

# **THE HOME CARE BOOK**

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**A PARENT'S GUIDE TO CARING  
FOR CHILDREN WITH PROGRESSIVE  
NEUROLOGICAL DISEASES**

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**JEANNE M. BORFITZ, RN,C; FNP  
MEREDITH MARGOLIS, PH.D**

*A Publication of:*  
**NATIONAL TAY-SACHS & ALLIED DISEASES ASSOCIATION, INC.**

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#### **PARENTS:**

**YOU ARE STRONGLY ENCOURAGED TO READ THE INTRODUCTORY SECTIONS BEFORE PROCEEDING WITH THE MAIN PARTS OF THE TEXT.**

This handbook is a compilation of suggestions, ideas, knowledge, and even wisdom, generated by a diverse group of individuals who have one thing in common: by necessity, each has become an expert in the care of their child with a progressive neurological disease.

We view this publication as a forum for sharing information and exploring options. In addition to providing pragmatic advice based on direct experience, it is meant to give parents new ways to think for themselves and to use their own creativity, ingenuity and assertiveness. It is meant, as well, to be seen as a work-in-progress. The pages that follow represent a beginning. We hope, over time, to add new insights and generate new ideas. To do so effectively we look to you for reactions and suggestions.

This care manual is intended to offer some practical assistance for those who have chosen to care for their children at home. It is written for parents who are ready to deal with explicit information and are seeking answers to the question: "How do I manage all of this?"

*We are aware that some of the information may be difficult to read. You may not be ready to know "too much." You may find that it is easier to skip around and read just the information that pertains to your child "now." It is important to recognize how much you are ready for and to proceed in a way that is comfortable for you.*

*"We were given names of parents in the area who were caring for children with this type of disease. In the beginning we rejected this type of support. We were in a state of disbelief. We did not know what the future would hold and we were afraid to talk with others who knew too much."*

**"WE TOUCH  
EACH OTHER,  
AND TEACH  
EACH OTHER.  
PART OF THE  
HEALING IS  
TO SHARE."**

The information in this book is not intended to be “disease specific,” but rather to focus on the wide spectrum of problems affecting children with any of the progressive neurological diseases. Although it is written by National Tay-Sachs & Allied Diseases, Inc. parents, it can be a useful resource for any parent caring for a neurologically impaired child. While much of the information is written with the younger child in mind, some can be applied to the older child or adult as well. In addition, it is important to keep in mind that the age of onset, progression and life expectancy in different progressive neurological diseases vary significantly. Your child may never reach or may have never reached the stages that are discussed. Therefore, some of the suggestions may not be applicable to your child’s condition.

This publication is *not* a substitute for professional medical care. It is meant to augment the information, guidance and care you and your child receive from your physician and other health care professionals. Ultimately, the most important goal of this publication is to let parents know that they are not alone. If we can all somehow reach out and help one another, then we are doing something significantly greater than surviving our own tragedy.

*“Isolation is one ‘condition’ that we can do something about.”*

We can also be comforted by the fact that through this experience of helping one another, our children have made a major contribution. **This is part of their legacy.**

All of the information contained within this publication has been contributed by parents who have lived the day-to-day experience of caring for a seriously ill child at home. Our primary goal in organizing the material was to make it readily accessible and understandable to other parents.

In these pages we discuss both the medical issues you may face in caring for your child as well as the psychological and social aspects of this experience. The medical section is presented in a “systems” format, according to the body system one is dealing with. This was done to make it easier for parents to target the specific information they may need at any one time. The “cardiac” system was purposely excluded as heart problems do not commonly affect our children.

Where pertinent, we have included a sampling of anecdotal information and suggestions from parents about specific treatments, medications and equipment they have found to be particularly useful. We do not endorse any individual product or procedure and are aware that these suggestions represent only a small fraction of the range of therapeutic options that are available. The health care professionals with whom you work will help guide you in developing an appropriate treatment plan for your child.

Throughout the text, you will notice that we have also interspersed personal quotes from individual parents. We do so in an effort to explore the wide spectrum of thoughts, feelings and reactions surrounding the experience of living with and caring for a child with a progressive neurological disease. Each individual handles his or her situation in a unique and personal way. We as parents need the freedom to feel our feelings and register our reactions. By reading other parents' diverse reactions, we hope you will come to understand that there is no right way or wrong way to handle this challenge that life has set before you—just your way.

While, by necessity, some of the material presented in this publication is technical in nature, we have attempted within the text to define and demystify the medical terminology. In addition, we have included an Appendix with a Glossary, a list of Medical Abbreviations, and a Resource List, all of which we hope will further assist you in understanding and utilizing the information provided.

**Note:** *The names of all our children in this publication have been changed to maintain confidentiality.*

We would like to thank everyone who made this book possible. Many thanks to the wonderful nursing, recreation, occupational therapy, and physical therapy staff at Northampton Nursing Home for your input. To Allen C. Crocker, M.D., thank you for your time in reviewing this material as well as your valuable comments and suggestions.

Most of all, we would like to thank all of the NTSAD parents who responded to our questionnaire. Your input was invaluable to the writing of this book. We are indebted to you for your honesty in sharing your personal stories and including your practical knowledge and experiences.

**PART** **1** **ONE**  
*Psychological and Social  
Issues Facing Parents and  
the Family*

**COPING**

Caring for a child with a fatal progressive neurological disease is an immense challenge which requires energy, ingenuity and all the coping skills one can muster. For many parents, as painful as the child's eventual death is, the various steps along the journey—from hearing the initial diagnosis to watching the child's inevitable decline—can be equally difficult.

It is probably true that you and those close to you will “never be the same” after going through this experience. It is also true that this does not have to be a solely negative change. Positive changes can and do result from extraordinary situations.

**“PARENTING A SPECIAL-NEEDS CHILD CAN BE, AT TIMES, AN ALMOST INSURMOUNTABLE CHALLENGE, AND THE STRAIN OF CARING FOR THE CHILD AT HOME IS DIFFICULT FOR ANYONE, OTHER THAN THE PARENTS, TO REALIZE.”**

*“Karen’s life was far from normal in the number of her days or the extent of her abilities. Yet I have often said she introduced me to some of the most exceptional people I have ever known. They, and she, enriched my life in many ways.”*

**WHO TAKES CARE OF THE CARETAKER WHEN THE CARETAKER’S TAKING CARE?**

Probably the most important thing to remember when trying to cope with the care of your child is that you need to take care of yourself, too. Your sick child will not benefit from having an exhausted, burned-out parent; nor will the other members of your family.

So, what should you do? On a regular basis make some time just for YOU. It doesn't matter how much time you have. Even if it's just a few hours each week, getting away from your responsibilities at home will replenish your energies. Do something that you enjoy: go for a walk, go to a movie, have lunch with a friend. You will be amazed at how revitalized you will feel after such a simple activity, and how much better you'll be able to cope with your situation when you return.



**RESPITE CARE: WHEN A FEW HOURS ARE NOT ENOUGH**

Sometimes, however, getting away for an hour or two is simply not enough. At these times, respite care is a good option to consider.

Generally, respite care is care provided for your child outside of your home for periods ranging from several days to several weeks. Today, some communities have a nursing facilities that can provide respite care, but it is best to check out the availability of these services before you actually need them. If you have the choice of more than one facility, select the place that “feels best” to you. If you are having difficulty in locating a nursing facility that offers respite care, contact your doctor, Visiting Nurse Association, local hospice, or other parents in your area.

And, if no formal facility exists in your community, don't lose heart. Several families have been able to develop “creative and informal” ways of finding respite care. We have heard of groups of families forming to help parents take care of their special children. Relatives may also be able to lend an occasional hand.

*“Respite is like a shot of adrenalin. It allows you to regain your energy so that you can get through all of this.”*

Having located a place or source for respite care, the next step is to make the transition as easy as possible for you and your child. One parent suggests writing a “Care Plan” to give to those who are temporarily caring for your child. This Plan might include: pertinent medical information; instructions regarding playing, feeding, comforting, sleeping; and helpful hints for solving specific problems. If possible, use a computer to record your Plan so that you can update it easily.

*“When I first decided to leave David at a nursing home for extended respite, I knew that giving a report to a nurse and filling out some general information forms were not enough for me. I had to know that certain crucial information was accessible to all who cared for my child. Only then could I feel comfortable about leaving. With this in mind, I developed the Parent Written Care Plan.”*

#### **RELATING: YOU TO OTHERS AND OTHERS TO YOU**

Many, if not most, relationships undergo a significant change as a result of the birth of a child with a progressive neurological disease. The reality is that caring for your sick child is an extremely time-consuming activity, and friends and family may not be able to understand the scope of the demands that have been placed on you. Many people may seem at a loss for words. You, too, may feel uncomfortable sharing what you and your child are going through. As a result, you may find yourself cut off from the support of others around you.

Experience shows, however, that families who stay involved with the outside world seem to fare better than those who isolate themselves. If at all possible, try to maintain your outside connections. Keep in mind that those around you may really want to help but simply may not know how or what to do. You will probably be able to sense who among your friends and family is able to be there for you; don’t be afraid to reach out and give them a chance to help.

*“Parenting a special-needs child can be, at times, an almost insurmountable challenge, and the strain*

*of caring for the child at home is difficult for anyone, other than the parents, to realize.”*

*“Due to the level of involvement Michael’s care required of us, we kept a low profile. Many of our relationships were cut off, as a result of our own doing or the other persons involved. Learning to deal with people including friends’ and families’ reactions to his ever changing and deteriorating condition was painful for us, as parents.”*

*“As Michael’s situation worsened, we were fortunate to have a few friends and relatives that were supportive and were not afraid to discuss or deal with our situation.”*

For many families, grandparents are an invaluable resource. While grandparents, too, are mourning the loss of their dreams, many have found that helping their own children through this difficult time has made them feel useful and needed. When grandparents are able to spend some time taking care of their sick grandchild, parents are better able to use that time to be together or to be with their other children. If you have parents or in-laws nearby, and they are willing to be involved, call on them. They can be a wonderful source of support to you.

*“Kate loves hard surfaces (like my parents’ kitchen table). The smooth surface enables her to manipulate herself better. My parents will sit on either end of the kitchen table (the long way). They put Kate on her stomach. They put their hands open on the bottom of her feet and she will push off them and repeat this until she ends up at the other end of the table. Then she repeats the other way.”*

#### **WHEN YOU’RE FEELING OVERWHELMED**

Even with the most supportive family and friends, sometimes you will feel overwhelmed. Caring for a child with a progressive neurological disease is stressful, emotionally painful, physically exhausting, and sometimes, just overwhelming. At those hard-to-cope times, it is especially helpful to be able to talk to a supportive listener. Good listeners can be found in many places. Some parents find it particularly comforting to speak to other parents. They will tell you that it’s simply a relief to be able to vent your feelings to someone who knows exactly what you’re going through. Other parents seek professional counseling. In fact, many parents have found

therapy to be extremely helpful at some point (or several points) in their child’s life.

Whatever avenue or avenues you choose to take, recognizing when you’re feeling overwhelmed and seeking appropriate help is one of the most important coping mechanisms you can develop.

*“I spoke many times with a social worker from Boston Floating Hospital. He was very supportive. He phoned often and when I brought Tim to Boston every few months, we would speak at length. [Tim’s sister] saw a child psychologist after the death of her brother to make sure that she was dealing with it all. I most certainly believe that this type of support is needed. We all need help at some time or another.”*

*“It was always therapeutic for me to talk to another parent in the Parent Peer Group of NTSAD. So many times we would be faced with making a decision and would feel like we had virtually no guidance from the doctors. It was at these times that discussing other parents’ solutions to similar problems was particularly helpful.”*

#### **MARITAL ISSUES**

Any traumatic experience which impacts on the two members of a couple can affect the marital relationship. While, in some cases, sharing the tragedy of having a child with a serious illness can bring a couple closer together, more often the experience places a significant strain on the marriage. Problems present in the marriage before the birth of a child with a progressive neurological disease may well be exacerbated with the added burdens on the family.

One common source of marital difficulties is anger. It is a natural reaction for parents of a child with a progressive neurological disease to feel angry: angry at God, angry at the doctors, angry at just about anyone.

*“I didn’t feel guilty about our daughter—I felt angry. I waited until I was married to have a child (unlike all of my siblings.) I planned to have this child. I was going to be the best mother in the world—and I felt cheated. I also became angry at every healthy child’s parents.”*

Sometimes this anger becomes misplaced and is taken out on those closest at hand; understandably, that closest person may well be the

spouse. It is important to keep in mind that the anger is often a reaction to the loss, or the impending loss, of the child. If each member of the couple can recognize the underlying feelings of frustration and helplessness, and hear past the anger, they may be able to work toward pulling together rather than tearing each other apart.

*“I sometimes have to remind myself that despite everything that is happening to us right now, we still have each other. That’s why we married in the first place—to be together.”*

Disappointment and unmet expectations are another cause for strain in the marital relationship. Sometimes one parent may feel that the other “just can’t deal” with a seriously ill child. She or he may feel overburdened and abandoned. The unfortunate reality is that both parents are hurting, leaving limited energy available for comforting each other. When one needs comforting the most, the other may be least able to “be there.”

*“When we found out about Elizabeth he dove head first into drug and alcohol abuse. I took care of everything with little financial support from him and no emotional support.”*

*“There are times when I want my husband to leave because rather than offer me support, he adds to my pain. I feel like I’m going through this alone.”*

Finally, numerous couples express concern over their sexual relationship. The emotional pulls and physical demands of caring for a seriously ill child over an extended period of time often impact on a couple’s desire or ability to maintain their sexual relationship. Sometimes just recognizing that this is a common reaction to particularly stressful situations can help couples in understanding and dealing with these issues.

*“A wedge seemed to come between my husband and me, come bedtime. My thoughts focused only on my ill son and I could not seem to enjoy my husband’s sexual advances.”*

*“Laura was up a lot in the middle of the night. I would get extremely tired and lost interest in lovemaking.”*

It seems that the best way to protect your marriage during this difficult time is to make every effort to keep the lines of communication open. This means that each of you is free to share feelings, thoughts or concerns without fear of blame or of overwhelming your partner.

At the same time, each may want to recognize that some of your “comforting needs” may best be met outside of the marriage. Family members, friends, clergy, counselors, or other parents can be an alternative source of support and caring during this stressful time. Parents must also respect each other’s differences and recognize that no two people mourn in the same way. While you may feel better being very much involved in your child’s care, your partner may need to be more removed.

There is no right way to deal with a tragedy such as this, and while you may not always be “in sync,” if you talk about your feelings and concerns openly and explore solutions together, you could emerge from this experience with a stronger, more understanding relationship.

*“How do I cope? I talk and talk and talk about Sandra and my feelings to any and everyone who’s interested.”*

*“The greatest preventative measure against marital stress, for us, was to talk, talk some more, hug, and every once in awhile, have a really good, earth-shattering cry.”*

## SIBLING ISSUES

Caring for a child with a progressive neurological disease requires immense time and energy. If you have other children as well, life often feels like a juggling act with too many balls in the air. Many parents worry about the toll that their sick child’s condition will take on their healthy children. Surely they will be affected by this experience just as you will be. However, with guidance and love, many families find that their other children emerge with a sensitivity and caring about others that makes them very special indeed.

Based on their own experiences, parents of children with neurodegenerative diseases have offered some suggestions regarding the care of siblings.

- Try to include your healthy children in what’s happening. When feasible, even allow them to participate in their sibling’s care.
- Be honest and answer questions directly. If you are comfortable talking about your sick child and his or her illness, chances are your other children will respond similarly.



- Try to reassure your healthy children that they are OK: they cannot “catch” this illness like other diseases; and, equally important, they had no role in causing it, despite any angry feelings they may have had towards their sibling.

- Resist temptations to “deify” your sick child because he or she never “misbehaves” as your healthy children sometimes do.

*“One parent told me about her healthy son and her son with Tay-*

*Sachs disease. The healthy son felt that he was always getting in trouble, while Andrew was getting so much more attention and never did anything wrong. Andrew’s mother’s solution to this problem was to “admonish” Andrew for doing things such as coughing or “eating” too rapidly through his G-tube. The sibling rivalry died down when her healthy son felt on a more equal level with his brother.”*

Although your time and energy are at a premium, it is important to remember that your other children

need you too. They need to feel free to be themselves and not feel constantly pressured to be “good” because Mom or Dad has all that he or she can handle. Open communication is especially important now between you and your other children.

The reality of these diseases is that sometimes you may not be spending as much time with your healthy children as you are with your sick child. Give your well child the freedom to discuss his or her feelings about this and other issues. And give yourself the freedom to ask for help if you feel you need it. Many families find that professional counseling by a family therapist during this stressful time can be an important support in helping you and your children deal with the loss of a child or a sibling as well as the loss of a more “normal” family life.

### COMMUNICATING WITH HEALTH PROFESSIONALS

Establishing a good working relationship with the health care professionals who are involved in your child’s care is essential if you are to manage your child’s illness effectively. Being able to talk frankly with your child’s doctor

about the medical issues that arise will ultimately lead to better care for your child, as well as help you to feel more in control. So to begin with, you will want to choose a doctor with whom you feel comfortable.

*“One of the first things I did after Kathy was diagnosed was to find a new pediatrician, someone I felt I could talk to and who would be there to help us through. I actually called and interviewed several before I made my choice. I was very straightforward about my needs and expectations. Not only did I find a good doctor, the experience was a very empowering one for me. I felt I had taken control in what up until then had seemed a totally out-of-control situation.”*

### ESTABLISHING A GOOD WORKING RELATIONSHIP

What does a good working relationship mean? It means, for example, that you feel free to ask your doctor for the information you need in terms that you can understand. It means, as well, that you feel free to tell your doctor what you know about your child. In reality, you are the expert on your child; no one knows more about your child than you. In fact, many

of these progressive neurological diseases are so rare that your doctor may not have seen or treated a case before. Ultimately, the best care becomes a collaborative effort between the family and the medical professionals.

### HANDLING COMPLEX INFORMATION

In the course of caring for your child, you may well be learning to perform procedures that are new to you. Make sure that you understand the instructions you are being given regarding your child’s care. If you are unsure, don’t hesitate to ask the health care provider to explain or demonstrate again. Some parents find it quite helpful to take along a tape recorder during doctor’s visits so that they can refer to the taped instructions later on.

*“Very little information was given to me at the time of the initial diagnosis. In fact, the information that was available was so involved that I had to use a medical dictionary to understand its contents.”*

*“When we received the initial diagnosis we were too stunned to ask the right questions or even remember exactly what was said. We called later that evening and asked to have another conference, this time bringing a tape recorder. We felt that*

*we could relax a bit and really listen better since we weren’t so intent on memorizing every word. This technique worked so well for us that we got in the habit of taking a recorder along on most doctor’s visits.”*

### FORGING A PARTNERSHIP

One area of extreme importance in maintaining an effective working relationship between you and your health care provider is ensuring that you are involved in the decision-making process regarding your child’s care. Many medical problems can be handled very differently, depending on your feelings and those of the health care professionals with whom you are working. It will empower you to be an active participant in the decisions that are being made and may well help you later on with the reassurance that you did the very best you could do for your child.

*“I had a very serious problem at one point. The doctor had ordered, without my knowledge or consent, to withhold food from my son. It seems that Jimmy had thrush, which had been overlooked. The doctor thought that Jimmy had lost his ability to swallow and ‘thought it best just to let him go.’ I demanded the doctor return to the hospital to rescind that order. I was outraged*

*that such a decision was made without my consent.”*

One problem area that can arise between parents and health care professionals relates to the terminal nature of these diseases. Most doctors and other medical workers are used to aggressively treating any medical problems that occur. For many parents, however, the major focus is not to prolong life but to make their child’s days as comfortable as possible. Sometimes it is hard for health care professionals to “switch gears” and take an approach more focused on comfort than cure. Open and ongoing communication about this issue is important.

*“Sara had gotten to the point where she had a lot of mucus in her throat most of the time. One of her nurses kept aggressively suctioning her to the point where she would bleed, moan, and try her best to turn away. Despite my protests, the nurse continued, explaining that she needed to suction like that to prevent more serious illnesses. It wasn’t until I finally asked Sara’s doctor to write it in her records that this nurse was comfortable with suctioning Sara less frequently, and more gently.”*

Keep in mind that the doctors and other health professionals you are

working with are there to **help you** care for your child. If you feel that you are spending too much time fighting, or that you do not have the control over your child’s care that you want, perhaps it is time to search for other medical personnel. Having caring physicians and other health care professionals “in your corner” can make a world of difference.

*“If you ever need to talk to your doctor, stand between him and the door. If he tries to walk around you, change doctors.”*

### WHEN YOU NEED OUTSIDE HELP

Caring for your child at home can be a rewarding experience, assuring you that your child is living the best life possible under the circumstances. It can also, however, be demanding, stressful, and emotionally draining. There probably will be times when you feel that you simply cannot care for your child on your own. At these times, several options may be available to you to help “lighten the load.” Some of these include: respite care (see Coping), home health care, hospice, and placement. (*Consult Additional Resources Section in the Appendix.*)



When evaluating the alternatives that are available to you, it is important to check whatever insurance coverage you may have. Unfortunately, insurance policies are sometimes written in such vague or complicated language that it is difficult to know exactly what help might be provided. Or, as is often the case, a policy may provide for quite extensive hospital stays but have no specific provisions for home or hospice care. If you are faced with either of these scenarios, it is important for you to sit down with an executive from your insurance company and talk frankly about what lies ahead. It is possible

that you can work out a solution that is of mutual benefit for both you and the insurance company.

*“My insurance provided generous hospitalization benefits. Since I was intent upon keeping Maggie at home, I asked that instead of paying exorbitant hospital fees, would they consider reimbursing some home care? After numerous discussions, the insurance people finally realized that it would be cheaper **for them** if Maggie was at home rather than in a hospital. A side benefit, of course, was that we would be happier too. They agreed to pay for as much home nursing care as was needed.”*

#### HOME HEALTH CARE

Home health care aides can help in numerous ways. A health care aide can free you to do the things you cannot get to when you are caring for your child. An aide can provide a “mini-medical check-up” in your home. No matter how well you know your own child, there are times when a nurse or other health care professional may detect a specific medical problem and may be able to suggest helpful solutions. For some parents, just having an experienced health care professional in the home alleviates some of the stress, allowing them to feel more comfortable and confident in caring for their child.

It should be noted, however, that parents do give up some privacy when someone else, anyone else, is in the home. This is really a personal preference issue: to some the benefits of at-home care outweigh the loss of privacy; for others, it is preferable to take care of the child themselves, rather than work with another caretaker.

*“Our home aide was a life-saver for me. She was so helpful. Sometimes what was most important was that she just assisted with other tasks around the house so I could care for Ellen undisturbed.”*

*“We had nursing care during the day so that we both could work. Having nursing care for one night a week was a blessing for us, though. Even though our insurance provided for more coverage, we felt that we wanted some family time without anyone else around. Also, we wanted to take care of Andy ourselves.”*

#### HOSPICE AND OTHER OUTREACH SERVICES

Your local hospice program or Visiting Nurse Association may also be able to provide health care assistance in your home. The mission of these organizations is to give help and support to families coping with terminal illness. Again, some families welcome this kind of help, and others may not be comfortable with it.

*“Hospice and my family cared for Josh. We were assigned caregivers for four hours a day, two days a week. They were trained professionals who took care of Josh. They would run and pick up prescriptions, run to the ER with me and would assist any way they could. They helped us with the funeral arrangements for Josh and have continued to support our family even today. They became our friends, a part of*

#### PSYCHOLOGICAL & SOCIAL ISSUES

*Josh’s life. I am truly grateful to them for what they did.”*

#### STATE FUNDED PROGRAMS

Many states have specialized programs in place to help families with children like ours, so don’t forget to check into this possibility as well. Different states provide different benefits, but many can assist you in getting low-cost help in your home, or even providing respite care at no charge. If dealing with bureaucracies sounds like just another headache for you right now, perhaps a friend can help. So many times others ask us if there is anything they can do for us. What a valuable gift it would be for someone to help by researching state-provided services for you. And by all means, be sure to check with NTSAD. Resource files are continually being gathered and updated and are available to any interested parents.

#### PLACEMENT

There may come a time when you consider placing your child in a nursing facility, particularly if you have no insurance covering at-home help, and few other resources or alternatives are available to you. The decision to place a child is not

an easy one, but sometimes it is simply the best one for a family.

*“I just couldn’t handle it anymore. The strains on our marriage and life were unbearable. I’d had enough.”*

*“For me, placement has allowed me to get on with my life. I have recovered from the strain of caring for him at home.”*

*“We have the ideal set-up. The nursing home is near my home and job. It has become a second home for the rest of the family and the staff have become our friends. And the time I spend with him now is just in holding him and loving him.”*

If you are considering placement as the best alternative for you and your child, there are several sources available through NTSAD that may help you in making your decision. It may be especially helpful for you to talk to other parents who have placed their children. Members of the NTSAD Parent Peer Group (PPG) make themselves available to talk to other families of children with progressive neurological diseases. These PPG members are simply parents like you who have lived through or are currently going through a similar situation. In talking with other parents, you can express your concerns and

questions and learn from their experiences. They are an invaluable source of information as well as understanding.

*“Talking with other families who had decided on placement helped me to feel better about my decision. Since I have gone through this experience, I have been able to help other families as well.”*

### FACING DEATH

One issue that all parents of a child with a neuro-degenerative disease must eventually face is the death of their child. While scientific information and medical experts may describe the typical course of these diseases, each individual case is different, and it is often difficult to accurately predict when or how a child will succumb to his or her illness. Several families have faced situations where health care professionals have predicted a child’s death as imminent many times (and sometimes many years) before the actual death occurs.

One way to cope with this uncertainty is to make as many arrangements as you feel comfortable with ahead of time. Decisions such as burial place, type of memo-

rial service and the people you want to be involved can be made in advance and will save you the added burden of taking care of these arrangements at the time of your child’s death. Many funeral homes are receptive to making advance arrangements and can assist you in this effort.

*“I made no funeral arrangements prior to his death. However, I wish I had. I let others pick out the place for him to be waked, and the people who took care of his arrangements were terrible. I guess I would tell others that, as difficult as it is to face, you should try to make some arrangements or choices earlier on.”*

### INTERVENTION: DECIDING WHEN AND HOW

At some time in your child’s life you may have to confront the issue of the “Do Not Resuscitate” (DNR) order. Doctors and hospitals are wary of not doing everything possible to save your child’s life (including performing tracheotomies and utilizing ventilators or respirators) unless you have expressly stated that you do not wish these interventions to be used.

It is extremely difficult to make the decision to use “no heroic efforts” to save your child. Even the decision to do everything possible can be a troubling one. In either case, it helps to have the understanding and support of the health care professionals involved with your child. And in all cases, it helps to remember that these are very personal and individual decisions. There are no right or wrong answers, only your answers, and indeed your answers may change over time. You are free to change your mind and alter your decisions.

*“We have always known that we would do everything possible to keep Tammy alive, never giving up on the hope that research will result in a treatment becoming available.”*

*“We never discussed this [DNR] until one day when he was about 2½, and had to be admitted to the hospital. He was very weak. He was having trouble eating and drinking. We knew at this point it was not possible for him to have any quality of life, and it was at this point we made that decision. It was very difficult to make, and prior to his getting this sick I would not have been able to make that decision.”*

*“I have discussed the DNR with both her pediatrician and neurologist. I don’t know how we’ll feel when actually faced with sticking with this decision. Our feeling in making this decision is that it is selfish to keep her alive by extraordinary means. It is not life to be on a machine and unaware. When Rebecca’s time comes I pray to have the strength to let her go. For what it’s worth, her pediatrician is very good at reminding me of these type of decisions and helping me to stick with it.”*

### WHEN THE TIME COMES

Parents are often curious as to just how a child does eventually die. While textbooks on these diseases state that respiratory infections of some kind are usually responsible, many parents report that at some point, “my child’s body just seemed to stop working. He stopped breathing and didn’t start again.” Interestingly, often a cessation of breathing (apnea) has occurred previously (sometimes numerous times) but there’s “just something different” about the last time. Perhaps the difference is that the parent is more ready and less willing to intervene at that point to

prolong the child’s life. Mercifully, most such deaths are as peaceful as we would wish for our children, and for ourselves.

*“The last two weeks of Daniel’s life were quite different from what we were accustomed to during any other time in his life. Daniel’s digestion completely slowed down to the point where he could not digest his formula or even water for that matter. Danny’s temperature was below normal, approximately 96 degrees, also something we had not seen before. It was during this period that he stopped breathing again, but for the first time we did not intervene. Daniel died at 3 a.m. exactly two months before his fifth birthday.”*

### GUILT

When a child is born or diagnosed with a devastating disease, it is natural to want to “look for someone to blame.” Often, it seems, parents end up either blaming themselves in some way or feeling bad about something they did or did not do for their child.

*“As his mother, I felt so bad I had put Tommy through so many tests. Sometimes I still torture myself*

*with my thoughts. I’ve stopped doing that now. But if I could go back, I would have stopped all unnecessary tests.”*

*“I felt guilt when I was told that one in four were the odds for my children being born with Niemann-Pick Disease. Gregory was my fourth pregnancy, and my first pregnancy resulted in an abortion.”*

*“Guilt was experienced with hindsight that we failed to fully enjoy her more outgoing abilities while she still had them.”*

*“Only rarely did I lose patience and that is what I regret, that I did at all.”*

*“The guilt I have at times is not exercising Kathy enough.”*

As is apparent, the guilt reaction runs the gamut from things over which we truly have no control to our simply being less than perfect people. Whatever the source, rational or not, guilt can be a heavy burden and destructive force. Many parents have mentioned that reading specific books has been helpful to them as they try to deal with and work through their feelings of guilt. Others have been helped by talking over their feelings with other parents, family members, friends, clergy, or professional therapists.

**NOTE TO PARENTS:**

You are strongly encouraged to read the introductory sections of this publication before consulting the following text.

This medical section will hopefully provide you with additional insights into caring for your child. Please keep in mind that the information presented is meant to complement the care and guidance you receive from your doctor and health-care providers. It is not a substitute for professional medical attention.

**HEENT: HEAD, EYES, EARS, NOSE AND THROAT**

**HEAD**

For several reasons, many children with progressive neurological disorders have enlarged heads. An increased head circumference, which is generally noticed during a routine physical exam, may be one of the first indicators that something is wrong.

The first year and a half of life is the period of greatest head growth, because the bones of the head are not yet fused. Head growth begins to slow down as the sutures (lines between the bones) in the head begin to fuse and the fontanelles (soft spots) close.

As the size of the cranium grows, the head becomes increasingly heavy. Holding the child for extended periods becomes tiring because of the weight, especially if your child tries to “extend” (arch) backwards. You may find yourself shifting your holding positions and accommodating for the head size when you position your child for sitting or laying down.

Some positioning problems exist because of the weight of the head and the child’s inability to move. When your child lays on his or her

**“THERE WERE SO MANY THINGS THAT HAD TO BE DONE FOR MY CHILD: MEDICATIONS, TUBE FEEDINGS, EXERCISING AND OTHER NECESSITIES OF DAILY CARE. THEN THERE WERE ALL THE WAYS IN WHICH I COULD ‘STIMULATE’ HIM THAT THE THERAPIST SUGGESTED. THERE WAS SO MUCH THAT I HAD TO DO, WHEN ALL I WANTED TO DO WAS TO SIT AND HOLD HIM AND LOVE HIM.”**

**PART TWO**  
*Medical Aspects of Home Care*



side, the ear must be checked periodically for pressure areas and soreness. Using a sheepskin under your child’s head, with a soft cloth over it to absorb moisture is helpful (see *Dermatology section*).

When buying clothing for your child with an enlarged head, you should look for shirts, sweaters etc. with large or stretchy neck openings to comfortably accommodate the head. It may be helpful to remind well-meaning friends and relatives about this special need if they are buying gifts for your child.

**EYES**

One of the earliest signs of a potential neurological problem is the presence of nystagmus which is a rapid, exaggerated, involuntary eye movement. Although this may be present to a lesser degree with a healthy newborn, exaggerated movement is an indicator of a neurological abnormality.

The presence of a “cherry red spot” on the retina (back of the eye) is a diagnostic sign in Tay-Sachs and some of the other progressive neurological diseases. This can be seen by a trained professional using special equipment for examination of the eyes.

Parents may begin to have concerns about their child's vision, especially if the child is not tracking (following with the eyes) by a certain age, or does not seem to be focusing on a familiar face or object.

Many children with progressive neurological disorders may be visually impaired. They may have partial vision or may progress to a complete loss of sight. A test called "Visually Evoked Potential" can help to diagnose the type of visual impairment your child has. In this test, brain waves are measured while a strobe-type light is flickered in front of the eyes.

Since it is difficult to measure precisely how much vision a child has, it is important to provide lots of visual stimulation for your child (*see Stimulation, Play, Communication section*). Many local voluntary and governmental agencies provide wonderful services and support for visually impaired children. You may want to investigate the services in your area.

The most common eye infection in all children is conjunctivitis ("Pink Eye"), which produces yellow

mucus and a red eye. An eyelash can also cause redness, so you may consider looking for a foreign body irritating the eye before you assume it is conjunctivitis. Conjunctivitis may be an early signal of an ear infection, so it is important to have any persistent redness of the eye evaluated. Be aware that the discharge may be contagious, so precautions need to be taken not to infect others. Antibiotic drops or ointment are generally used to treat conjunctivitis.

#### EARS

Ear infections are very common in all children. While these infections are not necessarily more frequent in our affected children, they do occur and require attention.

Middle ear infections (otitis media) have the potential to cause hearing loss. It is especially important to maintain optimal hearing function in our affected children since so many of them are or become visually impaired.

The more obvious signs of an ear infection are fever and discomfort, but with our children it is helpful to learn some of the early signs. As mentioned above, an ear infection

may first present as a conjunctivitis. Other early signs may be: the persistence of a cold with discolored nasal mucus for more than one week; unusual fussiness; loose stools; and vomiting. Middle ear infections are treated with oral antibiotics.

External infections of the ear canal (otitis externa) are less serious, but are perhaps more frequent with our children because of chronic drooling which can enter the ear. It is important to change moisture laden cloths under the face and ear frequently. These infections can be painful; they may or may not cause fevers. External ear infections are treated with ear drops.

#### NOSE

Because these children cannot blow their noses, they often have thick dried secretions in the nose. Carefully using a nasal or ear syringe with some saline solution will help to remove the discharge. If your child has persisting nasal drainage which is discolored, he or she should be evaluated for an ear infection, as mentioned above. This may also indicate a sinus infection, which is treated with an oral antibiotic.

#### THROAT

Tonsillitis should be considered if your child is uncomfortable, has a foul odor on his or her breath, or more pronounced drooling and fever. When the tonsils are greatly enlarged, they have the potential for blocking the airway in children who have little or no head control. One family used a neck brace to support their child's head during episodes of tonsillitis. Suctioning may be indicated to help to handle secretions. Tonsillectomy is sometimes indicated in cases of recurring tonsillitis. Tonsillitis may be caused by the streptococcal bacteria ("strep" throat) which requires antibiotic treatment.

#### DENTAL CARE

Regular dental visits and good oral hygiene are very important. Try to find a dentist in your area who has experience with special needs children. The Americal Dental Association may be able to provide you with a list of dentists with this specialty.

Teeth and gum problems are common in children who do not chew or use their oral muscles in a normal way. Oral care should include brushing the teeth at least twice a day with water or mouth

wash. If you decide to use toothpaste, use it sparingly. Try to avoid foods with sugar if your child is eating by mouth. Dental flossing should also be attempted on a regular basis, if possible.

If your child is not being fed by mouth, you should regularly moisten the mouth with a toothbrush or swab. Children often enjoy chewing on the toothbrush; if they are not orally fed, this may provide the only opportunity to chew. A child can chew on a cold wet washcloth as well, and the cloth can be rubbed on the gums, tongue and teeth which can be especially comforting during teething. Stimulating the mouth on a regular basis helps to maintain the swallowing reflex, which enables the child to handle his or her own secretions for as long as possible.

One common problem among these children is the presence of a high arched palate (roof of the mouth). Secretions and food can get caught in this area and are not easily removed with the tongue. Care should be taken to see that the area is clean and moist, as dryness can cause fissuring (cracking) of the palate, which is painful and can be difficult to heal.

Lemon and glycerine swabs or Toothettes are useful in cleaning the palate. If there are dried crusty secretions, you can try a solution of half hydrogen peroxide and half water on a swab to clean the mouth and palate.

Enlargement of the gums (gingival hypertrophy) may occur if the child is on long term Dilantin therapy (an anticonvulsant). Gentle brushing of the gums is important to keep them healthy.

#### RESPIRATORY SYSTEM

##### COMMON RESPIRATORY PROBLEMS

Lung problems go hand-and-hand with the progressive feeding problems that develop in most children with neuro-degenerative diseases. As these children begin to have difficulty swallowing, their gagging and coughing can lead to "mini" or major aspirations (inhaling liquid into the lungs) and bronchospasm (spasm in the windpipe). These conditions can cause wheezing, which is a whistling sound made by air going through a narrowed passageway. In addition, these children often have a lot of mucus in their lungs resulting in congestion.

Two families mentioned medications which they used to help decrease the amount of secretions in the mouth. They are Scopolamine® patches (an antihistamine used for motion sickness) and Artane® (an antispasmodic). Ask your health care provider about these options. Both families felt that these medications helped a great deal.

*“John has been like a new person since he went on Artane. The side effect of Artane is dryness of the mouth and it has controlled the drool and phlegm very effectively.”*

Children with neuro-degenerative diseases commonly have a condition called reflux, which causes the stomach acids to be regurgitated up into the esophagus. Reflux is a common precipitator of respiratory problems and can also cause wheezing (see Feeding section).

These children may also develop asthma, a condition in which there is spasm of the airways, causing wheezing. In addition to the previously mentioned physiologic triggers (reflux, aspiration, etc.), asthma may also have environmental causes. Typical allergens such as dust, mold, cats, dogs,

etc., can aggravate your child’s asthma and may further complicate the picture.

Of course the main respiratory condition that is on all of our minds is pneumonia, which can be very serious in our children. Pneumonia is a lung infection in which sections of lung develop severe inflammation. Children with progressive neurological disorders are certainly at increased risk for developing pneumonia with any respiratory infection because of their relative lack of mobility: they often have poor cough reflexes and more shallow breathing patterns because of decreased activity levels. Swallowing difficulties also make them more susceptible to aspiration, which can cause pneumonia.

*“With every cold or flu, with every fever, I ask myself, ‘Is this it?’ It is on my mind until he is well again.”*

Physical therapy of the chest (Chest P.T.), and frequent position changes (e.g. turning side to side in bed and alternating with time sitting in the chair) are beneficial in dealing with congestion. These measures loosen the chest secretions, thereby stimulating the cough reflex. Chest P.T. involves clapping and vibrating the hands over the lobes of the lungs.

Injury can be caused if the percussion (clapping) is done incorrectly, so it is necessary to get accurate training, and to feel comfortable about what you are doing. During this procedure, the child is held downward and prone (face down). This position helps to facilitate the removal of the mucus. Elevating the foot of the bed (postural drainage) to help promote drainage of secretions can also be beneficial. The use of an expectorant (e.g. Robitussin®) was also recommended to help loosen secretions in the lungs.

*“When my son is congested, I encourage him to cough by saying ‘cough’ and then I mimic a cough. He eventually does cough, and I give him lots of positive reinforcement. Who knows if he really gets it. I’d like to think that he does.”*

#### SUCTION AND UPDRAFT MACHINES

Because of the ongoing potential for respiratory problems in our children, two important pieces of equipment should be considered for the home: a suction machine and an updraft machine.

If a child does have intermittent wheezing, he or she may need an updraft machine, which delivers



bronchodilator medication (opens up the airway) in an efficient way.

Having a suction machine at home, although it may actually never be used, will give you peace of mind and may save you an unnecessary, frantic trip to the emergency room. A suction machine is especially convenient to have around during colds and to suction out the extra secretions from the back of the cough that your child may have difficulty swallowing. Make sure

that you receive thorough instructions on how to suction and that you are comfortable with the procedure.

#### GASTROINTESTINAL SYSTEM

##### FEEDING AND NUTRITION

Children with progressive neurological diseases almost always develop difficulty with feeding. Initially, they may be able to breast

feed or take a bottle; although they may have difficulty “latching on” and the mouth may have to be held in a certain way to improve the grip.

It is important to monitor the caloric and nutritional value of foods that are given orally. Baby food may not provide enough calories as the child grows. You may have some questions about your child’s nutritional condition: Is my child’s weight O.K.? Is my child getting enough liquid? How many calories a day does my child need? Having a nutritionist available for periodic consultation is helpful for both oral and tube-fed children.

Oral feedings can be a joyful, special time. This may be a time for lots of vocalizations, especially if your child is very hungry. This can be a good time to encourage language development by increasing the child’s understanding of certain words and his or her anticipatory response. For example, saying “ready” and repeating the names of favorite foods on a consistent basis before the spoon is introduced are good techniques.

*(See “Stimulation, Play, Communication” section.)*

Feedings can be incorporated into family meals as a special time for all to be together. The children enjoy hearing familiar voices and seeing familiar faces, if they are able. This encourages sibling bonding and helps to establish a sense of “normalcy.”

#### POSITIONING FOR FEEDING

Since these children generally do not progress to sitting up on their own, they are usually fed in our arms or in their strollers or other seats that give support to the head and neck. Correct positioning prevents the children from arching or extending, and helps to “break” their high tone (muscle tension), enabling them to be more relaxed during feeding.

They need to be fed in an upright position and the head needs to be in midline, not extended back, in order to help prevent aspiration and reflux. (*See G-E Reflux section.*) When not being held, your child can be propped up to a sitting position with lots of pillows and stuffed animals for comfort and support.

Accepting the fact that your child needs special equipment is a gradual process. Because very young children can “get by” with

strollers for transportation for quite some time, the use of a special seat for feeding (e.g., a Tumble Form seat) in a restaurant may present the first situation in which you will face the issue of dealing with “special equipment” in public. (*See Dealing with Special Equipment section.*)

*“I felt like everyone’s eyes were on us in a restaurant, saying ‘What’s wrong with that child? He should be sitting up by now.’ In the beginning, I used to block out their stares. It gradually got easier.”*

#### FEEDING DIFFICULTIES

There may be a period of time during which your child eats and drinks normally. Eventually, problems with swallowing affect the eating process, and taking anything by mouth becomes difficult. The child may start to cough or gag during feedings. He or she may develop a poor suck. He or she may begin to aspirate food which can cause pneumonia, or have “mini” aspirations which can precipitate bronchospasm, and cause wheezing. (*See Respiratory section.*)

The mouth muscles are designed for sucking during infancy, and as the child grows, the muscles adapt

for eating. In the neurologically impaired child, the muscle patterns may never progress past the sucking stage.

Liquids generally present the first feeding problem. It can be helpful to thicken liquids with gelatin, baby cereal, oatmeal or bran flakes. Thick It, a commercially available product used to thicken fluids, may also be used. When eating strained foods, the children often will begin a “chewing” style of eating, again because of lack of coordination. It is helpful to use a vinyl coated baby spoon to protect your child’s teeth. Helping them to purse the lips by squeezing the cheeks together may make chewing and swallowing easier.

Your child may open his or her mouth more easily if he or she smells the foods first (and you can also encourage understanding of language if you say what the food is prior to introducing it). Another way of getting the mouth open is to apply firm pressure on the corner of the jaw and manipulate the area (also a useful technique for dental care). You can encourage chewing by placing your hand along the lower jaw bone and moving it up and down.

To desensitize a hypersensitive mouth and thus aid in swallowing, one family suggests playing inside and outside the mouth with toys and a NUK® brush (a dental stimulator with soft plastic protrusions on the end of it). A Toothette (foam pad on the end of a stick used for dental care) can be used as well.

Another family suggests using a nipple designed for babies with cleft palate. This is available free of charge from Mead-Johnson. One parent suggests a NUK cross-hatch nipple. Cutting off the top or widening the opening of a regular nipple also works. The child can then “suck and chew” out the thickened liquids. A First Years Infa-Feeder was also recommended for feeding thickened meals. Evenflo makes a similar product.

Invariably, the entire feeding process eventually becomes very time consuming. The children need to be fed slowly to prevent aspiration, and they tend to eat slowly. Feeding can be very tiring for our children, and they may need to rest during the meal. By experience, we have learned that it is also important to feed them on time. If the child is very hungry, the resulting irritability may increase tone (muscle

tension) and reflux (see below) and thus further complicate the process.

Very often the feedings are done with the child in our arms. Although this is a very special bonding time, the increased length of the feedings, and the child’s increased weight can combine to make this method of feeding an exhausting experience for parents. Having another adult around to help with feedings makes the process easier.

#### G-E REFLUX

Reflux is a common problem affecting many children (also called G.E.R. or gastro-esophageal reflux). In reflux, the acid from the stomach regurgitates up the esophagus because of poor muscle tone of the sphincter muscle at the end of the esophagus.

In children with progressive neurological disorders, who are already beginning to have difficulty swallowing liquids, this chronic regurgitation of stomach acids further compounds the problem.

Chronic reflux causes irritation of the esophagus resulting in “heartburn”, an inflammation of the esophagus (esophagitis) or a much more serious problem, ulcers of the esophagus.

Pain from reflux may result in irritability and an increase in muscle tension. As mentioned earlier, irritability (e.g., from hunger) seems to precipitate reflux, so it is important to anticipate the child’s needs as much as possible to avoid this cycle of complications.

Although reflux can be somewhat controlled by certain medications, feeding in the upright position, and by elevating the head of the bed on blocks, it is apt to remain a chronic problem. Positioning your child on his or her side (sidelying) when in bed, is the best bed position to help prevent aspiration from reflux or vomiting.

#### NASOGASTRIC AND GASTROSTOMY TUBES

Chronic reflux, exhausting and difficult feedings, and problems with dehydration from lack of adequate fluid intake are often the precipitating factors leading to the decision to begin tube feedings. A video esophagram can be a helpful test in determining whether or not tube feedings are necessary. In this test, an x-ray “video” of the esophagus is taken while the child is eating.

There are generally two methods of tube feeding to choose from. Some parents choose a nasogastric tube (N-G tube) in which a tube is inserted into the child's nose and threaded into the stomach.

*"We chose this option because we felt that we did not want to put him through the gastrostomy operation since his health was so bad."*

The advantage of the N-G tube is that it is a non-surgical procedure and can easily be removed (it is usually replaced once a month). The disadvantage is that the N-G tube can be irritating to the nose and can involve some risk (the tube may inadvertently slip into the lungs). It needs to be taped to the nose to prevent dislodging. It is essential to check for proper placement prior to starting a feeding by placing the end of the N-G tube in water to check for air bubbles. If air bubbles are present, the tube is in the lungs. The N-G tube then needs to be reinserted properly. While many parents learn to handle this procedure, proper instructions and training are very important. You will want to give yourself a chance to feel comfortable with N-G tube insertion before attempting it on your own.



Some parents choose gastrostomy tube (G-tube) placement, which is a surgical incision into the stomach through the abdomen. In conjunction with gastrostomy tube placement, a fundoplication is usually done to help the reflux problem. Fundoplication is a surgical procedure in which the stomach is "enveloped" around the end of the esophagus, thus preventing stomach contents from "refluxing" up into the esophagus.

A parent may choose a traditional G-tube, made out of silastic or latex, or a "button" which lays flat against the abdomen. Insertion of

the "button" may have to wait until the original incision is healed.

The decision to place a G-tube or use an N-G tube is a big one, as it is often the first major intervention in our children's lives. Indeed, some families may decide not to tube feed at all. One struggles with the desire to protect the children from such intervention, while trying to find ways to make them most comfortable.

*"We discussed the idea for months. It was a most difficult decision. We were strongly against medical intervention that would prolong*

*Matthew's life. Yet the part that was so painful was that we did not want to be responsible for killing him, since feeding Matthew had become a risky activity."*

*"Feeding had become so difficult; he was losing weight and he had so many respiratory problems. We prolonged it for as long as we could. But once the crisis of the surgery was over, we knew that we had made the best decision."*

*"When my husband and I came to the full realization of what was going to happen to our son, we decided not to place an in-dwelling gastrostomy tube. This decision was made in private without counseling from anyone. It was made over a period of months, as we witnessed the progression of this devastating disease. We decided that to the best of our knowledge, when our child could no longer eat without surgical intervention, he would be permitted to die."*

**TUBE FEEDINGS**

Most parents use a traditional G-tube pump for the regular feedings which can be run by battery for some time after being unplugged. A gravity fed bag can be used for traveling; however, there is a danger that the formula will go in too

quickly, so it must be monitored carefully. Another method is called a bolus feeding (a certain amount going in all at once) in which a 50cc syringe is refilled with food and given evenly over an hour or so. This technique is handy for traveling, or in case of electrical failure.

You may choose a traditional formula or you may make your own in a blender from table food. The following recipe was made by a parent each morning for her sixteen month old and used throughout the day. The recipe was given to her by a nurse who researched G-tube feedings.

It is important that both the non-commercial and commercial feedings be evaluated periodically

for correct caloric and nutritional content as the child grows. It is also important to give plenty of "free" water (plain water), and to flush out the tube with water after each feeding to prevent blockage. Your health care provider can assist you with this information.

Even when a G-tube is placed, it is important to continue some level of oral feedings for as long as possible to maintain the swallowing ability as well as to stimulate the sense of taste and smell. Considering, however, that oral feedings at this stage may be very difficult and that adequate nutrition is provided by tube feedings, "tastes" of food are sufficient to maintain these skills.

INGREDIENTS	CALORIES	CARBOHYDRATE	PROTEIN	FAT
1/2 CUP COOKED CREAM OF WHEAT	68	15	2	
1 JAR BABY BEEF	100	1	14	5
4 OZ. ORANGE JUICE				
1/2 JAR BABY CARROTS	17	3.5	.5	
6 TBSP. NON-FAT DRY MILK	166	23.33	16.22	.37
2 SLICES WHOLE WHEAT BREAD	136	30	4	
3 TBSP. CORN SYRUP	180	45		
3 TBSP. LITE CORN OIL	450			50
1 3/4 CUPS WATER	-	-	-	-

Another method of stimulating the sense of taste is to use a foam swab (Toothette) dipped in flavored water. One occupational therapist calls this “gum on a stick” because children like to chew on it.

#### CONSTIPATION

Children with neuro-degenerative disorders eventually develop problems with bowel movements. This is due to changes in the nerve cells of the intestines that result in a decrease in gastric motility (movement of the stool through the intestine) and is complicated by the relative inactivity of our children. Certain medications can also aggravate the problem.

It is not uncommon for a child to go several days without a bowel movement and not feel uncomfortable. However, severe constipation can cause a great deal of discomfort, and measures should be taken to prevent this problem.

As it becomes more and more difficult for your child to accept adequate amounts of fluid by mouth, severe constipation can result. Once tube feedings become the main source of nutrition, the problem with constipation generally subsides, although this is not always the case.

It is important to include plenty of “bulk” or “roughage” and fluids in the diet of an orally fed child. Fresh fruits, vegetables, and grains such as bran flakes should be incorporated into the diet. As liquids become more difficult to take, they can be thickened with bran flakes. If prune juice is used, keep in mind that it can cause diarrhea when given in large amounts.

For G-tube feedings, one parent recommends a soy-based formula to help with constipation. Another suggests adding one tablespoon of vegetable oil to the food or three tablespoons of wheat germ. Ensure is a formula enriched with fiber. Natural laxatives, such as Metamucil®, are generally better for the child as they provide bulk which helps to stimulate the bowel. One family recommends Malt-Supex powder be added to each feeding.

Even though our children can’t get exercise in a traditional way, exercising the arms and legs passively (someone else moving the arms and legs) may be helpful in improving digestion. Massage of the abdomen along the large colon (up the right side, across the top, down the left side) may help to move the stool along.

It is also not unreasonable to place the child on the potty chair after feeding to take advantage of the gastrocolic reflex, or urge to have a bowel movement at this time. Thus gravity is allowed to help with this normal process. The child can be held on the potty chair in a slightly bent forward position. Of course the child will enjoy any positive reinforcement after a job well done!

It is best to try to prevent constipation by the above measures. Once the child is constipated, there are several ways to deal with the problem. Fleets® enemas for children are good. The containers can be cleaned and reused by making your own solution of water and liquid soap (one tablespoon soap and fill with water). Mineral oil enemas (the plastic containers can also be cleaned and refilled with mineral oil) or glycerine suppositories can be used as well. Stool softeners are also available under various names. And stimulating the rectal area with a lubricated cotton swab or thermometer may encourage a bowel movement.

Occasionally it is necessary to disimpact (manually remove) the rectum of stool. Use a latex glove with lubricating jelly (or a finger cot). You will want to make sure

that your finernail is short before performing this procedure.

Continued severe straining during bowel movements may cause hemorrhoids, rectal fissures or even rectal prolapse (where the rectum actually herniates outside the anus). It is important to keep constipation under control to avoid these complications. If the rectum does become inflamed with hemorrhoids or fissures, ask your health care provider for a hemorrhoidal cream with steroid (Anusol®-HC) which provides a soothing, anti-inflammatory effect.

#### ADMINISTERING MEDICATIONS

Medications for children generally come prepared as liquids for oral use. To make them more palatable, they are generally sweetened, artificially colored, and have alcohol added. You may prefer to shop around for over-the-counter medicines for children that are prepared without sugar or alcohol (e.g., cold or pain medications).

Because of these additives in liquid medications, you might ask about getting medications prescribed in pill form. Pills can be crushed with a mortar and pestle (found in gourmet food stores) and given in a feeding tube or by mouth in apple

sauce or some other food. The pills can also be soaked instead of crushed. Some pills may be difficult to dissolve, so you may have to go back to the liquid form.

Consider rectal suppositories as another means of administering medications. Tylenol® (acetaminophen) is available by suppository. Also available are glycerine suppositories for constipation. Other specific medications mentioned by parents are Tigan®, Phenergan® and Dulcolax®. Ask your pharmacist if a particular medication your child needs is available in suppository form.

When giving medication via feeding tube it is important not to mix it with the feeding. If the feeding has to be stopped for some reason or another problem occurs, you will not know how much of the medication was absorbed. It is also important to flush the tube with water after administering a medication through the tube to ensure that the whole dose of medication enters the stomach.

#### ORTHOPEDICS

##### COMMON ORTHOPEDIC PROBLEMS

Most of the orthopedic problems that affect our children are those

that result from high tone (muscle tension) and lack of weight bearing (standing).

##### HIP PROBLEMS

Hip subluxation (partially out of the hip socket), which can lead to dislocation, can occur at an early age. This problem is generally identified during an infant physical examination. To treat this condition, the child is positioned with the legs spread apart or “abducted”. This can be accomplished either manually by proper holding techniques (e.g., holding the child with your arm between the legs to keep them apart), by pillows wedged between the legs, or by the use of a special brace. Dealing with special problems like these has prompted several parents to explore unique and creative solutions.

*“My child was given a brace called a Pavlik Harness® at six months of age for his subluxed hip. It strapped over the shoulders, attaching to the thighs to hold them in abduction. The straps dug into his shoulders, causing discomfort and abrasions. He cried constantly while in it. My mother and I designed a “vest” that attached to the thighs in a much more comfortable way for my child.”*

Parents should not feel inhibited about designing any equipment that may make their child more comfortable and make life easier. Should you find a helpful and creative solution to a problem, please share your experience with NTSAD so that other parents can benefit from this information.

If your child's hip problem progresses to dislocation, and your child is not walking or standing, it may not be a problem. In the absence of weight bearing, there is usually no discomfort from a dislocated hip.

#### CONTRACTURES

Another common orthopedic problem is contractures. With this condition, the joints remain slightly flexed (bent) and eventually are no longer able to completely return to their normal position. This condition inevitably occurs over time. The best way to prevent or limit contractures is to continue a regular physical therapy program which includes exercising the joints, ligaments and muscles with passive exercise techniques. Some of the common contractures are: wrist drop and foot drop, as well as contractures of the knees.

Various splints can be worn during parts of the day or during the night to help prevent the progression of contractures. Ankle-foot orthotics (AFO Splints) can be used to prevent foot drop. These splints generally are made from a thin plastic material, extend over the heel and foot up to the mid calf and keep the foot at a 90 degree angle. They are worn over a sock, but you can put a sock and shoe over it as well. Surgery can also be done to correct tight heel cords that can cause the foot to point downward. Hand splints can be worn to help keep the thumbs abducted (spread apart). Neoprene® Sof-Splints allow for normal thumb movement and help to prevent cortical thumb (when the thumb crosses in over the palm).

#### SCOLIOSIS

Curvature of the spine (scoliosis) can also be attributed to high tone, which gradually causes the spine to curve towards the area of greater tone. If the curve becomes severe, it can cause discomfort as the hip and lower rib cage begin to meet and the internal structures become compressed. A body jacket is the one answer to help stall the progression of scoliosis and give the child relief from discomfort.

*"A body jacket was just one more intervention that I could not deal with. I couldn't imagine him being comfortable in that hard plastic. I also had trouble with the idea of the plastic coming between our bodies while we were snuggling. But he was clearly becoming uncomfortable as the scoliosis progressed. The body jacket definitely helped. His scoliosis actually improved and he is now more comfortable. I just take it off when I want to snuggle with him."*

#### PHYSICAL THERAPY

The ongoing involvement of a physical therapist in your child's care is very helpful in preventing various problems and in monitoring existing ones. Early intervention programs may provide this service. In some states, supportive services such as physical therapy are included within the scope of special education under Entitlement Law 94-142. Check your state or local programs to determine what services are available in your region.

#### DEALING WITH SPECIAL EQUIPMENT

One of the most difficult transitions we must make as parents of special

needs children is to begin to use adaptive equipment which finally identifies our children as being "different." For a while, we can get by with traditional car seats and strollers, but eventually, as our children grow, the traditional equipment no longer provides the support the children need.

Here again a parent's ingenuity can provide successful solutions. As an interim measure, one parent used a "twin" stroller with a front and back seat. By putting the back of the front seat down she made a "longer" stroller which worked very well for transporting her child. Graco makes a good twin stroller. It can be further adapted if need be.

Eventually, however, most children require special equipment. The first transitional piece of equipment is usually a wheelchair. No longer can a parent go to the mall "unnoticed." This is a big, brave step, but if we are to function and be part of the world around us, it is one that many of us learn to take and somehow learn to cope with. (See *Positioning for Feeding* section.)

*"With our children, we must continually accept with reluctance."*

#### WHEELCHAIRS, RAMPS, CAR SEATS

Adaptive wheelchairs provide good head support and can be adjusted to the contour of the child's body, providing comfort as well as helping to slow the progression of orthopedic problems. Some of the smaller wheelchairs, such as the Mulholland Travel Chair®, convert to a car seat which is an added convenience. Other stroller-type wheelchairs that convert to car seats

include: Snug Seat and Tumble Form Carrie Seat® and Carrie Rover® (the stroller part). A good car seat that has been recommended by parents is the Evenflo 7-year Car Seat®.

As the child gets heavier and more difficult to carry to and from the house, parents may want to consider installing a wheelchair ramp. Because of new accessibility laws, many carpenters are becoming familiar with building



codes for adding access ramps to existing structures. Increasingly attractive and “less obvious” ramps are becoming more commonplace. While building a ramp can be an expensive project, funding assistance may be available from various sources.

**SOME THOUGHTS ABOUT FINDING THE NECESSARY RESOURCES TO HELP MEET THE GROWING NEEDS IN YOUR FAMILY:**

Many of us find it very difficult to ask for help. But this is an extraordinary experience we are living through, with extraordinary demands placed on our time, energy and resources. Along the way, we will all be faced with the challenge of learning new skills; one of those challenges may be learning to ask for, or simply to accept, help.

There are organizations out there which are willing to help families with sick children. Indeed, some are specifically dedicated to that goal. When you are confronted by a special need, research the facilities and services available in your area. Check with the NTSAD office to see if their resource files can provide any leads. You may want to ask other parents for guidance as well; many of us have become, by

necessity, expert detectives in seeking out valuable resources. And by all means, when friends and family want to help, let them. In the end, not only you will get some much needed assistance, but those who offer that helping hand will feel good about being useful.

**NEUROLOGY**

**SEIZURES**

Many of our children have seizure disorders of one kind or another. Although undesirable, seizures may not be as damaging to a child as we might expect, as long as the seizure is not prolonged. A prolonged seizure can cause anoxia (loss of oxygen) to the brain.

Seizures usually can be controlled with medication. The kind and amount of medication may change over time. An increase in the frequency or severity of a child’s seizures may be due to a progression of the disease or may indicate that the dose of the anticonvulsant is not high enough for his or her weight. Optimum doses of medication can be monitored in part by blood levels drawn on a periodic basis. In some cases, vigorous efforts to control seizures may not be justified.

**DEALING WITH FEVERS**

Fevers tend to go quite high in our children and can accelerate quickly because of the instability of the fever center in the brain. High fevers need to be treated aggressively, because they can precipitate seizures.

You need to know how to read a glass thermometer. It is best not to rely on digital thermometers, as they can be inaccurate. Rectal temperatures are most accurate (three minutes). Axillary (under the arm) temperatures take too long (ten minutes).

Always have acetaminophen (Tylenol®) on hand for fevers. Never use aspirin for fevers because of the risk of Reye’s Syndrome. Have the acetaminophen available in both suppository form and liquid form so that you have options for use. Use the acetaminophen for any fever over 102° F. (Ask your health care professional if he or she wants it given sooner than this.) Fevers under 102° F should be closely monitored for sudden elevation. With a very high fever, over 104°F, you can try a sponge bath (not tub bath) using tepid water (lukewarm). Lay the moistened cloths on the child’s forehead, armpits and groin. Another method to control high

fevers is “fanning” the body with a hand-held fan or piece of paper to promote evaporation. If you use an electric fan, make sure it is across the room from the child. Keep your child covered with a sheet or light blanket so as not to chill the body too quickly.

**IRRITABILITY: COMFORTING YOUR CHILD**

One of the common symptoms of neurologically impaired children is irritability. Irritability can be caused by neurological immaturity or the disease progression itself. It is often compounded by related conditions such as reflux, orthopedic problems or other discomforts. Your child may “grow” out of a stage of extreme irritability as he or she matures. If irritability is ongoing, and you sense your child is experiencing discomfort, speak to your health care provider. A specific problem could be identified and corrective measures taken. For example, esophagitis (inflammation of the esophagus) caused by reflux can be very uncomfortable for the child. A fundoplication may correct the reflux problem (*See G-Tube section.*)

One of the many challenges we must face is how to console an irritable child. The challenge is not only in finding ways to console your child, but also in finding ways to take care of yourself, as the child’s irritability can be ongoing and very stressful.

*“When my son was in for an asthma admission, prior to G-tube insertion, I recall one night that I’ll never forget. The only thing that would console him was having me walk him up and down the halls. I did this for six hours straight, holding him in my arms with an IV pole in tote.”*

Singing, humming, or just the sound of a familiar voice can help. Music is soothing also. Sometimes the child just needs to hear a continuous familiar sound. One thing that usually works is motion: holding, rocking or walking the child.

*“We had our washing machine in the kitchen. When my son became fussy, I would put him on top of the washer in his infant seat, and put it on spin. The vibration would always console him.”*

(If you decide to try this method, make sure that the seat is secured somehow to the washer and that the child is strapped into the seat.)

It is important not to overstimulate a child if he or she has a tendency to become irritated. For example, many children get extremely upset when their faces are touched. Loud or sudden noises, although enjoyed by some children, may cause irritability in others. Sometimes a child can be kept more comfortable with medications to alleviate discomfort or encourage sleep.

*“There were times when I would become so frustrated because of the constant irritability that I would just resort to giving him a medication for sleep. Nothing else seemed to work. I had to convince myself that this was O.K. to do.”*

**SLEEPING PROBLEMS**

Sleeping difficulties are a common problem with these children. They may be up several times a night for various reasons: muscle spasms, reflux discomfort, nasal congestion. They may not take naps during the day. It may not be possible for your child to “cry himself to sleep.” As the crying becomes more intense, muscle tone, reflux or wheezing can increase causing greater discomfort. Occasionally, just patting the child on the back will work, but



often he or she will have to be picked up and held, rocked or sung to. If sleeping problems are chronic it may be helpful to consider giving a sleeping medication on an occasional basis. Chloral Hydrate is probably the most useful sleeping medication available for our children. Some suggestions: Providing a warm, comfortable bed is essential. Water mattresses are excellent for

our children. Sheep skin pads and egg crate foam pads also work well. If your child has difficulty going back to sleep at night, try to arrange medications and treatments so that he or she does not have to be awakened. If the child does wake up, do the diaper change, turning, treatment or medication during that time, if at all possible.

### DERMATOLOGY

#### GOOD SKIN CARE: AVOIDING PROBLEMS

Good skin care is especially important for many reasons. Our children are relatively immobile and are susceptible to skin breakdown and pressure sores. Frequent turning and position changes are essential. Body prominences, such as hips, shoulders, shoulder blades, etc., should

be checked regularly for redness or irritation (pressure areas). If a pressure area shows up, the child should be kept off that area until normal circulation resumes. Any skin breakdown should be monitored for signs of infection (redness, swelling or discharge).

Because our children are in diapers, frequent changing of wet diapers is important, both for asthetic reasons and because prolonged exposure to moisture can cause rashes.

Good skin care around the G-tube (Gastrostomy tube) site is important to help cut down on drainage, prevent infection and to keep the child more comfortable. The area can be cleansed with a solution of half hydrogen peroxide and half water. This will sanitize the area and help to debride (remove) any buildup of secretions. If there is an area that looks irritated at the stoma site (opening for the G-tube to enter the stomach), an application of a silver nitrate stick will cauterize the area, promoting healing. This causes momentary discomfort, but the subsequent healing from the application should reduce the drainage at the site. If there is chronic drainage and redness at the

site and no obvious sore spot, the irritation may be a reaction to the material the G-tube is made of. In this case, it may be worthwhile to consider changing the type of material (e.g., latex to silastic). (*See G-Tube section.*)

### BATHING

Bathing is a special time to be with your child, although it does require some organization and planning. The bathing room (bathroom or kitchen) should be warm. A time should be set aside and things should be relaxed, without too much interruption (e.g., phone). The temperature of the bath water should be tested first by you (using your elbow) to make sure it is not too hot. Soap should be used sparingly; too much can cause skin dryness and itching. Talcum powder should not be used at all, as it can be inhaled and may cause lung irritation.

One family recommends using dotted jersey gardening gloves for handling a slippery child during bathing. This is especially important when taking the child in and out of the tub. The gloves also provide some nice tactile stimulation as you are washing your child. Using a Water Pic shower massage

was also recommended to provide tactile stimulation.

A regular kitchen sink may be fine for bathing your child for awhile (the height is usually good). You may want to consider installing a longer sink to accommodate your child as he or she grows.

Eventually your child may outgrow any sink, and you will need to try tub-bathing. Since lifting a heavy child in and out of a regular bathtub can be difficult, bathroom renovations may become necessary. If financially feasible, consider installing an elevated bathtub with a hand held shower. This makes it easier for an older child to be bathed in a bath seat, and lifted in and out of the bathtub. One family had this type of tub installed. When it was not being used for the child, a movable step was placed in front of the tub, giving it the appearance of a "Roman Bath." They also installed a sink with a long counter, making an excellent changing table.

Good bath supports will make life easier. When your child is very young, you can place him or her on a bath sponge, or a "Tubby"

inflatable raft-type cushion. An inflatable bath pillow will help to support the child's head. This is especially helpful with children who have enlarged, heavy heads. Rifton makes an excellent bath seat. Columbia makes another. One family designed their own seat with PVC piping and lawn chair-type webbing from a hardware store.

## BEHAVIOR

### STIMULATION, PLAY AND COMMUNICATION

Families are wonderfully creative in finding ways of interacting with their children. Since many of our children are visually impaired, we look for learning or creating ways to encourage stimulation of the other senses (*see Eye section*). We all know what a gift it is to hear our children laugh or see them smile, and we all will do just about anything to get those responses!

As they get older and the neurological system has a chance to mature, our children may become less irritable and more open to play. The chronically irritated child who hates to have his or her face touched, and has never smiled, may grow out of this behavior.

Sensory stimulation activities are varied and are open to much creativity, depending entirely on your child's specific situation. Basically, anything that will stimulate your child's sense of sight, hearing, taste, smell, and touch, as well as sense of motion, is included in these activities. The senses should be stimulated whenever possible, even if there is some sensory impairment. We can never be totally sure of the level of our children's vision or hearing impairments.

Part of play is also learning. Although it is difficult to measure what our children are capable of learning, we shouldn't assume that they are unable to learn.

*“Robert has been in an integrated program at our public school for a few years now. I know that he has ‘learned’ counting on a number of occasions. When I fold the laundry with him, I ‘count’ the socks, undershirts, etc., over and over again. He really seems to enjoy this as if he understands what the numbers mean.”*

One way to incorporate “cause and effect” learning into your child's play is to practice anticipatory skills by saying “ready” prior to intro-

ducing the stimuli. You might want to try this technique while feeding your child.

### VISUAL STIMULATION

Bright flashing lights, contrasts of lights and darks, intermittent introduction of stimuli, and movement of stimuli can serve to encourage vision.

Several of the newer baby mobiles are made with black and white designs. Black and white are good colors for early visual development; then progress to red and other bright and shiny colors. Shiny pom poms and pinwheels are great! Shiny or colored tape can be applied to less interesting toys and objects to encourage tracking. Toys with lights, especially flashing lights, are also appealing.

Toys are available that can be adapted with large, easy to operate switches so that children with neurologic impairments can turn them on and off (Switch toys). Mirrors (made of soft material, not glass) can be taped to the bed or play areas.

### AUDITORY STIMULATION

Our children seem to enjoy lots of sounds and will often “complain” if

it is too quiet. Provide tapes with both soothing and more lively music, depending on the mood you are attempting to create. Some children enjoy sudden, loud noises (dropping pots and pans or pretending a loud sneeze). This is one activity that siblings greatly enjoy helping with. Provide as many types of sounds as you can think of for variation: music boxes, radios, “Walkman-type” tape players (make sure that the volume dial is fixed somehow to prevent accidental turning up of the volume), stuffed animals and other toys that make sounds. There are several battery-run musical instruments (e.g., keyboards) that your child can “play” while you place his or her hands on the instrument. This will also encourage “cause and effect” learning. Keep in mind that some children become very irritable when exposed to loud and sudden noises. Sensory stimulation activities need to be adapted to each child's needs.

If your child is to be away from you (e.g., during hospitalizations or respite care) you might consider a tape of familiar family voices singing, laughing and talking, or the dog barking. There are lots of tapes available that have various

sounds your child may enjoy (different animal sounds, ocean sounds, etc.)

### OLFACTORY STIMULATION

The sense of smell can be stimulated by utilizing vials of food flavorings (peppermint, vanilla, orange, etc.) or by just having your child smell the family's dinner. And, of course, throwing in words like “spaghetti sauce” may encourage learning. Likewise, if your child is able to, tastes are encouraged so that the sense of smell is connected with the sense of taste. Smelling and tasting have the added benefit of stimulating the appetite and aiding in digestion.

Note: If your child tends to have seizures, you might try to discover if anything in particular seems to bring them on. Certain colognes, for example, have been known to precipitate seizures.

### TACTILE STIMULATION

Any physical contact that you have with your child will stimulate his or her sense of touch. Providing tactile cues as a part of routine daily activities allows your child to participate more fully in his or her life. You can have your child hold the toothbrush before mouth care,

touch a special toy before playing with it, or feel an item of clothing before dressing.

Look for things that provide a variety of textures for your child to touch with his or her fingers or for you to rub on the child's skin. Many “special” toy catalogs (*see Resources in Appendix*) have toys designed with various textures. You can make your own texture board or blanket (or have a special friend or relative make it for your child). Any rough, smooth, ribbed, rubbery, sticky surface will do. You may want to experiment with liquids and other textures as well (play dough, finger paint, shaving cream, pudding!). The possibilities are endless, and your child will enjoy the new experience.

One way of encouraging cause and effect is to provide tactile “cues” for your child when you or someone else greets him or her. For example, help him or her to touch your ring or watch as you repeat your name.

### VESTIBULAR STIMULATION (MOVEMENT)

Children generally love to be held, bounced, walked or rocked. Sometimes, this is the only thing that works with an irritable child (*see Irritability section*). Some children also



enjoy more vigorous movements like swinging or rocking. Twirling around, however, may upset your child because of immaturity of the vestibular system (which gives us our sense of balance).

It is important to provide a variety of “places” in which your child can play and learn. Bean bags are great because they mold to the child (and siblings enjoy them as well). Hammocks can be hung in a common room and are easy to take down when not in use. Make sure that your child is safely secured in the hammock and that you are standing next to him or her. Hammocks provide motion and vestibular stimulation.

Corner chairs, which are triangularly shaped, provide good support for sitting and can be easily adapted to fit your child. For smaller children, a baby carrier (e.g., Graco Courier Soft Baby Carrier) provides good head support and enables you to take your child around with you as you work around the house. A carrier also allows for greater accessibility outdoors. Prone standers (e.g., by Mulholland) allow the child to “stand up” supported, freeing the arms for movement. A seat low to the ground with wheels allows the

child to be easily moved and to be “where the action is” if other children are around. (You can order a low, removable base on wheels for a Tumble Form seat.)

Many parents report that waterbeds are great and provide a range of vestibular bouncing and rolling sensations. Waterbeds can be particularly helpful during periods of fussiness. Children with poor head control need to be watched carefully while on a waterbed, however, to avoid suffocation.

Provide cues for your child before moving him or her. Say “We’re getting out of the chair now” and touch his or her arm. Or say, “Ready” or “1-2-3.” This helps your child to be part of the activity. It also avoids startling the child.

*“I always told him what I was doing and even though he may not have understood me, he knew the routine, he trusted me and approaching him this way always seemed to minimize his fears and seizures.”*

One thing that many of us have learned through the years is that the total care of our children has many dimensions, and that we do not have to continually meet needs that others perceive as important.

*“There were so many things that had to be done for my child: medications, tube feedings, exercising and other necessities of daily care. Then there were all the ways in which I could ‘stimulate’ him that the therapist suggested. There was so much that I had to do, when all I wanted to do was to sit and hold him and love him.”*

Each of us and each of our children are unique and we need and deserve the “space” to exercise our own judgments in our own ways.

#### COMMUNICATING WITH YOUR CHILD

As parents we are usually very sensitive to a child’s subtle signs. We can tell if the child’s cry is one of pain or boredom; if the child’s face is showing discomfort or hunger. We are the experts in interpreting the needs of our children.

Vocalizations from our children are little gifts. Smiles and laughs are even greater gifts. Vocalizations can be encouraged by repeating the sound after it is made by the child. Encouraging laughter is usually one reaction we all seem to strive for. Be it throwing a cookie sheet on the floor to make a sound or faking a loud sneeze, or anything

else, we’ll go to great lengths to get a laugh from our children.

Use every opportunity to look for signs of communication from your child, and make an attempt to reinforce any attempt that your child makes to communicate. A signal may be as simple or subtle as a tense muscle. These signs may initially be random, but if you give the signal a meaning, perhaps your child will learn to give it a meaning as well.

*“I would talk a lot to him and reassure him. He responded to me and totally trusted me. He knew my voice, my touch, my smell...as he lost some senses, others would become more pronounced...don’t underestimate what they can do –and adapt!”*

DEFINITION OF TERMS:

**ABDUCT:** To draw away from the middle of the body.

**ANOXIA:** Deficiency of oxygen.

**ANTI HISTAMINE:** A medication used to treat allergies.

**ANTISPASMODIC:** A medication used to relieve spasms.

**ASTHMA:** Difficulty breathing, and wheezing, caused by spasm of the bronchial tubes or by swelling of their mucous membranes.

**BOLUS:** A mass of food given at once.

**BRONCHOSPASM:** Spasmodic narrowing of the bronchial tubes.

**DISLOCATION:** The displacement of any part of a bone from its normal position in a joint.

**ESOPHAGITIS:** Inflammation of the esophagus.

**EXTENSION:** A movement which brings the members of a limb into or toward a straight condition, for example, straightening out of the arm; opposite of flexion.

**FISSURE:** An ulcer or cracklike sore.

**FLEXION:** The act of bending or condition of being bent in contrast to extension.

**GASTROESOPHAGEAL REFLUX:** A return or backward flow of stomach acids or contents up into the esophagus.

**GASTROCOLIC REFLEX:** Peristaltic wave in the colon induced by the entrance of food; this will often produce to urge to have a bowel movement.

**HEMORRHOID:** A mass of dilated veins in the anus, which can become inflamed, causing itching or pain.

**HYPERTROPHY:** An increase in size of a structure or organ which does not involve tumor formation.

**NYSTAGMUS:** Involuntary, cyclical movement of the eyeball.

**PASSIVE EXERCISE:** Muscular exercise without any effort on the part of the patient.

**PNEUMONIA:** Inflammation of the lungs caused primarily by bacteria, viruses, and chemical irritants.

**RECTAL PROLAPSE:** Protrusion of the rectal mucosa through the anus.

**SCOLIOSIS:** Curvature of the spine.

**SEIZURE:** A sudden attack of involuntary muscular contractions and relaxations.

**SUBLUXATION:** A partial or incomplete dislocation of a joint.

**WHEEZE:** A whistling or sighing sound resulting from narrowing of the respiratory passageway.

ABBREVIATIONS

COMMON MEDICAL ABBREVIATIONS:

q	every
qd	every day
qod	every other day
q wk	every week
q4h	every four hours (q6h, q8h, etc.)
bid	twice a day
tid	three times a day
qid	four times a day
e	with
s	without
p	after
ac	before meals
pc	after meals
pt	per tube
gtts	drops
prn	as needed
po	by mouth
P.T.	physical therapy
R.O.M.	range of motion

SPECIAL EQUIPMENT

- ▶ Tumble Form Carrie Seat and Carrie Rover
- ▶ Mulholland Travel Chair
- ▶ Snug Seat
- ▶ Evenflo 7 Year Car Seat
- ▶ Rifton Bath Seat
- ▶ Columbia Bath Seat
- ▶ Mulholland Prone Stander
- ▶ Corner Seat
- ▶ Graco Courier Soft Baby Carrier
- ▶ Mead Johnson - Cleft Palate Nipple
- ▶ First Years Infa-Feeder

National Tay-Sachs and Allied Diseases Association was founded in 1956 by a small group of concerned parents committed to the eradication of Tay-Sachs and related genetic disorders. These parents shared a common tragedy; each had a child affected with a rare, fatal genetic disease.

Over the past four decades, NTSAD has grown in size, scope and stature. NTSAD services now encompass 40 genetic diagnoses, including adults affected by a rare Adult Onset form of Tay-Sachs disease. Fueled by a partnership of dedicated volunteers, gifted professionals and a distinguished Scientific Advisory Committee, NTSAD programs of Public and Professional Education, International Laboratory Quality Control, Carrier Screening, Family Services, Advocacy and Research serve a diverse and international constituency.

NTSAD's Parent Peer Group, a Family Services program, links families affected by Tay-Sachs and the allied diseases to a unique lifeline of information, resource and support. The Parent Peer Group is coordinated by volunteers who are themselves parents of affected children. A newsletter, family directory and national telephone network, all funded through NTSAD, connects hundreds of Parent Peer Group families. An extensive lending library is also available to families. Additionally, NTSAD sponsors an Annual Conference and family scholarship program.

Today, NTSAD is recognized as a leading, non-profit, voluntary health organization. For further information on NTSAD, please call or write:

**NATIONAL TAY-SACHS AND ALLIED DISEASES ASSOCIATION, INC.**

2001 Beacon Street, Suite 204  
 Brookline, Massachusetts 02146  
 (617) 277-4463  
 (617) 277-0134 FAX

**TAY-SACHS AND THE ALLIED DISEASES**

Tay-Sachs and the allied diseases are rare, fatal genetic diseases that primarily affect young children.\* These disorders are characterized by a common biochemical defect: the inability of the body cells to dispose of certain metabolic waste products. The waste products gradually accumulate in the cells of the affected children, causing a variety of debilitating symptoms and ultimately death, usually by early childhood. To date, a cure remains to be found for Tay-Sachs and the allied diseases:

- GLYCOSPHINGOLIPIDOSES
- LANDING'S DISEASE (GM 1 GANGLIOSIDOSIS)
- TAY-SACHS, SANDHOFF DISEASE (GM 2 GANGLIOSIDOSIS)
- FABRY'S DISEASE (TRIHEXOSYLCERAMIDOSIS)
- GAUCHER'S DISEASE (GLUCOSYLCERAMIDOSIS)
- NIEMANN-PICK DISEASE (SPHINGOMYELINOSIS)
- METACHROMATIC LEUKODYSTROPHY (SULFATIDOSIS)
- KRABBE DISEASE (GALACTOSYLCERAMIDOSIS)
- FARBER'S DISEASE (LIPOGRANULOMATOSIS)
- MUCOPOLYSACCHARIDOSES (MPS)
- HURLER SYNDROME (MPS I-H)
- SCHEIE SYNDROME (MPS I-S)

- HUNTER SYNDROME (MPS II)
- SANFILIPPO SYNDROME A,B,C,D (MPS III A,B,C,D)
- MORQUIO SYNDROME (MPS IV)
- MAROTEAUX-LAMY SYNDROME (MPS VI)
- SLY SYNDROME (MPS VII)
- OLIGOSACCHARIDOSES
- MANNOSIDOSIS
- FUCOSIDOSIS
- ASPARTYLGLYCOSAMINURIA
- MUCOLIPIDOSES (ML)
- SIALIDOSIS (ML I)
- I-CELL DISEASE (ML II)
- PSEUDO-HURLER POLYDYSTROPHY (ML III)
- ML IV
- ADRENOLEUKODYSTROPHIES
- A-BETA-LIPOPROTEINEMIA
- BATTEN'S DISEASE
- CANAVAN'S DISEASE
- CEREBROTENDINOUS XANTHROMATOSIS
- CHOLESTERYL ESTER STORAGE DISEASE
- PELIZAEUS MERZBACHER DISEASE
- POMPE'S DISEASE
- REFSUM'S DISEASE
- TANGIER DISEASE
- WOLMAN'S SYNDROME

\* The exception being Late Onset Tay-Sachs disease: a rare disorder affecting adults that is commonly misdiagnosed as multiple sclerosis or similar muscular/neurological disorders. Additionally, symptoms of manic-depressive psychosis are present in nearly 40% of affected persons. A newly recognized disorder, the course of Late Onset Tay-Sachs disease is not completely known. It appears however, that life expectancy is probably not reduced.

**ADDITIONAL RESOURCES**

- ALLIANCE OF GENETIC SUPPORT GROUPS**  
 35 WISCONSIN CIRCLE, SUITE 440  
 CHEVY CHASE, MARYLAND 20815  
 1/800/336/GENE
- CHILDREN'S HOSPICE INTERNATIONAL**  
 700 PRINCESS STREET, LOWER LEVEL  
 ALEXANDRIA, VA 22314-2265  
 800/242-4453
- COUNCIL OF REGIONAL NETWORKS FOR GENETIC SERVICES (CORN)**  
 CORNELL UNIVERSITY MEDICAL SCHOOL  
 1300 YORK AVE., GENETICS Box 53  
 NEW YORK, NEW YORK 10021  
 212/746/3475
- MARCH OF DIMES BIRTH DEFECTS FOUNDATION**  
 1275 MAMARONECK AVENUE  
 WHITE PLAINS, NEW YORK 10605  
 914/428/7100
- NATIONAL HOSPICE ORGANIZATION**  
 1901 NORTH MOORE STREET, SUITE 901  
 ARLINGTON, VA 22209  
 703/243-5900
- NATIONAL INSTITUTE OF NEUROLOGICAL DISORDERS & STROKE (NINDS)**  
 NIH BUILDING 31, ROOM 8A16  
 BETHESDA, MARYLAND 20892  
 301/496/5751
- NATIONAL ORGANIZATION OF RARE DISORDERS (NORD)**  
 P.O. Box 8923  
 NEW FAIRFIELD, CONNECTICUT 06812  
 1/800/999/NORD

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