healthcare providers are unaccustomed to a parent/caregiver "calling the shots", but stay calm, respectful and act as a team player and advocate. For example, tell healthcare providers inexperienced with your child or family member, "This is what happens with my child...this is what to expect...this is what happened last time when..." in order to help direct the team to understand the unique cues and presentation of your/your child's mitochondrial disease. Collaboration with or availability of a phone consult with the Mito specialist is also very helpful. Remember to be patient; you may be asked to repeat things many times. Offer specific written summaries, 2-3 pages long; the house staff will appreciate this help. Do not be afraid to offer your insight.

As a last resort, if there are multiple services involved in the care and there is disagreement about the kinds of care needed, then you can INSIST on a family team meeting where a consistent plan must be agreed upon by all physicians and you, and this plan must be communicated to all involved.

How can we prevent infections?

Prevention is obviously key and we want to PREVENT illness as much as possible. Again, most Mito patients do not have a specific immunodeficiency (although some do), but rather infections, viruses, and illness all have such a high energy cost and prolonged recovery.

Dr. Korson recommends that vaccinations should be up to date - including seasonal flu vaccine as well as the pneumococcal vaccine. Good hand hygiene is essential both at home and in school. Some parents even provide the classroom with hand sanitizers to emphasize how important this is. Avoid unnecessary exposure. This might involve having the school (classroom teacher or nurse) notify you if illnesses are "going around." Depending on the situation, this allows parents to keep the Mito child home if needed. Dr. Korson also has a fairly low threshold for the use of antibiotics and may use them earlier and more often to prevent infections from getting worse. When the illness is caused by a virus then good nutrition, hydration and fever management (alternating Tylenol and Motrin) are even more important.

Other issues: Fever, fluids

It is essential to keep a fever down in all cases. High body temperatures make everything in the body run faster and exacerbate fatigue and all other symptoms. It is not unusual for Mito patients to have a lower baseline body temperature than 98.6. As a result, a fever, or elevated body temperature, depends on "normal" body temperature unique to that patient. Know what that is for your child and watch for other symptoms like clammy, flushed or pale skin, lethargy, increased breathing (panting, out of breath), or seeming "out of it...not right." Be sure to let hospital personnel and your physician know this. The numbers on the thermometer may not tell the whole story. Be aware of how you or your child reacts to fever. Some people have a dramatic decline with even a slight fever. When in doubt, call the doctor.

Fluid management is one of the cornerstones to supporting the Mito patient during an illness. The amount of fluids to give a Mito patient with an infection is going to be individual. Often, 1¼ or 1½ times maintenance fluids are recommended. If there is a question of maintaining nutrition, 5% dextrose can be added to a normal saline IV solution; if there is documented problem of a breakdown of fats, then a 10% solution of dextrose might be used, but again these are all dependent on the individual. The specific recommended protocols are also listed in the MitoAction physicians' guide.

Adult patients with Mito may also have trouble finding a primary care physician willing to be as involved as necessary. PCP's need to be the gateway between the patient, the hospital and the specialist and ideally are the first stop for the patient who is ill with a cold or flu-like symptoms. However, although primary care doctors can help manage minor illnesses, do basic labs, etc. they may not be equipped to handle all your issues. This continues to be an ongoing issue for many adult Mito patients in particular. Seek the support of patient advocate organizations or parent-to-parent networks, such as the Massachusetts-based Federation for Children with Special Needs.

Have a plan in advance about what to do when and if you, your family member or your child gets sick. Ideally, when the Mito patient develops a fever, you can call your physician with an already established threshold and plan in place. Don't wait for a crisis: be proactive!

Submitted by, Joanne Turco, RN, MS & Cristy Balcells RN MSN