
Suggestions for Caring for a Child With MLD

1. Nutrition

Nutrition is very important for a child with MLD. If a child is properly nourished, he or she will be in a much better position to combat viruses, bacteria and infections. In our experience, the best way to feed a child who has MLD is through a gastrointestinal tube, commonly called a G-tube. This is a tube that is inserted by a surgeon directly into the child's stomach, allowing the child to receive nourishment without having to swallow. This is very important for children with MLD, because as their muscles weaken, it becomes harder for them to swallow, and they run the risk of inhaling food into their lungs, or aspirating. This can frequently result in pneumonia.

Parents can sometimes have very strong feelings about whether to have a G-tube placed in their child's stomach. Sometimes parents are reluctant to do it because they feel that it will be a sign that they are giving up on the child. However, G-tubes can be easily removed, and the hole can close in a matter of hours. They can also be used for supplemental feeding, such as when a child is sick, or for giving medicine. We have found that it is generally better to have a G-tube placed earlier, when the child is healthy, rather than waiting until they are undernourished.

To determine your child's nutritional needs, you need to consult your child's doctor or a nutritionist. The nutritionist will determine what the child's proper weight and height should be for his/her age. If the child is fed through a G-tube, the nutritionist will most likely recommend a formula that is specifically made for G-tubes (sometimes called an "enteral formula"). These formulas usually have all of the vitamins and nutrition that a child requires and ensure that the child is not ingesting "empty" or useless categories. *Please see the separate pediatric G-tube formulas document for a listing of various enteral formulas.* Keep in mind that a child can react differently to different formulas, so if one formula doesn't work, try another.

2. Pain & Symptom Management

Medications

Children with MLD often have difficulty with swallowing, seizures, tight muscles, nerve spasms, or reflux. All of these conditions can be addressed with medication, and you should work with your child's doctor to develop a medication regimen for your child. Keep in mind that medications that work well for a period of time may need to be adjusted after awhile, and we work with our doctors regularly to adjust medication levels depending upon the boys' needs. Below is a listing of the medications our boys take:

- **Reglan** helps the stomach to empty and can help stop reflux. Since children with MLD cannot move around, it takes longer for their stomachs to empty. If their stomachs are not working efficiently, this can lead to reflux and vomiting. If your

child has issues with this, Reglan may help. Another option to consider is adjusting the child's g-tube formula, feeding continuously or using a pre-digested formula such as Peptamen Junior PreBio.

- **Prevacid** also helps with reflux and reduces the amount of acid in the stomach, making the stomach less irritable.
- **Baclofen** is a muscle relaxant and is commonly used. Some children have baclofen pumps surgically inserted in order to help with pain management. However, children can develop a tolerance to Baclofen and higher doses may not be effective. Consider using Dantrium (see below) in conjunction with Baclofen in order to avoid this.
- **Dantrium** is similar to Baclofen in that it is a muscle relaxant, but it can also have the effect of enhancing the effectiveness of the Baclofen.
- **Inderol** is generally used for high blood pressure or headaches. It was prescribed to our boys by our neurologist, who asked if the boys would tense up, arch their backs and get very upset at times. We were amazed that he knew this! The doctor said that the hypothalamus portion of the brain, which regulates the flow of hormones, is often damaged in neurologically impaired children. The hypothalamus can trigger a release of hormones/chemicals into the body causing the child to experience extreme emotions for no apparent reason. Inderol helps to control this release of hormones.
- **Robinul** is used to control drooling. If children have a hard time swallowing, saliva can accumulate in their mouths, causing them to gag or drool. Robinul reduces the amount of saliva that is produced but a side effect can be dry mouth.
- **Klonopin or Clonazepam** is an anti-seizure medication that our boys receive in small amounts three times per day.
- **Elavil** is a drug that helps to control random neurological impulses. We found that our boys were experiencing extreme sensitivity in their hands and feet and would quickly pull away whenever they were touched in those areas. Elavil helps to manage those impulses so they are not so startling.
- **Motrin and Tylenol** are used to manage pain and muscle tightness. We alternate using Motrin and Tylenol since Motrin can be hard on the stomach.
- **Pulmicort** is a steroid that is inhaled into the lungs through the use of a nebulizer and helps to reduce any inflammation. As MLD children cannot move and run around, they are susceptible to pneumonia and breathing issues. Pulmicort can reduce any inflammation. Our boys use Pulmicort once a day when they are healthy and 3-4 times per day when they are sick.
- **Miralax** is a laxative that helps maintain regular bowel movements. It is now available over-the-counter and does not require a prescription. Miralax does not have to be given every day (it may result in excessive bowel movements!), so you may want to experiment with the amount given and how often you give it. For instance, our boys used to get ½ capful every other day. Please make sure to give your child plenty of water during a 24-hour period while they are on Miralax, or it can actually gel together and block the colon even further.

- **Tylenol 3 or Tylenol with Codeine** can be used for muscle relaxation and pain relief. It is best to give this in the evening or other times when the child does not need to be alert.
- If your child's muscles are very tight, **Botox** therapy can be very helpful. Botox is now commonly used for cosmetic procedures, but has been used for cerebral palsy patients for many years. Christopher's arms used to be very tight and clenched. His doctor gave him Botox injections in his arms, and coupled with the short-term use of arm braces, he no longer has the tightness in his arms that he used to have. A key point to note is that it is very important to go to a doctor who is skilled and experienced in administering Botox since the location of the injection and the amount given can make a difference in the success of the treatment.

Some of these medications can be made into a liquid form while others are available in pills. If only pills are available, we grind them up into a powder using a mortar and pestle (available at cooking stores). We then mix the powder with other liquid medication or water and administer it with a syringe through the G-tube. However, different drugs may require specific handling, so please consult your physician for guidance on how to administer each medication.

3. Durable Medical Equipment

A wide variety of medical equipment can be found through Abilitations (see www.abilitations.com or call to request a catalog). Some of this equipment can also be provided by a home health care service or hospice. E-bay can also be a good source of used adaptive equipment.

Respiratory

- A **suction machine** is an essential piece of equipment. Since MLD children have difficulty swallowing or can be more prone to vomiting or aspiration, a suction machine is used to suction mucous from the nose and mouth. If a child is sick with a cold or pneumonia, deep suctioning can be used to remove mucous deep in the throat. Additionally, it is helpful to use when brushing teeth. We have several suction machines throughout the house so we can quickly grab one if a child begins to gag, vomit, or needs to have mucous removed from an airway. Battery-operated suction machines are also available.
- A **nebulizer machine** allows a child to inhale medicine such as Pulmicort (a steroid for the lungs) or Albuterol (helps open up the passageways in the lungs). These medications are necessary because the lungs often get weak from not expanding as much as a child who can move, run, or speak. Both of these medications are commonly used by children with asthma or cystic fibrosis.
- A **bi-pap machine** is used to provide extra air when the child breathes in to help the lungs fully inflate. We noticed a significant improvement in lung function with both of our boys when they began wearing their bi-pap machines at night. This machine is similar to a C-pap machine which is used by people who have sleep apnea or problems snoring. With a bi-pap



machine, the child wears a mask that covers his nose. Bi-pap machines also help prevent pneumonia by keeping the lung airways open. These machines are relatively small and can be rented.



- The **Airway Clearance Vest** is a vest that hooks up to a machine with vacuum hoses and vibrates against the child's chest. It provides intense respiratory therapy within a short period of time and works by loosening the mucous from a child's lungs. The vest is typically used for about 20-60 minutes each day. It is very expensive (approximately \$16,000) and can only be obtained with a prescription so you cannot find used units available for sale on sites such as e-Bay. However, Hill-Rom, the manufacturer, will allow a 30-day trial and will also work extensively with insurance companies to try to get insurance coverage for the vest.
- Another piece of equipment that we use extensively is a **pulse oximeter**. This is placed on the child's finger and measures the amount of oxygen in the blood and the heart rate. We have found this very useful in determining if the boys are getting sick, since they usually have elevated heart rates and lowered oxygen levels when that occurs. These can also be expensive, and good machines can run \$320+. You can talk to your doctor about which model may be best for you, although they can be found on [eBay](#) or [Amazon](#). For an example see http://www.amazon.com/Fingertip-Oximeter-Nonin-Onyx-9500/dp/B000MOYHWQ/ref=sr_1_20?ie=UTF8&s=hpc&qid=1205517343&sr=1-20.

Movement and Mobility

- A good **wheelchair** is essential. Since most children with MLD have limited use of their limbs, a manual wheelchair is usually the best choice. Our boys have a [Convaid Safari Tilt](#), which can be reclined to 45 degrees. The wheelchair also folds up easily (like a stroller) so it can fit in a trunk or the back of a van. Additional accessories can be purchased for the wheelchair, such as a padded seat, a padded backrest, additional headrests and back extensions as well as a sun shade. If you plan to transport your child in a wheelchair in your car or in a school bus, be sure to get the crash-tested transit model so that the child can be safely secured to the car or bus.



- **Ankle foot orthoses (AFOs)** are braces that are worn on the feet and are good for ankle support when standing in a stander as well as for keeping the heels stretched. Children with MLD seem to get very tight heels, and often have to have their heel cords surgically cut. AFO's prevent this from happening and allow a child to wear shoes and have his or her feet look "normal".
- A **stander** is basically a board that a child is strapped to which can then be moved into a vertical position so the child is "standing". A stander helps with hip and bone development by having the bones bear weight and can also serve to provide a good stretch for the muscles. We have a Rifton stander, and we really like it because only one adult is needed to put a child in the stander. Some standers are very complicated and require two adults. You can see a picture of the Rifton stander at this website: <http://www.rifton.com/products/standing/supinestanders/index.html>
- If you transport your child in the car and the child is not in a wheelchair, a **car seat or booster seat** will be necessary. Our boys weigh approximately 50 lbs. and in our vehicle, we use a Graco booster seat that can be reclined. We also slightly recline the seats in the car that the boosters sit on. This prevents the boys' heads from "flopping" forward and keeps them more relaxed.

However, if your child needs more support, several companies make special needs carseats, and Adaptive Mall (<http://www.adaptivemall.com>) has a good selection. These seats can be expensive and often cost \$600 or more.

- If you have access to a pool or body of water, **swimming** can be wonderful. In water, the child's body seems much lighter and movement is easier, not to mention it can be very relaxing. Our boys use a neck collar and a kick board with special straps while in the pool, which allows them to easily float and be more stable. These items can be purchased through www.abilitations.com.

- **Neck Collar**



- **Kick Board with straps**



Body Positioning

- We have found that our boys respond very well to vibration. When they are laying down on their bean bag, we have them lay on top of a **full length vibrating massage pad** (made by Homedics and cost about \$40). See [Amazon.com](http://www.amazon.com) or click this link to purchase:
<http://www.weatherconnection.com/product.asp?itmky=158780&model=MMSQ-100&cat=5&subcat=158718&sortby=&filtermfq=&page=>
- We also use a **small vibrating massage pad** on the boys' beds. We wrap the massage pad in a cotton pad (see purchase information in *Miscellaneous* section) and then put the pad under their legs. The pad has an auto shut-off after one hour so we do not need to worry about having the boys get "hot spots" while they sleep. We use the Heat Plus Massager Heating Pad by Sunbeam but do not use the heat function. See http://www.amazon.com/Sunbeam-Health-Home-Massage-Heating/dp/B00075M1VO/ref=sr_1_10?ie=UTF8&s=hpc&qid=1205516787&sr=8-10 for purchasing information.
- During the day, we often have the boys sit in a [Love Sac](#). This is essentially a **very large bean bag**, but instead of being filled with beads, it is filled with memory foam pieces. The Love Sac comes in many sizes and the covers are removable and can be washed in the washing machine. We have a Super Sac which is very large, but can easily accommodate both boys comfortably as well as an adult in between. We often lay the long vibrating massage pads on top of the Love Sac to help with comfort. The Love Sacs can be expensive, so you may want to look for a store near you or check on eBay.
- Use a **Memory Foam Mattress Topper** to minimize bed sores or "hot spots" from the massage pad on the skin. We have found that John and Christopher do not move much when they are sleeping and have a tendency to get hot spots on their skin, but the memory foam mattress topper has completely stopped that.



- Using risers to slightly elevate the head of the child's bed can help with breathing and swallowing. See: <http://www.lnt.com/product/index.jsp?productId=2256040&cp&kw=riser&origkw=riser&sr=1> . These cost about \$9.99 per set.



Bathing & Hygiene

- A **bath chair** allows the child to remain stable while he is getting a bath or shower, plus it saves your back! We have the Leckey bath chair similar to the one pictured below. You can also get an additional platform that fits under the bath chair and raises it to approximately 3-1/2 feet high. See <http://www.adaptivemall.com/lecbatchair.html>



- Even though your child may not eat by mouth, caring for your child's teeth and mouth are very important and bacteria can easily build up in the mouth. Swab your child's mouth twice per day using a **disposable toothette** (see http://www.amazon.com/Sage-Toothette-Disposable-Oral-Swab/dp/B0000DK4KH/ref=pd_bbs_sr_2?ie=UTF8&s=hpc&qid=1205528248&sr=8-2) that has been dipped in an anti-bacterial solution such as Crest Pro-Health Rinse, Biotene, or Chlor-Hexadine. If your child has difficulty swallowing, place your child on his side, with the chin tucked in to help avoid aspiration and to encourage him to spit out the anti-bacterial solution. You may also want to use a suction machine while you are doing this. Bacteria may also build up on the tongue and cause bad breath, if this is the case, use a **tongue brush** (we prefer

the Tung Brush see <http://www.tungbrush.com/>) to scrape the tongue and remove bacteria.

Miscellaneous

- Use **rubber lined cotton pads** (see www.discountpads.com) and put them on the child's bed or any surface he is sitting or laying on to save on laundry. These are good for catching leaks from diapers, spit ups, etc. Our boys lay on these and we put them over the vibrating pads to prevent them from getting dirty and to protect them from getting sores from laying on a vibrating motor too long.

4. Medical Team & Insurance Management

Insurance

In addition to private insurance, some other insurance options may be available:

1. A **pediatric hospice** service can provide medication, tube feeding formula, or other services. Depending on your state or insurance company, the insurance company may pay the hospice a flat amount per month for these services. To qualify for hospice, generally the insurance company requires that a doctor write a letter stating that the child may not have a life expectancy of more than six months. While that may sound pessimistic, children with MLD are often hospitalized and have many medical issues so it can be reasonable to assume that their life expectancy may be short. Most insurance companies do not penalize you or cancel the services if the child uses hospice services for more than six months.
2. Within the United States, most hospitals have **pediatric social workers**. These social workers are trained in knowing what programs are offered by the state, federal government, or local programs. Social workers can help to identify programs that your child or family may be qualified for.
3. A nonprofit group called **SNAP (Special Needs Advocates for Parents)** can work with parents to confront medical insurance companies and help families of special needs children get the most from their benefits. You can read more about them at <http://www.snapinfo.org/ourservices.html>.

Managing your child's medical care can easily become a full-time job, but it can be easier if you choose a primary care physician who has the ability to be the "leader" of your child's medical team. The coordination effort helps to make sure that everyone is informed and that the doctor advocates for the proper care both with other doctors and the insurance companies. Members of your team may include:

- A **primary or family care doctor** who is willing to work with you and your family is essential. Because of the medical issues associated with MLD, it is vital to have a doctor who you have a good relationship with and who is familiar with your child and willing to advocate for him or her. You may need to talk to doctors on the phone and interview them to make sure that they are willing to work with your child.

- A **physiatrist** is a doctor of physical medicine; this person often serves as the “general contractor” for all of the therapy or care that a disabled person receives. However, these doctors are often very busy and carry a high patient load. While a physiatrist is good for discussing various therapies, it is critical to have a strong primary care doctor who can deal with colds, hospitalizations, etc.
- Since MLD primarily affects the nervous system, a **neurologist** may be involved with your child. Often they do not provide treatment, but may be involved in the initial diagnosis or may be consulted if additional nervous system issues are noted.
- **Therapists** who specialize in physical therapy, occupational therapy (fine motor skills) and speech therapy may work with your child, depending upon his or her abilities. These therapists can be consulted to help with proper positioning, determining methods of communication, or other ways of helping your child. Keep in mind that extensive therapy is beneficial for restoring function, but since MLD is a degenerative condition, it may not be possible to restore function (unless a child has undergone treatment such as stem cell transplant) due to damage to the brain or nervous system. However, consulting with therapists can be beneficial to get ideas on how to help your child improve his or her quality of life.
- Even though your child may be tube fed, visiting a **dentist** is very important so that no mouth sores or severe dental problems develop. Additionally, if your child is young, a dentist can help to make sure that the baby teeth fall out (or are removed) to avoid impacted teeth or other serious problems.
- An **ophthalmologist** (eye doctor) or **neuro-ophthalmologist** (an eye doctor who is also familiar with how the nervous system works) can help to determine your child’s visual capabilities. Because eyes often degrade with MLD children, these doctors can help with glasses or other treatment as needed.
- A **pulmonologist** is a doctor of the lungs and respiratory system. Because children with MLD are immobile and cannot run around and expand their lungs like other children, they are often susceptible to pneumonia and lung problems. A pulmonologist can assess your child’s lung health and make recommendations for maintaining or improving lung function.
- A **gastroenterologist** specializes in stomach and digestive issues. This doctor may be involved in placing a G-tube or other type of feeding tube and can be consulted for vomiting or reflux concerns.

5. Education

Educational services are available through the school system, although services can vary greatly among school districts. Within the United States, if your child is younger than three years old, therapy services can generally be provided by an early intervention program offered through your county or state. Once your child turns three, generally the school district will begin providing these services. Your school district will work with you to complete an Individual Education Plan (IEP), which will contain goals that the school will work with your child to achieve. Depending upon your child’s capabilities, your child may also be assigned an aide to help him or her throughout the day. For our boys, we have found school to be beneficial, especially from a social perspective. Currently, John

and Christopher are in separate kindergarten classes, and their classmates enjoy being with them and helping John and Christopher is a coveted “job” in the classroom. The boys typically attend school with a nurse, who rides the bus with them and also helps them with their classroom activities.

Another alternative that we have used through the school district is **homebound education services**. Because our boys seem to get sick very easily and colds can quickly turn into pneumonia, the school district provided services to them at home for a period of time. Although it was a new concept for our school district, we worked out a plan during the boys’ IEP (individual education plan) to make this happen. The boys received about 45 minutes of education and therapy from the school four days per week.

Other services that may be available through the school district include access to someone who is an expert in the areas of **adaptive equipment and assistive technology**. We had an individual from the school district evaluate our boys to determine which equipment would be best to enhance their communication and education. These services can also be available through a rehabilitation hospital or Easter Seals.

6. Other Resources

- **Exceptional Parent magazine** is for parents of special needs children and is very helpful in highlighting and discussing resources that are available. See <http://www.eparent.com> for subscription information and online resources.