

Daily Living

with MPS and Related Diseases



A joint effort between the MPS societies of Canada and the United States



*The Canadian Society
for Mucopolysaccharide
& Related Diseases Inc.*



Pictured on the cover: Maddy (MPS I), Matthew (MPS III B), Andre and Ashton (ML II)

Introduction

This comprehensive booklet represents the first collaboration between the MPS societies of Canada and the United States. Just as MPS and related diseases know no boundaries, neither does the wealth of information provided in this booklet. Our goal is for this to be a booklet written by you, the parents of children with MPS. We recognize that there is so much information and support provided by parents for parents, above and beyond the medical, and we want it available for all to share. Thank you to all the parents who sent ideas and suggestions. Because of you, parents of children with MPS have additional resources, those never-before written and helpful daily living guidelines.

We recognize that not everyone has need for all the information contained in this booklet, either now or perhaps ever. This is a reference booklet with sections to be read and referred to when needed.

The booklet is organized in sections beginning with that initial period after diagnosis, “Getting Information,” then how to talk to people important in your life – your other children, relatives and friends. “Asking for Help” is difficult for most people and more so if you have a child with special needs because you don’t know if you can repay the generosity of others. People appreciate being asked for help, as they often don’t know what to do but want to offer their support. The remainder of the booklet focuses on behavior, feeding, toileting and sleeping, and ends with home modification ideas.

We hope you find the information in this booklet to be thought-provoking and useful. We would appreciate receiving any feedback, including additional suggestions for potential future editions. You should evaluate the suggestions in this booklet in the context of your individual situation and consult appropriate professionals before implementing them. The Societies and those responsible for submitting ideas for this booklet cannot be held responsible for any negative consequences resulting from your implementation of these ideas.

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Table of Contents

Getting and Recording Information	4
General	4
Phone Calls	5
Emotional Responses of Siblings	7
Fear and Anxiety	7
Anger and Justice	7
Jealousy	7
Embarrassment, Guilt and Shame	8
Loss and Sadness	8
Loneliness and Isolation	8
Emotional Intelligence	8
Talking to Family and Friends	9
Sharing Information About Your Child	10
Dealing with Strangers and Inquisitive Children	11
Strategies for Dealing with Strangers	11
Teasing	12
Asking for Help	13
Should You Ask for Help?	13
Government and Charitable Organizations	13
Family and Friends	13
How Others Can Help	14
Behavior	15
Understanding the Before and Why Behaviors Occur	15
Strategies for Dealing with Difficult Behaviors in the Home	15
The Unexplainable	16
Behavior without Development Delay	16
Behavior with Development Delay	16
Socialization	17
Communication	17
The Classroom	17
Respite Care	17
Networking	17
Medication	17
Wander Prone Children	18
Gifts and Toys	19
Chew Toys	19
Other Toys and Other Activities	20
Gift Suggestions	20
Adapting Toys and Games for Your MPS Child	21
Feeding and Swallowing Difficulties	22
Stages of Progression in MPS Diseases	22
The Role of a Speech and Language Therapist	22
Videofluoroscopy	23
Cervical Auscultation	23
Management	23
High-Risk Foods	23

Safe Eating Techniques	24
Fluids	24
Ways to Control the Swallowing Rate.....	24
Stages of Safe Eating Techniques	25
Meal Ideas and Recipes	25
G-Tube Feeding – Pros & Cons	25
Elimination.....	25
Constipation, Diarrhea, Incontinence	26
Constipation	26
Diet Changes and Natural Remedies	26
Medication	26
Diarrhea.....	26
Medication	27
Incontinence.....	27
Diaper Rash.....	27
Diapers in General	27
Lifting	28
Sleeping	29
Troubleshooting.....	29
Sleep Apnea/Airway Obstruction	29
Medication.....	29
Sleeping Safely – Modifications to the Bedroom	30
Dealing with Insomnia in General	30
Home Modifications	31
Outside the Home	31
Accessibility Possibilities	31
General Home Safety Ideas	31
Bathroom.....	32
Kitchen	33
Bedroom	33
Living Area	34
Safe Room Or Sensory Room	34
Travel	35
What to Pack	35
Airplane Travel	35
Car Travel	36
Resources	37

Getting and Keeping Information

GENERAL

Dealing with your child's diagnosis will most likely be the hardest task you will encounter during your lifetime. Many parents learn that obtaining, deciphering, and monitoring information is their best weapon to combat the powerless feeling that comes with their child's diagnosis. As a result, you will have to learn how to find and manage information like you never have done so before. This information will be useful in alleviating your own fears about the future and adequately managing the care of your child. You will often be required to know what needs to be done for your child and to inform healthcare providers of your child's medical history. Parents have shared the following tips on how they obtain and manage their child's medical information:

- There are no stupid questions. If you don't know something, ask.
- You can and should request information. Educate yourself on how to request access to information and records and what your rights are. Sign forms to have medical information released to you regularly from your doctors.
- The MPS societies have a collection of journal articles available to you. There are many links to MPS-related topics on MPS society Web sites.
- Don't isolate yourself. Find a support group and join it. Subscribe to newsletters relevant to your child's condition. If you have a computer, search the Internet for support groups and associations such as MPS Societies. If you don't have a computer, go to your local library and have the librarian help you find this information.
- Ask your health care professionals for information resources. Seek second opinions on medications and surgeries, but keep in mind that there are very few doctors who are experts in the MPS field.
- It may not be possible to collect all the medical information you need before you meet with someone or have an appointment. Have faith that you will pick up the knowledge and skills with time and experience.
- Don't keep things in your head. Write a summary about your child's medical and developmental history and keep it current. This will be helpful when answering questions during appointments, making phone calls or choosing services.
- Request copies of clinic notes after you've seen the doctor. Ask your doctor for a letter after each visit, summarizing what was discussed. Keep these records in a notebook for easy access.
- Ensure any verbal information you've received is accurate by taking notes, then summarizing them to the information provider (i.e., your doctor) at the end of a meeting or phone call. It may help to have a friend/support person with you, asking for explanations or clarification.
- Involve your child in asking for information and organizing the information you have in a way that works for you.

PHONE CALLS

- Although information from phone calls may be informal and impersonal, whenever possible keep track of the information you receive. Mark the call on your calendar or day book.
- First, ask yourself if a phone call is the best way to raise certain issues or questions. If you feel you have a lot of information to give or need to communicate in detail about personal issues, it may be more appropriate to request a meeting or an appointment.
- Make a list of your questions and concerns before you call. This will ensure that you don't hang up realizing you forgot to ask the most important question. It also will help you keep the phone call on track.
- Keep a notebook or record sheet ready for notes and additional questions that arise from the call.
- If phone calls make you nervous, practice the call with a friend to make sure what you're asking is clear.
- Devise various ways of asking the same question if you don't feel you are getting the information you need.
- Make your calls during the time of day you are at your best, and when you are least distracted. Try to make calls when your children are at school or daycare, or are sleeping. Don't start a difficult task when you are tired or rushed.
- Always record the name of the person with whom you are speaking. Note the date and time of your conversation.
- Use open-ended questions that encourage the other person to provide you with useful information. Avoid questions that only need a 'yes' or 'no' response. For example, instead of asking your doctor if your child's insomnia will improve, ask how you or your doctor might be able to improve your child's insomnia. Skip the first question (will it improve)...and go right into how. Ask questions that begin with Who, What, When, Where, Why and How.
- Instead of asking "Will you be able to find my child an ear, nose and throat specialist," ask,



Damien (MPS IV) and Natasha (MPS IV) and family

"What can an ear, nose and throat specialist do for my child? Who can you refer us to? When can you do this?" and so on.

- Record notes during or after the phone call. If new questions arise from the call, note them so you have the list handy for the next call.
- Summarize the main points of the call. Make sure you have the information you need, and clarify any points that are not entirely understandable. Note any items that require action and follow-up.
- Be prepared to leave a voice message. Specify the times when it would be convenient for the person to call you back.
- You may want to send a letter confirming your conversation after the phone call. Include any follow-up plans: Who is doing what? Are there any deadlines involved? Has any agreement been made on a time and date for follow-up? This also applies to schooling and daycare issues as well as medical appointments.
- After the call, ask yourself: Were all of my questions answered? Is there someone else I need to speak with? Is there something else I need to do to get additional information?

Getting and Keeping Information

- When you must leave a message on voice mail, be clear and specific with your message, indicating:
 - Who you are
 - When you are calling
 - Why you are calling
 - Your phone number(s)
(remember to read your number clearly and slowly and repeat the number)
 - When they can call you back
- Some parents have found the call record sheet below to be useful for documenting phone calls.

PHONE CALL RECORD SHEET		
Date / Time / Who Called	Person / Organization / Position	Phone / Fax / E-mail / Address
April 15, 2001 3 p.m. <input checked="" type="checkbox"/> I called. <input type="checkbox"/> I was called.	Dr. Smith, Ophthalmologist Honeybrook Clinic	416-555-1212 (phone) 133 Westmount St. Suite 208
Notes: Time for Julia's annual exam. I left a message with the receptionist.		
Follow-up:		
Date / Time	Person / Organization / Position	Phone / Fax / E-mail
Notes:		
Follow-up:		

APPOINTMENT LOG													
Doctor	Phone	Jan	Feb	Mar	Apr	May	Jun	Jul	Aug	Sep	Oct	Nov	Dec
Smith	875-1111			*8th-2:15			*						
Jones	875-2222					*							
Clark	875-3333	*11th-3:00						*					

Insert * in month you are to return for next appointment. Insert appointment date and time once appointment is made.

Emotional Responses of Siblings[†]

Other than yourself as a parent and your child affected with an MPS condition, your child's diagnosis will have the greatest impact on his siblings. It is important to not minimize or underestimate that impact. Many siblings share common feelings. Though these feelings are not evident in every child and do not remain the same over time, parents who are able to recognize these feelings and responses can help their children develop constructive ways to cope with them.

FEAR AND ANXIETY

Preschoolers may wonder "Did I make this happen?" Repeatedly reassure your preschooler that their sibling's health condition is not their fault.

School-aged siblings are more aware of differences between people and responses of others. That awareness can be harnessed to increase their knowledge of MPS that can be transferred to others. Children will often take pride in knowing something and sharing information with others. Devise scenarios at home to help your children practice responses to strangers, neighbors, friends and even other family members. The confidence they develop in their increased knowledge of their sibling's condition will greatly reduce their anxiety.

Adolescents are often more concerned about fitting in with peers and may feel embarrassment if their family is perceived to be different. Teens should be encouraged to develop their independence and should learn that their identity is not wrapped up in the validation of their friends. Educating their friends and making them familiar with their sibling is a good way for adolescents to minimize their anxiety.

If your unaffected children have fear and anxiety because of a real or perceived threat to their safety (due to aggressive or unpredictable behavior by your MPS child), you should address these fears head on by taking action to ensure their safety and developing coping strategies to address perceived threats.

ANGER AND JUSTICE

Sibling anger is usually generated from the loss of parental attention. Sometimes, it will be expressed outwardly in behavior, rather than verbally.

Though feeling angry, some siblings will try to protect parents from their negative feelings. However, these children will eventually have to find a release for all this repressed emotion. Sometimes this will express itself through inappropriate responses to a situation, such as over-reacting to a seemingly minor incident.

Younger siblings may state that the division of chores is unfair or unjust, whereas older siblings might state that it isn't fair that they don't have a sibling with whom they can relate.

Parents should validate these feelings of anger and frustration so they don't get "bottled up." Siblings can be assisted in developing insight and constructive ways of coping with their situation. Explaining and demonstrating that anger is an acceptable emotional response that can be channeled into healthy expressions will help siblings and parents.

JEALOUSY

Healthy siblings might feel less special after a diagnosis is made because their parents are spending more time with their MPS sibling. For these children, spending additional short amounts of time or having some special "ritual" with either or both parents may help restore their own sense of "specialness." At particularly stressful times, increased attention from a grandparent, aunt, uncle or family friend also can help.

A teen also may experience jealousy of their peers whose lives seem less complicated. With open communication and validation of their feelings, a teen's reaction of jealousy can decrease over time.

EMBARRASSMENT, GUILT AND SHAME

By middle childhood, siblings become aware of other people's responses to their brother or sister, and young children may experience embarrassment for the first time. Young children may experience a conflict between a will to protect their sibling and the need to be accepted by peers.

By adolescence, siblings have often developed empathy for their affected brother or sister, but may also have developed guilt about their own good health and opportunities in life.

Listening to your children and helping them to understand they are not bad people for feeling angry or embarrassed can help offset feelings of guilt and shame.

“Every time I thought about the stress I was experiencing, and complained to myself, I immediately felt incredibly guilty. How could I complain when ‘Maddy’ was going through so much? Guilt became part of my life...”

(Fleitas, 2004)^{† †}

LOSS AND SADNESS

For very young children, loss will most likely result from sudden changes in family life. This will probably be expressed in behavior or reactions to situations that may be quite inconsistent with their emotions. For example, the loss of a favorite toy may suddenly become the focal point for frustration and sadness. So what may appear to the parent to be an easily resolved problem (there it is, under the couch) can evolve into a catastrophe for the child with bottled-up sadness who has finally found a tangible way of expressing it.

When children can empathize, they become aware of suffering of parents and siblings; but until a child is able to understand the sibling's chronic health condition, they also may be worried about

developing their sibling's condition. When parents explain more about MPS and encourage children to talk and describe their feelings, they tend to cope better with the losses in their life. Siblings may cope by continuing to ask questions and seeking reassurance.

When small achievements are celebrated, the sibling relationship will be linked to positive experiences which can offset the sense of loss or grief.

LONELINESS AND ISOLATION

Just as parents often benefit from sharing their experiences with other parents, so too do siblings. Other siblings may be people who children can express emotions to.

“It was eerie, actually. The doctor looked through me as if I was a plant or something. He just talked to my mom. There I was, scared out of my wits...my sister asleep in bed with a high fever, and all those tubes and equipment...I felt angry and confused...and pretty lonely, too. She's my best friend.”

(Fleitas, 2004)^{† †}

EMOTIONAL INTELLIGENCE

Siblings may learn to gain praise and validation by being helpful and compliant. By middle-childhood, siblings may be contributing significantly by helping parents with daily chores or with the care of their MPS sibling. This attunement means that siblings develop skills and abilities that enrich their lives and the lives of those around them in advance of their friends. Yet it is important that siblings do not feel the need to compensate for family difficulties all the time.

In all, understanding the emotional responses of siblings requires patience and understanding on the part of the parents. As children go through various stages of emotional development, parents need to not only take cues from their child, but guide their child in their emotional development. Open communication is the key in helping siblings of children with MPS cope with their situation.

Talking to Family and Friends

Volunteer information...they may not ask.

There is no right or wrong way to talk to family and friends about your child with MPS. You will find that some family members and friends are very receptive, whereas others will be in denial. Some will be supportive and will want to help wherever possible, while others will be sympathetic, yet not comfortable being with your child. Some will never accept your child's diagnosis. You should prepare yourself to encounter all of these situations and to keep an open mind. You may be surprised by what you find. Some people who take more time than others to come to terms with your difficult situation can end up being your best advocates.

Sometimes a family member's silence does not necessarily indicate indifference. Many people want to help, but are concerned they will offend you by offering. Some family or friends may, in fact, wish to know more about MPS, but are worried that they may come across as prying. Thus, it is important for you to set the tone. If you are comfortable answering questions about your child's condition, let people know. Keep some literature and the MPS Society's Web site address handy. Remind people that you are happy to answer their questions.

If there are times you do not want to talk, or if there are certain issues you do not wish to discuss regarding your child's illness, let your family and friends know that as well.

Some suggestions from parents for talking to family and friends are:

- Talk about your child's condition in a normal, everyday manner. Be open. Discourage pity. Emphasize the positive things, such as your child's easy disposition, good health, sense of humor and loving nature.
- Don't expect family to automatically learn about MPS and help you deal with it. They also must grieve. In some ways, this can be harder for them since they are not around your child as much as you are. Let them grieve and understand it may take a long time. Don't give up on them as this is a gradual process. Provide them with information and access to the Society's Web site so they can work through it on their own terms.
- Include your MPS child in all family activities that are safe to do so. Be frank and speak openly about your expectations.
- Send copies of the Society's newsletter to your family and invite them to MPS Society activities.
- Ask family members to join the Society.
- Tell your local family members about your diagnosis in person. For those who are out of town, call them and talk. Keep everyone updated through e-mail or holiday letters.
- Tell family and friends how they can help. (See Asking for Help, page 13.)

Sharing Information

About Your Child

Effectively sharing information about your child with MPS may be challenging. Parents have made the following suggestions to assist you in this process:

- Don't keep information to yourself. Let others know what your child's needs and capabilities are.
- There's no rush. Share with others as you feel comfortable, in a way that feels right for you. This may mean phone calls, handwritten notes or a brochure.
- It is okay to be emotional. Do not apologize for being upset.
- Know and respect that everyone has a different reaction to the same situation. Some people readily accept information while others may take some time to digest it.
- Talk to friends, family, other parents and health care professionals about ways to share information. Make sure to speak with several people. You will find that each person will give a different perspective and offer a variety of suggestions.
- Involve your child as much as possible. Teach her how to communicate information about herself.
- Emphasize who your child is rather than what is "wrong" with him. Your child is a person who has MPS; he is not defined by the disease. Focus on the person and not on the disability. For example:
 - Let people know what your child's interests are and are not.
 - Explain your child's strengths, as well as areas where your child will need assistance – and just how much assistance is needed.
 - Discuss how to effectively communicate with your child. Remember to let people know if your child has any hearing, language or visual impairment, as well as the extent of such impairment.
 - Write up key information about your child and her care and keep it handy for caregivers, teachers or anyone who spends time with your child.
 - If possible, share information through photos or videotapes.
 - Create a personal profile of your child, including photos, a list of favorite activities, food preferences, dislikes or anything else you think is appropriate and pertains to your child. Think of it as an instruction manual for your child.

Person-Focused Description

Suzanne is an energetic and happy child who has Sanfilippo. She enjoys videos and chew toys. She is easily distracted and prone to wandering, so she requires careful supervision. Suzanne loves to eat, but needs some assistance with feeding.

Disability-Focused Description

Suzanne is a Sanfilippo child. She is extremely hyperactive. She requires constant supervision.

Dealing with Strangers and Inquisitive Children

The more people are aware and knowledgeable, the better the world will be.

The answer to the question: “How to deal with staring strangers or inquisitive children?” is best answered with the very simple maxim: “Do what you’re most comfortable with.”

Some MPS families have sent in suggestions, ranging from the subdued: “Ignore them,” to downright funny: “Tell them more than they will ever want to know.” Overall, it’s your call. Just because a stranger is staring at your child or offering unsolicited advice, does NOT mean you have to apologize, reply or even acknowledge what may very well be an intrusion.

For some families, questions and stares are opportunities to spread the word about MPS and special needs children in general. For others, it is just one more straw for their already burdened backs. As several parents have pointed out, often strangers will comment in front of their child as though their child is not even present. As one parent said, it is important to be your child’s voice.

In general, if you have the time, energy and desire to do so, engaging the questioner in a brief discussion about MPS can be positive. Many people ask questions because they really do want to know. Others are simply rude people who have nothing better to do with their time than pass judgment on others. Often, families of special needs children will feel obligated to respond politely to the rudeness of others. However, you have every right NOT to be apologetic and should NOT feel guilty.

Remember that your dignity and the dignity of your child are not bestowed upon you by strangers.

STRATEGIES FOR DEALING WITH STRANGERS

- Ignore them or try to explain the situation.
- Kill them with kindness. Respond with a smile and a kind comment.



Maggie (MPS III A)

- Use the opportunity to vent some frustration in a positive way, possibly opening the door of enlightenment.
- Be prepared for stares and questions by smiling and answering. It helps spread MPS news and awareness.
- Ask people, “Do you know what MPS is?” If they reply “no,” politely ask them to keep comments to themselves unless they want to know what MPS is.
- Take along booklets and brochures wherever you go.
- Tell people that your child’s behavior is not voluntary, but is part of the disease process. Be matter-of-fact and don’t respond in anger.
- When dealing with stares, say something like, “Thank you for noticing our beautiful daughter. If you have any questions about her, we’d be happy to answer them.”

- Have the MPS Society Web site or phone number available. Explain that your child has a rare genetic disorder that is devastating to all affected; if you would like to know more and would like to help find a cure, please go to ___ or call ___. Consider making business cards that read, "Want to know more?" then list the Society's Web site.



TEASING

More often than not, your child will not tell you they are being teased about their condition. Although some teasing may be playful, generally it can be demoralizing and devastating. Your child may express their stress by displaying such symptoms as sleeping and eating problems, becoming withdrawn or aggressive, or complaining of physical problems such as headaches or nausea. If you think your child is being teased, you should take action promptly, not only to stop the teasing, but to help your child cope with future incidents. Because the physical features of MPS children will alter as the disease progresses, they are more vulnerable to stares, comments and questions. It is best to be prepared.

- Sometimes teasing is "accepted" by the silence and ignorance of supervising adults in your child's life. Talk to and educate teachers, friends and neighbors about your child's illness.
- Role play teasing situations with your child, each of you taking turns being the "teaser" and the "teasee." Work on likely scenarios and humorous comebacks.
- If your child is still able to walk, teach him or her to walk (and talk) assertively. Shoulders back, head up.
- Find out if other children in your child's school are being teased. Encourage your child to pass on strategies for standing up to teasing.
- Involve yourself with your child's parent/teacher association (or other relevant group) and make teasing and bullying issues a priority.
- Contact the principal, as well as your child's teacher, if there is any threat of violence.

Asking for Help

As it is said in the airplane as you depart, if an emergency situation arises, you must put on the oxygen mask first before anyone else. In order to make yourself better equipped to deal with your child and others, you must take care of yourself as well.

First thing first: it is okay to ask for help. You may not want to ask for help, and you may feel bad about doing it. You may want to ask for help but aren't sure what to ask for. Whatever category you fall into, remember that asking for help does not indicate weakness or laziness on your part.

Asking for help is probably the hardest. At first you really don't know what you need and then you feel bad about asking.

SHOULD YOU ASK FOR HELP?

That is really your decision. If it helps, consider that you are asking for help on behalf of your child and not on behalf of yourself.

I learned to open my mouth...because my child needed it and could not ask for himself.

Recognize that having a child with MPS can be exhausting. You can better help your child and your family if you have some rest for yourself. Running yourself ragged helps no one. Depending on the severity of your child's illness, his life may be shortened. Therefore, quality of life may be just as important as quantity of life.

GOVERNMENT AND CHARITABLE ORGANIZATIONS

Although varying from country to province and state to municipality, government programs are available to give aid to families with special needs children. Take advantage of these programs. They were developed for your benefit. If you find a program isn't available, contact your local

government representative and suggest that it be developed. If you receive no response, contact local television and radio stations and newspapers and explain the situation. Sometimes a little media attention creates tremendous results.

Many organizations, such as Easter Seals, March of Dimes and the Shriners, offer programs for special needs children. These programs vary, depending on your location. Contact a local office of the organization, or consult the Internet or your local library for suggestions on such organizations. Once you've found suitable programs, prepare yourself for an onerous application process. Keep detailed records of your child's condition, progress and prognosis. Ask teachers, care attendants and your doctor what other special needs children have in the way of aid. This sounds like a daunting task, but it is worthwhile.

FAMILY AND FRIENDS

Having family and friends ready, willing and able to help you is a mixed blessing. On the one hand, it's wonderful to have those close to you be supportive. On the other hand, make sure you don't ask for anything they aren't yet ready to handle. The more your family and friends know about your child with MPS, the better they will be prepared to help you. Some day-to-day activities may be very simple and straightforward to you, but daunting to someone who has had only brief encounters with your child.

We take our daughter with us to as many places as we can. That way people see her and have a better understanding of what it's like to take care of her. The offers to help then usually pour in.

It's important that family and friends get to know you, your family and your child with MPS. In doing so, they may be able to offer suggestions you may not have considered. People get a great feeling when they've helped one another!



Alena (MPS VI)

HOW OTHERS CAN HELP

When someone says, "Can I help?" what do you do? Do you freeze? How can they help? What can you ask for? It is helpful to have some ideas ready for this question. Some suggestions from MPS families on ways people can help are:

- Be there when you want to talk, to lend an ear when you need to vent.
- Make a donation to the Society in your child's name, such as on their birthday.
- Ask a friend to accompany you for an outing, particularly to help watch the kids. Children with severe MPS II and MPS III can be extremely hyperactive and may need shadowing.
- Write advocacy letters on your behalf.
- Allow their children to associate with your child with MPS. If your child is no longer very active, perhaps their children can sit with your child and watch videos, or play with your child.
- Ask them to take your unaffected children on an outing.
- Pick up groceries.

Family and friends also can provide:

- A voucher for babysitting, house cleaning, lawn mowing, etc.
- Relaxation music tapes/CDs
- A gift certificate for a massage or a facial at the local beauty salon
- A night away at a hotel or bed & breakfast
- Tickets for a movie or restaurant meal; including babysitting
- Carpet cleaning service
- Credit account at the local pharmacy
- Grocery gift card

Behavior

Not all children with MPS have behavior problems. However, parents of children with behavior problems are on a constant quest for information and strategies to cope with behaviors. Strategies do exist to make life a little more manageable for families.

Be sure to think creatively and obtain assistance from both professionals and respite workers about the most beneficial criteria when dealing with a child who has behavioral problems.

Children with MPS may display hyperactive, “motor driven,” non-stop behavior. This is when you see children who are on the go with a very short attention span, flitting from object to object, subject to subject, often with no apparent purpose. The children often are non-compliant with instructions and have no sense of danger. Physical aggression, occasionally with property destruction, is fairly typical and one of the most difficult aspects for parents to deal with and for doctors to treat.

UNDERSTANDING THE BEFORE AND WHY BEHAVIORS OCCUR

Physiological factors: Hunger, fatigue, illness, medication (side effect), language delay, boredom, inability to expel energy, fear and anxiety.

Environmental factors: Noise, lighting, size of area, unfamiliar setting, activity level.

Situational factors: Time of day, transitions, type of activity, demands/requests.

Interactional factors: Physical interference (being touched or pulled), being hurried, invading your child’s social space.

STRATEGIES FOR DEALING WITH DIFFICULT BEHAVIORS IN THE HOME

Once you have tried to determine the root causes of difficult behaviors, the next step is developing strategies to deal with these behaviors. Some families might find the help of a behavior consultant useful.

A consultant can assist with strategies to best suit your family and home environment. Being proactive and having a positive approach to situations will help to alleviate stress in your family and children.

- Set aside a room or area of the house within hearing distance of parents. Redesign the room to be safe for your child to play without constant supervision. (See Home Modifications chapter, Safe/Sensory Room, page 34.)
- Learn to live within your child’s world instead of trying to force your child to conform to yours.
- Entertainment can be provided by mounting a TV up on a shelf out of arms’ reach or mounted on the wall.
- Remove all breakables and furniture with pointed edges. Replace the furniture with a bean bag chair or a mattress without the bed frame.
- Provide safe chew toys, music, books, photo albums, tape recorder, balls for bouncing, and puzzles that are appropriate for your child.
- Create a Dutch door. This will keep your child safely in the room and at the same time provide a clear view outside the room.
- Install safety gates in doorways, especially in the kitchen.
- Investigate the wide range of safety items, now available in most children’s toy stores, to determine what can make your day-to-day living a little easier.
- Although it is difficult not to spoil or give in to children who have been through a lot (i.e., surgeries, doctor appointments, etc.), remember that all children will develop attitudes and expectations from you. If they feel they have an opportunity to

exploit your reluctance to set boundaries (bed time, chores, homework), they will.

- Securing reliable respite workers is essential for families facing the many challenges behavior difficulties present. This includes acquiring semi-skilled, informed babysitters for short times away from home. Out of the home respite with family members or respite care facilities can provide longer relief. (See page 17 for more information on respite care.)
- Above all, do what works for you. If you find that your child is happy watching hours of 'Barney,' adopt it as one of your strategies!

THE UNEXPLAINABLE

Screaming, pacing and crying episodes are likely signs of anxiety and are difficult to manage. If there are certain times of the day these behaviors occur, try to maintain routine and eliminate any additional stress for your child and yourself. This may mean avoiding unnecessary outings. There are some medications that might help your child through these difficult times. Discuss these episodes with your child's physician.

Parents report that children with MPS III may experience periods of frequent crying, often for no apparent reason. After ruling out all potential sources of pain, such as teeth, joints and constipation, you are left questioning the cause. It is not unusual for parents to feel helpless because you hate to see your child in misery and are not able to help. Some parents have found that giving Ibuprofen can help, but check with your doctor before giving any medication.

BEHAVIOR WITHOUT DEVELOPMENTAL DELAY

By giving in to children, you will unintentionally teach them that it is okay to manipulate you through their difficult behavior. Children learn quickly that a parent might give in to a public temper tantrum if the parent is easily embarrassed.

Use momentum and games to distract your uncooperative child (i.e., "Who can get dressed faster?").

It's important to be consistent, have clear expectations and set boundaries during these times. Lots of encouragement and reinforcement are needed to establish independence. Use verbal cues, such as "Good job," "I'm very proud of you," etc.

Whenever possible, activities should be thought out and prepared ahead of time. This will minimize stress. However, your child may be at a point where no amount of preparation will help and the best way to cope is to go with the flow.

BEHAVIOR WITH DEVELOPMENTAL DELAY

If your child with MPS is restless and needs to move, initiate a physical activity such as bouncing a ball, running around outside or going to the park. If other children are at the park and your child tends to be aggressive, be prepared to shadow your child and block behaviors before they occur. Try to redirect your child to a more appropriate area or activity away from other children. Some days this will work, whereas other days it simply will not. To avoid over-stimulation, scout neighborhoods for less occupied playgrounds or go to the local park during a quieter time of day when your child can wander at will. Investigate indoor playgrounds and ask when they have quieter times.

On the days when your energy is low, perhaps going for a car ride will satisfy your child. Share parenting responsibilities – one parent gives the other a break and vice versa.

Investigate local community centers and learn what programs are offered for children with special needs. Your child is entitled to attend programs. Some programs will provide one-on-one workers, but these workers may require some extra training from you.

SOCIALIZATION

When a child has behavior problems, socialization can be difficult and heartbreaking. However, once other children are taught how to approach and associate with your child, wonderful things can happen.

- Talk to children about MPS, and about strategies to assist them with communication and socialization with your child. This will give them a better understanding of your child's behaviors and help to relieve some of their own anxieties.
- Encourage other children to verbally remind your child to use nice touching and not to hit.
- Children should leave a space (about an arm's length) around your child so he does not feel crowded.
- If your child has hearing loss, he may not know if someone is nearby until seeing the person. Your child should be approached from the front so as not to startle.
- Teach children to put their hands out as a greeting. If your child grabs another child's arm or hair, teach them to put their hand on top of your child's and tell him to "stop" or "let go."
- Help children to learn that behaviors are not intentional or malicious. By using patience, there will be many rewards.

COMMUNICATION

Children who have impaired language will require different approaches to communication. Provide picture symbols for children with verbal impairment. Teach them functional sign language if they have the motor skills to sign. Prepare children for transitions (moving from one activity to another) by using photos. If you will be going to the mall, show pictures of your home, the car and the final destination.

THE CLASSROOM

For information on strategies for integration in a regular classroom, see Tatjana Smrekar's article, "Child & Youth Worker," on the Canadian MPS Society's Web site (www.mpssociety.ca). Information also can be found on the National MPS Society's Web site (www.mpssociety.org) under "Educational Strategies." Topics such as explaining the individual education plan process, parents' rights and suggestions for integrating into the classroom are covered.

RESPITE CARE

Implement breaks from behavior problems by allowing for adequate respite. Respite allows families to regroup to better deal with their situation. Incorporating regular respite breaks for you is essential. Hire respite workers to come the time of day most helpful to you, such as dinner time. Once a routine is established, you can use this time to cook, spend time with siblings, take a peaceful walk or take a nice hot bath. Don't forget to save some respite time for your partner.

Having people constantly in your home can be an adjustment. But the advantages outweigh the disadvantages. Don't be afraid to explore options for respite care.

NETWORKING

Identifying community and professional resources is essential in helping to assure a higher quality of life for your child and family. The Society can provide you with information as well as emotional support by putting you in touch with other families who have experienced similar circumstances.

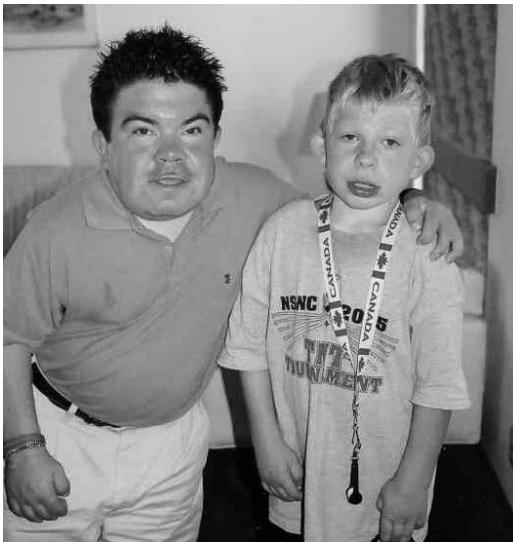
MEDICATION

For information about medication and behavioral issues in children with MPS, see the full conference transcript of Use of Medication in MPS Diseases, by Dr. Ed Wraith, which can be found on the Canadian MPS Society's Web site at www.mpssociety.ca.

Wander Prone Children

It's all about anticipation...you have to anticipate their every move, be one step ahead of them...

Children are curious by nature and are easily distracted by sights and sounds around them. Because of this, many children are prone to wander off, following something more interesting than the delights offered by a grocery shopping trip with mom. Although most children have wandered at times, children with severe MPS II and MPS III are particularly prone to it, and their speed should never be underestimated. Trips to the mall can become real adventures. See Home Modifications, page 31, for ideas on minimizing wander-related risks in your home. Following are general ideas for such potential adventures as grocery shopping, walking to the park or getting out of your car.



Simon (MPS II) and Nicklas (MPS I)

- Dress your child in bright colors so you can easily see her.
 - Dress your child in overalls so you can hold on to her.
 - Have your child wear an ID bracelet or necklace. This may be difficult though, as many children pull them off.
 - Write your child's name, address and phone number somewhere on their clothing (labels in T-shirts, etc). This can be done with a simple water-proof marker.
 - Use a Harla flex (Velcro leash) to attach your child's wrist to your belt loops.
 - Sew simple belt loops onto the back of a vest or your child's shirts. This will give you something to grab on to in case your child tries to dart away.
 - If your child does run away in a store, don't hesitate in starting the search and informing as many of the staff as possible right away. Have as many people as possible do a grid-circle search starting from outside lanes and moving to the center. Check the toy section.
 - Hold onto your child at all times, especially when exiting the car.
 - Bring a friend or a responsible young adult with you for day trips and shopping.
 - Keep everything you need readily available. Put car keys on a belt clip, and before going to a store put folding money or ATM cards in a shirt pocket. That way you don't have to get out your wallet and be distracted or let go of your child's hand.
 - Learn the places your child is prone to wander to and avoid them; distract your child as you pass by; or go there first and let your child expend energy early in the trip.
- Research child locator programs in your area. Police stations often have kits you can prepare and keep on file in case your child is lost.
 - You are required by law to restrain your child in a vehicle. To ensure your child does not run away, some families find a multi-purpose harness useful when walking. Research the child-restraint laws in your area, as some locales do not allow use of the multi-purpose harness.

Gifts and Toys

Plastic hammers, tools, rubber ducks, my arm...

Children with MPS are no different than other children in their need to play, relax and grow. It is important to remember that toys can present choking hazards or other risks (breakage, causing cuts or splinters) for all children. Parents suggest asking the following questions when buying or making toys for children with MPS:

- Is the toy durable? Consider the strength and size of the child. If breakable, can the toy's parts present a choking hazard?
- Is the toy easy to use? How much strength is required to activate it? Is the toy complex or easy to understand?
- Is the toy adjustable or adaptable to the child's needs, or are there restrictions on where the toy is being used (for example, if your child has limited mobility)?
- Is your child an active participant during the toy's use, or is the toy something to be watched (for example, a mobile)?
- Does the toy have multi-sensory appeal? Lights, movement, sound, texture, taste, scent?
- Does the toy involve other players? Can it be played alone as well?
- Can the toy be washed?
- Visit Discovery Toys (www.discoverytoysinc.com) and Lamaze Baby Toys (<http://lamaze-baby-toys.bava.biz/>) for a wide selection of safe, durable toys for children of all ages, sizes, strengths and limitations.

CHEW TOYS

Parents of children with severe MPS II and MPS III should be prepared to invest in chew toys – you can never have too many! It is important to keep all chew toys clean, as bacteria grows quickly and will thrive in some chew toys, particularly those made from cloth.

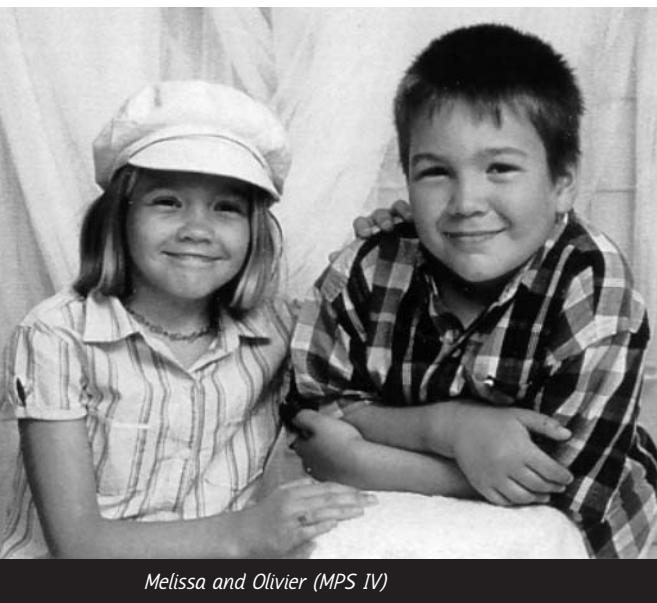
Below are some parents' suggestions for chew toys and their maintenance:

- Make a basket of safe chew toys. When one is dropped, take a fresh chew toy and place the dropped one in a "to be cleaned box."
- For kids in day care or school, suggest to the school that the children have their own toy basket. This will prevent, or at least minimize, children chewing on each other's toys.
- Use a simple washcloth; it is absorbent and easy to wash.
- Attach baby teething toys and pacifiers to strollers.
- Use rings from a ring toss toy, rubber chew toys made for dogs, cloth diapers and small cloth dolls.
- Chewy Tubes (www.chewytubes.com) are oral motor devices designed to develop biting and chewing skills.
- Use clear tubing from a wine-making store (the tubing is sterile; hardware tubing is not). Cut a piece 4 to 5 inches long, put string through and tie a clip on the end. Pin to clothes.

Again, the hazard of choking cannot be overstressed. Legos and other small-pieced toys must be avoided, particularly if you have an active child who is prone to chewing toys.

OTHER TOYS

- Duplos
- wooden puzzles with large pieces
- Thomas the Tank Engine pieces
- plain wooden blocks
- toy air planes
- picture books with big bright words and illustrations (also good for chewing).



Melissa and Olivier (MPS IV)

OTHER ACTIVITIES

If your child is very active and needs constant, supervised activity, consider:

- Slides and other indoor playground equipment in the basement.
- Swimming. Make sure your child is wearing an approved life jacket.
- Trips to the park. Try to go when it's less busy.
- Riding a bike can be dangerous and difficult to supervise closely. Consider buying a tandem bicycle and riding with your child. Make sure you and your child wear approved helmets.
- Build a safe playroom where all the toys are safe for your child (see Home Modifications, page 31).

GIFT SUGGESTIONS

- Colorful mobiles may be found at gift shops, baby shops, department stores or you can make your own.
- Wall posters. It's a good idea to get them laminated.
- Pillow cases featuring brightly colored animals or the child's favorite characters.
- Large multi-display photo frames. Pop in photos of the child's favorite people (family, friends, pets).
- Picture frames to mount and display your child's artwork.
- Flexitube lighting. These are clear ropes containing colored lighting strips. The lights can "chase" or change color and can be mounted on the wall or ceiling. Contact specialty lighting stores for information and prices.

Gifts for the Active Child

- Thomas the Tank Engine Bop Bag – from toy and department stores. This product stands about one meter (3 feet) tall when inflated and returns to an upright position when bopped.
- Large rubber balls – now come in fantastic bright multicolor or feature cartoon characters (Tweetie, Sesame St., etc) – found in department stores.
- Sand play toys – buckets, waterwheels, etc.

Gifts for Less Active Kids

- Aquaduck – beanbag for floating in the pool.
- Bubble blowing kit.
- Hand puppets are a fun way for siblings, parents and friends to interact with your child. Homemade is best.
- Small photo album filled with favorite people.
- Sesame Street dolls that sing and talk.
- Foot bath/massager – from electrical and department stores.
- Wind-up musical TV from toy stores.



Joe (MPS III A)

Miscellaneous Gifts

- Good quality wash cloths.
- School accessories (lunch box, drink containers).
- Swimming accessories (bathing cap, buoyancy aids, bright towels).
- Bath toys and vinyl bath books from toy/baby departments.
- Cardboard books (also great for chewing).
- Cassette player for child's room.
- Videos cassettes and DVDs.
- Carton of appropriate brand and size diapers.
- Bibs. For those who can sew try making bibs shaped like a triangular scarf – far more attractive than conventional bibs. Velcro fasteners are a good idea.
- Accessories to make little girls' feel pretty (hair scrunchies, bubble bath, powder, pretty socks, etc).
- Baby play frame.
- Vinyl toys to chew on.
- Electric toothbrush.

ADAPTING TOYS AND GAMES FOR YOUR MPS CHILD

Doll Houses

Glue or Velcro furniture pieces in place so your child can play with the dolls without “redecorating” the house. Securely attach the house to a table, using a C-clamp for stability, or place the doll house on the floor.

Puppets

Make a puppet using a sock. Attach facial features with Velcro (beware of potential choking hazards of small pieces). Make a puppet stage using a cardboard box.

Books

Your child's hands may have difficulty grasping smaller objects as the disease progresses. To make page turning easier, glue small felt spacers or tabs on each page, or insert page dividers so your child can turn pages independently. Also consider removing the binding of the book and placing the pages into clear plastic sleeves.

Playing Cards

Make or buy large-sized playing cards. Laminate them, or paste them to poster board. Fashion a cardholder out of a clean Styrofoam tray or the lid of a shoebox by turning the tray or lid upside down and cutting slits into the back.

Feeding and Swallowing Difficulties

Swallowing is a significant problem for many children with MPS. In addition to narrow airways, MPS children have physical disabilities requiring more support with feeding and swallowing as their coordinated movement is affected. This will inevitably have a direct impact on their feeding skills. Feeding is a complex process involving a combination of actions and coordination to ensure that breathing and swallowing do not occur at the wrong time.

(See also Safe Eating Techniques, page 24.)



Kerrin (MPS III A)

SWALLOWING AND THE STAGES OF PROGRESSION IN MPS DISEASES

There are three stages involved in the normal swallowing process: oral, pharyngeal (swallow) and esophageal (food moving to the stomach). A breakdown can occur at any or all three of these stages.

In individuals with MPS, the oral stage results in reduced chewing ability and longer mealtimes. Pharyngeal difficulties are displayed initially by inconsistent swallowing, borderline aspiration and/or chronic aspiration. This is followed by increased

pharyngeal difficulties resulting in recurrent aspiration of fluids and solids.

Clinical symptoms of a swallowing difficulty and aspiration include recurrent chest infections or history of frequent upper respiratory infections, regular spiking temperatures, choking or coughing during feeding, and inability to manage oral secretions.

THE ROLE OF THE SPEECH AND LANGUAGE THERAPIST

Once a swallowing difficulty (dysphagia) has been identified, the individual should be referred to a speech and language therapist for an assessment of the problem. Speech and language therapists have specialist knowledge in the anatomy and physiology of the head and neck structures. Speech and language therapists, in addition to being trained to assess verbal and non-verbal communication and speech and language development in individuals, also can provide input on developing feeding and drinking skills. This includes giving advice on positioning during feeding, which food types and textures are appropriate, the use of feeding utensils and facilitating communication during feeding. Other medical factors also need to be carefully considered as well as the nutritional and safety aspects of the swallow. The speech and language therapist works in liaison with a multi-disciplinary team.

VIDEOFLUROSCOPY

The Videofluoroscopic Swallow Study is a high-tech device that is often used to get a closer look at a person's swallowing reflexes. This also may be used to assess apparent swallowing difficulty. It gives a clear view of all stages of swallowing and can detect aspiration if it occurs. It also can identify the impact that different food textures and positioning strategies may have on the safety of the swallow. The examination lasts about 10-15 minutes and is simply an X-ray of how well the swallowing mechanism is working.

CERVICAL AUSCULTATION

This refers to a method whereby the different acoustic sounds heard during respiration and swallowing can be determined. It can be very effective in accurately diagnosing dysphagia. Despite being a new technique, it has been confirmed as effective by further videofluoroscopy examinations.

MANAGEMENT

The speech and language therapist will agree on an appropriate treatment plan with your family designed to help manage dysphagia. Factors considered in the management of a feeding disorder include modifying the diet, avoiding high-risk textures, slowing the rate of feeding, using thicker liquids (thickening powder rather than naturally thick drinks), reducing volume (aspiration may increase with amounts taken) and non-oral feeding such as gastrostomy (making a new opening to the stomach from the abdomen, created by a surgical procedure).



Matthew (MPS VII)

HIGH-RISK FOODS

High-risk foods include stringy, fibrous textures such as pineapple, runner beans, celery, lettuce, vegetable and fruit skins, including beans (broad, baked, soy), black-eyed peas and grapes. Other foods are mixed consistency foods, including cereals that do not blend with milk, such as muesli; minced meat with thin gravy; soup with lumps; crunchy foods like toast, flaky pastry, dry biscuits and chips; crumbly items such as bread crusts and pie crusts; hard foods like boiled and chewy sweets and toffees, nuts and seeds; and husks including corn and granary bread.

Safe Eating Techniques

Eating is a complicated process and may become problematic for some children with MPS, particularly as they regress and lose skills. If your child is having difficulty swallowing safely and you're concerned whether he or she is receiving adequate nutrition, consult your child's doctor and set up a meeting with a speech pathologist, occupational therapist and dietician. This multiple disciplinary team will determine problem areas and will make the necessary recommendations to ensure your child's safety, comfort and adequate nutrition.

FLUIDS

Natural gravity causes fluids to run down the throat very quickly. We use our natural instincts to control the rate of swallowing through our tongue, cheeks and mouth mechanics. Some children with MPS have difficulty controlling the rate at which the fluid runs down their throat.

Ways to control the swallowing rate are:

- sippy cups
- baby bottles
- straws
- various types of cups on the market
- thickeners

Depending on the severity of swallowing difficulty, liquids can be thickened with a modified cornstarch product, such as "Thick it." Thickening drinks to nectar, honey or pudding consistencies allows for safe swallowing. Thickeners can be obtained from specialty food shops or drug stores.

Chewing food and moving it around the mouth with your tongue ultimately creates a bolus (a small lump of food), ready for swallowing. As skills are lost, the ability to coordinate chewing action, to



Matthew (MPS III A)

move food around the mouth, and to transfer it safely to the back of the throat to be swallowed is also lost. Oral spillage, coughing and aspiration are all signs of choking risks and steps should be taken to alleviate these risks.

There also are risks when moving the head into an extended position, opening the airway and leaving your child with the inability to fight gravity and the movement of food/drink to the back of the throat.

SAFE EATING TECHNIQUES

- Proper seating and head positioning are essential. Some children with MPS have poor trunk and head control. Sitting in a wheelchair or a feeding chair will offer trunk and head support and assist in aligning the body's midline.
- Cut up food into small, bite-size pieces
- Give small amounts of food at a slow rate
- Use verbal cues – “eat, slow, chew”
- Use a safe sipping cup (many types are available)

When small, bite-size pieces of food become a problem and a child shows signs of discomfort such as coughing, sputtering, oral spillage or aspiration:

- Mash or puree food
- Give small amounts at a time
- Thicken liquids to nectar consistency
- Continue to use verbal cues such as “good chewing”

If chewing and swallowing becomes too difficult, consider:

- Pureeing all food
- Thicken the food as necessary (soup consistency)
- Thicken all liquids
- Feed at a manageable rate. Slow down and give your child time to complete transitions.

MEAL IDEAS AND RECIPES

Breakfast: pureed French toast, oatmeal, cream of wheat with whole milk or cream to increase calories, yogurt, applesauce.

Lunch: egg salad with mayonnaise, well-cooked egg noodles/sauce puree, pureed chunky soup, tuna, salmon.

Dinner: combine mashed potatoes, pureed chicken/veal/beef and vegetables, top with gravy.

G-TUBE FEEDING

At some point you may be faced with the decision of whether or not to use a g-tube to feed your child. This is a very personal decision and there is no right or wrong answer. Only you can make the decision. Here are a few pros and cons to consider:

Pros

- Increases safety
- Ensures proper nutrition
- Maintains weight
- Improves your child's quality of life
- Reduces stress around feeding issues
- Reduces your time so you can get other things done

Cons

- Interferes with the natural progression of the disease
- Prolongs the life of a very ill child
- Increases the risk of aspiration if your child experiences reflux, receives too much food, or is in the incorrect body position
- Often performed after the child is too ill to benefit from it. Talk with your doctor about the timing for g-tube placement.

ELIMINATION

Whether your child is eating orally or by g-tube, it is imperative that he is having regular bowel movements. If not, complications can arise such as impacted bowels, bowel perforation, improper digestion and painful gas. If you have concerns or questions about your child's bowel habits, consult with your child's doctor or dietician.

Constipation, Diarrhea, Incontinence

Children with MPS often experience problems with incontinence, diarrhea and constipation. These problems may be severe. As your child grows, she may not only lose skills, but muscular control and strength. This will affect digestion as well as elimination. In many cases, children with severe MPS who previously had been toilet-trained will lose that skill. In others cases, children are not able to be fully trained. It's important for parents and other caretakers to understand that these problems are part of the progression of the MPS diseases, and they should address the situation without getting emotional.

As your child's condition regresses, you will notice that constipation and diarrhea will occur with increasing frequency. As your child becomes less active and mobile, constipation will overtake diarrhea.

CONSTIPATION

If your child is experiencing constipation, you must address it quickly before your child experiences bowel obstruction. Use a natural approach to keeping your child regular whenever possible. Laxatives are dependency forming and should be used in consultation with your child's doctor.

Diet Changes and Natural Remedies

Some parents have found the following items to be helpful. However, you should first check with your doctor.

- beans
- juice
- raisins
- apples
- two parts water, one part prune juice
- Kayro syrup
- baby food spinach
- oatmeal
- raw vegetables

Medication

As stressed above, use these only on the advice of your child's physician.

- Miralax
- Citrucel
- milk of magnesia
- Enulose
- suppositories (silicone, Dulcolax)
- stool softeners (such as Lactulose)
- mineral oil
- enemas
- Pathmark Sienna C Plus laxative

DIARRHEA

Children with MPS may experience extreme episodes of diarrhea. It is crucial that you ensure your child is properly hydrated. Children with MPS can dehydrate quickly and are not able to tell you they are dehydrated. The Canadian Pediatric Society offers the following as some signs of dehydration:

- decreased urination (less than four wet diapers in 24 hours)
- no tears
- dry skin, mouth and tongue

- sunken eyes
- grayish skin

Continue to offer your child food and drink, but avoid giving your child sugary foods. Also, avoid giving your child medication without the approval of a doctor. In many cases, diarrhea can be lessened by reducing or eliminating milk products in your child's diet. *If you feel the diarrhea is severe, or is accompanied by frequent vomiting, seek medical attention for your child.*

Some parents of children with MPS have suggested the following remedies for chronic diarrhea:

- papaya enzyme tables
- cream of wheat
- white grape juice
- yogurt
- bananas
- applesauce
- rice

Medication

- Immodium
- Metamucil

INCONTINENCE

Many children with severe MPS can be toilet trained, but will more often than not lose this skill as their condition worsens. Due to the physiology of the disease, many children with MPS cannot be trained at all. Parents should be aware that the progression of the various forms of MPS varies from child to child and that, for the most part, an inability to be toilet trained does not necessarily reflect a behavioral issue. Diapering is a reality for most children with severe MPS. Nonetheless, consult with your physician if you are having problems toilet training your child.

Below is some advice from parents of children with MPS:

- Take a towel with you so if you need to change a child in a public place they don't have to lay on the bathroom floor.
- Try to have children eat properly, from all food groups.
- Always have a change of clothes wherever you go, in addition to diapers, wet naps and blankets.

DIAPER RASH

Diapering will be a necessity for many children with MPS. Because of prolonged diapering, children will be more vulnerable to rashes. In many cases, children will be on antibiotics. Because of this, their natural bacteria is disrupted, leaving these children more prone to yeast infections which are aggravated by diapering. Consult your child's doctor if rashes become severe or appear to be unaffected by over-the-counter ointments such as zinc oxide. Putting baking soda into your child's bath also may help prevent rashes, although excessive bathing will only irritate your child's skin further.

DIAPERS IN GENERAL

Many states in the United States and provinces in Canada offer programs to help offset the cost of diapers. Because these programs can change, consult your doctor or your government's Web site for information on such programs. If you do not have Internet access, or are unsure where to begin your search, consult your local library for resources. Other sources of information are local homes or hospices for the elderly or the disabled. Many charitable organizations also offer financial assistance. Some insurance carriers will cover diapers and other related expenses. In some cases, you may need to ask your child's doctor to write a prescription for diapering.

Lifting

As the disease progresses, a child with MPS requires more frequent lifting from cars, baths, wheelchairs, beds, etc. Following are some suggestions to reduce the amount of stress on your back:

- Lift with your legs, not with your back.
- Plan the lift. The best way to avoid injury (to you and your child) is to minimize sudden, unplanned twisting, reaching, bending, stepping-over, ducking. Knowing where you're going to lift, and how, is very important.
- Make sure the pathway is clear. Remove toys, pets, other children, shoes, etc. before you lift.
- Position your feet. Feet should be shoulder-width apart and pointing in the direction you're traveling.
- Carry your child as close to you as possible. This will minimize the weight you're carrying.
- Bend your knees, keeping your back straight, head and shoulders up. Essentially, you're squatting. This keeps the stress of the lift evenly distributed.
- Tighten your stomach muscles as you lift. This takes stress off your back.
- **DO NOT TWIST YOUR BODY TO CHANGE DIRECTIONS!** If you're turning, turn with your feet.
- Carefully lower your child, keeping your knees bent and your back straight.
- If more than one person is lifting, make sure you are continually communicating with the second person, what direction you are going (whose left and whose right) and count out the lift.



Garrett (MPS II)

Sleeping

I always said if I could come up with a bed that smells, sound and feels like a car, I would be rich!

Sleeping difficulties are extremely common in children with MPS. This may mean that your child has problems falling asleep, or may wake frequently throughout the night. If your child isn't sleeping, chances are you aren't getting much sleep either.

There are several issues to consider that are connected to the insomnia your child may be experiencing. Using medications, making the child's room more conducive to sleeping, and protecting the wander-prone child who is up at 2 a.m. when everyone else is sleeping should all be addressed.

It is important to determine whether your child's sleeping problems are related to other health issues, such as sleep apnea or airway obstructions common to children with MPS. As with any other medical issue, it is important to consult your doctor to determine whether these or other issues are causing sleeping problems. Often your doctor will refer your child to an ear, nose and throat (ENT) specialist. If your doctor has not done this, request a referral as eventually all children with MPS should meet with one.

TROUBLESHOOTING

Keeping a sleep diary for your child for at least a week or two will help when you consult with your doctor or sleep specialist. Note when your child goes to bed, how long it takes your child to fall asleep, how often your child wakes throughout the night, and any naps your child may take during the day. Keep notes on whether your child experiences breathing problems or appears congested in any way. Also record the condition of your child's room, such as dark, noisy or drafty, and the type of bed. All of this information will prepare you for questions your doctor or sleep specialist may have.

SLEEP APNEA/ AIRWAY OBSTRUCTION

A very common and severe problem for many individuals with MPS is airway obstruction. This can be caused by narrowed or blocked nasal passages, a large tongue, or enlarged adenoids and tonsils. If a child is not getting adequate oxygen intake at night, sleep is disturbed, leading to frequent naps during the day. Surgical removal of tonsils and adenoids, or giving oxygen at night helps alleviate this problem. Your ENT can help assess this problem and may recommend a sleep study. Other possible treatments include opening the airway with nighttime continuous positive airway pressure (CPAP), and bi-level positive airway pressure (BiPAP).

MEDICATION FOR SLEEP

The decision to aid your child's sleep through medication is a difficult one and should involve your family physician and/or a sleep specialist. Children with MPS often take several medications and thus, it is important to ensure there are no interactions among sleeping aids and other medications.

Although you may not want to medicate your child, in some cases it may be necessary. Parents have to sleep, as do the other members of the family, including your child. As with other issues, it is important to stay informed by talking with other families and your doctors. Many families have found melatonin to be useful. Melatonin is available by prescription in Canada, and over the counter in the United States. It is important to know that melatonin may aid your child's sleep, but will not lead to normal sleep. A fact sheet about melatonin can be found at www.mpsociety.org.

Many MPS families have reported success with certain prescription medications, some of which are listed on page 30. Please speak with your doctor before giving your child any medication.

- Amitriptyline
- Benedryl
- Chloral hydrate
- Clonidine
- Clorazepam
- Lorzapam
- Trazodone
- Trileptal

SLEEPING SAFELY – MODIFICATIONS TO THE BEDROOM

Children with severe MPS II and MPS III may be very active. Having a bedroom that is safe and secure reassures parents that if the child awakens at night, he or she cannot hurt himself or wander through the house. By creating a safe environment, you can also place your child in this room whenever you need to make a phone call, cook dinner or simply use the bathroom. Although home modifications are discussed in more detail on page 31, below are some suggestions from families about strategies for making a child's bedroom safe:

- Put a mattress and box springs on the floor so the child won't fall far if he does fall out of bed.
- Put a crib mattress and single bed mattress up against the wall on two sides of the bed so the child won't hurt himself hitting the wall with head or legs.
- Make a two-part door so you can open the top-half to check on the child. Close and lock the bottom half so the child can't get out of the room during the night.
- Remove all hard furniture.
- Place only soft, safe toys in the room.
- Suspend the TV from the ceiling.

- Cover all wires from lamps, TVs, etc. with tubing and nail them to the walls so your child cannot pull on them.
- Place a baby monitor in your room to listen for your child at night.
- Put a childproof lock on the door knob.
- Put a baby gate in the doorway of the room.
- Use a white noise maker or music; the sounds may help your child fall asleep.
- Lava lamps, fiber-optic or Christmas lights, and water fountains (anything visual with movement) may be helpful for the more regressed child.

DEALING WITH INSOMNIA IN GENERAL

- Always have a bedtime routine, such as having a warm bath followed by putting on pajamas and singing songs.
- Turn off all lights.
- Let your child walk around his room or the house a little before bedtime.
- Rub your child's hands, legs and back.

Consult the Canadian and American MPS Society Web sites for articles (such as below) and links to information about sleeping problems and children with MPS.

Sleep-Disordered Breathing: Causes and Treatments.

The Connection: Summer 2001. Frank P. Manginello, MD, FAAP, and Mary T. Carbone, MD, FAAP. *Exceptional Parent Magazine*, July 2000

Home Modifications

Making changes to your home to accommodate the needs of your child with MPS can reduce daily stress, make children happier and simplify the parent's job. At some point, most families will need to make changes to their homes. Below are some innovative ideas that families have discovered and found effective in their situations.

MODIFICATIONS OUTSIDE THE HOME

- Wide walkways with slip-resistant surfaces and no abrupt changes in level
- Ramping to porches and doorways making sure the ramp is built at the proper slope to be safe
- Ramping inside a garage for adverse weather situations. A canopy to cover the outside ramp also is helpful.
- Porch lifts if ramping is not practical or desirable
- Hand rails adjusted for the height of your child
- Fenced outdoor play area with gates and secure
- A safe porch or deck where children can go outside independently. Make sure the gate and lock to the stairs are very secure, or install a railing around without yard access.

ACCESSIBILITY POSSIBILITIES

A one-level home is ideal for accessibility. However, wheelchair lifts for stairs or vertical home lifts that require a shaft can be used in multilevel homes. Putting common activity rooms on one floor and bedrooms on another is helpful. For example, remodel the rooms on the main level to create an accessible bedroom and bathroom or build a main floor addition to add a bedroom and bathroom.

GENERAL HOME SAFETY MODIFICATIONS

- Light switches lowered or raised depending on your child's height
- Light switches moved to the outside of your child's room so she cannot constantly flip the switch
- Lever-type door openers
- Child-proof door knob covers
- Outlet covers
- Slide locks on bi-fold closet doors
- Alarm system that sounds when door is opened
- 36" wide doors for wheelchair access and wider hallways
- Keyed locks on all outside doors or place locks high on doors out of reach, such as chain locks. If you decide to do this, **MAKE SURE** you can open the door in the event of an emergency.
- Reduced amount of furniture, such as coffee tables, to make the home roomier for wheelchairs and to decrease tripping hazards
- Tempered (plexiglass) glass in windows
- Heavy curtains and rods that will stand up; copper tubing is one suggestion
- Hardwood or vinyl floors for ease of wheelchair use and ease of cleaning. Low-pile commercial carpets with high-quality underpad also can stand up to wheelchair traffic.

- Handrails at your child's height, using closet dowels
- Elimination of throw rugs
- Gates or locked doors at top of stairways to prevent falls
- Carpeted stairs to soften falls
- Locked cabinets for personal care and home care products



DJ (MPS IV)

BATHROOM MODIFICATIONS

Bathroom safety is unique for each family depending on your child's level of functioning. Accidents easily can occur in a bathroom as you struggle to bathe a very slippery and active child. Innocent items like hand soap, toilet paper and shampoo can be irresistible to your child and may need to be located in a locked cabinet or placed out of reach. When addressing bathroom safety, consider not only the child's safety but the safety of the caregiver as well. It is of utmost importance that you care for yourself so you can provide the care your child deserves. An improper lift in or out of the tub can cause serious back strain and injury. Following are some bath time/bathroom suggestions:

- Place a lock or hook on the outside of the bathroom door out of your child's reach to keep her out of the bathroom and out of trouble when unsupervised.

- Install safety latches on cupboards and cabinets or have personal care items on a high shelf out of your child's reach (also try to place cabinets so care products are easy to access by the caregiver).
- Use "no tears" baby products.
- Provide safe chew toys or a clean wash cloth to keep your child's hands busy so she is less likely to grab personal care products.
- Place rubber mats on the bottom of tubs and showers, as well as non-skid throw mats, on the bathroom floor.
- Put a plastic stool or lightweight plastic chair in the tub or shower so your child can sit. Use a rubber mat so the stool or chair does not slip. Cover the plastic stool/chair with a wet towel for comfort and to prevent slipping.
- Turn the thermostat of the hot water heater down to a safe temperature to prevent burns.
- Take the covers off the faucets so the child cannot turn them.
- Cover the faucet with an inflatable infant faucet cover.
- Move faucets to a level out of reach of your child.
- Provide a sturdy stool with steps or a ramp so your child can safely reach the sink for hand washing, teeth brushing and toileting.
- Purchase inflatable bath pillows if your child is having trouble sitting unassisted.
- Place the bathroom on the main floor of the home or on an accessible level, if remodeling.
- Use a 36" door with the door swinging out to maximize available bathroom space. A sliding door also works well.
- Provide sufficient clear floor space for easy wheelchair access and for one or two caregivers to assist with personal care.
- Install grab bars in bath and toilet areas if your child is able to understand how to use these items. They also can be helpful to the caregiver.

- Install a barrier-free, roll-in shower on a grade, allowing water to flow to the drain. There is no lip or edge on this type of shower so a bath chair can be rolled in and out with ease.
- Install additional equipment in a shower, such as a hydraulic bath lift, ceiling lift or bathtub seat, grab bars, a thermostatically controlled hand-held shower attachment on a vertical rod, or high and low mounting brackets.
- Expand the shower to accommodate a bath chair and the caregiver.
- Move a changing table for diapering and dressing to the bathroom. One that adjusts in height can save an enormous amount of back strain. Velcro straps attached to the table keep your child safe if the caregiver needs to step away.
- Add a chute from the bathroom to a garage or basement for easy disposal of diapers.
- Install a therapy tub (air tub).
- Acquire a portable Hoyer lift that can be used in other rooms as well.

KITCHEN MODIFICATIONS

- Gates on doorways to kitchen area
- Dutch door (half door that can be open on top half and locked securely on the bottom half and child can be seen but cannot get out)
- Locks on all drawers and cabinets
- Magnetic locks on fridge or other fridge lock system (such as Velcro)
- Step stools for shorter children
- Remove knobs off the stove when not in use
- Locked drawer for knives and all sharp, dangerous utensils
- Restaurant booth that has a fastened bench and table to prevent tipping chairs or tables
- Dishes in lowered cabinets
- Up and down faucets for ease of operation

BEDROOM MODIFICATIONS

- Vail bed
- Mattress on the floor so a child cannot hurt himself if he falls out of bed
- Padding on the walls or pillows against the wall, especially next to the bed
- A single bed with high foot and headboard, and custom-built side that folds down and folds up for sleeping
- A hospital bed is safe and appropriate for some children
- Sleeping bags or tucking covers tightly around them to keep children covered at night and also may keep them in bed (use a double or queen-size sheet on a single bed and tuck in very securely)
- Kelli quilt – a weighted blanket used for sleeping
- Heavy blanket sleeper or extra socks and sweatshirt so your child won't get cold if the covers are thrown off
- Television and VCR or DVD combo on a ceiling mount
- Dutch door
- Hook or lock on the bedroom door from the outside so you know your child is unable to roam the house unsupervised at night
- Baby monitor
- Toys attached to a wall-mounted board so your child cannot throw them
- Collages of favorite pictures and photos on poster board and fastened under plexiglass

LIVING AREA MODIFICATIONS

- Remove knick knacks and plants; replace with safe items that your child enjoys.
- Remove coffee tables and end tables with sharp edges and corners or cover sharp edges with corner protectors.
- Replace table and floor lamps with ceiling or recessed lighting.
- Secure entertainment center within a cabinet with door locks.
- Mount the television on the wall or hang from the ceiling.
- Build a television into a wall at an out-of-reach height.
- Put an acrylic panel over the buttons on the front of the TV.
- Move pictures higher on the wall.
- Pad fireplace hearths with carpet or cushions when not in use. Be sure to move all flammable materials away from area when a fire is burning.
- Build a decorative iron railing around the fireplace and hearth area.



Bryce (MPS II)

SAFE ROOM/SENSORY ROOM MODIFICATIONS

Some families find it is worthwhile to have one room for your child that is a safe place, full of enjoyable activities. This can make life easier and more enjoyable for the entire family. Some ideas for creating a safe room are:

- TV and VCR/DVD out of reach
- Cassette or CD player to play your child's favorites and quiet soothing melodies
- Bean bag chairs or other comfortable, safe furniture
- Sensory lights, such as lava lamps, fish tank or bubble tubes, string lighting, and fiber optic lamps all behind a plexiglass cover or on a high shelf
- Projector that displays shapes and colors on ceiling, floor and walls
- Tactile panels – a variety of textures to touch and explore fastened to a board and mounted on a wall
- Manipulative panel – a variety of switches, dead bolts, other latch hooks fastened to a board and mounted on the wall
- Ball pool
- Christmas tree lights in netting or strings hung on the wall (twinkling or blinking lights may not be appropriate if your child is prone to seizures)
- Plenty of safe, manipulative and chew toys within reach

Please keep safety in mind whenever you make changes in your home. You might consider involving an occupational therapist, finding more information on the Internet, or asking questions to a MPS chat group. Sometimes magazines such as *Exceptional Parent*, or a catalogue of products for special needs children and adults, such as "Flaghouse," can give you the inspiration you need to find a solution.

When planning to remodel or modify your home, consult with professionals, such as architects and reliable builders. Some of these professionals specialize in accessibility problems and may suggest novel solutions to your very unique problems.

Travel

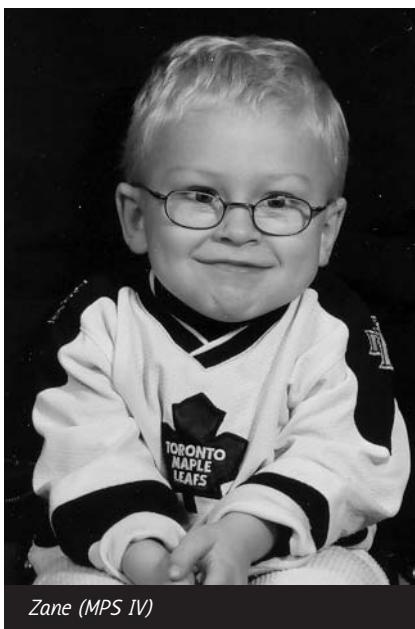
Travel is a challenge for any family with children. Traveling with a child with MPS can be even more of a challenge. But with some preparation, a pinch of optimism and maybe a Barney tape or two, a vacation can be successful. Remember to re-evaluate your travel strategies as your child's illness progresses. A trip you may have made a year ago now might not be possible. Take breaks during long trips and give your child one-on-one attention. If your child is prone to wandering, ensure that her clothing has labels sewn into them with contact information. It is very easy to become separated in a crowd.

WHAT TO PACK

- CD and/or DVD player
- toys and snacks
- stroller
- pillows (for sleeping as well as to protect your child from thrashing), and extra slip covers
- hand sanitizer
- diapers/wipes
- changing pad
- extra sets of clothes
- sippy cups
- specialized utensils
- juice/water
- chew toys
- other toys that, if thrown, will not break or harm someone in its path

AIRPLANE TRAVEL

If you are traveling by air, inform the airline that you'll need boarding assistance, and explain that your child will need/be in a wheelchair or stroller. Call the airline a day or two before your flight to confirm that your requests have been processed. Arrive early so you can find good seating close to the boarding gate.



Zane (MPS IV)

If you must check a wheelchair, make sure your name and address are clearly marked on it. If the wheelchair or scooter is battery-operated, attach the instructions for battery removal. Many airlines provide wheelchair assistance getting on and off the plane.

While on the aircraft, remember the space constraints of the cabin. Take extra care for yourself, your child and those around you when lifting your child between the seat and wheelchair. Many flight attendants are trained to assist with this. Arrange seating on the plane so you and your party are beside and in front of your child

to minimize your child's disturbance of other passengers.

Before landing, remind the flight attendant that you will need your child's wheelchair to be brought to the gate. They will be able to radio ahead. In some cases, it may be easier to rent equipment than to travel with it.



Jack (MPS II) and family

CAR TRAVEL

- When your child outgrows his car seat, try a Gorilla car seat that provides ample support for larger children with special needs.
- Place a waterproof pad underneath your child in the car seat.
- Hang toys from a strap placed over the headrest on the seat in front of your child.
- Take breaks on long trips and walk around and stretch.
- Use a seatbelt cover that your child can open if given a few minutes.
- To prevent your child from unbuckling a car seat, place a sock with the toes cut off over the seatbelt, or turn the seat belt buckle upside down and then hook it.
- Rent a van with extending and turning seats, and/or a hydrolift.
- Travel by Winnebago to visit nearby friends and family.

RESOURCES

Associations

www.mpssociety.org

United States National MPS Society

The National MPS Society exists to find cures for MPS and related diseases. The Society provides hope and support for affected individuals and their families through research, advocacy and awareness of these devastating diseases.

www.mpssociety.ca

The Canadian Society for Mucopolysaccharide & Related Diseases Inc. Provides support to families affected with MPS and related diseases and links to additional resources.

www.rarediseases.org

National Organization of Rare Diseases (NORD)

Unique federation of voluntary health organizations in the United States dedicated to helping people with rare "orphan" diseases and assisting the organizations that serve them. NORD is committed to the identification, treatment, and cure of rare disorders through programs of education, advocacy, research and service.

www.goldinfo.org

Global Organization of Lysosomal Diseases (GOLD)

GOLD is an international collaboration dedicated to improving the lives of everyone with a lysosomal disease. The Web site has information about lysosomal storage diseases and discussion forums.

www.lpaonline.org

Little People of America

Little People of America, Inc., is a nonprofit organization that provides support and information to people of short stature and their families.

www.cord.ca

Canadian Organization for Rare Disorders provides information on rare diseases.

www.cacl.ca

Canadian Association for Community Living (CACL)

CACL is an association that promotes participation and meaningful lives for people with intellectual disabilities.

www.seattlechildrens.org/sibsupp/

Sibling Support Project

This project supports the development of peer support and education programs for brother and sisters of people with special health and developmental needs.

"Sibshops" The Sibling Support Project

Children's Hospital and Medical Centre

P.O. Box 5371 CL-09

Seattle, WA 98105

206.527.5712

Syndrome-Specific Resources

www.mpsldisease.com

Patients, families, caregivers and health care professionals can find information about the disease, support programs and online resources to manage the challenges associated with MPS I.

www.hunterpatients.com

Resource center for the MPS II community to access information about the genetics, diagnosis and management of MPS II, as well as information about the drug development process.

www.maroteaux-lamy.com

Provides education and information about MPS VI for families, educators and healthcare providers.

www.lysosomallearning.com

Learn more about lysosomal storage disorders and the science behind the diseases. Health care providers can get detailed clinical information on lysosomal storage disorders, including incidence, pathogenesis, diagnosis, treatment options and more.

Product Resources & Helpful Information

www.adaptivemall.com

Adaptive Mall

Special needs equipment and products

www.mpsforum.com

MPS Forum is an independent parent-run message board for families with a child with MPS or related disease.

www.dragonflytoys.com

Dragonfly sells specialized toys and allows users to search by type of toy based on the child's developmental level.

www.flaghouse.com

Flaghouse Canada Inc. has a wide range of products for children and adults with special needs, including therapy and positioning equipment, toys, switches and living aids.

www.esmerel.or/canada

Esmerel's List of Canadian Disability Resources is dedicated to Canadian disability resources.

www.easterseals.org

Easter Seals provides direct financial assistance to families for the purchase of equipment needed for mobility and/or communication. Contact Easter Seals and ask for the regional office in your area.

If you have private insurance, you may be able to submit receipts and be reimbursed for part or all of the cost of certain items. Ask your insurance provider – this insurance may be through your employer.

Health

www.canadian-health-network.ca

Canadian Health Network supports Canadian residents in making informed choices about their health by providing access to multiple sources of credible and practical e-health information.

www.cihr-irsc.gc.ca/e/193.html

Canadian Institute of Health Research is Canada's premier research funding agency.

United States Government Resources

www.cms.gov

Center for Medicare and Medicaid Services

www.ssa.gov

Social Security Administration

www.os.dhhs.gov

United States Department of Health and Human Services

www.ncd.gov

National Disabilities Council

www.wrightslaw.com

Wrights Law: Special Education Laws and Advocacy

www.snapinfo.org

Special Needs Advocate for Parents

Canadian Government Resources

Information and Referral Service for the Government of Canada

1.800.622.6232

This toll-free line provides information on how to contact various programs or services offered by the Government of Canada.

www.canada.gc.ca

This Web site provides links to the various departments, contact information and government publications.

www.ccr-aadrc.gc.ca/disability/

Families who have children with disabilities are allowed certain income tax credits. Revenue Canada produces a guide, "Information Concerning People with Disabilities," and has a Web site with information for people with disabilities.

Government Funding

Check with your provincial government or member of Parliament for programs that provide funding for respite care, special equipment, income support and health travel grants.

Sources:

[†] Reprinted with permission of Wendy Bulthuis, MSW, RSW "Siblings – We're Important Too!" (a presentation for the Canadian MPS Society, July 11, 2004)

^{††} Fleitas, J. (2000). "When Jack Fell Down...Jill Came Tumbling After: Siblings in the Web of Illness and Disability." *The American Journal of Maternal/Child Nursing* 25(5): 267-273.

